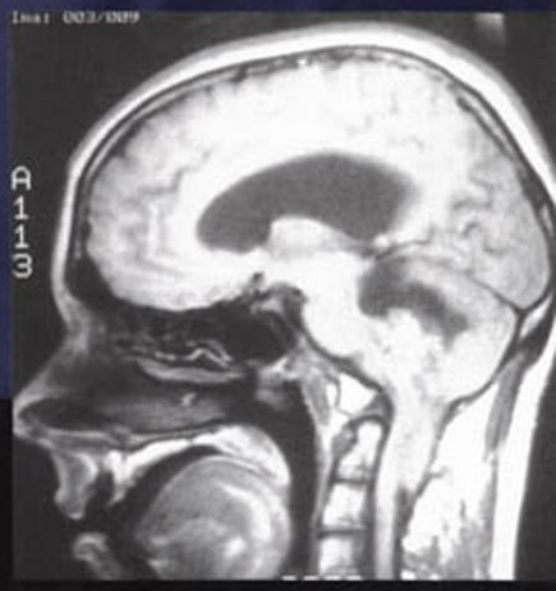
OPERATIVE TECHNIQUES *in* PEDIATRIC NEUROSURGERY



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Operative Techniques in Pediatric Neurosurgery

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This book is dedicated to our wives,
Elizabeth, Connie, and Barbara,
and to the children whose care we hope to improve.

CONTENTS

PREFACE

The authors of these chapters are experienced neurosurgeons. Many have developed reputations for their expertise in performing the operations they describe. They have illustrated techniques they have found to be useful over the years. In many instances, the authors describe "little" techniques; yet these "little" techniques may contribute importantly to the success of an operation, e.g., the use of nasolacrimal stents in the treatment of sincipital encephaloceles.

In some instances, the authors describe their rationale for these techniques and in others they simply describe them as being effective. Other experienced, excellent neurosurgeons may prefer different techniques than the ones described. There are often multiple ways to surgically treat a condition effectively. We would not want readers to infer that the techniques described herein were written in stone. Authors frequently use the term "should" throughout the text; that term should be interpreted as being the author's opinion but not as constituting a "standard of care."

Advances in neuro-imaging, navigational techniques and technology have substantially altered the operations we have used to treat children with neurosurgical disorders in the past 15 years, and have made it possible for us to treat many children who would not have been treatable previously. Within the next 15 years, advances in in-utero surgery may improve the prognosis for children with spina bifida; deep brain stimulation may improve the outlook for those with movement disorders; radiosurgery may treat those with mesial temporal lobe seizures; and gene therapy may help both brain tumors and congenital metabolic disorders. Yet, both now and then, personal, compassionate, careful care will be as important as any technological innovations. Both now and then, the goal will be to treat our patients as if they were our own children, which, in a way, they are.

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Congenital and Developmental Disorders

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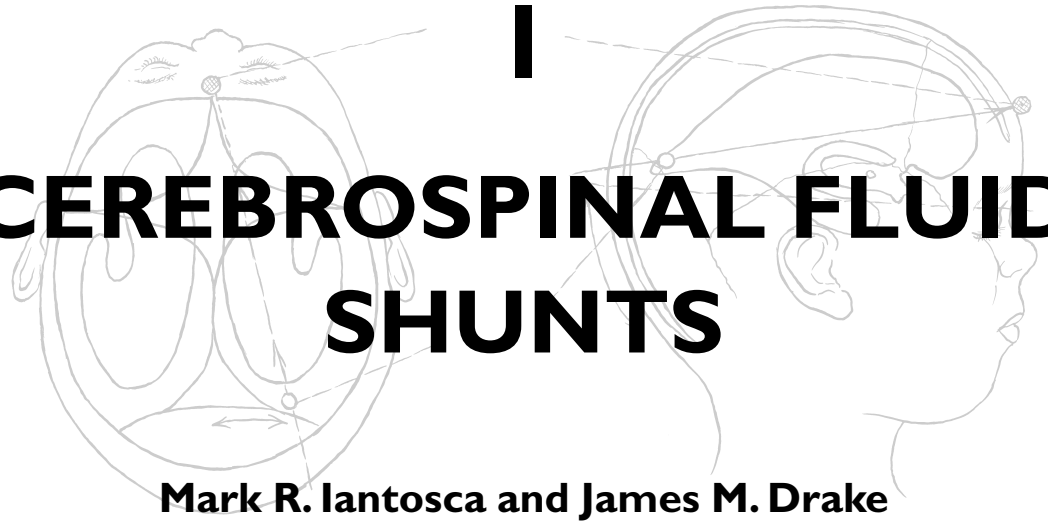
CEREBROSPINAL FLUID SHUNTS



Mark R. Iantosca and James M. Drake

Hydrocephalus is the most frequent neurosurgical problem encountered in the pediatric age group. With an incidence of 1 in 2000 births, it occurs in nearly one third of all congenital malformations of the nervous system. The treatment of hydrocephalus was revolutionized following the introduction of valved shunt systems by Nulsen and Spitz in 1952 and the subsequent development of silicone shunt systems by Holter and Pudenz. Cerebrospinal fluid (CSF) shunting procedures constitute approximately half of most pediatric neurosurgical practices, with roughly half of these procedures performed for revision of an existing shunt. Clearly, despite recent advances in shunt design, they continue to present a technical challenge for the neurosurgeon treating children with hydrocephalus.

INDICATIONS FOR CSF SHUNT INSERTION

Infants and children who present with ventriculomegaly and clinical evidence of elevated intracranial pressure clearly require a CSF diversionary procedure. Whereas conventional CSF shunting procedures have long been the main treatment option in these children, dissatisfaction with their long-term outcome and significant failure rates has resulted in a resurgence of interest in the earliest form hydrocephalus treatment: ventriculostomy. Patient groups exhibiting appropriately high success rates following third ventriculostomy are still being defined, however, and probably represent a minority of patients with hydrocephalus. CSF shunting procedures are likely to remain the treatment of choice for most children with symptomatic ventriculomegaly.

The decision to treat ventriculomegaly becomes more difficult in the patient whose symptoms are minimal, absent, or difficult to assess because of the patient's age. Radiographic progression of ventriculomegaly over time, even if asymptomatic, is generally considered an indication to treat, unless it is secondary to cortical atrophy associated with anoxia, syndromic malformations, or degenerative/metabolic disease. Less agreement exists, however, about the treatment of asymptomatic stable ventricular enlargement. The inconsistent use of the terms *arrested* and *compensated* hydrocephalus adds to this controversy. Patients with stable ventricular enlargement resulting from age-related CSF absorption abnormalities (e.g., benign extraaxial fluid collections of infancy) or venous occlusive disease (e.g., pseudotumor cerebri, achondroplasia) generally are not considered for treatment unless radiographic or clinical progression is clearly documented. Children with stable clinical and radiographic findings over the age of 5 years can be monitored closely for evidence of subtle abnormalities in intellectual development. It is important to weigh the known long-term risks of shunt complications against the largely unknown impact of asymptomatic hydrocephalus in this group of patients. Younger children, particularly those under the age of 3 years, cannot be evaluated by standard measures of intellectual development. Many authors recommend treatment of asymptomatic patients with moderate to severe ventriculomegaly in this age group to protect against the impact of ventriculomegaly on future intellectual performance. Unfor-

tunately, imaging studies and invasive testing, including infusion tests, radionuclide studies, Doppler screening, intracranial pressure monitoring, and magnetic resonance (MR) spectroscopy, to date have been unable to predict reliably which of these children may be at risk for intellectual decline.

PREOPERATIVE ASSESSMENT

Initial Presentation

The diagnosis of hydrocephalus is based on clinical and radiological features. Infants most commonly present with symptoms of irritability, delayed development, vomiting, and headache and on examination have increasing head circumference and a bulging fontanel. MR imaging has the best diagnostic utility in terms of establishing the anatomy and likely pathology of potential obstructive lesions. Computed tomography (CT) is usually sufficient for follow-up of previously diagnosed patients with existing shunt systems and preliminary or urgent evaluations of undiagnosed patients. Ultrasound is more practical in critically ill, premature infants with intraventricular hemorrhage or in infants with myelomeningocele the etiology of which is not in doubt. Moderate to severe ventriculomegaly usually can be visualized easily using any of these techniques. In patients with milder ventricular enlargement, evidence of transependymal flow of CSF usually suggests that the process is more acute. Other signs of progressive hydrocephalus—such as enlargement of the temporal horns, dilatation of the third ventricle, and effacement of the sulci—are less specific. If doubt exists, careful observation with serial images, rather than subjecting the patient to the known risks of shunt failure, is prudent.

Shunt Failure

An appreciation of the epidemiology and presentation of shunt failure resulting from mechanical or infectious complications is essential in determining appropriate shunt hardware and insertion techniques. The most common time for a shunt failure is in the first 6 months following insertion. Overall 1-year failure rates approach 40%. Most centers report infection rates on the order of 5 to 10%. These infections usually present within 2 months of shunt insertion, indicating that most infections occur during shunt surgery itself. Although the mechanism of shunt contamination seems relatively straightforward, numerous studies instituting procedures aimed at risk reduction have failed to demonstrate consistently that these interventions led to lower infection rates in some centers.

A detailed history, including all the previous shunt surgeries, is mandatory. Previous notes from the operating room (OR), from whatever hospitals the surgery was done, should be sought. The relevant imaging, which should be related to the patient's clinical status at the time they were obtained, also should be carefully studied. This entire process often requires creating a spreadsheet or log to keep track of the multiple interventions. All previous culture reports also should be sought, looking for an unrecognized or partially treated infection. Current imaging should be complete, including plain radiographs of the shunt equipment.

Examination of the site of the shunt equipment implantation may provide confirmatory evidence of shunt dysfunction. Although pumping of the shunt reservoir is a time-honored technique, in fact, it is often misleading. A patient in whom a reservoir fills slowly may simply have small ventricles; however, shunts whose reservoirs remain umbilicated for prolonged periods, or even permanently often are blocked proximally. A reservoir that is quite difficult to depress or that refills apparently instantaneously frequently indicates a distal obstruction. Fluid collecting around the shunt, particularly if it firmly distends the skin, is progressive, and tracks along the distal catheter, is often a sign of shunt occlusion. When shunts fracture, CSF often continues to track along the fibrous sheath. In this scenario, often a small amount of fluid and a space where the shunt has come apart can be felt. It may be difficult, however, to distinguish an empty sheath from the sheath containing shunt tubing, particularly if the shunt has been implanted for some years or the tract is calcified or distended with fluid under high pressure. A fluid thrill sometimes can be felt at the site of a distal catheter disruption with pumping of the proximal reservoir.

Infection must be ruled out in all patients presenting with early shunt malfunction (within 2–6 months of insertion) and in patients with clinical features suggesting infection (e.g., meningismus, fever, elevated white cell counts, abdominal pain). Exclusion of infection is achieved most conclusively by sampling the CSF, usually from the shunt reservoir itself. CSF samples from patients with isolated distal shunt infections often are sterile, however, making abdominal ultrasound or CT necessary to rule out intraabdominal abscesses or "pseudocysts." These studies are especially important for patients presenting with predominantly abdominal symptomatology. All children with suspected shunt infection should have a thorough history and physical examination to rule out other common febrile illnesses, such as otitis media and

viral gastroenteritis, which can be indistinguishable from a shunt infection. Likewise, urinary tract infections always should be considered in myelomeningocele patients.

OPERATIVE PLANNING

Shunt Selection

No data are available to determine which particular shunt should be recommended; in fact, a recent randomized trial on CSF shunt design comparing a standard differential pressure valve to both siphon-limiting (Delta valve, Medtronic PS Medical, Goletta, CA) and flow-limiting valves (Orbis-Sigmam NMT, Boston, MA) failed to show any difference in terms of overall shunt failure. Important considerations must be taken into account, however, when assessing the individual patient: age, weight, skin thickness, head size, size of the ventricles, pathogenesis of hydrocephalus, acuteness of the illness, presence of internal lines or gastrostomy, tracheotomy openings, status of the distal drainage site, and plans for further surgery.

For example, a premature infant with thin skin stretched further by a rapidly expanded head cannot accommodate adult-sized equipment without the risk of skin erosion. The presence of intraventricular hemorrhage in this patient group also may increase the risk of early obstruction following implantation of a valve with a narrow flow-limiting orifice. Conversely, in patients with large ventricles and large skulls with fused sutures, placing a flow-limiting or siphon-reducing device may decrease the risk of subdural hemorrhage. Ventricular configuration and asymmetry also may dictate the choice of insertion site.

In addition to patient characteristics present at the time of initial shunt insertion, it is important to consider future treatments that may affect shunt function. For example, endoscopic fenestration of a loculated ventricular system when the shunt is inserted may be indicated to decrease the number and complexity of future shunt systems. Patients who have elaborate shunt configurations with several connections, or multiple separate shunts, suffer higher rates of shunt failure than those who have simple shunts. Metallic shunt components or magnetic programmable valves should be avoided in patients expected to require multiple MR imaging studies to avoid degradation of the images and unnecessary reprogramming of the valve. Finally, if a patient is scheduled to have further intraabdominal surgery, for example, colostomy closure or bladder reconstruction/augmentation, the preferred distal drainage site may be altered.

For most routine cases, the surgeon probably should choose familiar shunt equipment and use the same system consistently. Current evidence does not support the superiority of any particular valve design, and so the individual surgeon's technique and ability to evaluate outcomes are likely enhanced by this approach. In this situation, we prefer a two-piece system with a nonflanged ventricular catheter, connected to a flat-bottomed valve with a reservoir, and integrated open-ended distal tubing.

Preoperative Preparation

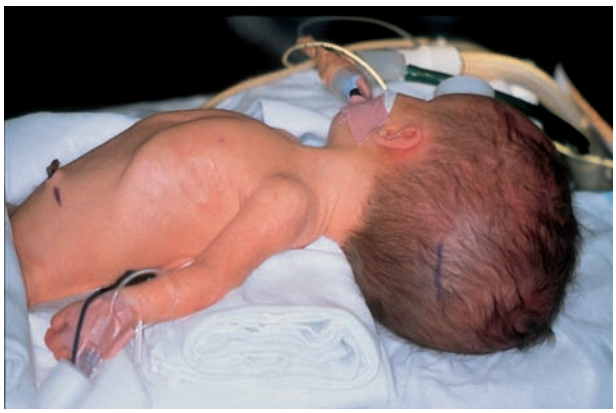
Shunt surgery is often regarded with some disdain by staff and trainee neurosurgeons alike; it has the highest failure rate of any neurosurgical procedure, and nothing is less forgiving of any technical errors than a shunt operation. It is well known that shunts frequently fail as a result of tissue occluding the proximal or distal catheter. Avoidable complications, such as intraparenchymal ventricular catheters, extraperitoneal distal catheters, and spontaneously disconnected or migrated shunts, have occurred on virtually every neurosurgical service. Shunt surgery should command great respect, require meticulous attention to detail, and be carried out in a skilled and expeditious fashion.

Body wash and shampoo the night before and again just prior to surgery using an antiseptic solution (e.g., chlorhexidine) are recommended. A number of meta-analyses have shown that prophylactic antibiotics are effective, and they are strongly recommended. Likewise, all shunt equipment is soaked in antibiotic-containing solution prior to insertion. For patients in whom an abdominal trochar is being used, the bladder should be emptied either by a Crede maneuver or by urinary catheter. Hair removal has never been shown to decrease the risk of infection; however, the hair can be clipped (not shaved) in the OR if so desired to assist with skin closure and application of postoperative dressing.

INTRAOPERATIVE TECHNIQUE

Ventriculoperitoneal Shunt

The patient should be positioned so there is a flat plane between the upper and lower incision sites, facilitating subcutaneous passage. For an occipital burr hole, this means rotating the head to the opposite side, and extending the neck, usually with a rolled towel (Figs. 1–1A and B). The site of the burr hole and abdominal incisions should be selected and marked before draping and before



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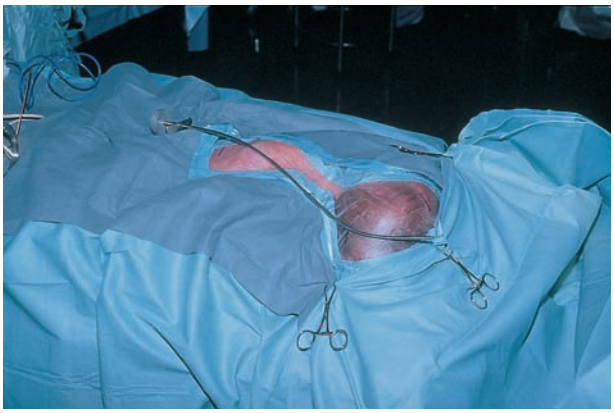

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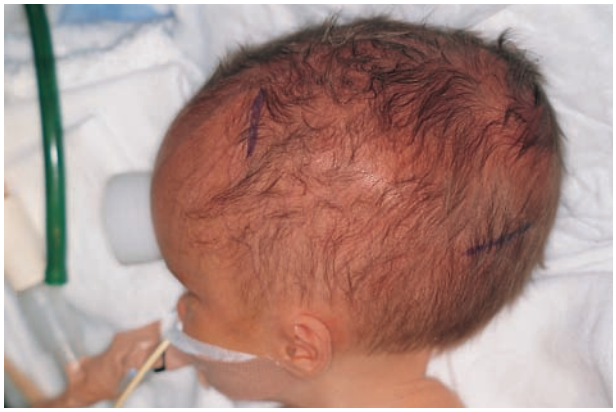

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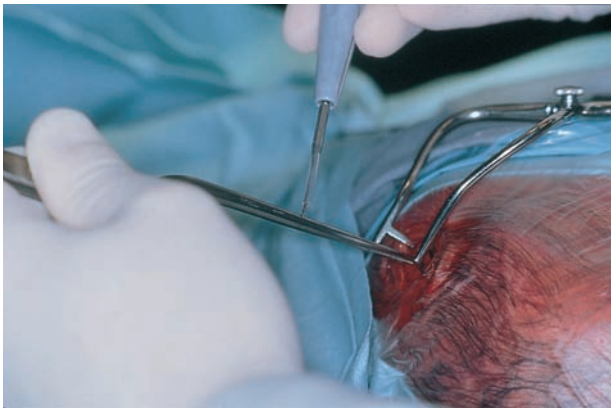

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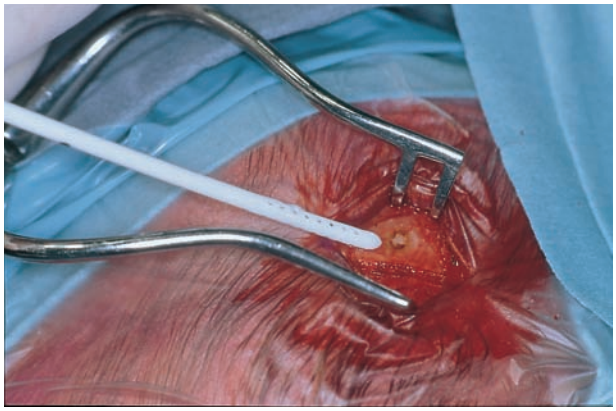

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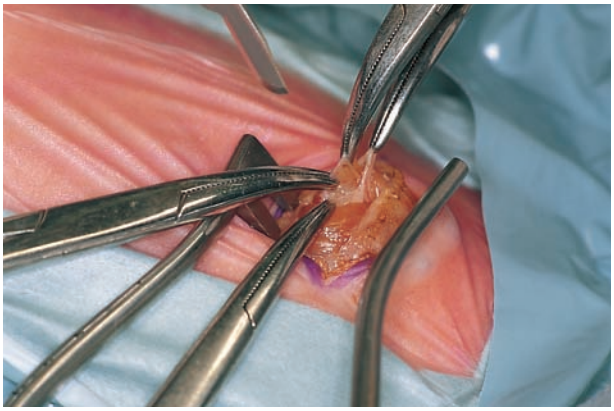

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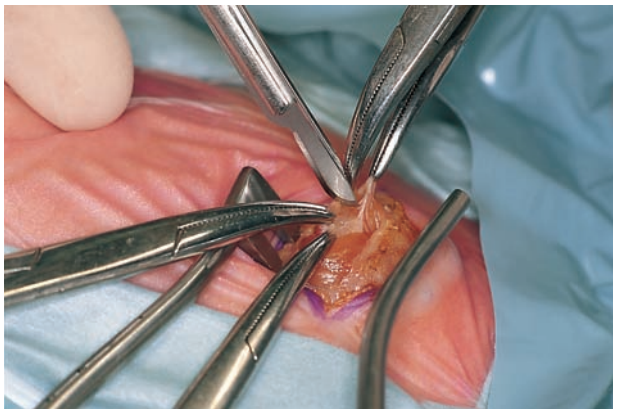
FIGURE 1–1. A: Patient positioning and marking of incisions for ventriculoperitoneal shunt. **B:** Draped patient with passer, illustrating planned subcutaneous path of the shunt. **C:** Proposed incisions for occipital or frontal shunt placement, with plastic syringe cap used for landmark. **D:** Monopolar cautery applied to bayoneted forceps. **E:** Result of **(D)**, a small dural opening for the ventricular catheter. Sequential steps in accessing the peritoneal cavity; **F:** Elevation of peritoneal layer.

the surface landmarks are obscured. Placement of a surface marker such as an electrocardiographic (ECG) electrode that can be palpated through the drapes may assist in placement of the ventricular catheter (Fig. 1–1C).

The issue of frontal versus occipital burr hole has never been resolved. We generally prefer an occipital shunt placement if the occipital horns are sufficiently enlarged because it allows insertion of the entire shunt system without an intervening neck incision. Usually, occipital burr holes are placed on the flat part of the occiput 3 to 4 cm from the midline along the course of the lambdoid suture (Fig. 1–2A). Patients with a Dandy-



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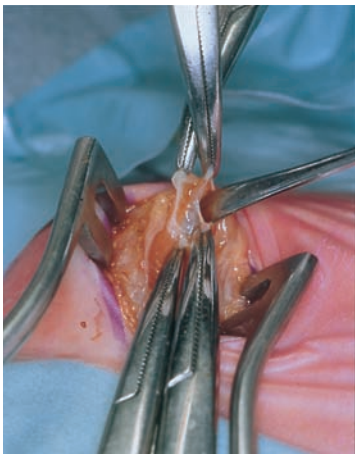

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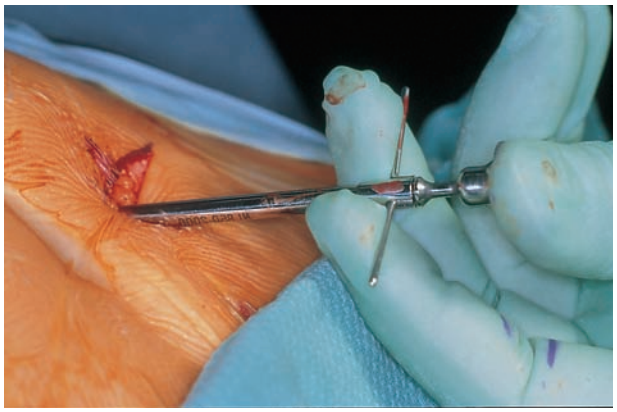

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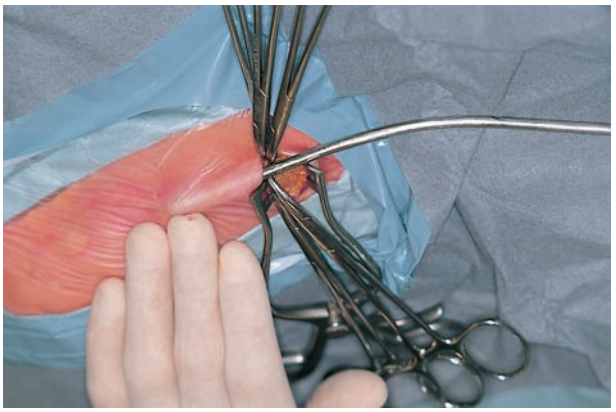

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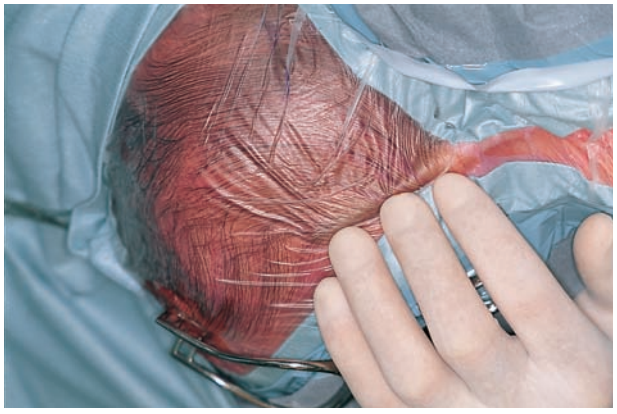

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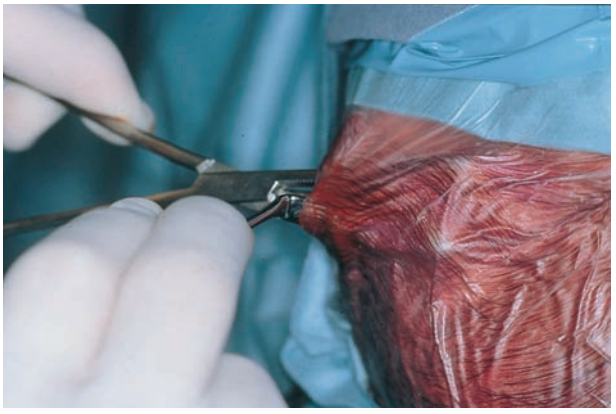


L

FIGURE 1–1. (***continued***) **G:** Sharp opening. **H:** Probing peritoneal cavity with blunt instrument to confirm entry. **I:** Alternative peritoneal access using an abdominal trochar. **J:** Insertion of tunneling instrument initially angled posteriorly to ease passage over the chest. **K:** Passer rotated to angle anteriorly in the neck to pass over occiput. **L:** Blunt dissection with a hemostat to create a subcutaneous "pocket" for the valve.

Walker malformation or large arachnoid cysts of the posterior fossa frequently have abnormally high transverse sinuses. These children should be evaluated preoperatively by MR imaging to guide appropriate modification of the burr-hole placement. Frontal burr holes generally are placed along the coronal suture 2 to 3 cm from the midline (Fig. 2–2B). Anterior burr-hole placement may be preferable when endoscopic catheter placement or other procedures (e.g., septostomy, cyst fenestration) are planned.

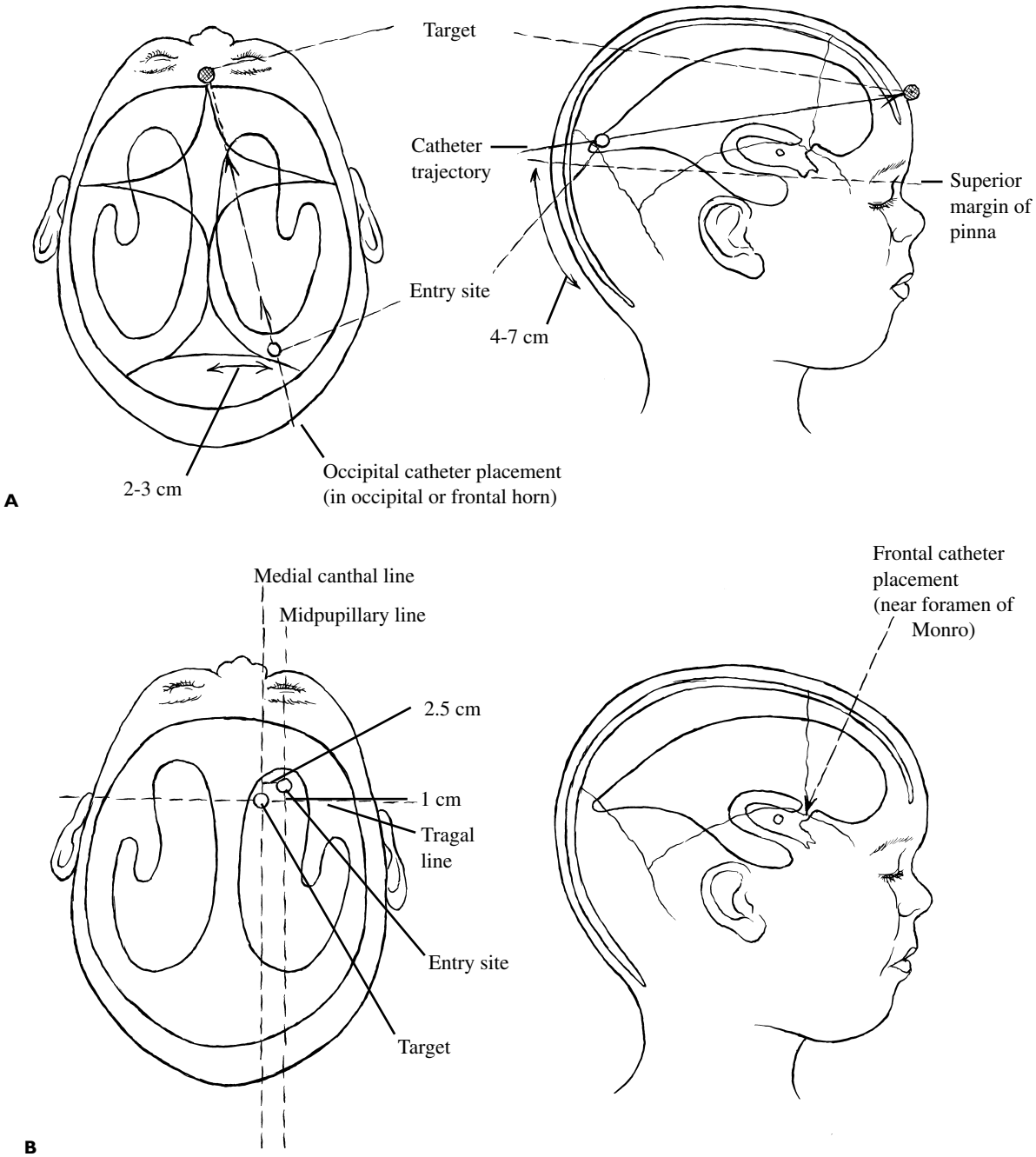


FIGURE 1–2. A: Standard landmarks for placement of an occipital ventricular catheter. The entry site is placed 4 to 7 cm above the inion and 2 to 3 cm off midline, just above the lambdoid suture. Alternatively, the lambdoid suture itself may be used in small infants. The insertion site should be above the superior margin of the pinna. The catheter is directed toward the midline at a point superior to the glabella (along the anterior hairline), which is marked preoperatively with an object that can be palpated through the drapes [i.e., syringe casing cap or electrocardiographic (ECG) sticker]. This trajectory generally prevents insertion into the temporal horn. Depth of insertion is determined from the preoperative imaging to place the catheter tip either in the frontal or occipital horn, avoiding placement in the atrium adjacent to the choroid. **B:** Standard landmarks for placement of a frontal ventricular catheter. The entry site is placed over the midpupillary line, generally 2.5 to 3 cm off the midline, 1 cm anterior to the coronal suture. The catheter is directed toward the nasion, along the intersection of lines through the medial canthus and the tragus. This trajectory is generally obtained by insertion at a right angle to the surface of the brain. The depth of insertion is determined from the preoperative imaging to place the catheter tip near the foramen of Monro.

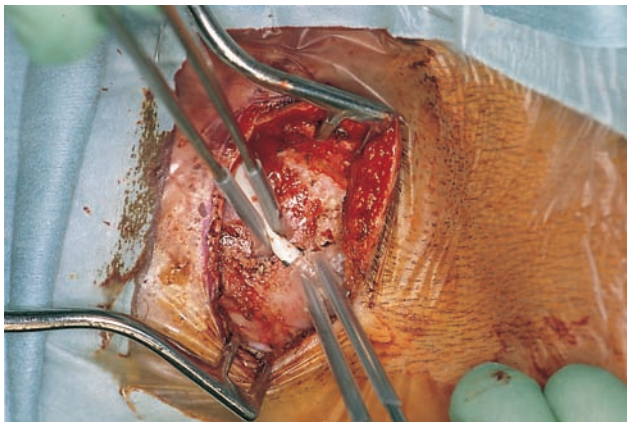
The skin is prepared meticulously with iodine- or chlorhexidine-based solution. Disposable, adhesive drapes are used to cover entirely the patient and the operating table, except for a small band of skin from the burr hole site to the abdomen (Fig. 1–1B). A transparent, adhesive sheet is applied to cover the remaining area of exposed skin. Small skin incisions are adequate and generally preferable (Fig. 1–1C). They are carefully planned to avoid placement over underlying hardware. The burr hole need not be a standard size, and a twist drill is adequate unless a burr hole reservoir or intraoperative ultrasound is being used. In infants, an opening between the splayed coronal or lambdoid sutures is all that is required. The dura should not be opened widely, particularly in patients with thinned cortical mantles, where a wide dural opening may allow CSF to escape around the ventricular catheter into the subcutaneous tissues, promoting a CSF leak and subsequent infection. Low-power monopolar coagulation applied to a small brain needle or bayoneted forceps will create a dural opening sufficiently large for the typical ventricular catheter (Figs. 1–1D and E). The pia matter is carefully cauterized and nicked with a fine-tipped bipolar forceps.

Simultaneously, an assistant can open the abdominal incision. Various methods (small laparotomy versus trocar insertion) and sites of insertion (midline versus lateral) are all acceptable, depending on the surgeon's preference and experience. Paraumbilical and upper midline sites are most common. During an open laparotomy, the peritoneum is elevated and sharply incised to avoid injuring abdominal organs (Figs. 1–1F and G). To verify that the peritoneum, and not just the preperitoneal space, has truly been opened, a blunt dissector is passed well into the abdominal cavity (Fig. 1–1H). A pursestring suture around the peritoneum tends to prevent the omentum from herniating but is not absolutely necessary. We prefer to use abdominal trocars in "virgin" bellies from a paraumbilical location. Opening the rectus sheath through a tiny incision and visualizing the posterior wall of the sheath facilitate placement. The posterior sheath is then "picked up" by the tip of the trocar, and the tip is angled inferiorly and off the midline to avoid hitting the great vessels (Fig. 1–1I). A gentle "pop" is felt as the peritoneum is penetrated. A blunt instrument also can be passed along the trochar sheath to verify peritoneal entry.

Care must be taken during subcutaneous tunneling of the shunt tubing. If the metal tube is too deep, the chest or even the posterior fossa can be entered. One must be particularly careful in patients who have had an occipital craniectomy because it is possible to pass the device into the bony defect accidentally. If the tunneling device is too superficial, a skin laceration, which may be unrecognized initially, can occur. A gentle curve to the tunneling instrument allows the tip to be directed posteriorly when coming over the anterior chest into the neck and then by rotating 180 degrees, angling the tip anteriorly toward the occiput (Figs. 1–1J and K). Usually, significant resistance is felt at the posterior nuchal line. Firm pressure, ensuring that the pointed central stylet has not backed out, and guarding against plunging, usually will allow this fascia to be penetrated. If excessive force is necessary, a separate incision in the neck is advisable. Likewise, passing to a frontal burr hole usually requires an intervening incision over the occiput. There appears to be no logical reason to tunnel down the back of the patient. Not only is this cumbersome, but over time it also results in the formation of a fibrous cordlike subcutaneous scar. The resulting "bowstring" is unsightly, limits neck mobility, and often persists even after the shunt hardware is removed. Insertion of the rigid tunneling device can compress the chest of a small child significantly. If the passer is left in place too long, the anesthetist may note an increase in airway pressure. This instrument also can tear the scalp, particularly in small infants, when trying to bring the straight tunneling tube around the curved skull.

The peritoneal catheter, with or without the attached valve, then is passed through the tunneling device, attaching suction to the distal end, and irrigating. Care is taken to avoid contact between the shunt and the patient's skin as well as the surgeon's gloves by using instruments and clean gauze to handle components, and covering incisions with additional sterile towels (Figs. 1–3A and B). The valve then should be attached and irrigated to fill it with fluid, usually the antibiotic solution soaking the shunt equipment. It is not necessary to test the opening pressure of the shunt in the operating room. Merely handling the valve will change its performance characteristics for days, and air bubbles also can affect these measurements. Care must be taken to ensure that the valve is inserted in the proper orientation and lies flat beneath the scalp.

The ventricular catheter trajectory is determined according to external landmarks. From a frontal burr hole, traditional landmarks for the foramen of Monro or the intersection of the planes through the medial canthus and the tragus (or simply being perpendicular to the skull) are used (Fig. 1–2B). From the occipital location, a target at the midpoint of the forehead, just at the normal hair line, will ensure that the catheter proceeds into the frontal horn instead of the temporal horn (Figs. 1–1C



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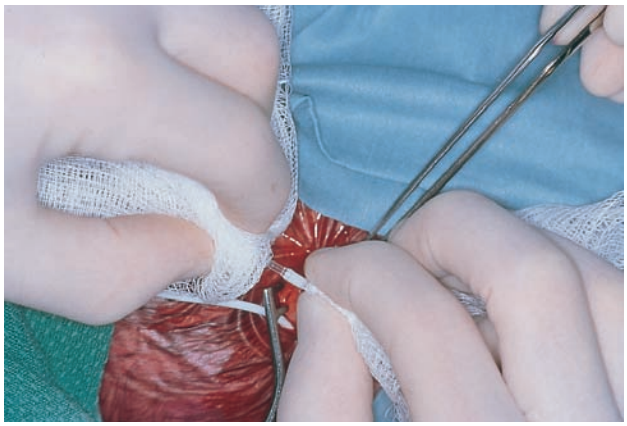


B

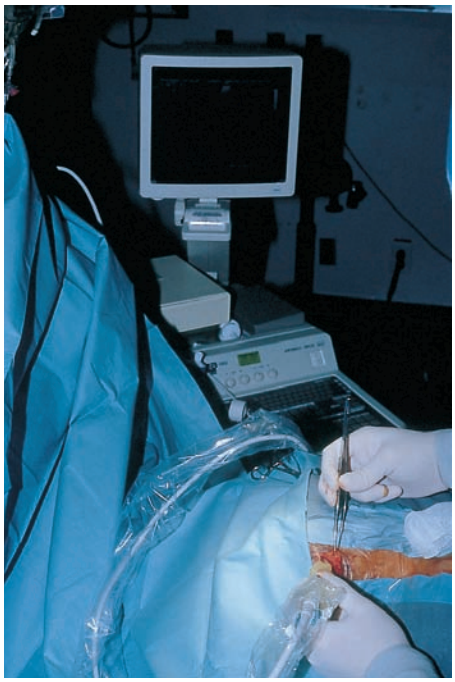
FIGURE 1–3. Shunt contact with the patient's skin or surgeon's gloves is avoided by the use of blunt-tipped or sleeved forceps **(A)** and clean gauze **(B)**.

and 1–2A). There is, however, no proven ideal location for the ventricular catheter. Recent evidence suggests that frontal or occipital locations are better than locations in the body of the ventricle or in the third ventricle. Cannulating small ventricles is easier from a frontal location. In these patients, either ultrasound, or even stereotaxis, may assist with successful ventricular cannulation. We routinely use ultrasound either through the shunt burr hole or in infants through the open fontanel (Fig. 1–4A). Fiberoptic endoscopic stylets are also available for shunt placement (Fig. 1–4B). The impact of endoscopy-assisted ventricular catheter placement is the subject of an ongoing randomized, prospective clinical trial. With these techniques, the surgeon is as certain as possible that the catheter is in good position at the end of the case, and not in one many "unusual" sites (e.g., sylvian fissure, quadrigeminal cistern).

The ventricular catheter usually can be felt to "pop" once the ependyma is breached, with a concomitant gush of CSF. If uncertainty exists, gently irrigating the catheter may show pulsatile CSF flow into and out of the catheter. Withdrawing vigourously will simply draw brain tissue into the catheter and obstruct an intraparenchymal shunt. Although there is no universally accepted limit on the number of passes, after two, we prefer to use ultrasound. A small amount of fresh blood that clears is not unusual and is one reason to recommend a separate ventricular catheter, allowing blood and debris to be cleared prior to attachment to the valve. Extensive hemorrhage should prompt copious irrigation until the CSF clears. Installing a narrow-orifice high-resistance valve in this setting will likely result in rapid occlusion.

A number of methods can be used to get the ventricular catheter to approximate a right angle at the burr hole. Simply bending the catheter, using the forces of the brain, burr hole and dura is often adequate, but the inherent stiffness of the catheter tends to move the tip in the opposite direction. In patients who have large ventricles and thin cortical mantles, the catheter can assume an almost straight trajectory, resulting in a final position against the ventricular wall or septum. Right-angled guides avoid this, but they are frequently awkward to use. Additionally, many of these guides are manufactured of hard plastic that will erode the scalp of small infants. When attaching burr-hole reservoirs (usually with contained valves), the ventricular catheter must be withdrawn and then readvanced. The attachment site is usually below the cortical surface, where it becomes adherent, and losing the catheter at a subsequent revision is quite possible.

When using a flat-bottomed valve, a pocket must be created along the distal path (Fig. 1–1L). This pocket must be exactly along the course of the catheter, or the valve will bind when attempting to slide it along, which can be troublesome, particularly if the ventricular catheter is already connected. When attaching the ventricular catheter to the valve, one should avoid using metal instruments directly on the tubing forcefully because they can lacerate the tubing, thereby causing subsequent leakage or disconnection. We use forceps or hemostats fitted with Silastic sleeves or clean gauze sponges to grasp the hardware (Figs. 1–3A and B). Similarly, when tying the catheter over the connector, it is vital to have the tie directly over the neck of the connector, tight enough not to allow spontaneous disconnection, or, alternatively, not too tight to lacerate the tubing. The valve system then is placed into its pocket by gently pulling on the peritoneal catheter from below. The shunt system then should be



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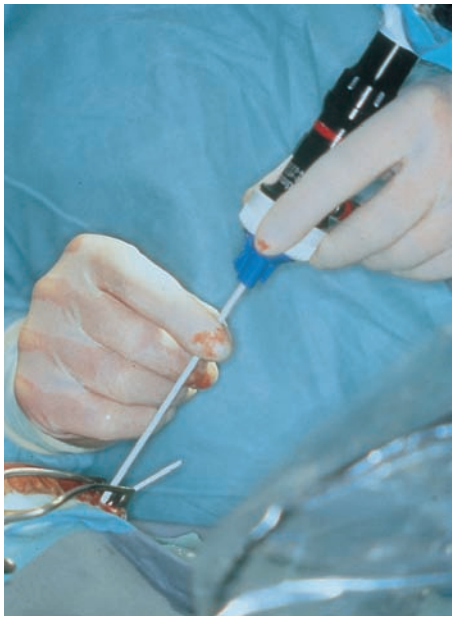

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FIGURE 1–4. Intraoperative aides for the placement of ventricular catheters include ultrasound **(A)** and fiberoptic endoscopy **(B)**.

secured to the pericranium to prevent subsequent migration. Posterior fossa catheters are particularly susceptible to migration and are difficult to secure. Three-way connectors in this site also seem to come under excessive stress with neck motion, frequently resulting in disconnection.

Once in place, the system should be checked to ensure that it is flowing, either spontaneously or with gentle pumping of the reservoir. If there is any doubt, the system should be disconnected to verify that both ends are patent. The distal catheter then is inserted into the peritoneal cavity. If resistance is encountered or the catheter is spontaneously backing out of the abdomen, it may be coiling up in the preperitoneal space. Once intraperitoneal placement is ensured, a pursestring peritoneal suture is placed, and the abdominal layers are reapproximated with absorbable 3–0 or 4–0 suture.

Skin closure is critical. The abdominal incision is closed in subcuticular fashion with a 4–0 or 5–0 undyed absorbable suture. Normally, the scalp incision is closed in two layers, a 3–0 or 4–0 absorbable galeal suture and a running nonabsorbable 3–0 or 4–0 monofilament, to achieve careful apposition of the skin edges. Any CSF leak predisposes to wound breakdown or infection. The fragile skin of premature infants may fray and permit CSF leakage through large needle holes.

Ventriculopleural Shunts

Pleural shunts are a second choice. Contraindications include previous chest surgery and adhesions, active pulmonary disease including infection, or patients with borderline pulmonary function in whom a significant pleural effusion might push them into respiratory failure. Infants are more likely to develop a significant effusion temporarily. The pleural space can be entered at a variety of sites. Along the anterior axillary line, in the fourth to sixth interspace (below the pectoralis muscles) is often a convenient site. A muscle-splitting approach along the upper border of a rib (to avoid the neurovascular bundle) will reveal the translucent pleura and the lung moving with ventilation.

The pleura is opened sharply, as with the peritoneum. There is no need to ask the anesthetist to collapse the lung; it will move away slightly as atmospheric pressure enters the chest cavity. The distal catheter then is introduced gently, with care taken to guide it along the chest wall, not into the lung parenchyma. The catheter may need to be cut to length to avoid putting excess tubing, even allowing for growth, into the chest. A pursestring suture is placed in the intercostal muscles and secured around the catheter while the anesthetist provides a Valsalva's maneuver to expel air and reinflate the lung adequately. Rapidly closing these muscles with a few sutures will avoid subsequent air entry into the chest.

A small pneumothorax will be seen on the mandatory postoperative film. It will resolve over the next few days; the CSF usually will accumulate as a small pleural effusion, especially in infants. These patients need to be monitored for any evidence of respiratory distress, and serial chest radiographs should be taken. Usually, the intrapleural fluid disappears over the next several weeks. In patients in whom the pleural fluid progressively accumulates, leading to respiratory distress with significant shift of the mediastinum, percutaneous drainage of the fluid and moving the distal tubing to another site are required.

Ventriculocardiac Shunts

Cardiac shunts are the third choice among the distal sites, due to the serious complications of cor pulmonale and shunt nephritis. Catheter embolization is also a possibility. As the child grows, these shunts tend to block as they pull out of the right atrium, often necessitating several revisions for growth-related failure.

The shunt tip should be placed in the right atrium just above the triscuspid valve. There are a number of ways of achieving this. Entrance to the jugular vein usually is achieved through the common facial vein, which is tied proximally, and held with a stay suture distal to the venotomy site. The catheter then is advanced down the jugular vein into the superior vena cava, which is much easier to do on the right side. Percutaneous methods into both the jugular and subclavian vein also have been described. Positioning the tip can be done under fluoroscopy or ultrasound. Fluoroscopy is useful because on occasion the catheter can be seen traveling out of the subclavian vein. Alternatively, one can use the tip of a saline-filled catheter as an ECG lead and look for a change in P-wave polarity when the right atrium is entered.

Shunt Revision

The surgery for shunt revision differs little from an initial shunt insertion, but a few points are important. Unless removal of the shunt because of an infection is planned, the patient should be prepared and draped as for a shunt insertion, including upper and lower incisions, even if one strongly suspects one or other end of the shunt, because these suspicions can be wrong.

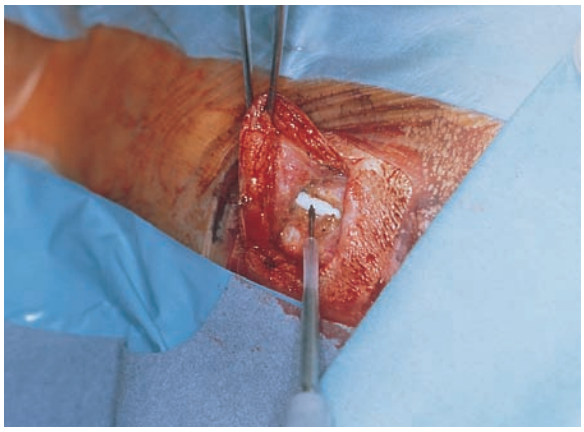
Normally, we explore the upper end of the shunt first because both ends can be tested from the same location, and piecemeal replacement of the lower end from an abdominal site using connectors is discouraged. Once the skin is incised, cutting cautery can be used to expose the shunt hardware easily and with minimal bleeding (Fig. 1–5A). Care must be taken with burr-hole reservoir systems, particularly where there is a tie on the ventricular catheter that resides below the pial surface. The proximal catheter may be quite adherent and is easily lost in the parenchyma if it separates from the valve. These lost catheters are extremely difficult to find. We recommend ventriculoscopic retrieval rather than searching blindly in the parenchyma. For this reason, we rarely recommend burr hole systems. The equipment then should be inspected carefully for signs of damage, CSF egress, or infection.

Disconnecting the ventricular catheter from the valve will allow patency of the upper end to be determined as well as the opportunity to take a CSF sample. Slow drops from the upper end often indicate an incomplete but clinically significant ventricular catheter obstruction, as evidenced by the "gush" of high-pressure CSF when the catheter is replaced. If there is any doubt, the catheter can be attached to a manometer or clear Silastic tubing to demonstrate CSF pulsations and ICP (Fig. 1–5B).

Connecting the clear Silastic tubing or manometer to the valve and watching for spontaneous drainage assesses distal catheter patency. The distal system may need to be irrigated, but if flow is poor, the lower end should be replaced. It is possible to reopen the same lower end incision and, using the cutting cautery, to expose the tubing within a fibrous tract. Stay sutures on the tract allow the tubing to be removed from the abdomen, and a new catheter can be passed down the same tract, avoiding a separate laparotomy.

If the upper end of the shunt is occluded, the ventricular catheter should be gently removed and a new catheter introduced in rapid sequence, being careful not to lose too much CSF because ventricles will collapse. Again, the standard landmarks are used; as with an initial shunt insertion, ultrasound, endoscopy, or stereotaxis can be used as aides. It is preferable to use the metal stylet to direct the catheter, although with unusually small ventricles, sliding the limp catheter down the old tract may suffice. One has no control over the trajectory of the catheter without the stylet, and astonishing catheter positions can result.

If the ventricular catheter is adherent, gently rotating the catheter may free it. Otherwise, the metal stylet can be advanced down the lumen to the tip and cautery applied to the metal stylet while rotating the ventricular catheter (Fig. 1–5C). Extremely adherent catheters probably should be left in place rather than produce a serious intraventricular hemorrhage. If hemorrhage does occur, manifested as frankly bloody CSF, the ventricle should be irrigated copi-



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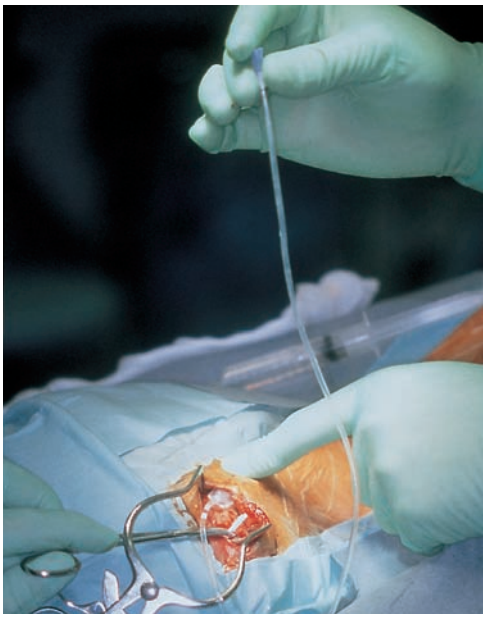

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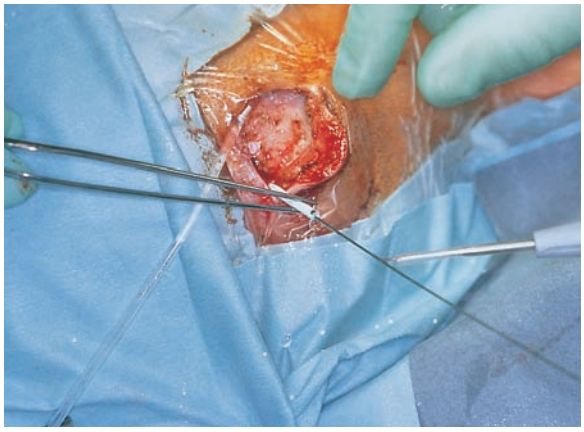

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FIGURE 1–5. Additional techniques for shunt revision. **A:** Monopolar cautery used to expose shunt hardware. **B:** Clear Silastic tubing used to assess intracranial pressure (ICP) and distal catheter patency. **C:** Monopolar cautery applied to catheter stylet to free adherent proximal catheter.

ously with warm irrigation fluid. Failure of the CSF to clear should prompt placement of an external ventricular drain and postponement of the shunt revision.

Lower-end obstruction is less common, and its cause always should be sought. Distal slit valves may accumulate debris, which forms a column inside the shunt, eventually blocking the slits. Unclogging the tip or just cutting it off may suffice. If the peritoneal catheter has fractured or is too short, we recommend replacing the entire aging system rather than piecing it together. The latter may result in further disruption in short order. Connectors should not be placed anywhere along the catheter distal to the skull in growing children because they adhere to surrounding tissues and eventually fracture. If the problem is the valve, or if one is changing a valve onto the same peritoneal catheter, care should be taken when pulling the peritoneal catheter up into the wound and then pushing it back down from above. To do so may cause the catheter to kink or coil out of the site, impeding shunt flow. It is preferable to expose the valve rather than to extract and reinsert it blindly.

POSTOPERATIVE MANAGEMENT

An occlusive dressing, which will resist attempts by small children to remove them, is recommended for 48 hours. Positioning in the postoperative period is also important. In patients with large ventricles, early mobilization may risk a subdural hemorrhage. Conversely, deliberately placing patients with high-resistance valves in an upright posture may promote CSF drainage and prevent subcutaneous collections.

The postoperative hospital stay is typically 2 or 3 days. Prophylactic antibiotics normally are given intravenously preoperatively and occasionally for a few doses postoperatively. Prolonged antibiotic treatment in the postoperative period in an uncomplicated shunt patient is unwarranted. Shunted patients typically have immediate resolution of acute symptoms. In infants, a sunken fontanelle with standard valves is typical. Low-pressure headache can occur in older patients, particularly if the hydrocephalus is long standing. In the absence of some particular problem intraoperatively, an initial postoperative CT or MR image is unlikely to be helpful. Normally, patients are seen in follow-up at approximately 3 months postoperatively for a CT or MR imaging. Because the ventricles usually require 1 year to attain their final size, follow-up imaging is usually obtained 1 year postoperatively as well.

EDITOR'S COMMENTARY

Although shunt operations are the most common neurosurgical operations in the U.S., they are frequently not given the meticulous attention to detail they require, and therefore have been traditionally associated with complication rates that frustrate both patient and neurosurgeon. This chapter contains many important principles to reduce those complications. Considerable effort should be given to position the ventricular catheter tip anterior to the foramen of Monro, away from the choroid plexus. It is important to remember that at least one-third of children will have no ventriculomegaly when their shunts malfunction; replacing ventricular catheters in these children may be facilitated by injecting air via a spinal needle through the old ventricular catheter to provide a ventriculographic target for the new catheter. Plural shunts may be inserted with the trocar used to insert peritoneal catheters. Atrial shunts are now often inserted via the percutaneous techniques used to insert central venous catheters. In the future, shunts may be impregnated with antibiotics to decrease infections and may be designed to prevent overdrainage, but their insertion and follow-up will still require meticulous attention.

PEARLS

In these authors' experience:

- When deciding whether hydrocephalus needs treatment, one should use ICP monitoring in difficult cases, and treat when there is a progressive problem. Repeated assessments over time may be necessary.
- When choosing shunt equipment, the surgeon should become familiar with one system and use it consistently. Realize that for first shunt insertions, current evidence does not favor any particular shunt design.
- When monitoring shunted patients, baseline images should be obtained at 3 and 12 months, and great caution should be exercised in concluding that a child's disconnected shunt does not need to be fixed.

SUGGESTED READINGS

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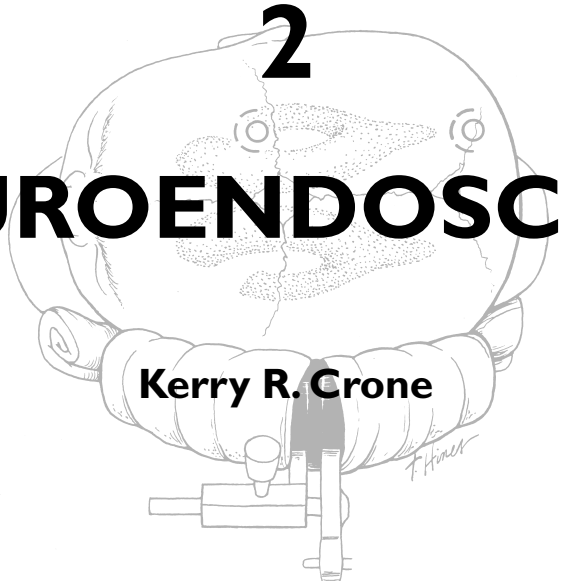
2

NEUROENDOSCOPY



Kerry R. Crone

OVERVIEW

Neuroendoscopy has emerged as a valuable technologic adjunct in the treatment of many neurosurgical conditions. Endoscopically guided shunt placement, membrane fenestration, and tumor management are the most common procedures performed using a neuroendoscope.

SURGICAL INDICATIONS

To date, most neuroendoscopic procedures involve the fenestration of intraventricular cysts or membranes and the biopsy or removal of intraventricular tumors. In most patients, obstructive hydrocephalus accompanies these conditions and produces symptoms of increased intracranial pressure, commonly including headache and vomiting, although accelerated head growth may be the only presenting symptom in neonates or infants.

Ultrasound or computed tomography (CT) usually is performed as a screening study to diagnose these conditions. Characteristic images demonstrate hydrocephalus alone or accompanied by intraventricular tumors or cystic membranes. Magnetic resonance imaging (MRI) should be performed prior to any neuroendoscopic procedure because it provides the best anatomic detail of the surrounding vasculature and adjacent brain structures. Preoperative contrast ventriculography should be performed in cases of complex hydrocephalus to evaluate communication between compartments within the ventricular system. No additional diagnostic studies are needed prior to neuroendoscopic surgery.

INSTRUMENTATION

Essential components for performing surgery include an endoscope, either rigid or flexible, light source, camera, viewing monitor, irrigation pump, and instrumentation for dissection and coagulation. Typically, the components are assembled on a wheeled cart, or they may be manufactured in a unitized system. Rigid endoscopes have traditionally been favored over fiberscopes because of their superior optics and multiple ports for irrigation and suction. Rigid endoscopes are difficult to maneuver through the ventricles when performing multiple fenestrations and require strategic targeting of the initial burr hole. Fiberscopes provide greater maneuverability with less strategic planning but compromise optical clarity and image quality. In many procedures, both types of endoscope may be used.

Although the instrumentation for performing endoscopic surgery is limited, specific instruments are available to perform biopsy, to dissect, and to coagulate tissue. Cup forceps are essential for tissue biopsy and for grasping foreign bodies within the ventricular system (Fig. 2–1A). Small balloon catheters are available to enlarge small fenestrations created in the septum pellucidum, the floor of the third ventricle, or walls of obstructing cysts (Fig. 2–1B). Fiberoptic lasers and monopolar electrodes may be used as energy sources to coagulate tumors or membranes and to dissect tissue. Both have been used successfully, but the higher cost of laser fibers and the concern about heat dissipation from the laser have favored the emergence of a monopolar electrode as the preferred device in neuroendoscopic surgery. Additional

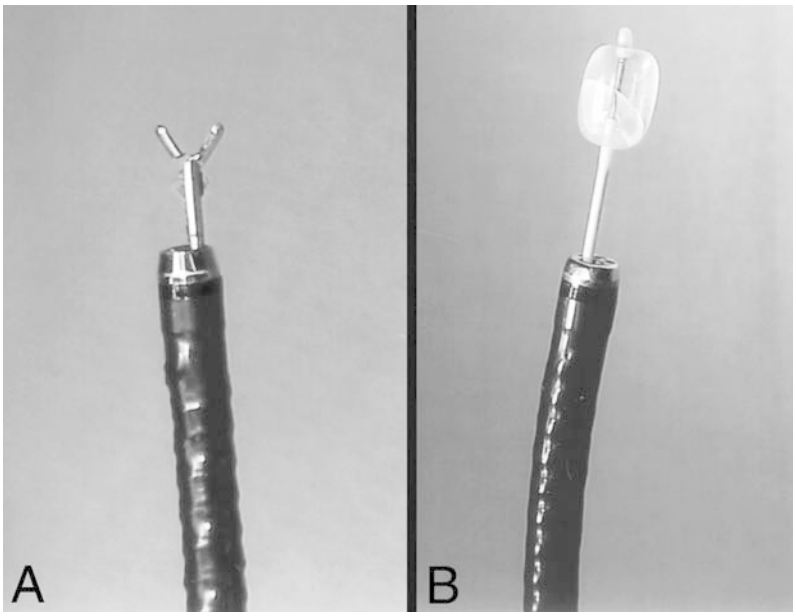


FIGURE 2–1. A: Cup forceps used for biopsy. **B:** Inflated balloon catheter used to enlarge fenestrations.

instrumentation consisting of scissors, probes, and various configured forceps are available, but they have limited use and are awkward to use. Bipolar devices have been manufactured, but they depart from the traditional design used in conventional neurosurgery, giving them limited benefit.

INTRAOPERATIVE TECHNIQUES

The most important factor in planning an endoscopic procedure is the location of the burr hole. For lesions in the anterior half of the ventricular system, the burr hole should be created anterior to the coronal suture. For lesions in the posterior half of the ventricular system or temporal horn, the burr hole should be positioned at the parietal eminence (Fig. 2–2). Navigational guidance systems may be necessary when the ventricles are small or when the targeted lesion requires a precise point for entering the ventricular system or cerebral parenchyma.

Patient position becomes important once the entry point has been selected. A supine position is optimal for lesions in the anterior half of the ventricles, whereas a prone position is necessary when entering the posterior part of the ventricular system or the posterior fossa. The head is usually placed in a cerebellar or soft doughnut headrest when the patient is positioned supine. If the patient is placed in the prone position, a cerebellar headrest is used. Three-point cranial fixation is rarely necessary unless an open craniotomy is planned and the endoscope is to be used as an adjunct surgical device.

Medications recommended for neuroendoscopic procedures include prophylactic antibiotics and intravenous steroids. Arterial lines, large-bore intravenous lines, and urinary catheters are seldom necessary in endoscopic procedures. Following routine intubation, a balanced anesthetic technique consisting of an inhala-

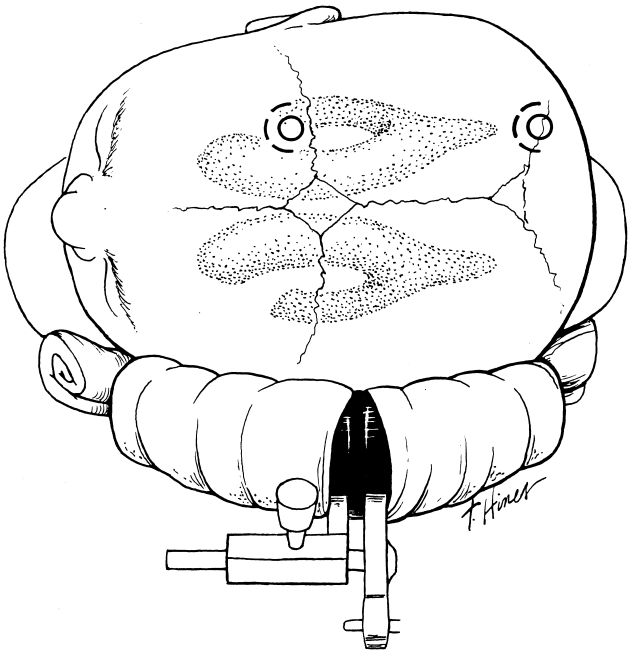


FIGURE 2–2. Patient position demonstrating entry sites for the coronal and parietooccipital approaches. (Printed with permission from Mayfield Clinic.)

tional agent and short-acting narcotic is used. The patient is mildly hyperventilated until the ventricle is cannulated. When the endoscope is introduced, homeothermic lactated Ringer's solution is used for irrigation. Ringer's solution is preferred over NaCl 0.9% because it is nearer the normal physiologic composition of cerebrospinal fluid.

Shunt Placement

Creation of a ventriculoperitoneal shunt is a common procedure in pediatric neurosurgery. There is considerable controversy regarding the optimal position for entering the ventricular system. My preference is to place the ventricular catheter from the frontal or coronal approach. A small pen neuroendoscope may be used for optimal positioning of the intraventricular catheter.

The patient is positioned supine on the operating table with a small roll placed beneath the shoulders to elevate the chest. The head is turned 90 degrees to the left side so that the nasal–occipital line is parallel to the floor. A small doughnut roll is placed beneath the head. If shaving the hair is the surgeon's preference, the right frontal region and a small area of the right parietal region may be shaved clean of hair for a frontal approach. A standard surgical preparation of the skin is used, followed by draping the field with sterile sheets. Skin incisions are infiltrated with a preparation of lidocaine and epinephrine. A small curvilinear incision is made in the right frontal region. The burr hole is created with a high-speed drill sufficiently large to accommodate the reservoir of the intraventricular catheter. An integrated valve with distal catheter is tunneled beneath the skin first to the parietal incision and then to the abdomen.

The dura mater is incised and the arachnoidal membrane is coagulated with bipolar cautery. The neuroendoscope is inserted into the proximal catheter and passed through the brain into the lateral ventricle (Fig. 2–3). The neuroendoscope then is passed through a precut slit in the ventricular catheter to view the ventricular system (Fig. 2–4). Normal ventricular landmarks should be identified for proper orientation. Once the surgeon feels familiar with the orientation, the catheter with the endoscope is moved in unison toward the foramen of Monro. Any abnormal findings should be noted and may include deposits of hemosiderin from a previous hemorrhage, thickening of the ependyma from previous infection, or thin transparent cystic lesions not identified on radiographic imaging. The association of these findings with shunt malfunction is being investigated. To date, patients with dense hemosiderin deposits appear to experience early malfunction, most likely related to free-floating fragments obstructing the proximal catheter or valve. Patients found to have membranes may demonstrate ventricular loculations. After the tip of the endoscope is navigated to the choroid plexus, the proximal catheter is deposited to rest just above this near-contact point. The endoscope is

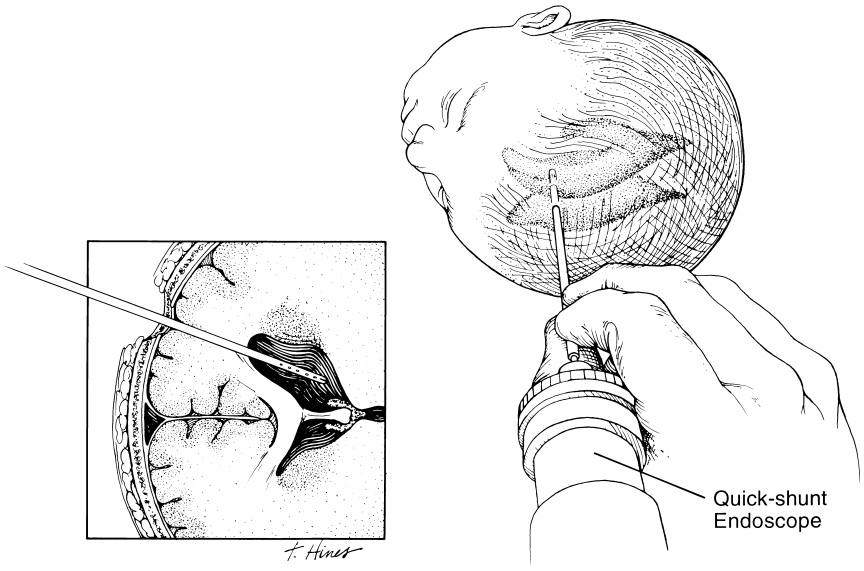


FIGURE 2–3. Ventricular catheter inserted into the lateral ventricle. The quick-shunt endoscope substitutes as a stylet for the ventricular catheter. *Inset:* Coronal view shows correct placement of the catheter and stylet. (Printed with permission from Mayfield Clinic.)

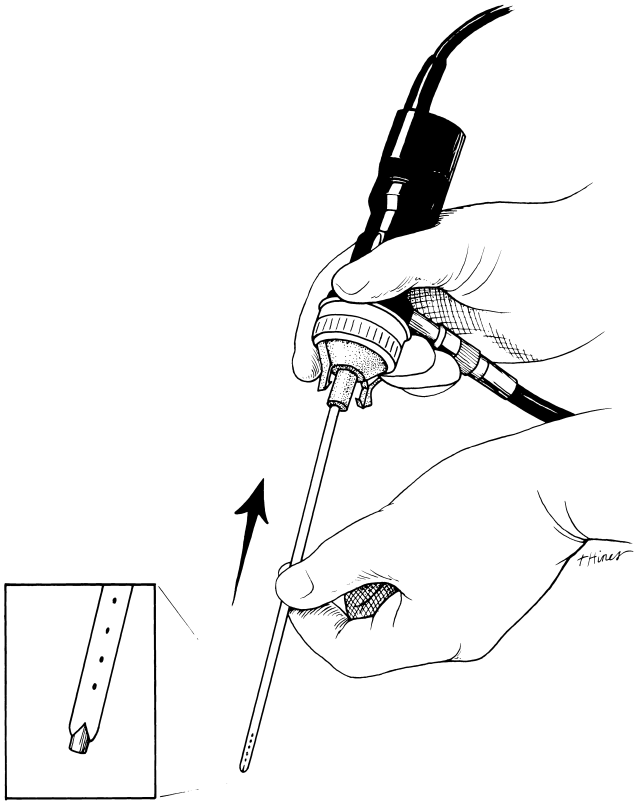


FIGURE 2–4. Tip of the endoscope protrudes minimally through the slit-ended ventricular catheter. (Printed with permission from Mayfield Clinic.)

withdrawn and the catheter is attached to the reservoir and valve with 3–0 silk ligatures. Cerebrospinal fluid flow is observed from the distal end of the shunt before the distal catheter is inserted into the abdominal cavity. After the reservoir and valve assembly are secured to the pericranium with 3–0 silk suture, the skin incisions are closed in an anatomic multiple-layer fashion.

Third Ventriculostomy

Endoscopic third ventriculostomy has attracted the most attention of all neuroendoscopic procedures. The ability to treat obstructive hydrocephalus without the insertion of a diversionary shunt is appealing to the surgeon, patient, and parents. Despite the current interest in third ventriculostomy, the long-term patency of the fenestration has not been clearly established. In 1923, Mixter performed the first endoscopic third ventriculostomy. Additional reports, including many recent publications, showed early high patency rates, but the long-term success of third ventriculostomy is uncertain.

Ideal candidates for third ventriculostomy are school-age children or young adults with late-onset hydrocephalus from aqueductal stenosis. Success rates in these groups have been reported to be as high as 85%. Less success is seen in obstructive hydrocephalus resulting from hemorrhage or infection, but neither condition is a contraindication to performing endoscopic third ventriculostomy.

It is essential to perform MRI in the preoperative planning for third ventriculostomy because the size of the third ventricle and massa intermedia and the position of the basilar artery are best demonstrated by MRI (Fig. 2–5). The third ventricle should measure at least 3 to 4 mm to accommodate the endoscope, and the floor should not be obstructed by an enlarged massa intermedia. The basilar artery lies beneath the mammillary bodies, leaving a corridor for the fenestration between these structures and the more anterior infundibular recess.

The patient is positioned supine on the operating room bed with the head in 0-degree rotation. Following the administration of general anesthesia, the scalp is prepared in the usual fashion and draped with sterile sheets. Following infiltration of the skin with lidocaine and epinephrine, a 2-cm linear incision is made 2.5 to 3.0 cm from the midline just anterior to the coronal suture. The coronal suture is identified, and a 1-cm diameter burr hole is created (Fig. 2–6). The dura is incised and coagu-

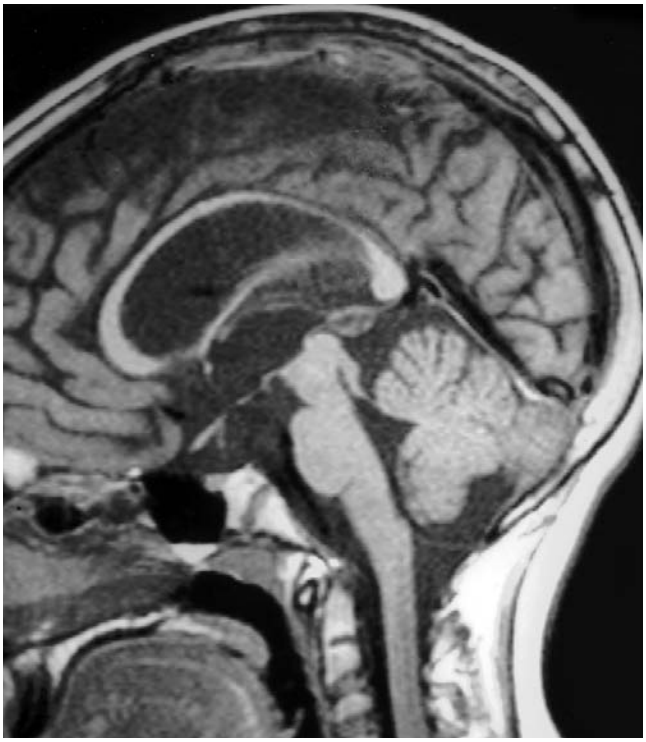


FIGURE 2–5. Preoperative sagittal MRI shows the basilar artery and the third ventricle. The third ventricle should measure at least 3 to 4 mm to accommodate the endoscope.

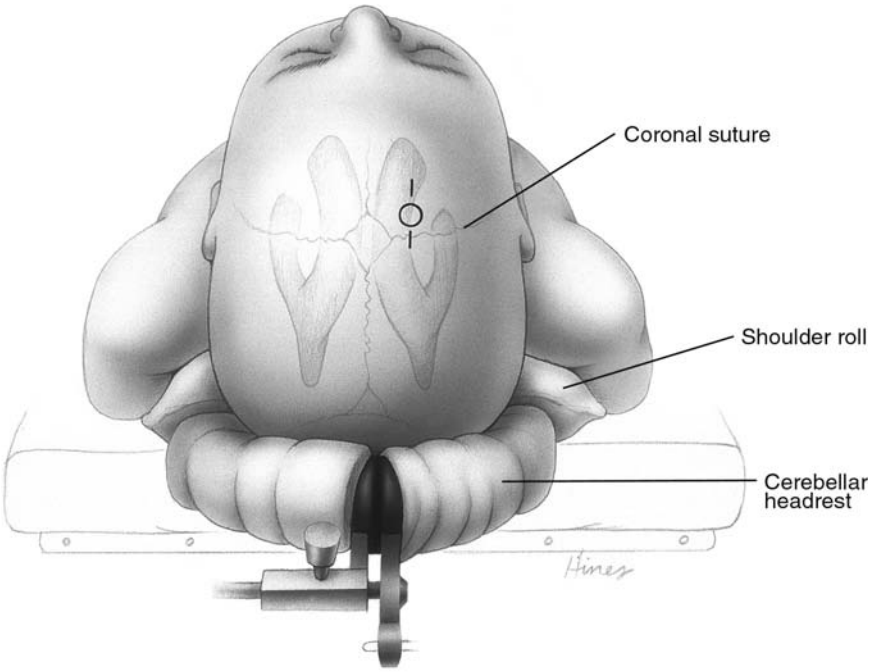


FIGURE 2–6. Patient is positioned supine on a horseshoe cerebellar headrest with a small roll placed under the shoulders to elevate the chest 10 to 15 degrees. A 3-cm vertical incision, based on the coronal suture, is made 2.5 cm from the midline. A 1-cm burr hole is created slightly anterior to the coronal suture. (Printed with permission from Mayfield Clinic.)

lated. A peel-away sheath introducer is guided into the ventricular system. The stylet is removed, and the endoscope is inserted through the sheath into the lateral ventricle. After the proper orientation is achieved, the lateral ventricle should be inspected and the foramen of Monro identified (Fig. 2–7). The endoscope is advanced into the third ventricle, and the floor of the third ventricle is inspected to identify the infundibular recess anteriorly and the mammillary bodies posteriorly (Fig. 2–8A). If a steerable fiberscope is used, the tip of the endoscope may be maneuvered posteriorly to inspect the aqueduct of Sylvius (Fig. 2–8B). A stenotic aqueduct will appear small compared with a normally sized aqueduct (Fig. 2–9A-B).

If the floor of the third ventricle is transparent, the tip of the basilar artery can be identified easily. The floor will be perforated anterior to the vascular complex. If the floor is translucent or opaque, the fenestration should be placed just posterior to the infundibular recess to reduce the risk of vascular injury (Fig's. 2–10A and B).

Several methods have been advocated to fenestrate the floor of the third ventricle. Blunt perforation may be achieved by bringing the endoscope into contact with the floor of the third ventricle and then gently applying constant pressure until the endoscope passes into the interpeduncular cistern. Alternatively, a monopolar electrode or thin laser fiber may be used to cauterize and perforate the floor. A balloon catheter then is inserted into the fenestration and inflated to widen the opening (Fig. 2–11). Once the surgeon is satisfied with the fenestration, the endoscope is slowly withdrawn, and the entry tract is inspected for any site of hemorrhage. It is unnecessary to insert an external ventricular drain. The burr hole is covered with a thin titanium plate, and the skin is closed in layers.

The patient is observed for 24 hours following surgery and then discharged home. A cine MRI scan is

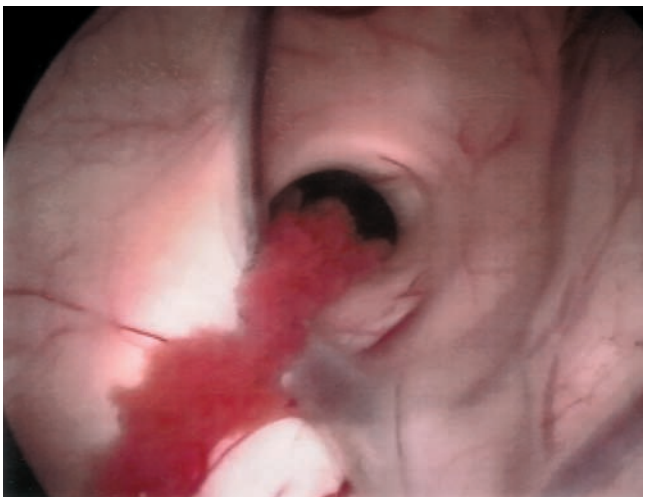
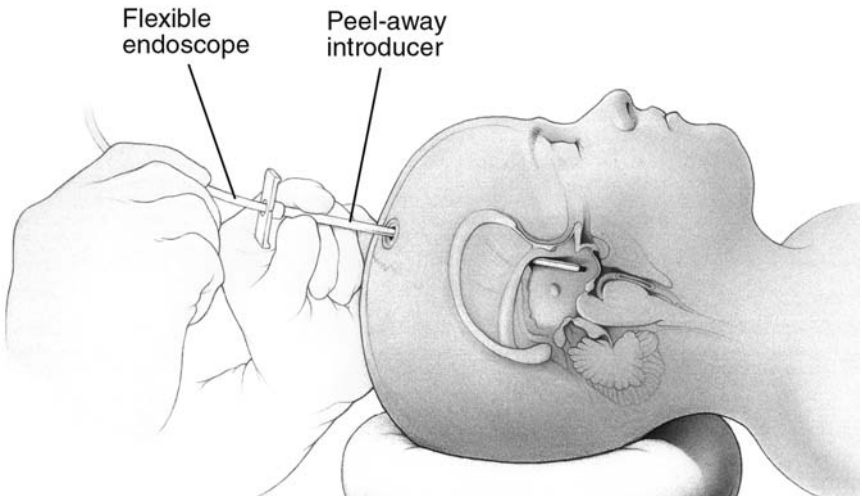
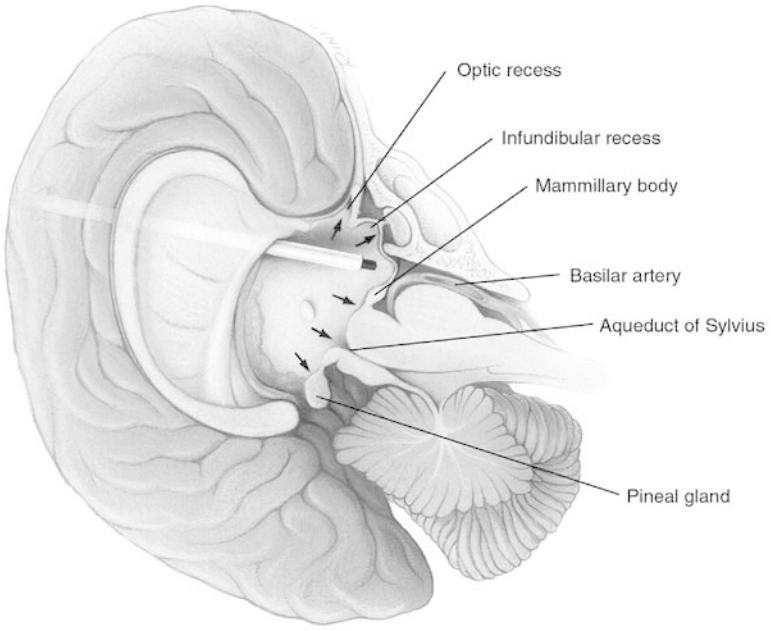


FIGURE 2–7. Endoscopic view (right) of foramen of Monro, partially obscured by the choroid plexus.



A



B

FIGURE 2–8. **A:** The dura is incised and coagulated to permit entry of the introducer. A 12.5-French peel-away sheath introducer is used to cannulate the lateral ventricle. The flexible endoscope is inserted through the cannula and guided through the foramen of Monro to enter the third ventricle. (Printed with permission from Mayfield Clinic.) **B:** The tip of the flexible endoscope can be moved to visualize the optic recess, infundibular recess, mammillary bodies, aqueduct of Sylvius, suprapineal recess, and, in rare instances, the roof of the third ventricle. (Printed with permission from Mayfield Clinic.)

obtained in 4 to 6 weeks to access flow through the fenestration (Fig. 2–12).

Compartmentalized Hydrocephalus

The presence of cystic areas or septations within the ventricular system may arise from previous hemorrhage, infection, or trauma. The placement of a conventional shunt system to treat this condition often resulted in loculated areas that do not communicate with the ventricle. Prior to neuroendoscopic surgery, treatment often required multiple shunt catheters joined together in a complex fashion to drain each compartment. Alternatively, blind fenestration of the loculated area has been advocated. Imprecise methods and techniques often failed to penetrate the cyst wall. Application of neuroendoscopy to manage this condition results in a reduction of shunt revisions and simplification of the complexity of the shunt system.

Management of compartmentalized hydrocephalus requires the most strategic preoperative planning of any

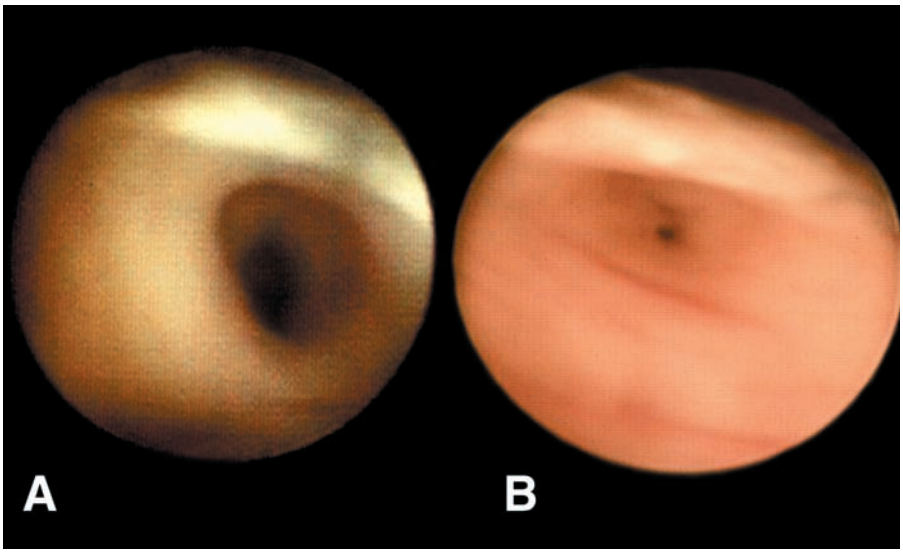
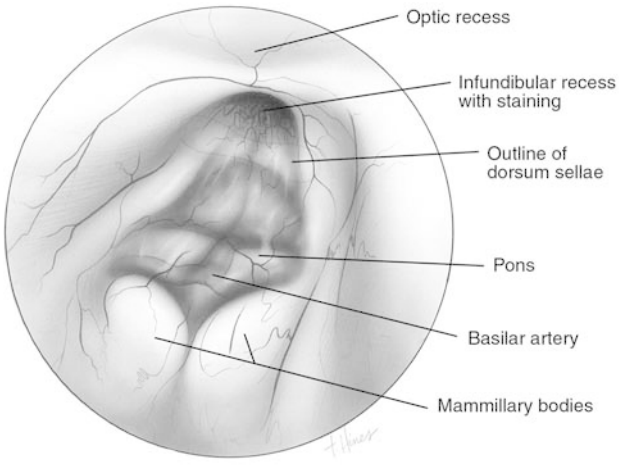


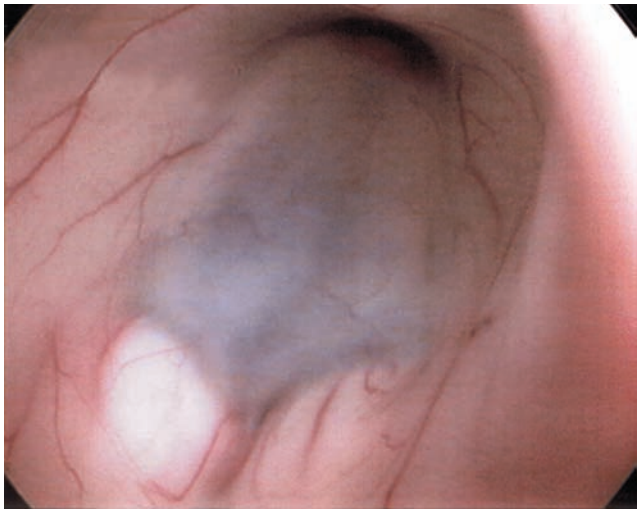
FIGURE 2–9. Normal **(A)** and a stenotic **(B)** aqueduct of Sylvius.

neuroendoscopic procedure. The anatomy of the compartments should be delineated before performing the operative procedure. An MRI should be obtained to identify vascular structures adjacent to cystic membranes or septations. A CT ventriculogram also may be needed to identify the loculations and to assist in selecting the burr hole (Fig. 2–13). Once the anatomy has been identified, consideration of the following principles will guide the surgical approach. In the treatment of a unilaterally dilated isolated ventricle, the burr hole should be created to cannulate the smallest ventricle. It is easier to work from the smallest to the largest ventricle, and potential damage to adjacent cortical structures is diminished. If multiple loculations are present, the largest loculation is chosen because it will permit the greatest maneuverability within the ventricular system.

Patient position is determined by the location and extent of the loculations. In most situations, the patient



A



B

FIGURE 2–10. **A:** Floor of the third ventricle. **B:** Floor of the third ventricle attenuated between the infundibular recess anteriorly and the mammillary bodies posteriorly. Typically, the infundibular recess is stained with a multitude of small blood vessels. Structures that may be visible beneath the attenuated floor include the clivus, dorsum sellae, pons, and the basilar artery. (Printed with permission from Mayfield Clinic.)

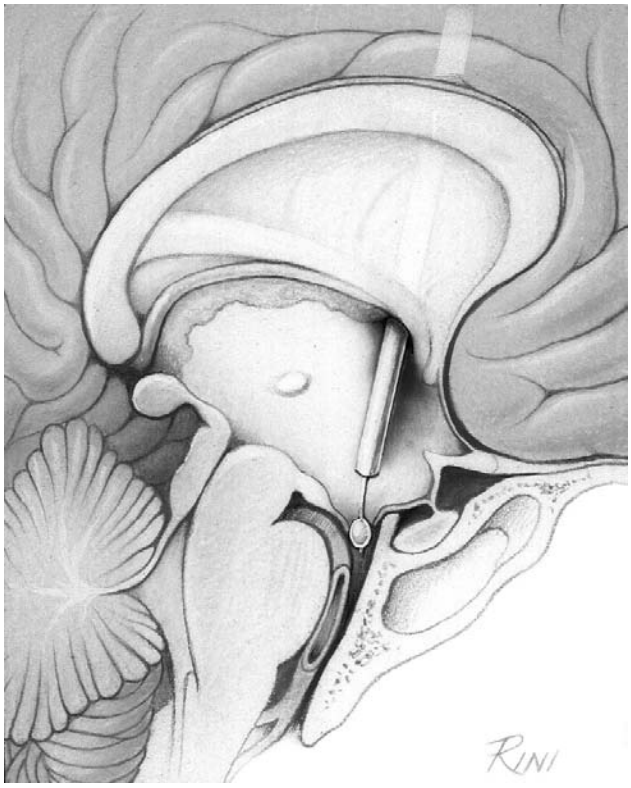


FIGURE 2–11. Small Fogarty balloon may be advanced through the endoscope and should be inflated when the epicenter of the balloon is aligned with the fenestration. (Printed with permission from Mayfield Clinic.)

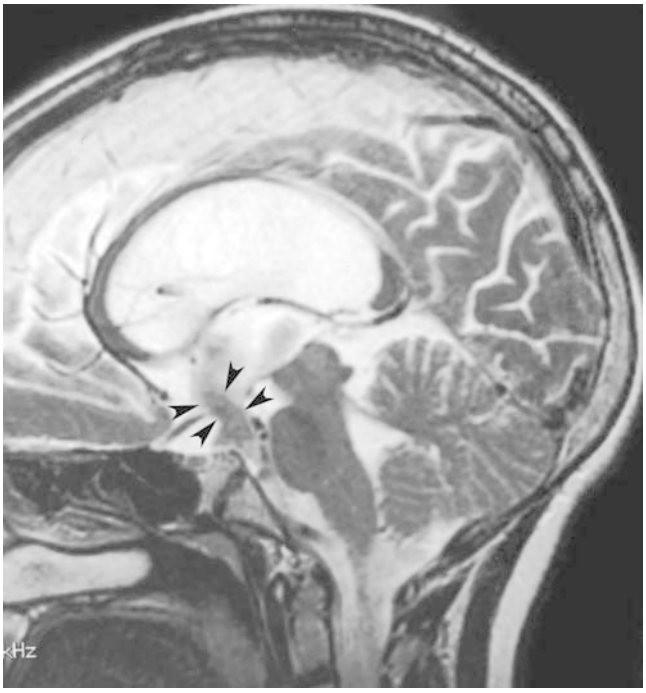


FIGURE 2–12. Phase-contrast cine MRI scan demonstrating flow jet through the fenestration (*arrowheads*).

will be positioned supine or prone on the operating room table with the head supported by a cerebellar headrest. A strip shave of the hair is performed at the site for the burr hole(s). If intraoperative ultrasound is used, the anterior fontanelle or site for a small trephine must also be prepared and draped into the sterile field. Either a rigid or flexible endoscope may be used for the procedure, but if multiple loculations are present, the flexible endoscope provides greater maneuverability to navigate through the ventricular system and loculations.

With the patient properly positioned and the head prepared and draped in a sterile fashion, a burr hole with a diameter of 12 mm is created. The dura is incised and the pia–arachnoid coagulated with bipolar cautery. A peel-away sheath with introducer is inserted into the ventricular system or largest loculation. A frameless navigational system or ultrasound may be necessary to assist in guiding the introducer into the proper location. The stylet is withdrawn from the introducer and the endoscope is inserted into the ventricle or loculated space. If the ventricle is cannulated, specific landmarks such as the choroid plexus should be identified for orientation, which will assist in navigating the endoscope. The endoscope is directed into the appropriate location with the cyst membrane or septation identified.

The membrane may be translucent or opaque, thin or thick, with a variable degree of vascularity. If uncertainty exists in differentiating a thickened opaque membrane from a cortical surface, gentle irrigation may cause the membrane to flutter or move, whereas a cortical surface should remain unaffected. The endoscope should be moved near the membrane and the monopolar electrode or laser fiber advanced through the scope to contact the membrane. The membrane should be incised with cutting current or a low-wattage laser in continuous pulse mode. The tip of the endoscope or a balloon catheter may be used to widen the fenestration, which should be at least 1 cm in diameter. Blood vessels within the cyst wall are coagulated to devascularize the cyst. This procedure also effectively widens the fenestration by constricting the cyst walls.

When multiple cysts are present, ultrasound may be useful in navigating the endoscope through the ventricular system. Although frameless guidance systems may assist in targeting the initial approach to the ventricle, this should not be used after the loculation has been fenestrated because significant brain shift may occur, ren-

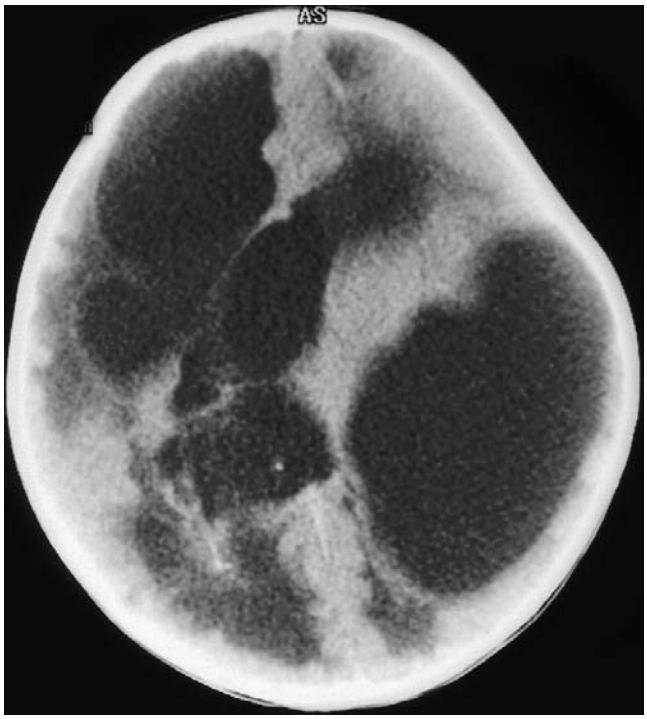


FIGURE 2–13. CT ventriculogram depicting multiloculated hydrocephalus.

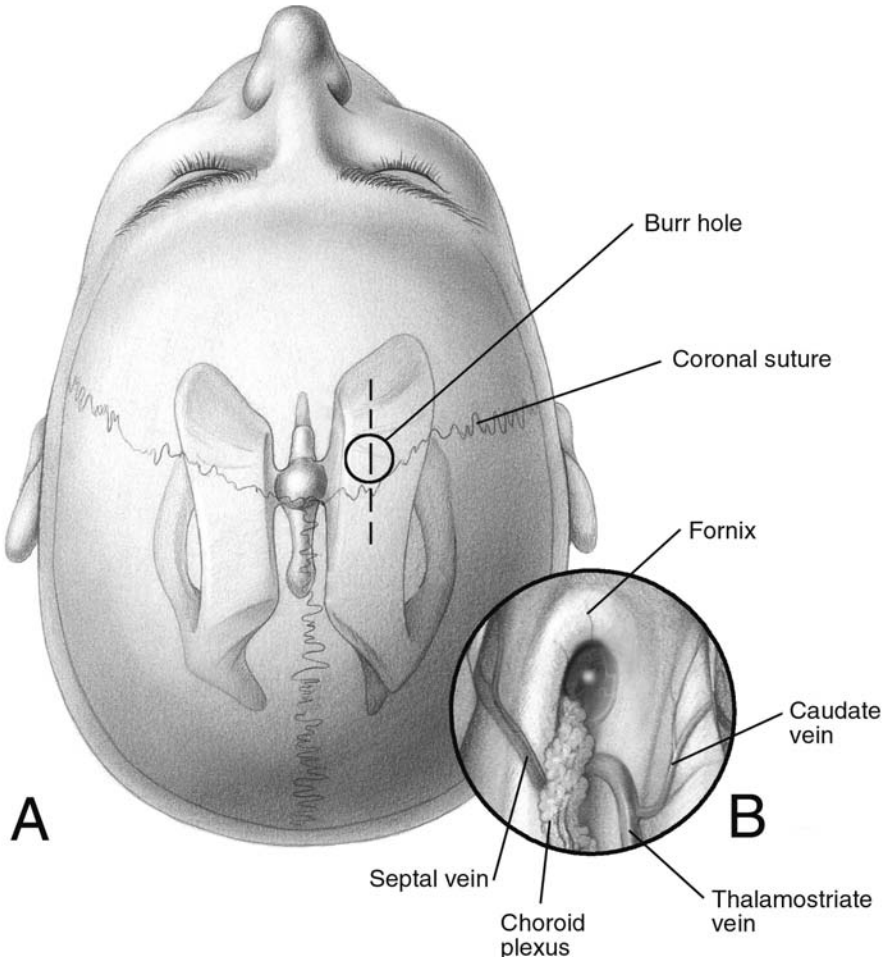
dering previous coordinates inaccurate. An external ventricular drainage catheter should be left in the ventricle if bleeding has occurred during the procedure. This catheter allows drainage of blood products and assists in monitoring any additional bleeding. The catheter may be accessed the following day to perform a postoperative ventriculogram to determine the success of the fenestration. Closure of the burr hole and skin is similar to that used in an endoscopic third ventriculostomy.

Tumor Management

Intraventricular tumors can be safely biopsied or excised on the basis of their size, vascularity, and location. Most tumors can be approached through a frontal or occipital burr hole. Tumors larger than 3 cm in diameter are best managed through conventional surgical techniques because endoscopic devices for rapidly debulking larger tumors either have not been developed or are not adequately tested to determine their utility. The risk for intraventricular hemorrhage is increased for vascular tumors so that preparation for open craniotomy should be made before attempting endoscopic removal.

Tumors in the anterior portion of the third ventricle and frontal horn to the midbody of the lateral ventricle are best approached from a burr hole placed at least 5 to 6 cm from the midline just anterior to the coronal suture. Lesions in the posterior third ventricle or midbody of the lateral ventricle are best approached from a more anteriorly placed burr hole, 4 cm from the midline near the hairline or 10 cm superior to the orbital ridge. Tumors posterior to the midbody or in the atrium of the lateral ventricle or temporal horn are approached from a burr hole created 4 cm from the midline and 6–8 cm superior to the nuchal ridge. The entry site may be altered if frameless navigational systems are used to target the entry point precisely.

The following information describes the technique for removing a colloid cyst from the third ventricle. Following the administration of general endotracheal anesthesia, the patient is positioned supine on the operating room table with the head placed in a cerebellar headrest. A small roll is placed beneath the shoulders to elevate the chest approximately 10 degrees. A strip shave of the hair is performed, and the head is prepared and draped in a standard fashion. The skin is incised, and a burr hole is created 6 cm from the midline and 2 cm anterior to the coronal suture (Fig. 2–14A). The dura and pia–arachnoid are incised and coagulated. A peel-away sheath introducer is inserted into the ventricle. The stylet then is removed, and the endoscope is inserted. A rigid endoscope is best suited for tumor removal because of multiple working channels and its superior optics. After the endoscope is passed into the ventricle, the colloid cyst (green) is usually seen filling the foramen of Monro (Fig. 2–14B). Occasionally, the cyst may be hidden by the choroid plexus covering its surface (Fig. 2–14C). A monopolar electrode or thin-laser fiber is passed through one working channel and is brought into contact with the cyst wall. Frequent bursts of electrocautery or laser energy are delivered until an opening is made in the cyst. Colloid material may erupt spontaneously from the cyst if the material is gelatinous. Continuous irrigation should be used with either energy source to dissipate heat within the ventricular system. A small-suction catheter introduced through the working port may be used to remove additional colloid material, whereas solid material is removed with small-cup forceps. The entire capsule then is coagulated. Before the endoscope is withdrawn, the third ventricle and aqueduct are inspected to ensure that there is free flow of cerebrospinal fluid. Solid tumors are managed in a similar fashion, with the exception that any surface vessels are coagulated prior to debulking the tumor.



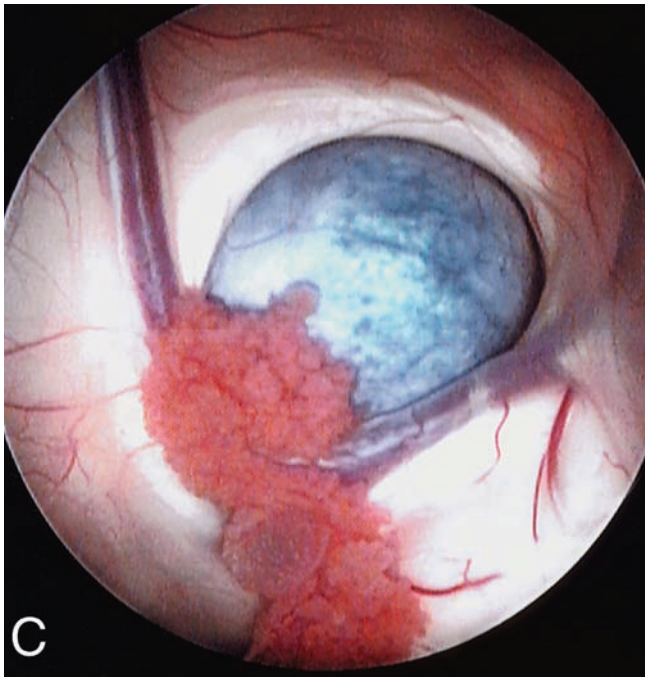


FIGURE 2–14. A: Endoscopic approach for colloid cysts. A vertical skin incision is made and a burr hole created 6 cm from the midline and 2 cm anterior to the coronal suture. (Reprinted with permission from the *Atlas of Operative Microneurosurgery,* Volume 2, in press.) **B:** Endoscopic view depicting a colloid cyst filling the foramen of Monro. **C:** Colloid cyst partially obscured by the choroid plexus.

COMPLICATIONS

The potential for hemorrhage, cerebrospinal fluid leak, or infection is similar to that with conventional neurosurgical procedures. Hemorrhage poses the greatest risk to the patient. Access to the source of bleeding is limited by a burr hole, and endoscopic instruments are not available to manage brisk hemorrhage. Continuous irrigation is successful in stopping most bleeding, but on rare occasion an open craniotomy may be necessary.

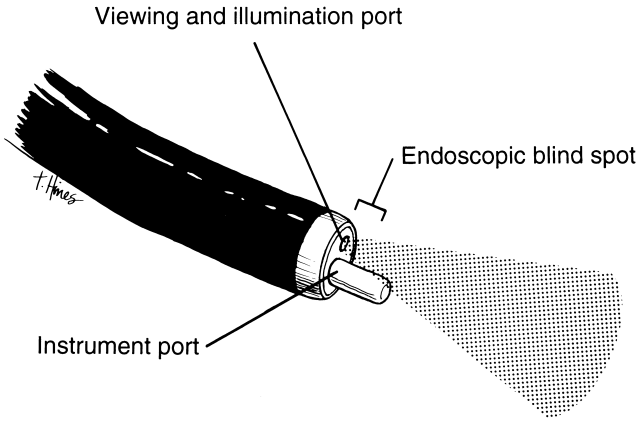


FIGURE 2–15. "Endoscopic blind spot," which occurs when instruments may project several millimeters beyond the end of the scope before they are seen by the camera, resulting in potential damage or hemorrhage of critical structures. (Printed with permission from Mayfield Clinic.)

Trauma to normal brain tissue may result from endoscopic instruments or thermal damage from energy sources. Extreme caution should be taken when instruments are inserted through the endoscope. The instruments may project several millimeters beyond the end of the scope before they are visualized on the monitor. Instruments projected into this "endoscopic blind spot" may damage adjacent brain tissue, resulting in hemorrhage or neurologic disability (Fig. 2–15). Thermal energy from a laser or monopolar electrode must be dissipated into the surrounding cerebrospinal fluid. Irrigation with warmed lactated Ringer's solution is satisfactory to prevent this thermal energy from causing infarction of adjacent tissue. Care must be taken to avoid accumulation of excessive fluid within the ventricle as intracranial pressure could increase, resulting in bradycardia, neurologic damage, or death.

SUMMARY

Endoscopy has benefited neurologic surgery greatly. Neuroendoscopy provides the surgeon with a safe, effective means of treating obstructive hydrocephalus by performing a third ventriculostomy, removing intraventricular tumors, and fenestrating intraventricular cysts. Additional therapeutic applications for neuroendoscopy will likely emerge with greater technical advancements. To date, many articles have reported the advantages of minimally invasive surgery in reducing operating times and hospitalizations.

EDITOR'S COMMENTARY

In the past decade, neuroendoscopy has become an essential tool in the treatment of hydrocephalus, cysts and intraventricular masses. There is a learning curve with these techinques, a curve that begins with patient selection, continues into decisions as to which membranes can be safely fenestrated, and includes decisions about when these procedures should be aborted because of bleeding or inadequate visualization. As Crone points out, if you cannot see, endoscopy is difficult (and risky). In this editor's (ALA) experience, the use of endoscopes within ventricular catheters has been of little help in positioning them optimally; endocsopes are not needed for big ventricles and in small ventricles the catheter's position within the ventricle is not easily adjusted after the anatomy is visualized. In performing endoscopic third ventriculostomies, I strongly favor perforation with a monopolar cautery, having encountered serious arterial bleeding on one occasion when the ventricular floor was perforated with the endoscope itself. Neuroendoscopy clearly has the potential to avoid many shunts and to minimize surgical trauma. It is accompanied by restricted ability to suction and coagulate, but if used with great care, its potential substantially outweighs its risks.

PEARLS

In this author's experience:

- Knowledge of normal endoscopic intraventricular anatomy is essential before attempting complex interventional endoscopic procedures.
- Vision can be obscured easily by any amount of bleeding; therefore, the risk of bleeding must be minimized.
- Irrigation is essential in endoscopic procedures to clear the lens of any blood or debris and to minimize the heat generated from energy delivery systems. The volume of irrigation that is used must be monitored closely to prevent the development of elevated intracranial pressure.
- The surgeon should consider aborting the procedure if unfamiliar anatomy is encountered or if the endoscopic system malfunctions and a suitable replacement is not available.

SUGGESTED READINGS

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3

THE CHIARI MALFORMATIONS

W. Jerry Oakes

CHIARI I MALFORMATIONS

The Chiari I malformation (CIM) is the most subtle form of hindbrain herniation. It is generally not associated with congenital neural-tube defects and consists of caudal migration of the cerebellar tonsils through the foramen magnum a variable distance. When cerebrospinal fluid (CSF) movement is severely restricted, it commonly causes syringomyelia.

With the introduction of magnetic resonance (MR) imaging and the increased ease of visualization of the craniocervical junction, our ability to diagnose this condition has increased exponentially. At the same time, the safety of craniocervical decompression has increased, making operation a much more attractive option. The most difficult problems facing the clinician currently are what symptoms justify operation and defining the line between a truly pathological state and an innocent anatomic variant.

Surgical Indications

Surgery for a patient with a CIM (Fig. 3–1) should be considered if the patient is symptomatic from spinal cord or medullary compression, if there is an associated syrinx, or if significant pain is thought to be produced from the lesion.

Pain is currently the most common presenting symptom of a patient with a CIM. It is most easily appreciated when the patient is old enough to communicate effectively and consistently complains of sharp pain in the craniocervical junction or occiput induced by activities that cause abrupt increases in intracranial pressure (ICP). Children who are noted to begin laughing and then suddenly to grab the back of their neck in pain are diagnosed easily. Initiating activities may include coughing, vomiting, bowel movements, strenous physical exercise, or anything associated with a Valsalva maneuver. Less clear are cases of patients who have life-altering dull pain in the occipital region without any initiating factors; however, even within this group, I have seen dramatic improvements following surgical decompression.

Symptoms specific to the compressive aspect of the problem are many and range from life-threatening alterations of basic functions (e.g., bradycardia, apnea), to motor disturbances (e.g., myelopathy), to cranial nerve signs (e.g., dysartharia, dysphagia, strabismus). This is the least common form of presentation today, but its life-threatening nature demands our attention.

When present, the simplest to appreciate and most objective indication for surgery is the presence of an appropriate syrinx. Although there are numerous other causes of syringomyelia, hindbrain herniation is the most common. The syrinx usually is located in the cervical or upper thoracic region. The longer the skip area between the craniocervical junction and the syrinx, the more uncomfortable the clinican should become to attribute the cavity to the hindbrain CSF dynamic disturbance.

Occasional patients will have a syrinx without an obvious cause (trauma, neoplasm, arachnoiditis) and have unimpressive caudal tonsillar descent. Even in this group, craniocervical decompression has been effective in re-

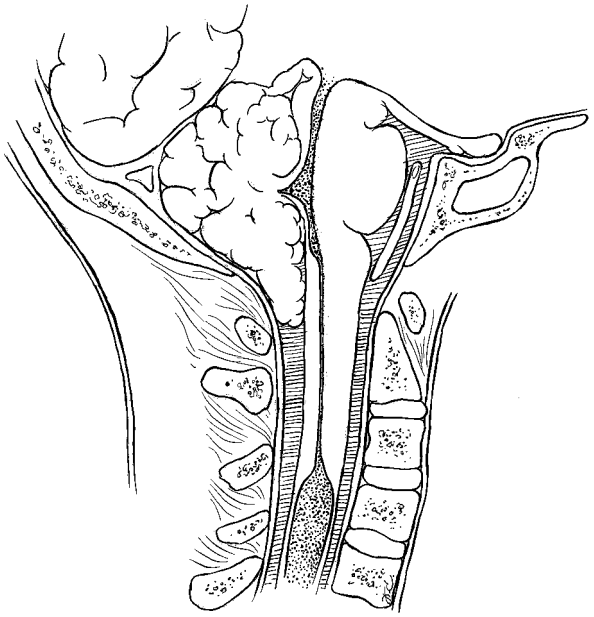


FIGURE 3–1. Midsagittal drawing of the craniocervical junction of a patient with a Chiari I malformation. Note the descent of the cerebellar tonsil to the upper edge of C-1 and the presence of a large syrinx beginning at C-3 and extentding inferiorly. The C1-2 region is spared.

solving the intramedullary CSF collection. Great care must be exercised to search exhaustively for other causes before assuming this to be the case.

Relative contraindications for a Chiari decompression include raised ICP, cervical instability, and significant upper cervical ventral compression.

Preoperative Evaluation

Evaluation of a patient for a CIM should begin with a complete history and physical examination, including a neurologic evaluation. Special attention should focus on the optic fundi for papilledema, pain on neck motion, scoliosis, leg- or foot-length discrepency, loss of pain or temperature over the trunk and an asymmetric abdominal reflex. Sensory testing in small children can mislead even experienced clinicians. Temperature assessments over the trunk are generally more reliable than testing the child with a sharp point.

Today virtually every patient being evaluated for a CIM undergoes MR imaging. Exceptions are children with pacemakers, cochlear implants, or some other technical reason why MR imaging should not be performed. A small subpopulation of patients may need general anesthesia to obtain an adequate study. The detail and ease of MR imaging are truly impressive to anyone who has had experience with the pre-MRI era. No other radiographs come close to the precision of MR imaging in delineating the anatomy of the craniocervical junction or for the evaluation of syringomyelia. Rarely is useful information obtained with contrast enhancement, and it can generally be avoided. Much enthusiasm accompanied the introduction of cine-MR imaging to evaluate CSF flow in this region. We have had this capability for several years and still are awaiting critical information to be obtained using this modality that would be helpful and that would alter the decision based on the routine MR imaging. This includes the presence of CSF behind the tonsils in patients with large syrinxes who were effectively treated with posterior fossa decompression. MR imaging should include images of the cervical and thoracic spinal cord. On occasion, the syrinx from a CIM may skip the cervical region and appear only in the thoracic region.

Some evaluation of ventricular size is necessary. This information can be obtained by either MRI or computerized tomography (CT). Hydrocephalus or raised ICP should be considered prior to any consideration of posterior fossa intervention.

The bony anatomy of the craniocervical junction can be seen easily on routine radiographs of the cervical spine. To the extent the child will voluntarily move the neck, flexion and extension films will help to ensure preoperative stability. These films should be done cautiously or under fluoroscopic control if neck pain is present. The art of persuading a frightened child to move the neck through the extremes of flexion and extension is routinely performed in pediatric centers without difficulty.

Ventral compression by the dens or some other tissue in the region is increasingly recognized as a contributing factor in the formation of hindbrain hernias. Special attention should be given to this assessment. As with many other areas of medicine, ventral compression is not simply present or absent, but rather it may be some gradation along that spectrum from no abnormality to full impaction. If significant ventral compression of the dens into the medulla is present, consideration should be given to addressing this issue primarily and then reassessing the remaining factors. The assessment of ventral compression is done with sagittal MRIs. Flexion and extension MRIs can be obtained in the cooperative child but are technically difficult.

Operative Planning

As previously stated, hydrocephalus and raised ICP should be addressed before any consideration of surgery

for the posterior fossa pathology. Posterior fossa decompression in the presence of hydrocephalus is dangerous and has little likelihood of resolving the CSF flow problem and avoiding a shunt or third ventriculostomy. Cervical instability may be addressed at the same time as the Chiari decompression, but more care will be necessary in positioning and fixating the patient during positioning for the procedure. Ventral compression of sufficient magnitude to warrant primary consideration is generally done by transoral resection of the dens and then careful reevaluation of the craniocervical stability. Once these three issues are resolved, operative planning for the Chiari decompression can begin.

From the preoperative sagittal MRI, the extent of tonsilar descent can be assessed along with the position of the torcula and the degree of any brainstem displacement. The general condition of the skin and, in particular, the skin in the area of the incision should be evaluated. Open draining wounds, as from an insensitive foot ulcer, should be healed before this or any other elective procedure can be considered.

Intraoperative Technique

A successful operation starts with proper positioning. The head is held in a pin head-holder, taking care to avoid a shunt, if present, or excessive pressure if the sutures are not fused. The patient is turned prone and the neck is flexed. Flexion is not extreme, and two fingers should be inserted easily between the chin and chest. The head of the bed is elevated until the operative field is parallel to the floor (Fig. 3–2), which decreases venous pressure and facilitates hemostasis. Once positioned, the midline occipital hair is clipped but not shaved. I use a chest Doppler both to detect air embolus and to listen to the heart rate. Small fluctuations in the heart rate occur with manipulation of vital medullary areas and can be appreciated easily.

The initial exposure begins in the midocciput and extends to the spinous process of C-2 (Fig. 3–3A). It is rare that C-2 must be removed, and leaving the muscular and ligamentous attachments to C-2 minimizes postoperative pain and instability. The avascular plane between the nuchal muscles is identified by a small amount of fat that separates them. With the monopolar current or periosteal elevator, the muscle insertions on the occiput are removed. In the area of the posterior arch of C-1, care is taken to avoid the frequent bony spina bifida and inadvertant injury of the underlying neural tissue. Young infants will have significant cartilage in the posterior arch that can easily be penetrated by using monopolar current or an instrument. With the muscle and other soft tissue reflected laterally, exposing the midocciput to the foramen magnum and the posterior arch of C-1, a decision is made as to the extent of the lateral

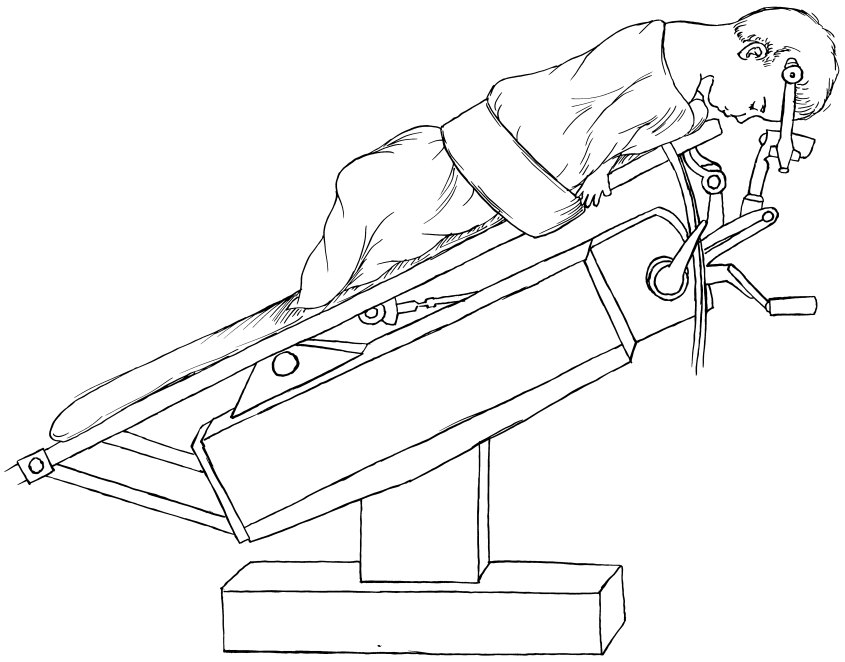
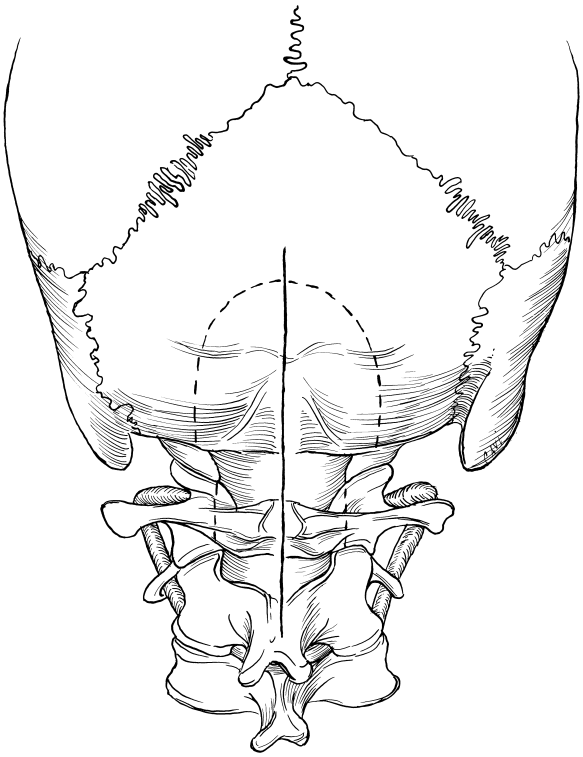


FIGURE 3–2. Position of a child on the operating table. The head is held in a pin head rest with the neck flexed. The head of the table is elevated to decrease venous pressure.



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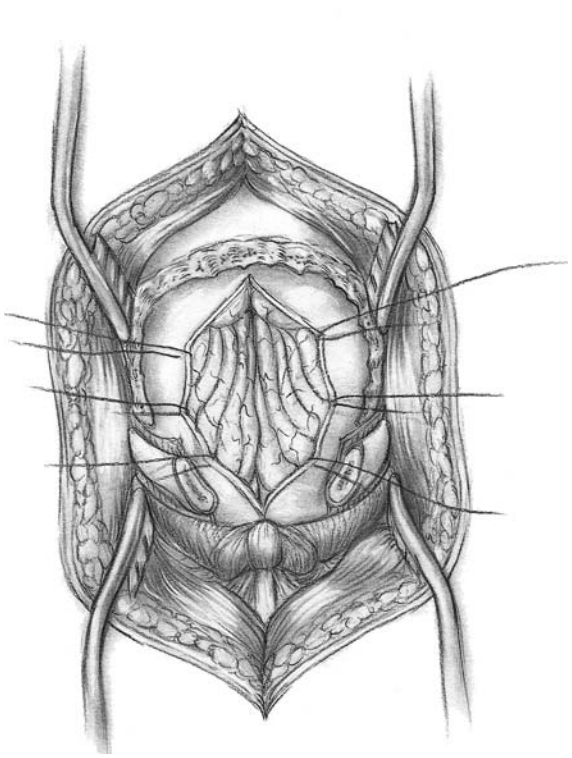

B

FIGURE 3–3. A: Depiction of the skin incision and typical bony removal required for a Chiari I malformation decompression. Note that the upper extent of the skin incision is well below the muscle insertion and that the lower extent is to the spinous process of C-2. The suboccipital craniectomy is limited. The arch of C-1 is removed, but C-2 with its soft tissue is left intact. **B:** Intraoperative view with the bony removal accomplished. The dura is opened in the midline beginning in the cervical region. The cerebellar tonsils are caudally displaced to the level of C-1.

exposure. This is a midline operation, and there is no need for extreme lateral exposure. When both the foramen magnum and posterior arch of C-1 begin to turn vertically, exposure of soft tissue must be halted. Alternatively, the width of the spine from the MR imaging and the exposure necessary can be measured. There is little reason for a vertebral artery injury to occur with this procedure. In addition, the artery is encased in a venous plexus that will transmit an arterial pulse to warn the observant operator of the approaching structure. Burr holes are placed on each side of the midline and connected with a drill or rongeurs. The horizontal width of the foramen magnum opening should approximate the height of the exposure. The posterior arch of C-1 is removed with rongeurs. There is no place for a posterior fossa craniotomy that is replaced; this is a decompressive operation.

At this point, some surgeons would use ultrasound of the underlying neural structures to look for intradural movement. If movement with pulse and respiration is present, an argument can be made not to open the dura. These advocates argue that the bone is the primary constricting force and, with its removal, the problem is solved. In addition, if the dura is left intact, any intradural injury can be avoided and chemical meningitis minimized. My practice is to use ultrasound and, unless appreciable movement is present, to perform an intradural inspection and dural grafting. The dura is opened beginning under the C-1 arch and cautiously extending cephalad. A circular sinus at the foramen magnum is sometimes present as well as a poorly delinated sinus over the dorsal posterior fossa. The younger the patient, generally, the more significant are these venous structures. At times, these structures can be serious technical challenges. Using cautery on the outer dural leaf will increase rather than decrease sinus bleeding. Control of both inner and outer leaves of the sinus is necessary to gain control and can be accomplished by using bipolar cautery or suturing if necessary. Preservation of the arachnoid insofar as possible is important at this point to protect the subarachnoid space from blood contamination (Fig. 3–3B). At times, with a tight foramen magnum, lateral relaxing incisions are necessary in the dura at the point of maximum constriction. Hemostasis is secured before any further manipulation is performed. With the dura opened over the area of bony and dural constriction, the arachnoid is

opened off the midline and the intradural contents are inspected. On occasion, with chronic tonsillar herniation, the tips of one or both tonsils may be blanched or gliotic. The fine reticular vascular pattern may be lost. With magnification and adequate illumination, the tonsils are separated carefully and a search is done for adhesions or arachnoid veils over the foramen of Magendie. When the avascular floor of the fourth ventricle is appreciated, the exploration is terminated. I do not favor obex plugging with any material resection of any neural tissue, or placement of a fourth ventricle to subarachnoid stent on any primary decompression. Adequate establishment of free CSF egress out of the fourth ventricle has a high likelihood of success. All three additional maneuvers have an associated risk that seems unwarranted with the initial procedure.

A periosteal graft is harvested above the nuchal attachment and cut to fit the dural opening. A watertight dural closure is attempted, and the soft tissues are closed in layers.

Postoperative Management

Patients who are clinically stable preoperatively are generally extubated in the operating room and monitored in an intensive care unit overnight. Children are generallly ready for discharge by the third day; minor analgesics are administered. Nausea and persistent vomiting may delay some discharges. Without some other clinical indication, no postoperative imaging is necessary, and patients are simply monitored clinically. Patients with a significant syrinx are exceptions. These patients have cervical or thoracic MR imaging repeated in 4 to 6 months to assess the effectiveness of the decompression. Patients with scoliosis also should have their curve monitored by taking serial measurements.

Operative complications from this procedure should be exceedingly few, with permanent serious problems occurring in less than 1 to 2% of patients. Efficiency with regard to the resolution of preoperative symptoms should be greater than 90% of patients. When pain is the preoperative symptom, a lack of resolution is much more likely to be an indication of poor patient selection rather than a failure of the procedure. Syrinx resolution or significant reduction is quite likely within the 4- to 6-month window of observation.

The greatest challenge with this problem is selecting children for operation who are truly symptomatic from their MR imaging finding or who are at risk of serious consequence if the hindbrain hernia is left to express the natural history of the condition. Resisting operation in children with inappropriate symptoms, moderate cerebellar ectopia, and anxious families demanding the lesion be "fixed" is a test of character for the surgeon.

CHIARI II MALFORMATIONS

The Chiari II malformation (CIIM) (Fig. 3–4) is seen almost exclusively in patients with neural-tube defects. These patients present generally with more serious symptoms, signs, and MRI findings than patients with CIM. Their care and treatment are more difficult and demand a higher level of expertise. With our increasing experience in managing this lesion, the role of truly normalized ICP has become clearer.

Surgical Indications

Categories of presentation for this condition are generally grouped into brainstem disturbances, spinal cord (syrinx) problems, and neck pain with the extreme finding being opisthotonus. On rare occasions, a patient may primarily exhibit cerebellar dysfunction, but this is unusual without other, more distressing symptoms. Cerebellar disturbances, both truncal and appendicular, are quite difficult to detect in a small child confined to a

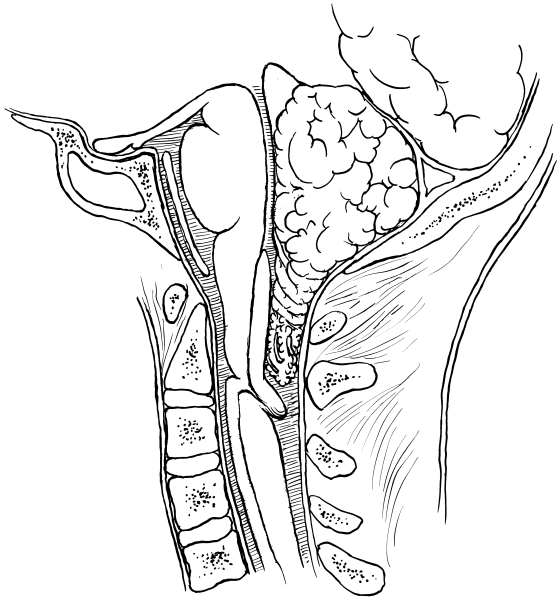

FIGURE 3–4. Midsagittal drawing of the craniocervical junction of a patient with a Chiari II malformation. Note the caudal displacement of the cerebellar vermis, the numerous additional malformations, and the position of the torcula.

wheelchair, which may be a factor in the limited number of patients who present with these findings.

The brainstem presentations occur primarily in infants and are potentially life threatening. Difficulties with swallowing, inspiratory wheezing from laryngeal stridor, poor palatal motion, tongue fasciculation and atrophy, and periodic apnea all may be seen. These problems may be present at or near birth and, if present then, are associated with a poor prognosis with or without therapy. Many of these brainstem symptoms can have insidious presentations as pneumonia representing aspiration from disorganized swallowing or as the infant who is "so good he never cries" because he can not generate a vocal response. All these serious problems are indicative of a life-threatening situation and should be evaluated expeditiously.

Syringomyelia is commonly seen with myelomeningoceles. When present, it should be considered an indication of altered CSF circulation from the CIIM and an indication for intervention. The syrinx itself may be found in evaluating progressive scoliosis, altered bladder function, increasing leg weakness, or simply incidentally.

Epsiodic neck pain or opisthotonus is a more urgent problem that demands explanation. The tempo of evaluation and treatment should reflect this urgency.

In addition to these clinical presentations, a position of the hindbrain hernia at or below C-4 is a strong indication of eventual symptomatic presentation, and patients with this finding should be monitored closely or considered for decompression before the onset of severe symptoms.

Preoperative Evaluation

First and foremost, the ICP must be normalized in patients with CIIM symptoms. Patients with modest ventricular enlargement and no shunt should be shunted. Patients with a shunt in place and "no change," "stable," or relatively small ventricles should have the shunt tapped, ICP monitored, or the system explored surgically before any consideration is given to a Chiari decompression. If the system is explored, it is insufficient to see some flow from the ventricular catheter and assume the system is functional. The catheter also must be free without intraventricular fixation. If the ventricular catheter is adherent, soft tissue must be present within it and will cause intermittent or partial obstruction. Even with small ventricles, endoscopic placement sliding down the old catheter tract can be done successively. Endoscopic inspection of the lumen of the catheter also can be helpful. Numerous children with "unchanged" ventricles or a negative shunt tap have inadequate shunt function and have been permanently injured during a posterior fossa decompression.

The imaging evaluation should include an MR image of the craniocervical junction and cervical spine to evaluate the possibility of a syrinx. Some assessment of the ventricular system is necessary, and comparison with other brain images when the patient was asymptomatic may prove useful. A shunt series on more than one occasion has demonstrated a disconnection with unimpressive ventricular enlargement. In each case, shunt revision has relieved the new symptoms immediately. Flexion and extension lateral cervical spine films are unlikely to demonstrate instability, but this must be excluded, especially in young infants. Ventral compression of the brainstem may be assessed on the MRI and should be specifically evaluated and excluded.

Interest has increased regarding spinal-cord fixation possibly causing symptoms at some distance from the point of fixation and even causing brainstem symptoms and signs. Evaluation of the postmyelomeningocele closure tethered cord syndrome (TCS) rests with clinical criteria alone. Virturally all MR images of patients following a myelomeningocele closure will have evidence of dorsal fixation at the closure site. I still consider posterior fossa decompression before any attempt at untethering for the aforementioned presentations.

A host of additional evaluations can be performed, including brainstem-evoked responses, somatosensory evoked responses, barium or other swallowing evaluations, pulmonary function with CO_2 infusion, and electromyography. Each of these procedures has advocates, but no procedure has reliably provided critical information and therefore none is performed routinely.

Operative Planning

Once a decision concerning shunt function is satisified and no other concerns are present, Chiari decompression is contemplated. Before the procedure is performed, the MRI of the posterior fossa is reviewed. The position of the torcula, extent of the medullary kink, extent of the cerebellar vermis caudal displacement, and presence of a syrinx all should be assessed.

The torcula can be quite low and abut the foramen magnum. There is no justification for opening into this venous sinus because one forgot to assess its position. Typically, the medullary kink will be even with or a segment below the caudal extent of the vermis. The lower aspect of the vermis usually has choroid plexus marking its position. Occasionally, the choroid can be seen on the MRI. The relationships between these structures are assessed to determine the extent of the laminectomy. The position of the syrinx in relation to the spinal segments is also determined.

Intraoperative Technique

As with CIM patients, the infant or child is positioned prone with the neck flexed and held in a pin fixation device. The younger the child, the less pressure is exerted on the skull. With care, even infants may be positioned in these devices, but little compression is placed against a mobile, fragile skull with an open fontanelle and unfused sutures. Again, the shunt is avoided with the pin sites. The patient is secured to the table and supported with wide tape to support the body's weight as the head of the bed is elevated 20 to 30 degrees. The planned incision is marked by palpating the spine. Rarely is there need for exposure above the foramen magnum, and the bone in this area is usually left alone. Most compression occurs at C-1 and to a lesser degree at C-2. The distance from the foramen magnum to C-1 is usually much greater than is normally seen, which must be remembered during the soft-tissue exposure. Routinely, the cervical laminectomy is extended to the level of the medullary kink (Fig. 3–5A).

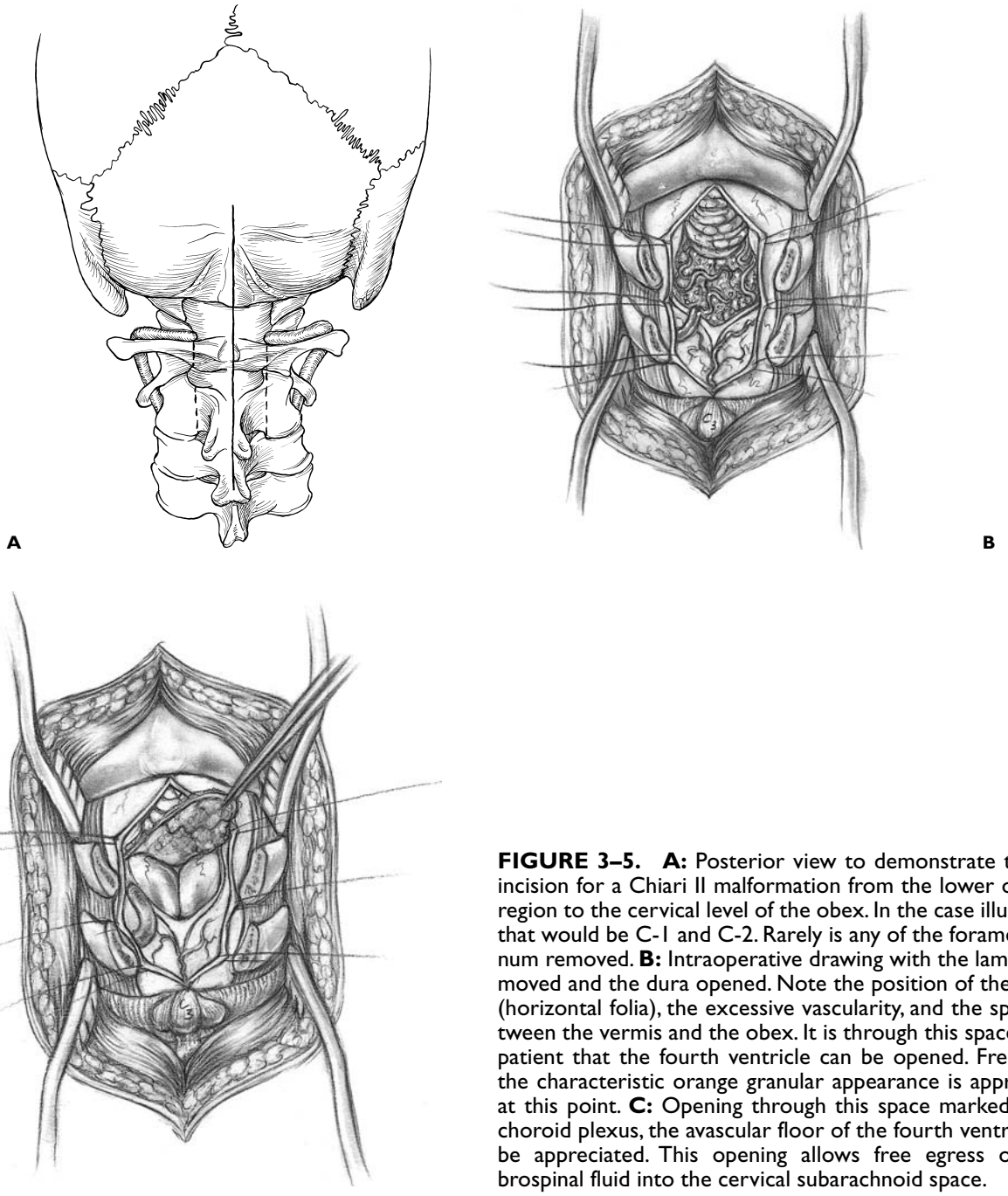

FIGURE 3–5. **A:** Posterior view to demonstrate the skin incision for a Chiari II malformation from the lower occipital region to the cervical level of the obex. In the case illustrated, that would be C-1 and C-2. Rarely is any of the foramen magnum removed. **B:** Intraoperative drawing with the laminae removed and the dura opened. Note the position of the vermis (horizontal folia), the excessive vascularity, and the space between the vermis and the obex. It is through this space in this patient that the fourth ventricle can be opened. Frequently, the characteristic orange granular appearance is appreciated at this point. **C:** Opening through this space marked by the choroid plexus, the avascular floor of the fourth ventricle can be appreciated. This opening allows free egress of cerebrospinal fluid into the cervical subarachnoid space.

More recently, I have limited the exposure to the caudal vermis. The smaller the soft-tissue and bony exposure, the less likely any spinal instability or deformity postoperatively. Spinal instability is of special concern when the syrinx is large and encompasses most of the cervical area.

Again, there is little need for lateral exposure. This a midline operation. The dura is opened in the midline and tented laterallly. Ultrasound may be helpful in determining the anatomy. With the dura open, the arachnoid is opened and clipped to the dura. Significant subarachnoid scarring is frequently present, and determination of cerebellar vermis versus medullary kink may be quite difficult (Fig. 3–5B). For this reason, some surgeons would terminate the procedure with the bony removal or simply by opening and grafting the dura. I favor communication of the fourth ventricle with the subarachnoid space to reestablish CSF egress (Fig. 3–5C).

The tissues in this area are ischemic and as a result may have developed an exaggerated vascular pattern; this may mask the identification of the vermis. At times, the vermis will retain the horizontal folial pattern, and which may help in the dissection. Lateral entry into the fourth ventricle may be easier at times, with extension to the midline secondarily. Injury to a compromised medulla can be avoided by not sacraficing significant vascularity during the dissection and precisely identifying the plane between the vermis and the medulla. The dura is grafted with the pericranium in a redundant fashion, and the soft tissues are closed to prevent a pseudomeningocele.

Postoperative Management

Depending on the degree of exposure necessary and the condition of the patient preoperatively, the patient will be extubated in the operating room and watched in an intensive care unit overnight. Patients are discharged 3 to 5 days after surgery.

EDITOR'S COMMENTARY

Since the advent of MR scans, it has become apparent that many children have asymptomatic CIMs. Unless they have an associated syrinx, they should be followed clinically rather than operated upon; no one knows their likelihood of developing the symptoms or a syrinx over time. The venous lakes in dura at the foramen magnum that Oakes describes can be difficult to control. His technique of controlling that bleeding by suturing the dural edges is a good one. Options for dural grafting include periosteum, cadaver dura or pericardium, fascia lata, and Goretex; the optimal material is unknown. Limited "tonsillectomies" have been performed; data are not available to determine if they facilitate syrinx drainage. A single operation is sufficient in 90 to 95% of CIM cases. The morbidity of CIIM operations is higher, partly because of substantial bleeding due to inadvertent entry into the torcula (the bleeding site can rarely be clipped so must be sutured) and because of brainstem injury caused by trying to open into the fourth ventricle through the vermis rather than under its caudal end. The need for CIIM re-operations will probably be reduced by Oakes' admonition to prove shunt function before any decompression.

PEARLS

In this author's experience:

- Hydrosyringomyelia is never a diagnosis in itself. It is always caused by another pathology. Surgical decision making should be postponed until the *cause* of the syrinx is determined.
- With the availability of MR imaging, increasingly younger patients are presenting for surgical evaluation with minimal symptoms and marginal caudal descent of the tonsils. Without clear Chiari I-related symptoms or a syrinx, caution should be exercised in recommending surgery.
- Chiari II symptoms are life threatening. Early surgical intervention in the symptomatic infant or child is the only method of altering the natural history of the problem.

(*continued*)

PEARLS (*continued*)

- It is always wise to determine the position of the torcula in planning the surgical approach for patients with Chiari II malformations. I question the need to remove any occipital bone.
- Shunt malfunction is assumed to be the cause of all Chiari II symptoms until good shunt function is proven. A "normal" or "unchanged" CT is an inadequate reason to rule out shunt malfunction.

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4

ENCEPHALOCELES, MENINGOCELES, AND DERMAL SINUSES

David F. Jimenez and Constance M. Barone

Although relatively uncommon in North America, the surgical correction of sincipital encephaloceles presents the neurosurgeon with a significant challenge. This chapter discusses surgical principles necessary to obtain an optimal surgical repair and results when managing these lesions. Additionally, the chapter introduces similar surgical principles necessary to successfully manage occipital encephaloceles and cranial dermal sinuses.

ANTERIOR CRANIAL FOSSA ENCEPHALOCELES

Although sincipital encephaloceles are relatively uncommon in North America, they present the surgeon with a complex set of management issues. Ideally, these lesions should be treated by a craniofacial team with interest and experience in their management. The vast majority of patients present with normal intelligence and development, but because of the nature of the lesion, moderate to severe facial disfigurement is frequently seen and may be associated with psychosocial problems. The complexity of the surgical correction is dictated by the extent of associated craniofacial deformities. Besides resection of the encephalocele, other important issues that need to be addressed include correction of hypertelorism or telecanthus, vertical dystopia, and nasal deformities as well as management of nasolacrimal duct obstruction and epiphora. The input of a plastic craniofacial surgeon is essential for a successful outcome.

Surgical Indications

Most of these lesions are evident at birth. Depending on the severity, they may present as a barely noticeable mass at or near the nasion or as a larger lesion with extension into the midface and orbits. Although the diagnosis is made clinically, at least a computed tomography (CT) scan should be obtained to delineate the bony involvement, the extent of the lesion, and the presence of associated neurologic anomalies. Hydrocephalus must be ruled out before any surgical correction is attempted. If available, a magnetic resonance (MR) scan also should be obtained to demarcate fully the extent of the encephalocele and to assist in preoperative planning.

Operative Planning

At least two large-bore peripheral intravenous lines are inserted, given the potential for significant blood loss. An arterial line is inserted to monitor blood pressure, blood gases, coagulation profile, hematocrit, and serum electrolytes. If the procedure is done in small children or infants, a warming blanket should be used to ensure proper

temperature regulation and normothermia. A long anesthesia circuit should be used to allow the surgical team access to the patient's head on all sides. A single dose of antibiotics (first-generation cephalosporin) should be given within 45 minutes prior to starting the procedure. If the operation lasts more than 6 hours, a second dose then is administered. Given the need for facial exposure and possible injury to the eyes, ocular shields should be inserted with ophthalmic lubricating ointment.

Intraoperative Techniques

These procedures are done with the patient under general anesthesia in a supine position, on the horseshoe with the head slightly extended to gain access to the entire anterior cranial fossa and face. The entire scalp and face should be prepared with povidone–iodine solution. The scalp is infiltrated with lidocaine 0.25% with epinephrine 1:400,000 for adequate hemostasis. The head need not be shaved except for a small area where the bicoronal incision is to be made. The hair is draped out of the working field with stapled towels posteriorly and 4 × 4 surgical sponges anteriorly. The shape of the incision is the surgeon's choice, although many surgeons prefer a zigzag-type bicoronal incision because it provides better postoperative cosmetic results. The upper face is draped into the surgical field above the upper lips, placing povidone–iodine soaked cottonoids into both nostrils. A standard bicoronal scalp flap then is elevated using monopolar electrocautery (needle tip, low wattage) to create a bloodless subgaleal dissection plane. A separate pericranial flap is elevated from the most posterior aspect of the incision, extending laterally to the superior temporal lines and anteriorly to the supraorbital rims and nasofrontal suture (Fig. 4–1). Care should be taken to prevent injury to the supraorbital nerve and vessels.

Intracranial Approach

Described herein is the standard and currently accepted method for approaching sincipital encephaloceles. The procedure may be performed alone, followed later by the second stage of the procedure or as a combined single-stage approach. Most craniofacial centers currently perform a single-stage correction of these lesions.

Following elevation of the bicoronal scalp and pericranial flaps, burr holes are strategically placed to elevate a bifrontal craniotomy. We have found it unnecessary to create multiple holes to raise the bone flap and only one or two located under the temporalis muscle below the superior temporal line (Fig. 4–2). The bone flap does not need to be large and should extend as close as possible to the supraorbital rims. In adolescents and adults, the frontal sinus should be preserved and not included with the craniotomy.

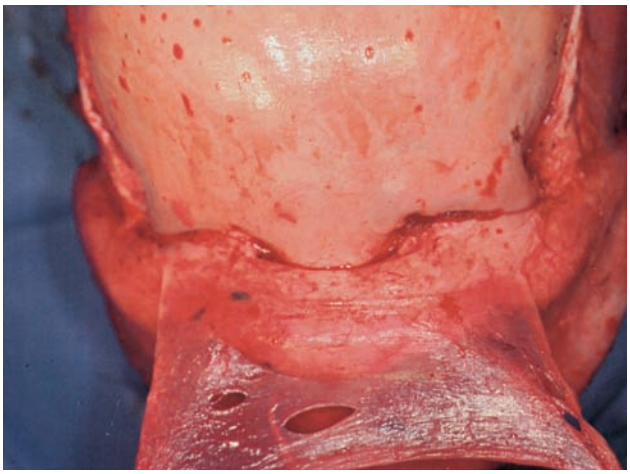


FIGURE 4–1. Elevation of a separate pericranial flap is extremely important for proper closure. The distal end of the pericranial graft can be cut, and a free graft may be used to recreate dural closure and obliterate the bony defect. The remaining vascularized pericranial graft then is inserted under the dura and above the anterior cranial fossa. The dura is closed in a watertight fashion using a running interlocking braided nylon suture.

Following elevation of the bone flap, the dura is exposed and in most patients with nasofrontal and nasoethmoidal encephaloceles, the neck of the lesion can be identified easily (Fig. 4–3). For basal encephaloceles, exposure of the neck requires posterior dissection over the cribriform plate or planum sphenoidale. The dura is opened over the frontal lobes, immediately above the supraorbital rims. The sagittal sinus is doubly ligated with 00 silk and cut between dura openings (Fig. 4–4). The dural incision then is taken posteriorly to include the falx cerebri. Elevation of the frontal lobes should expose the neck of the encephalocele. Bipolar electrocautery is used to cauterize the cortical surface of the neck, and suction can be used to amputate the lesion flush with the cranial base (Fig. 4–5). As much of the gliotic, herniating mass as posible should be resected from the bony canal.

A free pericranial graft then is used to close the dural defect in a watertight fashion with 0000 braided nylon (Fig. 4–6). A vascularized pericranial graft then is laid over the dura and the cranial fossa floor. Often, following resection of nasofrontal encephaloceles, large areas of

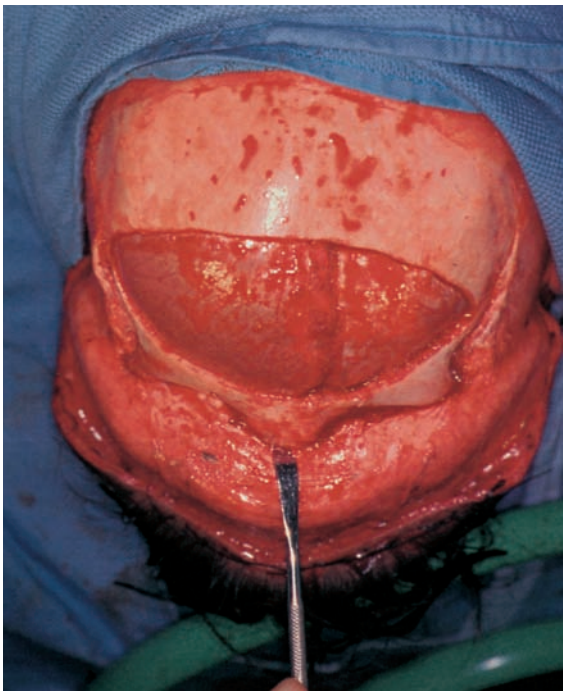


FIGURE 4–2. Elevation of a bifrontal craniotomy may be performed with a footed attachment of a high-speed drill (Midas Rex B5). The craniotomy is extended as inferiorly as possible to improve visualization of the encephalocele.

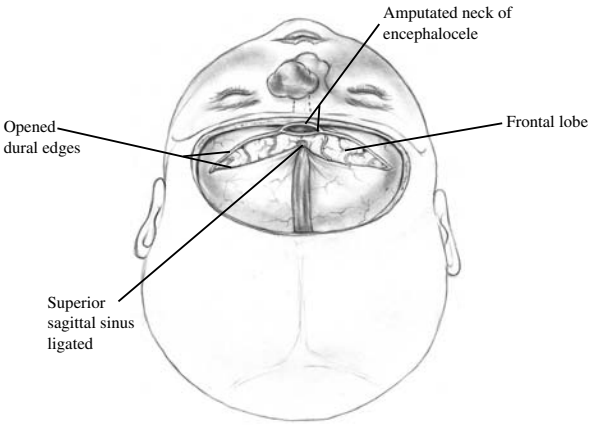


FIGURE 4–4. Following bifrontal exposure, the dura on either side of the sagittal sinus is opened. The sinus is doubly ligated with 2.0 silk sutures. After transection of the sinus, the frontal lobes and the encephalocele are exposed. The neck of the lesion is transected flush with the floor of the anterior cranial fossae using bipolar electrocautery or ultrasonic aspiration.

dead space are created at the resection site. We prefer to use vascularized pericranial grafts to manage these spaces. As previously described, a large pericranial flap is elevated, the blood supply of which is based on the supraorbital vessels. The lateral edges extend along the superior temporal lines, and the posterior edge extends several centimeters behind the coronal suture. The grafts

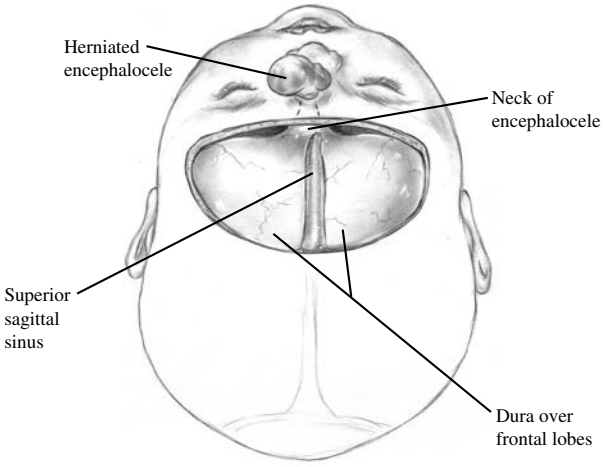


FIGURE 4–3. The classic approach to sincipital encephaloceles requires a bifrontal, intradural exposure to the neck of the encephalocele. A bifrontal craniotomy is created, thereby allowing exposure of the floor of the anterior cranial fossa and the area of the foramen cecum.

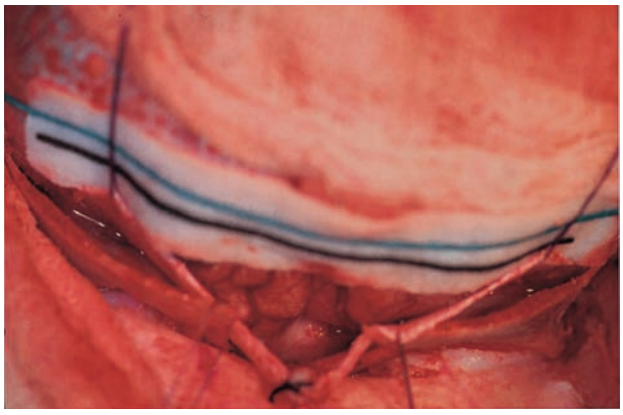


FIGURE 4–5. The area of the glabella also may be resected to improve visualization of the encephalocele. Intraoperative photograph shows removed glabella, ligated sagittal sinus inferiorly, and neck of the encephalocele. A cottonoid has been placed over the frontal lobes.

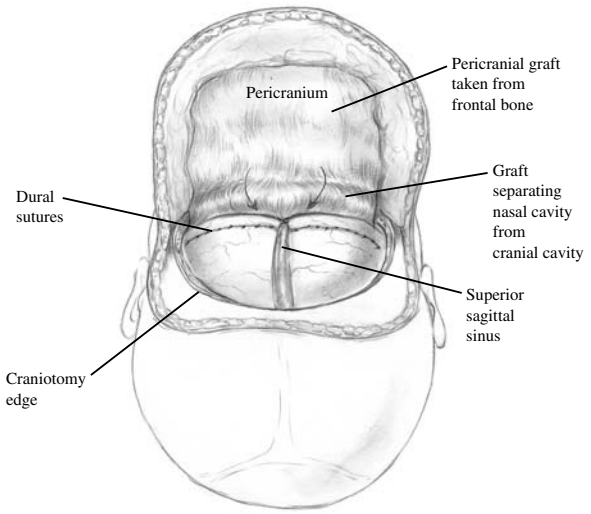


FIGURE 4–6. Following resection of the encephalocele, the dura is closed in a watertight fashion, and a vascularized pericranial graft is used to isolate the nasal and cranial cavities.

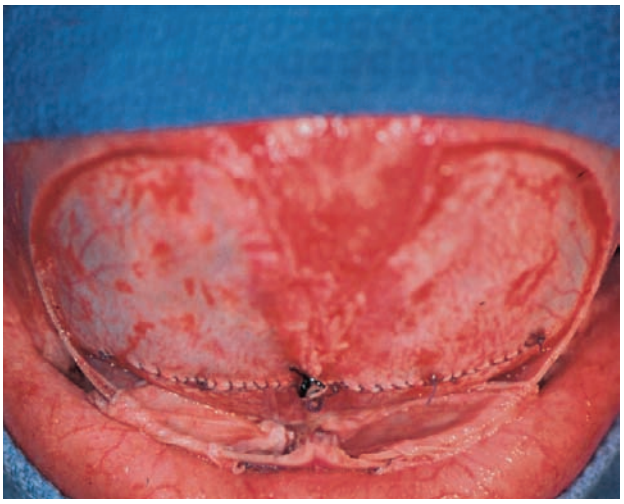


FIGURE 4–7. Dural closure in a 3-month-old infant following resection of nasoethmoidal encephalocele. Braided nylon is used to close the dural opening. Note the placement of the lateral pericranial grafts under the orbital surface of the frontal lobes.

can be divided longitudinally into thirds. The medial third is used for reinforcement of the dural patch graft and coverage of the nasal bone grafts. The lateral thirds are brought down into the resection site through small osteotomies located in the superior medial orbital walls (Fig. 4–7). The grafts are used to line the walls and fill the cavities. This maneuver is particularly important if entrance into the nasal cavity occurs during the resection of the encephalocele. If there is a large bony defect, an autologous split-thickness calvarium can be harvested to reconstruct the bony defect. Proper dural closure is followed by replacement of the bone flap, which is rigidly fixated with microplates.

Although previously unreported, in our series, we have found a high incidence of nasolacrimal apparatus blockage in patients with frontal encephaloceles. Early surgical experience resulted in several nasolacrimal duct infections following resection of the encephalocele without treatment of the blockage. Currently, we proceed with nasolacrimal duct cannulation prior to resection. A punctal dilator is used to probe and dilate the superior and inferior puncta, followed by insertion of the Crawford guidewires.

These guidewires are attached to a long Silastic tube. The guidewires are guided medially and then inferiorly to exit in the nasal cavity under the inferior turbinate. The ends of the Silastic tube then are ligated, cut, and left in place for 6 months. Once removed, a patent nasolacrimal system should be present, thereby relieving the symptoms associated with obstruction. We have found this management protocol to be extremely helpful in the care of these patients.

A subgaleal drain is left in place for 24 to 36 hours postoperatively. If the patient is hyperteloric, hypertelorism correction can be carried out prior to closure of the bicoronal incision. Preferably, radial osteotomies are carried along the superior, medial, inferior, and lateral anterior orbital margins. Following resection of the widened nasal bone, medial orbital transposition osteotomized orbits is done along with medial canthopexy.

Extracranial Approaches

In the past, strictly extracranial approaches to sincipital encephaloceles have proven to be unsatisfactory for treatment of these lesions. Although the extracranial part may be resected easily proper dural closure is sometimes difficult to obtain, leading to a higher incidence of cerebrospinal fluid (CSF) leaks and infection. Currently, there is consensus among craniofacial and neurologic surgeons that, given current available techniques, endoscopic approaches to repair encephaloceles are not warranted and are associated with greater risk of infection, meningitis, inadequate resection, and CSF leaks.

Our method of choice for treating nasofrontal, nasoethmoidal, and nasomaxillary encephaloceles involves

modification of the aforementioned methods. We have found it unnecessary to perform a bifrontal craniotomy to do an adequate repair (Fig. 4–8). A standard bicoronal flap is elevated as described. Using a high-speed drill (Midas Rex, Ft. Worth, Texas) a straight bit (C1) is used to create a small nasofrontal craniotomy. The osteotomies are made along the lateral edges of the nasal bone, medial to the orbits, and superiorly across the root of the glabella. This section of the nasal bone is carried out distally to the end of the nasal dorsum. The unit is elevated with a small elevator, and the entire unit is removed, thereby exposing the neck of the encephalocele and unroofing its bony canal (Fig. 4–9). The dura is incised at the level of the nasofrontal suture, and the neck of the encephalocele is resected. Resection is carried out directly posteriorly reaching the anterior margin of the cribriform plate (Fig. 4–10). Elevation of the dura from the anterior fossa is carried out circumferentially around the bony defect. A free pericranial graft is sutured to the surrounding dura. Fibrin glue may be used to seal the defect further. If fibrin glue is not available or if one chooses not to use it, a spinal drain may be inserted preoperatively and left in place for several days during the postoperative period. A vascularized pericranial graft then can be used to compartmentalize the cranial and nasal cavities (Fig. 4–11). The herniating mass then can be resected and delivered through the nasal opening, even if it extends into the orbit or maxillary sinus. If the skin overlying the herniating mass is of normal texture, it is left alone and dermapexy sutures are placed for proper contouring. If the skin is grossly abnormal, it is resected and properly closed. Hypertelorism may be corrected us-

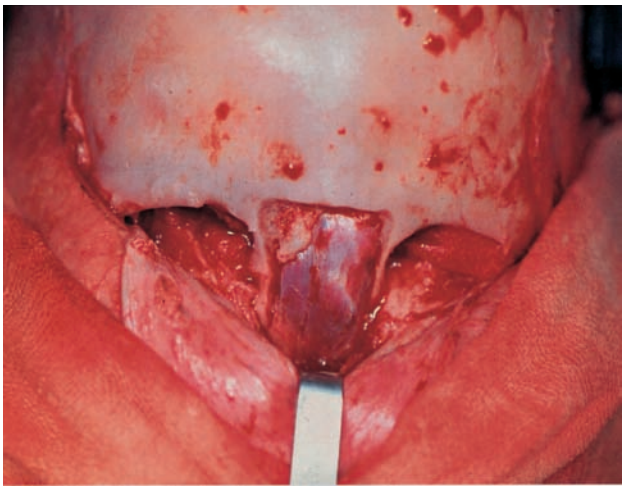


FIGURE 4–9. Adequate exposure of the neck of the encephalocele is obtained through the "nasional" craniotomy. A bicoronal flap has been elevated and displaced anteroinferiorly to expose the orbital rims and nasal area. Following elevation of the craniotomy, the dura overlying the encephalocele is seen and easily exposed.

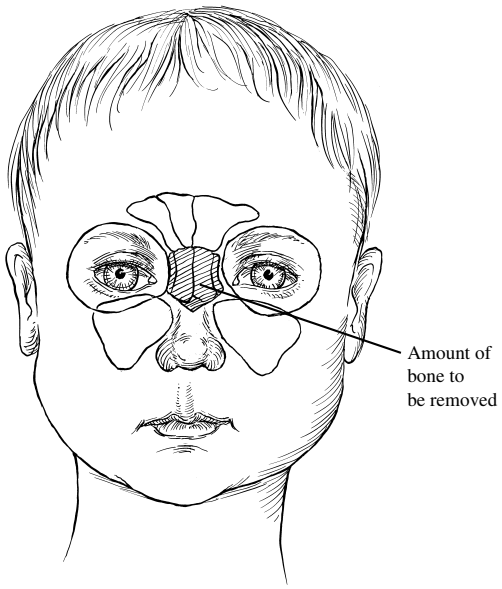


FIGURE 4–8. Our method of choice for repairing nasofrontal encephaloceles does not require a bifrontal craniotomy. The nasional craniotomy involves removal of the inferior-most aspect of the glabella as well as the nasal bones, as demonstrated in this diagram.

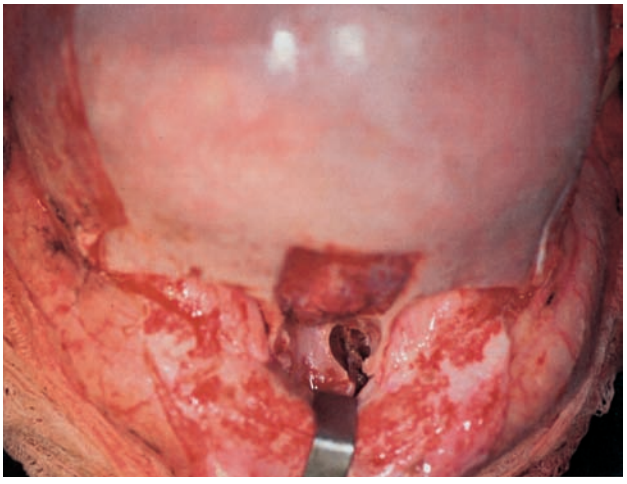


FIGURE 4–10. Following dural opening, dissection of the neck is extended posteriorly at the level of the nasofrontal suture to reach the cribriform plate. Complete amputation of the encephalocele as well as the bony defect that allowed extension of the encephalocele into the left orbitomaxillary complex are seen.

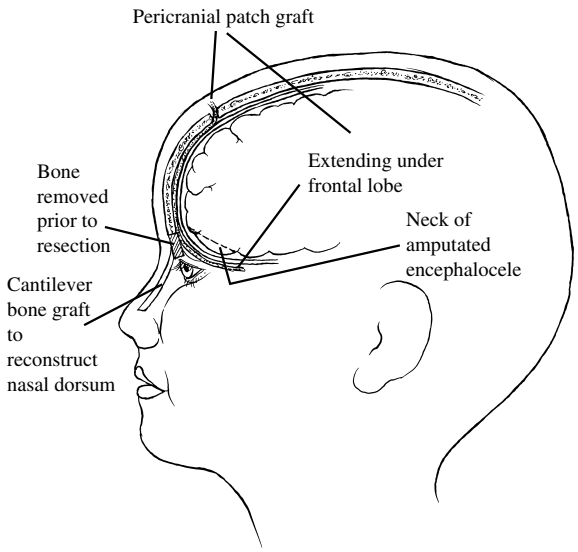


FIGURE 4–11. Following resection of the encephalocele, a vascularized pericranial graft is used to isolate the cranial cavity from the nasal cavity. If the bony defect is large, it may be reconstructed with autologous calvarial graft to obtain adequate cosmesis. Small defects need not be grafted. A cantilever bone graft may be used to reconstruct the nasal dorsum.

ing this approach without the need for bifrontal craniotomy. The small bone graft is rigidly fixated in situ to obtain proper frontonasal contour (Fig. 4–12).

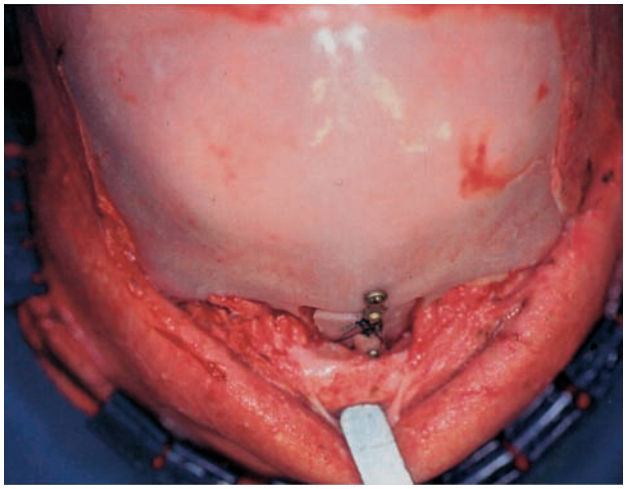


FIGURE 4–12. Following resection of the encephalocele and adequate dural closure, the bone graft is rigidly fixated with miniplates. If necessary, dorsal nasal reconstruction may be achieved by using a free calvarial cantilever bone graft. The top of the graft is seen attached to the center of the bone graft.

OCCIPITAL ENCEPHALOCELES

Preoperative Management

A MR scan of the brain as well as magnetic resonance angiography (MRA) should be obtained prior to attempting resection of the encephalocele. This type of encephalocele is often in close association with dural venous sinuses, and the complex venous anatomy associated with some meningoceles should be well delineated before closure and repair. Cerebral angiography is not routinely performed. Unless the sac is ruptured and there is concomitant CSF leakage, repair of the encephalocele may be done at the earliest possible time (Fig. 4–13). Given the risk of bleeding, at least two large-bore intravenous lines should be inserted. An arterial line should be in place to monitor blood pressure and to follow blood chemistries during the procedure. Several units of packed red blood cells should be available at the beginning of the operation. It is imperative to avoid hypothermia in neonates, and thus, a warming blanket should be used to maintain normothermia. The patient is positioned prone on an appropriately padded horseshoe headrest. Care must be taken to avoid injury to the eyes from improper positioning and undue ocular pressure. A single dose of an appropriate antibiotic should be given

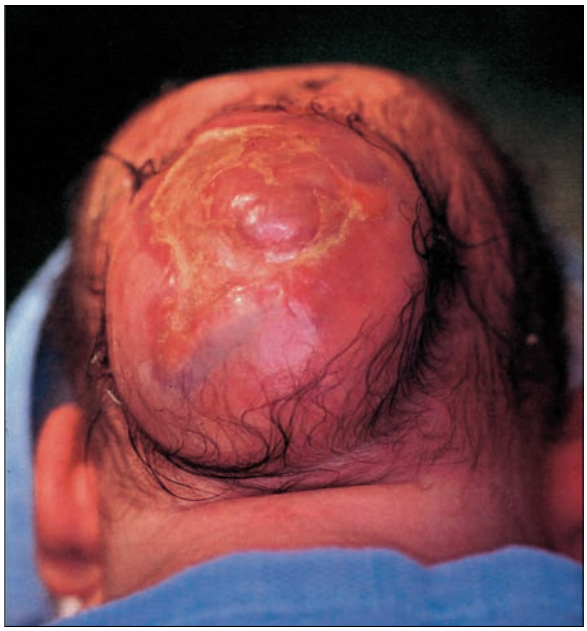


FIGURE 4–13. Newborn infant with typical suboccipital encephalocele. Irregular edges of improperly epithelized skin are noted over the encephalocele. The lack of CSF leakage is allowed for elective closure the day after birth.

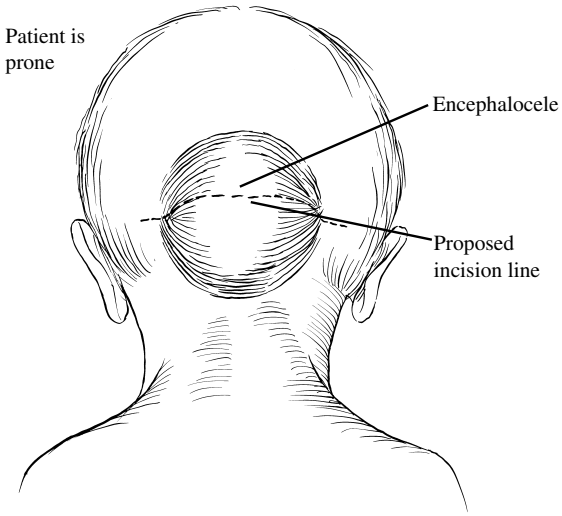


FIGURE 4–14. A transverse incision is made to gain access to the encephalocele mass. A vertically oriented incision would be preferable if the encephalocele extends into the upper cervical area.

prior to skin incision. Preparing the surgical field can be done with povidone–iodine solution.

Intraoperative Management

A transverse incision is preferred for occipital encephaloceles (Fig. 4–14). Vertical incisions should be considered in cases where the encephalocele is located low in the posterior fossa or when there is extension into the upper cervical spine. We prefer to use fine-needle electrocautery at a low wattage level to minimize bleeding. A plane usually can be developed subcutaneously around the encephalocele sac. Circumferential dissection is carried out in all directions toward the bony edges (Fig. 4–15). Following incision of the sac, the contents are explored (Fig. 4–16). A pure meningocele can be repaired

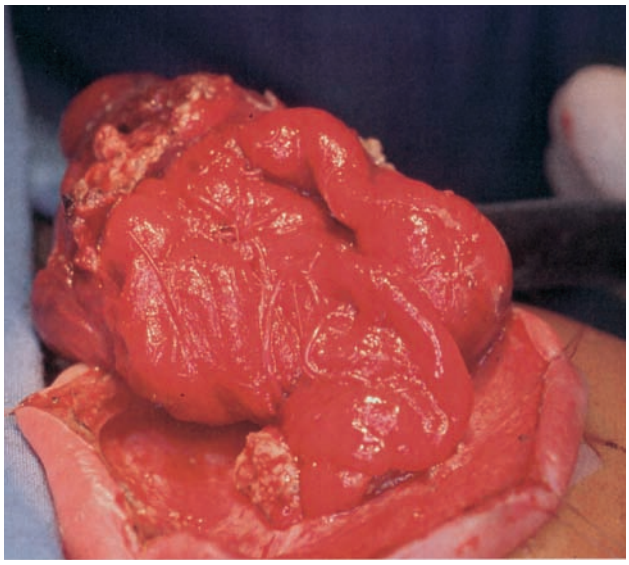


FIGURE 4–16. After opening the sac, inspection of its contents often reveals disorganized, gliottic, nonfunctional tissue. When this is the case, a decision can be made to amputate the mass.

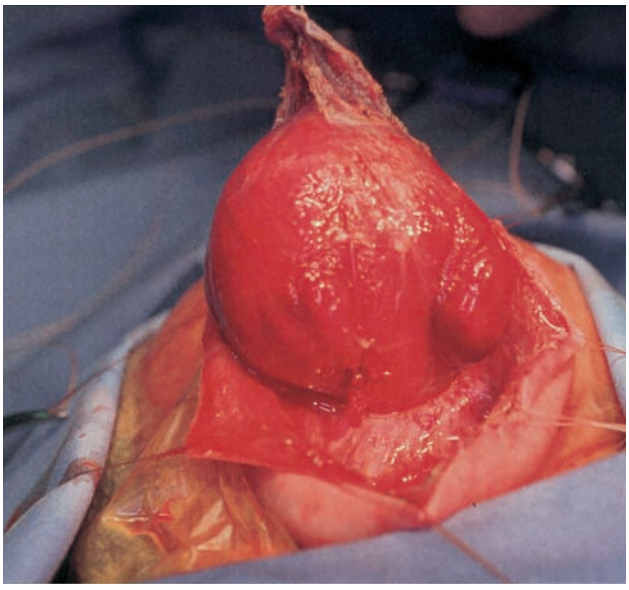


FIGURE 4–15. Following skin incision, the encephalocele sac is dissected circumferentially and held by its apex with a pair of hemostats.

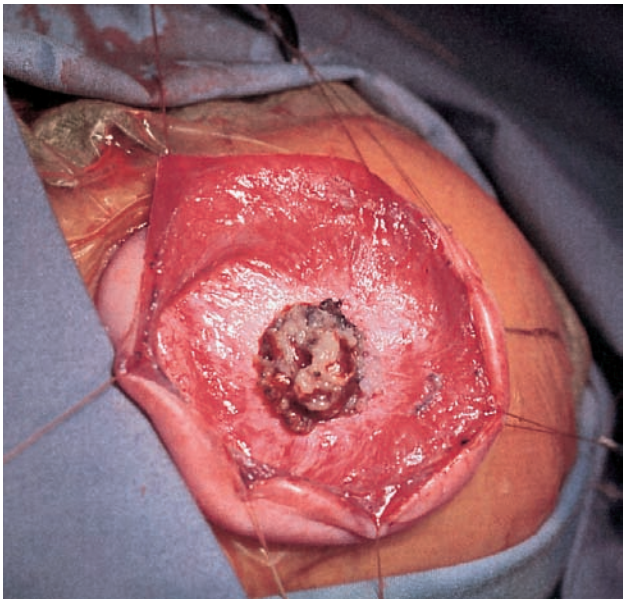
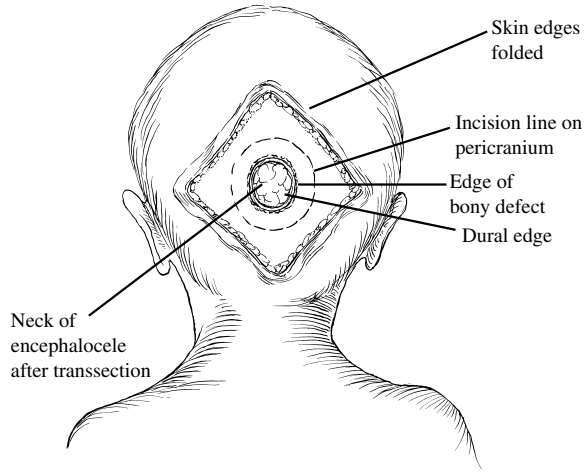


FIGURE 4–17. Occipital encephalocele has been amputated and resected flush with the occipital bone.

easily simply by folding over and suturing the dural edges at the base of the stalk. If neural tissue is found, careful inspection should be done. If the tissue is grossly dysplastic (as in most cases), amputation of the herniating mass can be done by using bipolar electrocautery and sharp dissection at the base of the encephalocele (Fig. 4–17). A watertight closure of the surrounding dura can be done by enfolding the dural edges and using 4.0 braided nylon (Fig. 4–18).

In some instances, inspection of the herniated cerebral tissue will reveal relatively normal neural tissue. In such cases, an attempt should be made to preserve as much tissue as possible (Fig. 4–19). The use of an expansion cranioplasty may be ideal to create the necessary room to accommodate the herniating mass. Following adequate exposure and dural closure, circumferential osteotomies are created on the bone surrounding the encephalocele (Fig. 4–20). A calvarial donor graft may be obtained from the normal surrounding cranium (Fig. 4–21). Radial osteotomies on the donor graft will allow proper contouring (Fig. 4–22). The expansion cranioplasty is finished by securing the donor graft in situ with microplates or miniplates (Fig. 4–23). Excellent results can be obtained using this technique (Fig. 4–24).



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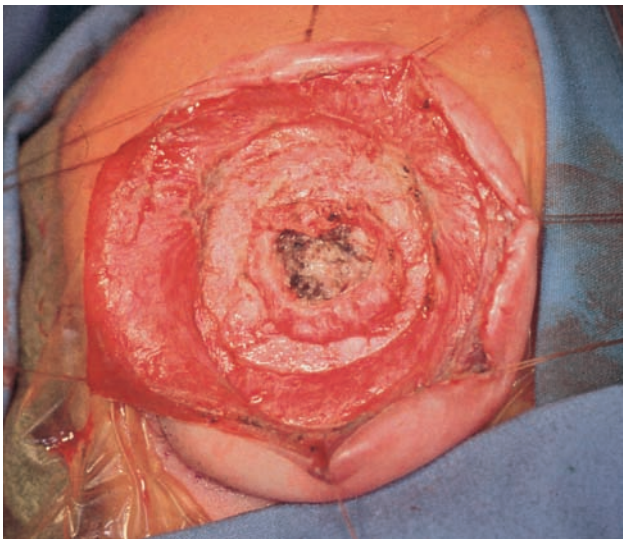

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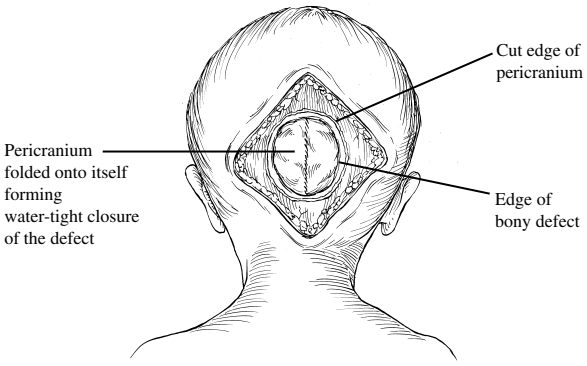

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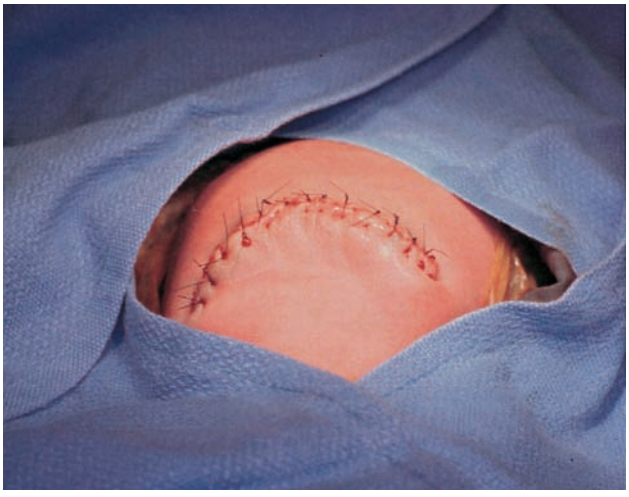


D

FIGURE 4–18. **A:** A watertight graft is created. To obtain adequate closure, the surrounding pericranium is used to create a neodura. Dotted lines indicate the proposed site for pericranial incision. **B:** A surrounding cuff of pericranium has been created and will be used to develop a new dural closure. **C:** Final dural closure is obtained by infolding the surrounding pericranium edges and suturing it with interlocking nonabsorbable suture. **D:** Skin closure is done with interrupted nylon sutures. In the vast majority of cases, primary closure can be achieved without the need for transposition flaps.

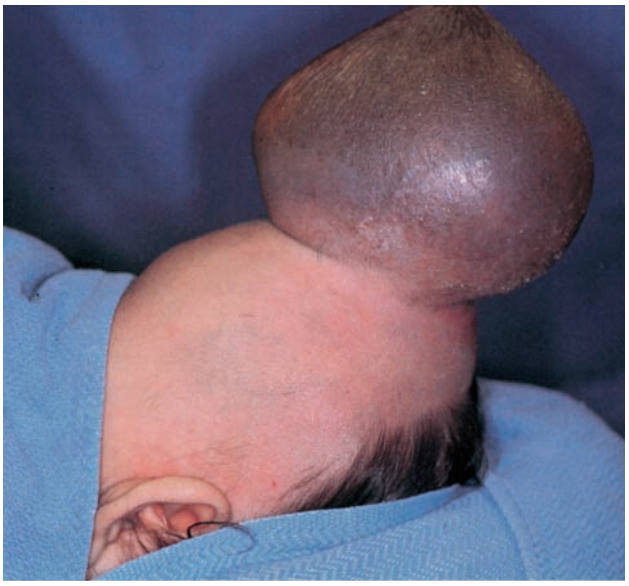


FIGURE 4–19. Newborn male infant with a large occipital encephalomeningocele located 7 cm above the torcula.

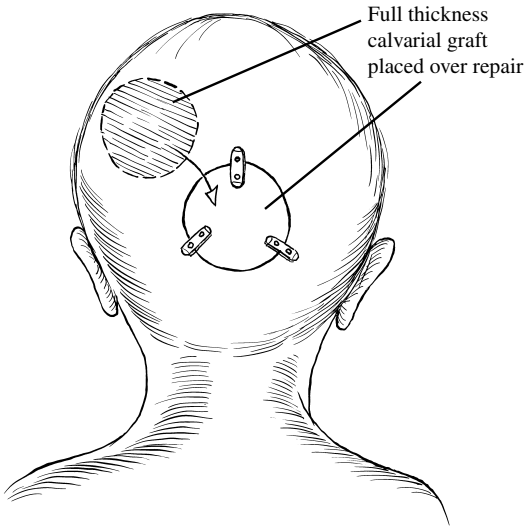


FIGURE 4–21. An excellent method to close the bony defect overlying the encephaloceles involves creating a full thickness craniotomy in the adjacent calvaria and transposing it to the bony defect. The dura is of the donor site will induce osteosynthesis and closure of the defect.

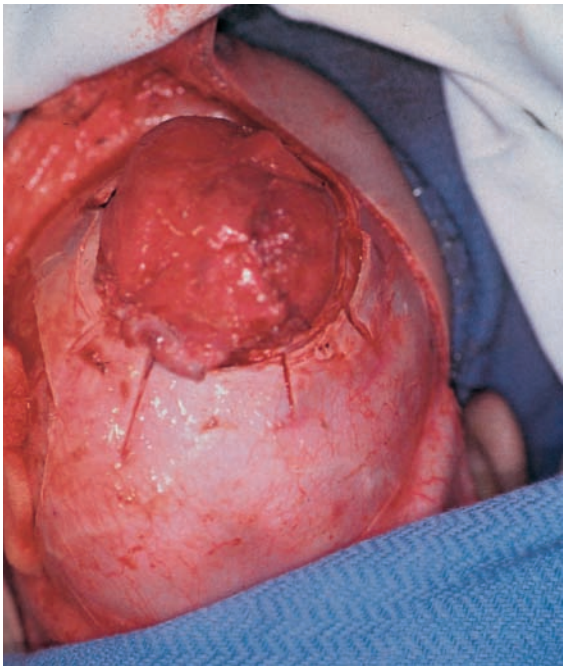


FIGURE 4–20. Following sac opening and drainage of cerebrospinal fluid, inspection of the encephalocele revealed well-organized cortex. Dural closure has been done with free pericranial graft. Circumferential osteotomies allow for calvarial expansion.

DERMAL SINUS TRACTS

Surgical Treatment

The surgical principle governing the management of any dermal sinus (cranial or spine) is that the tract and any associated dermoid tumors need to be removed prophylactically following diagnosis. Furthermore, excision of the tract should include its full extradural and intradural extensions (Fig. 4–25). The goal is to remove these lesions before they become infected or meningitis develops. Simple excision of the sinus tract is inadequate because these tracts are lined with epithelium and incomplete resection may lead to subsequent meningitic episodes or deep tumor development. If excision of these lesions is delayed until they become symptomatic (infected), subsequent management will be more difficult, creating a less than optimal final result. If the patient presents with an infected tract, meningitis, or hydrocephalus, these conditions should be treated and controlled prior to surgical resection of the sinus.

Nasal lesions should be approached in consultation with a plastic surgeon. Our approach is to perform a single-stage procedure, although some authors advocate staged operations in which a craniotomy is done first and the nasal component is resected later. We see no advantage to such an approach. Given that nasal tracts originate in the nose and extend through the foramen cecum,

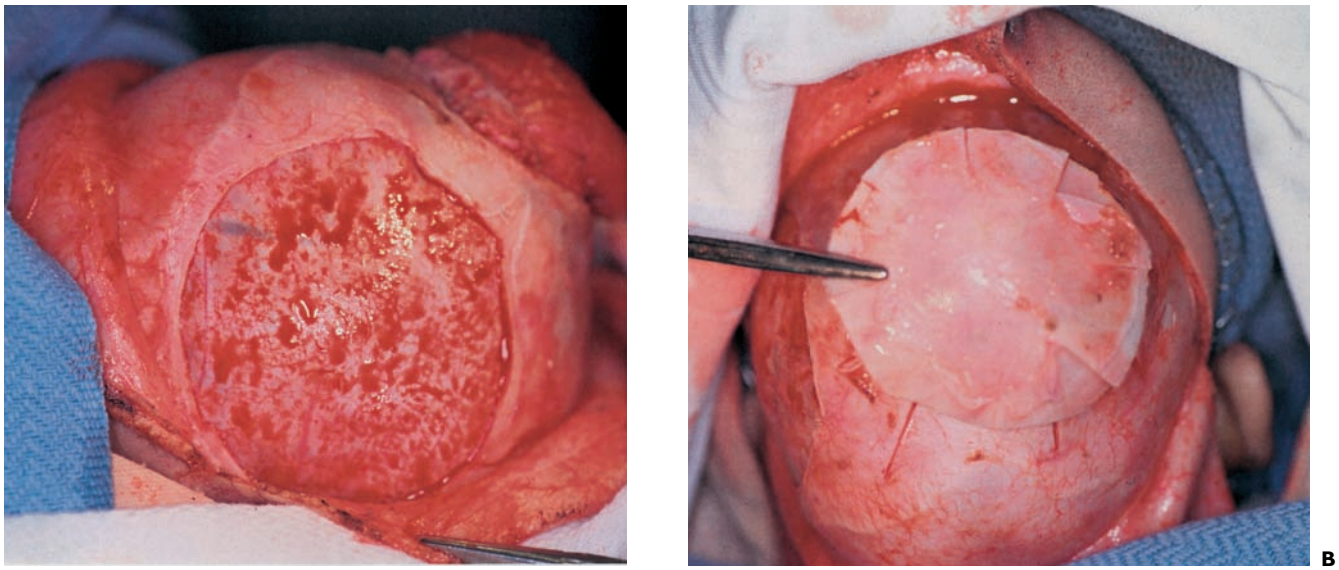


FIGURE 4–22. **A:** Lateral view of the donor site craniotomy anterior to the lambdoid suture. **B:** Donor graft with radial osteotomies that allow appropriate rounding and contouring of graft.

a midline partial rhinotomy and a frontal craniotomy may need to be done to resect these lesions completely. Intracranial dermoids are removed, the dura is closed, and the tract is amputated below the level of the bone. The entire tract should be removed from the skin and internasal structures entirely to prevent future recurrence or infection.

Intraoperative Management

Following induction of general anesthesia, a limited vertical midline incision is made in the nasal area incorporating all associated dermal pits and cutaneous fistulas (Fig. 4–26). The tract is dissected circumferentially along its path from the surrounding subcutaneous tissue and fol-

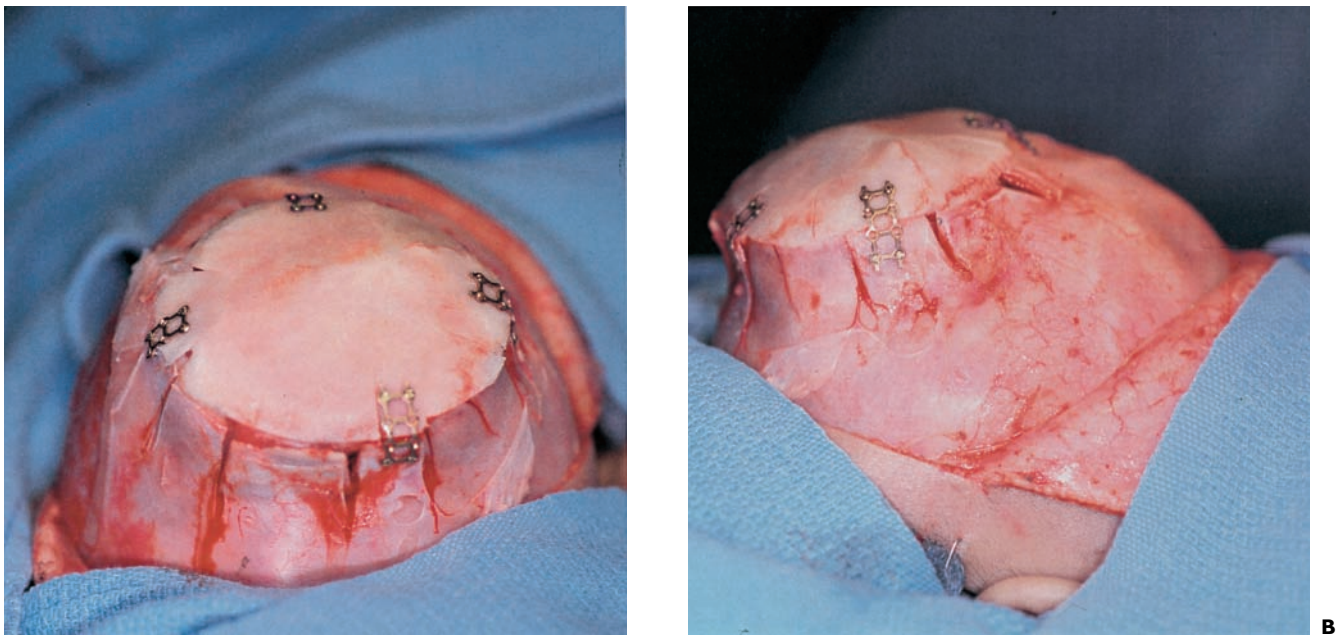


FIGURE 4–23. **A:** Donor bone graft has been contoured and shaped using Tessier rib benders and a high-speed drill (M12-Midas Rex). The graft is rigidly fixated with microplates. **B:** Lateral view of the expansion cranioplasty for closure of occipital encephalocele.

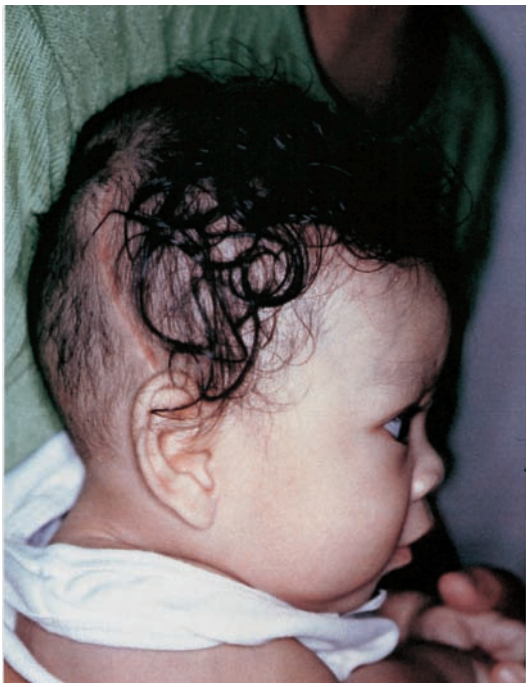


FIGURE 4–24. Three-month postoperative view of patient following closure of encephalocele using an expansion cranioplasty.

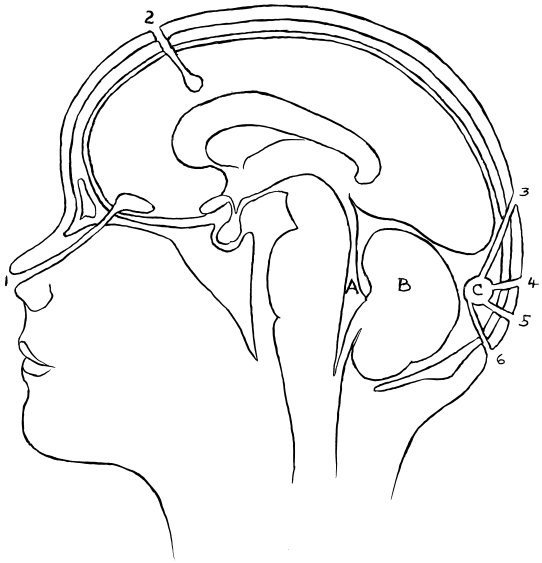


FIGURE 4–25. Cranial dermal tracts may extend into the cranial cavity along the midline and from multiple directions. The most common types include the nasal (1) and suboccipital (5) types. In addition, frontal and parietal tracts with intraparenchymal dermoid extension (2) may be found. Occipital dermal tracts extending into the infratentorial compartment (3, 4), also may be seen. In addition, dermoids may be found at the level of the fourth ventricle (A), cerebellum (B), and extracerebellar compartment (C).

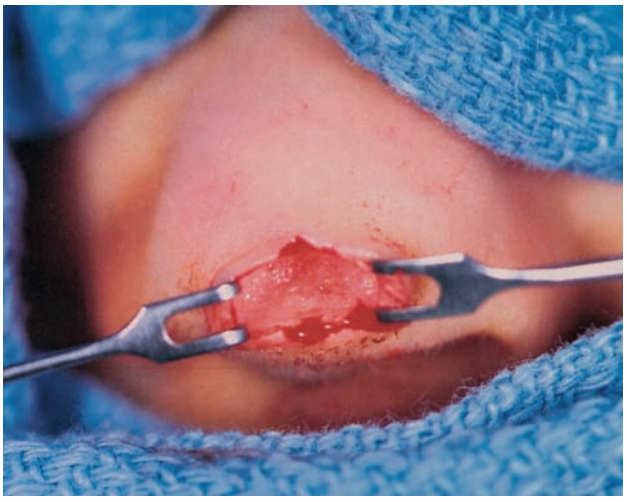


FIGURE 4–26. Nasodermal tracts are commonly associated with dermoids at either terminus of the tract. This 10-month-old infant presented with an enlarging nasal tip mass. A circumferential, vertically oriented incision is made at the nasal tip. The cyst is opened, and the contents are removed, and followed by complete excision of the cyst wall.

lowed to the point of attachment with the skull base (Fig. 4–27). If the tract penetrates the nasal bones, they are fractured and separated vertically over the dorsum of the nose at the nasofrontal suture. Periosteal integrity is maintained at the nasomaxillary junction and directly over the lateral aspect of the nasal bones. A bicoronal scalp flap is

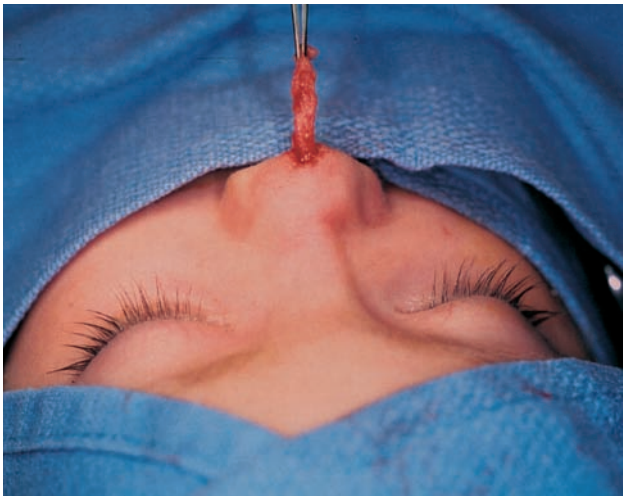


FIGURE 4–27. Following resection of the dermoid cyst, the dermal sinus tract is dissected cranially. The use of tension on the tract aids in delineating the direction of the sinus tract. Tenotomy scissors can be used to skeletonize the tract. A preoperative magnetic resonance scan aids in determining whether the tract extends into the septum or dorsal to it.

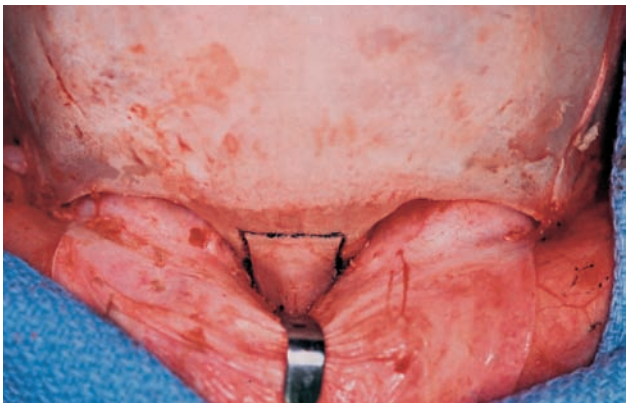
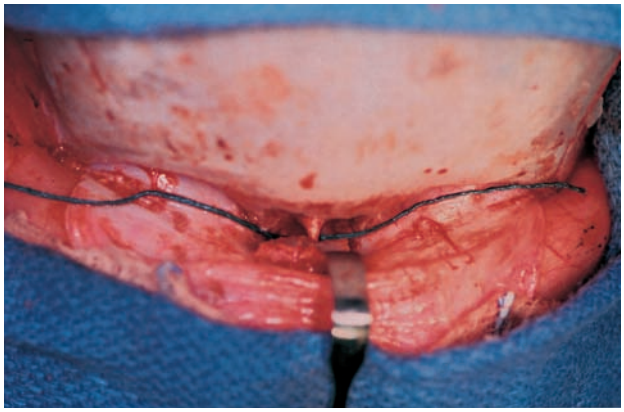


FIGURE 4–28. Our approach of choice to the extension of the dermal sinus tract includes developing a bicoronal flap and exposing the supraorbital rims bilaterally as well as the nasion. Because the lesion will extend into foramen cecum, we perform a mini-"nasional" craniotomy by removing the lower-most part of the glabella and the nasal bone. This exposure will be sufficient for most dermal sinus tracts unless a large dermoid is present intracranially, in which case a frontal craniotomy will be necessary.

elevated, and subpericranial dissection is carried out to the nasofrontal region. This maneuver allows further tract dissection in that area. If intracranial extension is suspected, the neurosurgical team proceeds to create a limited frontal craniotomy. Our method of choice is to elevate the nasal bones and the inferior glabella as a unit as previously described under the section on sincipital encephaloceles (Fig. 4–28). This maneuver allows the surgeon to follow the tract into the dura for complete excision (Fig. 4–29). If, however, a large dermoid tumor is found intracranially, a larger frontal craniotomy may be necessary. Following complete excision of the nasal dermal tract, a free pericranial graft may be used to cover any dural defects (Fig. 4–30). The bone graft is fixated rigidly using miniplates (Fig. 4–31). Excellent postoperative results, with minimal facial scarring, have been obtained (Fig. 4–32).

When approaching occipital dermoids, plans should be made for a posterior fossa craniotomy. Regardless of where the skin dimple may be located (above the inion, below the inion, upper cervical region), the tract most commonly penetrates the bone below the torcular Herophili. Fortunately, the major venous sinuses are not involved or associated with these tracts. Incisions should be vertical to allow full exposure of the posterior fossa.



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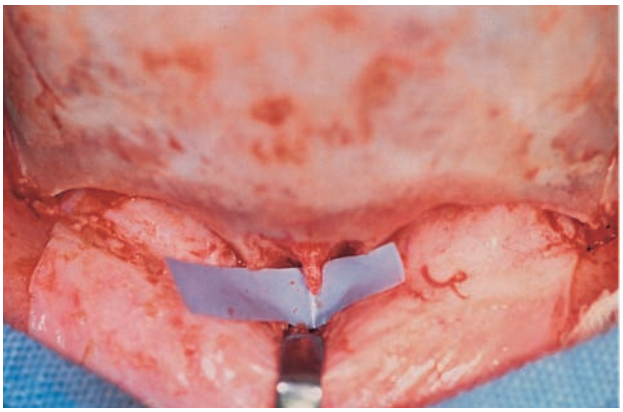

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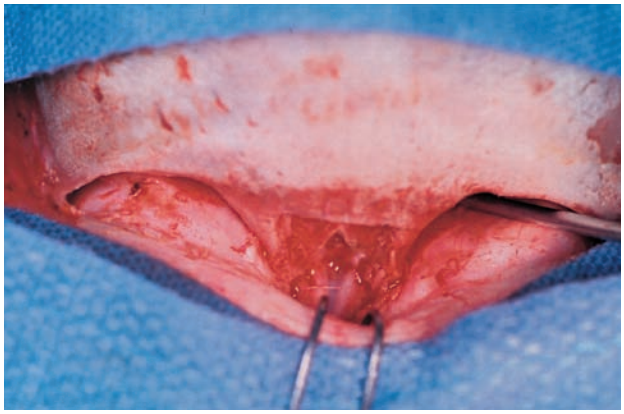

C

FIGURE 4–29. **A:** Following removal of the small bone flap, the tract is easily seen entering the dura at the level of the foramen cecum. **B:** The entire nasal extension of the dermal tract has been excised except for its dural attachment. **C:** In this patient the dermal sinus tract was followed into the dural leaves, thereby removing the epithelial contents that may lead to meningitis or future tumor formation.

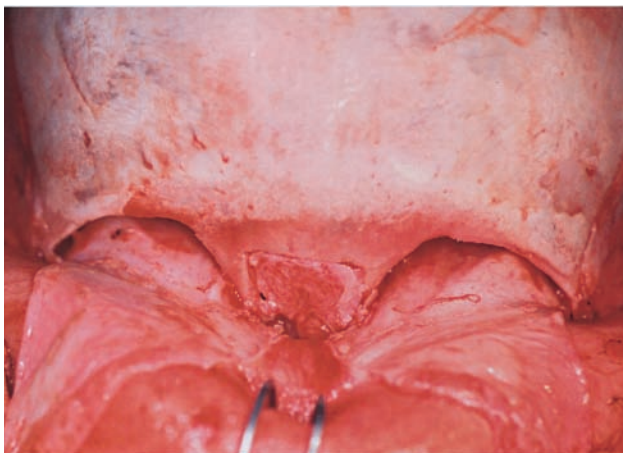

FIGURE 4–30. A free pericranial graft is used to isolate the nasal cavity from the opened dura. Simple interrupted braided nylon sutures are used to secure the graft in place.

An elliptical incision is made around the skin dimple, and circumferential dissection along the tract is done. Care should be taken to keep the tract intact and not to leave any remnants behind. After reaching the bone, a local circumferential craniectomy should be performed to keep the tract intact and to expose the dural side of attachment. If dural penetration is found, or if a dermoid is known from preoperative studies to be present, the dura is opened and intracranial resection of the lesion is carried out. Posterior fossa dermoid tumors usually are located on the vermis between the cerebellar tonsils or even in the area of the fourth ventricle.

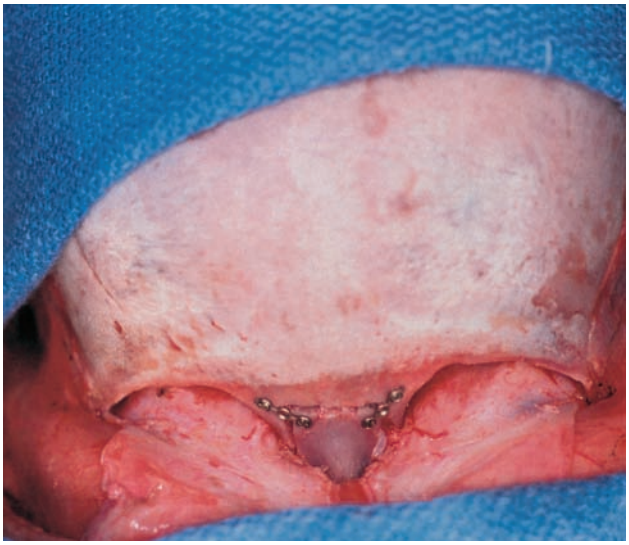

FIGURE 4–31. The bone graft is replaced and rigidly fixated with 1.3 titanium miniplates and screws. Prior to scalp closure, a subgaleal drain is left in place for 24 hours.

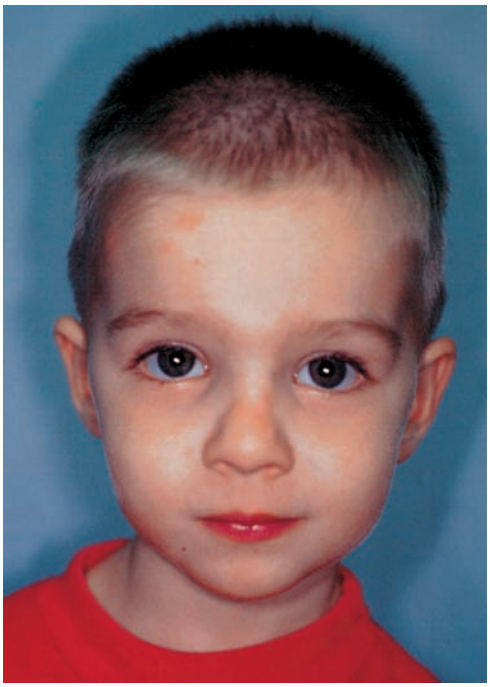

FIGURE 4–32. Postoperative view of patient following complete excision of nasal dermal tract using the nasional craniotomy technique.

Prognosis

Patients with completely resected asymptomatic tracts and dermoid tumors have an excellent prognosis. Incompletely resected tracts place the patient at higher risk for development of meningitis, tract infection, and cystic tumor development. Every attempt should be made at achieving gross total resection of these lesions.

EDITOR'S COMMENTARY

Because of the rarity of sincipital encephaloceles in the U.S., few neurosurgeons have the authors' experience in their management. Consideration should be given in large, complicated sincipital encephaloceles to having them treated in a center with such experience. Their management necessitates the joint involvement of a neurosurgeon and craniofacial surgeon. When treating basal encephaloceles that have a large bone defect, it is probably advisable to fill in the defect, either with calvarium or methylmethacrylate. Otherwise the dural graft may slowly herniate downward through the defect, particularly if hydrocephalus is present. The authors' suggestion to perform MRA before operations for occipital encephaloceles is a good one. These lesions, when large,

may contain large arteries and veins that are easier to deal with if their presence is known beforehand. The need for an expansion cranioplasty is exceedingly uncommon. The suggestion to cover the occipital skull defect with either pericranium or skull is a good one; the skull defect often persists otherwise, particularly if it is greater than 2 cm. The potential need for such a graft should be considered when the incision is planned.

PEARLS

In these authors' experience:

- If there is evidence of nasolacrimal duct obstruction in a patient with sincipital encephalocele, the duct should be cannulated with Silastic tubes and left in place for 6 months to prevent dacryocystitis and reestablish duct patency.
- Sincipital encephalocele patients with ventriculomegaly should undergo ventriculoperitoneal shunt placement prior to encephalocele repair to prevent postoperative CSF leaks.
- Every attempt should be made to preserve "normal-functional" brain and occipital encephaloceles; however, most often, the tissue is fibrous, gliotic, and nonfunctional and therefore can be amputated easily.
- Dermal sinus tracts should be excised "prophylactically," and total excision of the tract and associated dermoids should be done to prevent recurrence.

SUGGESTED READINGS

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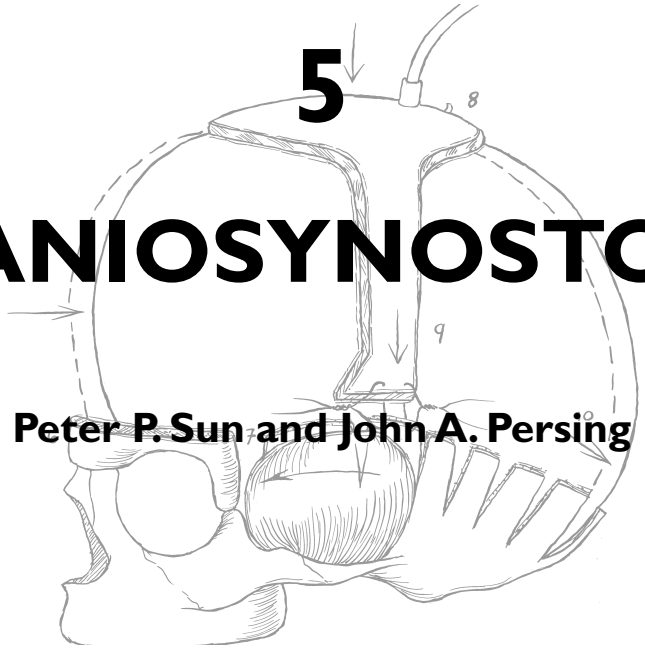
5

CRANIOSYNOSTOSIS



Peter P. Sun and John A. Persing

The treatment of craniosynostosis has evolved dramatically over the last 50 years. Linear-strip craniectomies were the first treatment approaches used to release synostotic cranial sutures. This approach was effective in young children who had not yet developed prominent compensatory cranial vault and base abnormalities. In children who had developed these abnormalities, however, incomplete correction of the skull deformities was seen regularly. Various techniques using foreign interposing material or application of caustic agents to the dura were used to prevent refusion of the suture, the mechanism thought to account for the variable success of the strip craniectomy procedures. Further modifications were developed in reshaping hypoplastic areas of the skull, such as the orbits, to improve the overall appearance. These techniques improved results, but it became clear that the existence of compensatory bulging skull deformities continued to constitute significant residual deformities. To achieve a normal skull form, bilateral and eventually holistic craniofacial approaches have been developed to correct all significant abnormal dimensions. Currently, surgical approaches focus on releasing the synostosis, reshaping the bone, and achieving as normal a correction as possible with direct reconstruction of compensatory deformities.

SURGICAL INDICATIONS

The diagnosis of primary nonsyndromic craniosynostosis is based on the characteristic skull shape: Metopic synostosis causes trigonocephaly, unilateral coronal synostosis causes an anterior plagiocephaly, bilateral coronal synostosis causes brachycephaly (turribrachycephaly), sagittal synostosis causes scaphocephaly, and lambdoid synostosis causes a posterior plagiocephaly. Combinations of these pathologies may be evident if the individual forms occur in combination with another. The deformity is usually present at birth and becomes more exaggerated over time. Skull-shape abnormalities also can result from deformational forces such as compression or positional molding, which are much more common than true craniosynostosis. The overwhelming majority of posterior plagiocephaly is from molding. The skull shapes in these situations differ distinctively from the characteristic shapes of craniosynostosis. Positional (posterior) plagiocephaly is characterized by a parallelogram-shaped head when viewed from above and typically improves as the child develops motor skills to mobilize the head. Radiographic evaluations assist in the diagnosis. The characteristic skull shapes of craniosynostosis are easily recognized on computed tomography (CT) and bony bridging across a suture can be seen. Three-dimensional (3-D) CT reconstruction provides particular clarity in defining the full extent of the deformity.

The recommendation for surgery in craniosynostosis is based on aesthetic and functional considerations. Prominent craniofacial deformities from craniosynostosis can affect a child's psychosocial development significantly. For many parents and surgeons alike, the psychosocial benefits of early corrective surgery outweigh the low risk of modern craniofacial surgery. In addition, there is also evidence that single-suture synostosis can have an effect on intracranial dynamics, and children with uncorrected simple craniosynostosis must be monitored actively for signs and symptoms of elevated intracranial pressure.

The timing of surgery is not standardized, although there is general agreement that an early operative proce-

dure can capitalize on the ameliorating postoperative effects of brain growth on overall skull shape. Compensatory deformities cease to progress after surgery, and remaining minor asymmetries are more likely to be normalized with growth, including skull-base asymmetries. The operation is easier to perform in an immature skull, and there is less postoperative risk if the patient is not yet ambulatory. Consequently, surgery is advised as soon as the infant is best able to tolerate the stresses of surgery, usually at age 3 to 9 months, depending on the magnitude of the surgical intervention and the child's general health. At this age range the calvarial bone has also developed sufficient thickness to undergo surgical remodeling and fixation. If intracranial pressure (ICP) elevations or rapidly progressive deformities are discovered earlier, prompt treatment is indicated at that time. There is some evidence that operating after the age of 1 year may expose the child to a greater risk of mental insufficiency. Most surgical procedures in our unit are done between 3 and 6 months of age. This appears to be an appropriate compromise between the use of the brain's growth to ameliorate skull shape and the ability to operate safely in the more complex procedures and still provide stability of bony support to the restructured skull.

PREOPERATIVE MANAGEMENT

A multidisciplinary evaluation is performed preoperatively in patients with craniosynostosis. Genetic counseling and testing assist the family in understanding the full spectrum of potentially associated problems. Neuropsychologists assess and follow the child's developmental status when there is clinical suspicion of developmental delay (or, in older children, psychological stress). Visual acuity, and the presence of papilledema, diplopia, or strabismus are tested by an ophthalmologist. A social worker or psychological therapist may be instrumental in assessing the family's expectations.

Radiographic evaluation centers on the head CT with 3-D bony reconstruction, which delineates the full extent of the deformity for presurgical planning. Attention is also given to the cerebrospinal fluid (CSF) spaces and ventricular size to rule out coexisting hydrocephalus. When a modified prone position is planned to gain simultaneous access to the anterior and posterior portions of the skull, cervical spine films are taken to exclude the presence of craniocervical abnormalities that might lead to brainstem or spinal cord injury during extension of the neck. In patients with syndromic synostosis or multiple suture involvement, magnetic resonance (MR) imaging is also appropriate to rule out an associated Chiari malformation. If a craniocervical abnormality is discovered, the patient would not be placed in the modified prone position but would be operated on serially on the same day with a two-stage operation, initially with the patient in a supine position and then in a prone position to correct both posterior and anterior abnormalities of the skull.

Standardized photographs are taken preoperatively and serve as a baseline for outcome assessments. Longitudinal follow-up for at least 5 years is recommended. Early assessments of surgical efficacy are not complete until the facial skeleton has undergone additional development, particularly in patients who have asymmetric skull deformities such as unilateral coronal synostosis. With these developments in the facial skeleton, either amelioration of any postoperative deformity or recreation of the preoperative deformity may be seen.

PREOPERATIVE PLANNING

The major intraoperative concern in craniosynostosis surgery is the potential for blood loss and air embolism, which must be anticipated *preoperatively* by securing adequate amounts of blood for the patient and arranging appropriate monitors for surgery. All patients undergoing surgery receive a central intravenous line, large-bore peripheral venous lines, arterial line, Foley catheter, precordial Doppler, and end-tidal CO_2 monitor. The patient is also volume expanded with a 10 to 15 cc/kg crystalloid intravenous bolus prior to skin incision to decrease the risk of air emboli and cardiovascular collapse. Detection of possible air emboli by the Doppler or end-tidal CO_2 is immediately followed by copious irrigation of the field, Trendelenburg positioning, and waxing of all bone edges. Rarely is the blood loss acute and rapid, although it may occur from tears of the sagittal or transverse sinuses. Extreme care must be taken when dissecting the sinus under a nonfused suture because it is regularly adherent. A more common observation is that there is a continuous low-grade, small amount of blood loss that occurs throughout the operative procedure. Because the circulating blood volume of the infant is relatively meager, even small amounts of blood loss assume greater importance. The surgeon can minimize blood loss by injecting local anesthetic with epinephrine in the anticipated incision line to achieve meticulous homeostasis on opening, dissecting in the supraperiosteal plane to minimize blood loss from the bone, and waxing all edges of bone when cut. Prompt replacement of fluid losses is also critical to maintaining hemodynamic stability. Controlled hypotension also can be employed intraoperatively to minimize blood loss.

Nevertheless, blood replacement is routine. Intraoperatively, the coagulation status and platelet count are monitored. A continuing dialogue between the surgeon and anesthesiologist is helpful for optimal intraoperative management.

INTRAOPERATIVE TECHNIQUES

Surgical technique is tailored to the type and severity of craniosynostosis as well as to the age of the patient. Various techniques have been described for each type of craniosynostosis. The occasional patient younger than 6 weeks of age with a mild deformity may be treated adequately by a limited suture resection, particularly those with sagittal synostosis. In most cases, however, suture resection alone will not yield a satisfactory result. The compensatory skull shape and associated fronto-orbital abnormalities produced by the fused suture have become the predominant deformities. Therefore, *actively* reshaping the deformities with a more extensive reconstruction frequently provides the best results. This includes supraorbital rim advancement in conjunction with bifrontal craniotomy in cases of metopic and coronal synostosis, for which the deformity involves the fronto-orbital region.

Initial Exposure

No hair is shaved in any patient. The prepared area includes the orbital region in patients with coronal or metopic craniosynostosis where the deformity extends to the orbits. Care is taken to protect the cornea with shields or lubricants. In this way, the reconstruction can be evaluated by pulling the scalp back over the skull and orbits intraoperatively and examining the overall fronto-orbital soft-tissue profile. A coronal incision line is used for all patients (Fig. 5–1A). We extend the inferior portion obliquely posterior to the ear and then angle it anteriorly to the vertex of the skull (Fig. 5–1B). This incision allows simultaneous access to the anterior and posterior aspects of the skull, regardless of the skull deformity, and conceals the incisional alopecia. The incision should be beveled in the direction of the hair follicles to avoid amputating hair shafts and creating a wide scar. Dissection is done in the supraperiosteal plane to minimize blood loss, because stripping of the periosteum leads to significantly more bleeding from the bone. Leaving the periosteum attached to the underlying bone also assists in holding the bone together if it fractures during later remodeling. In metopic and coronal synostosis, exposure of the supraorbital region is needed for the supraorbital rim osteotomy. In these cases, an incision is made in the periosteum paralleling the rim. Dissection proceeds directly on the bone 1 cm above the supraorbital rim (Fig. 5–1C), thus allowing the supraorbital groove and its nerve to be visualized and separated. Subperiosteal dissection is carried to the level of the nasofrontal suture medially and the frontozygomatic suture laterally, exposing the entire supraorbital rim in preparation for the orbital rim osteotomy. Care is taken not to detach the medial canthus. The temporalis muscle is exposed in a suprafascial fashion and not detached from the underlying bone.

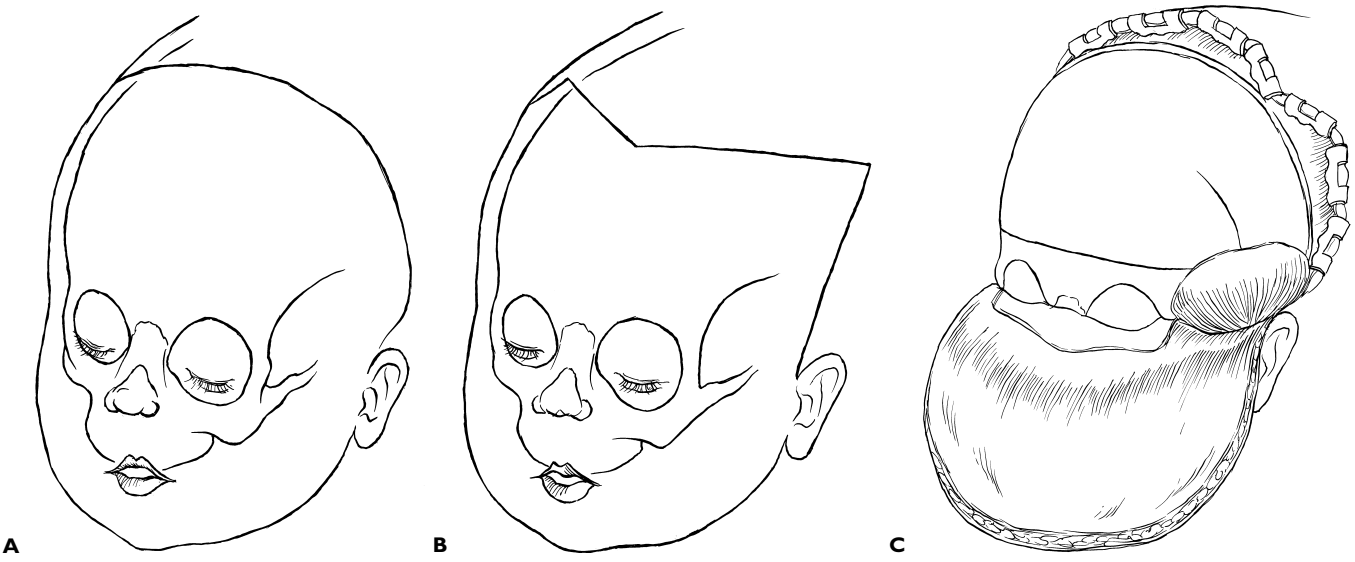


FIGURE 5–1. Scalp incisions. **A:** Bicoronal skin incision. **B:** Stealth incision. **C:** Subperiosteal dissection in the supraorbital region beginning at 1cm superior to the orbit.

Orbit Osteotomy

The orbital osteotomy is performed after a bifrontal craniotomy, which extends approximately 5 mm above the supraorbital rim (Fig. 5–2A). The dura is separated from the anterior frontal fossa to the level of the cribriform plate dural attachments and also dissected off the lateral greater sphenoid wing in the anterior temporal fossa in preparation for the osteotomies. A side microreciprocating saw or osteotome then is used to perform an orbital roof osteotomy approximately 5 to 10 mm posterior to the orbital rim. Malleable retractors are

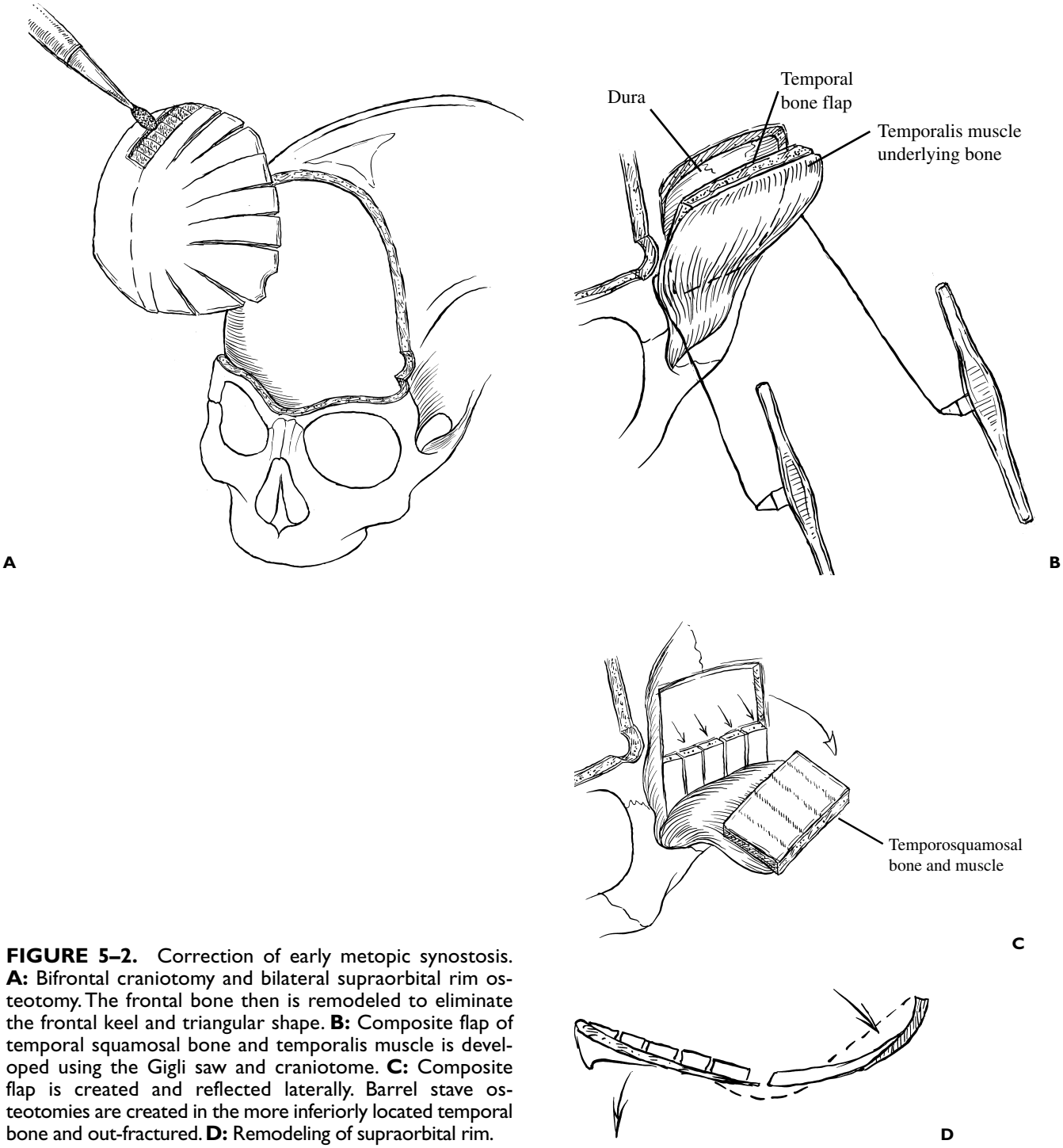


FIGURE 5–2. Correction of early metopic synostosis. **A:** Bifrontal craniotomy and bilateral supraorbital rim osteotomy. The frontal bone then is remodeled to eliminate the frontal keel and triangular shape. **B:** Composite flap of temporal squamosal bone and temporalis muscle is developed using the Gigli saw and craniotome. **C:** Composite flap is created and reflected laterally. Barrel stave osteotomies are created in the more inferiorly located temporal bone and out-fractured. **D:** Remodeling of supraorbital rim.

used to protect the brain and orbital contents. The osteotomy extends medially just anterior to the cribriform plate and laterally through the lateral wall of the orbit to the frontozygomatic suture. An osteotome is used to complete the fractures at the level of the frontozygomatic suture and the frontonasal suture. Both supraorbital rims are removed as a single unit for later contouring.

Temporal Region

Postoperative temporal hollowing has been a cosmetic problem after detachment of the temporalis muscle and orbital advancement in the treatment of coronal and metopic craniosynostosis. Consequently, a composite temporalis advancement was devised. Instead of dissecting the temporalis muscle off the bone, a composite of the temporal squamous bone and muscle is elevated in an osteoplastic fashion. After a bifrontal craniotomy above the temporalis, two 2.0- to 2.5-cm vertical incisions are made into the temporalis muscle and squamous temporal bone: anteriorly at the greater wing of the sphenoid and posteriorly at the level the external auditory canal. The sphenoid wing usually needs to be partially resected with a rongeur. After the dura is freed of the inner table of the cut bone, a Gigli saw is slid down the two vertical grooves and used to undercut the base of the squamous temporal bone while preserving the overlying temporalis muscle (Fig. 5–2B). This composite myoosseus unit then is subsequently attached to the advanced orbital rim to prevent postoperative temporal hollowing.

Bone Remodeling Techniques

The techniques for reconstruction of the skull differ for different age groups. For patients younger than 1 year of age, the bone is thinner and more malleable. An abnormally convex surface may be made flat by performing radial osteotomies into the center of the bone. In an area that is abnormally flat and needs to be made more convex, radial wedge resections along the radial osteotomies are made. The bone then can be remodeled appropriately by gentle bending with a Tessier rib bender. Cranial defects are consistently regenerated during this time. The development of absorbable plates and screws now permits the use of rigid fixation in this age group. Permanent metallic rigid fixation is not preferred because endosteal displacement of hardware and the possibility of limiting brain growth are to be avoided. In patients older than 1 year of age, the bone of the skull is thicker, much more brittle, and more difficult to remodel. The best reconstructive strategy may be switching different osteotomy segments, substituting, for instance, in metopic synostosis, an appropriately contoured parietal skull for the triangle-shaped forehead. The more triangle-shaped forehead then may be weakened by creating channels or kerfs in the undersurface of the skull and with gentle "green stick" fracturing of the bone to a more flat configuration to serve as the posterior frontal skull. In some cases, an appropriate contour can be found by dividing an osteotomy segment and rotating the two pieces. If, however, there is no suitable substitute for the forehead bone, remodeling of the original frontal bone may be done by cutting it into smaller slats and remodel-

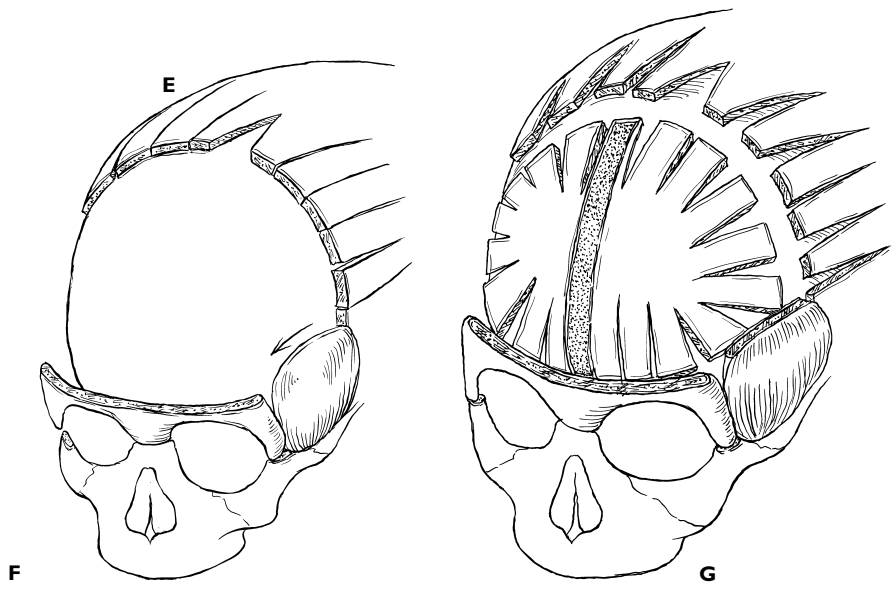


FIGURE 5–2. (*continued*) **E:** Parallel osteotomies in the parietal region for lateral expansion. **F:** The supraorbital rim is advanced and fixed to the glabella. The composite flap is then brought forward to the advanced orbital rims. **G:** Reshaped frontal bone is attached to the underlying dura and supraorbital rim.

ing these using kerfs or selectively placed weakening points (channels) on the endocranial surface of the bone. The thicker bone cannot be remodeled satisfactorily as one piece with radial cuts or wedge resections. Sizable cranial defects should be filled with bone grafts because they will persist.

TREATMENT FOR INDIVIDUAL FORMS OF CRANIOSYNOSTOSIS

Metopic Synostosis

The surgical objectives in patients with metopic synostosis are to reduce the prominence of the midline forehead bone, to release the synostosis at the metopic suture, to reduce to normal the trigonocephaly over the forehead region, to advance the supraorbital rims laterally to a normal projection, and to advance the temporal myoosseous units forward to prevent temporal depression.

In children younger than 1 year, the patient is placed supine, and the coronal incision is made to expose the frontal and orbital regions. A bifrontal craniotomy from just anterior to the coronal suture to 1 cm above the supraorbital rim is performed. The anterior fontanel is usually patent; therefore, craniotomy is based on the anterior fontanel as an entry point, followed by a bilateral supraorbital rim osteotomy with an oblique cut at the frontozygomatic suture (Fig. 5–2A). The supraorbital rim is remodeled to a more convex shape with restored lateral frontal projections, and the frontal bone is remodeled to eliminate the frontal keel and to achieve more lateral frontal prominence (Figs. 5–2A and D).

In the temporal region, the composite resection is performed. The remaining basal squamosal temporal bone is split into vertical "barrel staves" and out fractured, thereby increasing the temporal flare (Fig. 5–2C). The remodeled supraorbital rim is advanced and fixed to the glabella, taking care to achieve the desired frontal nasal angle of approximately 20 to 30 degrees. The advancement is anchored by placing the lateral frontal bone of the supraorbital rim in front of the lateral zygomatic process (Fig. 5–2F). The composite temporal bone and muscle flap are brought forward and attached to the advanced orbital rims with absorbable plates. The advancement achieved is usually between 12 and 17 mm. At the coronal suture, adjacent bone is removed, and the parietal region is expanded by parallel osteotomies (Fig. 5–2E). The reshaped frontal bone is attached to the supraorbital rim (Fig. 5–2G). Occasionally, a prominent frontal keel exists without significant orbital retrusion or temporal narrowing. These cases can be treated simply by burring down the keel without an extensive reconstruction.

In children older than 1 year, who have more mature bone, the frontal graft can be remodeled by splitting it into vertical slats with kerfs cut on the undersurface (Fig. 5–3). Alternatively, a second bone graft can be harvested with a biparietal craniotomy to substitute for the malformed frontal bone. The biparietal bone provides a smooth, round forehead without the need for extensive reshaping. With this biparietal bone segment, the proper amount of flare and flattening of the forehead usually is achieved. This segment should be cut to equal the height of the forehead which is usually 4.5 to 5.5 cm. Split calvarium may be needed to fill in bone gaps.

No effort is made to correct the hypotelorism during early surgery. It is usually mild and partially self-correcting. If hypotelorism is pronounced in an older child, however, correction may be warranted. The supraorbital rim can be widened by splitting it at the midline and fixating an intervening free bone graft. By age 9 to 12 months, the lateral intercanthal distance should be 85 to 100 mm. Additionally a 3-cm inferior nasofrontal osteotomy with a wedged bone graft can be performed to increase the medial intercanthal distance. Of the various types of craniosynostosis, metopic synostosis has the most consistently favorable results after surgery. Reoperation is less common than after early supraorbital advancement and frontal reshaping.

Unilateral Coronal Synostosis

Treatment goals for patients with unilateral coronal synostosis are to remove the stenosis of the coronal suture, to provide symmetry to the forehead, to advance the supraorbital rim on the side of the fused suture, to normalize the shape of the orbits, to reduce the compensatory bulging on the contralateral frontal bone, and to advance the temporalis myoosseous unit ipsilateral to the fused suture to prevent hollowing of the temporal region following orbital rim advancement. The associated nasal deviation is not corrected because it usually improves with further development.

In children younger than 1 year, the patient is placed in a supine position, and a bicoronal incision is made. A bifrontal craniotomy is performed, taking care not to detach the temporalis muscle (Fig. 5–4A). A bilateral orbital roof osteotomy with lateral cuts at the frontal zygomatic sutures follows. The orbital rims not only are recessed in unilateral coronal synostosis, but they are asymmetric. A drill is used to contour the orbits to symmetry and the orbital bar is outfractured with weakening

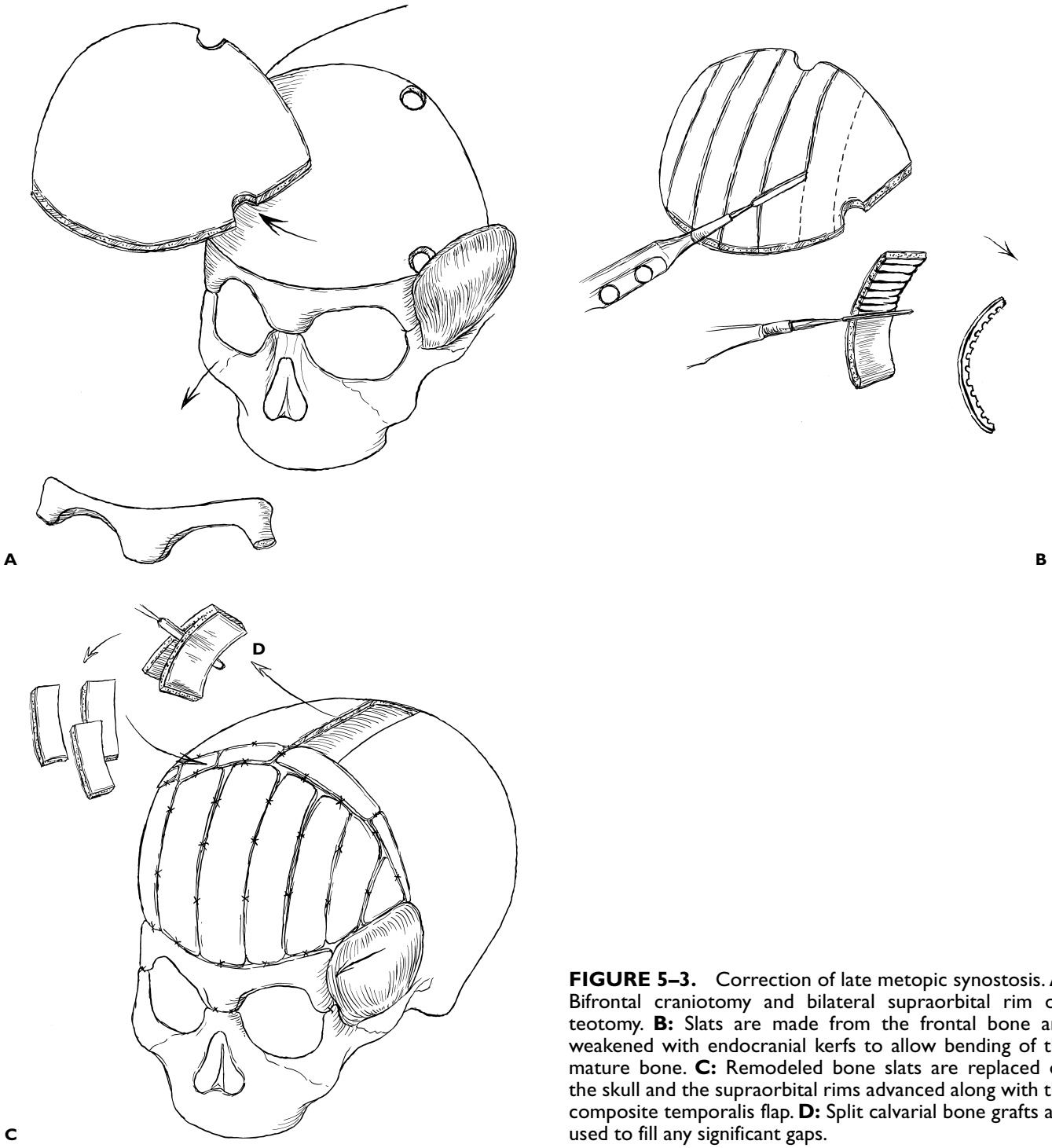


FIGURE 5–3. Correction of late metopic synostosis. **A:** Bifrontal craniotomy and bilateral supraorbital rim osteotomy. **B:** Slats are made from the frontal bone and weakened with endocranial kerfs to allow bending of the mature bone. **C:** Remodeled bone slats are replaced on the skull and the supraorbital rims advanced along with the composite temporalis flap. **D:** Split calvarial bone grafts are used to fill any significant gaps.

kerfs to restore the normal projection (Figs. 5–4B and C). The ipsilateral pterion is resected to the frontosphenoid suture, and a composite temporalis flap is elevated ipsilateral to the fused suture. The reshaped symmetric supraorbital rim is advanced, secured at the midline and sutured laterally in front of the frontal process of the zygoma (Fig. 5–4D). The advancement should aim for overcorrection. A composite temporalis flap is created on the ipsilateral advancement side and is attached to the advanced supraorbital rim. The bifrontal bone graft is contoured to increase the projection on the flattened side and to reduce the projection of the bulging side. The re-

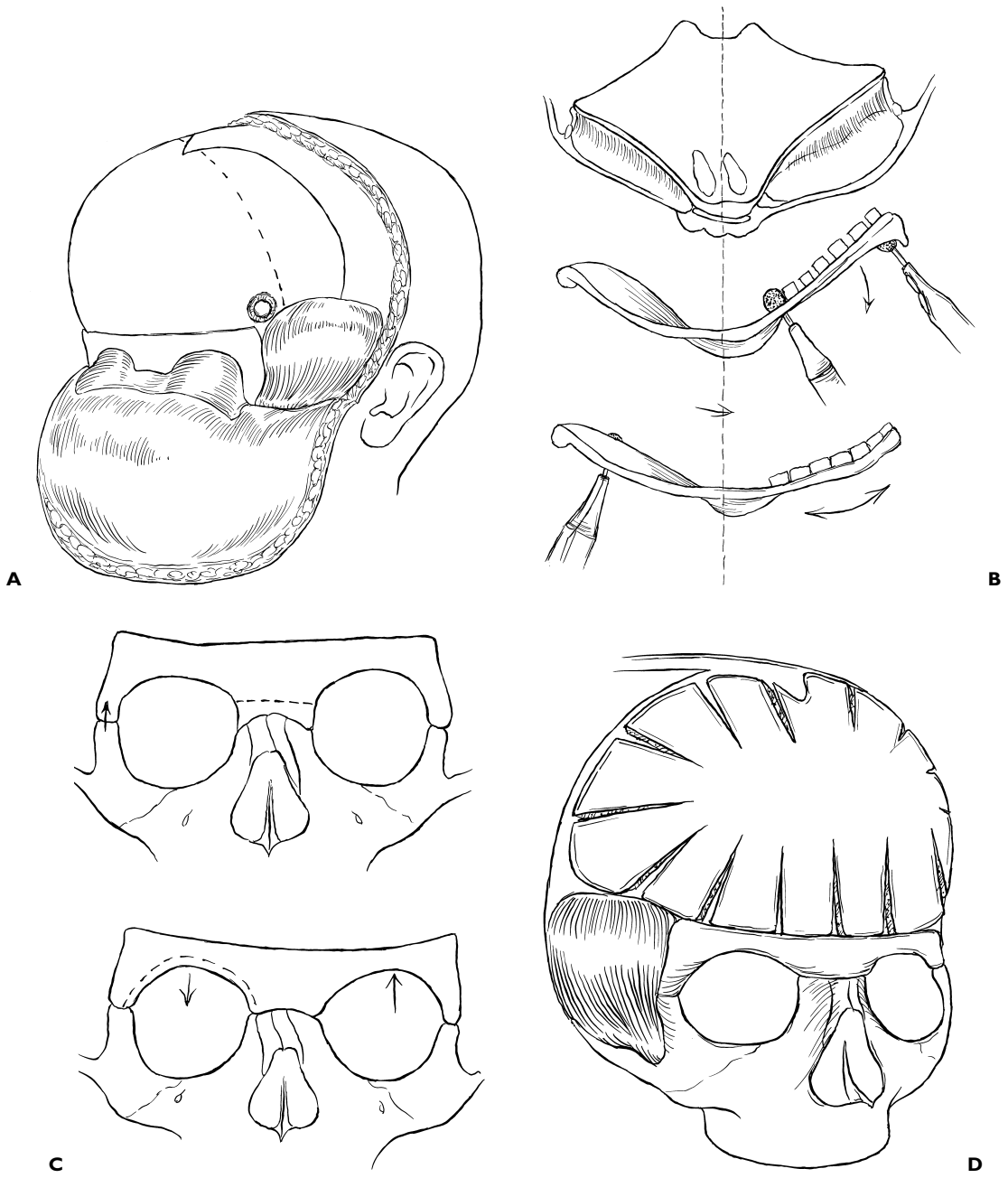


FIGURE 5–4. Early correction of unilateral coronal synostosis. **A:** Bifrontal craniotomy and bilateral supraorbital rim osteotomy. **B, C:** The supraorbital rim is contoured to equalize the frontal projection and orbital dimensions. **D:** The contoured supraorbital rim is advanced with the composite flap.

modeled frontal bone is attached to the advanced supraorbital rims.

In children older than 1 year of age, a bilateral orbital rim advancement is generally performed as well. Occasionally, the osteotomy of the orbit extends into the body of the zygoma, creating a C-shaped orbital advancement (Figure 5–5). This is done when it is perceived that the anterior projection of the zygoma is deficient ipsilateral to the fused suture. This situation is not usually characteristic of nonsyndromic unilateral coronal synostosis. Parietal bone grafts are harvested to insert into the zygoma and support the advancement. Care is taken to ensure orbital symmetry by contouring the inner aspects of the orbits. Temporalis composite flaps are attached to the advanced orbits. The bifrontal bone graft is reshaped by splitting it into slats with underlying kerfs, or a separate

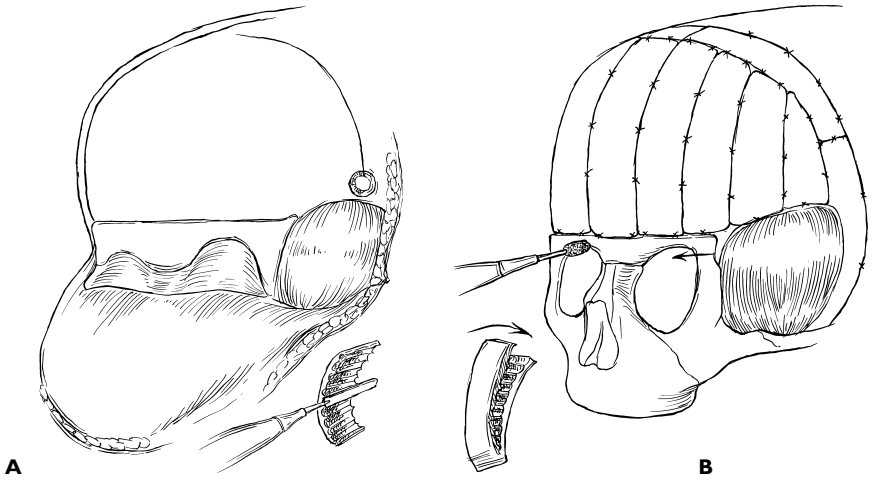


FIGURE 5–5. Late correction of unilateral coronal synostosis. **A:** Bifrontal craniotomy with composite flap outlined. **B:** Supraorbital rim may be advanced ipsilaterally by a C-shaped orbital osteotomy; the contralateral orbit is contoured to equalize the orbital dimensions.

biparietal bone graft is harvested to recreate the forehead, as in metopic synostosis.

Good to excellent results have been achieved in about 70 to 80% of early unilateral coronal synostosis reconstructions using various methods of supraorbital advancement and forehead reconstruction. The difference in techniques mostly involves the method of supraorbital advancement. The bilateral approach outlined above is designed to avoid the orbital asymmetries, temporal hollowing, and inadequate supraorbital rim advancement, which typifies the unsatisfactory outcomes.

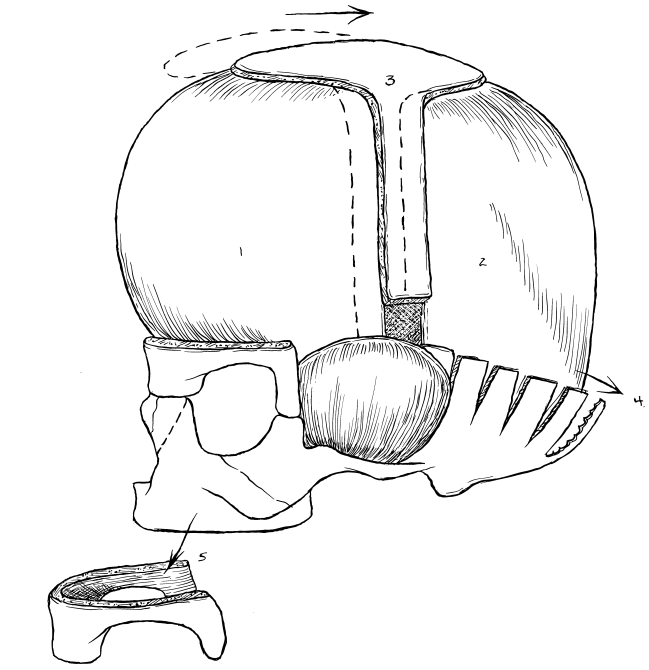
Bilateral Coronal Synostosis

The treatment of bilateral coronal synostosis skull deformities is challenging. It is directed at increasing the anteroposterior dimension of the skull, reducing the superior-inferior dimension of the skull, releasing the fusion of the coronal sutures, and advancing the supraorbital rims bilaterally. ICP monitoring is regularly used.

In children younger than 1 year of age, with bilateral coronal synostosis, because specific anterior and posterior skull deformities are characteristic of this form of craniosynostosis, a modified prone position is elected. Once appropriate preoperative imaging has excluded any craniocervical abnormalities, appropriate candidates are placed in a modified prone position on a well-padded headrest; a beanbag usually is used to support the body position. A combination of foam supports and the anterior portion of a small Philadelphia collar allows support of the head with the neck extended to visualize simultaneously the anterior and posterior aspects of the skull. At this point, coronal incision is carried out. Supraperiosteal dissection is carried out anteriorly to the level of the superior orbital rims and posteriorly to the inferior nuchal line. Visualization of the skull deformity is now evident.

Bilateral frontal and parietooccipital craniotomies are performed. Particularly careful epidural dissection is performed posteriorly over the sinus as the craniotomy extends just beyond the inion. Multiple burr holes frequently are made with a handheld burr to ensure adequate separation prior to the craniotomy. The craniotomy leaves a strip of bone over the vertex and two lateral struts from the vertex to the temporal squamosal region (Fig. 5–6A). Barrel stave osteotomies are made in the occipital region and fractured posteriorly to increase the anteroposterior dimension of the skull. Bilateral supraorbital rim advancement and bilateral temporalis composite advancement are performed. The struts extending from the vertex are reduced in height and shifted posteriorly. Wires are passed between the inferior tip of the struts and the posterior basal temporal bone and gradually cinched down to reduce the height of the skull (Fig. 5–6B). ICP monitoring is used during this time to guide the rate and degree of height reduction so that the pressure does not extend beyond 20 mm Hg except transiently. It must return to normal rapidly in a matter of seconds. Typically, 1 to 1.5 cm of height reduction can be achieved. The frontal and parietooccipital bone flaps are reshaped to be more convex. The frontal bone is attached to the supraorbital rim and the parietooccipital graft is left floating posteriorly. The parietooccipital bone is solely attached to the dura, not to surrounding bone.

In children older than 1 year of age, the height reduction and barrel stave osteotomies are performed using a technique similar to that used in the younger patient. The height reduction, however, is less and re-



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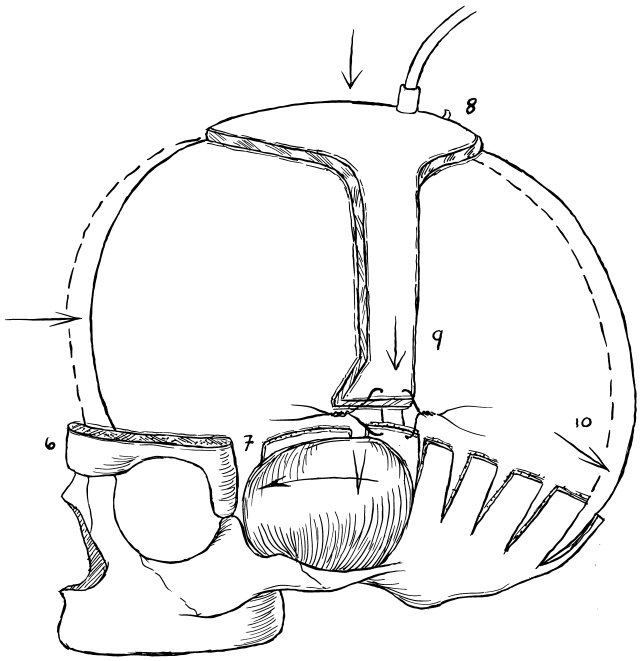

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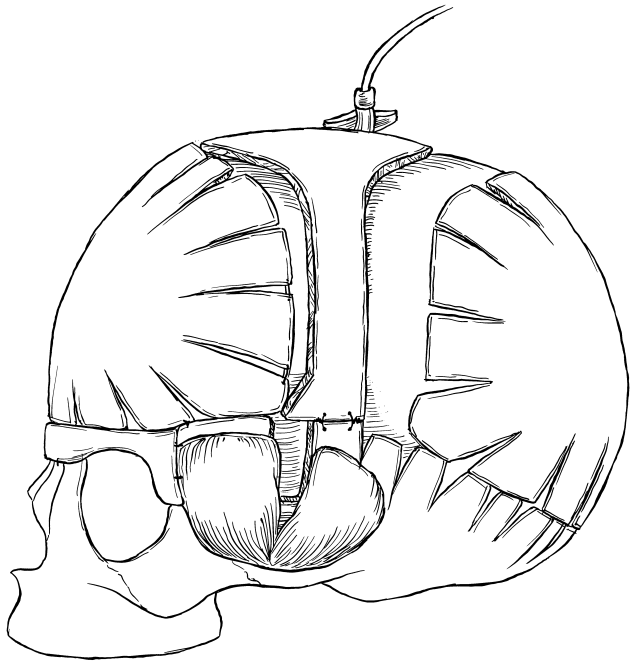

C

FIGURE 5–6. Early correction of bicoronal synostosis. **A:** Osteotomies: *1,* bifrontal craniotomy; *2,* biparietal occipital craniotomy; *3,* struts severed inferiorly with the amount of bone removed equal to the anticipated height reduction; *4,* barrel stave osteotomies in the occiput; *5,* bilateral supraorbital rim osteotomy. **B:** Height reduction: *6,* bilateral supraorbital rim advancement; *7,* composite flap advancement; *8,* ICP monitor; *9,* dynamic height reduction by slowly cinching wire struts; *10,* occipital prominence achieved. **C:** Completed reconstruction.

quires a much longer time of ICP accommodation. Bilateral C-shaped orbital osteotomies can be performed to advance the lateral orbits as well if there is pronounced malar retrusion as seen in some forms of syndromic coronal synostosis. The bone flaps require the usual slats and kerfs to be remodeled (Fig. 5–7). Bony defects should be filled in by bone grafts.

Turribrachycephaly has been notoriously difficult to correct, and reoperations are common in attempts to achieve an acceptable skull shape. Several other effective techniques have been proposed, and all represent variations of a 360-degree circumferential calvarial vault remodeling. More limited procedures fail to address adequately the salient height and width deformities.

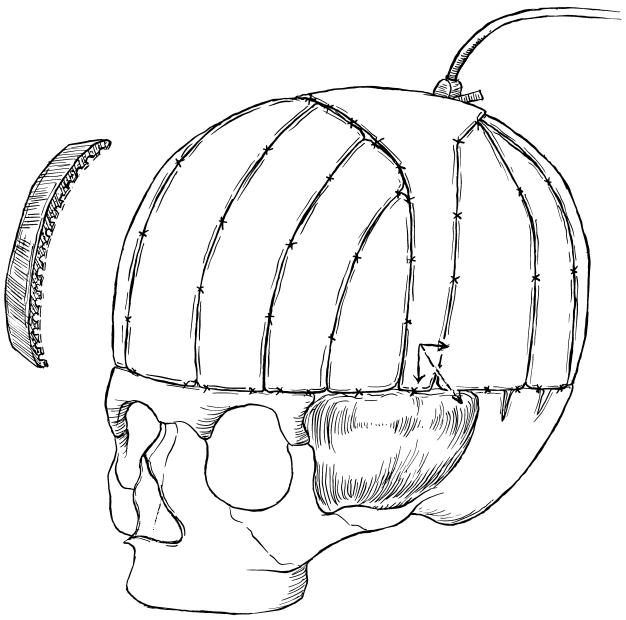

FIGURE 5–7. Late correction of bilateral coronal synostosis using slats remodeled with kerfs.

Sagittal Synostosis

Treatment goals of a patient with sagittal synostosis are to release the stenosis of the prematurely closed sagittal suture, to increase the mediolateral dimension of the skull, and to reduce the anteroposterior dimension of the skull. Locally prominent compensations usually occur in the superior frontal and the occipital regions, both of which regularly require direct reshaping.

In children younger than 1 year, the patient is placed in the modified prone position and a coronal incision is made to expose the entire vault. Supraperiosteal dissection is to the superior orbital rims anteriorly and just inferior to the inferior nuchal line posteriorly. Bifrontal, bioccipital, and individual parietal craniotomies are performed (Fig. 5–8). The extent of the frontal and occipital bone flaps reflects the areas of compensatory frontal bossing and occipital keel. The ridge over the sagittal suture usually is shaved at this point to achieve a more normal contour with the adjacent bone. Barrel-stave osteotomies are performed in the temporal regions and out-fractured to increase the width from the level just posterior to the supraorbital rim between the greater wing of sphenoid and the petrous temporal bone. The bifrontal and bioccipital bone grafts are reshaped to a flatter plane with radial osteotomies and attached to the glabella and inferior occiput. The bifrontal bone is attached to the glabellar region in a more posteriorly angled position to eliminate the frontal bossing, and the lateral frontal-supraorbital triangular overlap is resected. The vertex bone is shortened approximately 1.5 cm and wired to the attached bifrontal and bioccipital bones, thereby shortening the anteroposterior dimension. The two parietal bone grafts are reshaped to a more convex form and sutured to the dura. Wide separations of 1 to 2cm are left around the parietal bone grafts to reduce the likelihood of refusion.

As an alternative, young patients (i.e., younger than 3 months) with mild scaphocephaly and minimal frontal bossing can be treated effectively with a more limited "clam-shell" craniectomy procedure. A bicoronal incision is made with the patient in the prone position in a cerebellar headrest. Supraperiosteal dissection is carried anteriorly to the coronal suture and posteriorly to the inion. Large temporoparietal bone flaps are created by removing a 2.5-cm C-shaped rim of bone anteriorly at the coronal suture, superiorly next to the fused suture, posteriorly in front of the asterion, and then out-fractured at the temporal base on both sides. The strip of fused suture left on top then is removed, leaving the outfractured temporoparietal flaps like an open clam shell. Prominent occipital keels can be resected or remodeled at the same time.

In children older than 3 years, large bone flaps may not be reshaped easily. Consequently, serial bifrontal bifrontoparietal, biparietal, and occipital bone grafts are made (Fig. 5–9). Underlying kerfs are made to reshape the bones more readily. The frontal bone is angulated posteriorly to eliminate the frontal bossing. If the frontal bossing is severe, the bifrontoparietal or biparietal bone graft may be used for the forehead instead, in which case the segment should correspond to the height of the forehead (4.5–5.5cm). Along with the posterior angulation of the forehead, one or two serial flaps is shortened so that there would be a 1.5- to 2.0-cm reduction in the overall anteroposterior dimension of the skull. Barrel-stave osteotomies are performed in the temporal region and outfractured.

The more extensive calvarial remodeling procedures are obviously longer, involve greater blood loss, and require more invasive monitoring. They have minimal morbidity and better cosmetic results than the more limited procedures in most patients, however.

Lambdoid Synostosis

In a patient with lambdoid synostosis, the fused suture abnormality is usually unilateral. Rarely does bilateral lambdoid synostosis occur in isolation. In unilateral lambdoid synostosis, the major goals of treatment are to

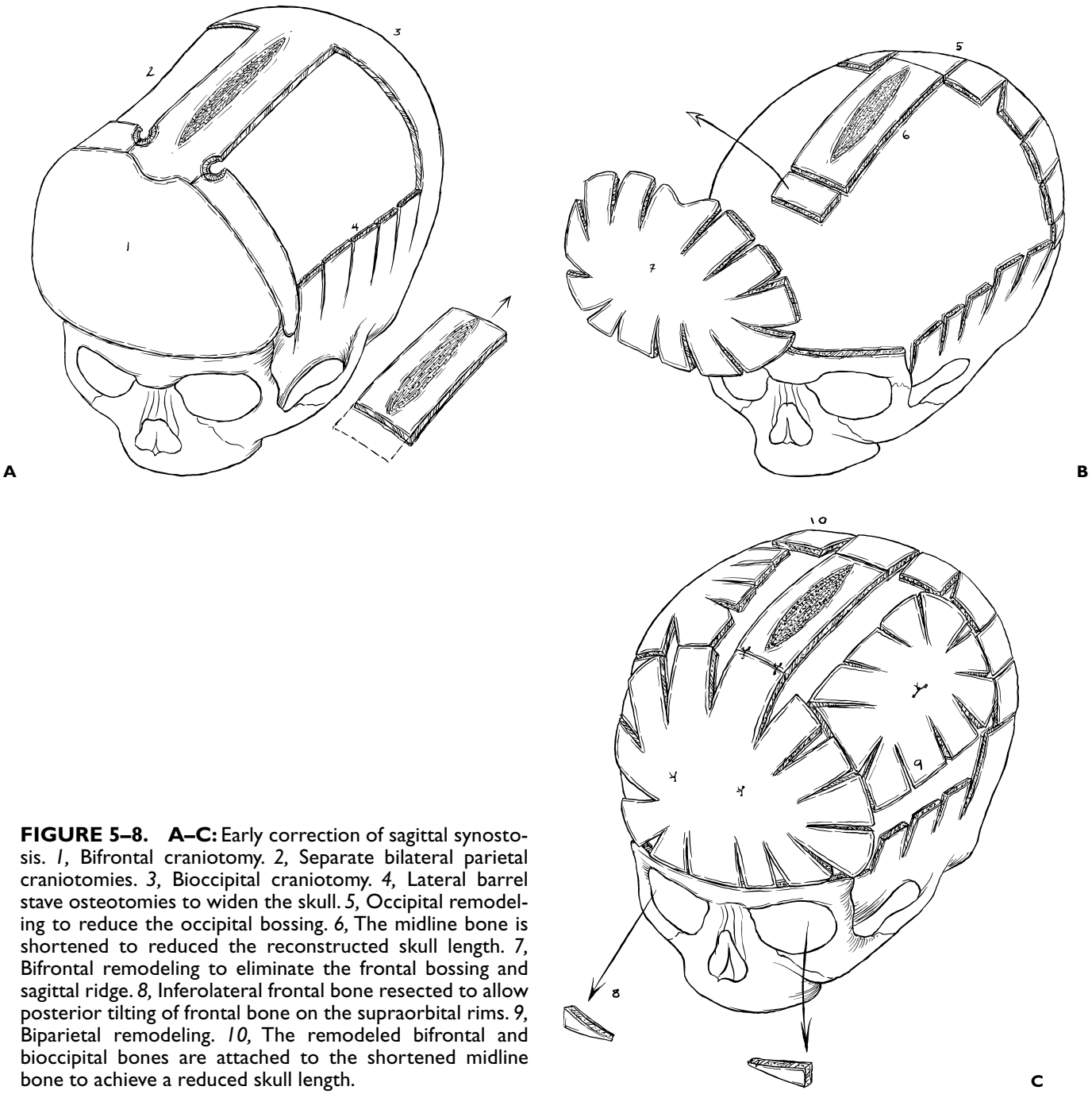

FIGURE 5–8. A–C: Early correction of sagittal synostosis. *1,* Bifrontal craniotomy. *2,* Separate bilateral parietal craniotomies. *3,* Bioccipital craniotomy. *4,* Lateral barrel stave osteotomies to widen the skull. *5,* Occipital remodeling to reduce the occipital bossing. *6,* The midline bone is shortened to reduced the reconstructed skull length. *7,* Bifrontal remodeling to eliminate the frontal bossing and sagittal ridge. *8,* Inferolateral frontal bone resected to allow posterior tilting of frontal bone on the supraorbital rims. *9,* Biparietal remodeling. *10,* The remodeled bifrontal and bioccipital bones are attached to the shortened midline bone to achieve a reduced skull length.

make more convex the flattened parietal and occipital bone adjacent to the fused lambdoid suture and to reduce the bony abnormality of protuberant occipital bone contralaterally. In bilateral lambdoid synostosis, the occiput is flattened bilaterally, and the goal is to increase the convexity posteriorly and bilaterally. Both can be approached similarly.

In children younger than 1 year, the patient is placed in a prone position, and a coronal incision is carried out posterior to the ear. A biparietal occipital craniotomy is performed superior to the inion. Careful epidural dissection to avoid the torcula and transverse sinuses is performed prior to the craniotomy. Barrel-stave osteotomies are performed in the basiocciput and out-fractured on

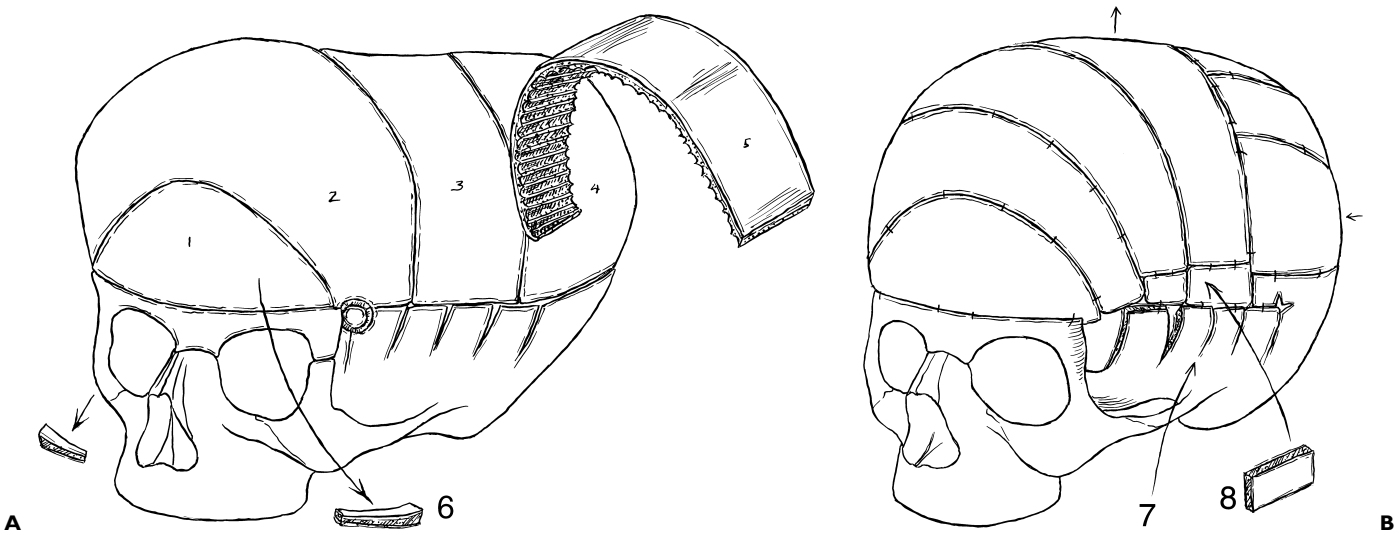

FIGURE 5–9. Late correction of sagittal synostosis. **A:** Serial bone grafts: *1, 2, 3, 4, 5.* Bifrontal bone tilted posteriorly following removal of lateral frontal wedges *(6)*; **B:** Lateral barrel stave osteotomies *(7)*. Bone grafts inserted to increase parietal convexity *(8)*.

the flattened side to increase projection and in-fractured on the bulging side (Fig. 5–10). The biparietal occipital bone flap is reshaped by radial osteotomies to achieve more convexity and symmetry. To maintain a convex posterior projection, absorbable plate fixation is useful, and the lateral head position in the early postoperative period is beneficial. Skull helmets may be fashioned to assist in this procedure. In bilateral lambdoid synostosis, the barrel staves all are out-fractured, and the biparietal occipital flap is reshaped to gain convexity.

In children older than 3 years, basiocciput barrel-stave osteotomies and biparieto-occipital bone flaps are similarly performed; however, an additional biparietal bone flap may be harvested to substitute for a rounded occiput, and the biparietal occipital bone flap may be used to cover the parietal areas.

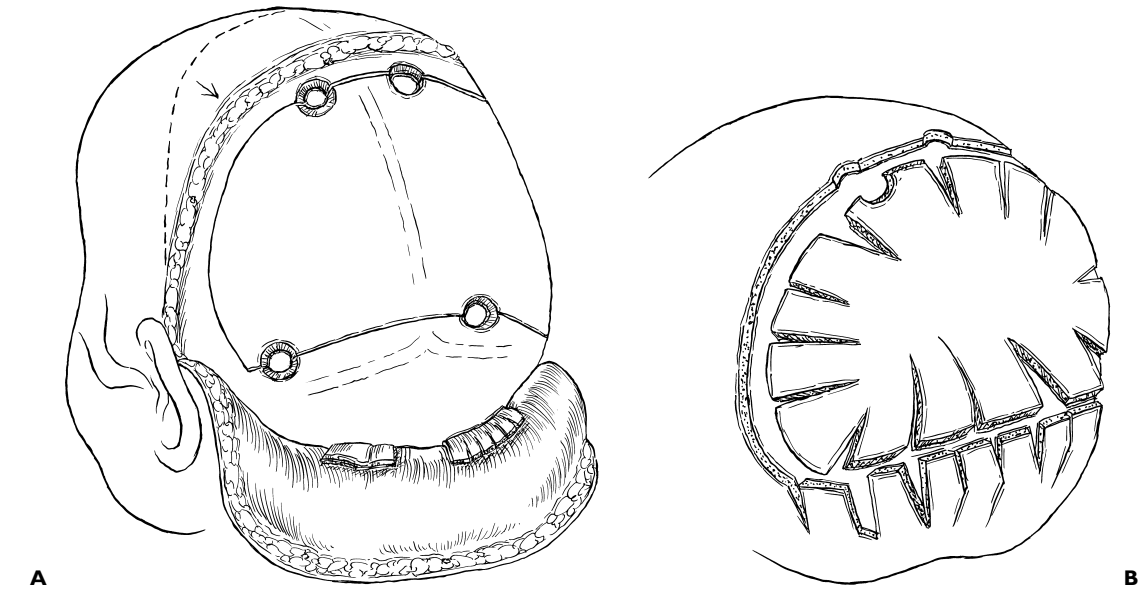

FIGURE 5–10. Correction of lambdoid synostosis. **A:** Biparietooccipital bone flaps. **B:** Asymmetry recontoured.

POSTOPERATIVE MANAGEMENT

Surgery for craniosynostosis is generally quite safe. Complications are minor and usually are related to concerns about blood loss. Blood loss should be monitored in the intensive care unit setting postoperatively. As the child ages, the loss in circulating volume underneath the galea may be proportionately less than in a younger child and will not be of the same concern. Due diligence is important in all patients who are undergoing surgery. Postoperative monitoring with frequent serial hematocrits to determine the stability of the blood (circulating) volume in the early postoperative period is usually most appropriate. We have not used drains postoperatively on a routine basis largely because of concerns related to infection. We use preoperative antibiotics to try to diminish the likelihood of infection; to date, however, no wound-site infections have been encountered. Neurologic complications are rare.

EDITOR'S COMMENTARY

Although the management of craniosynostosis is one of the more common problems faced by the pediatric neurosurgeon, the optimal treatment strategies for the different types of synostosis remain a topic of spirited controversy. For example, in the management of sagittal synostosis in infants, many surgeons perform simple "suture-ectomy", others add various techniques for modest remodeling, and others perform major cranial vault reconstructions. Similarly, for nonsyndromic coronal and metopic synostosis, issues regarding the timing of intervention and the extent of the surgical procedure that is needed to achieve good cosmetic results are subjects of debate. Because the primary indication for intervention in most cases of single-suture synostosis is a cosmetic one, efforts to minimize morbidity assume particular importance. There is even more interinstitutional variability in the treatment of children with multisutural synostosis, a group that poses the greatest long-term management challenges. Notwithstanding these areas of controversy, most pediatric neurosurgeons now agree that the more complex synostotic problems are best managed using a multispecialty team approach, which is often provided in the context of a multidisciplinary craniofacial center.

PEARLS

In these authors' experience:

- Not all progressive skull deformity is craniosynostosis; deformational (positional) plagiocephaly must be ruled out.
- Even unilateral cranial suture fusion results in bilateral skull deformities.
- Holistic reshaping of the skull, when dealing with scaphocephaly and turribrachysephaly, is superior to serial staged anterior and posterior procedures.
- Active remodeling of the skull and manipulation of skull dimension (anteroposterior or superior-inferior) are more effective in achieving normal skull form than is passive remodeling relying on brain growth alone.
- An inadequate correction of synostosis skull deformity exposes the patient to more cumulative surgical risk by making reoperation more likely.

SUGGESTED READINGS

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6

CRANIOFACIAL SYNDROMES: OPERATIVE TECHNIQUES

James Tait Goodrich

Surgical treatment of the various craniofacial syndromes has undergone a number of technical changes in the last several years. Various techniques, including strip craniectomies, lateral canthal advancement, facial augmentations, and partial to total calvarectomies, were developed to treat these congenital disorders. Over the last 15 years, our practice led us to use almost exclusively the bandeau/forehead reconstruction techniques developed by our French colleagues, Tessier, Marchac, and Renier. These techniques allow the surgical team to deal with the multiple anomalies that can occur in craniofacial syndromes, including restricted brain growth, asymmetric foreheads, and orbital dystopia.

Craniofacial syndromes almost always involve a premature fusion/sclerosis of one or more cranial sutures. As a result, the patient can develop a number of craniofacial problems. If the coronal sutures are involved, typically a brachycephaly occurs, with fusion of both coronal sutures leading to a tall and flattened forehead. Retrusion of the orbital bandeau is not uncommon and usually causes a bandlike constriction over the orbits. If one side is more involved than the other, unequal growing planes result, leading the child to develop an orbital dystopia caused by an asymmetric orbital rims. In more severe syndromes, such as kleeblattschädel (cloverleaf skull), multiple sutures are affected and lead not only to facial and calvarial asymmetry but also to restriction in brain growth. To address the multiple problems seen in craniofacial syndromes requires correcting the facial asymmetry and orbital dystopia, allowing brain growth, and providing an aesthetic, symmetric alignment of the forehead and face.

SURGICAL INDICATIONS AND PREOPERATIVE EVALUATION

Typically, children with craniofacial syndromes are diagnosed at birth or at least by 3 months of age; the rare exception is oxycephaly, which is diagnosed at a later age (>3 years). The neonatalogist or pediatrician usually makes the first diagnosis. Once the craniofacial syndrome is recognized and diagnosed, a series of diagnostic studies can be done that are helpful in clarifying the operative approach to be taken. A routine skull series, in most cases, will identify the affected sutures and determine the presence of increased intracranial pressure (ICP), typically seen with the presence of thumbprinting or digital markings on the skull. Three-dimensional (3-D) reformatted computed tomography (CT) has proven extremely helpful in the preoperative planning as well as for documenting the extent of suture synostosis. Hydrocephalus is not uncommon in these disorders and also can be evaluated by CT scan. The presence or absence of a Chiari malformation also can be evaluated. The head circumference is measured from birth onward to assess head growth. It is not unusual to have restricted head growth in multiple suture closure. As part of the

preoperative evaluation, the patient and family are seen by all members of the craniofacial team, which includes the neurosurgeon, plastic surgeon, pediatrician, pediatric neurologist, geneticist, social worker, and child-life specialist. As we have become more aware of the genetic influence in these syndromes, we involve the genetics team at the beginning to work up chromosome markers and to help with family counseling. After each team member has seen the patient and family, a conference is held, and the surgical recommendations to the family are planned.

There has been much discussion over the years about the child who has both severe craniosynostosis and midface retrusion (e.g., in Crouzon's syndrome) and the timing of the surgery to correct the calvarium and the midface. Ortiz-Monestario has been one of the strongest advocates for doing the monobloc advancement, in which the forehead and midface are both advanced early, that is, shortly after birth. Others have believed the risks of infection and blood loss and the severity of surgery to be too high and advocate doing the procedures in a two-stage fashion. The forehead and orbital regions are corrected shortly after birth, with the midface done later, at about age 8 to 9 years, depending on the facial maturity and presence of permanent dentition. The arguments for both sides are extensive and beyond the scope of this chapter.

Timing of Surgery

In craniofacial surgery, timing is extremely important. Several factors should be evaluated in making a decision about when to operate. Initially, we believed these children should be operated on as soon as possible, even as early as 2 to 3 weeks of age; however, we now prefer to wait until the child is 4 to 6 months of age to allow a more mature hematologic system and a larger blood volume for the anesthesiologist. In addition, the calvarial bone is firmer and has a more mature matrix, allowing better contouring and placement. The 6- to 12-month age period is critical in the developing child. This period is one of rapid head growth, and this growth assists in the final remodeling of the child's head and face. Therefore, it is critical to use this period of rapid growth to assist in the reconstruction. In certain situations in craniofacial syndromes, however, premature suture fusion and restricted head growth have made it necessary to perform surgery earlier to allow adequate brain growth and to preserve vital brain function. In particularly severe cases [e.g., kleeblattschädel (cloverleaf skull)], we have done calvarial expansions as early as 2 weeks of age. This group of children not uncommonly (>80%) will need later surgery for aesthetic corrections; in addition, they have the propensity to restenose, again requiring a calvarial expansion.

Hydrocephalus

In children who present with severe cloverleaf deformities at birth, almost all (and in our experience all) children develop hydrocephalus early. CT is obtained for these children shortly after birth, usually within the first 48 hours. The presence of enlarged ventricles merits a rapid, early placement of ventriculo-peritoneal shunt. We elect to place the shunt through an occipital burr hole so that the anterior and middle portions of the skull are easily accessible for the later calvarial reconstruction. An additional helpful step is to elevate the pericranium as a second layer and place the shunt under this layer. If the pericranium is closed over the shunt, this will provide an additional layer of tissue to protect the shunt apparatus during later surgical reconstruction.

PREOPERATIVE MANAGEMENT

Children with craniofacial syndromes often have other systemic problems that need to be evaluated. Routine ultrasounds are done of the organ systems to rule out any abnormality of the heart and kidneys. In children with multiple suture fusions, hypothalamic abnormalities also may be present; for this reason, routine endocrine examinations are done prior to surgery. We have the pediatrician do a well-baby checkup the day before the child is to be admitted to the hospital. If the child comes in with a fever and a workup reveals no active bacterial infection (normal white blood cell count with no shift), we proceed with the surgery. If the child has a upper respiratory infection but no signs of congestion, we also will proceed. If the child appears septic or an active bacterial source is identified, surgery is cancelled and rescheduled. Because these children often require blood transfusions, we offer the family the opportunity to provide donor-directed blood. Our experience is that more than 80% will take advantage of this option.

Intracranial Pressure Monitoring

In the child who presents with multisutural synostosis and marginal signs of increased ICP, a helpful adjunct test is ICP monitoring. The work by Marchac and Renier has been extremely useful in determining the ICP levels

that are clinically significant. Their work has shown that children with ICP readings greater than 15 mm Hg merit serious consideration for a calvarial expansion.

In our center, we admit the child for a 2-day inpatient stay in the intensive care unit (ICU). A lumbar drain is placed at the L4–L5 level, left in situ, and connected to a transducer. A graph is kept at the bedside with readings taken and recorded every half hour. Notes are kept of when the child is awake or asleep and during rapid eye movements (REM) sleep. Acceptable pressure readings are up to 10 mm Hg, ranges of 10 to 15 mm Hg are suspect, and persistent readings of greater than 15 mm Hg are consistent with abnormal ICP. An alternative technique for measuring ICP is using an epidural transducer placed through a small trepanation between the dura and skull. The same criteria for length and timing apply here as for measurements.

INTRAOPERATIVE TECHNIQUES

Anesthetic Techniques

General anesthesia with paralytic agents is used in all cases. Inhalation agents that increase ICP are avoided. Because of potential movement of the head, the airway must be secure, and because of potential blood loss, all patients require at least two large-bore intravenous 20-gauge lines or larger. Arterial lines are placed for monitoring blood gases, hematocrit, and electrolytes during the procedure. A Foley catheter is placed to monitor urine output. We do not routinely use steroids or anticonvulsants. Antibiotics are used (oxacillin 50 mg/kg) beginning with a preoperative dose and carried out for 24 hours. Our anesthesiologists will volume-load the child with crystalloid solutions at the beginning of the case. This hemodilutes the blood, thus helping to reduce blood loss. "Tanking up" the patient also reduces the risk of air embolism.

Airway Management

In children with severe cloverleaf anomalies, in particular those with Pfeiffer's syndrome, the cranial-base hypoplasia and midface anomalities cause significant airway obstruction. These children are typically obligate oral-airway breathers. For children with midface retrusion, abnormal palates, and large tongues, a tracheostomy is sometimes placed, usually within the first week of life, to prevent airway obstruction.

Ophthalmological Considerations

Children with severe cloverleaf deformities typically have shallow orbits and, as a result, present with severe orbital proptosis. An early oculoplastic consultation is extremely important for discussion about how to protect the globes and prevent keratopathies. Tarsorrhaphy, conjunctival flaps, and other measures are often necessary to protect the globes until the forehead and midface advancements can be accomplished.

Operative Position

The patient is placed in a supine position with the head resting in a horseshoe headrest. In cases where a total calvarial removal is planned, the head is flexed more forward, and the U-shaped headrest is used in place of the horseshoe. Some surgeons prefer the sphinx position, in which the child is placed prone and the head is hyperextended, with the child resting on a bolster or beanbag. If this position is to be used, preoperative radiographs of the cervical spine need to be done to rule out any congenital abnormalities. This position is contraindicated in the child with a Chiari malformation. The draping is done so that the head is fully exposed from the nasal tip to vertex to inion. A 180-degree access to the head and facial region is required so that no stands are placed to either side of the patient's head. The anesthesia team is placed parallel to the patient's side at foot level. The nursing team is placed on the opposite side at the foot of the patient. A small Mayo stand is placed over the patient's abdomen and a second mobile stand is placed to the side for surgical trays. Because multiple teams are involved in a staged fashion, several surgical trays are needed. Our nursing team has found it beneficial to keep a large table in the background to hold the various tray setups. As each surgical team comes into the field, their instruments are placed on the mobile table, and the required working instruments are placed on the Mayo stand.

Initial Exposure: Skin Incision and Flap Elevation

A bicoronal incision is performed and carried from ear to ear behind the hair line. Over the last several years, we switched to the stealth, or zigzag incision. Making several curves in the incision reduces the hair parting over the incision when the hair is wet and also seems to reduce the hypertrophic scarring that occurs over the temporalis

muscle. In most children, it is not necessary to shave the hair; instead, the incision can be carried through a parted hairline. In some cases, hair is shaved at a width of about 1 cm to allow placement of the incision and convenience in the later closure. The skin flap is elevated separate from the pericranium and carried down to the orbital rims bilaterally. Both frontozygomatic sutures must be exposed. In cases where a total calvariectomy is to be performed, the posterior flap is carried down to the inion. The pericranium then is elevated as a second layer and also carried down to the orbital rims. The neurovascular bundle is identified at the supraorbital notch and opened. There is often a thin rim of bone over the notch that can be easily opened with an osteotome. The dissection then is carried further around and under the orbital rims. At completion of the skin incision and forehead flap, both orbital rims and frontozygomatic sutures are exposed. Posteriorly, the inion and lambdoid sutures are exposed.

The temporalis muscles are elevated from their insertion point at the temporal line down to the level of the zygoma. The belly of the muscle is not incised because to do so would cause atrophy and later a cosmetic deformity ("dimpling" in the pterion region). The temporalis muscle is elevated using a monopolar needle tip cattier, winged out on its base in what I call a "fish-belly" fashion.

The orbital rims are dissected further until the nasion suture is exposed. Laterally, the orbital rims are dissected to the attachment of the lateral canthal ligament. We rarely detach this ligament except in severe cases of orbital dystopia. If the ligament is to be detached, an identification suture is placed through the ligament and then cut on the side closest to the orbital wall. This suture is helpful for locating and reattaching the ligament at the end of the procedure.

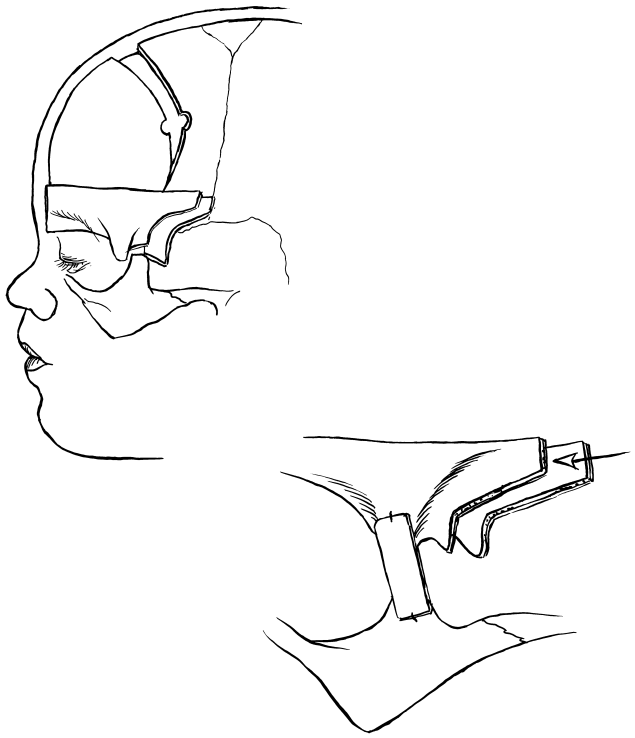

FIGURE 6–1. Orbital bandeau advanced in a tongue-in-groove fashion with the forehead position forward. Tongue-in-groove principle is shown in the lower insert. Sometimes an overlying bone graft is placed over the zygomatic defect to avoid a stepoff.

Craniotomy and Craniofacial Reconstruction

We use two types of reconstruction in surgically correcting craniofacial anomalies. In children with only brachycephaly, we elevate the original orbital bandeau and then reshape it prior to replacement. The bandeau should be over advanced at the initial placement to allow for adequate brain growth. If placed in its normal position, the child will rapidly develop a "waistbanding" or constriction just over the orbits. In cases where the bandeau is too deformed, a new bandeau is harvested from over the calvarial vertex. In a brachycephalic child or one with a cloverleaf deformity, the forehead unit is almost always deformed. To correct for this, a new forehead is marked out with a Marchac template. Figure 6–1 shows the osteotomy cuts; intraoperatively, these will be marked out with methylene blue. The team first makes the decision as to which type of craniotomy (partial or total calvariectomy) is to be performed, and then the appropriate marks are mapped.

Figure 6–2 illustrates a case in which the original bandeau was elevated and a new forehead was elevated from the right parietal region. The technique for elevating these units requires a round burr (e.g., Midas Rex M-33, Medtronic-Midas Rex, Fort Worth, TX) and a foot-plated craniotome (e.g., Midas Rex B-5). In cases where the anterior fontanelle is still open, we use this as our entry point for the craniotomy. The new forehead is marked and elevated first. By taking this piece first, the surgeon can use this entry point for further dural dissection. Superior cuts along the orbital rim are made, and the remaining original forehead unit now is elevated as a single piece, taking care not to injure the sagittal sinus (Fig. 6–3). Once this forehead unit is off, the frontal lobes are retracted gently, exposing both orbital roofs and bilateral sphenoid wings. Osteotomies are made just behind the bandeau and car-

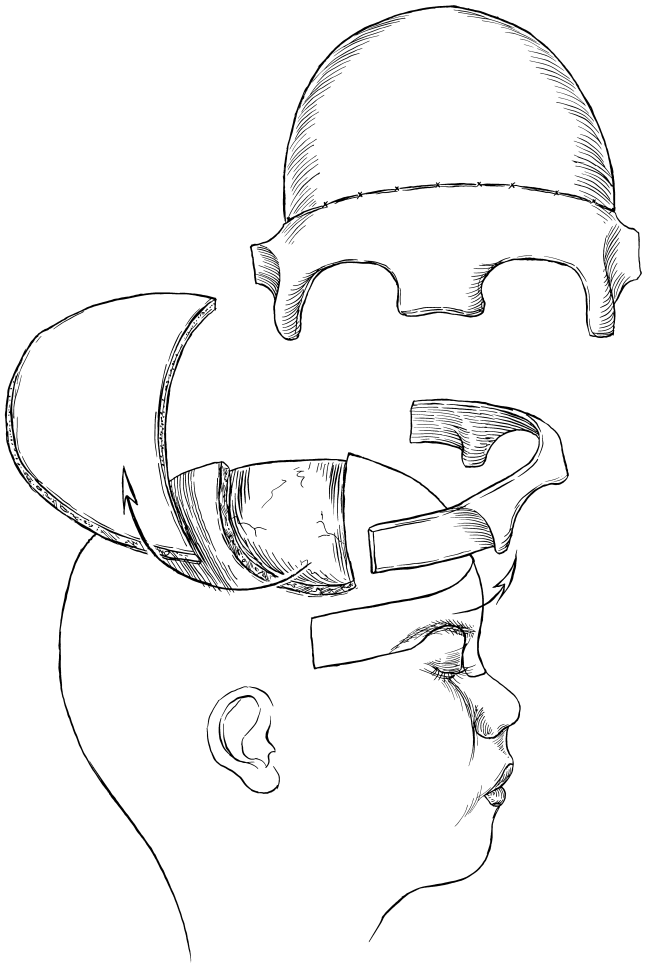

FIGURE 6–2. Artistic reconstruction where the bandeau has been elevated and a new forehead has been harvested from the right frontotemporal region. These two units then are combined into a single unit as shown in upper insert.

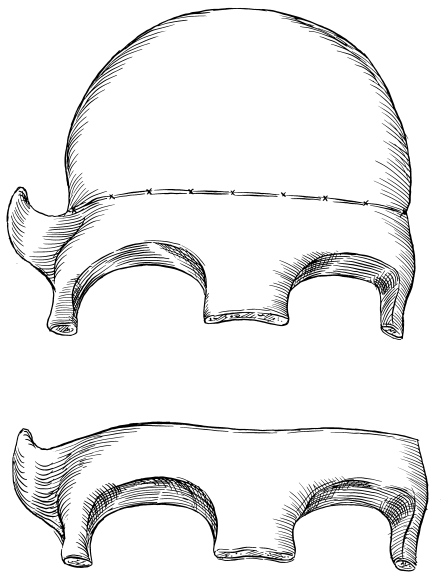

FIGURE 6–3. The principle behind the orbital bandeau and floating forehead is shown here along with a representation of where the osteotomy cuts are made to elevate each of the units.

ried over both orbital roofs and in front of the cribiform plate. The zygomatic arch is cut at the frontozygomatic suture line, and the nasal bone is cut at the nasion suture.

Once the bandeau is freed, it is handed off to the plastic surgery team, who re-constructs a bandeau and forehead unit. In the past, we typically used miniplates to reconstruct this unit. We have abandoned the use of miniplates and wires now, however, because of several cases of unacceptable migration of this material into the dura and, in one case, into the brain. We now use only absorbable sutures (e.g., vicryl 3.0) in placing the bone units. Once the bandeau and forehead have been handed off to the plastic surgeons, the neurosurgery team uses this interval to inspect for any dural or sinus tears, which must be meticulously closed. The bandeau and forehead are now replaced in a symmetric fashion and tongue-in-groove to the temporal/squamosal bone. If there is an asymmetry in the squamosal bone, radial cuts can be made and the bone green-sticked and out-fractured. A bone strut is placed from the top of the forehead to the skull (Fig. 6–4). The two open areas, lateral to the strut, are now reconstructed with the remaining bone in a mosaic fashion. A Tessier rib bender is helpful in modeling the bone. Split lamellar bone grafts are harvested from the inner table to patch any remaining defects. In our experience, bone defects larger than 1.5 to 2.0 cm will not close, particularly in a child older than 1 year of age. At this point, we leave no bone defects because split bone grafts can be harvested easily by using a sharp, small osteotome.

In craniofacial cases with multiple suture closures, the craniotomies are carried out more posterior to below the inion. Typically, the nuchal muscles must be detached from their insertion. We do this by elevating them with the paracranial layer. When elevating the bone, great care must be taken not to disrupt the transverse or sagittal sinus. In children with cloverleaf skulls, the sinuses commonly have invaginated into the bone, forming deep grooves. It is easy to transect the sinus at these grooves, and so careful dissection must be done in these areas (Figs. 6–5 and 6–6). Children with increased ICP typically have multiple digital impressions into the bone. Gentle and tedious dissection over these impressions will

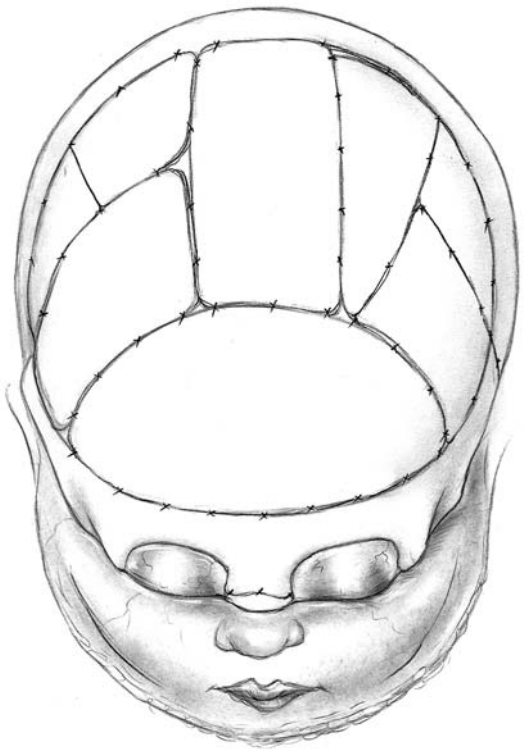

FIGURE 6–4. Artistic reconstruction showing the bandeau replaced (and advanced as necessary in a tongue-in-groove fashion). A strut has been placed from the new forehead back to normal skull. This key structural support keeps the forehead from collapsing backward when the closure is done.

reduce the risk of dural tears and injury to the sinuses. Sometimes large, diploic emissary veins go from the dura, through the bone, to the scalp. These emissary veins not only can bleed heavily but also entrain air emboli; therefore, caution must be exercised in obliterating them.

Closure Technique

Once the bone units have been repositioned and stabilized, the operative field is copiously irrigated to remove bone dust, debris, and other potential sources of infection. The gutters that formed where the skin flaps have been folded over need particular attention because a great deal of debris can collect here.

The pericranium is reelevated and tacked into position with several absorbable sutures. The temporalis muscles are reattached to the pericranium. This maneuver is extremely important because if this muscle is not adequately repositioned, bitemporal dimpling will result postoperatively, resulting in an unacceptable hourglass configuration to the face.

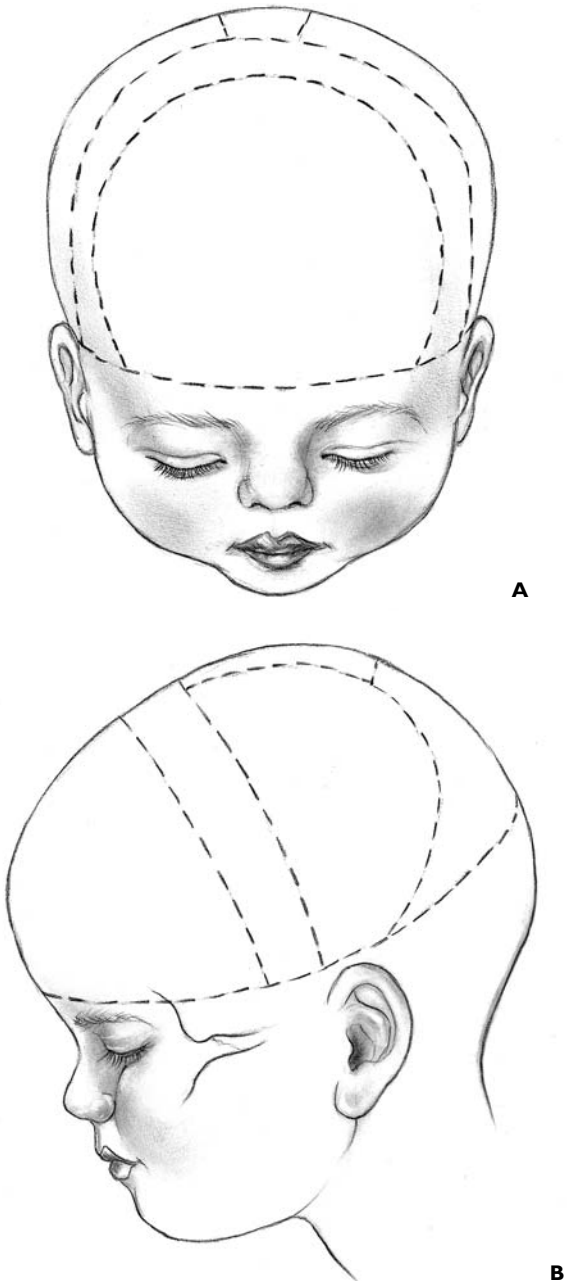

FIGURE 6–5. A child with a kleeblatschädel deformity (cloverleaf skull). **A:** To correct the aesthetics and also to allow for adequate brain growth, the calvarium is removed in total. **B:** Osteotomies for the new forehead and bandeau have been marked out. The forehead was selected by using a Marchac template (Leibinger) that is marched over the skull until the best contour is found.

The skin flap is reapproximated with a subgaleal closure. In the past, we placed a subgaleal drain for 24 hours

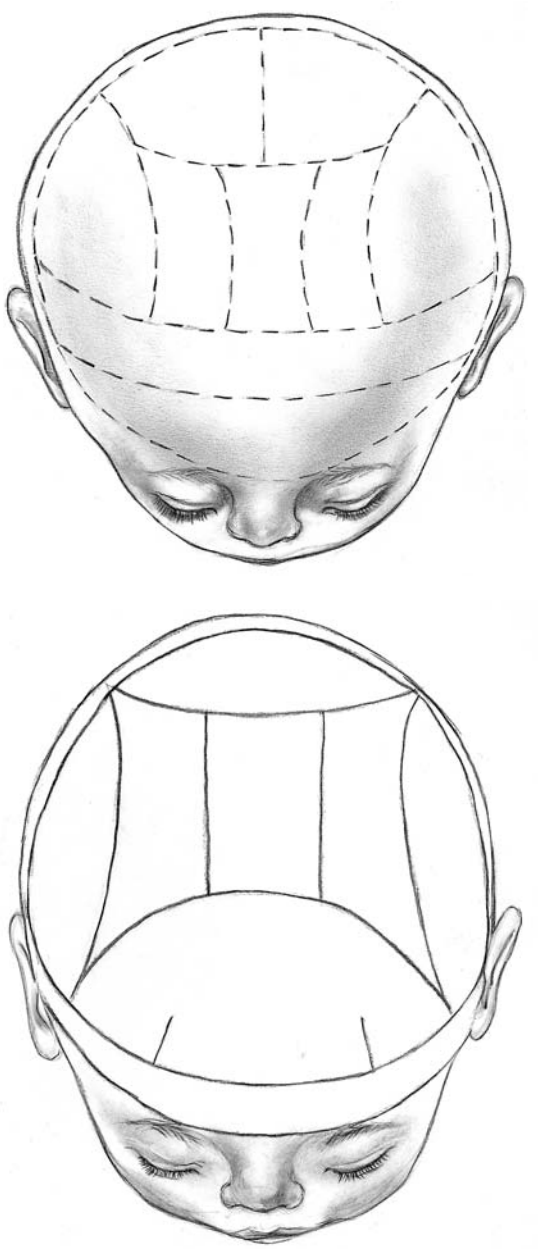

FIGURE 6–6. The top figure shows a vertex view of this patient with the osteotomies marked out. The bottom figure shows the bandeau and forehead placed into position and a lateral mosaic reconstruction of the rest of the calvarium done. The calvarium expansion accomplished and the aesthetics are immediately evident.

but now no longer do. In our experience, an unacceptable amount of blood was lost through the drain. Skin closure is done with a galeal closure using 4.0 dyed vicryl sutures and a 4.0 undyed vicryl suture placed in a subcuticular fashion. Steristrips are applied to the skin edges. A snug but nonbinding fluffy dressing is placed over the entire calvarium.

POSTOPERATIVE MANAGEMENT

The child is cared for in the pediatric ICU for a 24-hour postoperative period. Appropriate analgesia is provided to reduce the stress for the child. In the rare case where the child has lost more than a full circulating volume of blood, clotting factors are analyzed and replaced as necessary. After 24 hours, the child is fed by mouth. The child is positioned by the nursing team to lie only on the back so that no undue pressure is applied to the forehead. Restraints or sedation can be used as necessary to assist in keeping the child appropriately positioned. On postoperative day 3 to 4, the patient not uncommonly will develop fever in the 38° to 39°C range. A routine fever workup is done but rarely is a source found, and it is believed that these fevers are due to the blood resorption that occurs under the scalp flap. The child typically is discharged on day 6 or 7, assuming he or she is eating and has resumed normal behavior.

COMPLICATIONS

The single most dreaded complication is infection, particularly infection to the harvested bone. Osteomyelitis is devastating to the child because all of the infected bone will need to be removed. Fortunately, this complication is rare, occurring in fewer than 2% in most series. It cannot be emphasized enough that sterility is crucial, in particular the irrigation done at the end of the case to remove debris and bone dust, which can be a source of potential infection.

Risk of injury to the neural structures remains low in all reported craniofacial series. It is the responsibility of the neurosurgical team that no direct injury occur to the brain while the calvarial remodeling is under way. In our service, a member of the neurosurgical team is present throughout the entire surgery (i.e., when the plastic surgeons are working) and participates in the closure.

Significant blood loss, despite careful attention to detail, is always a possibility because these patients are so small. We routinely request from the family donor-directed blood, obtained 1 week before surgery. Two units are more than adequate in 95% of the cases. The single most common cause for excessive blood loss is a tear in the sagittal sinus. When recognized, these tears must be repaired by the neurosurgical team as quickly as possible. The team also must be prepared for air embolism if a major tear occurs, although a clinically significant air embolism is unusual.

Leakage of CSF and potential meningitis can be devastating to the patient. For this reason, we are extremely

meticulous in the closure, always checking for CSF leaks. It is often helpful to have the anesthesiologist provide a sustained valsalva maneuver before the craniofacial reconstruction to check for dura tears or leaks.

EDITOR'S COMMENTARY

The treatment of children with syndromic craniosynostosis is substantially more complex than in nonsyndromic cases for a variety of reasons. First, a substantial percentage of affected patients develop hydrocephalus during infancy or early childhood. Second, there is a high incidence of associated facial and general systemic anomalies, which must be taken into consideration during operative planning. Third, even after an apparently successful initial operation, patients are at risk for recurrent cranial growth restriction as a result of progressive multisutural synostosis. Accordingly, long-term follow-up of affected children is advisable. The optimal timing of intervention and the details of the surgical approach remain controversial. However, a point of almost uniform agreement is that the complex, multisystem nature of these syndromic cases requires a comprehensive multidisciplinary approach to patient management.

PEARLS

In this author's experience:

- In dealing with craniofacial syndromes, the surgical team must be prepared to work up all the medical systems to rule out any unexpected medical or anatomical anomalies such as cardiac, renal, and Chiari malformations. Preoperative appreciation of any underlying malformations is critical for reducing postoperative morbidity due to these anomalies.
- Eye/ocular protection is extremely important in those syndromic children with shallow orbits, protruding globes, and lack of adequate eyelid coverage.
- Blood loss can be extremely high in some cases, particularly in surgeries involving the midface region. Volume expansion in the child with crystalloid solutions prior to the start of the surgery reduces the incidence of air embolism and helps expand the vascular volume without blood products.
- The single greatest cause of morbidity in craniofacial surgery is blood loss and resultant anoxia. Constant monitoring of blood counts both during surgery (via arterial line monitoring) and the immediate 48 hours postoperatively is key to reducing morbidity. If the child loses more than 1.5 to 2.0 times his or her circulating blood volume, the addition of other blood factors, such as platelets and fresh frozen plasma, should be considered.
- Children with midface abnormalities (e.g., Pfeiffer, Crouzon, Apert) should always be considered to have abnormal airways. Fiberoptic intubations and delayed postoperative extubation should always be considered in the airway management of these patients.

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7

MYELOMENINGOCELES AND MENINGOCELES

J. Gordon McComb and Mark A. Mittler

MYELOMENINGOCELES

Patients with myelomeningoceles are often referred to as having spina bifida; however, the term *spina bifida* refers to a midline defect in the mesenchyma-derived tissues and does not include the neural tube (NT), which is the significant element. It is better to refer to these congenital lesions as neural tube defects (NTDs), which, from a practical, clinical standpoint, can be subdivided into two groups: *open* and *closed.*

With an open NTD, visible neural tissue is present and cerebrospinal fluid (CSF) drains either continuously or intermittently from the lesion. The entire central nervous system (CNS) is affected with anomalies that include the Chiari II hindbrain malformation, polymicrogyria, and other lamination defects of the cerebral cortex. Hydrocephalus is commonly present, and more than 90% of these newborns require CSF diversion for control of its progression. The widespread CNS anomalies may relate to the early and continuous loss of CSF from the open NTD.

With a closed NTD, no neural tissue is exposed, and CSF does not drain from the lesion. With few exceptions, such as some forms of posterior cervical meningoceles, the deformity of the CNS associated with a closed NTD is limited exclusively to the lower spinal cord.

Open NTDs can be subdivided into three types. The most common is the *myelomeningocele,* which is cystic in appearance with a dorsally displaced neural plaque or placode setting atop a collection of CSF. The second type, much less common, is that of *myeloschisis* (Fig. 7–1), wherein the open NT is "plastered" against the anterior wall of the spinal canal; there being no cystic structure. The third, and quite rare, is the hemi-myelomeningocele (Fig. 7–2) wherein the spinal cord is split with one portion being an open NTD and the other closed.

A cranial ultrasound, if convenient, can be done preoperatively to gauge the ventricular size at that time. The ventricles are usually only mildly to moderately dilated, and rarely is the hydrocephalus marked as the loss of CSF at the site of the open NTD prevents the hydrocephalus from becoming progressive unless there is complete aqueductal stenosis.

Intraoperative Techniques

Anesthesia and Positioning

Advances in anesthetic management have made operating on a newborn a routine matter, with the incidence of complications becoming ever lower. The two areas that tend to count for most problems are heat loss and blood loss. There are several ways to keep the temperature of the newborn at the appropriate level. An important factor is to monitor the infant's temperature and to make adjustments before the temperature drifts out of normal range. Because blood loss with most open NTD repairs can be kept to a small amount with judicious attention to hemostasis, usually only one peripheral intravenous line is needed. When a more extensive procedure is planned, such as a kyphectomy, a second intravenous and even an arterial line should be considered. When the blood loss reaches approximately 10% of the circulating blood

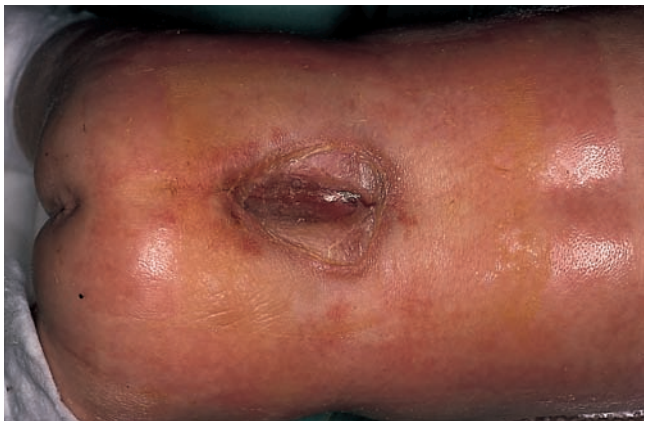


FIGURE 7–1. A newborn with myeloschisis. Note that there is no cystic component to this open NTD. The open neural tube is displaced and lies anteriorly on the spinal canal. CSF drains from the central canal at its rostral end. Other than having less tissue to close, this defect has the same extensive CNS malformation as a myelomeningocele.

volume, a decision needs to be made about possible transfusion, depending on how much more blood loss is expected before completion of the repair. The infant is paralyzed for induction but not thereafter, which allows one the ability to stimulate suspected functional nerve roots, if needed.

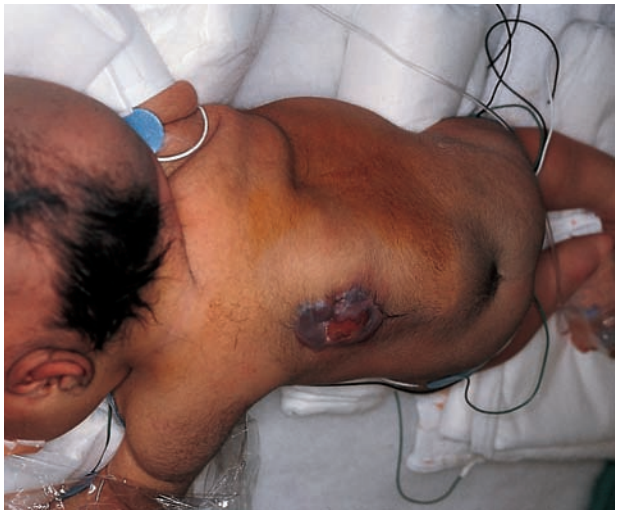


FIGURE 7–2. A newborn with a hemimyelomeningocele. This open NTD is thoracic in location and to the left of midline. The spinal cord is split with one portion being open and the other closed. The left lower extremity has minimal motor sensory function, while the right was normal. The hemicord on the right is fully ensheathed with dura mater while the left hemicord had an exposed neural placode. The malformation of the CNS was the same as if this infant had a full myelomeningocele.

If only the open NTD is to be addressed, the patient is placed prone, arms above the head, and towel rolls are placed under the upper chest and pelvis (Fig. 7–3). The lesion is cleaned with only normal saline or Ringer's lactate, taking care to avoid agents that could damage exposed neural tissue (Fig. 7–4). An iodine-impregnated adhesive drape with a large hole in the center is used to secure the towels and is placed on the intact skin at some distance from the defect.

We have not routinely inserted a CSF diverting shunt in an infant at the time of repair of the open NTD unless the hydrocephalus is marked. If both are to be done together while avoiding the need to reprepare and drape the infant, it is necessary to rotate the torso to have access to the peritoneal cavity as well as the back and head. The peritoneal tubing is directed medial to the scapula and enters the abdominal cavity from the flank. Visualization is enhanced with either higher-power loupe magnification or with an operating microscope. The only additional adjunct is the availability of a nerve stimulator.

Surgical Approach

The goal of operative repair is to preserve the neural function and prevent infection, primarily by obtaining a good skin closure over the defect. The manner in which the open NTD is closed can prevent long-term secondary complications, such as an enlarging dermal inclusion cyst (Fig. 7–5), and may diminish the later effects of tethering. These lesions come in a spectrum as to size, shape, and complexity; no two are alike. A degree of flexibility is needed in the technique to obtain the best closure. The overwhelming majority of these lesions can be simply closed without the need for myofascial flaps, rotational full-thickness skin flaps, partial-thickness skin grafts, or flank-relaxing incisions. At the beginning of the procedure, no tissue is excised that might be used for closure at the end. The dysplastic epithelial tissue adjacent to the neural plaque provides a better source of skin than a partial-thickness skin graft taken from elsewhere. Newborn tissue has a remarkable healing capability if the blood supply is adequate and infection does not intervene.

At the beginning of the procedure, CSF is often obtained and sent for a cell count with differential, gram stain and culture to be used as a baseline for future comparison, if needed (Fig. 7–6). The neural plaque is incised circumferentially, care being taken to exclude any epidermal or dermal elements that could subsequently develop into an inclusion cyst (Fig. 7–7).

In many cases, it is possible to close the open NTD by bringing the lateral edges of the plaque to the midline

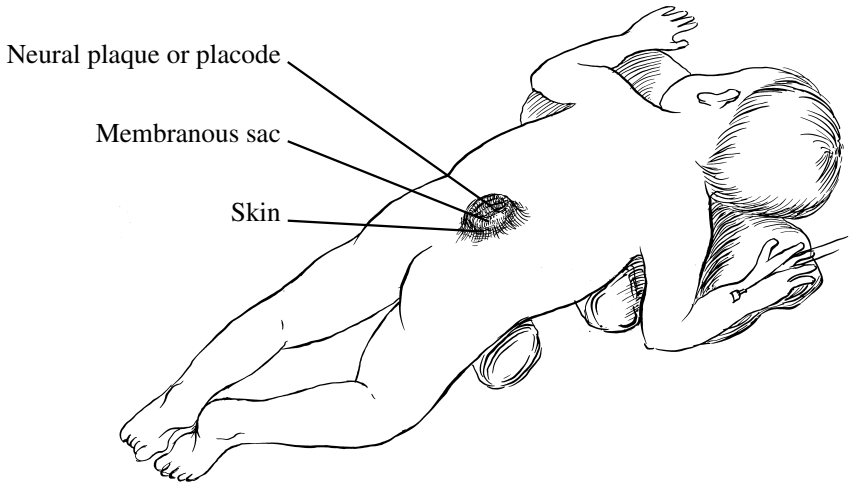


FIGURE 7–3. Salient features of the myelomeningocele. Patient is positioned for surgery with a small transverse roll under hips and another roll under chest.

and suturing the pial surface of the placode together with a fine, running, absorbable suture (Fig. 7–8). The configuration of some neural plaques make it difficult to obtain "a meaningful" closure of the NT. Any adhesions to the overlying meninges at the rostral end of the defect are divided. The dysplastic dura mater is circumferentially incised, taking care to have more than enough tissue to cover fully the repaired NTD and attached nerve roots with excess dura mater to spare because this might decrease the degree of adherence and thus later symptomatic tethering (Fig. 7–9). The dura mater is separated from the deep fascial layer beneath, brought together in the midline, and closed with a fine running, absorbable suture (Figs. 7–10 and 11).

No attempt is made to mobilize the deep fascial layer, with or without muscle attachments, because to do so increases operative manipulation, tissue disruption, and blood loss without improving the repair or decreasing the incidence of skin dehiscence, wound infection, and CSF leakage. Occasionally, the gap in the deep fascial layer is small, in which case it can be approximated, assuming it does not compromise the neural structures in the spinal canal. If there are bony protuberances from the malformed posterior vertebral arch elements, these are separated from the surrounding soft tissues and removed to make a smooth surface, thereby decreasing the chance of possible compromise to the overlying skin.

The skin is separated and undermined to a variable degree, depending on the size of the defect, at the abnor-

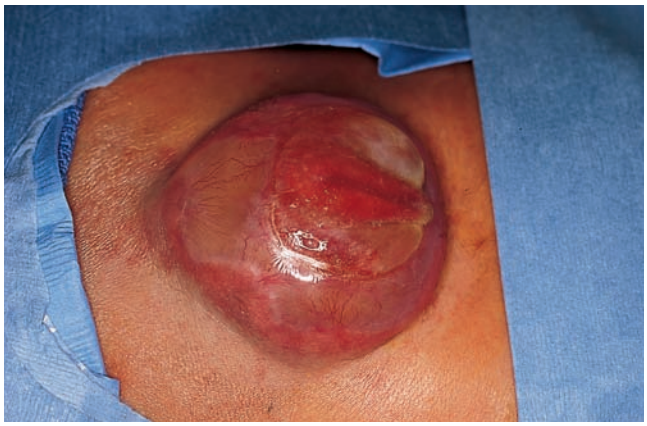


FIGURE 7–4. Typical lumbosacral myelomeningocele which was prepped with normal saline prior to beginning the closure.

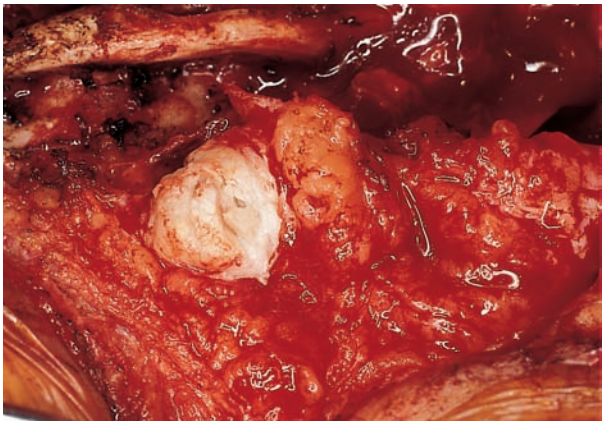


FIGURE 7–5. A dermal inclusion cyst at the site of a previously repaired myelomeningocele.

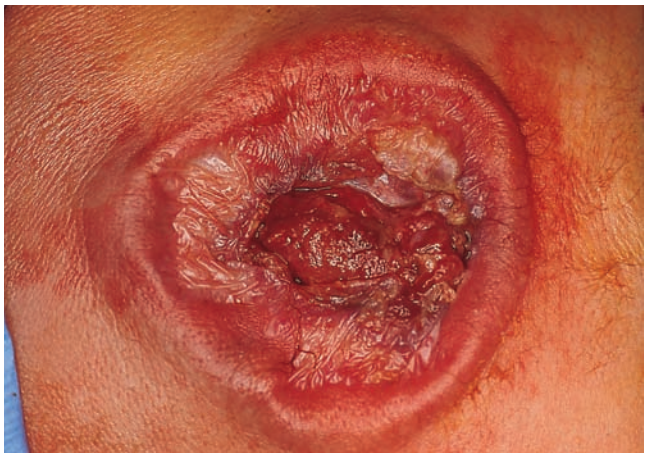
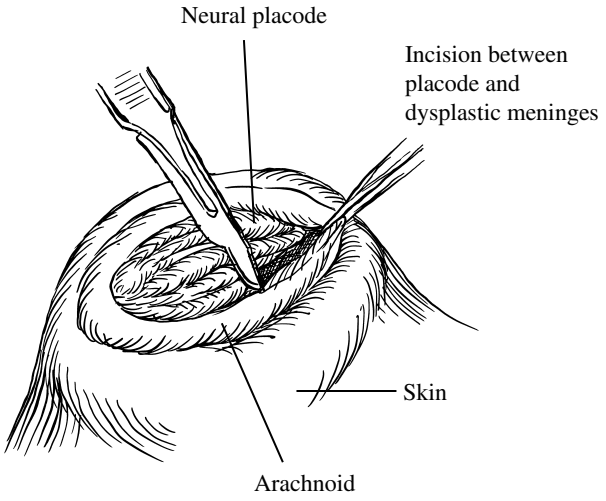


FIGURE 7–6. CSF has been drained from the sac and a sample sent for cell count with differential, gram stain and culture.

mal junction between the cutaneous layer and the underlying deep fascia (Fig. 7–12). Skin closure is preferably done in a transverse rather than a vertical direction because to do so usually produces a better cosmetic result and keeps the incision further separated from the anus. However, whichever direction provides the least tension is generally chosen (Fig. 7–13). The superficial fascia is approximated to whatever degree possible, often with a gap between the two sides in the midportion of the defect, where it is widest (Fig. 7–14).

Redundant skin is removed only when the superficial fascia of the skin is approximated to the maximum degree possible, with one or two sutures at the point where the gap is the widest (Fig. 7–15). The amount excised from either side of the defect may be different, depending on the quality of the tissue. If necessary, all the dysplastic skin, including the thin membranous epithelium and arachnoid located just lateral to the plaque, is retained. This tissue readily heals and creates an intact epithelial barrier. At either end of the incision, the excess



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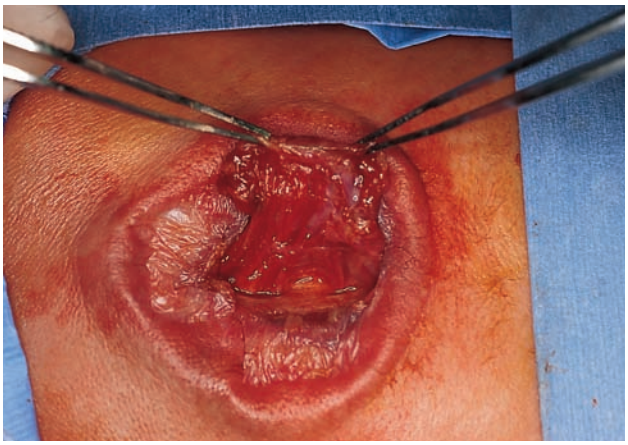

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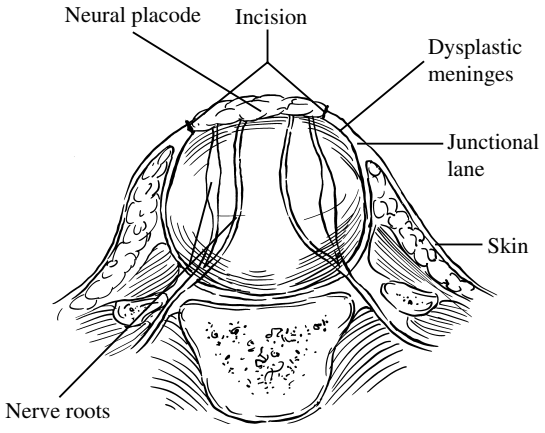

B

FIGURE 7–7. **A:** Initial incision with #15 blade scalpel. Incision is made between the neural placode and membranous sac. **B:** The neural plaque is circumferentially freed from the surrounding dysplastic tissue with care being taken not to include any epithelial or dermal elements. Note the nerve roots attached to the neural placode. **C:** Placode separated from the membranous sac. CSF has been drained and the placode collapses toward the spinal cord.

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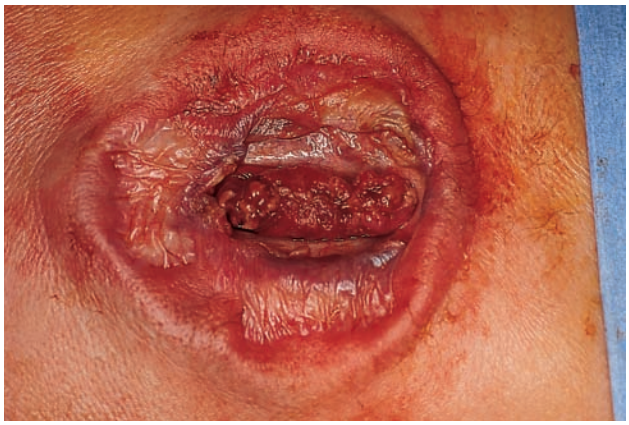

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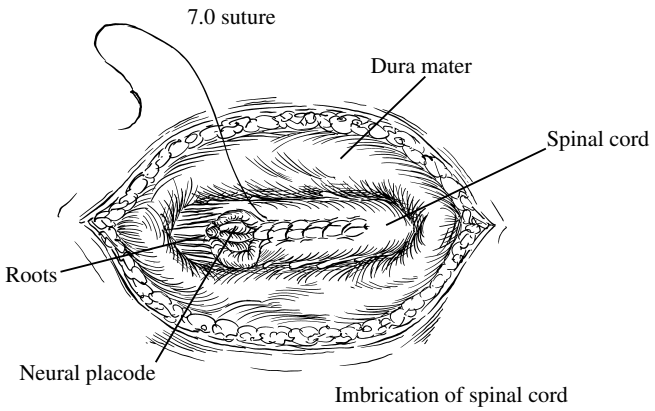

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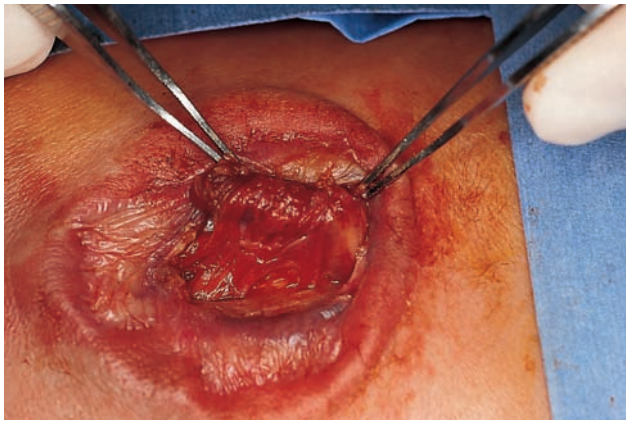


C

FIGURE 7–8. A, B: The lateral edges of the neural placode are brought to the midline and closed with a fine absorbable running suture. This reconstitutes the neural tube and hopefully decreases the area for scarring and subsequent tethering. **C:** The re-constituted neural tube is shown with the nerve roots attached.

skin or "dog ears" are excised. At this point, the skin edges on either side of the wound should lie opposite one another with the minimal tension possible. Additional interrupted absorbable sutures are placed to approximate the subcutaneous tissue further. The cutaneous layer then is closed using a fine running, absorbable suture or Steri-Strip.

Additional Operative Procedures

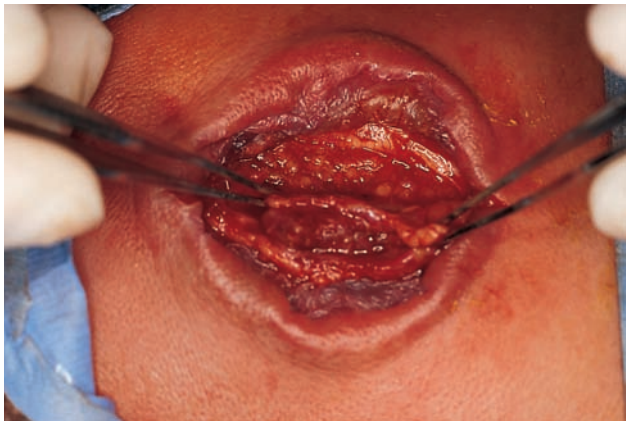
Repair of a myeloschisis is similar to that of a myelomeningocele, the only difference being less dysplastic dura mater, superficial fascia, and skin available for closing the defect. In this situation, every available bit of tissue counts. It is also necessary to undermine the skin surrounding the lesion more extensively.

A hemimyelomeningocele makes its presence known. It is located on one side of the midline, usually in the thoracic region, and is associated with a marked motor or sensory discrepancy deficit between the lower extremities (Fig. 7–2). This form of an open NTD is always associated with a split-cord malformation wherein the two cord segments are in separate dural sheaths with a bony spur between. In addition to repair of the open hemicord, the bony spur should be removed and the cord untethered at the time of the repair. In this situation, it would be helpful to perform magnetic resonance imaging (MRI).

Although infrequent, other NTDs can be seen in conjunction with a myelomeningocele. The most common are a split-cord malformation with the two hemicords within the same dural sheath and a neurentenic cyst.

Occasionally, the newborn might also have a marked gibbus deformity, which is usually seen in the context of a lumbothoracic or thoracic myelomeningocele, often with a complete loss of neurologic function below the level of the defect. In this situation, it is best to resect one or more vertebral bodies to reduce or eliminate the gibbus. Because this additional operative manipulation results in a more significant blood loss, transfusion should be anticipated.

If the patient is to have a shunt placed at the same time as the open NTD is repaired, it is recommended that the shunt be placed first because theoretically there should be less chance for contamination of the shunt hardware and resultant infection. Once the shunt is in place, the patient can be partially repositioned to gain



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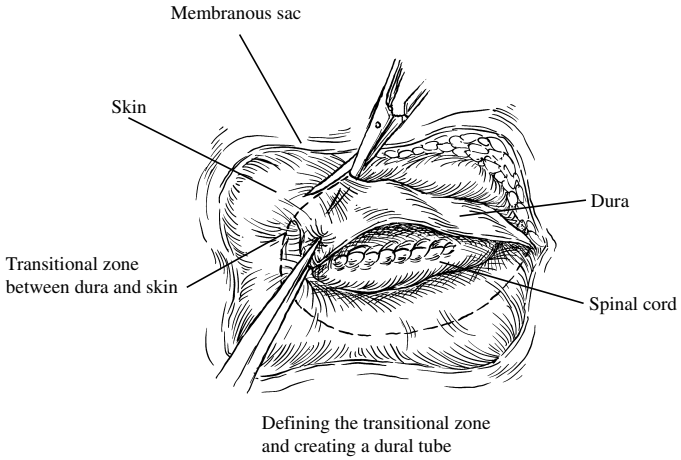

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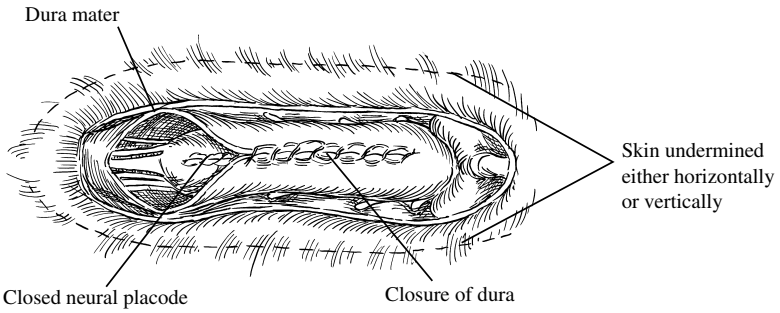


C

FIGURE 7–9. **A:** The dysplastic dura mater is being circumferentially incised and separated from the surrounding superficial fascia. **B:** Scissors are used to free up the dura, which will later be closed over the spinal cord. Note that remnants of the membranous sac are still attached to the skin margins. Some advocate excising this membranous sac, but it can be used like skin in closing large lesions, so we leave it in place until confirming that the skin will close without it. **C:** The next layer to be closed is the dura. We close this using a running suture in the midline. The goal of this closure is to create an enclosed space for CSF to circulate around the spinal cord.

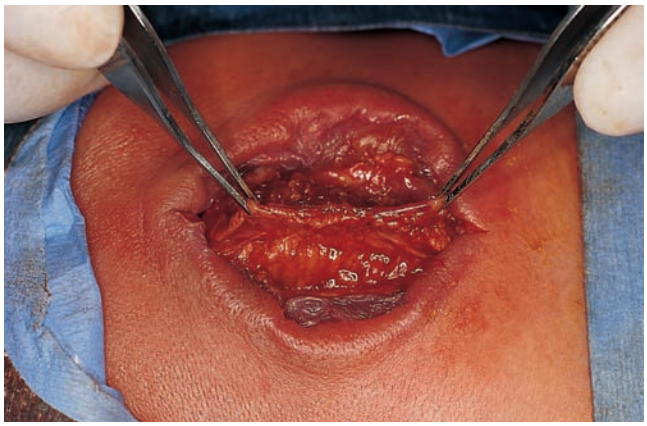


FIGURE 7–10. This shows completion of the dissection of the dura mater so as to be able to bring it together in the midline with ample room to enclose the neural elements.

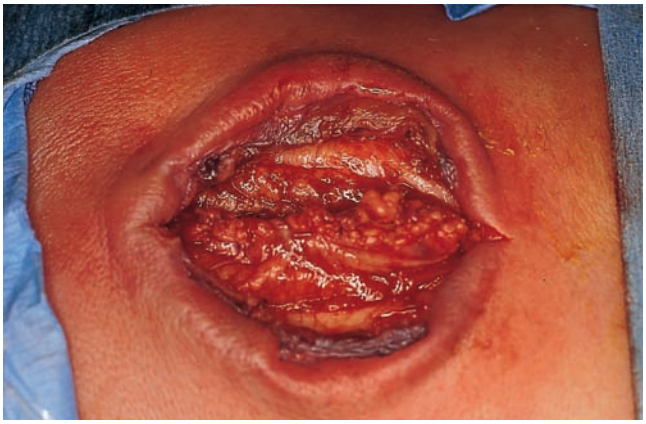


FIGURE 7–11. The dura mater is approximated with a running fine absorbable suture.

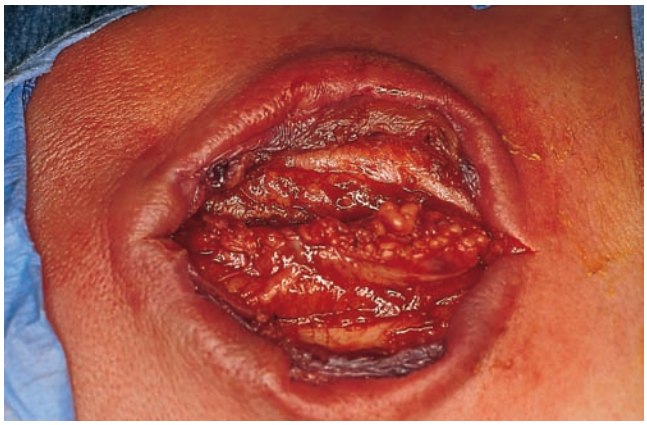


FIGURE 7–12. The skin and superficial fascia are undermined to the extent that the superficial fascia can be approximated.

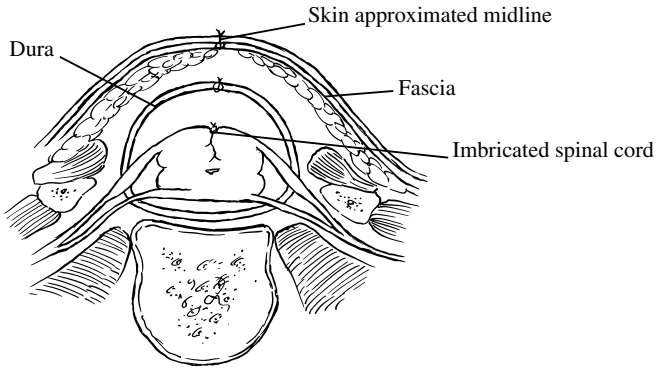
better access to the myelomeningocele without having to reprepare and drape the field.

Postoperative Management

The patient is kept prone most of the time to avoid soilage of the wound with urine or feces. The infant may be placed on his or her side for feeding. It also may be necessary to place the patient supine to catheterize the bladder intermittently in the presence of significant residual urine. In the past, we have placed a barrier drape or "mud flap" with an adhesive band covering the buttocks, thus separating the site of repair from the anus and the bladder. This technique has proved ineffective in enough situations that we have abandoned the practice. We now remove the dressing on the first postoperative day. This allows frequent observation and cleaning of the incision site when it is contaminated by urine or fecal material. These infants are usually continually dribbling urine and feces because of a lack of normal sphincter innervation, thus making it hard to keep them and the wound clean. If fluid begins to build up at the operative site, it indicates increased resistance to CSF circulation, and the patient needs to be shunted, even if the ventricles have not enlarged further with a follow-up cranial ultrasound.

To protect the repair site, the skin is prepared and tapped with a 25-gauge butterfly needle and CSF is aspirated, which can be sent for cell count, Gram stain, and culture, as indicated. By removing the fluid collection, it will protect the wound from dehiscence until a CSF diverting shunt is inserted.

The complications of repair of an open NTD include infection, dehiscence, and CSF leakage, which often occur together. Once again, CSF leakage can be avoided by tapping and removing any collected CSF and then ventricular CSF diversion. Obviously, an infection is treated by using the appropriate antibiotics, whereas dehiscence usually responds to local wound care. Only rarely is it necessary to bring the newborn back to the operating room to repair the wound. Problems with



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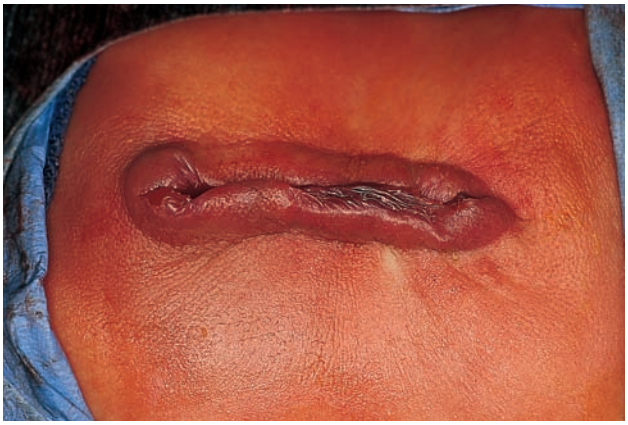

B

FIGURE 7–13. A: The superficial fascia is re-approximated. The goal of this layer is to take all tension off the next layer, the skin. The end result of this re-approximation often is extra skin. The edges are trimmed to neatly come together and the membranous sac is discarded along with the extra skin. If the skin does not come together, the membranous sac is used in place of skin for those small regions. **B:** Intraoperative view of **(A)**. No dysplastic skin has been excised at this point.

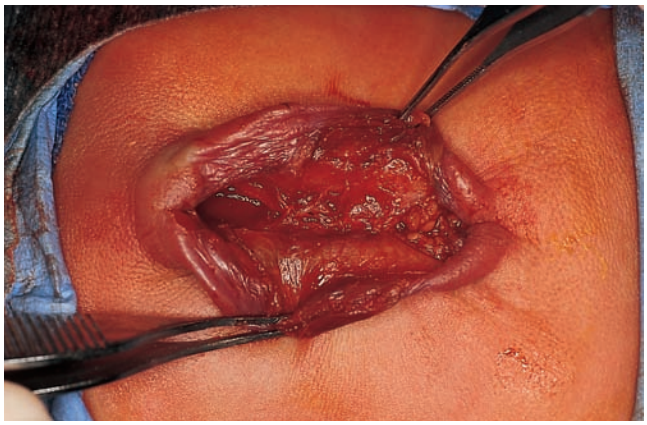

FIGURE 7–14. This shows the redundant skin reflected and the superficial fascia approximated. It is only at this point that the redundant skin is excised.

wound healing can be a major factor result in prolonging the hospital stay.

We recently reviewed almost 200 cases of newborns who underwent repair using the simple closure technique as described already. Our total complication rate for infection, dehiscence, and CSF leakage was 6%, which compares favorably with other reported series using more complex and involved techniques. In addition to the usual aseptic technique, the best way to avoid complications with closure of an open NTD is not to compromise the blood supply to the skin at the incision site.

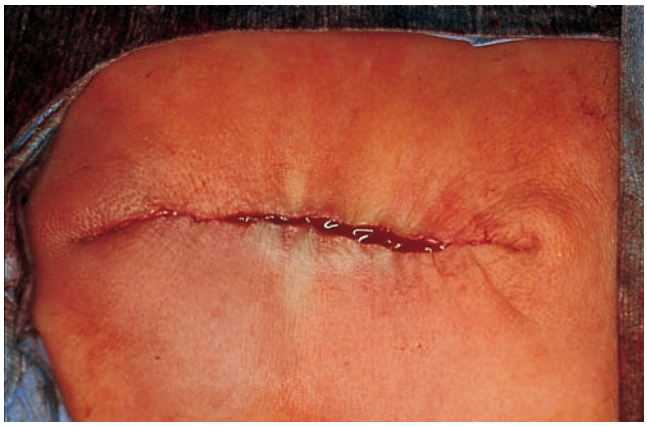

FIGURE 7–15. Redundant and dysplastic skin has been excised and the superficial layers are approximated with absorbable sutures. In spite of the lesion being quite large, it has been possible to fully close the lesion with full thickness skin. The skin edges can be further opposed with Steri-Strips or with a fine running absorbable suture which makes it less likely that CSF could leak at the site of repair.

Neurologic function postoperatively in the lower extremities should be quite similar to that preoperatively and is rarely more than a dermatome different between the sides. Because of the presence of a neurogenic bladder, the infant, on occasion, can develop overflow incontinence with significant bladder distention. Inspection and intermittent catheterization for residual urine are part of the postoperative management scheme.

A small percentage of infants, probably 5% or less, will develop symptoms referable to the Chiari II malformation such as apnea, difficulty with handling secretions, being able to feed, vocal cord paralysis, etc., which relate to dysfunction of the lower cranial nerves and brain stem. If the patient has not been shunted a CSF diverting shunt should be inserted without delay. If the patient has already been shunted it is important to make certain that the shunt is functioning well. If a shunt is in place and working well it would be necessary to do a posterior fossa decompression without delay as early operative intervention can reverse the symptoms in some of these patients.

In the postoperative period, the other members of the multidisciplinary group, especially those from urology and orthopedic services, need to evaluate the infant and institute appropriate measures.

MENINGOCELES

Spinal meningoceles, by definition, are a protrusion of the dura mater and accompanying arachnoid membrane through a defect in the spinal column with the spinal cord remaining within the confines of the spinal canal. Meningoceles are an infrequent and heterogeneous group of cystic lesions. For the purpose of discussing surgical repair, the lesions can be divided into those that are posteriorly located, from the cervical to the sacral region, and those that are anterior to the sacrum. Even more rare are meningoceles anteriorly or laterally located in the cervical, thoracic, or lumbar region. These meningoceles require operative intervention but are rare, particularly in the pediatric age group, and are not discussed here.

Probably most, if not all, posterior meningoceles are not "pure" in that they contain, either singularly or in combination, aberrant nerve roots adherent to the wall of the herniation, occasional ganglion cells, and a glial stalk that may represent a diverticulum from the central canal of the spinal cord. Even though fragmentary neural elements are beyond the confines of the spinal canal, the spinal cord is not.

Posterior Meningoceles

Preoperative Management

These lesions are more commonly found in the lumbosacral region and are fully covered with skin, albeit dysplastic on occasion; thus, they have no exposed neural tissue and are not leaking CSF. Repair can be done electively, although many neonates undergo surgical correction prior to discharge from the newborn nursery.

On rare occasions, the dysplastic skin may be torn during delivery, resulting in CSF leakage, in which case the defect should be repaired shortly after delivery. All these infants should have MRI of the entire spinal cord to exclude other NTDs, whereas those with a cervical meningocele also should undergo MRI of the head because in this group there appears to be a transition between open and closed NTD, with some of these infants having a Chiari malformation and hydrocephalus. The cervical meningoceles are also more likely to be associated with an accompanying split-cord malformation, neurentenic cyst, and encephalocele. Computerized tomographic (CT) studies and CT myelography may be needed on occasion to sort out a complex malformation. Most of these infants need no special care prior to operative correction other than to protect the skin overlying the meningocele from pressure.

Intraoperative Techniques

Anesthesia and Positioning

Because it is unlikely that the need to stimulate nerve roots with repair of meningoceles will arise, the infant may be kept paralyzed. In the rare occasion where stimulation is indicated, most anesthetic agents allow rapid recovery to the level where stimulation can be used.

If the lesion is lower on the spine, the patient is placed prone with rolls under the upper chest and pelvis and the head turned to one side. In the cervical region, the patient's head is placed in a neutral position in a horseshoe. The horseshoe is thoroughly padded to prevent any chance of pressure injury to the face. The amount of draping should be kept light enough so as not to injure the facies as well.

Visualization can be done with the unaided eye or loupe magnification for the superficial portion of the procedure. It is helpful to have the magnification and lighting of the operating microscope to see within the spinal canal in most cases.

Surgical Approach

Having enough skin and fascia for closure of the meningocele is rarely a problem (Fig. 7–16). The meningocele can be incised in either a transverse or a vertical fashion, with a transverse repair usually providing a better cosmetic appearance. If a more extensive exploration of the spinal canal is contemplated, a vertical incision makes visualization easier. A glial stalk adherent to the side or dome of the sac is often encountered (Fig. 7–17). Although the cystic component of the meningocele can be large, the sac usually narrows to a relatively small opening in the dorsal aspect of the spinal canal (Fig. 7–18). If visualization is not adequate, a limited one or two level laminectomy is undertaken.

The glial stalk is traced to the dorsal surface of the spinal cord and amputated. If the defect is relatively large, such as with a myelocystocele that can accompany a cervical meningocele, the pial edges of the defect are opposed with a fine running, absorbable suture to decrease the chance of postoperative tethering (Fig. 7–19). The spinal cord is explored to ensure that no additional tethering remains and there are no other associated NTDs. In the sacral region, the filum terminale can be identified and divided if there is any evidence of tethering.

The dura mater is easily closed with a fine running, absorbable suture. The deep fascia is closed with larger interrupted or running absorbable sutures. The excess skin is resected so that the edges lie opposed to each

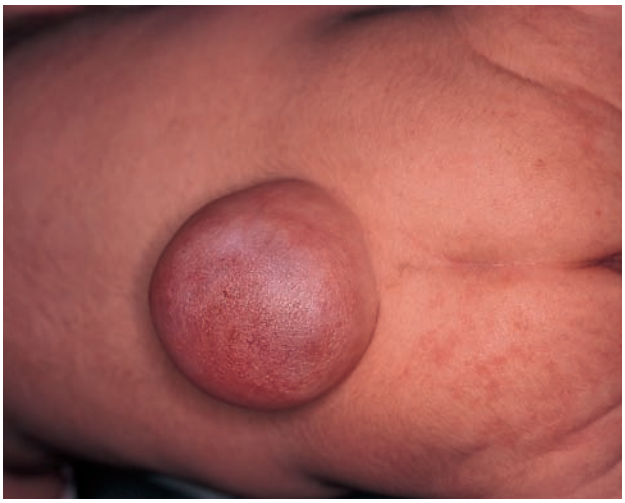

FIGURE 7–16. Typical lumbosacral meningocele in a newborn. The overlying skin is normal. The neonate had no neurologic deficit. The meningocele was repaired and the patient has remained completely normal.

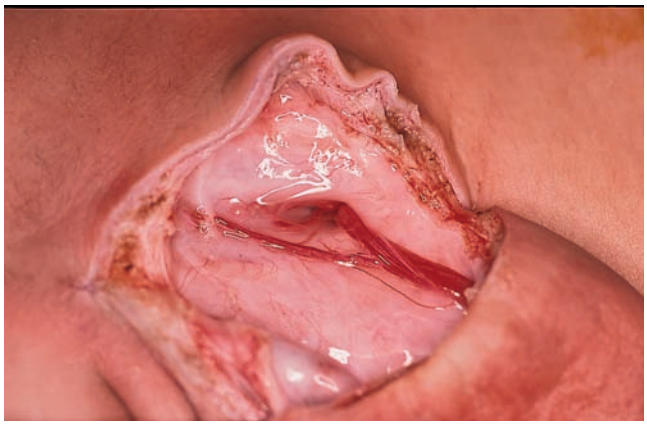

FIGURE 7–17. After opening the meningocele dome, a fibroglial nodule, densely adherent to the meningocele sac was present. This was resected flush with the spinal cord to minimize subsequent tethering.

other tension free. Additional subcutaneous and cutaneous absorbable sutures can be placed for the final approximation. The use of skin sutures reduces the chance of a CSF leak.

If a preoperative MRI study has shown the presence of a Chiari malformation in conjunction with a posterior cervical meningocele, it probably would be best to do a bony decompression at the same time as repair of the meningocele. In this case, a vertical incision would be preferable to allow adequate access to the foramen magnum.

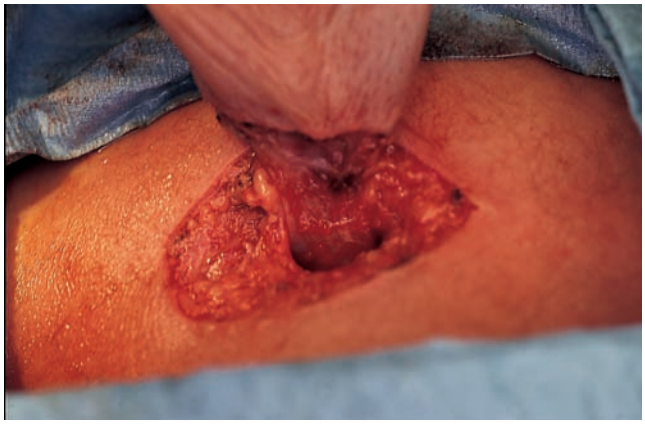

FIGURE 7–18. Although the cystic component to the meningocele can be large, it usually narrows to a relatively small opening into the dorsal aspect of the spinal canal. It is necessary to visualize the dorsal aspect of the spinal cord to make certain it is not tethered. If the opening narrows to such an extent that adequate visualization is not possible, a limited laminectomy is done to provide for further exposure.

Postoperative Management

Most infants with repaired meningoceles need no special care and can be discharged soon thereafter. A patient with a posterior cervical meningocele who has a Chiari malformation, encephalocele, or progressive hydrocephalus would be the exception. Cranial ultrasonography would be used to assess ventricular size and, if progressively enlarging, a CSF diverting shunt inserted. If CSF accumulates at the site of repair, it can be aspirated intermittently with a fine butterfly needle until a CSF diverting shunt is inserted. This problem should not be a factor for those lesions located in the lumbosacral region.

Anterior Sacral Meningoceles

Preoperative Management

The anterior sacral meningocele is a truly occult lesion because there are no visible abnormalities. These lesions frequently are associated with rectal abnormalities (including an imperforate anus) malformation of the uterus and vagina, duplication of the renal pelvis and ureters, bony pelvic and vertebral abnormalities, a dermoid, teratomas, or hamartomas associated with the cyst (Fig. 7–20). The embryologic abnormality of the bony anterior sacral elements allow herniation of the dura mater into the pelvis, leading to the development of an anterior sacral meningocele that will slowly enlarge over the course of decades, displacing the rectum, bladder, and uterus, leading to difficulty with bowel and bladder function as well as dystocia. Diagnostic evaluation includes plain radiographs of the spine that show the characteristic crescent or scimitar deformity to the sacral region (Fig. 7–21). Pelvic and abdominal ultrasonography easily detects the presence of a cystic lesion. MRI is indicated in all cases to define further the location of the spinal cord and the presence of other NTDs, the site of communication to the anterior sacral region, and the nature of the cystic mass within the pelvis and whether it contains solid components. CT myelography can define further the size and location of the CSF fistula if there should be any question on the MRI studies (Fig. 7–22). Depending on the presence and nature of the abnormalities of the alimentary, urologic, and reproductive systems, other diagnostic studies may be necessary. Aspiration of the fluid contents of the anterior sacral meningocele via the rectum or vagina is contraindicated as it may result in CSF leakage or meningitis.

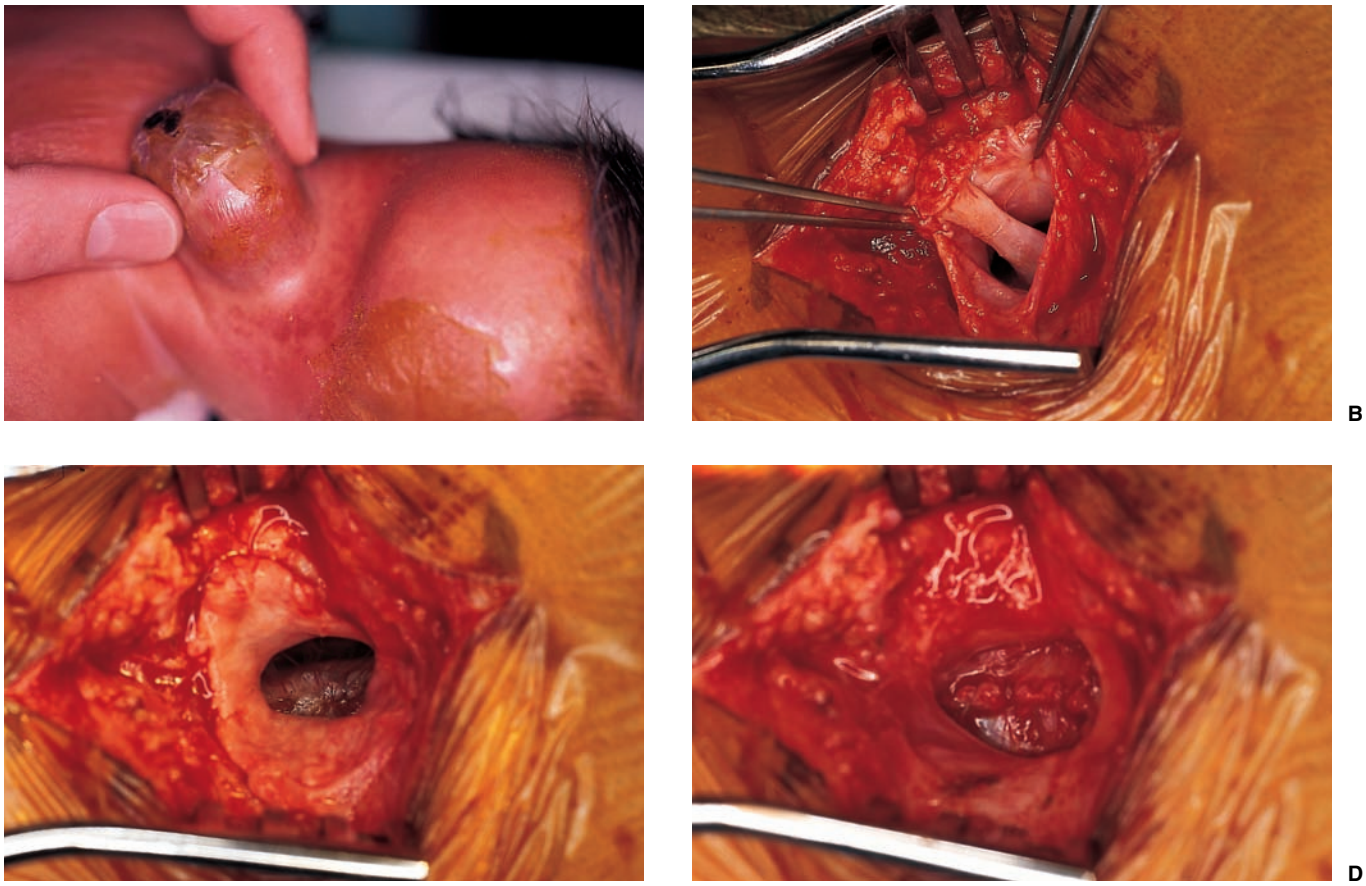

FIGURE 7–19. **A:** This newborn has a cervical meningocele in which the dome is covered with an eschar rather than being fully epithelialized. **B:** A thickened fibroglial stalk ran from the dorsal surface of the spinal cord to the dome of the meningocele. **C:** The stalk was resected flush with the spinal cord. The pial edges were then approximated in the midline to decrease the chance of significant scarring and tethering to the overlying dura mater. **D:** Closure of the dura mater. The skin was subsequently closed with layers of absorbable sutures.

Intraoperative Techniques

Although these lesions are being detected more frequently and at an earlier age, these patients tend to be older and, as a result, heat loss and blood loss are less significant factors than in the newborn period. The patient is placed in a position similar to that of repair of a myelomeningocele or a lumbosacral meningocele. Anterior sacral meningoceles are best approached posteriorly through the sacrum. As indeed they are truly occult lesions, there are no abnormalities of the overlying skin, and so there is no need to resect any abnormal tissue in this location. A midline incision is made, and the subcutaneous tissue is divided. The deep fascial layer may or may not be intact, and often a defect exists in the posterior sacral bony elements. At times, it may be necessary to remove some bone for adequate visualization. The dura mater is opened posteriorly in the midline and retracted laterally with stay sutures to allow exposure to the anterior aspect of the spinal canal. The dura mater surrounding the fistulous connection is isolated, ligated, and divided. The anterior portion of the meningocele sac within the pelvis, if it contains only CSF, need not be excised because it will shrivel once the access to CSF is eliminated. Prior to the last two steps, the lesion is carefully inspected to ensure that there are no solid components that would constitute a dermoid or teratomatous component in addition to the CSF-filled meningeal sac. If a solid tissue component is encountered, an attempt is made to resect as much as or all the abnormal tissue as possible. If this should not prove feasible, the anterior meningocele is disconnected from the spinal subarachnoid space at the end of the thecal sac and both lesions are closed using running absorbable sutures. Markers such as hemostatic clips can be placed within the sac as well as on the margins of the remaining isolated intrapelvic portion of the meningocele for subsequent identification and removal at the time of surgically correcting the rectal abnormalities. If nerve

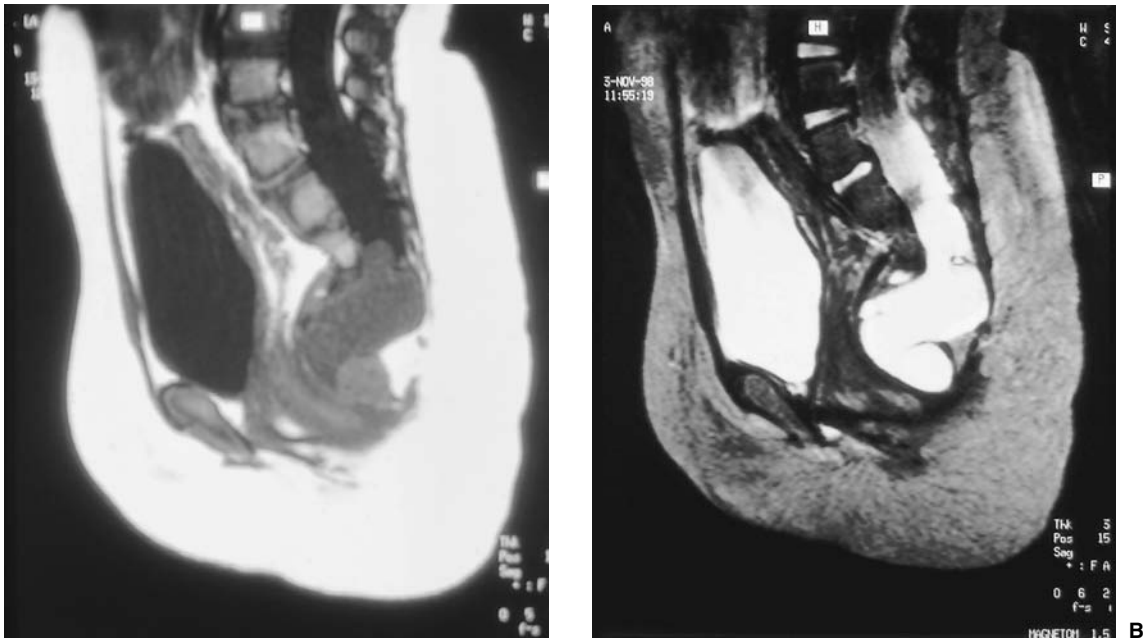

FIGURE 7–20. A T1-weighted image (**A)** showing the lower end of the thecal sac which contains CSF. Just beneath this is a large dermoid which becomes bright on the T2-weighted image (**B)** as does the CSF. This infant was born with an imperforate anus and required a diverting colostomy. Because of the known association between rectal anomalies and closed NTD, an MRI study was obtained. This showed the presence of this anterior sacral meningocele and associated dermoid tumor. From a spinal canal approach, much of the dermoid was resected and the thecal sac closed. At the time of the anal reconstruction the remainder of the dermoid was removed. The distal sac had been closed at the time of the first resection and metallic clips applied to the edges for easy identification at the time of the second procedure.

roots are noted to enter the tract to the anterior sacral meningocele, they are preserved and the dural opening plicated after packing oxidized cellulose or gelatin foam with or without fibrin glue into the tract surrounding the nerve roots. The sac itself is not divided. After dural plication, the additional oxidized cellulose is placed on the subarachnoid side of the tract. If the spinal cord is low lying and appears to be tethered, the filum terminale is also divided. If other NTDs are present, they also can be addressed during the same operative procedure.

Many infants with anterior sacral meningoceles have rectal abnormalities that require an initial diverting colostomy right after birth. Repair of the anterior sacral meningocele from a posterior approach should be performed before any subsequent anterior abdominal procedure to repair the rectal anomalies to minimize the risks of meningitis and CSF leakage. At the time of the anterior abdominal exploration, any remnant of the cyst is explored to ensure that no solid component harboring tumor remains. If the anterior sacral meningocele is diagnosed during a woman's pregnancy, it is prudent in most cases to deliver the fetus by C-section at or near term and repair the woman's meningocele postpartum.

Postoperative Management

Wound healing usually proceeds rapidly in the pediatric population. As CSF circulation is rarely a problem and one can usually get a good dural and fascial closure, progressive buildup of fluid at the operative site is uncommon. Keeping the patient flat will reduce the hydrostatic pressure on the closure.

If the anterior sacral meningocele is repaired posteriorly and no anterior operations are needed, follow-up with ultrasonography or an MRI or both may be indicated to ensure that the fistulous connection was obliterated at the time of surgical repair and that no solid components harboring dermoid or teratomatous elements exist.

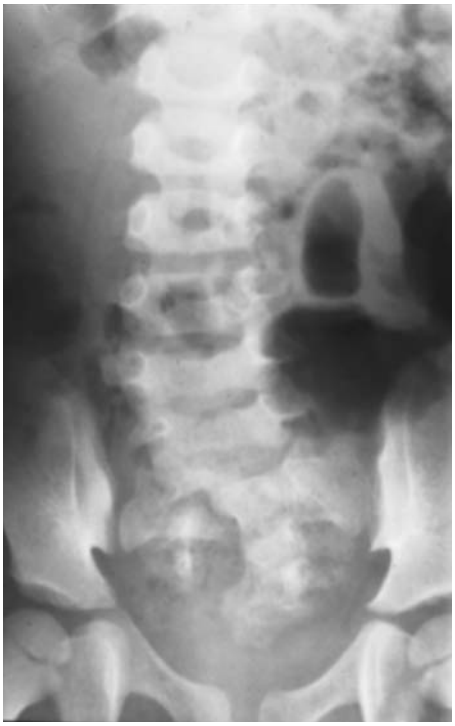

FIGURE 7–21. Plain radiograph showing the typical crescent or scimitar deformity that occurs in the sacral region in the presence of an anterior sacral meningocele.

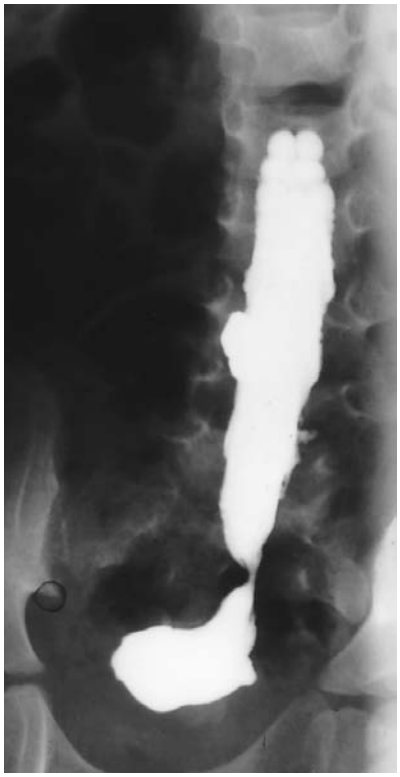

FIGURE 7–22. A CT myelogram showing the narrow neck connecting the distal end of the thecal sac with an anterior sacral meningocele.

EDITOR'S COMMENTARY

Surgical techniques used to close myelomeningoceles and meningoceles have changed minimally in the past 25 years. The principles of early timing of operations, closure of dura over the neural placode, and skin coverage of the defect are unchanged. Pediatric neurosurgeons still do not have the techniques to bridge the neural gap across the myelomeningocele nor to prevent subsequent cord tethering. The authors describe a straightforward closure techique. The 6% complication rate associated with the technique attests to its usefulness. Other neurosurgeons use a vertical closure so that later tethered cord operations can use the same incision. Others develop semicircular leaflets of lumbodorsal fascia that are approximated at the midline to minimize CSF leakage if the dural closure weakens and minimize CSF infection if a superficial infection develops. If corpectomies are needed to treat severe congenital kyphosis, it is difficult to stabilize the realigned vertebral bodies; sutures tend to pull out. Mini-plates and screws probably provide the best stability and can be supplemented with a postoperative brace. Techniques for the in-utero closure of myelomeningoceles were not demonstrated in this chapter because such operations are currently performed on an investigational basis.

PEARLS

In these authors' experience:

MENINGOCELES

- The entire spinal axis need to be imaged to exclude the presence of additional NTDs.
- Posterior lumbosacral meningoceles are the more common of this form of NTD and usually carry an excellent prognosis for normal neurological development.
- Chiari malformations and hydrocephalus are only found with some posterior cervical meningoceles.
- At the time of surgical repair of the meningocele, it is important to explore the spinal cord to make certain that it is fully untethered both ventrally and dorsally. It may be necessary to do a limited one- or two-level laminectomy to accomplish this.
- Anterior sacral meningoceles produce a characteristic scimitar deformity of the sac.

MYELOMENINGOCELES

- This is a open NTD in which the entire CNS is affected and is associated with a Chiari II malformation and hydrocephalus.
- The overwhelming majority of these lesions can be simply closed without the need for myofascial flaps, rotational full thickness skin flaps, partial thickness skin grafts, or flank relaxing incisions.
- At the beginning of the procedure, no tissue is excised that might be used for closure at the end.
- It is important to exclude any epidural or dural elements from the neural placode that could subsequently develop into an inclusion cyst.
- It is often possible to convert the placode into a tube by bringing the lateral edges to the midline and suturing the pial surface together.
- Bony protuberances from the malformed posterior vertebral arches should be removed.
- If the patient has not been shunted and CSF builds up at the repair site, intermittent aspiration of the fluid can protect the wound until a CSF diverting shunt is inserted.

SUGGESTED READINGS

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8

SPINA BIFIDA OCCULTA

W. Jerry Oakes

Spina bifida occulta (SBO) is a complex of congenital disorders of the spine that are united by a common etiology and embryologic error, clinical presentation, and occurrence of more than one expression of pathology in a single patient. The seven pathological entities include the tethered spinal cord (TSC) from a thickened, fatty infiltrated filum terminale; lipomyelomeningocele; dermal sinus tract and dermoid; split cord anomaly (diastematomyelia); terminal syrinx; neurentic cyst; and meningocele manqué. A patient frequently expresses more than one pathological manifestation of disease, such as a lipomyelomeningocele coexisting with a TSC and a terminal syrinx; in addition, these problems may coexist with a myelomeningocele. In this case, the filum terminale may be a significant point of fixation below a myelomeningocele, or a myelomeningocele may occur on only one hemicord (hemimyelia) at the site of a split cord anomaly. Without the understanding that lesions are frequently seen together, incomplete or partial operations may be done, and the patient may continue to express the natural history of the unoperated portion of the pathology.

The purpose of all operations in this group of conditions is to release points of fixation of the neural tissue and to remove compressing masses. The points of fixation may be caudal, as from a thickened filum terminale; dorsal, as from meningocele manqué; or, occasionally, ventral at the site of a split cord anomaly or a neurentic cyst.

INDICATIONS AND PREOPERATIVE EVALUATION

The indications for operation on almost all these lesions are simply their presence. Little controversy exists regarding the prophylactic removal of dermal sinuses or dermoids, split cord anomalies, TSC from a thickened filum terminale, or neurentic cysts. Meningocele manqué is generally diagnosed accompanying some other process and may be difficult to confirm with imaging. Terminal syringomyelia, which expands the spinal cord, is generally addressed at the onset of symptoms or before a significant deficit occurs. The only real controversy surrounds lipomyelomeningoceles. A number of well-respected authors (primarily from Europe) advocate a conservative approach until and unless a progressive neurologic deficit develops. This is primarily not a question regarding the natural history but rather concern regarding the risk of an extensive procedure on a complex lesion in an asymptomatic infant. Therefore, justification for taking on the surgical risks rests in the experience of the surgeon. Some surgeons would allow the magnetic resonance (MR) imaging appearance of the lesion to influence operability. In the United States, much less controversy exists, and lipomyelomeningoceles usually are recommended for operation at the time of diagnosis.

Prior to operation, patients require a detailed assessment of their neurologic function and the anatomy of their lesion. In small infants, this examination may be challenging, but indirect findings can be seen readily. Loss of muscle bulk and unusual postures of a joint imply neurologic weakness. Seeing the extremity in use may be as much as can be expected in a 6-month-old infant.

Some objective measures of neurologic function may be helpful, such as electromyography (EMG) or urodynamics. If a urologic deficit is suspected, documentation of the preoperative situation may be important in assessing a postoperative deficit. Simply assessing the rectal wink reflex and obtaining a history of a strong urinary stream are inadequate to assess bladder function. I personally rarely use EMG to assess motor function but rely heavily on urodynamics.

Routine radiographs of the involved area, ultrasound in young infants, and MR imaging to determine

the anatomy of the lesion when the acoustic window is inadequate are routinely performed. On occasion, the ultrasound or MR imaging may be difficult to interpret and computerized tomographic (CT) myelography may be helpful.

Contraindications

There are few clear reasons not to recommend operative intervention. In the case of serious life-threatening associated congenital anomalies, where survival is doubtful, surgery to prevent neurologic deterioration is not logical. Even in the setting of significant intellectual debility, maintaining useful lower extremities and bladder control and minimizing neurogenic pain are powerful reasons to proceed with surgery.

The timing of operation is a different consideration. If the infant is medically stable and the natural history of the lesion is progression, albeit at an unpredictable rate, I see little argument for postponing intervention and usually proceed within a few weeks to days of diagnosis.

INTRAOPERATIVE TECHNIQUE

All procedures are performed with the patient prone and bolsters placed under the iliac crest and chest to allow free abdominal excursion (Fig. 8–1). Intraoperative monitoring with somatosensory evoked potentials (SSEPs) or EMG can give the surgeon some reassurance but are not used in our clinic.

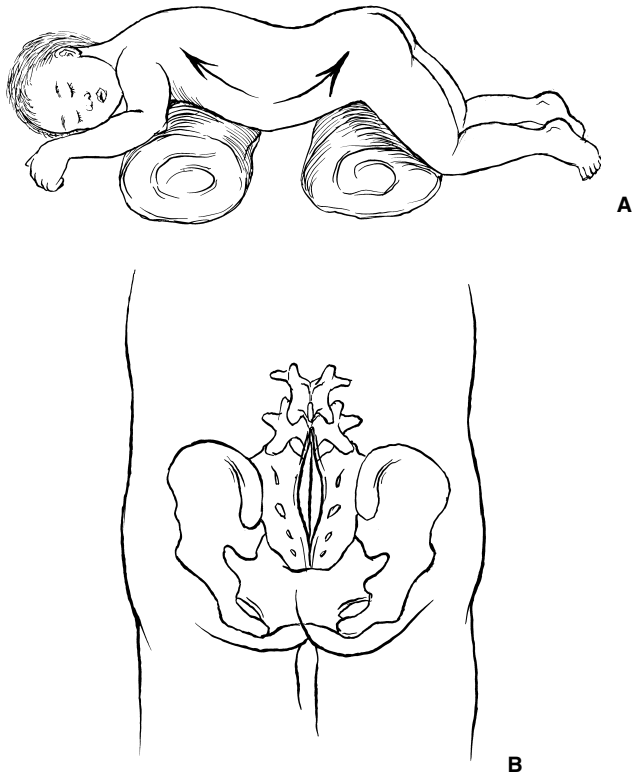

FIGURE 8–1. A: Position of infant ready to undergo spinal exploration. Bolsters placed under the iliac crest and chest allow free abdominal excursion. **B:** Position of the midline sacral incision in its relationship to the underlying spine.

Tethered Spinal Cord

Section of the filum terminale for the TSC should be done in the cul-de-sac of the sacral subarachnoid space. This requires a midline incision from the L-5 spinous process to the midsacrum and a laminectomy of S-1 and S-2 (Fig. 8–1B). The fat infiltrated filum frequently can be seen through the dura. The periostium in small children will appear as a separate layer under the lamina. The inexperienced surgeon might assume this to be the dura and, on incising this structure, confuse it with the epidural fat.

The dura is opened in the midline, and the filum is separated from the exiting nerve roots. The filum can be identified as a midline structure with a characteristic vessel on its under surface. It usually exits the dura dorsally in the midline at the point of termination of the subarachnoid space. Most importantly, nerve roots have a predictable striation pattern or banding that occurs approximately every millimeter. This pathognomonic finding is absent with the filum. Almost every pathological filum will have fat within it; however, on occasion, fat also may appear within the roots. Following the structures to their point of exiting the dura will add confidence, with neural elements coursing ventral and lateral as opposed to the dorsal midline filum. Neural structures are separated from the filum (Fig. 8–2A), which is coagulated and cut. The sectioning is done first distally and then, a second time, cephalad to the first to allow a specimen to be taken and sent for pathological examination. This examination will confirm the structure sectioned and, with a section removed, the likelihood of the two ends reuniting is remote. All this is done with little or no blood contamination of the subarachnoid space to minimize postoperative adhesions.

When the filum is more robust and occupies much of the intradural space (Fig. 8–2B), care must be exercised to section the filum below the last exiting nerve

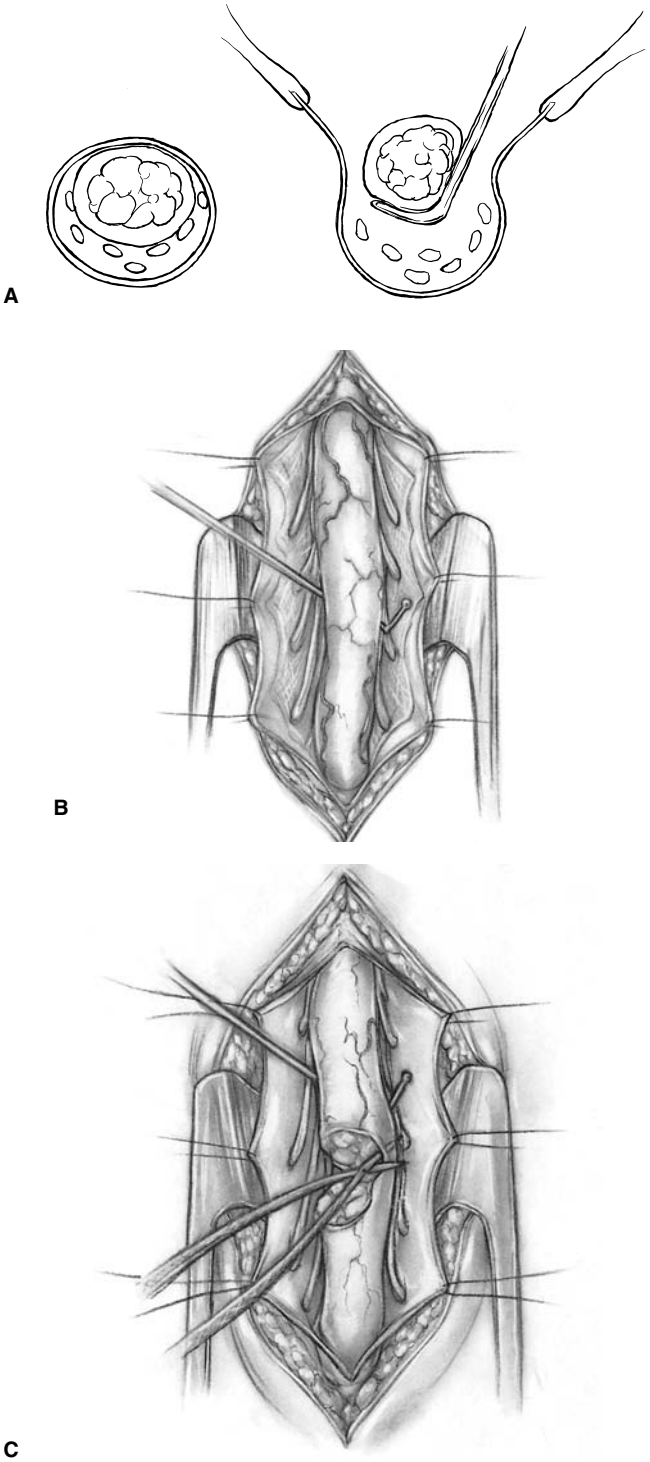

FIGURE 8–2. A: Cross-sectional view of the intradural contents of the sacrum. The large dorsally positioned and fat infiltrated filum is being separated from the adjacent roots. **B:** Intraoperative view of the sacral intradural contents. The nerve hook is separating the thickened filum prior to sectioning. **C:** Same view as **(B),** with the distal filum being sectioned.

root. The filum must be rotated from side to side to visualize the undersurface and to ensure that no roots remain adherent (Fig. 8–2C). The dura and soft tissues are closed in the usual manner. Alternative techniques include replacing the S-1 and S-2 laminae and visualizing the filum endoscopically.

Lipomyelomeningoceles

Lipomyelomeningoceles have two pathological aspects: the fixation of the spinal cord and the mass effect of the fat. The fixation occurs caudally from a thickened filum and either caudally or dorsally at the point the fat exits the dura; this is the primary pathological process. The fat coming into the caudally situated spinal cord will need to be debulked to allow visualization of all points of fixation, but it should not be considered a neoplasm in which the goal of surgery is to remove all neoplastic tissue. Leaving small amounts of fat adherent to nervous tissue is quite acceptable and in fact desirable. The goal of removal of all the fat is unnecessary and dangerous to the continued function of the nervous elements.

The patient is positioned prone, as with all other procedures (Fig. 8–3A). A midline skin incision is made over the subcutaneous fatty mass. Skin flaps are elevated off the mass and reflected laterally. The fatty mass is dissected circumferentially from the lumbodorsal fascia. As the edge of the mass is lifted and freed, the neck of the mass protruding through the fascial defect can be seen. All adhesions between the neck and the edge of the fascia should be opened. Redundant fat is removed from the subcutaneous mass (Fig. 8–3B). The fascia cephalad to the neck of the lesion is opened in the midline, and the paraspinal muscle is dissected off the most distal intact spinous process and laminae. Frequently, caudal to the bone is a band of tissue constricting and compressing the upper aspect of the dural neck (Fig. 8–3). The band should be cut and a segment of bone cleared to expose dura within the spinal canal. With the soft tissue reflected laterally, the exposed dura is opened in the midline above the lesion. The spinal cord and its junction with the fat are inspected. Beginning laterally, the dura is opened at its junction with the fat neural complex; this is the most important aspect of a successful procedure. If visualization of this junction is obscured by bone, the bone is removed. If it is obscured by redundant fat, the fat must be debulked, which can be done with a defocused CO_2 laser, a routine microsurgical technique, or (as I prefer) with an ultrasonic aspirator. Great care must be exercised in this maneuver to avoid neural injury (Fig. 8–4A). The

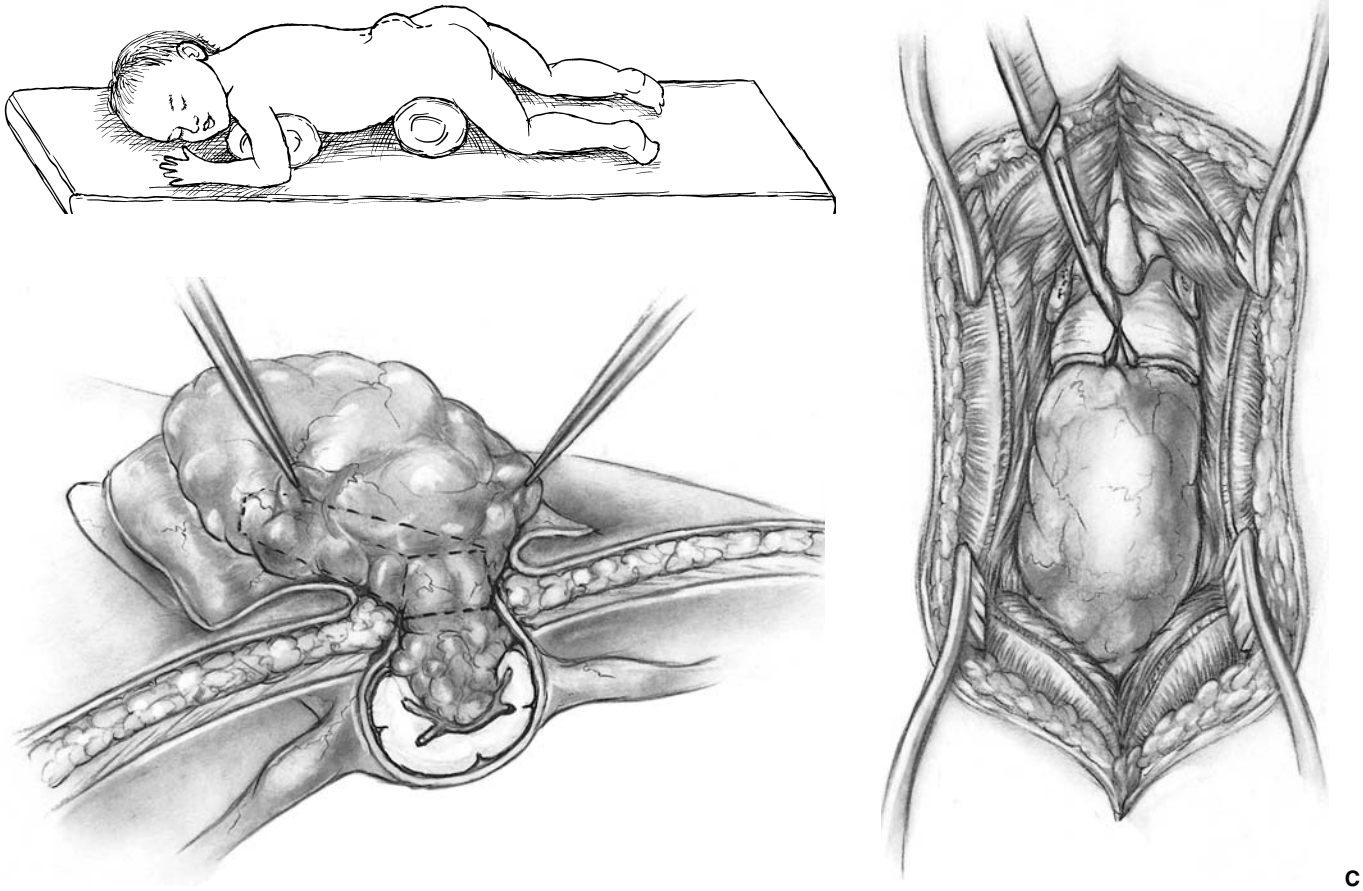

FIGURE 8–3. A: Infant with a typical lumbosacral lipomyelomeningocele is positioned for surgery. **B:** Oblique cutaway view of a lipomyelomeningocele. The skin has been reflected off the subcutaneous lipoma. Extension of the lesion into the dorsal cord is obvious. The forceps are elevating the lipoma so the neck of the lesion can be dissected. **C:** Operative exposure of a lipomyelomeningocele. Note that immediately above the lipoma and below the last intact laminae there is a constricting band that has been partially cut. This structure can apply significant pressure to the underlying cord as it angles dorsally to emerge from the dural defect.

roots here leave the cord in an unpredictable fashion coursing caudally, horizontally, or even cephalad. They may lose the regular segmented orientation and be bunched in large groups or have long skip areas. Sensory roots coming from the dorsal aspect of the cord and hugging the inner aspect of the dura are most vulnerable to injury. Slowly, as the junction of the dura with the fat–neural complex is opened, the intradural anatomy is revealed. Frequently, before each cut in the dura, fat will need to be debulked. Routine microsurgical technique is associated with troublesome bleeding, with the cut end of the vessel retracting into the fat. Ultrasonic aspiration tends to preserve these vascular structures, allowing them to be coagulated and cut prospectively. Low suction and power settings are effective with this process.

The surgeon works along each side and eventually enters the cul-de-sac of the subarachnoid space. A search for the filum is made; when found, it is sectioned. With the dural attachments completely freed, the neural elements should fall against the ventral surface of the dura. If not, the cause of the continued dorsal or caudal tension should be found and resolved.

A situation that complicates the exposure is rotation of the neural elements so that the fat appears to come more from the lateral rather than strictly the dorsal aspect of the cord. This may position intact cord rather than fat in the midline and makes one side of the dural opening somewhat simpler and the other much more difficult. If the neural elements leave the cord above the plane of the dural tube, the likelihood of injury is signifi-

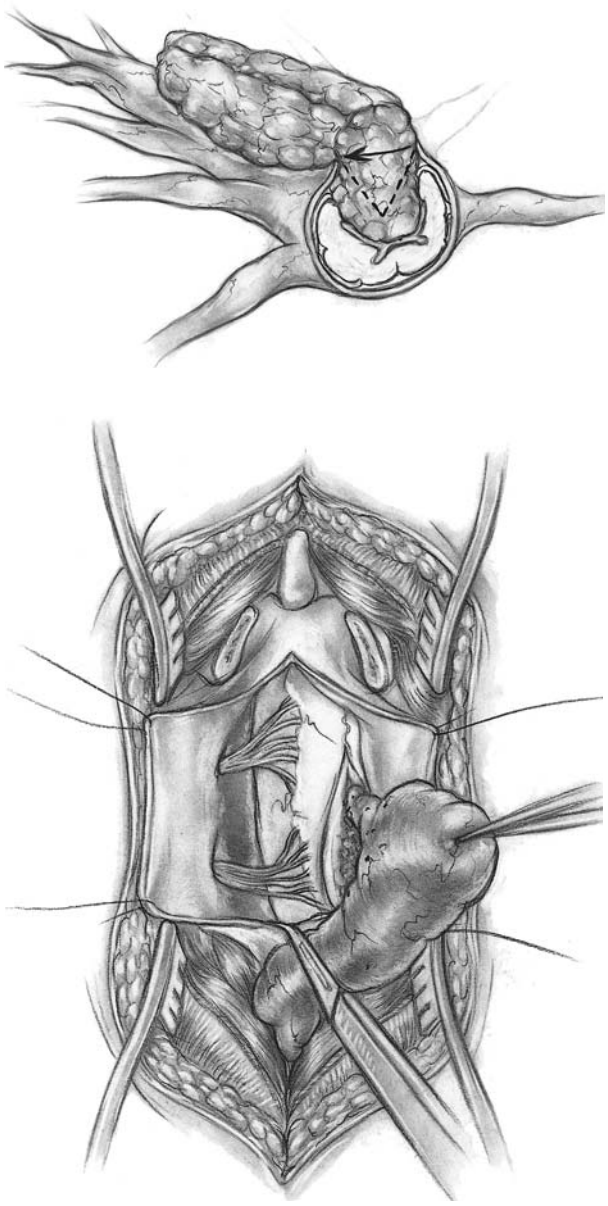

FIGURE 8–4. A: Cutaway view to demonstrate a dorsal lipoma with a larger subcutaneous component. This superficial mass must be removed to allow visualization of the intradural structures. Care must be taken in excising the subcutaneous mass to avoid all neural injury. The cord and sensory roots may be quite close to the dural insertion. **B:** Intraoperative view showing a hooked knife incising the critical interface of the dural insertion on the lipoma. The superficial fat can be debulked further at this point to increase visualization.

cantly greater. Lesions with a primarily caudal exiting of the fat, rather than dorsal exiting, also seriously complicate the situation. Reoperations for these lesions increase the risk of injury.

With the neural elements disconnected from the dura, a judgment is made as to how much additional fat should be removed (Fig. 8–4B). Ideally, the fat comes symmetrically from the dorsal aspect of the cord, allowing the neural tube to be reconstructed with small inverted sutures (Fig. 8–5). This may minimize postoperative adhesions. The dura is closed with a dural graft in a redundant fashion or primarily if the cerebrospinal fluid (CSF) space is large and the soft tissue is closed in layers in the usual manner. If a significant subcutaneous lipoma has been removed, a drain may be used above the fascia. It is essential to achieve a meticulous dural closure to minimize the risk of wound CSF leakage or pseudomeningocele formation.

Adequate hemostasis at each step protects the subarachnoid space from blood contamination and lessens the chance of postoperative adhesions necessitating reoperation. The risk of damage to the cord from extensive coagulation must be considered here.

A special and difficult situation is the cystolipomyelomeningocele, a caudally fixated lipomyelomeningocele associated with a terminal syrinx (Fig. 8–6). Generally, it is approached in same manner; however, the syrinx is opened to the subarachnoid space in an area that is likely to remain patent and minimize neural damage. Dissecting the exiting roots from the caudal fat is challenging. Closure of the dura is also a major problem because fat routinely infiltrates the dura even laterally. It is important to clearly delineate this lateral dural border in order to acheive secure dural graft placement and water tight dural closure.

Dermal Sinus Tracts and Dermoids

Dermal sinus tracts are epithelial tunnels that connect the skin surface with some deeper layer. In the spine area, they ultimately project to the embryologic center (central canal) of the spinal cord. Rarely can CSF be seen to come from them, but they still provide a conduit for bacterial contamination of the CSF with resultant meningitis. These lesions are uncommon and should not be confused with the common coccygeal pits that are of no clinical importance. Both MR imaging and ultrasound are inadequate to predict intradural extension of a dermal sinus tract.

If significant epithelial remnants are present, they continue to act as dermal or epidermal tissue and produce a mass composed of desquamated epithelium. These can occur along the course of the dermal sinus tract or within the spinal cord (Fig. 8–7). If a dermoid becomes infected, it will produce an abscess. The goal of surgery for these lesions is to disconnect and remove the

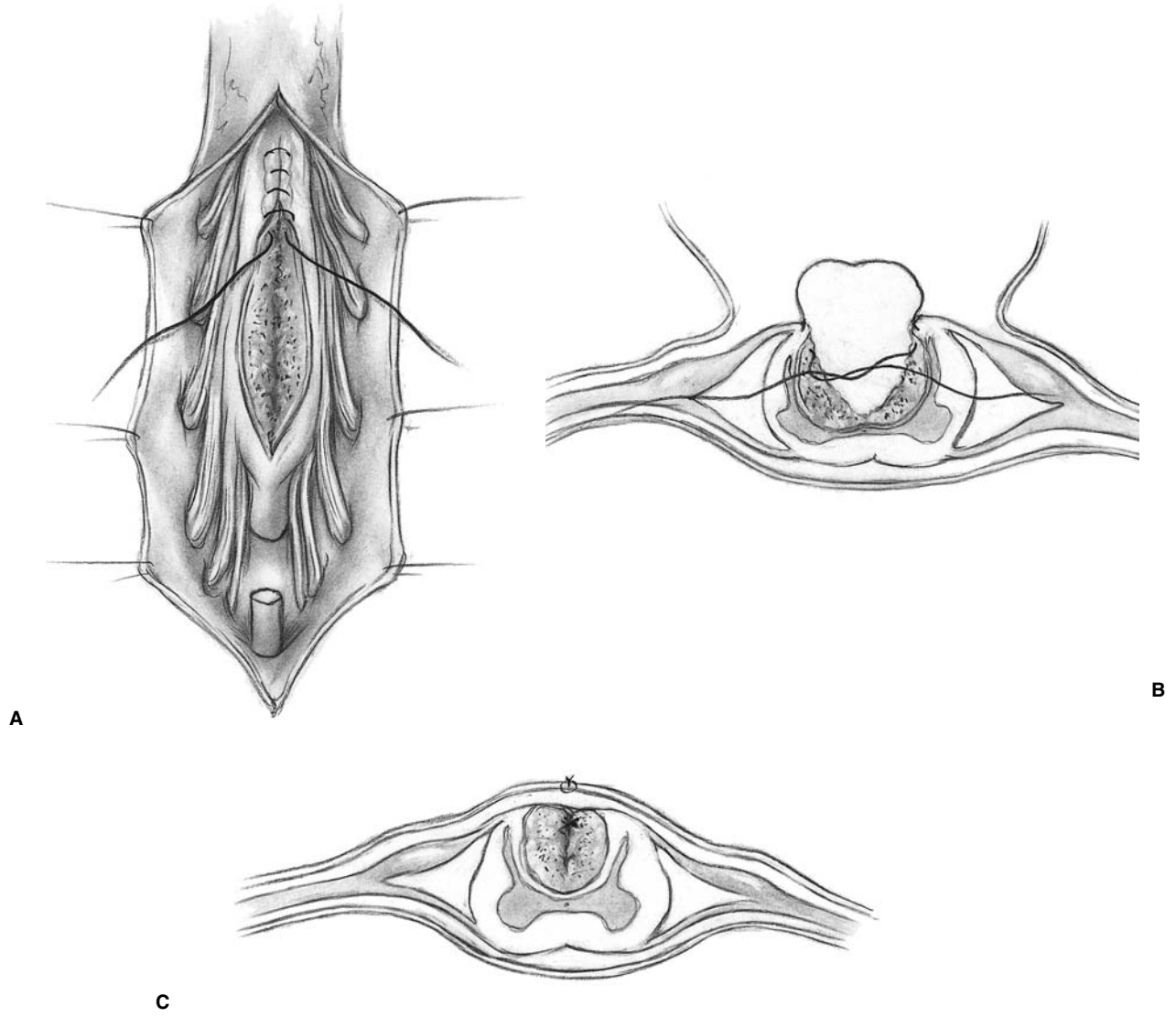

FIGURE 8–5. A–C: Intradural view with the bulk of the lipoma removed and inverted pial sutures being used to reapproximate the dorsal neural tube. Note that the filum has been sectioned.

epithelial elements, including an intramedullary extension if it were present. The typical location near the lumbosacral junction is associated with a cephalic subcutaneous course, making skin preparation above the tract important at the time of excision.

The mouth of the tract is incised and the tract followed. If it penetrates the lumbodorsal fascia, an intradural inspection is prudent. Small accumulations of pearly desquamated epithelium appear as "beads on a string." In small infants, cutting the cartilaginous spinous processes with stout scissors and then separating the lamina with a small retractor sometimes avoids a laminectomy. If the bone is more completely formed, a laminectomy or formal laminotomy is necessary. As the surgeon follows the tract within the dura, a sharp upward course frequently occurs, with the tract ending in the conus. Without clear evidence of a mass within the spinal cord, there is no justification for performing an exploratory myelotomy. The dura, bone, and soft tissues are closed in the usual manner.

Split Cord Anomalies

Split cord anomalies or diastematomyelia constitute a complex congenital anomaly of the spinal cord in which

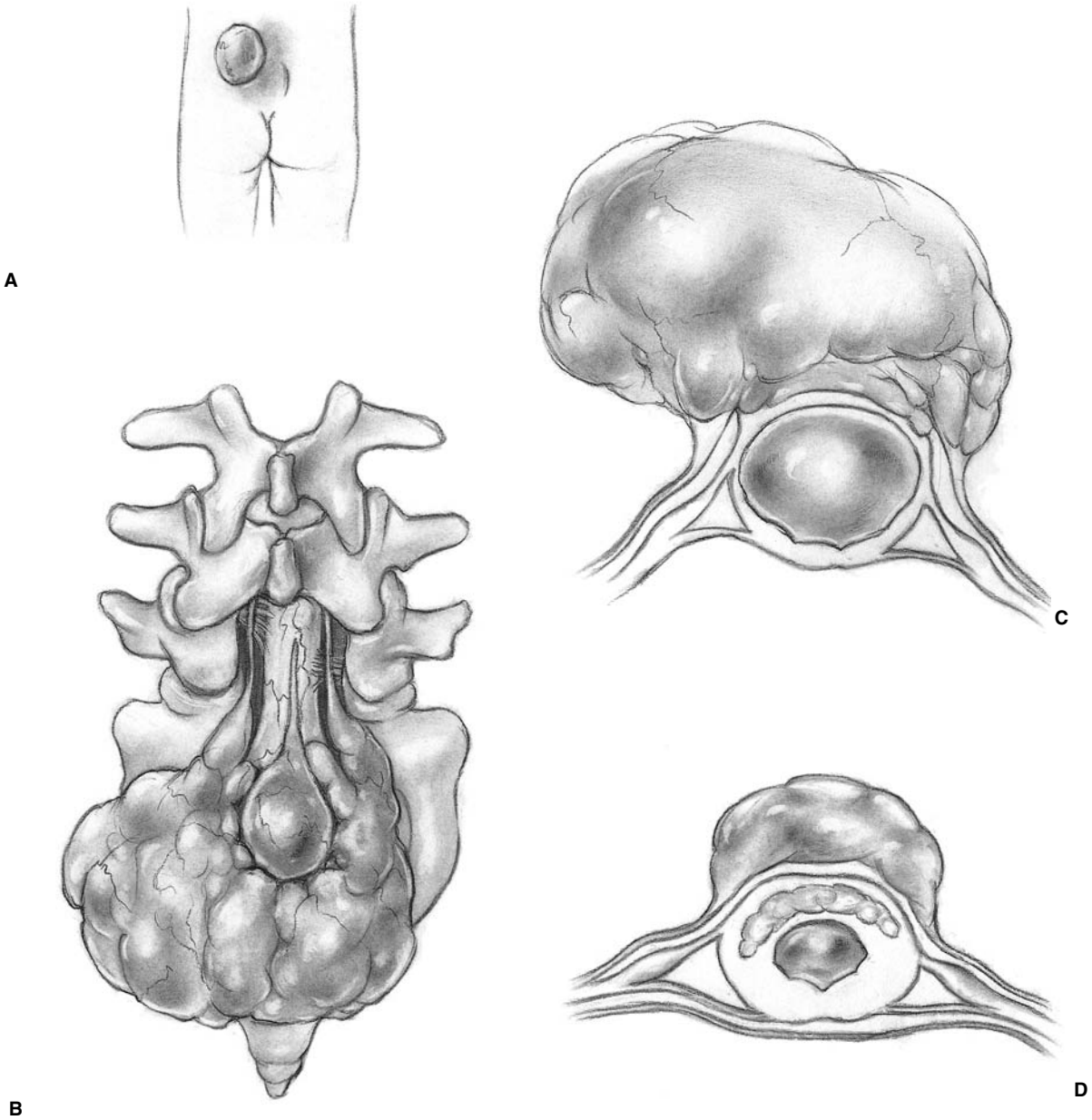

FIGURE 8–6. A–D: Cystolipomyelomeningocele. These lesions frequently have the terminal syrinx present as a bluish and somewhat transparent area just off the midline of the skin. In this case, the lipoma involves the caudal cord and dura. It must be disconnected distally while preserving the roots coursing through the fat. The syrinx is drained to the reconstituted subarachnoid space and the dura is closed. Again, the goal is not to remove every aspect of fat.

the two hemicords may or may not be separated by a median septum. Slightly more than half of patients have a telltale area of focal hirsutism marking the involved area (Fig. 8–8A). The two hemicords may unite below the median septum, or they may remain separate. Commonly, the filum is thickened and applies traction on the distal

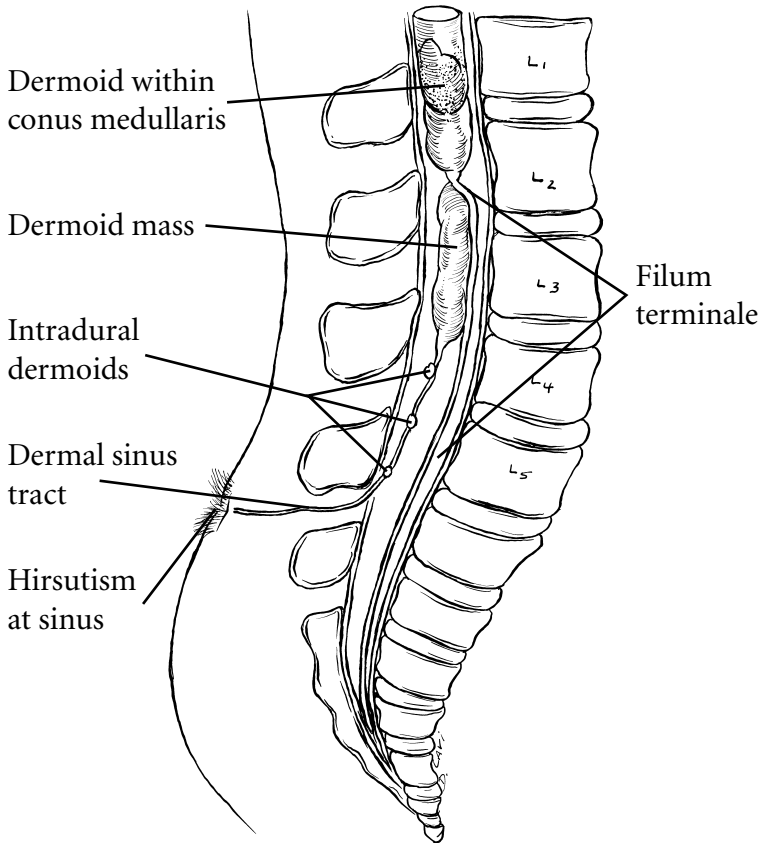

FIGURE 8–7. Midsagittal cutaway to demonstrate a lumbosacral dermal sinus and various locations of dermoids. The smaller accumulations along the filum are sometimes referred to as *beads on a string.*

cord. If the two hemicords remain separated, each may have a significant filum that must be sectioned. On the medial aspect of each hemicord, aberrant nerve roots, which serve no neurologic function, may exit the dura dorsally or occasionally ventrally. These bands (meningocele manqué) must be sought and sectioned to allow full spinal cord release. All these factors (filum, median septum, and medial aberrant roots) are potentially points of fixation and should be released if they are present. A median septum, either fibrous or bony, may project either cephalad or caudad (Fig. 8–8B) and may be rotated in the axial plane.

In planning the operation, the surgeon must have exposed the entire length of the spinal cord separation and the filum as it exits the conus. A midline incision is made in the skin, and the dorsal bony elements are exposed. Laminotomy, in an attempt to reconstruct the bone, should not be attempted. Median aberrant roots may penetrate the dura and attach to the under surface of the lamina. Removing the bone without first detaching these spinal cord connections can traumatize the cord. In addition, if a bony median septum is present, removal of the adjacent lamina without first disconnecting the septum may leverage this spicule into the cord causing harm. The laminae in the area of the median septum are almost always dysmorphic and act as an internal landmark for the most pathological levels. This bone is removed with rongeurs, taking care to remove each bite cleanly. If a bony median septum is present, the laminae around it are first removed; with the dura intact, the bone is drilled down as much as possible toward the base. The dura is incised circumferentially around the septum (Figs. 8–8C and D). This dural opening then is extended superiorly and inferiorly along the entire length of the cord separation. The medial aspect of each hemicord is inspected for exiting aberrant roots; if present, they are cut. Frequently, large vessels will penetrate the dural sleeve or the dura dorsally; these, too, are disconnected. With the hemicords gently separated, the dural sleeve is coagulated at its base and removed flush with the ventral surface of the dura (Fig. 8–8E). Additional bone removal may be necessary where the septum unites with the vertebral body, which obviously should be done in a controlled manner. There is little need to close the ventral dural defect. Hemostasis of the dural cuff may be difficult but should be approached systematically. With the septum and dural cuff removed, the ventral aspect of each hemicord is inspected with a right-angle hook and points of fixation released. The area of the connection between the conus and the filum is exposed, the filum

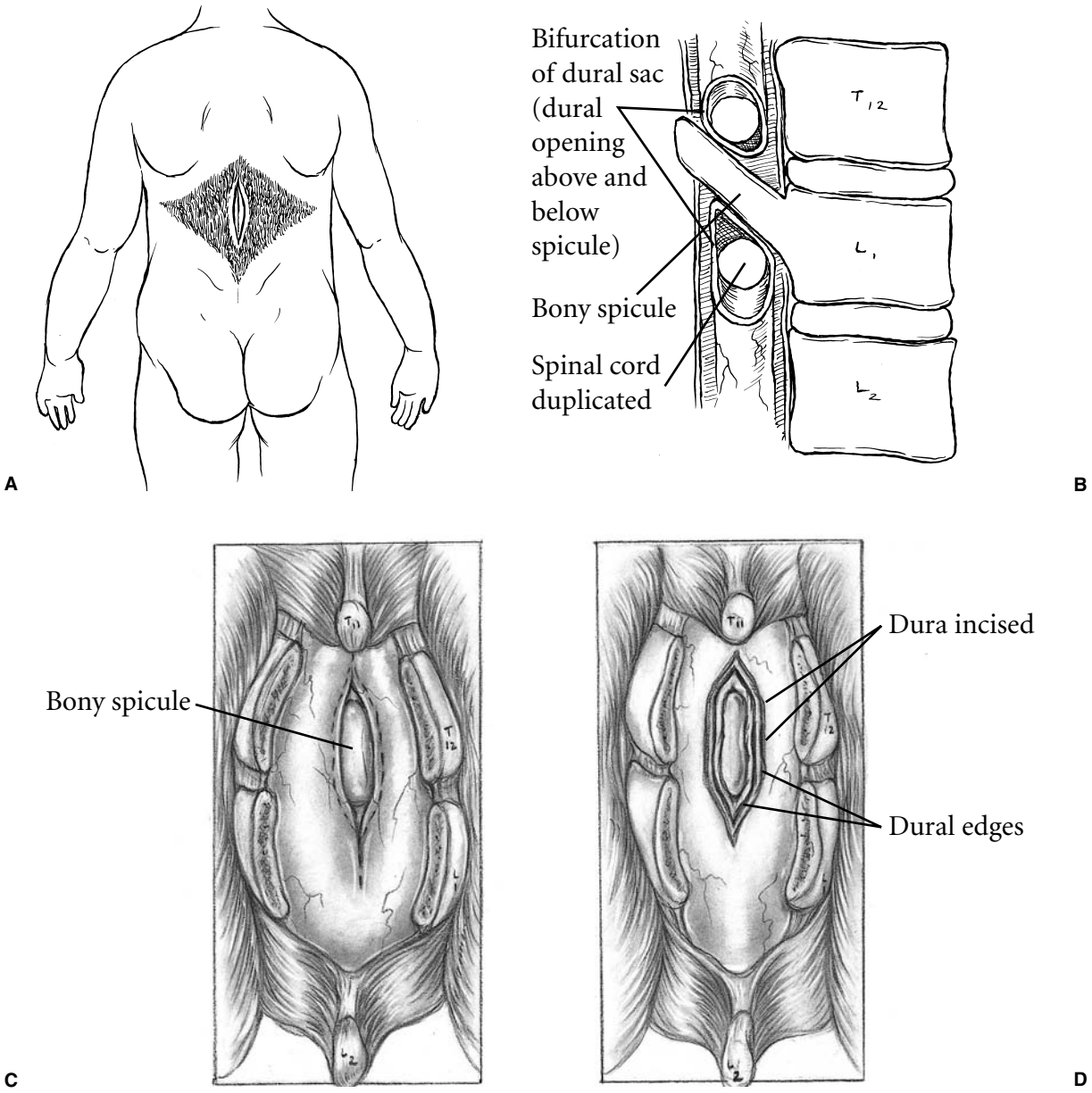

FIGURE 8–8. A: Infant with focal hirsutism at the lumbodorsal junction. The incision to expose the pathology is indicated. **B:** Midsagittal view of a split cord anomaly with one hemicord removed to show the cephalic course of a bony median septum. The dural sleeve also is appreciated. **C,D:** Dorsal intraoperative view with the dura intact and then incised to show the line of the dural incision. (*Figure continued next page.*)

separated, and a section removed. The dura is usually closed primarily.

Neurentic Cyst

These lesions, which can involve the spine, are composed of respiratory or gut epithelium that is misplaced. They may occur within the spine extradurally, within the intradural extramedullary compartment, or within the substance of the cord. They can be quite difficult to treat because the target tissue that needs removal is the wall of the lesion. The mucinous contents will reaccumulate if the lining is not removed. If the lining is adherent to the cord, removal can result in a significant neurologic injury. Judgment regarding the extent to which the surgeon

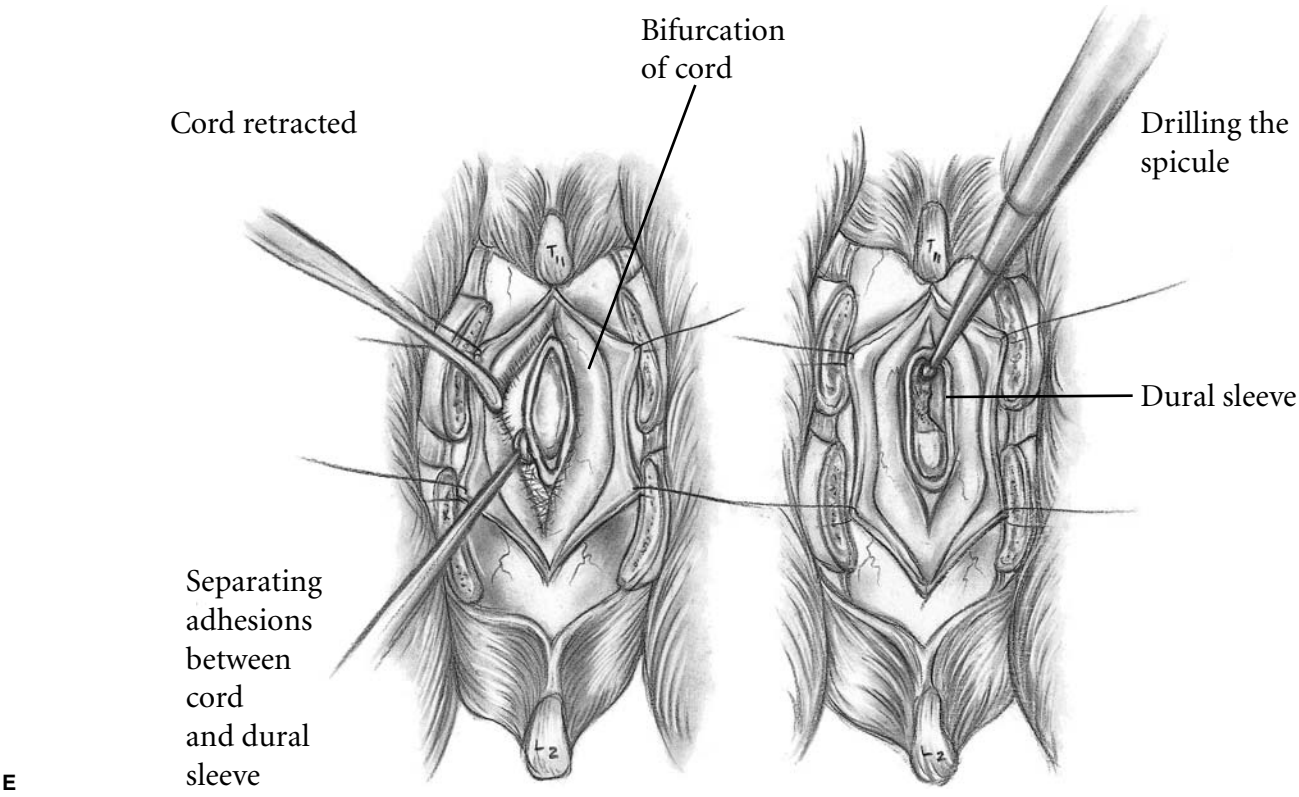

FIGURE 8–8. ***(continued)*** **E:** Similar view as **(C)** and **(D)** demonstrating the inspection of the dural sleeve looking for aberrant roots fixating the cord to this structure. Once all adhesions are disconnected, the remainder of the bony septum is drilled flush with the level of the vertebral body and the redundant dural sleeve is removed. **F:** Once the median septum is removed, the cephalic aspect of the filum is exposed for sectioning. Care is taken to do this well below the takeoff of any roots.

is willing to jeopardize the cord plays a significant role. The lesion may extend ventrally through a vertebral body defect (butterfly vertebrae), which complicates removal from a dorsal approach. Each lesion has unique characteristics, making generalization difficult. Total excision of the wall with preservation of neurologic function is the goal. Care must be taken to avoid subarachnoid space contamination with the cyst contents. Reoperation is rarely associated with total removal, making the initial procedure the only reasonable time to accomplish a cure.

Meningocele Manqué

The term means false meningocele and has been taken to refer to one of two situations. A partial thickness midline skin lesion (cigarette burn) associated with bony spina bifida can have fibrovascular strands fixating the dorsal surface of cord. These may attach to the partial thickness skin, making manipulation of the skin lesion painful as it applies traction on the dura and cord. Simply exploring the cord and disconnecting the strands and formally closing the dura will treat this lesion successfully.

The other situation where the term *meningocele manqué* is applied refers to the aberrant roots that come from the medial aspect of a split cord anomaly and generally course dorsally. They are nonfunctional and attach to the dura and then may terminate on the undersurface of the laminae. The therapy for both lesions is simply to cut and excise the fixating structures. More recently, the possibility of ventral fixation by these aberrant roots has been raised; this, too, should be sought.

Terminal Syringomyelia

This is the last of the pathological expressions of SBO and is the most poorly understood. Generally, CSF-like accumulations of fluid within the distal third of the spinal cord are termed terminal *syringomyelia.* They can be associated with any other form of SBO but are seen not infrequently with lipomyelomeningocele, split cord anomalies, and TSC. Treatment is to communicate them with the subarachnoid space with a small stent, such as a tympanostomy tube or other small shunt tubing. In general, I anchor the tubing to the pia with a fine stich. This requires a myelotomy, which never can be performed without a neurologic price, however small. In my experience, there is no need to shunt the syrinx fluid extradurally.

The more difficult question is not how to treat the lesion but rather which lesions should be treated and when. Small fluid accumulations that do not expand the cord and have a thick area of spinal cord dorsally should be left alone because more injury than benefit will occur from treatment. Lesions that markedly expand the cord so that only a thin rim of neural tissue is present dorsally should be stented as part of the procedure for the other associated findings. Most patients lie between these two extremes. My own approach is not to consider treatment of any terminal syrinx that does not significantly expand the size of the spinal cord. In a lesion that does expand the cord, the thickness of the dorsal cord that must be incised is considered along with the course of the patient over time. If the syrinx is clearly expanding on serial MR imaging, dorsal myelotomy is appropriate. If only a single MR image is available, the lesion is moderate in size, and the patient has no clear symptoms, then watching with serial imaging is more appropriate.

POSTOPERATIVE MANAGEMENT

Following operation, urinary drainage must be ensured. Constant or intermittent catheterization may be necessary. Assessment of postvoiding residual urinary volume may help to determine bladder integrity. Adequate nutrition must be maintained and bowel impaction avoided. Patients are generally nursed flat to lessen the likelihood of a CSF leak or pseudomeningocele. The period of maintaining this position is a function of the integrity of the closure. A small infant with normal dura submitted to sectioning of the filum may do quite well with little or no time spent lying flat. Multiply operated adults with fatty infiltration of the dura and extensive operative exposure may require weeks before adequate healing occurs and they are able to sit without a pseudomeningocele occurring. In general, 3 to 5 days are adequate for most wounds, but exceptions occur on both extremes.

Pain control in all patients is critically important. Patient activated pain control gives back an aspect of control that is lost by many children and adolescents. We have successfully used this option in children as young as 6 years of age. A special statement should be made for adequate pain relief in small infants. Although respiratory depression is a concern with most narcotic drugs, the alternative of not giving adequate pain relief is inappropriate. By assessing the infant's demeanor and resting pulse and knowing the extent of the procedure adequate pain relief can be safely given.

Patients with extensive procedures over the spinal cord, especially when intraoperative hemostasis has been a problem, should be observed in an intensive care unit. Procedures over the cauda equina with little blood loss can be observed routinely in a hospital room with the family assisting in the comforting process.

Generally, the development of a pseudomeningocele can be handled by time without reoperation, as long as skin integrity is maintained. Outside the setting of a terminal syrinx or a true mass lesion (neurentic cyst), postoperative imaging is not used routinely.

Avoidance of Complications

Most of the advice that could be included in this section has already been given or is obvious. The thoughtful sur-

geon will be able to avoid complications by obtaining images of the patient's pathology that clearly outline the anatomy that will need to be dealt with, providing adequate exposure of all the pathology, proceeding from normal into abnormal tissue, and minimizing the trauma to the nervous tissue while removing all tension and mass effects. Neural elements can be damaged by vascular injury and resultant ischemia more easily than by direct manipulation. Therefore, the surgeon should try to minimize the contamination of blood and other fluids in the subarachnoid space, leave as much intact pial surface over the cord and roots as possible to minimize postoperative adhesions (arachnoiditis), achieve adequate dural closure without constricting the intradural contents, use dural grafts freely if any question exists regarding compression of the neural elements, avoid significant dead spaces, and close soft tissues without hematomas or large areas of nonviable tissue damaged by thermal or electric dissection that can both act as a source of infection and poor healing. Although these principles seem self-evident to an experienced surgeon, the challenge is to put them into action.

EDITOR'S COMMENTARY

The advent of MRI has led to increased recognition of the occult forms of spina bifida and has facilitated early diagnosis, often in otherwise asymptomatic patients. Because patients with tethering lesions often demonstrate progressive deterioration over time, many pediatric neurosurgeons favor proceeding with untethering procedures shortly after diagnosis. A notable area of controversy involves the management of patients with lipomatous malformations, which exhibit a high risk of recurrent postoperative tethering with late functional deterioration that often necessitates a technically difficult reoperation. Some neurosurgeons therefore defer the initial untethering procedure until progressive symptoms and signs occur, rather than operating prophylactically. This chapter nicely emphasizes the fact that many patients with SBO have multiple tethering lesions (e.g., a midline bone spur, meningocele manqué, and thickened filum terminale), and such "tandem" lesions should be anticipated and appropriately treated.

PEARLS

In this author's experience:

- Care must be taken while removing the lamina during SBO surgery. Dorsal bands originating from the dura or spinal cord may insert on the undersurface of the lamina.
- All patients with some form of caudal agenesis or hypogenesis should have imaging of the spine to rule out a tethered spinal cord.
- When exploring the spine at a higher level than the conus (e.g., for split cord malformation), the MR scan should be studied closely to assess the need for division of the filum terminale. If this is deemed necessary, the filum may need to be exposed through another incision or an extension of the same incision.
- In the presence of a large syrinx, simple untethering of the spinal cord is not sufficient. In such cases, the syrinx should also be stented to the subarachnoid space.
- To be considered pathological (i.e., tethered), a conus at a normal level should be accompanied by objective findings of SBO, such as lamina defects, cutaneous signatures, neurological defects, or all three referable to dysfunction of the lower spinal cord or conus.
- The surgical exploration of lipomyelomeningoceles is technically demanding of precise knowledge from the surgeon. These lesions should be operated on by an individual with special knowledge and expertise in this area. Of all the lesions in pediatric neurosurgery, this lesion is worthy of referral to a recognized expert because long-term normal function is likely to result from exploration by a knowledgable individual and surgical misadventure is likely to result without this expertise.

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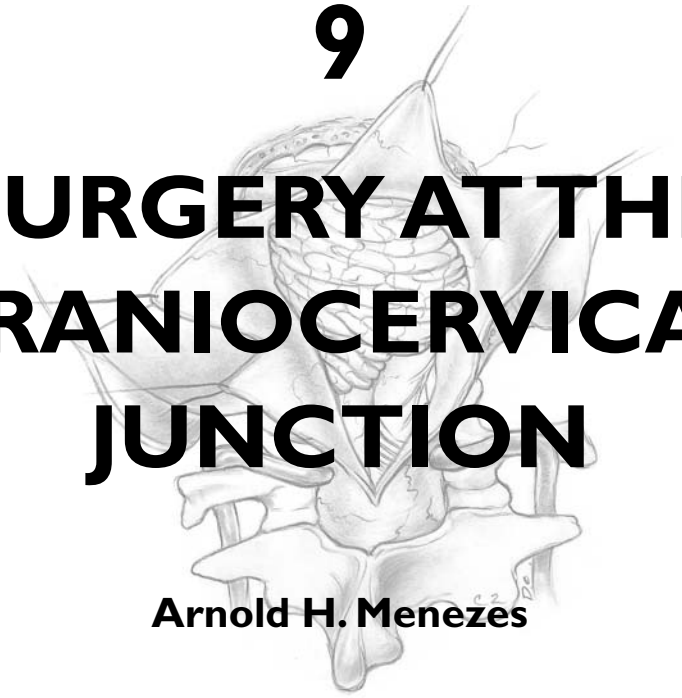
9

SURGERY AT THE CRANIOCERVICAL JUNCTION



Arnold H. Menezes

The pathology of abnormalities at the craniocervical junction is extensive. A complete knowledge of the bony anatomy, embryology, and biomechanics of the craniocervical junction is necessary to understand the etiology of the abnormalities in this area and, thus, to plan their treatment. Congenital, developmental, and acquired lesions arise at the craniovertebral junction to produce changes that ultimately affect the neural structures. The mainstay of decompression of the posterior fossa and upper cervical spine has been by the dorsal route, at times with a fusion; however, the morbidity and mortality of such procedures, especially when the indication is a ventrally situated abnormality, are high. Operative procedures based on an understanding of the surgical pathology of this region have been developed and are outlined in Table 9–1.

PREOPERATIVE EVALUATION

Congenital abnormalities at the craniocervical border are now being recognized with increasing frequency at birth and require a careful understanding of the pathophysiology to undertake management. Congenital torticollis as a result of atlas assimilation and rotary subluxation of the atlantoaxial articulation are not uncommon. Similarly, syndromic disturbances, such as the Goldenhar's syndrome, Klippel-Feil syndrome, fetal warfarin syndrome, and Conradi's syndrome, are but a few examples that may be associated with craniocervical abnormalities and cervicomedullary dysfunction.

The diagnostic imaging of these infants is the same as that of the young child and the adolescent: plain radiographs, computed tomography (CT), and magnetic resonance (MR) imaging. The factors that influence specific treatment are (1) the reducibility of the lesion, the most important factor, which implies restoration of anatomic relationships of the craniospinal axis using position as well as traction; (2) the mechanics of compression and the direction of encroachment; (3) the etiology of the lesion, such as syrinx, Chiari malformation, vascular abnormalities, or basilar invagination; and (4) the presence of epiphyseal growth plates and ossification centers in congenital abnormalities. Figure 9–1 outlines the decision tree for treatment of craniocervical abnormalities. In the neonate as well as the young infant younger than the age of 2 years, craniocervical immobilization is the mainstay of treatment and is accomplished with custom-built orthoses that are changed at 3-month intervals. At the age of 2 years, the child is reassessed for surgical management.

Halo-Ring Placement

Indications for halo traction in the pediatric population are (1) instability and correction of alignment; (2) stabilization during surgery for the cervical spine and the cran-

TABLE 9–1.
Surgical Approaches to the Craniovertebral Junction in Children

- Approaches for decompression
  - Ventral
    - Midline
      - Transoral-transpharyngeal
      - Transpalatal-transseptal
      - Lateral rhinotomy
      - LeForte I with maxillary "down fracture"
    - Lateral
      - Lateral transcervical extrapharyngeal
      - Preauricular infratemporal
  - Lateral
    - Lateral transcervical with translocation of vertebral artery and partial resection of occipital condyle
  - Dorsal
    - Midline posterior fossa and upper cervical posterolateral decompression
- Fusions
  - Atlantoaxial
    - Interlaminar
    - Modified Gallie/Brooks
    - Transarticular C-2-C-1 screw fixation
  - Occipitocervical
    - Dorsal interlaminar
    - Titanium loop
    - Plates with screws or cables

iovertebral junction; (3) immobilization of the craniocervical region and cervical and upper thoracic spine; and (4) prevention of potential instability. Contraindications for halo-ring placement are (1) age, because it should be avoided in infants younger than 2 years old; (2) scalp infections; (3) inexperience of the surgeon; (4) the presence of bone-softening syndromes; and (5) cardiovascular or respiratory abnormalities or infants prone to seizures.

Before placement of the halo ring, the occipitofrontal circumference must be measured and an appropriate pediatric crown halo obtained. It is important that the pediatric pins be allowed to fixate perpendicular to the cranium and at least a 2- to 2.5-cm space be available between the halo ring and the scalp for cleansing as well as for ease of placement of the halo-immobilizing vest. It is important to assess the skull thickness by age as well as by obtaining a head CT scan. The halo ring is placed with the patient under intravenous sedation or general anesthesia. If the latter is used, a cervical collar must be placed around the neck to correct the instability, and paralyzing agents should be avoided. The head is placed on a positioning board that also supports the upper trunk. The crown halo is placed over the cranium, ensuring that it is slightly below the cranial equator. Positioning pins are fixated temporarily with suction cups to the scalp to maintain the halo ring temporarily in place. This allows cleansing of the scalp with 10% PVP-iodine and local infiltration of the pin sites. In children younger

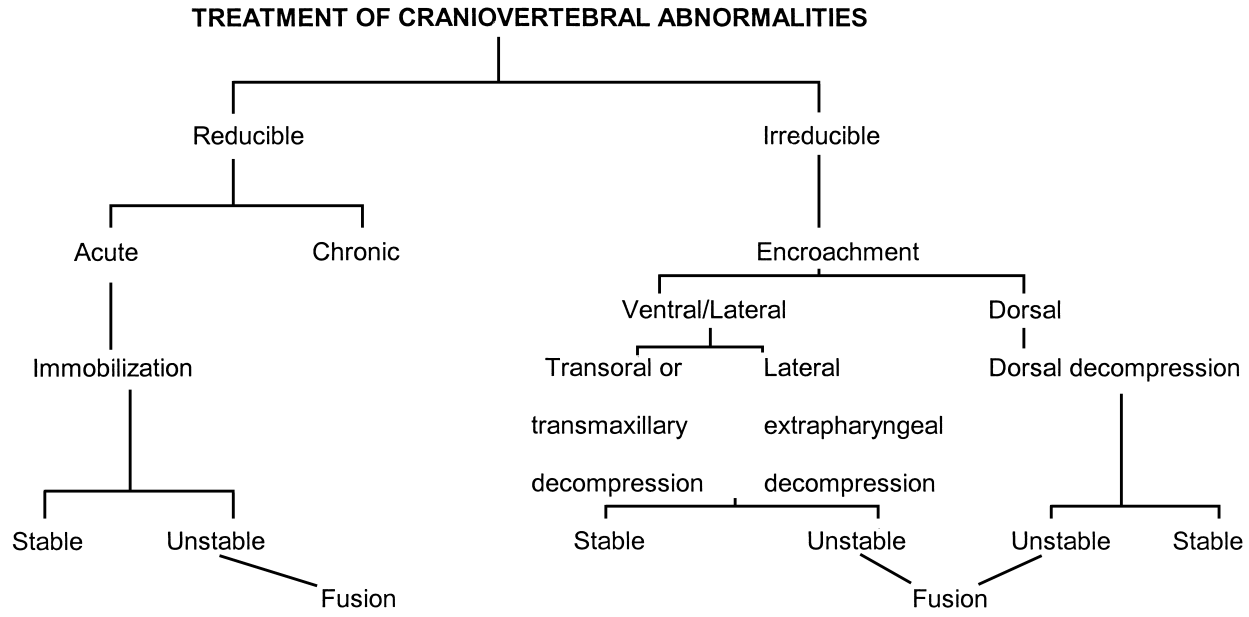


FIGURE 9–1. Treatment of craniovertebral abnormalities.

than the age of 4 years, 8 to 10 pediatric pins are used. In patients 4 to 8 years of age, 6 pins are recommended. Four pins can be used in patients older than 8 years. These consist of one frontal and one parieto-occipitotemporal pin on each side. These pins are screwed in place, and iodine ointment is placed at the "business" end of the pin; the pin is advanced to touch the scalp. All pins must be brought to position on the scalp before being tightened. Tightening is done with diagonal pins tightened to the recommended pressures. Pressures recommended below in children less than 2 years of age are that which is generated between the thumb and index finger, usually about 1 to 1½ pounds of torque pressure. In patients 2 to 4 years of age, 2 pounds of torque pressure is recommended and 4 pounds in those between the ages of 4 and 6 years. In children between the ages of 6 and 10 years, 4 to 6 pounds of torque pressure is used. The pins are tightened, and a hex nut that accompanies the halo pin is locked into position. Again, at the end of 48 hours, this position is rechecked with the patient under sedation. A bucket handle is attached to the crown halo and placed in a neutral position. The traction that I use in the operating theater is 2 to 3 pounds for a child at 2 years of age and 3 to 4 pounds of traction pressure in a patient between the ages of 2 and 4 years.

The complications of halo-ring and halo-pin placement are (1) penetration of the skull and perforation; (2) injury of the supraorbital and supratrochlear nerves; (3) dislocation of the upper cervical spine as a result of excessive traction; (4) respiratory embarrassment resulting from laryngeal injury; (5) dislodgment of the ring from the head; (6) local infection and cellulitis; (7) loosening of the pins; and (8) os odontoideum formation.

It is extremely essential that halo-ring traction placement in the pediatric population be only carried out by an experienced team comprising a pediatric neurosurgeon/orthopaedic surgeon, radiologist, intensive care nursing staff, and physicians conversant in the problems mentioned. Cervical spine radiographs must document the effects of the traction immediately as well as at close intervals, at times on a daily basis.

TRANSORAL–TRANSPALATOPHARYNGEAL DECOMPRESSION OF THE CRANIOCERVICAL JUNCTION

Indications

The transoral–transpalatopharyngeal approach to the craniovertebral junction is the most frequently used route for ventral decompression of the cervicomedullary junction. It is indicated only in non-reducible ventral extradural osseous compression (Fig. 9–2) of the cervicomedullary junction and at times with extradural tumors, such as chordoma and osteoblastoma. It is contraindicated in most

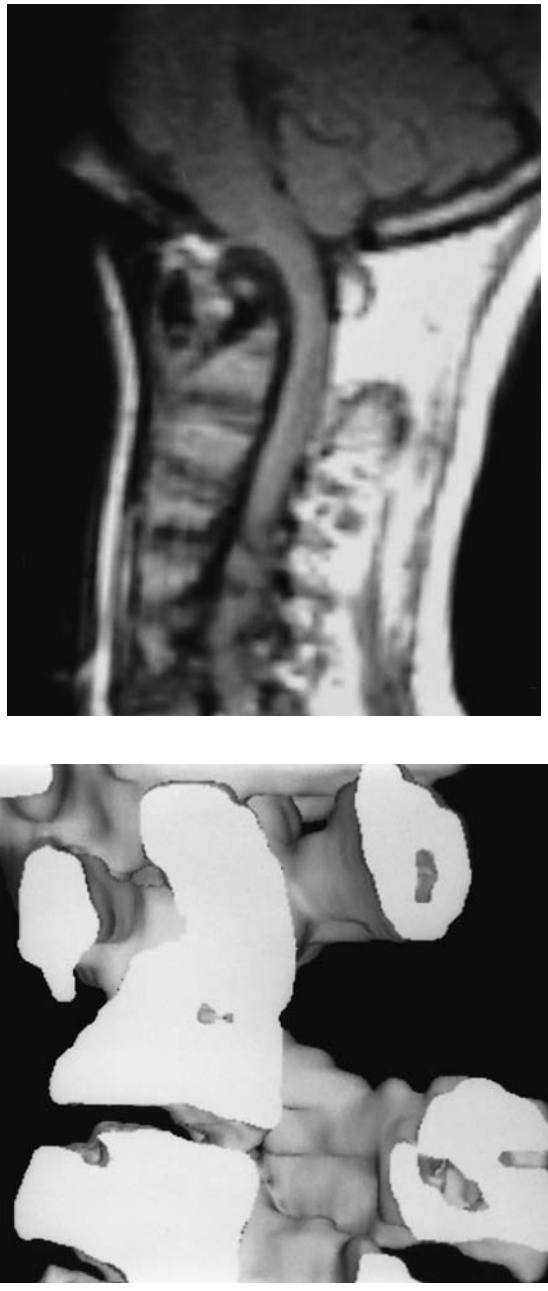


FIGURE 9–2. **A:** Midsagittal T_1-weighted MR image of the craniocervical junction in a 10-year-old boy with fixed atlantoaxial dislocation causing severe ventral cervicomedullary compression. **B:** Three-dimensional CT scan of the upper cervical spine and craniocervical junction with midsagittal visualization of the fixed atlantoaxial dislocation and severe compromise of the spinal canal.

cases of intradural pathology or in extradural pathology that is more than 15 mm from the midline.

The nutritional status of children undergoing transoral decompression of the craniocervical junction must be evaluated. The achievement of a high caloric intake is feasible in some children with nasogastric tube feedings or at times with intravenous hyperalimentation. This is particularly important in the preoperative and perioperative management because oral intake is not permitted during the first postoperative week. It is also important to be able to achieve a working distance of 2.5 cm between the upper and lower incisor teeth in the young child to make the transoral route feasible. If this is not possible, a median mandibular split with midline glossotomy may be essential to the transoral operation. We prefer not to use the LeForte I dropdown maxillotomy in children below the age of 8 to 10 years because of disturbance to the anterior growth plate at the nasal septum. If a procedure is called for to the sphenoid, ethmoid, or lower clivus, a transpalatine approach can be used. It is important to bear in mind that even though lesions may appear to be irreducible, the stabilization of cervical traction is essential during the ventral transoral operation because of potential instability as well as that which may be realized after the destabilization of the ventral decompression and anesthesia.

Oropharyngeal cultures are obtained 3 days before a surgical procedure. No antibiotics are given unless pathological flora are present. If the lower cranial nerves are compromised, it is imperative that preoperative assessment of the respiratory function as well as swallowing mechanism be made to determine whether a tracheostomy is needed at the time of the transoral operation or immediately thereafter.

Intraoperative Technique

The child is transported to the operating theater in skeleton traction on a fracture bed with an MR imaging compatible halo at 5 to 6 pounds of traction in the older child in whom preoperative traction is possible. In the young child, the halo ring is applied after general anesthesia is obtained. This is achieved with a cervical collar in place and mask induction, followed by fiberoptic oral endotracheal intubation made through the mask; subsequently, the halo ring is applied. Nasal endotracheal intubation is avoided because it disrupts the integrity of the high nasopharyngeal mucosa, which is the avenue of approach to the craniocervical border. The head then rests on a padded Mayfield horseshoe headrest; traction is maintained over a pulley bar at the weights recommended. In an older child, that is, between the ages of 12 and 16 years, an awake intubation is made, and the patient is positioned. The awake patient is examined to ensure that no neurologic change has occurred during positioning, after which general endotracheal anesthesia ensues. In the last 8 years in our institution, a tracheostomy has not been performed in a child or an adult undergoing transoral–transpalatopharyngeal approach to the craniocervical junction. Following intubation, a gauze packing is placed to occlude the laryngopharynx and to prevent secretions and blood from draining into the stomach.

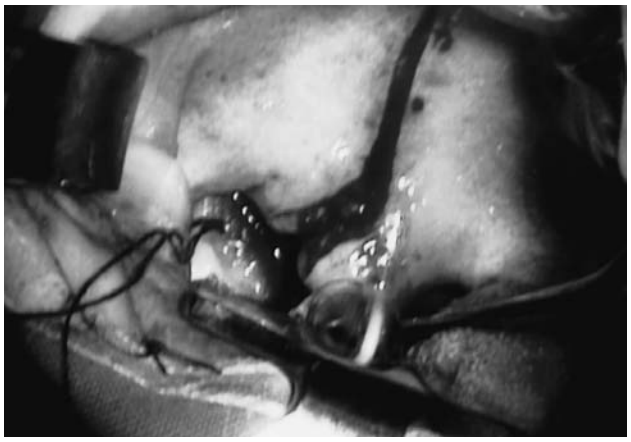
A modified Dingman self-retaining mouth retractor secures the endotracheal tube and allows exposure of the oral cavity as well as the pharynx (Fig. 9–3A). In procedures that involve the foramen magnum and clivus, it is essential to split the soft palate to provide the necessary exposure.

The operating microscope provides magnification and a concentrated light source. The oral cavity is cleansed with 10% povidone iodine and hydrogen peroxide and then rinsed with saline. A midline incision is made into the soft palate extending from the hard palate to the base of the uvula and deviating to one side. Stay sutures are applied to the incised soft palate, retracting the flaps to either side exposing the high posterior nasopharynx down to the C-3 vertebral level (Fig. 9–3B). The posterior pharyngeal wall is first topically anesthetized with 2.5% cocaine and the median raphe with 0.5% lidocaine solution with 1:200,000 epinephrine.

A midline posterior pharyngeal incision is made from the midclivus to the upper border of C-3. The pharyngeal flaps are retracted to either side with stay sutures, and the longus colli and longus capitis muscles are dissected free of their osseous-ligamentous attachment to expose the lower clivus, the anterior arch of C-1, and the body of C-2. Lateral exposure is limited to 1.5 cm to either side of the midline in a child to preserve integrity of the hypoglossal nerve superiorly, the eustachian tube orifice, and the vertebral arteries in descending order.

For an inexperienced surgeon working in the transoral area, it is extremely important to recognize that congenital abnormalities with a foreshortened clivus mandate that there is platybasia and that the clivus is far from the hard palate. This situation necessitates removal of the caudal hard palate to achieve exposure with the clivus.

The anterior arch of the atlas and the caudal clivus are removed using a high-speed drill. The soft tissue ventral to the odontoid process is resected with rongeurs (Fig. 9–3C). The odontoid process then is removed in a rostral caudal dimension using the diamond burr; the pannus then is excised (Fig. 9–3D).



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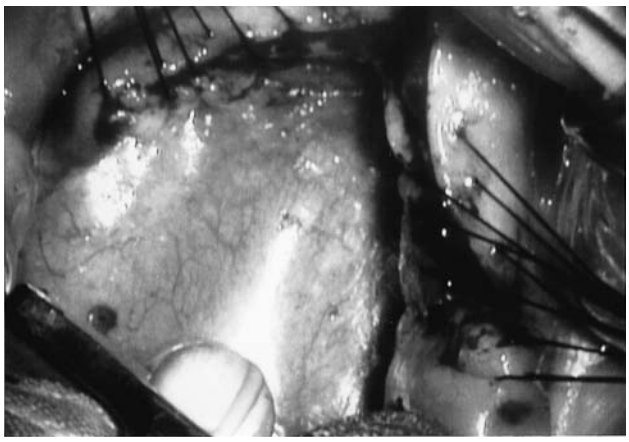

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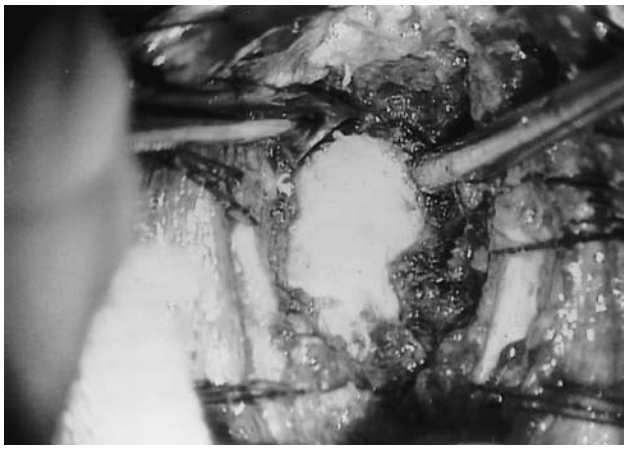

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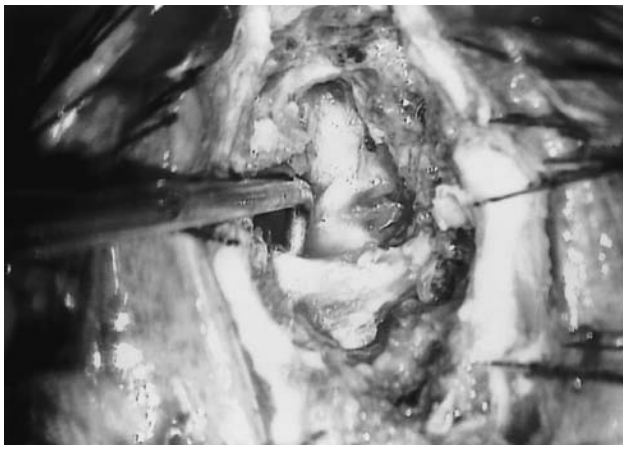

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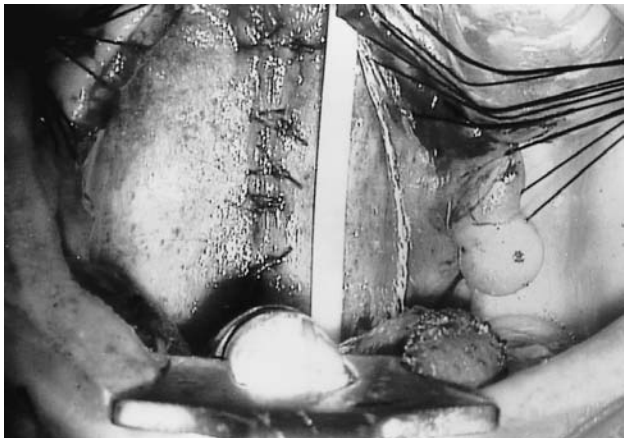

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FIGURE 9–3. **A:** Operative photograph through the microscope with Dingman mouth retractor in place. The endotracheal tube and the tongue are depressed with the tongue blade. The cheek retractors improve the illumination and visualization. The incision in the soft palate starts at the hard palate and proceeds to the base of the uvula and skirts to one side. **B:** The leaves of the soft palate are held apart with stay sutures to expose the mucosa of the high nasopharynx. **C:** The posterior pharyngeal musculature as well as the longus colli and longus capitis muscles are swept to either side of the midline to expose the odontoid process after the anterior arch of the atlas has been resected. Note the apex of the odontoid process here. **D:** The odontoid process has been resected and a right angle separator identifies the transverse portion of the cruciate ligament. **E:** The longus colli and longus capitis muscles have been approximated as is the posterior pharyngeal wall. A nasogastric feeding tube is visualized.

Pannus signifies instability. It should be resected but not beyond the limits of the exposure. Bipolar cauterization will allow shrinkage. Division of the odontoid process at its base and downward traction should be avoided. It is tempting to proceed with this; however, the retained fragments of odontoid tip are testimony to inadequate operative procedure and an inexperienced surgeon. Depending on the bulging of the dura into the wound, the tectorial membrane may or may not be divided. The cruciate ligament usually is visualized at the caudal aspect of the bony exposure. It is wise to leave the transverse portion of the cruciate ligament intact for partial stability of the atlas.

Initially, I used median-nerve sensory evoked responses, but they were not useful—an especially important consideration in children with severe cervicomedullary compromise. The somatosensory evoked responses usually cannot be obtained.

If a tumor such as chordoma is encountered, direct visualization of its extent is needed before piecemeal resection is done. Undue traction must be avoided because a pedicle onto the vertebral artery or a major branch may be present.

Resection of an intradural lesion requires relieving cerebrospinal fluid turgidity by having previously placed a lumbar subarachnoid drain. The dura is opened in a midline fashion, extending up as high as necessary into the ventral posterior fossa. This cruciate dural incision is made by converting the vertical incision to a cruciform one below the foramen magnum, thus avoiding the circular sinus. Closure demands that an attempt be made to bring together the leaves of the dura in a watertight fashion insofar as possible. If the dura has been violated, it is closed and backed with fascia harvested from the external oblique aponeurosis, after which a fat pad is placed before the posterior pharyngeal wall is closed.

The longus colli and longus capitis muscles are approximated in the midline with subsequent anatomic layered closure of the posterior pharyngeal musculature and the posterior pharyngeal mucosa. I use 3–0 polyglycolic suture. A soft feeding tube is inserted through the nose into the stomach and allows feeding in the early postoperative phase (Fig. 9–3E).

Closure of the soft palate is done by bringing the nasal mucosa together with interrupted sutures, and subsequently the muscularis and the oral mucosa are approximated with a through-and-through interrupted vertical mattress suture of similar strength. The patient is maintained in a Philadelphia collar and turned over for the fusion procedure, most often carried out using the same anesthetic (Fig. 9–4B).

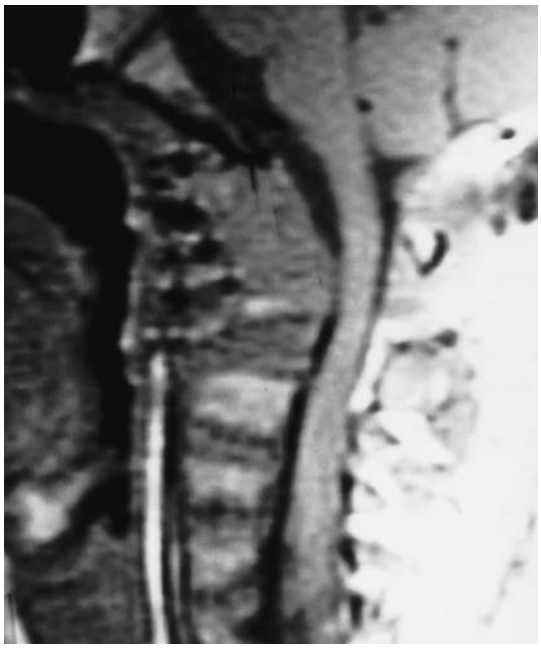


FIGURE 9–4. A: Postoperative T1-weighted MR image of the craniocervical junction demonstrates the ventral osseous decompression of the cervicomedullary junction. **B:** Three-dimensional CT scan of the craniocervical junction in the frontal plane reveals the resection of the odontoid process into the midportion of the axis body.

Postoperative Management

No oral intake is permitted for the first 5 days. Nasogastric tube feedings are initiated on the day after surgery and are followed by a gradual increase in feeding until a regular diet is resumed 15 to 18 days after the surgery. If the dura was opened, intravenous antibiotics (cefotaxime, metronidazole and methicillin) are administered and spinal drainage maintained for 10 days after the operation. The endotracheal tube is removed only after lingual swelling has receded, usually after an average of 3 to 4 days. Table 9–2 outlines the possible complications of this procedure and the treatments.

LATERAL TRANSCERVICAL EXTRAPHARYNGEAL APPROACH TO THE CRANIOCERVICAL JUNCTION

Indications

This approach has been of interest to surgeons who fear contamination of the oral cavity. The advantage is the ability to proceed with a ventral fusion if this is necessary and

TABLE 9–2.
Perioperative and Delayed Complications in Transoropharyngeal Craniovertebral Junction Surgery and Their Management

Complication	Prevention and Management
Perioperative complications	
Unnecessary transoral procedure	Preoperative reduction if possible
Damage to eustachian tubes and hypoglossal nerves	Limit lateral exposure to 2 cm from midline
Too small an oral exposure	May need to split mandible and tongue
Inability to reach clivus due to platybasia	Divide soft palate and possibly hard palate; intraoperative fluoroscopy
Lost! cannot reach or resect odontoid or epidural masses	Fluoroscopy; start resection at the rostral end
Persistent bleeding	Pannus and arterial bleeding must be controlled with bipolar coagulation; bleeding from circular sinus needs Avitene and Oxycel; else clip both leaves of dura
CSF leakage; intra-arachnoid lesion	Preoperative lumbar drain; attempt dural closure; fascial graft and fat pad and plasma glue; CSF drainage × 1 wk. Triple antibiotics × 10 days
Delayed complications	
Severe tongue swelling	Decadron; intermittent release of tongue depression; retain dental guards in children
Meningitis	CSF examination and lumbar drainage; no oral intake; antibiotics; close leak
Palatal dehiscence	Immediate reclosure
Pharyngeal dehiscence	<1 wk - reclosure; >1 wk hyperalimentation and antibiotics
Neurological worsening	Check alignment - traction: meningitis; abscess? retained lesion? vascular compromise?
Retropharyngeal abscess	Check for osteomyelitis and meningitis; extrapharyngeal drainage
Delayed pharyngeal bleeding	Secondary infection; rule out osteomyelitis and vertebral artery erosion with false aneurysm; MRI and angiography
Velopalatine incompetence	Usually appears 4–6 mo postoperatively; pharyngeal retraining; may need pharyngeal flap

CSF, cerebrospinal fluid; IV, intravenously; MRI, magnetic resonance imaging.

also the potential for placing C-2 to C-1 lateral mass screws. This procedure is difficult, however, because it goes into a narrowed pyramid at the apex with restriction of exposure.

There is a high risk to injury of the hypoglossal and the glossopharyngeal nerves and difficulty in visualizing deep midline structures of the true craniocervical border. Likewise, the clivus is poorly visualized using this approach.

Intraoperative Technique

The awake patient undergoes fiberoptic intubation through a nasal endotracheal route. The oral cavity is kept free of any tubes. The same precautions are taken as with the transoral route regarding anesthesia and intubation.

The cervical incision starts behind the ear, proceeds over the mastoid process, and extends 1.5 cm below the angle of the mandible toward the midline above the hyoid bone. An inferior extension of this at the level of the sternomastoid muscle converts the transverse incision into a T. The head is turned to the left in the case of a right-handed surgeon. The extent of the vertical limb of the incision depends on the amount of cervical spine that needs to be exposed. The incision traverses through the subcutaneous tissue and the platysma. Subcutaneous dissection allows mobilization in a subplatysmal plane of the superficial fascia. The inferior division of the facial nerve is identified, and facial nerve dissection into the parotid gland may be needed for mobilization. The superficial draining veins into the jugular vein are dissected free and ligated prior to their entrance into the common facial vein or the jugular vein. Superficial branches of the facial nerve are protected by staying deep to this vein.

The deep fascia at the anterior border of the sternomastoid muscle is incised allowing for visualization of the carotid sheath. Dissection is made anterior to the vascular structures when the midline and the opposite side of the craniocervical junction are necessary. The submandibular salivary gland may be elevated. Resection of the submandibular salivary gland has no consequences if its duct is sutured properly to prevent a salivary fistula. The nodes in the carotid and the digastric triangle are removed. The posterior belly of the digastric is traced to its tendon, where it is transected and transfixed with a suture for subsequent reapproximation.

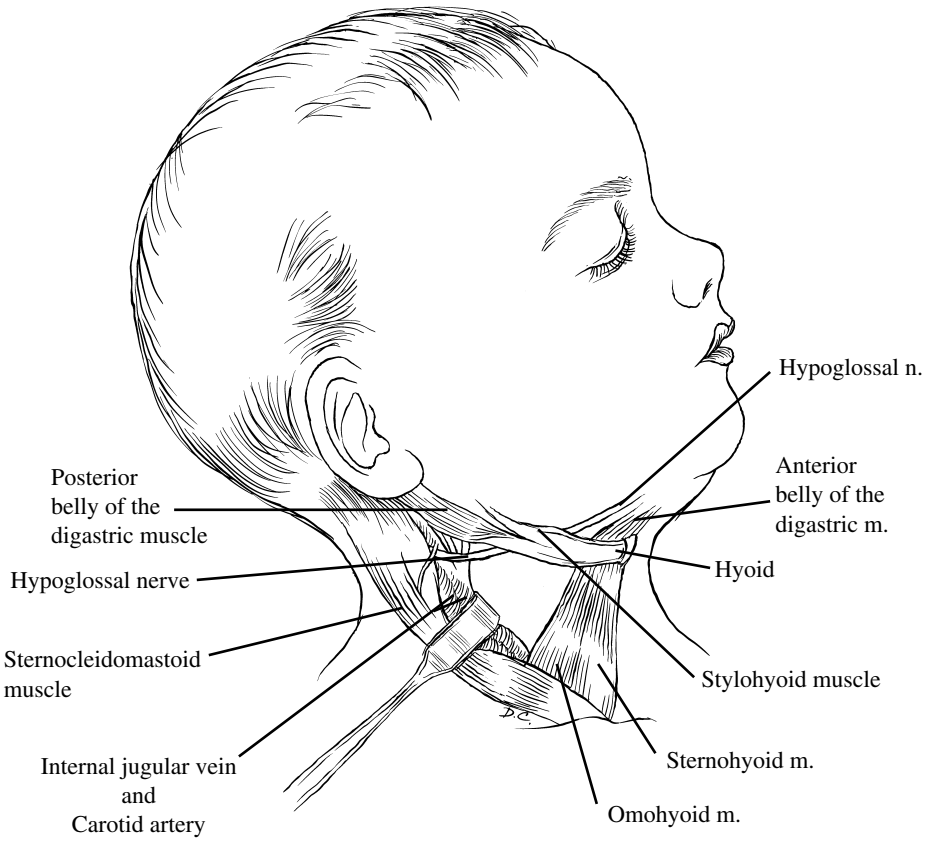


FIGURE 9–5. Dissection for the lateral extrapharyngeal approach to the craniocervical junction. The vascular compartment, together with the sternocleidomastoid, is retracted laterally, and the pharynx exposure is made above the hypoglossal nerve.

The stylohyoid muscle is divided allowing for medial retraction of the laryngopharynx. Care is taken to identify the hypoglossal nerve as it swings between the external carotid artery and the internal carotid artery at the greater cornu of the hyoid bone. This nerve may be mobilized superiorly, taking care to preserve the descendens hypoglossi. The retropharyngeal space is reached by using blunt dissection (Fig. 9–5). The prevertebral fascia is incised in a vertical fashion to expose the longus colli muscles. The operative procedure here is carried out as with the transoral route once the prevertebral space is reached. After decompression has been accomplished, fusion may be carried out using a tricorticate iliac crest graft or a fibular strut interposed between the caudal clivus and the inferior aspect of the axis vertebrae.

For closure, the cervical wound is approximated in a layered fashion, and the digastric tendons are approximated with 3–0 Neurolon sutures.

POSTEROLATERAL, FAR LATERAL-TRANSCONDYLAR APPROACH

The posterior lateral approach has been labeled the lateral suboccipital approach, the transjugular approach, the extreme lateral and dorsolateral suboccipital transcondylar approach, and the extreme lateral transcondylar approach. These approaches vary slightly; all represent an attempt to expose the lower clivus, the foramen magnum, the craniovertebral junction, and the upper cervical spine without retracting the neural structures. An extensive amount of posterior and lateral squamous–occipital bone is resected including the lateral rim of foramen magnum and the posterior aspect of the occipital condyle (Fig. 9–6).

Depending on the need, the posterior arch of the atlas vertebrae as well as the posterior aspect of the foramen transversarium and the lateral atlantal mass may be resected. This approach gains control of the extracranial and intracranial vertebral artery, thus allowing the ability to work in front of the brainstem and the cervical spinal cord. A fusion construct may be carried out also.

Indications

An indication for posterolateral approach with transcondylar exposure of the ventrolateral craniocervical border is the presence of intradural neoplasms located anterior to the brainstem and cervical spinal cord. In the adult, aneurysms of the vertebral artery and the proximal basi-

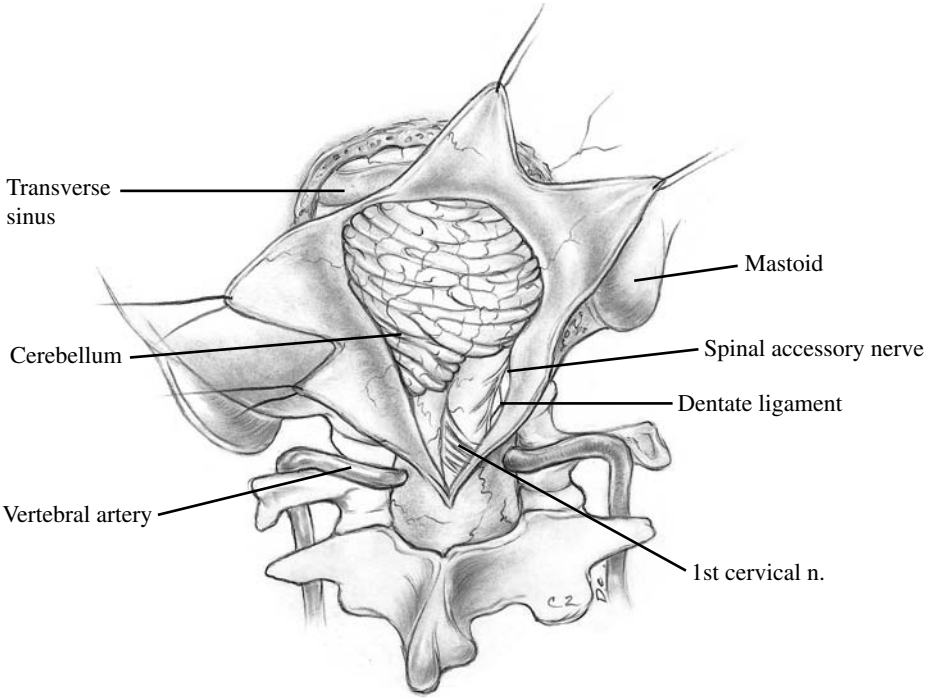


FIGURE 9–6. Exposure obtained with the posterolateral-transcondylar approach to the ventral cervicomedullary border. Note the extent of the occipital craniectomy going above the transverse sinus and exposing the sigmoid sinus in addition. The medial aspect of the occipital condyle has been resected as has been the posterior atlantal arch and the dorsal bony compartment of the foramen transversarium. The vertebral artery is thus mobilized. The dural incision is made to have control of the vertebral artery as well as gain access in front of the brainstem.

lar artery trunk may be approached in this manner. Frequently, congenital bony abnormalities at the craniovertebral border cause posterolateral compression of the medulla and may require this procedure as an adjunct to the posterior decompression, at which time a fusion also is accomplished. Thus, it is important for the treating physician to be conversant with the technique of vertebral artery decompression.

Intraoperative Technique

I prefer the prone position with the patient's head turned slightly to the side of the exposure and fixed in a multi pinned headrest secured to the operating table to allow for 45-degree rotation of the table during the operation. The lateral position allows the cerebellum to fall away from the site of the operative field, and tilting of the head upward provides venous drainage without the risk of air embolism.

An inverted U-shaped skin incision or an inverted hockey-stick incision is made, starting at the mastoid process and proceeding beneath the superior nuchal line to come down the midline. A cuff of fascia is maintained at the superior nuchal line, enabling a tight closure of the muscle at the end of the procedure. The paraspinal muscles are split along the spinous processes, and dissection is carried out laterally to the site of the lesion. The paraspinal musculature then is retracted with fish hooks and weights. A self-retaining retractor never should be used because the cervical musculature will bunch up and block the avenue of approach. Care is taken when dissecting the lateral mass of C-1 because of the possibility of injury to the vertebral artery.

The occipital bone is removed to include the condylar fossa; the posterior rim of foramen magnum goes lateral to the occipital condyle. The posterior arch of the atlas, including the transverse process and the lamina of C-1, is exposed. It is important to expose the transverse sinus as well as the sigmoid sinus to the jugular bulb. At times, a mastoidectomy expands the exposure. The vertebral artery then is elevated from the sulcus arteriosus and dissected free from the transverse foramen of the atlas to its penetration into the atlantooccipital membrane. Transposition of the freed vertebral artery allows further exposure into the posterior fossa and upper cervical spine. The transverse foramen of the atlas is removed from its dorsal aspect, and the vertebral artery is unroofed and dissection is made downward to the axis. The extreme lateral resection of bone into the condylar fossa and sometimes into the medial aspect of the occipital bone is key to being able to approach the front of the brainstem and cervical cord without retraction (Fig. 9–6). It may be necessary to remove the posterior medial aspect of the occipital condyle and the lateral mass of the atlas vertebra. If less than half of the occipital condyle is removed, no instability should result. Dissection into the anterior portion of the occipital condyle should be avoided to prevent injury of the hypoglossal nerve.

The dural incision is curvilinear and extends from the sigmoid sinus to the lateral cervical exposure. This incision then runs medial to the vertebral artery, and several secondary incisions are made perpendicular to the primary incision to allow for retraction.

The intradural vertebral artery is encountered immediately, and the dentate ligament is then sectioned. The cervicomedullary junction now can be rotated upward and away from a ventrally located lesion. Several specifics must be addressed during closure. The hemostasis must be exact. Mastoid air cells are carefully waxed, and a fat pad may be necessary to occlude this area. Meticulous dural closure is needed. The paraspinal muscle closure must be performed in a layered anatomic fashion to prevent leakage of cerebrospinal fluid. Instability does not occur if only the medial aspect of one occipital condyle has been resected; however, if a significant portion of the lateral mass of C-1 is removed and a facetectomy made, postoperative instability will result, which mandates fusion on the opposite side. A fusion on the same side can be made by spanning the operative defect and extending one vertebral level below it; for this, instrumentation is crucial.

Complications of Posterolateral Approach

The far lateral transcondylar approach as described by Sen and Sekhar as well as Al-Mefty is fraught with complications of cerebrospinal fluid leakage and craniocervical instability. In the series by George and co-workers, 3 of 14 patients died. For this reason, I prefer the posterolateral exposure, which provides a satisfactory muscle closure; most neurosurgeons are familiar with this route, which has proven safe and effective.

INSTABILITY AT THE CRANIOCERVICAL JUNCTION

The mechanical definition of instability is an abnormal or unexpectedly large motion that occurs in response to an implied load. It also encompasses the further load application beyond a certain critical load that leads to increased motion without any additional further load. There is load sharing at the craniocervical junction between the osseous structures, the ligaments, and the muscles. The criteria used to detect instability at the craniovertebral junction in children include (1) the presence of predental space of more than 5 mm in patients below the age of 8 years and more than 3 mm in children above 8 years of age; (2) separation of the lateral atlantal masses of more than 7 mm on the open-mouth view, suggesting a Jefferson fracture and the possibility of disruption of the transverse portion of the cruciate ligament and also possibly signifying a congenital abnormality of bifid anterior and posterior arches of the atlas; (3) vertical odontoid-clivus translation of more than 2 mm, indicating injury to the occipitoatlantal ligament; (4) a minimal gap between the occipital condyles and the lateral facets so that the occipital condyles on a lateral radiograph of the cervical spine are never visualized (bare occipital condyles indicate an occipitocervical dislocation); (5) abnormal craniocervical motion dynamics; and (6) an abnormal relationship between the spinal canal and foramen magnum, which is pathological except for widening that may take place between flexion and extension between the occiput, C-1 and C-2.

Posterior Fusions for Reducible Lesions

Indications for craniocervical fixation and fusion were outlined earlier in this chapter. More than 1200 patients have undergone craniocervical fusion at the author's institution over a 20-year span with a 98.6% success rate. This series is divided into congenital, developmental, and acquired lesions. Table 9–3 outlines the indications for

TABLE 9–3.
Indications for Occipitocervical Fusion

Congenital
Anterior and posterior bifid arches of C-1
Absent occipital condyles
Developmental
Severe reducible basilar invagination
Unstable dystopic os odontoideum
Unilateral atlas assimilation with chronic rotary luxation of occiput to C-1 to C-2
Acquired
Traumatic dislocation of occiput to C-1 (especially vertical and posterior occipitocervical)
Complex CVJ fractures of C-1 to C-2
Reducible rheumatoid cranial settling
After transoral CVJ decompression
Cranial settling in ankylosing spondylitis, psoriasis, pseudogout, Down's syndrome, inflammatory ileitis
Inflammatory disease: chronic Grisel's syndrome
Primary malignancies affecting the CVJ (e.g., chordoma of clivus and occipital bone, plasmacytoma, osteoblastoma, chondroma, neurofibromatosis)
Secondary metastatic disease affecting the CVJ (e.g., breast metastasis)

CVJ, craniovertebral junction.

TABLE 9–4.
Indications for Atlantoaxial Arthrodesis

Congenital
Absent odontoid process
Dystopic os odontoideum
Absent posterior arch of C-1
Developmental
Morquio's syndrome
Goldenhar's syndrome
Conradi's syndrome
Spondyloepiphyseal dysplasia
Acquired
Down's syndrome
Trauma
Postinfectious
Malignancy

occipitocervical fusion, and Table 9–4 encompasses the indications for atlantoaxial arthrodesis.

TECHNIQUE OF CRANIOCERVICAL FUSION

Preoperative cervical traction, if possible, is obtained using crown halo with traction at 6 to 7 pounds to support the head. In the older child (i.e., 10 to 12 years of age), traction never should exceed more than 15 pounds for craniocervical abnormalities. The child is brought to the operating room in halo traction whenever possible, which is often feasible in children older than 10. Otherwise, the halo traction is placed after general anesthesia. The intubation is carried out as previously outlined for halo placement.

The child then is placed prone on the operating table with traction being maintained (Fig. 9–7). The halo pins and ring rest on a modified Mayfield-Kees horseshoe headrest, and traction is maintained over a pulley bar to allow for dynamic motion during the operative procedure. Fixation of the halo ring is to be avoided because this does not allow dynamic motion during the operative procedure and may permit a "snaking" effect. The chest wall is elevated on laminectomy rolls or a Wilson frame. Lateral radiograph documents the optimum position for reduction, which is guided by preoperative dynamic studies. Cervical traction must be maintained during the operation.

The posterior scalp and cervical regions are prepared, as is the area for harvesting of donor bone. I prefer rib graft to iliac crest bone graft. If an intradural procedure is planned in conjunction with the fusion, then cephalothin-sodium is administered starting 12 hours before the procedure and continuing for 48 hours after the procedure given every 6 hours.

A midline incision from the external occipital protuberance to the spinous process of the fifth cervical vertebrae is made. A subperiosteal exposure of the squamous–occipital bone and the posterior arches of the upper three cervical vertebrae is obtained using sharp dissection alone. The axis vertebrae may be stabilized with a towel clip passing through the spinous process if instability is present. This should be done during the dissection of the paracervical musculature. Stabilization of the operative exposure is obtained by placing the angled cerebellar retractors at 90 degrees to each other. This

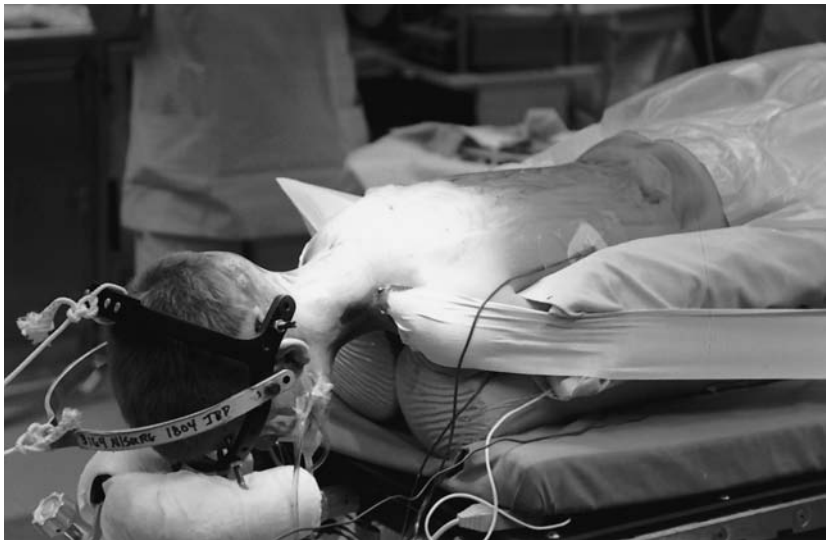
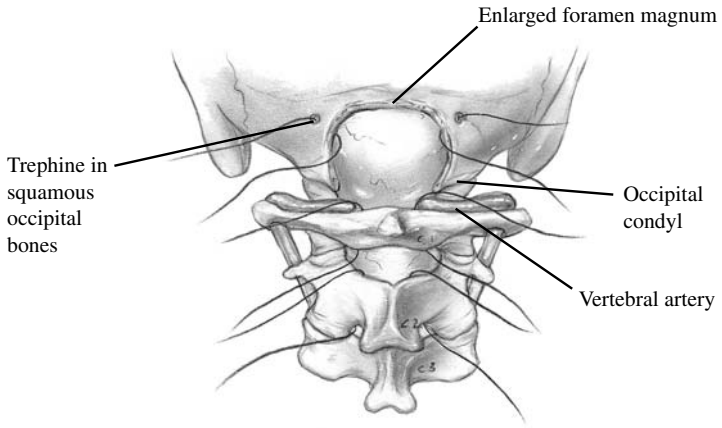


FIGURE 9–7. Operative photograph of child positioned prone for dorsal occipitocervical fixation. Note the crown halo resting on the padded Mayfield horseshoe headrest with traction maintained in a neutral position. The posterior rib cage as well as the iliac crest are exposed for possible harvesting of donor bone.

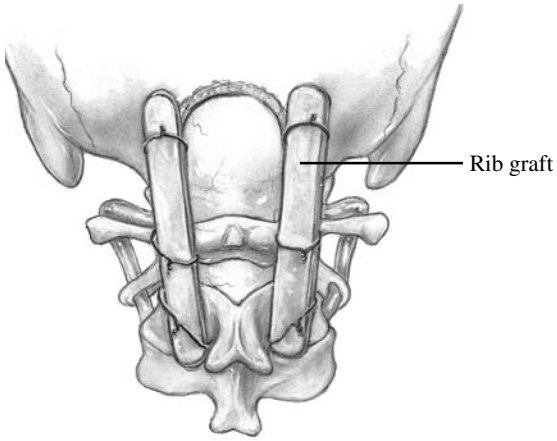
fixes and stretches the muscle-bone relationships to prevent motion at the craniocervical joints.

Occipitocervical Fusion

A craniectomy excises the posterior rim of foramen magnum, and the bone is used in the fusion construct. This ascends upward for 1.5 cm and 1 cm to either side of the midline, thereby eliminating the exoccipital ridge and allowing ease of passage of cable from a laterally placed occipital trephine to the midline (Fig. 9–8A). Trephines then are made 2.5 cm to either side of the midline and about 2 cm above the foramen magnum in the squamous–occipital bone. Soft titanium cable is passed from the occipital trephine to the midline craniectomy gaining purchase of the occipital bone. Sublaminar cable is passed beneath the axis and the atlas vertebrae and the donor bone harvested. A full-thickness rib graft is removed starting at the head of the rib and extending out laterally. This natural curvature, once reversed, allows good bone apposition between the occiput and the upper cervical vertebrae (Fig. 9–8B). Decortication of the laminae and the spinous process as well as the occipital bone is essential at the recipient site. The donor bone is secured to the occiput, the atlas, and the axis vertebrae by



A (1)



A (2)

B

FIGURE 9–8. A: Technique for dorsal occipitocervical arthrodesis with rib graft. **B:** Operative photograph of completed dorsal occiput C1–2 fusion with bilateral rib grafts and titanium cables.

passing the cable through the graft to anchor it into position. Remaining excess bone then is packed as matchstick slivers into the crevices at the donor-recipient interface. It is mandatory that the bones not be tied to each other because fracture will occur as the child grows, which will lead to neurologic sequelae.

Over the past 7 years, immediate fixation of the occiput to the upper cervical spine has been achieved with a contoured threaded titanium loop custom fitted to the occiput and the upper cervical vertebrae and anchored in place with titanium cables (Fig. 9–9A). It is imperative that the superior horizontal limb of the contoured loop instrumentation be fixated and supported by cables superiorly and laterally to the occiput. This construct is always supplemented by bone (Figs. 9–9B and C). In a few patients, bone was harvested from the occiput; however, this membranous bone offers less advantage than the marrow bone from the rib or iliac crest.

Craniocervical instability after "wide posterior decompression" will tax the surgeon's ingenuity to gain purchase in the occiput laterally as well as with the atlantoaxial facet lateral masses. In addition, associated atlas assimilation, which is often present with abnormal vertebral artery anatomy, complicates the situation. Here, transarticular screw fixation between C-2 and C-1 becomes hazardous. Hence, angled plates anchored to the facets at C-2 and C-3 below and superiorly to the lateral occiput become attractive.

The wound closure is made in anatomic layers with an interrupted 0 polyglycolic suture. The child is maintained in cervical collar until the effects of general anesthesia and endotracheal intubation have receded. Following this, halo vest immobilization is accomplished. This immobilization is required for 6 months after occipito–atlantoaxial arthrodesis. Less prolonged immobilization results in nonunion, union in an abnormal position, additional cranial settling with subsequent increased neurologic deficit, or fracture of the construct.

Dorsal Atlantoaxial Arthrodesis

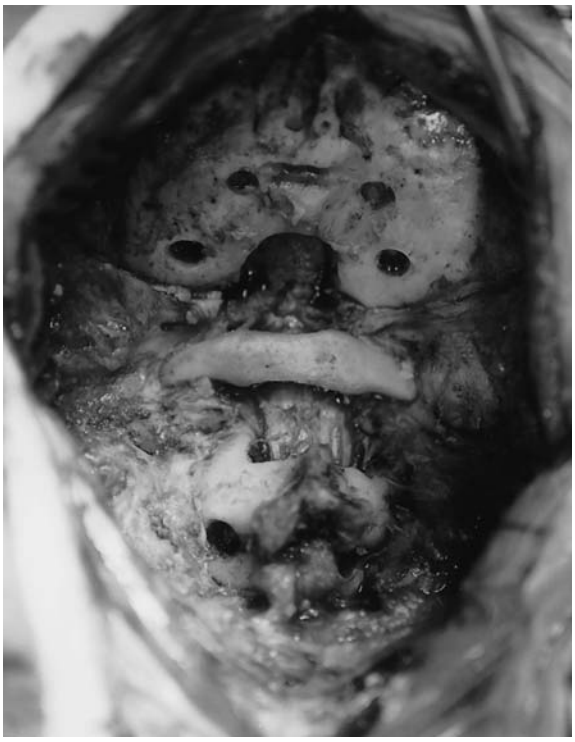
The technique for dorsal atlantoaxial arthrodesis is similar to that of occipitocervical fixation except the grafts do not extend to the occiput. Interlaminar, circumlaminar fixation with rib grafts fulfills the criteria mentioned previously as requirements for craniocervical arthrodesis (Fig. 9–10). If ventral atlas displacement has occurred, the posterior construct must be given added strength by extending it down to the C-3 level. Clamping or cerclage between C-1 and C-2 posterior arches incites upward migration of the odontoid process and leads to basilar invagination and cranial settling. In younger children, this will lead to dorsal lordosis as the child grows. In addition, the wires cut through the bone and may penetrate the dura when the wires fracture. Thus, each graft should be individually anchored to the dorsal recipient surface of the atlas and the axis vertebra and, at times, a third cervical vertebra if needed.

An alternative that I have used is a fusion construct that is modified between a Gallie wire technique and the modified Brooks technique. For this, the iliac crest must be of satisfactory thickness to provide a full thickness of tricorticate bone. The superior surface of the laminae and spinous processes of the axis is prepared by smoothing the upper border and also by creating a notch at the junction of the laminae and the spinous processes to seat the cable (Fig. 9–11A). The donor bone is harvested in a rectangular fashion to provide seating over the spinous process and lamina of C-2 vertebrae dorsally, and a notch is made to accommodate the spinous process of the axis vertebrae. The superior surface of the graft is notched to allow seating of the inferior portion of the posterior arch of C-1. A lateral notch at the waist is provided to hold the bone graft using the cable.

The cable is passed beneath the lamina of C-1 and the bone graft is accommodated between the dorsal aspects of the C-1 and C-2 laminae. Forward motion of this bone piece is prevented by a notch that creates a fit between C-1 and C-2 as well as by correct placement of the holding cable. This loop must pass underneath the lamina of C-1 and then is brought over the graft to hook under the spinous process of C-2. A snug fit is secured by the anterior aspects of the free cables to be brought around the bone graft and secured behind it (Fig. 9–11B). Postoperative halo vest immobilization is maintained for at least 3 months. The actual extent of immobilization is guided by appropriate radiographs of C-2. Table 9–5 illustrates the perioperative and delayed complications of dorsal decompression and occipitocervical fusion and management of the same.

Posterior Atlantoaxial Facet Transarticular Screw Fixation

Posterior screw fixation rigidly couples the facets of the atlas and the axis vertebrae in obtaining immediate internal fixation. This technique has the largest separation in the circumference of the C1–2 cervical articulation and thus prohibits rotation and translation. It must be used in conjunction with bony arthrodesis to obtain a three-point fixation. A rigid coupling then obviates the necessity for postoperative halo brace immobilization.



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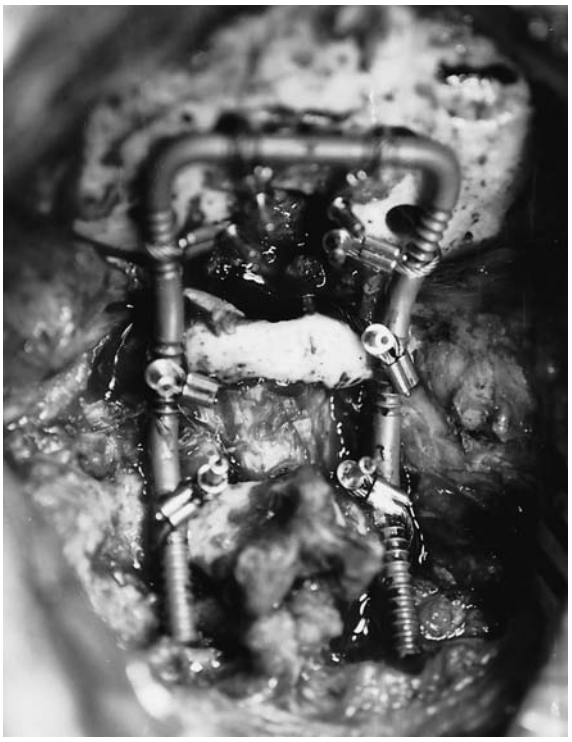

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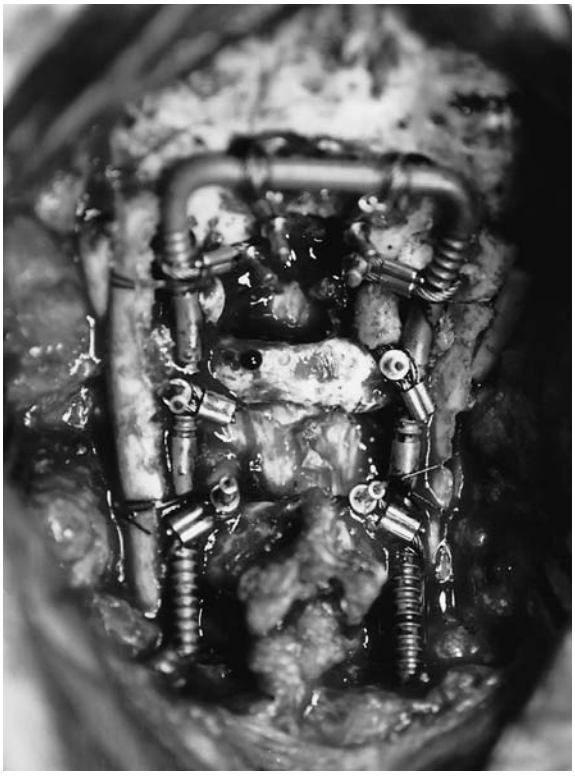

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FIGURE 9–9. **A:** Operative view of exposure of squamous-occipital bone and the posterior bony arches of C-1 and C-2. Note the midline craniectomy and the occipital trephines with a horizontal groove made above them to accommodate the transverse portion of the oncoming occipitocervical titanium loop. **B:** The custom-contoured threaded occipitocervical titanium loop instrumentation is secured to the occiput as well as the lateral most portion of the laminae of C-1 and C-2 with cerclage titanium cables. **C:** Completed construct of dorsal occipitocervical fixation with titanium loop instrumentation and autologous rib grafts packed against the occiput and the dorsal aspect of the laminae of C-1 and C-2.

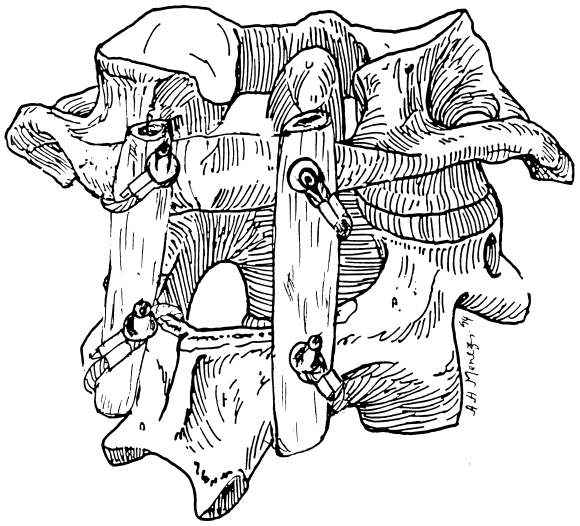
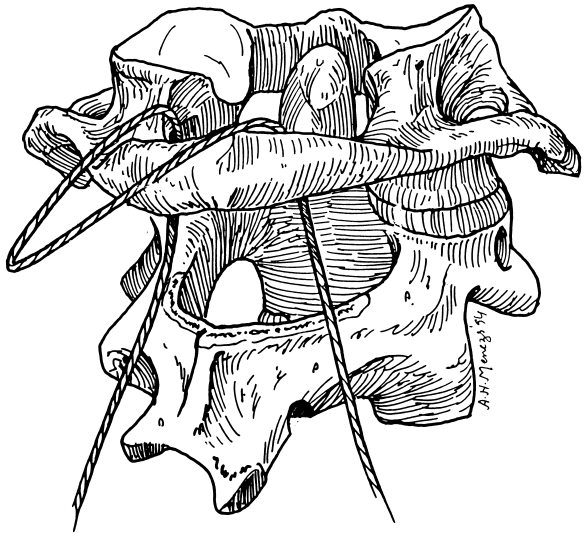


FIGURE 9–10. Dorsal atlantoaxial arthrodesis with bilateral interlaminar rib graft fixation.

Transarticular screw fixation is contraindicated in the young child (i.e., <10 years of age). It is also not possible in 18 to 23% of the normal population because of a high-riding vertebral artery in the groove at C1–2. It is contraindicated in incomplete C1–2 reduction and in children with atlas assimilation. The main problem with transarticular screw fixation of the atlas and axis vertebrae is injury to the vertebral artery, which can be silent and is more common with noncannulated screw fixation. In a series of 61 patients by Crockard, vertebral artery injury occurred in 5 patients, screw breakage 5, malposition of the screw in 17, and a nonunion of the fusion mass in 8. In 14 patients, graft reabsorption occurred, and there was incomplete reduction in 14 others.

Thus, it is important that patients to undergo transarticular screw fixation be chosen carefully. In the pediatric population, this procedure should be limited to patients with an absent posterior arch of C-1 when a previous decompression has been accomplished, in dystopic os odontoideum, and in children with Down's syndrome between the ages of 8 and 16 years.

Before transarticular screw fixation in a child is considered, a three-dimensional CT of the craniocervical junction must be obtained and reconstructions accomplished in a sagittal plane at the level of the facet (Fig. 9–12A). The smallest-diameter screw that is available is 2.5 mm. Thus, the C-2 pedicle must be capable of accommodating this without difficulty. The superior facet joint of C-2 and the lateral mass of C-1 must be aligned properly to obtain an adequate screw purchase. I prefer to use a cannulated lag screw 3.5 to 4 mm in diameter in a child between the ages of 14 and 16. In younger children, the anatomical dimensions of the facet–pedicle dictate the size of the screw and may be anywhere from 2.5 to 3.5



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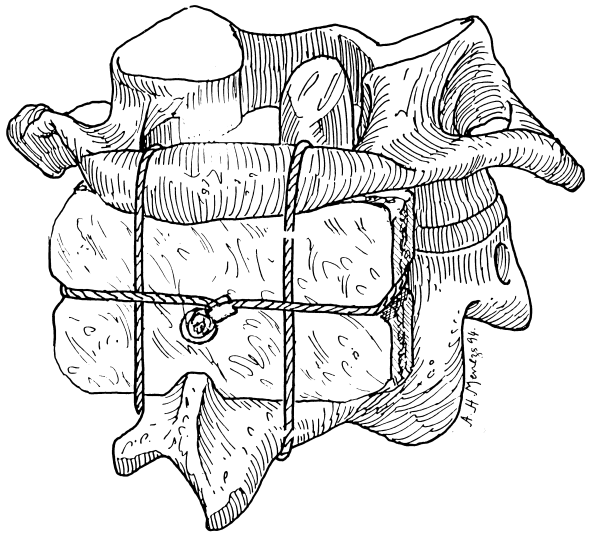

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FIGURE 9–11. A: Cable placement for modified Gallie-Brooks fusion technique. Note the smoothing of the superior aspect of the lamina-spinous process of the atlas as well as the notch created at the base of the spinous process of the axis vertebrae. **B:** A large block of tricorticate iliac crest has been fashioned to fit in the interlaminar space between the atlas and the axis vertebrae. A notch accommodates the spinous process of the axis, and the cables run in the grooves at the base of the spinous process as well as the waist of the rectangular graft. This supposedly acts as a spacer and provides compression for pustule induction.

TABLE 9–5.
Perioperative and Delayed Complications of Dorsal Decompression and Occipitocervical Fusions: Management

Complication	Prevention and Management
Perioperative complications	
Problems with intubation	Preoperative evaluation; awake fiberoptic intubation; else tracheostomy
Worsening neurological status	Patient should be positioned awake, if possible; traction must be used; no head holder
Unstable CVJ	Do not allow muscle paralyzing agents; stabilize C-1 and C-2 with towel clip during dissection
Bulging dura	Preoperative CSF drainage
Persistent venous bleeding	Make sure it is not from low-lying torcula, else clip dural leaves
Inability to decompress cervicomedullary junction	Use magnification and high powered drill to remove foramen magnum; no rongeurs
Cannot stabilize	Choice of fusion procedure; grafts must be anchored; in total destabilization, fuse occiput to C-2 with contoured loops if needed
Failure to awaken	Secondary to preceding problems; must obtain head CT to detect clot
Delayed complications	
CSF leakage	Preoperative assessment of hydrocephalus and drainage; poor dural or graft approximation; will need reoperation if persists after CSF diversion
Loss of alignment in halo/brace	Replace in traction and then halo vest; graft resorption occurs at 3–4 mo and requires refusion
Wound infection	Needs careful hemostasis; perioperative antibiotics; antibiotics, debridement and closure
Delayed worsening after removal of brace	Pseudoarthrosis; broken fusion or ventral pathology uncorrected; possible hydrocephalus, infection, or hydromyelia
Nonunion	Construct must be solid at placement; occipitocervical fusion requires 6 mo halo immobilization
Complications related to halo	Constant care
Loose pins	Retighten initially
Infected pins	Local care; replacement
Penetration	Remove halo; MRI; CSF examination; antibiotics; use collar until reassment
Halo falls off	Use molded brace if needed

CSF, cerebrospinal fluid; CT, computed tomography; CVJ, craniovertebral junction; MRI, magnetic resonance imaging.

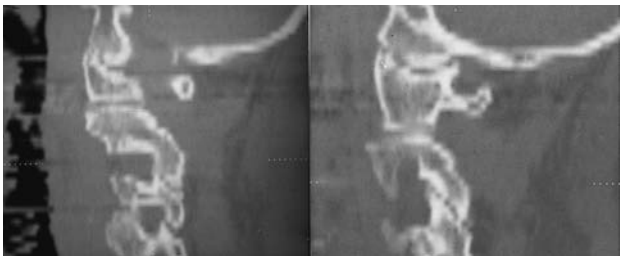
mm in diameter. The length of the transarticular screw averages 38 to 40 mm.

Intraoperative Technique

Controlled flexion of the patient's neck is required to obtain the proper trajectory for insertion of the drills and screws. This should be parallel to the posterior surface of the spine. Lateral fluoroscopy monitoring is used to assess spinal alignment during the positioning for the drilling as well as screw insertion. A posterior cervical incision is made to assess the atlas and axis vertebrae. The incision should extend to the spinous process of T-1. Skin preparation should extend to the upper thoracic level in case a longer incision or percutaneous access is required for drilling.

The atlas and axis vertebrae are aligned by using manual reduction, and the vertebrae are exposed in a subperiosteal manner to the lateral aspect of the facets (Fig. 9–12B). A wire may be passed around the ring of C-1 for traction. The spinous process of C-2 may be able to be pulled up toward the base of the occiput using a clamp on the spinous process. The pedicle of C-2 and the C1–2 articular surfaces are directly visualized and exposed. The ligamentum flavum attached to the pedicles and the laminae are removed. A fine dissecting instrument then is placed into the atlantoaxial facet joint. The C-2 nerve root is exposed, and the veins are cauterized to allow direct visualization of the pedicle of C-2 and the C1–2 facet joint.

The drill enters the caudal aspect of the C-2 inferior facet 3 mm lateral to the medial edge of the C2–3 facet (Fig. 9–12B). The posterior cortical plate of bone is pene-



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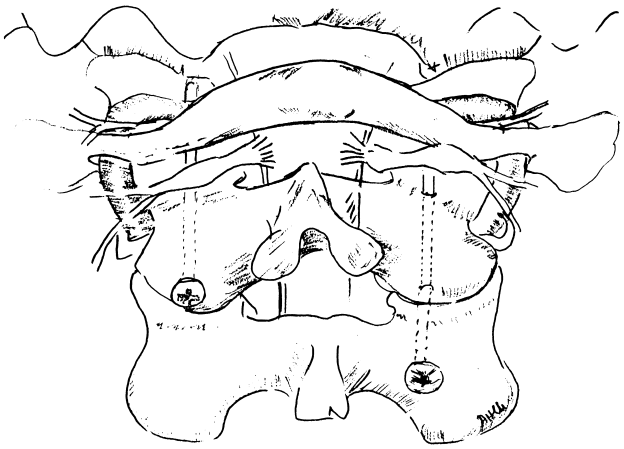

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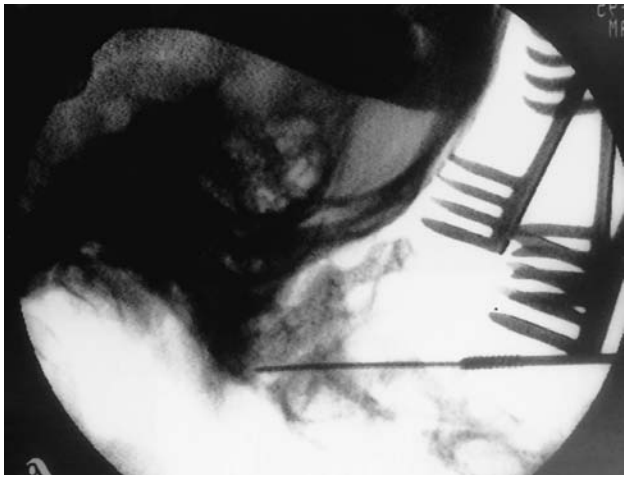

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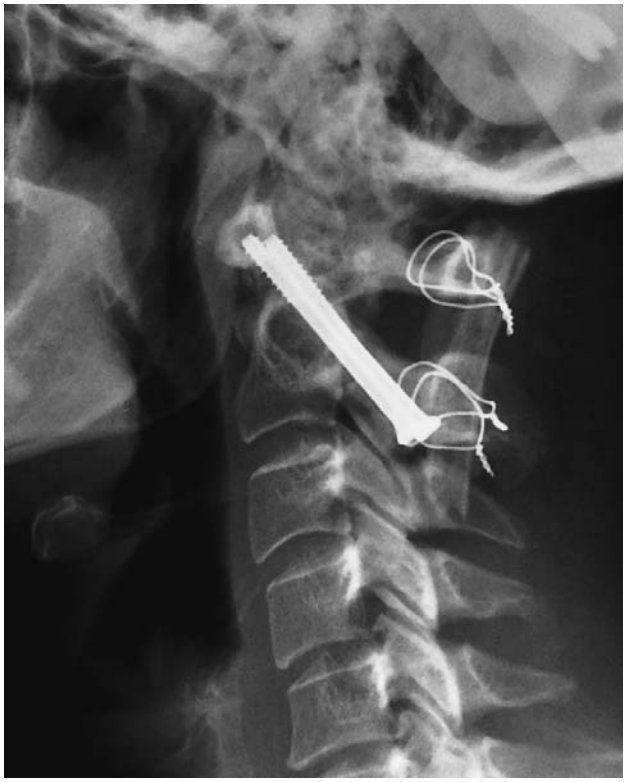

D

FIGURE 9–12. A: Parasagittal reconstruction through the plane of the axis pedicle. This determines the thickness of the anatomic structure as well as the presence of a vertebral artery groove that may hamper consideration of screw fixation. **B:** Exposed dorsal craniocervical junction. The position of the C-2 nerve roots as well as the ganglion and the facet joint should be appreciated. Note the vertebral artery loop and the position of the transarticular screws. **C:** Intraoperative lateral craniocervical fluoroscopy in the prone position. The K-wire has been drilled through the pars interarticularis of the axis into the lateral atlantal mass. The aim is to the anterior cortical border of the atlas arch. Note the threaded lag screw being advanced over the K-wire. **D:** Lateral craniocervical radiograph made 4 days after bilateral transarticular C2–1 screw fixation with interlaminar rib graft fusion. Pathology was grossly unstable dystopic os odontoideum.

trated with a bone awl or high-speed drill. The drill trajectory is directed to the dorsal cortex of the anterior arch of C-1 on lateral fluoroscopy, which is monitored throughout the drilling procedure (Fig. 9–12C). In the anteroposterior direction, the drill is placed through the central axis of the C-2 pars interarticularis. A sagittal trajectory of about 5 to 10 degrees medially is required. Typically, this is about true sagittal orientation; entry of the drill into the facet joint and then into the lateral mass of C-1 can be directly visualized. I prefer to use a K-wire for drilling and a cannulated screw. On the other hand, a noncannulated screw can be used. The length of the screw is measured off the K-wire. A tap may be passed over the K-wire, over which a self-tapping screw is placed. The position of the guidewire must be monitored to prevent it from advancing. I prefer to have the guidewire stopping short of the anterior cortex of the atlas by about 4 mm. Once the screw is seated in the bone of C-1, the K-wire is removed with a reversible drill. As the screw crosses the joint space into C-1, the atlas and

axis become rigidly coupled. The vertebral "lock" is easily appreciated. I prefer to place the guidewires bilaterally prior to insertion of the screws so that the image of the screw does not obliterate the guidewire on lateral fluoroscopy. A bilateral interlaminar or interspinous fusion then should be accomplished. Percutaneous drilling and screw insertion are possible, but I believe the pediatric population is better served by using direct exposure of the atlantoaxial joint and direct guidance of the K-wire for insertion. Postoperative immobilization is carried out with a Miami J collar.

It is important to carry out neurologic evaluation for patients who have undergone transarticular screw fixation because of the possibility of damage to the C-2 nerve root and potential vertebral artery injury and neurological sequelae; however, 90% of vertebral artery injuries go unnoticed. Thus, use of transarticular screw fixation should only be carried out by persons experienced in surgical procedures in this area.

EDITOR'S COMMENTARY

In this chapter, Menezes highlights the significant complexity of the craniocervical junction and its surrounding anatomy. There is no question that the approach to lesions and pathology in this region are done so with a significant amount of trepidation. The complex anatomy involves both the anterior and posterior cervical skull base with as complex an anatomy of brainstem, cranial nerves, and vasculature than anywhere else in the body. The optimal approach to lesions in this area is best accomplished with adequate imaging and understanding of the underlying pathologic processes. To reiterate, young children are often best treated with craniocervical immobilization while, in children greater than 2 years of age, consideration for surgical intervention can be planned. It is clear that any operative intervention requires adequate visualization, and this should be approached aggressively, particularly in the transoral approach. Decompression or excision of the pathologic lesion is most often just the beginning of the surgical intervention where fusion and stabilization are required. The surgeon should be prepared to perform the posterior fusion either following the anterior or a lateral approach. Lastly, because of the motion arm and significant stressors at the upper cervical spine, even a straightforward posterior C1,2 fusion may require halo fixation for a 3–6 month period to gain adequate fusion. As Menezes points out, to avoid halo placement following a C1,2 fusion transarticular screw fixation should be considered if the pedicle is of adequate diameter to allow for a minimum of a 2.5 mm screw. A longer incision is often necessary in these instances, or through a transcutaneous apporoach combined with a smaller incision to gain adequate visualization and angle for screw placement.

PEARLS

In this author's experience:

- It is not uncommon for children with craniocervical abnormalities to present with basilar migraine, dysphagia, sleep apnea, the syndrome akin to the central cervical spinal cord dysfunction, and scoliosis.
- When there are unexplained neurologic symptoms and signs associated with craniocervical abnormalities, vertebral angiography should be performed with dynamic motion studies.
- Crown halo application in children requires six to eight pins placed beneath the cranial equator. Finger tightening of the pins is used below 2 years of age. At 5 years of age, 4 pounds torque pin pressure is used. The "adult" 8 pounds torque present is applied past age 12 years.

SUGGESTED READINGS

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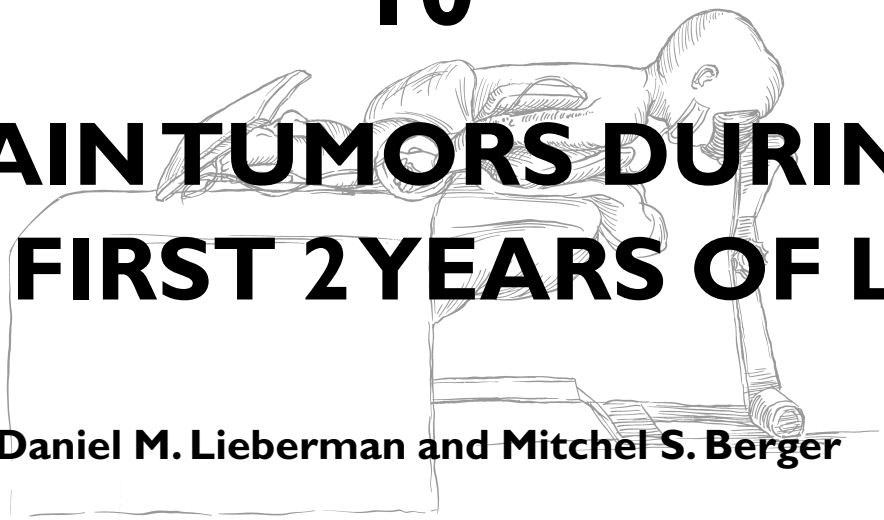
Neoplasms

10

BRAIN TUMORS DURING THE FIRST 2 YEARS OF LIFE



Daniel M. Lieberman and Mitchel S. Berger

Brain tumors that arise in infants during the first 2 years of life may be congenital, developmental, or identical to the lesions that affect older children or adults. Although they are rare, affecting only 1.1 of every 100,000 liveborn or stillborn infants, brain tumors are the most common solid tumors occurring in young children. Of all intracranial tumors affecting infants, most are astrocytomas (32%), primitive neuroectodermal tumors (PNETs, 25%), ependymomas (12%), or choroid plexus papillomas (10%) (Fig. 10–1). An aggressive surgical approach that maximizes the extent of surgical resection is essential to the treatment of brain tumors during an infant's first 2 years of life because radiation therapy, the most effective adjuvant treatment for older children and adults, is harmful to the immature nervous system.

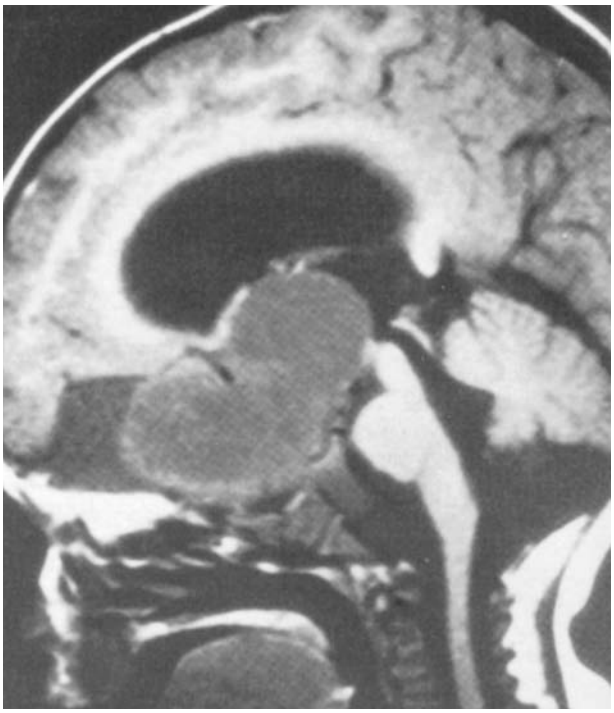
SURGICAL INDICATIONS AND PREOPERATIVE EVALUATION

Surgical resection or biopsy is the initial therapy of choice for infants with brain tumors of all types except diffuse pontine or hypothalamic glioma without exophytic components, optic-nerve glioma in infants with excellent vision or neurofibromatosis, or subependymal giant-cell astrocytoma without symptomatic ventriculomegaly in a child with tuberous sclerosis. Magnetic resonance imaging (MRI) performed with and without gadolinium contrast agent is indicated in nearly all cases. In our experience, sedation is the most important variable in obtaining high-quality diagnostic magnetic resonance images. To avoid hypoventilation-induced increases in intracranial pressure, sedation is accomplished most safely by using general anesthesia and controlled ventilation. Computed tomography done without contrast agent is useful in screening for intracranial masses in meganencephalic infants who have no neurologic deficit; if a mass is found in such patients, however, MRI should be performed.

PREOPERATIVE MANAGEMENT

Nearly all patients require some degree of preoperative management for increased intracranial pressure, associated hydrocephalus, or seizures. Increased intracranial pressure may result from mass effect of the tumor and hydrocephalus caused by obstruction of cerebrospinal fluid drainage. Corticosteroids relieve increased pressure from the mass effect. The timing of extraventricular drainage for hydrocephalus is determined by the patient's clinical condition; children who are obtunded or comatose undergo emergency extraventricular drainage. Normal intracranial pressure varies according to age: Physiological intracranial pressure in a neonate (<2 mm Hg) is lower than that in infants up to 1 year of age (1.5 to 6 mm Hg) and in turn is lower than that in young children (3–7 mm Hg).

Preoperative management of seizures begins with routine use of antiepileptic drugs. In the event the patient has controlled, nongeneralized seizures and is not being medicated already for seizure prophylaxis, an intravenous loading dose should be given in the operating room at the time of surgery and continued postoperatively. Patients with intractable seizures who are not taking antiepileptic drugs should be given them preoperatively and the pa-



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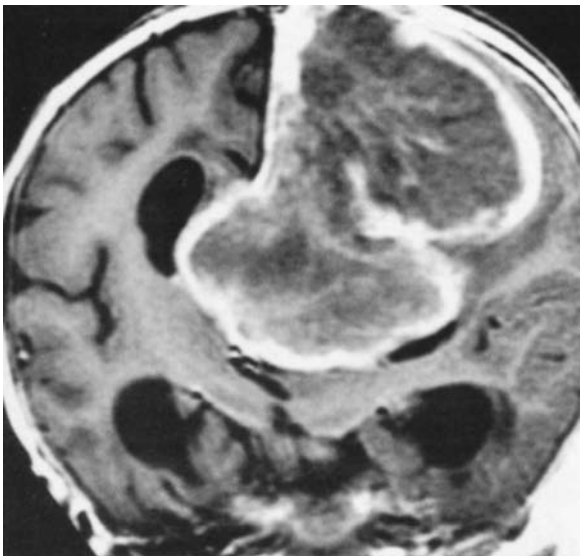

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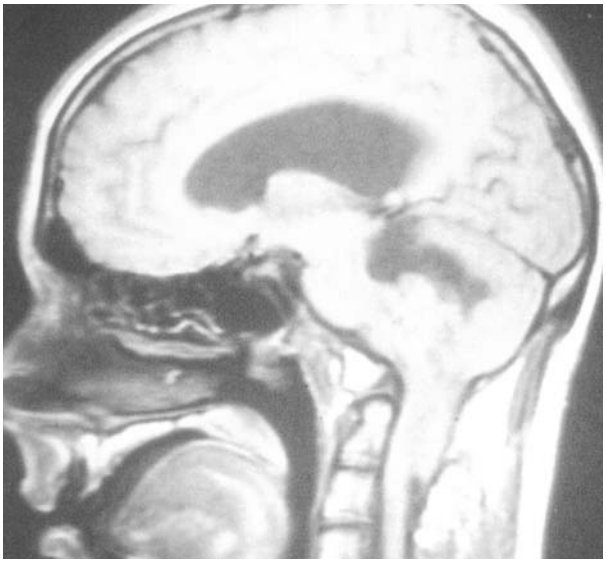

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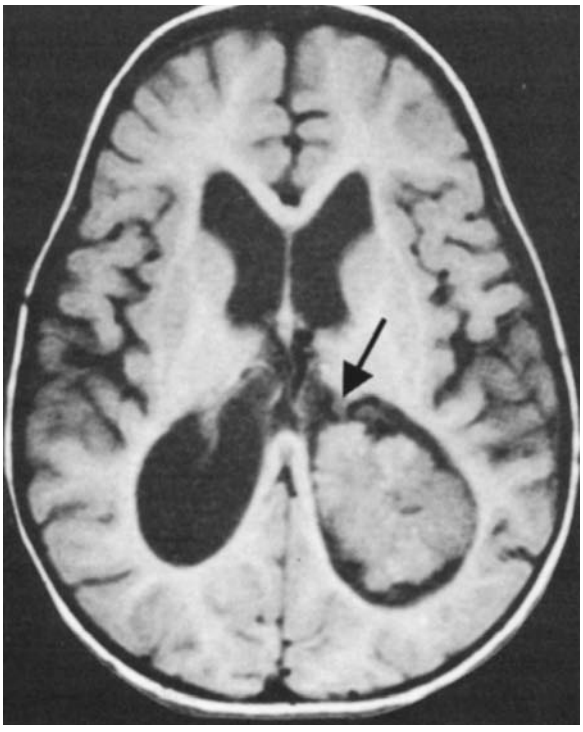

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FIGURE 10–1. Examples of the typical appearance of **(A)** astrocytoma, **(B)** primitive neuroectodermal tumor (PNET), **(C)** ependymoma, and **(D)** choroid plexus papilloma on magnetic resonance images (MRIs). Astrocytomas are often midline and may be cystic or solid. In our experience, the classic pilocytic astrocytoma consisting of a cyst and an enhancing mural nodule occurs in the minority of cases. Although astrocytomas have variable intensity on MRI sequences, most have decreased signal on T1-weighted sequences and increased signal on T2-weighted sequences **(A)**. The high nuclear-to-cytoplasm ratio found in PNETs make them hyperdense on computed tomography, whereas their signal intensity tends to be isointense or hypointense to the brain on T1-weighted sequences on MRI. Most PNETs contain cysts or calcification, and contrast enhancement patterns are variable **(B)**. Ependymomas have variable intensity on MRI sequences; however, they are often differentiated from PNET by their tendency toward invasion.

tient's response monitored. If seizures are refractory to antiepileptic drugs, even though the patient is verified to have received a therapeutic concentration, an electroencephalogram should be performed preoperatively to lateralize and help localize the epileptogenic focus, and preparations should be made for the intraoperative mapping of seizure foci by means of electrocorticography or evoked potentials. In contrast, there is no indication for intraoperative electrocorticography for patients whose seizures are well controlled because the tumor is nearly always the focus in these cases, and tumor resection is likely to leave the patient seizure free.

INTRAOPERATIVE TECHNIQUES

Approaches for specific tumors are well described in the accompanying chapters of this atlas. Here we emphasize unique aspects of intraoperative techniques in the care of infants.

Anesthesia and Positioning

In the operating room, warming pads, heated fluid bags, and an elevated room temperature are used to maintain the patient's normal physiologic temperature. Intravenous access is established by using an angiocatheter when possible; however, a saphenous vein cut-down sometimes is required. The urinary bladder is catheterized by using a pediatric urinary bladder catheter without a balloon.

Patients are positioned appropriately for the operation, with the limbs partially flexed and all pressure points padded (Fig. 10–2). The patient's head is placed on a foam headrest or padded doughnut-shaped cushion because the skull of children younger than 2 years of age may deform or fracture if pin fixation is attempted. The endotracheal tube then is fastened into place because even slight pressure may dislodge it during the operation. Special care is taken to cover the endotracheal tubing with towels to prevent the skin drapes from adhering to the tubing and perhaps inadvertently dislodging the endotracheal tube when the drapes are removed.

Surgery

After superficial skin incision of the scalp, bleeding is prevented by direct pressure from the surgeon. The incision is continued by using electrocautery (Bovie) or a contact laser. Alternatively, the Shaw scalpel, which uses heat for hemostasis, may be used to incise the skin and subcutaneous tissues. Moistened towels are placed along the edges of the scalp incision and secured in place with Michele clips; in addition to protecting the tissue, these sponges make any ongoing bleeding sites apparent. Rainey clips are generally too large for use in infants; when they do fit, they apply such excessive force that they may cause necrosis in delicate infant tissues. If the fontanelles remain open, craniotomy may be performed without the necessity of a burr hole by using the fontanelle to gain access to the epidural space. In young infants, bone often can be cut with scissors; however, a pneumatic drill with a side-cutting bit is often necessary.

FIGURE 10–2. Careful padding is critical for protection of infants during surgery. For posterior fossa lesions, we use the prone position and support the head with a horseshoe rest. After securing the endotracheal tube, the infant is placed prone and the axilla, shoulders, arms, and knees are padded. The horseshoe headrest is adjusted so that its arms are lateral to the lateral canthus without contacting the eyes, and the neck slightly flexed. After the infant is in the correct position, the surgeon looks from beneath the headrest a second time to ensure that the eyes are neither under pressure nor in contact with the headrest.

Functional Brain Mapping

Mapping of functional brain is a useful adjuvant when operating on tumors involving Rolandic cortex, supplemental motor areas, or the corona radiata and internal capsule (Fig. 10–3). Current (2–16 mA) is applied to the brain by using a constant-current generator and stimulating probe, bipolar forceps, or grid. Because the sensitivity of the brain to electric stimulation depends on the anesthetic state of the patient, paralyzing agents are not used or are reversed, and anesthesia is maintained by using inhaled agents at 50% concentration with narcotic

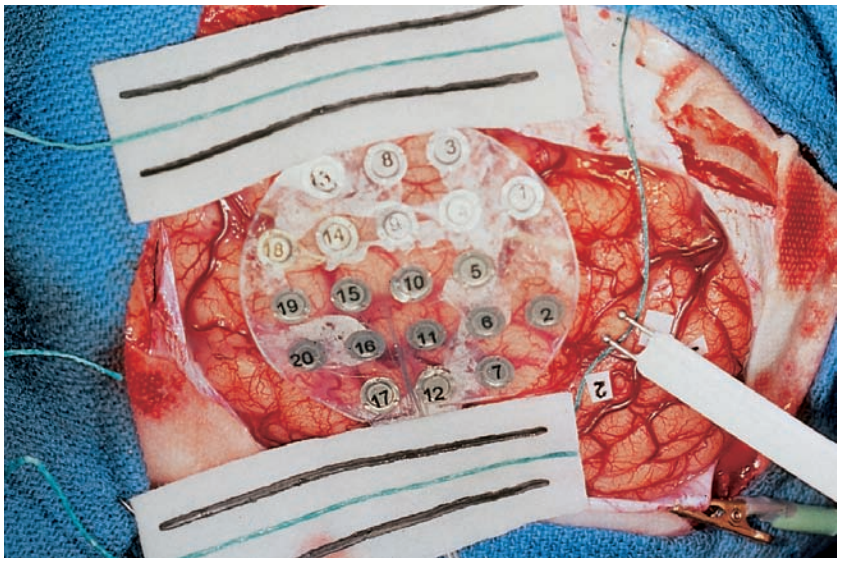

FIGURE 10–3. Techniques and equipment for intraoperative brain mapping in infants are similar to those used with older children and adults; however, the cortex in infants is sometimes difficult to excite electrically. We have observed motor responses after direct stimulation in infants, such as this 6-month-old child with intractable seizures and tuberous sclerosis **(A)**. In the event the cortex cannot be stimulated directly, identification of the central sulcus is possible using evoked potentials. Pyramidal neurons in the postcentral gyrus are oriented such that they have a natural dipole that is relatively positive anterior to the central sulcus and negative posterior to the central sulcus. In response to peripheral stimulation, generally stimulation of the median or ulnar nerves, electrodes spanning the central sulcus therefore will record a shift in potential from negative to positive. In this case, electrodes 1 and 2 were anterior to the central sulcus, and electrodes 3 and 4 were posterior to the central sulcus **(B)**.

infusion. However, the motor cortex in children younger than 2 years of age is often resistant to electrical stimulation. In such a case, the central sulcus still can be identified through motor or sensory evoked potentials, but it may not be possible to map subcortical pathways. Speech mapping is not necessary in infants or children until they are 2.5 years of age because, in the event of injury, speech function redistributes to the contralateral hemisphere.

Electrocorticography

For children who have seizures refractory to medication, an epileptogenic focus often is found outside the tumor mass. Thus, tumor resection in patients with medically refractory seizures does not necessarily affect the patient's seizure disorder. To identify seizure foci outside the tumor for these patients, subfrontal, temporal, and interhemispheric subdural grid electrodes are inserted and monitored for 5 to 7 days. During that time, the motor strip also may be identified with stimulation. Tumor resection then may accompany resection of any additional epileptogenic foci. In addition, electrocorticography should be repeated after tumor resection to search for seizure foci that become more apparent after removal of the tumor. In our experience, this paradigm—in which seizure foci identified by using electrocorticography are removed from children with astrocytoma—results in a child who is seizure free without using antiepileptic medication in 90% of cases.

Surgical Navigation

Image-guided navigation is a useful adjuvant to neurosurgery for infants, particularly infants undergoing attempted gross total resection of an astrocytic tumor. Fixation of the skull during surgery for registration is

prerequisite to the use of all navigational systems. Although some pediatric neurosurgeons use pediatric pins at 20 pounds of tension in children from age 1 to 2 years, we have eschewed pins for children younger than 2 years old and have used alternate strategies for head fixation. If the operation requires that the patient be placed in a prone position, the head may be placed on a padded doughnut-shaped cushion and then taped to the bed to minimize movement. If the operation requires that the patient be in a supine position, the skull may be held in position by the Sugita six-point head holder with nonpenetrating pins; the caps of vacutainer vials removed and placed over the pins works well for this purpose.

The operating team attempts to disrupt the head position minimally during the scalp opening. Then four small divots are drilled in the skull at the margins of the anticipated craniotomy; these divots are to be used as internal registration landmarks. After this point, the head position is no longer critical because the navigation system can be reoriented easily by registration using the internal landmarks drilled on the surface of the skull.

Tumor Removal

Most tumors are manageable with resection performed using bipolar electrocautery and controlled wall suction; however, some tumors are sufficiently intractable that they require the use of the ultrasonic aspirator. Minimizing blood loss during tumor resection is essential, and the contact yttrium aluminum garnet (YAG) laser may be helpful in this regard. For extremely vascular tumors, such as hemangioblastoma, we have used the argon-beam laser. Close communication with the anesthesiologist is essential during the removal of vascular lesions because it may be necessary to interrupt the operation and allow fluid or blood administration to prevent hemodynamic instability.

CLOSURE

The bone flap is replaced and fixed into position. Although wire or suture fixation of the bone flap has been used successfully for years by many pediatric neurosurgeons, these methods may permit sinking of the flap over time, potentially resulting in a cosmetic defect. We often use titanium plates or silk suture for fixation of the bone flap. Although we do not believe the use of metallic plates is contraindicated in infants, even though substantial growth of the skull is anticipated, it is encouraging that absorbable plates and screws made from polyglycoxylic acid are now available and hold considerable promise.

The use of subgaleal drains is optional and usually is unnecessary when meticulous hemostasis is used during the opening. The galea is closed as a separate layer by using nondyed absorbable suture, and the skin is closed with a running stitch of nonabsorbable suture. Staples should not be used in young children because children tolerate them poorly.

POSTOPERATIVE MANAGEMENT

Children are monitored postoperatively in the intensive care unit. Antiepileptic drugs are continued postoperatively. Serum levels are checked and maintained in the higher aspect of the therapeutic range. It is important to obtain a postoperative MRI study within 48 hours of surgery, before substantial tissue granulation or inflammation occur, because these events confound interpretation of the scan. Careful attention must be given to the serum hemoglobin and electrolyte values. Intravenous fluids are administered at 50% maintenance until the child is taking fluids normally.

Extraventricular drainage of cerebrospinal fluid is generally continued for 3–5 days, during which the height of the drainage system is increased by 5 cm each day, if tolerated by the patient. Normal intracranial pressure varies according to age; for example, physiologic intracranial pressure in a neonate (<2 mm Hg) is less than that for infants up to 1 year of age (1.5–6 mm Hg), which in turn is less than physiologic intracranial pressure in young children (3–7 mm Hg). If symptomatic hydrocephalus or subgaleal fluid collection persists at the end of this observation period, the cerebrospinal fluid is diverted chronically by using a ventriculoperitoneal shunt.

EDITOR'S COMMENTARY

Brain tumors in infants present a number of challenges. First, these lesions are often large, and in view of the small blood volume in these young patients, life-threatening blood loss is a concern during resection. Second, many lesions arise in midline structures (e.g., hypothalamic gliomas) and may not be amenable to extensive resection. Third, the sequelae of adjuvant radiotherapy are extremely high in this age group. The goals of treatment have generally consisted of achieving a gross total resec-

tion, if this is safely feasible, and deferring irradiation by using chemotherapy in malignant tumors and incompletely resected or inherently unresectable midline gliomas. Unfortunately, for malignant tumors, long-term disease control is poor, even with intensive induction chemotherapy. Strategies that are being pursued to improve these results include the use of highly intensive maintenance chemotherapy with stem cell support and the administration of reduced dose/reduced field irradiation after induction chemotherapy.

PEARLS

In these authors' experience:

- Surgical resection or biopsy is the initial therapy of choice for infants with brain tumors of all types except (1) diffuse pontine or hypothalamic glioma without exophytic components; (2) optic nerve glioma in a patient with excellent vision or in the setting of neurofibromatosis; and (3) subependymal giant cell astrocytoma in a patient with tuberous sclerosis and without symptomatic ventriculomegaly due to obstruction of the foramen of Monro.
- Maximizing the extent of surgical resection of a tumor is critical in infants because gross total resection may prevent the need for adjuvant radiation therapy, which is extraordinarily damaging to the immature nervous system.

SUGGESTED READINGS

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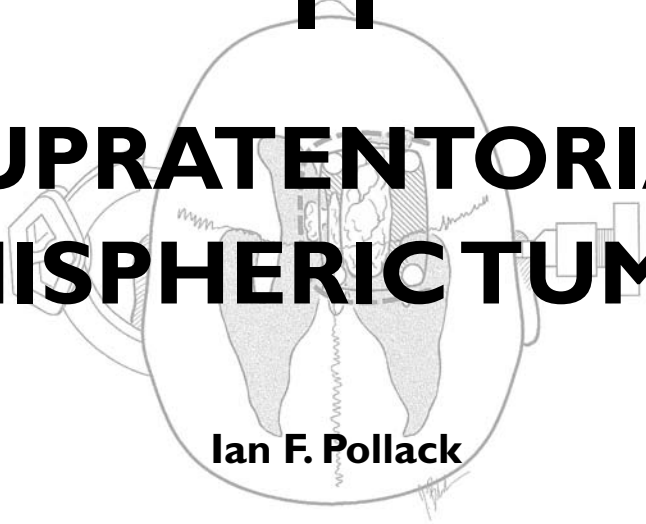
11

SUPRATENTORIAL HEMISPHERIC TUMORS



Ian F. Pollack

Supratentorial tumors account for 40 to 60% of childhood brain tumors and, of these, approximately 60% involve the cerebral hemispheres. In children, as in adults, most such lesions are gliomas. A distinguishing feature is the histologic distribution of these tumors; whereas malignant gliomas account for the bulk of intraparenchymal lesions in adults and in children, the overwhelming majority of such tumors are low-grade gliomas. In addition, meningiomas, which are among the most common supratentorial tumors in adults are rare in children. Another important characteristic of virtually all types of pediatric hemispheric tumors is the strong association between resection extent and outcome, a factor that highlights the importance of obtaining extensive surgical resection when it is safely achievable.

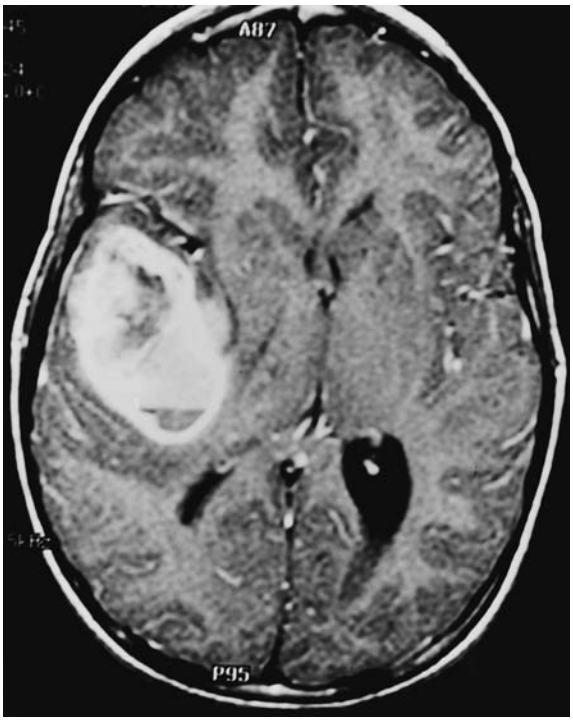
SURGICAL INDICATIONS AND PREOPERATIVE EVALUATION

Cerebral hemispheric tumors characteristically present with seizures and focal neurological deficits, such as hemiparesis, hemisensory deficits, or aphasia, depending on the site of the lesion. In high-grade tumors, the symptoms and signs often progress rapidly, whereas in low-grade lesions, seizures may be present for years without any objective neurologic impairment. Headaches and vomiting, the hallmarks of pediatric infratentorial tumors, are less common with supratentorial tumors because typically cerebral lesions are detected before they produce significantly increased intracranial pressure (ICP) from hydrocephalus or local mass effect. Exceptions to this pattern of presentation are often seen in young children and in adults with intraventricular lesions, who may present with tumors that are quite large. Such patients often manifest symptoms and signs of increased ICP from local mass effect or from obstruction of the ventricular system with resultant hydrocephalus.

Computed tomography (CT) or magnetic resonance imaging (MRI) are usually the only diagnostic studies needed to establish the presence of a supratentorial hemispheric tumor. Characteristic imaging features of these lesions are reviewed in the accompanying textbook and are summarized in Figure 11–1. Other diagnostic studies are rarely needed preoperatively. Lumbar puncture should specifically be avoided because of the risk of herniation and death. Angiography is indicated only for tumors that have unusual vascularity, in which preoperative embolization may be entertained (e.g., for large extracortical tumors, such as in Figs. 11–1G and H) or if concern is raised that the lesion may be a vascular malformation. Neuraxis imaging is needed for only a subset of supratentorial tumors, such as primitive neuroectodermal tumors (PNETs) and perhaps ependymomas and malignant gliomas, and is best reserved for the postoperative period after a histologic diagnosis has been obtained. If indicated, it can then be combined with imaging of the head to provide an assessment of the extent of residual disease.

PREOPERATIVE MANAGEMENT

Most children referred for neurosurgical evaluation have already had a CT or MRI, but if a good quality study has not been performed, we obtain a high-resolution MRI



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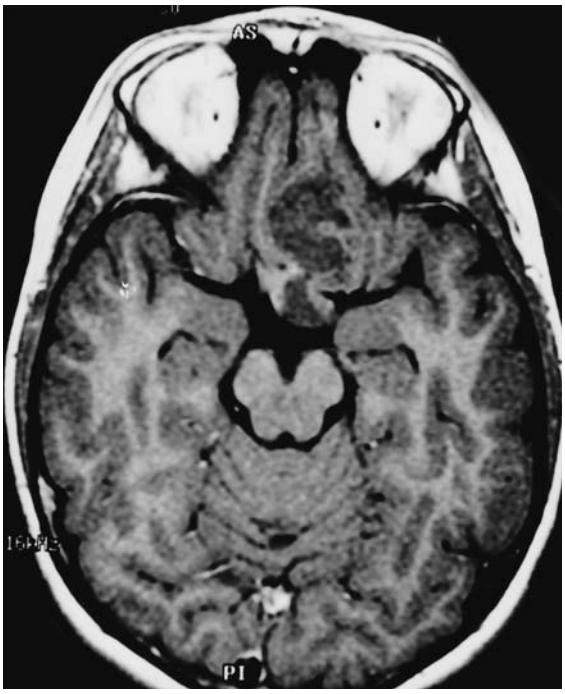

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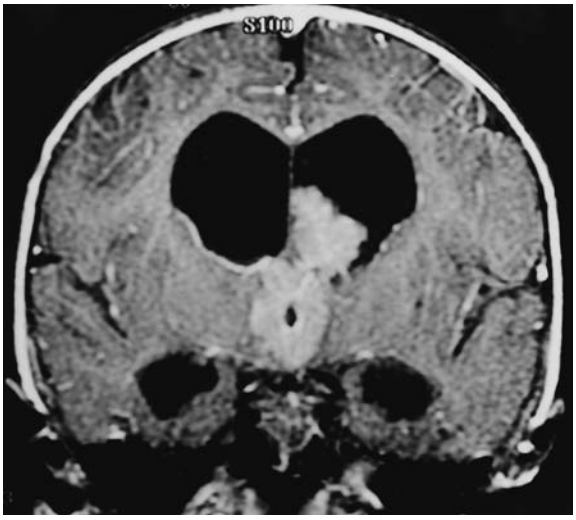

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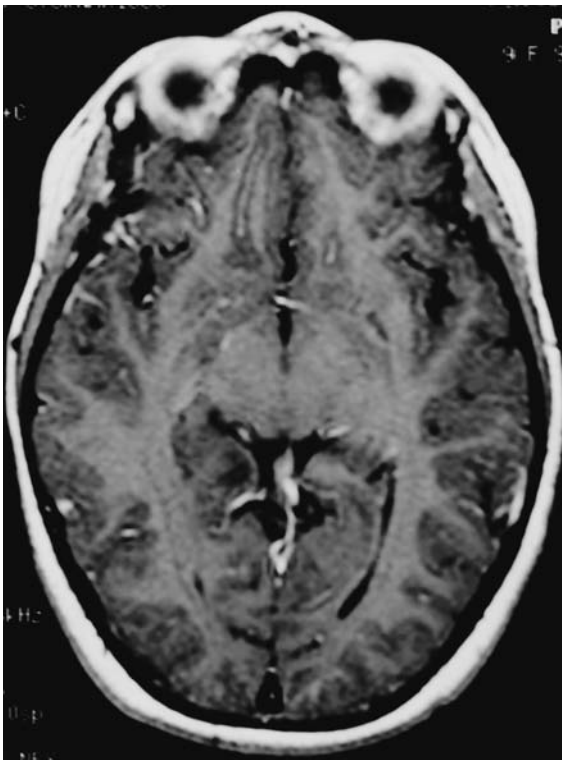

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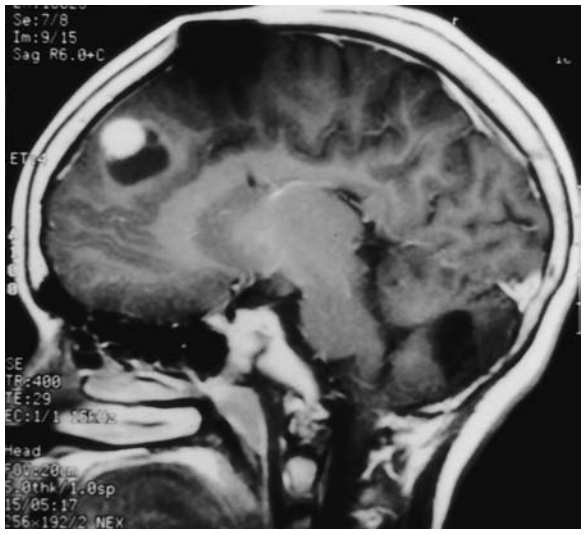
FIGURE 11–1. Characteristic imaging features of several common supratentorial hemispheric lesions in a variety of locations. **A:** Temporoparietal malignant glioma. **B:** Low frontal benign neuroepithelial tumor. **C:** Lateral and third ventricular choroid plexus papilloma. **D:** Occipital and posteromedial temporal nonpilocytic astrocytoma.

examination, in some cases using a format suitable for frameless stereotaxy, which for small or deep-seated lesions or tumors in functionally important cortex helps both to localize the lesion and to assist with operative planning. The timing of operative intervention is determined largely by the condition of the child. Patients who present with obtundation from a large mass lesion undergo resection on the day of admission. Children who have a large lesion but are minimally symptomatic undergo surgery the next available operating day. Smaller



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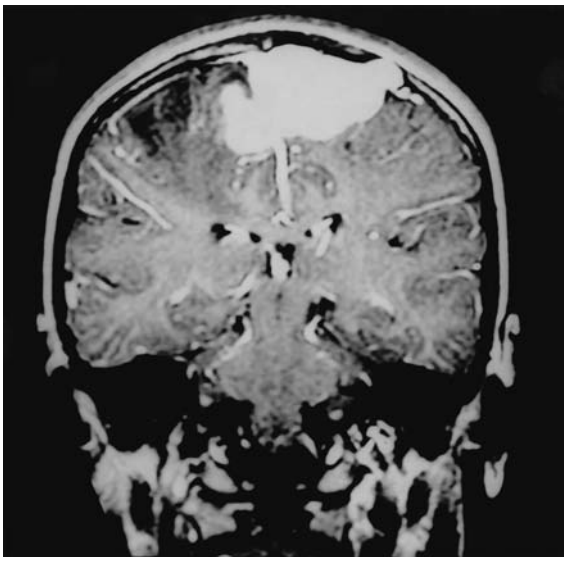

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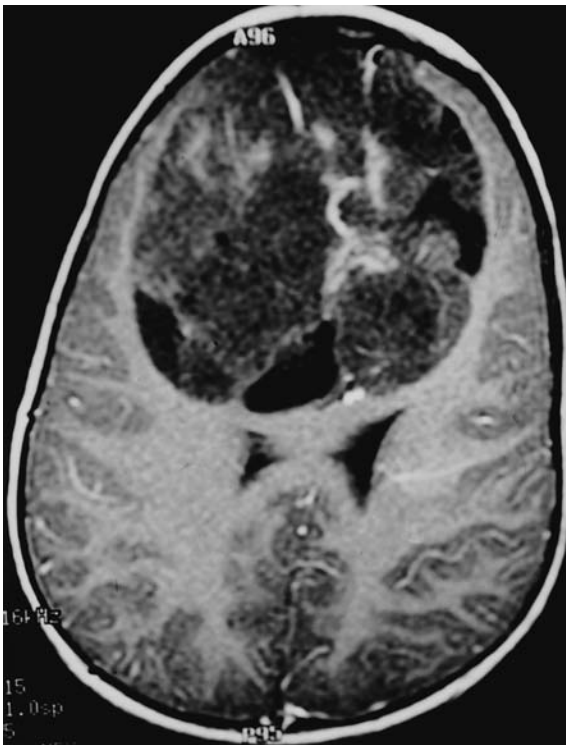

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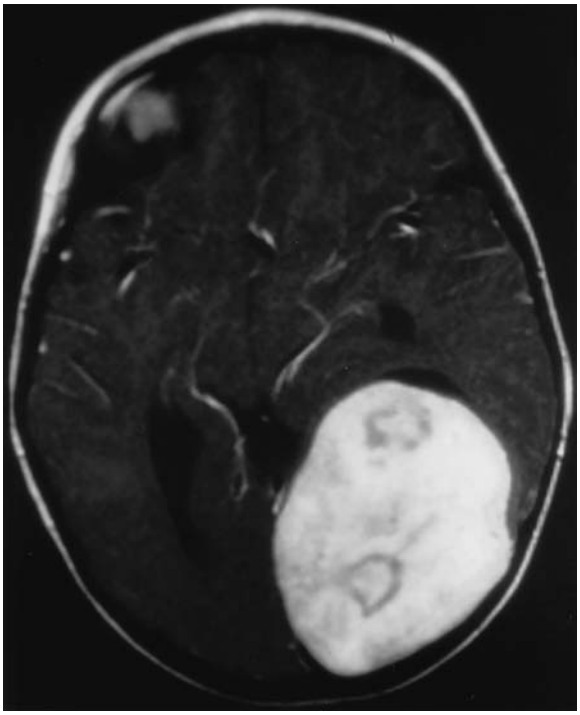


H

FIGURE 11–1. (*continued*) **E:** Posterior frontal pilocytic astrocytoma. **F:** Bihemispheric primitive neuroectodermal tumor. **G:** Intradural extracortical bihemispheric chondrosarcoma. **H:** Intradural and extradural myofibroma.

lesions that present with seizures and minimal mass effect are treated on a more elective basis.

Corticosteroids are generally begun on admission in children with large tumors or are administered preoperatively in patients with smaller lesions at a dose of 1 to 6 mg every 6 hours (for dexamethasone), depending on the child's size. These doses are continued intraoperatively. Because children with hemispheric tumors may be at risk for seizures during the perioperative period, anticonvulsants generally are begun preoperatively, even if the child has not previously had a seizure. In view of the growing trend toward minimizing or avoiding shaving the patient's hair before the surgical procedure, an antibacterial shampoo the night before surgery and on the

morning of surgery often is used to decrease skin and hair flora.

OPERATIVE PLANNING

In general, surgical intervention lays the foundation for the treatment plan by providing tissue to establish the histologic diagnosis and, if feasible, achieving cytoreduction. When selecting the optimal approach to a supratentorial hemispheric lesion, it is critical to develop a clear plan of the goals of the operation (e.g., biopsy, reduction of mass effect, major cytoreduction, treatment of hydrocephalus), which are influenced by the growth characteristics of the tumor as depicted by CT or, preferably, MRI. Ideally, for well-circumscribed lesions, a gross total resection should be the operative goal if it can be achieved without inordinate risk. This is feasible for most pilocytic astrocytomas, even if they arise in subcortical regions; for choroid plexus papillomas; for many superficial nonpilocytic astrocytomas and benign neuroepithelial tumors; and for some superficial high-grade gliomas.

Conversely, for some poorly circumscribed high-grade gliomas and nonpilocytic low-grade gliomas that cross the midline or extensively invade the deep nuclei and other critical brain regions, extensive resection may not be feasible without unacceptable morbidity. In some cases, a percutaneous CT- or MRI-guided stereotactic biopsy may be preferable to an extensive open operation with limited tumor removal. For large lesions with significant mass effect, however, a radical subtotal resection may be of value in stabilizing the child in preparation for adjuvant therapy.

Fortunately, a number of adjuncts for preoperative and intraoperative planning have become available during the last several years that have facilitated extensive removal of lesions previously thought to be unresectable or resectable only with substantial morbidity (Fig. 11–2). Frame-based or frameless stereotactic guidance systems allow precise localization of the tumor (Fig. 11–2A), which permits the surgeon to choose an approach to the lesion that minimizes manipulation of functionally critical brain and, potentially, increases the safety of aggressive tumor removal. Ultrasound is also useful in providing real-time feedback on the location of the lesion, which avoids problems with intraoperative brain movements that limit the accuracy of stereotactic techniques after the tumor resection has been initiated. Functional mapping approaches also have a role in the resection of lesions situated in or beneath critical brain regions. For example, cortical stimulation techniques are useful for identifying speech and motor areas, whereas somatosensory evoked potential recordings are useful for delimiting the primary sensory cortex and central sulcus (Fig. 11–2B). Functional MRI offers another way for localizing critical areas before beginning a tumor resection (Fig. 11–2C); this information can be integrated with stereotactic techniques to delineate precisely important loci around the tumor. Finally, in patients with intractable seizures in association with cerebral cortical lesions, electrocorticography provides a means for defining areas of epileptogenic cortex in and around the tumor to optimize the chances for postoperative seizure control (Fig. 11–2D).

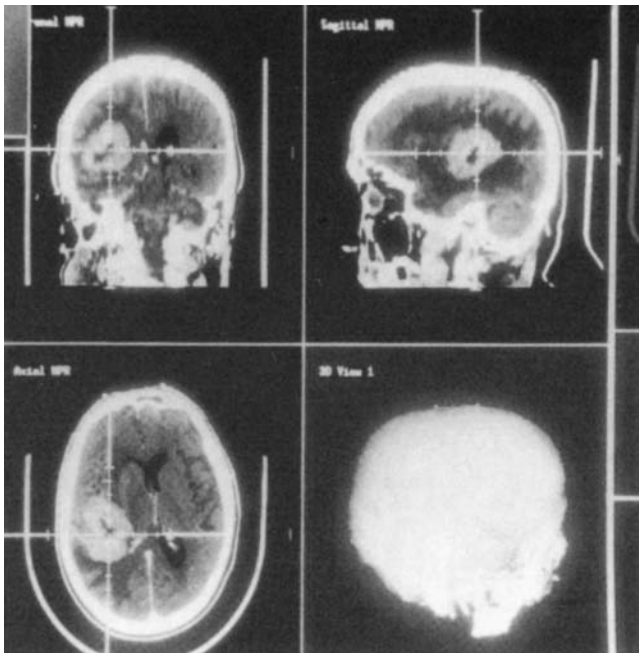
INTRAOPERATIVE TECHNIQUES

Anesthetic Techniques and Positioning

For patients who are appropriate candidates for open resection, the anesthetic technique, positioning, and surgical approach are determined both by the location of the lesion and the type of intraoperative monitoring that is planned. The anesthetic technique generally consists of a mixture of fentanyl, vecuronium, nitrous oxide, and isoflurane. The approach is modified somewhat depending on the type of monitoring being used. For example, somatosensory evoked potential monitoring requires a reduction in the levels of inhalation agents, whereas motor mapping requires a limitation in the level of paralysis; the other agents are adjusted accordingly. For tumors that arise in potential speech areas, awake craniotomy techniques are useful in older children for localizing critical regions. Because these techniques are often not feasible in young children, we generally perform preliminary extraoperative speech mapping using strip or grid electrodes before embarking on a resection in these areas. Achieving reliable monitoring while keeping the patient appropriately anesthetized requires that the anesthesiologist be aware of the surgeon's plans preoperatively.

Other common features of the surgical preparation include insertion of a urinary drainage catheter, arterial line, and, in cases in which significant blood loss is anticipated, a central line and sizable peripheral intravenous lines to facilitate expeditious replacement of blood and clotting factors, if needed. Mild hyperventilation (to a pCO_2 of 30 to 35) is instituted if increased ICP is a concern. Prophylactic antibiotics are administered during the skin preparation and every 6 to 8 hours during the procedure. Corticosteroids and anticonvulsants are also continued intraoperatively.

For situations in which a ventricle is partially trapped by tumor and the risk of perioperative hydrocephalus is a concern, a ventricular catheter may be



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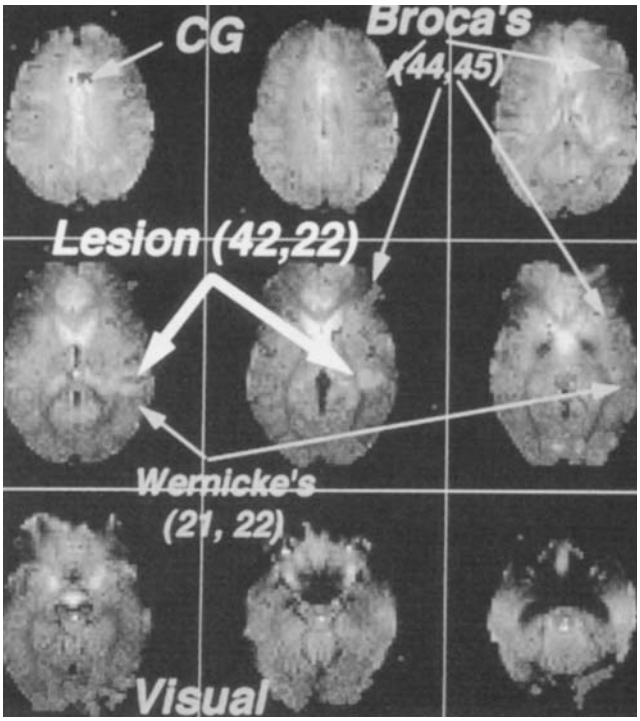

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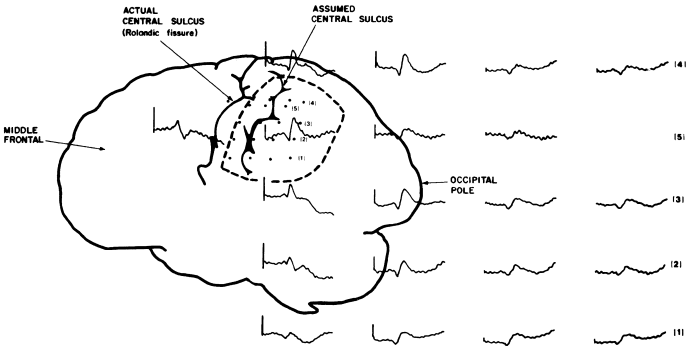

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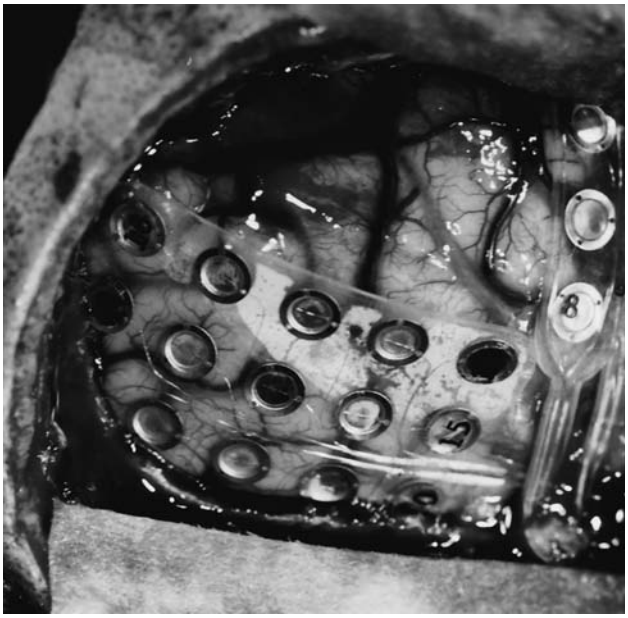

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FIGURE 11–2. Preoperative and intraoperative adjuncts that facilitate tumor resection. **A:** Stereotactic localization using frameless navigation allows planning of the operative trajectory and facilitates anticipation of the tumor margins during the course of the resection. **B:** Intraoperative somatosensory evoked potential mapping of the central sulcus, which, as illustrated here, is often displaced or distorted by a nearby tumor. In this case, the data recorded from a strip grid positioned over the lesion, both posterior and anterior to the presumed central sulcus, all demonstrated a classic N_{20}, P_{30} complex, suggesting that this region was actually parietal cortex. Only when the recording strip was advanced anterior to the lesion was an N_{20}, P_{30} inversion consistent with the motor cortex observed. The lesion was resected with the patient suffering no motor deficits. **C:** Functional magnetic resonance imaging mapping of speech functions, which as illustrated here, are in close proximity to a dominant parietal tumor. **D:** Extraoperative functional mapping and electrocorticography using subdural grids and strips and intraoperative electrocorticography are useful for defining functionally significant cortex in and around a tumor as well as identifying perilesional seizure foci.

placed either before the initial exposure or after the craniotomy has been completed, depending on the planned surgical approach. The ventriculostomy is attached to a drip chamber, which is kept elevated or closed until the dura is exposed; if the dura is tight, the anesthesiologist can allow cerebrospinal fluid to drain out in a controlled fashion.

The positioning used for tumor resection is commonly supine or lateral decubitus, depending on the operative trajectory. The prone position is used for occipital tumors. A three-pin head fixation device is generally used for children older than 2 years, whereas a horseshoe headrest is commonly used in younger patients. Care should be taken to avoid excessive tightening of the headholder, particularly in young children, because this can lead to an iatrogenic depressed skull fracture. Attention must also be directed at padding all potential pressure points and avoiding extremes of neck rotation or flexion that may place traction on the brachial plexus or compression of the jugular veins.

If frameless stereotactic localization is to be used, the device is at this point registered to the patient's craniofacial surface anatomy and any fiducials that may have placed during a preoperative localizing study. This facilitates planning of the subsequent incision and bone flap for small or deep-seated lesions.

Initial Exposure

We currently avoid shaving large areas of the head but instead clip or shave a 1- to 2-cm strip along the planned incision line and then scrub the hair and scalp with an antibacterial soap before formal skin preparation. The skin incision is determined by the location of the lesion (Fig. 11–3 corresponds to the exposures used to expose the lesions illustrated in Fig. 11–1). A question mark or C-shaped incision is used for temporal lesions (Fig. 11–3A), extending upward from the zygoma, with the base less than 1 cm anterior to the ear and the superior end curving either anteriorly or posteriorly and then anteriorly in a gentle arc, depending on the position of the tumor. The anterior portion of the incision extends toward the midline, but it is kept behind the hairline. A bicoronal incision beginning 1 cm anterior and superior to tragus of the ear on each side is used for low frontal lesions (Fig. 11–3B) or if bihemispheric exposure is required (as for the lesions depicted in Figs. 11–1F and G). A C-shaped incision is used for posterior frontal, parietal, occipital, or intraventricular lesions to preserve the predominant sources of vascular supply to the skin flap (Fig. 11–3C and D). Alternatively, a linear incision may be used for smaller lesions (such as the one illustrated in Fig. 11–1E), particularly if stereotactic guidance is being used to minimize the extent of the bone opening. In general, the incision is performed in layers, using a no. 15 blade to incise the skin and a needle-tip cautery to incise the galea and periosteum. With the latter maneuver, care must be taken to avoid thermal injury to the skin, but if it is properly executed, this approach facilitates scalp exposure with minimal blood loss; skin clips are rarely required. The scalp flap then is reflected subperiosteally off the underlying calvarium and retracted using skin hooks attached to rubberbands, which are secured to the surrounding drapes.

As with the skin incision, the site and extent of the craniotomy are determined by the location and size of the tumor and, if indicated, may be guided using stereotactic techniques. The latter approach enables the surgeon to limit the extent of the craniotomy in selected cases. Conversely, if intraoperative functional monitoring, mapping, or electrocorticography are to be performed, the extent of the craniotomy must be increased accordingly. At least two burr holes are made at the periphery of the planned exposure using a high-speed drill (e.g., a Midas Rex M8 bit). For convexity lesions, the exact orientation of the burr holes is determined by the surgeon's preference. For more lateral, medial, anterior, or posterior exposures, the location of the burr holes is more critical (Figs. 11–3 and 11–4).

For a temporal craniotomy (Figs. 11–3A and 11–4A), one of the burr holes is made at the pterion and another low over the temporal bone to facilitate lateral and inferior exposure; one or two additional burr holes are made above the superior temporal line, depending on the extent of superior, anterior, or posterior exposure needed. For a low frontal exposure (Figs. 11–3B and 11–4B), I generally make two small anterior burr holes just above the orbital rim medially and laterally and, in some cases, also place a third burr hole at the posterior limit of the planned exposure. The anterior bone cuts then can be carried down to the frontal base or, if needed, continued into the orbit to allow an orbitofrontal craniotomy.

For a medially situated craniotomy to expose a medial frontal or parietal lesion, burr holes are made over the midline at the anterior and posterior limits of the exposure; great care must be taken to avoid injuring the sagittal sinus during this step. Additional burr holes then are made over the convexity, depending on the amount of lateral exposure needed. A similar approach is taken in preparation for a transcallosal exposure (Figs. 11–3C and 11–4C), for which I generally use a craniotomy bone flap that is approximately 5 cm in anterior–posterior length and is situated two thirds in front and one third behind the coronal suture, which provides a familiar trajectory

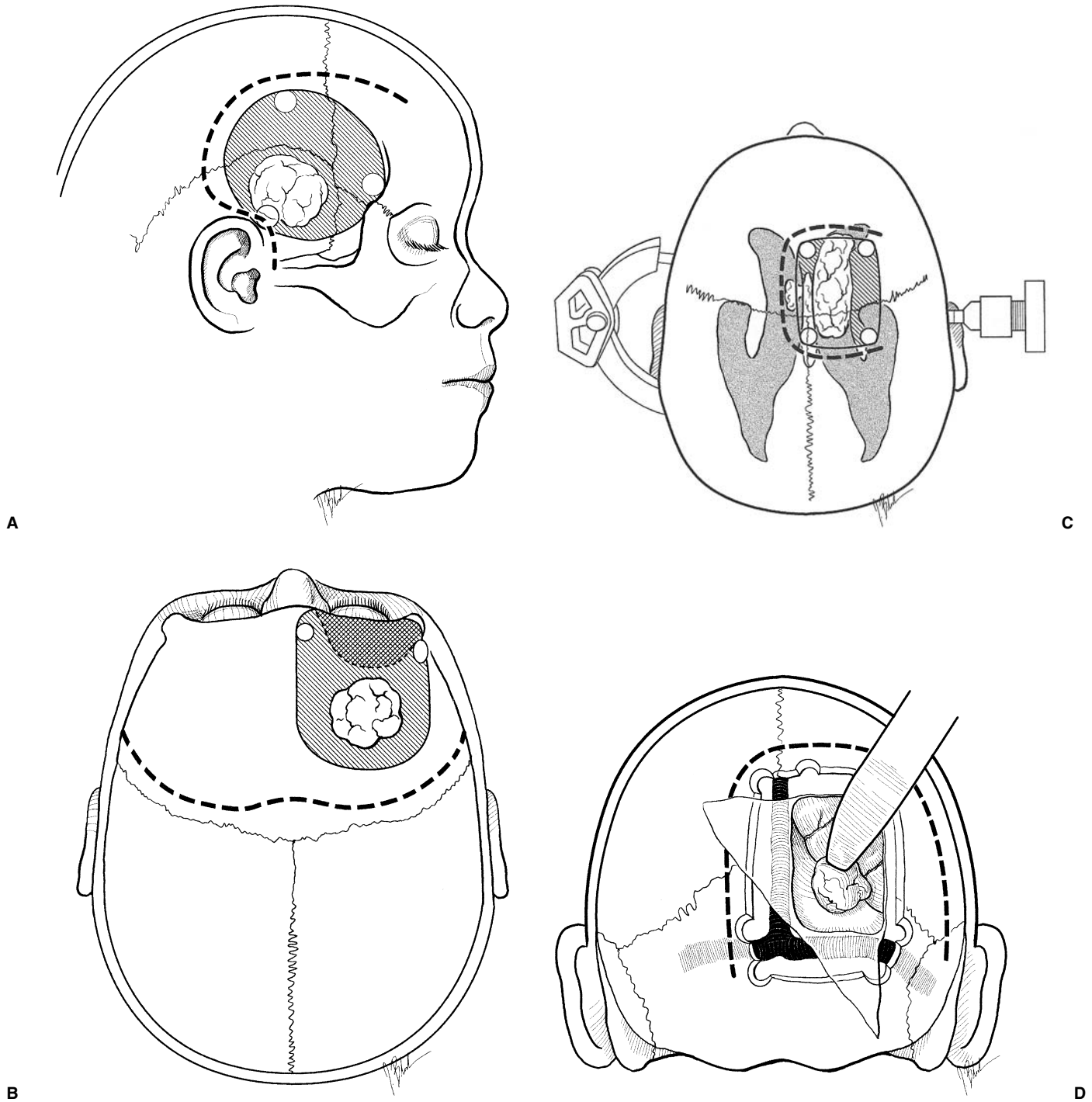
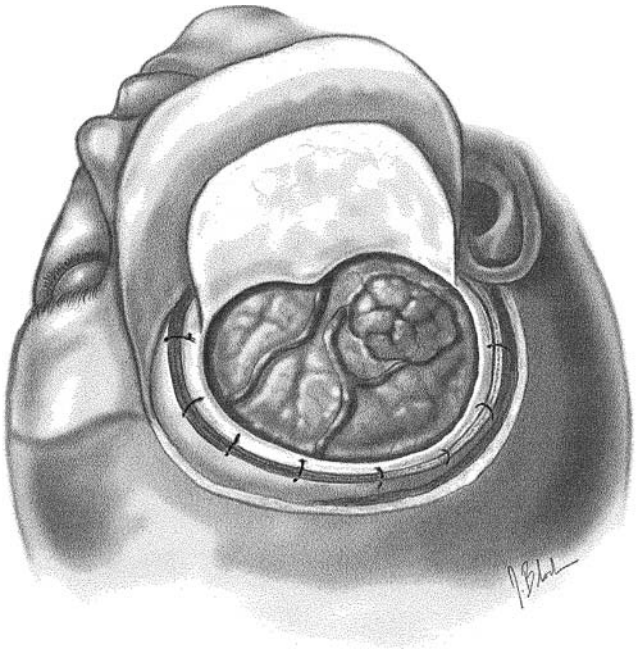


FIGURE 11–3. Skin incisions (indicated as *dashed lines*) and operative approaches for supratentorial hemispheric lesions in various locations. **A:** Temporal lesion. **B:** Low-frontal lesion. **C:** Intraventricular lesion. **D:** Occipital lesion.

to the lateral ventricle and foramen of Monro and minimizes the risk of venous infarction from interruption of draining veins from the cortex during the subsequent intradural exposure. If a bihemispheric approach is needed (as for the lesions illustrated in Figs. 11–1F and G), burr holes are placed laterally on both sides in addition to the midline holes, which allows elevation of separate bone flaps on each side and minimizes the risk of injuring the sagittal sinus during subsequent removal of the bone. Finally, if a low posterior exposure is planned for an oc-



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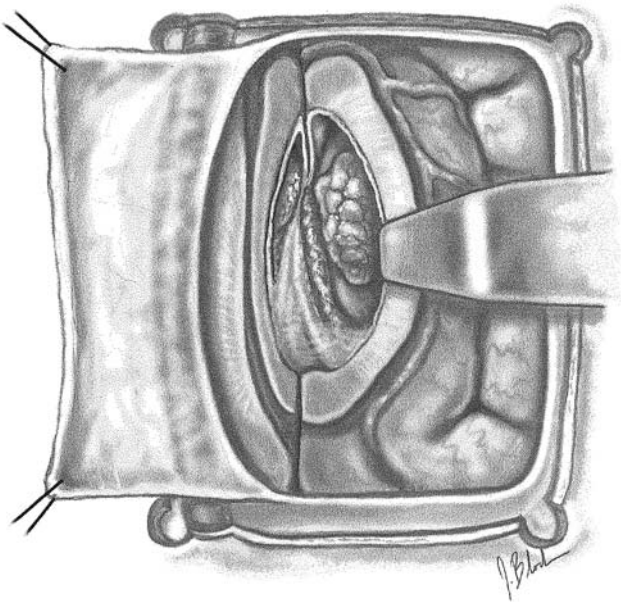

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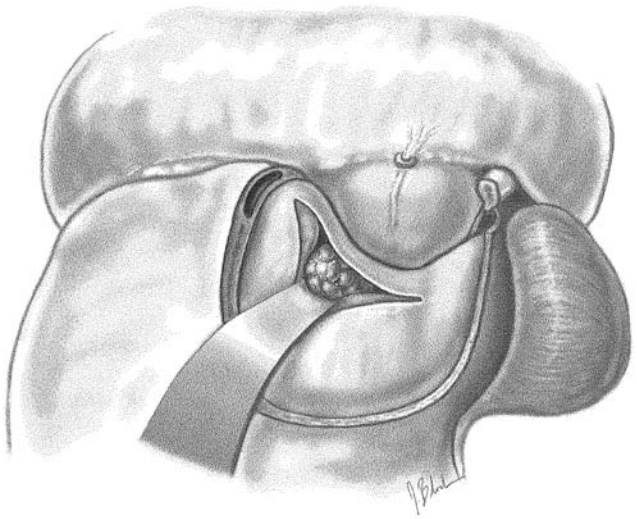

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FIGURE 11–4. Craniotomies and dural openings for supratentorial hemispheric lesions in various locations. **A:** Temporal lesion. **B:** Low frontal lesion. **C:** Intraventricular lesion. The craniotomy and dural opening for an occipital lesion are depicted in Figure 11–3D.

cipital or posterior hippocampal lesion (Fig. 11–3D), not only the sagittal but also the transverse sinus must be visualized and protected, and the medial and inferior burr holes must be planned and executed accordingly.

An important caveat in the removal of large extracortical tumors is the need for generous exposure to facilitate resection and reconstruction of the surrounding involved dura, if needed. This is particularly critical for those lesions that arise in the vicinity of a major venous sinus (e.g., Figs. 11–1G and H) because control and potentially reconstruction of the sinus on either side of the tumor may be required.

If frameless stereotactic techniques are to be used during the tumor resection, I generally register a series of secondary landmarks around the site of the planned craniotomy at this time. This step is performed because slight movement of the head occasionally may occur during the next (craniotomy) step, particularly if a horseshoe headrest has been used, which can degrade the accuracy of the primary localization. The location of the secondary points can be rechecked after the craniotomy and, if movement has occurred, the head cam be repositioned or the orientation of the stereotactic map in relation to these points readjusted accordingly.

The dura is dissected from the undersurface of the bone, and the burr holes are connected using a craniotome bit (e.g., a Midas Rex B5 bit). If an orbitofrontal approach is required for an inferior-medial frontal lesion, the intraorbital cuts can be made using a Midas B5 bit with a B attachment (or alternatively with a reciprocating saw), which allows the orbitofrontal bone piece to be cracked off from the posterior orbital roof as a single segment.

Usually, if the craniotomy has been situated appropriately, no additional bone removal is required prior to dural opening. An exception occurs with inferior-medial temporal lesions or if a transsylvian exposure is anticipated. In these cases, inferior exposure can be enhanced by using a rongeur to resect the temporal bone down to the middle fossa floor, and transsylvian exposure can be facilitated by resecting the lateral sphenoid ridge using a combination of rongeurs or a high-speed drill.

Dural Opening

Prior to the dural opening, I generally place tacking sutures circumferentially around the craniotomy site, if possible. The dura then is opened in a C-shaped, U-shaped, or stellate fashion. If indicated, stereotactic or ultrasound guidance may be used at this point to localize the tumor in relation to the planned opening. For most laterally situated lesions, the exact orientation of the dural opening is not critical as long as sufficient exposure is obtained. For medially situated frontal or parietal lesions or for cases in which a transcallosal exposure is planned, however, a U-shaped, medially based dural opening is preferred to facilitate exposure up to the falx. In the latter case, care must be taken to avoid inadvertent injury to cortical veins draining into the sagittal sinus or to the sinus itself. Similarly, for low occipital exposures, the dura is opened in a C-shaped fashion, based medially and inferiorly, and a relaxing incision is made toward the junction of the sagittal and transverse sinuses with the torcular to permit maximal inferior and medial exposure.

If the dura is tense to palpation, mannitol (0.25 g/kg) is administered before the dural opening; if a large cyst is present, it is aspirated. Both measures are designed to prevent the brain from herniating through the dural opening. If the dura remains tense despite these measures and other anesthetic techniques have been optimized (e.g., the head is elevated slightly, compression of the jugular veins has been avoided, and mild hypocarbia has been maintained by slight hyperventilation) and a functioning external ventricular drain is in place if hydrocephalus is present, the dura is opened immediately over the lesion and internal debulking of the tumor is performed to achieve preliminary decompression before completing the dural incision. Once the dura is opened, the brain surface is protected using cottonoid patties or biological collagen sponge.

Surgical Approach

For extracortical tumors, the lesion is readily apparent on opening the dura and is easily distinguished from the surrounding brain. Such tumors are uncommon in pediatric neurosurgery, but may include meningiomas, dural-based sarcomas (Fig. 11–1G) and metastases, myofibromas (Fig. 11–1H), and occasional gliomas. Because these lesions are often extremely vascular, preoperative embolization is commonly done. If this is not feasible, extra effort must be directed intraoperatively to identify and occlude feeding vessels arising from the bone or dura to minimize blood loss. The tumor capsule then is coagulated and incised, and the plane between tumor and brain is identified. The lesion is debulked and resected as described below. If the lesion is entirely extracortical, a complete resection can be achieved without violating the underlying brain surface.

For superficial cortical and subcortical tumors, the most direct trajectory to the lesion is usually appropriate. In many cases, the involved gyrus is immediately apparent because it is enlarged, discolored, or firm to palpation. If these changes are not detected, stereotactic localization, if used, can help to identify the site of the tumor. If these techniques are not available, ultrasonography may be used to target the corticotomy. Finally, for lesions arising in or around critical brain regions, such as the somatosensory cortex, functional mapping may be performed at this time in conjunction with stereotactic or ultrasonographic techniques to choose a trajectory to the lesion that avoids violating functionally significant cortex. Once the entry site has been selected, the pial surface is coagulated and incised over the tumor and the resection is performed as outlined later. Some surgeons prefer a transsulcal approach, although I generally prefer to place my corticotomy over the most superficial point of the gyrus to avoid injury to the sulcal vessels. The corticotomy then is deepened through the gray and white matter until the tumor is reached. The length of the cortical exposure is extended as needed to provide adequate exposure of the tumor, and self-retaining retractors are placed over cottonoid patties to protect the brain along the corridor of the exposure.

For deep subcortical tumors, stereotactic or ultrasonographic techniques and, if indicated, functional

mapping techniques are useful for selecting the safest approach to the tumor. These strategies are particularly valuable for lesions that arise from or extend into the thalamus and basal ganglia (Fig. 11–5). The approach to these deep subcortical lesions is also influenced substantially by the predominant direction of tumor growth. Lesions that grow medially and encroach on the lateral ventricle can be approached transcallosally or transfrontally (Fig. 11–5A), whereas tumors that extend laterally in the nondominant hemisphere may be approached through the insula after the sylvian fissure has been opened (Fig. 11–5B). Laterally extending lesions within the dominant hemisphere and tumors that arise more posteriorly within the thalamus may be reached using a posterior parietal approach behind the sensorimotor cortex and above the angular gyrus (Fig. 11–5C). Selected lesions also can be reached through an occipital transtentorial trajectory through an opening in the pulvinar (Fig. 11–5D). Tumors that project anteriorly and laterally can be reached from a paramedian frontal trajectory, provided that care is taken to avoid injury to the motor pathways.

For lateral ventricular tumors, the predominant direction of tumor growth and the pattern of vascular supply determine the optimal approach. For lesions that arise in the body or frontal horn of the lateral ventricle, which often have vascular pedicle at the region of the foramen of Monro, I use a transcallosal approach to obtain tumor exposure (Fig. 11–6); some surgeons prefer a transcortical trajectory. The corpus callosum is identified by its striking white color (Fig. 11–6B), which is readily distinguished from the grayish hue of the overlying cingulate gyri, which often are closely opposed in the midline and must be separated to provide callosal exposure. Division of cortical veins entering the sagittal sinus is avoided if possible, but if these hamper the exposure, I occasionally divide an anterior vein if it is small and have encountered no sequelae when this maneuver is used. Lateral retraction on the hemisphere is maintained using cottonballs rather than retractors to minimize the likelihood of cortical injury. The intraventricular tumor then is exposed through a 2- to 3-cm opening in the corpus callosum. Regardless of the side of initial ventricular entry, it is generally possible to achieve adequate biventricular exposure by extending the callosotomy or fenestrating the septum pellucidum. Both maneuvers are often needed for lateral ventricular tumors that extend bilaterally via the foramen of Monro (Figs. 11–1C and 11–6C). Additional exposure of an associated third ventricular tumor component can be obtained, if needed, using a subchoroidal exposure.

Lesions arising in the temporal horn and receiving their blood supply from the anterior choroidal artery are approached by a 2- to 3-cm incision in the inferior temporal gyrus. Finally, tumors arising in the atrium of the lateral ventricle may be approached using either a middle temporal gyrus or posterolateral parietal corticotomy, which allows early control of feeding vessels from the thalamogeniculate and posterior choroidal arteries, respectively. Large lesions may require a combination of approaches. As noted later, control of the vascular pedicle is an essential step before beginning the tumor resection to minimize blood loss, and the approach(es) should be chosen to facilitate this maneuver.

Tumor Resection

Most extracortical tumors show extensive attachment to or invasion of the meninges, which (if safely feasible) should be removed along with the tumor to minimize the risk of recurrence. The large size and dense vascularity of many of these tumors mandate that arterial and central venous monitoring and adequate intravenous access be obtained before beginning the resection and that appropriate blood and clotting factor replacement be provided expeditiously because death from hypovolemia or coagulopathy has historically been a significant risk. As noted earlier, the identification and occlusion of superficial feeding vessels to the lesion using embolization, operative coagulation, or a combination thereof, are essential steps prior to the debulking of large vascular lesions. The central component of the lesion is resected using the ultrasonic aspirator, large-bore suction, or ring cautery. I usually obtain a frozen-section diagnosis at an early stage of the removal of most tumors (including each of the groups discussed later) if the diagnosis is in question, both to ensure that the pathologist has an adequate specimen to examine and to guide other intraoperative management. For example, if the intraoperative diagnosis of an extracortical tumor is sarcoma, we proceed with insertion of a mediport and performance of a bone marrow aspirate under the same anesthetic and begin our staging workup in the immediate postoperative period.

After the central component of the tumor has been resected, the plane between tumor and brain is progressively delineated as the tumor capsule is mobilized inward, and cottonoid patties are placed on the brain surface to maintain the exposure. For tumors that are deeply invaginated into the brain, the process of internal debulking and capsule mobilization may need to be repeated sequentially until the full extent of the tumor is delivered and resected.

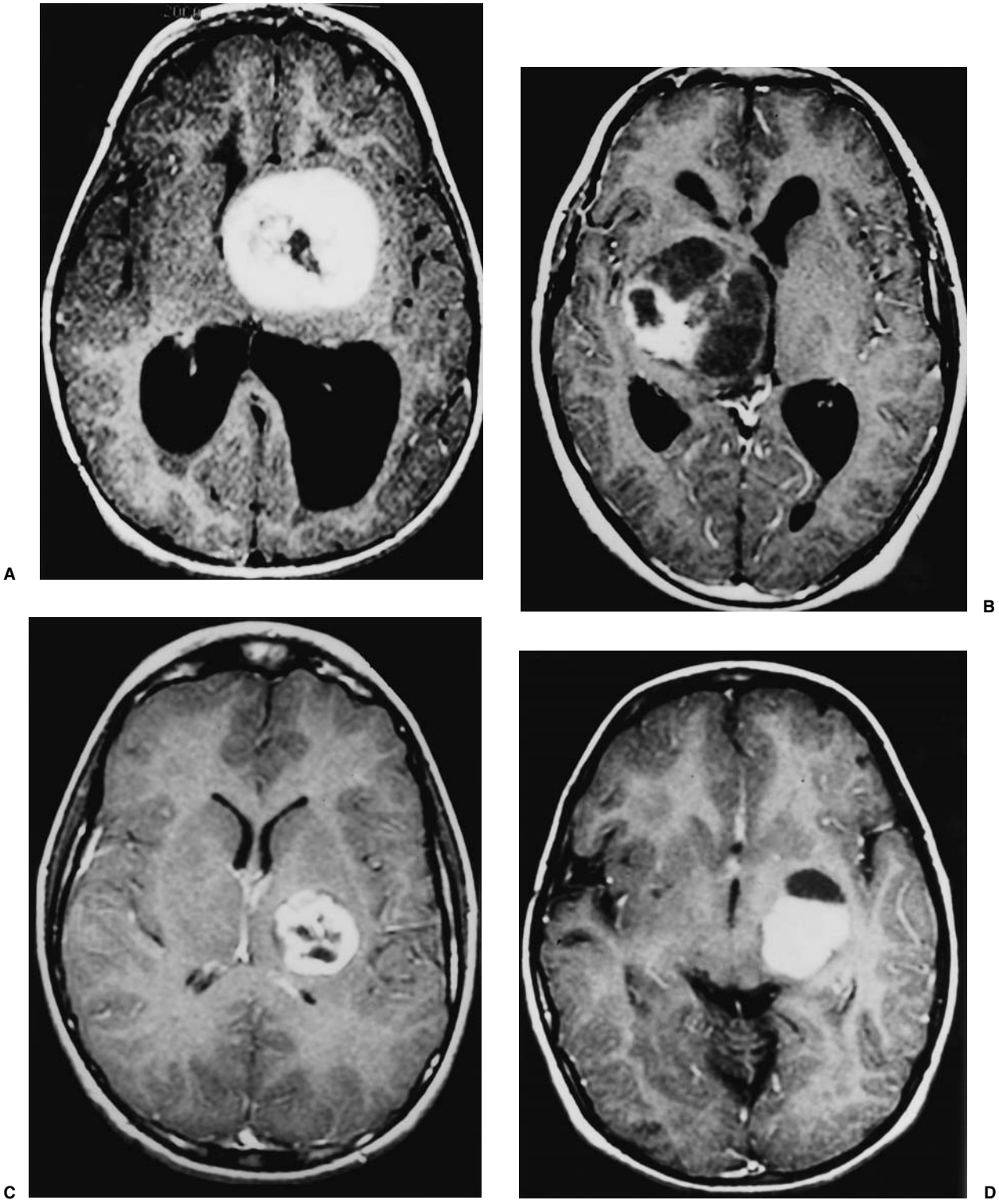
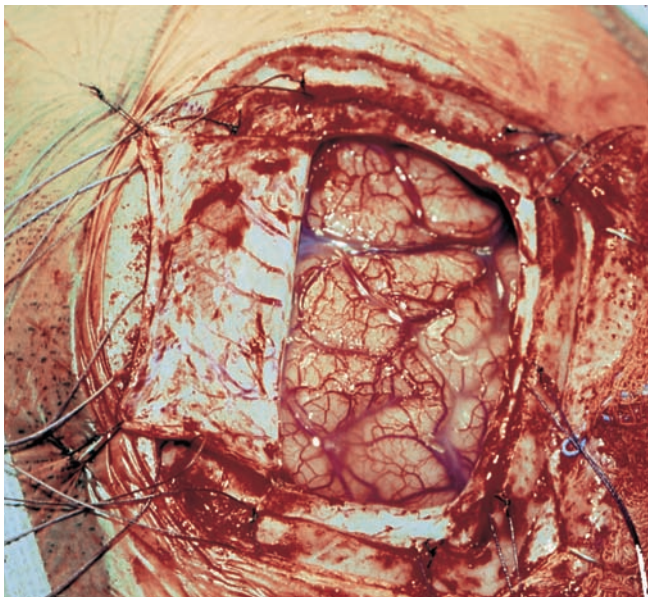


FIGURE 11–5. Subcortical tumors reached by a variety of trajectories. **A:** This huge, partially intraventricular, dominant hemisphere thalamic pilocytic astrocytoma was resected transcallosally, which facilitated opening of the ventricular system and fenestration of the septum pellucidum. **B:** This partially cystic, laterally projecting, nondominant hemisphere thalamic astrocytoma was resected through an opening in the posterior sylvian fissure. **C:** This posterolaterally projecting dominant hemisphere ependymoma was resected through a posterior parietal approach, avoiding the speech and sensorimotor pathways. **D:** This dominant hemisphere pilocytic astrocytoma was resected by means of an occipital transtentorial approach through the pulvinar.



A

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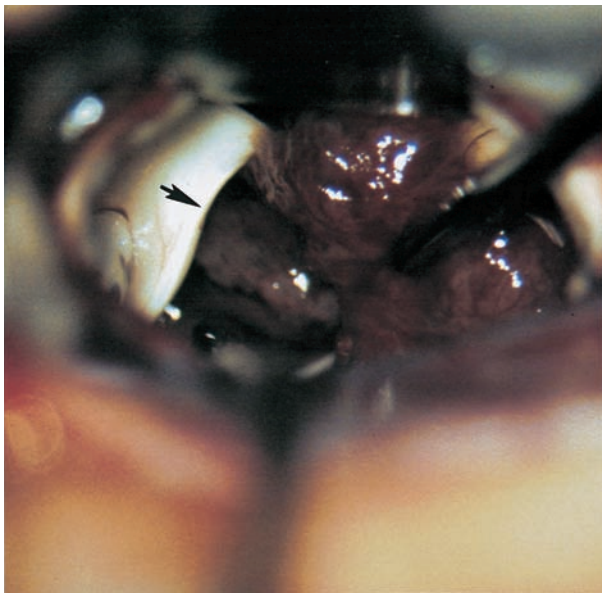

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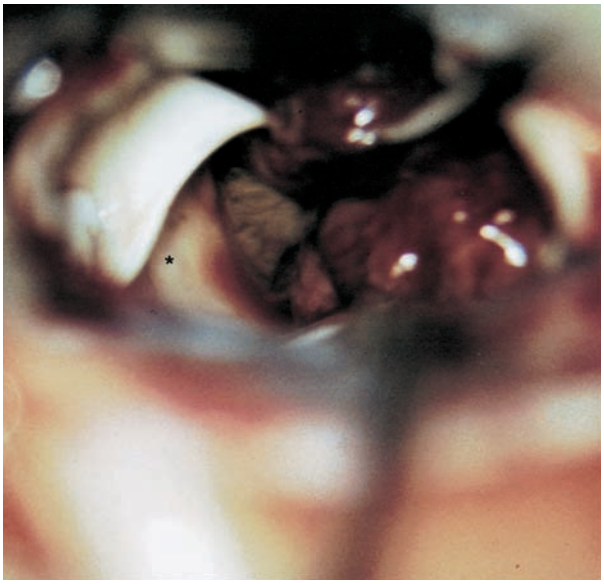

D

FIGURE 11–6. Operative exposure for resection of lateral and third ventricular choroid plexus papilloma. **A:** Dural opening for transcallosal approach. **B:** After the hemisphere was retracted laterally (toward to the right), the corpus callosum can be visualized as a glistening white structure. The corpus callosum is divided for length of 2 to 3 cm between the pericallosal arteries. The tumor is seen bulging through the partial opening in the corpus callosum. **C:** Within the lateral ventricle, the purplish brown tumor is seen growing through the widely dilated right foramen of Monro (*arrow*). **D:** Following coagulation, shrinkage, and partial debulking of the lesion, the ependyma of the third ventricular floor (*asterisk*) is apparent.

The mode of resection of cortical and subcortical tumors depends on the growth characteristics of the tumor. Many pilocytic astrocytomas and ependymomas have a well-delineated peritumoral plane through which the neoplasm can be separated from the surrounding brain after the central portion of the mass has been debulked. In such cases, the tumor pseudocapsule is coagulated and retracted inwards away from the surrounding brain until the bulkiness of the tumor limits further mobilization. Vessels extending between the tumor and cor-

tex are coagulated and divided, which facilitates internal debulking, and cottonoid patties are placed in the plane between tumor and brain. The central portion of the tumor then is debulked. As progressively deeper portions of the tumor are reached, the sequence of pericapsular dissection, placement of patties, and internal debulking is repeated as many times as needed to allow circumferential delineation of the lesion with minimal retraction on the surrounding brain.

For most nonpilocytic gliomas and high-grade gliomas, no such plane between tumor and brain is observed, and the resection must proceed cautiously from the inside outward, until a boundary between tumor and normal brain is approached. In relatively silent areas of the brain, it may be feasible to resect the tumor circumferentially until normal-appearing white matter is encountered, but in more critical regions, this maneuver is potentially hazardous. In such cases, intraoperative functional mapping may enhance the safety of the resection, but in cases in which the tumor is found to infiltrate functionally important brain, a gross total resection may not be feasible without unacceptable morbidity. An important caveat in attempting to achieve a gross total resection concerns the management of cystic tumors. For lesions with a well-defined mural nodule in which the cyst lining is nonenhancing and gliotic, resection of the wall is unnecessary, and primary attention is focused on removing the nodule. In contrast, for lesions in which the wall is thick and enhancing, removal of this component of the tumor is essential, if feasible, to optimize the chances for disease control.

For highly vascular intraventricular tumors, such as choroidal plexus tumors, it is important to avoid attempts at resecting the tumor until the arterial supply has been coagulated and divided. If the large size of the lesion precludes early control of the pedicle, I generally try to shrink the individual lobules of the tumor progressively using the bipolar cautery until the lesion can be mobilized away from its feeders. These are then coagulated and divided. The tumor can be debulked using bipolar cautery and microscissors, removing the tumor in lobules or groups of lobules (Figs. 11–6C and D). A patty should be placed behind the tumor over the dependent portion of the ventricle to avoid letting blood or tumor fragments slip back out of the field into the cavity of the ventricle, where they may be difficult to retrieve. Often, the venous drainage of the tumor is encountered at the end of the resection, when the tumor has been reduced in size sufficiently to allow its free mobilization within the operative field. This venous pedicle should be sought and then coagulated and divided to avoid bleeding that may result in an inaccessible area if a draining vein is inadvertently avulsed.

Closure

After the tumor has been removed or debulked and preliminary hemostasis has been achieved, several Valsalva's maneuvers are performed to confirm that the field is dry. Minor oozing from the tumor cavity or from areas of unresectable residual disease can often be controlled with gentle pressure from a cottonball soaked in half-strength (1.5% in saline) peroxide. Persistent oozing after this maneuver mandates a close inspection of the operative field for the bleeding source(s). The cavity may then be lined with oxidized cellulose at the surgeon's discretion. Hemostatic material adds an element of security in terms of bleeding control, but it complicates interpretation of postoperative imaging studies and is best avoided. The dura is closed in a watertight fashion; autologous pericranium, allograft fascia lata or dura, or xenograft pericardium make good dural substitutes if primary dural approximation is difficult. If the frontal sinus has been exposed during a low frontal approach, this is covered with a vascularized pericranial graft that is secured to the undersurface of the frontal dura.

As noted earlier, I routinely use dural tacking sutures at the margins of the craniotomy. The bone is then secured using 2-0 absorbable or nonabsorbable sutures; a central 4-0 dural tacking suture is used to minimize the risk of epidural fluid collection. Temporalis muscle and fascia (if incised), galea, and skin are closed in layers with absorbable sutures, and the head is wrapped.

POSTOPERATIVE MANAGEMENT

Patients are generally monitored in the intensive care unit for the first night after surgery. If a significant tumor resection has been achieved, which is generally the goal in pediatric tumors, corticosteroids are tapered over a period of 3 to 5 days. External ventricular drainage, if used, is generally weaned over a period of 3 to 7 days by gradually elevating the drip chamber. Most patients tolerate removal of the ventriculostomy, although some patients with intraventricular tumors, such as choroid plexus papillomas, will exhibit signs of communicating hydrocephalus several days later because of impaired cerebrospinal fluid absorption and will require insertion of a ventriculoperitoneal shunt.

Patients are maintained on anticonvulsants for at least 1 week postoperatively. The use of longer courses of postoperative anticonvulsants in patients without a history of seizures is of uncertain benefit. In patients with a history of preoperative seizures, the duration of postoper-

ative anticonvulsants is largely empirical. I generally discontinue the anticonvulsants several months after surgery if the child has remained seizure-free postoperatively.

A critical issue in the postoperative management concerns the importance of postoperative imaging to determine the extent of resection. For virtually all types of pediatric hemispheric gliomas, the extent of residual disease is a major predictor of outcome. In this context, if the initial operation was undertaken with the goal of achieving a gross total tumor removal and postoperative imaging discloses that a potentially resectable lesion has been incompletely removed, there is at least some rationale for embarking on another attempt at gross total resection several days later, before proceeding with any adjuvant therapy. Similarly, in rare cases in which the tumor resection has intentionally been staged (e.g., the large, highly vascular, bihemispheric PNET shown in Fig. 11–1F), an initial postoperative scan is useful for planning the second-stage tumor removal.

Finally, in children with malignant lesions, further adjuvant therapy will need to be initiated once the child has recovered from surgery and has adequate healing of the cranial incision. Details regarding adjuvant therapy for the different tumor types are discussed in the accompanying textbook.

EDITOR'S COMMENTARY

It is important for the neurosurgeon to know preoperatively the correlation between extent of resection and prognosis for virtually all the tumor types that might potentially be present. The differential diagnosis may be extensive, but that knowledge often alters the extensiveness of resection. As Pollack states, the prognosis of most–but not all–pediatric brain tumors is increased by extensive resections, a correlation that is not true for adults with gliomas. Some pediatric brain tumors are exceedingly vascular and 1–2 blood volumes may be lost as the tumor is being resected. In such cases, it is often appropriate to discontinue the resection after an appreciable portion of the tumor has been resected and remove the residual tumor a week or so later after the child is in more stable condition. Pediatric brain tumors are rare and are ideally operated on by neurosurgeons who do the operations frequently. Recent data indicate that operative morbidity is lower and the likelihood of extensively removing the tumor is higher if the operation is done by a neurosurgeon who does these operations frequently. Operative mortality should be 1% or less and permanent neurological morbidity approximately 10%.

PEARLS

In this author's experience:

- There is a strong association between resection extent and outcome for virtually all types of pediatric hemispheric tumors, including malignant lesions.
- An important consideration in selecting the optimal approach to a supratentorial hemispheric lesion centers around developing a clear plan of the goals of the operation (i.e., biopsy, reduction of mass effect, major cytoreduction, and/or treatment of hydrocephalus), which are influenced by the growth characteristics of the tumor as depicted by CT or, preferably, MR imaging.
- For well-circumscribed hemispheric tumors, a gross total resection should be the operative goal if it can be achieved without inordinate risk.
- A number of adjuncts have become available during the last several years that facilitate radical removal of lesions previously thought to be unresectable or resectable only with substantial morbidity.
- After a gross total or radical subtotal resection, low-grade gliomas can be managed expectantly; adjuvant therapy is reserved for the small percentage of tumors that progress. In contrast, high-grade lesions require intensive multimodality treatment following surgical resection.

SUGGESTED READINGS

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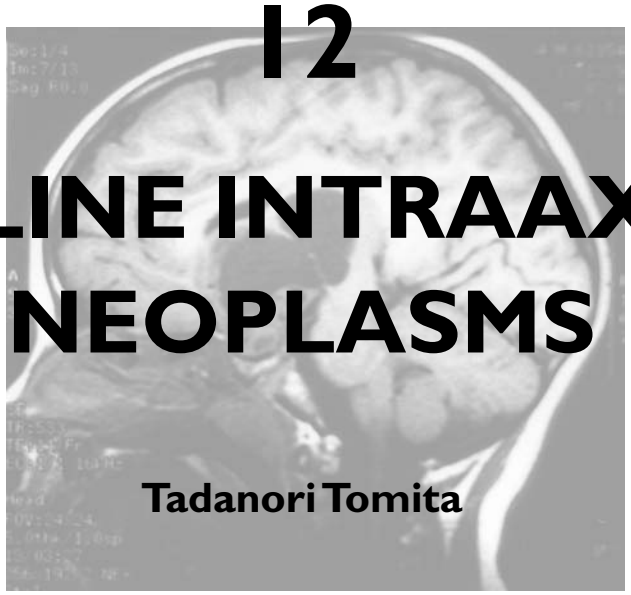
12

MIDLINE INTRAAXIAL NEOPLASMS



Tadanori Tomita

The third ventricle is a common location for pediatric brain tumors. In this location, various types of tumors can occur, and the histological distribution differs between the anterior and posterior portions of the third ventricle. Chiasmal–hypothalamic astrocytomas and craniopharyngiomas are the main tumors in the anterior third ventricle of children, whereas germ cell tumors and pineal parenchymal tumors dominate in the posterior third ventricle of children. Differential diagnosis of tumor histology and advanced neuroimaging studies are important in selecting an appropriate treatment plan. Surgical resection of tumors located in the midline is often challenging; thus, selecting an appropriate surgical approach and technique is critical. This chapter focuses on craniopharyngioma and pineal region tumors.

CRANIOPHARYNGIOMA

Surgical Indications and Preoperative Evaluation

Craniopharyngiomas originate in or above the sella turcica, and the cystic components extend in various directions: the third ventricle superiorly, the posterior fossa posteriorly, under the frontal lobe anteriorly, and into the middle cranial fossa laterally. In childhood craniopharyngiomas, calcifications and cyst formations are almost invariably present. Children often present with visual disturbances, endocrine deficits, and hydrocephalus. Hydrocephalus is the result of the suprasellar cyst, which often penetrates through the floor of the third ventricle and fills the third ventricle. Because the cyst wall is composed of neoplastic tissue, residuals of not only the solid portion but the cyst wall as well will guarantee recurrence if tumor resection is incomplete; however, as a result of the proximity of the optic pathway, hypothalamopituitary axis, and the circle of Willis, total resection is difficult and often is complicated by postoperative neurologic and endocrine deficits. Therefore, some prefer limited resection followed by radiation therapy (RT); however, I recommend at least an attempt at complete resection within a safe margin because of the uncertain responsiveness of craniopharyngiomas to RT and concerns of latent radiation toxicity in children.

Both computed tomography (CT) and magnetic resonance imaging (MRI) are routinely obtained preoperatively. For the disclosure of calcification, CT is superior to MRI; however, detailed information in regard to multiplicity of cysts and their extension are better appreciated by multiplanar MRI images. Multiple noncommunicating cysts are shown in a different intensity on MRI. Angiography is seldom necessary. Noninvasive magnetic resonance angiography provides sufficient information in regard to the relationship with the arterial system of the circle of Willis.

The location of the solid tumor, the extension of any cyst and its relationship with carotid arteries, anterior cerebral arteries, and the basilar artery should be noted on neuroimaging. Careful review of the MRI scan may disclose the position of the optic nerve. On sagittal MR images, the chiasm and the anterior communicating artery are elevated in the case of a postfixed chiasm (Fig. 12–1), whereas they are located just above the tuberculum sella in a case of prefixed chiasm (Fig. 12–2). As

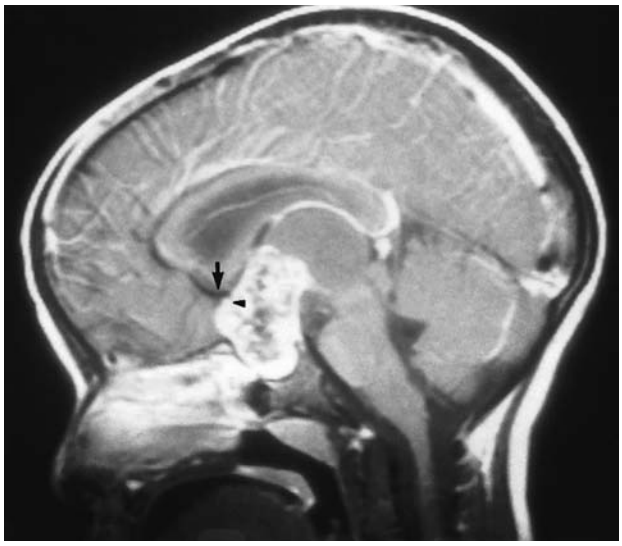


FIGURE 12–1. Midsagittal MRI showing a partially calcified craniopharyngioma with a large cyst occupying the third ventricle. Note that the anterior cerebral artery *(arrow)* and optic chiasm *(arrowhead)* are elevated (postfixed chiasm).

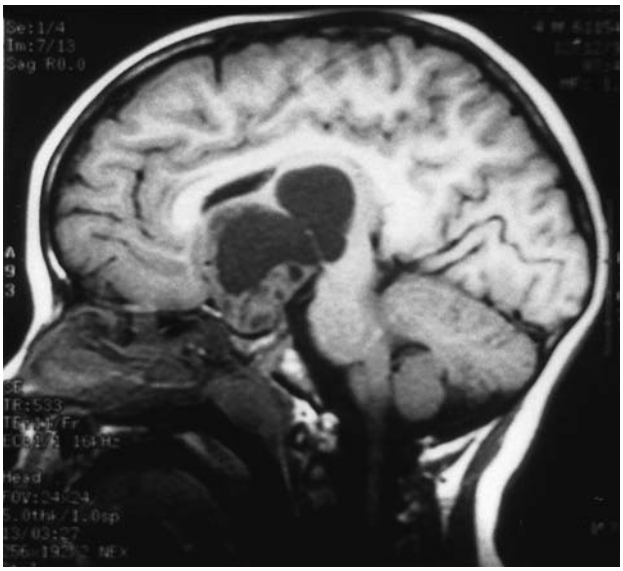


FIGURE 12–3. Midsagittal MRI showing a massive craniopharyngioma extending into the third ventricle from the sellar region through the disrupted floor of the third ventricle.

stated, the extension or presence of the cyst in the third ventricle tends to cause hydrocephalus. Craniopharyngiomas of this type already have broken through the floor of the third-ventricle, directly exposing their capsule in the third ventricle cavity (Fig. 12–3). On the other hand, some children with craniopharyngiomas may have an intact floor of the third ventricle (Fig. 12–4). It is important to use preoperative neuroimaging to distinguish these two groups; both a interhemispheric transcallosal approach and a lamina terminalis approach would traumatize the floor of the third ventricle in the later group.

Ophthalmologic examinations should be conducted carefully to identify visual deficits and should include ophthalmoscopic examination to disclose the presence of optic atrophy or papilledema. Pupillary responses to light, particularly a swinging flashlight test, discloses afferent deficits, if any are present. Also, formal tests for visual acuity and field need to be done preoperatively.

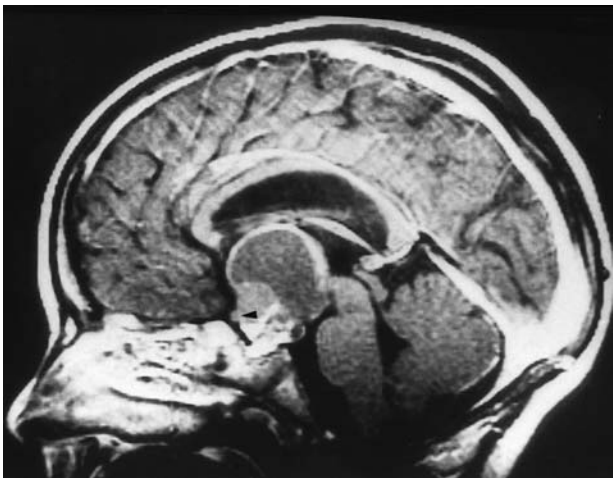


FIGURE 12–2. Midsagittal MRI showing a cystic craniopharyngioma in the suprasellar and third-ventricle location. Note that the optic chiasm *(arrowhead)* is depressed (prefixed chiasm).

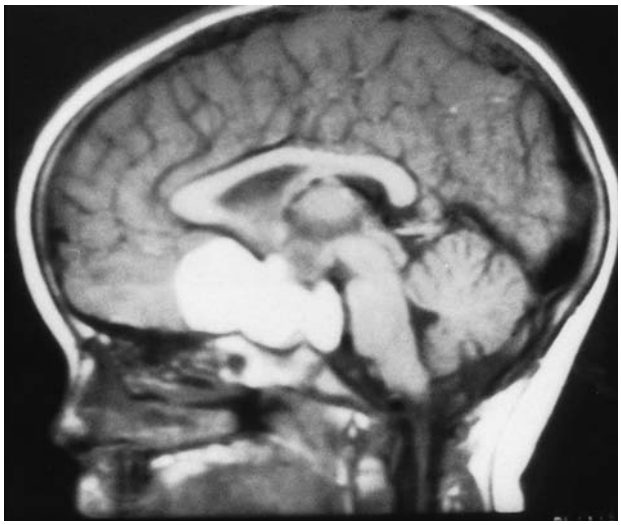


FIGURE 12–4. Midsagittal MRI showing a large cystic craniopharyngioma lifting the third-ventricle floor with an extension to the subfrontal region and the posterior fossa.

Endocrine investigation also is done preoperatively. Important endocrine studies include serum level of thyroxine (T_4) and thyroid-stimulating hormone, serum morning cortisol level, serum electrolytes, urine specific gravity (particularly the first morning urine) and prolactin level. Baseline insulin-like growth factor I (IGF-I) may be helpful. For pubescent children, preoperative luteinizing hormone and follicle-stimulating hormone together with testosterone for boys and estradiol for girls may be helpful. They are of interest from an academic point of view; however, they are not mandatory from a surgical and anesthesia point of view.

Preoperative Management

Once blood samples are drawn for endocrine study, corticosteroid hormone therapy is initiated. The usual stress dose of hydrocortisone is 30 mg/m^2 per day, and the maintenance dose is 5 to 10 mg/m^2 per day; however, most children with craniopharyngioma present with relatively acute symptoms resulting from optic nerve compression or hydrocephalus. In these circumstances, they usually receive a high dose of dexamethasone, 2 to 10 mg every 6 hours, depending on patient size and the severity of the presenting symptoms. If hypothyroidism is noted, thyroid hormone needs to be replaced.

Operative Planning

First, the goal of surgery is determined: biopsy alone, cyst drainage alone, or radical tumor resection. Biopsy or cyst drainage can be attained by stereotaxic or ventriculoscopic procedures. If major cytoreduction and decompression are the goals of surgery, however, the optimal approach to intrasellar, suprasellar, and third ventricular craniopharyngiomas must be selected. Intrasellar craniopharyngiomas (Fig. 12–5), although rare in my experience (only 3 of 40 childhood craniopharyngiomas), are best approached transsphenoidally. Most craniopharyngiomas are accessible through a pterional–subfrontal approach, but in special cases, such as primarily third-ventricular craniopharyngioma, the interhemispheric transcallosal approach is used.

An important consideration at resection of the craniopharyngioma is to attain a relaxed brain and a better angle to the sellar and suprasellar region. Hydrocephalus associated with craniopharyngioma may be favorable for the operating surgeon because a relaxed brain is achieved by intraoperative decompression of the ventricle and provides effortless brain retraction during skull-base manipulation. The presence of hydrocephalus makes it easier to manipulate the third ventricular mass through an enlarged foramen of Monro when the transventricular route is used.

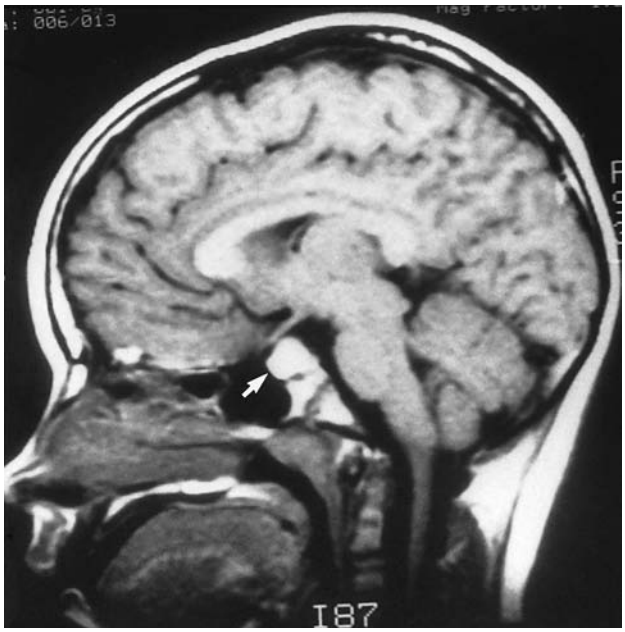


FIGURE 12–5. Midsagittal MRI showing an intrasellar craniopharyngioma *(arrow)*. A transsphenoidal approach is suitable for tumor resection.

Intraoperative Technique: Ventriculostomy

A burr hole is made just anterior to the coronal suture on the midpupillary line of the right side. The dura is coagulated and incised. Ventricular puncture is done by aiming toward the nasion in the coronal plane and the tragus of the ear in the sagittal plane. In older children, the distance from the outer table of the skull to the foramen of Monro is 6.5 cm. When the tumor cyst is occupying the third ventricle, ventriculoscopy is used to inspect the ventricular space. The cyst walls often show yellow–green discoloration. If feasible, I would puncture the cyst to drain the cystic content through a ventricular catheter under ventriculoscopic control.

Positioning

In most cases of craniopharyngioma, I use a combination of pterional and subfrontal approaches from the right side. For this approach, the patient is orally intubated and placed in a supine position. A three-pin head fixation device is used for older children, but a horseshoe headrest is used for infants and younger children with a

thin skull. The head is rotated toward the left side about 15 degrees, and the neck is extended slightly backward.

Initial Exposure

The scalp incision is made in a bicoronal fashion anteriorly and superiorly to the tragus of the right side and extending along the coronal suture to the superior temporal line of the left side. The skin flap is turned subperiosteally. The right temporal muscle is sectioned vertically up to the zygomatic process by using cutting cautery. Anteriorly, the superior temporal line is cut, and the temporal muscle is turned with the scalp flap. At the orbital ridge, the superior orbital foramen and nerve are identified. The caudal portion of the supraorbital foramen is chiseled away, and the superior orbital nerve is turned forward with the scalp flap without sacrificing it. The pericranium is sharply dissected at the superior orbital ridge, and the periorbita is separated from the roof of the orbit. The scalp flap needs to be turned forward sufficiently to expose the glabella.

Craniotomy

For a fronto-temporo-orbital craniotomy, four burr holes are made. One is made directly on the pterion, and is enlarged anteriorly and posteriorly to expose the frontal and temporal dura. Then the pterion is removed caudally as much as possible. Another burr hole is made at the keyhole (frontozygomatic junction), and both the frontal base dura and periorbita are exposed. The third burr hole is made on the glabella, and the fourth burr hole is made parasagittally in the posterior frontal bone of the right side.

The fronto-temporo-orbital craniotomy is performed as follows: Craniotomy is made using a craniotomy from the temporal burr behind the pterion to the posterior frontal burr hole and then to the burr hole at the glabella. The burr holes at the pterion and the keyhole are connected (Fig. 12–6). Subsequently, an orbitotomy is carried out between the key hole and glabella burr holes. A Gigli guide is placed through these burr holes in the anterior cranial fossa, allowing placement of the Gigli saw. By using the Gigli saw on the orbital roof from the anterior cranial fossa to the orbit, the orbitotomy is completed (Fig. 12–7). During that time, the orbital contents need to be protected by a spatula, and the orbitotomy is made as posterior as possible. A free bone flap, which includes the frontal, temporal, and orbital rims and the anterior orbital roof, is attained (Fig. 12–8). Once the craniotomy is completed, the sphenoid ridge is removed epidurally from the pterion as medially as possible.

Dural Opening and Tumor Exposure

The dura is incised along the frontal base extending over the Sylvian fissure to the temporal region. Once the dura is open, the posterior inferior frontal lobe is elevated along

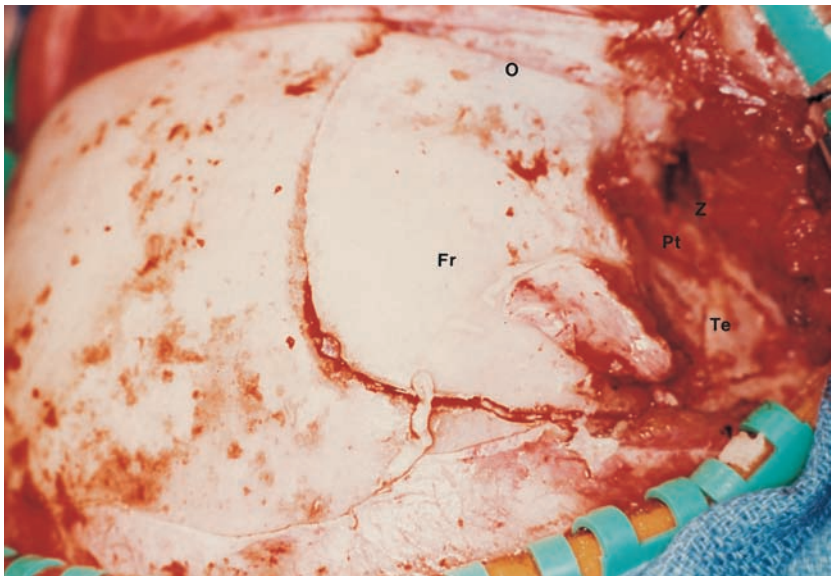


FIGURE 12–6. Right fronto-temporo-orbital craniotomy is shown. O, orbital ridge; Pt, pterion; Z, zygomatic arch; Fr, frontal bone; Te, temporal bone.

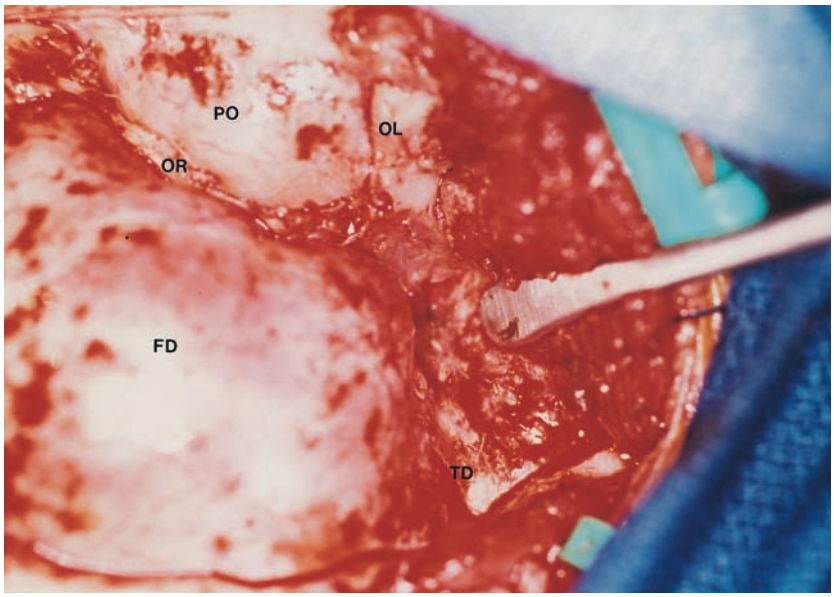


FIGURE 12–7. After fronto-temporo-orbital craniotomy is performed, frontal (*FD*) and temporal dura (*TD*) is exposed. The periorbita (*PO*) is exposed anteriorly after the orbit is unroofed. OR, posterior edge of the orbital roof; OL, lateral wall of the orbit.

the sphenoid wing. Further medially, the anterior clinoid process and, subsequently, the planum sphenoidale are identified. Posteriorly to the planum sphenoidale is the arachnoid of the suprasellar cistern. Once the cistern is opened and the cerebrospinal fluid (CSF) is drained, the relaxed brain allows further elevation of the frontal lobe with gentle retraction. Then the medial sylvian fissure is opened. Once the suprasellar and sylvian cisterns are open, the right and left optic nerves, the optic chiasm, and the right internal carotid artery are in the surgical view.

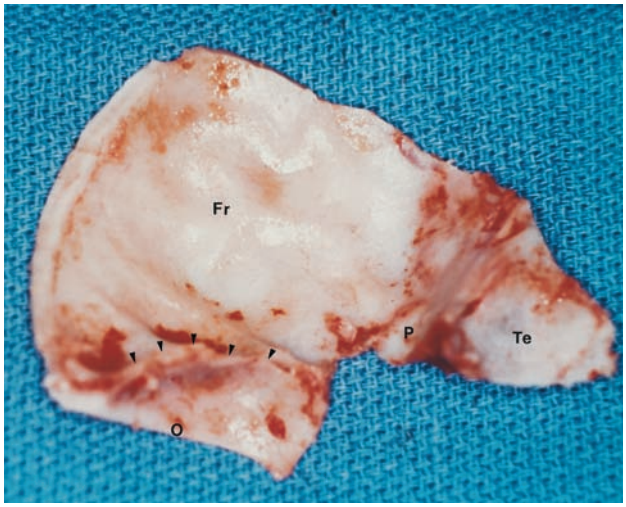


FIGURE 12–8. Inner surface of a free bone flap of the fronto-temporo-orbital craniotomy is shown. It includes the orbital roof (*arrowheads* indicate the posterior edge of the orbital roof). Fr, frontal bone; Te, temporal bone; P, pterion; O, superior orbital ridge.

The carotid artery is present lateral and inferior to the right optic nerve. Further elevation of the gyrus rectus of the right side allows for identification of the lamina terminalis and the A1 segment of the right anterior cerebral artery and anterior communicating artery posteriorly.

Neural and vascular structures are inspected under microscopic magnification once the chiasmal cistern is exposed. The nature of the optic chiasm determines whether it is fixed posteriorly *(postfixed)* or anteriorly *(prefixed)*. In the case of postfixed chiasm, the craniopharyngioma capsule is present in the prechiasmatic space (Fig. 12–9). This enlarged prechiasmal space becomes the primary access to the tumor. Also, the space between the right optic nerve and the carotid artery (opticocarotid space) is an important access site for the tumor resection (Fig. 12–10).

In a case of prefixed chiasm, the prechiasmal space is virtually absent; therefore, the lamina terminalis is opened, and the tumor is removed through it. The lamina terminalis is somewhat pale, bulgy, and less vascular. One should take care not to traumatize the anterior communicating artery, which is located just above the lamina terminalis. Any intrasellar mass can be removed subsequently by reducing the retrochiasmal mass, which often creates an enlarged prechiasmal space. The widened opticocarotid space of the ipsilateral side provides an important access site to the tumor under the chiasm (Fig. 12–11). Tumor under the chiasm, or deep in the anterior sella turcica, is often difficult to remove when the prechiasmatic space is absent. The right anterior clinoid process is removed through an epidural approach, which, once removed, gives further access to these regions.

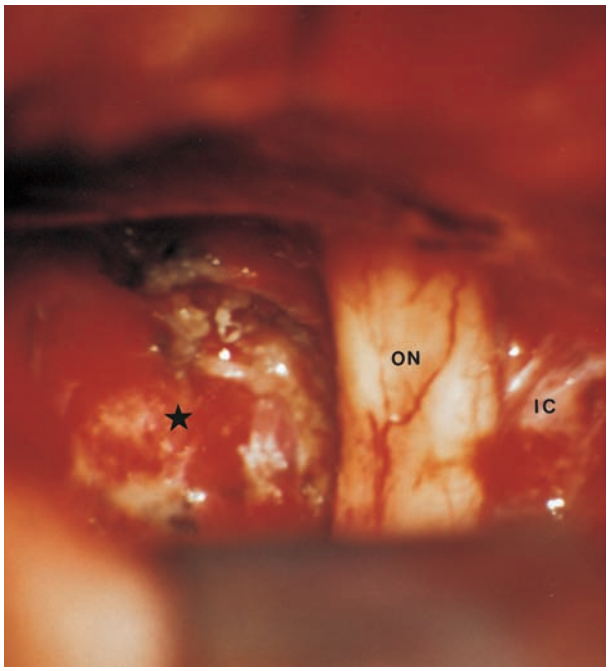


FIGURE 12–9. Craniopharyngioma capsule (*asterisk*) is exposed through the prechiasmal space. IC, the internal carotid artery; ON, optic nerve.

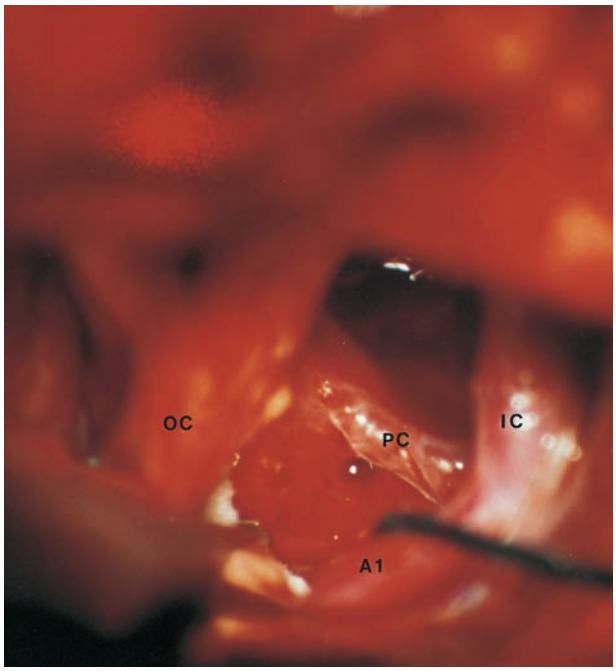


FIGURE 12–11. The optic nerve and chiasm (*OC*) is displaced, and the right opticocarotid space is widened. Note the large access to the region under the optic chiasm. The posterior communicating artery (*PC*) is visualized, together with the internal carotid artery (*IC*) and anterior cerebral artery (*A1*).

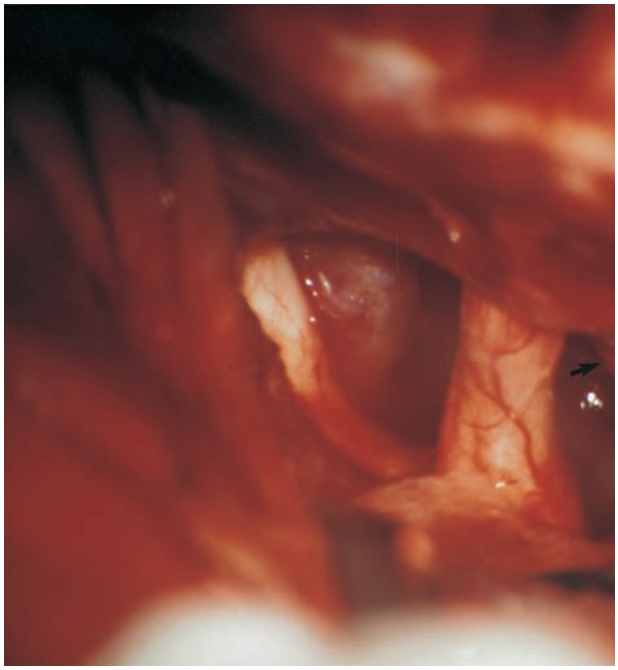


FIGURE 12–10. Following resection of the prechiasmal craniopharyngioma, both optic nerves and the left carotid artery are shown. The opticocarotid space is shown next to the right optic nerve (*arrow*).

Tumor Resection

Once the craniopharyngioma capsule is opened, internal decompression is achieved, and the cystic contents are aspirated. The solid portion, which consists of brittle, calcified tissue, is removed in a piecemeal fashion. Some areas are densely calcified, and piecemeal removal may be difficult. Forcible removal of large, calcified tissue through the prechiasmal space or retrochiasmal space will traumatize the highly sensitive optic structures. I use the carbon dioxide laser to dehydrate a highly calcified tumor and then break it into small pieces for removal. It is important to debulk the intracapsular content as much as possible. The cystic contents of a third ventricular cyst can be drained once the solid component in the suprasellar area is reduced.

Once sufficient internal decompression is achieved, the tumor capsule is gently and meticulously separated from the optic and vascular structures. In a case of postfixed chiasm, the capsule is separated from the ventral surface of the chiasm. The right optic nerve is also separated away from the tumor at its ventral surface. The internal carotid arteries are always separable from the

craniopharyngioma at primary resection. The tumor capsule is gently separated from the carotid artery through the interoptic space and optic carotid space. Laterally and inferiorly to the carotid artery is the right third nerve, which should be identified. A further opening of the sylvian fissure facilitates tumor separation from the third nerve. The left optic nerve moves into the surgical field once the internal decompression is achieved and is carefully separated. The left internal carotid artery is identified by gentle retraction of the tumor to the right side and is separated from the tumor capsule. The tumor capsule is coagulated by bipolar cautery, which not only reduces the size of the capsule and tumor but also makes the capsule firmer. The tumor capsule is gently retracted continually while separating it from the surrounding structures. The pituitary stalk is usually present behind the solid portion and may be displaced in either direction. The appearance of the pituitary stalk has a distinctive reddish color and has vertically oriented blood vessels along its course; however, it may be totally replaced by the tumor. Although preservation of the pituitary stalk is ideal, in most cases this is difficult because of its involvement by the tumor. If the pituitary stalk needs to be sectioned, this should be done in the most distal portion.

Although the ventral chiasmal surface is easily separated from the tumor, the tuber cinereum is always adherent to or replaced by the tumor. The trajectory of the microscope needs to be changed to inspect the ventral surface of the chiasm. The fronto-temporo-orbital craniotomy provides a better angle for that purpose. Continuous gentle retraction of the capsule brings the tuber cinereum into the surgeon's view. Under direct vision, the tumor capsule is separated from the tuber cinereum. It is important not to break the continuity of the capsule to attain complete removal of the craniopharyngioma. If the capsule breaks during the retraction, that portion of the capsule will migrate back into the third ventricle and become difficult to resect. Craniopharyngiomas, which extend posteriorly and compress the brainstem, can be removed by gentle retraction because the basilar artery and the brainstem are separated from the tumor capsule by the Lillequist membrane.

In a case of prefixed chiasm, the primary route for the resection is though the lamina terminalis (Fig. 12–12). One should confirm that the tumor is present directly in the third ventricle following internal decompression. The tumor resection is carried out as already described. The portions of the tumor from the third ventricle and the retrochiasmal region are removed with relative ease, but resecting the tumor under the chiasm and

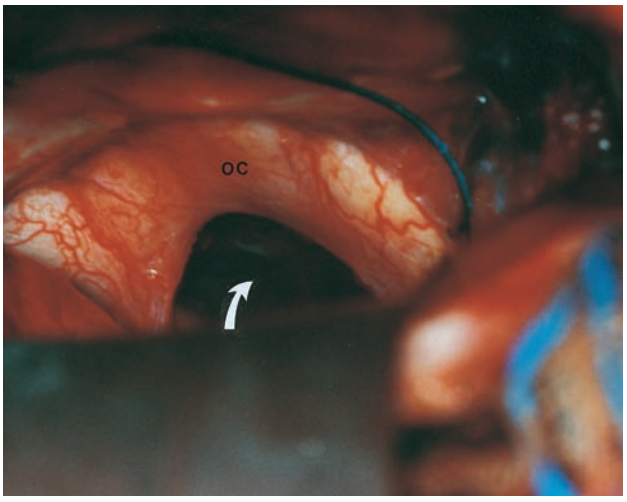


FIGURE 12–12. In a case of prefixed chiasm (*OC*), the open lamina terminalis (*arrow*) provides entry to the third ventricle.

the sella turcica is often difficult. Following removal of craniopharyngioma from the third ventricle, the prechiasmal space may be widened; however, often this space is not enough for tumor resection from these areas. The opticocarotid space needs to be used in such a case. If this does not suffice, I would resect the anterior clinoid process extradurally and the lateral portion of the planum sphenoidale of the right side by means of a high-speed drill, which exposes the proximal portion of the optic nerve, thus creating the prechiasmal space intradurally.

Alternate Approach

An interhemispheric transcallosal approach is suitable for craniopharyngiomas in the third ventricle, which invariably present with hydrocephalus. I use this approach as a combined approach with the pterional–subfrontal approach or as a primary method for resection of a primarily third-ventricular craniopharyngioma.

The patient is placed in a supine position with the head slightly turned toward the left side. The scalp incision is made behind the coronal suture from the right temporal to the left superior temporal line sufficient to expose the posterior frontal bone to within approximately 6 cm of the bregma. The interhemispheric entry is primarily anterior to the bregma, dependent on the location of the bridging veins. A 7 by 5 cm craniotomy is made 1 cm posteriorly from the bregma and 6 cm anteriorly from the bregma, extending. 1 cm to the left and 4 cm to the right of the midsagittal suture. Once the craniotomy is done and the superior sagittal sinus is verified,

the dura is incised in a medially based, C-shaped fashion. Several bridging veins anterior to the coronal suture can be coagulated and sectioned with impunity; however, one should avoid sacrificing major veins coming from the posterior frontal lobe. Dissection of the arachnoid around the vein allows some relief to the vein while turning the dural flap to the contralateral side and entering the interhemispheric fissure.

The frontal lobe is gently retracted laterally, and the interhemispheric fissure is enlarged. At this time, the drainage of the ventricular CSF contributes to brain relaxation. I generally avoid the use of intravenous mannitol for brain relaxation at the resection of craniopharyngiomas or other suprasellar tumors because this agent confuses the picture of increased urinary output associated with diabetes insipidus during and following the surgery.

At the depth of the interhemispheric fissure, the corpus callosum is identified. Bilateral pericallosal arteries run along the course of the corpus callosum. The arachnoid membrane of the pericallosal cistern is opened, and these arteries are separated. The body of the corpus callosum exposed between the pericallosal arteries is coagulated and sectioned longitudinally in a 3-cm length. At the depth of the corpus callosum, the ependyma is opened and the lateral ventricle entered. In some cases, the cavum septum pellucidum may be well developed, and it is confusing whether the cavity is the cavum septum pellucidum or the right or left lateral ventricle. Identification of the choroid plexus and the foramen of Monro confirms the position.

Craniopharyngioma with a greenish capsule often protrudes through the enlarged foramen of Monro. The cyst capsule is coagulated and incised and its contents aspirated, causing the cyst to collapse. Subsequently, the surrounding third ventricle walls are identified. The roof of the third ventricle is always free from the tumor. The tumor capsule is shrunk with bipolar cautery. The tumor capsule and its contents are removed in pieces and microsurgically resected. Extreme care not to traumatize the fornix during the manipulation in the third ventricle is essential, particularly when resecting the tumor from the anterior wall of the third ventricle and the chiasmal region. At that time, the fornix is stretched because the anterior foramen of Monro needs to be displaced anteriorly. The upper portion of the bilateral wall (thalamus) of the third ventricle is free from tumor invasion; however, at the lower portion, craniopharyngioma is intimately adherent to the bilateral hypothalamus and is directly continuous to the suprasellar mass. The hypothalamic wall in that area is covered by gliotic tissue. The tumor capsule is sharply dissected from these gliotic structures. On the other hand, the mammillary bodies and the aqueduct of Sylvius usually are not involved by the tumor. Extremely anteroinferiorly in the third ventricle, the optic chiasm is visualized and separated from the tumor. By this approach, the portion of the tumor beneath and anterior to the chiasm is not visible and further tumor resection is not possible without jeopardizing the visual pathway. Also, the carotid artery is not well visualized. Therefore, it is safer to resect the remaining tumor through the subfrontal–pterional approach described previously.

When the craniopharyngioma is localized in the enlarged sella turcica, a standard transsphenoidal approach is best. In children, the sphenoid sinus is well pneumotized after 8 years, but the nasal structure is not well developed. Therefore, surgical exposure has some limitation with this approach, and it would be wise to have a pediatric ear, nose, and throat (ENT) specialist available to provide access by this route.

Postoperative Management

Patients are observed in the continuous care unit. Physical and neurologic status are observed and recorded. Hourly intake and output measurements, urine specific gravity, and 6-hour serum sodium and osmolality (every 6 hours) are monitored. If diabetes insipidus is evident, vasopressin is started.

I prefer to obtain immediate postoperative CT, which provides the information regarding residual calcified lesion, presence or absence of intracranial hemorrhage, and the status of hydrocephalus. If an external ventricular drain is placed during the tumor resection, blood-tinged CSF is drained at the drainage pressure of 10 cm H_2O for several days; subsequently, it is used strictly for intracranial pressure (ICP) monitoring, which is correlated with patient's symptoms and interval changes in ventricular size by follow-up CT scans. In most cases, the external ventricular drain is removed at the bedside in 5 to 7 days.

PINEAL REGION TUMORS

Surgical Indications and Preoperative Evaluation

In the pineal region, tumors of variable histology occur: pineal parenchymal tumors, germ cell tumors, and glial tumors. Pineal parenchymal tumors include pineoblastomas and pineocytomas. Germ cell tumors include teratomas, germinomas, embryonal cell carcinomas, choriocarcinomas, endodermal sinus tumors (yolk-sac tumor) and

mixed tumors. Glial tumors originate from the surrounding neural tissue, such as the quadrigeminal plate, midbrain, and thalamus. Ependymoma can occur from the ependymal lining of the third ventricle. Also, the third ventricle choroid plexus can give rise to choroid plexus papilloma and meningioma.

Germ cell tumors have a strong preponderance for males. Teratomas are benign and require total resection. There may be some malignant components even in benign teratoma; thus, the entire tumor sample needs to be examined by the pathologist. Neuroimaging studies and specific tumor markers such as alpha-fetoprotein and beta human chorionic gonadotropin (HCG) are important for preoperative differential diagnosis. Certain malignant germ cell tumors respond to chemotherapy, which should be considered preoperatively. Germinomas respond well to RT or chemotherapy, which provides excellent cure rates. On the other hand, malignant nongerminomatous germ cell tumors need extensive chemotherapy. Often following chemotherapy, the tumor size decreases and the tumor vascularity diminishes, making resection of the tumor safer and more effective at a second-look operation. Concomitant anterior and posterior third-ventricle solid lesions, in my experience, guarantee their histologic nature to be malignant germ cell tumors. Pineoblastomas are common in younger childhood, and the prognosis is often poor; however, maximum cytoreduction is required for better tumor control.

Preoperative Management

Although some recommend biopsy either sterotaxically or by means of ventriculoscopy, others raise concern about hemorrhagic complications caused by these biopsy procedures. Also, biopsy samples are not large enough to represent entire pathological pictures.

Pineal-region tumors are almost invariably accompanied by hydrocephalus. A ventriculoperitoneal shunt, a common method to treat hydrocephalus associated with pineal region tumors, may cause shunt dependency and potential intraperitoneal dissemination of malignant tumors. Third ventriculostomy is a preferred procedure for controlling hydrocephalus associated with a pineal-region tumor. At the time of access to the lateral ventricle, CSF can be collected for cytologic studies and identification of tumor markers. Also, it may be possible to perform tissue biopsy though a ventriculoscope. When the patient needs surgical resection of the pineal region tumor, establishment of a ventriculostomy and subsequent external ventricular drainage helps during intraoperative and postoperative care.

Operative Planning

Preoperatively, neuroimaging needs to be reviewed carefully as to the size, location, and extension of the tumor to choose the most advantageous surgical approach (Figs. 12–13, 12–14, 12–15, and 12–16). There are multiple approaches to the pineal region, the most popular being the occipital transtentorial approach and the infratentorial supracerebellar approach. I prefer the occipital transtentorial approach because it provides a wider surgical field and allows more vertical angles from the dome to the bottom of the tumor. The vein of Galen is present usually over the dome of the tumor; thus, it is secured under direct vision during manipulation of the tumor. The portion of the tumor that extends laterally beyond the edge of the tentorial opening or to the lateral ventricle is difficult to visualize through the infratentorial approach. Tumors located in the caudal quadrigeminal cistern or precentral cistern are difficult to access when using an infratentorial approach unless the culmen of the vermis is depressed to a significant degree. Also, when the patient is placed in a prone position, as in the occipital transtentrial approach, it is less fatiguing to the surgeon because the surgeon is looking down onto the surgical field instead of looking up, as is the case when the patient is in a sitting position for an infratentorial approach.

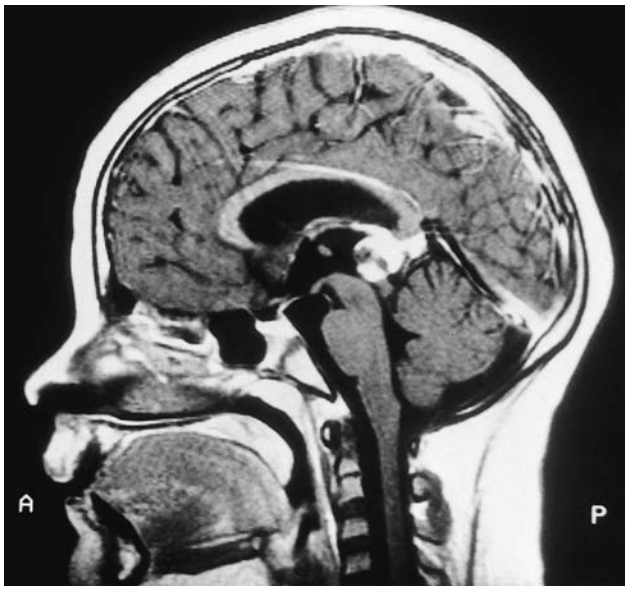


FIGURE 12–13. Midsagittal MRI after contrast infusion showing a pineoblastoma.

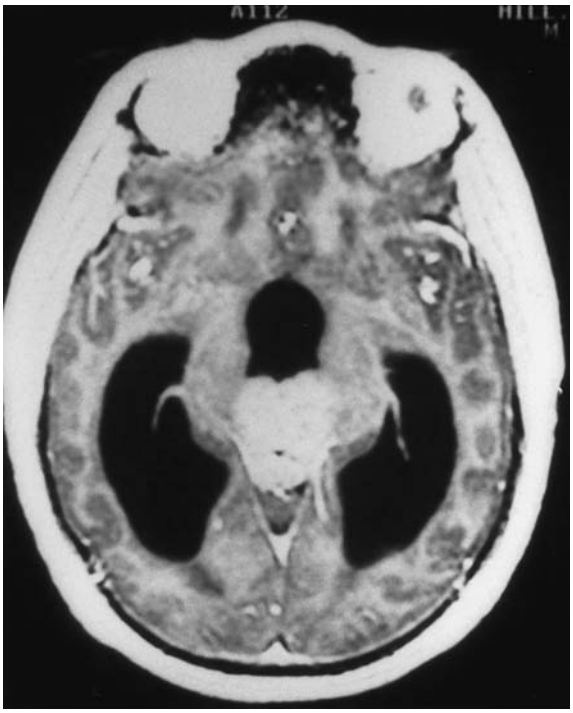


FIGURE 12–14. Axial MRI after contrast infusion showing a large germinoma.

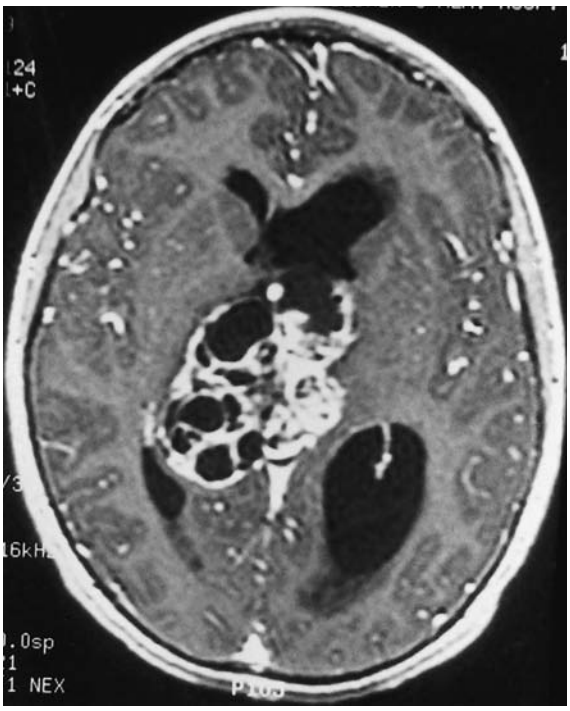


FIGURE 12–16. Axial MRI after contrast infusion (same case as Fig. 12–15) showing a heterogeneously enhancing teratoma. Note that the tumor extends further laterally into the lateral ventricle.

Intraoperative Techniques

Ventriculostomy is performed in the same fashion as that described previously. I place a burr hole in the posterior frontal location with the patient in a supine position.

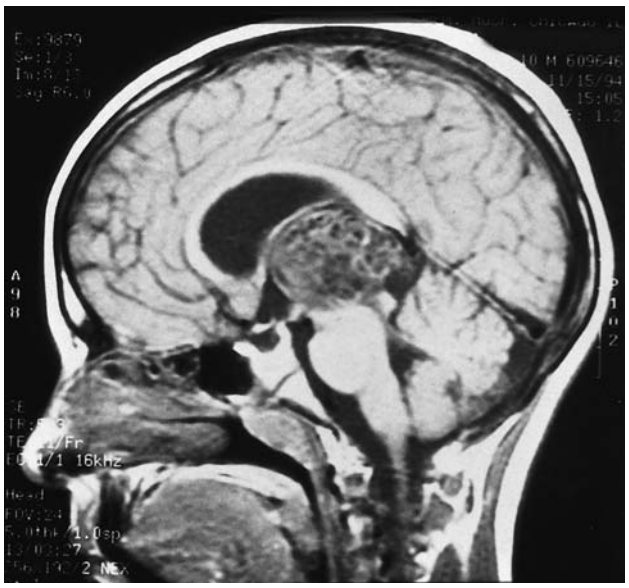


FIGURE 12–15. Midsagittal MRI showing a large teratoma in the pineal region extending into the third ventricle.

This frontal ventriculostomy is done to inspect the third ventricle if indicated and also for the purpose of performing third ventriculostomy if indicated.

For an occipital transtentorial approach, the patient is in a prone position. A three-pin fixation device in children older than 5 years secures the head, with the head turned approximately 15 to 30 degrees toward the contralateral side of the occipital craniotomy (Fig. 12–17). Turning the head provides the natural gravitational fall of the occipital lobe without forcibly retracting it. In young children, however, the prone position is obtained with the head in a neutral position on a well-padded horseshoe headrest. Although the gravitational fall of the occipital lobe is not gained in these cases, intraoperative control of the hydrocephalus by means of ventriculostomy will minimize the occipital lobe retraction.

Initial Exposure

A craniotomy usually is performed on the right side (i.e., the nondominant side); however, when tumor extends to the left lateral ventricle, however, the craniotomy is made on the left side. A parasagittal incision on the left side is made 1 cm laterally to the sagittal plane from the inion to approximately 5 cm rostral to the lambda; then it is turned

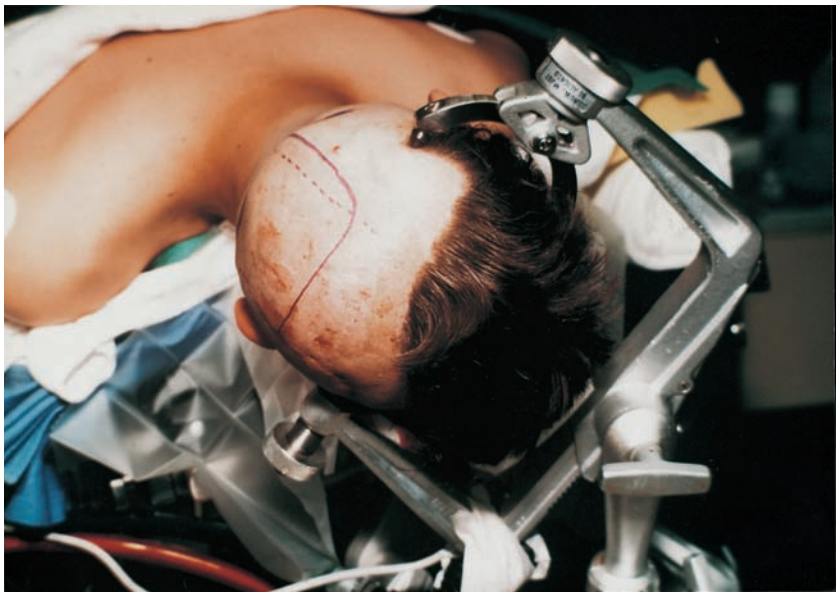


FIGURE 12–17. A position for an occipital transtentorial craniotomy is shown. The patient is placed in the prone position. The head is turned 30 degrees contralaterally and secured with a three-pin fixation device. The dotted line indicates the midsagittal plane; the solid line indicates the placement of skin incision.

toward the right side laterally toward the posterior parietal region to a length of about 10 cm. When the scalp flap is turned subperiosteally, the occipital and parietal bones are exposed. About 5 cm of the posterior sagittal suture, the lambda, and lambdoid suture of the right side is in the surgical field. Although the size of the craniotomy is decided according to the appearance of MRI findings, I usually set the lambda in the center of the sagittal axis of the craniotomy, which is usually about 10 cm long.

Craniotomy

Three burr holes are made parasagittally crossing the midline: one 5 cm from the lambda rostrally, one at the lambda, and another 5 cm caudally just above the torcular Herophili. At that time, the superior sagittal sinus should be preserved carefully. The fourth burr hole is made on the lambdoid suture, approximately 5 cm from the midline. The craniotomy is performed using a Midas Rex craniotome, and a 6 × 10-cm free bone flap is obtained. The superior sagittal sinus is located in the medial portion of the bone opening; however, the torcular Herophili and the lateral sinus are not in the surgical view.

Dural Opening and Tumor Exposure

The dural opening is done in a C-shape, based medially next to the superior sagittal sinus (Fig. 12–18). Bridging veins usually are not present in the occipital location. The most posterior vein is present in the posterior parietal region. At the rostral and caudal corners, a 1-cm dural incision is made both anteriorly and posteriorly and parallel to the sagittal sinus to avoid the incision

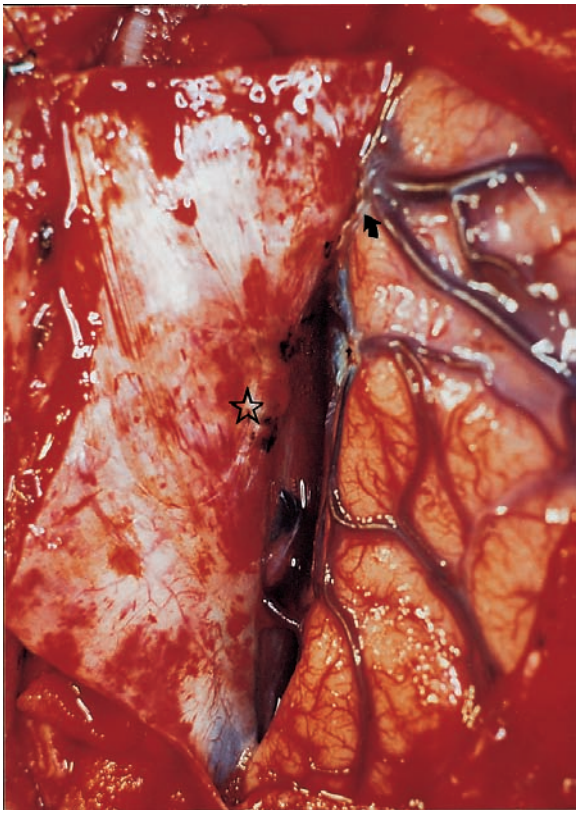


FIGURE 12–18. The dura is turned medially over the superior sagittal sinus. The most caudal bridging veins are noted in the upper corner (*arrow*). The position of the lambda is at the midportion of the dural opening (*asterisk*).

spreading into the sinus during turning of the dural flap medially. At that time, the most rostral bridging vein needs special care to avoid excess tension. The arachnoid is separated from the bridging vein, which allows more retraction of the parietooccipital lobe during the interhemispheric approach.

Arachnoid granulations present between the superior sagittal sinus and the arachnoid membrane at the interhemispheric fissure should be coagulated and sectioned. Gradually, the parietooccipital lobe is retracted laterally. If the brain is tight, CSF is drained from the external ventricular drain. When the ventricle is small (e.g., in the presence of a shunt), intravenous mannitol is administered (0.25 to 0.5 g/kg of body weight) to relax the brain. Entering down the interhemispheric fissure along the falx and reaching its depth, the corpus callosum can be identified anteriorly and the tentorium posteriorly. The surgical microscope is essential for magnification and illumination. At the junction between the falx and the tentorium is the straight sinus. The splenium of the corpus callosum distinguishes itself as a whitish, smooth avascular structure (Fig. 12–19). The vein of Galen is present behind the splenium of the corpus callosum.

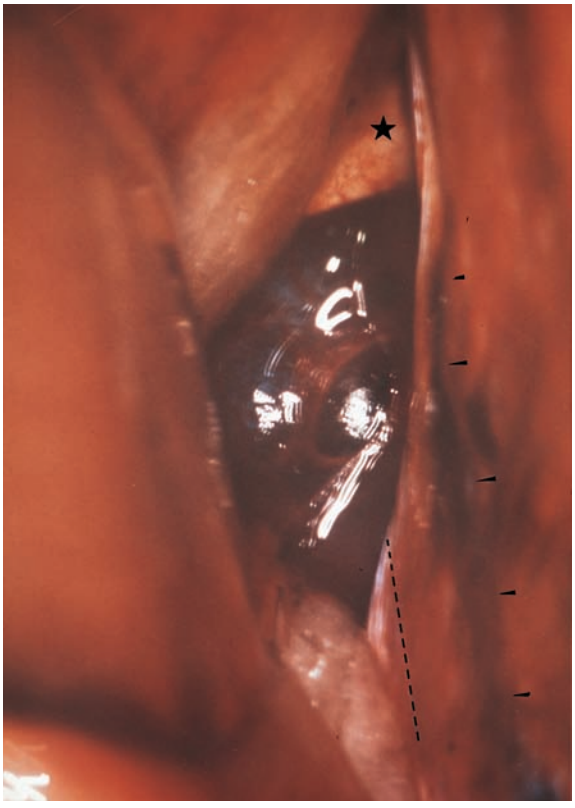


FIGURE 12–19. The corpus callosum is shown in the upper corner of the left-side interhemispheric approach (*asterisk*). A large malignant germ cell tumor is exposed in the pineal region. The dotted line indicates the site for tentorial opening which is placed about 1cm lateral to the straight sinus (*arrowheads*).

The longer the interhemispheric fissure exposure, the greater the angle of trajectory. The microscope trajectory is shifted posteroinferiorly, and the cloudy, thick arachnoid membrane of the quadrigeminal cistern is observed. Once this arachnoid membrane is opened, the tumor capsule protruding from the quadrigeminal cistern is identified. To expose the caudal quadrigeminal cistern, a tentorial section is made, providing entry into the posterior fossa. A 2.5-cm tentorial section is done approximately 1 cm laterally to the straight sinus on the right side, parallel to the sinus. The tentorium may be vascular and thick and is first cut with a no. 11 blade knife and coagulated with bipolar cautery. Once the tentorium is sectioned and the arachnoid is opened further, the superior vermis comes in the surgical field. The precentral vein is present anteriorly to the superior vermis entering into the straight sinus. The quadrigeminal cistern can be opened further toward the left side once the precentral vein is coagulated and sectioned.

When the splenium is draping over the third ventricle tumor, a small portion of the splenium may need to be removed with suction. Usually, in such cases, the splenium is thin, and if the incision is limited to 1 cm, I have never had any neurologic sequences. To expose a tumor extending into the lateral ventricle through the choroidal fissure, a further retraction of the posterior cingulate gyrus laterally and a section of the splenium are needed. In the lateral ventricle, the choroid plexus is coagulated, and the tumor is gradually brought out of the lateral ventricle.

Tumor Resection

The vein of Galen and its tributary veins need to be identified. The splenium of the corpus callosum is gently lifted, and the bilateral internal cerebral veins are identified. The vein of Rosenthal of the right side is present laterally to the tumor capsule. Placing the brain retractor tip deeper onto the isthmus of the cingulate gyrus will expose the anterior portion of the vein of Rosenthal and lateral portion of the tumor capsule. The only arterial structure in this location is the posterior medial choroidal artery, which is coagulated and sectioned. The tumor capsule is coagulated and opened. Internal debulking and gentle traction of the tumor capsule make it easier to separate Galen's veins from the tumor capsule. A cottonoid strip is placed between the vein and tumor capsule. For the dome of the tumor or the anterior third ventricle, the micro-

scope trajectory is almost parallel to the roof of the third ventricle. For the posterior dome of the tumor and the quadrigeminal plate, the trajectory is parallel to the precentral cistern.

In general, to excise the tumor, the right side wall of the tumor is separated from the right thalamus. Then the superior dome of the tumor is separated from the vein of Galen and the internal cerebral veins. Posteriorly, the tumor is lifted and the quadrigeminal plate is identified. Thus, the posterolateral portion of the tumor is secured in the surgical field. Once the tumor is gradually brought posteriorly and the cottonoid strip between the thalamus and right side tumor wall is placed farther forward, the third ventricle is entered. Above the superior dome of the tumor, the internal cerebral vein is gradually separated from the tumor capsule. Posteriorly, by lifting the posterior dome of the tumor, the floor of the third ventricle comes into the surgical field just anterosuperior to the superior colliculi. At the floor of the third ventricle, the entry of the aqueduct of Sylvius is also visualized. The third ventricle is protected by placement of a cottonoid strip. Once reduced by internal decompression, the tumor is gently retracted toward the surgical field, and the vein of Rosenthal of the left side is visualized. It is separated from the tumor capsule, and then the left wall of the third ventricle is gradually separated from the tumor. Once the four corners of the tumor are isolated from the wall, floor, and roof of the third ventricle, the tumor is gradually brought out and delivered. The foramen of Monro of both sides is visualized next to the column of fornices (Fig. 12–20). Cottonoid strips are placed in the third ventricle and tumor-resection cavity for hemostasis for several minutes.

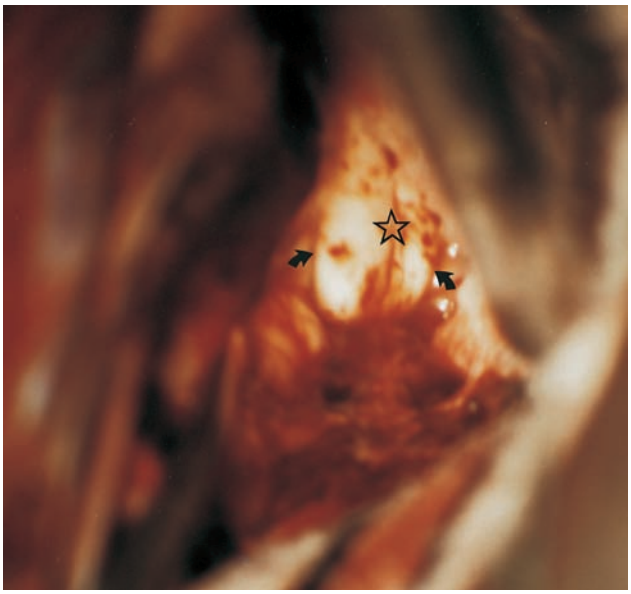


FIGURE 12–20. The third ventricle space after removal of a pineal teratoma. Note the right and left foramena of Monro (*arrows*) and the columns of the fornices in the center (*asterisk*).

Tumor resection is done in general by using bipolar cautery, ultrasonic aspirator, and microsurgical instruments. A teratoma is well encapsulated, and a small teratoma can be removed in toto. In a large teratoma, however, it is often impossible to separate the entire capsule from the surrounding structures without internal decompression. In teratomas, some areas are densely calcified, whereas others are as firm as leather. In such cases, I use a carbon dioxide laser to vaporize the tumor because the ultrasonic aspirator will be of little help. Germinomas are granular and fibrous and often well encapsulated. Other malignant germ cell tumors are often extremely vascular. These tumors would be best treated by preoperative chemotherapy, followed by a second-look surgery when the tumor is smaller, largely necrotic, and much less vascular. Pineoblastomas have a pseudocapsule and are necrotic but quite vascular; however, they are often easily suctioned away.

During manipulation of a pineal region tumor, it is not uncommon to encounter a hemorrhage from a lacerated vein of Galen. The hemorrhage is often brisk but can be controlled with gentle pressure by temporary application of gel foam and compression with cottonoid, which can often maintain the continuity of the vein.

Closure

The tumor-resected cavity and the third ventricle are inspected for any residual tumor by changing the angles of the microscope trajectory. At the anterior end of the third ventricle, the fornices, and the foramen of Monro of the right and left side are observed. Laterally, the right and left thalami are observed. Often, open entry to the aqueduct of Sylvius is observed at the posterior floor of the third ventricle.

Hemostasis is confirmed by valsalva maneuvers. I do not leave in the ventricle any hemostatic agents, such as Gelfoam or oxidized cellulose, because to do so would result in postoperative hydrocephalus. The ventricle is thoroughly irrigated with sterile normal saline, and the dura is repositioned and closed in a watertight fashion. Following placement of multiple dural tack-up sutures at the craniotomy edge, the free bone flap is repositioned. I use 00 nonabsorbable sutures to secure the bone flap. The skin is closed in two layers.

Alternate Approach

An infratentorial supracerebellar approach is suitable for open biopsy or for a small pineal tumor in a patient with small or slitlike ventricles. I use a posterior fossa craniotomy for this approach with the patient in a sitting position. The sitting position is obtained with a three-pin fixation device to the head with the neck flexed slightly forward. To detect and minimize air emboli during a sitting position, a precordial Doppler monitor, placement of a central venous line into the right atrium, and the use of posterio-end-expirectory pressure are important.

The skin incision is placed in the midline from 2 cm above the inion to the C-2 spinal process. Midline dissection is attained, and the squamous portion of the occipital bone, the foramen magnum, and the C-1 arch are exposed. Accurate distance between the torcular helophilli and the foramen magnum is provided by a measurement on the midsagittal MRI. A burr hole is made on the midline just below the torcular. At that time, the dura on the both sides of the bony ridge is first exposed, and then the midline bony ridge is removed with a rongeur because the internal occipital protuberance is well developed, even in young children. The posterior fossa dura is separated using a Penfield no. 3 dissector through the burr hole. The soft tissue at the foramen magnum is sharply dissected, separating the pericranium from the posterior fossa dura. A posterior fossa craniotomy is done using a Midas drill. Superiorly, the edge of the craniotomy is parallel to the lower edge of the lateral sinus; inferiorly, the craniotomy crosses the foramen magnum. At the upper edge of the craniotomy, the bone is removed up to the superior edge of the lateral sinus on both sides.

The dura is incised in a C-shape based rostrally toward the lateral sinus. Dural opening is done at the superior two thirds of the posterior fossa dura. The lower dura would suspend the cerebellum during the depression at the supracerebellar approach in a sitting position. The surgical microscope provides optimal visualization and illumination. There are always several bridging veins from the cerebellum to the straight sinus. Already stretched by the gravity, any downward retraction of the cerebellum would aggravate and avulse them from the sinus. These veins are coagulated and sectioned away from the sinus, which allows further gravitational fall of the cerebellum from the tentorium. The tentorial opening is in the surgical field. Subsequently, the superior vermis needs to be depressed, and the microscope trajectory is changed. Anteriorly, the thick arachnoid of the quadrigeminal cistern is visualized and widely opened. The precentral vein is present in the cistern, which may be displaced to either side. Most of the time this vein needs to be coagulated and sectioned to obtain direct access to the tumor. Further dissection of the arachnoid provides a wider view of the pineal region. The vein of Galen is present at the dome of the tumor and noted entering into the straight sinus. The veins of Rosenthal are seen laterally. Posteromedial choroidal arteries and minute blood vessels present on the posterior wall of the tumor capsule are coagulated. To expose the superior colliculi, it is necessary to depress the cerebellum further, although often it is still not well visualized.

The tumor capsule is opened, and biopsy is done of the inner contents. Subsequent internal debulking of the tumor allows easier separation of the tumor capsule from the surrounding veins. I usually separate the lateral wall of the tumor on both sides and then circumferentially move upward, where the vein of Galen and the internal cerebral veins are separated from the tumor. The internal cerebral veins are covered by the velum interpositum at the roof of the third ventricle. I use bipolar cautery to gradually separate the tumor capsule from the venous structures. Once significant internal decompression of the tumor is attained, it is possible to place cottonoid slips into the third ventricle by sliding the cottonoid between the thalamus and the lateral wall of the tumor and between the internal cerebral veins and the dome of the tumor. Subsequently, the tumor is gently lifted and the floor of the third ventricle is identified. The tumor is gradually brought out posteriorly and delivered. Following hemostasis from the third ventricle, cottonoids or other hemostatic agents are removed.

The dura is coagulated in a watertight fashion. The bone flap is repositioned and secured to the edge of the bone in multiple sites with 00 nonabsorbable ligatures. The muscle layers are closed with 000 absorbable sutures. The skin is closed in two layers.

Postoperative Management

Immediate postoperative CT or MRI is obtained to identify residual tumor and to assess intracranial hemorrhage and ventriculomegaly. Postoperatively, patients are monitored with external ventricular drain output drainage pressure at a 10 cm of H_2O for several days. The drain is then converted to an ICP monitor for 2 days. During that time, patient's symptoms and the interval changes in ventricular size on follow up CT are closely mentioned. If the hydrocephalus is under control, the external ventricular drain is removed. If hydrocephalus is persistent, either third ventriculostomy or shunt placement is performed. About 80% of our patients with pineal tumors are free of hydrocephalus following tumor resection.

EDITOR'S COMMENTARY

The surgical management of midline tumors challenges the skills of even the most talented microsurgeons. Children with craniopharyngiomas in particular should ideally be cared for in centers with experience in all the treatment options available, and treatment should be individualized. Intracavitary, stereotactic and conformal radiotherapy techniques represent a complement rather than a competitor to surgical resections, in view of the high morbidity of gross total resections. With regard to pineal turmors, most neurosurgeons now agree that a histological diagnosis is necessary if the identity of the lesion is in question. In such cases, some surgeons favor proceeding with a stereotactic biopsy, whereas others prefer a craniotomy because of unfavorable results with hemorrhage or sampling error. Most germinomas respond well to adjuvant therapy and do not require extensive resection, although the optimal radiation dose and fields, and the role of chemotherapy remain points of controversy. Malignant germ cell tumors often show an incomplete response to adjuvant therapy and "second-look" surgery is often pursued to biopsy and remove areas of residual disease. The optimal management of pineal parenchymal tumors remains uncertain because of the rarity of these lesions; at present, pineoblastomas are generally treated after maximal resection with similar protocols as other high-risk primitive neuroectodermal tumors.

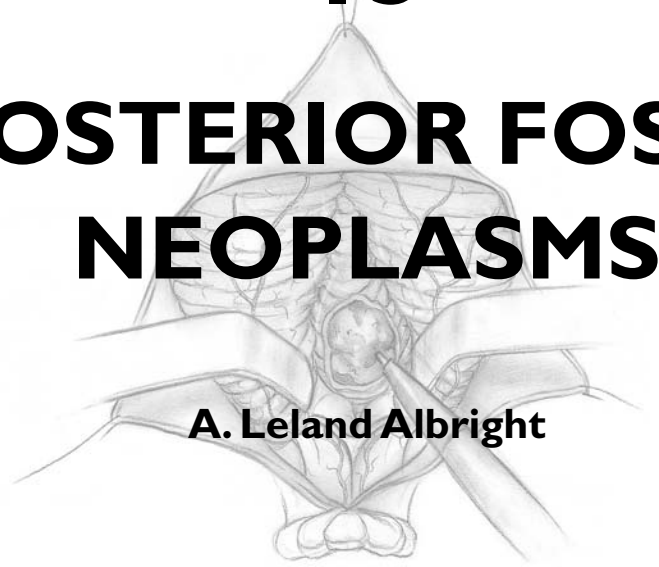
PEARLS

In this author's experience:

- The histologic nature of a pineal region tumor of children is highly predictable based on the gender, age, neuroimaging appearance and tumor markers; however, if in doubt, confirm histology. If biopsy is needed, the author recommends it be done through craniotomy.
- Hydrocephalus is the primary cause of symptoms. It is treated by the third ventriculostomy, when the CSF studies such as cytology and tumor markers are performed.
- Germinomas and malignant nongerminomatous germ cell tumors are chemosensitive. Chemotherapy should be incorporated in the treatment protocol.
- Pineal region tumors can be removed with minimum morbidity through supratentorial or posterior fossa approaches. Surgeons should be familiar with advantages and disadvantages of both approaches.

13

POSTERIOR FOSSA NEOPLASMS



A. Leland Albright

The common posterior fossa tumors in children include medulloblastomas, juvenile pilocytic astrocytomas (JPAs), ependymomas, and brainstem gliomas. Craniotomies are almost always indicated for medulloblastomas, astrocytomas, and ependymomas: to make a tissue diagnosis, to remove tumor and thereby improve prognosis, and to treat the associated hydrocephalus. Craniotomies are probably never indicated for diffuse brainstem gliomas but occasionally are indicated for focal enhancing brainstem gliomas.

PREOPERATIVE CARE

Corticosteroids are begun at admission to improve symptoms of hydrocephalus and to reduce postoperative edema. Prophylactic anticonvulsants are not needed. Ideally, the scalp is cleansed with an antibacterial shampoo the night before and the morning of craniotomy.

If the preoperative scan demonstrates the presence of a posterior fossa tumor with hydrocephalus, hydrocephalus can be treated effectively by an external ventricular drain (EVD). If the child is alert and craniotomy is planned for the next day, we begin corticosteroids and do not insert the EVD until just prior to craniotomy. Precraniotomy shunts almost never are indicated. If the child is drowsy from hydrocephalus and is admitted late in the day, I usually do the EVD and craniotomy that night; however, if additional time is needed to obtain a good-quality scan or a more experienced surgical team in the operating room, it is reasonable to insert the EVD that night and defer the craniotomy until the next day.

It is important not to administer preoperative sedatives until shortly before the operation, if at all. After their administration, the child must be monitored closely by pulse oximetry, vital signs, and neurologic checks.

INTRAOPERATIVE TECHNIQUES

A complete description of the anesthetic management during posterior fossa operations can be found in Chapter 69 of the companion textbook, *Principles and Practice of Pediatric Neurosurgery*.

Monitoring

Monitoring for posterior fossa craniotomies includes an electrocardiogram and pulse oximetry, blood pressure monitored by an arterial catheter, and urinary output. Intravenous access at two sites using catheters of 22 gauge or larger (depending on the size of the child) is indicated. It is more important to have two good intravenous catheters than to have central venous access. The utility of Doppler monitoring during posterior fossa craniotomies is debatable. Dopplers frequently detect significant air embolism when craniotomies are performed with the patient in the sitting position, but the use of that position has declined. The likelihood of detecting significant air embolism if the child is in the prone position is small. Because significant air embolism is infrequent if the child is in the prone position I do not routinely use Doppler monitoring. I have not found

monitoring of somatosensory-evoked potentials or brainstem-evoked potentials to be helpful and infrequently use either.

External Ventricular Drains

I prefer to insert EVDs in the right frontal region at the junction of the pupillary line and the coronal suture. I prefer the frontal site because the anatomic coordinates used during insertion of the catheter allow reliable placement of the catheter in the frontal horn of the ventricle and, to a lesser extent, because the frontal position of the catheter postoperatively is more comfortable for children than are posterior catheters. I make a 5- to 7-mm linear incision, perforate the skull with the M8 bit on a Midas Rex drill (Medtronic, Fort Worth, TX), coagulate and open the dura, and then insert a long (30 cm) EVD catheter for the appropriate length into the lateral ventricle. That length, usually 5.5 to 6.5cm, can be determined from measurements made from preoperative scans. The distal end of the catheter is tunneled several centimeters inferiorly toward the temporal region where it exits and is connected to a closed drainage system. I remove 10 to 20 mL of cerebrospinal fluid (CSF) at this time and then close the system until the craniotomy is being performed.

Alternatively, some neurosurgeons prefer to insert the catheter posteriorly, either through an occipital burr hole 6 cm above the inion and 2.5 cm lateral to the midline, with a trajectory toward the ipsilateral medial canthus, made after the child is in the prone position, or through a burr hole made in the parietal region at Keen's point. In my experience, catheters inserted at Keen's point have not been as well positioned in the frontal horn as have catheters inserted frontally, and catheters inserted occipitally have been uncomfortable postoperatively as the child lies supine. In addition, there may be more difficulties cannulating the occipital horn than the frontal horn.

A few neurosurgeons do not insert EVDs before removing posterior fossa tumors but prefer to make an occipital burr hole so that the ventricle can be cannulated if the child develops acute hydrocephalus postoperatively. That technique does not allow CSF drainage during the operation, and it assumes that if acute hydrocephalus develops, the ventricle can be cannulated under emergent conditions. I prefer the more controlled conditions that the EVDs provide.

Positioning

The head of an infant 1 year of age or younger usually is stabilized on a large foam block (Fig. 13–1) or on a pediatric horseshoe headholder, ideally padded with gel. For children 1 to 2 years of age, I use either the horseshoe headholder or pediatric pins in the Mayfield holder and I use adult-sized pins in children 4 years of age and thereafter. It is important to remember that not all 1- to 2-year-old children have skulls thick enough to hold pins and that the skull of a child may be thinned by chronic hydrocephalus; in such patients, less pin pressure may be tolerated before the pin causes a focal skull indentation or depressed fracture. Pin pressures of 20 to 30 pounds are used in children 1 to 3 years of age and increased progressively thereafter to the 60-pound pressures used for an adult skull when the child is 12 years of age or older.

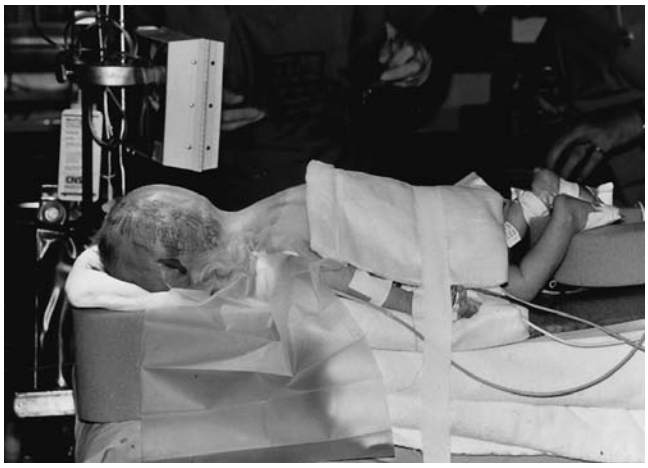


FIGURE 13–1. Infant positioned prone on foam blocks prior to posterior fossa tumor removal.

Once the child is in the prone position, I move him or her to be at the edge of the operating table on my side, and then I angulate the head approximately 30 degrees away from me and flex the neck about 30 degrees (Fig. 13–2). After the head position is finalized, it is important to auscultate the chest to ensure that the neck flexion did not advance the endotracheal tube into the right mainstem bronchus. Once the patient is in the correct position, the surgeon can stand comfortably by the side of the table without leaning over it, and his or her right hand can be held comfortably at the surgical site for several hours if need be, with the assistant standing to the left.

Alternatively, some surgeons prefer to keep the child in the center of the operating table and use the quadrascope so that the primary surgeon on one side of the table and the assistant on the opposite side have approximately equal access to the child. The sitting position for craniotomies was used commonly until the past decade, but its disadvantages of air embolism, hypotension, and fatigue of the surgeon's arms outweigh the advantages of better blood and CSF drainage, and its use has declined substantially.

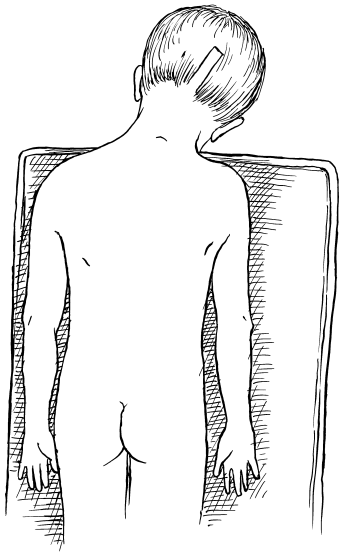


FIGURE 13–2. Patient positioned prone at the surgeon's edge of the operating table with the neck flexed 30 degrees and angled away from the surgeon 30 degrees. Scalp has been trimmed of hair in a 5-mm strip from inion to C-2.

Scalp Preparation

No longer does scalp preparation involve shaving the entire head. Rather, a strip of scalp 5 to 7mm wide can be trimmed along the line of incision with scalp clippers, thereby decreasing the potential of staphylococcal infection, which has been reported after shaving. The operative site is prepared with an iodine-containing solution and alcohol. The solution, Hibiclens, carries a label stating that it is not to be used around the scalp or face because of the risk of conjunctivitis if the solution enters the eye.

Exposure

Exposure of midline tumors begins with a midline incision from the inion to C-2. I control subcutaneous bleeding with the coagulating needle-tip cautery; alternatively, clips can be applied to the skin edges. The cautery can be used to dissect periosteum off the occipital bone laterally and down to the foramen magnum, and then to dissect soft tissues away from C-1 and the space between the foramen magnum and C-1. As the periosteum is dissected off the occipital bone, veins often are divided where they enter the bone and can bleed substantially. Their bleeding can be controlled by electrocautery or bone wax. If a Doppler is used and air embolism is detected by its characteristic washing-machine sound, treatment includes flooding the operative field with saline, increasing the venous pressure by lowering the head of the bed or compressing the jugular veins, and aspirating air from a central venous catheter if one is available. Subcutaneous tissues are held apart with cerebellar retractors.

Pediatric neurosurgeons increasingly perform an occipital craniotomy rather than a craniectomy, which leaves a concave depression in the skull and a potential site for trauma. For a craniotomy, a single burr hole can be made in the midline at approximately the level of the torcula (obviously taking considerable care not to enter it), or two burr holes can be made on either side of the midline in the region of the transverse sinus (Fig. 13–3). These burr holes can be made safely with an M8 bit on a Midas Rex drill; I have reservations about making burr holes over the torcula with powered burr-hole makers. The traditional exposure is by an occipital craniectomy, removing bone with Kerrison and Leksell rongeurs from the foramen magnum upward until the inferior aspect of the transverse sinus is exposed.

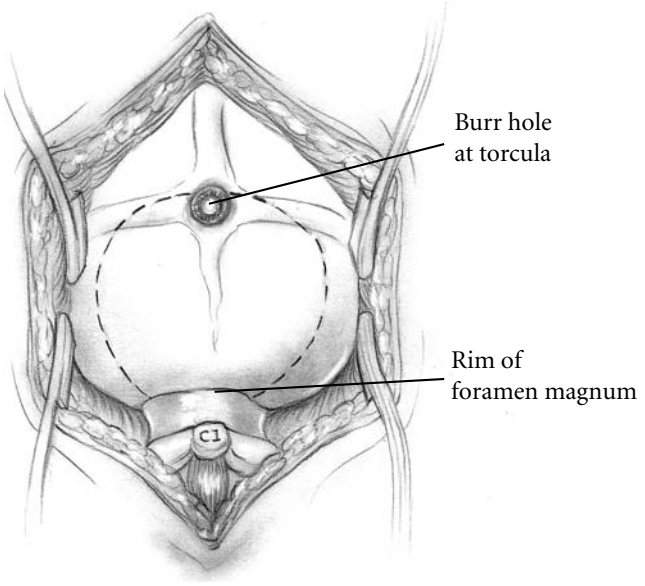


FIGURE 13–3. Exposure of the occipital bone, foramen magnum, and C-1. A single burr hole can be made with a Midas Rex M8 bit (not an automatic-power burr-hole maker) at the torcula, or two holes can be made at the transverse sinus. After dura is stripped away from the overlying bone and foramen magnum, the craniotomy flap is made with a B5 bit along the dashed lines, and holes are made in the bone for its later reapplication.

Dura is dissected away from the undersurface of the occipital bone with a Penfield 3 or Adson periosteal dissector and away from the undersurface of the foramen magnum; then the craniotomy is performed using a high-speed drill. Holes are made in the bone margins for its reapplication later in the case. I usually begin removing CSF from the EVD as the burr hole is being made and drain the CSF until the dura is slack before opening it.

The dura is opened in a Y-shaped manner, opening it initially laterally over the cerebellar hemispheres and then coming across the midline while the occipital sinus is temporarily occluded by hemostats (Fig. 13–4). I prefer not to occlude the sinus with Weck clips because of the artifact they cause on postoperative scans. Once the midline dura is opened, I occlude the midline occipital sinus with a permanent suture; this technique is more reliable than electrocautery. I then retract the superior triangle of the dura superiorly. The inferior midline dural opening is made down to C-1 immediately lateral to the midline sinus, and additional dural tackup sutures are inserted to retract the dura laterally, reflecting it over cottonoid strips that are kept moist during the tumor resection to minimize drying and shrinking of the dura. It is not necessary to remove the lamina of C-1 to remove most posterior fossa tumors (Fig. 13–5).

TUMOR RESECTION

Midline Tumors

Arachnoid over the foramen magnum is opened. I insert a 0.75 × 0.75-inch cottonoid patty over the dorsal aspect of the cervical cord to minimize blood from migrating down the spinal axis, and then elevate the most caudal aspect of the vermis or tumor and inspect the floor of the fourth ventricle, at least its caudal aspect, and insert a 0.5 × 2.0-inch cottonoid strip upward onto the floor of the ventricle to minimize its risk of being traumatized. To facilitate access to midline tumors, it is often helpful to divide the most caudal 5 to 10 mm of the vermis. If the tumor appears to be a medulloblastoma, I obtain a specimen for frozen-section diagnosis because that diagnosis may alter the extent of subsequent resection if the tumor invades the floor of the fourth ventricle.

The medial aspects of the cerebellar hemispheres are held laterally by retractor blades affixed to a stable system, often the Greenberg system, secured to either the Mayfield headholder or to a Bookwalter bar attached to the rail on the side of the operating table. Under microscopic magnification, the ultrasonic aspirator is used to remove the tumor; lasers are of little use in removing posterior fossa tumors. I insert the aspirator into the nidus of the tumor and remove the central two thirds to

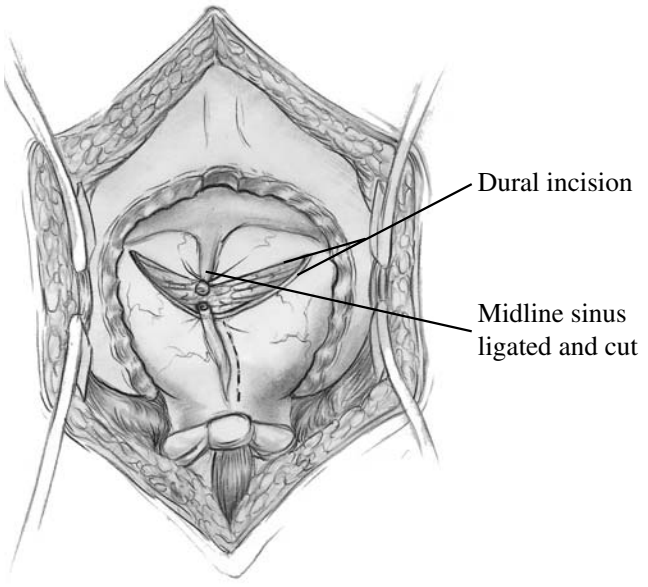


FIGURE 13–4. Dura is opened in a Y-shaped manner. Initial dural openings are diagonal over the hemispheres lateral to the midline. The midline sinus is temporarily occluded with hemostats and then is divided with scissors and ligated.

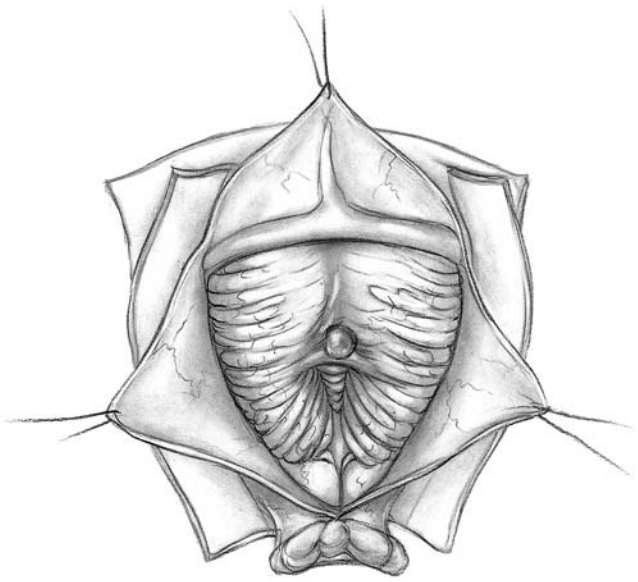


FIGURE 13–5. Dural leaflets have been retracted, exposing the posterior and inferior aspects of the cerebellum.

three fourths of the medulloblastoma and then use the aspirator to resect the periphery of the tumor, usually superiorly first so that the aqueduct is exposed and covered with a 0.5 × 0.5-inch cottonoid patty. Next the lateral margins of the tumor are dissected off the hemisphere and peduncle, and lastly the inferior portion is removed toward the brainstem. The tumor is gently retracted away from the cerebellum with either the suction tip or a cottonoid patty, and the aspirator is used to develop the tumor-normal interface (Fig. 13–6). If the tumor is a medulloblastoma and seems to infiltrate the brainstem, the resection should shave the tumor down to the ventricular floor but not below it. There is no evidence that 100% resection of a medulloblastoma is associated with better survival than a 99% resection, leaving 1% in the brainstem. Approximately 15% of medulloblastomas are exceedingly vascular. For these tumors, primarily two removal techniques are used: Remove a bit of tumor, then stop and obtain hemostasis, or use the aspirator and work rapidly until the tumor is removed, stopping only if bleeding from a substantial vessel is encountered. I prefer the latter technique. After the obvious tumor is removed, I insert a cottonoid patty, apply the suction tip to it, and gently wipe the margins to see whether any tumor remains. After tumor removal is complete, the walls of the resection cavity may be lined with Surgicel.

If the tumor is an ependymoma and enters the floor of the ventricle, I usually break scrub, talk to the parents, and confirm that a resection to remove the tumor completely may cause cranial nerve deficits. Because residual ependymomas are likely to recur, the higher morbidity associated with complete resections may be acceptable, although certainly not at any possible neurologic cost. If the ependymoma is in the cerebellopontine angle, it usually can be dissected off the cranial nerves; if it encases the lower cranial nerves unilaterally, I try to dissect it off them, knowing that the attendant morbidity may require a gastrostomy and tracheostomy postoperatively, at least temporarily.

A Valsalva maneuver is often performed to ensure that no occult venous bleeding is occurring. The dura is closed, initially with a single central suture to bring the dural leaflets together, and then with a running 4-0 polygalactin (Vicryl) suture for a water tight closure (Fig. 13–7). If the dura will not close completely primarily, small defects may be closed using a piece of deep cervical fascia; larger defects may be closed with lyophilized dura. I secure the craniotomy flap back in place with 2-0 Vicryl sutures (Fig. 13–8) and close the wound in layers with absorbable suture and the skin with subcuticular suture. Subcutaneous drains are not needed.

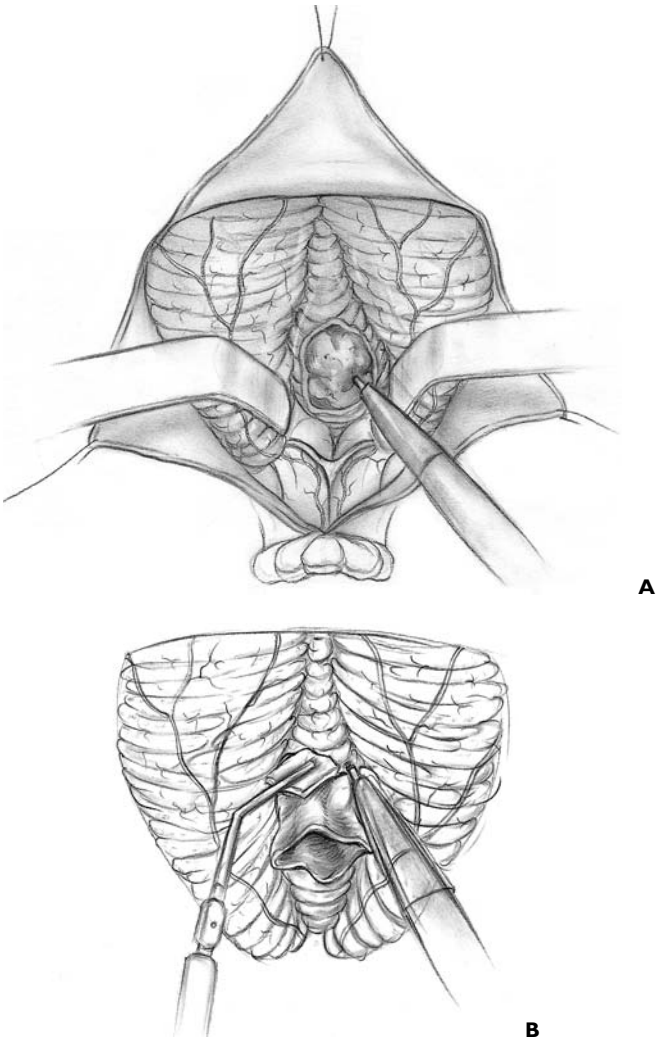


FIGURE 13–6. A, B: Cerebellar tonsils are held laterally with retractor blades. The core of the tumor is removed with an ultrasonic aspirator, which is then used with suction and cottonoids to develop the tumor–normal interface.

Cerebellar Hemispheric Tumors

Many JPAs have an associated cyst. The vertical incision is made between the midline and mastoid, centered over the maximal tumor volume. If dura over the tumor is taut despite CSF drainage, dura can be punctured and a blunt Cone needle inserted into the cyst to remove cyst fluid until the dura is soft. The dural opening for hemispheric tumors is triangular or quadrangular. Unless the tumor comes to the cortical surface, a corticotomy is needed to approach the tumor. There seems to be no difference in morbidity between horizontal and vertical cortical openings in the cerebellar folia; I use whichever will give me the most direct access to the tumor.

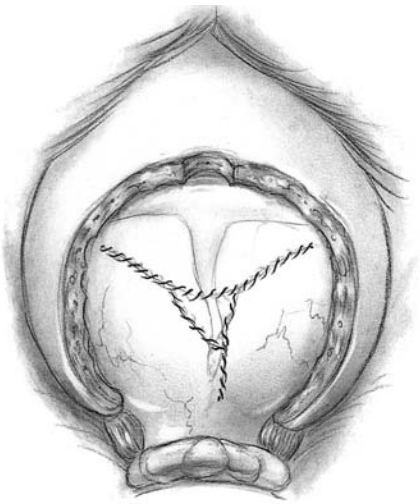


FIGURE 13–7. Dura closure, primarily by approximation of dural edges, supplemented with a central graft of cervical fascia if needed.

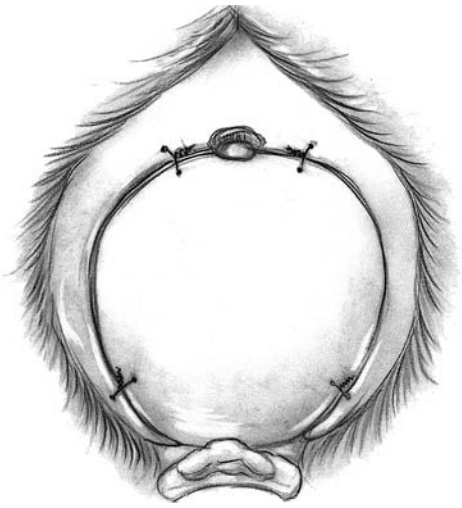


FIGURE 13–8. Craniotomy flap replaced and secured with 2–0 polygalactin sutures.

Thinned-out cerebellum over the tumor is teased away from the tumor with a Penfield 1 dissector and held away with blades of a self-retaining retractor system. The tumor may be removed by using the ultrasonic aspirator or traditional instruments, such as a Penfield dissector and bipolar cautery. The ultrasonic aspirator is particularly helpful in removing large JPAs to debulk the tumor's interior before its periphery is approached. The tumor boundary of JPAs is variable; a distinct demarcation between tumor and cerebellum is present in some regions, but in other areas the boundary is less distinct. If the tumor is a JPA, meticulous attempts to remove the tumor are indicated because it is surgically curable. Most cyst walls are membranous and translucent and do not need to be excised; however, if the rim of a cyst enhances on preoperative scans, the rim should be either removed or biopsied to determine whether it contains tumor.

Focal Brainstem Gliomas

Focal tumors in the midbrain are usually JPAs and often are treated better by stereotactic biopsy and focal irradiation than by resection. Resections are occasionally appropriate because of concerns about the effects of radiosurgery on adjacent structures or because of cysts associated with the tumors. In such cases, dorsal mesencephalic tumors are approached through a supracerebellar, subtentorial route, as for a pineal region tumor. The tumors often bulge dorsally into the quadrigeminal cistern and are entered there with the ultrasonic aspirator, using techniques similar to those for a cerebellar astrocytoma, on a much smaller scale. Focal brainstem gliomas are similar in their consistency to cerebellar astrocytomas, but they also have regions where the distinction between tumor and brainstem is difficult to ascertain, where risks of resections are high.

Focal enhancing tumors rarely occur in the pons, but they do occur in the medulla. Solid tumors in this location may be treated with radiosurgery. Those with associated cysts, especially those with a lateral enhancing nodule, may be resected. The tumors may be approached by a retromastoid craniotomy retracting the cerebellar hemisphere either posteriorly or superiorly or approached by a posterior midline exposure. In either case, the resection is often done with the small aspirator tip, working between the inferior cranial nerves; postoperative nerve paresis is common, at least temporarily. I have not found the intraoperative monitoring of lower cranial nerves to reduce the risk of a postoperative nerve palsy.

POSTOPERATIVE CARE

For the first 3 days after posterior fossa craniotomy, several symptoms are common, including headache, poor appetite, nausea, vomiting, and low-grade fever. We treat postoperative pain with intravenous morphine every 1 to 2 hours, vomiting with Zofran, and fever with acetaminophen.

Corticosteroids are given in constant doses for 3 days postoperatively, tapered off over 7 days. It is common to give histamine blockers with corticosteroids, but I am

aware of no literature documenting their benefit in children. Postoperative prophylactic antibiotics have not been shown to reduce the small risk of a wound infection.

The EVD is positioned 10 to 15 cm above ventricular level for 2 days and then elevated by 5 cm daily, to 25 to 30 cm above ventricular level, clamped for 12 to 24 hours, and removed if the child remains asymptomatic. In the few children in whom symptoms of hydrocephalus recur, treatment includes insertion of a new EVD catheter for an additional 5 to 7 days. If hydrocephalus persists after the second EVD catheter, we treat the hydrocephalus with an endoscopic third ventriculostomy or a shunt. While the EVD is in place, if there is a CSF leak through the incision, the incision is oversewn and the height of the EVD is lowered to promote additional CSF drainage for 2 to 3 days; then gradual elevating of the EVD is resumed.

Risks of operation include death (1%), wound infection (1%), ventriculitis/meningitis (2%), CSF leak (1%), cerebellar signs (transient 25%, permanent 10%), and the pseudobulbar posterior fossa syndrome (15%). Postoperative shunts are needed in 25% of patients. Pseudomeningoceles are due to either inadequate dural closure or inadequate CSF drainage and are better prevented than treated. Treatment includes lowering the EVD, compression dressings, and occasional aspiration.

The posterior fossa syndrome of mutism, whining, and uncooperativeness may develop 24 to 72 hours after operation. Its occurrence does not appear to be closely related to the extent of vermis division. The syndrome's cause is unknown but has been attributed to edema of the dentato-rubro-thalamic tract. No effective treatment of the syndrome is known, and improvement occurs over the following weeks and months.

In the occasional circumstance when a pin site is found postoperatively to have caused a focal skull indentation, most often in younger children, the site can be closed with a suture, and the small depressed fragment may not need to be surgically elevated. When such focal fractures and indentations occur, they raise the possibility of an underlying epidural hematoma, which will be detected on the postoperative scan.

Postoperative scans are obtained ideally within 24 hours after operation to determine whether residual tumor is present. If the scan shows less than 1.5 cm^2 residual medulloblastoma, the residual usually can be treated with irradiation and chemotherapy; if more than 1.5 cm^2 is present, reoperation or adjunctive radiosurgery should be considered. If the tumor is an astrocytoma and there is definite residual tumor, it can be treated by reoperation or radiosurgery. If the tumor is an ependymoma and there is more than 1.5 cm of residual tumor, reoperation should be seriously considered because of the lower response of ependymomas to irradiation or chemotherapy.

EDITOR'S COMMENTARY

Surgical therapy plays a central role in the management of most infratentorial tumors. For patients with cerebellar astrocytoma, gross total resection of the tumor is often curative and adjuvant therapy is usually not required. Ependymoma is also largely a "surgical disease", because obtaining a gross total resection is the single factor that strongly influences overall survival; patients with residual disease frequently experience disease recurrence, despite the use of adjuvant therapy. However, achieving an extensive resection is often challenging because these lesions may invade the fourth ventricular floor or infiltrate around the cranial nerves and lateral brainstem, which complicates obtaining a gross total resection without significant morbidity. Provided that the family understands the risks involved, an attempt at complete resection is generally warranted if this can be achieved without unacceptable morbidity. In contrast, the management goal in children with medulloblastoma is to obtain a "nearly total" resection, leaving a focal layer of tumor within the brainstem, because adjuvant therapy is quite effective and outcome is not measureably improved by more aggressive resections. For the above tumor types, the use of intraoperative neurophysiological monitoring when resecting lesions in and around the fourth ventricular floor and cranial nerves remains controversial (even among surgeons at our institution). Although selected focal brainstem tumors are amenable to surgical resection, most brainstem gliomas are diffusely infiltrative and malignant, and do not warrant surgical intervention.

PEARLS

- Hydrocephalus associated with posterior fossa tumors can be managed safely and effectively with EVD and tumor resection in 75% of cases.
- If children present in stupor or coma from a posterior fossa tumor and associated hydrocephalus, immediate EVD and tumor removal are indicated because if only an EVD is performed and the child does not awaken, the tumor may be the reason.
- Operations done with the patient in the Concorde position have fewer risks than those done in the sitting position and are more comfortable for the surgeon.
- Posterior fossa craniotomies can be performed safely and can avoid the postoperative concavity and skull defect of craniectomies.
- The midline cerebellar sinus must be occluded during dural opening; otherwise, substantial bleeding may occur.
- Vermian splitting should be minimized.
- Extent of tumor removal correlates with outcome for juvenile astrocytomas, medulloblastomas and ependymomas, and extensive tumor removals should be attempted.

SUGGESTED READINGS

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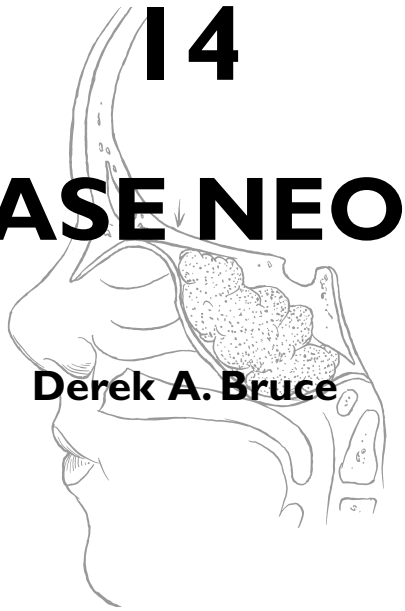
14

SKULL-BASE NEOPLASMS



Derek A. Bruce

There is no single surgical approach to the skull base in children and the various approaches have been discussed elsewhere (Bruce, 1999). The most frequent lesions of the skull base in children are central and anterior in the frontal fossa, air sinuses, or clivus. The most common surgical approach is an extended subfrontal approach or a modification of this to focus on unilateral lesions. This is the procedure, with its various modifications, that is discussed in detail here. Areas that are accessible through an extended subfrontal approach or a modification include the orbit, sinuses (frontal, ethmoidal, sphenoidal, maxillary), nasal cavity, frontal fossa, pterygoid fossa, infratemporal fossa, and clivus. This approach has many advantages: a cosmetically invisible incision; the ability to reconstruct the face, orbits, and nose as well as the forehead; access to a large area of the skull base; and ready access to cranial bone for splitting to aid in the reconstruction. In addition, this approach under the frontal lobes minimizes brain retraction and allows both intradural and extradural access if necessary. It can be performed at almost any age of childhood and is appropriate for the treatment of many pathological lesions in this area.

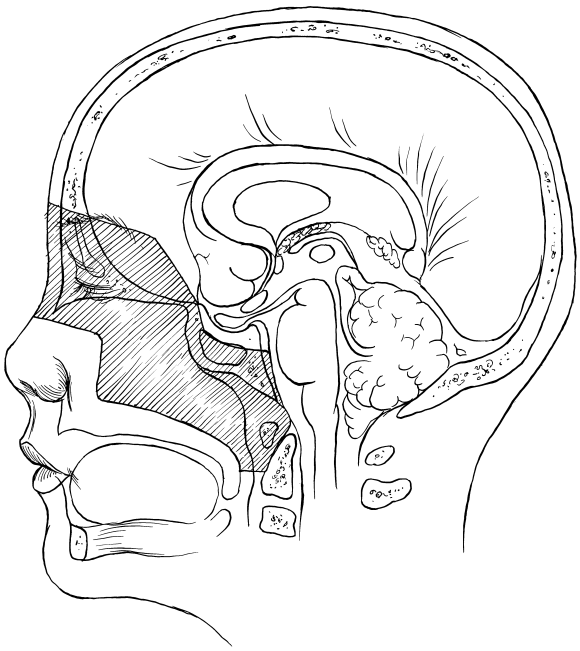
The extended subfrontal is appropriate for midline and paramedian lesions but also for bilateral orbital and cavernous sinus lesions (Fig. 14–1). An extended orbitofrontal approach is used for unilateral orbital and anterior middle fossa lesions (Figs. 14–1C and D).

The primary step in deciding which operative procedure will be best is to identify the epicenter of the tumor. Because the tumor grows from this center, an approach that gives access to the epicenter is most likely to enable complete surgical excision. If the tumor is of extradural origin, the first operative procedure should be by an extradural approach.

The extended subfrontal approach is not adequate for tumors that extend superiorly and posteriorly to the pituitary fossa because it is difficult, if not impossible, to see around the angle produced by the pituitary fossa. The use of the endoscope is of limited value unless the tumor is soft and easily aspirated with the sucker. (Lesions in this area require a Lafort 1 approach through the midface.) The remainder of the clivus can be accessed in cases where the tumor involves the sphenoid sinus as well as the clivus and posterior nasopharynx (Fig. 14–1B). The lateral extension of this exposure is also limited superiorly by the carotid arteries and the cavernous sinus. Inferior to the cavernous sinus, the lateral exposure is good. A final drawback to this approach is that, if the dura of the middle fossa has been eroded by the tumor, it is impossible to achieve a primary watertight repair. A pericranial or other flap must be used, which is a limitation of any approach to this area. Thus, as part of the surgical planning, an idea about the status of the middle fossa dura must be obtained from the imaging studies and an appropriate flap for transfer to the skull base planned. This approach can be combined with a facial degloving or maxillotomy approach to give better access to the facial structures, maxillary sinuses, nasal cavity, and upper jaw (Fig. 14–2). If maxillectomy and upper jaw resection are required, reconstruction below the level of the orbital floor can rarely be done as a primary procedure, and a temporary prosthesis is used to maintain the soft-tissue alignment.

PREOPERATIVE CONSIDERATIONS

Preoperative plain photographs of the patient are helpful to achieve the best approximation to the preoperative ap-



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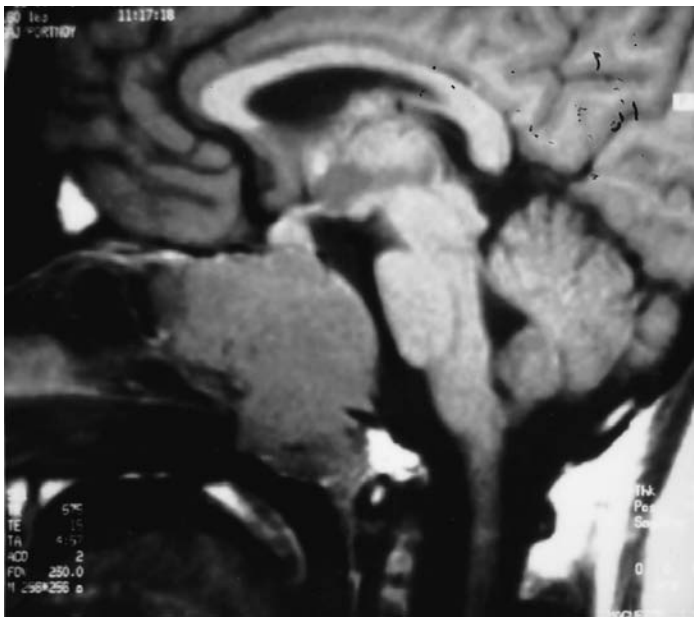

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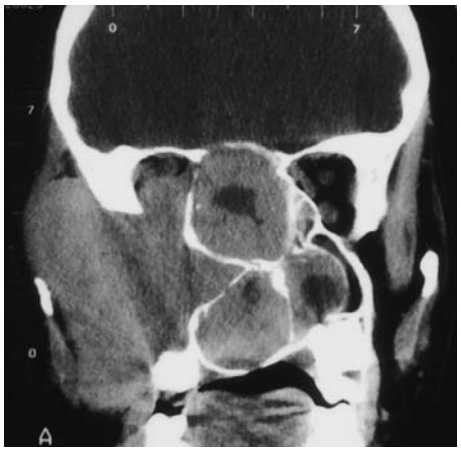

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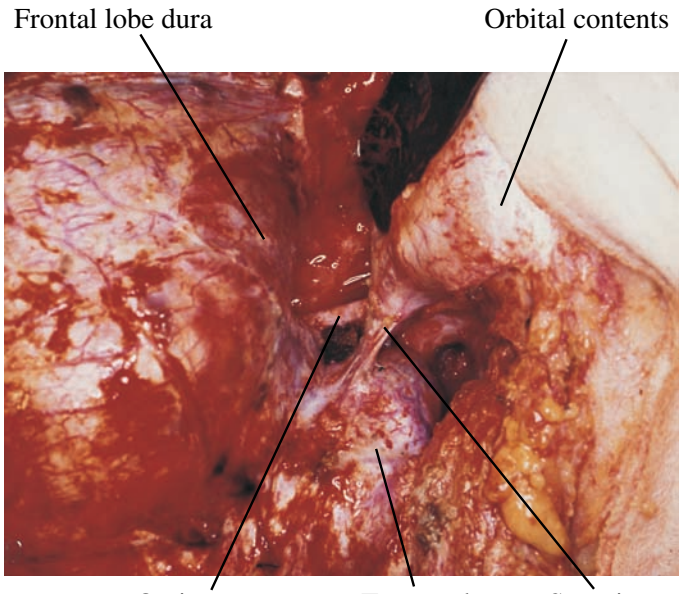
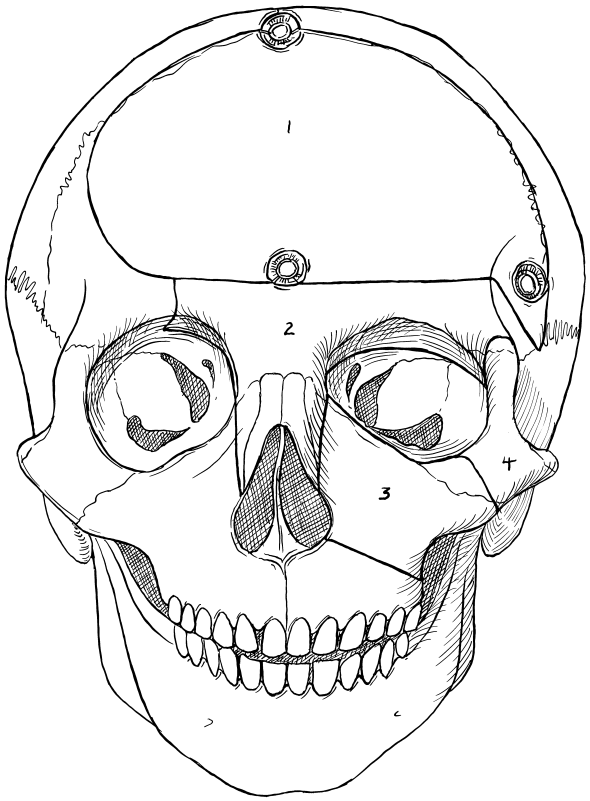


D

FIGURE 14–1. Location of tumors that are candidates for an extended frontal operation. **A:** Midline tumors that are best approached by this technique include lesions involving the frontal base, the ethmoid and spenoid sinuses, nasal cavities, retropharyngeal area, and clivus. **B:** Sagittal T1 image from a magnetic resonance imaging (MRI) scan showing a giant cell tumor replacing the clivus, elevating the pituitary gland and filling the sphenoid sinus and retropharynx. This tumor is an excellent candidate for the extended frontal approach. **C:** Anteroposterior computed tomography (CT) scan after contrast showing a massive angiofibroma involving the nasal cavity, medial and lateral pterygoid spaces, right maxillary sinus, ethmoid sinuses, and right orbit. This would be approached using an extended frontal approach with associated removal of the right zygoma, permitting a right subtemporal approach to the pterygoid space. This tumor was removed completely, and the only incision was the coronal one. **D:** Postresection of unilateral fibrous dysplasia of the right orbit and lesser wing of the sphenoid bone. The optic nerve and superior orbital fissure are completely skeletonized, and the dura covering the frontal and temporal lobes is exposed. The anterior clinoid has been resected.



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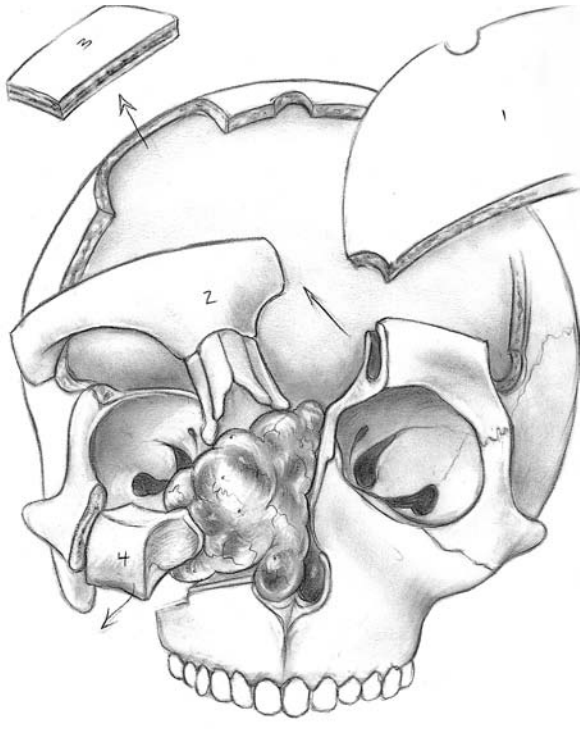

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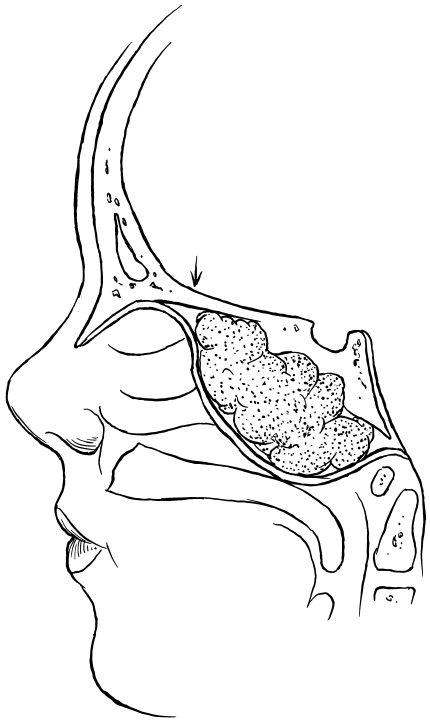
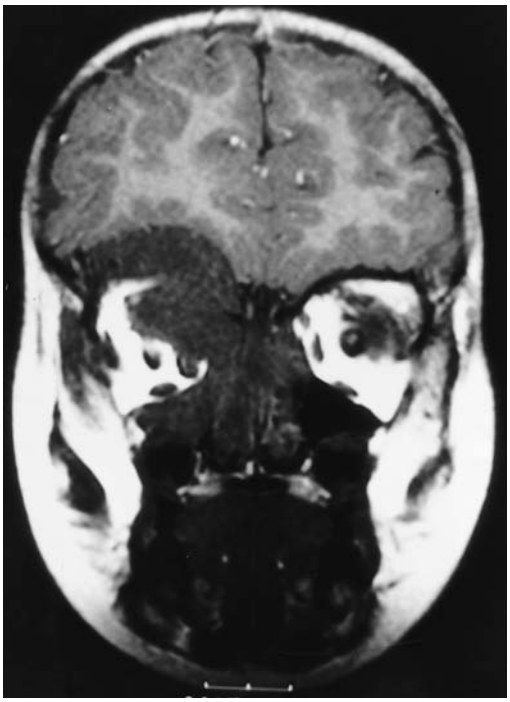


C

FIGURE 14–2. Craniotomy and facial osteotomies for access using an extended frontal approach and maxillotomy. **A:** Craniotomy incision is marked out, in this case extending into the left middle fossa. Osteotomy cuts are shown for removal of (1) craniotomy flap, (2) the midline bandeau and nasal bone, (3) the maxilla, and (4) the zygoma. **B:** Drawing of removal of (1) craniotomy flap, (2) frontal bandeau and nasal bone, (3) additional craniotomy for split cranial bone grafts, and (4) right maxilla swung laterally. **C:** Arrow on lateral view shows the position of the midline skull base cut to free the bandeau, just anterior to the crista galli.



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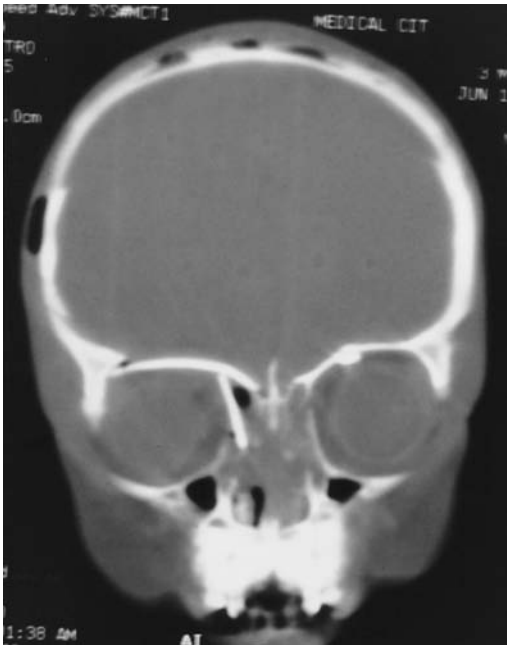


B

FIGURE 14–3. Tumor of right orbit and postoperative bony reconstruction of the orbit. **A:** Coronal MRI scan without gadolinium showing low-grade myxosarcoma invading the intracranial space and right orbit in a 3-year-old boy. **B:** Postoperative coronal CT scan showing the repositioning of the cribriform plate (this was anterior to the tumor and olfaction was preserved). Also shown is the split bone reconstruction of the right orbital roof and medial orbital wall.

pearance during the reconstructive phase of the operation. Often one orbit is involved and occasionally both by tumor. Reconstruction of the orbit is difficult (Fig. 14–3) and often results in enophthalmos or exophthalmos. Keeping as much as possible of the orbital roof and walls intact during the initial bone resection helps facilitate reconstruction.

Neuroimaging studies usually consist of computed tomography (CT), magnetic resonance imaging (MRI), and magnetic resonance angiography (MRA) to evaluate both soft-tissue and bone involvement. Angiography is done depending on the appearance of the tumor, its suspected vascularity, and the degree of compression of the carotid arteries seen on MRA. If the tumor appears extremely vascular, preoperative embolization of the extracranial supply is helpful to decrease blood loss. Surgery should be done within 24 hours of the embolization, if possible, especially if there is additional vascular supply from the internal carotid branches, because these vessels will enlarge over time and may be difficult to control during surgery. Preoperative radiation is rarely recommended because of tissue-healing problems that can occur postirradiation. For highly malignant tumors (e.g., rhabdomyosarcoma) or extensive tumors that are known to respond to chemotherapy or radiation (e.g., olfactory neuroblastoma), presurgical chemotherapy can be valuable for reducing the size of the tumor and to increase the chances of complete resection.

Antibiotics are given at the induction of anesthesia, not before. Nasal cultures may be used to select antibiotic coverage, although we usually do not do this in children. The most frequently used antibiotic is cefotaxime (Claforan). The hair is usually washed the night before with Hibiclens shampoo.

OPERATIVE TECHNIQUE

Following the induction of anesthesia and insertion of monitors, the nasal mucosa is injected with 0.25% bupivacaine hydrochloride (Marcaine) plus 1:200,000 epinephrine. The sublabial region is injected if a sublabial incision is likely to be required. The endotracheal (ET) tube needs to be well secured and in older children usually is wired to the front teeth. The location of the ET tube must be compatible with any potential surgical access requirements. The conjunctival sack is covered with lacrilube, and the lids are sutured closed to protect the cornea. Preparation is with Betadine soap and lotion and includes the entire head and face, including the mouth, if a sublabial incision is required. The nasal cavity is also prepared with Betadine lotion. The hair is parted along

the incision line and held in place with rubber bands. The incision is made in a zigzag configuration, well behind the hairline, usually extending from ear to ear. The zigzag incision is cosmetically better. The scalp flap is reflected anteriorly in the subgaleal plane, keeping the pericranial layer intact if possible. Once the orbital rims are identified, a decision is made as to how the pericranial flaps should be based. This decision depends on the locus of the tumor and the extent of dural involvement. The usual configuration is shown in Figure 14–4. The flaps are brought into the frontal area through the burr holes in the lateral aspect of the frontal bone. The advantage of

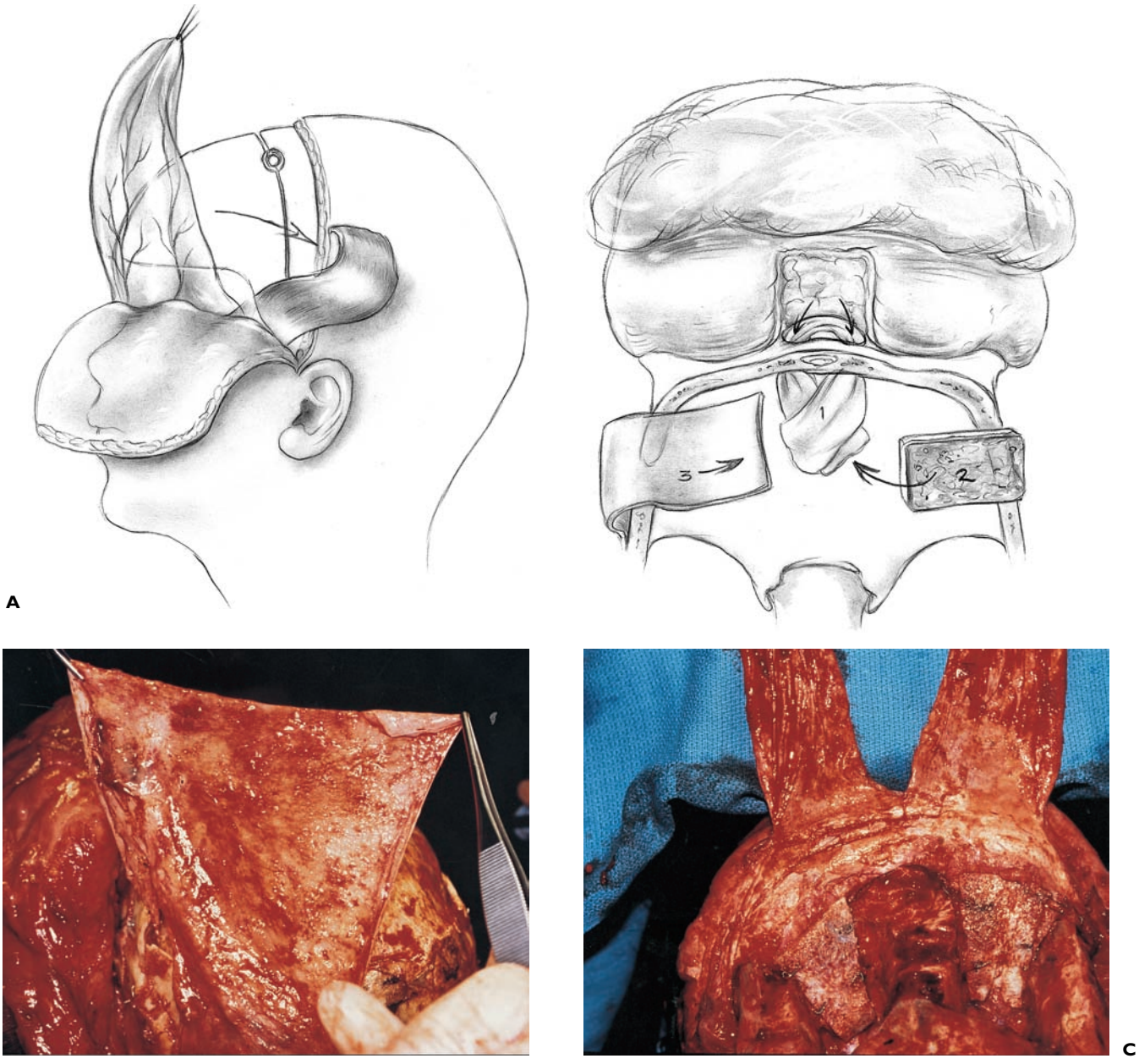


FIGURE 14–4. Pericranial flaps. **A:** The pericranial flaps which can be based anteriorly or laterally. The second drawing shows the site of entry of the flaps into the intracranial space; either through a lateral burr hole or through two medially located drill holes. 1, Galeo-frontalis flaps over ethmoid. 2, Cribriform bone graft added. 3, Pericranial flap over cribriform bone graft. **B:** Operative photograph of a large pericranial flap based on the left temporalis muscle. This is large enough to cover the whole cranial base of the frontal fossa or to swing into the sphenoid sinus to cover the temporal lobe. **C:** Operative view of the reconstructed orbital roofs in a teenaged patient with bilateral optic nerve encroachment by fibrous dysplasia. In this case, two pericranial flaps have been prepared based frontally.

this configuration is that the flaps are left attached to the temporalis muscle, and the blood supply is likely to be better preserved than when the flaps are based frontally. In addition, the temporalis muscle can be reattached to the lateral orbit to avoid a cosmetically disfiguring temporal indentation. If the pericranial flaps are brought in frontocentrally, they often produce noticeable bulging in the medial epicanthal area, which usually resolves over several months and has not required a second operation for correction of the cosmetic deformity.

A subperiorbital dissection is made in each orbit. If the supraorbital nerve is enclosed within a bony canal, a small osteotome is used to open the canal and spare the nerve. The subperiorbital dissection is carried superiorly, laterally to the lateral rim of the orbit and mesially. Mesially, the lacrimal duct must be identified and preserved along with the two heads of the medial canthus. The duct and lacrimal sac are positioned just posterior and lateral to the insertion of the heads of the medial canthal tendon. We have had more successful repositioning of the medial canthus since we started leaving it attached to the nasal bone and cutting out a small piece of the nasal bone on either side of the midline with the canthus connected to it (Fig. 14–5).

The bifrontal bone flap is now outlined. The size of the flap depends on the amount of bone required for reconstruction and the extent of the tumor. The more posteriorly and laterally the tumor extends, the larger the frontal flap. The frontal craniotomy flap also may be extended into one middle fossa if the tumor is predominantly unilateral in the orbit or lesser wing of the sphenoid. A single posterior burr hole is placed over the sagittal sinus as far posterior as necessary (Fig. 14–2A). In children younger than 10 years of age, there is rarely a frontal sinus; thus, a frontal midline burr hole is optional. If one is made, it should be placed at least 4 cm above the nasion to preserve the strength of the bone of the bandeau. As many burr holes as necessary are placed to be able to free the dura and to avoid tearing it with the craniotome. The freeing of the dura usually is done using a Gigli saw guide. The frontal flap then is excised with the craniotome, trying to bevel the edge as much as possible. The greater the bevel, the better the fit when the bone is reattached.

An epidural dissection is now made in the midline to free the dura entering the foramen caecum. The dura is freed laterally over the orbital roofs, medially to the lateral margins of the cribriform plate (Fig. 14–6) and posteriorly to the lesser wing of the sphenoid bone. The nasal dissection is carried below the nasion down to the junction of the nasal bone and the nasal cartilage. The cartilage is freed from the nasal bone, keeping the muccosa of the

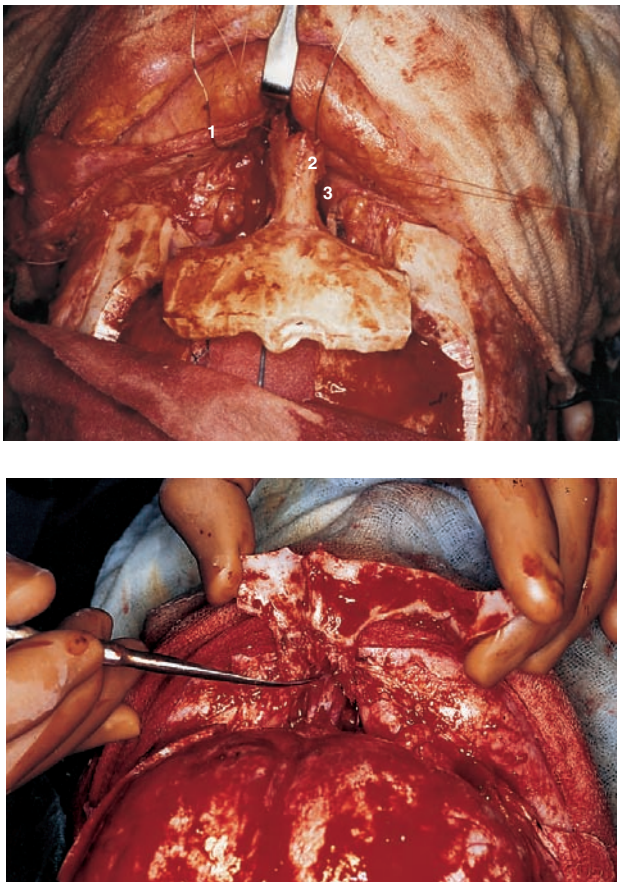


FIGURE 14–5. Removal of the orbitonasal bandeau. **A:** Demonstration of the freed bandeau showing the location of the medial canthus and the lacrimal sac and duct. 1, Medial canthus. 2, Area where bone is removed and canthus attached. 3, Lacrimal sac and duct. **B:** Dissection of the under surface of the bony bandeau from the nasal mucosa.

nose intact, if possible. Lateral saw cuts are made through the frontal bone and orbital roof. Posterior cuts are made along the medial aspect of the roof of the orbit, lateral to the cribriform plate (Fig. 14–6A). The more posteriorly the roof can be cut, the easier the reconstruction of the orbit. The ability to preserve the orbital roof depends on it not being invaded by tumor. The medial anterior skull base with the cribriform plate is left intact, and the anterior midline saw cut is made anterior to the crista galli. The anterior bandeau with the nasal bone can now be removed (Figs. 14–5A, B and Fig. 14–6B). If there are frontal sinuses within the bandeau, they must be cranialized. The nasal cavity is closed at the end of the procedure with a pericranial flap.

If the tumor does not invade the base of the skull, it is possible to leave the olfactory nerves intact by removing the midline skull base with a small cuff of

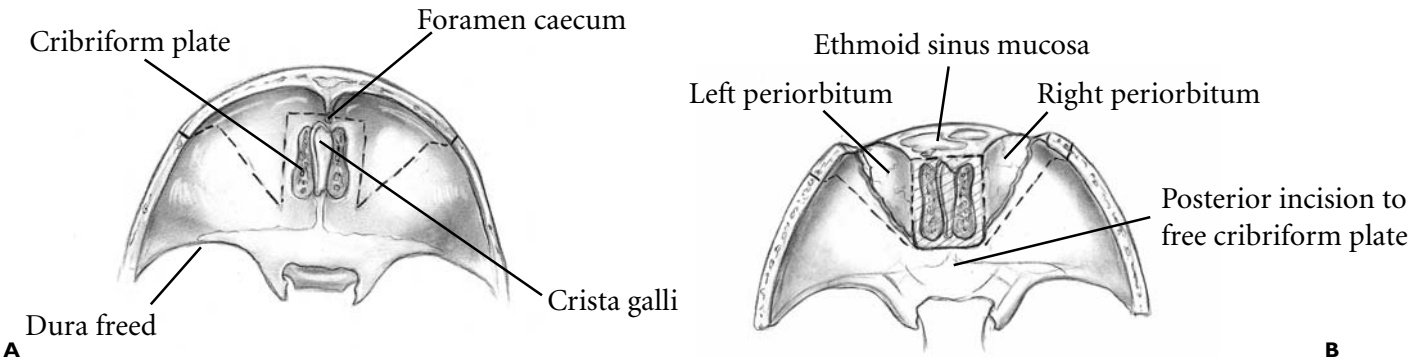
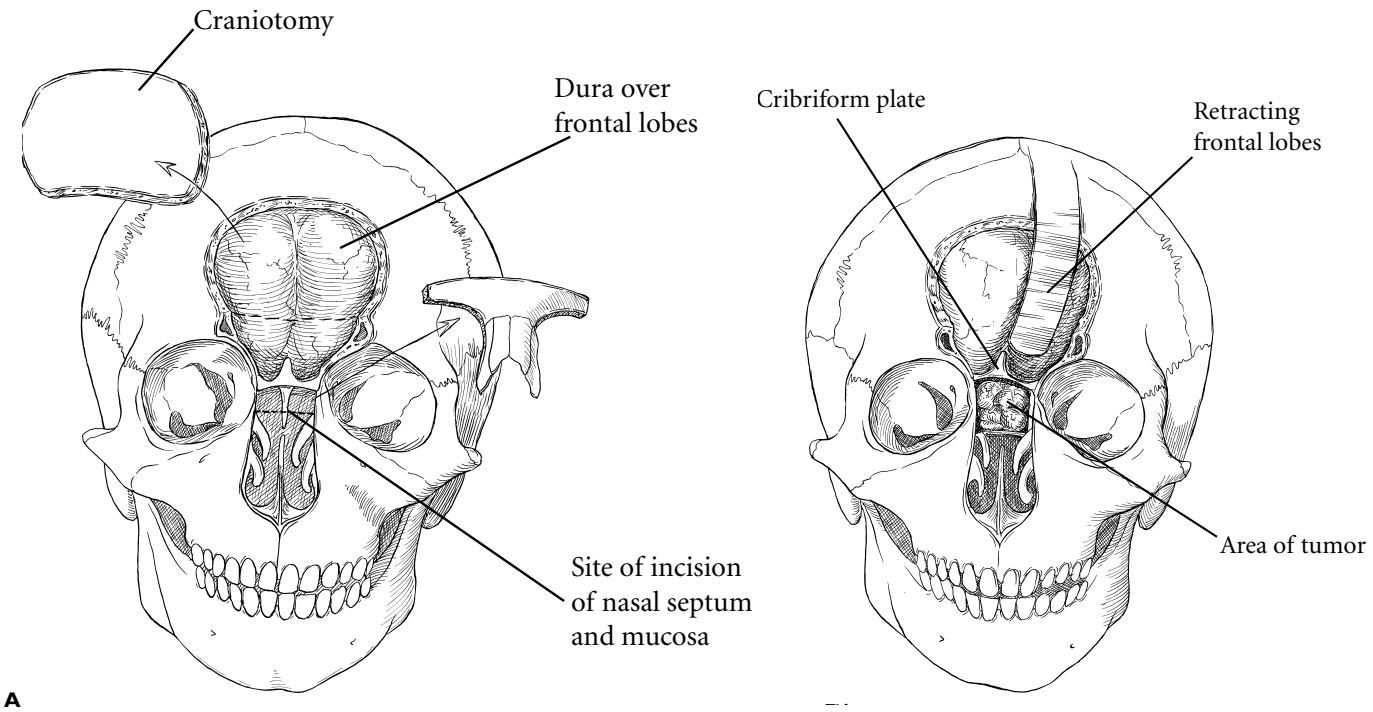


FIGURE 14–6. Illustration of the frontal skull base to show the location of the osteotomies used to remove the frontonasal bandeau. **A:** The outline in ballpoint is the area contained between the lateral and medial cuts through the orbital roofs. The anterior cut is shown going through the foramen caecum. The dark shaded area is the cribriform plate. **B:** The bandeau has been removed, leaving the cribriform plate and exposing the periorbitum bilaterally and the ethmoid sinuses. The location of the posterior cut to save olfaction is shown posterior to the cribriform plate.

nasal mucosa and septum attached. In the adult, this leaves a rather bulky area to retract, and it is not clear how well olfaction is preserved. In children, however, the extra volume of tissue is small and indeed olfaction is maintained (Fig. 14–7). The technique is to free the dura at the posterior margin of the cribriform plate and divide the bone in a transverse direction with a small osteotome. Because the periorbitum has been freed from the medial orbital wall, the location of the optic nerves can be visualized within the orbit and the transverse cut across the frontal fossa made, anterior to their exit from the apex of the orbit (Fig. 14–6B). If the skull base is invaded by the tumor, the cribriform plate dura must be freed from the bone and any defects in the dura sutured closed, either primarily or with a patch graft of pericranium. If there is dural involvement by tumor, the involved dura must be resected and the defect patched with pericranium or another substitute. When the base of the skull is invaded by tumor, the remainder of the bone of the skull base is removed using a small rongeur. The intraorbital optic nerve is identified, and the bone is removed carefully to avoid damaging the nerve. The dura at the posterior aspect of the anterior fossa, covering the optic chiasm, is thin and is easily torn while bone is being removed. If this occurs, it is difficult to suture this area, and the use of tissue glue over a small piece of Gelfoam makes a good seal.

The exposure to the superior pole of the tumor is now complete (Figs. 14–8A and B). When the tumor is large enough to present in the lateral pterygoid space, the temporalis muscle is reflected inferiorly and the zygomatic arch is divided to maximize the lateral exposure of this area. If the inferior pole of the tumor must be addressed in the maxillary sinus, a second sublabial incision is made in the upper gum to access the maxillary sinus, hard palate, and nasal cavity. In children, a Weber-Ferguson incision is rarely required. Any intracranial epidural tumor is removed first. If there is dural invasion at any point, the dura is excised and replaced. Posteriorly in the frontal fossa, it is difficult to find dura that is thick enough for suturing, and the graft often is attached using only tissue glue. The ethmoid and sphenoid tumor is now resected by going around the outside of the tumor and coagulating any feeding vessels. The tumor is debulked in most cases because it is impossible to do a block dissection of the clivus and tumor. As the superior portion of the tumor is resected, the floor of the pituitary fossa is immediately posterior to the optic nerves, and if the bone is deficient here, the gland itself can be easily damaged (Fig. 14–8B). The tumor dissection in this area must be gentle and careful. Continued dissection posteriorly and inferiorly allows resection of the tumor of the clivus down to the foramen magnum. The posterior pharyngeal mucosa usually can be preserved and the tumor removed from it. In patients with large tumors that invade or erode the lateral sphenoid sinus, the carotid arteries cannot be displaced laterally at the dural ring and are easily damaged if the bone covering them has been eroded. The location of the carotid can be identified using a small pencil Doppler. The dura covering the medial aspect of the middle fossa may be deficient, and it is important that the operator recognize the arachnoid and mesial temporal structures and not continue the resection into brain tissue. If the arachnoid is intact, it is extremely important to avoid opening it because the risk of a cerebrospinal fluid (CSF) leak will be greatly increased. This finding of absent middle fossa dura has occurred with large pituitary tumors and angiofibromas. At the time of closure, a large peri-



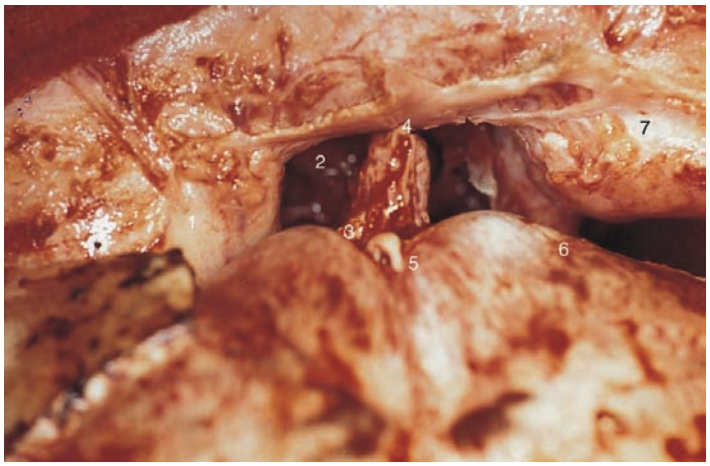


FIGURE 14–7. Saving olfaction. **A:** Drawing of anterior operation after removal of the craniotomy and orbiyonasal bandeau. The cribriform plate is shown at the superior aspect of the nasal septum and the dotted line shows the locus of the incision into the nasal mucosa and septum. **B:** Frontonasal region and cribriform plate being retracted to get exposure of the tumor **C:** Operative photograph of the cribriform plate complex after saving olfaction. The dura that was in the foramen cecum is well seen. In the depth of the central cavity is the tumor in the posterior ethmoids and sphenoid sinuses. 1, Left orbit. 2, Region of ethmoid sinus and nasal cavity and tumor. 3, Bone of the cribriform plate. 4, Superior nasal septum and mucosa. 5, Dura from the foramen caecum. 6, Frontal dura. 7, Right orbit.

cranial flap is brought in to cover the temporal lobe and line the cavity. There is nothing to suture to, and the graft is held in place with tissue glue.

Once the posterior and superior portions of the tumor are removed, the tumor within the maxillary sinus, inferior orbit, and nose is removed. In cases of malignant tumors, the maxilla, upper jaw, and orbit all can be resected if they are involved by tumor. When such a massive resection is needed, autologous bony reconstruction is rarely possible because of the lack of tissue to cover the grafts. Usually, adequate reconstruction of the orbit is possible, and the grafts can be covered by rotating the temporalis muscle into the face to cover the orbital grafts. The maxilla and jaw usually are replaced by a temporary prosthesis to maintain the soft tissue in the correct position and to support the upper airway. The most

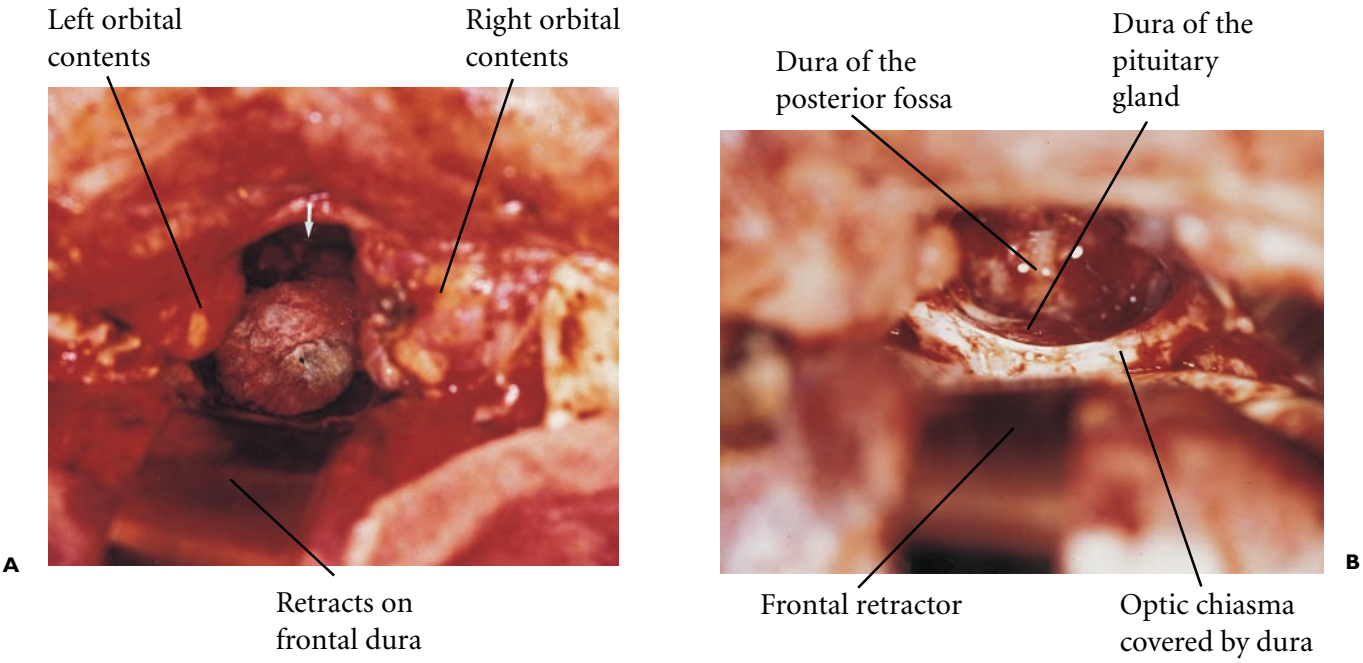
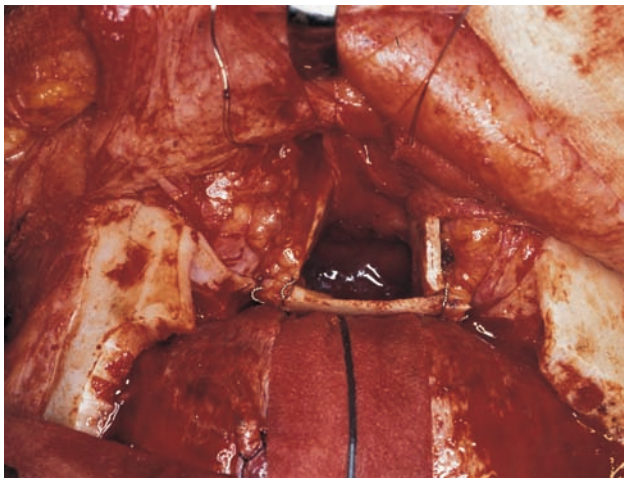


FIGURE 14–8. View of tumor, optic chiasma dura, and pituitary fossa dura after the anterior skull base is removed. **A:** Appearance of the dome of a giant cell tumor of bone after skull-base removal. The left and right orbital contents are seen covered by periorbitum. Frontal lobe retraction is minimal. Anterior to the tumor is the nasal cavity. Arrow, dome of tumor in sphenoid and ethmoid sinus. **B:** Appearance after tumor resection. The dura covering the optic chiasma is in the anterior aspect of the field. Immediately posterior to this, the red area is the dura covering the pituitary gland. The most posterior white, shiny tissue is the dura of the posterior fossa. The clivus was destroyed by the tumor.

likely areas to leave residual tumor are superior and posterior to the posterior clinoid process. This area can be visualized by using the endoscope. The second area is the posterior pharyngeal mucosa.

In most cases, the maxilla and nasal bones are not removed, and the reconstruction involves the orbits, skull base, and sometimes the anterior maxillary wall. The bone for reconstruction is harvested from the inner table of the frontal bone flap and, if necessary, from a second more posterior craniotomy flap. The first area to be reconstructed is the orbit and then the medial orbital wall, roof, and floor, if necessary (Fig. 14–9). The next area is the skull base and the clivus if it has been resected. When the skull base is replaced, it is critical to avoid any pressure on the optic nerve or chiasm by the replaced bone. When the clivus needs reconstruction, a single bone graft is fashioned that will cover the clivus and frontal fossa. (Fig. 14–9). When the cribriform plate and olfaction have been spared, the midline reconstruction stops at the posterior margin of the cribriform plate bone. The nasal mucosa attached to the cranial bone is resutured to the upper nasal septum. The pericranial flap is now used to cover the intracranial area to seal off the intracranial space from the nasal cavity and sinuses (Fig. 14–9B); again, this is sealed with tissue glue. The nasal bandeau is reattached and the nasal cartilage sutured to the nasal bone. The medial canthi then are reattached by passing an awl transnasally and wiring the canthi, which is still attached to the small island of bone, back into position. The tendency is to position the canthi too anteriorly. Nothing is done to eradicate or fill the posterior pharyngeal space, which is left after tumor resection. Some of the accumulated fluid will drain through the sphenoid antrum into the nose. We have never had this area become infected. The two critical aspects of the closure are to get the correct orbital volume such that there is neither enophthalmus or exophthalmus and to seal the intracranial space from the extracranial air spaces. Once the basal reconstruction is complete, the skull flap is replaced usually using PDS suture. The burr holes are filled in with bone dust saved during the opening of the craniotomy. The temporalis muscle is reflected anteriorly to avoid any temporal dents, if possible, and the skin is closed with Vicryl in the galeal layer and usually 5′0 monocryl in the skin. No drains are left, the hair is washed at the end of the procedure, and polysporin ointment is applied to the incision. If there is a sublabial incision, it is closed with catgut.



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B

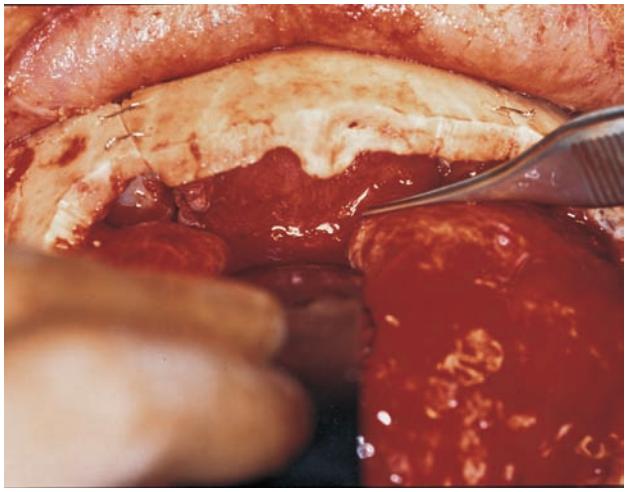

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FIGURE 14–9. Bony reconstruction. **A:** Bony reconstruction of the skull base and medial orbital walls (the cottonoid is on the frontal dura). **B:** The reconstructed skull base is shown. The bandeau has been replaced. We would not use wire to hold the bone in place now; rather, we would use either PDS suture or dissolvable plates. **C:** View from the intracranial space after the pericranial graft is in place over the reconstructed skull base.

POSTOPERATIVE CARE

We do not leave drains and do not place any dressings on the incision. The ET tube usually is removed at the end of the procedure because there is rarely encroachment into tissue below the palate when this approach is used. In infants in the first year of life the ET tube is usually left in place because, at this age, infants are nasal breathers and the nasal passages are likely to be blocked or poorly patent following such surgery. This approach is rarely used in infants because of the location of the common tumors in this age group. In rare cases where it is used, the children usually present with nasal obstruction and thus have already learned to breath by mouth. Nonetheless, it is safer to leave the ET tube in place until the infant is fully awake and clearly capable of managing the airway.

When the pituitary gland has been displaced by the tumor or when the surgery involves manipulation of the pituitary dura, diabetes insipidus (DI) can occur. Thus, careful recording of urine output compared with intake, frequent urine specific gravity measurements, and serum sodium and osmolality are necessary. In addition, daily body weights should be taken. It is fairly common to have some degree of inappropriate antidiuretic hormone (ADH) production after any subfrontal procedure. Thus, the oscillation between too much and too little ADH can make the postoperative fluid balance difficult to control. Hyponatremia and decreased serum osmolality are a greater risk to the patient than moderate hypernatremia. The risks of brain edema and seizures are much greater in the former than in the later state. Therefore, the use of desmopressin (DDAVP) in the postoperative period must be controlled carefully and should occur only when the presence of DI is

well documented and the serum sodium is above 150 mg/100 mL. The lowest dose necessary to increase urine concentration and stabilize the serum sodium is the ideal dose. If urine replacement has been the treatment for the increased urine output, there should be no further intravenous replacement of urine output after the dose of DDAVP is given; otherwise, a sudden drop in serum sodium can occur with resultant brain swelling, seizures, or both.

Because of the extensive bifrontal scalp flap and potential bleeding into the tumor cavity, retropharynx, and sinuses, the hemoglobin and hematocrit must be monitored for the first 24 hours in case additional blood replacement is necessary. Also, disturbances of the clotting factors can occur postoperatively and should be checked at least once in the first 24 hours and again if excessive bleeding occurs. Significant postoperative swelling almost always occurs, and the eyes become closed. The children and parents should be informed of this problem prior to surgery to know that it is the normal response, and not of concern. Drainage from the nose, especially when the tumor has involved the sinuses, is common. This drainage will be glucose positive because it is serum, not mucous. The risk of CSF leakage should be easily judged based on the intraoperative findings. In most cases, the dura is not entered other than at the cribriform plate, and it is rare that lumbar drainage is necessary because the glucose-positive drainage is usually not CSF. Routine lumbar drainage is not advised either intraoperatively or postoperatively. During surgery, it is rarely necessary and, when used, makes it harder to identify small dural tears. In the postoperative period, it encourages formation of an epidural dead space, and entrance of air from the nasal cavity into the epidural space and it can result in retroalar or transtentorial herniation from excess drainage. Lumbar drainage should only be used in cases where CSF leakage is seen at surgery and cannot be satisfactorily stopped, such as with deficient middle fossa dura.

EDITOR'S COMMENTARY

The skull base is the site of origin of a diverse group of benign and malignant tumors that require highly individualized approaches to treatment. It is important to recognize which lesions are best managed by aggressive cranial base resections and which are more appropriately controlled using nonsurgical approaches. After a decision is made to proceed with resection, a thoughtful deliberation regarding the optimal surgical approach is mandatory, which should take into account the pattern of growth of the tumor as well as the relevant anatomical features of the developing craniofacial skeleton. Despite significant advances in the surgical management of many of these lesions, certain tumor types remain difficult to cure, particularly chordomas and aggressive sarcomatous lesions, highlighting the need for further improvements in the adjuvant treatment of these neoplasms.

PEARLS

In this author's experience:

- Surgery may be a major therapy for many of the less common malignant tumors, especially low-grade sarcomas.
- Most of the surgically treatable and potentially curable benign lesions are extradural in origin, and the first surgical approach almost always should be by the extradural route.

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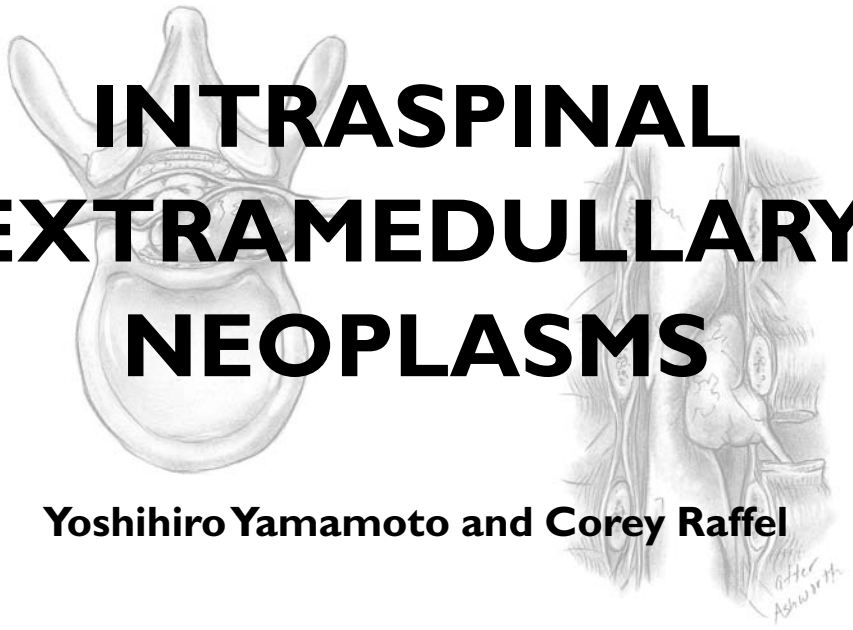
15

INTRASPINAL EXTRAMEDULLARY NEOPLASMS



Yoshihiro Yamamoto and Corey Raffel

In children, tumors of the spinal canal constitute a smaller proportion (5 to 10%) of tumors of the central nervous system than in adults. The various types of tumors that occur in children also differ from those seen in adults. Intradural extramedullary tumors, such as meningiomas, schwannomas, and neurofibromas, the most common tumors in adults, are rarely found in children.

The most common acquired, nontraumatic cause of paraparesis in children is compression of the spinal cord by malignant epidural tumor deposits. These deposits differ from those found in adults in the histologic and biologic features of the tumors involved, the location and direction of compression, the degree of bone involved, and the general medical condition of the patient.

Table 15–1 summarizes the various types of neoplasms, both intramedullary and extramedullary, found in the spinal canals of 635 pediatric patients in 10 large series published between 1953 and 1990 (see Yamamoto and Raffel, 1999, for references). Myxopapillary ependymoma of the filum terminale and cauda equina, a rare neoplasm that appears in children as a extramedullary lesion, is underrepresented in these previous series.

Thirty-five percent of neoplasms in the spinal canals of children are located extradurally. Table 15–2 summarizes the tumor pathologies that caused epidural compression of the spinal cord in children in four series (see Yamamoto and Raffel, 1999 for references). It has been suggested that 3 to 5% of children with a systemic malignant tumor have a compressed spinal cord sometime during their illness. In many of these children, compression of the spinal cord is the first sign of malignancy.

The rarest spinal lesions in children are diffuse subarachnoid, or leptomeningeal, tumors, which usually stem from the dissemination of a posterior fossa tumor, such as a primitive neuroectodermal tumor. Tumors of the posterior fossa rarely present with spinal symptoms; nonetheless, about 20% of primitive neuroectodermal tumors in the posterior fossa have already disseminated in the cerebrospinal fluid at presentation. Leptomeningeal tumors cannot be treated surgically. A surgical approach is indicated only if the patient's diagnosis is unknown.

PREOPERATIVE EVALUATION AND SURGICAL INDICATIONS

Signs and Symptoms

In patients with extramedullary spinal tumors, weakness and pain are the most common symptoms. The weakness is usually of the upper motor neuron type and is associated with increased tone and hyperactive deep-tendon reflexes. If the conus medullaris or cauda equina is involved, the weakness may be flaccid. In infants and toddlers, the weakness may be subtle and therefore difficult to detect.

Pain is an important early symptom in children with spinal tumors. Pain occurs most commonly in the back

TABLE 15–1.
Tumor Type in 635 Pediatric Patients in 10 Large Series[a]

Location/Tumor Type	*No. of Patients (Percent)*
Intramedullary	189 (29.7)
Astrocytoma	114
Ependymoma	50
Lipoma	25
Intradural extramedullary	156 (24.6)
Dermoid	39
Neurofibroma	28
Schwannoma	20
Meningioma	17
Epidermoid	14
PNET[b]	30
Hemangioepithelioma	8
Extradural	219 (34.5)
Sarcoma	67
Neuroblastoma	64
Teratoma	35
Metastasis	29
Ganglioneuroma	19
Lymphoma	5
Other	71 (11.2)

[a]For references, see *Principles and Practice of Pediatric Neurosurgery.*
[b]PNET, primitive neuroectodermal tumor. Some of the teratomas, neurofibromas, and dermoid tumors arose from both intradural extramedullary and extradural compartments.

and is reported by 28 to 59% of patients. Depending on the level of the lesion, some children have radicular pain radiating into the arm or leg or around the chest wall. Radicular pain is reported most often by patients with intradural extramedullary and extradural lesions. In a child with known malignant disease, back or radicular pain requires immediate evaluation to determine whether the spinal cord is compressed. Similar pain in a child with no known malignancy also indicates a careful evaluation to determine the cause.

Sensory disturbances are also common in patients with spinal tumors but are difficult parameters to determine in the young patient.

Bladder or bowel dysfunction is also common, occurring in about half of patients with severe compression of the spinal cord. A loss of bladder control in a toilet-trained child should raise the suspicion of a spinal lesion.

An abnormal curvature of the spine occurs in about one fourth of children with spinal tumors and may be the initial sign that indicates a lesion. In any child with a progressive abnormality of spinal curvature, the diagnosis of intraspinal neoplasia should be considered.

TABLE 15–2.
Epidural Tumor Type in 246 Pediatric Patients in Four Large Series[a]

Tumor Type	*No. of Patients (Percent)*
Neuroblastoma	64 (26.0)
Ewing's sarcoma	52 (21.1)
Rhabdomyosarcoma	31 (12.6)
Osteogenic sarcoma	29 (11.8)
Lymphoma[b]	19 (7.7)
Undifferentiated sarcoma	12 (4.9)
Germ cell tumors[c]	12 (4.9)
Leukemia	7 (2.8)
Wilms' tumor	4 (1.6)
Other	16 (6.5)

[a]For references, see *Principles and Practice of Pediatric Neurosurgery.*
[b]Hodgkin's and non-Hodgkin's.
[c]Embryonal cell carcinoma, endodermal sinus tumor, and teratoma.

Radiological Studies

Because of the increased anatomic detail gained without the need of a lumbar puncture, magnetic resonance (MR) imaging with and without intravenous contrast is preferred over computed tomography (CT) myelography to identify spinal cord neoplasms. Avoiding a lumbar puncture eliminates two important risks: (1) bleeding from thrombocytopenia or coagulopathy associated with some malignant diseases, and (2) accelerating the rate of neurologic deficit through the loss of cerebrospinal fluid. In our experience, MR imaging is at least as sensitive as myelography combined with CT, and it provides superior anatomic details of tumors both inside and outside the spinal canal. Administration of Gd-DTPA aids in the identification of intradural–extramedullary nodules as small as 2 to 3 mm and easily depicts the leptomeningeal spread of tumor.

Plain radiographs may help to define the region of the spine to be scanned because they reveal abnormalities in half of patients with spinal cord tumors. Intradural extramedullary tumors can thin or cause sclerosis of the pedicles. A tumor that extends through a neural foramen enlarges that foramen, which is best visualized on oblique films. An enlarged foramen indicates a tumor of the nerve sheath or a paraspinous tumor that enters the canal at that level. Malignant tumors of the bone and epidural metastases erode bone and may cause a vertebral body to collapse. Paraspinous masses may be seen on plain radiographs in association with an enlarged foramen or bony destruction. Although MR imaging is usually the initial imaging study done, plain radiographs may be helpful in defining bony anatomy and alignment and also

can serve as a baseline for comparison if the patient develops scoliotic changes after treatment.

Previously, myelography with water-soluble contrast material was used extensively in the evaluation of suspected intraspinal lesions in children; however, sudden deterioration after a lumbar puncture has been reported in patients with intraspinal neoplasms. Therefore, myelography should be reserved only for patients who require an emergent study when MR imaging is not immediately available. Other occasions include a patient with severe scoliosis, which makes it difficult to interpret MR findings.

In such cases, CT scanning after intrathecal water-soluble contrast administration is an important adjunct to myelography for imaging spinal tumors. Because a small amount of contrast medium may pass around a high-grade block seen on a myelogram, a CT scan can often define the upper extent of the lesion without the need for a second injection of contrast material above it. CT scanning with intrathecal contrast is more sensitive than myelography alone in detecting small drop metastases and lesions in the epidural space. CT scans delineate the bony anatomy especially well; thus, they are the best choice for determining the degree of bone destroyed by an invasive tumor.

Surgical Indications

Surgical decompression is indicated for patients with severe compression of the spinal cord, regardless of the type of tumor; for symptomatic patients newly diagnosed with sarcomas; for patients with symptomatic compression who have no diagnosis; and for patients whose neurologic function deteriorates during nonsurgical therapy.

The treatment of intradural extramedullary tumors is primarily surgical. These tumors are usually nerve sheath tumors or meningiomas, which are separated from the surrounding tissue by a distinct margin. This margin allows total excision in almost all patients. Some nerve-sheath tumors extend into the extradural space, through the neural foramen, and into the paravertebral soft tissue. In these cases, the surgeon may need to carry out a second extraspinal resection to remove the remaining tumor.

Extradural spinal neoplasms causing spinal cord compression, mostly "small cell blue" tumors, such as neuroblastoma and lymphoma, can be treated successfully with chemotherapy or radiation therapy. On the other hand, most sarcomas require operative decompression. The goal of decompression is not to remove all tumor, but to decompress the cord. Adjuvant therapy is indicated in almost all instances.

PREOPERATIVE MANAGEMENT

Each patient must be evaluated carefully for possible instability of the spine caused by destruction of the spinal column and paraspinal structures. This evaluation should be done both clinically and radiologically. If the patient has a known primary malignancy, preoperative staging with radiological studies is required to assess possible metastatic lesions in other organs. Bleeding tendencies associated with malignant disease should be carefully corrected before surgery.

OPERATIVE PLANNING

For a patient with significant spinal cord compression, perioperative use of methylprednisolone can be considered, although there are no scientific data to support this to date. Intraoperative monitoring of somatosensory-evoked potentials (SSEP) is used by some surgeons, but there is controversy about whether the monitoring can give up-to-the-moment feedback and whether the surgeon must modify the procedure according to changes in the SSEP. We do not usually use SSEP at our institute. If the lesion is expected to be extremely vascular, blood for a transfusion should be prearranged according to the patient's body weight. We often use intraoperative ultrasound to localize intradural extramedullary lesions as well as intraoperative radiography to confirm the level of bone work.

INTRAOPERATIVE TECHNIQUES

For small children, minimizing blood loss during any surgery is crucial. We always use a hot knife (Shaw Scalpel, Oximetrix, Mountain View, CA) and a bipolar coagulator of an appropriate size. Respecting anatomic dissection planes is helpful in preserving normal vasculature as well as minimizing blood loss. Laminectomy is usually carried out using different shapes of rongeurs, kerrison punches, or a high-speed drill with diamond bits (Figs. 15–1A and B). Care must be taken to avoid compressing the spinal cord further. Laminotomy can be done using the Midas Rex (Medtronic Midas Rex, Fort Worth, TX) with the footed attachment (Fig. 15–1C). This must be done carefully so as not to compromise the already restricted spinal canal.

Intradural Extramedullary Tumors

The surgical approach for intradural extramedullary tumors in children does not differ from that used in adults. Most meningiomas, neurinomas, myxopapillary ependymomas, and dermoids are operated on through a laminectomy either with or without laminoplasty (see section on surgical complications). Intraoperative ultrasound can be useful in localizing the extension of the tumor and minimizing the levels of the laminectomy. The microscope and microdissection tools are indispensable for preserving normal neural structures. In some instances, electrophysi-

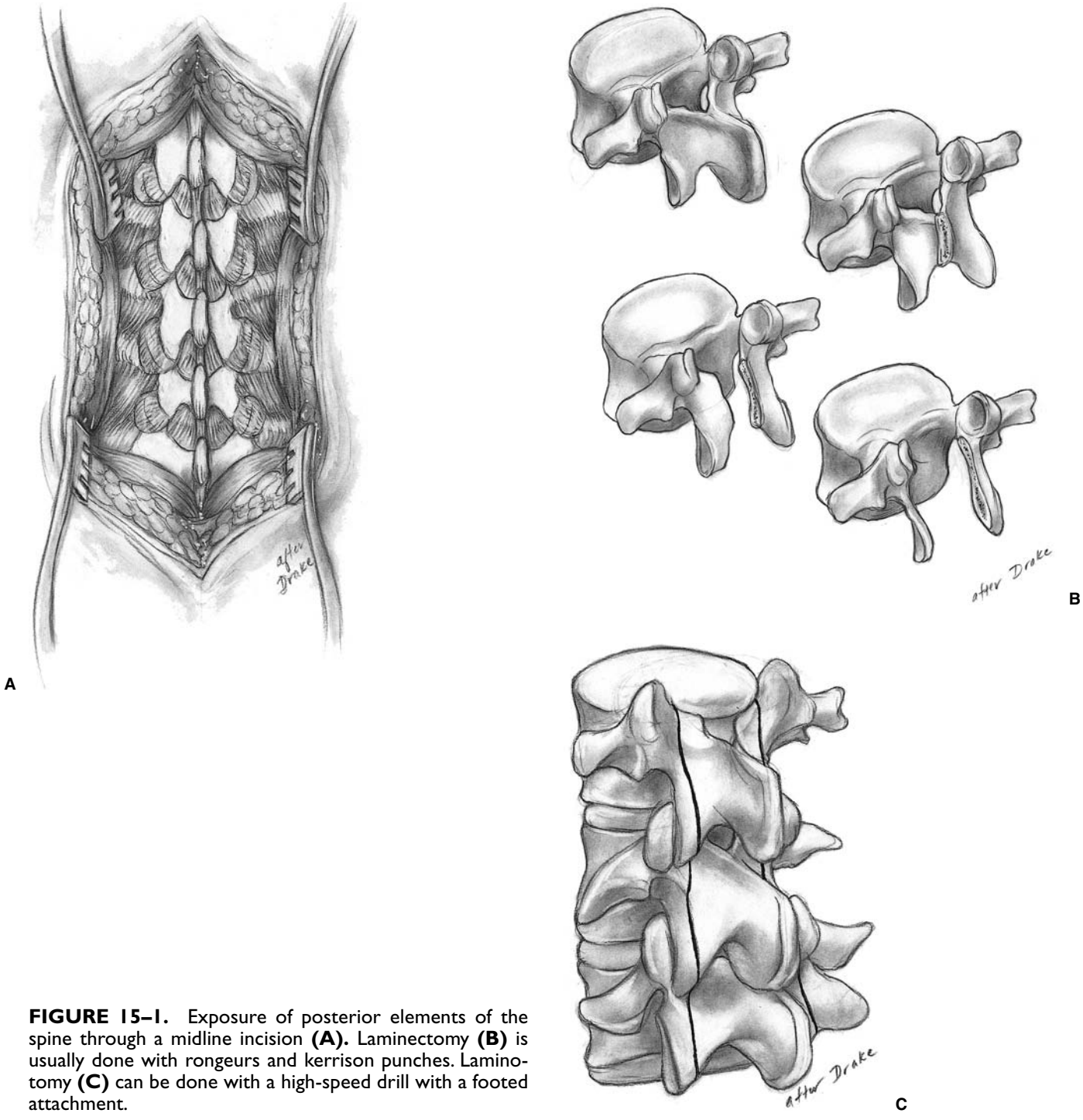


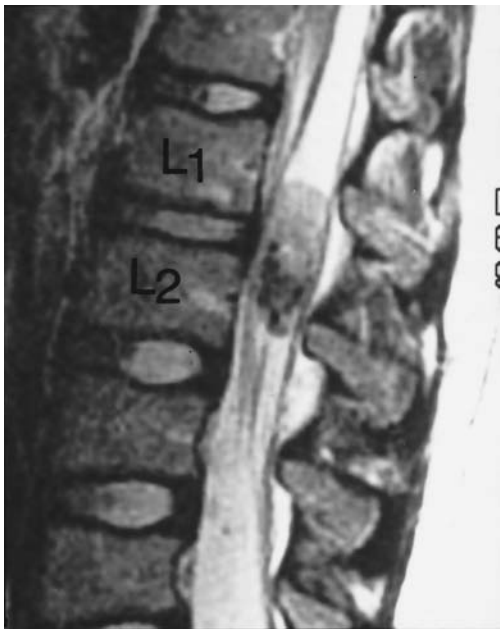
FIGURE 15–1. Exposure of posterior elements of the spine through a midline incision **(A).** Laminectomy **(B)** is usually done with rongeurs and kerrison punches. Laminotomy **(C)** can be done with a high-speed drill with a footed attachment.

ologic monitors, such as SSEP to monitor dorsal column function during dissection or electromyography to identify nerve roots, offer important information. Myxopapillary ependymomas of the filum terminale and conus medullaris also can be removed surgically. Meticulous attention should be paid to preserving the tumor capsule because any spillage of tumor cells into the subarachnoid space can be a source of recurrent tumor or spinal dissemination. Radiation therapy of disseminated myxopapillary ependymomas has been unsatisfactory.

In our case 1, a 13-year-old boy presented with an acute onset of low back pain and leg spasm after he "plopped" himself down on a chair in a movie theater. MR imaging of his spine revealed an inhomogeneously enhancing lesion at the conus medullaris (Fig. 15–2A). Laminectomy of L1-3 was carried out, and intraoperative ultrasound confirmed the location of the extramedullary lesion. Gross total removal of the lesion was achieved under the microscope without violating the tumor capsule (Fig. 15–2B). Pathological examination showed a myxopapillary ependymoma with focal intratumoral hemorrhage. The postoperative MR study of the total spine with and without gadolinium enhancement showed no tumor dissemination or residual tumor. The patient left the hospital without pain or neurologic deficit.

Benign Extradural Tumors

Children with neurofibromatosis sometimes present with progressive scoliosis. Workup should be directed to identifying the neurofibroma or other neoplastic process of the spinal cord or roots. If the lesion is causing a neurologic deficit or deformity, or if it increases in size on serial imaging studies, most neurosurgeons recommend surgical removal of the lesion (Fig. 15–3). Another



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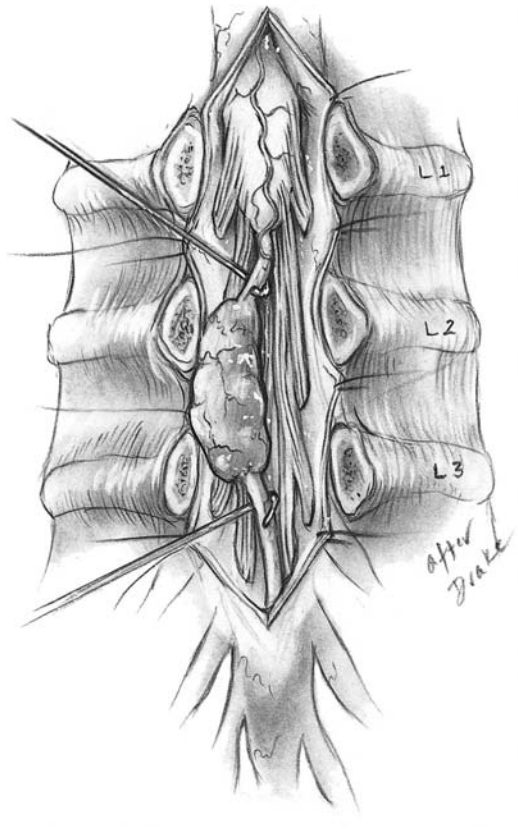
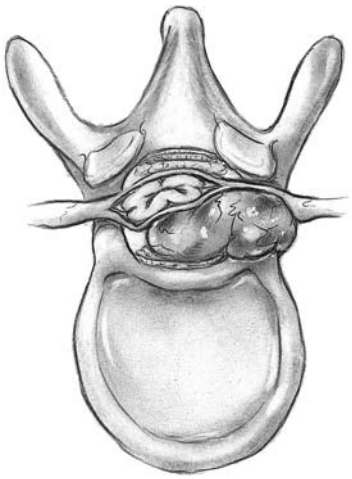


B

FIGURE 15–2. This 13-year-old boy developed an acute onset of low back and bilateral leg pain when he "plopped" himself down on a chair in a movie theater. Neurologic examination revealed no motor weakness or sensory changes. MR imaging of the spine **(A)** showed an irregularly enhancing lesion at the L1-2 level. Gross total removal of the lesion was achieved through a posterior approach without violating the capsule. The lesion was attached to the filum terminale **(B)**. Pathologic examination showed the tumor to be a myxopapillary ependymoma.



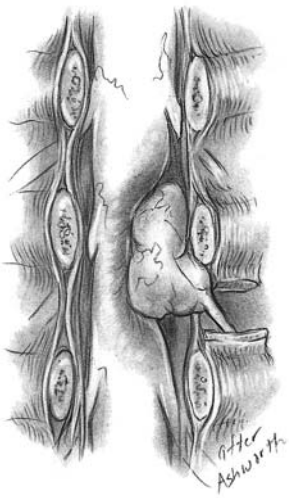


FIGURE 15–3. The typical appearance of a neurofibroma originating from the dorsal nerve root and expanding the foramina.

indication for surgery is to rule out a neurofibrosarcoma developing in the preexisting neurofibroma when the lesion is progressively enlarging or showing worrisome imaging characteristics.

The origin of the Adamkievicz artery should be suspected if the lesion is on the left side at the level of T-10 to L-2. This location sometimes requires a preoperative angiogram to identify the artery. The artery should be protected to prevent an anterior spinal cord infarct.

In case 2, a 3-year-old girl, whose father had neurofibromatosis type 1, was found by her parents to have progressive scoliosis (Fig. 15–4A). A large plexiform neurofibroma of T8 through 10 on the right side was found on CT and MR scans (Figs. 15–4B thorugh D). The MR scans showed inhomogeneous enhancement of the lesion, suggesting possible sarcomatous transformation. A costo-transversectomy was done to approach this lesion lateral to the thecal sac and to remove the extracanalicular plexiform neurofibroma. Pathological examination showed no evidence of sarcomatous changes. The patient developed progressive worsening of the scoliosis, requiring instrumentation with Harrington rods.

Malignant Extradural Tumors

Usually, epidural compression in children with malignant tumors is caused by direct extension of the tumor through the vertebral foramen without significant involvement of bone. Thus, the mass compressing the cord is lateral to the vertebral body and can be reached easily by a posterior approach, either a laminectomy or laminotomy. The goal of decompression is to relieve the pressure on the spinal cord and is accomplished through the removal of as much tumor as possible. A gross total resection can be done only rarely.

Case 3 involved a 5-month-old male infant who was brought to his pediatrician with a history of chronic cough. His neurologic examination was completely normal. A chest radiograph showed a large mass in the posterior mediastinum on the right side (Fig. 15–5A), and a CT scan clearly showed the mass extending through the neural foramen into the spinal canal. The patient's vanillylmandelic acid (VMA) level was elevated, giving a probable diagnosis of neuroblastoma. MR studies of the spine with T1-weighted images showed the mass filling more than 50% of the canal at the T-10 level with tumor extending to T-9 and T-11 levels (Figs. 15–5B and C). A laminectomy was done to expose the epidural tumor (Fig. 15–5D), which was removed until the foramina and paraspinal muscles were reached. Pathological examination confirmed this tumor to be neuroblastoma. Later, the main bulk of the lesion was gross-totally removed by a pediatric surgeon.

POSTOPERATIVE MANAGEMENT

After surgery, the child should be closely monitored in the pediatric intensive care unit for worsening of cord

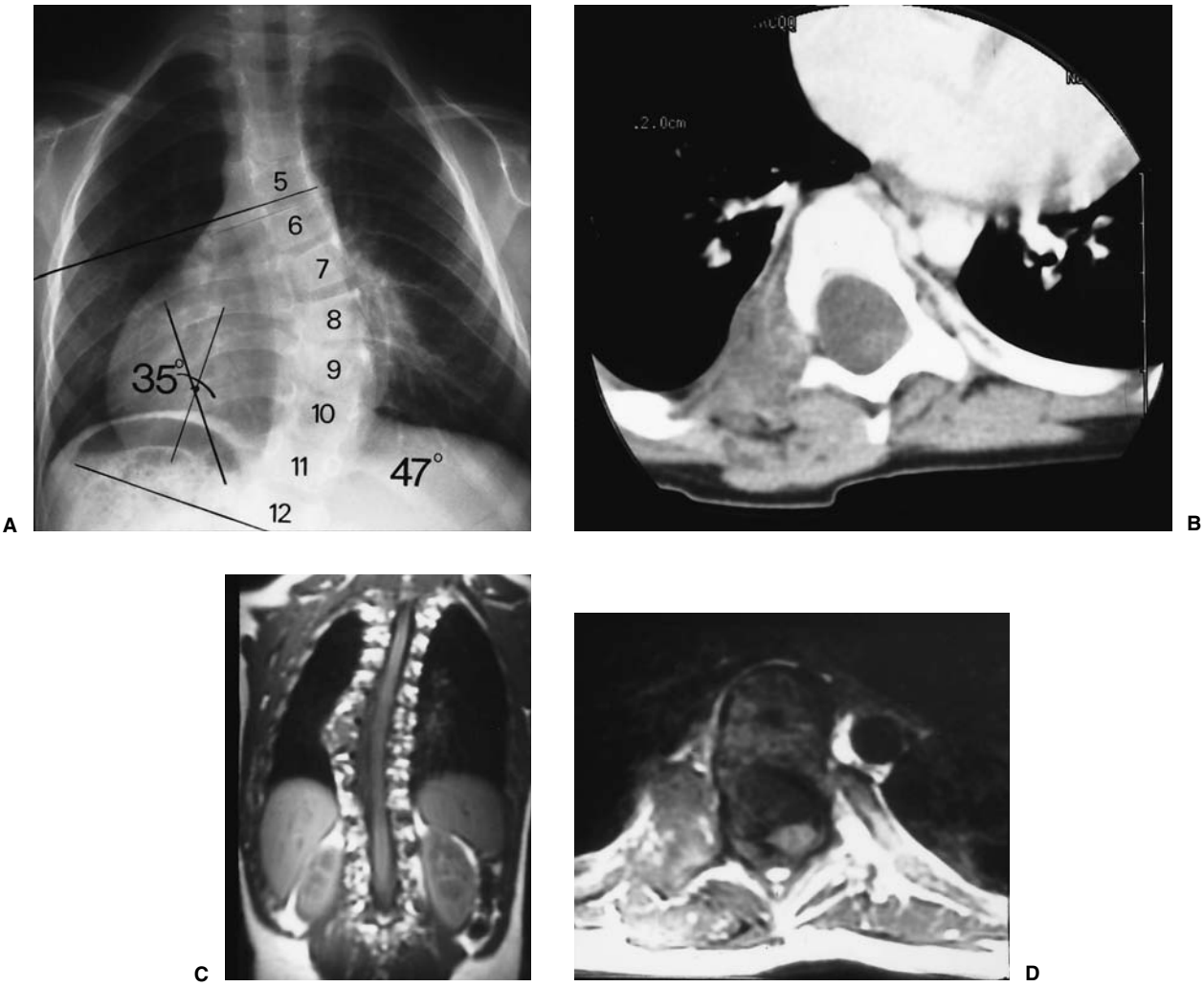
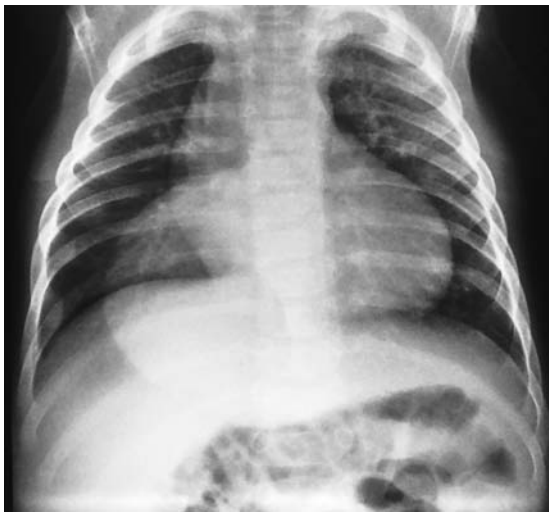


FIGURE 15–4. This 3-year-old girl's father had a history of neurofibromatosis type 1. Her parents noted her progressive scoliosis. The spine radiograph viewed from the back **(A)** showed a 35-degree right-side convexity curve. It also showed thinning of the pedicle at T-7, T-8, and T-9 on the right greater than on the left. Her body CT scan **(B)** and MR images **(C,D)** revealed an epidural mass extending into the paraspinous area at multiple levels from T-7 down to T-10. This lesion was removed subtotally through a costo-transversectomy approach. Pathologic examination confirmed a plexiform neurofibroma without evidence of sarcomatous transformation. The girl developed progressive scoliosis after the resective surgery and required fusion with instrumentation.

compression signs, which could be due to postoperative bleeding. If the child has underlying malignancy, coagulation studies should be repeated, and abnormalities should be corrected as needed.

A careful histologic examination must be made of neurofibromas removed from patients with neurofibromatosis, as some of these tumors contain malignant elements. If indicated, postoperative MR studies should be done within 48 hours after surgery to minimize postoperative changes and to identify any residual tumor. Only rarely did we reoperate to attempt more extensive resection because most pediatric extramedullary lesions cannot be cured with surgery alone. Spine radiography usually is repeated periodically for patients with questionable spinal stability or progressive spinal curve.



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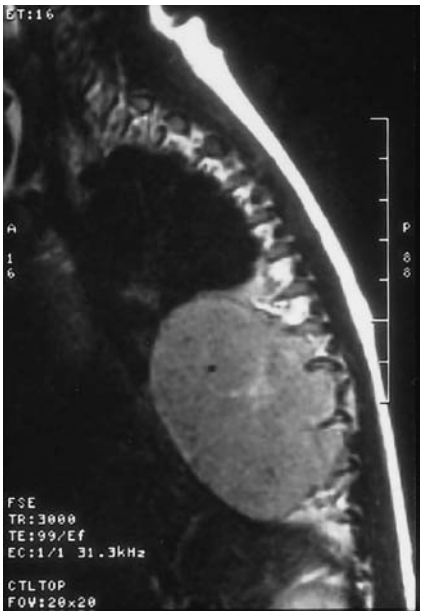

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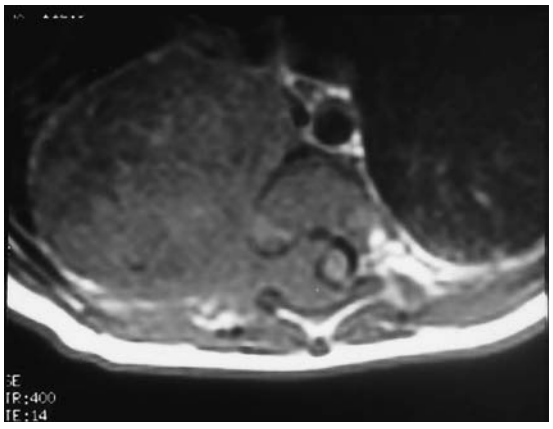

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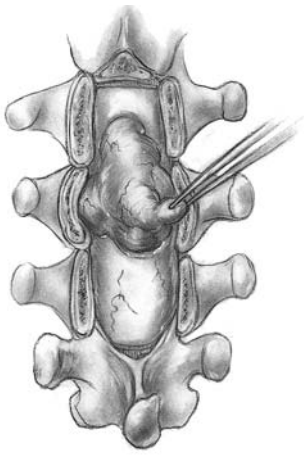

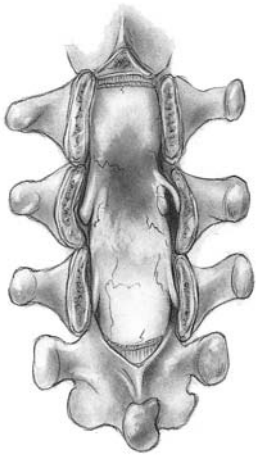

D

FIGURE 15–5. A 5-month-old boy presented to his pediatrician with a chronic cough. A chest radiograph showed a large right paraspinal and posterior mediastinal mass **(A).** His VMA level was elevated, giving a diagnosis of probable neuroblastoma. The T1-weighted image of the MR study showed the mass filling more than 50% of the canal at the T-10 level, with tumor extending to T-9 and T-11 levels **(B,C)**. Laminectomy was done to expose the epidural tumor, which was removed until the foramina and paraspinal muscles were reached. Pathological study confirmed this tumor to be a neuroblastoma. **(D)** The schematic appearance of the tumor before and after partial removal. Later, the main bulk of the lesion was gross-totally removed by a pediatric surgeon.

COMPLICATIONS OF TREATMENT

Although the immediate complications of surgical decompression are usually negligible, a delayed deformity of the spine regularly occurs. In early series of patients with intraspinal tumors treated with laminectomy, cervical kyphosis, anterior subluxation, and disabling scoliosis were reported as late complications. The incidence of spinal deformity after multilevel laminectomy is related to the patient's age and to the spinal level of the laminectomy.

The use of laminotomy to prevent deformity is controversial, and further study is needed to determine the effect of this technique on late spinal deformity. Other authors, however, are not convinced of the superiority of laminotomy.

Spinal deformity is also a complication of radiation therapy used to treat epidural tumors. A higher rate of deformity is associated with a young age at the time of radiation, doses of radiation greater than 20 Gy, and asymmetric radiation fields.

Because of the spinal deformity seen in patients undergoing laminectomy or radiation therapy, it is not surprising that combining radiation with laminectomy predisposes patients to the late occurrence of deformity. Thus, in children with an epidural tumor without evident cord compression or neurologic deficit, both surgery and radiation therapy should be avoided. If possible, a child with an epidural tumor should undergo chemotherapy alone.

EDITOR'S COMMENTARY

Most intradural extramedullary tumors in children are benign lesions that are best treated with surgical resection, the notable exception being a patient with neurofibromatosis with multiple small, asymptomatic lesions, which are best managed expectantly. Subarachnoid tumors usually represent dissemination of disease from a primary site elsewhere within the neuraxis and surgical intervention is rarely indicated for these metastases. For extradural lesions, surgery is indicated if the diagnosis is unknown and no primary lesion is apparent, if the patient has progressive neurological deterioration during adjuvant therapy, or if the primary tumor is known to be poorly responsive to adjuvant therapy. The management of patients who manifest symptoms of spinal cord compression from a tumor that is known to be sensitive to adjuvant therapy remains controversial. In some centers, such patients are treated initially with chemotherapy, reserving surgery for those that fail to respond or who deteriorate neurologically. In other centers, urgent tumor debulking is performed before adjuvant therapy. When surgery is performed, a posterior approach is often preferred, because most childhood lesions arise from transforaminal extension of a paraspinal lesion. Patients require long-term postoperative monitoring both for signs of disease progression and for evidence of progressive kyphosis.

PEARLS

In these authors' experience:

- Spinal cord compression by malignant tumors in children is most often caused by direct extension of the tumor through the neural foramina.
- In the absence of severe spinal cord compression, most so-called small cell blue tumors, such as neuroblastoma and lymphoma, can be treated successfully with chemotherapy or radiation therapy.
- Most sarcomas require operative decompression.
- Indications for operation include tumors that severely compress the cord, regardless of type, and undiagnosed tumors that have spinal cord compression as the initial symptom.
- The goal of decompression is not to remove all tumor, but to decompress the cord. Adjuvant therapy is indicated in almost all instances.

SUGGESTED READINGS

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16

INTRASPINAL INTRAMEDULLARY NEOPLASMS

Cheryl A. Muszynski, Shlomo Constantini, and Fred J. Epstein

Recent microsurgical progress, including advancements in instrumentation, have fostered a more aggressive, successful surgical approach to intramedullary spinal cord tumors. Throughout nearly two decades, the senior neurosurgeon–author, Fred J. Epstein, M.D., has used a radical surgical approach to intramedullary gliomas on more than 200 children and 150 adults. Based on his experience and that of others, it is now an established, exciting fact that with most pediatric spinal intramedullary tumors, gross total resection can be safely accomplished. It is imperative, however, to note that early diagnosis and treatment improve postoperative outcome.

PREOPERATIVE EVALUATION

Early Indicators

The pathological type of intramedullary tumor present is reflected in the individual patient's clinical course. Benign neoplasms typically manifest symptoms over a period of several months to years, whereas children with malignant neoplasms develop symptoms over a brief period of a few weeks to months. When a patient has a benign ependymoma that has hemorrhaged, an exception to this clinical pattern occurs as signs and symptoms develop over a brief period of hours or even a few days, similar to the time frame of a malignant tumor.

Common indications for surgery include (1) motor regression, such as refusing to stand and reverting from walking to crawling; (2) local pain along the spinal axis escalating in severity at the spinal level directly over the tumor; (3) gait abnormalities reflecting lower extremity weakness; (4) dysesthesias; (5) progressive kyphoscoliosis; and (6) torticollis with corroborative radiographic images.

Diagnosis

Sensitivity and specificity make magnetic resonance imaging (MRI) the current diagnostic procedure of choice except in patients with severe scoliosis or adjacent metallic implants, which produce disturbing artifacts. In its disclosure capacities for intramedullary, rostral, or caudal tumor-associated cysts, the T1-weighted MRI is most informative; the T2-weighted MRI is reserved for assessing associated edema and for providing complementary information. In that the administration of gadolinium (Gd-DTPA) enhances the solid component of some intramedullary spinal cord tumors, delineating gadolinium-enhancing regions of the tumor from adjacent cord edema, preoperative evaluation of all patients with intramedullary spinal cord tumors must include Gd-DTPA scan.

Specifically, most tumors of pediatric patients are low-grade astrocytomas and gangliogliomas. When treated

with aggressive resection, approximately 85% of patients may experience long-term, progression-free survival even without additional treatment. The following serves to detail the surgical procedure used for resection of spinal intramedullary tumors.

SURGERY

Patient Positioning and Room Arrangement

The patient is placed in the prone position. The body lies on longitudinal gel rolls on both sides, thereby allowing free chest movements and preventing abdominal compression, which would result in increased venous pressure. For cervical and upper thoracic operations, the hands are secured on the sides; for lower thoracic or lumbar procedures, they may be elevated and positioned on armboards. All pressure points must be adequately padded and cushioned. The legs are slightly elevated and wrapped with elastic bandages. All patients should have a perioperative indwelling urinary catheter.

For *cervical* operations, the gel rolls are advanced to the edge of the bed and elevated slightly at their upper ends to create space for the chin, thus reducing the amount of reverse trendelenburg required to reach a horizontal cervical spine when the head is flexed. The skull is immobilized to the operating table using a pin-fixation device (Fig. 16–1).

The surgeon(s) and assistants stand on both sides of the patient. The microscope and the CO_2 laser are on one side of the table. One corner of the room is devoted to the evoked potentials team and equipment. The ultrasonic aspirator is brought in from either side. The ultrasound screen should face the operating surgeon.

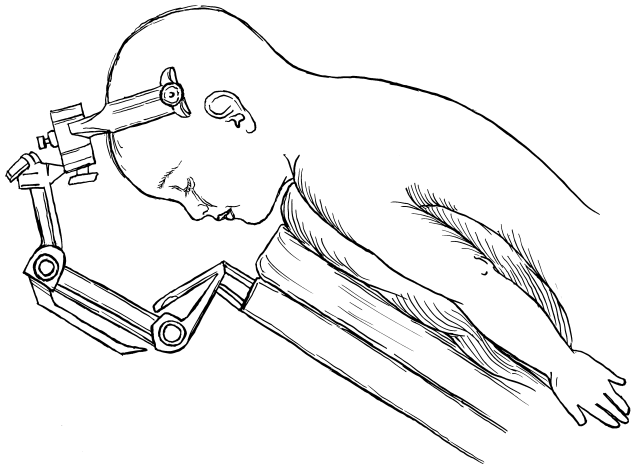

FIGURE 16–1. The patient is positioned prone on chest rolls, the head is immobilized in the skeletal fixation device and the neck flexed.

Soft-Tissue Opening

Before scrubbing, the exact location of the spine level(s) of interest is calculated by counting either from a landmark below (i.e., the anterior superior iliac spine) or from one above (i.e., the C-7 spinous process). In the thoracic region, verification by a localizing radiograph is recommended. Following skin cleaning, the midline and a few perpendicular lines are drawn with methylene blue or an indelible marker.

The epidermis and dermis are opened with a number 10 blade. The rest of the skin is opened with the monopolar cutting instrument to the level of the fascia. The skin must be opened one level above and below the planned span of the laminotomy or laminectomies. It is important to identify the fascia and to divide it in the precise midline for easy identification when one later closes the wound (Fig. 16–2). Two self-retaining cerebellar retractors are inserted, and hemostasis is obtained with the bipolar coagulation device. The spinous processes and the laminae are exposed in the standard subperiosteal fashion until the medial facets are identi-

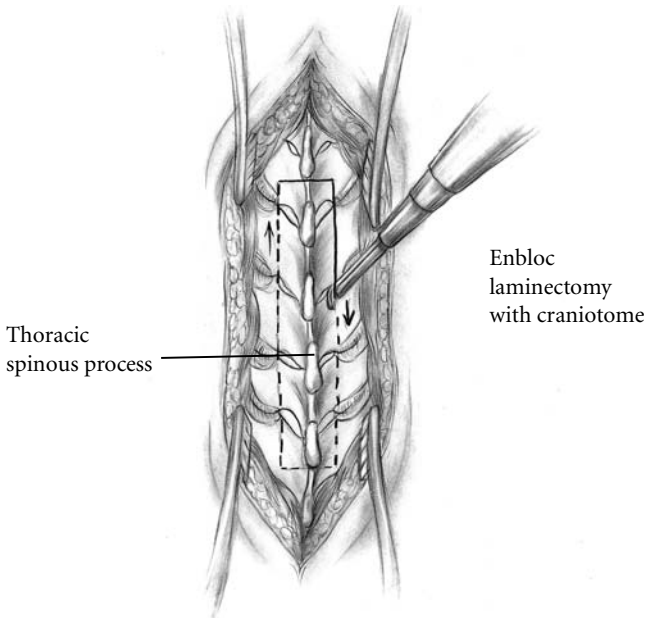

FIGURE 16–2. Exposure of the laminae and spinous processes. The muscles are retracted to the sides with self-retaining retractors. Note the craniotome underneath the lamina, medial to the facet joint.

fied. The self-retaining retractors are repositioned to a deeper level, and adequate hemostasis is obtained.

Osteoplastic Laminotomy

The interspinous ligaments and the ligamentum flavum are resected in the midline above and below the planned span of the laminotomy flap. The epidural fat is exposed at these levels. The laminae are marked symmetrically with methylene blue at points just medial to the corresponding facet joints. The Midas Rex instrument (or any other high-speed device) is applied at the epidural plane to cut the laminae at a slightly diagonal level. The laminotomy flap is fully released and preserved. Bleeding epidural veins are carefully coagulated, and thin strips of Gelfoam are applied to the lateral gutters to assist in the tamponade of epidural oozing.

Next, ultrasound is used to verify that the bony exposure is adequate to locate the intramedullary tumor (Fig. 16–3). If it is necessary to improve the exposure, the laminotomy is expanded in the appropriate rostral or caudal directions. It is unnecessary to extend the laminotomy or the dural opening over rostral and caudal tumor-associated cysts; the latter are typically decompressed after the solid portion of the tumor has been debulked. Both rostral and caudal epidural electrodes are inserted (Fig. 16–3), secured, and then connected to the monitoring unit.

Dural Opening

The dura is best opened with a number 11 blade after it has been elevated with a dural hook (Fig. 16–4). Further opening is executed with the blade on top of a grooved dental tool. A small dissector is used to peel the dura from the adjacent arachnoid mater. Retraction sutures are applied to the cut dural edges and suspended by hemostat clamps (Fig. 16–4). The operating microscope is brought into play. At this stage, ultrasound identifies rostral, caudal, or intratumoral cysts for determining that portion of the solid tumor with the greatest anterior-posterior dimension. Next, intradural monitoring electrodes are placed.

Midline Myelotomy

As the bulk of the underlying mass lesion usually distorts the spinal cord, obscuring its normal anatomy, it is thor-

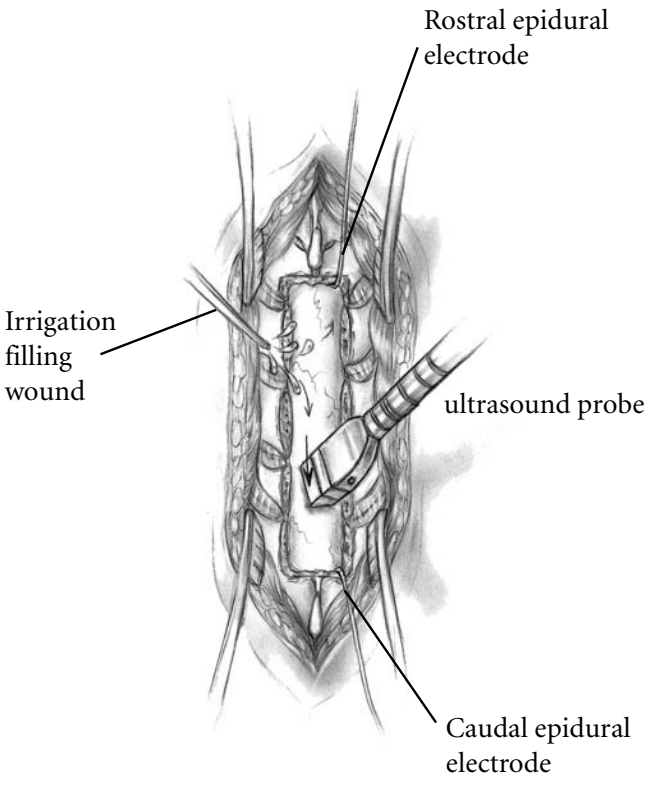

FIGURE 16–3. The ultrasound probe is applied to the saline-filled wound for locating the rostral and caudal ends of the tumor.

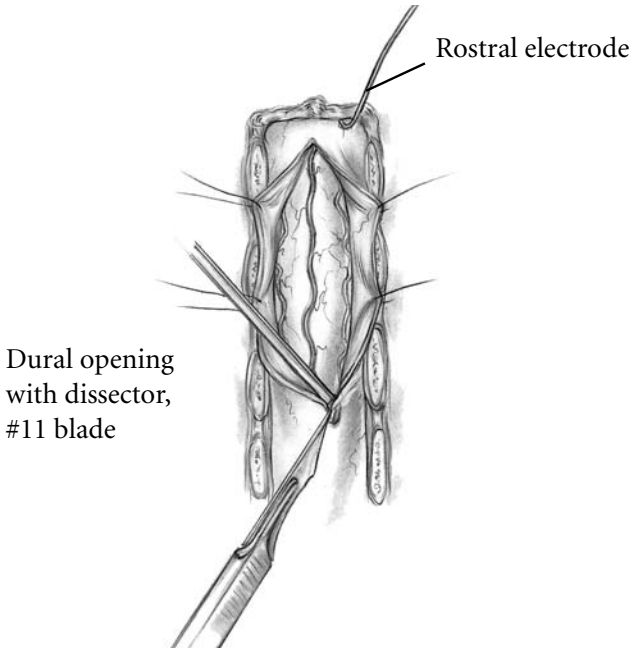

FIGURE 16–4. The spinal cord with the cut dural edges is retracted laterally to each side.

oughly examined. Both of the dorsal root entry zones are identified in an attempt to locate the precise midline. Often the midline raphe may be identified by the small diagonal vessel typically situated at the medial surface of the posterior columns. Next the midline dorsal surface of the cord is meticulously bipolar coagulated. In general, coagulation of posterior surface veins is risk free. The myelotomy incision is best initiated with an arachnoid knife in that region of the cord in which the thickest portion of the tumor exists (Fig. 16–5). Any bleeding vessels are coagulated carefully with the irrigating bipolar.

The edges of the myelotomy incision then are reflected laterally with either the plated bayonet instrument or two fine microdissectors (Fig. 16–6). Following this, the myelotomy incision is extended rostrally and caudally with microscissors.

Next, fine pial traction sutures are applied to open the lips of the myelotomy incision further. Usually, the intramedullary tumor tissue first is encountered at a depth of 1 to 2 mm. After the tumor tissue is identified, the posterior columns are gently reflected laterally with the plated bayonet instrument, exposing the lateral aspects of the tumor.

Tumor Removal

The excision of a solid, noncystic astrocytoma is initiated at the midportion (rather than at the rostral or caudal poles) of the neoplasm. When there are no associated cysts, there is no definite rostral or caudal demarcation of the tumor. In addition, because the poles of the neoplasm are the least voluminous, their removal may prove the most hazardous because of the associated risk of disruption of normal neural tissue. When rostral or caudal cysts exist, one of the cyst–tumor junctions is identified first. After the cyst is entered, inspection of the cavity will localize the rostral or caudal aspect of the neoplasm extending into it. After each of the cyst–tumor junctions is identified, the myelotomy is extended in the midline between the previously placed incisions (Fig. 16–7).

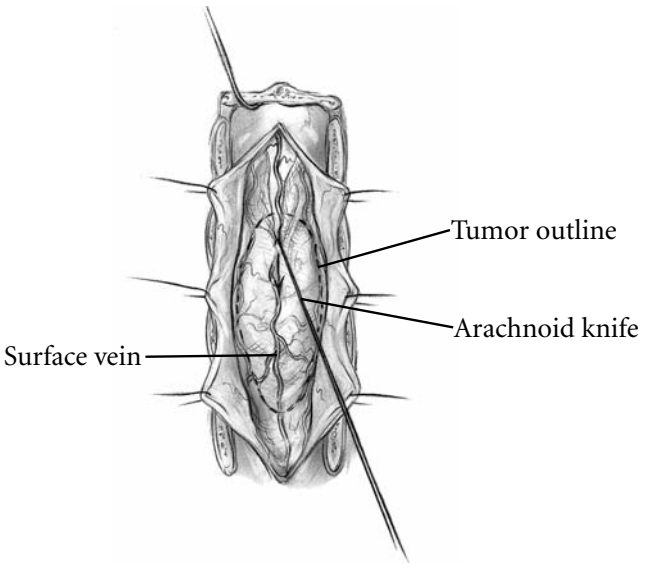

FIGURE 16–5. An arachnoid knife is used to divide the coagulated pia mater.

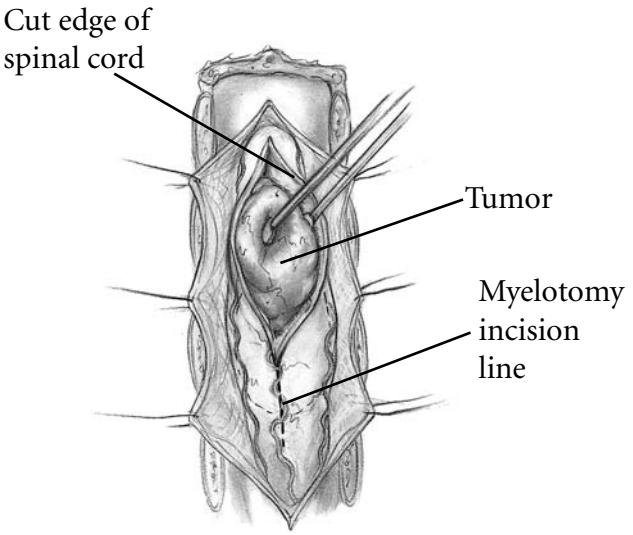

FIGURE 16–6. The plated bayonet laterally displaces the lips of the myelotomy incision, exposing the underlying tumor. Arachnoid-pial sutures help to better expose the intramedullary tumor.

The bulk of the tumor is removed most easily and safely by using either the ultrasonic aspirator or the laser, with minimal traction forces on the adjacent, normal white matter. It is preferable to resect the tumor from an inside-to-outside direction, without attempting to find a cleavage plane at the glial–tumor interface. Intraoperative ultrasound is used to monitor the degree of resection. After the tumor is debulked with ultrasonic aspiration, the bipolar coagulation–suction technique is used to resect the visible residual tumor fragments (Fig. 16–7).

Ependymomas are usually red or dark gray, typically with well-defined glial-tumor margins. The principle of an initial central debulking is no longer used for this type of intramedullary spinal cord tumor because ependymomas invariably having a true cleavage plane separating them from the adjacent cord substance. Therefore, following the myelotomy, the caudal-most lateral aspects of the neoplasm are identified, and the planes of cleavage between the tumor and adjacent tissues are developed.

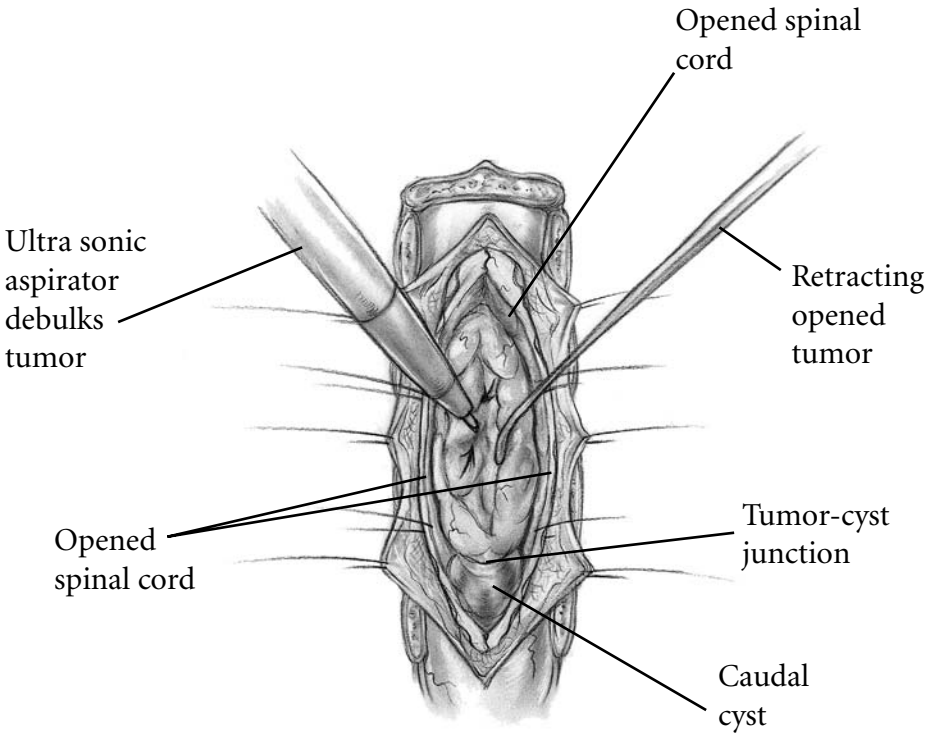

FIGURE 16–7. Resection technique for an intramedullary astrocytoma. The ultrasonic aspirator, which is positioned opposite the microdissector, demonstrates the recommended "inside-out" method of resection of these types of infiltrating gliomas.

Progressive resection in a caudal-to-rostral direction then is accomplished (Fig. 16–8). Because the ventral aspect of the neoplasm is typically adherent to the anterior median raphe, this segment of the ependymoma must be removed carefully—employing sharp dissection under high magnification—to avoid injury to the anterior spinal artery. Because of the ependymoma's noninfiltrating nature, this tumor is considered a "surgical disease."

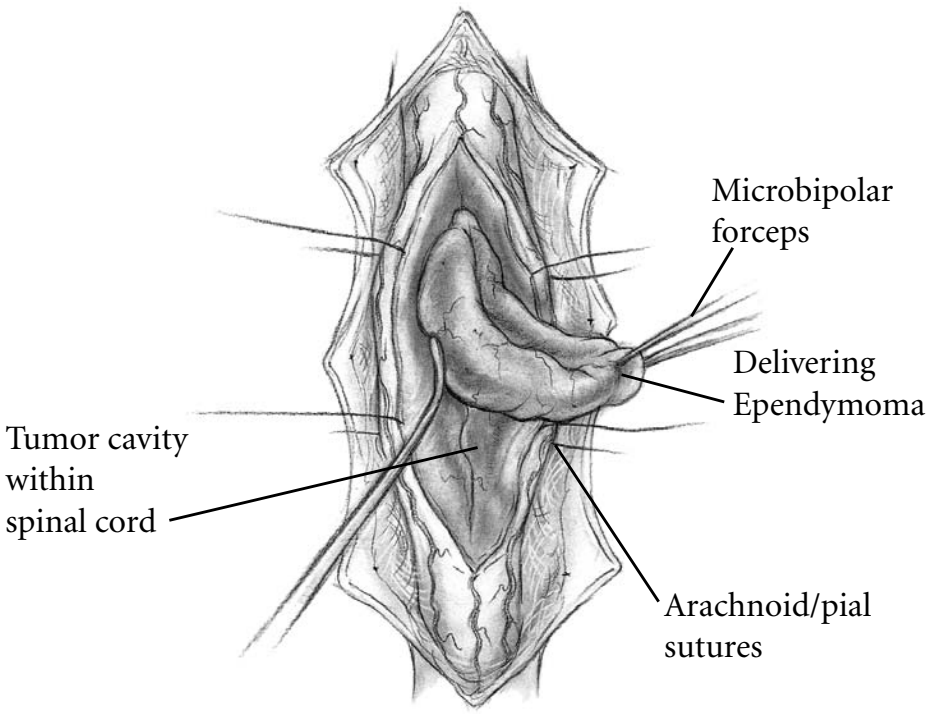

FIGURE 16–8. Resection technique for an ependymoma. A pair of microbipolar forceps are shown grasping the ependymoma. The forceps are positioned opposite the microdissector, demonstrating the caudal-to-rostral method of ependymoma dissection.

Closure

After tumor removal, the pial traction sutures are removed, and the cord is brought into a relatively normal position. The pial surfaces are not sutured together. The dura usually is relaxed at this stage and typically can be closed primarily without the need for duraplasty. Holes are drilled at each laminar level for resecuring the bone into place; 2-0 PDS sutures are used for this purpose. The spinous processes of the osteoplastic laminar flap are trimmed to prevent perforation of the fascia after closure. To achieve closure without tension, the fascia and muscles are released both superficially from subcutaneous tissues and deeply from the bony elements. If tension still exists, fascial-relaxing incisions are indicated. The musculofascial layer is closed with tightly knotted figure-of-eight permanent sutures. For optimal wound closure, the skin itself is closed in three layers (the deep subcutaneous level, the superficial layer, and the skin surface) with absorbable sutures. Small-caliber drains are left between the fascia and the skin until they no longer drain any fluid.

A pressure dressing is applied to the wound. With low thoracic or lumbar incisions, the patient is kept at bed rest for at least the first 72 postoperative hours. With cervical and upper thoracic incisions, the patient typically is mobilized within the first 48 postoperative hours.

SPECIAL CONSIDERATIONS

Intraoperative Neurophysiological Monitoring

Intraoperative neurophysiological monitoring has proven invaluable and indispensable in customizing the surgical technique to an individual patient's baseline electrophysiologic signals, monitoring motor evoked potentials (MEPs) and sensory evoked potentials (SEPs). The capacity to update every 2 to 3 seconds provides optimum feedback to the surgeon during the vital tumor resection. Joint position sense is preserved postoperatively when SEPs are preserved intraoperatively. If baseline SEPs are normal, and are lost intraoperatively, a resultant transient loss of joint position sense has been observed postoperatively. Experience has shown that spinal epidural SEPs are more reliable in predicting postoperative function than the cortical ones. The senior neurosurgeon author (FJE) has used such monitoring in more than 300 procedures for intramedullary spinal cord tumor resection.

Current experience confirms that epidural MEPs are reliable in predicting postoperative neurologic status. Specifically, less than a 50% reduction of intraoperative epidural MEPs has portended a transient monoparesis or paraparesis postoperatively. A more profound postoperative neurologic deficit is associated with an amplitude reduction of more than 50%, with the potential for significant recovery much less a certainty. Although scientific documentation of the threshold for motor recovery is unavailable, dissection beyond a 50% decline in the MEPs is not advised. If such a fall in potentials occurs, dissection is halted to await the recovery of potentials.

The function of the motor pathways from the subcortical white matter to beyond the neuromuscular junction in each of the patient's extremities is reflected in muscle MEPs, with the evoked potentials particularly useful in the conus medullaris and cauda equina regions, two areas over which the epidural MEPs are nonobtainable. One documented limitation of muscle MEP monitoring is its inherent sensitivity to the effects of anesthetic agents. Our intramedullary spinal cord tumor patients are therefore anesthetized with a combination of propofol, fentanyl, and nitrous oxide; no muscle relaxants or halogenated agents are used.

In general, we have observed that intraoperative muscle MEPs tend to decrease before any changes are noted in epidural MEPs. More specifically, when muscle MEPs are lost without any significant change in epidural D-wave amplitudes, a transient postoperative motor deficit invariably results. Therefore, the loss of muscle MEPs are now recognized as a "warning sign" of subsequent change in epidural MEPs.

Prior to tumor resection, we consider the patient's baseline signals and preoperative clinical status to outline criteria for "significant signal changes." Often, during dissection of an intramedullary tumor, a minor (i.e., <30%) decrement in epidural MEPs occurs. Irrigation of the wound with tepid Ringer's solution often reverts the signals to baseline status; therefore, the decrement may be a reflection of local ionic environmental changes occurring during tumor resection rather than disruption of motor pathways, as in a "true" loss of signals. For this reason, we start and stop the dissection, as indicated, during the course of tumor resection.

When the laser is used for more than 20 consecutive seconds, often an adverse, probably thermal, effect is manifested through a decrease in amplitude and an increase in latency. The dissection then is interrupted for the irrigation of the resection cavity with Ringer's solution at a temperature of 30 to 33 °C, returning electric activity, in most cases, to "baseline" within 30 to 90 seconds.

Because a consistent correlation between intraoperatively acquired neurophysiological data and postoperative function exists, we rely on monitoring for precise decision making during resection of intramedullary spinal cord tumors.

Postoperative Complications

Paralysis, the major complication following resection of an intramedullary spinal cord tumor, is closely related to the patient's preoperative condition. The risk is less than 5% for a patient without any preoperative motor deficit(s), whereas those with preoperative motor dysfunction face a relatively greater risk. Thus, it is imperative for patients with known intramedullary tumors to undergo surgery prior to the manifestation of significant neurologic deficits. Avoidance of this complication may be achieved by carefully following the intraoperatively acquired electrophysiologic signals.

Impaired joint position sense, a serious functional disability, is another potential complication. When this occurs, extensive postoperative physical and occupational therapy often is required for successful adaptation. Avoidance of this complication may be achieved by carefully placing the myelotomy in the posterior median raphe.

Spinal deformity in the form of scoliosis and kyphosis is yet another potential postoperative complication. The gravity of this complication is underscored in some reported cases of kyphosis where the condition caused spinal cord compression and progressive myelopathy.

It is vital to understand postoperative spinal deformity because appropriate treatment and prognosis differ from that of recurrent intramedullary spinal cord tumors. Although scoliosis is not usually the causative factor of spinal cord compression, it may indeed exacerbate a preexisting neurologic disability. Therefore, close evaluation by an experienced orthopedic surgeon is seriously advised. Spinal fusion may be viewed as more urgent for this group of patients than in those with idiopathic deformities.

Recently, the impact of osteoplastic laminotomy versus a simple laminectomy on progressive postoperative kyphoscoliosis was evaluated. Within the limited length of follow-up, it was concluded that replacement of the bone in osteoplastic laminotomy may minimize the incidence of progressive postoperative kyphoscoliosis (personal communication, S.C.), and use of osteoplastic laminotomy is recommended whenever possible.

Hydrocephalus is more likely to manifest after resection of malignant rather than benign intramedullary spinal cord tumors. For patients presenting with cervical astrocytomas with an associated rostral cyst and thickened cervicomedullary junction leptomeninges, it can be deduced that the thickening caused fourth-ventricular outlet obstruction, resulting in hydrocephalus. Hydrocephalus complicating an intramedullary spinal cord tumor caudal to the cervicothoracic junction strongly suggests the presence of malignant dissemination with obstruction of cerebrospinal fluid pathways. Early detection of this complication may be achieved by performing postoperative, surveillance brain computed tomography scans on individuals who have undergone resection of malignant intramedullary spinal cord tumors.

Postoperative Management

Abundant evidence emphasizes the deleterious effects of radiation on the immature nervous and osseous systems. Furthermore, radiation may promote the future development of a second malignant tumor. Left untreated by radiation, low-grade tumors in children possess little ability to transform into high-grade ones. Because pediatric patients with benign intramedullary spinal cord tumors treated with radical surgery may have long-term, progression-free survivals without adjuvant treatment, we recognize pediatric intramedullary low-grade tumors as a surgical disease, both at the time of presentation and again at recurrence.

No clear evidence points to either radiation or chemotherapy as improving the outcome of low-grade astrocytomas of the central nervous system. In addition, the overall effect of radiation therapy on grade II astrocytomas has not been well documented. It is felt that patients with recurrent high-grade intramedullary astrocytomas should consider radiation and chemotherapy only if there is no option for surgery, even as glioblastomas invariably progress.

Serial MRI scans are performed to monitor patients carefully with minimal residual disease. Reoperation of substantial residual tumor may be proposed after having evaluated the patient's previous electrophysiologic signals. With the effects of chemotherapy and radiation therapy so unclear, use of these modalities is reserved for rare occasions only.

CONCLUSION

Overall, great strides have been made in the treatment of pediatric intramedullary spinal cord tumors. Viewed as inoperable only a few decades ago, today they are not

only operable but also frequently curable. Expeditious diagnosis and treatment ensure a reasonable likelihood of long-term success.

EDITOR'S COMMENTARY

The senior author (FJE) has been a leading figure in shaping the state-of-the-art management of intramedullary spinal cord neoplasms. The majority of such tumors are low-grade lesions, which can be extensively resected with appropriate techniques, with favorable long-term results. Nonetheless, the potential certainly exists for serious morbidity with such operations, particularly in children that present with severe neurological impairment. The role of functional monitoring as a way of avoiding rather than merely detecting irreversible neurologic injury continues to evolve. In contrast to the majority of pediatric spinal cord tumors, which can be viewed as a surgical disease, the small percentage of tumors that are malignant require a multimodality approach to management.

PEARLS

In these authors' experience:

- Radical surgery for the removal of pediatric spinal intramedullary neoplasms may be performed with acceptable morbidity.
- Intraoperatively measured spinal epidural MEPs are reliable predictors of postoperative neurological outcome.
- Postoperative functional performance is directly related to the patient's preoperative neurological status.
- It is imperative to diagnose and to treat these patients as early as possible.

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Trauma

17
REPAIR OF SKULL FRACTURES

Dennis Johnson and Paul M. Kanev

Twenty percent of children admitted to the hospital with head injury have a skull fracture. About 25% of skull fractures are depressed, and about 10% of those will be associated with dural laceration; 20% will be compound.

DEPRESSED SKULL FRACTURES

The indications for elevation of depressed skull fracture have been discussed by Luerrsen and include cranial deformity, dural laceration, contamination, compression of the brain, and mass effect. Elevation of depressed fractures neither decreases the incidence of posttraumatic epilepsy nor substantially improves associated neurologic deficits.

Principles of technique include adequate surgical exposure, decontamination, protection of the brain, and skin closure. *Adequate exposure* means a skin incision that allows ample working space to elevate the fracture. Small, chic incisions can lead to additional injury to the surrounding skin by power instruments, bone elevators, and retraction.

Satisfactory decontamination includes the use of betadine or Hibiclens shampoo of the scalp and removal of all particulate matter. The time-honored neurosurgical technique of shaving the scalp has not proven efficacious. Shaving is time consuming, abrades and inoculates the skin with bacteria, and is not associated with a lower incidence of infection, although closing hairless skin is simpler, quicker, and requires less skill and patience. Sonic water irrigation and bacitracin irrigation solution are supplemented by intravenous antibiotic prophylaxis.

The brain also must be protected from any additional injury caused by periosteal elevators. The heel of the curved Adson periosteal elevator can damage the underlying brain if used inappropriately in the dissection (see later discussion).

Skin incisions generally incorporate any accompanying laceration unless the wound is a puncture. The hairline is always respected. S-shaped incisions allow wide exposure with self-retaining Gelpi retractors and facilitate extensive mobilization of the skin to close skin defects (see Goodrich). Wide undermining is essential in sliding the margins together; releasing the galea about 4 cm back from both edges of the incision can be useful.

Three basic techniques are used to elevate the skull: dent pulling, craniotomy extraction, and prybar elevation. Each has its place in neurosurgical armamentaria. *Dent pulling* is a term taken from the automotive repair business. A screw is placed at the centerpoint of the depression. The screw is "noosed" with an 18-gauge wire, and upward pressure is exerted to elevate the fracture (Fig. 17–1). Alternatively, a small hole at the centerpoint of the depression gives purchase for a sturdy hook to be inserted, and the hook provides a good handle to pull up the fragments (Fig. 17–2).

Another option is two obliquely drilled channels directed toward each other to allow a sharp towel clip to grip the fragment for extraction (Fig. 17–3). This technique is not well suited to depressions 1 cm or larger or more because the brain is too closely applied to the undersurface of the depressed bone to avoid injury.

Simple or prybar elevation is best suited for depressions in neonates and infants (<1 year of age). A slot can

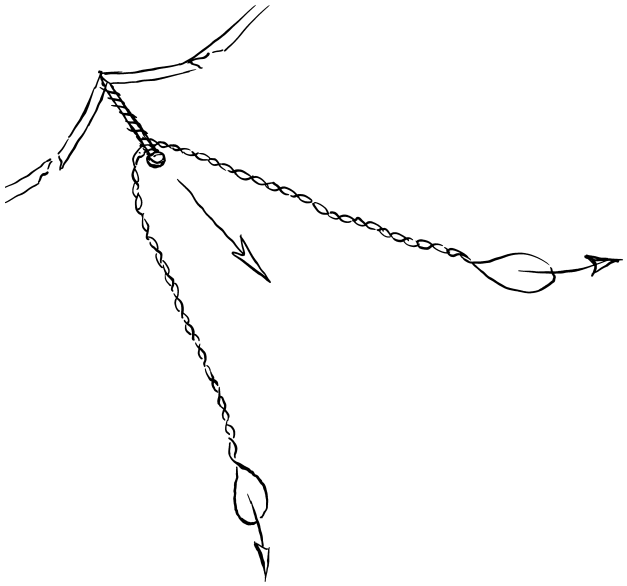

FIGURE 17–1. Dent pulling.

be made in the bone at the margin (i.e., at the beginning of the slope) of the depression by a sharp, curved Adson elevator. Use controlled pressure downward and a side-to-side rocking motion similar to the action of the ancient tumi (Fig. 17–4). Alternatively, the edge of the depression can be rongeured away for a point of access. A straight Adson then is slipped through the slot, parallel to the surface of the dura, and edged carefully down the slope of the depression. If the diameter of the depression is short (<3 cm), a curved Adson works best; the tip of the curved Adson can be brought to bear near the deepest part of the depression and levered upward. Care is taken not to drop the heel or curve of the elevator on to the underlying brain. To erase the tissue memory of the depression, elevation of the fracture must be exaggerated and then allowed to settle into place.

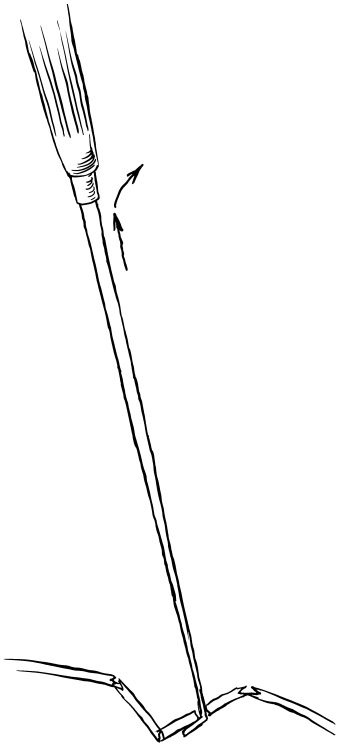

FIGURE 17–2. Inserting a hook to pull up bone fragments.

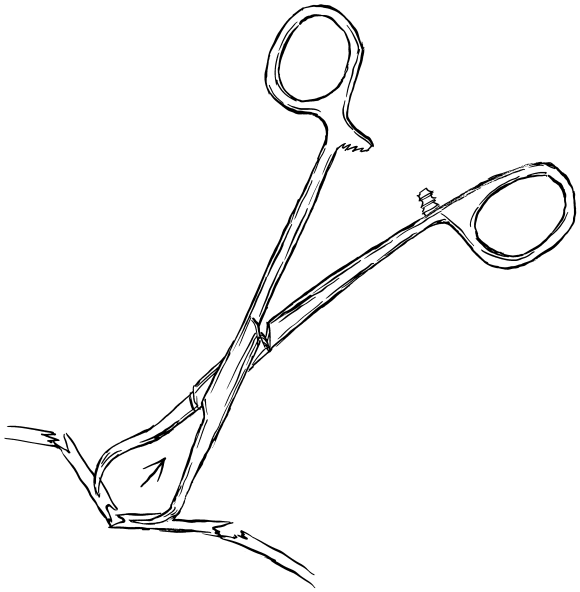

FIGURE 17–3. Two channels are drilled toward each other, and a sharp towel clip is inserted to grip the fragment for extraction.

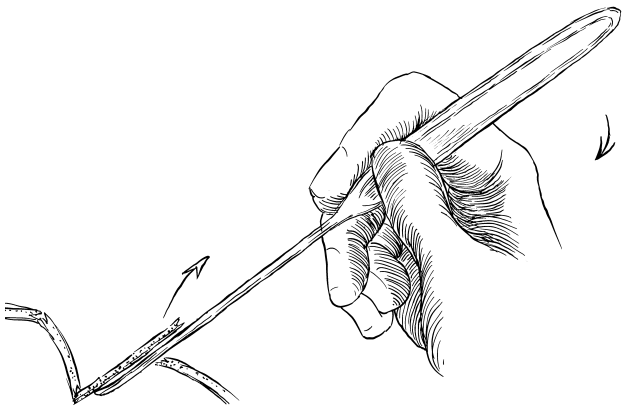

FIGURE 17–4. Simple or prybar elevation.

In older children or fractures depressed more than 1 cm, the full expanse of the depression is elevated as a craniotomy plate (Fig. 17–5). Care is taken to preserve the attachment of the periostem over the surface of the plate and at the hinge of the craniotomy. A good blood supply averts resorption of the plate. Extracting the fragmented plate precludes inserting instruments below the bone against the brain to "pop" up the fracture. The plate then can be separated into fragments and replaced. Microplates and screws tend to loosen and migrate. If fixation is deemed necessary, the authors prefer 22-gauge stretched wire with the twisted ends tucked into the crevices between bone fragments. Absorbable suture (2-0 Vicryl) and absorbable plates and screws are also available. With modern neuroimaging, intradural exploration is seldom necessary.

GROWING OR UNHEALED FRACTURES OF CHILDHOOD

Operative exposure is essential to satisfactory closure of unhealed fractures. The entire length and breadth of the fracture and a 2-cm penumbra must be exposed to find the margins of the dural defect. The margins of the dural defect may extend well beyond this penumbra. A pseudodura is exposed overlying the surface of the brain at the fracture site, but the true dura lies retracted well back beneath the margin of the defect. The pericranium provides the most physiologic dural grafting material, but the osteoblastic potential of free pericranial grafts will be less than those on a vascularized pedicle. Grafts based on the temporalis muscle can produce pain during chewing. The pseudodura and dura must be separated carefully from the overlying bone margins before the bone is removed to expose the dural margins. Injudicious dissection simply keeps the dura just beyond reach. If 3 mm or more thickness of the bone is removed, it is split like a child splits a cookie to lick out the icing (Fig. 17–6A).

The edge of the fracture is often thicker than the subjacent skull and is particularly suited to splitting. Thin, straight, and curved osteotomes are preferable to an oscillating saw. The saw wastes too much bone in sawdust and burns the osteoblastic cells in the bone. In the child with an unusually thin skull, there are three alternatives: Peter/Paul exchange, primary closure by advancing the edge of the fracture, and split rib. We use even less than mesh or methacrylate because harvesting the rib is associated with considerable postoperative discomfort, and the resulting scar is unaesthetic. Mesh and methacrylate will loosen over time, are subject to foreign-body reaction, and do not expand with the growing child. *Peter/Paul exchange* involves transplanting bone from a part of the skull that has normal underlying dura (Peter) to the defect (Paul) (Fig. 17–6B). There is no need to "repay" Peter because the bone will reform over the normal dura in children younger than 5 years of age.

Advancing the edges of the fracture follows the same principle. A 2-cm swath of bone on either side of the fracture is removed, the dura is repaired with pericranium, and the two pieces of bone are brought together and wired over the defect, leaving half the original defect on either side of the newly created plate (Fig. 17–7). Because the underlying dura has osteogenic potential, new bone will form in the "outside," marginal defects.

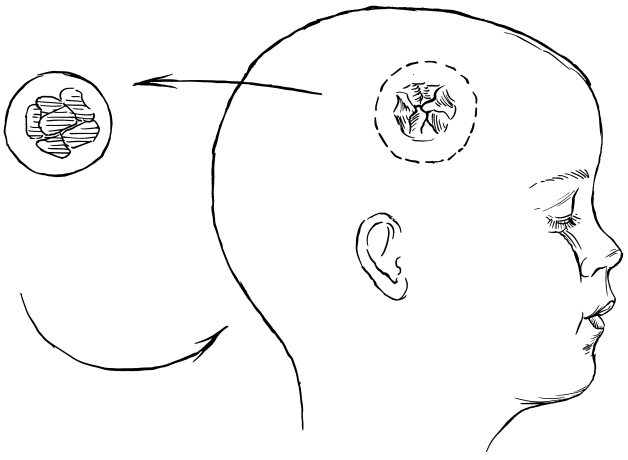

FIGURE 17–5. A craniotomy plate.

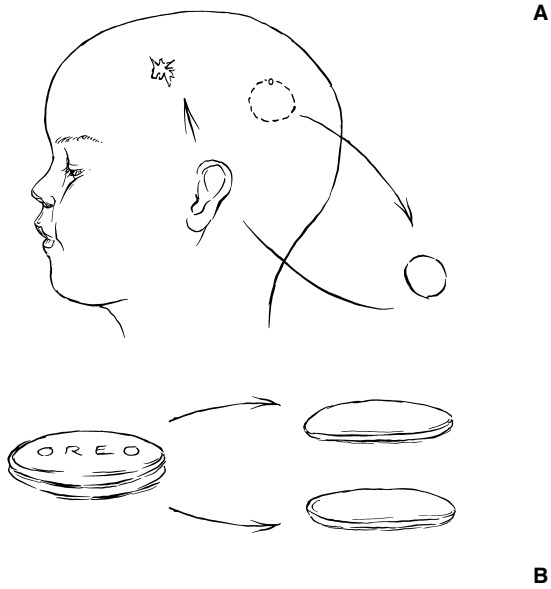

FIGURE 17–6. **A**: The bone margins are separated from the dura. **B**: Peter/Paul exchange.

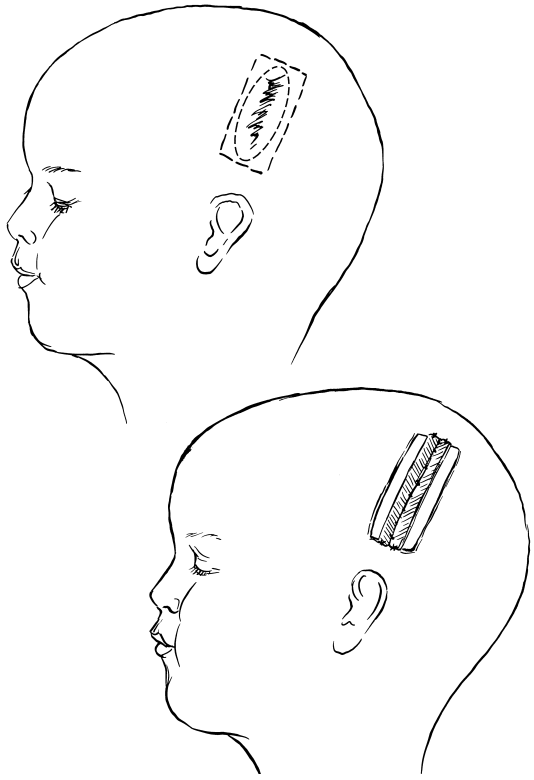

FIGURE 17–7. Advancement of the edges of the fracture and repair of the dura with pericranium.

A word of caution is in order. Not infrequently, the underlying porencephalic cyst is backed by mild communicating hydrocephalus. Subtle expansion of the skull after surgical repair may be reason enough to shunt the underlying hydrocephalus.

FRACTURES INVOLVING THE FRONTAL OR ETHMOID SINUSES

A simple, nondisplaced fracture through the frontal or ethmoid sinuses does not justify surgery. If pneumocephalus is present and the edges of a frontal fracture are displaced, repair of the sinus is indicated to reduce epidural contamination and to decrease the prospect of intracranial mucocele. The back wall of the sinus is removed and the mucosa are carefully stripped. The frontal ostium then is obliterated with cautery and "stuffed" with a tightly rolled "cigarette" of dry Gelfoam. This cranialization of the sinus automatically fills the sinus space with dura and brain. The ethmoid sinus is too complex to obliterate but should be blocked from the cranium by overlaying a wedge of bone into the fracture and overlying a strip of vascularized pericranium. A tear in the overlying dura mandates a separate closure of the tear to prevent cerebrospinal fluid (CSF) leakage. Fibrin glue with CSF drainage may be necessary for tears that extend far posteriorly. By draining the CSF temporarily through

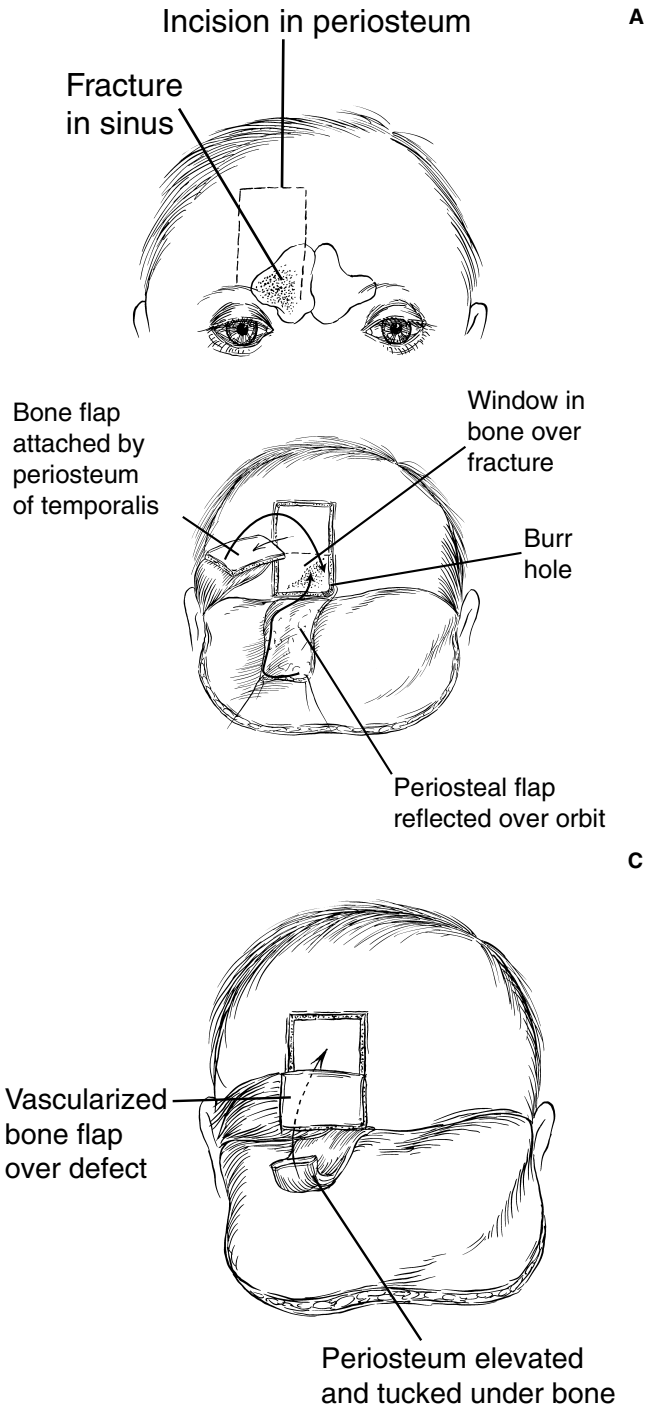

FIGURE 17–8. A–C: Skull-base repair.

lumbar puncture, a dry field will expedite firm application of the glue to the defect and the pericranium.

SKULL-BASE REPAIR

Repair of the skull base is seldom necessary because the fractures inevitably heal, and most CSF fistulas cease spontaneously. For those that do not respond to elevation of the head or lumbar drainage, direct repair of the dura may be necessary. Otorrhea usually is associated with a dural tear and associated fracture overlying the antrum. The skin incision straddles the external auditory meatus, and an initial intradural approach will localize the tear (brain often has slipped into the tear). The dural margin does not often lend itself to direct repair, and the angle of gentle exposure does not permit easy access with needle and needle holder. Pericranium attached laterally and flapped medially over the laceration provides one side of the sandwich; a similar pericranial flap provides the other, extradural side (Fig. 17–8). The sandwich is filled with fibrin glue and allowed to "set" with continuous lumbar drainage of CSF.

Fractures of the frontal base traverse the cribriform plate or violate the ethmoid sinuses. In an attempt to preserve olfactory fibers that perforate the cribriform plate, the extradural and intradural subfrontal dissection is purposefully limited. Fibrin glue and postoperative CSF drainage are the mainstays of this repair.

EDITOR'S COMMENTARY

A common problem in pediatric trauma, there are many methods for the repair of these fractures. New materials such as resorbable plates and mesh may be particularly useful for the comminuted fragmented fracture. The editor had used resorbable mesh epidurally and along the outer cortex to act as a "sandwich" with crushed cancellous bone and/or hydroxyapitite between the plates for a construct. This method has been quite useful for the repair of skull defects that have not adequately healed following trauma repair. We have likewise been finding the usefulness of these materials to replace rib graft and methylmethacrylate. It is the low skull base frontal ethmoidal fracture that can be particularly problematic in regard to CSF leaks and repeated bouts of meningitis. This editor has found that at times, an intradural repair with placement of a graft along the inner surface in conjunction with the vascularized pericranium graft extradurally and fibrin glue may be necessary for adequate sealing of the CSF leak (described by the authors).

PEARLS

In these authors' experience:

- Dent pulling is not well suited to depressions 1 cm or larger because the brain is too closely applied to the undersurface of the depressed bone and therefore may be injured.
- Prybar elevation is best suited for depression in neonates and infants.
- Craniotomy elevation is best suited for older children or fractures larger than 1 cm.
- Split rib grafting may cause considerable postoperative discomfort and scarring at the donor site.

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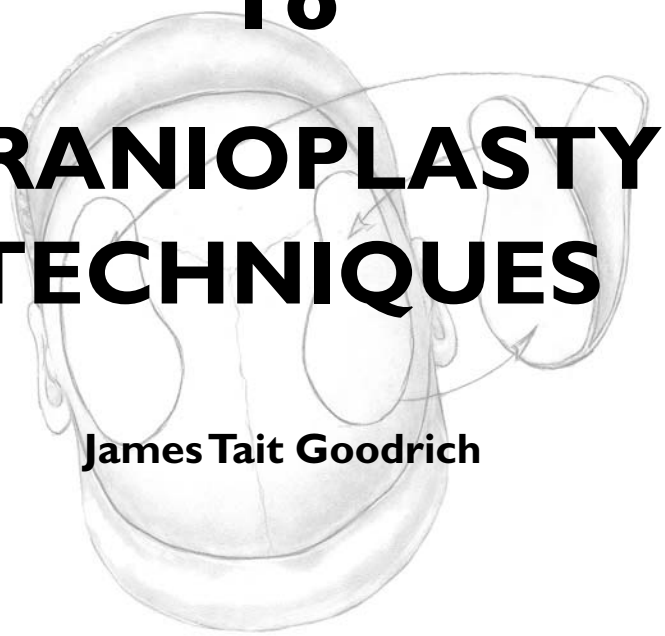
18

CRANIOPLASTY TECHNIQUES



James Tait Goodrich

A cranioplasty is a procedure for repairing a defect of the calvarium. A successful cranioplasty is one in which the cranial defect is repaired and then heals and grows with the child in the anatomically and aesthetically appropriate fashion. The location of the defect is extremely important. Defects in the frontoglabellar region require much more attention to detail than a defect behind the hairline. Contours, healing scars, alignment of eyebrows, orbital dystopia, and the like are all important factors.

SURGICAL INDICATIONS AND PREOPERATIVE EVALUATION

In the pediatric population (and often in the adult population), the indications for a cranioplasty are repair of a skull defect; these defects are most commonly a result of trauma or tumors of the skull and brain (Fig. 18–1). Children are active, and, lacking the "normal" senses of the adult, rarely appreciate a defect in the skull. A simple writing instrument, such as a pen or pencil, can become a lethal weapon in the hands of a playful child with a skull defect. A simple fall on the jungle gymn can lead to extraordinary damage to the brain in a child without adequate skull coverage and protection. The surgical indications for repair of a skull defect are straightforward: repair of any significant open bone defect where there is a risk of potential penetration of the brain.

The preoperative evaluation of a child with a skull defect is also straightforward. A plain skull series is done to outline the defect and to allow calculations of the area that needs to be repaired. If available, three-dimensional (3-D) computed tomography (CT) is extremely useful in outlining the defect and helpful to the family in the preoperative evaluation. A routine axial CT is important in outlining any underlying brain injury, the presence of encephalomalacia, and to rule out any sequestered debris or potential infectious material. An axial CT also can provide additional information about bone thickness and the availability of material for split-thickness calvarial grafts.

PREOPERATIVE MANAGEMENT

Most cranioplasties are done in elective situations and typically are delayed until 4 to 6 months after the original injury. Any evidence of infection at the injury site should be ruled out. Routine preoperative blood screening also should eliminate any potential of blood infection or sepsis. Any evidence of infection or sepsis is a definite contraindication to surgery. In cases where the child will require a helmet, we have our child-life specialist meet with the family to discuss the helmet requirements, such as when it must be worn and its purpose. Not uncommonly, we have the child wear the helmet for a couple of weeks preoperatively to become accustomed to it. In the younger child, it is important to appreciate the social stigma associated with wearing a helmet, especially in school, and these psychosocial problems should be addressed early.

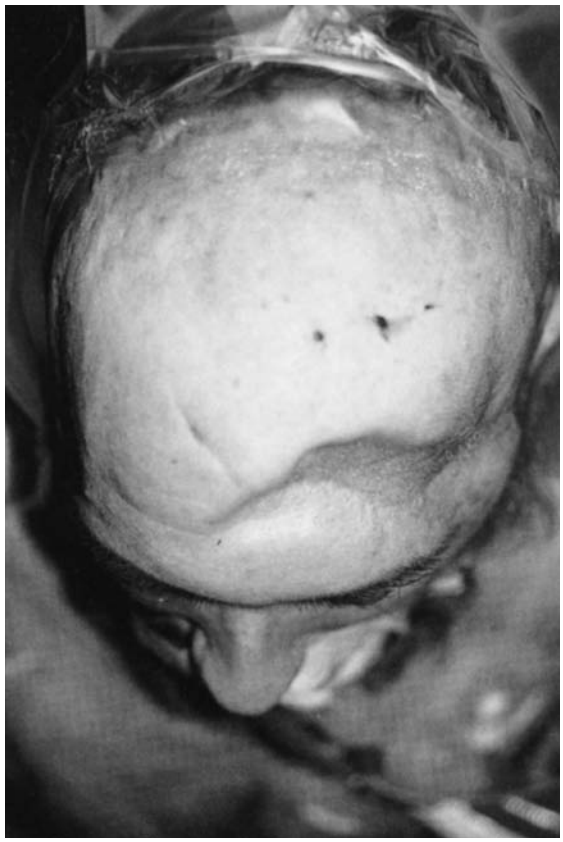


FIGURE 18–1. Intraoperative view of a patient with a large left frontal region skull defect secondary to severe head trauma. This patient presented 6 months after the injury with a large pulsatile cranial defect.

OPERATIVE PLANNING

Cranioplasties often are done in association with plastic surgery colleagues. The input of the plastic surgeon can be extremely useful, particularly for defects that involve the orbit and forehead regions. As part of the operative planning, we often include conferences with the radiologist, plastic surgeon, anesthesiologist, and child-life specialist to provide input on what each team can contribute to the case. In cases with extensive postoperative rehabilitation and physical therapy planned, we also ask the physical therapy and rehabilitation team to join the conference. Often, many questions are asked about the length and intensity of therapy and the restrictions implied by a cranioplasty; these issues are best addressed in the conference.

INTRAOPERATIVE TECHNIQUES

Anesthetic Techniques and Positioning

General anesthesia with an endotracheal tube is almost always used in cranioplasty repairs. Occasionally, a laryngeal mask is used in cases of short duration (i.e., less than 2–3 hours). In patients with large bone defects (i.e., greater than 8 cm), the potential need for a blood transfusion must be considered and blood made available for these cases. In cases that involve the face and orbit, the endotracheal tube must be well secured because there is often a good deal of movement of the head during the surgery to evaluate for aesthetic contours during reconstruction. We routinely give antibiotics (oxacillin 25 mg/kg of body weight) as soon as anesthetic is induced and well before the skin incision is made.

The positioning of the patient should be such that access to the repair site is generous. In cases where bone is to be harvested, both sites need to be in the prepared field. We have found the horseshoe head rest the most reliable in frontoorbital and vertex approaches. For defects of the lateral regions, the head is turned to the appropriate side and then rested on a soft donut cushion.

Skin Incision

Most cranioplasties are done in areas of previous injury or surgery. Therefore, a skin incision from the previous surgery is present. In the operative planning, the original scar can be incorporated in the incision and extended as necessary. All incisions should be planned in such a fashion that no vascular pedicle is comprised; close (<3 cm) parallel incisions always should be avoided. The vascular supply to the scalp arises from the base of the skull and ascends the scalp in a palisade fashion; therefore, knowledge of this anatomy and the various vascular anastomoses always should evaluated in the surgical planning. A common error is made when the skin incision is too close to the forehead hair line. A forehead incision should be placed either in a natural skin crease or at least 2.5 cm behind the hairline, even more if possible. We prefer to make wavy-type incisions, if possible, to prevent later hair parting over the incision when the hair is wet. Curvilinear incisions over the temporalis region also seem to reduce the incidence of hypertrophic scar formation that often occurs in this area.

Preparation of the Cranial Defect

The preparation of a cranial defect for repair remains the same in all cases no matter what the source of the defect. The surgical bed must be clean and free of debris of any kind. Both the surgical bed and the overlying surgical flap must be well vascularized. If these conditions are not present, neither the flap nor the implant will heal.

In the case of repairs in the vicinity of the frontal sinuses, it is extremely important to cranialize the sinus or obliterate it with a pericranium flap. In elevating the skin flap, the pericranium is brought up as a second layer with its pedicle kept intact, thereby developing a vascularized pedicle flap that can be easily rotated into a number of different positions. Bone edges to be incorporated in the repair have to be "freshened," that is, cleaned and debrided of any scar tissue or debris. The bone edges should be burred to provide a fresh bleeding edge. The use of bone wax along the freshened bone margins is discouraged because this material retards bone bridging and can provide a nidus for later infection. We use thrombin-soaked Gelfoam with pressure to stop the bleeding. Only in cases of large emissary veins where bleeding is difficult to control do we use bone wax, and then as little as is necessary. Meticulous hemostasis and gentle handling of soft tissues are always obligatory.

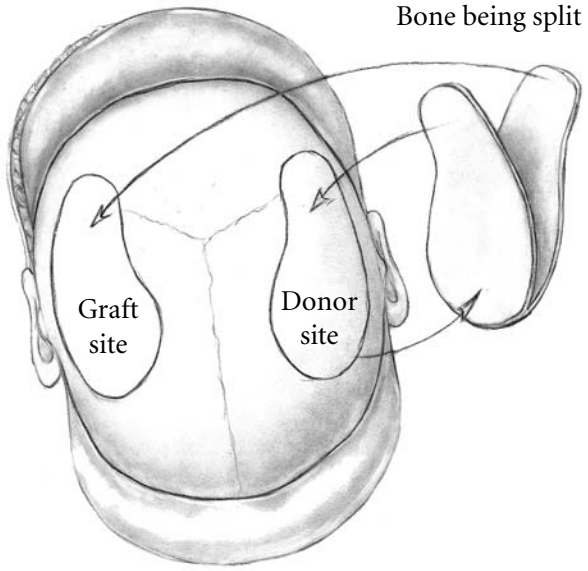


FIGURE 18–2. An artistic reconstruction on obtaining a split calvarial bone graft. In this case (see Fig. 18–1), the defect is over the right parietal temporal region. A mirror image bone is harvested from the contralateral side and elevated. Once the bone is split, the inner table will go back to the donor site, and the outer table is placed in the defect.

Techniques for Cranioplastic Reconstructions

Autologous bone always should be considered first as the primary repair material in cranial defects, particularly in a growing child. Autologous bone heals most naturally, will grow with the child, and has the lowest incidence of infection.

The surgeon has three types of autologous bone available for reconstruction: (1) calvarium, (2) costochrondral rib grafts, and (3) iliac crest. Each has advantages and disadvantages.

In cases in which a bone graft is needed to close a bone defect, the best source of autologous bone remains the calvarium. The skull has a natural mirror image that allows the surgeon to harvest contralateral bone and that often possesses a contour that closely matches the defect (Fig. 18–2). Because the skull is bilamellar, it can be split along the diploë, generating two pieces of bone, one to be placed in the defect and the other back at the donor site (Fig. 18–3 through 18–8). Using a soft malleable piece of metal, a template of the defect is designed and applied to the contralateral skull in various positions until an area is located that closely matches the defect to be repaired. The donor graft is always made 4 to 5 mm larger than the defect to allow for the bite of the craniotomy drill and gives additional trimming room for positioning the graft. The flap is elevated using standard craniotomy techniques with copious irrigation so that the heat generated by the craniotome does not burn the bone. As discussed already, the use of wax on bleeding bone edges is discouraged.

Once the bone flap has been harvested, it can be split in any of a number of ways. In the young child (i.e., younger than 2 years), a freshly sharpened osteotome handled much like a knife allows one to slice the bone apart (Fig. 18–5). In the child older than 2 years of age, the bone usually has become thick enough to split with fine, thin, high-speed bits, such as the C-1 attachment for the Midas Rex drill. Depending on the width of the flap, curved osteotomes and reciprocating saws with flexible blades are also helpful in splitting the bone (see Figs. 18–3 and 4).

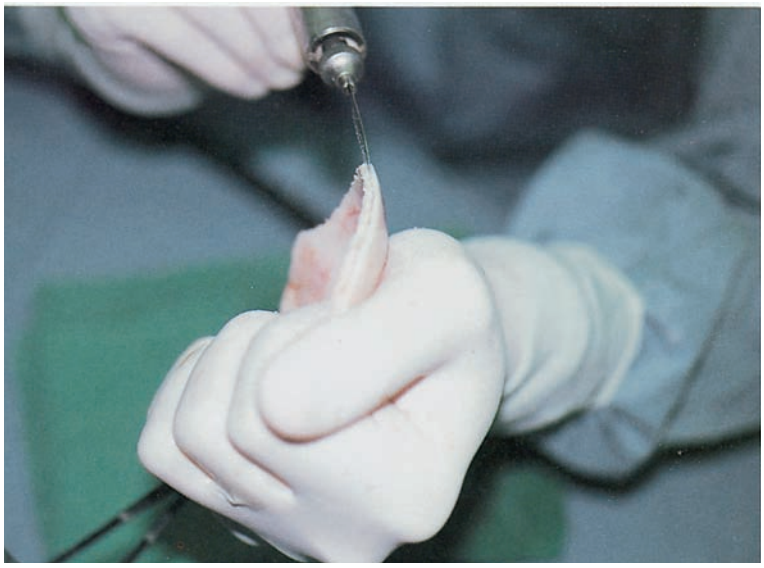


FIGURE 18–3. Intraoperative view showing the technique of a split-graft harvest. In this case, an oscillating (or reciprocating) saw is used with a flexible blade and the saw blade with naturally followed the diploë if no undue pressure is placed on it. Copious irrigating is key to keep the bone from overheating and burning.

The thickest bone for harvesting is over the frontal and parietal boss regions. The thinnest bone (and hence least useful) is over the temporal regions—considerations to be kept in mind when harvesting bone. It is always desirable to create large bicoronal exposures when the need for large grafts is expected.

Fixation of bone plates in the pediatric population has been a recent focus of discussion. We try not to use fixation miniplates in children younger than 3 years of age; despite being "mini," they can be felt easily through the skin, which is disconcerting to the child and parents. We also have had personal experience of unacceptable plate migrations such that at later reoperation, we found that the plates had migrated intracranially and in two cases were actually within the dura or brain parenchyma. With rare exception, we now use only sutures (e.g., Vicryl or Neurolon) to stabilize the flaps. The only exception is the occasional instance in which exceptional stability of the repositioned flap is required, as in repairs around the orbits. If the flap repair is being done well behind the hairline and the child is older than 3 years of age, the use of miniplates is acceptable.

Once the bone flap has been replaced, the edges are contoured using a high-speed burr so that they are not

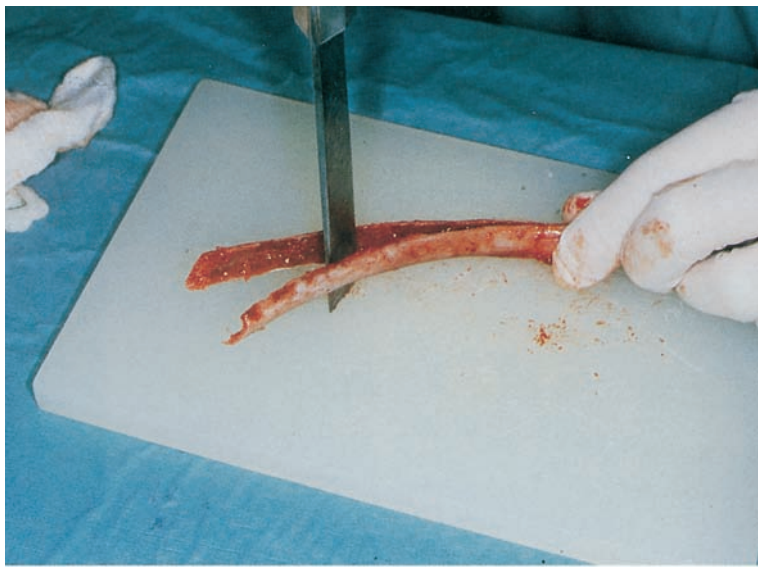
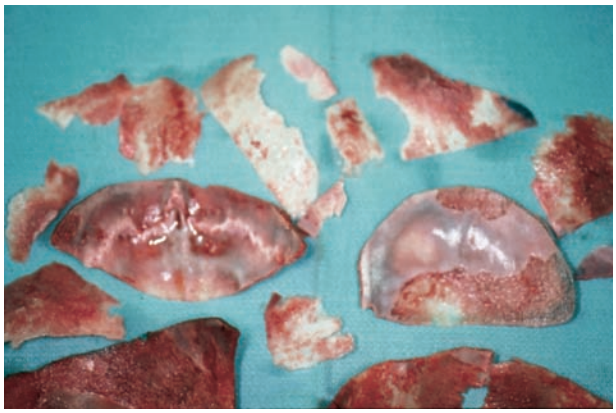
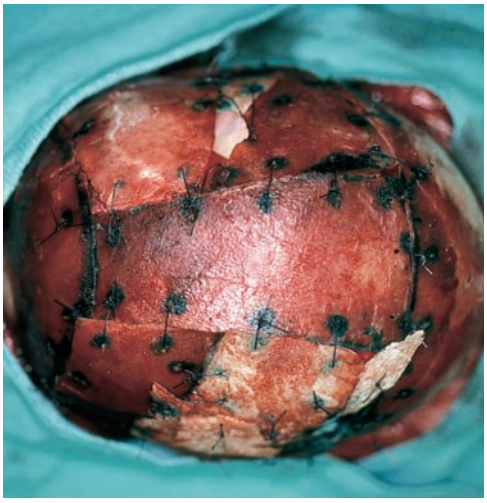


FIGURE 18–4. Illustrated here is the technique of using side-by-side osteotomes to follow the diploë and, in turn, splitting the bone.



A



B

FIGURE 18–5. **A:** In the young child split calverial bone grafts can be easily obtained by splitting the bone with a sharp osteotome or a fine cutting tip such as a Midas Rex C-1 attachment. **B:** Illustrated here is a complete reconstruction on a craniofacial case where multiple pieces of split calvarial bone grafts have been used to provide for a complete bone covering of the calverium.

palpable through the skin. In the infant, the bone can be contoured easily using a Tessier rib bender. Radial (fan-like) osteotomies also can be used to provide additional contouring. In the older child with a thicker, less malleable skull, the pericranium is left attached to the bone when the craniotomy is done. A series of radiating osteotomies is created on the inner surface of the bone that incorporate the inner table and extend well into the diploë without cutting through the outer table. The bone then is fractured using the pericranium as a hinge. This is a useful way to tailor the contours of hard bone.

The use of split-thickness grafts has many advantages and remains our main source for repair today. The infection rate is low—0% in my experience, and the risk of rejection is nil. The natural bony union that results grows with the child, an important consideration in the younger patient.

Costochrondal rib grafts have long been used in various types of cranioplasties. Ribs can be obtained from the patient in any of a number of surgical positions. Anteriorly, the fourth through the sixth ribs are easily obtained; posteriorly, the eighth through the tenth ribs are accessible below the scapula; and in the lateral position, with the patient's arm abducted, the fourth through the eight ribs are most accessible.

The craniotomy site is exposed and prepared in standard fashion with fresh bone margins. The area is measured and the graft size determined. The rib grafts then are harvested. An incision is made directly over the ribs and carried down to the rib periosteum with a monopolar cautery using a needle tip. The periosteum then is separated from each rib using a Key-type periosteal elevator. In elevating the rib, the neurovascular bundle and underlying periosteum and pleura are left intact and gently stripped away using a pigtail rib separator. In determining how much rib is to be removed, beyond what is required to fill the defect, it is extremely important to allow an additional 5 to 8 mm for bending and contouring. Each end of each rib is cut, removed from the field, and split longitudinally with a sharp osteotome, yielding two pieces that can be bent and contoured with a Tessier rib bender. Ribs are malleable and fracture easily; therefore, the splitting must be done slowly and carefully. A useful technique is to use the osteotome like a knife blade and slowly slice through the rib. This gentler technique results in a rib that is more stable and easier to shape. The ribs then are positioned at the donor site and shaped to fit the size and contour of the defect. It is desirable to create a graft whose height and contour are slightly exaggerated because with time there is typically some collapse and resorption of the graft. The rib is the most malleable of the materials available, and so it is particularly applicable to reconstruction around the orbits and over the forehead.

The following are some key points to keep in mind when harvesting ribs and using them in reconstruction:

- Never take more than two adjacent ribs; otherwise, a "flail" chest might develop.
- Always keep the rib periosteum intact. This will allow a rudimentary rib to form in the donor site.
- If the pleura is violated and a pneumothorax occurs, place a no. 10 French red rubber catheter over

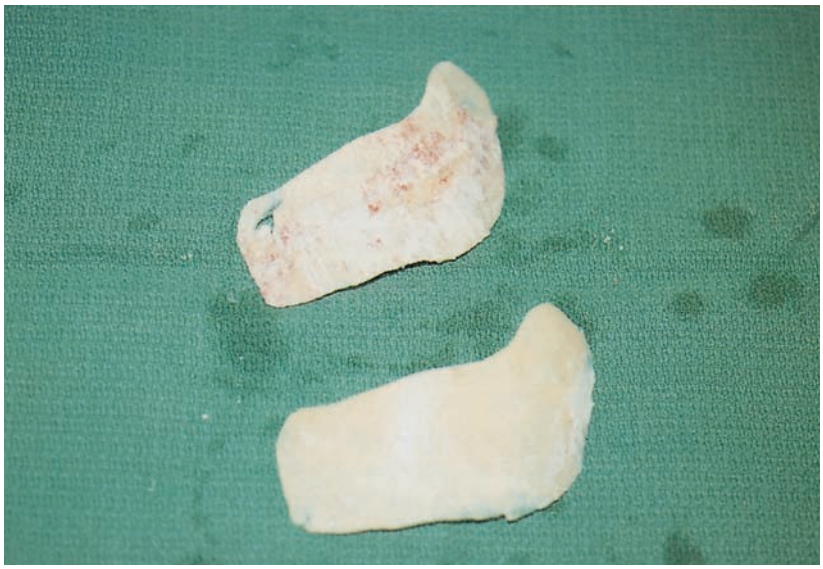


FIGURE 18–6. The calvarial bone after split showing the two units of obtained, one for the donor site and one for the graft site.

the repaired pleura defect during a Valsalva's maneuver. The red rubber catheter is placed in a low water-sealed suction apparatus with 20 mL of water pressure applied. Once a chest radiograph shows the pneumothorax to be resolved, the tube is removed and an occlusive dressing is placed over the wound. This tube usually can be removed in the recovery room.

- In shaping the ribs at the donor site, exaggerate the contour to allow for the resorption that will naturally occur.
- In the female child, the best exposure for taking the rib is through an inframammary incision. Such scars are well hidden and heal nicely.

Despite the usefulness of rib grafts, given the problems with resorption and the "washboard" effect that can occur with lattice-type reconstruction, we reserve this technique only for cases in which calvarium is not available. Rib grafts look best in the initial 2 to 3 years; 10 years after repair, the aesthetic effects are much less gratifying.

The iliac crest can provide a fair amount of bone without entailing significant morbidity to the child. The major problem with this bone source is that the available bone is limited in quantity and possesses minimal contour. In the young child (i.e., <3 years of age) the bone is fibrous and less "osseous" and can be quite difficult to shape and mold. The anatomic location can lead to complaints of pain at the belt line in the older child. Despite these reservations, the iliac crest remains an adequate source of bone in situations where calvarium or rib bone is not available.

To harvest iliac bone, the iliac crest is identified. A linear incision then is made 1 to 2 cm below the crest. This location is chosen to avoid excessive scar formation and the painful "belt-level" scar. The incision is carried down through the subcutaneous tissue to the muscle. The muscle aponeuroses of the internal and external oblique muscles are incised and the transversalis fascia exposed. This fascia then is excised from the internal table of the iliac bone. The abdominal contents are retracted medially. By following the subperiosteal plane and depending on the size of the patient, up to 4 to 5 cm of iliac crest can be exposed. Using osteotomes and an oscillating saw, a premeasured unit of bone is harvested. The site is then packed with Surgicel or Gelfoam soaked in thrombin. Bone wax is applied to any large emissary veins in the bone. A closed Jackson-Pratt drain is tunnelled subcutaneously away from the surgical site and removed after 24 hours. Fascial layers are reapproximated and closed, and a multilayer closure is accomplished. Because the harvested iliac bone is bilamellar, it can be split, yielding two units of bone for the cranioplasty. Iliac bone is cancellous, like calvarium, and entails less resorption than rib. In addition, this bone is revascularized much more rapidly than rib and so is more rapidly incorporated into the calvarium. The disadvantages are mainly that iliac bone is more brittle, less malleable, and less amenable to contouring than rib. Iliac crest is not a good source of bone in regions such as the vicinity of the orbit where aesthetic considerations are important.

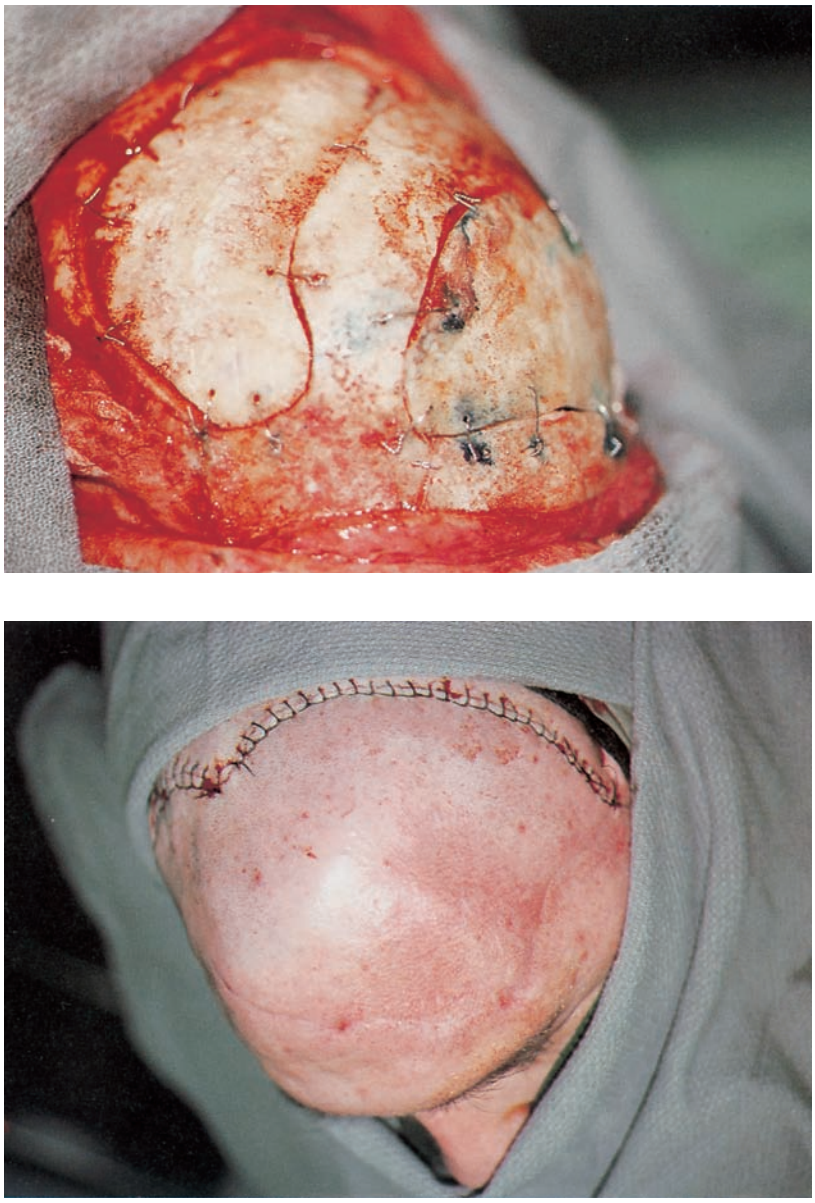


FIGURE 18–7. A, B: The two split pieces of bone have been placed in position at the graft site and the donor site. Full bone coverage on both sides has now been completed, and this bone, being autologous, will grow with the child.

Implantable Cranioplasty Materials

Neurosurgeons are continuing to search for the ideal material for repair of bony defects in the skull. This ideal material needs to be inert, malleable, sterile, durable, biocompatible, and inexpensive. In the infant and child, another factor is important: the ability to grow with the child. To my knowledge, no material has been produced that meets all these criteria.

Several problems are seen with foreign material implanted in the growing child. Because metal implants conduct heat and cold, they are uncomfortable during weather changes. Acrylic resins, which are not incorporated biologically, always remain "foreign" and do not grow with the child. Foreign materials of any type degrade with time and entail increased risk of infection. Probably most disturbing to the pediatric neurosurgeon is the return of a child several years after repair whose "foreign" donor material is now free floating, no longer in contact with the bone margins, and providing none of the intended protection and stability. For the sake of completeness, however, some of the cranioplasty materials available to the neurosurgical community are discussed.

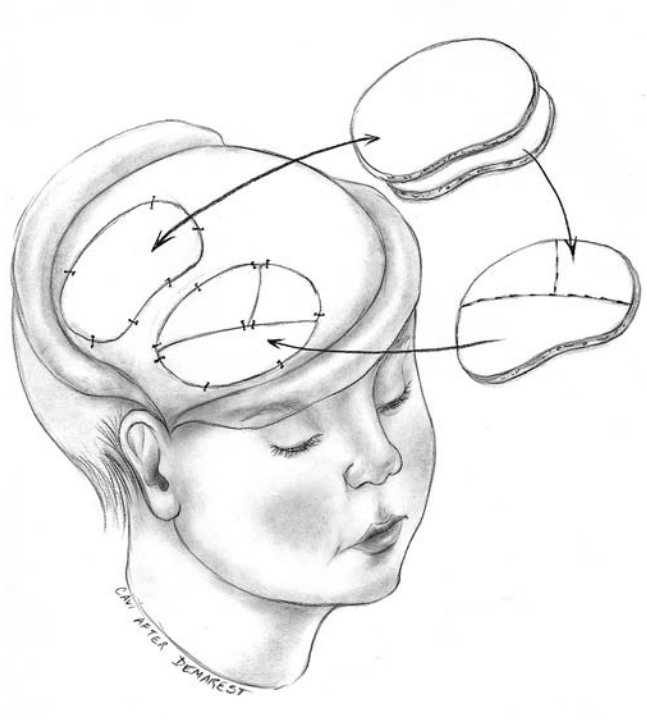


FIGURE 18–8. Illustrated here is the use of "curved" bone being harvested from the right frontotemporal bone to fill in a traumatic defect that occured over the right frontoorbital region. A mirror image piece of bone is harvested, split, and then contoured to fill into the defect site.

Methyl methacrylate remains the most used cranioplastic material in the adult neurosurgical community. When combined with a wire mesh, it becomes durable, malleable, and amenable to shaping as needed. The graft site is prepared as described already. The bone edges are freshened and any fibrotic material removed. The bone edges should be bleeding slightly, with any heavier bleeding controlled with Avitene or Gelfoam (not bone wax). Any dural tears must be repaired. The edges of the craniotomy are prepared with a "stepoff" edge using a squared-edged burr. The stepoff along the bone edge prevents the wire mesh from slipping and penetrating the dura. On this ledge, the shaped contoured wire mesh is placed. The contour of the mesh always should be slightly lower because the methyl methacrylate will be placed on top the wire. The wire mesh used is made of titanium rather than the formerly used stainless steel (Fig. 18–9). With the increased use of magnetic resonance imaging (MRI) and CT, the fact that titanium causes less artifact on scanning has become a significant advantage.

Methyl methacrylate is provided as a powder and a liquid catalyst, which are mixed to form a paste that has a putty-like consistency; if applied when too liquid, it will run off the field. Using a moistened gloved finger and paying close attention to desired contours and height, the paste is applied in layers. Any rough edges are smoothed out before the material sets. Under no circumstances are any of the edges of the wire mesh left exposed; failure to cover these edges may lead to erosion through the skin edges. As the methyl methacrylate dries, it becomes extremely exothermic, and so constant irrigation must be applied until the flap is completely dry; otherwise, the dura and underlying brain will be burned. A useful surgical trick is take a chunk of the methyl methacrylate paste and hold it in the hand while irrigating the plate, which allows the operator to gauge the temperature of the paste as it dries. Once the methyl methacrylate has completely set, a check is done for any rough or sharp edges. These edges are smoothed down with a high-speed round burr before closure. Copious irrigation is crucially important to remove dust and debris.

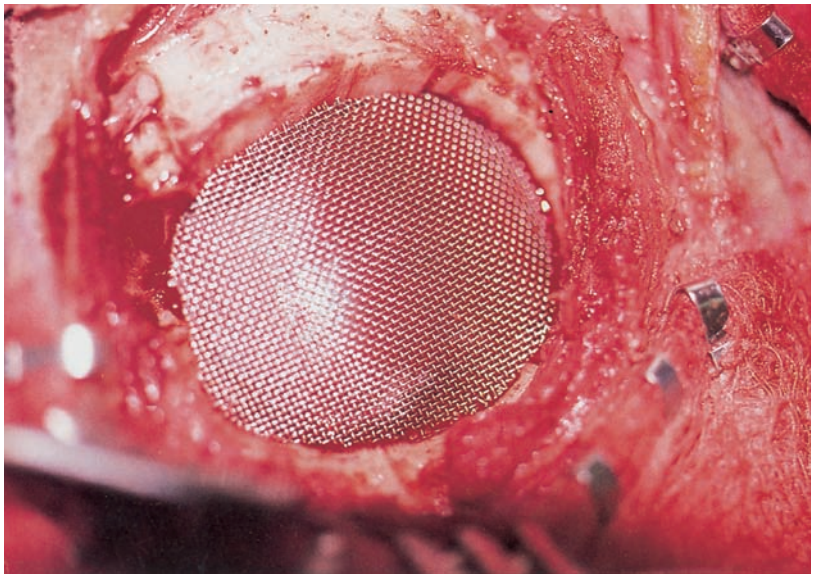


FIGURE 18–9. A craniotomy site prepared with the defect exposed and a titanium wire mesh in place prior to placement of the acrylic material. The mesh has been contoured to match the skull outline, and the edges of the mesh have been placed on a ledge of bone with no edges protruding up. The contour of this mesh has to be lower than the normal contour of the skull because a layer of methylmethacrylate will be placed on top. Not reducing the profile of the mesh will generate too high a cranioplasty.

A number of substitute materials have been proposed as the "ideal" bone replacement. One of these is a substitute material frequently used in maxillofacial surgery: hydroxyapatite. Hydroxyapatite is derived from native sea coral, which is triphosphorylated under high pressure, and then reaminificated to form a matrix resembling that of bone. When placed within a region where there is bone, the matrix is filled by ingrowth of native osteocytes. Hydroxyapatite is available in blocks that can be shaped or contoured with high-speed drills. Special contoured pieces can be ordered that are shaped on computer-assisted design–content addressable memory (CADCAM) devices. A paste form of hydroxyapatite has been used, mixed with saline and then applied to the defect, allowed to dry, and hardened. The theoretical advantages of this material led to its initial enthusiasm. No donor graft was required, contoured pieces could be obtained, and the ingrowth of normal bone appeared to make this material ideal for the growing child. Experience has shown, however, that these ideals could not be realized in practice. The material is hard to shape and extremely brittle—rendering it more susceptible to fracturing in children, who are less cautious in contact behavior than adults. Finally, bone ingrowth appears not to have been as extensive as one would like. Despite all these reservations, there may still be clinical indications for use of this material. In particular, this paste may have the most promising long-term application, although its present expense remains prohibitive.

Cadaver bone as a cranioplasty material is again being used. The principle behind cadaver bone is to provide a matrix for new bone ingrowth. The best source of bone appears to be that which has been freeze-dried; chemically treated or irradiated bone undergoes severe denaturation of base proteins, increasing the risk of rejection and resorption. Even bone that has been freeze-dried undergoes significant resorption, which can be uneven, leading to aesthetically undesirable outcomes, particularly when applied to the orbitofacial regions. Cadaveric material should be considered only when there is a lack of autologous donor sources.

POSTOPERATIVE MANAGEMENT

Postoperative care and management principles for a patient with a cranioplasty are straightforward. Sterility and a clean wound are key to reducing postoperative infection. In children who tend to be aggressive in removing head dressings, we often apply a chin strap to the turban portion of the dressing. Full head wraps are almost always the rule in my practice; this is done to keep away prying, dirty fingers. The head dressing should be snug but not so tight as to compromise the vascular supply to the wound. The use of drains remains unresolved in cranioplasty. We have stopped using them in our practice because our incidence of infection is lower without them; however, this point remains open to discussion, and the placement of drains following cranioplasty still should be dictated by the surgeon's preference until more conclusive data are available.

Antibiotics are used for 48 hours after the cranioplasty (oxacillin 25 mg/kg of body weight). We never routinely use steroids or anticonvulsants in these cases.

The child is mobilized as soon as reasonably possible. The parents are counseled about care of the wound, the use of helmets (if required), and the time it takes the bone or implant to heal. In the toddler or young ambulating child (younger than 3 years), we request that the child be observed closely for prospective falls in the first 3 months after surgery. In the 4 to 6 months after surgery, they do not need to be as attentive, but serious falls still need to be avoided. In our experience, the patient's siblings bring most of the problems in handling and care. Younger siblings tend to be more aggressive in playing and handling of the child; therefore, parents or caretakers need to be cautioned about this potential behavior. We do not routinely obtain postoperative radiological studies unless clinical problems arise.

EDITOR'S COMMENTARY

The timing of cranioplasties in children of less than five years of age is sometimes indeterminate because of a child's potential to fill in some skull defects spontaneously. Defects of 2 cm often fill in on their own but larger ones are unlikely to fill in satisfactorily. We agree with Goodrich that calvarial bone is usually the optimal autologous bone source and with his views about alternative materials. His admonition about blood loss is important; dissection of dura away from the margins of a skull defect can be both difficult and bloody. The size of skull defects themselves generally do not increase after cranioplasties are performed; the adjacent calvarium should grow normally so that the size of the defect filled in with methylmethacrylate should not enlarge unless there is underlying hydrocephalus. I (ALA) have had good results combining tantalum mesh with methylmethacrylate, implanting a piece of mesh about 5 mm larger than the bone just below the defect, then drilling a shelf in bone around the defect edges, and filling in the defect with methacrylate. The cranioplasty will not migrate externally because the mesh is larger than the bone opening, and will not migrate inward because of the bone shelf around the periphery.

PEARLS

In this author's experience:

- Autogenetically harvested material (e.g., skull, rib, iliac crest) is normally the best replacement material to use in a cranioplasty. This type of material provides a lower infection risk and will grow with the child.
- There is no contraindication to replacing open contaminated bone in a pediatric head trauma case. As long as the bone is debrided and cleaned and sterilized in diluted betadine solution, the replacement of this bone does not increase the risk of infection.
- In replacing bone, the use of rigid fixation helps reduce resoption of bone and reduces the risk of slippage or displacement.
- For craniofacial reconstruction, the use of calvarial bone remains the best harvest site; this bone has the lowest risk of infection and the lowest risk of resorption and provides the best area for finding appropriately contoured bone.
- It is best to minimize the use of foreign materials such as bone wax and Gelfoam when replacing bone flaps. Bone wax in particular reduces bone healing and can act as a nidus for bacteria and potential infection.

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19

SUPRACLAVICULAR APPROACH TO BIRTH-RELATED BRACHIAL PLEXUS INJURY

John P. Laurent

The role of pediatric neurosurgeons in the treatment of infants with birth-related brachial plexus injuries has increased steadily over the last 10 years. Previously, these children were referred to orthopedic and plastic surgeons, neurologists, and physical therapists.

Symptomatology of birth-related brachial plexus injuries is usually first observed by the obstetrician, midwife, or infant's family shortly after birth. Obstetric factors related to brachial plexus injury at birth include a multiparous mother, prolonged labor, high birthweight, and vertex delivery with shoulder dystocia. Brachial plexus injuries have been seen in premature births and cesarean sections. These injuries are more common with breech deliveries; this presentation is relatively rare compared with vertex presentations.

INDICATIONS AND PREOPERATIVE EVALUATION

Most children who present with birth-related brachial plexus injuries demonstrate classic Erb's type upper plexus lesions of roots C-5 and C-6. The arm is clinically limp, the shoulder abducted with its upper part internally rotated, the elbow extended, and the forearm pronated with flexed wrist and fingers. Other levels of involvement can include C-7, which causes triceps paresis and some insufficiency of wrist and finger extension. A C-4 root lesion causes phrenic nerve palsy with accompanying respiratory problems. In contrast to radiculopathies, brachial plexus neuritis, and tumors among adults, the birth-related brachial plexus syndrome does not include pain. Patients with classic Klumpke's syndrome, a variety of brachial plexus injury rarely seen today, present with flexion in the elbow and extended wrist without finger movement; this syndrome involves the C-8 and T-1 roots, often with avulsion. The presence of Horner's syndrome is diagnostic of C-8 and T-1 level lesions. Dysfunction of the dorsal scapular (rhomboid muscle) nerve or the long thoracic (anterior serratus muscle) nerve indicates a root-level injury. A lesion of the suprascapular (supraspinatus and infraspinatus muscles) nerve indicates an upper-trunk lesion. Combined total plexus lesions from C-5 to T-1 constitute the remaining cases.

The entire plexus is involved at birth in most infants, with the neurologic level definitely established at age 6 to 8 weeks. Upper-plexus injury may be present without lower-plexus involvement, but lower plexus injury usually is not seen without some degree of accompanying upper plexus damage.

Clinical neurologic evaluation allows assessment of the progression of a birth-related plexus injury. Accurate motor assessment of an infant is difficult; observation and reward-induced movements are the primary means of evaluating muscle strength and range of motion. Preoperative and postoperative electrodiagnostic testing [electromyelography (EMG), somatosensory-evoked response, and upper-extremity H-reflexes] are helpful in confirming the diagnosis, but these tests are not regarded as definitive indicators for surgical intervention. Intraoperative electrodiagnostic testing (EMG and somatosensory- and motor-evoked responses) is invaluable in delineating lesions on the operating table. Multiple muscles must be assessed in the effort to ensure future "good" functional outcome from surgery.

The ability to use the limb is the best indicator of function, and, when possible, the Mallet scheme is invaluable. A diagnostic workup in an infant or young child should include careful inspection of the injured extremity from a distance and observation of how the child uses it: Does he or she reach for objects? If so, how are the objects handled? Finger and hand moisture (*autonomic function*) may be observed or may be evaluated either by iodine test or by Wickler's test. In addition to movement, muscle atrophy, Horner's syndrome, and withdrawal from pinprick should be assessed. Any or all these tests may be difficult in an overweight newborn.

INTRAOPERATIVE TECHNIQUES

Anesthesia and Positioning

The patient is placed supine on the operating table. Anesthesia is induced, avoiding paralyzing agents, and less than 2.0% fluorane levels are used. Bilateral surface skull electrodes for electroencephalograms are placed 3 cm from the sagittal suture and 8 cm from the coronal suture on the parietal region of the skull. Bifrontal skull electrodes also are placed. Needle electrodes are positioned at the base of the skull, over the C1–2 region. The reference electrode is the *inion.* Dermal electrodes are placed on the ipsilateral rhomboid, infrascapular, suprascapular, both heads of the deltoid, both heads of the biceps, triceps, extensor carpi radialis, flexor carpi radialis, adductor pollicus minimus, abductor pollicus brevis first interosseous, and adductor minimus V muscles. The head is turned to the contralateral side. The ipsilateral arm is extended, lowering the clavicle on the affected side. Identification of the sternal notch, the acromiun process, and the position of the clavicle permits palpation of the brachial plexus in the sleeping patient. Following thorough cleaning of the surgical site, both anterior cervical neck areas as well as both lower extremities are draped in a specially adapted clear plastic drape that permits assessment of upper extremity muscle movements during direct stimulation of nerve of the brachial plexus (Fig. 19–1).

Exposure

A curvilinear incision is made on the lateral border of the sternocleidomastoid muscle to the medial portion of the clavicle and then carried laterally 1 cm below the clavicle to the axillary junction. Evidence of prior subcutaneous injury with xanthochromic staining of scar tissue, dense scar tissue of ruptured muscles, thick fascial-layer injury, induced fatty degeneration, increased vascularity, and attempted regrowth of neural tissue will blanket the normally loose adipose–fascial plane over the brachial plexus. No distinct surgical planes of cleavage will be present.

Dissection and Exploration

The internal jugular vein is carefully dissected to its junction with the subclavian vein under the clavicle. The carotid artery is palpated medial to the jugular vein. The external jugular vein is ligated and coagulated at its junction with the internal jugular vein. A large transverse vein superior to the scalene anterior muscle is usually present and is ligated to permit access to the distal phrenic nerve. The entire length of the phrenic nerve is carefully dissected from the anterior surface of the scalene anterior muscle and noted to be intact morphologically. The proximal phrenic junction with the C-4 nerve root is identified. The C-4 root is explored to its neural foramen and its exit from the spinal cord. The distal C-4 nerve is dissected to expose the three distal branches, which are freed to their entrance into the colli muscles of the neck. Identification of the XI cranial nerve distal branches into the sternocleidomastoid muscle and exposure of the XII cranial nerve's hypoglossal division are made to prevent injury to these structures. The C-5 nerve-root contribution to the phrenic nerve is exposed, and the C-5 nerve root is dissected to its neural foramen. Dense postinjury scar tissue is usually adherent to a neuroma. The overlying thinned, scarred, ruptured platysma and cervical muscles are identified.

The omohyoid muscle is transected at its medial origin near the internal jugular vein and distal transection as it passes inferior to the clavicle. Removal of a postinjury scarred fat pad usually (95%) reveals a fusiform neuroma. Proximal exposure of the neuroma will expose the previously dissected C-5 nerve root. Identification of

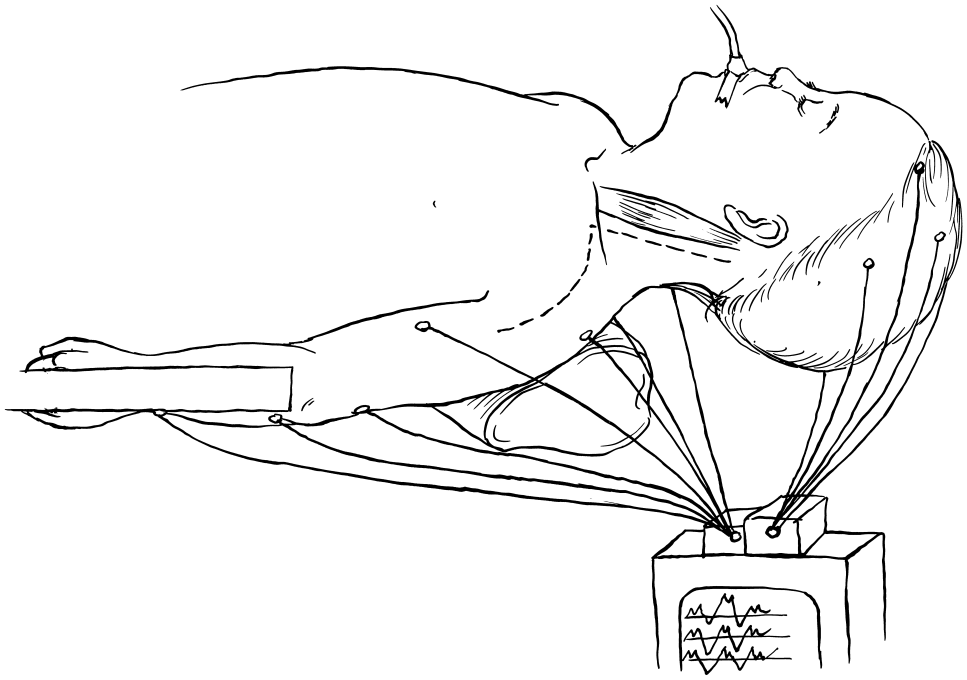

FIGURE 19–1. Operative layout for brachial plexus operation. Note the placement of electrodes and the extension of the arm.

the suprascapular peripheral nerve to the infrascapular and suprascapular muscles as it exits from the upper trunk permits exposure of the distal portion of the neuroma. Vascular loops placed around the nerves and around the neuroma are for retraction purposes. Before proceeding with the distal dissection of the neuroma, the subclavicular area structures (subclavian artery and vein) need to be identified. The scar tissue surrounding the distal brachial plexus divisions and cords and peripheral nerves near the clavicle is removed. Vascular loops are placed around the posterior division of the upper trunk, the upper trunk, the anterior-division musculocutaneous nerve complex, the posterior division of the middle trunk, middle trunk, and lower trunk of the brachial plexus (Fig. 19–2).

Medial–inferior exploration of the brachial plexus exposes the lower trunk of the brachial plexus. The subclavian artery extending toward the carotid artery, the pleura of the lung, and the entrance of T-1 nerve root into its neural foramen are identified. Vascular loops are placed around the T-1 nerve root and the subclavian artery. A constant large vessel originating from the subclavian artery and crossing over the medial portion of the lower trunk is hemiclipped, coagulated, and transected. A fibrous plane of tissue separates the lower trunk from the middle trunk. Dissection in this plane frees the lower trunk and permits identification of the posterior–interior portion of the middle trunk. Proximal–medial exploration of the inferior portion of the middle trunk permits exposure of the C-7 nerve root at its entrance into the neural foramen. A vascular loop is placed around the C-7 nerve root. Exposure of C-6 is noted as it enters its neural foramen. A vascular loop is placed around the C-6 nerve root.

The neuroma is carefully dissected free from its scarred muscle attachment and adipose connection. Neuroplasty and external neurolysis are performed on the total brachial plexus exposure, exposed peripheral nerves, and the neuroma. The brachial plexus is untethered.

Intraoperative Monitoring

Neuroelectrophysiologic testing for nerve action potentials, muscle action potentials, and evoked parietal

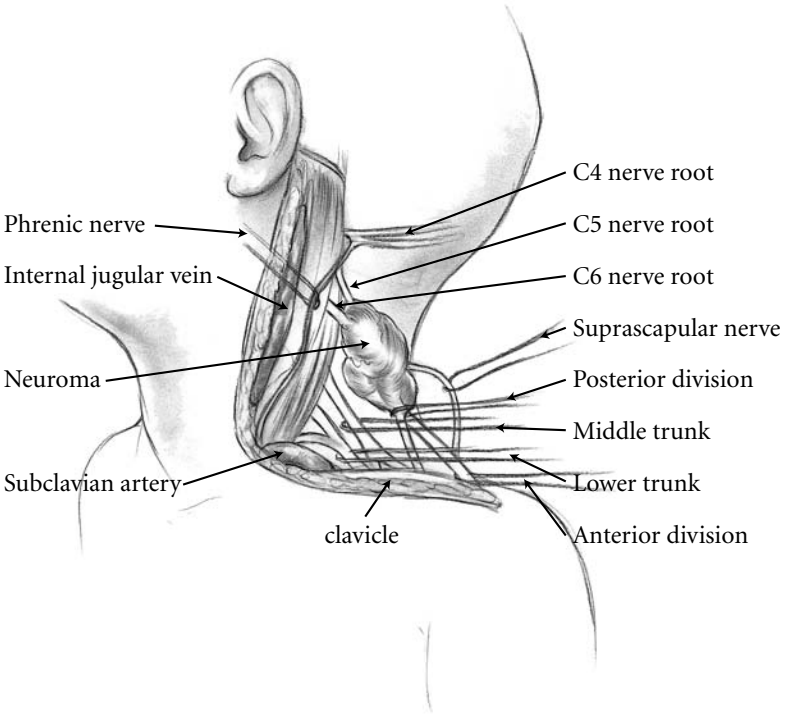

FIGURE 19–2. Representation of an actual operation for an upper brachial plexus neuroma from birth injury. Note the vascular bands placed around individual nerve roots and vascular veins placed around all distal branches that are dissected in the supraclavicular area.

responses are completed at this time using the previously placed electrodes on arm, forearm, hand, and skull. A bipolar hand-held electrode specifically designed for intraoperative use, with directional polar capabilities, is placed on nerve roots, trunks, divisions, cords, and peripheral nerves of the brachial plexus. Initially, the suprascapular nerve is stimulated, directed toward, and measures the muscle action potential amplitude, latency, and wave form of the infraspinatus/supraspinatus muscles. The next step is directed stimulation of the posterior division of the upper trunk/axillary nerve toward and measurement of the muscle action potential amplitude, latency, and wave form of the deltoid muscle, biceps, and triceps muscles, followed by the anterior division–musculocutaneous complex toward and measurement of the muscle action potential amplitude, latency, and wave form of the biceps, wrist, and finger flexor muscles. This is followed by the middle truck/radial nerve toward and measurement of the muscle action potential amplitude, latency, and wave form of the triceps, wrist, and finger extensor muscles, which is followed by the anterior division of the middle truck toward and measurement of the muscle action potential amplitude, latency, and wave form of the extensor muscles of the wrist and fingers and lower truck/ulnar nerve complex toward and measurement of the muscle action potential amplitude, latency, and wave form of the flexor and adductor muscles of the hand, finally followed by the posterior division of the lower trunk toward and measurement of the muscle action potential amplitude, latency, and wave form of the interosseous muscles of the hand. Electric bipolar stimulation of the nerve roots directed toward distal muscles helps to identify ruptures and nonconducting neuromas (Fig. 19–3). The C-5 nerve root is proximally stimulated, at its entrance to the neural foramen, to attempt electrically to induce conduction in axons across the upper trunk to the suprascapular nerve, the posterior division of the upper trunk to the middle trunk/axillary nerve, and the anterior division–musculocutaneous complex and to measure the muscle action potential amplitude,

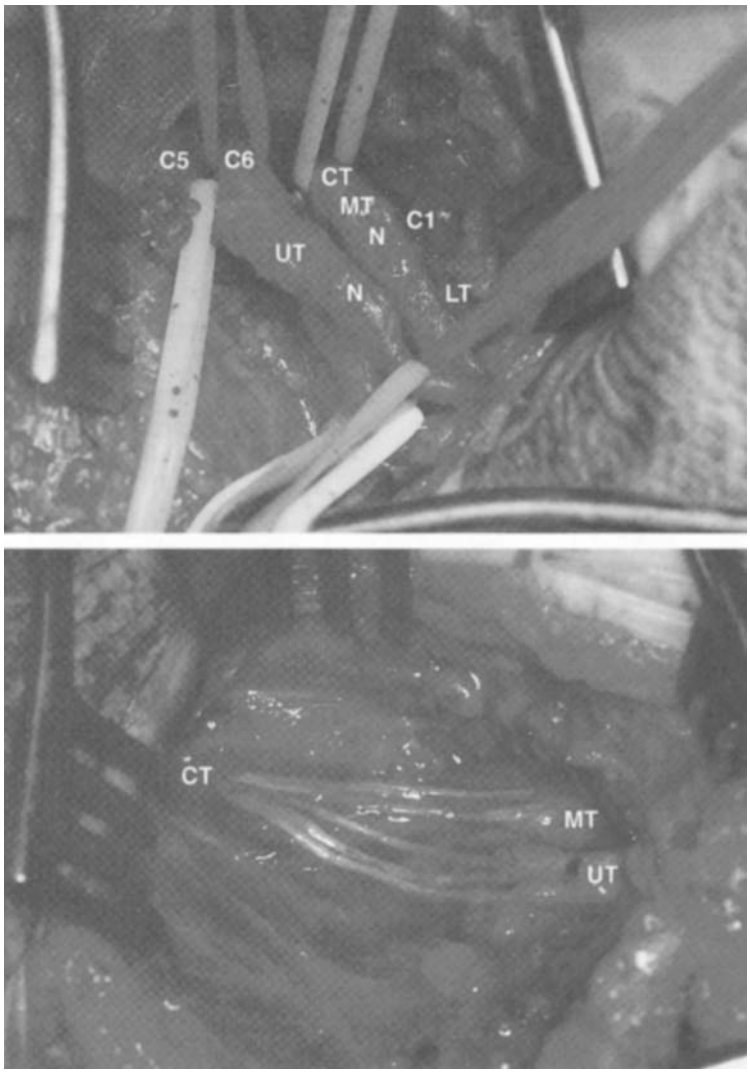

FIGURE 19–3. A: Exposure of the C-5, C-6, C-7, and C-8 nerve roots; upper trunk (*UT*) and division; and middle trunk (*MT*) and lower trunks (*LT*) shows a neuroma (*N*) in both the upper and middle trunks. Intraoperative electrophysiologic data demonstrated a lack of conduction through the upper trunk neuroma, decreased conduction through the middle trunk neuroma, and avulsion of the C-6 nerve root. **B:** Multiple sural nerve cable grafts from the C-5 nerve root to the transected upper trunk and to the side of the middle trunk are depicted.

latency, and wave form of the individual muscles respective of their nerve supply. The C-6 nerve root is proximally stimulated, at its entrance to the neural foramen, to attempt induced conduction in axons across the upper trunk to the posterior division of the upper trunk to the middle trunk/axillary nerve, the upper trunk/cord to musculocutaneous, and median nerve, and to measure the muscle action potential amplitude, latency, and wave form of the individual muscles respective of their nerve supply. The C-7 nerve root is proximally stimulated, at its entrance to the foramen, to attempt to induce electrically conduction axons across the middle trunk/radial nerve and the anterior division to the upper trunk to the median nerve to measure the muscle action potential amplitude, latency, and wave form of the individual muscles respective of their nerve supply delineated in the preceding description. The C-8 nerve root is proximally stimulated, at its entrance to the foramen, to attempt to induce electrically conduction in axons across the lower trunk/ulnar nerve and posterior division of the lower trunk to measure the muscle action potential amplitude, latency, and wave form of the individual muscles respective of their nerve supply. The T-1 nerve root is proximally stimulated, at its entrance to the foramen, to attempt to induce electrically conduction in axons across the lower trunk/ulnar nerve and posterior division of the lower trunk to measure the muscle action potential amplitude, latency, and wave form of the individual muscles respective of the nerve supply. To access nerve-root continuity to the spinal cord (to rule out avulsions), evoked action potentials are recorded over the parietal region of the brain and C1–2 cervical spinous process. By reversing the polarity of the stimulating electrodes so that the current is directed toward the spinal cord, each nerve root, at its entrance to the neural foramen, is stimulated. Each nerve root (C-5, C-6, C-7, C-8, T-1) requires multiple 3-

minute recordings to determine whether the nerve root is intact to the spinal cord.

Using the preceding procedures, certain surgical parameters are determined for neuromas. End-to-end interpositional sural nerve (auricular, C-4) grafts are placed if no conduction is elicited across the neuroma. A side (partial)-to-side (partial) bypass graft is placed when conduction is present across the neuroma. Ruptures are treated by interpositional end-to-end grafts. Avulsions are treated with neurotization from an intact nerve root.

POSTOPERATIVE MANAGEMENT

The child's arm is immobilized in a sling for 2 to 3 weeks after surgery, at which time full physical therapy is instituted. Sling and dressings are changed daily by the parents. Follow-up depends on patient proximity to the hospital, but it is usually every 6 months up to the age of 3 years and yearly thereafter. Additional trauma to the shoulder joint must be prevented. The parents should be instructed to avoid abduction and posterior projection of the shoulder and must support the limb when holding the child. Sensory feedback is important for future use of the limb. Electric stimulation has no documented value.

The child's attainment of a functional hand and arm and the ability to feed itself are the final measurements of recovery. Recovery of muscle contractions without functionality is considered a disability, and evidence of electrical activity in muscles and nerves is not a predictor of functional muscle return. The operative window for birth-related brachial plexus trauma is similar to that for traumatic brachial plexus injury occurring later in life, that is, 4 to 6 months postinjury if no functional recovery is observed. Of interest in this pediatric population is the fact that brain maturation has yet to be accomplished, making reeducation possible, an advantage not enjoyed by the similarly injured adult population.

EDITOR'S COMMENTARY

The true challenge in children with brachial plexus injuries is the timing of the decision to proceed to surgery or to continue to follow the child that is progressing. Clearly, a nonfunctional limb by 4 months of age is concerning and a likely candidate for surgical intervention. It is the child at 6 months of age who continues to improve though still having a significant amount of deficit who represents the dilemma and the controversy. The author has been noted to espouse early surgery. Others, later. Obviously, non-intervention is preferable, if possible, though weighing the deficits with the child's plasticity makes early intervention desirable. In this editor's experience (PDA), holding off surgery until 6–9 months of age does not lessen outcome in this group of children. In relation to the surgery, the editor uses a transverse cervical incision using the child's skin creases to hide the scar. The flexibility of the skin in the infant is such that the anatomy from C4 to subclavicular region can be fully appreciated with a more appealing scar. In relation to the decision of grafting, the point made with regard to side-to-side neurorraphy is an important one. Data in laboratory studies have shown that there is no difference in graft response compared to end-to-side. Obviously, conduction, no matter how little across the neuroma, should be preserved with the use of side-to-side graft.

PEARLS

In this author's experience:

- 99% of children will have a neuroma that, in 95% of cases, occupies the upper trunk and occasionally the middle trunk. Lower trunk neuromas are rare.
- A large vessel will be seen transversing the C7–8 junction of nerve roots. This is not the transverse cervical; rather, this is a direct vessel from the subclavian artery that lies deep to the omohyoid muscle.
- In most cases, the omohyoid muscle is entirely ruptured as a result of the stretch injury and cannot be used as an identifying landmark.

SUGGESTED READINGS

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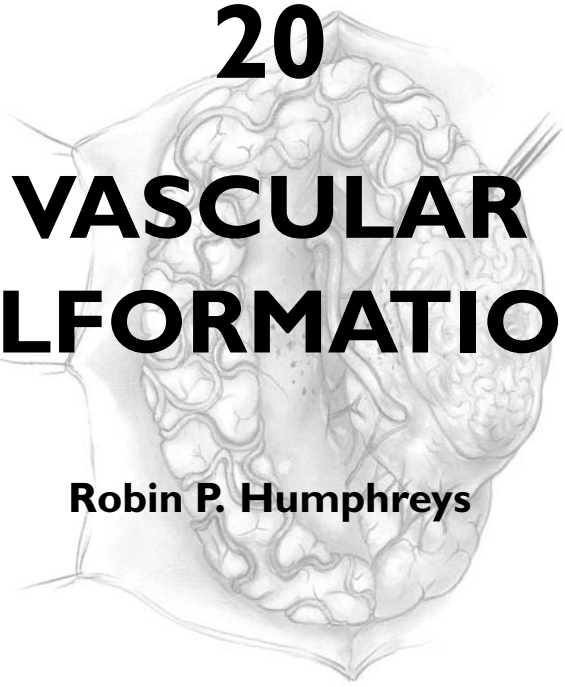
Vascular Disease

20

VASCULAR MALFORMATIONS



Robin P. Humphreys

The presenting symptom of eighty percent of arteriovenous malformations (AVMs) in children is spontaneous intracranial hemorrhage. The hemorrhagic stroke in children, in particular, often has a devastating presentation that demands early intervention. It is now much easier to define the primary cause of the brain hemorrhage and to map a treatment plan that is best suited for it and its attendant phenomena. The operative evacuation of a cerebral clot is no longer the only strategy for a child's impaired neurologic features. Clot removal may be one component of the therapy, but today's technology has resulted in more effective treatments for AVMs in children.

Because the operation for an AVM can be tedious and protracted, it is desirable to plan the procedure as one that is elective. Thus, if only subarachnoid or intraventricular hemorrhage has occurred or if the intracerebral hemorrhage is small and the patient's condition is stable, interval surgery may be staged after a few days' wait. Sometimes when the patient's condition remains unchanged or even improved slightly, it is tempting to delay the operative intervention further, perhaps with a view to examine other, nonsurgical therapies; however, the startling incidents of hemorrhage as the presenting symptom for a child's AVM and the likelihood of repeat hemorrhage as well as the remarkable recoveries that the child may demonstrate immediately after the hematoma is removed all validate an operative solution for this type of stroke problem.

SURGICAL INDICATIONS AND PREOPERATIVE EVALUATION

Successful AVM management is dependent on the AVM's location, size, hemodynamics; the patient's clinical condition; and the treatment method selected. The treatment goals should be to preserve life and neurologic function while achieving complete removal of the AVM and preserving the normal cerebral circulation.

A certain number of children, without warning, will experience the onset of a severe, generalized headache that is not representative of any organic pathology within the cranium. Computed tomography (CT) imaging usually sorts out these cases at the primary, referral level. When the CT scan demonstrates a cerebral hemorrhage from a child's ruptured AVM, blood clot and perhaps calcification are seen; and on enhanced study, the dilated feeding and draining vascular channels and large blood-containing varices may be seen (Figs. 20–1 and 20–2).

Magnetic resonance imaging (MRI) is capable of defining the nature of the hemorrhage as well as the involved vasculature. In reality, however, the urgent nature of the child's condition often mitigates against the consumption of time necessary for a properly performed MRI. Additionally, the surgeon should not rely on the vascular anatomy as revealed on the MRI to inform one fully of its abnormalities. Most AVMs that bleed are small, especially in children. The tiny feeding and drain-

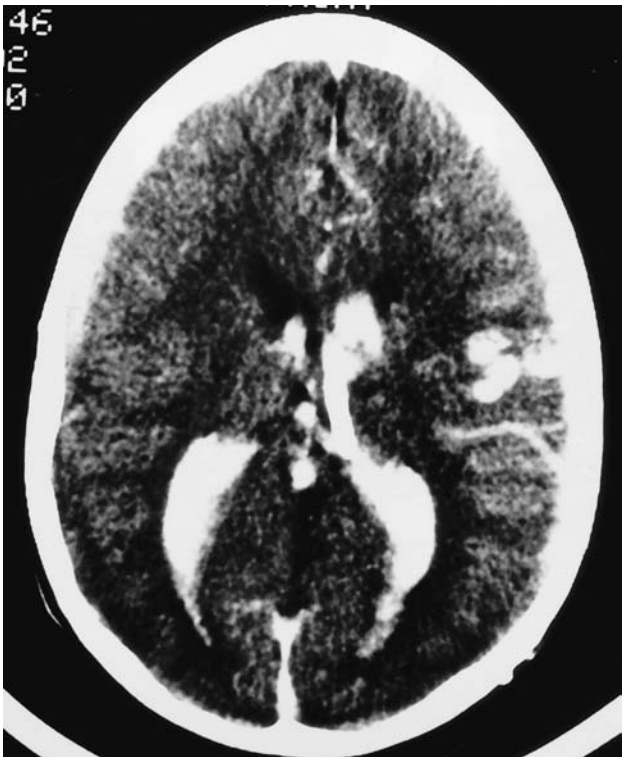


FIGURE 20–1. This CT scan outlines intraventricular and subcortical blood in the left hemisphere. A serpentine enhancing vessel is shown in the anterior parietal region. This study does not confirm whether it is arterial or venous.

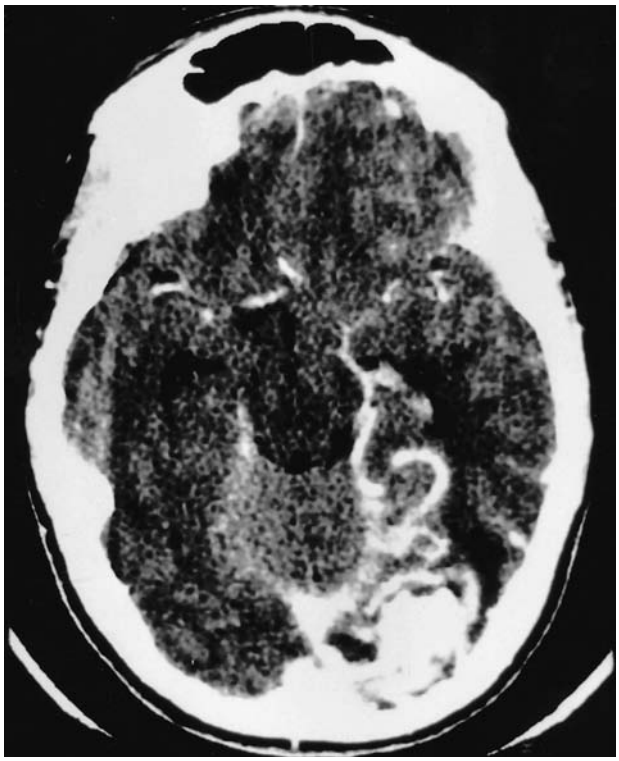


FIGURE 20–2. This left occipital arteriovenous malformation proved to be fed by a large branch from the posterior cerebral artery.

ing vessels associated with a nidus of varying size may be completely obscured on MRI, particularly when there is blood clot present in addition. The MRI is a most valuable adjunct in the elective situation, especially for small lesions adjacent to the ventricular wall or within the diencephalon or brainstem. The valuable information obtained from this imaging technique could change the type of therapy.

Selective carotid and vertebral arteriography, with superselective views if necessary, remains the standard of investigation in a child with an AVM. The numbers and location of the arterial feeders can be examined by films processed with subtraction and magnified techniques (Fig. 20–3). Seldom is a malformation served only by one arterial channel. There may be only one major artery contributing to the lesion, but the surgeon should anticipate any number of minor vessels coming from pial or deep perforating sources, especially in relation to the underlying ventricle. On the other hand, the major venous drainage from an AVM in a child is frequently through a solitary large cortical vein, or one ending in the deep venous system. This venous outflow is usually precisely defined on arteriography.

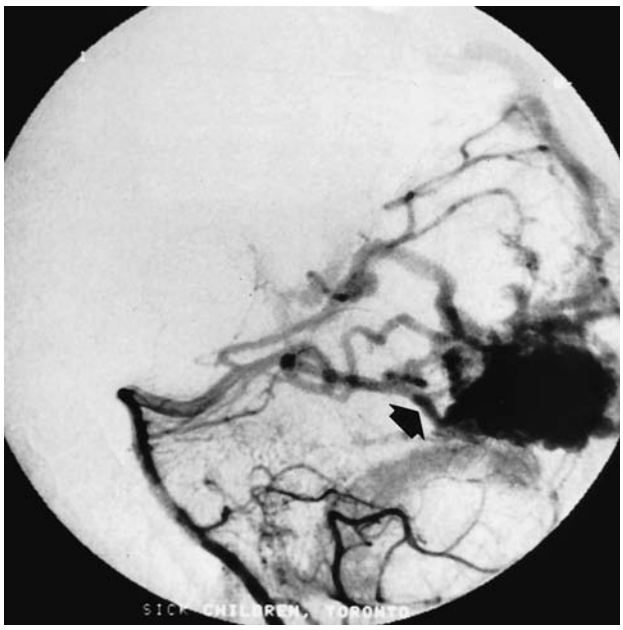


FIGURE 20–3. The direct arterial supply to the occipital arteriovenous malformation is demonstrated (arrow). (Same case as Fig. 20–2.)

PREOPERATIVE MANAGEMENT

If a child's AVM is to be treated in an elective fashion or is part of a staged program in which first the clot is removed and then, after an interval, the malformation is excised, the child should be monitored in a typical neurosurgical acute care fashion.

Cerebral vasospasm is seldom a problem in children with AVMs, but rebleeding during the waiting interval may be. Thus, antispasm, antihypertensive pharmacologic measures usually are not required. If the clot is large, it is advisable to initiate an anticonvulsant drug because of the threat of delayed seizures. If the operation possibly can be delayed for 3 to 7 days after the hemorrhage, the surgeon will be rewarded with a properly prepared patient whose relaxed brain contains a partially liquefying hematoma.

A certain percentage of patients will arrive in a precarious condition because of the size and location of their brain hematoma. The latter may be sufficiently large to obscure vessel abnormalities on CT scan. Under these circumstances, the surgeon has no option but to perform a craniotomy for clot removal. As the cause of the child's cerebral hemorrhage is still unknown at this stage, the surgeon should confirm that it has not arisen from an undiagnosed coagulopathy. With alleviation of brain compression, the stage is properly set for an interval, elective arteriogram.

The urgency is not the same for the 20% of patients whose presenting symptom is other than spontaneous hemorrhage. This group has been investigated in a traditional fashion for seizures, headaches, and perhaps developmental delay. The malformation has been discovered during the testing or even during elective surgery. The clinical challenge is to determine in advance whether surgical obliteration of the malformation will influence the seizure events, headache, or developmental problem and whether, therefore, it should be undertaken.

Anesthetic Techniques and Positioning

With appropriate lines placed and controlled ventilator cycles, the patient has adequate brain relaxation for tissue exposure and dissection. Induced hypotension is not usually required for pediatric AVM surgery, nor is cerebral perfusion pressure breakthrough a frequent complication. Cerebrospinal fluid (CSF) evacuation through a lumbar drain to facilitate brain relaxation is often difficult to achieve, especially in the young child. Finally, the surgeon should decide with the anesthetist whether mannitol is to be administered during the craniotomy.

The patient's body position should be such that maximum comfort is ensured to all the major pressure points and extremity joints in the event that surgery lasts for several hours. This is especially the case with regard to positioning of the shoulders, arms, and cervical spine. The table is flexed appropriately to elevate the head slightly above the heart, and the headrest position should ensure that the area of brain to be explored is parallel to the floor.

If stereotactic localization of the lesion is not to be used, the surgeon must rely on traditional surface landmarks of the skull (and thus the brain) to plan a generous flap. These take into account not only the three-dimensional size and location of the AVM but also the associated hematoma, especially if it extends away from the malformation. No one can be faulted for being overly generous with the scalp and bone flaps. If a localizing technique has been used to isolate a small malformation, especially one located in critical areas of the brain, then the overall exposure and bone flap can be reduced accordingly.

The Dura Exposed

Arriving at the dura, the surgeon may be somewhat concerned that it feels snug or is bulging. If the anesthetic management to this point is ideal, the surgeon should expect that there would be a degree of bulkiness to the brain, even when the hematoma volume is minimal.

If peripheral dural tack-up sutures are part of the surgeon's regular procedure, they should not be put in until the end of the procedure. Doing so beforehand runs the risk of impaling the brain or entering surface vessels of the malformation.

It is unusual for a child's AVM to obtain arterial supply from the external carotid system. Hence, there are few concerns when the dura mater is opened over the lesion, except around the region of the major draining vein. This structure is often so engorged that it becomes lightly adherent to the inner layer of dura, from which it can be separated by means of blunt dissection as the dura is reflected (Fig. 20–4).

With the dura opened (and it should not be reflected more than necessary to visualize the malformation), the surgeon will notice that the brain "pouts" through the opening, indicative of its engorged stage. Unless there has been fresh bleeding, this minimal herniation usually resolves on its own.

Intradural Strategies

A variety of surface hallmarks may guide the surgeon to the site of the lesion. The most common is a generous

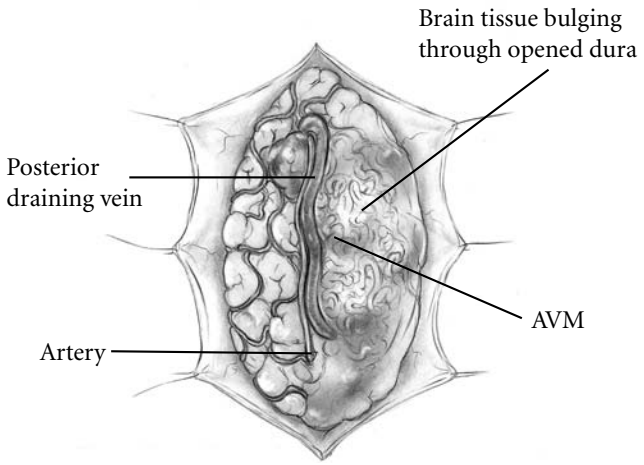


FIGURE 20–4. The AVM and surrounding brain tissue bulge through the dural opening. It is not immediately clear where the supply arteries are in relation to the posterior venous structures.

arterialized draining vein that arises from a sulcus and heads to a major venous sinus. The most reliable surface markings are the veins because, regardless of their origin, at some point they all cross the brain's surface (Fig. 20–5). Other clues include the characteristic coil of vessels spread across the pial surface, thickened, opacified, and sometimes icteric arachnoid that crosses a sulcus to encase the juxtaposed gyri (even when a vascular abnormality is not obvious), or any vessel that does not maintain the typical arborized appearance of a normal cortical artery. Sometimes a lateral hemispheric AVM may not be found curled on the cortex. Instead, it may be hidden subcortically and bare, its only surface landmark a usually straight and feeding artery that runs a nonanatomic course and then dives into a sulcus.

The site for the first incision into the brain is either through a normal-appearing gyrus at the margin of the malformation (Fig. 20–6) and remote to any known critical structure or through an area of cortical discoloration that indicates hematoma. Even in the urgent situation when clot evacuation is mandatory, the surgeon gains nothing by passing an exploratory brain needle in search of the hematoma with the hope that the clot will trickle forth. Instead, a corticotomy is created over the hematoma, which usually has a jelly-like consistency. Evacuation of the hematoma lessens brain tension and can provide the surgeon with some important intracavity anatomic information about the location of the deep

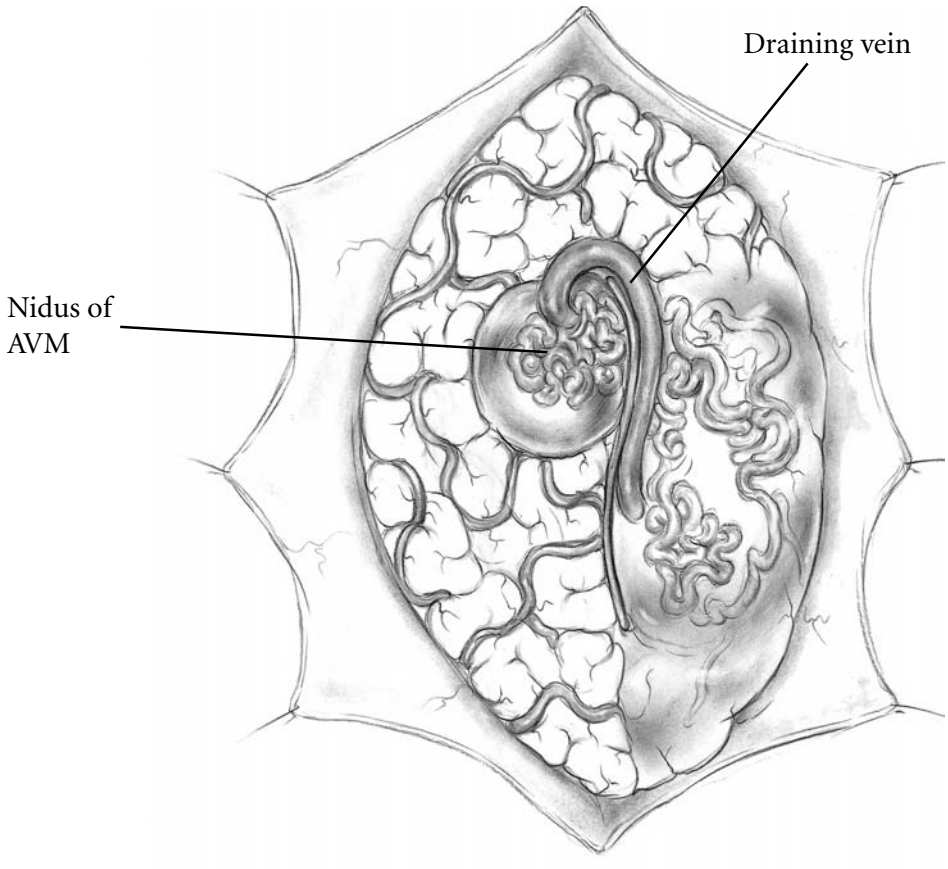


FIGURE 20–5. Part of the nidus of the malformation is quite apparent, as is the draining vein, which is heading off to the nearest major venous collecting system.

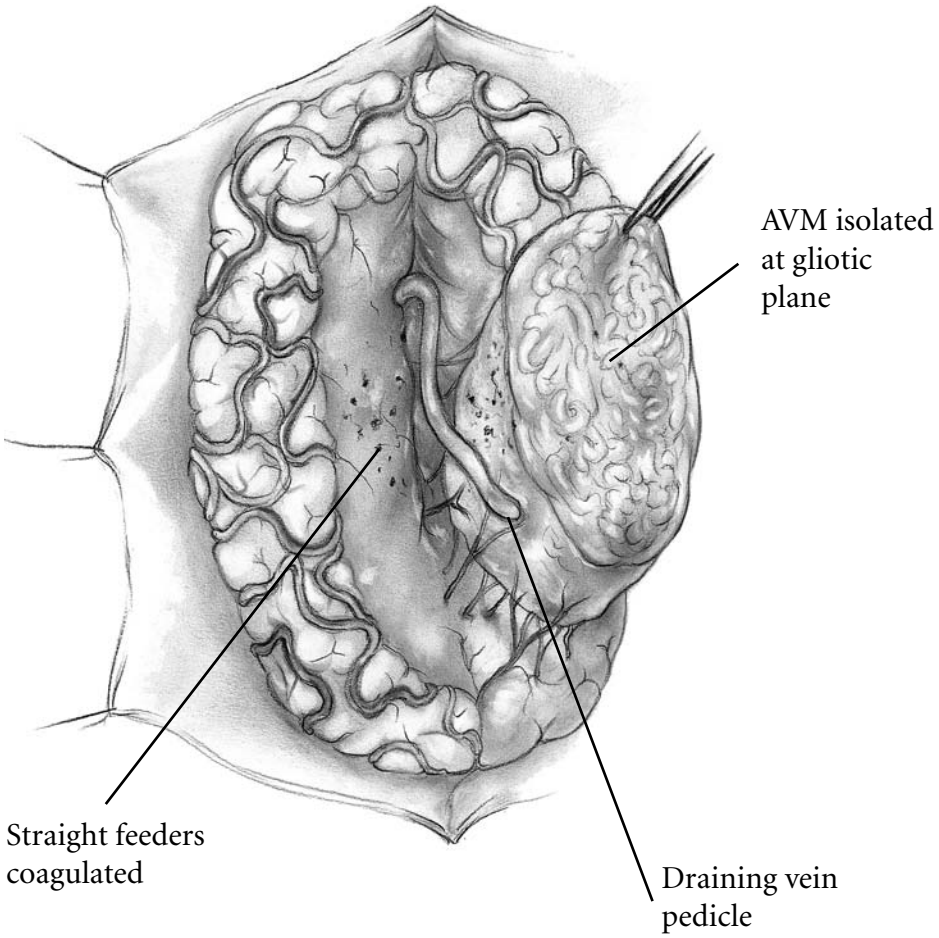


FIGURE 20–6. Circumferential dissection proceeds through the gliotic plane allowing the surgeon to look for the "straights and avoid the coils." The major draining vein is protected as a pedicle until the end.

components of the malformation. Indeed, finding the malformation hanging from either the roof or the wall of the clot cavity sometimes rewards the surgeon. Almost always, the blood clot remote from the malformation can be easily removed. If the surgeon encounters adherent clot that is resistant to suctioning, it should be left alone because it represents hematoma entwined in the malformation.

The surgeon should not be discouraged if, during the course of clot evacuation, the adjacent ventricle is entered. This is useful happenstance because the CSF not only serves to wash the bed, but it also confirms the location of the ventricle and the fact that the malformation undoubtedly has some intraventricular connections.

All too frequently children's malformations have only one major draining vein. The surgeon does not have the "luxury" of sacrificing this vein early in the procedure and using it as a "handle" to manipulate the lesion. Hence, the identified draining vein should be protected until the end of the operation. All dissection should proceed remote from it. The vein usually has excellent integrity so that, as the dissection proceeds circumferentially, the brain incision can approach within a few millimeters of its walls.

"Look for the straights and avoid the coils" should be the maxim for dissecting around the malformation. There may be a fairly generous glial plane about the malformation that makes recognition of the borders of the lesion clear. Anytime a red coil or knuckle of a vessel appears, the surgeon should back away because he or she is entering the lesion itself. Instead, the surgeon should locate the straightened arteries and veins serving the malformation, dissect these, and divide them after coagulation. This is the site of the appropriate margin of the lesion.

Once the incision has been taken through the cortex, the white matter dissection can be achieved easily with a gentle blunt dissector and a microscopic suction device. Two independent suction systems and a coagulating forceps must be available. There is debate about the size of vessels that can be reliably coagulated and those that require clip obliteration. A selection of small homeostatic clips always should be available to the surgeon. In using them, however, the surgeon should be aware that even

the smallest clips have a propensity to get in the way of further dissection, to be aspirated up the suction device, or to become snarled in the cotton packs.

Managing the AVM Nidus

Because the classic AVM has a defined wedge shape, there is a tendency early in the dissection to perform tissue dissection in an "ice cream cone" fashion, which often leads the surgeon directly into the deep portion of the malformation. The surgeon must be disciplined to maintain fairly vertical walls around the malformation until almost at the ependymal surface of the ventricle. At that stage, the deep dissection can become a little more "pointed."

The blunt dissector used to enter through the pial surface tells the surgeon, especially within the first centimeter of dissection from the surface, whether there are obstructions to its normal sweeping motion as it passes through the cortical gray and subcortical white matter (Fig. 20–6). Such obstructions usually represent traversing blood vessels. There are always more vessels, usually quite small ones, entering and leaving the malformation than the arteriogram would suggest. Moreover, they do so in an irregular fashion so that there may be several such channels clustered in one area, whereas tissue slightly more remote is free of vasculature.

It serves no purpose to place coagulating forceps blindly on the topside of a vessel with the intent of coagulating and cutting it. There may be another vessel directly beneath it, especially with veins, which are distended and somewhat floppy and frequently have a tiny artery on their deep surface. Hence, the surgeon runs the blunt dissector along the shaft of the straightened vascular structure, eventually frees it with a blunt hook beneath it, and then applies coagulation and division. The blunt hook determines whether there is another vascular channel running parallel to the observed one or whether the vessel about to be taken has a right-angled branch on its deep side.

Even in expert hands, the circumferential dissection around an AVM can be slow and tedious. If a surgeon recognizes that no progress is being made in one zone, then he or she should move 180 degrees away and start at a remote site to complete dissection there. The strategy can also be used if relatively minor bleeding is encountered from the lesion. It can be packed off, and the dissection can proceed elsewhere. Such a strategy also continues to reinforce to the surgeon the magnitude of the lesion and to provide different perspectives on the lesion's inward passage. At all times, the surgeon should be reassured that if the dissection stays within the glial plane about the lesion, minimal, if any, additional harm will be created for the patient.

For retracting the malformation, cotton strips should be laid on the malformation as it is dissected free. The surgeon should avoid using too many such strips because they not only add to the overall bulk of the exposure and tissue turgor, but they also may become coated with blood, curl, and dry up or misinterpreted for the malformation at a later stage in the dissection.

Like a gentle column of smoke coming off the top of a mountain peak, the apex of the malformation has wisps of vessels running to the ventricular wall and usually engaging the choroid plexus. These represent the choroidal supply to the deep component of the lesion. Hence the interior of the relevant ventricle must be visualized, the lush and engorged choroid plexus isolated, and eventually the malformation disengaged from it. These steps are most easily achieved by dissecting back along the body of the choroid plexus until it adopts a normal character. It can be freed from the ventricular wall, the vessels within it easily coagulated and divided, and then the choroid plexus and apex of the malformation rolled out of the ventricle.

No attempt should be made to isolate and sacrifice the draining vein until the entire malformation has been mobilized from its deep connections and the lesion rolled out of its bed (Fig. 20–7). If there is any suggestion of deep connections that persist or tissue dissection that has not been completed, this must be accomplished before the vein is sacrificed. When the malformation has been disengaged entirely from its parenchymal connections, the bed in which it lies has no oozing vessels, and the irrigating fluid returns almost colorless. If the major draining vein is still arterialized, it is because of the presence of a small artery that runs on its deep surface. Hence, with the lesion entirely mobilized, the dissection proceeds from one side to the other beneath the vein until the "culprit" artery is found. With this done, the last vascular leash is coagulated and divided, and the specimen is removed.

As the dissection proceeds through the gliotic tissue planes, the bed that remains after the malformation has been removed should be yellow–white and avascular. Sometimes a small component of the nidus may have broken free, may not ooze, and may be stuck within one of the walls. Such a component can be sought with blunt dissection. This component can be traced backward until its straight, feeding vessels are encountered, coagulated, and divided. It is not usually necessary to leave any hemostatic agents within the bed. While irrigating the wound, the surgeon should have one last look at the inte-

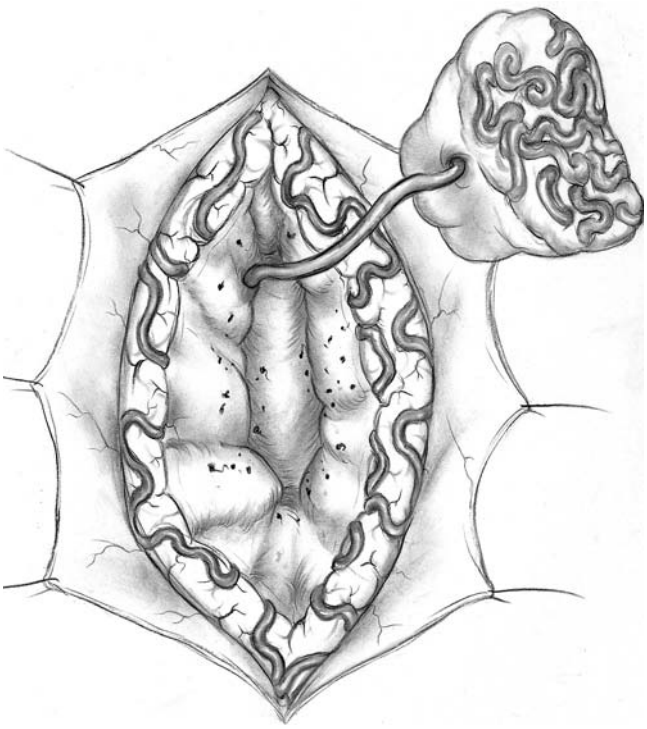


FIGURE 20–7. The AVM is rolled out of its bed and remains connected only by the draining vein.

rior of the ventricle to ensure that neither bleeding nor a clot remain behind.

Technique for the Cavernous Angioma

Most of the principles that have been elaborated for excision of the cerebral AVM can be relied on to assist the surgeon with removal of a cavernous malformation. There is not usually the level of anxiety associated with a "cavernoma" because of high-flow fistulae and a number of knurled, torturous vessels. Frank arterial bleeding resulting from rupture of a nidus is not the usual experience with an angioma. Moreover, the hematoma or softened, icteric brain usually serves as a good entree to the cavernous lesion. In ideal circumstances, the malformation retains its shape and substance, having the character of a black raspberry of whatever size. The surgeon can rely on his or her favorite microscopic dissecting tool to skirt about the perimeter of the lesion to dissect it completely free.

The trap in this particular surgery is one's inability to identify the cavernoma if it was destroyed at the time of the hemorrhage. In addition, the surgeon may be uncertain as to what margins to create if a cavernoma lies within the brainstem, spinal cord, or eloquent cortex. In most circumstances, the safety zone is reached when one encounters normal-appearing white fiber bundles at the margin of the angioma.

Finishing the Operation

Once the malformation dissection has been completed, bleeding ceases in its bed, and the brain is considerably relaxed. Given that the operation was undertaken, the surgeon should be satisfied with nothing less than total excision, even if two stages are necessary to achieve it. If a deliberate decision has been made to stage the procedure, with a second attempt planned after an interval, the surgeon should discontinue the first procedure when the operative region is neither bleeding nor swelling. The child then is allowed to recover and, if necessary, to undergo further study with repeat arteriography. One or two strategically placed metallic hemostatic clips will permit identification of the area of surgery and serve as coordinates for the further radiologic and surgical examination of any malformation that remains. Proximal vessel ligation can be dangerous and is an inadequate solution; subtotal excision is messy, and the shunt remains served by the newly recruited contributors.

The bed from which the malformation and hematoma have been evacuated will resemble those of a fresh, unperturbed exposure of white matter. There should not be any round, vascular "blisters" appearing here and there in the white substance. If these are present, residual malformation requires additional definition and removal. With the expectation that there is no further oozing within this bed, the brain should be relaxed considerably, and the dura is closed in a standard fashion. The bone flap also is replaced. It is usually satisfactory to close the scalp wound with typical substances, but if metallic staples are inserted, one must be prepared for their radioopaque interference with the interpretation of an early postoperative arteriogram.

POSTOPERATIVE MANAGEMENT

The need for a child to return to a postoperative critical care unit is dictated by that child's condition prior to the operation. At the least, the patient will be managed in such a unit for the first overnight stay and then transferred to the neurosurgical ward the following morning. If the surgeon is satisfied that a complete excision of the AVM has been achieved, then the only limitations to the child's early

mobilization from bed and initiation of a rehabilitation program will be the degree of preoperative neurologic deficit. Usually, there is no impediment to a passive physiotherapy program beginning until the child can participate actively in his or her own rehabilitation plan.

A fundamental decision that must be made during the early postoperative phase is when the postoperative arteriogram should be obtained. The purpose of this study is to determine whether the child has residual nidus or early draining veins; if none is present, the patient is regarded as cured with the risk of hemorrhage eliminated. Some arguments favor completing this study during the patient's first hospital admission so that if residual malformation is defined, it can be removed at a second procedure at this time. The contrary argument is that early arteriography, by virtue of residual tissue swelling or brain manipulation, may be blinded to a small vascular remnant, especially if the lesion is small.

EDITOR'S COMMENTARY

Children with AVMs are cared for ideally in centers that treat several AVMs per year and that have the capability to do all of the procedures that may be needed in caring for children with these lesions—including angiography, embolizations, operations and radiosurgery. The morbidity and mortality of AVM surgery varies according to the AVM (Spetzler) grade, although outcomes of AVM resections in pediatric and adult patients with similar grade AVMs have not been compared in large series. The surgical principles described herein have been developed from years of experience in treating AVMs and will minimize the morbidity of excising these lesions. We concur with obtaining angiograms soon after a child's admission; a few AVMs do rehemorrhage within 48 hours after the first bleed and if an operation is needed acutely, knowledge of the vascular anatomy is reassuring. There seem to be fewer indications for preoperative embolization of AVMs in children than in adults. At operation, excision of the AVM nidus is akin to resection of a particularly vascular neoplasm; one works entirely at its periphery until the lesion is completely free of vascular and glial attachments. If postoperative angiography at six months demonstrates a small residual nidus, we usually treat it with stereotactic radiosurgery, which has excellent rates of obliteration of minimal residual lesions.

PEARLS

In this author's experience:

- Spontaneous intracranial hemorrhage is the means by which 80% of children will declare their malformation. For those children presenting with hemorrhage or epilepsy, 80% will require an operation.
- Whenever possible the surgery for a child's AVM should be planned as an elective procedure.
- The tissue dissection around the AVM takes advantage of the malformation's gliotic plane. When working through that tissue, the surgeon, with respect to the vasculature, should "look for the straights and avoid the coils."
- The use of adjunct endovascular embolization in a child's AVM should be customized case by case. In a few patients with traditional brain AVMs, endovascular techniques may serve as a prelude to excision of the malformation.
- The venous angioma often lies beside a cavernous angioma. Regardless, it must be respected as part of the normal cerebral venous circulation and thus left unperturbed.
- A postoperative survelliance angiogram is mandatory. To avoid a false negative result, it should be obtained after an interval when tissue swelling and operative vascular distortions have resolved.

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21

MOYAMOYA DISEASE

Yoshiharu Matsushima

In 1979, we developed an operation called encephalo-duro-arterio-synangiosis (EDAS) for the treatment of moyamoya disease in childhood. In moyamoya disease, intracranial and extracranial arteries easily form anastomoses spontaneously, and this tendency is increased by adding surgical interventions. EDAS is an operation that uses and favors these characteristics of the disease. By performing this operation, we could eliminate all the disadvantageous factors associated with surgical methods introduced before EDAS. The following are the characteristics of EDAS:

1. Operative procedure is simple, and no special technique is required. Because the operation time is short, the requisite duration of anesthesia is short and carries a lower risk of accidents.
2. There is almost no possibility that preoperatively existing extracranial-to-intracranial vascular anastomoses are injured. Because the periphery of the donor scalp artery is not discontinued, and therefore blood flow is not stopped either during or after operation, extracranial-to-intracranial collaterals already formed at the periphery of the donor scalp artery are left intact.
3. Because EDAS does not require procedures like compression to the brain or transient blockade of the bloodstream, which are unfavorable for the brain with reduced blood flow, no neurologic deterioration can occur postoperatively.
4. As shown in Figure 21–1, arteries that can be used as donor arteries [anterior branch (aSTA) and posterior branch (pSTA) of the superficial temporal artery (STA) and the occipital artery (OA)] are distributed over the entire cranial area. Therefore, the operation site can be selected with no limitation according to the findings of cerebral angiography, neurologic symptoms, computed tomography (CT) findings, electroencephalographic (EEG) recordings, and regional cerebral blood flow; the operation can be done even in sites where muscle is absent.
5. There is no possibility of forcing blood flow into the brain unnecessarily, especially in conditions in which acute elevation of blood pressure is undesirable, because the brain forms spontaneous anastomoses according to its demands.

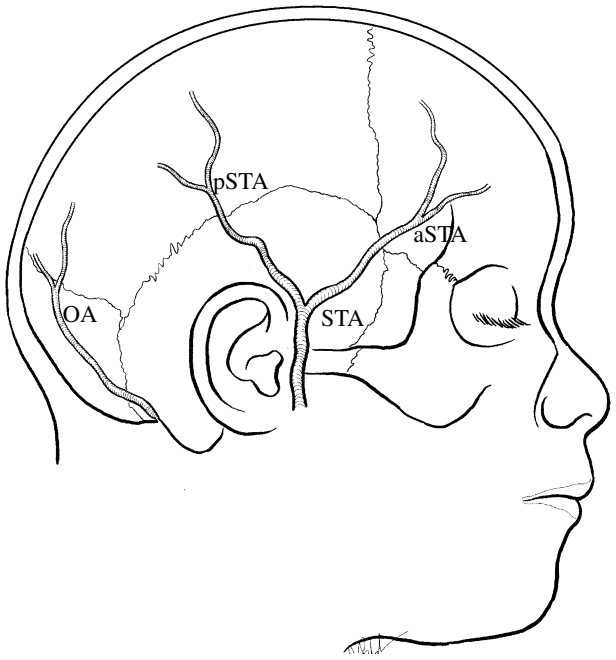

FIGURE 21–1. Arteries available as donor arteries. STA, superficial temporal artery; aSTA, anterior (frontal) branch of the STA; pSTA, posterior (parietal) branch of the STA; OA, occipital artery.

6. Blood flow of transdural anastomosis that existed before the operation increases as a result of a remarkable postoperative increase in blood flow of the dura mater.

INDICATIONS AND TIMING OF OPERATION

The goal of the treatment of moyamoya disease is to reach "a condition under which sufficient blood is supplied to the brain through the external carotid arterial system" before an oligemic state—unfavorable to the developing brain and manifested by repeated ischemic attacks—causes irreversible damage to the young brain. We believe this operation can be performed at any stage of the disease in patients who have had repeated ischemic attacks, except for those in which the brain has not yet stabilized after recent occurrence of transient ischemic attack (TIA), reversible ischemic neurologic deficit (RIND), or infarction.

We recommend using the anterior and the posterior branch of the STA as the donor arteries on the dominant hemisphere in patients who have onset of the disease who are younger than 5 years of age or whose cerebral function deteriorates rapidly. Otherwise, what we consider to be the standard operation, using the bilateral posterior branches of the STA as donors, should be sufficient.

INTRAOPERATIVE TECHNIQUES

Skin Incision

The course of the donor artery is determined through palpation or with the help of a Doppler device (making reference to the course of the donor scalp artery as shown by the cerebral angiograms) and is marked on the scalp with marking ink. A shallow skin incision about 1 cm long is placed at the site immediately above and at the proximal side of the scalp artery, where the pulse can be palpated most strongly (Fig. 21–2). It is advisable to use a Feather no. 15 disposable operating knife here.

Then the assistant inserts a curved hemostat into the layer between the skin and the artery (between the adipose tissue and the aponeurosis) and spreads this layer over the artery, taking care not to injure the artery, and elevates the skin upward. The operator then incises the elevated skin between the blades of the hemostat. The edges of the incised skin are separated by placing hooks applied with tension by using rubber bands. We use an EDAS frame (see Fig. 21–5.) Hemostasis for dermal bleeding can be obtained by only the tension of these hooks.

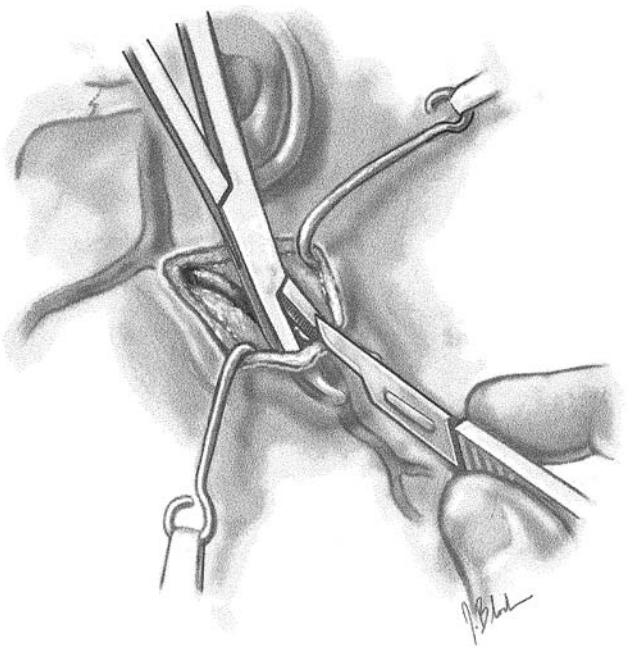

FIGURE 21–2. Skin incision.

Dissection of the Galea and Preparation of the Pedicle with the Donor Artery

The donor artery is exposed together with its attached galea for as far as possible (9 cm on average in our experience) within the range for which it can be traced safely. Two parallel incisions are placed in the galea 5 to 7 mm

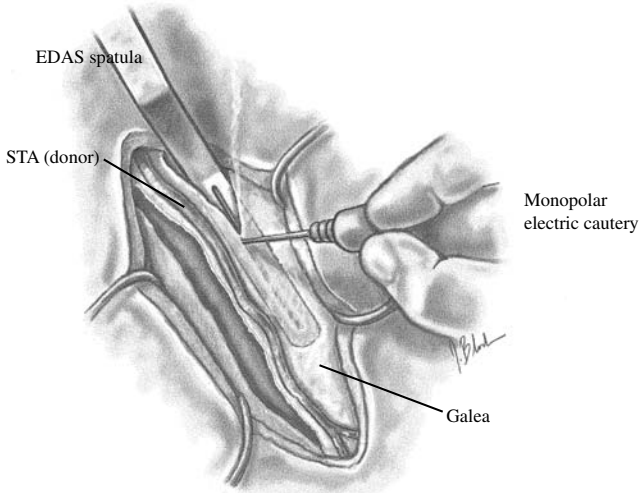

FIGURE 21–3. Encephalo-duro-arterio-synangiosis (EDAS) spatula and galeal incision.

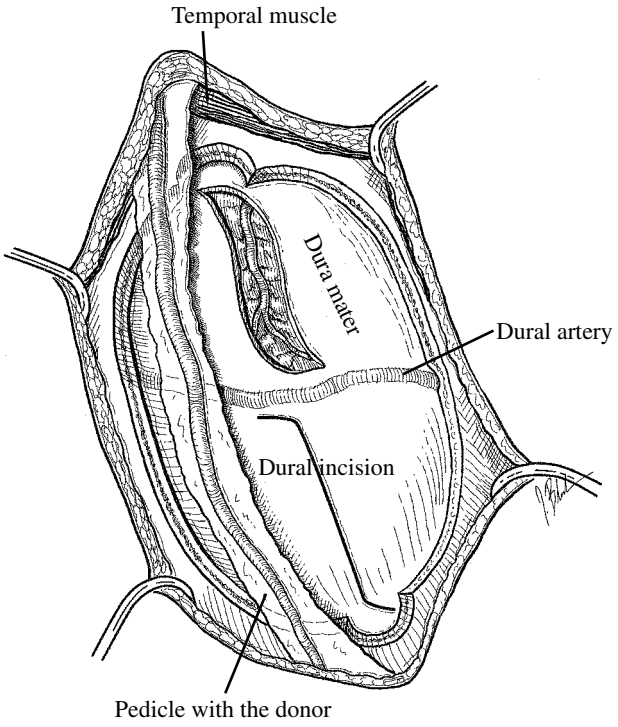

FIGURE 21–4. Opening of the bone window and dural incisions.

apart from and parallel to the scalp artery (Fig. 21–3) using a needle-shaped electric cautery. For these incisions we use a hard, nonconductive spatula with grooves, as shown in the figure. This spatula is inserted beneath the galea (above the fascia or periosteum), parallel to the donor artery, and incisions are made using a needle-tip electric cautery along the grooves of the spatula. With this method, the incision can be made quickly, producing no injuries to the adjoining tissue.

It is best to use an electric cautery in making these incisions. The edges become solid and thin-black-lined, which make subsequent procedures easier. Thus, a partially free bridgelike pedicle that possesses the donor artery on the band of the galea is prepared. In the preparation of the partially free pedicle, if the donor scalp artery bifurcates into thick branches at an acute Y-shaped angle, such branches are also placed on the galeal strip, together with the main donor scalp artery. Thus, the prepared partially free pedicle is separated from the periosteum or temporal fascia in the form of a bridge.

Opening of the Bone Window

Burr holes are made below the upper and the lower ends of the skin incision, namely immediately beneath the most proximal side and the most distal side of the partially free pedicle. After the two burr holes are placed, oval-shaped osteotomy, 7 to 8 cm long and 3 to 4 cm wide, is performed with these two burr holes used as both ends (Fig. 21–4).

Incision of the Dura Mater

The dura mater is examined closely, and a fine linear incision is made in the dura mater, taking care not to injure large dural arteries and transdural anastomoses. It is rec-

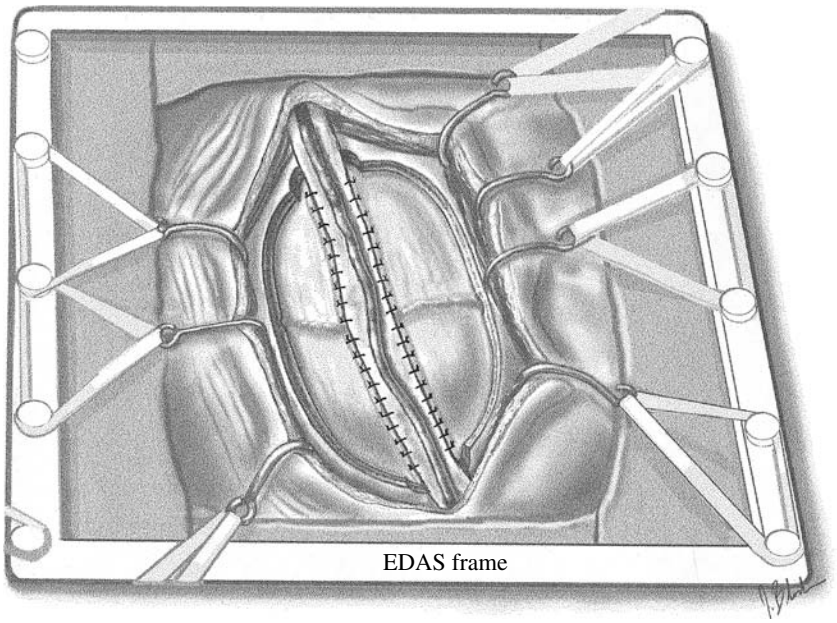

FIGURE 21–5. Closure of the dural opening by suturing the pedicle to the dural edges. EDAS frame in use.

ommended that large dural arteries be left in a bridge form (Fig. 21–4). It is convenient for suture of the partially free pedicle to angle both ends of the linear incision of the dura mater by 5 mm in length, as shown. Too meticulous hemostatic coagulation of the dural edge is not recommended here. In our experience, incision of the arachnoid here has proved unnecessary.

Suturing of the Pedicle to the Dural Opening

Here the partially free pedicle (the donor artery with the wing-shaped galea) is placed on the incised dura. Then the temporal muscle below the proximal pedicle is cut, and a rongeur is used on the cranium near the burr hole as necessary so as not to interfere with the blood flow of the donor artery. Next the dual wound is closed watertight by suturing the galeal edges of the pedicle to the margins of the opening of the dura mater (Fig. 21–5).

Closing the Wound

Tenting sutures are placed at several sites. When putting back and fixing the bone flap, it is required to prepare the bone flap so that blood flow of the donor artery is not hindered (Fig. 21–6). Finally, the wound is closed. In many cases, it is difficult to approximate the margins of the galeal wound; however, it is necessary to suture the galea by approximating as much as possible to minimize stretching of the skin wound later.

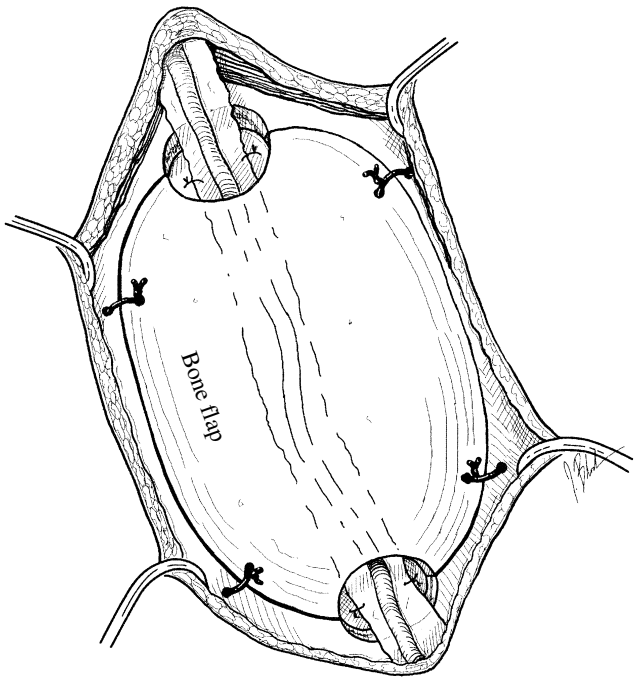

FIGURE 21–6. EDAS just before the wound closure.

POSTOPERATIVE MANAGEMENT

Recent clinical articles described the perioperative risk factors of postoperative ischemic complications. Sakamoto et al (1997) reported that patients who had omental transplantation and who had experienced more than one to four attacks of TIA per month preoperatively had more postoperative complications than patients who had direct anastomosis and only one attack in several months. Sato et al (1997) reported that three factors—the presence of low-density areas preoperatively, intraoperative urinary output, and postoperative reduction in hematocrit—were significantly correlated to the postoperative ischemic complications. On the basis of these findings, they concluded that it is necessary to infuse an adequate amount of fluid, maintain normocapnic and normothermic states, actively correct anemia, and give particular attention to perioperative care in patients whose preoperative CT shows low-density areas.

Administration of O_2 has been effective for TIA attacks; oxygen administration soon after hyperventilation was effective in eliminating the re-buildup phenomenon and hence in abolishing its symptoms. Concerning long-term prognosis, we concluded that the EDAS was significantly effective in decreasing the long-term deterioration in intelligence observed in the natural course of pediatric moyamoya patients.

MODIFICATIONS

In some cases, the donor scalp artery becomes tapered and so unpalpable while it is traced. In such cases, it is recommended that a partially free pedicle associated with relatively large galeal flap be prepared as shown in Figure 21–7b.

Professor Choi, in Korea, modified the EDAS as shown in Figure 21–7c. This modification should be recommended in cases in which the dural incision for EDAS reveals a thick vein or large sulcus below. Kashiwagi et al (1996) recently reported split-duro-encephalo-synangiosis (Fig. 21–7d). This procedure should be recommended when the frontal branch of the STA is small, difficult to follow, or becomes undetectable during the procedure. This procedure is useful also in reactivating an old and poorly revascularized EDAS. This procedure, combined

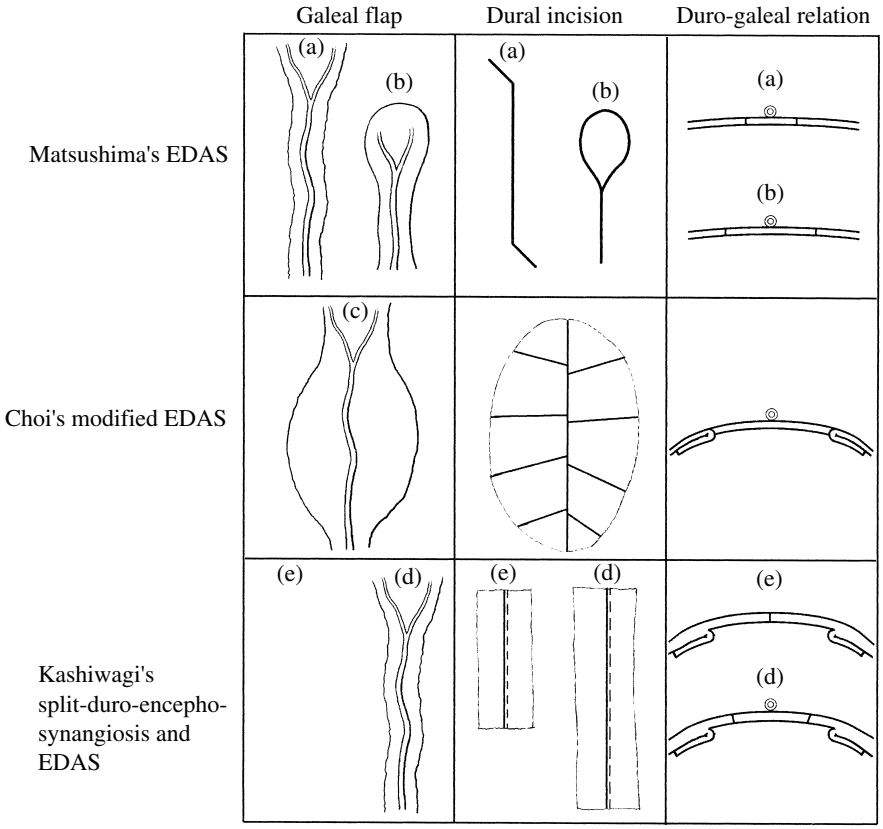

FIGURE 21–7. Modifications and others. **a, b:** Standard Matsushima's EDAS; EDAS often is used when the donor artery is the occipital artery or the anterior (frontal) branch of the STA. **c:** Professor Choi (Korea's) modified EDAS: In this procedure, a cut dural flap is turned inward beneath the dura mater. **e, d:** Kashiwagi's procedure is used to reinforce the EDAS. Kashiwagi's original split-duro-encephalo-synangiosis: Dural incision is made only to the outer layer of the dura mater first and then to the inner layer, which is turned inward beneath the dura mater.

with the author's EDAS (Fig. 21–7e) also would be useful in cases in which revascularization to a wide cerebral area is necessary.

EDITOR'S COMMENTARY

The encephalo-duro-arterio-synangiosis (EDAS) in the treatment of moyamoya disease has been eloquently described by the author. Because the indirect revascularization approaches described have at least comparable results to the direct anastomoses, in this editor's experience (PDA), the EDAS is quite preferable, particularly in young children. The use of a long arterial strip remains important to maximize the exposure of the STA to the surface of the ischemic brain. Because of the facility of the procedure, bilateral revascularization under the same anesthesia is often recommended as appropriate. We have preferred to place these children in 3-point fixation being able to turn the head 180 degrees within the 3-point fixator at the completion of the initial side in order to treat the often bilateral problem with the same anesthetic. In these instances, the more ischemic side is treated first. While the author again highlights that it is unnecessary to open the arachnoid for revascularization, it has been this editor's experience that direct fixation of the vessel to the pial surface after arachnoid opening improves the percentage and extent of revascularization. The editor would also reiterate the usefulness of supplementing the EDAS with burr holes in the frontal and parieto-occipital areas in order to further enhance dural arterial anastomoses to the ischemic tissue. Lastly, optimization of the perioperative environment is essential in order to avoid intraoperative ischemic events. Central access with adequate hydration and maintaining normocapnea cannot be stressed enough in order to avoid intraoperative ischemic complications.

PEARLS

In this author's experience:

- Establishment of a good relationship with the patient and the family is very important. The family should be educated that ischemic attacks may occur at any time in this disease.
- When performing EDAS, it is necessary to make a long donor arterial strip (approximately 9 cm) and a big bone flap (3 to 4 $\times$ 7 to 8 cm). The arachnoid should not be opened. It is better to perform this procedure without a microscope.
- The greatest postoperative problem are ischemic attacks, usually induced by crying in response to uneasiness and dermal pain during postoperative management or examination.

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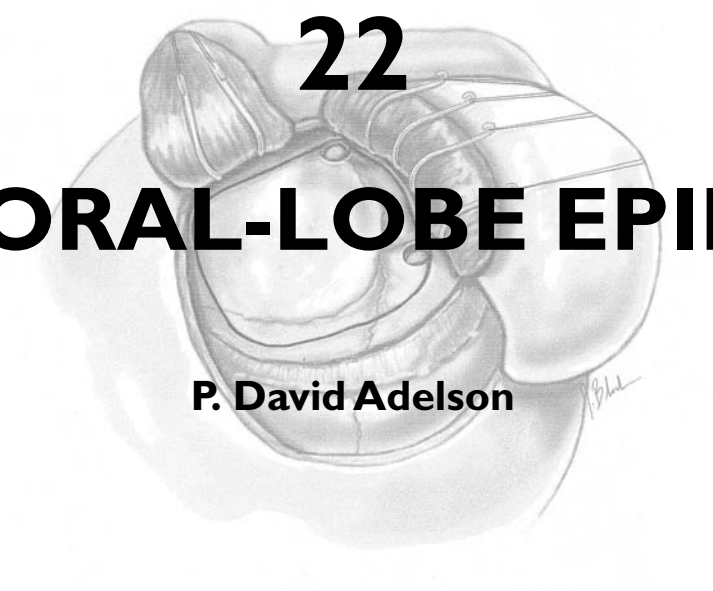
Functional Disorders

22

TEMPORAL-LOBE EPILEPSY



P. David Adelson

Epilepsy affects approximately 0.5 to 1.0% of the population, and its onset most frequently occurs during childhood. In young children, uncontrolled seizures adversely affect intellectual maturation and the development of normal social behaviors, which may be a result of prolonged and often toxic levels of anticonvulsant medication or the constant abnormal electric activity during development. Because intractable seizures in children have a "malignant" natural history with eventual declines in both intellectual and behavioral functions, surgery has become an acceptable early treatment modality for these patients.

The temporal lobe and its medial structures, the amygdalohippocampal complex, are the most common source of medically intractable complex partial seizures in adults and are the origin for approximately 30% of such seizure disorders in children. Historically, temporal-lobe epilepsy has been the most common surgically treated form of epilepsy because its distinct pathology and accessible location, low morbidity of surgery, and excellent outcomes. The percentage of temporal-lobe resections in most pediatric series exceeds 56%. The determination of intractability and the evaluation of surgical candidacy in children can be done in a systematic and effective way to identify which patients would benefit from surgical intervention when they have failed medical management.

Several techniques have been used for temporal lobectomy for epilepsy. The original description by Penfield, later modified by Falconer, consisted of an en block resection of the temporal lobe and the medial structures. Falconer initially described the successful use of his technique for temporal lobectomy, specifically for the treatment of epilepsy in children, almost 30 years ago. This author prefers a two-stage resection similar to that described by Spencer, which involves an anterolateral cortical resection followed by a microscopic removal of the medial structures. This method allows limited resection of the anterior temporal lobe and yet provides adequate access to the medial basal structures. Often the best method is that which the surgeon is comfortable with and performs most commonly. This description is meant as a guide to the anatomy and one approach that provides a low incidence of morbidity with excellent outcomes.

SURGICAL INDICATIONS AND PREOPERATIVE EVALUATION

The goal of the surgical evaluation is to characterize the seizures, define the epileptogenic zone by lateralizing and localizing them to a particular region, and, finally, determine the relationship of the epileptic focus to the eloquent cortex. This is achieved through the formalized evaluation process, which attempts to obtain concordant data as to the origin of the abnormal region. The initial phase is noninvasive and includes a battery of tests to define the seizures, their semiology, and their electrophysiologic dysfunction. Besides electroencephalography (EEG), a neuropsychologic battery, imaging with magnetic resonance (MR), and some type of functional imaging are used to obtain a better understanding of the preoperative anatomical and functional anomalies. These imaging studies, in conjunction with the EEG, often are able to define the seizure syndrome and its etiology. If necessary, further evaluation may include EEG and videotelemetry to characterize the seizure semiology or amobarbital (Wada) testing, where needed, for language and memory localization. Concordance between the EEG and imaging as indispensible for defining the epileptogenic zone and for making the decision to proceed with a surgical intervention.

Imaging

With the further refinement and improvement in neuroimaging capability and resolution, particularly with the development of new sequencing modalities (e.g., fluid attenuated inversion recovery (FLAIR) for MR, lesions and subtle structural differences between normal and abnormal cortical areas have been increasingly defined that have been correlative with the epileptogenic zone with increased sensitivity (Fig. 22–1). Because epilepsy is a *functional* disorder of the brain, defining the anatomic abnormalities may not be sufficient to define the epileptic focus fully. In these instances, functional neuroimaging may detect the generalized and focal functional abnormalities and their relationship to the EEG and anatomic imaging. These imaging modalities have been useful in temporal-lobe evaluation and are believed to have a significant role in the evaluation of patients with extratemporal epilepsy as well. The MR can be used for MR spectroscopy (MRS) and functional MR (fMR); the efficacy of positron emission tomography (PET) and single-photon emission computerized tomography (SPECT) also have had a major impact on the evaluation of patients with temporal lobe epilepsy.

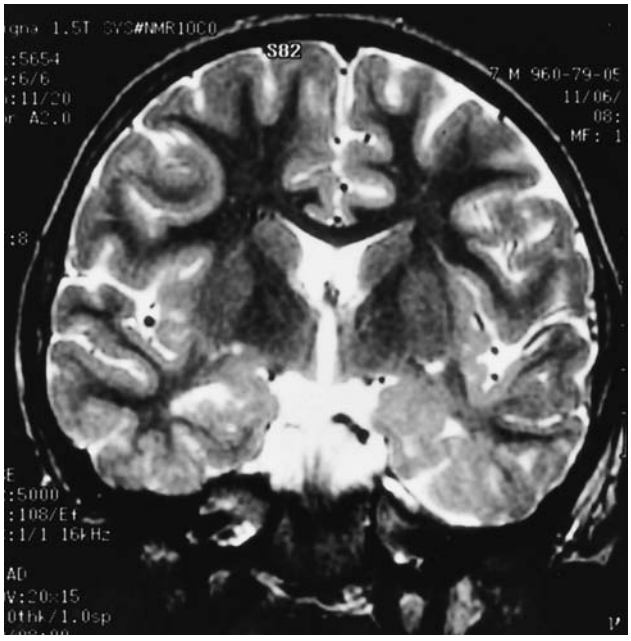


FIGURE 22–1. Coronal MR (T_2-weighted) scan of a 3-year-old boy with a 2-year history of complex partial seizures intractable to medication. Previous scans were reportedly normal. A cortical signal abnormality is noted in the left inferior temporal, parahippocampal gyri and the medial structures.

With the improved evaluation techniques now available, many pathologic and physiologic changes that were previously undetectable can be identified, which can further support the decision for surgical intervention when medical therapy has failed. Many young children would have previously had a prolonged observation period prior to surgical resection with little medical efficacy. With modern neuroimaging techniques, a child with a clear cytoarchitectural, neoplastic, or metabolic abnormality that correlates with the seizure syndrome can be identified and a focused surgical resection performed without unnecessary prolongation. These new technologies also have decreased the need for invasive monitoring with intracranial electrodes in some adult series by 50% and in children in up to 90% by identifying those patients who would have had a previously undefined epileptogenic zone, with the necessary concordant data defining their epileptic focus before implantation. These patients were able to benefit directly from a focal surgical resection without a second operation.

SURGICAL ANATOMY OF THE TEMPORAL LOBE

There are two sources for a temporal-lobe seizure focus: the medial basal portion and the lateral neocortex. The medial basal portion includes the fusiform and the parahippocampal gyri as well as the amygdalohippocampal complex. Anteromedially, the parahippocampal gyrus curves in front of the midbrain to form the uncus; immediately posterolateral to the uncus is the entorhinal cortex. Medial to these structures lies the hippocampal formation. When approaching from lateral to medial along the parahippocampal gyrus, the subiculum forms arising from the parahippocampal gyrus. Directly medial to the subiculum are the fields of the cornu ammonis (CA1–CA4) and the dentate gyrus. In medial temporal sclerosis, the greatest damage occurs in Sommer's sector (CA1 and prosubiculum) and the end-folium (CA4 and hilus). The other areas, such as granule cell layer are CA2, also are severely damaged. The only region not as badly affected is the subiculum, although there is some damage.

Inside the temporal horn of the lateral ventricle, the body of the hippocampus is covered by the alveus, which posteriorly, in the tail of the hippocampus, forms into the fimbria of the fornix. Within the ventricle and superomedial are the choroidal fissure and the choroid plexus, which lie in the roof of the temporal horn and have a vascular supply from the anterior choroidal artery. The lateral cortex includes the superior and medial gyri and the anterior part of the inferior temporal gyrus. Any of these

ls ____
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cortical regions also may support an epileptic focus either due to a lesion or architectural abnormality but may also include eloquent or functionally important tissue, especially in the dominant hemisphere.

The vascular supply to the temporal lobe consists of multiple perfusing vessels. There is a lateral and superolateral supply from the middle cerebral artery as it arises from the sylvian fissure. The inferior surface of the temporal lobe is supplied by the posterior cerebral artery with the P2 and P3 segments traversing the posterior portion of the ambient cistern to reach the inferior part of the temporal lobe. The P2 segment also supplies part of the hippocampus and parahippocampal gyrus via the small Ammon's horn arteries medially. The anterior choroidal artery also contributes to the medial vascular supply as it extends posteriorly from the internal carotid artery through the choroid plexus within the temporal horn and supplies part of the cascade of microvasculature to the hippocampus and parahippocampal gyrus (Fig. 22–2).

Cranial nerves III and IV extend from the brainstem within the ambient cistern anteriorly along the edge of the tentorium. Pupillary dilatation is a harbinger of uncal herniation because of third-nerve compression by the uncus anterior to the hippocampus. Injury to these nerves during a temporal lobectomy can be minimized by maintaining the integrity of the pia–arachnoid border of the superior compartment.

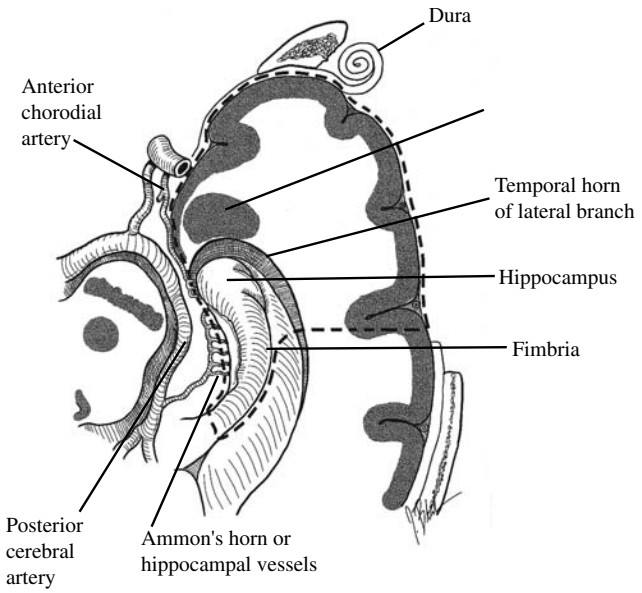


FIGURE 22–2. The P-2 segment of the posterior cerebral artery provides part of the arterial supply to the hippocampus via the small Ammon's horn arteries medially. The anterior choroidal artery also contributes to the medial vascular supply as it extends posteriorly to the choroid plexus within the temporal horn. The planned resection is mapped in the dashed lines.

INTRAOPERATIVE TECHNIQUE

Anesthetic Techniques and Preoperative Medications

The anesthetic issues for patients undergoing a temporal lobectomy depend on whether intraoperative electrophysiologic studies will be performed. In general, the anesthetic technique includes variable combinations of inhalation agents, narcotics, and paralytics. After induction, anesthesia often is maintained using nitrous oxide along with a continuous fentanyl infusion. There is usually no contraindication to paralysis for the standard procedure, and vecuronium is usually the drug of choice. Other inhalation agents, such as isoflurane, may be used at minimal dosages if necessary but may impact on the intraoperative EEG. Benzodiazepines and barbiturates also are usually avoided because of their long-acting effect on the EEG. "Awake" craniotomies are rarely done in children except in the older, mature child. In these instances, a combination of local injections of a long-acting anesthetic, such as marcaine hydrochloride, and a general, short-acting amnestic medication, such as propofol, is continued throughout the initial intervention until the eloquent area is exposed. Once the patient's cooperation is needed, the infusion can be discontinued and the intraoperative functional testing can proceed. Overall, we prefer to map these functional areas extraoperatively with grid and strip electrodes in a planned two-stage procedure with the presumed craniotomy adequately placed to expose these areas. Mapping of the epileptogenic focus then can be performed and cortical stimulation algorithms used to define the areas of eloquent cortex prior to resection in a more relaxed setting.

Routine monitoring is used in all patients unless special circumstances or concerns exist (e.g., excessive blood loss from an arteriovenous malformation). The routine for the surgical preparation includes adequate venous access for medications and fluids; an arterial line for constant blood pressure monitoring; a urinary drainage catheter; and, in cases in which blood loss may be significant, a central venous catheter. Surface EEG leads also may be placed before positioning for intraoperative monitoring. Anticonvulsant medications are continued in full dosages throughout the intraoperative and postoperative periods.

Preoperative medications are dependent on the preferences of the surgeon and operative team. Antibiotics are given "preincision" and sometimes are continued at spe-

cific intervals during the procedure. The use of vancomycin and tobramycin or cefazolin has been described and probably has the same efficacy and outcome. Infection, although the most commonly reported complication, occurs only rarely. The use of preoperative steroids is also variable and often depends on the etiology of the seizures. As in standard tumor surgery, if there is a "lesional" component to the seizures and there is associated edema or mass effect, dexamethasone is often used at a dose of 1 to 10 mg every 6 hours. Some surgeons prefer to use steroids in all cases as a prophylactic measure for the swelling associated with surgical manipulation. Alternatively, a steroid regimen could be instituted postoperatively if there were any obvious neurologic deficits. Osmotic diuresis with mannitol also has been described to facilitate surgical manipulation of the cerebral structures; however, brain relaxation often can be achieved simply by drainage of cerebrospinal fluid and the use of mild hyperventilation ($PaCO_2$ of 30–35) without creating major fluid shifts.

Positioning and Initial Exposure

Patients are placed supine on the operating table with a roll under the ipsilateral shoulder and the head turned away, exposing the operative side. The head is positioned so that it is parallel to the floor with the vertex pointed downward 15 degrees, and the patient is placed in three-point fixation (Figs. 22–3A and B). The hair and scalp are scrubbed with an antibacterial soap, and a small strip is shaved along the planned incision. In this way, the need to shave this exposed area of the head is minimized. Following skin preparation, a question mark incision is marked out; inferiorly, it extends from 1 cm below the zygoma, just anterior to the tragus of the ear, and curves around the ear to the apex of the ear, which is its posterior extent. The incision then curves superiorly and anteriorly above the superior temporal ridge, staying within the hairline, and as far anteriorly as possible to optimize the anterior exposure. Infiltration of the skin with lidocaine and epinephrine can be used to assist hemostasis.

The skin incision is carried out with a no. 15 blade and carried through the subcutaneous tissues and the galea. Skin clips then are applied for hemostasis. Alternatively, the skin can be scored to the subcutaneous tissue and further incised using a needle-tip cautery to avoid the skin clips and to cause minimal blood loss. The scalp flap then is sharply dissected through the subgaleal tissue using the no. 15 blade and reflected anteriorly using fishhooks (Fig. 22–4). The periosteum is left intact on the bone, as is the temporalis muscle and its fascia. The temporalis muscle then is incised in a T-shaped manner

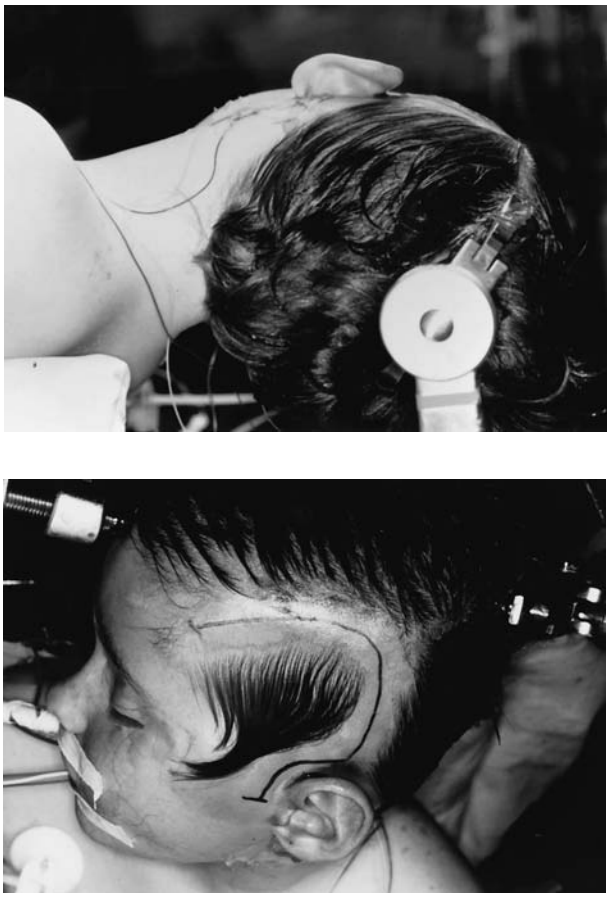


FIGURE 22–3. A: The head is placed in 3-point fixation with the vertex 15 degrees down toward the floor. **B:** The hair is minimally shaved along the proposed incision, which is marked out prior to draping.

with the vertical arm placed anteriorly so that approximately one third of the muscle is reflected anteriorly and two thirds reflected posterinferiorly using the fishhooks or suture. The superior muscle incision should leave a cuff of tissue attached to the superior temporal line for later reattachement (Fig. 22–5).

The craniotomy or bone removal does not need to extend much above the sylvain fissure but should permit generous exposure of the anterior temporal lobe. The optimal exposure facilitates visualization of the floor of the middle fossa, the sylvian fissure, and frontal operculum. Burr holes are placed in the low temporal region just superior to the zygoma and in the "keyhole" just superior to the pterion and sylvian fissure. The dura then is stripped and the craniotomy completed using a high-speed craniotome (e.g. Midas Rex). Often it is difficult to complete the circumferential craniotomy anteriorly because of the sphenoid wing; if it is scored, the bone flap can be fractured easily anteriorly to free it. For access to the low temporal

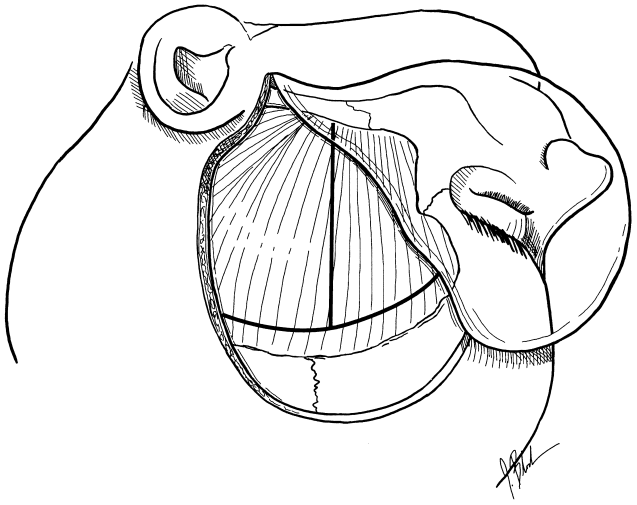


FIGURE 22–4. The scalp flap is reflected anteriorly using sharp dissection beneath the aveolar tissue and retracted using fishhooks. The temporalis muscle incision will be T-shaped, maintaining its vascular supply and a cuff of tissue superiorly along the superior temporal ridge for later reapproximation.

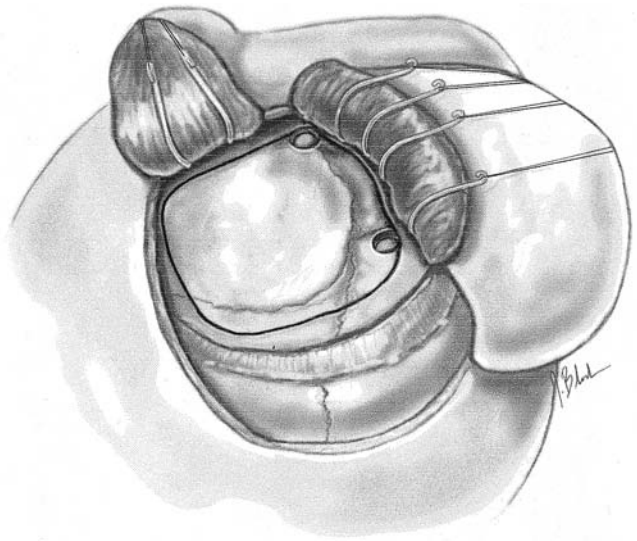


FIGURE 22–5. The muscle is incised with one third reflected anteriorly and the rest reflected inferiorly after being subperiosteally dissected. Burr holes are placed in the low temporal region just superior to the zygoma and in the "keyhole" just superior to the pterion and the craniotomy completed using a high-speed craniotome. The bone incision is brought as low and anterior as possible to minimize the amount of bone that will need to be later craniectomized. Note again that the superior arm of the temporalis incision should leave a cuff of tissue attached to the superior temporal ridge for later reattachment, which will completely cover the craniotomy defect at completion of the procedure.

region, the bone incision is brought as low and anterior as possible to minimize the amount of bone that will need to be later craniectomized (Fig. 22–5). The sphenoid wing also is craniectomized to facilitate access to the sylvian fissure following the dural opening. Bone wax is applied to the bony edges, and the middle meningeal artery often requires coagulation for hemostasis. A long, thin cottonoid can be placed anteriorly beneath the temporal muscle to diminish bleeding into the epidural space, and multiple dural stitches are placed along the periphery of the craniotomy to avoid epidural bleeding and postoperative hematoma formation.

A C-shaped dural incision, based anteriorly to maintain the middle meningeal arterial supply (Fig. 22–6), or a U-shaped inferiorly based dural incision is performed to gain exposure of the temporal lobe. The dura is rolled into a "tent roll" to keep it from drying out during the procedure, and low horizontal dural stitches help to lift and reflect the dura anteriorly for the optimal exposure of the anterior structures. The inferior flaps of dura then are tented upward using suture to gain adequate exposure of the floor of the middle fossa (Fig. 22–6). Once the dura is opened, baseline intraoperative electrocorticography (ECoG) can be obtained of the lateral neocortex and the medial structures using surface electrodes. In cases where an extraoperative grid was placed, the epileptogenic focus and areas of eloquent cortex are marked to correspond to the active surface electrode sites that either will require resection or will need to be avoided.

Anterolateral Temporal Lobe Resection

In the standard temporal lobectomy, the anterolateral temporal lobe resection is performed first; this extends 5 to 5.5 cm on the nondominant hemisphere and 4 to 4.5 cm on the dominant hemisphere at its most posterior extent as measured along the middle temporal gyrus from the temporal pole. To decrease further the incidence of neurologic (e.g., language or visual field) changes, the posterior extent can be lessened to 4 cm and 3 cm on the nondominant and dominant sides, respectively. Unless indicated otherwise by the preoperative studies (e.g., seizures arising from the superior temporal gyrus or a lesion involving the entire lateral neocortex), the resection is performed posteriorly and is limited to the lateral cortices of the middle and inferior temporal gyri and superiorly using a subpial technique just inferior to the superior temporal sulcus (Fig. 22–7). The pia first is coagulated and sharply incised, and the tissue dissections are deepened using suction aspiration and bipolar coagulation. The anterior extension will cross the superior

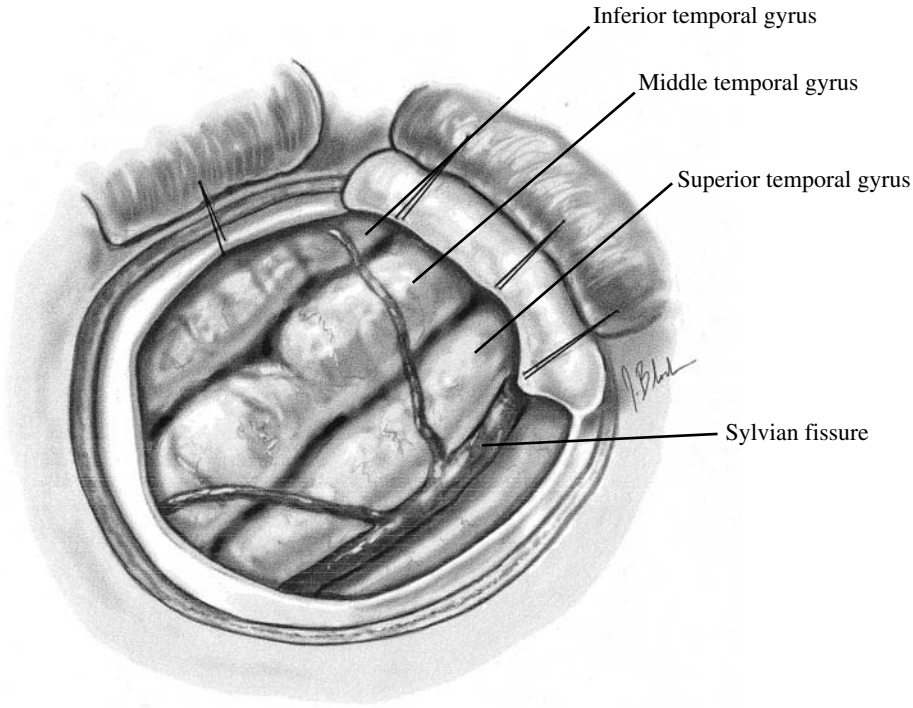


FIGURE 22–6. A C-shaped dural incision based anteriorly is performed to maintain the middle meningeal arterial supply and is rolled into a "tent roll" to keep it from drying out during the procedure. Low horizontal dural stitches anteriorly and inferiorly lift the dura for the optimal exposure of the anterior wall and floor of the middle fossa.

temporal sulcus into the superior temporal gyrus once the incision is anterior to the sphenoid wing. The resection is deepened superiorly and posteriorly until entry into the temporal horn of the lateral ventricle, which can be found through the white matter at the posterosuperior angle of the resection. The temporal horn of the ventricle then is protected using a cottonoid pledget to block access of blood into the ventricular system. Adequate inferior and anterior dissection is achieved when the floor and anterior well of the middle fossa are well visualized. The anterolateral resection of the temporal lobe then is completed by traversing the white matter by using a suc-

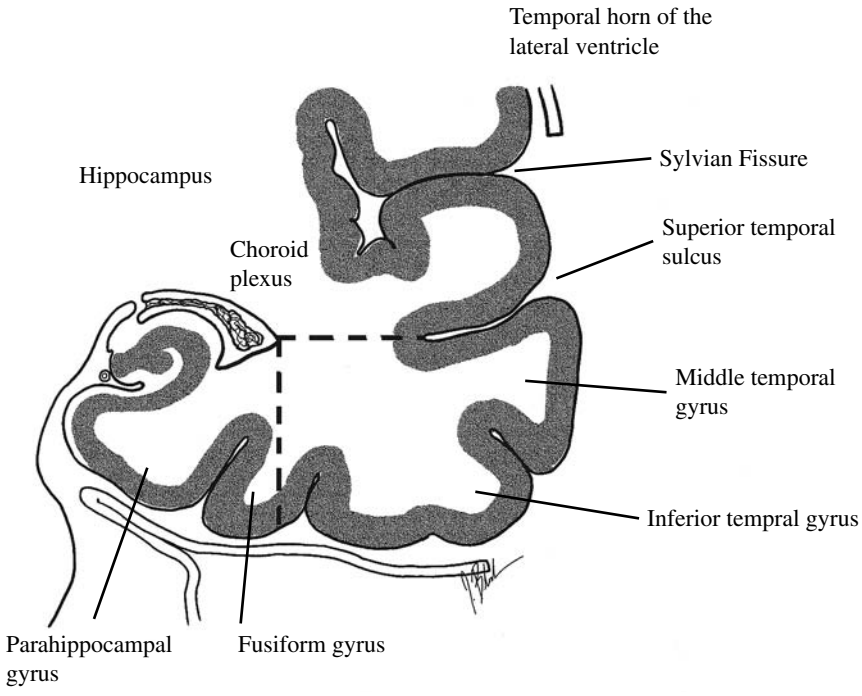


FIGURE 22–7. The resection is performed subpially along the superior temporal sulcus through the middle temporal gyrus. The tissue dissections are deepened superiorly and posteriorly until entry into the temporal horn of the lateral ventricle. The anterolateral resection of the temporal lobe is completed by traversing the white matter using the ultrasonic aspirator and subpially connecting the anterior and posteroinferior segments.

tion aspiration device (e.g., ultrasonic aspirator) and subpially connecting the anterior and posteroinferior segments through suction aspiration, coagulation, and incising sharply the pia using microscissors (Fig. 22–8). Once the lateral neocortex is removed, further aspiration is performed on the lateral part of the parahippocampal gyrus and uncus to make the second stage easier.

Medial Temporal Resection

The second stage consists of resection of the medial basal structures and is facilitated with the use of the operating microscopic. The hippocampal complex is identified by unroofing the temporal horn anteriorly (Fig. 22–8). The choroidal fissure is identified, and a cottonoid pledget can be placed over the choroid plexus and gently retracted superomedially for better visualization of the amygdalohippocampal complex. The commissural attachments between the hippocampus and the amygdala are incised from the ventricle anteriorly to the middle fossa floor, maintaining the integrity of the arachnoidal border between the superior and inferior compartments. The amygdala can be removed gently by using the ultrasonic aspirator at a low setting so as not to disrupt the sylvian fissure or its vessels. The hippocampal resection is initiated just lateral to the choroidal fissure by coagulating the alveus and accessing the subpial plane (Fig. 22–9). The hippocampus then is mobilized laterally, and the feeding vessels from the anterior choroidal and posterior cerebral arteries are coagulated as they pass through the pia–arachnoid into the hippocampal complex (Fig. 22–2). Care must be taken because these hippocampal, or Ammon's horn, vessels can be stretched at their attachment to the parent vessel and avulsed; attempts at hemostasis then may cause damage to the parent artery and the blood supply to the basal ganglia or the brainstem. Meticulous dissection is therefore necessary to identify, coagulate at a low setting, and sharply cut the perforating vessels to avoid injury to the parent vessel and an ischemic complication postoperatively. Once freed of the perforators from the deep vessels, and using gentle aspiration and a subpial technique, the hippocampus is elevated in an anterior-to-posterior manner and resected posteriorly in the tail of the hippocampus through the fimbria at the level of the tectum of the brainstem (Fig. 22–2).

Postresection and Closure

Hemostasis can be achieved by using bipolar coagulation along the remaining pial border, and occasionally thrombin-soaked Gelfoam can be placed in the medial resection site to avoid heat damage to the remaining perforating vessels. Oxidized cellulose often is used to line

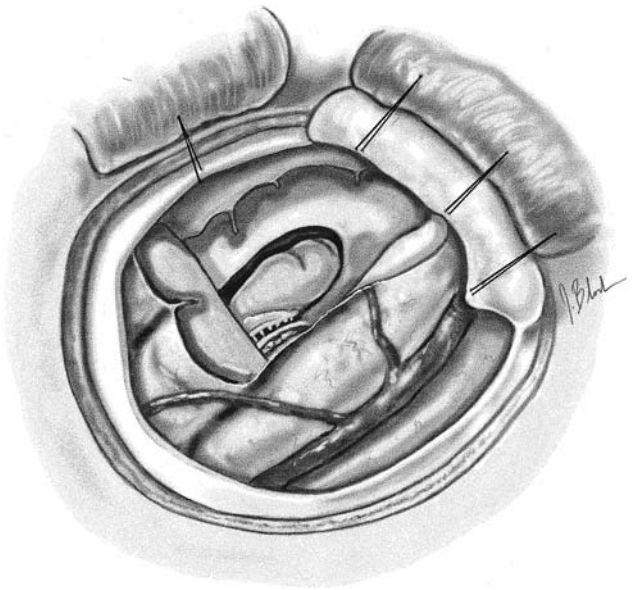


FIGURE 22–8. View of the mediobasal segments following the anterolateral resection and unroofing of the temporal horn of the lateral ventricle. As shown, the temporal horn is found via the white matter at the superoposterior angle of the resection.

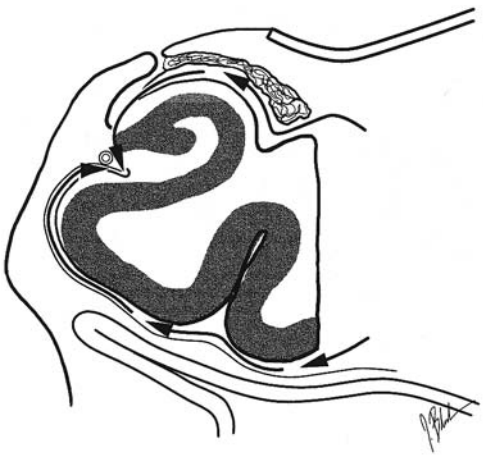


FIGURE 22–9. Once in the temporal horn, the choroidal fissure is identified. The white matter border zone lateral to the fissure is coagulated and incised, which gives access to the subpial plane and the perforating vessels. The hippocampus then can mobilized laterally and medially with suction aspiration in the subpial plane and eventually resected.

the resection borders, but it should be avoided in patients with tumors to avoid complicating the postoperative imaging results.

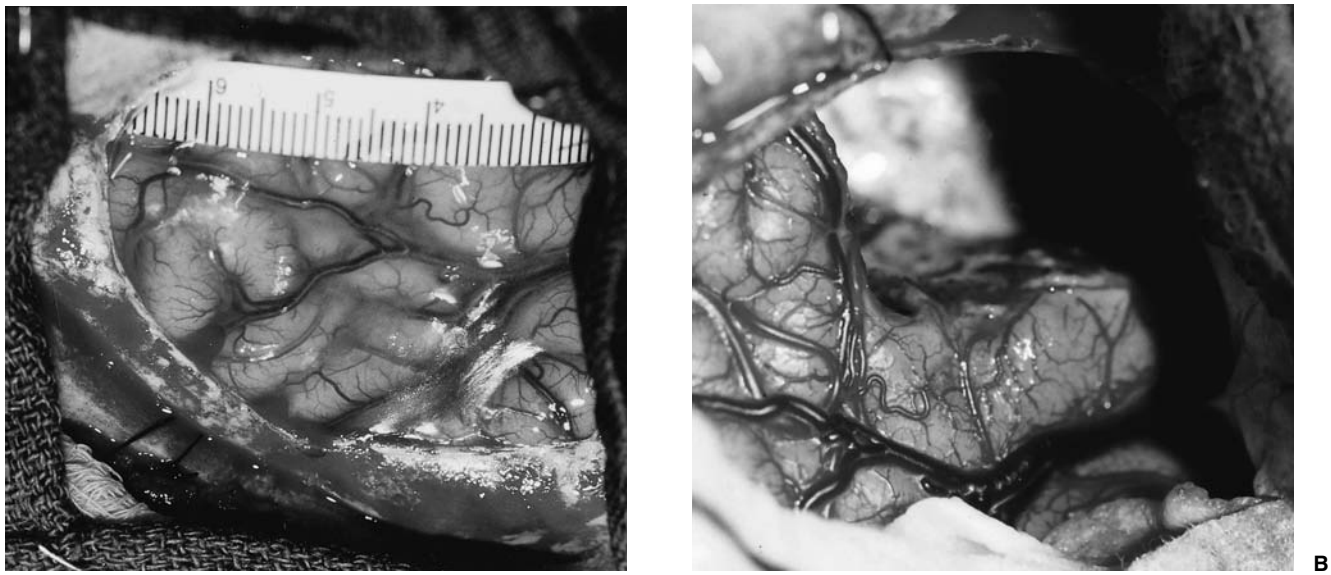
After the resection, ECoG can be done to ensue complete removal of the epileptogenic focus by placing the surface electrodes along the superior, posterior, an inferior borders of the resection cavity. Occasional interictal activity may be seen, but the overall activity usually is diminished, and the intraoperative EEG serves as a baseline for future studies. Clear epileptogenic activity seen on the ECoG at a focal area at the border of the resection site may be further resected, particularly if the preoperative studies indicated a lateral neocortical abnormality. In these instances, the lateral resection may not have adequately incorporated the complete epileptogenic zone.

Once the resections have been completed (Fig. 22–10) and hemostasis achieved, the dura is reapproximated and closed in a watertight fashion using interrupted or running absorbable or nonabsorbable suture. A graft is rarely needed, but pericranium or a small piece of Gelfoam may be adequate for repair of small defects or difficult closure areas. An antibiotic-impregnated piece of compressed Gelfoam conformed to the bony defect in the epidural space or a central dural tacking suture can be placed to minimize the risk of a postoperative epidural hematoma. The bone flap is replaced using a 2-0 nonabsorbable suture. The burr holes and the craniotomy then are completely covered with the reapproximation of the temporalis muscle and its fascia to the cuff of tissue at the superior temporal line. Because the muscle and the tissue cuff remain vascularized and the muscle is functionalize by returning it to its attachment, there is an excellent cosmetic outcome with little chance of temporalis atrophy and visualization of the burr hole and craniotomy defects. The temporalis, galea, and skin all are closed with absorbable suture, and the head is dressed sterilely along the incision. A firm wrap is applied to diminish swelling.

POSTOPERATIVE MANAGEMENT

The patient is observed in the intensive care unit overnight after surgery. In general, the postoperative medications depend on the surgeon's preference. Antibiotics are administered routinely for 24 hours; if corticosteroids are started, they can be tapered over a 4- to 7-day period. The preoperative anticonvulsant medication will continue postoperatively for at least 1 to 2 years and should be restarted immediately with adequate serum levels. An occasional seizure in the early postoperative period is not unusual and is probably secondary to cortical irritation from the debris, inflammatory reaction, and resolving blood products. These events are not prognostic of long-term outcome as long as they do not continue. Patients can be discharged when they are ambulatory and can tolerate a regular diet; this requires on average a 3- to 7-day hospital stay.

The postoperative evaluation of these children includes a routine postoperative follow-up 2 to 4 weeks af-



A B

FIGURE 22–10. A: Preoperative view. **B:** Postoperative view at the completion of a left temporal lobectomy. The floor of the middle fossa is well visualized and the superior temporal gyrus is intact, as is the sylvian fissure.

ter discharge. A more formal postoperative epilepsy evaluation that includes electrophysiologic, neuropsychologic, and anatomic evaluations (Fig. 22–11) is performed 6 months after surgery to assess the early results of the surgical intervention. Other follow-up visits are arranged for 1 and 2 years after surgery to decide whether discontinuation of medication is feasible.

COMPLICATION AND COMPLICATION AVOIDANCE

The percentage of surgical morbidity from temporal-lobe resections in general is low, on the order of 2 to 8%, with infection the most common postoperative complication. Local wound infections often can be treated simply with debridement and a course of pathogen-directed antibiotic therapy. Deeper infections of the bone or intracranial space may require more aggressive measures once medical therapy has failed. In the author's series of 70 temporal lobectomies, two superficial infections have occurred, one associated with electrodes and one without.

Neurologic deficits usually occur as a result of either vascular injury or as part of the resection. The classically described complications now can be avoided with the methods already presented in detail. A homonymous superior quadrantanopsia occurs when there is interruption of the optic radiations that loop into the temporal lobe prior to tracking posteriorly to the occipital lobe. This deficit is avoided by limiting the superior and posterior extent of the anterolateral resection. By not resecting the superior temporal gyrus, the incidence of language or verbal deficits, which can occur when language areas of the dominant hemisphere are resected, also are minimized. This problem rarely occurs in children younger than 9 years of age because language is not developmentally complete until 7 to 10 years of age. Also, the described technique limits the resection inferior to the superior temporal sulcus, which lessens the risk of developing manipulation hemiplegia because there is no interference and possible spasm of the sylvain vessels as can occur with en bloc resections, which has its subpial resection along the sylvian fissure. Motor or sensory deficits still can occur when there is manipulation and injury to the anterior choroidal artery and its perforating vessels to the internal capsule and basal ganglia or the posterior cerebral artery and its perforating vessels to the brainstem. Careful dissection, coagulation, and sharp incision of the Ammon's horn vessels as they enter the hippocampus will lessen the risk of stroke or ischemia to these structures. Postoperative high-dose steroids are usually effective in lessening long-term functional deficits.

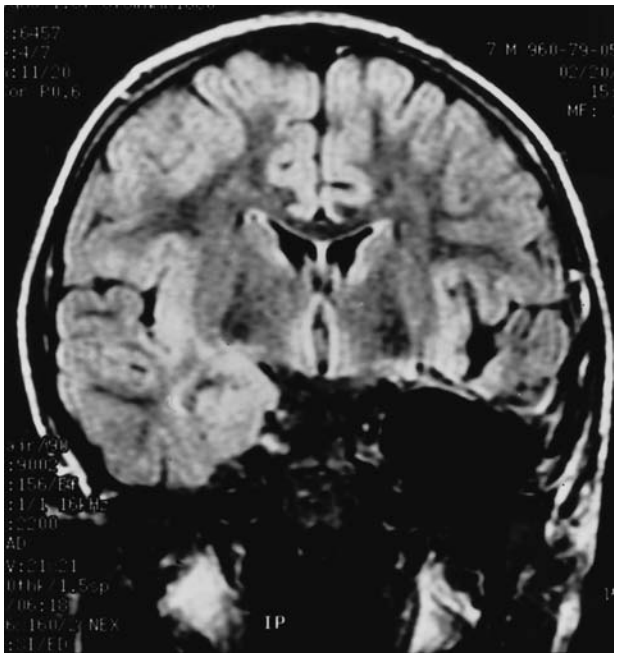


FIGURE 22–11. Postoperative MR (T2 weighted) image of the resection cavity following a left temporal lobectomy. Note again the preservation of the superior temporal gyrus and resection of the medial structures.

EDITOR'S COMMENTARY

The anatomy of the temporal lobe is complex. Neurosurgeons planning a temporal lobectomy are obligated to know the anatomy in the detail described in this chapter. The use of a frameless stereotactic guidance system may be helpful in obtaining and preserving anatomical orientation in these cases, particularly for less-experienced neurosurgeons. I (ALA) have performed both the en bloc resections described by Falconer and the two-stage procedure described in this chapter, and agree with Adelson that the latter is generally safer and the medial anatomy more easily defined. Throughout the chapter, Adelson repeatedly uses the word "avoiding", emphasizing that meticulous attention to the details of resection is necessary if the procedure is to have an acceptable complication rate. Complications cannot be avoided but can be minimized. The frequency of infection in these cases is partially attributable to the use of strip and grid electrode monitoring for several days before the resection is performed, and is probably unavoidable.

PEARLS

In this author's experience:

- Temporal-lobe epilepsy most often begins in childhood during the first decade.
- The amygdalohippocampal complex is the most common source of medically intractable complex partial seizures in the adult population, but only has an incidence of approximately 30% in children.
- Since the seizures have a focal onset and are accessible for surgical extirpation, temporal-lobe seizures remain the single most important subtype of seizures for successful surgical treatment.
- Intractable seizures in children have a "malignant" natural history, which eventual declines in both intellectual and behavioral functions.
- Patients with imaging abnormalities rarely "outgrow" their seizures.
- The preoperative evaluation localizes the epileptogenic zone through a concordance of multiple types of data that provide information on the structural and functionality of the tissue.
- In children, the pathologic substrates vary, but the epileptogenic zone is either lateral neocortical or mediobasal.
- Excellent surgical and seizure outcomes are now possible with little morbidity and virtually no mortality.

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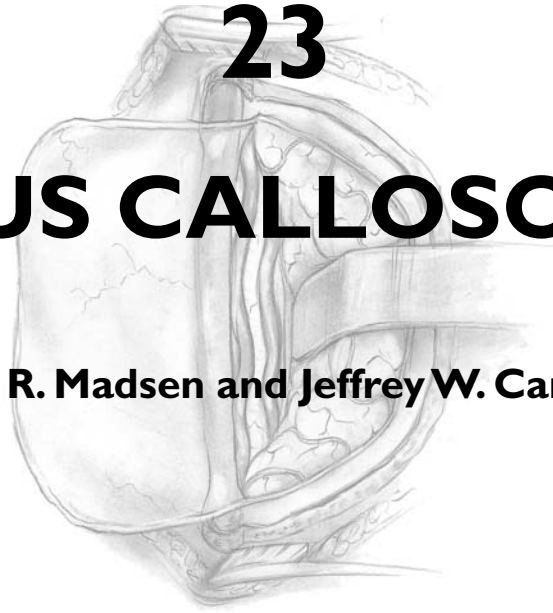
23

CORPUS CALLOSOTOMY



Joseph R. Madsen and Jeffrey W. Campbell

Corpus callosotomy originally was designed for medically intractable epilepsy patients with secondary generalization of focal seizures, in whom cortical resection of the seizure focus is not feasible. In addition, beneficial effects can be seen in children with generalized seizures in whom electroencephalography (EEG) demonstrates simultaneous bilateral sites of onset. Unlike other surgical therapies for epilepsy in which the epileptic focus is identified and removed, callosotomy interrupts the propagation of epileptic discharges, resulting in a truly functional procedure with the target being a physiological process rather than a pathological lesion. Conceptionally, severing the major connection between the hemispheres might logically reduce the generalization of a unilateral seizure focus. The benefit for patients with bilateral onset of epileptigenic activity is more difficult to explain, but it may result from global diminution of synaptic connectivity within the cerebrum. This "synaptoreduction" also may help to explain the observation that postoperative EEGs rarely demonstrate well-lateralized foci of seizure activity, which might be expected if corpus callosotomy simply prevents propagation of epileptic activity between the hemispheres.

The surgical technique is relatively straightforward and is associated with minimal morbidity and mortality. A few patients develop an acute disconnection syndrome characterized by mutism, nondominant left-arm and left-leg apraxia, inattention, bilateral Babinski signs, and urinary incontinence, although fortunately these symptoms typically resolve quickly. Postoperative hydrocephalus can largely be avoided by careful attention not to violate the ventricular system. Long-term seizure control in postcallosotomy patients is difficult to ascertain because without generalization, many postoperative seizures may be subclinical. Few studies report sophisticated outcome analysis and quality-of-life measures, which are likely superior to seizure frequency in assessing the efficacy of callosotomy. Recent series have been encouraging, however, with 56 to 100% of patients experiencing significant improvement in the frequency of generalized seizures. Although predicting which patients will benefit from a callosotomy remains difficult, it is clear that patients with "drop" attacks derive the most benefit from the procedure. The goal of this operation is not to make the patient seizure free but to improve quality of life and function.

SURGICAL INDICATIONS

The clinical indications for callosotomy continue to evolve with the development of newer investigational and therapeutic techniques. In particular, many centers are inserting vagal nerve stimulators (VNS) in most patients with seizures previously treated with callosotomy. The long-term outcome with this treatment strategy remains unknown, although many of these patients may undergo callosotomy secondarily if generalized seizures persist. Most centers consider callosotomy a safe and effective treatment for selected patients with intractable, debilitating, generalized seizures. In particular, atonic–tonic seizures, or "drop" attacks, are accepted as the most responsive seizure type. The role of callosotomy in partial epilepsy is controversial, with dramatically varying results in different centers.

At Children's Hospital in Boston, patients unresponsive to adequate trials of multiple antiepileptic medications with an absence of a resectable seizure focus are considered for callosotomy if there exist: (1) the presence of incapacitating generalized seizures causing drop attacks or interfering significantly with quality of life; (2) recurrent episodes of convulsive or nonconvulsive status

epilepticus; or (3) a desire to lateralize a frontal lobe focus where long-term EEG monitoring is inconclusive. The first two groups of patients are also good candidates for VNS, which we presently prefer as an initial procedure. Because roughly 50% of patients with VNS continue with intractable seizures, many patients may benefit secondarily from a callosotomy. Contraindications include an inadequate trial of antiepileptic agents, nondebilitating seizure pattern, or surgically accessible seizure focus. In addition, if there is a focal lesion that could lead to a new neuropsychologic deficit if combined with division of the corpus callosum, or if significant language function resides in both hemispheres, a callosotomy usually is not performed.

The presumed developmental potential of the child is our primary criterion for selecting a patient for a single-stage complete callosotomy rather than an initial anterior callosotomy. A partial callosotomy can be followed by a second procedure to complete the division of the corpus callosum if significant seizures persist. Whereas severely impaired children clearly have less morbidity with a single-stage procedure, the risks of a postcallosotomy disconnection syndrome often outweigh the advantage of a single-step procedure in patients who are expected to be able to read and live independently. There are no data to suggest that a complete callosotomy performed in two stages is more or less effective in dealing with seizures than a single-stage procedure.

PREOPERATIVE MANAGEMENT

The preoperative evaluation includes EEG monitoring to demonstrate sites of seizure origin as well as high-quality magnetic resonance imaging (MRI) to identify structural lesions that may be associated with epileptic activity. The MRI is also valuable for operative planning in defining the anatomy of the corpus callosum and its surrounding vasculature. A Wada test, consisting of intracarotid sodium amytal injection, is performed when there is a possibility of significant language function residing in the nondominant hemisphere for handedness, as seen with mixed or crossed language dominance. Ictal and interictal studies of metabolism or blood flow are used by some centers as confirmatory data to localize areas of seizure activity. Formal neuropsychologic evaluation should be obtained when possible.

As with all major surgery, the patient must be evaluated carefully for previous medical problems or physical findings that might complicate the intraoperative or postoperative course. A complete history and physical examination including appropriate blood work is vital to ensure the highest level of safety for the patient. Certain antiepileptic medications are known to interfere with coagulation, and some centers stop administering agents such as valproic acid several weeks prior to surgery. Careful discussion of the procedure, alternatives, possible complications, and likely results must be held with the parents and patients of sufficient age to participate in the decision.

INTRAOPERATIVE TECHNIQUE

Anterior callosotomy and single-stage complete callosotomy are performed with the patient under general anesthesia with the usual monitoring required for such a procedure, including adequate venous access, blood pressure monitoring, Foley catheter, and cardiac Doppler to detect air embolism. These children are often sensitive to narcotics and other central nervous system depressants even before the callosotomy is performed. As a result, we minimize the intraoperative and postoperative use of these medications. Intraoperative corticosteroids are not routinely used, although osmotic diuresis with mannitol or mild hyperventilation can minimize the degree of brain retraction required. Patients continue taking preoperative anticonvulsant medication except when that medication might interfere with coagulation. Prophylactic antibiotics are administered prior to the incision and every 6 hours during the procedure.

The patient is positioned supine with the head elevated and slightly flexed (Fig. 23–1). We use a 10-cm linear incision parallel, and 2.5 cm anterior to the coronal suture, centered 2 cm to the right of midline (Fig. 23–2). Two burr holes are placed on the midline, one located at or just posterior to bregma, the confluence of the coronal and sagittal sutures, and the other 5 cm anterior to bregma (Fig. 23–2). A trapezoidal or D-shaped cran-

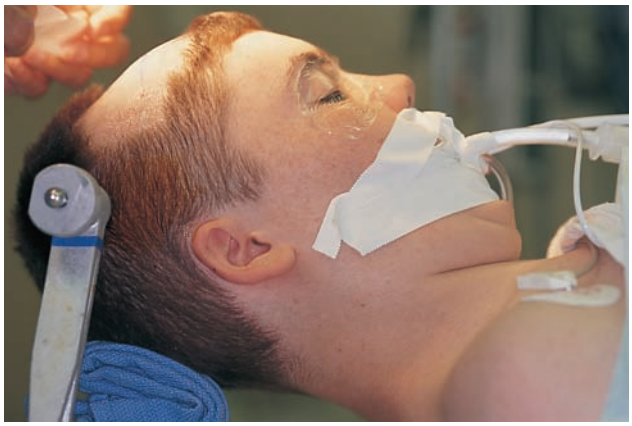


FIGURE 23–1. The patient is positioned supine with slight flexion of the neck. If the patient is older than 1 year, the head is placed in pin fixation.

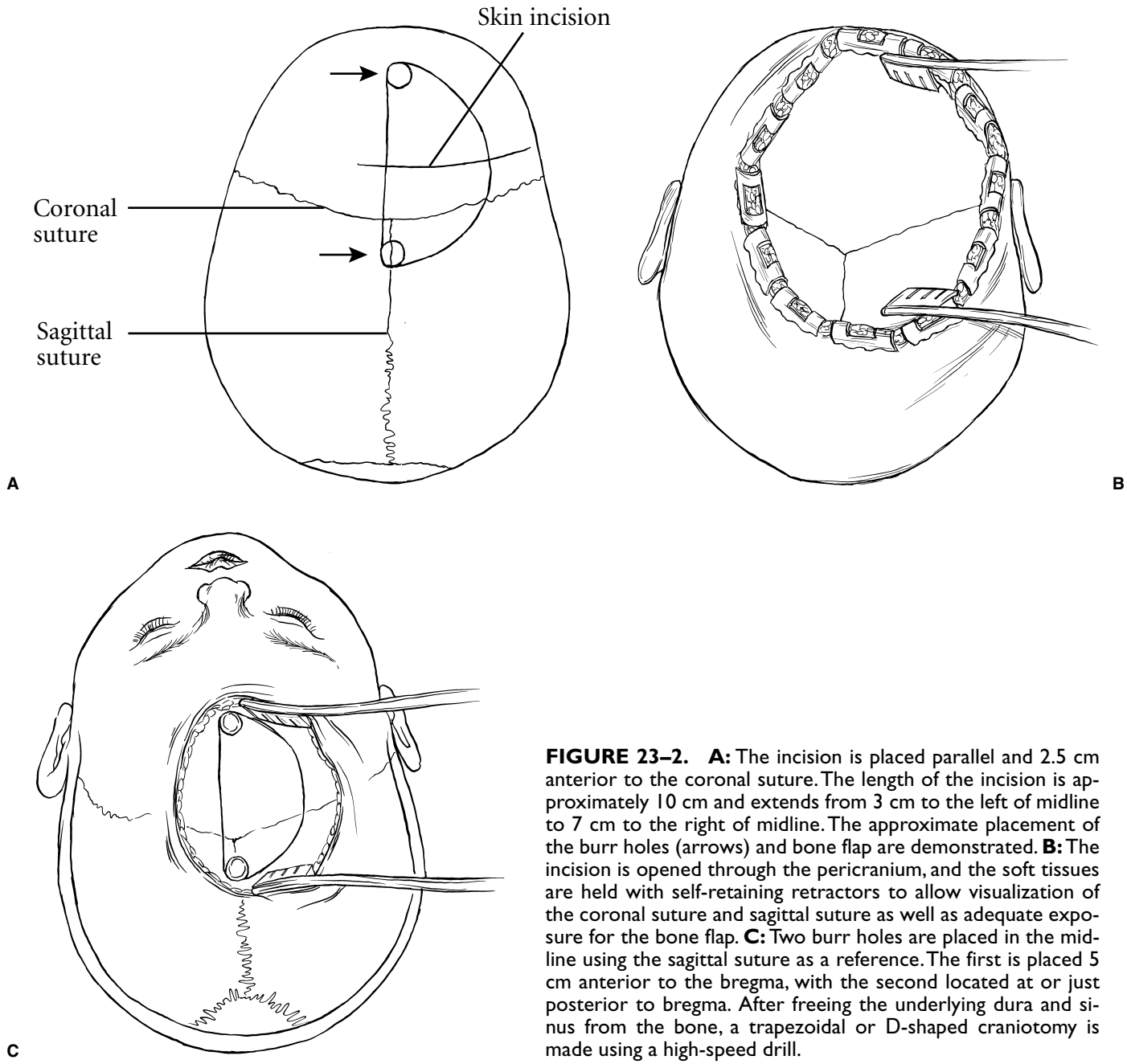


FIGURE 23–2. **A:** The incision is placed parallel and 2.5 cm anterior to the coronal suture. The length of the incision is approximately 10 cm and extends from 3 cm to the left of midline to 7 cm to the right of midline. The approximate placement of the burr holes (arrows) and bone flap are demonstrated. **B:** The incision is opened through the pericranium, and the soft tissues are held with self-retaining retractors to allow visualization of the coronal suture and sagittal suture as well as adequate exposure for the bone flap. **C:** Two burr holes are placed in the midline using the sagittal suture as a reference. The first is placed 5 cm anterior to the bregma, with the second located at or just posterior to bregma. After freeing the underlying dura and sinus from the bone, a trapezoidal or D-shaped craniotomy is made using a high-speed drill.

iotomy is made with a high-speed drill. To achieve true midline visualization of the corpus callosum, it is vital that the bone flap comes to or slightly past the midline of the sagittal sinus. If the sinus is firmly adherent to the bone, the edge of the craniotomy should be placed slightly to the *left* of the sinus to avoid inadvertent injury.

The dural opening is made parallel to the midline, at least 1.5 cm from the edge of the sagittal sinus (Fig. 23–3). Bridging veins should be preserved carefully during the dural opening, and venous lakes in the dura may need to be oversewn. Although an experienced surgeon can visualize the entire corpus callosum through a smaller bone opening, we prefer 5 cm of exposure to allow flexibility in avoiding the bridging veins that may be encountered. We have not found it necessary to perform preoperative angiography, although the MRI should be inspected for unusual venous anatomy, which may be encountered in the opening. After the dura is opened, the right frontal lobe is gently retracted away from the falx cerebri. A fixed retractor system, such as the Greenberg or Budde Halo, is used to provide careful constant retraction during the procedure. The retractor blades should be at least 1 to 2 cm wide and padded with Telfa or Biacol to avoid retraction injury to the frontal lobe.

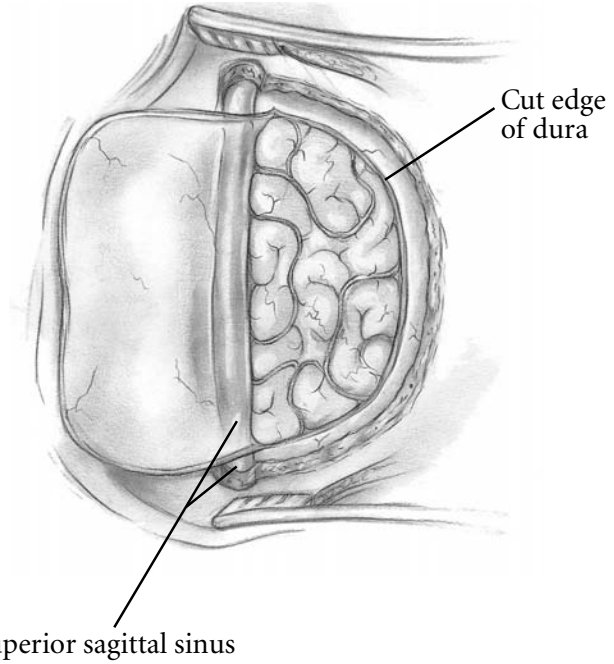


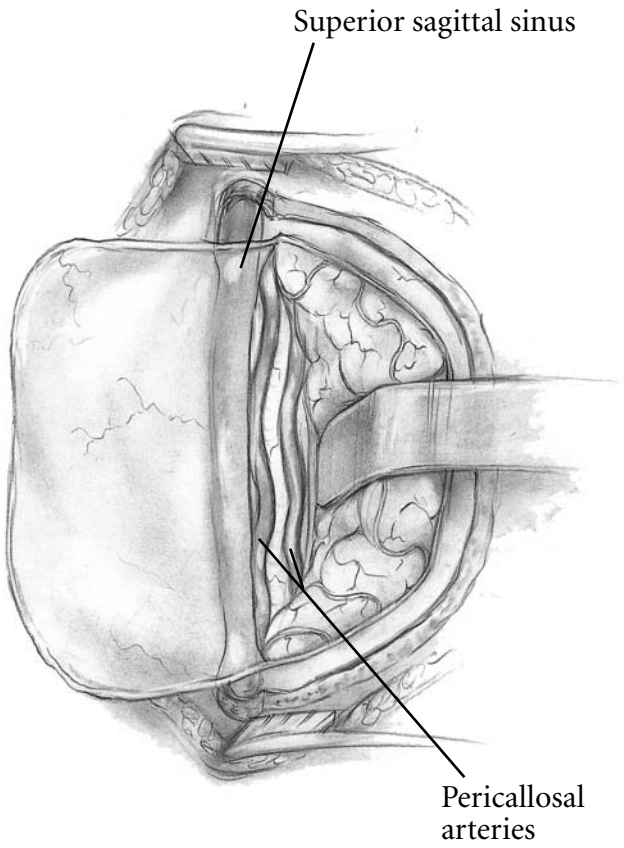
FIGURE 23–3. The dura is opened in a horseshoe-shaped based on the sagittal sinus. The width of the dural flap can be as little as 1.5 cm; the length should be close to 5 cm to allow flexibility around bridging veins.

The operating microscope is used from this point and improves visualization of the arachnoidal adhesions between the two cingulate gyri that are often encountered just deep to the falx. This dissection is continued until the entire corpus callosum and two pericallosal arteries are exposed (Fig. 23–4), being mindful not to tear superficial bridging veins with the frontal lobe retraction. The callosal marginal arteries and cingulate gyrus can be mistaken for the pericallosal arteries and corpus callosum, although the corpus callosum is significantly whiter and smoother than the cingulate gyrus. After exposure of the pericallosal arteries and corpus callosum, the left and right arteries can be identified by following small branches off these arteries to their respective hemisphere. The two arteries are dissected apart so that the actual callosotomy can be made between them. If this is not possible without significant manipulation of the arteries, the callosotomy can be performed lateral to both arteries, although staying on the true midline is more difficult. For a single-stage complete callosotomy, the entire corpus callosum is exposed from genu to splenium. For a partial anterior callosotomy, the posterior extent of the dissection can be determined either by measuring the distance back from the genu based on the anticipated length from the MRI or by direct visual inspection of the corpus callosum.

The dissection into the corpus callosum is made at the level of the coronal suture with suction aspiration, with the goal of not entering the ventricular system. Even in patients without a cavum septum pellucidum on MRI, there is usually a small space between the two leaves of the septum pellucidum that allows complete severing of the corpus callosum without entry into the lateral ventricle. This space is found by starting the superficial dissection directly in the midline, which is sometimes marked by a small vein on the corpus callosum. If the ependyma is seen, as evidenced by the darker color of the underlying ventricular fluid, manipulation of the tissue is avoided, and exploration can be done to either side in search of the midline cleft. Once the midline is identified, a small ball-tipped microdissector is inserted into the cleft and used in conjunction with suction aspiration for a rapid dissection of the corpus callosum while avoiding entry into the ventricular system (Fig. 23–5). If the ventricle is entered, a small piece of Gelfoam is placed over the entry site to prevent contamination of ventricular cerebrospinal fluid (CSF) with blood or debris.

Continuing the dissection anteriorly, the midline cleft ends in a small pit at the anterior border of the ventricular system. Remaining in the same plane, the genu of the corpus callosum is undercut, and the subarachnoidal surface of the genu is gently retracted inferiorly and posteriorly to separate it from the anterior cerebral arteries, which loop anteriorly. This allows complete division of the anterior corpus callosum to be done safely, removing all of the commissural tissue except for the anterior commissure, which is usually seen at this point in the dissection. A similar maneuver is used to remove the posterior corpus callosum. For a complete callosotomy, the midline cleft is followed to the posterior border of the ventricular system. The splenium is dissected in a fashion analogous to the genu, with completion of the dissection marked by visualization of the vein of Galen through an arachnoid sheath posteriorly. When a partial, anterior callosotomy is performed, the dissection is completed to the expected distance back from the genu.

After completion of the callosotomy, absolute hemostasis is obtained. We generally cut a piece of Gelfoam to the expected size and shape of the callosotomy and insert it into the defect. In addition to aiding in hemostasis, this is a convenient means for checking the length of callosum removed in a partial callosotomy on a postoperative MRI (Fig. 23–6). The retractors are removed, and the frontal lobe is inspected carefully for any sites of injury or bleeding. Absolute hemostasis can be confirmed by performing a Valsalva's maneuver or raising the patient's blood pressure. The dura is closed in a watertight fashion, and the bone flap is reattached using sutures or plates. We do not place dural tacking sutures unless there are particular concerns about a postoperative epidural collection. The galea and skin are closed in a routine fashion.



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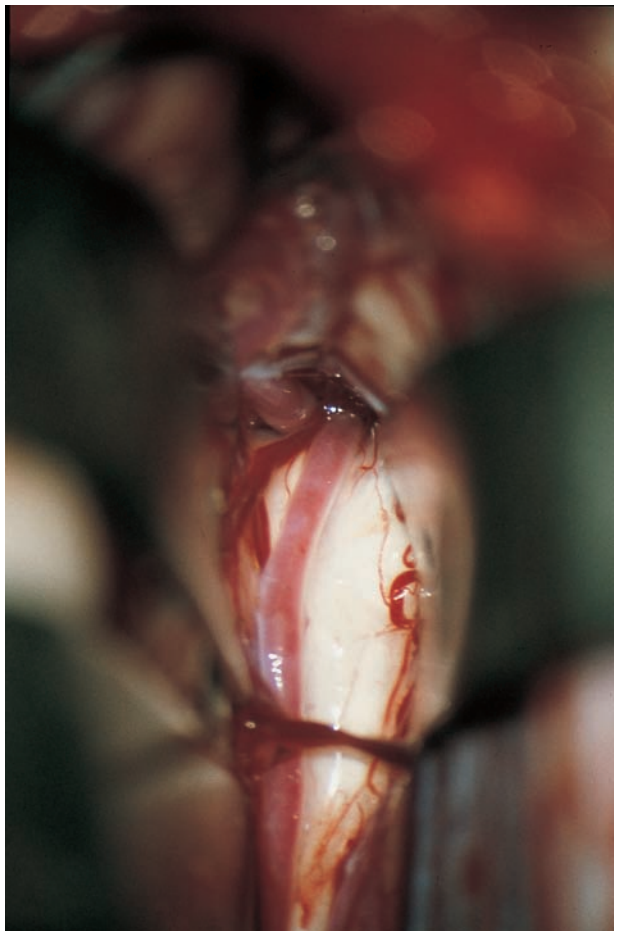

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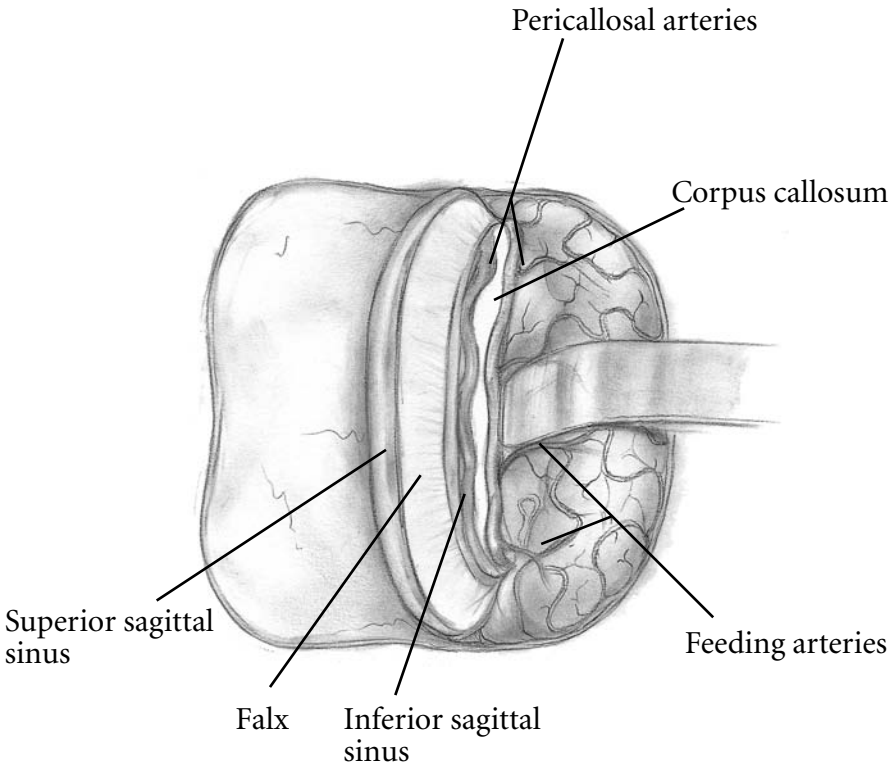
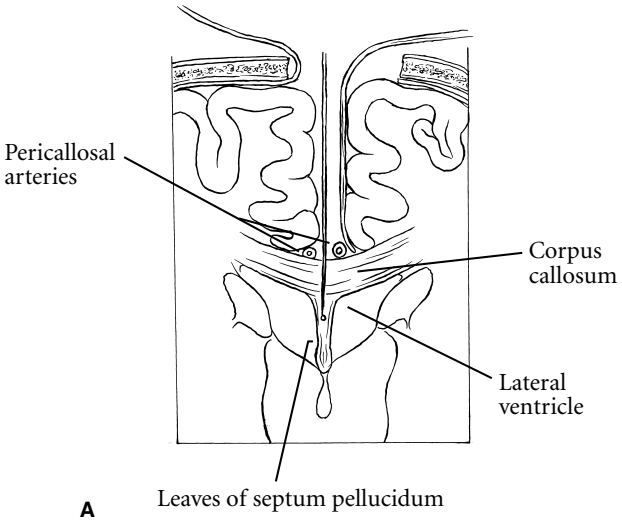


B

FIGURE 23–4. **A:** A fixed retraction system is used to retract the frontal lobe laterally until there is adequate visualization of the pericallosal arteries and corpus callosum. The microscope is used to aid in this dissection. **B:** The two pericallosal arteries are identified lying on the white corpus callosum. If there is any uncertainty as to which artery feeds which hemisphere, the small perforating arteries can be followed to the respective cingulate gyrus. **C:** The pericallosal arteries are separated gently to allow working space between them. In this photograph, the right pericallosal artery is out of view behind the retractor, allowing safer dissection of the corpus callosum.



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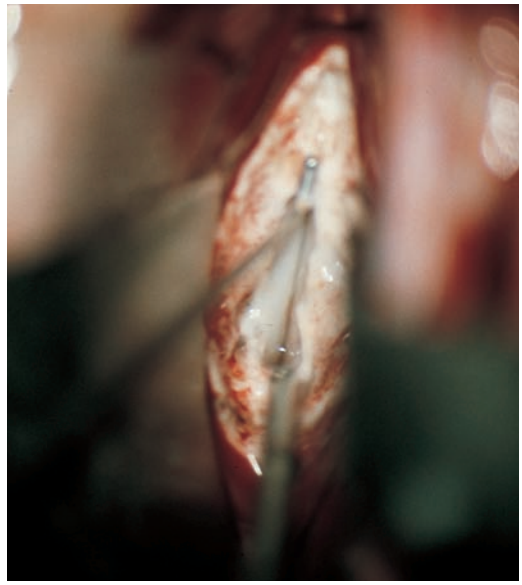

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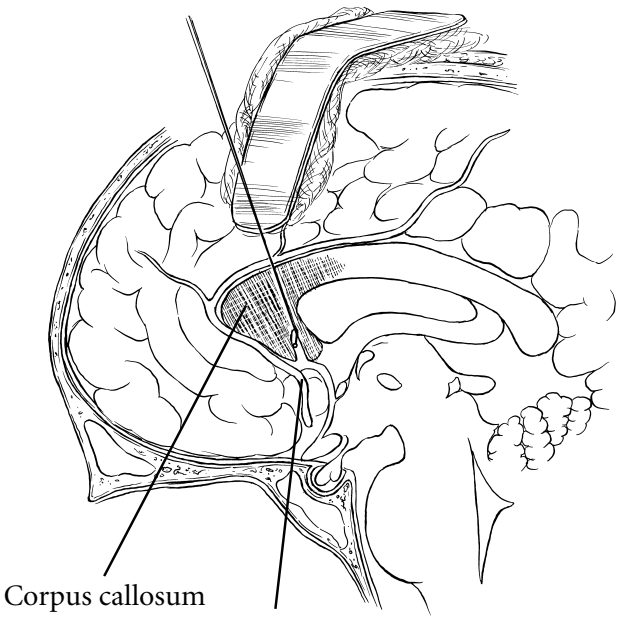

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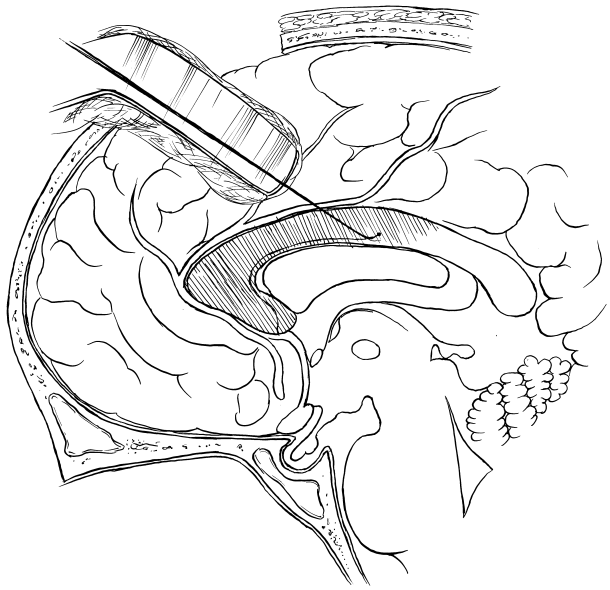
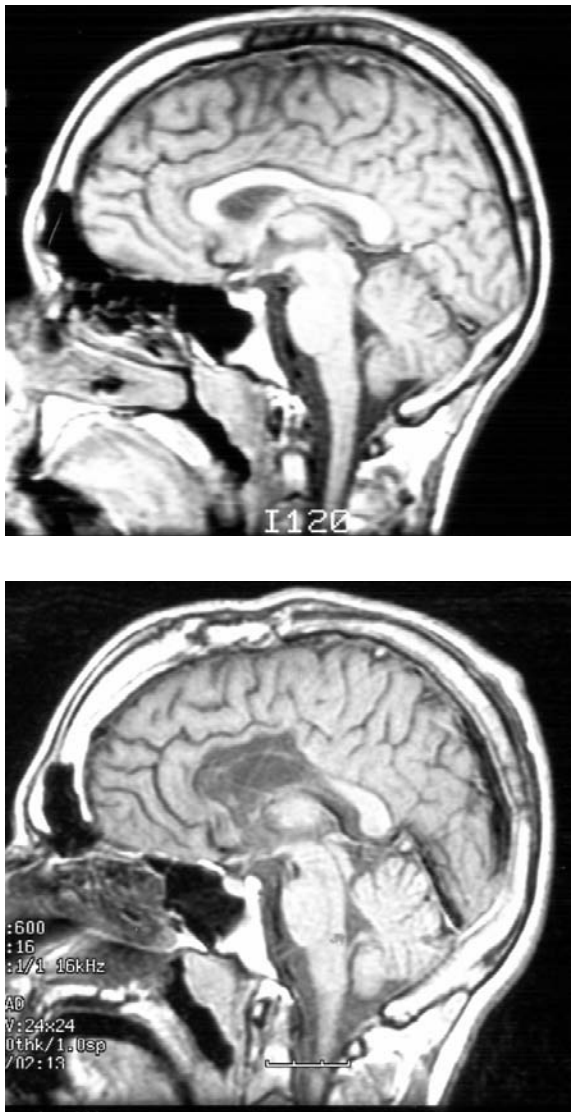


D

FIGURE 23–5. **A:** Careful dissection through the midline of the corpus callosum will lead into a potential space between the leaves of the septum pellucidum. A small ball-tipped microdissector can be inserted into this cleft to allow rapid sectioning of the corpus callosum without entry into the ventricular system. **B:** With this method, the dissection is carried anteriorly toward the genu. **C:** The genu is undercut until the subarachnoid space above the corpus callosum is reached, revealing the anterior cerebral arteries. **D:** The dissection is carried posteriorly toward the splenium. If a complete callosotomy is performed, the splenium is undercut until the pineal region is visualized.

POSTOPERATIVE MANAGEMENT

Patients are observed in an intensive care unit overnight so that any complications can be discovered quickly and appropriately treated. Postoperative seizures are treated with additional antiepileptics and benzodiazepines and are not thought to be predictive of the procedure's success. Any patient with a new or progressive postoperative deficit should undergo immediate neuroimaging to evaluate for either an ischemic or hemorrhagic complication. Routine postoperative imaging is generally not necessary except to evaluate the extent of callosal divi-



A

B

FIGURE 23–6. Preoperative **(A)** and postoperative **(B)** T1-weighted midline sagittal MRI demonstrating division of the anterior two thirds of the corpus callosum.

sion after an anterior callosotomy. Patients are continued on their preoperative anticonvulsant therapy with the hope of weaning them slowly if there is a significant decrement in seizures. Most patients remain on anticonvulsant medications indefinitely, even when generalized seizures significantly improve.

COMPLICATION AVOIDANCE

Disconnection Syndromes

Acute disconnection syndrome occurs to some degree in most patients following section of the corpus callosum. This is characterized by mutism, nondominant left-arm and left-leg apraxia, inattention, bilateral Babinski signs, and urinary incontinence. Other symptoms, such as visual or limb agnosia, confusion, bilateral grasp reflex, or competitive movements between the left and right hands, are reported. Whereas most of these findings are transient, left-sided apraxia can persist. Mutism may be marked but usually clears within several weeks. Quantitative studies of this syndrome have not been performed, and their frequency, usual severity, and time course are poorly defined.

In addition to the acute disconnection syndrome, dysphasia can occur in patients with mixed or crossed cerebral dominance for language. Dysphasia and difficulties with comprehension can be seen in right-handed patients with crossed dominance, whereas dysgraphia alone is more common in left-handed patients with crossed dominance. Furthermore, patients can have difficulty with speech output without actual dysphasia or dysgraphia, which is thought to be due to buccofacial apraxia. Interhemispheric sensory disconnection can occur following section of the posterior half of the callosum; in this situation, patients cannot verbally identify objects introduced to the sensorium contralateral to handedness. Fortunately, visual and tactile scanning allows both hemispheres to have adequate access to the patient's entire surroundings, greatly diminishing the potential disability of this deficit.

Memory deficits are noted in a minority of patients following callosotomy. Disorders of recent memory are more common when preoperative extracallosal injury extends to the fornix and its connection. Most new postoperative deficits involve difficulty with both encoding and retrieval of information that require interhemispheric cooperation. The *split-brain syndrome* refers to sensory disconnection and nondominant hand apraxia, when the patient cannot perform fine motor movements on command. At times, the patient may have great difficulty getting the two hands to work together on a task, such as using a knife and fork or folding a letter and inserting it into an envelope. These deficits in sensorimotor integration correlate with involvement of the posterior corpus callosum.

Limiting the extent of the callosotomy appears to reduce the risk of developing problems related to hemispheric disconnection. Therefore, most patients undergo an initial anterior callosotomy that is followed 6 to 12 months later with completion of the callosotomy if the partial section alone is not effective. When a significant disconnection syndrome is unlikely to be detrimental to the patient, a complete callosotomy remains a safe and efficacious procedure with reduced risks from multiple operations. At Children's Hospital, patients with severe mental retardation who meet the previously described indications for callosotomy undergo a single-stage complete procedure.

In addition, the age of the patient at time of surgery is predictive of long-term disability from hemispheric disconnection. Children born with agenesis of the corpus callosum or who undergo callosotomy at a young age seem to experience fewer sustained deficits referable to the disconnection. The remarkable plasticity of younger patients may be related to a critical period in development consisting of synaptic overproduction and redundancy that would favor the reinforcement of alternative neural pathways. These compensatory mechanisms appeared to become more limited in adolescence, when synaptic distribution assumes adult patterns.

Hydrocephalus

One early report on corpus callosotomy for epilepsy revealed an unacceptably high incidence of postoperative hydrocephalus. The authors conjectured that the violation of the ventricular system allowed blood and debris into the CSF, resulting in alterations of CSF absorption. They recommended careful microsurgical techniques to avoid entry into the lateral ventricle, and more recent reports described a significantly decreased incidence of this complication. Hydrocephalus is now considered a rare complication that is largely preventable by careful avoidance of the ventricular system

EDITOR'S COMMENTARY

With the advent of the vagal nerve stimulator (VNS), the hope, as with any new drug, was that this would eliminate the need for the corpus callosotomy. It has not. While clearly helpful with some patients (those with complex partial siezures), atonic or "drop attacks," in this editor's experience (PDA), have been absolutely recalcitrant to improvement with VNS. In these instances, we are now prospectively planning placement of the VNS with follow-up surgery for the corpus callosotomy. In addition, the authors have highlighted the issues of complete vs. partial corpus callosotomy well. In this editor's experience, patients with little language or cognitive capability rarely suffer a significant decline in function following a complete callosotomy and thus it is recommended as a first line procedure in these instances rather than performing the partial with the likely need for a second stage completion. Some have advocated moving the craniotomy more posteriorly when planning a complete callosotomy. This editor, similar to the authors, has not found it necessary. Staying for the most part anterior to the coronal suture still allows the surgeon the adequate exposure and access to visualize the splenium and the underlying structures.

PEARLS

In these authors' experience:

- Drop attacks are the seizures most effectively treated by corpus callosotomy.
- One-stage, complete corpus callosotomy may be indicated for patients with severe neurological deficits.
- Anterior two thirds callosotomy may be appropriate for patients who can read or are expected to be able to read.
- Wada testing may be useful for patients without clear left cerebral dominance and who are old enough or functional enough to tolerate the testing cognitively.
- A complete corpus callosotomy can be performed through a small craniotomy just in front of the bregma; however, a slightly larger craniotomy enables the surgeon to avoid bridging veins by changing the angle of approach.
- Finding exact midline within the corpus callosum allows dissection of the raphe between the ventricles, and in most cases, permits callosotomy without entry into the ventricular system.
- Discontinuation of antiepileptic drugs that may alter hemostasis, especially valproic acid, should be planned weeks in advance of surgery.

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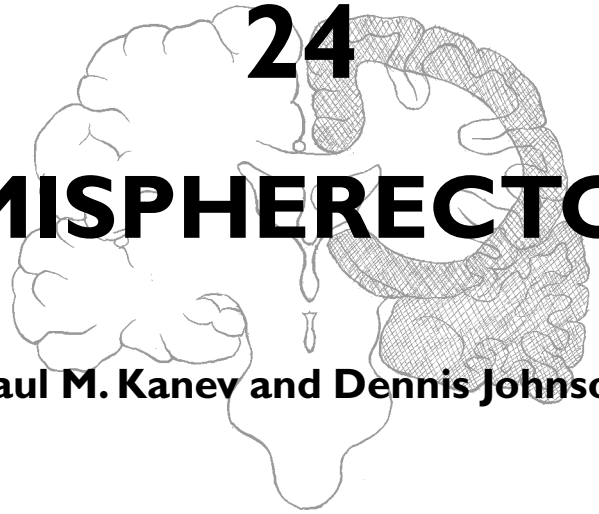
24

HEMISPHERECTOMY



Paul M. Kanev and Dennis Johnson

Walter Dandy completed the first hemispherectomy in 1928 when he attempted radical resection of a glioblastoma. Although surgical neurooncology containment was not achieved, the patient had acceptable postoperative neurologic function, and the foundation for anatomic resection of the cerebral hemisphere was introduced. In subsequent years, McKenzie (1938) and Krynauw (1950) reported the use of hemispherectomy in children with congenital hemiparesis who suffered intractable seizures. There was widespread acceptance of anatomic resection for seizure control until late hemorrhagic complications appeared, 3 to 20 years postoperatively. The mortality of chronic superficial hemosiderosis approached 33% in some series.

Numerous modifications of Dandy's surgical technique have been made to reduce the morbidity and long-term complications of hemispherectomy without reducing the long-term seizure control. Refinements have included the Adam's technique of suturing the dura to the falx, which reduced the subdural cavity in exchange for a large epidural space, and early postoperative spinal fluid shunting. After observing that hemorrhagic complications did not occur following multiple lobectomies, Rasmussen (1983) developed the now widely accepted technique of functional hemispherectomy. After temporal lobectomy and wide resection of rolandic cortex into the ventricle, cortical incisions were extended to the midline, disconnecting broad polar blocks of the frontal and occipital lobes, which retained their blood supply.

Other modifications to hemispherectomy have included hemi-decortication and hemicorticectomy. The technical evolution in hemispherectomy has been toward maximum conservation of cortical tissue. More recent refinements include intraventricular deafferentation procedures disconnecting large regions of cortex through infrasylvian and suprasylvian cortical windows. Endoscopic deafferentation in patients with congential porencephaly/infarct syndromes is on the horizon. We are pursuing the development of this technique in our laboratory.

Children appear quite sensitive to surgical manipulation within the body of the lateral ventricles; postoperative fever, lethargy, and inflammatory ventriculitis syndromes are common. We have used intraoperative ultrasound to tailor the frontal and occipital polar disconnections of modified hemispherectomy (MH) to avoid entrance into the ventricle and substituted central topectomy for rolandic resection. This chapter reviews the surgical techniques of hemispherectomy and highlights the use of intraoperative ultrasound to refine functional hemispherectomy resection.

SURGICAL INDICATIONS AND PREOPERATIVE EVALUATION

Children with intractable seizures are the best candidates for hemispherectomy. Generalized seizures are among the most common semiology, but complex partial, infantile spasms or jacksonian motor seizures may be encountered. The frequency of daily ictal events may range from several to 30 or more seizures. Seizures commonly begin in the first few months of life and are resistant to many anticonvulsant medications. In nearly all candidates for hemispherectomy, fine-dexterity hand movements are compromised. In select cases of patients suffering from

Rasmussen's encephalitis with progressive hemiparesis, hand dexterity and strength loss may be acceptable sacrifices for seizure control.

The etiology of intractable seizures in candidates for hemispherectomy includes hemispheric dysplasia, Rasmussen's encephalitis, cerebral hemorrhage or infarction, hemimegalencephaly, Sturge-Weber syndrome, schizencephaly, and porencephaly syndromes. Multiplanar magnetic resonance imaging (MRI) is the neuroradiological imaging study of choice and readily demonstrates the anatomic malformation of the affected cerebral hemisphere. The most common findings include encephalomalacia with an enlarged ipsilateral ventricle (Fig. 24–1). Other findings may include cortical dysplasias, vascular malformation, or schizencephaly. Single-positron emission computed tomography (SPECT) or PET identifies zones of abnormal cerebral blood flow and metabolism isolated within the involved cerebral hemisphere.

Preoperative evaluation is completed by the members of an integrated pediatric epilepsy program, including pediatric neurosurgeons, neurologists, and neuropsychologists. Multiple outpatient and video-telemetric electroencephalographic recordings are made to isolate ictal epileptiform discharges to a single cerebral hemisphere. Neuropsychology and Wada testing are completed in each patient older than 5 years of age to demonstrate that the normal cerebral hemisphere has capable memory and eloquent function. Intracarotid amobarbital injection of the effected cerebral hemisphere is helpful in predicting postoperative contralateral leg function. In children too young to complete language localization studies, left hemispherectomy can be performed with confidence that capable language dominance will evolve within the right cerebral hemisphere.

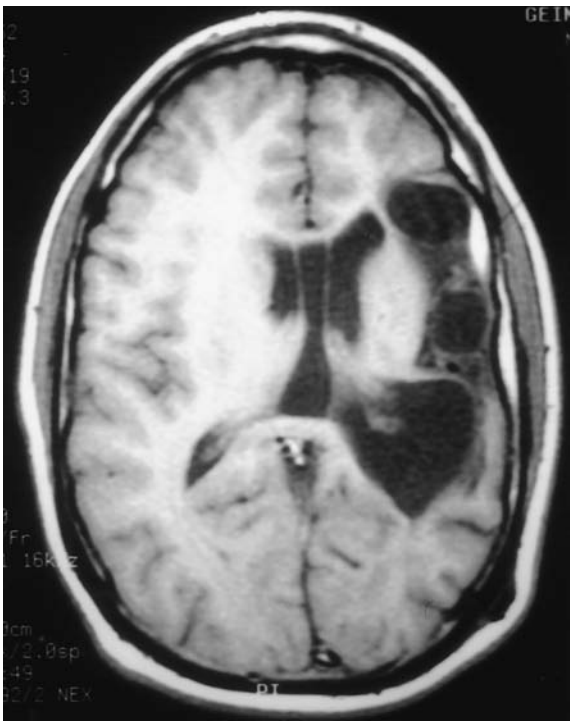


FIGURE 24–1. Preoperative axial T1 MRI sequence of an 18-year-old man with congenital right hemiparesis and intractable seizures. There is broad left middle cerebral territory infarction and mild ex vacuo left ventricular dilation.

PREOPERATIVE MANAGEMENT

Dexamethasone is started perioperatively (1 mg/kg daily up to 24 mg/day), and two units of donor directed packed red blood cells are prepared. The patient's usual anticonvulsants are given on the morning of surgery and are continued postoperatively for 1 year. In the first few days following surgery, drug levels can fluctuate widely and are closely monitored until corticosteroids are discontinued on the fifth postoperative day.

INTRAOPERATIVE TECHNIQUES

Anesthetic Techniques and Positioning

Children completing hemispherectomy do not have elevated intracranial pressure. Unlike patients with tumors or intracranial hemorrhage who have marginal cerebral perfusion, candidates for hemispherectomy are typically healthy and have normal cardiac indices. Deliberate volume expansion is well tolerated, and anticipation of blood loss during hemispherectomy warrants avoidance of hypovolemia. Mild hemodilution with 20 mL/kg of isotonic saline diminishes red blood cell loss during intraoperative bleeding and limits the volume of transfusion. Initial blood loss is supported with isotonic crystalloid solutions and then is replaced with donor-directed packed red blood cells when intraoperative hemoglobin drops below 7.5 g/dL.

Cerebral relaxation is achieved with mild hyperventilation; arterial pCO_2 is maintained at 28 to 30 mm Hg until the temporal lobectomy and cortical disconnections are completed. The remainder of the hemispherectomy is completed with eucarbia. Diuresis with mannitol or Lasix is generally not necessary to augment cerebral relaxation, even during resection of an enlarged hemi-

megalencephalic hemisphere. Avoidance of hypothermia through the use of warmed, humidified inspired air and warming circuits for fluids and blood products is critical to preserve platelet agglutination and avoid coagulopathy. A constant dialogue with the anesthesiologist is important in minimizing delays in fluid or blood product replacement.

After induction of general anesthesia, bladder, arterial, and central venous catheters are placed, and the patient is positioned supine. The ipsilateral shoulder is elevated, and the head is rotated sufficient for midline alignment in a neutral plane. We avoid the use of skull fixation, preferring support on a padded Mayfield horseshoe headrest, which allows intraoperative rotation or manipulation of the head for maximum visibility at the midline. A path of hair is shaved along the Dandy incision, and the scalp is prepared with Betadine scrub, alcohol, and Betadine solution. Intravenous nafcillin (50 mg/kg up to l.5 g) and gentamycin (l mg/kg) of body weight are administered before the skin incision is made, and nafcillin is continued every 6 hours for 24 hours following surgery. In the patient with a penicillin allergy, we substitute vancomycin, l0 mg/kg, slowly infused over 1 hour.

In most cases of hemispherectomy, the lateral ventricle of the affected cerebral hemisphere is enlarged. We mark skin coordinates and use intraoperative ultrasound to guide ventriculostomy placement if necessary to enhance cerebral relaxation. This is rarely necessary during modified or ultrasound-guided hemispherectomy because entrance within the temporal horn of the ventricle during temporal lobectomy drains a moderate volume of spinal fluid.

Ultrasound-Guided Functional Hemispherectomy

The temporalis muscle is divided along the path of the Dandy skin incision and elevated along a subperiosteal plane. A fronto-temporal-parietal bone flap is fashioned with a high-speed craniotome to within 0.5 cm of the midline and of the floor of the middle cranial fossa. Additional exposure of the anterior inferior temporal lobe is achieved with rongeur resection of the sphenoid bone. Frontal exposure continues across the sphenoid wing along the floor of the anterior cranial fossa. The posterior margin of the bone flap extends about 4 cm behind the pinna. Epidural tack-up sutures are positioned circumferentially along the margin of the bone flap. Based on the midline, the dura is opened along the perimeter of the exposure and, to minimize shrinkage, reflected under tension within moistened Telfa strips (Fig. 24–2).

Beginning within the superior temporal gyrus, an enbloc resection of the lateral temporal gyri is completed toward the collateral sulcus. In a coronal plane, within the inferior margin of the middle temporal gyrus, ultrasound guides entrance into the temporal horn (Figs. 24–3 and 24–4); the hippocampus then is uncovered from the pes toward the atria. We resect the mesial temporal structures, including the amygdala, hippocampus and the uncus with the CUSA using a subpial technique.

After entering the choroid plexus, penetrating branches of the anterior choroidal artery may also supply the posterior limb of the internal capsule and the adjacent thalamus. Bipolar coagulation of the choroidal fissure is avoided because injury of these vessels can lead to

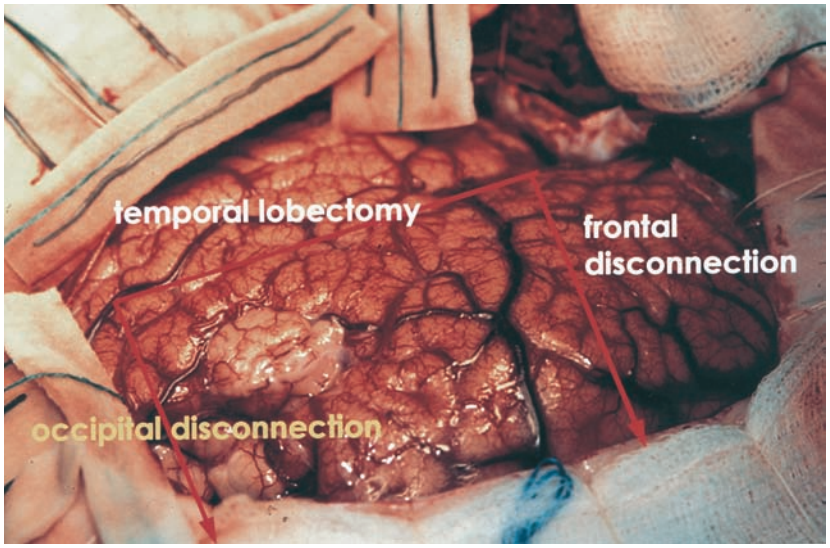


FIGURE 24–2. Intraoperative photograph of a large right craniotomy for ultrasound-guided hemispherectomy. There is parietal open schizencephaly. Arrows show the planned incisions for frontal and occipital disconnection and temporal lobectomy.

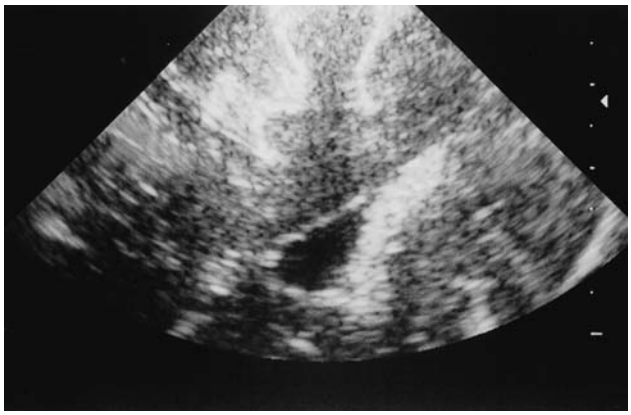


FIGURE 24–3. Intraoperative coronal ultrasound visualizing the atria of the lateral ventricle and adjacent hyperechoic choroid plexus.

flaccid hemiparesis or thalamic infarction. Every effort is made to preserve the mesial pia, protecting the cerebral peduncle, the third cranial nerve, and the perforators and larger vessels about the circle of Willis.

The occipital horn of the ventricle is identified by means of ultrasound, and the plane of occipital disconnection is selected to minimize or avoid further entrance into the ventricular system (Fig. 24–5). The cortical incision, which marked the posterior margin of the temporal lobectomy, is extended in a coronal plane along the lateral parietal-occipital cortex to the interhemispheric fissure, passing behind the occipital horn. Subpial resection performed with the ultrasonic aspirator divides the deep white matter and forceps major toward the interhemispheric fissure. Ultrasound helps gauge the depth of disconnection as the interhemispheric fissure is approached. Injuries to the vein of Galen tributaries immediately posterior to the splenium of the corpus callosum and the calcarine and distal posterior cerebral arterial supply to the occipital lobe are avoided by meticulous subpial resection. The central cerebral hemisphere is gently retracted to aid visualization at the depths of the cortical incision.

The frontal horn of the ventricle then is insonated along the lateral frontal cortex. A cortical incision in the coronal plane is made adjacent to the lateral sphenoid wing, passing in front of the rostrum of the corpus callosum, avoiding entrance within the frontal horn of the ventricle, and extends along the lateral frontal cortex toward the midline (Fig. 24–5). Through the preserved interhemispheric pia, the anterior cerebral vessels are visualized coursing about the rostrum of the corpus callosum. The internal frontal and frontopolar branches of the anterior cerebral artery are left intact. Again, ultrasound is a useful gauge of the depth of disconnection.

Topectomy of the insular and central cortices between the two disconnections begins with coagulation of distal middle cerebral artery branches as they cross the sylvian fissure. Subpial resection is accomplished by using the ultrasonic aspirator and extends across the lateral cerebral cortex toward the midline (Fig. 25–6). The topectomy is continued along the falx until the pia of the sulcus of the corpus callosum is reached. Maintenance of the subpial plane protects numerous veins draining toward the sagittal sinus and the path of the distal pericallosal arteries.

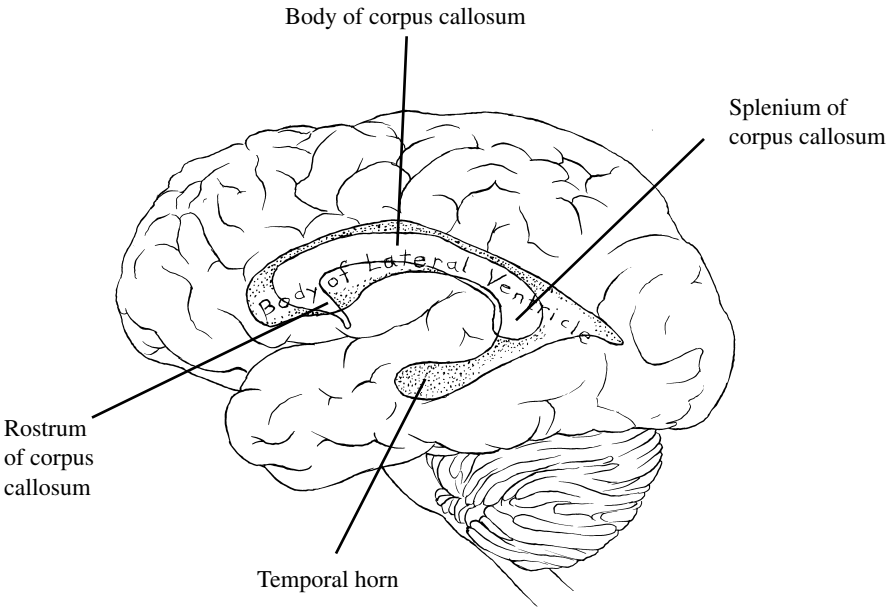


FIGURE 24–4. Relationships of the body, atria, and temporal horn of the lateral ventricle with the segments of the corpus callosum. The anterior wall of the frontal horn is adjacent to the rostrum of the corpus callosum, whereas the splenium is just posterior to the plane of the atria. The posterior extent of the occipital horn is easily visualized with intraoperative ultrasound.

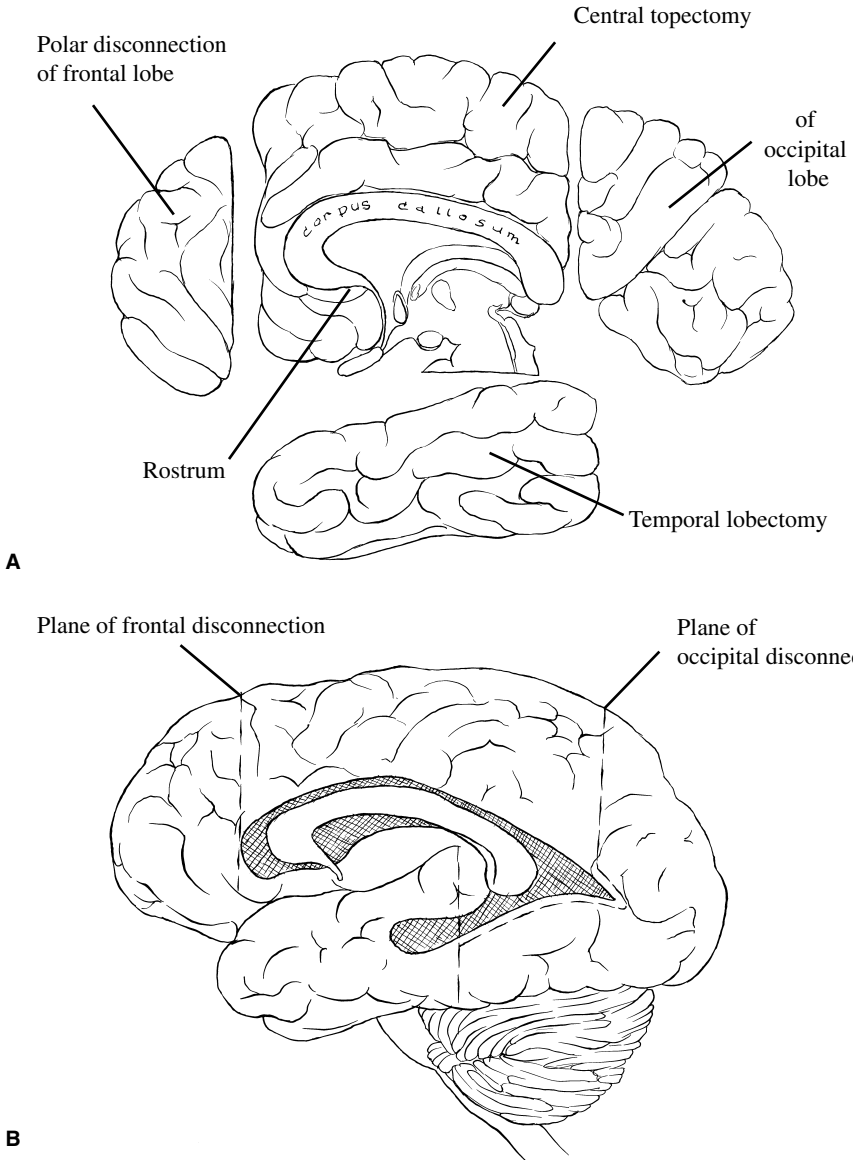


FIGURE 24–5. A, B: Ultrasound-guided modified hemispherectomy. The frontal polar cortex is disconnected from the lateral cortex to the interhemispheric fissure, crossing the plane of the rostrum of the corpus callosum in front of the frontal horn of the lateral ventricle. From the atria of the ventricle, the incision for occipital disconnection is marked to pass posterior to the splenium of the corpus callosum and the occipital horn of the lateral ventricle. Temporal lobectomy extends posterior from the temporal pole to the atria of the ventricle. Ventricle landmarks for disconnection are defined by intraoperative ultrasound.

After meticulous hemostasis is achieved with bipolar coagulation, all resection margins are sparingly lined with Surgicel. The dura is sutured primarily with continuous running no. 4-0 vicryl, augmented with pericranial grafts as needed. The remainder of the incision is closed in layers, without drains.

Modified Hemispherectomy

The MH technique developed by Rasmussen (1983) has gained wide acceptance. In contrast to the central topectomy discussed already during MH, resection of rolandic central cortex between the two polar disconnections is completed. There is wide entrance within the body of the lateral ventricle, followed by corpus callosotomy. Postoperative fever and inflammatory meningitis syndromes may be more common than they are after extraventricular resection, significantly prolonging hospitalization. Lethargy and irritability may reflect widespread ependymal contact with Surgicel and blood degradation products.

Anatomic Hemispherectomy

The patient is positioned in a similar fashion to the ultrasound-guided technique, with a horizontal neutral head position. In contrast to the Dandy incision, a midline incision is made from the inion just past the frontal hairline; a right angle incision extends from the midline toward the zygoma, completing the "barn-door" opening. An osteoplastic bone flap is fashioned with a high-speed cran-

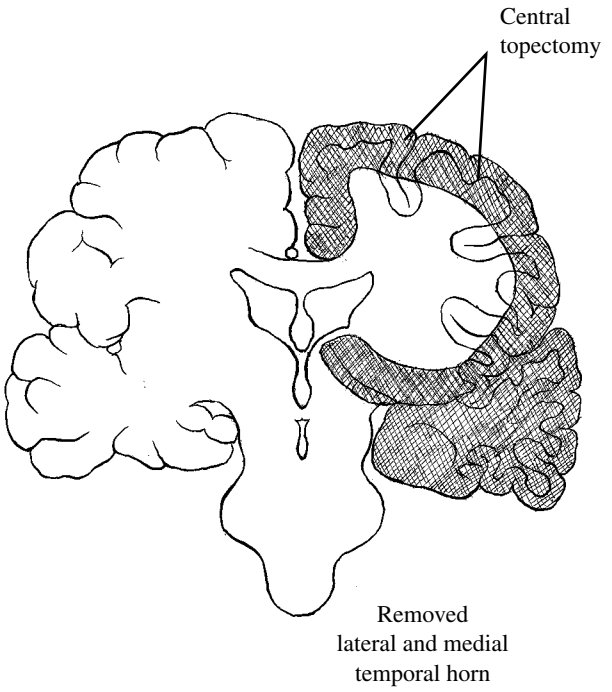


FIGURE 24–6. Coronal section of the cerebral hemisphere. Following temporal lobectomy and occipital and frontal polar disconnections, central topectomy of the cortical surface begins supra-sylvian. The aspiration of the cortex extends along the lateral cerebral surface to the midline and continues subpial along the interhemispheric fissure until the corpus callosum is reached.

iotome to provide maximum exposure to the entire cerebral hemisphere; multiple burr holes along the midline afford safety to the broad exposure of the sagittal sinus. The dura is opened along the perimeter of the bone exposure based on the midline. In cases of hemimegalencephaly, multiple cruciate dural incisions minimize retraction of the bulky temporal lobe and along the midline.

Aided by the operating microscope, the frontal operculum is gently retracted, dividing the olfactory tract and exposing the optic nerve and carotid artery. The arachnoid of the sylvian fissure is dissected similar to approaching a middle cerebral artery aneurysm, and gentle temporal lobe retraction exposes the initial segment of the vessel. After occlusion of the middle cerebral artery with an aneurysm and two titanium vascular clips just medial to its bifurcation, the vessel is divided between the distal two clips.

Splitting of the sylvian fissure allows sufficient temporal lobe retraction for identification of the junction of the posterior communicating and the posterior cerebral arteries (PCAs). The retraction then is reoriented, elevating the temporal lobe from the floor of the middle cranial fossa and tentorium edge, visualizing the lateral cerebral peduncle and the distal path of the PCA. Flow is occluded in the artery just beyond the perforators entering the midbrain, and the vessel is divided.

Following occlusion of the middle and posterior cerebral arteries, the surface of the cerebral hemisphere acquires a fine yellow pallor and becomes less engorged. The slackened hemisphere requires minimal retraction to reach the depths of the interhemispheric fissure; this process may be assisted with venticulostomy placement and spinal fluid drainage. The anterior cerebral artery is clipped and divided at the rostrum of the corpus callosum, carefully protecting the path of the contralateral vessel. Veins draining the lateral cerebral hemisphere and temporal lobe are then divided as they enter the sagittal and transverse venous sinuses.

The corpus callosum is divided with suction or the ultrasonic aspirator along its entire length from the rostrum to the splenium, entering the frontal horn, trigone, and body of the lateral ventricle. Cotton pledgets are gently inserted into the foramen of Monro to minimize blood entrance into the third and contralateral ventricle. The angle between the corpus callosum and head of the caudate nucleus is entered, establishing a horizontal plane of disconnection toward the insula and sylvian fissure. This plane is continued around the head of the caudate, forward and medial, crossing the deep white matter of the frontal lobe toward the interhemispheric fissure. Disconnection in the plane of the lateral wall of the ventricle emerges into the posterior insula. Dissection extends inferior into the temporal horn and toward the floor of the middle fossa, entering the collateral sulcus. The inferior temporal lobe is divided, passing forward to the temporal pole, allowing elevation of the occipital, frontal, parietal, and lateral temporal lobes en bloc. Great care must be exercised to avoid bleeding from the numerous veins that cross the interhemispheric fissure and pass along the inferior surface of the temporal and occipital lobes toward the tentorium.

The mesial temporal lobe structures are removed with subpial resection, carefully protecting the structures within the ambient cistern. Topectomy of the insular cortex with the ultrasonic aspirator completes the hemispherectomy. In cases of hemimegalencephaly or Sturge-Weber disease, the bulky cerebral hemisphere resists retraction and exposure within the ventricle. In these cases, piecemeal removal of the cerebral hemisphere is safer than en bloc removal.

After absolute hemostasis is achieved, the resection surfaces are sparingly lined with Surgicel. The dura is closed in a watertight fashion, and a subdural drain is brought out through one of the midline burr holes and a separate skin incision. Multiple central dural tack-up sutures are needed to reapproximate the dura upon the

osteoplastic bone flap. Spinal fluid is drained until crystalline, typically for 4 or 5 days.

POSTOPERATIVE MANAGEMENT

Patients are extubated in the operating room prior to transfer to the pediatric intensive care unit for 24 to 36 hours of monitoring. Dexamethasone is administered every 6 hours intravenously, 1 mg/kg daily, up to a maximum of 24 mg/day, and discontinued after 5 days, without taper. Oral intake is encouraged on the first postoperative day. Continuous morphine infusion, 0.03 to 0.05 mg/kg per hour, or intravenous toradol, 0.5 mg/kg every 4 to 6 hours, provides postoperative analgesia. These medications are replaced on the second postoperative day with Tylenol with codeine, 0.5 mg/kg of body weight. With the help of physical and occupational therapists, children are encouraged out of bed on the second day. Postoperative MRI is obtained in each patient before hospital discharge (Fig. 24–7). Postoperative scalp swelling is considerable and begins to subside by day 4, resolving within 14 days. Following ultrasound-guided functional hemispherectomy, hemorrhage, fever, irritability, infection or cerebrospinal fluid leaks are unusual, and most children are discharged on the fifth postoperative day. Inflammatory meningitis may occur following anatomic resection or MH and children are observed in the hospital until afebrile.

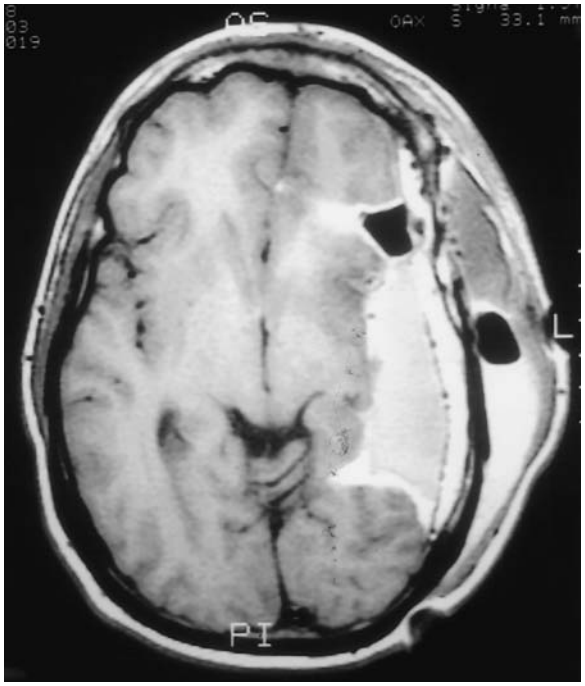


FIGURE 24–7. Postoperative axial T1 MRI following left ultrasound-guided hemispherectomy in an ll-year-old girl. Large subcutaneous fluid collections, disconnection of the frontal and occpital polar cortices, and broad central topectomy are present.

Postoperative speech or language deficits are not encountered. Immediately after surgery, there is dense contralateral hemiparesis. Weakness is greatest within the hand and wrist, in contrast to more proximal muscle groups. Most patients have recovery of antigravity shoulder, triceps, and biceps strength on discharge. Extension hand splints are warranted for nighttime restraint to minimize finger and wrist flexion contractures, and each patient requires enrollment in outpatient physical and occupational therapy.

Contralateral leg weakness is typical in the immediate postoperative period. There is rapid improvement in tone and muscle strength, and nearly all ambulatory patients can walk on hospital discharge. Assisted movements of the wrist can be anticipated within 1 month of surgery, and preoperative hand function will return within 4 to 6 months postoperatively. Patients can resume normal activities after 1 month postoperatively. We do not recommend protective helmets. Seizures in the early postoperative period are unusual. If they occur within the first 3 weeks of surgery, they do not influence the long-term success of hemispherectomy.

LONG-TERM FOLLOW-UP

Hemispherectomy is the most successful operation for control of epilepsy in children; most patient series report 80 to 90% long-term control of seizures. Anticonvulsants are continued postoperatively with careful drug-level monitoring. When patients have been seizure free for 1 year, medications are slowly withdrawn over 6 months. Long-term evaluation within a pediatric epilepsy program is critical for evaluation of seizure control, as is monitoring the neuropsychological consequences and long-term school performance following resection.

Although superficial hemosiderosis has been all but eliminated with modified surgical techniques, complications can occur anytime after resection, even into adulthood. Routine clinic visits to monitor headache and other signs and symptoms of increased intracranial pressure are important. Any child who develops headaches or breakthrough seizure activity should be promptly studied with MRI to investigate hydrocephalus or subdural or intraventricular hemorrhage. The incidence of hydrocephalus appears to increase proportionately to the extent of cortical and anatomic resection.

EDITOR'S COMMENTARY

With the increasing use of surgery for epilepsy in children, so has the use of cerebral hemispherectomy increased as more children who could benefit from the operation are being identified, evaluated, and treated. In the evaluation, unlike the authors, we have not been able to do Wada testing in children younger than 10 years of age. Similar to the authors, though, we have not encountered significant problems with language disorders postoperatively. As well, one should be careful with both PET and SPECT. Normal blood flow and metabolism does not always mean normal function in these children. Clearly, due to the plasticity inherent in the young child, the younger the child is at the time of operation, the greater the long-term benefit. With its increasing use, so has the number of modifications increased. The authors provide their methodology which, along with others, have strived to limit the intervention and resection and to optimize the disconnection. It has been this editor's experience (PDA) as well that limited surgical ventricular intrusion tends to lessen the hospital stay and the occurrences of complications.

PEARLS

In these authors' experience:

- Children are sensitive to manipulation in the body of the lateral ventricles, and postoperative fever, lethargy, and inflammatory ventriculitis syndromes may occur.
- In children with intractable seizures, multiplanar MRI is the study of choice for demonstrating malformation of the affected cerebral hemisphere.
- Postoperative speech and language disorders are not encountered, although dense contralateral hemiparesis, which is usually greatest in the hand and wrist, is found in all patients.

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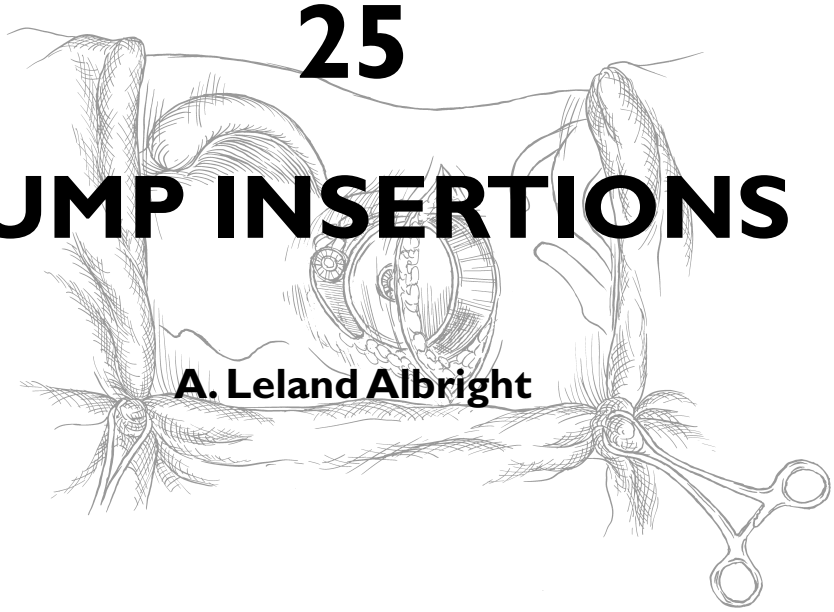
PUMP INSERTIONS



A. Leland Albright

In the past decade, pump insertion for intrathecal infusion of medication has become a relatively common pediatric neurosurgical operation. Pumps are similar to hydrocephalus shunts: Both are implanted devices with the potential to improve quality of life substantially, but both also become infected and malfunction and have catheter problems. As with shunts, meticulous attention to detail during pump insertion will reduce the frequency of complications.

INDICATIONS

Although some pumps are inserted into pediatric patients to treat severe pain, the vast majority are inserted to deliver intrathecal baclofen (ITB) to treat spasticity or dystonia. Briefly, the general indications for ITB in treating spasticity include improving function, facilitating care, and retarding or preventing the development of contractures. Although ITB often improves such abilities as communication and upper extremity function, it is not possible to predict with certainty the ways in which function will improve after spasticity is reduced. Most children or young adults in whom improved function is a goal have spastic quadriparesis, which causes stiff leg movements, limits upper-extremity range of motion, and increases the effort required to use either arms or legs. Patients in whom pumps are inserted to facilitate care are usually seriously retarded and require almost total care. Their spasticity interferes with leg abduction needed for perineal cleaning, knee extension needed to put on pants, and ankle dorsiflexion needed to put on ankle–foot orthoses or shoes. Pumps also are occasionally inserted to prevent or retard contractures, usually in children 4 to 10 years of age whose spasticity is already causing contractures, even if decreased spasticity is unlikely to improve function or facilitate care.

Pumps are considered for patients with moderate or severe spasticity, with at least an average muscle tone of 3 on the Ashworth scale. Many younger children will have been treated with oral medications and intramuscular botulinum toxin before pumps are recommended. A trial of oral medications is not a requirement before a pump is considered, however. Some patients have such severe spasticity when first seen, and are alert and cognitively functional, that the likelihood of them obtaining good relief from oral medications is minimal.

Most patients with dystonia are given oral medications before proceeding to a pump; but, as with spasticity, those with severe dystonia are unlikely to respond well to oral medications, and a trial of ITB may be the appropriate first treatment.

TEST (SCREENING) TRIALS

Pumps are rarely inserted without first confirming that the patient responds to ITB. Nowadays, screening trials for spasticity usually are done with a single dose of ITB. Children usually are not sedated for the lumbar puncture because many sedatives reduce their spasticity for several hours and confound interpretation of the test dose. If sedation is needed, intravenous propofol is ideal, providing excellent sedation for less than 1 hour. We apply EMLA cream to the skin of the lumbar region one hour before the lumbar puncture. When the lumbar puncture is done, it is important to measure the opening pressure. Many children with spasticity and cerebral palsy were born prematurely and sustained intraventricular hemorrhages that left them with ventriculomegaly, but they were not

shunted. If the opening pressure is 21 cm or greater, I obtain a head scan to evaluate the hydrocephalus. If their pressure is not measured and is higher than normal, their likelihood of developing a cerebrospinal fluid (CSF) leak along the intrathecal catheter is increased.

We often continue oral antispasticity medications during the screening trial because they are relatively ineffective. The standard test dose of baclofen is 50 μg; 80 to 90% of patients will respond to it. A clinically significant response is considered to be a 1-point decrease in the average muscle tone of the lower extremities. Tone is graded in the adductors, quadriceps, hamstrings, and plantarflexors before the injection and 2, 4, 6, and 8 hours afterward. Tone is maximally reduced 4 hours after injection. The test dose typically abolishes spasticity in the legs, so it is difficult to stand or walk until the effects begin to diminish. It is not possible to predict during the screening trial what function would be like during continuous infusion. Side effects of headache and vomiting are common during the screening and are related more to the lumbar puncture than to the baclofen. Patients who respond positively to the test dose can be implanted with a pump the following day or soon thereafter. If the response to the 50-μg dose is insignificant, doses of 75 or 100 μg are given. More than 95% of persons with cerebral spasticity will respond to some dose of baclofen.

Screening dystonia patients with baclofen differs considerably from screening those with spasticity. If patients have mixed spasticity and dystonia, we try a bolus dose first, but most patients with severe generalized dystonia do not respond to a bolus dose, although they often respond to continuous infusions. We therefore test them with an intrathecal catheter and infuse baclofen with an external microinfusion pump. The catheter is inserted in the operating room under sterile conditions. I first insert a 20-gauge spinal needle in the midline at L2-3 into the epidural space and then insert the 14 to 15 gauge Tuohy needle approximately 5 mm lateral to the midline, insert it into the thecal sac, and advance an intrathecal catheter through it to C5-7, confirming the position fluoroscopically. The guidewire in the catheter is withdrawn, and CSF flow is confirmed. The Tuohy needle is withdrawn, and fibrin glue is injected through the spinal needle into the epidural space to reduce the risk of a CSF leak along the catheter. Then the catheter is tunneled several centimeters laterally, where it exits the skin through a puncture. A pursestring suture is applied at the catheter exit site and a simple stitch at the midline. The catheter is occluded until the patient is on the hospital ward and then is connected to a microinfusion pump. The pump infuses baclofen, initially at a rate of 200 μg per day, and the rate is increased by 50 μg every 8 hours until dystonia in the upper and lower extremities is reduced by 25% or greater on the Barry-Albright dystonia scale, unacceptable side effects such as lethargy occur, or the infusion rate reaches 900 μg per day. Approximately 85% of patients will respond to the infusion, usually at rates of 300 to 600 μg per day. When the infusion trial is completed, if the patient has responded, the catheter exit site is sterilely prepared, the catheter is tied off at the point where it exits the skin, cut off 1 to 2 mm distal to the tie, tucked under the skin, and a stitch is inserted, all so that the catheter may remain in place and be used as the intrathecal catheter when a pump is inserted. If the patient does not respond to the infusion, the catheter is withdrawn and a stitch is inserted. Typically, we send patients home for 2 weeks or longer after the infusion screening trial to ensure that there is no evidence of infection before implanting the pump.

PUMP IMPLANTATION

Pumps are inserted with the patient under general anesthesia. We use prophylactic antibiotics during the operation and for 48 hours afterward and request that the operative region be washed with an antibacterial soap the night before and morning of operation. Patients usually are placed in the left lateral decubitus position with the right side up; this position is more comfortable for right-handed surgeons and avoids the gastrostomy tubes often present on the left side. Pumps are inserted in the left side if severe scoliosis toward the right narrows the space between the inferior rib margin and the iliac crest.

The abdomen, flank, and back are prepared; I prep the skin with Betadine scrub, alcohol, and Duraprep, a mixture of alcohol and iodine that reduces colonization for several hours postoperatively. A transverse incision is made at the abdomen, beginning about 1 cm below the inferior costal margin laterally and extending medially toward the midline for about 3.5 inches (Fig. 25–1). Using the coagulating cautery, subcutaneous tissues are opened down to and through the fascia over the lateral aspect of the rectus muscle, the intervening space, and the external oblique muscle (Fig. 25–2). Fascia is dissected off the muscle cephalad for about 1.5 cm, but mostly inferiorly, for some 8 to 9 cm, so that the pump can be inserted under the fascia without tension.

There is no age limitation to pump insertion; the limitation is size: I have inserted a 10-mL Synchromed pump into a 20-pound child, but I insert the 18-mL pump whenever it can be inserted without undue tension. It can be inserted in nearly all children weighing over 40 pounds without difficulty. In patients weighing 30 to 40 pounds, the smaller pump is needed perhaps half the time and in

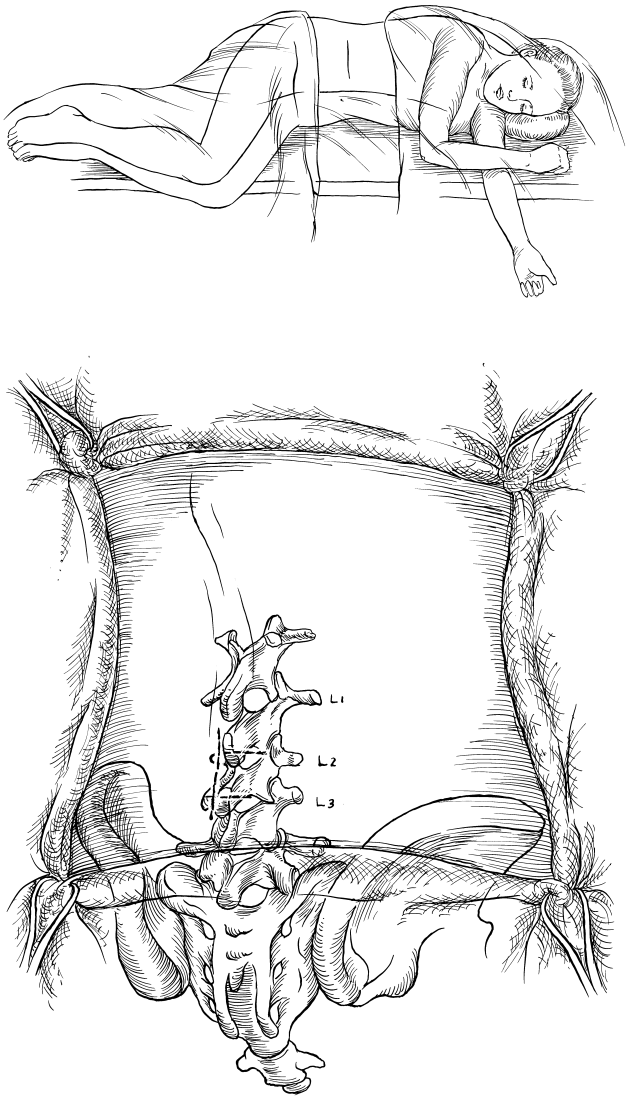


FIGURE 25–1. Transverse abdominal **(A)** and lumbar **(B)** incision sites. The anterior incision is 1 cm below the lateral costal margin.

nearly all patients weighing less than 30 pounds. Pumps are secured to the underlying muscle by either eyelets on the pump margins or by Dacron pouches. The eyelets are preferable: they do not tear as the pouches sometimes do, and they do not cause fibrosis, which makes later pump removal difficult. Pumps must be securely sutured to the surrounding tissue. If they are not, they can flip, so that the refill portal is posterior and unaccessible. If a Synchromed pump with a side port is used, I insert the pump so that the side port is caudal; in this way, the catheter attached to the side port is directed superiorly and laterally toward the back (Fig. 25–3).

At the back, I make a transverse incision about 3 cm long, beginning 2 to 3 mm to the left of midline and ex-

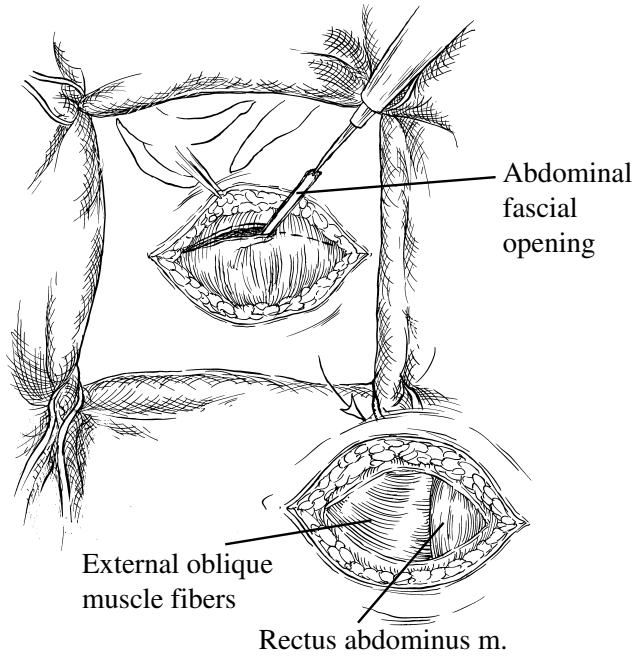


FIGURE 25–2. Abdominal fascia opening, with external oblique laterally and rectus abdominis medially.

FIGURE 25–3. The 8709 catheter has been attached to the pump, which is being inserted into the subfascial pocket. Suture eyelets are visible on the top of the pump.

tending to the right. Subcutaneous tissues and fascia are opened with the cautery, and a Weitlaner retractor is inserted. The underlying lumbodorsal fascia then is opened in the same transverse direction so that the catheter will be subfascial. Unless an intrathecal catheter is in place already, I insert a spinal needle in the midline into the epidural space for later fibrin glue instillation and then insert the Tuohy needle about 5 mm to the right of the midline, into the thecal sac (Fig. 25–4). The intrathecal catheter, generally the Medtronic 8709 catheter, is inserted with a guidewire and advanced cephalad. If the patient's spasticity affects the arms and legs approximately equally, as is usually the case, I advance the catheter to T4-6 in an attempt to get better arm relaxation. If the spasticity is predominantly in the legs, the tip is positioned at T8-10. Catheters inserted for dystonia usually are positioned at C5-7. Catheter position is confirmed. Fibrin glue or a blood patch can be injected through the needle in the epidural space at this time.

The intrathecal catheter can be connected to the pump either anteriorly or posteriorly. If the anterior site is used, the intrathecal catheter is tunneled subcutaneously to the abdomen and connected anteriorly to the stem on the side of the pump. The advantage of the anterior approach is the use of a single catheter around the back; the disadvantage is that the intrathecal catheter is of small diameter and may be more likely to perforate or break. Alternatively, the larger proximal 8703W catheter can be attached to the pump anteriorly, tunneled posteriorly, and connected there. The advantage of the two-catheter technique is that the larger anterior catheter never kinks or develops any other problems; the disadvantage is the potential for disconnection from the straight connector in the back. I now prefer the single catheter (8709) technique with the anterior connection.

Excess portions of both catheters, whose lengths are measured before they are used, are removed, and the two catheter ends are connected with a straight connector. Catheters are tightly tied to the straight connector with large sutures, whose ends then can be tied together to decrease the risk of disconnection. The intrathecal catheter is secured to paravertebral musculature with a Silastic sleeve (Fig. 25–5). The lengths of the removed catheters are measured so that the length of implanted catheters is known and their volumes

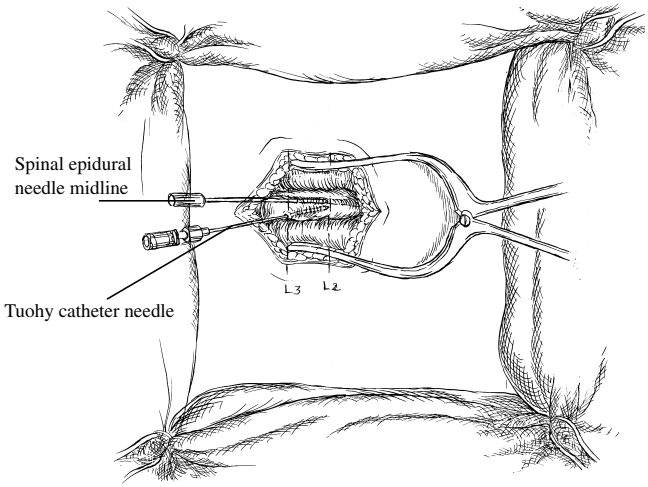


FIGURE 25–4. The lateral lumbar incision has been made, the lumbar fascia opened, a spinal needle inserted in the midline into the epidural space and a Tuohy needle paravertebrally into the thecal sac. The intrathecal catheter is inserted through the Tuohy needle.

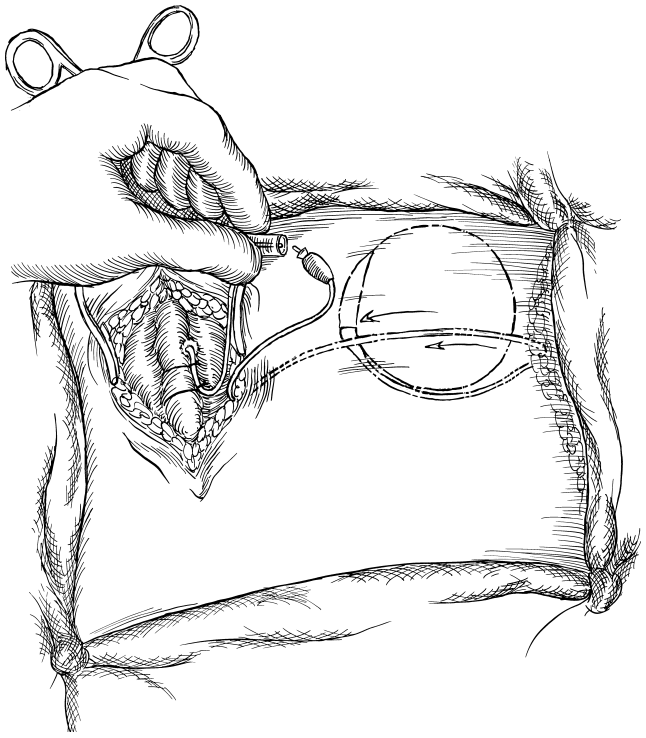
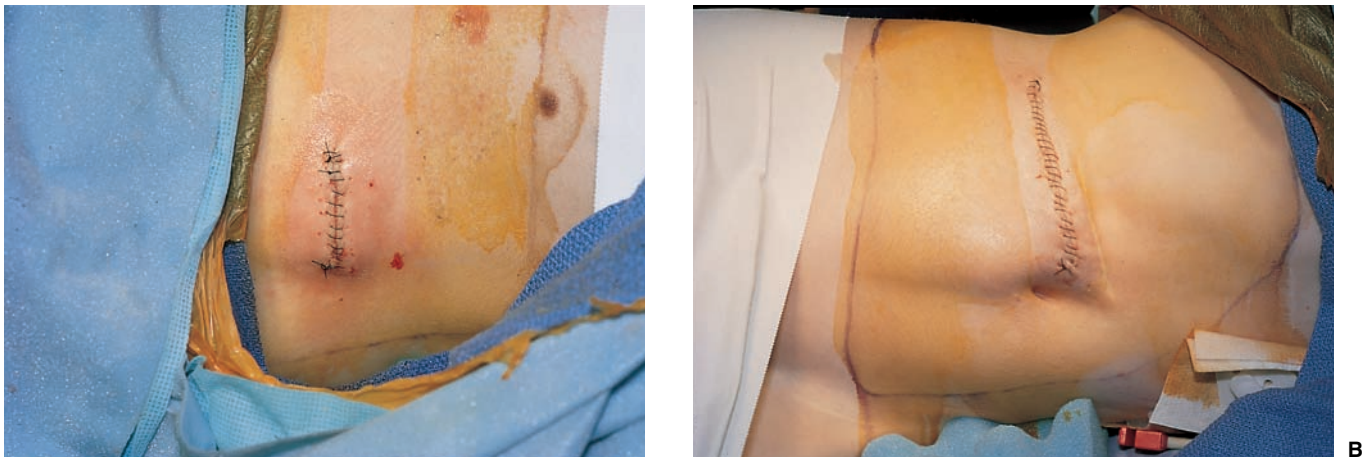


FIGURE 25–5. The two catheters are joined with a straight connector and secured with ties. The intrathecal catheter is secured to paravertebral muscle with a Silastic anchoring device.



A B

FIGURE 25–6. Final appearance after wound closures. **A:** Anterior. **B:** Posterior.

may be calculated. I irrigate both wounds with antibiotic-containing solution and then close the fascia over the catheter and over the pump with 2-0 Vicryl. The skin is closed with external running nylon suture (Fig. 25–6), bupivacaine is injected along the incision, collodion is applied to the incision to decrease CSF leaks, and dressings are applied.

POSTOPERATIVE CARE

Pumps are programmed soon after operation, usually either in the operating room or in the recovery room, or soon thereafter. An initial bolus of baclofen is given over 30 minutes, with a volume sufficient to fill the space from the pump reservoir to the end of the side port (about 0.25 mL), the catheters (about 0.15 mL), and an initial bolus to decrease tone (often 50 μg). When treating spasticity pumps are initially filled with a 500 μg/mL concentration but when treating dystonia 2000 μg/mL is used. The initial constant daily rate in treating spasticity is twice the amount to which the patient responded in the screening trial (100 μg per day if the patient responded to a 50-μg bolus). The initial rate in treating dystonia is usually 200 μg per day, although it may be higher if the patient did not respond in the screening infusion until receiving more than 500 μg per day.

Patients are kept "flat" for 48 hours and then are allowed out of bed, wearing a circumferential binder that compresses both wounds. Baclofen doses are increased daily, usually by 10 to 20%/day, during the 3- to 6-day hospitalization until the spasticity or dystonia are perceptibly improved. Sutures are removed at 14 days, later than usual because of concern that CSF may leak through the incision if sutures are removed earlier. We recommend that postoperative physical therapy be deferred for 1 month because of concern that earlier movement may cause a CSF leak or catheter problem.

EDITOR'S COMMENTARY

Children with movement disorders are ideally treated in multidisciplinary clinics since it is exceedingly difficult for individual practitioners to be familiar with the various movement disorders and the innovations in their treatment. In the past decade, the implantation of pumps for continuous intrathecal infusion of medication, particularly baclofen, has become a relatively common pediatric neurosurgical operation. During that decade, virtually everything about the operation has changed: the indications (from spasticity to either spasticity or dystonia); the pump battery (from lasting 4–5 years to lasting 7–8 years); the implantation site (from subcutaneuos to subfascial); the catheter (from the original two-piece system to a single catheter); and the catheter position (from T10 to C5-T6). Although the technology has changed substantially, the need for careful patient selection and meticulous operative technique are unchanged. Intrathecal baclofen has not helped children with athetosis or chorea. Additional studies are needed to evaluate other intrathecal medications, both to treat the common movement disorders and to determine more fundamental issues such as the sites of action of baclofen and the feasibility of intraventricular infusions. Intrathecal infusions of medications will probably undergo as many changes in the next decade as they have in the last one, so that the operation becomes even more common and the indications for treatment even broader.

PEARLS

In this author's experience:

- Higher catheter position seems to give better upper-extremity effects.
- Risks of infection can be decreased by preoperative skin washing, use of Duraprep, intraoperative antibiotics, antibiotic irrigation and instillation, but mainly by meticulous sterile technique and avoidance of CSF leaks.
- Risks of CSF leaks can be decreased by measuring the lumbar pressure to evaluate the presence of occult hydrocephalus when the test injection is performed, by an epidural blood patch or fibrin glue injection, by keeping the patients flat for 48 hours postoperatively, and by wearing a binder that compresses both incisions for 2 to 3 weeks when the patients are out of bed.
- Risks of pump "flipping" can be decreased by securing the pump to the adjacent fascia and muscle with multiple sutures and by avoiding CSF leaks.

SUGGESTED READINGS

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