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CHAPTER 12.1

Pediatric Neurosurgery

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Craniofacial Surgery

Surgical Considerations

Description: Craniofacial surgery is a broad term that refers to both cranial and/or facial reconstructive procedures for cranial dysostosis or craniofacial dysmorphism. Cranial dysostosis is the congenital maldevelopment of the cranial base and/or vault, 2° premature fusion of cranial sutures. More commonly referred to as **craniosynostosis,** the surgical correction of this disorder involves removal of the affected suture(s) and reconstruction of the cranial, orbital, or facial bones. The most common form of craniosynostosis—scaphocephaly—is caused by the fusion of the sagittal suture, which leads to a long and narrow calvarium. Other forms of craniosynostosis, in order of decreasing frequency, are coronal synostosis (brachycephaly), metopic synostosis (trigonocephaly), and lambdoidal synostosis. Deformational occipital plagiocephaly refers to flattening of the occiput 2° preferential sleep position and the resultant deformation of the skull. This condition is not a form of craniosynostosis and, despite the potential for significant flattening of the head, reconstructive surgery is not indicated. Crouzon and Apert syndromes are inherited craniofacial disorders associated with craniosynostosis and facial/orbital dysmorphism. The facial deformities common to Crouzon and Apert are shallow and misplaced orbits, exophthalmos, and midface hypoplasia. In each form of craniosynostosis, sporadic or predisposed, the abnormality is present at birth, but may not become recognizable until the rapid phase of brain growth, occurring in the 1st year of life, begins to accentuate the limitations on skull shape produced by the premature suture closure. In simple terms, the growth of the underlying brain drives the expansion of the skull, and closure of a suture produces reduced skull growth in the opposite direction.

Early recognition and correction of craniosynostosis results in the best cosmetic and neurologic outcome because, with release of the fused suture, the growing brain helps correct the abnormal cranial shape. Most procedures are scheduled during the 1st 6 mo of life; thus, the issue of blood volume and replacement becomes a critical factor for surgical and anesthetic consideration. The main principles of surgical treatment of craniosynostosis involve removal of the abnormal suture through a craniectomy or craniotomy, followed by reconstruction of the calvarium and/or orbit to overcome the cranial deformity and optimize the chance for normal cranial development. The surgery most often is done in conjunction with a pediatric neurosurgeon and a plastic surgeon. Patient positioning varies, depending on the approach to the craniectomy, and is generally prone for sagittal and lambdoidal synostosis, and supine for coronal and metopic synostosis. Another surgical principle important for synostosis surgery is to minimize intraop blood loss. The surgical team should make every effort to reduce blood loss during the procedure by infiltrating the scalp with 1:400,000 epinephrine, using point electrocautery, preserving the pericranium, and waxing the bone edges. The most common skin incision is a bicoronal opening that allows for access to the entire calvarium. The extent of the bone removal and reconstruction varies, depending on the type and number of sutures involved. Surgical correction of patients with Crouzon or Apert syndromes is often staged with correction of the cranial component, followed by a later procedure for the face, as described by Tessier and colleagues. Invariably, blood loss occurs from the scalp and bone, and the surgeon must remain mindful of the volume contained within the surgical field and readily communicate to the anesthesiologist when bleeding is felt to be either continuous or excessive. Injury to the underlying dural venous sinuses is rare, but the potential for catastrophic blood loss is great. Recent advances in endoscopy have lead to the development of minimally invasive techniques for craniosynostosis in some centers, and reports suggest that use of the endoscope reduces blood loss. Recombinant erythropoietin administered preop also has been studied in an attempt to reduce the need for intraop transfusion associated with repair of craniosynostosis. Subgaleal and epidural drains may be placed at the close of the procedure, and the patient is monitored closely in the PICU for postop bleeding that often requires additional transfusions in the 1st 24 h after surgery.

Usual preop diagnosis: Craniosynostosis (sagittal, coronal, metopic, lambdoidal); craniofacial dysmorphism; Apert syndrome; Crouzon syndrome

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Summary of Procedures

Position	Supine or prone (less common); or table 180°
Incision	Bicoronal, biparietal, Meisterschnitt, midsagittal
Special instrumentation	Midas Rex craniotome; absorbable plates and screws, Colorado needle
Unique considerations	↑ ICP and/or hydrocephalus may coexist. ↑ ICP is usually seen with multiple-suture synostosis; it also occurs in single-suture craniosynostosis, but is rare (< 5–10%). Hydrocephalus typically is seen in monogenic conditions (e.g., Apert, Crouzon). Most neurosurgeons shunt the hydrocephalus and treat the ↑ ICP prior to craniosynostosis surgery.
Antibiotics	Vancomycin (13–15 mg/kg) or ceftriaxone (25–50 mg/kg; avoid Ca^{++}-containing solutions) for cranial surgery. Vancomycin or cloxacillin and cefotaxime (25–30 mg/kg) for craniofacial surgery involving nasal sinuses.
Closing considerations	Watch for ↑ blood loss from the scalp. To prevent excessive blood loss, reapproximate only one portion of the scalp at a time.
EBL	Highly variable; must be minimized. Amount depends on preoperative condition, on the number of sutures involved, age of the patient, and magnitude of the repair. Operative injury to the superior sagittal sinus may be catastrophic if hemostasis cannot be achieved or volume loss is excessive.
Postop care	PICU. Postop Hct/Hb levels required. Blood transfusion often necessary in infants (< 10 kg).
Mortality	< 1–2%
Morbidity	Meningitis CSF rhinorrhea ↑ ICP (2° skull reshaping) Venous thrombosis Neurological injury: Rare
Pain score	1–3

Patient Population Characteristics

Age range	Newborn–young adult
Male:Female	1.2:1
Incidence	1/2000/yr
Etiology	Sporadic; heritable (monogenic and chromosomal syndromes); environmentally induced (amniotic bands, iatrogenic)
Associated conditions	Congenital defects (limbs, heart, brain, kidneys); hydrocephalus; encephalocele (sincipital/basal); fibrous dysplasia; craniometaphyseal dysplasia; holoprosencephaly

Anesthetic Considerations

Preoperative

Craniofacial surgery encompasses a wide variety of procedures, with the two most common being linear craniotomy for craniosynostosis (or premature closure of the cranial sutures) and reconstructive for congenital deformities of the forehead, orbit

ridges, and nose. Craniosynostosis usually manifests itself in the 1st yr of life; surgery for other congenital deformities of the face and skull usually is performed from ages 1–8 yr.

Respiratory	As a result of midfacial deformities, some children may present difficult intubations. Their airways should be carefully evaluated preoperatively and preparations made for a difficult intubation.
Neurological	Presenting Sx in infants with craniosynostosis include: progressive head deformation, progressively increasing irritability, crying, failure to eat, and failure to grow in head circumference. These Sx may be due in part to ↑ ICP. On physical examination, one or more of the cranial sutures are fused. Infants with other types of craniofacial deformity usually have no Sx related to their abnormalities.
Laboratory	Tests as indicated from H&P. Preop Hct, Hgb, platelets, INR
Premedication	For children, midazolam 0.5 mg/kg po (see p. D-1) generally provides satisfactory preop sedation after 30 min. For young children (< 5 yr) who refuse po meds, instillation of midazolam 0.3 mg/kg intranasally provides rapid amnesia, sedation, and easy separation from the parents.

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Intraoperative

Anesthetic technique: GETA using Pediatric circle. Forced air warmer. Warm room 75–80°F. Warm all fluids.

Induction	Standard pediatric induction (see p. D-1). Orotracheal tubes are preferred over nasotracheal tubes because the surgery may involve reflection of the scalp down over the eyes and nose, in which case a nasal tube would be in the way. Verify ETT placement (see p. D-2). Tape the tube firmly in place at one side of the mouth using benzoin adherent. When craniofacial surgery is performed, the surgeon usually places plastic corneal shields in the eyes or sutures the lids shut to protect the cornea from injury and may also suture the ETT in place.
Maintenance	Standard maintenance (see p. D-2). Muscle relaxation is usually provided. Maintain a near-normal BP unless mild hypotension is requested to limit blood loss. Maintain normal temp by keeping the OR warm (78°F) and using warming lights and blankets as needed. Ventilation is controlled to $PetCO_2$ = 35–40 mmHg with a mechanical ventilator from the start of anesthesia until the surgical wound is closed.
Emergence	No specific considerations. The ETT is removed at the conclusion of the anesthetic. Patient usually goes to the PICU for observation.
Blood and fluid requirements	Large blood loss iv: 18–20 ga × 1–2 NS/LR @ 4 mL/kg/h 5% albumin Fluid warmer Administer crystalloid via a continuous infusion pump or Volutrol. Blood is often necessary. It is advisable to begin transfusion early in course of surgery, to avoid getting behind. In smaller children, it is better to administer warmed blood by syringe, in 10 mL increments. Serial Hct determinations are useful. Blood salvage techniques may be used when large blood loss is anticipated.
Monitoring	Standard monitors (see p. D-1). ± Foley catheter ± Doppler ± Arterial line ± CVP line If operation is anticipated to last several hours, a Foley catheter should be inserted. If patient is semi-sitting, a Doppler ultrasound probe may be placed on the chest to monitor for air embolism. Invasive monitoring is often appropriate. ↑ K^+ and ↓

		$Ca^{++}$are most common following transfusions with blood or FFP.
Positioning	OR table usually rotated 180°. and pad pressure points. eyes.	Ensure access to airway by placing a Mayo stand over the chest area.
Complications	Major blood loss	
	VAE	Treatment of venous air embolism–Notify surgeon who will locate entry point and flood with saline, D/C N_2O, attempt to aspirate air from CVP, lower head of bed, compress jugular veins to decrease the rate of air entry, CPR as needed. PEEP is not recommended and may be deleterious. VAE more likely if dural sinus entered.

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Postoperative Considerations

Complications	Bleeding Extubation Hypovolemia	Major complications from these operations are uncommon.
Pain management	Parenteral opioids (see p. E-1). Avoid oversedation.	Fentanyl 1–2 mcg/kg q 60 min or morphine 0.05–0.1 mg/kg q 2 h.
Tests	Hct/Hb Coagulation parameters	Hct/Hb levels are necessary to determine adequacy of blood replacement.

Suggested Readings

1. Dahmani S, et al: Perioperative blood salvage during surgical correction of craniosynostosis in infants. *Br J Anesth* 2000; 85 (4):550–5.

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3. Fearon JA: Reducing allogenic blood transfusions during pediatric cranial vault surgical procedures: a prospective analysis of blood recycling. *Plast Reconstr Surg* 2004; 113(4):1126–30.

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11. Tessier P: Total facial osteotomy. Crouzon's syndrome, Apert's syndrome: oxycephaly, scaphocephaly, turricephaly. *Ann Chir Plast* 1967; 12(4):273–86.

Closure of Myelomeningocele

Surgical Considerations

Description: Myelomeningocele is a neural tube defect characterized by failure of the spinal cord to fuse posteriorly during primary neurulation. This results in an open neural placode joined to the incomplete epithelial defect, usually located in the thoracolumbar spine, and rarely in the cervical spine. Associated CNS conditions are hydrocephalus and Chiari II hindbrain malformation, both of which usually contribute more to long-term morbidity than the spinal cord defect itself. The presence of the myelomeningocele may be detected before birth by high-resolution ultrasound and/or elevated maternal serum alpha fetoprotein, as well as fetal MRI scans. The incidence of neural tube defects is declining in the United States due to maternal dietary folate supplementation and prenatal Dx and selective termination.

The fundamental goals of surgery are preservation of neural tissue, reconstitution of a normal intrathecal environment, and complete skin closure to prevent a spinal fluid leak. Despite a very thin parchment of dystrophic epithelium attached to the placode, most myelomeningoceles leak spinal fluid from the time of birth. Because of the risk of ventriculitis associated with the exposed subarachnoid space, closure of the myelomeningocele is recommended within 72 h after birth. Infants with neural tube defects have a higher incidence of other congenital anomalies, including hydronephrosis, malrotation of the gut, VSD or ASD, and craniofacial disorders. The neonate should be screened for these potential abnormalities before undergoing surgery and, in general, this can be accomplished within 24 h *(Print pagebreak 1158)* after birth. During the procedure, the child is in the prone position. The defect is dissected so that the various anatomic layers can be separated. The edges of the placode (spinal cord) are mobilized from the adjacent epithelium and often imbricated to form a closed tube. The laterally displaced dura is dissected from the fascia and closed over the spinal cord, thus reconstituting the elements of the spine, except for the lamina defect that is not reconstructed. An attempt is made to mobilize the lumbosacral fascia as a separate layer and the subcutaneous and skin layers comprise the final layer. In cases of large defects, local skin or myocutaneous flaps may be necessary to cover the spinal defect adequately. Progressive hydrocephalus usually presents within days to weeks after closure of the myelomeningocele, but 15% of patients will present at birth with significant hydrocephalus that requires early insertion of a VP shunt. Finally, in rare circumstances, prominent vertebral angulation, or kyphosis, at the defect could necessitate vertebrectomies to re-establish normal spinal alignment, usually at an older age.

Variant procedure or approaches: The efficacy of **intrauterine myelomeningocele repair** is currently being explored through a randomized multicenter trial, and the results may alter future approaches in favor of intrauterine closure if the incidence of hydrocephalus is reduced in these patients.

Usual preop diagnosis: Myelomeningocele; meningocele; myelodysplasia; spina bifida

Summary of Procedures

Position	Prone
Incision	Surrounding the defect, preserving skin that can be utilized in the closure. Use of plain lidocaine (0.25%) local anesthetic decreases blood loss.
Special instrumentation	Loupes or operating microscope (optional)
Unique considerations	Concomitant hydrocephalus, lower brain stem dysfunction. Need for blood replacement rare in straightforward cases. Latex precautions should be used in all cases. (p. G-1)
Antibiotics	Ceftriaxone (50 mg/kg iv), or if Ca^{++}-containing solutions are required use vancomycin (13–15 mg/kg iv, slowly),

Surgical time	1.5–3 h
Closing considerations	Skin closure may be complex and require rotation of flaps or aid of plastic surgeon.
EBL	Negligible–25 mL (in most cases)
Postop care	Neonatal nursery. Postop, child often nursed on stomach or side. Head size and head ultrasound are used to monitor for development of hydrocephalus, which may require shunting at a later date.
Mortality	Approaching zero
Morbidity	Meningitis/ventriculitis Hind brain dysfunction Wound infection CSF leak Apnea, vocal cord paralysis
Pain score	3–5

Patient Population Characteristics

Age range	Newborn (diagnosed at birth)
Male:Female	1:1
Incidence	1/1000 live births
Etiology	Congenital
Associated conditions	Hydrocephalus; lower extremity weakness; bowel and/or bladder dysfunction (neurogenic bladder) and/or hydronephrosis; scoliosis; Chiari II malformations; congenital cardiac anomalies ****NB**: Latex allergy is increased in this population. Latex precautions should be used beginning with the 1st operation. See Appendix G.

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Anesthetic Considerations

Preoperative

Myelomeningoceles are congenital abnormalities of the spinal cord that result in a saccular protrusion near the base of the spine. The sac, containing neural elements and CSF, can vary in size from very small to a volume that occupies the whole lower spinal region. The Dx may be suspected from maternal alpha-fetoprotein screening, fetal ultrasound, or prenatal MRI and is confirmed at birth. It is generally believed that immediate removal of the sac and covering of the defect with skin is desirable to preserve neurological function and avoid infections. These newborns, therefore, usually are brought to surgery within 24–48 h after birth.

Cardiovascular	May have associated congenital anomalies. **Test:** ECHO
Neurological	Although difficult to assess at this age, newborns may have motor and/or sensory deficits in the lower extremities, neurogenic bladder, and lower cranial nerve dysfunction. Most have an Arnold-Chiari malformation, which requires a ventriculoperitoneal shunt within days of the spine repair.
Renal	May have associated congenital anomalies. **Tests:** Renal ultrasound
Laboratory	Routine preop studies
Premedication	None necessary

Other	**NB** : Latex Precautions for all patients (beginning from birth). See p. G-1.

Intraoperative

Anesthetic technique: GETA using Pediatric circle; forced air warmer; Warm room to 75–80°F.

Induction	Before induction, the patient is placed in the supine position, and the back defect is protected by a sterile donut or rolls to prevent pressure on or rupture of defect. Patients usually will come to the OR with an iv in place, allowing for a standard iv induction. (See p. D-1). If not, a standard inhalational induction followed by establishment of iv access is indicated. See p. D-1. Sevoflurane is preferred because of its low blood-gas partition coefficient and absence of airway irritability, which allows a smooth, rapid induction. Oral ETT intubation usually follows muscle relaxation with rocuronium (0.6–1 mg/kg) or vecuronium (0.1 mg/kg). Atropine (0.05–0.1 mg) may be administered before intubation to reduce secretions and to prevent reflex bradycardia. Verify ETT placement and tape tube securely in place at one side of the mouth with benzoin adherent to facilitate prone positioning.
Maintenance	Sevoflurane 2–3% or desflurane 7% or less, with N_2O or air/O_2mixture to maintain arterial O_2sat at 95–97%. Depending on duration of operation, additional doses of rocuronium (0.3 mg/kg) or vecuronium (0.1 mg/kg) may be needed. Maintain a near-normal BP. Maintain normal temp by keeping OR warm (78°F) and using warming lights and blankets as needed. Ventilation is controlled to maintain $PetCO_2$= 35–40 mmHg with a mechanical ventilator or manually, from the start of anesthesia until surgical wound is closed.
Emergence	These patients almost always remain intubated for the 1st 24 h. The newborn is nursed in the prone or lateral positions for the 1st few days postop.
Blood and fluid requirements	iv: 22–24 ga × 1 D10 @ 2–4 mL/kg/h (newborn) D10 ¼ NS @ 4 mL/kg/h (> 24 h) Warm fluids. Administer crystalloid, usually D10 NS, via a continuous infusion pump, etc. Blood is rarely, if ever, necessary.
Monitoring	Standard monitors (see p. D-1).
Positioning	Prone with shoulders and hips on bolsters to elevate abdomen off operating table. and pad pressure points. Head turned to the side, which results in the eyes. ETT being furthest from the bed. tube placement by listening for bilateral breath sounds after repositioning.

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Postoperative

Complications	CSF leak Wound healing complications Infection Hydrocephalus Renal Failure
Pain management	Parenteral opioids (see p. E-3).

Suggested Readings

1. Cochrane D, Irwin B, Chambers K: Clinical outcomes that fetal surgery for myelomeningocele needs to achieve. *Eur J Pediatr Surg* 2001; 22(Suppl 1):S18–20.

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Surgical Correction of Spinal Dysraphism

Surgical Considerations

Description: Occult spinal dysraphism covers a spectrum of spinal anomalies generally related to defects in secondary neurulation, in contrast to a myelomeningocele, which occurs as a result of defective primary neurulation. These forms of congenital spinal defects are covered by intact skin and share the common pathophysiology of spinal cord tethering. Occult spinal dysraphism includes tight filum terminale, intramedullary lipoma, lipomyelomeningocele, split cord malformations (diastematomyelia), dermal sinus tracts, meningocele manque, neuroenteric cyst, and myelocystocele. Each of these lesions can result in a tethering, or stretching, of the spinal cord as the vertebral axis elongates during normal growth. The fixed spinal cord is stretched by the growing spine, resulting in neurological dysfunction, most likely as a result of reduced spinal cord blood flow. Excision of the lesion and release of the tethering elements is recommended to prevent either the onset or worsening of cord dysfunction.

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The surgical principles for correction of occult spinal dysraphism involve excision of the intradural lesion and release of the tissue element that is tethering the spinal cord, while preserving the normal neural structures. In cases of tight filum terminale, this is easily accomplished with little threat to the spinal cord or nerve root function, whereas excision of lipomyelomeningoceles may be extremely challenging procedures. Any tethering lesion needs to be carefully separated from the spinal cord and adjacent nerve roots without disrupting neurological function. Intraop neurophysiological monitoring may be a useful adjunct to monitor cord and nerve root function during the dissection and release of the tethering lesion. Following the intradural procedure, closure of the dura and myofascial layers are critical in prevention of postop spinal fluid leaks; and dural grafts may be necessary in some cases.

Usual preop diagnosis: Tethered spinal cord; fat filum terminale; lipomyelomeningocele; lipoma of the filum; diastematomyelia; spinal dysraphism; dermal sinus tract; caudal agenesis

Summary of Procedures

Position	Prone
Incision	Posterior midline centered over abnormality
Special instrumentation	Operating microscope; laser
Unique considerations	Blood replacement with loss of significant amount of blood in the infant (rare). Minimal latex exposure
Antibiotics	Ceftriaxone (50 mg/kg iv; do not use with Ca^{++}-containing solutions); or vancomycin (13–15 mg/kg iv slowly)
Surgical time	1.5–5 h (longer for diastematomyelia and lipomyelomeningocele)
Closing considerations	Surgeon often wants to test integrity of dural closure with Valsalva maneuver: sustained (10–20 sec) inspiratory pressure at 20–40 cmH_2O.

EBL	5–100 mL
Postop care	Patient often kept flat postop to protect dural closure.
Mortality	Approaching zero
Morbidity	Infection Neurological deficit Aseptic meningitis CSF leak Urinary dysfunction; Check preop renal function, renal U/S
Pain score	3–5
Other	Latex precautions See p. G-1.

Patient Population Characteristics

Age range	Newborn-adults
Male:Female	1:1
Incidence	Uncommon
Etiology	Congenital
Associated conditions	Ankle/foot deformity (talipes); neurologic impairment; neurogenic bowel/bladder; scoliosis; vertebral abnormalities; VACTERL/VATER association (vertebral, anal, cardiac, tracheoesophageal, renal, and limb anomalies); cutaneous anomaly over spine; syringomyelia; caudal agenesis; urinary anomalies; check abdominal ultrasound, renal function prior to OR

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Anesthetic Considerations

Preoperative

A variety of spinal abnormalities fall under the category of spinal dysraphism, the most common being a tethered cord or a lipoma of the spinal cord. In the case of the tethered cord, there may be a Hx of myelomeningocele repair at birth. Most patients range from 3–16 yr. of age.

Neurological	Presenting Sx in older children are usually pain in the lower back radiating into the legs and/or progressively worsening motor or sensory deficits in the anal region or involving the lower extremities (document carefully).
Musculoskeletal	Lower extremity foot deformity as well as sensory or motor deficits may be present and should be carefully documented.
Renal	Renal function may be impaired in patients with Hx of recurrent UTIs.
Laboratory	**Tests:** UA; BUN; Cr; others as indicated from H&P. Other tests as indicated from H&P.
Premedication	For children, midazolam 0.5 mg/kg po (see p. D-1) generally provides satisfactory preop sedation after 30 min. For young children (< 5 yr) who refuse po meds, instillation of midazolam 0.3 mg/kg intranasally provides rapid amnesia, sedation, and easy separation from parents.
Latex allergy	Latex precautions should be used in all patients even at 1st operation. Meningomyelocele patients may have developed a latex allergy (see p. G-1).

Intraoperative

Anesthetic technique: GETA

Induction	Standard induction (see p. D-1) with sevoflurane, N_2O, and O_2 This is followed by establishment of iv access, then ETT intubation with the use of a muscle relaxant (e.g., rocuronium 1 mg/kg or vecuronium 0.15 mg/kg). Sevoflurane is preferred because of its rapid, smooth induction. Tape the tube firmly in place at one side of the mouth, using benzoin adherent to facilitate prone positioning. Latex precautions.	
Maintenance	Standard maintenance (see p. D-2). Upon completion of placement of the dural graft, and before closure of the wound, the surgeon will want to check the integrity of the graft to eliminate any CSF leaks. The surgeon will ask that positive pressure be applied to the airway to at least 20 cmH_2O for 10–20 sec. If graft leaks are detected, they will be repaired and the test repeated.	
Emergence	The ETT is removed at the conclusion of the anesthetic. The patient may be nursed flat in the prone or lateral position for the 1st few days postop to lessen the chance of a CSF leak developing.	
Blood and fluid requirements	iv: 18–20 ga × 1 NS/LR @ 4–6 mL/kg/h	In children, administer fluids via a volumetric infusion set. Blood is rarely necessary.
Monitoring	Standard monitors (see p. D-1). ± Foley catheter ± UO	If the operation is anticipated to last several hours, or in the case of neurogenic bladder, a Foley catheter should be inserted.
Positioning	and pad pressure points. eyes.	Prone with the shoulders and hips on bolsters to elevate the abdomen off the operating table. Head turned to the side, resulting in the ETT being furthest from bed. tube placement by listening for bilateral breath sounds after repositioning.
Complications	Severe bradycardia Latex allergy	Manipulation of the spinal cord may produce ↓↓ HR reflexly. See p. G-1 for latex allergy considerations.

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Postoperative

Complications	Possible neurological deficits Infection CSF leak Urinary retention Incontinence	Major complications from this operation are uncommon, but include new neurological deficits from irritation of the spinal cord during surgery, localized infection, and CSF leak from the wound site.
Pain management	Parental opioids or PCA (see p. E-3–E-4).	

Suggested Readings

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Craniotomy for Vein of Galen Malformation

Surgical Considerations

Description: A vein of Galen malformation is a large, AV fistula between arteries, mainly of the posterior cerebral circulation, and a massively enlarged vein of Galen (deep venous drainage of the brain). Patients can present with high-output CHF (infants), progressive macrocephaly from hydrocephalus (infants, children, adults), or, rarely, IC hemorrhage in adult patients. Most cases present at birth or during the infant years, and immediate care is often directed toward stabilizing cardiac function related to CHF. Patients undergoing surgery with high output CHF carry the highest risk of periop mortality. Many patients will have some degree of accompanying hydrocephalus, but decisions regarding the placement of ventricular shunts are best postponed until the malformation has been treated, as hydrocephalus may improve with reduction in intracranial venous pressure. High output CHF is addressed by treating the fistula. Embolization via the arterial or venous system minimizes resistance or reduces flow in the fistula, improving cardiac function.

Treatment is directed at staged occlusion of the arterial feeders to the AV fistula and eventually thrombosis of the fistula itself from the venous side. Open microsurgical techniques, endovascular methods, or a combination of both may be used to reduce flow through the malformation. With reduction in the aneurysmal flow, CO will decrease, SVR will improve, and mixed venous PO_2 will decrease. Because of the high mortality associated with treatment by surgery alone and the advances in endovascular techniques, vein of Galen malformations currently are managed without direct primary surgical approaches. Surgery, as an option, is more likely to occur in the setting of staged or attempted embolizations. Subtemporal, midline occipital, or bilateral occipital craniotomies can be used to isolate and occlude arterial feeders to the malformation. Stereotactic radiosurgery has been used successfully as an adjunct to embolization for surgery.

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Variant procedure or approaches: Alternative approaches may access the torcular (confluence) of venous sinuses through a burr hole to allow direct retrograde placement of thrombogenic coils in the AV fistula.

Usual preop diagnosis: Vein of Galen aneurysm; IC hemorrhage; hydrocephalus; progressive neurologic deficits; current management focuses on embolization in the cath lab prior to surgical repair.

Summary of Procedures

Position	Lateral decubitus, Concorde (modified prone), or semisitting; for burr hole, lateral
Incision	Temporal or occipital; for burr hole, occipital
Special instrumentation	Operating microscope, microscopic instruments; intraop angiography; for burr hole, endovascular catheters and equipment; intraop SSEP monitoring
Unique considerations	Careful attention to blood loss in infants Central venous access and A-line mandatory
Antibiotics	Vancomycin (1 g iv slowly q 12 h for adults; 10–15 mg/kg iv slowly q 6 h for children); cefotaxime (1 g iv q 6 h for adults; 40 mg/kg iv q 6 h for children)
Surgical time	3–5 h
Closing considerations	Meticulous hemostasis; avoid ↓ BP or HTN (MAP 80–90 adults; 70–80, children).
EBL	< 250 mL
	Monitor for ↑ ICP (use of extraventricular drain) as a result of

Postop care	venous HTN 2° rapid occlusion of AV fistula. ICU × 1–3 d.
Mortality	Approaches 100% if high output CHF not resolved
Morbidity	Deep venous infarct: 5–10% Hydrocephalus Stroke Subdural hygroma Infection: Rare
Pain score	3–4

Patient Population Characteristics

Age range	Birth–3 yr (typically)
Male:Female	1:1
Incidence	Rare
Etiology	Congenital
Associated conditions	Other intracranial vascular malformations; high-output CHF

Anesthetic Considerations

Preoperative

Vein of Galen vascular malformations are rare congenital abnormalities representing < 1% of all malformations. They are usually diagnosed in infants because of high output CHF and may cause an abnormal increase in head size due to the aneurysmal dilation and obstruction of the dural sinus. The abnormal vasculature constitutes a high-flow shunt, much like an AVM. If left untreated, the morbidity and mortality are high; however, treatment with radiologic embolization is superior to surgical excision and also is associated with lower morbidity and mortality rates.

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Cardiovascular	Because these lesions constitute high-flow shunts through the brain, the infants are prone to develop high output CHF, which is fatal in more than 40% of patients. Newborns present with h/o CHF.
Neurological	Infants usually present with an abnormal head size, Sz disorder, or bizarre neurological signs, such as high-pitched crying, posturing, failure to eat or thrive, etc.
Laboratory	CT; MRI; cerebral angiography. The infant may need sedation and anesthetic management to obtain adequate diagnostic studies. Check renal function.
Premedication	For children, midazolam 0.5 mg/kg po (see p. D-1) generally provides satisfactory preop sedation after 30 min. For young children (< 5 yr) who refuse po meds, instillation of midazolam 0.3 mg/kg intranasally provides rapid amnesia, sedation, and easy separation from the parents.

Intraoperative

Anesthetic technique: GETA, with the goals being the same as those for IC vascular malformations.

Induction	Whenever possible, an iv induction is preferred. STP 2–3 mg/kg or propofol 1–2 mg/kg, fentanyl 2–3 mcg/kg, and rocuronium 0.6–1 mg/kg or vecuronium 0.1 mg/kg are satisfactory induction agents.

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Maintenance	Isoflurane ≥ 1%, sevoflurane 2–3% or desflurane ≥ 7%, with N_2O or air 60–70% to keep O_2sat = 95–98%. Depending on duration of operation, additional doses or NMBs may be administered as needed to maintain a single-twitch response to nerve stimulation. Maintain a near-normal BP. Ventilation is controlled to maintain $PetCO_2$= 35–40 mmHg with a mechanical ventilator or manually, from the start of anesthesia until surgical wound is closed. Mild hypothermia (32–35°C) may be requested by the surgeon. Normovolemia should be maintained.	
Emergence	Plan to leave ETT in place for at least 24 h postop; infant should receive controlled ventilation and sedation during that interval.	
Blood and fluid requirements	IV: 20–22 ga × 1–2 Central venous access = subclavian (no IJ, as may compromise cerebral circulation)	Replace blood as it is lost (cc for cc). Limit crystalloid fluid therapy to no more than 10 mL/kg above UO.
Control of brain volume	Same as for AVMs (see p. 21).	
Monitoring	Same as for AVMs (see p. 21).	
Control of BP	Goal = normal range for age Neonate: 55–70/40 (HR = 180) 1 yr: 70–100/60 (HR = 140)	BP should be kept in the normal range for the infant with close monitoring from an arterial catheter. If HR becomes excessive, esmolol infusion is useful.
Positioning	Same as for AVMs.	
Complications	Coagulopathy	If large volumes of blood are needed, a coagulopathy may ensue. Monitoring of coagulation status during surgery is mandatory.
	Hypothermia	After the aneurysm is surgically corrected, hypothermia needs to be continued for 24–72 h.

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Postoperative

Complications	Neurological deficits IC hemorrhage Heart failure
Pain management	Fentanyl 0.5–1 mcg/kg. $MSO_4$0.05–0.1 mg /kg.
Tests	Same as for AVMs (see p. 22). Patients will need a CT or MRI under anesthesia postop in the AM

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Ventriculoscopy and Third Ventriculostomy

Surgical Considerations

Description: Ventriculoscopy is the technique of intraop visualization of the lateral, 3rd, and, occasionally, 4th ventricles using fiber optic endoscopes inserted through standard cranial burr holes. The ventriculoscope permits direct inspection and limited navigation within the ventricle for both diagnostic and therapeutic purposes, and often is most commonly applied in the setting of hydrocephalus. In addition to CT scans, preop MRI exam of the brain is obtained to better depict the anatomy of the ventricular system, which is distorted frequently by congenital lesions. The enlarged ventricles produced by the hydrocephalus contribute to the safety and feasibility of most endoscopic approaches, enabling a variety of procedures. The endoscope can be used to fenestrate multicompartmental periventricular or arachnoid cysts, position ventricular catheters during shunt insertion, biopsy or, in some cases, resect intraventricular tumors. Neuroendoscopy generally does not help in the initial cannulation of the ventricle (a common misconception).

Endoscopic ventriculoscopy may be performed through either frontal or parietal-occipital approaches, with the patient typically supine with the neck slightly flexed. Standard small incisions similar to shunt insertions are used. A twist drill or burr hole is created and the ventricle cannulated by insertion of the shunt catheter or an introducer with a peel-away sheath (for larger endoscopes). Endoscopes vary in size from 1.1 mm, for use inside a standard shunt catheter, to larger endoscopes (12–14 Fr), equipped with working channels, for more complex intraventricular procedures. After the ventricle is "tapped" through conventional methods, the endoscope can be inserted and the ventricular anatomy identified. After the intraventricular anatomic landmarks—such as the choroid plexus—are recognized, the scope can be navigated to the site of interest. Smaller endoscopes are used to position the catheter in the optimal ventricular location during shunt placement or revision. Larger endoscopes equipped with channels for instrumentation are used for biopsy, tumor resection, cyst aspiration, or fenestration procedures. Most scope systems have a separate channel for fluid irrigation if minor bleeding or debris obscure visibility.

Intraop complications associated with neuroendoscopic procedures include: minor or major intraventricular hemorrhage; air entrapment (pneumocephalus); injury to paraventricular structures (basal ganglia, hypothalamus, brain *(Print pagebreak 1167)* stem); cardiorespiratory depression; and delayed arousal from anesthesia. Intraventricular hemorrhage is caused by direct or indirect injury to ependymal and extraependymal blood vessels. Fortunately, most bleeding encountered is minor, but may be sufficient to interfere with visualization and illumination of the ventricle. Cardiorespiratory depression and cardiac arrhythmia are due, at least in part, to phenomena attributed to ↑ ICP from excessive irrigation without equal extracranial egress, rate of fluid instillation, and/or nonisothermic irrigant irritating the hypothalamic nuclei adjacent to the 3rd ventricle.

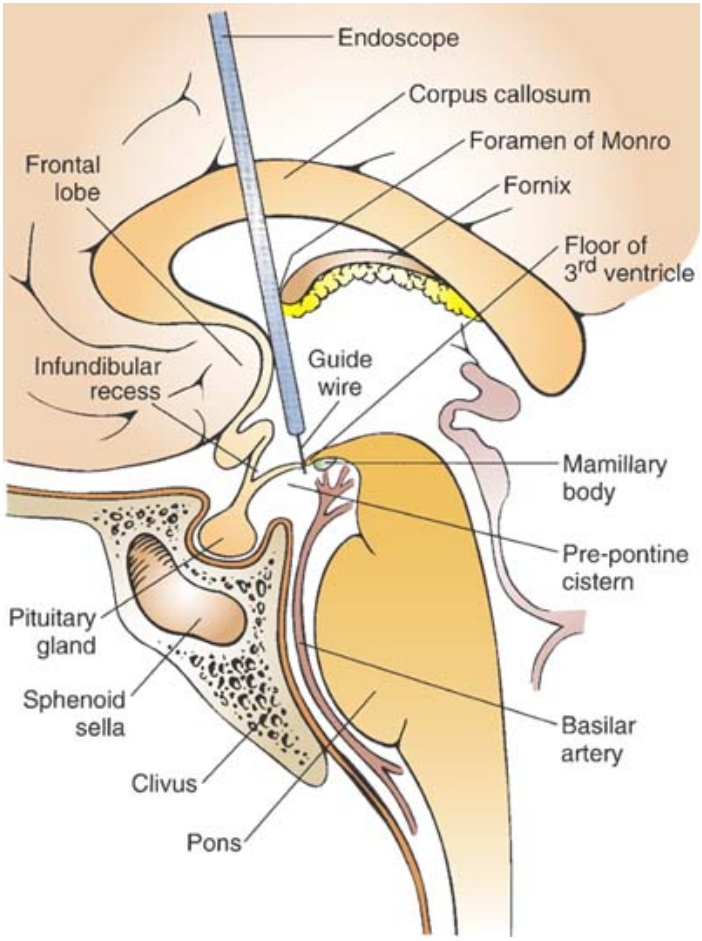

Figure 12.1-1. 1. Endoscopic 3rd ventriculostomy. The figure depicts fenestration of the floor of the 3rd ventricle by a blunt probe inserted through the endoscope.

Third ventriculostomy is one of the more common endoscopic procedures and refers to fenestration of the floor of the 3rd ventricle to create a communication between the 3rd ventricle and the basilar cistern (Fig. 12.1-1). The technique is most commonly applied to patients with obstructive, or noncommunicating, hydrocephalus, although broader indications are being explored. This form of hydrocephalus results from impaired CSF flow through the Sylvian aqueduct or 4th ventricle outlets. For patients with noncommunicating hydrocephalus, successful 3rd ventriculostomy allows CSF communication between the 3rd ventricle and the interpeduncular subarachnoid space, thereby alleviating the hydrocephalus and avoiding shunt placement. The fenestration is conducted first by direct visualization of the floor of the 3rd ventricle and then by perforation of the ependymal and arachnoid tissue between the mammillary bodies and the infundibular recess. The perforation can be dilated by inflation of a balloon catheter passed through the fenestration. Because of the proximity to the brain stem—in addition to the complications encountered with ventriculoscopy—3rd ventriculostomy carries the additional risk of mesencephalic injury, hypothalamic dysregulation, cranial nerve injury, and hemorrhage from the basilar artery and adjacent perforating vessels. Minor bleeding can be controlled easily by steady irrigation. In the event of excessive bleeding, conversion to an open craniotomy is unlikely to improve control of the hemorrhage. Patients also may be prepped for shunt insertion in the event the ventriculostomy is aborted because of unfavorable 3rd ventricular anatomy. A temporary extraventricular drain (EVD) may be left in place following some procedures to control CSF drainage postop and/or to allow assessment of ICP.

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Usual preop diagnosis: Obstructive hydrocephalus; shunt malfunction; loculated multicompartmental hydrocephalus; intraventricular mass; arachnoid cyst; retained catheter

Summary of Procedures

Position	Supine with head neutral; table 90° or 180°
Incision	Small frontal or parietal-occipital

Special instrumentation	Endoscope (1.1–6 mm diameter); video system
Unique considerations	Hydrocephalus, ↑ ICP; suspicion of latex allergy in patients with spina bifida (see p. G-1). Body temperature irrigation is used.
Antibiotics	Ceftriaxone (50 mg/kg q 24 h iv × 2; do not combine with Ca^{++} containing solutions); or vancomycin (13–15 mg/kg iv)
Closing considerations	Monitor for delayed intraventricular or subdural hemorrhage, ventricular collapse, acute hydrocephalus, CSF leak.
Postop care	Floor or ICU setting with cardiac and O_2sat monitoring × 24 h. Measurement of serum sodium. Fluid intake/output recording. CT to r/o hemorrhage and to evaluate ventricular volume may be necessary. EVD used in cases of 3rd ventriculostomy for draining CSF and testing ICP.
Mortality	0–7% (most series report 0%)
Morbidity	Intraventricular hemorrhage Acute hydrocephalus CSF leak/pneumocephalus Meningitis/ventriculitis Subdural effusions or hematoma Cranial nerve palsy Hemiparesis Diabetes insipidus SIADH Temperature dysregulation
Pain score	2–4

Patient Population Characteristics

Age range	Newborn–adult
Male:Female	1:1
Incidence	3/1000 live births (congenital only)
Etiology	Congenital and acquired
Associated conditions	Hydrocephalus; spinal dysraphism; Chiari malformation; posthemorrhagic hydrocephalus; arachnoid cyst

Anesthetic Considerations

Preoperative

Ventricular shunts are inserted to ameliorate hydrocephalus or cyst formations, which are either congenital or acquired.

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Cardiovascular	↑ ICP →↑ BP & ↓↓ HR (Cushing's response) **Tests:** As indicated from H&P.
Neurological	The most common presenting Sx is HA. If hydrocephalus is severe, Sx of ↑ ICP (>15 mmHg) (e.g., N/V, drowsiness, papilledema, Sz, and focal neurological defects) develop.
Laboratory	Tests as indicated by H&P, CT, and/or MRI.
Premedication	Usually not required; should be avoided in patients with ↑ ICP.

Intraoperative

Anesthetic technique: GETA with pediatric circle; forced air warmer.

Induction	If ↑ ICP, iv induction with STP (3–6 mg/kg) or propofol (1.5–3.5 mg/kg) is preferred, because of their ability to decrease cerebral blood volume and, hence, ICP. ETT intubation is accomplished with the use of a nondepolarizing NMB (e.g., vecuronium [0.1 mg/kg] or rocuronium [0.7–1 mg/kg]).	
Maintenance	Isoflurane <1% or sevoflurane < 2% inspired with N_2O/O_2 mixture to maintain O_2 sat 99%. Depending on duration of operation, additional doses of vecuronium (0.1 mg/kg) or rocuronium (0.2 mg/kg) may be needed. Maintain normal T in children by keeping OR warm (78°F) and using warming lights as needed. Ventilation is controlled mechanically (ventilator) or manually from the start of anesthesia until the surgical wound is closed. TV and frequency are adjusted so that $PetCO_2$ = 35–40 mmHg. Hyperventilation and hypocarbia are undesirable because they make cannulation of the ventricle(s) more difficult for the surgeon. Maintain normotension.	
Emergence	ETT is removed at the conclusion of the anesthetic. Prophylactic antiemetic (eg. ondansetron 0.1 mg/kg, max = 4 mg) should be given 30 min before extubation.	
Blood and fluid requirements	iv: 18–20 ga × 1 (22 ga for children) NS/LR @ 4–6 mL/kg/h	Administer crystalloid (via measured volume system in a child). Blood is rarely, if ever, necessary.
Management of ICP	Low normal $PaO_2 \geq 100$ Minimize fluids Isoflurane < 1% or Sevoflurane < 2% Hyperventilate to $PaCO_2$ = 25–30 mmHg ($PetCO_2$ = 20–25 mmHg)	Patients with malfunctioning VP shunts may be on the steep portion of the intracranial compliance curve such that any increase in intracranial volume may ↑↑ ICP. Transient increases in ICP are tolerated, provided that they are promptly terminated. Sustained increases in ICP > 25–30 mmHg are associated with severe neurological injury and poor outcome.
Monitoring	Standard monitors (see p. D-1).	
Positioning	Table turned 180° and pad pressure points. eyes.	Supine with a bolster under the shoulder on the operative side. The head, chest, and abdomen are prepped, so all anesthesia equipment and lines must be at the sides of the patient.
Complications	Infection Valve malfunction	Major complications from this operation are uncommon, but include infection at the valve site or in the tubing, and malfunction of the valve, either draining too little or too much CSF.

Postoperative

Pain management	Children < 2 yr: Tylenol suppositories (up to 40 mg/kg intraop, then 10–15 mg/kg q 4 h). Fentanyl 0.5–1 mcg/kg MSO_4 0.05–0.1 mg/kg	New shunts are painful as a result of subcutaneous tunneling.

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Pediatric Brain Arteriovenous Malformations (AVM)

Surgical Considerations

Description: An arteriovenous malformation is the most common cause of non-traumatic intracranial hemorrhage in childhood. It consists of direct connections between arteries and veins without an interposed capillary network and leads to a tangle of malformed vessels and channels (the nidus), carrying blood at arterial pressure. They may occur in any location of the brain, brain stem, or spinal cord; however, cerebral malformations are the most common. The arterial vessels deliver blood to the veins at systolic blood pressure, and that may cause bleeding. The lack of an interposed capillary network prevents local oxygen delivery leading to focal ischemia (steal) and seizures. This steal may result in a progressive neurologic deficit. In addition, venous congestion with arterial pressure blood may produce neurologic symptoms due to venous congestion and reduced perfusion to surrounding brain. AVMs can vary in size from lesions having only one feeding vessel to very complex lesions with multiple feeders and draining veins encompassing an entire hemisphere. It is estimated that 80–90% involve the cerebral hemisphere. Most are conical in shape with the apex near the ventricle. Small AVMs are more likely to exhibit spontaneous hemorrhage, resulting in headache, neurologic deficit, coma, and rarely death. Aneurysms within the nidus of the AVM (nidal aneurysm) or involving the feeding arteries are the most likely source of bleeding. The risk of hemorrhage is reported to be 2% per year. It is reported that after a hemorrhage the risk may be as high as 6% during the subsequent year. AVMs residing in the brainstem or cerebellum are more likely to result in permanent neurologic deficit or death if hemorrhage develops compared to those in the hemispheres. Recurrent headache or new onset seizure may herald the symptomatic AVM. However, the sudden onset of headache often followed by neurologic deficit should lead to an imaging study (CT or MRI) to demonstrate the lesion. If no lesion is identified, a lumbar puncture to confirm the presence of subarachnoid blood is indicated. Although CT angiography (CTA) or magnetic resonance angiography (MRA) is very sensitive in the diagnosis of AVM, four-vessel cerebral angiography is still the gold standard to confirm the presence and define the anatomy of these lesions. If the initial angiogram following a hemorrhage is nondiagnostic, the study should be repeated in 1–3 months, after resolution of hematoma and local compression of the vessels in the nidus. The ideal treatment of AVMs is complete surgical excision which removes the risk of hemorrhage, decreases or eliminates seizures and may alleviate or resolve neurologic symptoms of “cerebral steal” or venous congestion. A treatment decision for AVMs depends on size, location, and complexity, presence of associated aneurysms, as well as patient age, neurologic status, surgical expertise, and requires consideration of family and patient preference. The surgeon's decision to treat or observe a child with an AVM is complex with the ultimate goal of preservation of neurologic function. Some lesions situated in eloquent or deep regions of the brain are best observed or considered for interventional and/or stereotactic radiosurgery rather than craniotomy and resection. Angiography and embolization is most often performed under general anesthesia in the radiology suite. However, operating rooms have been constructed that can be used for complex angiographic procedures, open neurosurgical operations, magnetic-resonance imaging and image guided surgery. During angiography and embolization, physiologic monitoring (sensory and motor evoked potentials) is usually carried out. Careful blood pressure control is mandatory, and mild hypothermia (33° C) may be desirable. The duration of *(Print pagebreak 1171)* anesthesia is usually measured in hours, and post embolization the children are cared for in the PICU. Frequently, multiple episodes of embolization over weeks or even months are required for large or complex AVMs before the lesion is amenable to surgery or

radiosurgery. However, embolization as the sole treatment for AVMs is not recommended; rather it should be used as an adjunct to surgery or to decrease size in order to successfully treat with radiosurgery. Surgery is most often performed under GA. In older teenagers with lesions in eloquent cortex, especially those involving speech, awake craniotomy and direct brain mapping may be used. Obtaining adequate brain relaxation may be difficult using awake techniques. For most children GA is recommended. The use of image guidance helps in localization of the lesion allowing for more precise placement of the craniotomy flap and better localization of subcortical lesions. Intubation should be completed without the patient straining or becoming hypertensive so as to decrease the risk of hemorrhage. Mild hyperventilation is begun to decrease and maintain the $PaCO_2$ to 30–35 mmHg. A lumbar subarachnoid drain may be used to allow for brain relaxation and access to deep lesions. At least two peripheral intravenous lines, an arterial line, pulse oximeter, Foley catheter, and a CVP line are placed. The use of mild (33° C) or moderate hypothermia requiring central cooling (e.g. Innercool-type device) may be undertaken. Minimal isotonic fluids should be administered and the serum Na^+ is kept in the 140–150 mEq range. If needed, hypertonic (3%) saline is administered. Mannitol and/or Lasix may be given to aid in brain relaxation if a lumbar drain is not placed. The use of dexamethasone (1-2 mg/kg/d; max 16 mg/d) is controversial, but is used more often than not. Anticonvulsants are not routinely administered unless there is a history of seizures. Routine prophylactic antibiotics (eg. Ceftriaxone) are given 30–60 min before the skin incision. If intraop angiography is to be used, the catheter sheath is placed in the femoral artery and covered beneath a sterile dressing. The patient is carefully positioned on the table and a radiolucent headholder is applied to the skull. Evoked potential monitoring electrodes are placed and baseline potentials are established (special anesthetic techniques are required and motor paralytic agents are minimized or avoided). Image guidance is established and the patient is draped and padded to avoid pressure points, allow access to the femoral arterial sheath, and allow a clear line of sight to the image-guidance system. The skin incision is infiltrated with 1/4–1/2% bupivacaine with 1/200,000 or 1/400,000 epinephrine to minimize skin bleeding. After the elevation of the bone flap, either CSF drainage via the lumbar drain or hypertonic agents are used to allow for minimal brain retraction and/or manipulation during the approach and resection of the AVM. If planned temporary occlusion of a major feeding vessel is required, barbiturates may be administered. Surgery may be prolonged and intraop angiography performed on multiple occasions during the procedure. MAP is maintained at the preop resting levels or within a set range determined by the surgical team using SNP and other vasoactive agents. In order to prevent perfusion pressure breakthrough and cerebral swelling, hypertension is avoided. If the procedure is prolonged, the AVM very complex, or a subtotal or planned staged removal is the surgical outcome, then the patient should remain intubated and ventilated postoperatively. Strict control of MAP is mandatory, and the range is based on the patient's age, AVM complexity, location, use of and degree of hypothermia, and preop blood pressure. Intraop MRI scans are being studied at many institutions. MRI requires very careful planning to allow the patient's head to be moved or rotated into the MRI donut during the operative procedure. MRI eliminates the ability to perform evoked potential monitoring. The use of this advanced technology, albeit attractive, produces a complex and hostile operating room environment and increases the anesthesia workload. Its benefit for AVM surgery is as yet to be determined.

Summary of Procedures: See Chapter 1, Craniotomy for intracranial aneurysms, p. 5.

Anesthetic Considerations: See Chapter 1, Craniotomy for intracranial aneurysms, p. 6.

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