Chapter

35 Myelomeningocele and Hydrocephalus

Adam C. Adler

A four-year-old child presents for replacement of her ventriculo-peritoneal (VP) shunt. She has spina bifida diagnosed postnatally and has mild developmental delays. Over the last day, she was lethargic at home and "not acting herself" prompting a trip to the emergency department by her parents.

In the preoperative area, she is responsive but very sleepy. Vitals include: Temp 37.6^{0}C, BP 107/62, HR 67 bpm, SpO_2 100%. She is your next case to follow.

Two hours later, the nurse in the preoperative holding area calls you concerned that she seems "sleepier" and not very arousable. She is also concerned that the patient's heart rate is only 49.

What Are the Types of Spina Bifida?

Spina bifida is a broad term encompassing defects of the skin (spina bifida occulta), vertebrae, meninges (meningocele), and spinal cord (myelomeningocele [MMC]).

What Are the Predisposing Risk Factors Related to Spina Bifida?

Maternal folate deficiency has been associated with a two- to eight-fold increased risk for developing spina bifida. Other maternal factors include vitamin B12 deficiency, pregestational diabetes, obesity, and antiepileptic use (carbamazepine and valproic acid). History of previous pregnancy (with the same partner) complicated by spina bifida affords significant increased risk.

What Prenatal Studies Are Performed to Assess for Spina Bifida and the Degree of Neurologic Involvement?

Elevated levels of maternal serum alpha-fetoprotein are suggestive of spina bifida or anencephaly. Amniocentesis allows for measurement of alpha-fetoprotein in the amniotic fluid and fetal karyotyping for associated chromosomal abnormalities. When the diagnosis is suspected, ultrasound or fetal MRI can assess for neurologic defects and presence of a Chiari malformation. Ultrasound of the lower extremities is used to assess fetal leg and foot movement and associated limb deformities.

What Is the Incidence of Myelomeningocele (MMC)?

The incidence of MMC is around 0.5–1:1,000 live births.

What Are the Etiologies of MMC?

The exact etiology of MMC remains unknown. Failure of closure of the neural tube or mesenchymal closure of the caudal neuropore during development has been suggested. Exposure of unprotected neural tissue may result in traumatization of the spinal cord. In addition, it is thought that prolonged exposure of the spinal cord tissue to amniotic fluid throughout gestation leads to damage.

What Is the Defect That Occurs with a MMC?

A defect in the vertebral arches leads to protrusion of the meninges and spinal cord (neural tissue).

What Are the Most Common Locations of MMC Defects?

Most commonly, the defect occurs in the lumbar region, although it can occur at any location along the spinal cord. Anencephaly occurs when the cerebral portion of the primary neural tube fails to close.

https://doi.org/10.1017/9781108668736.036 Published online by Cambridge University Press

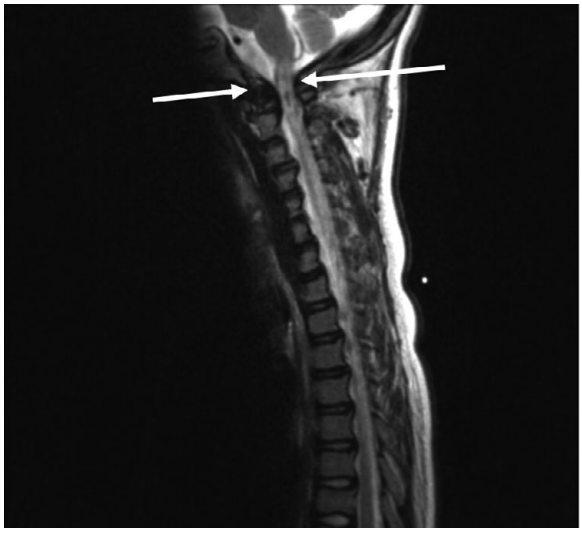

Figure 35.1 MRI of brain with herniation of cerebellar tonsil tissue at the level of the foramen magnum (arrows)

What Are the Potential Causes of Neurologic Morbidity in Patients with MMC?

Long-term sequelae relate to immobility leading to pressure-related ulcerations, venous thrombosis leading to deep venous thrombosis/pulmonary embolism, urinary tract infection from frequent bladder catheterizations, and issues relating to hydrocephalus and shunt function.

Hydrocephalus is common in children with MMC especially in the presence of Chiari malformations and may be related to obstruction of CSF flow by the herniated cerebellum (Figure 35.1).

What Are the Associated Neurologic Disabilities Attributed to a MMC?

Neurologic manifestations include: varying degrees of mental retardation, bowel and bladder dysfunction, and orthopedic-related disability. The degree of bowel and bladder dysfunction and extremity paralysis correlates with the level of herniated spinal cord tissue.

What Is the Rationale for Midgestation Fetal Repair of MMC?

Prolonged fetal exposure of the developing spinal cord to amniotic fluid is thought to contribute to significant neurologic morbidity. Fetal correction via a midgestational procedure has been shown to significantly improve neurologic function and reduce the morbidity from hydrocephalus and the Arnold–Chiari malformation. Fetal repair lends a significant reduction in the need for VP shunt, a major cause of morbidity for these patients. The considerations for fetal intervention are discussed in Chapter 46.

What Other Conditions Are Associated with a MMC?

Patients with spina bifida have a high incidence of Arnold–Chiari II malformation. The Chiari II malformation is herniation of the hindbrain (cerebellum and brainstem) through the foramen magnum. Fetal correction of MMC is associated with a reduced incidence of Chiari II malformations. Chiari II malformations are corrected by surgical decompression to remove the bone surrounding the hindbrain. The repair is performed in the prone position, is generally extradural and with the assistance of neurologic evoked potential monitoring.

What Is the Classic Allergy Triad in MMC Patients?

Patients with MMC are typically assigned an empiric latex allergy. Historically, latex-based products were used during the patient's surgical procedures in addition to the latex-based urinary catheters which led to sensitization and subsequent allergy. Latex is a rubber derivative. Additionally, a syndrome known as latex-fruit syndrome exists in which certain fruits (banana, kiwi, mango, pineapple, chestnuts, strawberry, and soy) produce a substance similar to that of latex which can also lead to allergic reaction. Approximately 30–50% of patients with latex allergy have cross allergic reactions to these fruits.

What Are the Signs and Symptoms of Patients with Chiari II Malformations?

Neonates with Chiari II malformations may present with swallowing difficulties, stridor, apneic spells, weak cry, or arm weakness. Older children and adults may present with bilateral limb weakness, muscle wasting, and sensory disturbances. Less commonly, dysphagia and ataxia can also be present.

https://doi.org/10.1017/9781108668736.036 Published online by Cambridge University Press

What Are the Anesthetic Issues Associated with MMC Repairs?

Children with large MMCs generally undergo surgical repair in the first days of life as the exposed neural tissue is at risk for infection. Large open MMC should be kept covered and moist to prevent significant evaporative losses. When intubating these children, care must be taken to avoid pressure on neural tissue in the supine position. Foam or gel donuts are used under the patient to avoid pressure on the exposed spinal cord.

What Are Evoked Potentials?

Evoked potentials provide testing of the continuity of major neurologic tracts in the brain and spinal cord. The tracts are evaluated by application of an external stimulus and measurement of the signals propagated by the neurologic pathway. Testing of evoked potentials is generally reserved for surgical procedures during which a high risk of neurologic pathway disruption exists.

There are four main types of evoked potentials monitored in the operating room setting: visual, auditory, somatosensory, and motor potentials.

- Somatosensory evoked potentials (SSEP) involve a stimulus applied to the extremities with signal propagation sampled at the cortex.
- Motor evoked potentials (MEP) involve stimulation of the motor cortex with measurement of signal propagation at the extremities.

SSEP and MEP monitoring allow for monitoring of the posterior spinal cord and dorsal columns (SSEP) and the anterior spinal cord and corticospinal tract (MEP) respectively.

Evoked potentials measure a baseline amplitude (signal strength) and latency (time between signal transmissions). Continuous monitoring throughout the case is performed with changes from baseline signifying a potential disruption of neural tissue. Decreases in amplitude of >50% or increases in latency of >10% are usually considered significant. Corticospinal tract assessments (MEP) are assessed intermittently to assure tract integrity especially after hardware placement.

Table 35.1 Changes to measured amplitude and latency expected with commonly used anesthetic agents

	Amplitude	Latency
Volatile agents	↓	↑
Nitrous oxide	↓	–
Propofol	↓	↑
Benzodiazepines	–	–
Ketamine	↑	–
Etomidate	↑	↑
Opioids	–	–
Barbiturates	↓	↑

How Do Anesthetics Affect Evoked Potentials?

Commonly used anesthetic drugs have varying effects on the monitoring of evoked potentials (Table 35.1).

Neuromuscular blocking drugs preclude the monitoring of motor evoked potentials. Volatile agents generally affect evoked potential monitoring on a dose dependent basis. Maintenance of a continuous plane of anesthesia is paramount. Major changes to infusion levels or bolus doses can cause large changes in evoked potentials.

During the Surgical Procedure, the Neuromonitoring Technician Reports Loss of Signals. What Are the Next Steps in Management?

- The first step is to be certain that all team members, especially the surgeon, are aware of the signal change.
- Change to total intravenous anaesthesia (TIVA) technique if not already in progress.
- Consider reversing the last step of the procedure when appropriate (removal of pedicle screw, distraction of spine, etc.).
- Increase perfusion pressure using vasopressors to elevate the mean arterial pressure (and hence the spinal cord perfusion pressure).
- Consider placing arterial line/central line if pressor infusion to be continued postoperatively.
- Transfuse packed red blood cells (PRBC) if anemic.
- Maintain normocarbia.

https://doi.org/10.1017/9781108668736.036 Published online by Cambridge University Press

- Reduce PEEP to optimize spinal cord perfusion.
- Maintain normothermia.
- Consider a wake-up test to assess neurologic function.
- Consider high dose corticosteroids.

What Are the Signs and Symptoms of Raised Intracranial Pressure?

Signs and symptoms of raised intracranial pressure (ICP) in children may include large head size, cognitive delays or regression, headache, incontinence, irritability, memory loss, decreased appetite, poor coordination, regression of milestones, seizures, visual disturbances.

What Are the Causes of Hydrocephalus in Children?

Hydrocephalus, or enlargement of the intracranial ventricles, is attributed to excess CSF production, disturbance of flow, or impaired reabsorption. Most commonly, the etiology of hydrocephalus involves impaired flow of CSF (obstructive hydrocephalus) and may be isolated or associated with other congenital or acquired malformations or tumors.

Prior to closure of the fontanelle, the infant skull can accommodate to ventricular enlargement. Once the fontanelle has closed, ventricular enlargement can lead to rapid and life threatening elevations in intracranial pressure if left untreated.

What Is Hydrocephalus Ex-Vacuo?

Hydrocephalus ex-vacuo refers to dilation of the ventricles due to surrounding brain atrophy and not the result of CSF flow obstruction.

Identify the Pathway from CSF Production to Reabsorption

CSF is produced by ependymal cells in the choroid plexus of the lateral ventricles. From there, CSF flows through the interventricular foramen into the third ventricle, through the cerebral aqueduct and into the fourth ventricle. The fourth ventricle, in communication with the spinal cord, allows CSF to flow in the subarachnoid space.

CSF is reabsorbed by the vascular system through dural venous sinuses in the spinal cord via arachnoid granulations.

What Is a Shunt?

A shunt is a valved conduit placed in the ventricle to allow drainage of CSF.

What Are the Indications for Shunt Placement?

Shunt requirement is based on symptomatology and/or the inability for CSF to flow properly.

What Are the Possible Locations for CSF Shunt Placement?

Shunts have their proximal end within the lateral ventricle (Figure 35.2). The distal end most commonly is placed in the peritoneal cavity. Other places for distal shunt placement are intra-pleural and intra-atrial. Intra-abdominal infection is the most common reason to avoid peritoneal shunt placement.

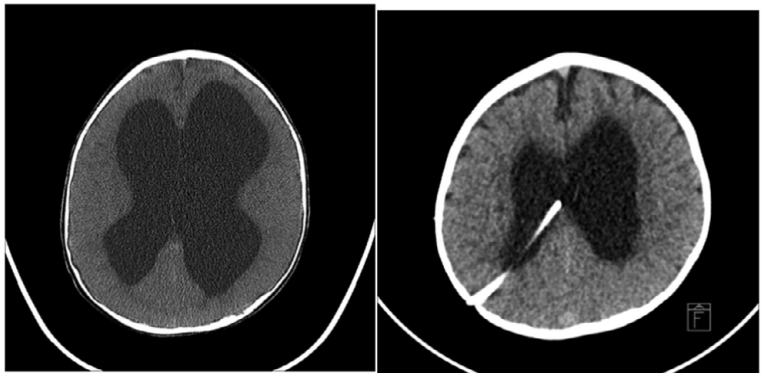

Figure 35.2 CT scan demonstrating hydrocephalus of the lateral ventricle pre (left) and post (right) decompression with a midline crossing VP shunt

https://doi.org/10.1017/9781108668736.036 Published online by Cambridge University Press

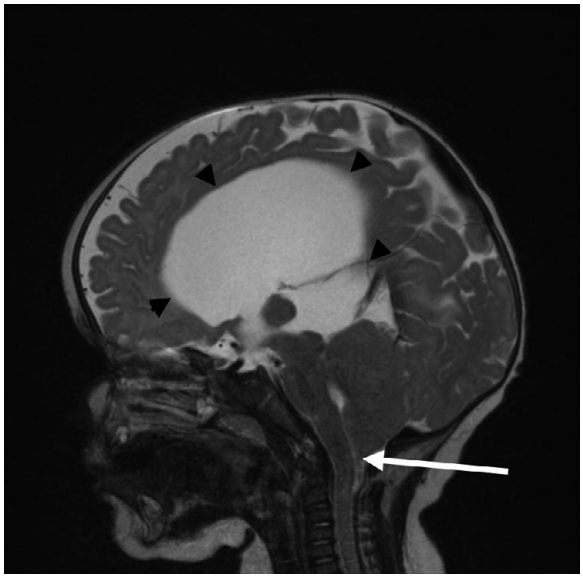

Figure 35.3 Brain MRI demonstrating enlargement of the lateral ventricle (arrow heads) and herniation of cerebellar tonsil (white arrow)

Describe the Symptoms and Signs Associated with Shunt Malfunction

Shunt malfunction in children with open fontanelle can be masked by enlargement of the head to accommodate the excess CSF (Figure 35.3).

In children with closed fontanelle, or those who have exhausted their compensatory mechanism, behavioral changes, somnolence, setting sun eyes, vomiting, and when severe, Cushing's reflex, may be present.

What Is the Cushing Reflex/Triad?

The Cushing reflex, described by famed neurosurgeon Dr. Harvey Cushing, is a physiologic response to elevated intracranial pressure. The reflex consists of hypertension, bradycardia, and an abnormal respiratory pattern. Hypertension may be a compensatory mechanism for preservation of cerebral perfusion in the setting of elevated ICP.

A Pre Op Nurse Calls You, Concerned for Bradycardia in a Patient Coming for VP Shunt Malfunction. What Is the Appropriate Plan of Action?

Bradycardia in this child may be the result of significant elevations in ICP. The neurosurgical team should be notified immediately, and the patient taken to the operating room emergently for decompression. CSF can be removed if an external ventricular drain is present. Drainage of CSF may be possible via needle insertion into a shunt with a reservoir by the neurosurgeon.

Suggested Reading

Adzick NS. Fetal myelomeningocele: natural history, pathophysiology, and in-utero intervention. *Semin Fetal Neonatal Med*. 2010;15(1):9–14. PMID: 19540177.

Fetal surgery for spina bifida: past, present, future. *Semin Pediatr Surg*. 2013;22(1):10–17. PMID: 23395140.

Hadley DM. The Chiari malformations. *J Neurol Neurosurg Psychiatry*. 2002;72 Suppl 2: ii38–ii40. PMID: 12122202.

Kumar A, Bhattacharya A, Makhija N. Evoked potential monitoring in anaesthesia and analgesia. *Anaesthesia*. 2000;55(3):225–41. PMID: 10671840.

Mitchell LE, Adzick NS, Melchionne J, et al. Spina bifida. *Lancet*. 2004;364(9448):1885–95. PMID: 15555669.

https://doi.org/10.1017/9781108668736.036 Published online by Cambridge University Press