Myelomeningocele

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KEY POINTS

- Myelomeningocele is the most common neural tube defect.
- Myelomeningocele is frequently associated with Chiari II malformation and obstructive hydrocephalus.
- Myelomeningocele requires surgical repair to preserve neurologic function and reduce the risk of central nervous system infection.
- Surgical repair is most commonly performed postnatally, though certain centres are now performing repairs in utero.
- Anaesthetic considerations for positioning, temperature management, and airway management are paramount.

INTRODUCTION

Neural tube defects during fetal development can result in long-term neurologic deficits. Myelomeningocele (also referred to as spina bifida aperta) is a neural tube defect that results in exposure of the meninges and spinal cord to the environment. The spectrum of neural tube disorders also includes meningocele, a defect of the spinal column resulting in protrusion of meninges posteriorly into a sac which is covered by dermis, and spina bifida occulta, a disruption in development of the posterior spine where there is no hernial protrusion of the meninges or spinal cord (Figure 1).

Myelomeningocele, the most severe form of spina bifida, is the most common neural tube defect and requires urgent surgical repair to avoid significant neurologic sequelae.[1] Most commonly, this surgery is performed postnatally; however, at some institutions, fetal surgery to repair the neural tube defect is being performed between 23 and 25 weeks gestation.[2] There are a number of anaesthetic implications of this condition, especially for postnatal repairs.

ETIOLOGY

Myelomeningocele is caused by a neural tube defect in the third or fourth week of fetal development. A failure of closure of the neural tube results in the posterior protrusion of a sac of meninges with an encapsulated spinal cord exposing them to the environment. It is the most common form of neural tube defect. Its incidence varies according to regions, ranging from 1 to 7 per 1000 live births.

The defect can occur at any level along the spinal column, but is most commonly found at lumbar or sacral regions. There is significant variability in the size and severity of the defects, with more cephalad defects typically being associated with worse neurological outcomes. Adequate maternal nutrition and folic acid supplementation have been found to reduce the incidence of neural tube defects.[3]

Newborns with myelomeningocele lack the natural protective tissue barrier for the spinal cord, predisposing the central nervous system to environmental exposure and/or trauma. This vulnerability can result in bacterial contamination of the cerebrospinal fluid and

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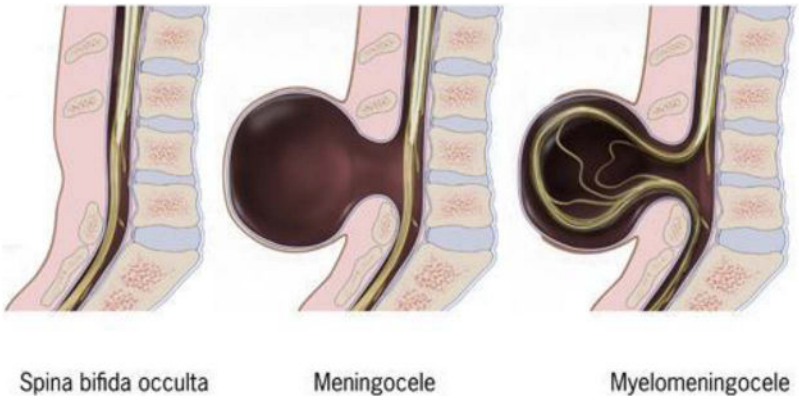

Figure 1. The spectrum of neural tube defects. Center for Disease Control and Prevention. Public Domain. July 24, 2023.

meninges. Myelomeningocele requires urgent surgical repair because failure to cover the defect increases the incidence of central nervous system infection or damage. Also, an earlier closure of the defect is associated with better neurological outcomes.[4] Long-term manifestations of neurological compromise related to myelomeningocele include loss of bowel and bladder function, inability to ambulate, and development of pressure ulcers.[5]

Most myelomeningoceles are associated with Arnold-Chiari II malformation (herniation of the cerebellum through the foramen magnum) and obstructive hydrocephalus. These additional problems can lead to symptoms of brainstem impairment, such as motor/sensory deficits and respiratory or oropharyngeal dysfunction.

Overall, Hispanics have the highest prevalence of neural tube defects, with a reported 3.8 per 10 000 live births, compared with 2.73 per 10 000 in Black populations and 3.09 per 10 000 in non-Hispanic white populations.[6] Additional risk factors for neural tube defects include family history with a recurrence rate of 3% to 8% after one affected pregnancy.[7]

POSTNATAL REPAIR

Postnatal repair of myelomeningocele is presently the most common technique worldwide and is currently the only option available in financially restrained areas. The infrastructure and workforce required for postnatal repair includes a neonatal intensive care unit for initial stabilization and subsequent management of the critically ill newborn, specialized neurosurgical and/or plastic surgeons with experience in neural tube defects, as well as anaesthesiologists experienced in the care of newborns.

PREOPERATIVE CONSIDERATIONS

The patients are typically stabilized in the delivery room and positioned in the prone or lateral decubitus position to remove pressure and contact with the defect. Preventing contact or pressure on the defect reduces the likelihood of infection and further damage to the exposed, delicate neural tissue. The neural tube defect is traditionally covered with a sterile moist gauze cover until surgical intervention is performed.

Immediately following delivery, intravenous access is obtained for routine administration of broad-spectrum antibiotic prophylaxis and for fluid and electrolyte repletion, if needed. Surgical correction is recommended to be performed within 48 to 72 hours to avoid severe systemic infection.[1]

All equipment should be latex free to avoid latex sensitization in this patient population. Spina bifida is strongly associated with development of latex allergy.

The patients commonly have associated hydrocephalus and can present with increased intracranial pressure. The severity and progression of intracranial hypertension will determine if the patient requires a ventriculoperitoneal shunt at the time of surgical correction.[8] The rate of ventriculoperitoneal shunt placement has been reported around 80% in patients with myelomeningocele, while some argue that a more conservative management of intracranial hypertension might be best.[9]

Preoperative laboratory assessment with a complete blood count, ABO typing/cross-matching, and basic metabolic panel should be performed prior to surgery. Any electrolyte derangements should be corrected, and blood should be available prior to surgical intervention.

Preoperative evaluation should include a thorough physical examination with special attention to baseline respiratory status of the patient. If the patient has associated hypoventilation or apneas related to brainstem involvement from associated structural

defect of the brainstem, they may be at higher risk of respiratory complications postoperatively.[10] Additionally, approximately 30% of patients will present with a congenital heart defect, most commonly an atrial septal defect or a ventricular septal defect. Cyanotic heart diseases like anomalous pulmonary venous return, tetralogy of Fallot, and hypoplastic left heart syndrome may also be associated with open myelomeningocele. Cyanotic neonates should always undergo an echocardiographic assessment. It is important to recall that approximately 20% of patients with myelomeningocele have additional congenital or chromosomal abnormalities.[11]

INTRAOPERATIVE CONSIDERATIONS

Positioning the patient prior to induction is critical to avoid further pressure and subsequent damage to the exposed neural tissue. The patient can be elevated with blankets leaving the defect free of pressure or contact with the operating table. In cases of very large defects, the patient can be placed laterally for induction. Ultimately, the patient will be placed prone for the procedure (Figures 2 and 3).

Standard monitors are used, including electrocardiogram, oxygen saturation, and noninvasive blood pressure monitoring.[12] Arterial lines are not usually required in the absence of hemodynamic instability. Discussion with the surgical team regarding their desire to use neuromonitoring modalities should take place prior to induction. This conversation is critical, as this would affect the anaesthesia induction plan if usage of neuromuscular blocking agents was anticipated and needs to be avoided.[13]

Maintenance of normothermia should be prioritized and temperature should be closely monitored. Given the large surface area of severe lesions, the patient can rapidly become hypothermic, especially if the positioning and preparation for surgery is prolonged. Hypothermia can cause physiologic derangement of numerous organ systems in these patients and should be avoided.[14] Temperature can be maintained with forced air and fluid warming devices, in addition to other radiant warming equipment.

Central venous access is not required if peripheral intravenous (IV) access is secured. Two peripheral IVs should be considered in case blood transfusion or volume resuscitation is required.

Most postnatal myelomeningocele repairs are performed with general endotracheal anaesthesia. Induction of anaesthesia can be performed intravenously or with inhaled anaesthesia.[15]

Of note, spinal anaesthesia has been performed for this procedure with hyperbaric tetracaine via direct injection of local anaesthesia into the neural tube defect.[16] In one case series, the spinal technique was performed on 14 neonates with 100% success rate. Supplemental intrathecal anaesthesia was required for longer procedures. Initial doses lasted about 90 minutes. The neonates received supplemental IV benzodiazepines as needed for agitation. Two patients had postoperative apnea/bradycardia episodes: one had history of central neonatal apnea prior to the procedure and another episode occurred after IV morphine administration.

During general endotracheal anaesthesia, the airway should be secured with an endotracheal tube prior to prone positioning. The anaesthesia team should be prepared for a difficult airway, as the size of the defect may preclude optimal positioning for laryngoscopy. Occasionally, the patient needs to be intubated in the lateral position.[15] Maintenance of general anaesthesia may be accomplished with total intravenous anaesthesia, with inhaled volatile agents or a combination of both. Extubation should be considered if the patient meets criteria. Of note, the patient will need to remain in the lateral or prone position to allow the defect to heal.[15]

Post-operatively, the patient should be transported to a neonatal intensive care unit for close neurological monitoring and evaluation for possible central nervous system infection.

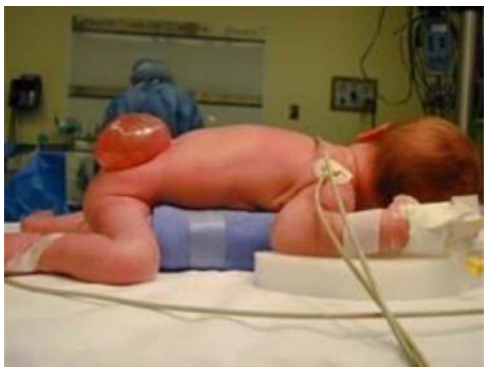

Figure 2. Original photo of child with myelomeningocele in the prone position prior to surgical repair.

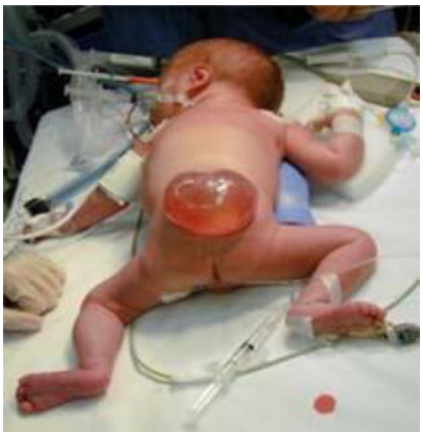

Figure 3. Original photo of child with lumbosacral myelomeningocele in the prone position prior to surgical repair.

PRENATAL SURGICAL REPAIR

Fetal surgery has been performed for myelomeningocele for decades. The procedure is performed mainly at maternal-fetal surgery centers.[4] In the United States of America, there are only a few major medical centres that offer fetal repairs of myelomeningocele. The procedure is performed between 23 and 25 weeks of gestational age and involves a maternal laparotomy, followed by a uterotomy and externalization of the fetus.[4] The exposed neural tissue is then returned to the spinal canal to avoid continued exposure to the amniotic fluid. Finally, the overlying skin is approximated after the fetus is internalized followed by closure of the uterus and abdomen.

The MOMS trial in 2011 was the largest trial comparing fetal intervention versus postnatal surgical correction of myelomeningocele. The results demonstrated that fetal intervention improved independent ambulation at 30 months (42% versus 21%) and reduced the rate of ventriculoperitoneal shunt placement (40% versus 82%). However, fetal intervention increased the incidence of maternal morbidity related to an increase in uterine dehiscence (10% versus 0%) and an increased incidence of preterm delivery (79% versus 15%).

Patients who were included in the original MOMS trial have been followed since their initial intervention. In 2020 a follow-up publication revealed that, at 6 years of age, patients who underwent the prenatal surgical repair showed persistent independent ambulation and mobility benefits compared to those who underwent the postnatal repair (29% versus 11%).[17]

The types of anaesthesia for patients in the MOMS trial consisted of general anaesthesia with epidural placement for the mother, and intramuscular injection of fentanyl and vecuronium for the fetus. The balance of long-term outcome benefits for the fetus versus increased morbimortality to the mother are continued concerns with intrauterine surgical repair.

SUMMARY

Myelomeningocele is the most common neural tube defect. Although fetal surgical procedures are available at specific centres, to date postnatal repair is the most common surgical intervention worldwide. The defect requires urgent surgical correction and there are several important considerations for the anaesthesia. Both general and spinal anaesthesia have been described. Positioning requirements for these patients can make airway management difficult and this must be anticipated. Temperature management can be difficult, especially if the defect is large. The patient's postoperative disposition is to a neonatal intensive care unit for close monitoring of central nervous system infection signs or symptoms and frequent neurological assessments.

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