Myelomeningocele

Protrusion of the spinal cord and its membranes through a defect in the vertebral column

ANESTHETIC CONSIDERATIONS:

- Considerations for the neonatal patient
- Exposed neural tissue
  - Hypovolemia 2° to underestimation of large fluid and blood loss from defect – replace all fluid deficits, including loss from defect; ensure x-matched blood available
  - Neurologic injury – place "doughnut" under defect to protect when supine
  - Hypothermia
  - Infection risk
- Arnold-Chiari malformation
  - Obstructive hydrocephalus
  - Aspiration risk 2° to brainstem dysfunction
  - Brainstem herniation
- Succinylcholine safe – does not elicit hyperkalemic response
- Latex allergy precautions
- Associated congenital anomalies

ANESTHETIC GOALS:

- Ensure correction of fluid deficits and blood loss
- Protect exposed neural tissue while positioning for intubation
- Avoid increasing ICP, maintain CPP
  - Avoid brainstem herniation and worsening neural tissue herniation through vertebral defect
- Ensure latex-free environment

HISTORY

- Birth Hx
- Neurologic deficits
  - Back pain, lower extremity weakness (progressive → flaccid paraplegia), spasticity, sensory deficits, bowel/ bladder dysfunction
- Associated congenital defects
  - Arnold-Chiari II malformation
    - Hydrocephalus (altered LOC, headaches, blurred vision (CN II palsy), diplopia (CN VI palsy), stridor (CN X palsy), N/V, neck pain, incontinence, seizures, presence/function of VP shunt
  - Cardiac defects
  - Klippel-Feil syndrome – congenital fusion of cervical vertebrae
  - Hydroureter/hydronephrosis – recurrent UTI/sepsis
  - Bladder exstrophy, uterine prolapse
  - MSK – scoliosis, hip dysplasia/dislocation, talipes equinovarus
- PSHx
  - Often multiple urologic and MSK procedures
- Allergies
  - Hx latex allergy (pruritus, rash, edema after wearing latex gloves or inflating toy balloons)

PHYSICAL

- Vital signs – HR, BP, RR, SpO2, T
- CNS
  - Hydrocephalus – level of consciousness, enlarged head, bulging fontanelles, dilated scalp veins, Macewen sign, papilledema, sunset eyes (failure of upward gaze), CN VI palsy, ataxia, spasticity
  - Spinal cord damage – muscle tone and strength (paralysis), sensation, anocutaneous reflex
  - Spine – cystic mass, skin with hair tuft, scoliosis
- HEENT – cranial sutures open vs closed
- Airway – stridor (CN palsy), standard airway exam
- CVS – intravascular volume status, heart sounds, murmurs
- Respiratory – respiratory distress, apneas,
- GU – associated congenital anomalies

INVESTIGATIONS

- **Labs**
  - CBC/D, lytes, urea, Cr
- **Imaging**
  - MRI spine – best radiographic test to confirm neural tube defects and/or spinal cord anomalies
  - CT head – to identify associated Chiari II malformation/hydrocephalus in myelomeningocele
  - Echocardiogram – if suspect cardiac defects

OPTIMIZATION

- **Protect exposed neural tissue**
  - Prone positioning; "doughnut" support under defect when supine
  - Cover defect with warm saline-soaked gauze to prevent injury, drying, and infection of spinal cord elements
  - Antibiotic prophylaxis

- **Replace fluid deficits**
  - Including losses from the defect (full strength balanced salt solution; NS)
  - Cyst rupture can lead to ongoing CSF leakage
- **Identify and treat hydrocephalus**
  - Consider need for VP shunt insertion/revision vs. posterior fossa decompression

ANESTHETIC OPTIONS

- **Regional anesthesia**
  - Case reports of spinal anesthesia (tetracaine) and midazolam sedation for myelomeningocele repair
- **General anesthesia**
  - Most commonly used

ANESTHETIC SETUP

- **Drugs**
  - Standard emergency drugs
- **Equipment**
  - Standard CAS monitors
- **Special**
  - Ensure x-matched blood available
  - Latex-free precautions

MANAGEMENT OF ANESTHESIA

- **Induction**
  - Avoid pressure on myelomeningocele sac during intubation
    - Protect sac by elevating on doughnut-shaped support
  - Avoid increasing ICP during intubation in presence of hydrocephalus
  - Succinylcholine safe – does not elicit hyperkalemic response
- **Maintenance**
  - Prone positioning
    - Ensure abdomen and chest are free to avoid pressure on epidural venous plexus (minimize bleeding) and allow adequate ventilation
  - Avoid long-acting NMBA – surgeon may need to use nerve stimulator to identify functional neural elements
  - To test that surgical closure of myelomeningocele sac tight enough to prevent CSF leakage – positive airway pressure to increase pressure in sac
  - Monitor for blood loss – may be insidious especially if large sac requiring significant undermining of subcutaneous space to ensure closure of defect
- **Emergence**
  - Typically extubated at end of case

DISPOSITION & MONITORING

- **Analgesia**
- **Oxygenation**
- **Positioning**
  - Maintain in prone position
- **Monitoring**
  - Monitor closely for development of ↑ICP
    - Brainstem dysfunction (stridor, apnea, bradycardia, cyanosis, respiratory arrest)
    - Hydrocephalus (lethargy, vomiting, seizures, apnea, bradycardia, CV instability)

COMPLICATIONS

- Progressive neurologic deficits
- Tethered cord
- Meningitis/ventriculitis
- Sepsis
- Anaphylaxis (latex)

OBSTETRICS

- Interactions between pregnancy and residual neurologic impairment with ongoing orthopedic and urologic complications post-repair of myelomeningocele
  - Recurrent UTI risk factor for PTL
  - Ileal conduits/stomas – intestinal and urinary tract obstruction, difficult c-section delivery
  - Greater immobility – pressure sores
  - Kyphoscoliosis + uterine enlargement – may compromise pulmonary function
  - Pelvic and lower limb anomalies/contractures – may obstruct pelvic outlet and warrant c-section
- Spina bifida occulta
  - Attempt to identify epidural space at site of lesion will likely result in dural puncture
    - Rarely a concern as lesion typically below site of epidural/spinal insertion
- Tethered cord syndrome
  - Low-lying and more posteriorly located spinal cord in occult spinal dysraphism and spina bifida *cystica* (not occulta)
    - ↑Risk direct needle trauma with spinal/epidural insertion; epidural safer than spinal
  - Obtain neurologic Hx and perform screening neuro/MSK exam if Hx of defective laminar arch
    - If spinal dysraphism suspected but no imaging available – prudent to avoid neuraxial anesthesia
  - Avoid prolonged lithotomy position
- Spina bifida cystica
  - Determine level of lesion and residual neurologic function below lesion

- Painless labor with complete lesion at/above T11
  - Risk of autonomic hyperreflexia with high thoracic lesions
- Determine presence/status of VP shunt; consult Neurosurgery
- Assess pulmonary function if scoliosis
- Determine baseline renal function
- Epidural space often abnormal; risk of inadequate epidural analgesia

- Neuraxial anesthesia may be considered in various forms of spina bifida with stable neurologic function provided that:
  - Full informed consent including patients informed limited data on risk of neurologic injury with neuraxial anesthesia in spina bifida
  - Consider neuroimaging to identify terminal portion of spinal cord, which typically lies at a vertebral level lower than normal
    - However, when there is negligible function of lower extremities and sphincters, concern for direct neural trauma to low-lying spinal cord not clinically relevant
  - Perform spinal anesthesia below level of conus or avoid in favor of epidural
- Note: majority of above considerations apply to any adult patient with spina bifida

PATHOPHYSIOLOGY

- **Definitions**
  - Neural tube defect (aka myelodysplasia) – abnormality in fusion of the embryologic neural groove that normally closes in the first month of gestation; failure of the canal end of the neural tube to close can result in:
    - Spina bifida occulta – defects of the vertebral arches leaves spinal cord unprotected although covered by skin; rarely neuro Sx, ↑risk posterior disc herniation
    - Occult spinal dysraphism – vertebral arch defect associated with less severe spinal cord anomalies (lipomas, cysts, bands); absence of or minor neurologic symptoms involving lower limbs, bowel, and bladder
    - Spina bifida cystica – failed closre of neural arch with herniation involving:
      - Meningocele – saclike herniation of the dura and arachnoid components of the meninges through the vertebral defect; neuro deficits, MSK anomalies
      - Myelomeningocele – saclike herniation of the dura, arachnoid, and pia components of the meninges and neural elements of spinal cord through the vertebral defect; neuro deficits, MSK anomalies
  - Arnold-Chiari II malformation
    - Caudal displacement of the cerebellar tonsils through the foramen magnum, caudal displacement of the medulla oblongata and cervical spine, kinking of medulla, and obliteration of cisterna magna.
- **Epidemiology**
  - Prevention of neural tube defects – antenatal folic acid, avoid antiepileptic agents if not essential
  - Spina bifida occulta in 10% of normal population
  - Occult spinal dysraphism associated with cutaneous stigmata (50%) and tethered spinal cord (70%)
  - Myelomeningocele is the most common form of non-occulta spina bifida; 0.5-1 in 1000 live births
  - Arnold-Chiari II malformation – 80-90% have associated hydrocephalus, 20% have associated brainstem dysfunction
- **Pathophysiology**
  - Spina bifida results from failure of neural tube closure during 4th week of gestation
    - Most commonly occurs in lumbosacral or sacral region; may extend to thoracic region
    - Detected on prenatal screening
      - Ultrasound
      - ↑Maternal serum AFP (5% false-positive rate); ↑amniotic fluid AFP more reliable
  - Consequences of neural tissue exposure
    - Trauma
      - Neural damage
      - Scarring of neural elements prior to or at time of closure can result in tethered cord; traction on conus medullaris
    - Spinal cord tethered caudally by sacral roots → back pain, neurologic Sx, urologic Sx
    - Leakage of fluid → hypovolemia
    - Heat loss → hypothermia
    - Infection → meningitis, ventriculitis
  - Associated Arnold Chiari II malformation
    - Hydrocephalus – most patients eventually require shunt (33% receive shunt during initial hospitalization)
    - Brainstem dysfunction – vocal cord paralysis → stridor, aspiration; sleep-disordered breathing; apnea; bradycardia, lack of coordination, spasticity
      - Vocal cord paralysis may be 2° to either pressure on brainstem (i.e., with hydrocephalus) or focal infarcts
      - Tx requires decompression of hydrocephalus – VP shunt, high cervical/posterior fossa decompression
        - Some children require a tracheostomy for long-term care
    - Potential for brainstem herniation
  - Recurrent bladder catheterization, surgical procedures, and hospitalization
    - Risk of sensitization to latex
- **Management**
  - Early Neurosurgical repair for closure of neural tube defects
    - Severe anomalies require surgical intervention within 24-48hrs of life to reduce risk of damage to and infection of exposed CNS tissue

REFERENCES

- Miller p.2589, 2671
- Barash p.1198-99
- Chestnut p.1046-48
- Anesthesia and Coexisting Disease p. 608-609