

Perioperative Implications**Preoperative Preparation**

- Isotonic fluid replacement to correct fluid deficits and electrolyte abnormalities
- Aspiration prophylaxis: H₂ antagonist, nonparticulate antacid, and nasogastric tube placement
- Antibiotics with adequate gram-negative and anaerobic coverage
- Opiate premedication for abdominal pain
- Baseline fetal heart rate tracing in parturient

Monitoring

- Standard.
- Urinary catheter.
- Consider invasive monitors if septic.
- Consider fetal heart rate monitor if fetus is viable and obstetrics team is immediately available.

Airway

- Full stomach precautions

Induction

- Rapid sequence induction with cuffed ETT.
- Anticipate hemodynamic instability with induction in septic or dehydrated pts.

- In parturient, avoid supine hypotension syndrome by positioning in left uterine displacement.
- Can consider regional anesthesia if pt is cooperative, hydration is adequate, systemic sepsis is absent, and high abdominal exploration is unlikely.

Maintenance

- Standard maintenance with adequate muscle relaxation.
- Evacuate stomach with oral-gastric or nasogastric tube.
- If using laparoscopic approach, anticipate hypotension with abdominal insufflation causing decreased venous return and cardiac output.
- In parturient, ensure fetal well-being by maintaining adequate maternal oxygenation, hemoglobin content, and hemodynamics.

Extubation

- Extubate when pt is fully awake and regains laryngeal reflexes.

Postoperative Period

- Pain control with PCA, oral opioids, and NSAIDs.
- PONV; Treat with ondansetron, metoclopramide, or promethazine.

- Continue antibiotics for 3-5 d for perforated appendicitis.
- Monitor for sepsis.
- In parturient:
 - Fetal heart rate monitor to ensure fetal well-being.
 - Consider tocolytic medication to prevent premature labor (22% of women in third trimester go into labor within 1 wk of surgery).

Adjuvants

- Laparoscopic versus open appendectomy: Laparoscopic approach tends to have shorter hospital stay, decreased postop pain, and decreased wound infection rate; however, it also tends to have a higher rate of intraabdominal abscess and hospital costs.

Anticipated Problems/Concerns

- Aspiration risk
- Hemodynamic instability due to dehydration and possible sepsis

Arnold-Chiari Malformation (Chiari Malformation Type II)

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Risk

- Arnold-Chiari malformation (Chiari Malformation type II or CMII) is found exclusively in pts with myelomeningocele.
- Myelomeningocele occurs in 0.6 of 1000 live births.

Perioperative Risks

- Vocal cord paralysis
- Respiratory distress
- Apnea
- Neurogenic dysphagia and pulmonary aspiration
- Hydrocephalus and increased ICP
- Congenital heart defects (37% of pts with myelomeningocele) including atrial septal defect, ventricular septal defect, anomalous pulmonary return, tetralogy of Fallot, bicuspid aortic valve, coarctation of the aorta, and hypoplastic left heart syndrome

Worry About

- Any symptoms of possible brainstem compression (stridor, hoarse voice, or difficulty swallowing) in a child less than 2 y with CMII must be urgently evaluated as a neurosurgical emergency.
- Nearly 21% of children with myelomeningocele will develop hindbrain, cranial nerve, or spinal cord compression by 3 mo of age, increasing to 33% by age 5 y.

- Of pts with symptomatic CMII, 15% die by 3 y of age.
- Mortality rate has improved with emergent surgical treatment in symptomatic pts.

Overview

- CMII is characterized by herniation of the cerebellar vermis, brainstem, and fourth ventricle through the foramen magnum in the setting of myelomeningocele.
- Commonly associated with hydrocephalus (90%) and syringomyelia (20-95%).
- Other variable abnormalities associated with CMII include dysplasia of the corpus callosum, enlargement of mass intermedia, abnormalities of the white and gray matter, hippocampal dysplasia, elongation of the pons, beaking of the midbrain tectum, and defects of the falx and tentorium.
- Symptomatic CMII is the leading cause of death in pts less than 2 y old with myelomeningocele.
- Symptoms differ relative to age of onset, with neonates presenting as a neurologic emergency and older children presenting with more subtle findings of hyporeflexia weakness, or headache.

Etiology

- Pathophysiology not completely understood

- Multiple theories of embryologic origin, including primary malformation or secondary abnormalities related to altered cerebral spinal fluid dynamics

Usual Treatment

- Early closure of myelomeningocele, usually within 72 h after birth, proven to cause upward movement of hindbrain herniation but does not appear to prevent lifetime occurrence of symptomatic CMII.
- Treatment of hydrocephalus more important than surgical decompression to prevent CMII symptoms, and, in any symptomatic CMII pt, physician must first rule out and treat hydrocephalus.
- Of pts, 20% will need surgical treatment including brainstem decompression via posterior cervical laminectomies at all involved segments and possible duraplasty.
- Unlike Chiari Malformation type I, suboccipital craniotomy often not necessary due to already enlarged foramen magnum.
- Intrauterine myelomeningocele repair shows evolving benefits of possibly preventing CMII and decreasing overall severity.
- Intrinsic brainstem dysfunction cannot be treated surgically.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CNS	Hindbrain herniation Hydrocephalus Increased ICP	Occipital headache, weak cry, irritability	Opisthotonus posturing, quadriparesis, hypotonia, ataxia, and down-beat nystagmus Increased head circumference; bulging anterior fontanelle	MRI Shunt evaluation
SPINAL CORD	Cervical myelopathy Syringomyelia	Altered dexterity, handwriting change, unable to provide self-care	Upper extremity weakness, spasticity, ataxia, hand muscle atrophy, scoliosis, back pain, and loss of sensorimotor function	MRI, SEP/MEP
HEENT	Cranial nerve X dysfunction	Stridor (must evaluate to rule out neurologic cause; do not assume viral upper-respiratory infection)	Vocal cord paralysis; altered gag reflex	Laryngoscopy (direct; fiberoptic)
RESP	PEAC Central and obstructive breathing disorders	Snoring, apnea, daytime sleepiness, daytime attention deficit	Cyanosis, bradycardia, death during painful experience	PSG, ENT evaluation
GI	Neurogenic dysphagia, aspiration, nutritional deficiency	Choking, regurgitation, prolonged feeding time, weight loss	Muscle wasting; aspiration pneumonia	Swallow study; direct laryngoscopy

Key References: Messing-Jünger M, Röhrig A: Primary and secondary management of the Chiari II malformation in children with myelomeningocele. *Childs Nerv Syst* 29(9):1553-1562, 2013; McClain CD, Soriano SG, Rockoff MA: Pediatric neurosurgical anesthesia. In Cote CJ, Lerman J, Anderson B, editors: *A practice of anesthesia for infants and children*, ed 5, Philadelphia, 2013, Elsevier, pp 510-532.

Perioperative Implications

Preoperative Preparation

- Present for multitude of procedures including myelomeningocele closure, cerebral spinal fluid shunt placement, shunt revision, brainstem decompression, and scoliosis correction.
- Assess for latex allergy (increased risk in pts with myelomeningocele).
- Preoperative evaluation including assessment of concurrent comorbidities and active medications, as well as a focused physical exam to assess level of consciousness, motor and sensory function, cranial nerves, and ICP.
- Laboratory evaluation including hemoglobin, type and cross, and electrolytes.
- Review of relevant imaging studies, including CT scans, ECG, and chest plain films.

Monitoring

- Standard ASA monitors with invasive arterial blood pressure.
- Monitor urinary output.
- If central venous access deemed necessary due to pt disease or limited peripheral IV access, avoid neck veins to decrease risk of altering cerebral blood flow or venous drainage.

- Consider TIVA to facilitate neurophysiologic monitoring when applicable.

Airway

- Endotracheal intubation required (naso, oro, or tracheostomy)
- Limit neck flexion during intubation in pts with brainstem compression

Preinduction/Induction

- Use opioids carefully because of their respiratory depressant effects and possible deleterious effects on ICP.
- Sedatives including midazolam appear safe under the direct supervision of the anesthesiologist.
- IV induction with propofol preferred over inhalational induction.
- Avoid hypoxia, hypercarbia, and coughing to limit further increases in ICP when elevated.
- Position pts with myelomeningocele carefully to avoid direct pressure on neural tube defect.

Maintenance

- General anesthesia with controlled ventilation.
- Consider TIVA for benefits of improved neurophysiologic monitoring and favorable effects on cerebral blood flow, cerebral metabolic rate, and ICP.

- When in the prone position, pt's head is rigidly fixed with pins or placed in a cerebellar head frame when pinning contraindicated. Avoid excess neck flexion.
- Carefully evaluate blood loss and management of IV fluid administration, and attempt to limit cerebral edema while maintaining hemodynamic stability.
- Carefully manage known exaggerated heat loss secondary to the disproportionately large head of the neonate.

Extubation

- Prepare for the need for postop mechanical ventilation.
- During emergence and extubation, carefully avoid large fluctuations in ICP and blood pressure.

Postoperative Period

- 24-h monitoring in ICU

Anticipated Problems/Concerns

- Problems related to surgery including hemorrhage, infection, vascular injury, nerve injury, and persistent symptoms of brainstem compression
- Apnea or airway obstruction due to respiratory center or cranial nerve damage
- Secondary cervical instability or kyphosis following cervical laminectomy
- Venous air embolism

Aspiration, Perioperative

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Risk

- Risk of aspiration: Approximately 3 per 10,000 anesthetics, 11 per 10,000 emergency and/or after-hours cases, and 29 per 10,000 emergency cases in ASA IV and V pts
- Loss of protective reflexes and sphincter function
- Obstructed or abnormal GI motility
- Increased gastric fluid volume; decreased pH
- Inadequate anesthesia leading to coughing and straining during airway manipulation or induction
- Trauma, emergency/night surgery, pregnancy, difficult airway, advanced age, long-standing diabetes mellitus, pain, analgesics, and ASA status >2
- Obesity: not an independent risk factor

Perioperative Risks

- Mortality after aspiration: 5%; higher if ASA >2 or if mechanical ventilation required for >24 h after the aspiration event

Worry About

- Of pts who aspirated, 20% had no risk factor: of these, 66% had difficult intubation
- Rapid-sequence induction may have deleterious effects on heart rate and blood pressure
- Clinical worsening may be delayed up to 24 h after the inciting aspiration event

Overview

- Prevention of aspiration best because there is no definitive treatment.
- Vast majority of pts with risk factor(s) do not aspirate.
- Consider aspiration in differential diagnosis of bronchospasm with hypoxemia.

Etiology

- Loss of protective reflexes: Sedation, neuromuscular disorders/relaxants, and altered mental status

- Obstructed or abnormal motility: Achalasia, gastroparesis, pain, and opioids
- Increased GI contents: Bleeding, obstruction, and feeds

Usual Treatment

- Suctioning or bronchoscopy if obstructing particles present
- Lavage and steroids not helpful; surfactant investigational
- Empiric antibiotics: Consider if pt is compromised, with fulminant course, or suspected high bacterial load due to bowel obstruction

Assessment Points

System	Effect	Assessment by History	Physical Examination	Test
HEENT	Awake intubation in difficult airway; cricoid pressure may distort anatomy and obstruct ventilation	Hx difficult airway, head and neck surgery/radiation	Airway exam	X-ray, CT scan, OR records as available
CV	Rapid-sequence intubation may lead to ischemia with tachycardia, hypertension/hypotension, or myocardial depression	Anginal Sx, exercise intolerance, Hx CHF, CAD age, sex, risk factors	S3, rales; displaced PMI	ECG and ECHO in selected patients
RESP	Rapid-sequence intubation may lead to bronchospasm	Hx pulm disease, wheezing with URI, smoking	Wheezing; prolonged expiratory phase	CXR, continuous pulse oximetry
GI	Abnormal sphincters, motility, acidity	Hx peptic ulcer disease, reflux Sx, diabetes, scleroderma, bowel obstruction	Abdominal exam for distention	
NEURO	Increased ICP leads to vomiting; depressed protective reflexes; muscle weakness		Neurologic exam	

Key References: Marik P: Aspiration pneumonitis and aspiration pneumonia, *N Engl J Med* 344(9):665–671, 2001; Tasch MD, Langeron O: Aspiration prevention and prophylaxis: preoperative considerations. In Hagberg CA, editor: *Benumof and Hagberg's airway management*, ed 3, Philadelphia, 2013, Elsevier, pp 265–279.