Occipital Encephalocele

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Risk

- Most frequent type of encephalocele in North America and Western Europe
- Incidence in North America: 1:3000–10,000 live births
- Worse prognosis compared with frontal encephaloceles

Perioperative Risks

- Seizures
- Hydrocephalus
- Cranial nerve deficits
- Poor feeding
- Spasticity
- Blindness

Worry About

- Positioning concerns
- Coexisting congenital anomalies, especially renal and facial
- Difficult airway
- Elevated ICP
- Body temperature changes
- IV access
- Blood loss
- Hemodynamic disturbances

Overview

- Herniation of brain, meninges, and/or CSF through a skull defect (cranium bifidum) that is usually covered with skin
- One of the three most common neural tube defects
- Cranial nerve deficits, poor sucking and feeding, spasticity, blindness, seizures, or developmental delay
- May be associated with hind-brain anomaly (Chiari III malformation), in which herniating occipital/cerebellar tissues distort the posterior fossa structures
- Associated conditions include:
  - Hydrocephalus (30–50%)
  - Corpus colossal abnormalities (18%)
  - Cerebral dysgenesis (13%)
  - Seizures
  - Meckel Gruber syndrome
  - Occipital encephalocele
  - Microcephaly
  - Microphthalmia
  - Polycystic kidneys
  - Ambiguous genitalia
  - Polydactyly
  - Cleft lip and palate
  - Other malformations

Etiology

- Unknown
- Isolated encephaloceles showing no familial inheritance
- Possibly a syndrome with an autosomal recessive pattern or inheritance
- Usually obvious at birth, with many diagnosed prenatally using fetal US or fetal MRI

Usual Treatment

- Requires surgical management, usually in infancy, by a pediatric neurosurgeon directed by the type of neural tissue protruding from the skull.
- Gliotic and malformed neural elements can be amputated.
- Staged repair may be necessary to return normal tissue to the cranial vault.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Difficult airway Difficult positioning Cleft lip/palate Microphthalmia Microcephaly	Obstruction and location Mass location Poor suckling Small head	Chin and tongue size Lips and mouth Small eyes	 CT, MRI
RESP	Respiratory failure Aspiration	Dyspnea Dyspnea	Tachypnea, desaturations Tachypnea, desaturations	CXR, CT, ABG CXR
CV	Congenital anomalies	Sweating, poor feeding, lethargy	Murmur, cyanosis	ECG, ECHO
GI	Dysphagia Aspiration	Emesis Respiratory failure	Feeding tube Tachypnea, desaturations	Swallow study Swallow study, CXR
CNS	Seizures Increased ICP	Irritability, lethargy Irritability, lethargy	Cushing triad	EEG, LP CT, MRI, LP
RENAL	Polycystic kidneys			US, lytes
MS	Polydactyly	Extra fingers and toes		

Key References: Alexiou GA, Sfakianos G, Prodromou N: Diagnosis and management of cephaloceles, *J Craniofac Surg* 21:1581–1582, 2010; Mahajan C, Rath GP, Dash HH, et al.: Perioperative management of children with encephalocele: an institutional experience, *J Neurosurg Anesthesiol* 23(4):352–356, 2011.

Perioperative Implications

Preoperative Preparation

- Thorough Hx and PE
- Availability blood products

Monitoring

- Arterial line.
- Two large IVs; consider a central line if IV access is inadequate.
- UP.
- Temperature monitoring.
- Prone positioning.
- Blood glucose.

Airway

- Potentially difficult mask ventilation and intubation; have difficult intubation equipment immediately available
- Supine positioning can be difficult; may need to secure the airway in a lateral position or with the pt elevated and heavily padded to avoid compression of the sac.

Preinduction/Induction

- Maintain spontaneous ventilation.
- Mask induction may be preferred.

Maintenance

- Sudden CSF leaks can cause severe hemodynamic instability and electrolyte imbalances.
- Sudden ICP changes can occur and can result in cardiac arrest.
- Maintenance of normothermia is imperative.
- Pneumocephalus can cause delayed wakening.

Extubation

- Extubate awake in the OR versus intubating in the ICU

Postoperative Period

- Monitor for hydrocephalus and increased ICP.
- Maintain normothermia.
- Maintain glucose control.
- Structural derangement of the respiratory control center can contribute to apnea.

Anticipated Problems/Concerns

- Ongoing concern for hydrocephalus; the pt may require a VP shunt.
- Ongoing concern for seizures; consider prophylaxis with anticonvulsants.
- Developmental delay is more common with sacs containing brain tissue; expect worse prognosis for occipital encephaloceles.
- Brainstem dysfunction can occur resulting in apnea and/or gastric aspiration due to a lack of gag reflex.
- Metabolic and electrolyte disturbances, hemodynamic instability, and septic shock can contribute to nonresuscitable cardiac arrest in these pts.