- Ear infections/hearing loss
- Choanal atresia

## Overview

- Clefts can be cranial or facial in origin and are defined as a gap in soft tissue and/or bone.
- Significant variability in severity exists with craniofacial clefts and subsequent implications for surgical and anesthetic management.
  - Can be unilateral or bilateral with multiple clefts occurring at the same time.
  - Can be as minor as a cutaneous manifestation or as extreme as skeletal malformations.
- Classically, clefts were described and classified by Paul Tessier; his classification uses facial meridians to describe the location of clefts, with a clock-face analogy numbering (0–14) to describe the locations with midline as (0). Facial clefts are numbered from 0–7, and cranial clefts from 8–14, in a counter clockwise rotation.

## Etiology

- Exact etiology is unknown, but most cases have associations to familial, genetic, and environmental factors.
- Possible theories include failure of the fusion process, failure of mesodermal growth/penetration, and/or disorder of migration of neural crest cells.
- Drug exposures in utero, including anticonvulsants, Accutane, and methotrexate, are associated with clefts.
- Infectious causes and exposure in utero.

## Usual Treatment

- Definitive treatment with surgical repair with surgical type depending on craniofacial cleft manifestation and severity of disease

## Assessment Points

| System | Effect | Assessment by Hx | PE | Test |
|---|---|---|---|---|
| HEENT | Craniofacial cleft<br>Craniofacial abnormality<br>Hearing loss<br>Chronic ear infections<br>Difficult airway | Fevers, previous history of difficult airway | Thorough airway exam | |
| RESP | Airway obstruction | Dyspnea, poor feeding and weight gain | Tachypnea | CXR |
| CV | ASD/VSD | Dyspnea<br>Lethargy | Heart murmur | ECG, ECHO |
| GI | Poor oral intake | Poor weight gain, malnourished, dry mucous membranes, poor skin turgor | Low weight for age<br>Dry mucous membranes<br>Sunken fontanels | Chemistry, serum albumin |
| CNS | Elevated ICP | Irritability, lethargy<br>Headache, seizures, vomiting | Papilledema<br>Bulging fontanels | Head CT, MRI |
| HEME | Anemia<br>Coagulopathy | Age, nutrition status<br>Bleeding gums, infections<br>Easy bruisability<br>Fatigue | | CBC<br>Type and screen/cross<br>PT, PTT |

**Key References:** Tessier P: Anatomical classification of facial, cranio-facial and latero-facial clefts, *J Maxillofac Surg* 4(2):69–92, 1976; Jackson O, Basta M, Sonnad S, Stricker P, Larossa D, Fiadjoe J: Perioperative risk factors for adverse airway events in patients undergoing cleft palate repair, *Cleft Palate Craniofac J* 50(3):330–336, 2013.

## Perioperative Implications

### Preoperative Preparation

- Assess for significant associated comorbidities such as elevated ICP and congenital cardiac disease.
- Particular attention to the airway examination.
- Assess for volume status because clefts can make feeding difficult, leading to malnourished and dehydrated pts.
- Communicate and coordinate with surgical teams on repair and surgical concerns.
- Weigh risks and benefits of premedication with anxiolytics because of risk of airway obstruction.

### Monitoring

- Consider large vascular access for blood loss in extensive surgical repairs.
- Consider arterial line for hemodynamic monitoring in complex repairs.

### Airway

- Conduct thorough airway examination and prepare in advance for a difficult airway.
- Mask ventilation and intubation can be difficult depending on severity and location of the craniofacial cleft.
- Airway devices including video scopes and flexible fiberoptic and laryngeal mask airways should be available.
- Consider presence of ENT surgeons during induction/airway management, especially if nasal passages are not patent or mouth opening is severely limited.

### Induction

- Consider maintaining spontaneous ventilation during induction with either inhalation agents or IV agents.
- Difficult mask ventilation is uncommon, but difficult intubation can be anticipated based on location of the cleft.

### Maintenance

- Be vigilant for accidental extubation and ETT obstruction or damage during the surgical repair.
- Monitor for blood loss and volume status and consider transfusion in complex craniofacial cleft repairs.

### Extubation

- Ensure throat packs are removed (if used) and the oropharynx is clear of blood.
- Elevated risk for adverse airway events, including laryngospasm, bronchospasm, obstruction, accidental extubation, and subsequent hypoxia.

### Adjuvants

- Infraorbital nerve blocks can provide postop analgesia for cleft lip repairs.
- Multimodal analgesia for postop pain control.

### Postoperative Period

- Pts may require ICU monitoring postoperative, particularly pts with syndromic diagnosis.

## Anticipated Problems/Concerns

- Monitor carefully for airway obstruction and postop bleeding in first 12–24 h.
- Monitor for blood loss and volume status.

# Craniosynostosis

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## Risk

- Occurs in 1:2000-2500 live births.
- May be spontaneous, syndromic, or familial, but it most commonly presents as an isolated abnormality; however, it can present as a component of a syndrome or genetic disorder in 15–40% of pts.
- Familiarity with associated head shapes can allow bedside diagnosis and differentiation from positional plagiocephaly. Rarely a CT scan will be required to differentiate plagiocephaly from synostosis.
- Craniosynostosis is diagnosed within the first months of life in most infants, but it can also present later.
- Surgical intervention should be performed during infancy, preferably in the first 6 mo of postnatal life, to prevent the further progression of the deformity and possible complications associated with increased ICP.

## Perioperative Risks

- Precipitous hemorrhage can occur with inadvertent dural venous tears or disruption of large emissary veins.
- Venous air embolism: The incidence is high (83%); however, the vast majority is clinically silent and not associated with hemodynamic compromise. Incidence is much lower with endoscopic procedures (8%).
- Intracranial hypertension, which is usually diagnosed by ophthalmic examination (papilledema) or CT scan or by clinical symptoms such as headache in older children, is more commonly seen in syndromic craniosynostosis involving multiple sutures (47%).
- Avoid hypothermia.

- Position pt carefully. Supply eye protection because syndromic forms of craniosynostosis can have proptosis that may prevent full eyelid closure. Infants undergoing procedures in the prone position are placed in a horseshoe headrest.

## Worry About

- Significant and rapid blood loss intraop
- Associated anomalies (if syndromic)
- Potential difficult airway (ventilation and possibly intubation)
- Monitoring for venous air embolism
- Management of increased ICP
- Difficulties upon extubation: OSA and significant airway edema

## Overview

- A disorder of skull development that occurs because of the abnormal fusion of one or more cranial sutures, the observed deformity relates to the affected sutures.
- Virchow (1851) was the first to describe the arrest of skull growth that occurs in a direction perpendicular to the affected suture.
- Most commonly associated syndromes are Pfeiffer, Apert, and Crouzon.
- Untreated craniosynostosis can lead to elevated ICP and disturbances in intellectual and neurologic development.
- Bilateral coronal sutures are more commonly affected, and there is often associated extremity anomalies (syndactyly) and midface hypoplasia.
- From both a cosmetic and neurodevelopmental perspective, optimal outcomes are achieved when these procedures are performed before 1 y of age, and earlier surgical intervention may translate to a less extensive operation.

## Etiology

- Biomechanical forces and genetically determined local expression of growth factors have been implicated. Spontaneous mutation of a syndromic gene is possible. The fibroblast growth-receptor pathway is most frequently involved.
- It can be inherited in an autosomal recessive or dominant pattern.

## Usual Treatment

- Surgical intervention is usually done during infancy, preferably in the first 6 mo of postnatal life.
- Surgery is usually performed in a specialized hospital.
- Principles of surgical intervention are not only to excise the fused suture but also to attempt to normalize the calvarial shape.
- It is important to differentiate from posterior plagiocephaly, which is not associated with a risk of head-growth restriction or increased ICP, and the treatment is nonsurgical, usually with position changes.
- Current surgical techniques include open calvarial reconstruction, minimally invasive strip craniectomy with the use of a postop molding helmet, minimally invasive strip craniectomy with spring implantation, and cranial distraction.

## Assessment Points

| System | Effect | Assessment by Hx | PE | Test |
|---|---|---|---|---|
| AIRWAY | Diff mask vent and/or intubation | Hx or previous mask ventilation or intubation | Facial symmetry<br>Size of mandible<br>Neck range of motion | Neck films may be indicated (Apert may have cervical fusion) |
| RESP | OSA | Apnea during sleep, snoring | Noisy breathing from the upper airway | Polysomnogram (apnea-hypopnea index)<br>Overnight pulse oximetry<br>Room air $O_2$ saturation |
| CV | CHD (ASD, VSD, tetralogy of Fallot) | Bottle feeds >30 min<br>Diaphoresis with feeds<br>Failure to thrive | Murmur | ECHO |
| MS | Diff IV access<br>Diff a-line access | | Syndactyly (Apert)<br>Fused elbows (Pfeiffer) | |
| CNS | Increased ICP | Irritable, vomiting, somnolence (if acute) | Papilledema | Ophthalmology exam, VEPS |
| HEME | Anemia (nadir at 3 mo of age) | | | Preoperative Hct, type/cross |

**Key References:** Haas T, Fries D, Velik-Salchner C, et al.: Fibrinogen in craniosynostosis surgery, *Anesth Analg* 106(3):725–731, 2008; Stricker PA, Fiadjoe JE: Anesthesia for craniofacial surgery in infancy, *Anesthesiol Clin* 32(1):215–235, 2014.

## Perioperative Implications

### Preoperative Preparation

- Laboratory testing should include CBC, type and screen, and coagulation profile. In open procedures also cross matching of blood.
- Midface hypoplasia and retrusion may cause OSA and postop airway obstruction such that postop mechanical ventilation may be indicated.
- Prepare for potential difficult airway. Children with severe airway obstruction may present with a tracheostomy.

### Monitoring

- At least two peripheral IVs, 22 gauge or larger when possible
- Arterial line for immediate detection of hypotension and frequent blood sampling
- Central venous catheter: Some centers routinely insert it for complex cranial vault reconstruction procedures to measure central venous pressure to guide fluid and transfusion therapy; others reserve their use for children in whom adequate peripheral access is difficult to obtain
- Precordial Doppler for detection of venous air embolism

### Airway

- Have several airway devices available, including LMA and videolaryngoscope if necessary.
- Consider using fiberoptic intubation for anticipated difficult airways, and even having a surgeon available in case an emergency tracheostomy is needed.
- Some anesthesiologists place a nasotracheal tube for infants in the prone position as a way to secure the airway better.

### Positioning

- Craniofacial procedures can be lengthy, and careful attention to proper positioning is important.
- If pt is supine, some surgeons prefer the eyes not to be taped closed because they are within the surgical field. Ophthalmic ointment should be applied in these cases.
- If pt is prone, the head must be positioned on the horseshoe rest, ensuring no pressure on the orbits or other pressure points.

### Induction

- An inhaled induction of anesthesia is most commonly performed. Children with signs or symptoms of significant acute ICP elevation may benefit from an IV anesthetic induction. Most elevation in ICP is chronic and asymptomatic.

### Maintenance

- Administer general inhalational or IV maintenance with muscle relaxant.
- Maintain constant vigilance to guide fluid administration. This includes direct observation of the surgical field and directing close attention to the invasive blood pressure and waveform, central venous pressure, response to fluid challenges, urine output, hemoglobin measurements and blood gas assessments, and systolic pressure variation.
- Use active warming techniques such as forced-air convection blankets, circulating warm water mattresses, overhead radiant lights, and fluid warmers.
- Mannitol may be required before calvarial removal if increased ICP.
- Use isotonic solutions for maintenance IV fluids.

### Extubation

- Most pts can be safely extubated at the end of the procedure.
- Infants who may require postop intubation and mechanical ventilation include those in the prone position for lengthy procedures with significant facial and tongue swelling and those with syndromic craniosynostosis who have significant preop obstructive sleep apnea.

### Adjuvants

- Using antifibrinolytic drugs such as tranexamic acid has been shown effective in reducing blood loss and periop transfusion requirements.
- The off-label use of recombinant activated factor VII has been described as a rescue measure, but it has been associated with significant thrombotic complications; therefore, its use should be reserved for life-threatening situations in which all other methods of achieving hemostasis failed.
- Cell salvage may allow for reduction of transfusion.
- The use of preop recombinant erythropoietin has been described; however, its current use seems infrequent and limited to select pts. When administered, this technique is often combined with acute preop normovolemic hemodilution and other techniques to maximize efficacy.
- Blood products should be immediately available in the operating room at the beginning of the procedure.

### Postoperative Period

- All children after open vault reconstruction are admitted to the ICU postop.
- Postop pain is usually not severe, and intermittent opioids together with acetaminophen provide satisfactory postop analgesia.