Assessment Points

Differentiation Between Croup (Laryngotracheobronchitis) and Epiglottitis

	Croup	Epiglottitis
Age	3 mo–3 y	1–7 y
Onset	Gradual	More rapid (usually <24 h)
Fever	Low grade	High
Cough	Characteristic barking	None
Sore throat	Occasional	Frequently severe
Posture	Any	Frequently sitting forward, mouth open, drooling
Airway sound	Inspiratory stridor	Inspiratory stridor
Voice	Normal	Muffled
Appearance	Nontoxic	Toxic
Seasonality	Peak winter, epidemic	Year-round

Key References: Jenkins I, Saunders M: Infections of the airway, *Paediatr Anaesth* 19(Suppl 1):118–130, 2009; Tibballs J Watson T: Symptoms and signs differentiating croup and epiglottis, *J Paediatr Child Health* 47:77-82, 2011.

Perioperative Implications

Airway

- Airway support with good mask fit and positive pressure ventilation can generally overcome obstruction from swelling of airway.
- Identification of larynx is generally routine, but a tracheal tube 0.5–1.0 mm diameter smaller than usual may necessitate having available extra-long or micro-laryngeal tracheal tubes.
- Tracheotomy rarely needed as therapy for these pts with current management and reserved only for unusual cases.

Induction

- Induction common when IV access has already been obtained.

Anticipated Problems/Concerns

- Symptomatic pts who require intubation of trachea need tubes 0.5–1.0 mm smaller in diameter than pts without croup.
- Pts who require tracheal intubation usually require sedative management to tolerate ventilation; this is often followed for development of leaks around the tracheal tube as a sign of improvement of edema; most pts improve within 2–4 d. When leak is present at 20–25 cm H_2O of pressure, extubation can be considered; complicated cases and pts with prolonged courses may benefit from examination of airway in operating room at time of extubation.
- Although a viral illness, some pts may acquire bacterial superinfection of airways and require antibiotic therapy.

Crouzon Syndrome

Geoff Frawley

Risk

- Represents approximately 4.8% of cases of craniosynostosis at birth.
- Birth prevalence of 1.6:100,000 births.
- Estimated prevalence in general population of Europe is 0.9:100,000.
- No race predilection.

Perioperative Risks

- Difficult BMV, difficult intubation, massive blood loss, arterial gas embolism

Worry About

- Difficult airway
- Intraop blood loss
- Inadvertent dural sinus injury
- Postextubation subglottic edema
- External facial fixation devices

Overview

- Crouzon syndrome is an autosomal dominant disorder characterized by craniosynostosis causing secondary alterations of the facial bones and facial structure.
- Common features include hypertelorism, exophthalmos and external strabismus, parrot beak nose, short upper lip, hypoplastic maxilla, and a relative mandibular prognathism.
- Synonyms: Craniofacial dysostosis type II, FGFR deficiency.

Etiology

- Due to mutation in FGFR2 gene on chromosome 10.
- Normal function of FGFRs is to restrain limb growth. FGFR mutations are hypermorphic, causing excessive cranial bone formation.
- Inherited in autosomal dominant fashion, but de novo mutations account for 50% of cases.
- High penetrance but variable expressivity.
- Male to female preponderance of 3:1.

Usual Treatment

- In neonatal period tracheostomy for UAO or ventriculoperitoneal shunt for hydrocephalus may be required.
- Posterior vault expansion may be carried out in first 6 mo to achieve cranial decompression of intracranial venous hypertension.
- Fronto-orbital advancement to protect orbitae from subluxation at 6–12 mo.
- Complex hypoplasia of cranial vault, orbits, and midface may require frontofacial advancement (Le Fort III osteotomy) and/or distraction osteogenesis with application of RED frame.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	UAO secondary to septal deviation, choanal stenosis, and nasopharyngeal narrowing Ocular hypertelorism and proptosis Cleft lip or palate rarely	Sleep apnea snoring, daytime somnolence	Hypoplastic maxilla, relative mandibular prognathism Mallampati scoring difficult in toddlers Exposure keratopathy of cornea	3D cranial CT planning Polysomnography (sleep studies) or overnight oximetry
CV	FGFs involved in cardiac cushion proliferation and valvulogenesis	Exercise tolerance or arrhythmias	PDA or ASD murmurs	ECHO
RESP	Choanal atresia	OSA snoring		Nasoendoscopy
HEME	No known bleeding diatheses			
RENAL	Nil reported			
ORTHO	Cervical fusion (18%), usually C2-C3, occasionally C3-C4, C5-C6 Scoliosis Subluxation of the radial heads Ankylosis of the elbows.		Reduced range of movement of cervical spine	Cervical spine lateral x-ray or craniocervical CT
CNS	Chiari malformation Cerebellar tonsil herniation (73%) Progressive hydrocephalus (47%). Intracranial hypertension	Headaches Seizures	Gait disturbance Paresthesia	Craniocervical CT and MRI
PNS	Mild to moderate mental retardation	Developmental delay		
MS	Usually normal		Metacarpophalangeal shortening	Hand x-ray

Key References: Stricker PA, Shaw TL, Desouza DG, et al.: Blood loss, replacement and associated morbidity in infants and children undergoing craniofacial surgery, *Paed Anesth* 20(2):150–159, 2010; Hughes C, Thomas K, Johnson D, et al.: Anesthesia for surgery related to craniosynostosis: a review. Part 2, *Paediatr Anaesth* 23(1):22–27, 2013.

Perioperative Implications

Preoperative Preparation
- Caution with sedative premedication in presence of OSA or intracranial hypertension
- Cooperation limited in younger age groups

Monitoring
- Invasive pressure monitoring warranted for major craniofacial surgery

Airway
- Difficult BMV and intubation
- Mandibular hypoplasia more prominent postmaxillary advancement and may worsen glottic view

Induction
- Upper airway obstruction common on gas induction may require NPA

Maintenance
- Protection of orbits and corneas
- Reliable venous access mandatory
- Risk of excessive bleeding, dural tears, and gas embolism during vault surgery

Adjuvants
- Antifibrinolytics, surgical hemostasis with topical agents, and cell salvage have been described to reduce transfusion requirements

Extubation
- RED frame impedes access to upper airway. Wire cutters and spanner required to be with pt at all times.

Postoperative Period
- Increased risk of upper airway obstruction on emergence and in PACU
- Difficult BMV with RED frame in situ

Anticipated Problems/Concerns
- Multiple surgeries in first year of life to reduce risk of hydrocephalus or intellectual impairment
- Upper airway obstruction with postop facial edema; may require ICU/PACU care overnight

Cryptococcus Infection

Pierre Moine

Risk
- In general population: 0.4–1.3 cases per 100,000; AIDS pts: 2–7 cases per 1000.
- Impact of cryptococcosis: Approximately 625,000 deaths each year worldwide.
- Underlying immunocompromised conditions and risk factors: AIDS, systemic lupus erythematous, prolonged treatment with corticosteroids, organ transplantation, advanced malignancy, hematologic malignancy, diabetes, sarcoidosis, cirrhosis, idiopathic CD4 lymphocytopenia, or use of immune-modifying monoclonal antibodies (alemtuzumab, infliximab, etanercept, or adalimumab).
- More and more pts with cryptococcosis are described as immunocompetent.

Perioperative Risks
- Respiratory insufficiency, severe ARDS
- Elevated ICP

Worry About
- Underlying immunocompromised, genetic, or other conditions

Overview
- Systemic mycosis and third most prevalent disease in HIV-positive individuals
- *Cryptococcus neoformans*/*C. gattii* typically infect immunocompromised persons, essentially HIV and transplant-recipient pts, but also pts who do not have underlying HIV infection or are not transplant recipients. These pts tend to have a delayed diagnosis compared with the HIV and transplant groups and are remarkably currently the highest risk group for mortality in resource-available countries
- Wide range of clinical presentations from asymptomatic respiratory colonization to dissemination of infection into any organ. In severely immunosuppressed pts, involvement of multiple body sites. Common sites for infection are the lungs and CNS
- Pulmonary cryptococcosis/cryptococcal pneumonia: Mainly underestimated, not often recognized, multiple clinical presentations—asymptomatic solitary or multiple nodules, lobar infiltrates, interstitial infiltrates, cavities, endobronchial colonization or masses, mediastinal adenopathy, hilar adenopathy, miliary pattern, cavitary lesions, or pleural effusions/empyema, pneumothorax, and life-threatening pneumonia with ARDS
- Cryptococcal meningitis/meningoencephalitis: Primary life-threatening infection, most frequent and most severe form. Mortality rate approximately 12%. Other CNS clinical manifestations: Cryptococcomas (abscesses) of brain, spinal cord granuloma, chronic dementia (from hydrocephalus)
- Laryngeal cryptococcosis: hoarseness, cough, or acute airway obstruction

Etiology
- Seven species are described in the *C. neoformans* species complex: *C. neoformans*, *C. deneoformans*, *C. gattii*, *C. bacillisporus*, *C. deuterogattii*, *C. tetragattii*, and *C. decagattii*. *C. neoformans* and *C. gattii* are the agents highlighted in cryptococcal meningitis fungal infection. Other species, *C. laurentii* and *C. albidus*, are reported.
- Cryptococcus species are encapsulated heterobasidiomycetous fungi. The presence of a polysaccharide capsule is considered one of the reasons for the virulence of the yeast, increasing its invasiveness, pathogenicity, and conferring resistance to the host.
- Cryptococcus infection occurs by the inhalation of infectious cells and is considered a primary pulmonary infection, which may lead to a disseminated infection, with a special predilection for invading the CNS causing meningitis, encephalitis, or meningoencephalitis.
- Skin/subcutaneous, ophthalmic, bone, and prostatic disease also occur. Any pt with a diagnosis of cryptococcosis should be investigated for disseminated disease.