Worry About

- Hypoglycemia in 0, 1, 3, and 6
- Cardiac function in 2, 3, 4, and 7
- Rhabdomyolysis leading to renal failure in 5 and 7
- Coagulopathy and cirrhosis in 4
- Renal function and platelet function in 1

Overview

- Glucose is the primary energy substrate for the majority of the tissues in the body (heart and brain can function well on ketones, but most other organs cannot). When a person is fasting or between meals, serum glucose (and thus intracellular glucose) is maintained primarily by the breakdown of glycogen in the liver.
- The GSDs result from mutations in genes involved in glycogen synthesis and breakdown, leading to the inability to use glycogen (in one way or another—usually tissue-limited).
- Ideally the NPO time should be limited, and for those associated with hypoglycemia, glucose should be monitored and periop glucose-containing fluids administered.
- The consequences of these mutations depend on (1) the tissue where the enzyme is expressed and (2) the effect of the mutation on the pathway. Whether it prevents the synthesis of glycogen, debranching of glycogen, phosphorylation/dephosphorylation of glucose, or other steps in glycogen breakdown.

Etiology

- Caused by homozygous mutations in enzymes involved in the synthesis or breakdown of glycogen.
- Autosomal recessive trait with a few case reports of compound heterozygotes who have mutations in two different enzymes in the pathway.
- Each heterozygous parent has a 50% chance of passing on the gene.
- Long-term prognosis depends on the enzyme affected (type of GSD): 0, Muscle cramping and occasional growth failure; 1, growth failure; 2, infantile—death by 2, juvenile—death by 30, adult—sometimes heart failure or sudden death in adulthood; 3, myopathy; 4, FTT and death by 10; 5, exercise-induced cramps; 6, growth retardation; 7, growth retardation; 9, delayed motor development and growth retardation; 11, pretty normal; 12, exercise intolerance; 13, exercise intolerance and muscle pain with aging.

Usual Treatment

- Several GSDs can be treated with liver transplant with excellent outcomes (1, 3, and 4).
- Several are managed with enteral cornstarch supplementation (1, 3, and 6).
- 2 (Pompe) may be treated with recombinant human α-glucosidase enzyme replacement therapy.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	In 2 (Pompe) macroglossia can occur		Limited ability to visualize glottic opening	
CV	Cardiomyopathy	Determine GSD type		ECG, ECHO
RESP	Decreased resp reserve (in 2)	Sleep apnea	Assess for sleep apnea	Sleep study
GI	Coagulopathy/liver failure in 1		Jaundice and hepatomegaly with a very firm liver raising concern for cirrhosis	LFTs and coagulation studies (PT/PTT/INR)
HEME	Platelet dysfunction (leading to decreased platelets) in 1	Easy bleeding and bruising	Ecchymosis	CBC with plts and plt function tests if possible
RENAL	Renal dysfunction in 1			Comprehensive metabolic panel (CMP) with calculated GFR
MS	Truncal obesity in 1 and 3		BMI	

Key References: Stuart G, Ahmad N: Perioperative care of children with inherited metabolic disorders, *Contin Educ Anaesth Crit Care Pain* 11(2):62–68, 2010; Fleisher L: *Anesthesia and Uncommon Diseases,* ed 5, Philadelphia, PA, 2006, Elsevier, pp 177.

Perioperative Implications

Preoperative Preparation

- Fasting should be minimized to the extent possible.
- Blood glucose should be monitored and periop glucose-containing fluids administered.
- The appropriate tests for GSD type should be performed; where risks are present, they should be managed appropriately (e.g., for decreased platelets or platelet function, the administration of platelets).
- For altered liver function, be aware that some medications may have altered metabolism.
- Conscious sedation is also possible; concern for sleep apnea syndrome.
- Assess individual pt based on systems approach and review relevant studies.
- For pts with decreased cardiac function, avoid myocardial depressants when possible.
- Avoid tourniqueting with type V; some evidence points to an association of MH with type V, so it is recommended to avoid MH-triggering medications (or to do an IVCT test).
- Similarly, there are case reports suggesting avoidance of propofol and sevofluorane in 2.
- There are theoretical preferences for some approaches, but the key factors of a successful outcome are attention to anesthetic technique and close monitoring.

Monitoring

- Standard ASA monitors
- For cases associated with hypoglycemia, glucose monitoring every 30 min (can use CGM)
- For cases associated with rhabdomyolysis leading to renal failure, consider intraop CKs.
- Bispectral index may be misleading in GSD 1.

Airway

- Anticipate difficulty intubating in 2 if macroglossia is present.
- Other diseases with skeletal muscle involvement can predispose pts to upper airway obstruction.

Induction

- As above in preparation

Maintenance

- Careful positioning in types 5 and 7 to avoid rhabdomyolysis

Postoperative Period

- Continuous pulse oximetry due to high incidence of sleep apnea in 2
- Continued glucose-containing fluids and monitoring for 0, 1, 3, 6

Anticipated Problems/Concerns

- Hypoglycemia in 0, 1, 3, and 6
- Cardiac dysfunction in 2, 3, 4, and 7
- Rhabdomyolysis leading to renal failure in 5 and 7
- Coagulopathy and cirrhosis in 4
- Renal function and platelet function in 1

Goldenhar Syndrome

Zulfiqar Ahmed

Risk

- Incidence: 1:5500 live births.
- Second most common facial birth defect after cleft lip and palate.
- Extracraniofacial anomalies can range from one anomaly (13%) to multiple affected organ systems (42%). No gender or side predominance was detected. Central nervous system, cardiac, and skeletal anomalies each occurred in more than 10% of cases. Surgical correction usually takes place in severe cases.
- Cleft palate occurs in 25% of pts

Perioperative Risks

- Difficulties with airway management
- Possible need for tracheostomy
- High risk of anesthesia overdose in premature and low-weight pts

Worry About

- Stabilizing the heart rate.
- Difficulty with airway management.
- Ensuring good mask fit. There are many sizes to choose from, and the mask must fit prior to induction. Mask may have to be changed after induction for better fit. The degree of inflation of the facemask cuff may be adjusted in order to ensure an appropriate seal.
- IV access.
- Associated anomalies, such as cardiac and or cervical spine malformation that may influence decisions.
- Cancelation of procedure owing to inability to intubate.
- Increased severity of microsomia. Pruzansky classification type III is associated with increased intubation difficulties.

Overview

- Craniofacial microsomia, also known as HFM or oculoauriculovertebral spectrum.
- Bilateral microsomia can occur.
- CNS, cardiac, and skeletal anomalies (expanded HFM spectrum) may occur
- Pulmonary, gastrointestinal, and renal deformities are less common.
- The majority of associated heart defects involve the outflow tract or septum. The increased frequency of cardiac anomalies with this condition suggests that abnormal development of the neural crest may result in both HFM and conotruncal heart defects.
- Children with HFM may have fused or hemivertebrae, resulting in limitation of neck flexion and extension and increasing the difficulty of intubation.
- There are positive correlations between the number of involved abnormal components and the degree of difficulty in visualizing the larynx in pts with both bilateral and unilateral microtia.
- Bilateral mandibular and auricular malformations increase the risk of difficult intubation.

Etiology

- Rare congenital abnormality
- May result from chromosomal abnormality or disrupted blood flow to the head in utero

Usual Treatment

- Removal of preauricular skin tags
- Remodeling, especially in the presence of orbital dystopia
- Orthodontic treatment
- Ear reconstruction
- Maxillary repositioning (Le Fort 1), mandibular advancement and soft tissue augmentation
- Mandibular distraction osteogenesis as required to facilitate subsequent intubations
- Rib grafting as required

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Mandibular hypoplasia	Facial asymmetry	Micrognathia, ear tags, OMENS	CT/MRI
CV	Conotruncal malformation	Dyspnea/poor feeding/delayed growth	Murmur	ECHO
MS	Fused or hem vertebrae	Neuromuscular changes	Limited neck flexion/extension	CT/MRI

Key References: Nargozian C, Ririe DG, Bennun RD, et al.: Hemifacial microsomia: anatomical prediction of difficult intubation, *Paediatr Anaesth* 9(5):393–398, 1999; Walker RM, Ellswood J: The management of difficult intubation in children, *Paediatr Anaesth* 19(Suppl 1):77–87, 2009.

Perioperative Implications

Preoperative Preparation

- Conduct craniofacial assessment OMENS:
  - Orbit: Orbital distortion.
  - Mandible: Mandibular hypoplasia.
  - Ear: Microtia, periauricular tags.
  - Facial nerve: Facial muscle hypoplasia.
  - Soft tissue: Hypoplasia or absence of the parotid gland and masticatory muscles (temporalis, masseter).
- Review detailed history with surgeon, radiologist, and parents/guardians.
- Be prepared to call for help early.
- Assemble ear/nose/throat team in case rigid bronchoscopy or tracheostomy is required.
- Determine correct ET tube size and depth, as changing the tube or having too short a tube can lead to complications.
- Discuss and plan all approaches and backup plans.
- Check and prepare all the instruments before bringing the pt to the OR.
- Determine severity of mandibular hypoplasia in radiologic reports to estimate the degree of difficulty of intubation.
- Plan for difficult IV access. Presence of preexisting IV may facilitate concurrent IV and inhalational induction.

Monitoring

- Arterial line may be required in the presence of cardiac or pulmonary morbidity.

Airway

- Make your first attempt the best attempt.
- Assess and plan mask ventilation.
- For pts who are difficult to ventilate, oral, nasopharyngeal, or LMA insertion can improve ventilation.
- Avoid higher peak pressure as much as possible.
- If direct laryngoscopy fails, quickly go to video-assisted technique.
- Conventional laryngoscopy with a flat curved blade, such as a Macintosh, will be less helpful in a pt with micrognathia, as even the normal-sized tongue cannot be compressed adequately into the mandibular space to reveal the laryngeal structures.
- Paraglossal approach: Use a narrow, low-profile, straight-bladed laryngoscope in a paraglossal manner. Advance the blade in the space between the tongue and the lateral pharyngeal wall or tonsillar fossa.
- Lateral approach: The straight axis is shorter and insertion of the ET may be aided by a stylet or use of a gum elastic bougie. This approach is also called the retromolar approach, far lateral approach, and right molar approach.
- LMA can be used as a conduit for fiberoptic insertion of an ET tube.
- Confirm LMA placement with a fiberoptic examination.
- Use an antisialagogue to decrease airway secretions.
- It may be appropriate to leave the LMA in situ so as to minimize manipulation. Have a clear plan to remove it if necessary. Can load up two ET tubes back to back on the fiberoptic scope to facilitate removal of the LMA.
- Use humidified oxygen and steroid prophylactically after difficult intubation to minimize edema.
- Multiple approaches for laryngoscopy may be required.
- Direct laryngoscope with various blade sizes; McCoy and Sward laryngoscope blades.
- Video-assisted airway devices: Fiberoptic scope, Glidescope, Truview, etc.
- Supraglottic airways; LMA classic, Proseal LMA, iGel and COPA, etc.
- Hopkins rod rigid bronchoscope.
- Use Tegaderm/Vaseline gauze or hold the mask with both hands to improve seal.

Postoperative Period

- Prolonged monitoring is recommended, especially when opioids are used to manage pain.
- Monitor pts for hemorrhage, regurgitation of gastric contents, hypoxic events.

Anticipated Problems/Concerns

- Temporomandibular joint malformation may make jaw thrust difficult.

Gonorrhea

Seth Eisdorfer

Risk

- The prevalence of gonorrhea is decreasing, with 106.1:100,000 as of 2013.
- Most common in people ages 15–24 y, in large urban areas, and among people with low socioeconomic status and/or low levels of education.
- Incidence higher in men; prevalence higher in women.

Overview

- Sexually transmitted disease.
- High incidence of coexisting chlamydial infection.
- Local infection: Purulent, profuse urethral discharge and possible epididymitis, prostatitis, or proctitis in men. It is often asymptomatic in women, but may have cervical discharge, vaginitis, salpingitis, or proctitis. Ascending infection may lead to PID.
- Disseminated infection: Fever/rash, tenosynovitis/arthritis (common), conjunctivitis (usually from autoinoculation), possible myopericarditis, and toxic hepatitis or perihepatitis (Fitz-Hugh-Curtis syndrome), rarely with endocarditis or meningitis.

Etiology

- *Neisseria gonorrhoeae*: Gram-negative intracellular diplococcus, usually found inside polymorphonucleocytes.
- Humans only natural hosts for *N. gonorrhoeae*.

Usual Treatment

- Dx gold standard involves the isolation of the organism by culture, testing for antimicrobial resistance.
- Test for other STDs, including syphilis and HIV; test partners as well.