

Case Study

- 6 y/o male, 50 lbs, 46 inches
- PMH: Duchenne's Muscular Dystrophy, Sickle Cell Anemia
- PSH: none
- Current Medications: Prednisone, Procainamide, Folic Acid, Hydroxyurea, Hydrocodone, Metoclopramide
- Procedure: T/A



Tonsillectomy and Adenoidectomy

Indications for tonsillectomy:

- 1) chronic or recurrent acute tonsillitis
- 2) Peritonsillar abscess
- 3) Tonsillar hyperplasia
- 4) OSA

Tonsillar hyperplasia may lead to chronic airway obstruction resulting in sleep apnea, carbon dioxide retention, cor pulmonale, failure to thrive, speech abnormalities, and swallowing disorders



Events leading to Cor Pulmonale

Long standing hypoxemia and hypercarbia→

Increased airway resistance→

Pulmonary arteriolar/venule constriction→

Pulmonary artery hypertension→

Right sided heart failure

- EKG shows RVH
- Chest X-ray shows cardiomegaly



Preoperative Evaluation

- Often have frequent infections – inquire about antibiotics, antihistamines, or other medications
- Symptoms of acute URI – postpone until symptoms have abated, usually 7-14 days
- A history of sleep apnea should be sought
- Physical examination – observe for the presence of audible respirations, mouth breathing, nasal quality of the speech, and chest retractions should be noted
- Oropharynx should be inspected for evaluation of tonsillar size to determine the ease of mask ventilation and tracheal intubation
- Measurement of coagulation parameters and hematocrit



Anesthetic Management

- Render the child unconscious in the most atraumatic manner possible
- Provide the surgeon with optimal operating conditions
- Establish IV access
- Provide rapid emergence so the patient is awake and able to protect the recently instrumented airway
- Antisialagogue – minimizes secretions in the operative field
- Avoid sedative premedication in children with OSA, intermittent obstruction, or very large tonsils
- Supraglottic area may be packed with petroleum gauze
- Consider cuffed endotracheal tube



Surgical Considerations

- Positioning – supine, shoulder roll, head extended (Rose position); surgeon at head of table
- Unique considerations – use of armored ETT prevents compression of tube by mouth gag; tube should be secured to lower lip in midline
- Surgical time– 30 to 60 minutes
- EBL– 25 to 200 mL
- Closing considerations – avoid hypercapnia on emergence to prevent vasodilation and resultant bleeding
- Postop care – lateral position; gentle suctioning



Complications

- Emesis – 30-65%
 1. Considering decompressing the stomach with an orogastric tube
 2. Zofran 0.10-0.15 mg/kg with or without dexamethasone
- Dehydration
- Postoperative hemorrhage – 75% occurs within 6 hours of surgery
 1. If re-exploration is necessary, patient is considered to have a full stomach
 2. monitor for orthostatic changes resulting from decreases in vascular volume
 3. Observe for frequent swallowing!
- Severe pain after tonsillectomy
- Acute postoperative pulmonary edema – encountered when airway obstruction is suddenly relieved
- Laryngospasm – treatment is with 100% oxygen, jaw thrust, CPAP

Procainamide

- Class IA antidysrhythmic
- Lengthens both the action potential duration and the effective refractory period reflecting sodium channel inhibition and lengthening of repolarization owing to potassium channel blockade
- Treatment of ventricular tachydysrhythmias
- Rarely used during anesthesia because of the propensity to produce hypotension



Metoclopramide

- Dopamine antagonist
- Increases lower esophageal sphincter tone and stimulates motility of the upper GI tract
- Gastric hydrogen ion secretion is not altered
- Blocks dopamine receptors in the CNS---produces extrapyramidal side effects
- Antagonism of dopamine agonist effects on the CTZ contribute to an anti-emetic effect
- Clinical uses: 1) preoperative decrease of gastric fluid volume, 2) production of antiemetic effect, 3) treatment of gastroparesis, 4) symptomatic treatment of GERD
- Inhibitory effect on plasma cholinesterase activity

Hydroxyurea

- Reduces the rate of painful attacks in sickle cell disease – breaks down cells that are prone to sickle, as well as increasing fetal hemoglobin content
- Fetal hemoglobin – able bind oxygen with greater affinity than the adult form
- Side effects: myelosuppression manifesting as leukopenia, megaloblastic anemia, and occasionally thrombocytopenia



Folic Acid

- Children with sickle cell disease will take 1mg of folic acid for life
- Folic acid produces healthy red blood cells and prevents anemia



Hydrocodone

- Vicodin
- Treatment of chronic pain



Prednisone

- Synthetic corticosteroid administered for its glucocorticoid effects
- Recommended when an anti-inflammatory effect is desired
- Low mineralocorticoid potency---limits sodium and water retention
- Side effects: suppression of the HPA axis, osteoporosis, increased susceptibility to bacterial or fungal infection
- Suppression of HPA axis – release of cortisol in response to stress, such as that produced by surgery, is blunted or does not occur
- corticosteroid supplementation in the perioperative period: Cortisol 25mg IV at the induction of anesthesia followed by a continuous infusion of cortisol, 100mg, during the following 24 hours
- Alternative glucocorticoid supplementation – for minor surgical stress, the daily cortisol secretion rate and static plasma cortisol measurements suggest that the glucocorticoid requirement is about 25mg Cortisol; if the postoperative course is uncomplicated, the patient can be returned the next day to the prior glucocorticoid maintenance dose

Sickle Cell Anemia

- Definition: hereditary hemolytic anemia resulting from abnormally formed hemoglobin (Hb S), resulting in the sickling & destruction of red blood cells.
- The sickling of RBCs is aggravated by the following conditions:
 - arterial hypoxemia
 - acidosis
 - intravascular dehydration
 - circulatory stasis
 - hyperthermia
 - hypothermia



Sickle Cell Anemia

- Usual Rx: oxygen, hydration w/IV fluid, pain control, sometimes exchange transfusions
- Pre-Op: Patients can be in acute crisis
 - Vaso-occlusive: causes pain, can result in infarction of organs & thrombosis
 - Aplastic: red cell production in marrow is either exhausted or prevented, resulting in marked anemia
 - Splenic sequestration: usually in children <6 yrs, resulting sudden entrapment of blood in the spleen, causing significant hypotension.
 - These pts are predisposed to anemia and infection

Sickle Cell Anemia

- Pre-op (con't)
 - These pts may have pulmonary complications such as:
 - Acute chest syndrome- a vaso-occlusive crisis resulting in fever, chest pain, dyspnea, tachypnea, and cough. This can lead to pulmonary HTN and sometimes even death.
 - Sickle cell lung disease: generalized pulmonary fibrosis & hypoxemia leading to cor pulmonale.



Sickle Cell Anemia

- These pts are predisposed to cholelithiasis, peptic ulcer disease, ischemic colitis, leg ulcers priapism, neurologic deficits, nephrotic syndrome, and inability to concentrate urine.



Sickle Cell Anemia

- What to do (Pre-op)
 - Optimize medical condition by treating acute crisis prior to surgery
 - Supplemental oxygen to optimize systemic oxygenation & reduce sickling
 - Optimize circulatory flow & hydration status w/ IVF
 - Check Hgb/Hct & consider exchange transfusions to decrease blood viscosity & increase oxygen carrying capacity (goal Hct 35-40% w/ 40-50% normal hemoglobin)
 - Check BUN/ Cr to screen for dehydration & renal abnormalities secondary to infarction

Sickle Cell Anemia

- What to do (Pre-op) con't
 - Control all infections; many of these pts have infected their spleens & are particularly susceptible to encapsulated organisms.
 - Manage pain in pts w/ vaso- occlusive crisis
 - Avoid pre-op meds that result in decreased respiration



Sickle Cell Anemia

- What to do (Intra-op)
 - Maintain FiO₂, even during regional anesthesia
 - Maintain adequate hydration status (CVP monitor may be useful in certain instances)
 - Keep pt normotensive to maintain perfusion & circulatory flow
 - Avoid hyper/hypothermia
 - Avoid acidosis
 - Tourniquets & vascular clamps may produce ischemia & promote sickling.

Sickle Cell Anemia

- Management:
 - Goal is to avoid sickling
 - Use regional technique if possible
 - Promote supplemental oxygen and IV fluids
 - Use warming blanket to prevent vasoconstriction; must avoid hyperthermia also, as it promotes sickling
 - Use pressors to support BP if necessary.



Sickle Cell Anemia

- Management (Post-op)
 - Similar goals as intra-op mgt: maintain oxygenation & perfusion, manage pain, prevent hypothermia
 - Close monitoring in post-op period; special attention to pulmonary complications, as acute chest syndrome in the post-op period is a significant cause of morbidity and mortality
 - Good pulmonary toilet is important
 - Early ambulation
 - Pain management in these pts can be quite difficult as many are narcotic-tolerant & respiratory depression should be avoided.

Duchenne Muscular Dystrophy

- Most common form of muscular dystrophy.
- 1 in 3500 male births.
- X-linked inherited type thought to be caused by deletion of a segment of DNA or a single-gene defect on short arm of the X chromosome.
- A protein called dystrophin is absent, which anchors the actin cytoskeleton of skeletal muscle fibers to the basement membrane through a membrane glycoprotein complex.

Clinical Manifestations

- Muscle weakness begins in pelvic girdle causing a “waddling” gait
- Diminished muscle strength produces ineffective cough, resulting in retention of secretions, pneumonia, and death.
- Smooth muscle dysfunction may cause megacolon, volvulus, cramping pain, and malabsorption in GI tract

- Cardiac involvement is as high as 95%.
- Chronic heart failure may occur in 50% of children.
- Treatment for cardiac dysfunction includes ACE inhibitors and B-adrenergic blockers.



- Pulmonary functions is compromised due to kyphoscoliosis.
- By age 12, most kids are confined to a wheelchair
- Only 25% live to age 21.



Anesthesia Concerns

- Preoperative premedication with sedatives or opioids is best avoided due to increased risk of aspiration.
- Intraoperative positioning may be complicated due to kyphoscoliosis or flexion contractures of extremities or neck.
- Increased risk for perioperative aspiration of gastric contents due to degeneration of GI smooth muscle with hypomotility and delayed gastric emptying.

Anesthesia Concerns

- Response to nondepolarizing agents, such as vecuronium, atracurium, and mivacurium, have been normal. Careful titration of short- or intermediate-duration NDMRs and may have prolonged recovery.
- Little data on response to anticholinesterases.
- Several reports of cardiac arrest after administration of succinylcholine, often associated with hyperkalemia, so Succ should be avoided.
- Controversy over whether they are susceptible to malignant hyperthermia.

Anesthesia Concerns

- Volatile anesthetics could damage the muscle membrane and cause rhabdomyolysis by releasing calcium from the sarcoplasmic reticulum.
- Volatile anesthetics may cause marked respiratory and circulatory depression.
- Regional or local anesthesia may be preferred in these patients.

- Some patients with vital capacities less than 30% may require postop mechanical ventilation.

