Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Cervical adenopathy Pharyngitis Enlarged tonsils with pseudomembranous appearance Otitis media Sinusitis Laryngeal edema	Difficulty swallowing Sore throat Headache Facial pain, ear pain Difficulty talking or breathing	Lymphadenopathy Tonsillar enlargement Spotty, erythematous tonsils Otitis media Tenderness over sinuses Drooling; tripod position Difficulty of breathing	CT Hx and physical exam Serologic test for EBV
RESP	Lung nodules Pleural effusions Hilar and mediastinal adenopathy	SOB Orthopnea	Decreased breath sounds Crackles, egophony	CXR CT
CV	HF	SOB, tires easily Edema	New murmur, crackles Pitting edema	ECHO ECG
GI	Liver dysfunction, Bowel obstruction, Bowel perforation, Tumors anywhere in GI tract	N/V Abdominal pain and discomfort Distention Swelling, tenderness over graft site	Jaundice Abdominal distention Tenderness over graft Rebound tenderness	LFTs Abdominal x-ray, CT US
RENAL	Renal insufficiency or failure	Decreased UO Swelling	Pitting edema, Crackles	BUN, Cr, lytes
ID	Mononucleosis syndrome Generalized lymphadenopathy Sepsis	Fatigue, fever	Elevated temperature	CBC, serology for EBV
CNS	Brain tumors	Headache LOC Seizure	Stupor, coma Seizure	CT, MRI

Key References: Friedberg JW, Aster JC: Epidemiology, clinical manifestations, and diagnosis of post-transplant lymphoproliferative disorders. In Freedman AS, Brennan DC, editors: Waltham, MA, 2015, UpToDate. www.uptodate.com/contents/epidemiology-clinical-manifestations-and-diagnosis-of-post-transplant-lymphoproliferative-disorders. (Accessed 01.06.16); Pinyavat T: Posttransplant lymphoproliferative disorder. In Houck PJ, editor: *Handbook of pediatric anesthesia*, New York, 2015, McGraw Hill, pp 166–168.

Perioperative Implications

Preoperative Preparation

- Difficult airway techniques; consider GE reflux precautions.
- Evaluate the need for blood products and specific antibiotics.
- Evaluate the function of the transplanted organs.
- Consider stress-dose steroids if receiving steroids.
- Consider side effects of the immunosuppressant medications.

Monitoring

- Consider invasive monitoring in the event of organ failure or mediastinal mass.
- Consider ICP monitor as indicated for CNS involvement.

Airway

- Consider awake fiberoptic techniques if upper airway edema or masses or mediastinal masses are present.

Preinduction/Induction

- Induction agents should be chosen based on organ function. Cyclosporine can potentiate the effect of succinylcholine.
- A mediastinal mass can compress the aorta and SVC, leading to significant hypotension if pt is supine. Consider sitting or semisitting induction.
- Consider lower extremity for volume resuscitation if a large mediastinal mass is present.

Maintenance

- Keep the pt breathing spontaneously in case of significant airway obstruction.
- If a mediastinal mass is present, keep the pt in semisitting position and turn to lateral or prone position if hemodynamics become compromised.

Extubation

- Risk of airway obstruction if airway is manipulated during surgery

Postoperative Period

- Airway edema can become a problem.
- Continue stress-dose steroids.

Anticipated Problems/Concerns

- Airway obstruction and hemodynamic compromise
- Dysfunction of transplanted organs
- Mental status change or increased ICP in CNS involvement

Prader-Willi Syndrome

Navil F. Sethna

Risk

- Prevalence: 1:25,000
- Incidence: 1:10,000-15,000
- Racial prevalence: None
- Gender predominance: Similar frequency in both sexes and all races
- Most common syndromic form of obesity, affects 350,000–400,000 individuals worldwide
- Annual death rate is 3% versus 1% in the general population, primarily due to respiratory arrest

Perioperative Risks

- Infantile hypotonia, hypoventilation, and breathing difficulty
- Potential for difficult intubation and aspiration risk
- Worsening of obstructive/central sleep apnea and abn ventilatory responses to hypoxia, hypercarbia, and bronchospasm
- Bradycardia, ventricular arrhythmias (PVCs)
- Postop resp insufficiency
- Potential risk of rhabdomyolysis with succinylcholine
- Aberrant thermoregulation: Hyperthermia and MHS-like syndrome
- Glucose intolerance or DM

Worry About

- Abn short and restricted neck mobility, limited mouth opening and difficult intubation
- Poor vascular access and intraop positioning
- Systemic and pulm Htn, conduction defects, RBBB cor pulmonale, and dilated cardiomyopathy
- Restrictive lung disease (obesity, kyphoscoliosis) and reactive airways

Overview

- Presents in two stages: Infantile central hypotonia, FTT, and delayed developmental milestones. Childhood stage presents with obesity (BMI >97th percentile in a child and ≥30% in an adult), skeletal abn (dysmorphic, short stature, short hands and feet, scoliosis), hypogonadism, and hypothalamic dysfunction.
- Restrictive pulmonary disease results from muscle weakness, obesity, and kyphoscoliosis. It starts in early childhood and is present in 80–90% of pts >30 y of age.
- CV system: Htn in 17–32%; myocardial hypotrophic hypokinetic syndrome.
- Central thermodysregulation: May develop hyperpyrexia.
- Cognitive problems: Mild to moderate mental retardation. Mean IQ in 60s–70s; some individuals have normal intelligence.
- Behavior problems of oppositional behavior, emotional lability, aggressive and violent behavior; obsession with food and compulsion to eat. Psychosis found in 5–10% of adults.
- High threshold for pain.

Etiology

- A complex genetic disorder; paternally inherited via 15q11–q13 deletion (65–70%), maternal uniparental disomy 15 (20–30%) and imprinting defect (1–3%). GH deficiency.

Usual Treatment

- Early intervention and education: Physical, occupational, speech, and behavioral therapies
- Weight and dietary management; low-calorie diet and regular physical therapy
- GH replacement therapy
- Nighttime CPAP for severe OSA

Assessment Points

System	Effect	Assessment by Hx	PE	Test
MS (Craniofacial)	Facial dysmorphia, poor mask fit	Snoring, nystagmus, viscous and sticky saliva	Dental crowding and caries Micrognathia, short neck with limited movement	Imaging scans
CV	Htn Pulm Htn Cor pulmonale Cardiomyopathy	Headache Exertional and at rest Dyspnea, exertional intolerance Dyspnea, exertional intolerance	High diastolic BP Lung rales Tachypnea, orthopnea, systemic venous congestion, gallop sounds	ECG, CXR, renal function ECG, CXR, ECHO ECG, CXR, ECHO
RESP	Alveolar hypoventilation Increased airway responsiveness Increased work of breathing Upper airway obstruction	Snoring and interrupted sleep, daytime somnolence, exertional dyspnea, wheezing	Fatigue, limited upper airway access, short neck, limited mobility of neck	PFT, room air ABG CXR Polysomnography for severe OSA Difficult airway scoring
ENDO (Diabetes I or II)	Increased risk for CVS, CHF and autonomic dysfunction	Hyperglycemia/hypoglycemia, osmotic diuresis	Dysfunction of CVS, renal and peripheral neuropathy	Periop blood glucose Other test related to end-organ involvement

Key References: Angulo MA, Butler MG, Cataletto ME: Prader-Willi syndrome: a review of clinical, genetic, and endocrine findings, *J Endocrinol Invest* 38(12):1249–1263, 2015; Lirk PC, Keller J, Colvin J, et al.: Anaesthetic management of the Prader-Willi syndrome, *Eur J Anaesthesiol* 21(10):831–833, 2004.

Perioperative Implications

Preoperative Preparation

- Owing to obsessive hyperphagia, only a well-supervised pt should be considered NPO.
- Oral metoclopramide and cimetidine.
- Assess airways difficulty, CV and pulm status, and blood glucose.
- Effective premedication to ensure a cooperative pt during awake/sedated intubation and induction of GA.

Monitoring

- Standard ASA monitors. Consider direct intra-arterial BP measurement if the noninvasive cuff does not fit. Continuous temp monitoring for instability.
- Frequent checking of ABGs, UO, and central venous or pulm artery pressure for major surgery.

Airway

- Elective awake intubation if difficult airway is anticipated; increasing neck circumference, a Mallampati score of ≥3, micrognathia, and limited mouth opening

Induction

- Gastric regurgitation due to delayed gastric emptying and hiatal hernia.
- Be prepared to manage a situation where ventilation and/or intubation are not possible. The degree of obesity is only one factor among others that makes visualization of the glottis problematic.
- Semisitting position improves FRC and preoxygenation.
- Slow IV induction with propofol and remifentanil or fentanyl with cisatracurium to facilitate intubation.

Maintenance

- Sevoflurane or desflurane with remifentanil infusion and cisatracurium. These inhaled agents are least soluble and allow rapid recovery from GA. No specific drug or combination is recommended; the aim is rapid emergence. Avoid long-acting opioids and substitute with IV NSAIDs and/or acetaminophen.
- Regional anesthetic techniques are desirable alone or to supplement GA and provide postop analgesia to reduce the need for opioids.

Extubation

- Decision is dictated by the severity of obesity, assoc risks such as OSA, and the extent of the surgical procedure. Early tracheal extubation is desirable.

Adjuvants

- Hydrophilic drugs (e.g., muscle relaxants are calculated by lean body mass). Lipophilic drugs (e.g., fentanyl) are calculated in mg/kg of body weight.

Postoperative Period

- Severe obesity is associated with more atelectasis during, immediately after, and for 24 h following GA as compared with nonobese pts. CPAP or BiPAP may be necessary to maintain patent airways, particularly during sleep and for those with severe OSA. These pts are highly sensitive to opioid-induced resp depression.

Anticipated Problems/Concerns

- Monitor for OSA and alveolar hypoventilation in ICU/PACU. Monitor hyperglycemia and/or hypoglycemia, hyperthermia, and arrhythmias.
- Early ambulation and thromboembolic precautions.

Preeclampsia

Shobana Bharadwaj | Lester C. Chua

Risk

- 2–8% of all pregnancies
- Nulliparous, or multiparous with previous preeclampsia/eclampsia Hx, advanced maternal age
- Increased with Hx of obesity, chronic htn, diabetes, renal disease, SLE, thrombophilia

Perioperative Risks

- Increased risk of fetoplacental or maternal deterioration necessitating (often operative) delivery.
- Increased risk of fetal death.
- Preeclampsia and eclampsia account for about 15% of maternal and perinatal deaths.

Worry About

- Hypertensive crisis leading to intracerebral bleed or LV failure.
- Increased interstitial volume leading to edema.
- Maternal hypotension producing placental hypoperfusion.
- Renal dysfunction progressing to acute renal failure.
- Thrombocytopenia may contraindicate regional anesthetic.
- Eclampsia (seizure in a severely preeclamptic pt) necessitating difficult tracheal intubation.
- Placental abruption.
- Risks associated with preterm delivery.

Overview

- Early onset (<34 wk gestation): High rate of recurrence, strong genetic component, high risk of adverse outcome.
- Late onset (<34 wk gestation): Higher incidence, maternal metabolic predisposition.
- Marked by Htn, proteinuria (spot urine protein/Cr ratio >0.3).
- Maternal hyperdynamic state with diastolic dysfunction, leading to acute cardiorespiratory deterioration.
- Proteinuria: Sign of deteriorating renal function and widespread endothelial damage.