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.

- Edema: Increasing total body water, proteinuria, Htn; lead to increasing interstitial edema and decreasing intravascular volume.
- Hematologic: Widespread endothelial damage often leads to thrombocytopenia.
- Epigastric/RUQ pain: Ominous sign of liver subcapsular edema and possible rupture. Delivery should be urgently effected.
- HELLP: Poor fetoplacental prognostic sign.
- Headache: Seizure may be impending.

Etiology

- Heterogenous disease of unknown etiology.
- Immune maladaptation causing placental angiogenesis dysfunction.
- Imbalance in circulating mediators of vascular tone and response (e.g., thromboxane vs. prostacyclin) from endothelial damage.
- Systemic inflammatory response from placental oxidative stress.
- Microangiopathy leading to endothelial change, platelet consumption, hemolysis.

Usual Treatment

- Prevention with daily low-dose aspirin beginning in second trimester has had limited success.
- Delivery becomes cure.
- In-hospital therapy: Antihypertensives, seizure prophylaxis with magnesium sulfate (therapeutic blood levels of 5–7 mg/dL), and support of maternal perfusion.
- Neuraxial analgesia for labor: Reduces catecholamine response to pain, increasing placental perfusion.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Edema		Airway exam	
CV	Systemic vasoconstriction Decreased intravascular volume Diastolic dysfunction		BP JVD Rales UO	ECG CXR ECHO Hct
RESP	Pulm edema	Dyspnea Chest discomfort	Rales/rhonchi Cyanosis	SaO_2 CXR ECHO
GI	Hepatic subcapsular edema	Epigastric/RUQ pain	Enlarged liver edge	LFT
HEME	Thrombocytopenia DIC	Easy bruising	Petechiae, gingival bleed Spontaneous hemorrhage	Platelet count Coag panel/TEG
RENAL	Increased capillary permeability	Weight gain	Nondependent edema Oliguria	Urinary protein Cr clearance Serum uric acid
CNS	Seizure Intracerebral hemorrhage Cerebral edema/posterior reversible encephalopathy syndrome	Headache Blurred vision Seizure	Retinal edema CNS exam Deep tendon reflexes	CT scan/MRI
OB	Decreased placental perfusion Placental abruption	 Vaginal bleeding	Fetal growth restriction FHR (lack of variability, late decelerations or bradycardia)	FHR monitoring BPP Doppler velocimetry FHR monitoring

Key Reference: Gogarten W: Preeclampsia and anaesthesia. *Curr Opin Anaesthesiol* 22(3):347–351, 2009; Chestnut DH, Wong CA, Tsen LC, et al. (editors): *Chestnut's obstetric anesthesia: principles and practice*, 5th edition, Philadelphia, PA, 2014, Elsevier.

Perioperative Implications

Antepartum Management

- Optimize maternal perfusion while lowering systemic diastolic BP to <110 mm Hg.
- Ensure therapeutic blood magnesium sulfate level.
- Maternal and fetal surveillance.
- Decisions on the timing/route of delivery; corticosteroids for fetal lung maturity if preterm.

Monitoring

- Consider intraarterial cath for extremes of BP.
- Consider transthoracic ECHO, CVP, or PA cath for oliguria/pulm edema.
- Fetal heart monitoring.

Airway

- Often difficult secondary to edema.
- Prepare for emergent airway.

Preinduction/Induction

- Neuraxial analgesia/anesthesia induces venodilation. Maintain maternal perfusion with judicious use of intravascular volume and (often) small, incremental prn doses of IV low-dose phenylephrine (by bolus or infusion) or IV ephedrine.
- Rapid-sequence induction of anesthesia, titrating IV antihypertensive drugs or rapid-acting opioids to blunt pressure response to intubation.

Maintenance

- Hemorrhage at delivery may lead to dramatic hypotension.
- Avoid ergot alkaloids.
- Titrate antihypertensive agents.

Extubation

- Extubate awake; control pressure response

Adjuvants

- Magnesium sulfate for seizure prophylaxis and increased UBF; IV antihypertensive drugs (most often hydralazine, labetalol, nifedipine, or nicardipine antepartum); rarely other inotropic support if demonstrable LV dysfunction; $PGE_1/PGF_{2\alpha}$ for uterine atony

Postoperative Period

- Risk for developing pulm edema, sustained Htn, stroke, VTE, seizures
- Effective postcesarean analgesia beneficial in BP control

Anticipated Problems/Concerns

- Maternal Htn causes maternal morbidity/mortality; maternal hypotension causes fetoplacental hypoperfusion.
- Eclampsia, intracranial hemorrhage associated with CNS residua, hepatic subcapsular hematoma.