- Under normal circumstances, central chemoreceptors in the brain stem detect decreased pH and respond by increasing the respiratory rate; in OHS, the ventilatory response is blunted.
- Episodes of nighttime acidosis due to sleep apnea lead to renal compensation with retention of bicarbonate.
- Nighttime apnea leads to hypoxia, causing hypoxic pulm vasoconstriction. This vasoconstriction, in turn, leads to pulm Htn as well as right ventricular failure and remodeling.
- The chronically low O_2 levels in the blood also lead to increased release of erythropoietin, causing polycythemia.

Usual Treatment

- Weight loss through diet and exercise (which is rarely successful) or bariatric surgery
- NIPPV
- Uvulopalatopharyngoplasty
- Tracheostomy

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Difficult airway access	Snoring	Poor visualization	X-ray of neck may be helpful
CV	Biventricular failure CAD	Dyspnea, poor exercise tolerance Angina, poor exercise tolerance	Venous engorgement, S_3 and S_4, dyspnea	ECG, ECHO, CXR ECG, stress ECHO, angio
RESP	Hypoventilation	Dyspnea, sleeping upright Poor exercise tolerance	Rapid shallow breathing, cyanosis	ABGs, Hct, CXR

Key References: Olson A, Zwillich C: The obesity hypoventilation syndrome, *Am J Med* 118:948–956, 2005; Chau EH, Lam D, Wong J: Obesity hypoventilation syndrome: a review of epidemiology, pathophysiology, and perioperative considerations, *Anesthesiology* 117(1):188–205, 2012.

Perioperative Implications

Preoperative Preparation

- Consider pulm function tests with bronchodilator to determine whether a reversible restrictive component exists.
- Assess for bronchitis/pneumonia, which can be improved with pulm toilet and antibiotic therapy.
- Assess myocardial and volume status using a central venous catheter or PA cath.
- Consider maintaining pt in a semisitting position to avoid sudden shifts of volume to central circulation and pulm edema.

Monitoring

- Consider an arterial line for frequent monitoring of ABGs.
- Maintain adequate respiratory volumes and pressures.
- Consider PAC or transesophageal ECHO to monitor filling volumes and wall motion.

Airway

- Awake intubation frequently required.
- Laryngoscopy can sometimes be facilitated by elevating the shoulders and head on a bolster.

Induction

- Do not expect to ventilate pt adequately by mask. Establish airway first.

Maintenance

- Pts may have to remain in reverse Trendelenburg position to allow adequate ventilation.

Extubation

- Perform with pt in sitting position without residual sedation.
- Ensure adequate tidal volume and consider preop levels of CO_2 retention in making decision to extubate, as a normal CO_2 level may not be attainable.

Adjuvants

- Regional anesthesia only if pt is able to maintain ventilation
- Residual sedation or narcosis may preclude early extubation.

Postoperative Period

- Consider prophylaxis for thromboembolism; early ambulation may minimize pulm and thromboembolic complications.
- Pts may be extremely sensitive to the respiratory depressant effects of benzodiazepines and narcotics.

Anticipated Problems/Concerns

- All those problems associated with morbid obesity apply to Pickwickian pts.
- Early ambulation may minimize pulm and thromboembolic complications.
- Prepare the pt for a possibly prolonged course of postop mechanical ventilation, especially after upper abdominal procedures.

Pierre Robin Sequence

Charles B. Cauldwell

Risk

- 1:8500-14,000 live births; PRS nonsyndromic in about 40% of cases.
- Syndromic PRS most commonly associated with Stickler, velocardiofacial, and Treacher-Collins syndromes.

Perioperative Risks

- Chronic airway obstruction, respiratory distress, hypoxia.
- Malnutrition due to feeding difficulties, GE reflux.
- Congenital heart defects with syndromic PRS.

Worry About

- Airway obstruction
- Difficult intubation

Overview

- An anomaly consisting of micrognathia (or retrognathia), glossoptosis (posterior displacement of the tongue), and varying degrees of airway obstruction as well as feeding difficulties; cleft palate may be present but is not diagnostic.
- Airway obstruction, which may be multilevel, can lead to hypoxia and /or cyanosis.
- Feeding problems associated with malnutrition, reflux, and aspiration.
- In nonsyndromic PRS, if hypoxia and malnutrition are overcome, obstruction may improve secondary to mandibular growth by the time the pt reaches several mo of age.

Etiology

- Nonsyndromic PRS associated with a defect in gene SOX9, a chondrogenic regulator.
- Syndromic PRS associated with multiple genetic syndromes involving multiple defective genes.

Usual Treatment

- Prone positioning, lavage feeding may treat 70% successfully.
- Nasopharyngeal airway is the next level of intervention,
- Glossopexy or mandibular distraction osteogenesis to relieve airway obstruction and allow growth.
- Tracheostomy for multilevel airway obstruction or failure of previous surgery.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Airway obstruction	Respiratory distress OSA	Micrognathia Stridor, retractions	Sleep study CT, MRI
CV	Congenital defects Pulm Htn	 Cyanotic episodes Tachypnea	Murmur Desaturation	ECG ECHO CXR SpO_2
RESP	Hypoxia Aspiration pneumonitis	Tachypnea	Retractions Stridor	SpO_2 CXR
GI	Failure to thrive	Feeding problems GE Reflux	Wt	Weight gain Reflux study
CNS	Hypoxia	Seizures Developmental delay		

Key References: Côté A, Fanous A, Almajed A, et al.: Pierre Robin sequence: review of diagnostic and treatment challenges, *Int J Pediatr Otorhinolaryngol* 79(4):451–454, 2015; Cladis F, Anand K, Grunwaldt L, et al.: Pierre Robin sequence: a perioperative review, *Anesth Analg* 119(2):400–412, 2014.

Perioperative Implications

Preoperative Preparation
- Avoid sedative premedication.
- Consider atropine as antisialagogue and to maintain heart rate.

Monitoring
- Pulse oximeter and precordial stethoscope are important.

Airway
- Intubation may be very difficult.
- Consider awake placement of an LMA or intubation in neonates.
- Airway management and intubation may become easier with age in isolated PRS.

Preinduction/Induction
- Spontaneous ventilation is recommended, usually inhalational induction.
- Consider oral or nasopharyngeal airway if obstruction occurs.
- Have difficult airway cart available, with multiple scopes and light wand.
- Consider use of LMA with fiberoptic bronchoscope and exchange catheter.
- Have surgeon in OR capable of performing rigid bronchoscopy and/or tracheostomy at induction.

Extubation
- Thorough evaluation before postop extubation in the OR. If extubation is chosen, pt must be fully awake and should recover in the ICU.

Adjuvants
- Muscle relaxants, if used, should be administered after intubation and reversed if extubation is planned. Minimize use of opioids intraop unless long-term intubation is planned.

Anticipated Problems/Concerns
- Airway obstruction during all phases of anesthesia is very common. Chronic airway may lead to opioid sensitivity intraop and postop.

Pituitary Tumors

Lauren K. Dunn | Edward C. Nemergut

Risk
- 10% of diagnosed brain neoplasms
- Peak incidence fourth to sixth decade of life

Perioperative Risks
- Related to specific hormone-related effects, including difficult airway management; cardiovascular complications (hypertension, coronary artery disease, cardiomyopathy); respiratory compromise (obstructive sleep apnea); and endocrine and lyte abnormalities

Worry About
- Airway management: Difficult mask ventilation and intubation, especially in acromegaly and Cushing disease
- Cardiovascular risk: Htn, CAD, cardiomyopathy
- Respiratory complications: Obstructive sleep apnea and postop ventilatory support
- Endocrine abnormalities: Acromegaly, hyperthyroidism, Cushing disease, panhypopituitarism, postop DI
- EleLyte abnormalities: Hypernatremia secondary to DI
- Rarely, management of elevated ICP

Overview
- Tumors classified by size (macroadenoma >1 cm vs. microadenoma <1 cm) and hormone secretion (functioning vs. nonfunctioning).
- Functioning tumors present with symptoms of hormone excess.
- Nonfunctioning tumors are more likely to be macroadenomas and present with symptoms of mass effect: headache, visual loss (bitemporal hemianopsia), and hypopituitarism.
- Pts rarely present with elevated ICP owing to obstruction of the third ventricle.

Etiology
- Disease and secreted hormones: Acromegaly, growth hormone; Cushing disease, ACTH; gonadotroph, FSH and luteinizing hormone LH; prolactinoma, prolactin; thyrotrophic, TSH
- May occur in MEN 1 syndrome with pancreatic and parathyroid neoplasms.

Usual Treatment
- Medical therapy for treatment of systemic effects of functional tumors: Acromegaly, somatostatin analog (octreotide, lanreotide), growth hormone antagonists (pegvisomant); Cushing disease, ketoconazole, metyrapone (block cortisol synthesis); prolactinoma, dopamine agonist (bromocriptine, cabergoline); thyrotropic, somatostatin analog (octreotide, lanreotide) or propylthiouracil
- Tumor resection via transsphenoidal approach (endoscopic endonasal or sublabial)
- Gamma knife radiosurgery