Morbid Obesity

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Risk

- Incidence in USA: Approximately 5% morbidly obese

Perioperative Risks

- Increased morbidity and mortality versus normal BMI from resp and cardiac issues

Worry About

- Challenging procedures: IV start, intubation, ventilation, epidural cath placement.
- Restrictive pattern of resp disease, hypoxemia, larger O_2 demand, small FRC; OSA is common, with associated cardiac issues.
- Htn: Systemic and pulm.
- DM.
- NASH.
- Reflux, hiatal hernia, and depression.

Overview

- Defined by BMI (weight in kg/height in m^2); >30 obese; >35 morbidly obese
- Cardiac and resp issues mainly due to size; large body mass to be perfused and oxygenated; increased cardiac strain and resp effort of breathing; OSA common; increased sensitivity to narcotics
- Depression common

Etiology

- Disputed role of genetics, mainly environmental and nutritional habits; essentially a form of severe malnourishment

Usual Treatment

- Medical treatment includes psychological counseling, along with decreased calorie consumption with increased exercise, if physically able.
- Surgical treatment includes gastric banding, Roux-en-Y, sleeve gastrectomy, or intestinal bypass.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Htn Pulm	Fatigue, dyspnea	Auscultation, increased heart size, ±rales	BP, ECG, CXR
	Htn Cardiac failure Coronary disease	Dyspnea, fatigue, syncope including JVP, peripheral edema, hepatomegaly, crackles	Auscultation, palpation, auscultation	CXR, ECG, ECHO ECG, ECHO
		Chest pain, SOB		ECG, stress ECHO Coronary angiogram
RESP	Restrictive disease	SOB, including resp rate, decreased exercise tolerance	Rapid shallow breathing, hypoxemia, large neck, redundant soft tissue in neck	PFT, ABG, CXR, Hg, pulse oxygen for room air saturation
	OSA	Hx of snoring, periods of apnea in sleep, nonrestful sleep, daytime somnolence and tiredness Different screening tests (STOP-BANG)	Large neck, redundant soft tissue in neck	Overnight sleep study for apnea hypopnea index $PaCO_2$ as predictor
NEURO	Depression	Hx	Question and answers, survey instruments	By psychologist and/or psychiatrist
HEENT	Potentially difficult intubation	Mallampati, upper lip bite test	Evaluation for large tongue, small interdental distance, limited ROM	
GI	NASH NIDDM	Hepatomegaly, icterus, ascites Polyphagia, polyuria, polydipsia	Palpation	LFT, PT, PTT, BUN, Cr UA, BS, GTT, HgA1c

Key References: Sinha AC: Some anesthetic aspects of morbid obesity, *Curr Opin Anaesthesiol* 22(3):442–446, 2009; Nishiyama T, Kohno Y, Koishi K: Anesthesia for bariatric surgery, *Obes Surg* 22(2):213–219, 2012.

Perioperative Implications

Perioperative Preparation

- All medications except for DM.
- Avoid sedation, unless benefit outweighs risk of postop resp depression.
- Consider prophylactic preop IVC filter placement if risk of DVT is high.
- Preop carbohydrate loading isoosmolar drink 2 to 3 h prior to surgery may benefit postop insulin resistance, nitrogen, and protein loss and decrease LOS.

Monitoring

- Routine with ± arterial cath if cardiac status dictates or ultra obese (BMI >70 kg/m^2 or weight >200 kg)
- If severe cardiac or resp disease, ABG
- UO
- Central venous access if peripheral access difficult, or CVP or pulm pressures need to be monitored for cardiac disease

Airway

- Position at 30-degree head elevated (HELP) to improve probability of intubation with direct laryngoscopy; BMV difficult in 10–15% and intubation difficult in 1–2%
- Minority of pts may need awake FOI
- Prepare for difficulty with multiple airway options, such as laryngeal masks and video laryngoscopes and short-acting drugs

Induction

- Preoxygenate with pressure support if possible; complete denitrogenation.
- Rapid sequence with cricoid pressure should be considered.

Maintenance

- Drug dosing: Lipophilic dosed to real body weight; lipophobic to IBW or LBM.
- Desflurane preferable due to complete and rapid recovery; avoid nitrous; TIVA has advantages if high risk of PONV.
- Consider opioid free or low opioid intraop; use such adjuncts as NSAIDs, acetaminophen, pregabalin, lidocaine infusions, α-2 agonists.
- Appropriate goal-directed fluid infusion based on deficit, losses, and UO.
- Ventilation: Start at TV 6–8 mL/kg IBW; RR 12–14/m; PEEP 8–10; adjust as needed, recruitment helps atelectasis, volume or pressure control.

Extubation

- Wide awake, no residual volatile agent, normocapnic, responsive with appropriate resp effort and partially sitting up; consider cyclodextrin for NMB reversal.

Postoperative Period

- Rapid placement on CPAP or BiPAP decreases atelectasis.
- Good analgesia with IV PCA, NSAIDs and local infiltration with LA and rapid mobilization helps resp function and decreases DVT.

Moyamoya

Francine S. Yudkowitz

Risk

- Occurs in both children and adults, peak age at 5 y and 40 y, respectively
- Female-to-male ratio of 1.8:1
- Highest incidence in Japanese and Asian populations; familial occurrence 10%

Perioperative Risks

- Stroke

Worry About

- Hypocarbia and hypercarbia
- Adequate cerebral blood flow
- Hypotension
- Hypothermia

Overview

- In Japanese, moyamoya means "puff of smoke," which describes the angiographic appearance of collaterals between internal and external carotid arteries.
- Chronic progressive cerebrovascular disease consisting of concentric stenosis or occlusion of the distal internal carotid arteries and large vessels of the circle of Willis with prominent basal collateral vessels.
- Histopathology shows eccentric intimal thickening by fibrous tissue, smooth muscle cell hyperplasia, and luminal thrombosis
- The most common presentation in children and adults is ischemic stroke.

- In contrast to children, adults may also present with intracranial hemorrhage.
- TIA, headache, and seizures are other presenting symptoms.
- Symptoms are precipitated by activities that involve hyperventilation that results in hypocarbia (e.g., crying, exercise).

Etiology

- Poorly understood but probably involves both genetic and environmental factors
- Autosomal dominant with low penetrance or polygenic mode
- Moyamoya disease (congenital): Vasculopathy without known associated risk factors, usually bilateral
- Moyamoya syndrome: Vasculopathy with other known associated conditions (e.g., sickle cell disease, neurofibromatosis, SLE, Graves disease, trisomy 21, prior radiation therapy to head or neck, brain tumors, and tuberculous meningitis)

Usual Treatment

- Medical:
  - Does not stop progression.
  - Antiplatelet agents (e.g., aspirin, ticlopidine).
  - Vasodilators (e.g., calcium channel blockers).
  - Rheologic therapy (e.g., pentoxifylline).
- Surgical:
  - Direct: Superficial temporal artery to middle cerebral artery (STA-MCA) bypass; also known as extracranial-intracranial bypass.
  - Usually done in adults, technically difficult in children.
  - Indirect: EDAS. Scalp or temporal artery is placed onto the arachnoid surface of the brain. Collaterals occur over time.
  - Usually done in children; combined with STA-MCA bypass in adults.
  - EMS: Temporalis muscle is attached to brain surface.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CNS	Decreased CBF Seizures	TIAs, strokes	Neuro deficits	CT/MRI/MRA EEG

Key References: Parray T, Martin TW, Siddiqui S: Moyamoya disease: a review of the disease and anesthetic management, *J Neurosurg Anesthesiol* 23(2):100–109, 2011; Chui J, Manninen P, Sacho RH, et al.: Anethetic management of patients undergoing intracranial bypass procedures, *Anesth Analg* 120(1):193–203, 2015.

Perioperative Implications

Preoperative Preparation

- Assess for associated abnormalities.
- Review of chronic medications. Continue antiseizure and calcium channel blocker.
- Premedication:
  - In children, anxiolysis may be beneficial to avoid hyperventilation from crying.
  - Caution with sedatives and opioids that may result in hypercarbia.

Monitoring

- Arterial line for BP monitoring and blood gas analysis

Induction

- Inhalation or IV.
- Avoid hyperventilation and hypoventilation.
- Avoid hypotension that will impair CBF.
- Avoid muscle relaxants that release histamine or cause hemodynamic changes.

Maintenance

- Balanced anesthesia or total IV anesthesia.
- Nitrous oxide use is controversial.
- Maintaining cerebral and systemic hemodynamics is paramount.
- Avoid cerebrovasodilators.
- Minimize increases in $CMRO_2$ with adequate levels of anesthesia during painful stimuli.
- Ensure adequate CBF by avoiding hypotension, hypocarbia, and hypercarbia.
- Maintain normovolemia to hypervolemia.
- Maintain normothermia with warming blanket if needed.

Postoperative Period

- Monitor for hypoventilation to avoid hypercarbia-induced neurologic symptoms.
- Provide adequate analgesia.
- Maintain normotension, normocarbia, normovolemia, and normothermia.

Anticipated Problems/Concern

- TIA/stroke
- Intracranial hemorrhage
- Infection

Mucopolysaccharidoses

Megan A. Brockel | James J. Fehr

Risk

- All forms of MPS are autosomal recessive except MPS II (also known as Hunter syndrome), which is X-linked recessive (only males affected).
- Estimated incidence in USA: 1:30,000.

Perioperative Risks

- MPS pts are at increased anesthetic risk (most complications are associated with airway obstruction), and surgery is associated with a high mortality rate.

Worry About

- Airway obstruction, difficult airway management, cardiac pathology, obstructive and restrictive lung disease, cervical instability, spinal canal narrowing with cord compression

Overview

- MPSs are a group of rare, inherited, progressive lysosomal storage diseases caused by a lack of lysosomal enzymes required to break down glycosaminoglycans, resulting in their accumulation in tissues and organs.
- Children may appear normal at birth but begin developing symptoms by 1 y of age.
- Diagnosis is made by clinical features and increased urine mucopolysaccharides.
- Typical clinical manifestations include coarse facial features, impaired vision and hearing, airway abnormalities, cardiac problems, pulm disease, organomegaly, cervical instability, spinal cord compression, joint contractures, growth impairment, and hernias.
- Some types are associated with cognitive impairment.
- Several different subtypes are described, with differing clinical manifestations, rates of progression, and anesthetic implications:
  - Type IH (Hurler): Considered the prototypic and most severe subtype of MPS I, it is characterized by coarse facial features and airway narrowing, leading to difficult intubation, cardiac involvement, hepatosplenomegaly, atlantoaxial subluxation, joint stiffness, and contractures.
  - Type IH/S (Hurler-Scheie): Characterized by macrocephaly, micrognathia, and mental capacity ranging from mild deficiency to normal intelligence.
  - Type IS (Scheie, formerly classified as type V): Characterized by mandibular prognathism, normal intelligence, aortic insufficiency, and joint stiffness.
  - Type II (Hunter): Characterized by coarse facial features and airway narrowing leading to difficult intubation, severe mental deficiency, valvular heart disease, hepatosplenomegaly, joint stiffness, and dwarfism.
  - Type III (Sanfilippo): Characterized by mildly coarse facial features and severe mental deficiency.
  - Type IV (Morquio): Characterized by mildly coarse facial features, aortic regurgitation, restrictive lung disease, atlantoaxial instability and narrowing of the spinal canal, and joint laxity.

Etiology

- Lack of lysosomal enzymes required to break down glycosaminoglycans results in their intracellular accumulation in tissues throughout the body, leading to progressive alteration of cellular structure and function.

Usual Treatment

- Life expectancy is decreased in pts with MPS but has improved with the introduction of HSCT and ERT, both of which have beneficial effects on pulm function.
  - HSCT must be performed early in the disease course (before developmental deterioration begins); it can prevent and/or reverse many clinical features of MPS (it appears to reduce airway complications in children treated at less than 2 y of age).
  - ERT is generally initiated later and also improves airway patency but does not cross the blood brain barrier and therefore cannot preserve CNS function.