

## Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Difficult airway anticipation Coarse facial features Small chin, corneal clouding	Pain	Neck ROM	Neck x-ray Neck US
CV	Aortic regurgitation Cardiomyopathy Difficult IV access	Poor exercise tolerance Angina Hx		ECG, CXR, ECHO
RESP	Restrictive lung disease Obstructive sleep apnea			PFT, sleep studies
GI	Hepatosplenomegaly Umbilical hernia, inguinal hernia			
CNS	Many have normal intelligence Cervical cord compression Hydrocephalus leads to increased ICP, deafness			CT/MRI
MS	Stiff joints Carpal tunnel syndrome Spondylolisthesis	Joint mobility	Decreased ROM of joints	

**Key References:** Clarke LA, Heppner J: Mucopolysaccharidosis Type I. October 31, 2002 [Updated July 21, 2011]. In Pagon RA, Adam MP, Ardinger HH, et al. editors: *GeneReviews®*, Seattle, WA, 1993–2015, University of Washington. <<http://www.ncbi.nlm.nih.gov/books/NBK1162/>> (Accessed 06.06.16); Nakayama H, Arita H, Hanaoka K: Anesthesia in a patient with Scheie syndrome, *Masui* 43(9):1385–1388, 1994.

### Perioperative Implications

#### Preoperative Preparation

- Anticipate upper airway obstruction.
- Excessive secretions (need for antisialagogue).
- Antibiotic prophylaxis for valvular heart disease.

#### Monitoring

- Routine

#### Airway

- Abnormal airway and short chin predispose to difficult airway.
- Large tongue and secretions leads to airway problems.

- Small size of ETT needed.
- Fiberoptic bronchoscopy.
- Use LMA and other supraglottic devices.

#### Induction

- IV access before induction
- Proper positioning with padding
- Spontaneous breathing

#### Maintenance

- Avoid myocardial ischemia.

#### Extubation

- Pt should be fully conscious, with intact airway reflexes.

#### Adjuvants

- Local anesthetics infiltration and regional anesthesia where needed

#### Postoperative Period

- Delayed emergence
- Pneumonia, bronchospasm, and laryngospasm apnea

#### Anticipated Problems/Concerns

- Management of difficult airway
- Cardiopulmonary problems common

## Schizophrenia

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### Risk

- Most common psychotic disorder with a lifetime worldwide prevalence of 1%
- Increased risk of suicide (5–10%)

### Perioperative Risks

- Marked by deterioration of function and self-care
- Exacerbation of psychosis with abrupt discontinuation of medications

### Worry About

- Pt being uncooperative, combative, or catatonic.
- Increased morbidity and mortality due to poorly controlled coexisting systemic disease and increased incidence of alcohol and substance abuse.
- Drug interactions and side effects:
  - Cardiogenic side effects include hypotension, tachycardia, prolonged QT interval, VFIB, and torsades-de-pointes.
  - EPS include muscle rigidity and laryngospasm.
  - Use of metoclopramide may worsen schizophrenic symptoms.

### Overview

- Schizophrenia is a psychiatric disorder that may be characterized by thought disorders, hallucinations, and fixed false beliefs.
- Antipsychotic medications are the mainstay treatment for schizophrenia.
- Antipsychotics have anticholinergic effects (dry mouth, blurry vision, urinary retention, constipation, tachycardia), histamine antagonism (sedation), and  $\alpha 1$  antagonism (orthostatic hypotension).
- First-generation antipsychotics have strong dopamine antagonism leading to EPS, such as tardive dyskinesia.
- Second-generation or atypical antipsychotics have serotonin antagonism and less dopamine antagonism leading to less EPS.
- EPS can be treated with anticholinergics such as benztropine 2 mg or diphenhydramine 50–100 mg.
- NMS is a rare but potentially fatal syndrome occurring after an increase in dosage of antipsychotic medications or abrupt D/C of dopamine agonist. The syndrome is marked by muscle rigidity, hyperthermia, altered consciousness, and autonomic instability. It is clinically similar to malignant hyperthermia and may be related to dopamine blockade.

- Autonomic instability presents as labile blood pressure, tachycardia, diaphoresis, incontinence, and flushing.
- Treatment of NMS includes hydration and cooling measures, IV dantrolene, and dopamine agonists such as bromocriptine.
  - Bromocriptine reduces mortality by 50% and is only available orally; thus NGT may be required.
  - Dantrolene is a skeletal muscle relaxant and will reduce heat production.
  - Benzodiazepines may also be used to alleviate catatonic symptoms.
- Avoid dopamine antagonists, such as metoclopramide, if NMS is suspected.

### Etiology

- Functional hyperactivity of dopamine transmission may play a role.
- Genetic and environmental factors are unclear and controversial.

### Usual Treatment

- Antipsychotic medications
- Psychotherapy
- ECT

Assessment Points				
System	Effect	Assessment by Hx	PE	Test
CV	QT, PR prolongation Torsades de pointe MI Postural hypotension Tachycardia	Dizziness Palpitations	Orthostatic hypotension Arrhythmia	ECG
RESP	Significant increased incidence of smoking		SOB, wheezing	
GI	Paralytic ileus (postop) Liver dysfunction due to meds			Abdominal x-ray LFTs
HEME	Agranulocytosis due to meds			CBC
ENDO	DM due to meds Hyperlipidemia due to meds			Blood glucose Lipid profile test
NEURO	Sedation EPS  1. Tardive dyskinesia  2. Akathisia  3. Dystonia 4. Parkinsonism		Somnolence  1. Choreoathetoid movements of head, limbs, trunk 2. Subjective discomfort causing agitation and restlessness 3. Slow sustained bodily contractions 4. Catatonia, rigidity, akinesia	
GENERAL	NMS	Antipsychotic use (usually increase in dose) or abrupt discontinuation	Hyperthermia, rigidity, autonomic instability, cardiac arrhythmia	WBC, body temperature monitoring, CK, UA (myoglobinuria)

**Key References:** Kudoh A: Perioperative management for chronic schizophrenic patients, *Anesth Analg* 101(6):1867–1872, 2005; Sukhminder B: Psychiatric diseases: need for an increased awareness among anesthesiologists, *J Anesthesiol Clin Pharmacol* 27(4):440–446, 2011.

### Perioperative Implications

#### Preoperative Preparation

- Hx may be unreliable or unattainable.
- Continue antipsychotic medications preop.

#### Monitoring

- Routine

#### Airway

- Routine considerations

#### Preinduction/Induction

- No specific technique clearly superior

#### Maintenance

- Hypotension.
- Tachycardia, arrhythmia.
- Increased risk of thermoregulation and hypothermia. Monitor temp and warm/cool pt appropriately.

#### Extubation

- Usual criteria

#### Postoperative Period

- Decreased reports of pain
- Increased incidence of severe postop ileus
- Increased risk of postop confusion
- Increased postop mortality

#### Regional Anesthesia

- While controversial, epidural analgesia may decrease incidence of postop ileus.

#### Anticipated Problems/Concerns

- Cardiac arrhythmia
- Hemodynamic instability and hypotension
- Hypothermia
- Potential for neurolytic malignant syndrome
- Disruption of the hypothalamic pituitary adrenal axis
- Medication side effects, including cardiac and extrapyramidal

## Scimitar Syndrome

Piedad Cecilia Echeverry Marín

### Risk

- Occurs in 1 to 3–5:100,000 live births. The incidence could be higher because of asymptomatic cases in adult population.

### Perioperative Risks

- Significant risk of pulm Htn, respiratory failure, and cardiac failure in the periop period.

### Worry About

- Intraop pulm Htn crisis, severe bleeding with hypovolemia, worsening left to right shunt, and in critical cases, reverse shunt right to left with acute cardiac failure and cardiac arrest.

### Overview

- Disease characterized by cardiopulmonary anomalies as partial or total anomalous pulmonary venous

return connection of the right lung to the inferior caval vein, leading a left to right shunt.

- Associated with other anomalies such as hypoplastic right lung, anomalous systemic arterial supply to the right lung with or without pulmonary sequestration, pulm Htn, dextroposition of the heart, heart failure, and atrial septal defect (with ostium secundum being the most frequent).

### Etiology

- Etiology is unknown, but in some pts, the anomalous pulmonary venous return has been coded genetically in the chromosome 4q12.

### Usual Treatment

- Depends on hemodynamic state and symptoms of the disease. The adult presentation usually is

asymptomatic, and the diagnosis is made by incidental findings.

- In children, clinical presentations are diverse. Symptoms in neonatal pts are severe and are associated with significant mortality due to severe respiratory insufficiency, cardiac failure, and pulm infections.
- When the L-to-R shunt and pulm Htn are significant, surgical correction is necessary to repair the anomalous venous return, ligation of collateral arteries, or lung segmentectomy.