

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.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Cardiac failure signs Anomalous pulmonary venous return, dextroposition of the heart	Poor breast feeding tolerance, poor physical activity tolerance, delay in growth and neurodevelopment	Jugular ingurgitation, hepatomegaly, low weight, heart murmurs at right side	ECHO, ECG Cardiac MRI
RESP	Hypoplastic right lung, recurrent infections, respiratory failure, pulm sequestration	Poor exercise tolerance, abnormal breathing, irritability, feeding inability, fever	Signs of respiratory distress, nasal flaring, decreased breath sounds at right side, hoarseness or wheezing if there is associated lung infection	CXR, ABG
IMMUNE	Chronic hypoperfusion Increased risk of infections	Recurrent respiratory symptoms, low weight	Fever, tachycardia, abnormal bronchial secretions	CBC
HEME	Chronic anemia Coagulation system is usually normal preop	Pallor, weakness, poor exercise tolerance	Tachycardia, pale skin, irritability	Hgb, Hct, blood type, and reserve blood components
GI	Prolonged fasting	Poor breast feeding tolerance, dehydration signs, low urine output	Lyte imbalance	Lytes
CNS	Neurodevelopmental delay	Psychomotor retardation, poor language development	Delay for crawling and walking	None

Key References: Orphanet. Scimitar syndrome. <http://www.orpha.net/consor/cgi-bin/OC_Exp.php?lng=en&Expert=185>, 2009 (Accessed 06.06.16); Rajaii-Khorasani A, Kahrom M, Mottaghi H, et al.: Scimitar syndrome: report of a case and its surgical management, *Ann Saudi Med* 29(1):50–52, 2009; Luna AM, Gonzalez G, Echeverry PC: Scimitar syndrome and anesthetic implications, *Rev Col Anest* 43(3):245-249, 2015.

Perioperative Implications**Preoperative Preparation**

- Anxiolytics like midazolam could help reduce anxiety but are contraindicated if pt is hemodynamically unstable or there are symptoms of cardiac failure.
- Avoid hypoxemia, pain, anxiety, dehydration, hypovolemia, severe anemia, and hypoxia. (FiO₂ higher than 35% because it raises L-to-R shunt.)

Monitoring

- Basic monitoring of ECG, noninvasive blood pressure, end tidal carbon dioxide, and temperature
- Pulse oximetry preductal and postductal (in neonates) and airway pressure
- Major surgery requires arterial line, venous central cath (to measure CVP and to instill inotropic drugs), urine output, and arterial blood samples.

Airway

- Tracheal intubation is essential.
- In cases of pneumonectomy, selective intubation and one lung ventilation could be useful but is not mandatory.

Induction

- Avoid hypotension during induction. Induction with inhalant agents takes longer than normal because of L-to-R shunt.

Maintenance

- Maintain cardiovascular stability, replace blood loss, correct lyte imbalance, and avoid acidosis, hypothermia, hypercarbia, hypovolemia, and severe anemia (Hb <7).
- Ventilation controlled by pressure is the best option with gentle airway pressure: low volumes (<8 mL/kg) and plateau pressure <30 cm H₂O.

Extubation

- Awake extubation if cardiovascular conditions, ventilation, and oxygenation are normal.

Adjuvants and Postoperative Period

- CXR to check central venous line position and to evaluate postop lung images.
- Although regional anesthesia is not contraindicated, be careful with epidural/caudal cath if pt will need postop anticoagulation.
- Pediatric intensive care should be available; ensure excellent pain management.

Anticipated Problems/Concerns

- Prolonged fasting is associated with hypovolemia and cardiovascular instability. Check fasting time and correct lyte imbalance.
- Respiratory signs of ventilatory failure require tracheal intubation and stabilization before surgery and suspect pulmonary infection that requires antibiotic use.

Scleroderma

Lee A. Fleisher

Risk

- Incidence: 9:1,000,000 per y.
- Prevalence: 300,000 Americans have scleroderma.
- Male:female ratio is 1:4; highest in young African-American women.
- More severe in Native Americans and African Americans.
- 10-y survival is 55–60%; presence of pulm Htn is a major prognostic predictor.

Perioperative Risks

- Severe hypotension secondary to hypovolemia
- Hypoxia secondary to pulm Htn and restrictive disease
- Failed intubation

Worry About

- GI reflux
- Obliterative vasculopathy leading to pulm Htn
- Restrictive lung disease

- Renal crises
- Intraop hypothermia-induced vasospasm

Overview

- Scleroderma, or systemic sclerosis, is a chronic connective tissue disease generally classified as one of the autoimmune rheumatic diseases.
- Targets skin, lungs, heart, GI system, kidneys, and MS system.
- Onset generally occurs between 25–55 y.
- Three features: Tissue fibrosis, vasculopathy of small blood vessels, autoimmune response.
- Two major classifications: Limited and diffuse cutaneous scleroderma.
- May have overlap syndromes with other rheumatic diseases.

Etiology

- Autoimmunity, genetics, hormones, and environmental factors may all play a role.

- Autoantibodies: Antitopoisomerase in diffuse forms, anticentromere in limited form.
- Twin studies suggest a limited genetic role.

Usual Treatment

- Treatment begins during early inflammatory stage, and strategies are target-organ specific, including antifibrinolytic agents, antiinflammatory drugs, immunosuppressive therapy, and vascular drugs.
- Treatment of symptoms including pain and reflux.
- Skin thickening can be treated with numerous experimental drugs or interventions (including D-penicillamine, interferon-gamma, mycophenolate mofetil, cyclophosphamide, photopheresis, allogeneic bone marrow transplantation).
- Surgical treatments include amputation and lung transplantation.