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. (FiO_2 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 H_2O.

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.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Cutaneous fibrosis		Masked facies Small oral aperture Atrophy of gums Hyperpigmentation	
CV	Pericardial disease Myocardial fibrosis Conduction abn	DOE CHF Arrhythmia Syncope	Rales	ECHO ECG, Holter
RESP	Fibrosing alveolitis Obliterative vasculopathy Pulm Htn	Dyspnea Nonproductive cough		CXR PFT Bronchoalveolar lavage ECHO
GI	Esophageal fibrosis/colonic dysmotility	Difficulty chewing Dysphagia Bloating Diarrhea	Weight loss	UGI/endoscopy
RENAL	Intrinsic renal vessel disease		Malignant Htn	Proteinuria Hematuria BUN or Cr
DERM	Cutaneous fibrosis		Fibrosis of limbs Sweating Atrophy and contractures Telangiectasis	
MS	Raynaud disease	Excessive cold sensitivity Pain	Cyanosis of digits	

Key References: Roberts JG, Sabar R, Gianoli JA, Kaye AD: Progressive systemic sclerosis: clinical manifestations and anesthetic considerations, *J Clin Anesth* 14(6):474–477, 2002; Dempsey ZS, Rowell S, McRobert R: The role of regional and neuroaxial anesthesia in patients with systemic sclerosis, *Local Reg Anesth* 4:47–56, 2011.

Perioperative Implications

Preoperative Preparation

- PPIs to reduce gastric acid.
- Consider metoclopramide for early disease; less effective for late disease.

Monitoring

- Invasive arterial monitoring relatively contraindicated in pts with Raynaud disease because of risk of digit ischemia, but ABG may be indicated.
- BP may be difficult because of reduced forearm blood flow.
- Consider PA cath in presence of pulm Htn.
- Skin temp may be significantly lower (1.5° C) than core temperature.

Anesthetic Technique

- RA may be preferable considering pulm problems, although it may be technically difficult.
- Regional technique may be associated with prolonged block in the presence of epinephrine because of severe vasoconstriction.
- Vasomotor instability may be seen.

Airway

- Pt may have severe decrease in oral aperture
- Consider awake FOB intubation.
- May require nasal intubation.

Preinduction/Induction

- Pt may be hypovolemic due to vasoconstriction.
- Consider volume expansion.
- May initially observe Htn, followed by vasodilation and hypotension.

Maintenance

- Usually requires mechanical ventilation because of restrictive lung disease.
- Intraop hypoxemia may develop secondary to pulm Htn.

Extubation

- Postop ventilation if significant pulm compromise.
- Pain control important to pulm status.

Anticipated Problems/Concerns

- Difficult airway
- Hypoxemia
- Hypotension

Scoliosis and Kyphosis

Chris C. Lee | Marie A. Theard

Risk

- Scoliosis is a lateral and rotational deformity of the spine that, when measured by x-ray, has a Cobb angle of >10 degrees. The incidence of this spine deformity is 1–4%.
- Most cases of scoliosis are idiopathic (70% of cases) with the infantile (<3 y) and juvenile (3–8 y) onset associated with higher morbidity and mortality than if developed later during adolescence (9–18 y). The F:M ratio for adolescent idiopathic scoliosis is 4:1, with severe curves occurring predominantly in females.
- Congenital scoliosis (1:1000 live births) in children is associated with bony abnormalities and GU malformations, or neural tube defects (meningomyelocele, spina bifida, syringomyelia). Failure of segmentation versus formation of part or all the vertebral body leads to development of this disease, with environmental influences like maternal alcohol abuse, IDDM, vitamin deficiencies, and hypoxia contributing to this defect.
- Neuromuscular scoliosis may be neuropathic (cerebral palsy and poliomyelitis) or myopathic (Duchenne muscular dystrophy).
- Other types of scoliosis include that found in neurofibromatosis type I (1:3000), Marfan syndrome, and osteogenesis imperfect.
- Kyphosis, a convex curvature of the spine (typically thoracic) usually occurs in older pts and is due to trauma (most common), degenerative disc disease, muscle weakness, inflammatory disease, postural changes or genetic. Congenital kyphosis, like congenital scoliosis, is caused by a failure of segmentation versus formation of part or all of the vertebral body.
- Scheuermann disease, with a prevalence of 8.3%, is the most common cause of juvenile kyphosis, defined by a Cobb angle ≥45. Males are more commonly affected. Mechanical factors and trauma have a role in the pathogenesis.

Overview

- Dx: The Cobb method is used to measure the severity of the curvature. A parallel line is drawn to the superior border of the caudal most vertebral body, which tilts to the concavity of the curve. A second line is then drawn parallel to the inferior border of the cephalad most vertebral body that tilts to the concavity of the curve. Perpendicular lines are drawn from these two lines, and the angle made by the intersection is measured as the Cobb angle.
- If left untreated, progression of this disease will lead to severe respiratory compromise (pts with curves >100 degrees).

Usual Treatment

- The treatment goal is prevention of curve progression and correction of the deformity.
- Observation or bracing techniques are used for skeletally mature with mild curves (<40–45 degrees).