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

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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).

For Cobb angles >55–60 degrees in the skeletally mature, spinal fusion with instrumentation of rods, pedicle screws, and/or laminar hooks are placed surgically to correct the deformity. Bone graft is applied to the fused area. Treatment between 45–55 degrees is not clear; however, most would recommend surgery once the Cobb angle reaches 50 degrees.

- Surgery may be approached posteriorly or via a combined anterior/posterior approach. The procedure may be staged to decrease morbidity and mortality. The anterior approach requires a thoraco-abdominal incision and retroperitoneal dissection. One lung ventilation may be needed for anterior exposure.

Assessment Points

System	Effect	Assessment by History	PE	Test
HEENT	Potential for difficult airway management	Prior difficult intubations, neck movement restrictions, glossal hypertrophy or aspiration risk secondary to DMD	Airway and neck exam	Cervical lateral spine, CT scan
HEME	Coagulation disorders	History of easy bruising or bleeding disorders		CBC, PT, PTT, platelet function, cross match, CMP
CV	Pulm Htn, cardiomyopathy secondary to underlying muscular dystrophies or mediastinal distortion	Functional status by exercise tolerance		ECG, ECHO, pulm arterial pressure
RESP	Restrictive pulm defect, severity of functional impairment related to curve severity	Functional status by exercise tolerance		CXR, ABG, spirometry, PFT with bronchodilator reversibility
GI	Poor nutrition	Feeding difficulty		Albumin, serum protein

Key References: Raw DA, Beattie JK, Hunter JM: Anesthesia for spinal surgery in adults, *Br J Anaesth* 91(6):886–904, 2003; Agabegi S, Kazemi N, Sturm P, et al.: Natural history of adolescent idiopathic scoliosis in skeletally mature patients: a critical review, *J Am Acad Orthop Surg* 23(12):714–723, 2015.

Perioperative Implications

Preoperative Preparation

- Respiratory dysfunction: Decreased chest wall compliance causing a restrictive pulm defect leads to chronic hypoxemia. Preop pulm function tests demonstrating VC <40% indicate the need for postop ventilation.
- CV dysfunction: Cor pulmonale may develop from chronic hypoxia and pulm Htn. An ECHO will help determine the degree of pulm Htn.
- Airway: Pre-existing arthritis in the cervical spine may necessitate an awake fiberoptic intubation or video laryngoscopy.
- Be aware of preop neurologic deficits.
- Consider administration of antifibrinolytics for surgery due to the significant risk of blood loss related to number of levels fused and length of surgery, and coagulopathy.

Preinduction/Induction/Maintenance

- Bronchodilators and pulm toilet may help to optimize the pt's respiratory status.
- Consider preop evaluation by the pain service for pain control.
- Induction with propofol or awake fiberoptic intubation.
- The use of succinylcholine should be avoided in pts with muscular dystrophies due to the risk of hyperkalemia causing cardiac arrest. Nondepolarizing muscle relaxants should be used sparingly due to the need for MEP and TCeMEP.
- Although IV induction agents can cause a slight reduction in amplitude and increased latency of cortical SSEPs, recording of SSEP or TCeMEP is still reliable.
- Maintenance of stable anesthetic depth is necessary to provide effective monitoring of SSEPs or MEPs. SSEPs and MEPs are mostly resistant to IV anesthetics. In the case of propofol, there is a decrease in SSEPs; however, rapid recovery after termination makes this an acceptable anesthetic for spine surgery. In the case of volatile anesthetics, it is recommended that low doses be used (<1 MAC), as higher doses cause a dose-dependent decrease in amplitude and increase in latency of SSEPs and MEPs. Nitrous oxide can also cause a decrease in amplitude of SSEPs, with less of an effect on MEPs. Hypothermia may also decrease the amplitude of SSEPs. Ketamine and etomidate may augment the amplitude of SSEP. Opioids have little effect on monitoring.
- For SSEP monitoring, an anesthetic technique with less than 0.5 MAC volatile anesthetic plus a propofol infusion with moderate/high dose opioid works well for these cases. For MEP monitoring, remifentanil/fentanyl combined with a propofol infusion is preferred. Ketamine and dexmedetomidine are also acceptable choices.

Monitoring

- Scoliosis correction is associated with major blood loss. Pts should be equipped with large-bore IV access and consideration given to strategies for reducing the need for blood transfusion (i.e., antifibrinolytic agents, preop erythropoietin, cell saver, preop autologous donation, intraop hemodilution).
- ASA standard monitors.
- Continuous blood pressure monitoring with an arterial line/CVP cath.
- SSEPs, MEPs and EMG, TCeMEP.
- BIS monitor to help asses anesthetic depth.
- Estimate blood loss from suction canister, cell-saver device, and sponges.
- Urinary cath.
- Consider O_2 sat pulse oximetry on big toes during anterior exposure of the lower lumbar spine to assess amount of iliac artery compression.

General Anesthesia

- Positioning for posterior fusion is prone with the abdomen free and in reverse Trendelenburg to reduce venous pressures at the surgical site and bleeding. Special care should be taken to ensure peripheral nerves are padded to prevent compression neuropathies, and eyes should be protected to avoid corneal abrasions. The potential for visual loss, though rare, is real; pts should be counseled preop regarding this risk.
- Thoracic approaches may require a lateral position with a double-lumen ETT for one lung ventilation. DLT position should be verified by fiberoptic bronchoscopy.
- Wake-up test allows intraop testing of the lower limb for motor function, allowing early detection of neurologic injury after instrumentation. The pt awakens from anesthesia and completes motor tasks on command. Wake-up test carries the risk of unintentional extubation or IV access loss if the pt becomes agitated.

Postoperative Period

- Postop ventilation may be necessary for some pts: Pts with neuromuscular disorders, severe restrictive pulm disease, congenital cardiac abnormalities, right heart failure, obesity and OSA, prolonged surgical procedure, pts who have a thoracotomy, and significant blood loss.
- Pain management requires a multimodal technique, including spinal/systemic opioids, local anesthetics, and NSAIDs.
- Ileus can be minimized with utilization of a multimodal pain management regimen.
- Fluid management with UO monitoring and replacement of blood loss is necessary.
- Pulm toilet and ambulation is beneficial to help decrease respiratory complications.

Anticipated Problems/Concerns

- Pulm compromise
- Corneal abrasions/vision loss (posterior ischemic optic neuropathy)
- Neurologic injury due to direct contusion by instrumentation, decreased spinal cord blood flow, distraction injury of the spinal cord, and epidural hematoma
- Vascular injuries/PE
- Superior mesenteric syndrome/acute kidney failure