Scoliosis

Scoliosis is a deformity of the spine resulting in lateral curvature and rotation of the vertebrae, as well as a deformity of the rib cage, which can cause severe restrictive lung disease, pulmonary HTN & cor pulmonale.

ANESTHETIC CONSIDERATIONS:

1. Pediatric patient (usually adolescent)
2. Considerations of scoliosis:
    - Etiology: idiopathic vs. neuromuscular disease / connective tissue disease / congenital syndrome
    - Potential difficult airway (C- or upper T-spine correction, assoc. conditions)
    - Potential neurologic damage with neck movement
    - Aspiration risk if muscular dystrophies (weakened bulbar muscles)
    - Severe restrictive lung disease with pulmonary HTN & cor pulmonale
    - Risk of post-op respiratory failure and need for prolonged ventilation post-op
    - Inability to generate effective cough to clear secretions or aspirations
    - Association with congenital heart disease (MVP, coarctation, cyanotic heart disease)
    - If non-idiopathic, cardiac conditions associated with neuromuscular disease, congenital abnormalities, connective tissue disorders, etc. (eg. cardiomyopathy, conduction defects, valvular lesions)
3. Considerations of surgical correction:
    - Potential for massive blood loss (transfusions, blood conservation strategies, antifibrinolytics)
    - Potentially long procedure with risk of hypothermia and airway edema
    - Difficult positioning: prone / lateral / both (also anterior approach)
    - Potential need for OLV if thoracic approach to anterior spine
    - Spinal cord monitoring: Requirement for SSEP / MEP & anesthetic interactions
    - Significant postoperative pain

ANESTHETIC GOALS:

1. Preoperative assessment and optimization
2. Balance perfusion pressure to spinal cord vs. need for mild hypotension to minimize blood loss
3. Avoid exacerbations of pulmonary HTN (hypoxemia, hypercarbia, acidosis, hypothermia, light anesthesia & pain)
4. Manage anesthetic agents to allow adequate monitoring of spinal cord integrity (SSEPs / MEPs)
5. Vigilance for life threatening complications of VAE or major vascular injury (aortic, vena caval or iliac vessel)
6. Avoid coagulopathy secondary to massive transfusion with attention to avoiding hypothermia, decreased Hct and decreased clotting factors
7. Optimize for postoperative wean from ventilation
8. Postoperative pain control

HISTORY

- Scoliosis history: etiology, duration, extent of disease, levels involved & previous imaging w/ Cobb angle
- Evidence of cardiorespiratory involvement: functional capacity, dyspnea including supine, ability to cough, recurrent pneumonias or respiratory tract infections, previous hospitalizations, previous evaluation of severity of respiratory involvement & evidence of pulmonary HTN / cor pulmonale, history of dependence on CPAP
- Associated conditions: CHD (valvular / shunt / obstructive / LVEF), neuromuscular disease, connective tissue disorder, document preexisting neurological deficits
- Swallowing and aspiration problems / Nutritional status
- Troubles with neck movement (limitations, paresthesias)?
- Surgical Plan – Approach (anterior/posterior, thoracoscopic, staged or single procedure, Vertical Expandable Prosthetic Titanium Rib or VEPTR) & monitoring to be employed (SSEP, MEP, intraoperative wake up)

PHYSICAL

- Vitals including RR, weight, height, supine SpO_2 on R/A
- Airway assessment with attention to cervical spine mobility
- Pulmonary: increased RR, decreased tidal volume, accessory muscle use, asymmetries, air entry & breath sounds
- Cardiac:
    - RVH & cor pulmonale (increased JVP, peripheral edema, hepatomegaly, +HJR, RV heave, loud P2, wide split S2)
    - Murmurs (AR, MR) – marfan's
    - Apical pulse displacement – cardiomyopathy, duchenne's
- Full neurologic assessment with attention to peripheral neuropathies

INVESTIGATIONS

- **Labs**
    - CBC for anemia or polycythemia, lytes, BUN, Cr
    - Coagulation profile (antiepileptic medications can interfere with coagulation esp. valproic acid)
    - Type and screen / Crossmatch
    - ABG (decreased PaO_2, increased or normal PCO_2, V/Q mismatch, increased A-a gradient)
    - PFTs (restrictive defect, reduced VC and TLC)
- **Imaging**
    - ECG for RVH, RAE
    - CXR for aspiration or lung disease
    - ECHO for pulmonary HTN, cor pulmonale, cardiomyopathy or CHD
    - Spine x-rays for Cobb angle, level(s) of scoliosis, neck involvement

OPTIMIZATION

- Pulmonary, Cardiology and/or ICU consult (especially if significant co-morbidities)
- If anticipate awake intubation, premedicate accordingly (topical LA, anticholinergic)
- Preoperative blood conservation strategies (autologous donation, EPO, Fe) & discuss with surgeon as to intraoperative blood conservation and with blood bank for potential need of large amounts of blood products
- Discuss with patient & family blood transfusions, ICU, postoperative ventilation, intraoperative potential for awareness given monitoring
- Discontinuation of NSAIDs preop
- Reflux prophylaxis
- Consider bronchodilators pre-op
- Premedication with tylenol and gabapentin or clonidine
- Cautious sedation if severe respiratory compromise or pulmonary HTN

ANESTHETIC OPTIONS
- Require GA with ETT
- Regional – option for post-op analgesia (in addition to multimodal Rx)
    - However, epidural may make neurologic assessment difficult, issues with spinal/epidural hematoma if patient receiving LMWH (DVT prophylaxis)
    - Intrathecal analgesia can be administered before surgical closure, but may not last the post-op pain that can last up to 4 days; Intrathecal morphine decreases transfusion requirement (if given by spinal post-induction and pre-surgery)

ANESTHETIC SETUP
- **Drugs**
    - Emergency drugs - phenylephrine, ephedrine, (norepinephrine, milrinone or NO if pulmHTN/cor pulmonale)
    - TIVA infusions ready
    - Tranexamic Acid
- **Monitors and Equipment**
    - CAS monitors + 5-lead EKG, temperature probe, nerve stimulator
    - Arterial line ± CVP (± PA catheter if severe cardiac compromise, but rarely used)
    - Volume access (at least 2 large bore iv's), cell-saver, blood warmers & rapid infuser
    - MEP & SSEP monitoring (and a neurophysiologist)
    - Forced air warmers & increased room temperature
    - Consider TEE or precordial Doppler
    - ETT type: SLT vs DLETT (or SLT + bronchial blocker) if thoracic approach, FOB as needed

NEUROPHYSIOLOGIC MONITORING

SSEPS
- Elicited by electrically stimulating a mixed peripheral nerve (post. tibial, peroneal, sural) and recording response at distant sites cephalad to level of surgery
- Functional integrity of somatosensory pathways determined by comparing amplitude and latency change of responses during surgery
    - Reduction in amplitude by 50% and increase latency 10% is considered significant
- Pathways: peripheral nerve, dorsomedial tracts of spinal cord, and cerebral cortex [proprioception and light touch]
- Can have normal SSEPs but paraplegic post-op (re: blood supply – hypoperfusion of anterolateral tracts but not dorsomedial tracts)

MEPS
- Monitoring techniques subdivided according to site of stimulation (motor cortex, spinal cord), the method of stimulation (electrical potential, magnetic field), and site of recording (spinal cord, peripheral mixed nerve, muscle)
    - Neurogenic (NMB OK) or myogenic (large amplitude but variable morphology) responses recorded
- Stimulation cranial to site of surgery causes prodromic stimulation of motor tracts in spinal cord and of peripheral nerve and muscle caudal to site of surgery
    - Perturbation leads to reduced amplitude and increased latency
- Potential adverse effects: cognitive deficits, seizures, bite injuries, intraoperative awareness, scalp burns, cardiac arrhythmias
- SSEP has become accepted standard of care (less affected by technical difficulties associated with MEP); however, MEP widely used and the two methods should be regarded as complementary

OTHER
- Also, ankle clonus test and Stagnara wake-up test are options
- Rarely used on their own since the introduction of MEP, but may still be used to confirm neurologic dysfunction in the presence of changes in MEP or SSEP waveforms
    - Involves intraoperative "awakening" after spinal instrumentation and requires a cooperative patient
    - Issues with wake-up include:
        a) Inadvertent extubation of the patient during movement in the prone position
        b) Air embolism during a deep inspiration
        c) Dislodgment of the instrumentation during violent movements

MANAGEMENT OF ANESTHESIA

Induction
- Intubation: awake vs. asleep? Breathing vs. apneic? Direct vs. FOB?
- Induction guided by extent of cardiac disease, pulmonary HTN
- Intubating dose of rocuronium should not interfere with MEPs; Succ is good but avoid in patients with muscle dystrophies
- Positioning
    - Anterior thoracic or thoracolumbar = lateral approach via thoracotomy or thoracoscopic
    - Prone = ensure abdomen free for ease of ventilation & to minimize epidural venous engorgement while protecting brachial plexus (avoid excess anterior flexion & abduction of arms, turn head toward abducted arm (if only one side out), ensure axilla free - can use evoked potentials to assess integrity of brachial plexus)
- Tranexamic acid: loading dose anywhere from 20 to 50 to 100 mg/kg, then 10 mg/kg/hr infusion

Maintenance
- Preservation of spinal perfusion pressure
- Consider TIVA with an opioid and propofol, or an opioid/propofol/low-dose volatile anesthetic (0.3-0.5 MAC)

- UAH Stollery uses propofol + remifentanyl or sufentanil or ketamine infusions
- In regards to neurophysiologic monitoring:
    - SSEPs
        - Inhalational agents and N_2O cause dose-dependent decrease in amplitude and increase in latency (N_2O 60% with Iso 0.5MAC compatible with effective SSEP monitoring)
        - All volatile anesthetics produce a dose related decreased amplitude & increased latency
        - IV agents cause less disturbance EXCEPT ketamine & etomidate which INCREASE amplitude
        - Acceptable evidence based practice:
            - Volatile anesthesia < 1.0 MAC + opioid with constant level of anesthetic maintained during critical monitoring periods (ie. no bolus)
            - TIVA with propofol & remifentanil — however prolonged propofol infusions (> 12h) are problematic (slow emergence & controversial in pediatric patients)
        - IV opioids cause small reduction in amplitude and increase latency
        - NMBs cause no changes
        - SSEPs can be reduced by hypotension
        - For each 1ºC reduction in body temp, amplitude decreased by 7% and latency increased by 3% (animal data)
        - SSEPs also altered by hypoxia, hypercarbia, HTN
    - MEPs
        - Cortical-evoked responses more prone to effects of anesthetic agents than spinal-evoked responses
        - Propofol ++suppress cortical-evoked (dose-dependent decrease in amplitude)
            - Depressant effect of propofol is diminished with ketamine,
                - the best TIVA may be an infusion of opioid, ketamine (at low doses), and propofol
        - Inhalational agents also ++suppress cortical-evoked
        - Least affected by opioids, midazolam, and ketamine
            - However, midaz and etomidate cause significant but smaller reduction in amplitude
            - Opioid response variable
        - Advisable to employ a soft bite block during MEP monitoring to prevent tongue biting and dental damage.
        - Should be avoided in patients with active seizures, vascular clips in the brain, and cochlear implants
        - NMBD are contraindicated if muscle response is being monitored but optional if spinal cord or peripheral nerve response is monitored
    - Neurologic injury or loss of monitoring:
        - Stop surgery & ensure not hypoxemic or hypercarbic
        - Increase BP to greater preoperative baseline
        - Check Hb & volume status
        - R/O surgical causes – direct trauma, compression, hematoma, ischemia related to excessive traction
        - Release distraction rods
        - UAH Stollery: SoluMedrol 30 mg/kg bolus then 5.4 mg/kg/hr for 23 hr and Zantac 50 mg IV q8h
- For blood loss:
    - Acute normovolemic hemodilution
    - Antifibrinolytics (e.g. tranexamic acid decreases blood loss >40%)
    - Cell saver
    - Autologous blood (as above)
    - Avoid hypothermia, avoid increased intra-abdominal pressure (epidural venous congestion), and tight BP control

Emergence
- A long-acting opioid such as morphine/ hydromorphone should be added near the end of the case for post-operative analgesia
- Preferable to be conscious and respond to command immediately after anesthesia
- However, may require post-op ventilation (see complications)
- Evaluate sending to ICU for wean vs. extubation in OR - factors to consider:
    - Pre-op cardiorespiratory function (VC < 40%, evidence of distress, pulmonary HTN, functional capacity, airway protection)
    - Length & positioning of procedure and possible airway edema
    - Volume shifts and intraoperative blood loss
    - Temperature and evidence of coagulopathy / bleeding
    - Adequacy of analgesia

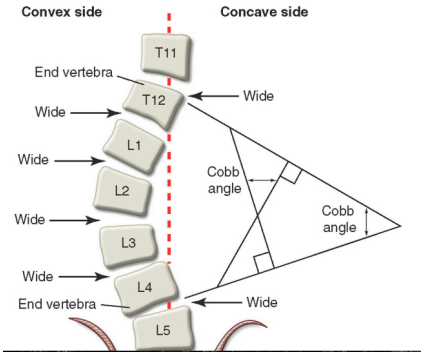
DISPOSITION & MONITORING
- ICU/obs bed

COMPLICATIONS
- Neurologic deficits/injury
    - Paralysis incidence 0.25-4% (caused by direct trauma, ischemia, compression, or hematoma; may be reversible with early detection)
- POVL
    - Rare after spinal surgery (≤0.1%)
    - May be caused by ischemic optic neuropathy (ION), retinal artery or vein occlusion, or cortical brain ischemia
- Respiratory Complications: atelectasis, infiltrates, hemo/pneumothoraces, pleural effusion, pneumonia, pulmonary edema, upper airway obstruction (occur more commonly when scoliosis is associated with mental retardation and developmental delay)
- Venous air embolism, PE
- CVA
- Post-op respiratory failure and ventilation
    - Patient factors
        - Pre-existing NM disorder, severe restrictive pulmonary dysfunction (pre-op VC<40% predicted), congenital cardiac disease, RVH, obesity
    - Surgical factors
        - Prolonged procedure, surgical invasion of thoracic cavity, blood loss>30 mL/kg
    - Post scoliosis surgery, the FVC and FEV1 decrease with a nadir at 3 days and are about 60% of preoperative values 7 to 10 days after surgery; It is not until 1 to 2 months after surgery that pulmonary function tests reapproach baseline values

PATHOPHYSIOLOGY
- Incidence of kyphoscoliosis is 4 per 1000 population

- M:F = 1:4
- 75-90% cases = idiopathic
- 10-25% cases are 2° to neuromuscular disease (e.g. DMD, poliomyelitis, CP, myelomeningocele), congenital syndromes, trauma, tumor or connective tissue disorders
- Curves can be described on the basis of age at onset, associated pathology, and anatomic configurations of the curve (e.g., single, double, or triple curves; amount of pelvic tilt; curve flexibility, as well as systems based on three-dimensional analysis)
- From an anesthetic perspective, a classification that gives some idea of the risk of adverse outcome, in particular respiratory failure, would be of clinical benefit
    - Children with scoliosis of early onset (<5 years) or with independent cardiac or pulmonary disease appear to be at increased risk of respiratory failure, whereas children with idiopathic scoliosis in whom the curve develops at adolescence appear to have minimal risk
- Measuring the "Cobb angle":
    - The end vertebra are the most superior and inferior vertebra which are least displaced and rotated and have the maximally tilted end plate
    - A line is drawn along the superior end plate of the superior end vertebra and a second line drawn along the inferior end plate of the inferior end vertebra
    - The angle between these two lines (or lines drawn perpendicular to them) is measured as the Cobb angle
    - In S-shaped scoliosis where there are two contiguous curves (double-major deformity) the lower end vertebra of the upper curve will represent the upper end vertebra of the lower curve
    - Cobb angle > 10° = scoliosis

Cote Figure 30-1:



- Severity of pulmonary impairment correlates with the magnitude of the thoracic curve:
    - Conventional thought: VC decreases when curve > 60 degrees and significant respiratory impairment occurs when thoracic scoliosis > 100 degrees
    - More recently it has been shown that children with adolescent idiopathic scoliosis may have pulmonary impairment disproportionate to the severity of the scoliosis and that significant respiratory impairment can occur well before the curve reaches 100 degrees
    - One review suggests that FVC falls below the normal threshold (80%) once the magnitude of the thoracic curve exceeds 70 degrees; FEV1 falls below the normal threshold once the main thoracic curve exceeds 60 degrees
    - Twenty percent of children with a thoracic curve of 50 to 70 degrees have moderate or severe pulmonary impairment (<65% of predicted)
    - Children with a structural cephalad thoracic curve, a major thoracic curve spanning eight or more vertebral levels, or thoracic hypokyphosis are at increased risk for moderate to severe pulmonary impairment
- Treatment:
    - Observation for Cobb angle < 25
    - Bracing for Cobb angle 25° - 40°
    - Corrective surgery considered when Cobb angle >50 degrees in thoracic or >40 degrees in lumbar or rapidly progressing curve (halts but does not reverse progression of respiratory and cardiac function)
- Pathophysiology:
    - Spine & rib cage changes cause changes in respiratory mechanics
    - Results in parenchymal defects & pulmonary vascular changes
    - Leads to decreased compliance & lung volumes which increases AW resistance & WOB
    - Causes increased V_d/V_t, alveolar hypoventilation, ineffective cough, recurrent pneumonias, & V/Q mismatching = hypoxemia
    - Eventually hypercapnia occurs as compensatory mechanisms fail
    - Pulmonary HTN occurs due to chronic hypoxia, hypercapnia, mechanical compression & impaired pulmonary vascular development
    - End-stage is cor pulmonale
    - If onset occurs at an early age (< 8 y/o), hypoplasia of the alveoli and pulmonary vasculature may occur = worse prognosis

Barash Figure 53-2:

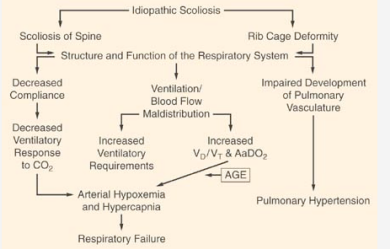


Figure 53-2. The factors in idiopathic scoliosis that contribute to respiratory function abnormalities and failure. VD, dead space volume; V_T, tidal volume; $AaDO_2$, alveolar to arterial oxygen gradient (Reprinted from Kafer ER: Respiratory and cardiovascular functions in scoliosis. Bull Eur Physiopathol Respir 1977; 13: 299, with permission.)

Cote:

Table 30-1 -- Classification of Scoliosis with Associated Key Anesthetic Risk Factors

Classification	Associated Issues with Scoliosis Surgery	↑ K^+ with Succinylcholine	Expected High Blood Loss	Respiratory Complication/Ventilatory Support
Idiopathic				
Infantile <3 years	Repeat operations, small size		√	√
Juvenile 3-9 years				
Adolescent 9-18 years	Regarded as "cosmetic" by patient—perfect result expected			
"Congenital"				
Bony abnormalities	"Acute angle" deformity: high risk of spinal cord injury, genitourinary malformations			
Neural tube defects				
Meningomyelocele, spina bifida, syringomyelia	Latex allergy, pressure sores, hydrocephalus, Arnold-Chiari malformation (avoid neck extension)			
Neuromuscular				
Neuropathic				
Upper motor neuron Cerebral palsy, cerebral hypoxia	Upper airway obstruction, recurrent pneumonia, postoperative pain management		√√	√√
Lower motor neuron Poliomyelitis				
Myopathic				
Progressive				
Duchenne muscular dystrophy	Cardiomyopathy, mitral valve prolapse, conduction abnormalities	√	√√	√√
Spinal muscular atrophy	Electrocardiographic abnormalities	√	√	√
Facioscapulohumeral muscular dystrophy	Hypertrophic cardiomyopathy, cardiac failure	√		
Other				
Friedreich ataxia		√		
Neurofibromatosis	Hypertension, other neurofibromas			
Mesenchymal				
Marfan syndrome	Mitral/aortic regurgitation			
Mucopolysaccharidoses (Morquio syndrome)	Atlantoaxial subluxation, difficult intubation			
Arthrogryposis	Difficult intubation, severe contractures		√	
Osteogenesis imperfecta	Small size			
Trauma				
Tumor				

Adapted from Goldstein LA, Waugh TR: Classification and terminology of scoliosis. Clin Orthop Relat Res 1973: (93):10-22.

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