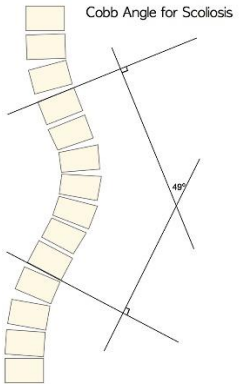
Neuromuscular Scoliosis

Definition of scoliosis

Spinal curve on the coronal plane of **at least 10 degrees**, measured by the Cobb method on AP X-ray.



Important terminology in scoliosis:

- Levo-scoliosis → curve to the Left
- Dextro-scoliosis→ curve to the Right
- Roto-scoliosis → scoliosis associated with significant Rotation

Types of scoliosis

A. Idiopathic scoliosis:

- Infantile scoliosis, age <3 years old, most cases spontaneously resolve
  - A/W ovarian abnormalities
- Juvenile scoliosis, age 3-10 years old
- Adolescent idiopathic scoliosis, age >10 years old, more common in females

B. Syndromic scoliosis:

- Common causes of syndromic scoliosis: NF 1, OI, Marfan syndrome, Ehlers-Danlos syndrome, Noonan syndrome, Prader-Willi syndrome, Down syndrome, Rett syndrome, Angelman syndrome, VACTREL association

C. Neuromuscular scoliosis:

- Results from disorders of the brain, spinal cord, or NM system (resulting in muscular imbalance)

Disorder	Incedince of scoliosis
CP (2 limbs involved)	25%
Mylodysplasia (lumbar level)	60%
SMA	67%
CP (4 limbs involved)	80%
Fredrech ataxia	80%
DMD	90%
Mylodysplasia (thracic level)	100%
Traumatic paralysis (< 10 years)	100%

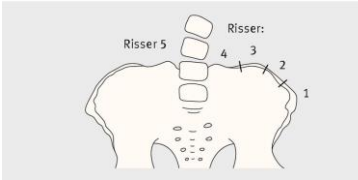
Patterns of scoliosis

Group-I curves: Double curves (thoracic **AND** lumbar), often in ambulatory patients, with minimal pelvic obliquity

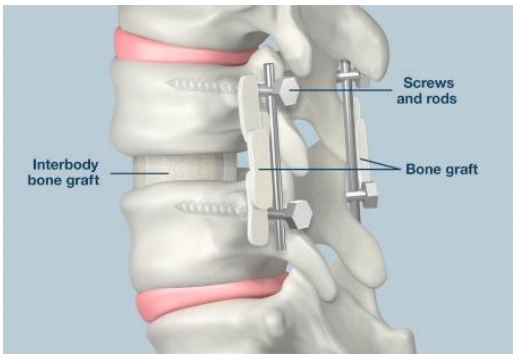
Group-II curves: Single curves (thoracic **OR** lumbar), often in quadriplegic patients, almost all display significant pelvic obliquity

Pelvic obliquity:

- Definition: Angulation of the pelvis to the horizontal plane
- Causes:
  - Supra-pelvic (spinal deformity)
  - Pelvic/infra-pelvic (lower limb contractures)
- Complications: Pressure injury, abnormal seating, hip pathology

Findings associated with significant growth remaining and potential scoliosis worsening	- Younger age - Premenarchal status - Tanner stage I or II - Risser stage 0 or 1 (measurement of ossification of the iliac crest) 
Differences between neuromuscular scoliosis & idiopathic scoliosis	1. Rate of progression: o Faster in children with neuromuscular scoliosis o Continues past skeletal maturity with neuromuscular scoliosis 2. Age of development (earlier [<10 Y/O] with neuromuscular scoliosis) 3. Risk of complications (more with neuromuscular scoliosis) 4. Management (refer to the management section)
Neuromuscular scoliosis & level of disability	The more the disability (e.g., GMFCS) → the less the ambulation → the more the risk of scoliosis & the more severe the scoliosis
Complications of scoliosis (particularly in CMC)	- Cosmetic and psychosomatic complications - Pain/irritability - Mobility/seating/care provision issues - Restrictive lung disease (Cobb ≥50°) - Cor pulmonale (Cobb ≥80°) - SMA syndrome **Complications of abnormal seating:** - Increased risk of reflux/sialorrhea/aspiration - Increased risk of pressure injury - Affects vision, communication & mobility - Compromises PO feeding
Physical examination findings in scoliosis	- Spinal asymmetry - Adam's test o Most sensitive o Asymmetric rib prominence - Neurological findings (if A/W spinal cord abnormalities)
Main investigation in scoliosis	AP/lateral spinal X-ray (severity and progression of scoliosis are measured according to the Cobb angle)
Treatment of scoliosis	1. Physiotherapy/seating/spasticity Tx 2. Brace 3. Posterior spinal fusion - Physiotherapy, seating, and bracing are less effective in neuromuscular scoliosis - **Posterior spinal instrumentation and fusion (PSIF)** is the MOST COMMON procedure

Posterior spinal fusion



Definition: Fusion of vertebrae (lumbar/thoracic) using screws, rods and bone grafts (in case of significant pelvic obliquity/risk of developing pelvic obliquity → instrumentation extends to the pelvis)

Indications:

1. Abnormal seating tolerance
2. Refractory pain/irritability
3. Restrictive lung disease
4. Cor pulmonale
5. Abdominal complications
6. Severe scoliosis with Cobb angle >90-100 degrees / rapid deterioration of curvature

Length of procedure/GA: 9-12 hours (up to 16 hours)

Risks:

- Pulmonary complications (MOST COMMON)
    - Difficult intubation and extubation (extubation may take place in PICU)
    - Risk of pneumothorax/pneumonia/aspiration pneumonia
    - May require post-op NIV/tracheostomy
- Significant post-op pain – requires involvement of APS
- Failed fusion (non-union, hardware breaking, loose hardware)
- Worsening of back pain
- Neurovascular injury/paralysis (transient/permanent)
- Bleeding & PRBC transfusion → can be significant / patient requires **iron-boosting protocol** pre-op
- SMA syndrome
- Blindness
- Prolonged recovery (may need establishing a new baseline post-op)
- Infection (mostly from bowel/bladder contamination)
- Mortality

Post-op care:

- Often requires 1-3 days **post-op PICU stay** before transfer to stepdown/inpatient ward (average length of hospital stay = 5-7 days)
- **Intensive rehabilitation** for 2-4 weeks after discharge that includes:
    - Pain management
    - Physiotherapy/improving seating tolerance
    - Nutritional rehabilitation
    - Establishing resp support

Post-op follow-up: 2 weeks, 6 weeks, 3 months, 6 months, 12 months, annually thereafter

Pre-op meeting with patient/family:
Should include discussion around:
- Risks, benefits, alternatives
- Importance of pre- and post-op optimization
- Realistic expectations

Who attends the meeting? Surgeon, PCC, PICU, PT, OT, RD, anesthesia, wound nurse

Pre-op follow-up: Every 6 months with a brace, every 4 months without a brace

Importance of pre- and post-op optimization
- Time frame: 6-9 months
- System-based

Neuro	**Red flags:** Uncontrolled seizures, uncontrolled dystonia, history of dystonic storming Optimizing AED/ketogenic diet within 3 months pre-op - Due to risk for increased seizure activity post-op Optimizing anti-dystonic medications within 3 months pre-op - Due to risk for increased dystonia/dystonic storming post-op
CVS	**Red flags:** Underlying cardiac condition, such as cardiomyopathy in DMD Echo within 3-6 months pre-op (to ensure adulate cord perfusion during procedure) In case of pre-existing cardiac condition → pre-op optimization of cardiac function, clearance and establishing post-op parameters by cardiologist
Resp	**Red flags:** Snoring/gasping/apneas in sleep, recurrent pneumonias/aspiration pneumonias, sialorrhea Optimizing resp status pre-op by optimizing secretion management and cough assist Sleep study within 6-9 months pre-op May require initiation of NIV pre-op
GI	**Red flags:** Uncontrolled GERD / feeding intolerance, constipation, low BMI Optimizing nutrition pre-op and post-op with the help of RD (improves post-op healing and decreases risk of post-op infections) - May require G-tube insertion at time of surgery for post-op tube feeding - May require TPN post-op in case of feeding intolerance Bowel management: - Pre-op with PEG BID x3 days - Post-op with PEG BID, daily bisacodyl +/- enema (due to use of opioids) Optimizing GERD management
Heme	**Red flags:** Anemia Patient requires **iron-boosting protocol** that involves iron supplementation and nutritional support with iron-rich diet pre-op and post-op (to restart supplement 7-10 days post-op)
GU	**Red flags:** History of recurrent UTI, AKI, CKD Keep indwelling catheter for 2-4 days post-op, close monitoring of in/out post-op

Bone health	**Red flags**: Non-weight bearing Optimizing Ca and vitamin D pre-op and post-op Patient requires x3 doses of IV bisphosphonate pre-op
Pain	**Red flags:** history of chronic pain, anxiety, parental history of pain catastrophizing Services to manage pain: - Pre-op: Transitional pain service - Post-op: Acute pain service (APS) followed by transitional pain service ➢ Not all patients are seen by TPS (based on surgeon performance) Gabapentin (if not initiated prior to surgery) may be initiated for 3 days pre-op and continued for 4 days post-po (average duration 7 days) (based on surgeon performance) Post-op: - **APS manages pain +/- bowel movements in the first 2-3 days post-op** - Patient may receive intra-thecal morphine intraoperatively (analgesic effect often lasts for ~24 hours) - Post-op, patient is started on IV opioid, which is transitioned to oral opioid shortly (often within 1-2 days) Scheduled PO opioid with a weaning schedule + PRN's (usually for 5-7 days in total including the weaning schedule and PRN's) - Scheduled NSAID (+PPI) + acetaminophen (usually for 2 weeks in total including the PRN's) - PRN diazepam/methocarbamol for the muscular spasms