# WELCOME


September 3-6, 2023

RIDING THE WAVES OF
Excellence
in SCI Care

# Spinal Cord Disorders

## in Children and Adolescents


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Shriners Children's Chicago



Thanks to Larry Vogel MD


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# Disclosure

Sue Mukherjee, MD discloses no conflicts of interest

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# LEARNING OBJECTIVES

**At the conclusion of this presentation, the learner will:**

1. Identify 2 unique etiologies for SCI in children.
2. Identify 2 unique musculoskeletal complications of SCI in children.
3. Discuss Management of hypercalcemia in an adolescent with a SCI
4. Identify different clinical presentation of Chiari I &II

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# Unique Features of Spinal Cord Injuries in Children and Adolescents

- **Uniqueness of pediatric SCI is based upon:**
  - the dynamic nature of **growth and development** in children and adolescents
  - the mutual **interactions of growth & development** with the manifestations & complications

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# Manifestations of SCI that are a consequence of young age

- SCIWORA = spinal cord injury without radiograph abnormalities
- Delayed onset of neurologic deficits
- SCI consequent to child abuse



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# Complications of SCI related to young age at time of injury

- Hypercalcemia
- Scoliosis
- Hip subluxation and contractures

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# Gender

| | Male | Female |
|---|---|---|
| 0-5 years | 51% | 49% |
| 6-12 years | 58% | 42% |
| 13-15 years | **70%** | 30% |
| 16-21 years | **83%** | 17% |
| 22+ | **80%** | 20% |

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# Etiology

| | 0-5 yrs | 6-12 yrs | 13-15 yrs | 16-21 yrs | 22+ yrs |
|---|---|---|---|---|---|
| **MVI** | 60% | 57% | 41% | 52% | 41.5% |
| Violence | 5% | 3.7% | 12.2% | 20.4% | 9.4% |
| Sports | 0% | 9.4% | 25.9% | 17.3% | 8.5% |
| Falls | 7.5% | 7.5% | 7.6% | 6.8% | 31.2% |
| Med/surg | 16.3% | 18.7% | 8.1% | 0.8% | 4.6% |
| Other | 11.2% | 3.7% | 5.1% | 2.9% | 4.8% |



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# Etiologies unique to pediatric SCI

- Lap-belt
- Birth injury
- High cervical lesions
  - Downs
  - Skeletal dysplasias
  - JRA



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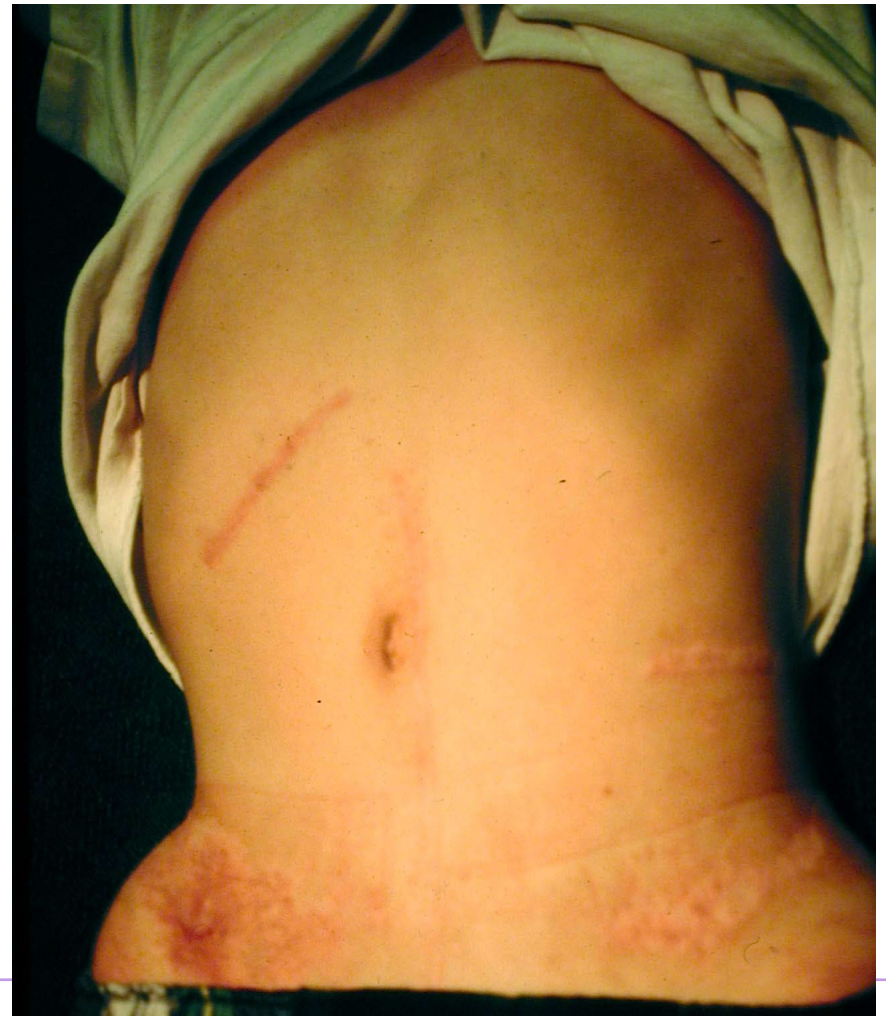
# Lap-belt injuries

- SCI
- Intra-abdominal injury
- "Seat-belt sign"
    - Abdominal wall bruising



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# Lap-belt injuries

- Intra-abdominal injury
  - Hollow viscus injuries in 33-40% of patients
    - Tears or perforations of small or large bowel
    - Less commonly seen are injuries to liver, spleen, pancreas, bladder and uterus



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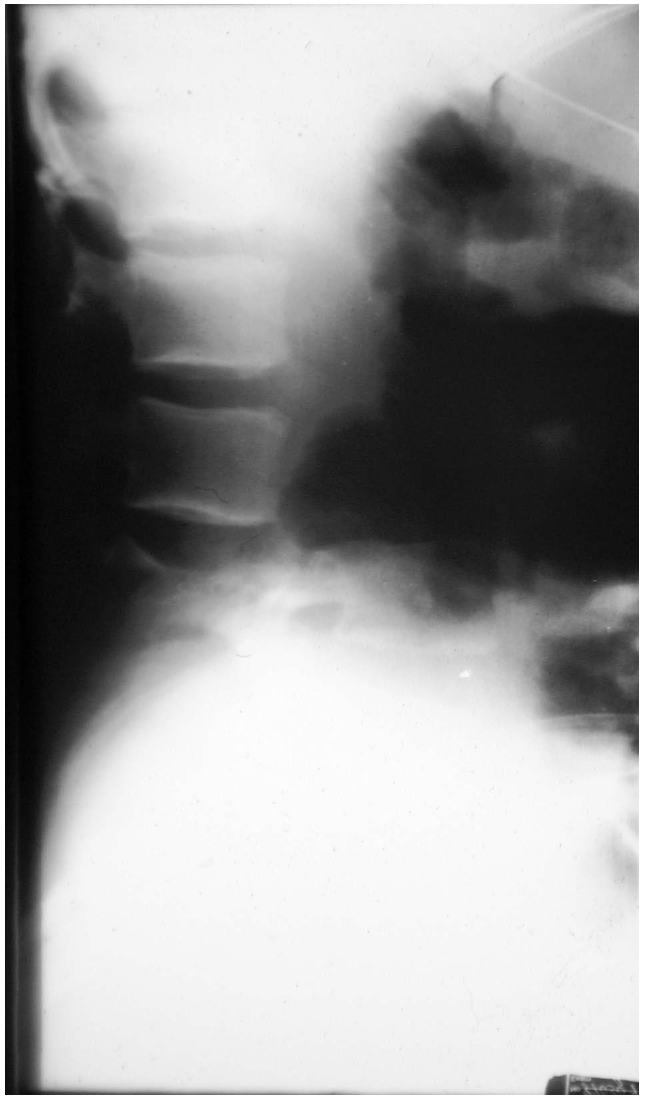
# Lap-belt injuries

- SCIWORA in 24-30%
- Level of vertebral injury = L2-L4
- Neurologic level varies
  - T6-T10 - ?related to vascular injury
  - Conus injury
  - Cauda equina injury



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# Lap-belt injuries

- Pathophysiology
  - Flexion/distraction forces with fixed anterior fulcrum = lap-belt above pelvic brim



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# Lap-belt injuries

- Prevention
  - Children < 40 lbs - Child restraints
    40-80 lbs - Booster seats
  - Children <2yo rear facing carseat

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# Neonatal Spinal Cord Injury

- Incidence = 1/60,000 births
- Upper cervical injury most common
  - caused by  torsion
- Lower cervical-thoracic injuries more commonly associated with breech deliveries
  - caused by traction



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# Neonatal Spinal Cord Injury

Associated injuries

- Brachial plexus injury
- Phrenic nerve damage
- Hypoxic-ischemic encephalopathy



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# Acute flaccid paralysis

- Acute flaccid myelitis
- Guillain Barre
- Toxic neuropathy



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# Acute flaccid myelitis

- Sudden weakness in one or more arms or legs
- Decreased muscle tone or absent reflexes
- Occasional involvement of cranial nerves
  - Facial weakness
  - Swallowing difficulties
  - Drooping of eyes



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# Acute flaccid myelitis

Confirmed case

- Acute onset of focal limb weakness AND
- MRI
  - Largely restricted to gray matter
  - Spanning one or more spinal segments

Probable case

- Acute onset of focal limb weakness AND
- CSF with pleocytosis

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# Acute flaccid myelitis

Etiologies

- Enterovirus D68, D71
- West Nile virus
- Herpes



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# Acute flaccid myelitis- AFM

Clinical presentation

- Acute onset of asymmetric limb weakness

Median age=7.6 years (5months – 20 years)

Prognosis

- 2/3 some improvement
- 1/3 no improvement
- No one fully recovered
- Typically bowel/bladder and sensation are unaffected

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# Neurological Impairment versus Age at Injury

| | Para | Tetra | Complete |
|---|---|---|---|
| 0-5 years | 55% | 45% | 80.7% |
| 6-12 years | 62.5% | 36.5% | 68.4% |
| 13-15 years | 47.6% | 51.9% | 55.6% |
| 16-21 years | 46.8% | 52.9% | 56.8% |
| 22+ years | 39% | 60.3% | 39.1% |

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# SCIWORA = Spinal Cord Injury WithOut Radiographic Abnormality

SCI without evidence of fracture or dislocation on:

- Routine spine radiographs
- Tomography
- CT
- Myelography
- Dynamic flexion/extension studies

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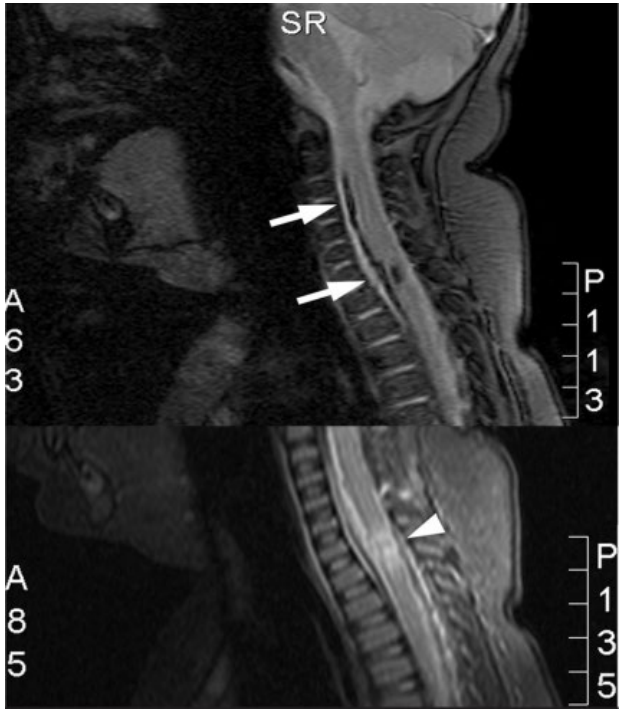
# MRI abnormalities noted in 65% of patients with SCIWORA

- Neural abnormalities
  - Complete cord disruption
  - Cord hemorrhage
  - Cord edema
- Extra-neural abnormalities
  - Rupture of anterior or posterior longitudinal ligaments
- Prognosis for neurologic recovery related to severity of MRI changes

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# SCIWORA

| | |
|---|---|
| 0-5 years | 64.2% |
| 6-12 years | 32.5% |
| 13-15 years | 22.3% |
| 16-21 years | 19.5% |

Ligamentous disruption/stretch



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# Delayed onset of neurologic findings

- 30 minutes to 4 days
- Approximately 25% of children with SCIs

- Many of the children may have transient neurologic symptoms such as subjective weakness or paresthesias



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# Bladder Management

Goals of bladder program

- Prevent urinary tract infections
- Preserve renal function
- Urinary continence
- Independence
- Socially acceptable

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# Intermittent Catheterization Program

- ICP initiated at approximately 3 years of age (when normally potty-trained)
  - ICP initiated earlier  if child experiences:
    - Recurrent UTI
    - Reflux/obstructive uropathy
    - Compromised renal function
  - Self catheterization is taught when child is ready, approximately 5 to 7 years of age

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# Bowel Management

- Goals
  - Complete and regular bowel movements
  - Short duration of bowel program
  - Aesthetics
  - Independence
  - Continence
  - Prevention of constipation or diarrhea

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# Bowel Management

- Bowel programs initiated at approximately 2-4 years of age
- Bowel program independently conducted by 5 to7 years of age

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# Components of bowel program

- Regularity - frequency and time of  day
- Privacy
- Conduct on toilet or commode
- Increase intraabdominal pressure
- Use of rapidly acting suppositories
- Manual extraction &/or digital stimulation
  - Interferes with independence
- Laxatives &/or stool softeners
- Diet

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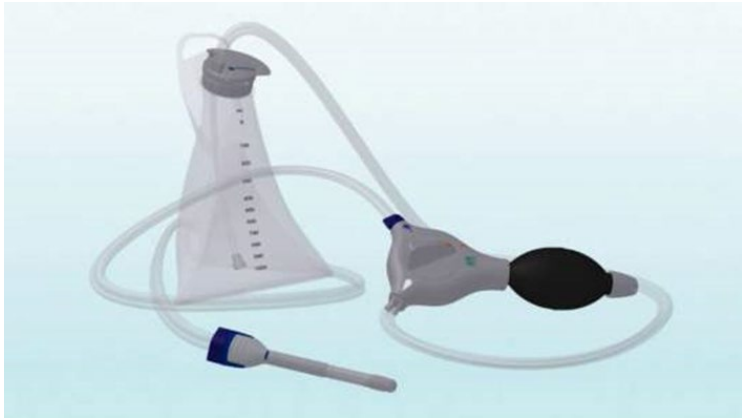
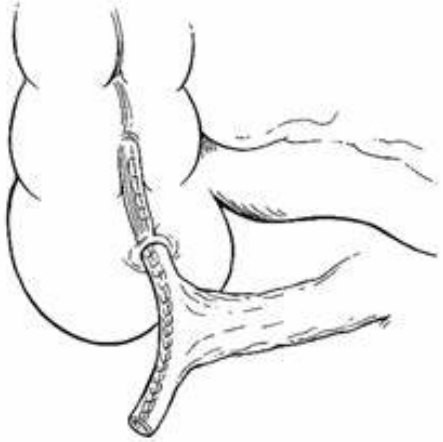
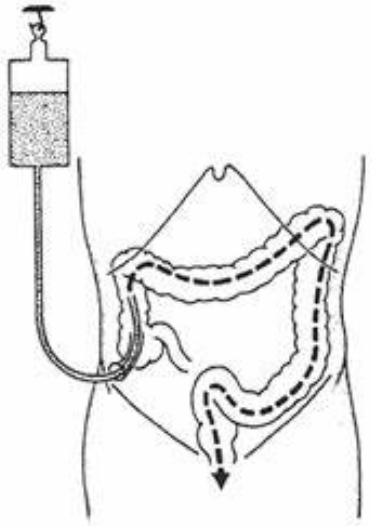
# Bowel Management Options

- Antegrade continence enemas
- Transanal irrigation
- Pulsed irrigation enemas
- FES at sphincter







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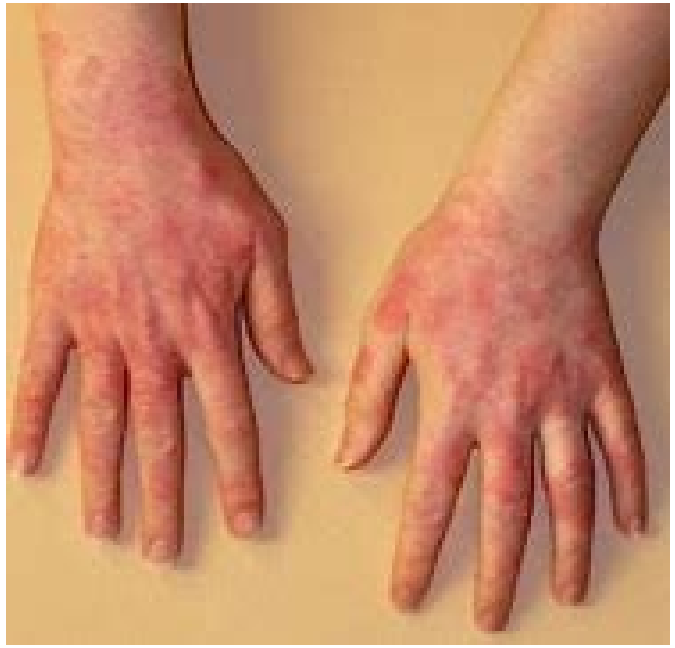
# Latex Allergy

- Type I allergic reactions
  - SCI
  - Myelomeningocele
  - Congenital GU anomalies
  - Health Care Workers



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# Latex Allergy Diagnosis

- History
  - including unexplained allergic reactions
- Skin testing
- RIA/RAST (blood test for antibody)
- ALLERGY=Positive Hx or Positive Lab



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# Latex Allergy Prevention

All patients

- Education
- Avoidance of all latex containing devices/materials

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# Latex Allergy
# Allergic Patients

- Medical alert IDs
- Autoinjectable epinephrine


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# Hypercalcemia

- Incidence=10-23%
- Most common in adolescent and young adult males with SCI
- Onset=1-12 weeks after SCI
- Less common since time to being upright and starting therapy is much quicker post injury than historical



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# Hypercalcemia Diagnosis

- Clinical
  - Abdominal pain, nausea and vomiting
  - Malaise, lethargy
  - Anorexia
  - Polydipsia, polyuria, and dehydration
- Calcium >10.2



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# Hypercalcemia Treatment

- Hydration with normal saline
- Pamidronate iv
- Vitamin D



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# Hypercalcemia Complications

- Nephrocalcinosis
- Urolithiasis
- Renal failure



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# Pulmonary Complications

- Atelectasis
- Pneumonia
- ARDS
- Aspiration
- Bronchitis/tracheitis
- Bronchospasm
- Respiratory failure

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# Respiratory failure

- Acute
- Chronic, particularly infants with high cervical lesions *increase Tidal Volume with growth
  - Excessive sleepiness during day
  - Investigations
    - Sleep studies
    - ABG



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# DVT

| | Acute | Rehab |
|---|---|---|
| 0-5 years | 0 | 0 |
| 6-12 years | 1.3% | 0.6% |
| 13-15 years | 5.7% | 2.2% |
| 16-21 years | 6.1% | 3.0% |



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# DVT Prevention

Anticoagulation

- Heparin SQ
- Warfarin
- Low molecular weight heparin
  - Monitor with anti-factor Xa levels



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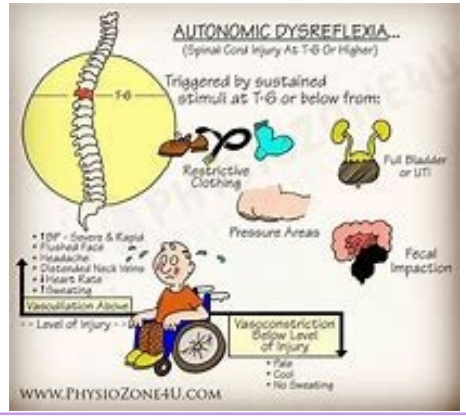
# Autonomic Dysreflexia

- Pathophysiology, manifestations, treatment, and complications similar to adult SCI except
- Blood pressure is lower in children and adolescents and increases with age - need to compare BP with child's baseline values
- Young children may not accurately articulate symptoms such as headache

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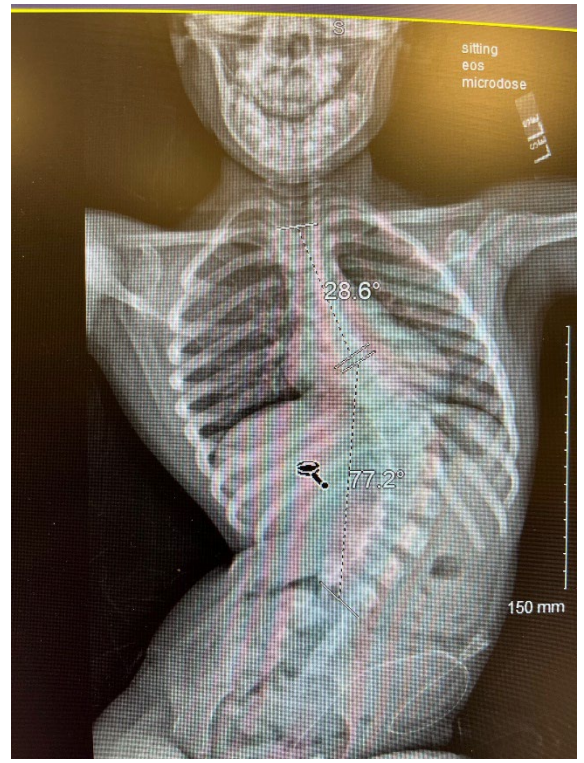
# Prevalence of scoliosis

Dependent upon age at injury

| Age at SCI | Prior to puberty | After puberty |
|---|---|---|
| Prevalence of Scoliosis | 98% | 20% |
| Need for surgery | 67% | 5% |

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# Orthosis

- Prophylactic to prevent development of deformity
- Advantage  may be effective in delaying surgery
- Disadvantage = may interfere with independence, ADLs and mobility, and may cause skin problems
- For curves  <20-40 : arrest/slow curve progression
- Sitting balance



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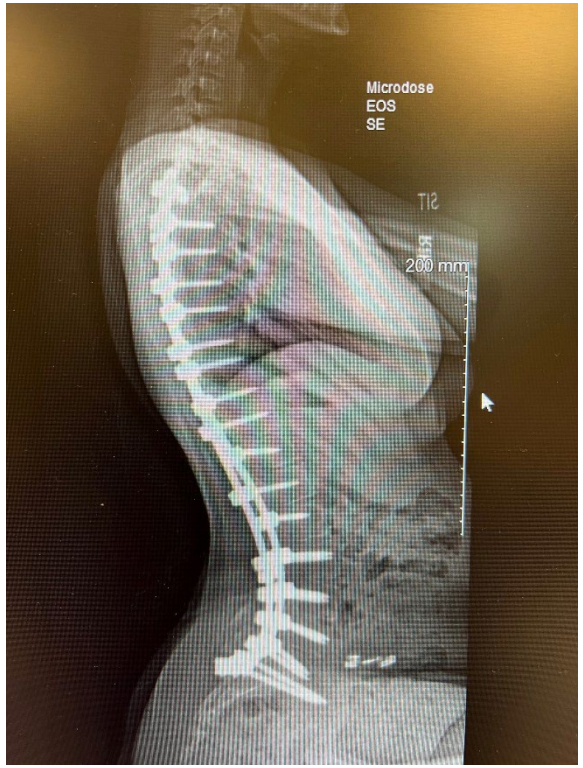
# Indications for Scoliosis Surgery

- Curves > 40 in a growing child
- Age > 10 years
- Rapidly progressive curve
- Functional problems or pain in a mature patient

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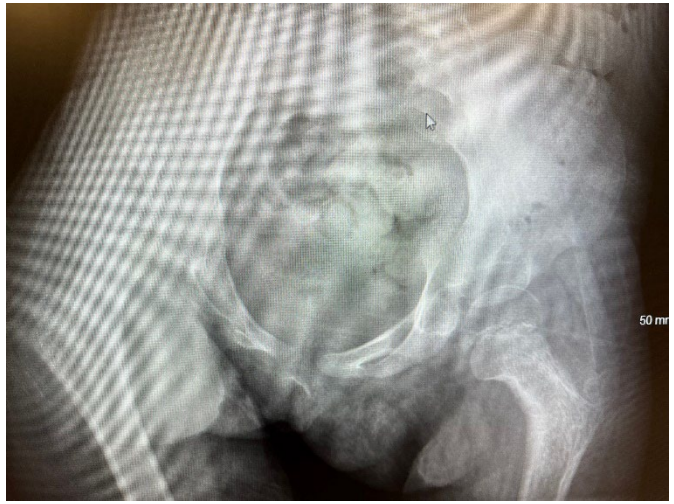
# Hip Subluxation/Dislocation

Incidence = 30-40%

Most common in children who are younger when injured

| Age when injured | Incidence of hip instability |
| --- | --- |
| ≤8 years | 62% |
| ≥9 years | 10% |





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# Neural Tube Defects

- Spina bifida and anencephaly
  - two most common forms of neural-tube defects
  - referred to as spinal dysraphism
- Spina bifida classified on whether neural tissue is exposed
- Fetal surgery or fetoscopic surgery is available



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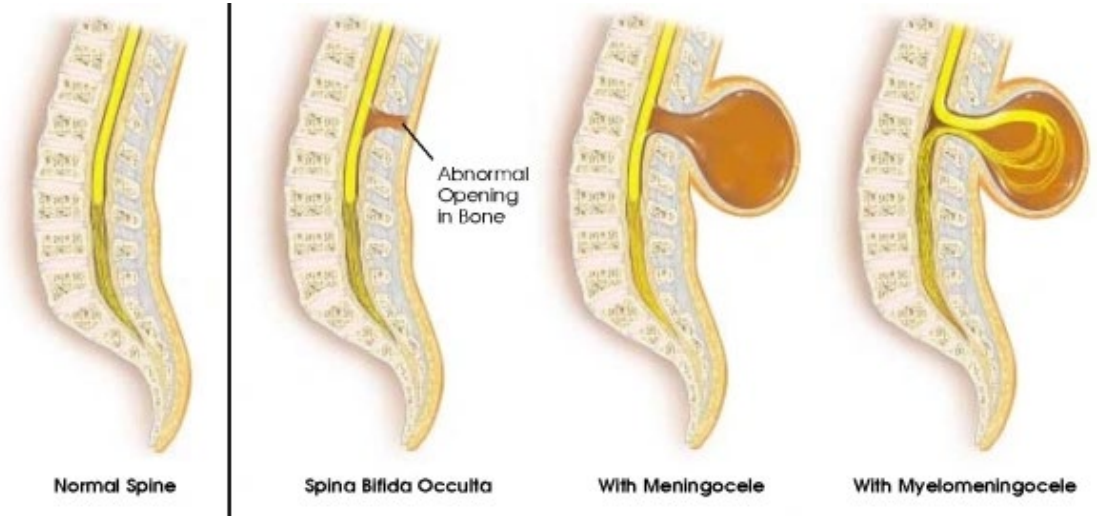
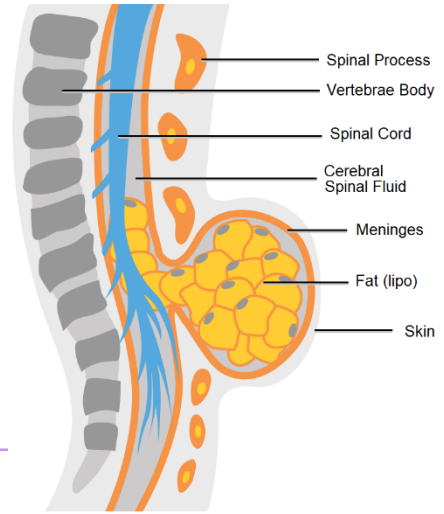
# Neural Tube Defects

- Myelomeningoceles
  - Open lesions that have either
    - absent skin covering
    - covered only by a thin membrane
- Spina bifida lesions with intact skin covering
  - occult spinal dysraphism
  - lipomeningocele

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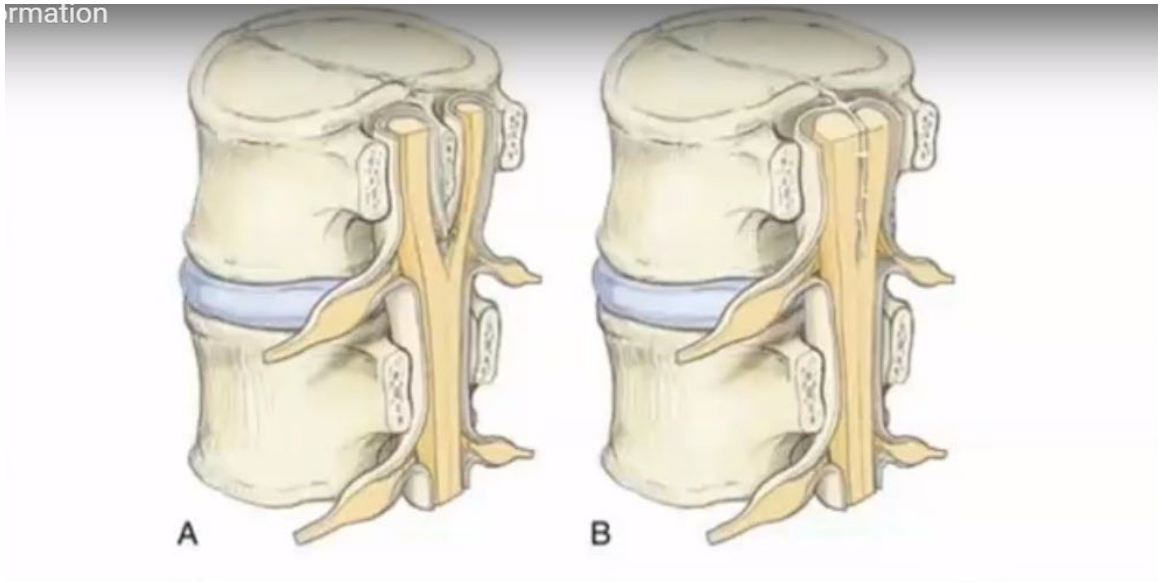
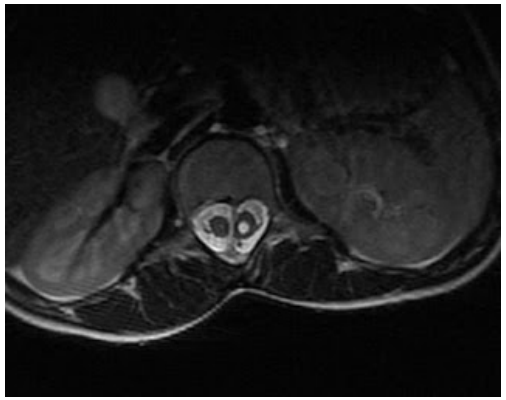
# Neural Tube Defects

- Occult spinal dysraphism
  - lipomeningocele
  - Diastematomyelia →
  - dermal sinus
  - meningocele
  - tight filum terminale
  - myelocystocele



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# Myelomeningocele Incidence

- 3.0 / 10,000  (1999-2000)
  - 3.8/10,000  Hispanic
  - 2.73/10,000 Non-Hispanic black or African American
  - 3.09/10,000 Non-Hispanic white



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# Myelomeningocele Etiology

- Both genetic and environmental factors
  - Maternal diabetes mellitus
  - Maternal use of valproic acid and carbamazepine
  - Maternal obesity, fever, and hyperthermia
  - Nutritional/dietary
  - All causes point to a role of FOLIC ACID availability at time of NT closure (21-28d embryo age)

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# Myelomeningocele

- Recurrence risk
- 2-5% if one affected sib
- 6-10% if two affected sibs
- Recommend 10x folic acid dose prior to conception = 4mg



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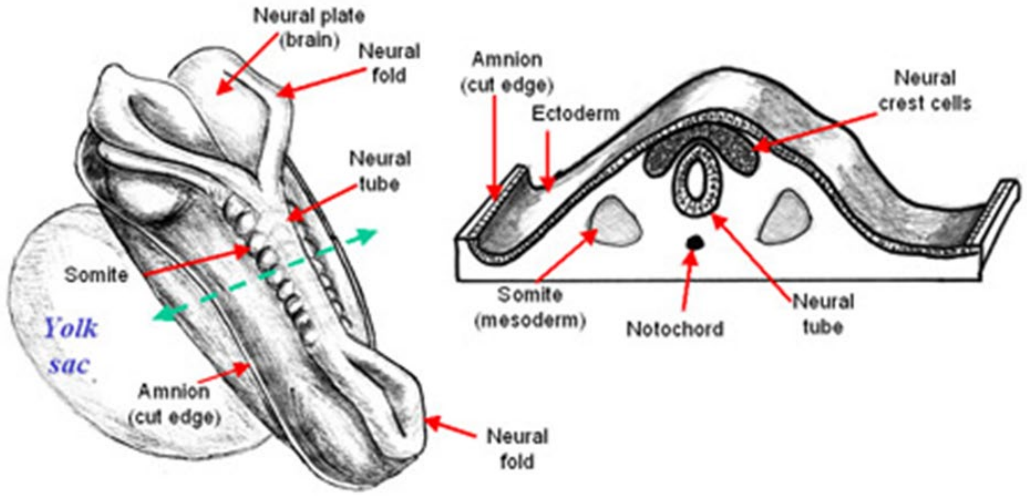
# Myelomeningocele Pathophysiology

- Neural-tube defects are caused by failure of the neural tube to close between the third and fourth week of gestation

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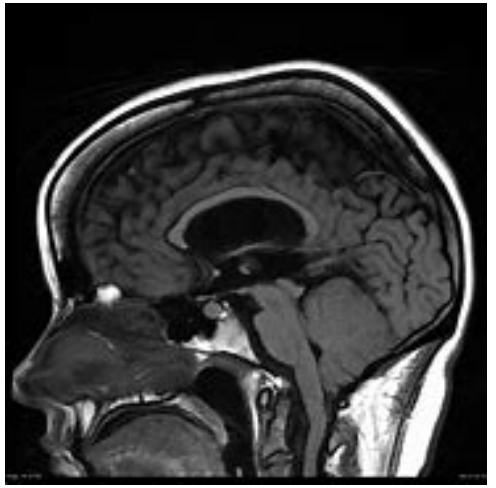
# Myelomeningocele Pathophysiology

- Brain
  - Hydrocephalus 70 to 90%
  - Chiari II Malformation
    - Small posterior fossa
    - Caudal displacement of cerebellar vermis & brain stem into cervical canal



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# Myelomeningocele Pathophysiology

- Spine and spinal cord defect can occur from the thoracic to the sacral levels
- Lumbosacral region most common
  - 66 to 75 %



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# Clinical Manifestations

- Approximately 30% of individuals with myelomeningocele have below normal intelligence
  - primarily perceptual motor abnormalities
  - normal verbal skills
- Hearing and visual impairments
- Seizures
- Clubfoot

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# Unique Clinical Manifestations

- Subtle CNS  deficits
- Disorders of visual-spatial organization
  defects in coordination and dexterity of hand function
- Cocktail chatter - excessive talking and superficiality of content
- LD and ADD



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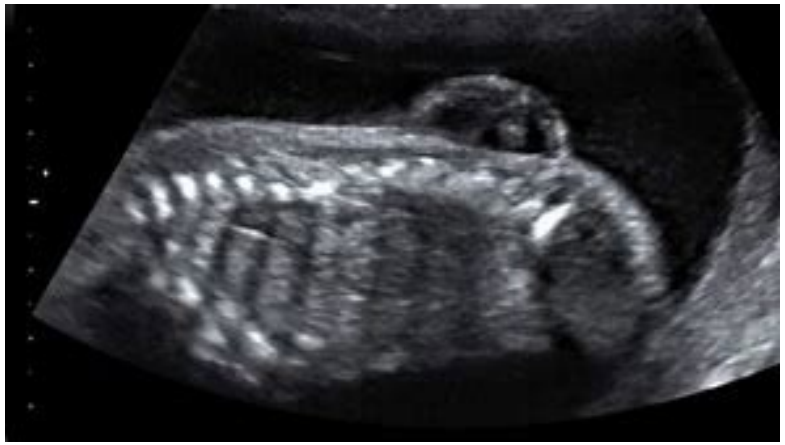
# Prenatal Diagnosis

- Alpha-fetoprotein
- Ultrasound –second-trimester anomaly scanning
- Amniocentesis



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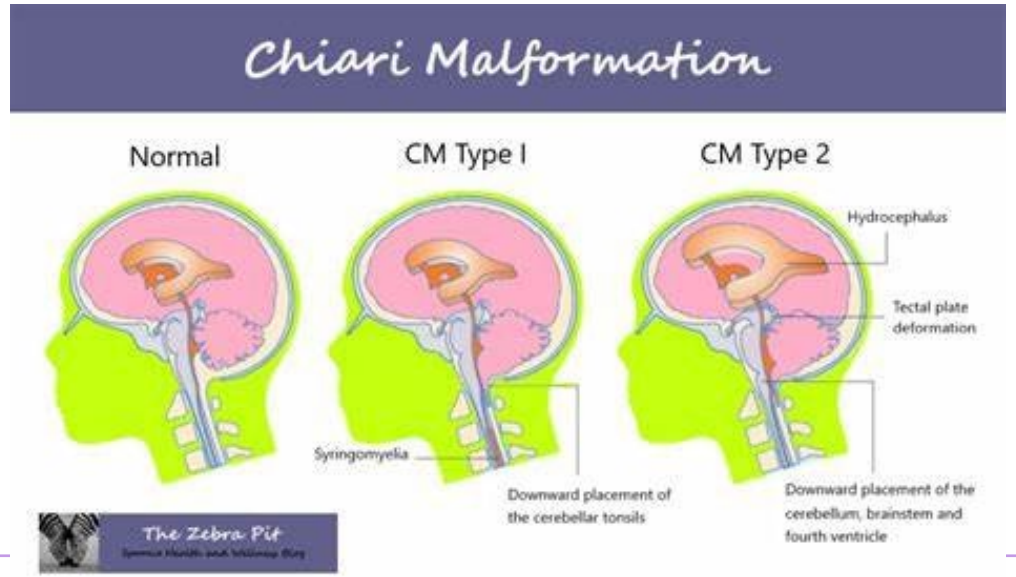
# Developmental Syringomyelia

- Chiari I malformation
  - Caudal displacement of cerebellar tonsils below the foramen magnum
- Most commonly involves cervical or cervico-thoracic cord

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# Developmental Sryingomyelia

- Orthopaedic abnormalities
  - Scoliosis
    - Left thoracic curve
    - Progressive curve on males
    - Abnormal neurological examination
  - Pes cavus
  - Charcot joints



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# THANK YOU!

# Q & A

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