Management of Spinal Deformities in Spinal Muscular Atrophy

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Disclosures

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  — I have no potential conflicts with this presentation.

• Brian Snyder
  — I have no potential conflicts with this presentation.
Spinal Deformities and the Lungs

- Chest wall deformities and scoliosis contribute to restrictive pulmonary disease.
- Pulmonary complications cause morbidity and mortality.
- Weak intercostal muscles and unopposed diaphragmatic function may result in the bell-shaped chest (parasol rib deformity).
- Symptoms include poor management of airway secretions, hypoventilation during sleep, poor chest wall development, recurrent pulmonary infections, skin pressure areas, and back and buttock pain.
Scoliosis

- Scoliosis occurs in greater than 50% of patients with SMA 1 and 2.
- Non-ambulatory patients are at greater risk for scoliosis.
- Pelvic obliquity and kyphosis are often associated with this spinal deformity.
- Because of the progression of the scoliosis and pulmonary compromise, early intervention is important.
Goals of Treatment

- Improve sitting balance/tolerance
- Decrease likelihood of decubiti, aspiration
- Relief of pain in hips and back
- Decrease need for assistance
- Eliminate use upper extremities for support
- Facilitate positioning/transfers
- Improve pulmonary function or pulmonary growth
Nonsurgical Management

• Careful observation for mild deformity.
• Orthotic management (avoid further constriction of thorax leading to impaired pulmonary function)
• Wheelchair seating systems to maintain sitting posture and accommodate pelvic obliquity.
• Orthoses may slow scoliosis progression; however, discontinue if there is progressive spinal deformity.
Surgery in Skeletal Immaturity (<10 years of age)

- Growing rod constructs without arthrodesis
- Distraction-based Systems:
  - Vertical Expandable Prosthetic Titanium Rib (VEPTR)
  - MAGEC Rods
- Guided Growth Systems:
  - Luque Trolley
  - Shilla
- Complications: infection, anchor displacement, laminar fracture, implant prominence, rod failure, premature arthrodesis, multiple surgical procedures
MAGEC Rod
MAGEC Rod
Surgery – Age 9 Years
2 Years Post Op
9 Years Post Op
13 Years Post Op
Surgery – Age 7 Years
Post Op
7 Months Post Op
6 Years Post Op
Surgery in Skeletal Maturity (>10 years of age)

• Posterior spinal arthrodesis, osteotomies to correct deformity, with segmental spinal instrumentation, pelvic fixation, and autologous / allograft bone graft.

• Complications: pseudoarthrosis, infection, functional deterioration, blood loss / transfusion, implant failure, thromboembolic phenomenon
Halo Traction

• Large rigid curves where spinal balance cannot be safely obtained via Anterior + Posterior procedure
• Halo-pelvic, Halo-femoral, Halo-gravity
  — Keep head/trunk elevated, sit up
• Traction applied before or between staged anterior and posterior procedure
• Must be able to tolerate traction
  — Normal Cervical spine – no instability
  — Monitor neuro status every shift
  — cranial n (esp Abducens), cervical chain


Safe Surgery

• Pre-op pulmonary / cardiology evaluation
• Total intravenous anesthetic technique
• Potassium supplementation
• Replace blood loss
• Cell-Saver
• Aminocaproic acid / Tranexamic acid
• Thromboembolic prophylaxis
• Steroid prep
• Malignant Hyperthermia Precautions
Spinal Cord Monitoring

- Somatosensory evoked potentials
- Motor evoked potentials
- EMG
Autologous Blood Transfusion

• Pre donation

• Cell-saver

• Constavac reinfusion
Pulmonary Management / Intervention

- Volume recruitment
- Ventilators
- Tracheostomy
- Mechanical insufflator / exsufflator
- Mucus mobilization devices
- Pneumococcal, influenza immunizations
Cardiac Management

• Evaluation: ECG, ECHO, Holter
• Intervention: angiotensin-converting-enzyme inhibitor (ACE inhibitor) i.e. enalapril
• Beta-blockers (carvedilol)
Gastroenterology / Nutrition

- Swallowing evaluation
- Diet control
- Supplementation
- Gastrostomy
- Pharmacologic
- Constipation management
- GERD management
Dietary Supplements

• Calcium citrate (better absorbed than Calcium carbonate)
  — Age 5 to 10  up to 600 mg./day
  — Age 11 to adult  more than 1300 mg./day  (in divided dosage)

• Vitamin D3 (better absorbed than D2)
  — Age 5 to 10  at least 800 I.U./day
  — Age 11 to adult  over 5000 I.U./day
Wheelchair Indications

• Prevent muscle fatigue
• Appropriate seating system
• Part-time use for long distance mobility; encourage short distance ambulation and transfers
Wheelchair Specifications

- Rigid seat and back
- Jay or Roho seating systems
- Appropriate trunk support, head control
- Tilt-in-space ➔ reclining
- Power assist modifications / controls
- Accommodate ventilatory support and growth adjustments
Musculoskeletal Issues in SMA: Appendicular Skeleton

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Contractures

- Loss of joint motion due to structural changes in muscles, ligaments, and tendons
- Contractures can occur at any joint
- Caused by weakness, decreased range of motion and prolonged positioning of extremity
Prevention of contractures

- Range of motion
- Splints and orthotics
- Equipment that help positioning
- Focus on all joints - emphasize joints that are tight
  - Incorporate stretching/passive ROM into daily routine
    - Diaper changes
    - Bath time
    - Before bed
Tendon Lengthening

- Surgical intervention indicated for fixed contractures that impair seating, mobility, function
- Contractures rarely occur in isolation
- Muscles that cross 2 joints most frequently affected
- **WARNING:** Weakens an already WEAK MUSCLE
Adduction contracture

- Limits ability to spread legs
  - interferes with toileting,
  - positioning in wheel chair,
  - standing,
  - walking (scissoring)

- Lengthen Adductor and Gracilis muscles
Hip flexion contracture

- **Ambulatory SMA 3**
  Fractionally lengthen psoas tendon @ pelvic brim to preserve hip flexion strength
- **Non-ambulatory SMA 2**
  Detach iliopsoas tendon from lesser trochanter

- Contributes to anterior pelvic tilt
- Fixed hip flexion contracture important to consider when fusing the spine to the pelvis
Hamstring Contracture

Limits ability to stand upright, Affects pelvic position when fusing spine to pelvis

Clinical characteristics

Clinical exam:
- Popliteal angle (>55°)
- Straight leg raise (<60°)
Fractional lengthening Hamstrings

- Supine position
- Single posterior incision above popliteal fossa

Tenotomy of Semitendinosus, Gracilis

Fractionally lengthen Aponeurosis of Semimembranosus, Biceps Femoris
Gastoc-Soleus muscle + Achilles tendon contracture

Toe walking during gait training/standing

- Younger patient without fixed contracture:
  - Serial cast / AFO

- Fixed contracture or severe foot deformity
  - Recession of gastrocnemius
  - Avoid Achilles lengthening
Osteopenia (low bone density) 

Increases Risk for Fragility Fractures
- Common in children who don’t walk
- Fractures caused by minimal trauma
- Common (reports up to 40%)
  - 40% femur, 26% tibia, 14% humerus, 9% clavicle
- Risk factors
  - Stiff joints
  - Decreased bone mineral density due to lack of weight bearing
  - Poor balance leading to falls
Factors Affecting Fracture Risk

Material
- Strength

Structural Geometry
- Load Capacity

Loading Condition
- Applied Load
Bone Mineral Density (BMD) accounts for the strength and stiffness of bone tissue.

Density = Mass of mineralized bone tissue
unit volume (gm/cm\(^3\))
Bone profoundly weakened when apparent on X-ray (osteopenia)

- Bone density reduced 30-50% before evident on radiographs
  - Corresponds to 40-75% reduction bone strength
Geometric Properties of Bone Are As Important As Bone Density

*Moment of Inertia*

A structure is more rigid when its mass is distributed away from the bending axis.
Ability of long bones to support or withstand load severely limited

- **Material** – Bone tissue
  - ✓ Severe osteopenia

- **Geometry** – Bone shape
  - ✓ Narrowed cross-section diameter
  - ✓ Thinned cortices
Clinical Presentation

- Osteopenia silent and progressive
- Routine radiography insensitive for detecting early bone loss
- Fracture often 1\textsuperscript{st} sign
Evaluation of bone density

- Uses detection system to measure transmission of X-rays through body
- Determines intensity that is transmitted
- Low radiation: 1-3 mrem

Dual Energy X-ray Absorptiometry (DXA)
Limitation: DXA Does Not Give True Density

- Bone Mineral Content divided by projected area of analysis: \( aBMD \, \text{gm/cm}^2 \)

- Fails to account for out of plane dimension
  - Affected by the SIZE of the bone (diminutive in SMA)

- Results reported as a Z-score = number of Standard Deviations different from age matched able bodied children, use height matched instead

\[ aBMD = \frac{\text{BMC}}{\text{Area}} \]
DXA Sites Evaluated

- Osteopenia appears initially @ sites comprised of trabecular (spongy) bone
- Trabecular bone metabolically more active than cortical bone
- Hip and spine contain greater proportion of trabecular bone
- However hip and spine may be abnormal in SMA
Limitations of DXA in Children with SMA

- Diminutive bone size – smaller than age matched peers
  
  ➢ Compare to height matched control rather than age

- Unreliable @ hip and lumbar spine:
  
  ✓ Hip – effected by flexion contractures, subluxation, previous surgery, hardware

  ✓ Spine – effected by pelvic contracture, scoliosis

- BMD lumbar spine unreliable predictor bone status @ proximal femur (Henderson Skel Radiol 26:544, 1997)
Lateral Femoral DXA to Assess BMD non-ambulatory Children

- Distal Femur most common site for pathologic fracture
- Distal Femoral Lateral aBMD predicts osteopenia & fragility fracture better than lumbar aBMD

- **Region 1:** distal femur metaphysis = trabecular bone
- **Region 2:** meta-diaphysis
- **Region 3:** diaphysis = cortical bone

Treatment reduced bone density (osteopenia)

- Passive weight bearing
  - Standing frame (>1.5hr/day x 5 day)
  - may improve bone density
- Maximize calcium and vitamin D supplementation
  - 2000 IU vitD3, 1500mg Ca divided over 3 meals
- If >2 non-traumatic fractures
  - Bisphosphonates: Alendronate (oral), Pamidronate (IV)
Treatment of Fractures

- Treat long bone fractures w/ surgical stabilization to maintain wt bearing potential and to facilitate transfers

  - Avoid prolonged cast immobilization
    - Early mobilization
Hip

- Instability
- Dysplasia
- Dislocation
Who is at risk for hip instability

- Risk for Hip & Spine deformity inversely correlated with physical function capability
  - Highest for SMA 1 and 2

- Hip subluxation / dislocation common non-walking children age 5-8 yrs

- Unilateral or bilateral

- Associated with Pelvic Obliquity
Pathophysiology

- Hips normal at birth
- Global symmetric weakness
  - Core and hip girdle muscles
- Ligamentous laxity
- Absence of weight bearing
  - Attenuates mechanical stimulus for growth and remodeling hip joint → Coxa Valgus + persistent femoral anteversion
- Associated with Pelvic Obliquity and Scoliosis (unclear which comes 1st)
Coxa Valgus: ↑ Neck Shaft Angle
Excessive Femoral Anteversion
Bone Morphology

Femoral Version
Acetabular Dysplasia:
Deficient postero-lateral coverage of hip
Posterolateral Migration of hip
Lateral Migration

Posterior Migration

Postero-lateral Migration
Center of Rotation Δ
Summary: Anatomical Changes

- Coxa valgus reduces efficiency (moment arm) hip abduction by gluteus medius (swing phase of gait)
- Upward and lateral migration femoral head out of acetabulum
- Maturation of acetabulum retarded by pressure from subluxed femoral head on lateral pelvic apophysis
  - Greatest when hip > 50% subluxed
- Posterior and superior acetabular deficiency
Physical Exam

• Young children – hip very unstable, pops in/out of joint as hip is abducted – adducted during diaper changes, toilet/hygiene

• Older children – dislocated hip becomes “stiff”
  — Symmetric loss of hip abduction ± flexion contracture
  — Asymmetric abduction = windblown hip deformity, pelvic obliquity, scoliosis

• *Physical exam alone unreliable to dx hip subluxation*
“Windblown” Hips

• Adduction one hip, Abduction other
• ↑ pelvic obliquity → ↑ risk unilateral hip dislocation
  — not seen in children with level pelvis
• Hip dislocates on side of elevated hemiplevis
• Relationship hip instability → pelvic obliquity → scoliosis
  — Hip subluxation occurs before scoliosis
  — ↑ pelvic obliquity → ↑ Scoliosis
  — Scoliosis less frequent in children without hip instability
  — Result of worsening muscle weakness?
Monitoring

- *Supine AP hips yearly* - screen for dysplasia
- *Standing AP* (if possible) – wt bearing provocative test for hip subluxation
- Evaluate whether hip reduces in Abduction
Monitoring

Migration index = width uncovered head / total width head

A/B > 40% = hip at risk

2D Lateral Migration Percentage

Perkins

Hilgenreiner

80%

20%
Monitoring

Acetabular index = angle formed by lateral corner acetabular roof with horizontal line

(> 30° = dysplasia)
Additional Imaging Studies: CT with 3D Reconstruction

- Assess deformity of femoral head
- Assess location of acetabular deficiency
- Images through distal femur to calculate femoral anteversion
  
  — (May be adversely affected by coxa valgus)
Treatment of subluxated hips

Controversial

• Few natural history studies untreated hips (Sporer & Smith JPO 2003)
  — >50% with hip instability
  — Few had pain at long term follow-up
  — Few had functional limitations with seating, positioning, hygiene
  — None had skin breakdown, ulcers
  — **BUT** Pelvic Obliquity & Scoliosis in 91% non-ambulatory children

• Recurrence of coxa valgus and hip subluxation reported after surgical treatment in young children
My Opinion

- No controversy for children who walk – Reconstruct hip if unstable
- Non-walking children:
  - Prevent hips from dislocating for comfort, maintenance pelvic alignment, improved sitting balance
  - May help prevent pelvic obliquity → scoliosis
  - Less complicated surgery children <10yrs
  - Primary disease persists – recurrent coxa valgus & subluxation may occur – revision may be required
Non-surgical treatment

- PT – maintains hip motion, promotes wt bearing (standers)
  - No evidence that prevents hip subluxation
  - However Standing ~ 2hr/day X5 day/wk helps maintain bone mass – may prevent fragility fractures
- Abduction bracing
  - No evidence that prevents hip dislocation long term
Indications for Surgical Reconstruction

- Child > 4 y/o, Migration > 40%  
  - <4 y/o, 96% will lose correction neck-shaft angle and rotation

- Child > 8y/o, Acetabulum shallow (acetabular index > 25°)  
  - Must address both sides of hip joint – acetabulum, femur

- **Comprehensive approach:**  
  - *Soft tissue release + Reconstruct both sides of hip joint – femur + acetabulum*
Varus Shortening Derotation Intertrochanteric Osteotomy (VDRO)

- **Do both hips** (even if only one hip subluxed)

- Unilateral VDRO may result in pelvic obliquity

- **Adduction osteotomy**
  - must have sufficient (>45°) abduction → adductor tenotomy first
Varus Derotation Osteotomy (VDRO)

- Lateral approach proximal femur,
- Set neck shaft angle: 100° non-walkers, 120° walkers
- Shorten femur, derotate femur (set anteversion ~15°)
- Rigid Hip Plate for fixation to allow early Range of Motion

- Remove 1-2 cm from Shaft → lengthens Hams, Rectus Femorus
- Recess iliopsoas tendon
- Medialize shaft To maintain Mechanical axis
Indications for Acetabular Osteotomy

- Shallow hip socket (Acetabular Index > 25°)
- Hips that do not reduce concentrically after VDRO

- Type of acetabular osteotomy depends on patient age (open vs. closed tri-radiate cartilage), ambulatory status, extent degenerative changes femoral head & acetabulum
Pemberton-Dega Acetabuloplasty

- Incomplete osteotomy above sourcil
- Hinges on tri-radiate

Reduce radius of curvature postero-lateral deficiency to contain hip

Interposed autograft stable
**Chronic Dislocated Hips in Young Adults**

- Leave out if painless
- *IF* Pain & deformity interfere w/ sitting & hygiene:
  - Procedures to relieve pain / improve ROM
  - Perform bilaterally if contralateral hip located to avoid pelvic obliquity (?)
Peri-operative Management

• Epidural anesthesia 3 days post-op
• Diazepam if muscle spasms
• A-frame long leg casts for 3 weeks
  — No SPICA casts
  — Be vigilant for skin breakdown, pressure ulcers
• Sitting, Active assisted ROM post-op day 2
• Chest PT, incentive spirometry for pulmonary toilet
• DVT prophylaxis older children (LMW heparin)
• Aggressive PT x 6 mos: strengthen hip girdle, trunk, quads, gait training (SMA 3)
Complications

• Depends on co-morbidities
  — Feeding/constipation Pulmonary

• **Implant fails** osteopenic bone
  — Use high angle blade plate

• **Decubitus ulcers** from cast

• **Fragility Fractures**
  — No SPICA casts