Orthopaedic Care in Duchenne Muscular Dystrophy

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Duchenne Muscular Dystrophy

- Progressive dystrophy leads to progressive muscular weakness and loss of function.
- Subsequent problems include joint contractures, spine deformity, low bone mineral density and fractures.
- These problems can be due to the progressive muscular dystrophy, secondary effects from weak muscles and the side effects of treatment (steroids).
- The problems vary as the disease evolves.
Aims of Orthopaedic Management

- Preserve motor function
- Minimize joint contractures
- Promote bone health
- Manage spine deformity
  - Apkon et al Pediatrics 2018
DMD - Progressive loss of function

- Difficulty with walking at end of first decade
  - Walk longer with prednisone and deflazacort
- Falling becomes common
  - Unable to get up independently
- Unable to climb stairs
- Ankle equinus initially helps stability but as it becomes severe limits function
- Quadriceps weakness
Contractures

- Splinting
- Physical therapy
- Occupational therapy
- Contractures
  - Mild tendoachilles tightness in 3-6 yo
  - Later- hip flexion and abduction contractures and progressive equinus
DMD- Historical surgical management near end of ambulation

- **Surgical Management**
  - Heelcord lengthening
  - Anterior transfer tibialis posterior tendon
  - Iliotibial band release
  - Hamstring and rectus femoris lengthening

- **Long leg casts, up walking immediately post-op**

- **KAFO’s with drop locks**
  - Prolongs 1 ½ years household ambulation
Severe equinovarus foot deformities in non-ambulatory child
Ankle function and foot position after TAL and anterior transfer of the Tibialis Posterior
Orthotics to improve function
- WREX
Low Bone Mineral Density

- Pathological fractures are common ~ 30%
  - risk when in wheelchair
- 2° to immobility, diet, steroids and ?
- Splint in position of comfort
- Bisphosphonates
DMD- Fractures

- 9 yo boy
  - on Prednisone
  - walking well
  - fell
  - # left femur
Was the child with a technique that allows weight bearing? If not, he probably won’t walk again.
10 yo boy on Deflazacort presents with onset of back pain
  - L4 vertebral compression fracture
DMD- 17 yo with hip fracture after fall out of wheelchair, treated by resection arthroplasty
Duchenne Muscular Dystrophy

- **Scoliosis**
  - > 90% incidence
    - Incidence is improved by steroids
  - 75% non-ambulatory at diagnosis
  - Peak progression- 13-15 years
    - 1-3° per month
DMD- Untreated Scoliosis

M. Sussman MD
Treatment
- Non-surgical methods may delay but do not prevent progression
  - bracing
  - lordotic positioning
  - wheelchair modification
    - Lateral trunk supports
Duchenne Muscular Dystrophy

- Pulmonary function
  - progressive decrease over lifetime
  - related to curve magnitude and weakness
  - FVC > 35% normal in 90% children with 30° scoliosis
  - Correlated with increased post-op respiratory complications (Miller et al 1992)
    - FVC < 35% - 50% risk
    - FVC > 35% - 11% risk
New Era- Steroids
Deflazacort vs. no treatment

% who did not develop >20° scoliosis

B Alman N Raza D Biggar
JBJS 2004
DMD

- **Surgical Indications**
  - Progressive scoliosis > 20-30° in pre-pubertal child not on steroids. Today more likely older child with progressive scoliosis
  - FVC > 35% normal is ideal but good outcomes can be achieved with more severe compromise with modern pulmonary management

- **Aim of surgery**
  - well balanced spine to provide comfortable seating and a better quality of life than would be possible with untreated scoliosis
  - possibly stabilize pulmonary function and improve longevity
DMD

- Surgical Technique
  - Fusion levels
    - T3 or higher to the sacropelvis - conventional
    - T3 or higher to L5
      - If pelvic obliquity < 15°
- **Sussman - 11 patients**
  - Early spinal fusion (<30-40°), consider fusion to L5 rather than to pelvis in mild curves
    - JPO, 1984

- **Gaine, Lim, Stephenson and Galasko - 85 patients**
  - Fusion to L-5, S-1 or the pelvis gave comparable results
    - JBJS-B, 2004

- **Mubarak, Morin, and Leach - 22 patients**
  - Instrumentation to L-5 is adequate for curves with pelvic obliquity < 10°
    - JPO, 1993

- **Sengupta, Mehidian, McConnell, Eisenstein, and Webb**
  - Fixation to L-5 is adequate if < 20°
    - Used pedicle screws at L-5
  - Greater blood loss and hospital stay with pelvic fixation
    - Spine 2002
DMD- Scoliosis

- 16 yo young man
  - 15° thoracolumbar scoliosis
  - Little spine growth remaining so unlikely to progress or need further treatment
DMD- Scoliosis

Conventional treatment in past
Fusion to L5

- RB - 14 yo
  - 28°, mild pelvic obliquity
15 y, used steroids
Current Standard

- Posterior spinal fusion and segmental instrumentation with pedicle screws
DMD

- Post-op
  - Wheelchair modifications
  - Bracing not necessary
  - Increased height may interfere with transportation
  - May feel unbalanced (leaning tower of Pisa)
  - Anterior thigh pain common but resolves over months
  - Excellent long term results
    - pseudarthrosis rare
DMD Spine Fusion - Results

- No good randomized controlled trials to evaluate effectiveness
- But available studies show a positive impact of spine fusion
  - Function
  - Sitting balance and tolerance
  - Back pain
  - Quality of life
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