Management of Spinal Deformities in Spinal Muscular Atrophy

- CURE SMA 2016 Annual Conference
- 30 June 2017

- Samuel R. Rosenfeld, M.D.
  - CHOC CHILDRENS HOSPITAL
  - UNIVERSITY of CALIFORNIA, IRVINE
Disclosure

- Consultant, MediCrea Spine

- I have no potential conflicts with this presentation
• 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, back and buttock pain.
• 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.
What are the 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.
Surgical Management 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
2 years post op
9 years post op
Surgery age 4 years
6 years post op
7 months post op
6 years post op
Surgical Management 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


Intrathecal Administration of Medication with Spinal Deformities

- Guided placement of intrathecal catheter: ultrasound, fluoroscopy, interventional radiologist
- Procedural sedation, cumulative effects of anesthesia
- Laminotomy, laminectomy to facilitate guided placement of needle, catheter
- Indwelling catheter, reservoir, infusion pump to deliver medications without repetitive dural puncture
- Planning long term intrathecal access at the time of spinal reconstructive procedures
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 Tranfusion

- 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
Please complete your conference survey at this link:

https://www.surveymonkey.com/r/2017SMAAnnualConference

Or fill out the paper survey in your conference folder.

- All participants who complete a survey by 10:30 am on Sunday July 2nd, will have their name entered into a raffle for a brand new iPad!
- The winners will be drawn and announced on Sunday, July 2nd at the Closing General Session/It’s a Wonderful Life.