VEPTR in the Management of SMA
A Case Based Discussion

Patrick J. Cahill, MD

The Children’s Hospital of Philadelphia
Perelman School of Medicine at University of Pennsylvania
Spinal Management of SMA – VEPTR

The Problems

Rib Deformity

Spine Deformity

Respiratory Muscle Dysfunction
SPINAL MANAGEMENT OF SMA – VEPTR

THE PROBLEMS

Orthopedic Surgery Fixes Structural Problems
Spinal Management of SMA – VEPTR

The Problems

Rib Deformity

Spine Deformity
Spinal Management of SMA – VEPTR

Scoliosis

• Incidence
  – In ambulatory patients
  – In non-ambulators

• Onset
  – Average 40° in 0-4 y/o
  – Average 83° in 14-15 y/o
Spinal Management of SMA – VEPTR

Scoliosis

• Pattern
  – C-shape most common but S-shape accounts for 1/3 of type III scoliosis

• Rate of Progression
  – 5-15°/y
  – 8°/y
    • Rodillo et al.
  – Twice as fast in non-ambulators
    • Granata et al.
    • Robinson et al. *Euro Spine J* 1995
Impact of Scoliosis on PFTs

- 4.7% reduction in vital capacity for every 10° increase in Cobb angle
- 3.3% reduction in peak flow for every 10° increase in Cobb angle

Spinal Management of SMA – VEPTR

Spinal Fusion

  - 16 pts. Treated with Harrington rods and fusion
  - PFTs improved slightly in immediate post-op period
  - PRO questionnaire: wheelchair comfort improved in 90%

- Fujak et al. *Arch Orthop Trauma Surg* 2012
  - 24 pts. Treated with fusion
  - Stable PFTs @1y post-op
  - **Scoliosis progressed 1°/y** over 6y post-op follow-up
Spinal Management of SMA – VEPTR

Spinal Fusion

• Does spine fusion slow pulm. decline?

• Chng et al. *J Paediatr. Child Health* 2003
  – FVC declined 7.7%/y pre-op
  – FVC declined 3.3%/y post-op
Spinal Management of SMA – VEPTR

Spinal Fusion

- Rib Deformity
- Spine Deformity
  - Spinal fusion
- Respiratory Muscle Dysfunction
Spinal Management of SMA – VEPTR
Spinal Fusion
Spinal Fusion

Spinal fusion is morbid

- 8-12h surgery
- 500cc-2L blood loss
- Mortality increases with PFTs<30% pre-op
Collapsing Parasol Deformity

A narrowing of the rib cage more pronounced at the upper ribs

Described by Campbell in 2007

Parasol Rib Deformity in Hypotonic Neuromuscular Scoliosis: A New Radiographical Definition and a Comparison of Short-term Treatment Outcomes With VEPTR and Growing Rods.

Livingston, Kristin; Zurakowski, David; Snyder, Brian; MD, PhD

Spinal Management of SMA – VEPTR

Rib Deformity

- Parasol Score = (T6 convex hemithoracic width/T6 concave hemithoracic width) × (T6 thoracic width/T12 thoracic width)
  - Livingston et al. *Spine* 2015
10 y/o female with history of type II SMA and progressive scoliosis

- Difficulty sitting

- 3 affected older siblings treated with PSF
Spinal Management of SMA – VEPTR

VEPTR Treatment

- Poor head control
- Pelvic obliquity
- areflexic
Treatment course

- EBL: 100cc
- Hospital discharge POD: 6
- 5 subsequent lengthening surgeries
Spinal Management of SMA – VEPTR

Spinal Fusion

Rib Deformity
- VEPTR

Spine Deformity
- Spinal fusion
- VEPTR

Respiratory Muscle Dysfunction
Spinal Management of SMA – VEPTR

**VEPTR Outcomes**

- **No immediate change in PFTs**
  - Mayer and Redding *JPO* 2009
    - All diagnoses
  - Tobert and Vitale *Am J Orthop* 2013
    - 3pt. Series of SMA
    - 28 mo. f/u

- **Increase in weight and Hb**
  - Skaggs, Sankar et al. *Spine* 2009
  - Caubet et al. *Spine* 2009
VEPTR Gantry

TREATMENT EVOLUTION
Spinal Management of SMA – VEPTR
Current Preferred Treatment

8 y/o male with Type Ib
left midline caudal
Two patients with 24 months' follow-up

Cobb angle, was $67^\circ$ (SD 10°) before implantation and $29^\circ$ (SD 4°) at 24 months

Throughout follow-up, both patients had no pain, had good functional outcome, and were satisfied with the procedure

No MCGR-related complications were noted
Spinal Management of SMA – VEPTR

Collapsing Parasol Deformity

Unknowns

- Timing of appearance
- Rate of progression
- Relationship to scoliosis
Spinal Management of SMA – VEPTR

Collapsing Parasol Deformity

Age 8y 11 mo.
Spinal Management of SMA – VEPTR

Collapsing Parasol Deformity
Spinal Management of SMA – VEPTR

Collapsing Parasol Deformity

Age 9y
Spinal Management of SMA – VEPTR

Collapsing Parasol Deformity

Age
15y