Does Rib-Based Distraction Control Curve Progression and Prevent Parasol Deformity of the Chest in Scoliosis Associated with Congenital Myopathy?

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Disclosures

- Smith: Synthes spine (Consultant; Royalties)
- Mickelson: Nothing to disclose
- D'Amato: Nothing to disclose
- Sawyer: Nothing to disclose
- Vitale: Biomet (Consultant, Royalties) Stryker (Consultant) AO Spine/CWSDSG (Research)

Background

- Congenital Myopathy:
  - Multiple forms
  - Progressive weakness of the chest
  - Variable prognosis
  - Scoliosis common and typically progressive
- Early Fusion and progressive weakness produces a parasol deformity of the chest.

Hypothesis

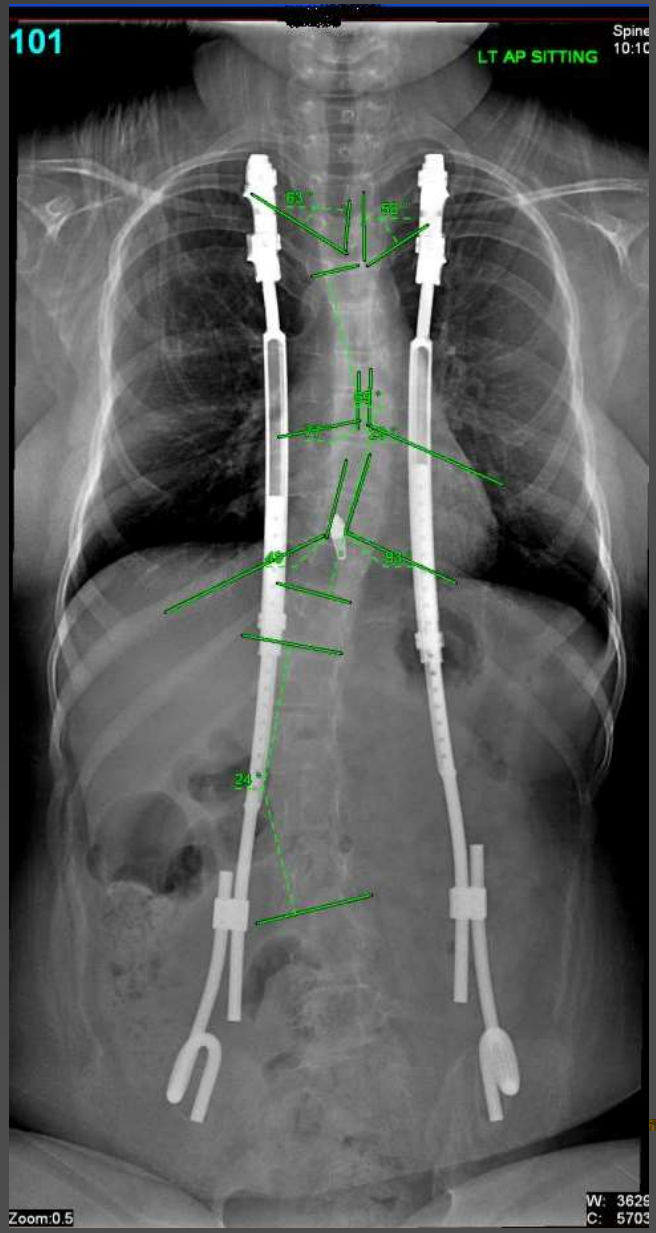
Rib-Based Distraction techniques can effectively control curve progression in scoliosis associated with Congenital Myopathy and prevent development of parasol deformity of the chest.

Methods

- IRB Approved Multicenter Retrospective Study
- CWSDSG Database
- Diagnosis of EOS associated with Congenital Myopathy
- Minimum follow-up of 9 months

Measurement of Scoliosis and Parasol Deformity

- Modified RVAD at T4, T8, T10
- Coronal and Sagittal Cobb Angles



Clinical Assessment

- Diagnosis
- Demographics
- Respiratory Status
- Cobb angles
- Adverse Events
- Follow-up

Results

- 14 Children with Congenital Myopathy treated with Rib-Based Distraction and complete data.
- Diagnosis:
  - SMA (8)
  - Merosin Deficient (2)
  - Myotonic Dystrophy, Congenital, Mitochondrial, Myotubular, (1 each)
- Mean Age: 6.03 years (2-9 yrs)
- Median Follow-up: 27 months (9-62 months)

Constructs

- Rib- Pelvis Bilateral: 10
- Rib – Pelvis Unilateral: 1
- Rib-Pelvis & Rib-Spine: 1
- Rib-Spine: 2

Results: Cobb Angles

- Coronal: 58° → 39°
- Sagittal: 51° → 39°

Results: Modified RVAD

- Eighty-four RVAD angles measured
- Thirty-five of 84 RVAD angles improved
- Main improvement seen at T-10 (improvements =16, patients=11)
- Eleven angles improved at T4 (patient=8) and 8 improved at T8 (patients=8).

RVAD Improvements with Rib-Based Distraction

	Count of Improvements per Level	Percentage of Improvements
T4	11	13.10%
T8	8	9.52%
T10	15	17.86%
total	34	40.48%

n=84

Adverse Events

- 4 total:
  - Infection
  - Wound Dehiscence
  - Persistent Pain
  - Hardware failure

Merosin Deficient Myopathy treated with bilateral rib-pelvis technique: 62 months after initial implant @ age 2+5

Conclusions

- Scoliosis associated with congenital myopathy can be effectively managed without early fusion
- Rib distraction techniques seem to reduce or prevent the development of parasol deformity
- Adverse events were acceptable for this population of children

Conclusions

Further study with larger numbers of patients is needed to confirm these findings.

A comparison with spine based distraction techniques and parasol deformities would be valuable.

Thank You