

Improvement of Pulmonary Function Measured by Patient-Reported Outcomes in Patients with Spinal Muscular Atrophy after Rib-based Growing Constructs

John Mueller, BA, Hiroko Matsumoto, MA, PhDc, Patrick Cahill, MD, Peter Sturm, MD, John Smith, MD, David P. Royce, MD, Michael G. Vitale, MD, MPH,
Children's Spine Study Group



Disclosures

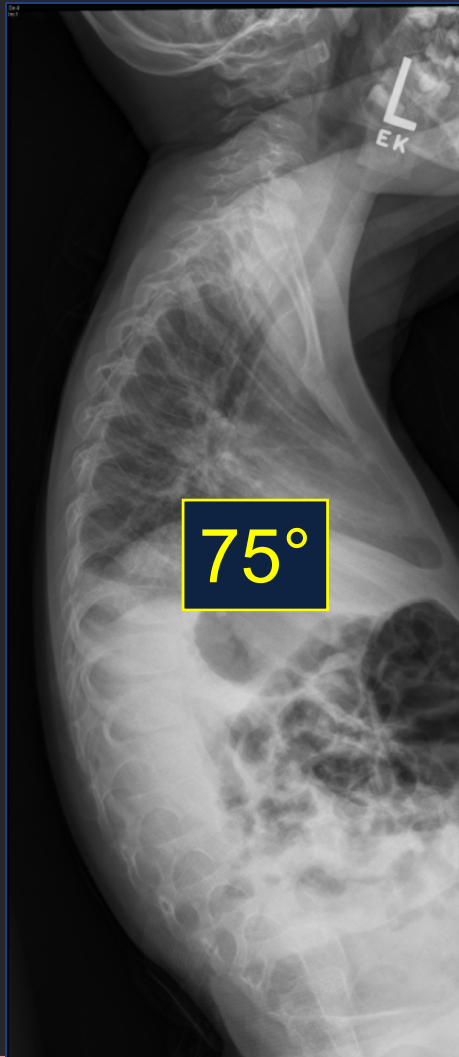
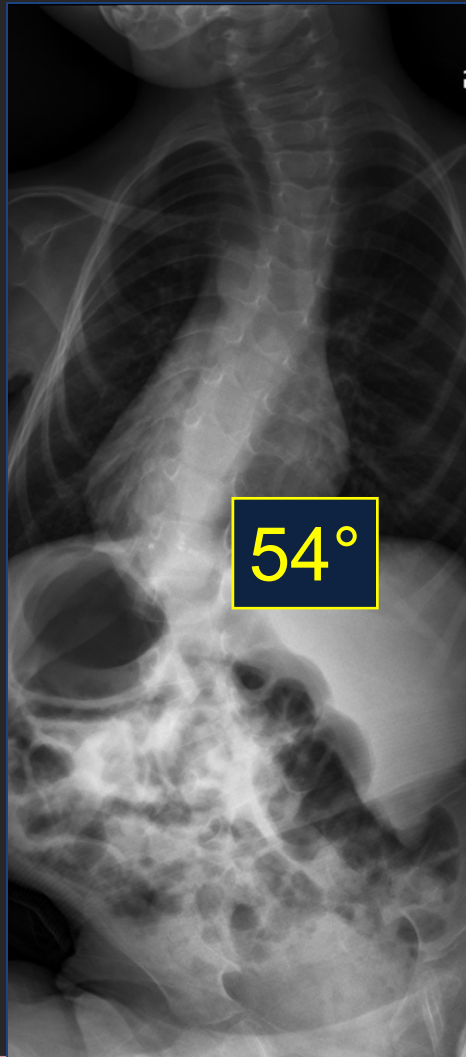
- John Mueller, BA: None
- Hiroko Matsumoto, MA, PhDc: American Academy for Cerebral Palsy and Developmental Medicine: Board or committee member; Children's Spine Study Group: Board or committee member; Scoliosis Research Society: Board or committee member
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- John Smith, MD: Biomet: Paid consultant; Chest Wall and Spine Deformity Research Foundation: Board or committee member; DePuy, A Johnson & Johnson Company: IP royalties; Paid consultant; Ellipse Technologies: Paid consultant; Globus Medical: Paid consultant; Spineguard: Paid consultant
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- Children's Spine Study Group: DePuy, A Johnson & Johnson Company: Research support

Introduction

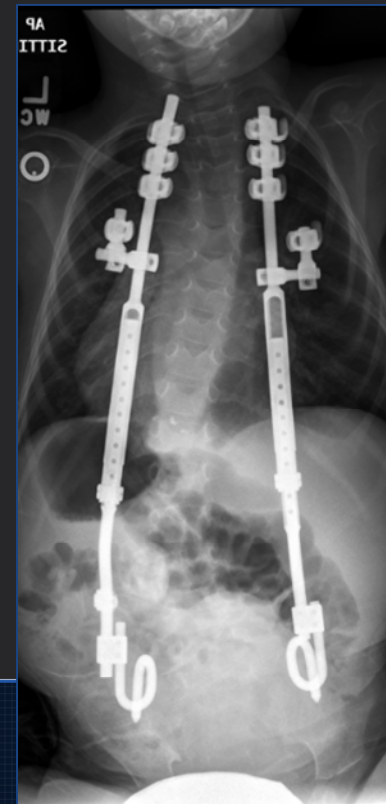
- **Spinal muscular atrophy (SMA) is characterized by motor neuron degeneration leading to progressive muscular weakness, secondary scoliosis, and restrictive lung disease**
- **One of the goals of Growth-Friendly Instrumentation is to decrease the rate of pulmonary function (PF) decline in patients with SMA**

5 yo Spinal Muscular Atrophy Type 2

C-EOS: N3+P2



- Significant pulmonary improvement post operatively



Patient OK:

Pulmonary Function in SMA

- FVC decline in SMA approximately 5-8% per year prior to surgery
 - Some evidence exists that the rate of decline decreases 2-4% per year following surgery (Chung 2003, Chua 2016)
- PFTs are difficult to obtain in young children
- The Early Onset Scoliosis Questionnaire (EOSQ-24) has been shown to be a valid patient-reported method of determining quality of life in patients with EOS and includes pulmonary domains

Purpose

- To evaluate changes in **self-reported** pulmonary function in SMA patients after rib-based growing construct implantation utilizing **the EOSQ-24 Pulmonary Function** domain

Methods

- The Children's Spine Study Group (**CSSG**) was queried for patients with **SMA treated with rib-based growing constructs** between 2005 - 2015 in this **multi-center retrospective cohort study**

EOSQ and PFT Collection Times

Both **EOSQ-24 PF domain scores** and **FVCs** were collected:

- 1 month preoperatively
- 1-year postoperatively
- 2-years postoperatively

Results

- **49 patients** met the inclusion criteria:
 - Average Pre-op Major Coronal Curve angle = 58.2°
 - 6.9 years of age at implant

EOSQ-24 PF	
Time Point	Patients Included
Pre-op	14
1-Year	10
2-Year Post-op	21

PFTs	
Time Point	Patients Included
Pre-op	17
1-Year	8
2-Year Post-op	13

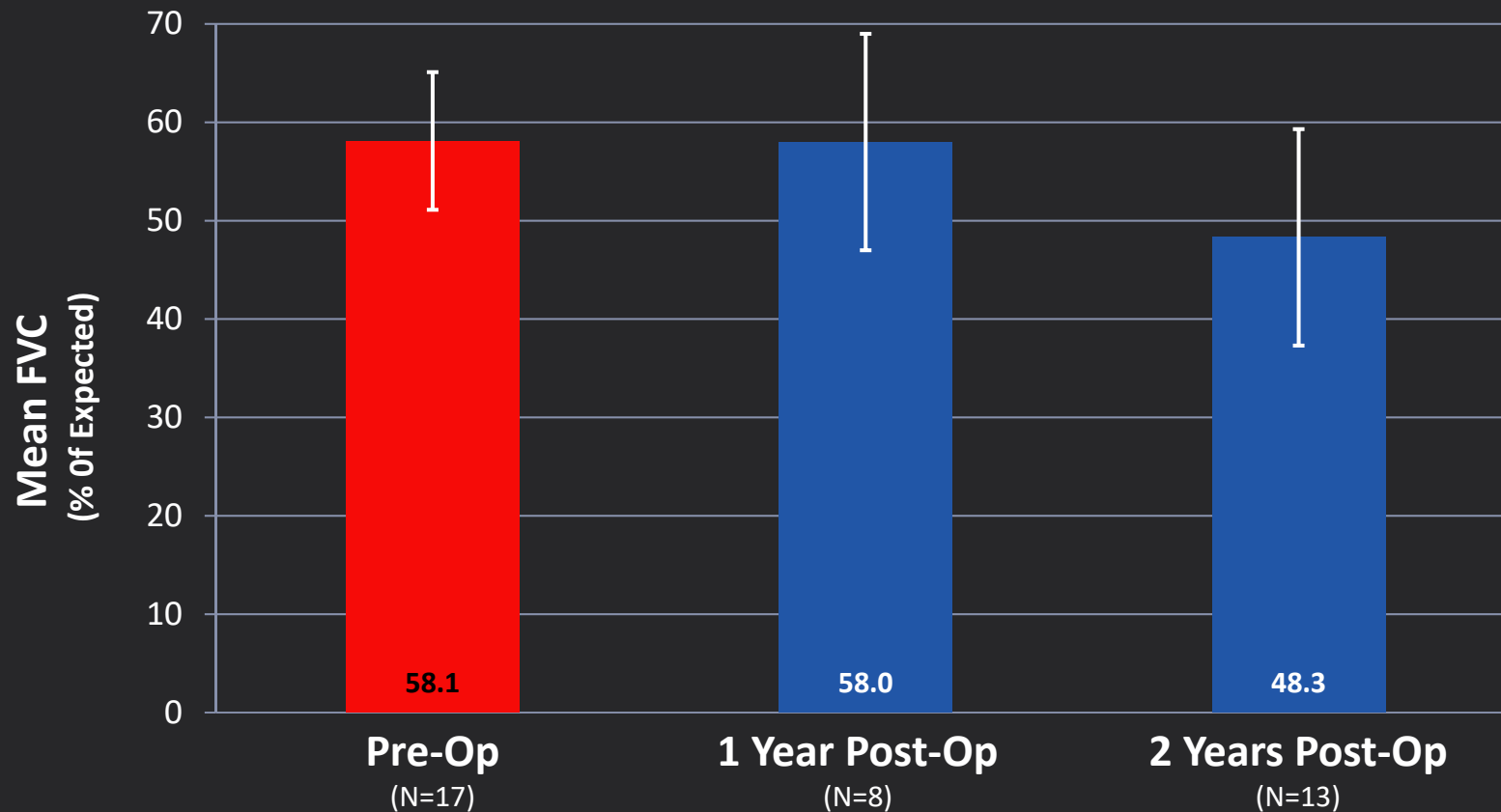
- Average initial major **coronal curve correction was 33.5%** (19.5°)

FVC Decreased at 2-year Follow-up

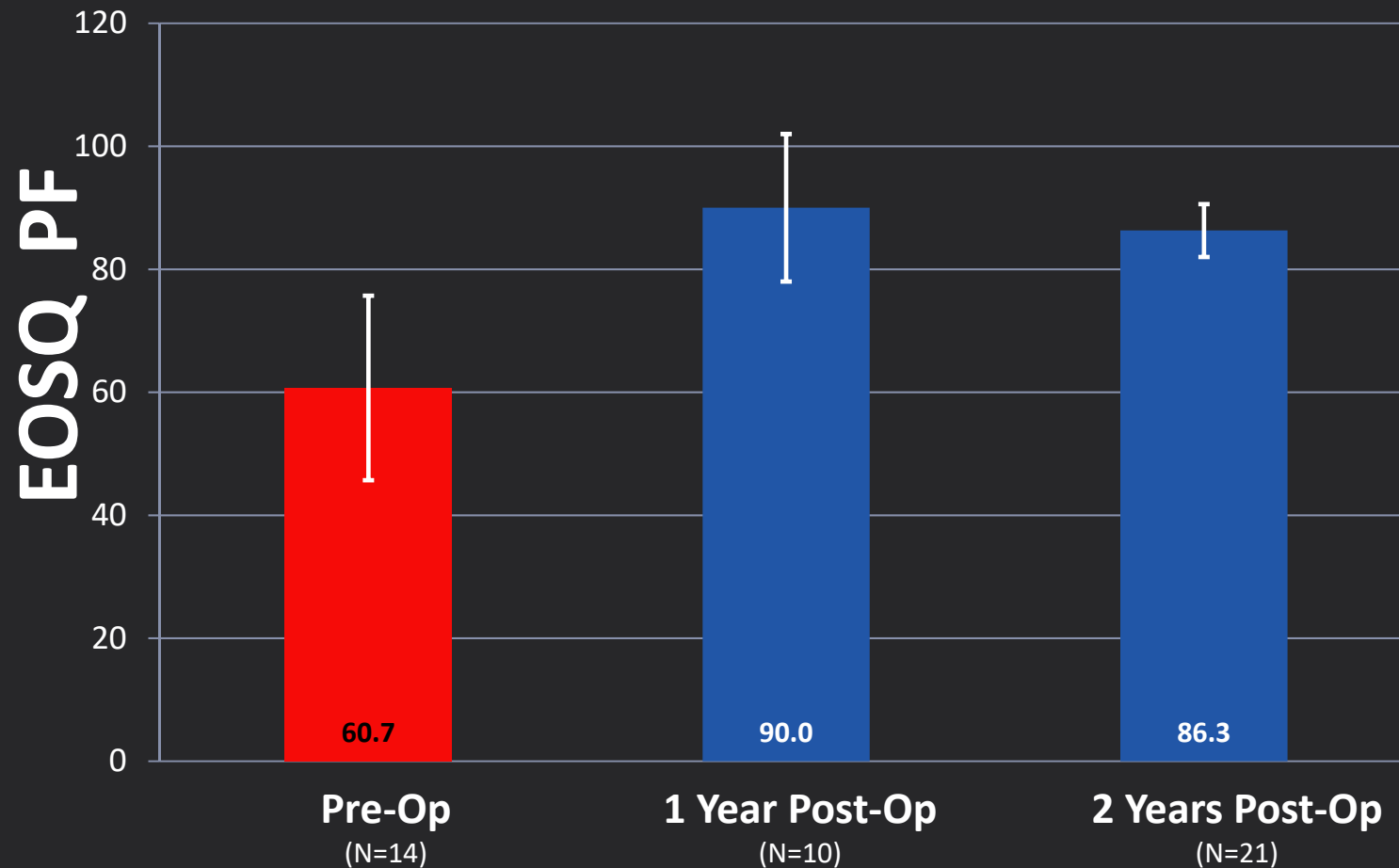
(Small Subset of Cohort)

(no change at 1 year post-op)

FVC Pre and Post Rib-based Growing Constructs Implantation



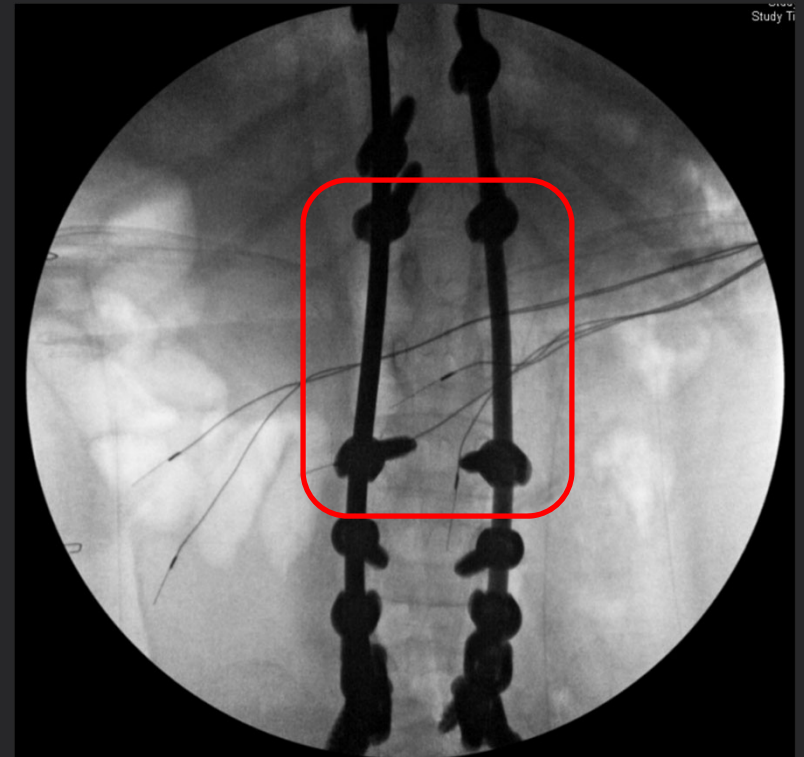
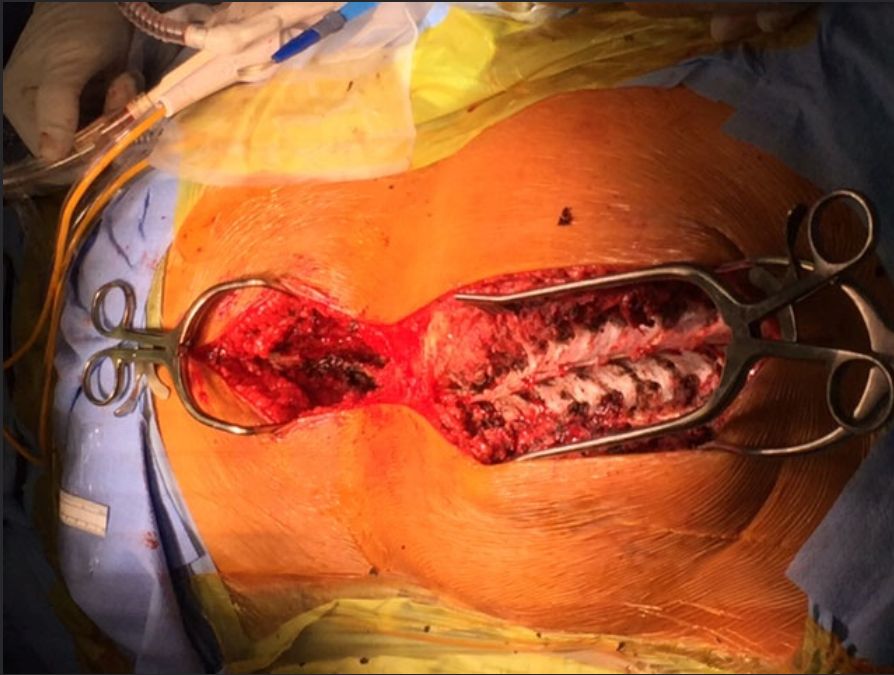
EOSQ-24 PF Scores Increased Postoperatively



Conclusions

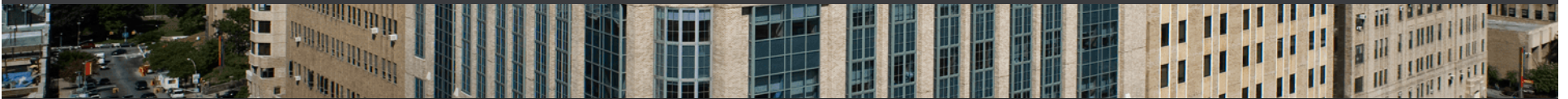
- Limited data on effect of growth friendly instrumentation on changing natural history of pulmonary demise in SMA EOS
- Pulmonary function in SMA patients measured by *patient-reported outcomes* demonstrated significant improvement after rib-based growing construct implantation
- ? Effect of Cognitive Dissonance

Remember “Skip Constructs” in SMA Fusion !



Do **not** expose/fuse the L1-L2 levels after the skin (SKIP) to allow continued delivery of intrathecal drug for his SMA

Thank you!



AMAZING
THINGS
ARE
HAPPENING
HERE

for kids



Hiroko Matsumoto

hm2174@cumc.columbia.edu



NewYork-Presbyterian
Morgan Stanley Children's Hospital



COLUMBIA UNIVERSITY
DEPARTMENT OF ORTHOPEDIC SURGERY
College of Physicians & Surgeons