

# Correlation of Pulmonary Function to a Novel Radiographic Parameter of Collapsing Parasol Deformity in Spinal Muscular Atrophy

---

---

Mitchell A. Johnson, BS; Suzanne Ho, BA; Didja Hilmara, BA; Nirupa Galagedera, BA; Robert M. Campbell, MD; **Patrick J. Cahill, MD**

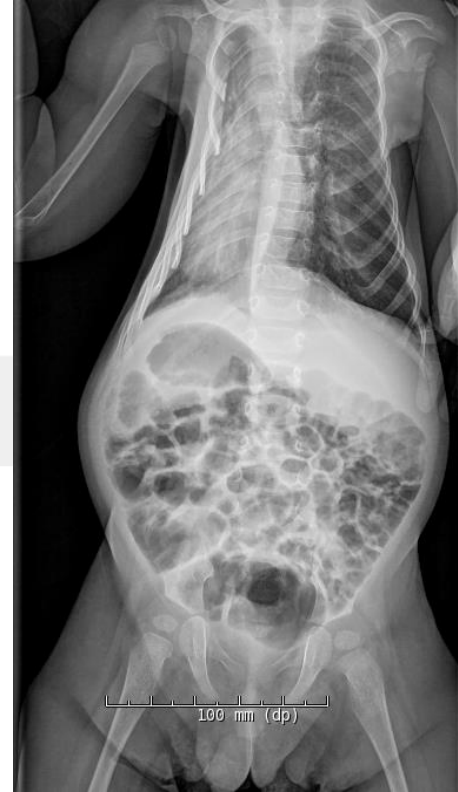


The Children's Hospital of Philadelphia  
Perelman School of Medicine at University of Pennsylvania

---

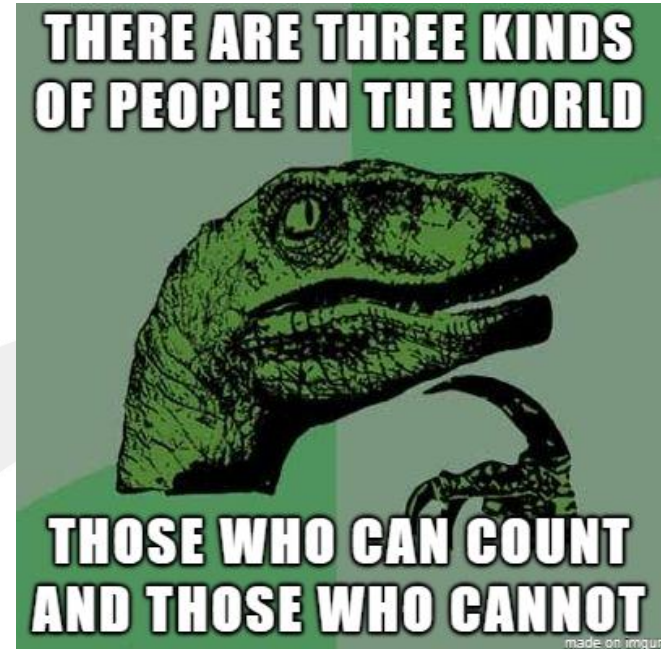
# Spinal Muscular Atrophy

- Degeneration of anterior horn cells
- Proximal muscle weakness
- Parasol deformity
  - Campbell 2007
- Pulmonary decline
  - Bridwell 1999, Brown 1989



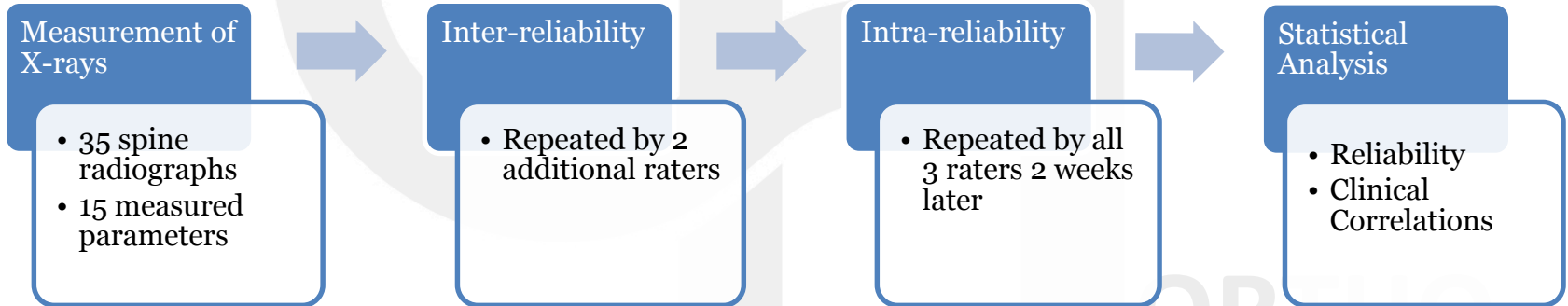
# Background

- Current assessment of rib deformity largely qualitative
- Previous literature does not address:
  - Vertical droop
  - Asymmetric deformity
  - Pulmonary function data
  - Measurements on non-operative or pre-operative patients



# Study Design

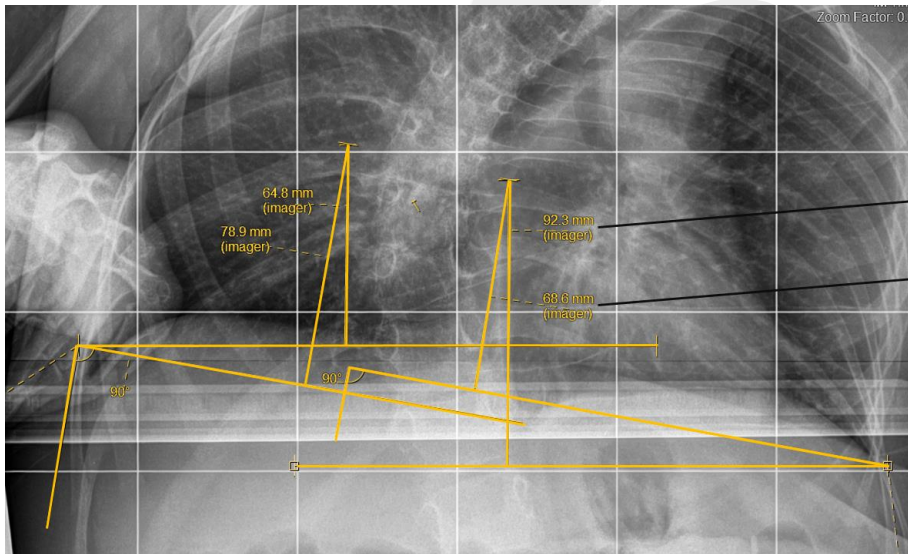
- Retrospective analysis of all patients with Spinal Muscular Atrophy diagnosis seen at single institution from January 2007 to January 2018 with usable scoliosis x-rays
- Measurement process:



## 15 Measured Parameters

### 15 Measured Parameters:

- Rib Vertical Displacement (R, L)
- Thorax Width (T6, Apical)
- Hemithorax Width at T6 (R, L)
  - All displacements measured both in x-y plane and normalized to vertebral endplate



Apical Vertical Displacement

Apical Vertical Displacement Normalized

- Rib Vertical Angle Difference
- Cobb Angle
- Pelvic Obliquity

ORTHO

# Patient Descriptive

---

- 35 patients
  - 22 male, 13 female
- Age:  $8.3 \pm 3.7$  years
- Weight:  $26.5 \pm 13.3$  kg
- Height:  $123.1 \pm 21.8$  cm
- BMI:  $17.3 \pm 4.7$
- Cobb angle:  $33.6 \pm 26.4^\circ$

# Statistical Analysis

## Part 1: Assessment of Inter- and Intra- Reliability of Measurements

- Intraclass Correlation Coefficient (ICC)
  - n= 28
  - ICC > 0.75 accepted as strong reliability

## Part 2: Correlation to Pulmonary Function Testing Data (Forced Vital Capacity and Forced Expiratory Volume)

- Pearson Correlation Coefficient
  - n= 21

# Results of Reliability Assessment

---

- 14 parameters demonstrate strong intra- and inter-rater reliability
- Intraclass Correlation Coefficient (ICC)
  - Intra-reliability= 0.86 – 0.99
  - Inter-reliability= 0.79 – 0.97



# Identification of Rib Collapse Index and Pulmonary Correlations

$$A = \frac{T6 \text{ concave hemithoracic width}}{T6 \text{ convex hemithoracic width}}$$

B= A normalized

$$C = \frac{\text{Apical concave hemithoracic width}}{\text{Apical convex hemithoracic width}}$$

D= C normalized

$$E = \frac{\text{Apical rib convex vertical displacement}}{\text{Apical rib concave vertical displacement}}$$

F= E normalized

## Rib Collapse Index:

$$G = B * F * \frac{\text{Apical thoracic width normalized}}{T6 \text{ thoracic width normalized}}$$

H= Rib Vertebral Angle Difference (RVAD)

I= Cobb Angle

- A-I correlated to Pulmonary Function Testing data
- **Rib Collapse Index**
  - Intra-rater ICC: 0.961 – 0.995
  - Inter-rater ICC: 0.937 – 0.952
  - FVC: R= -.607, p=.004
  - FEV1: R= -.528, p= .014

# Potential Implications

- Quantification of progression of parasol deformity that takes into account vertical droop
  - Guide timing of treatments and surgical intervention
- Ability to predict pulmonary function when testing difficult to perform due to respiratory status or age



## Conclusion

---

- We identified 15 parameters measured on x-rays that demonstrate strong intra- and inter- reliability
- We found a novel, reliable, clinically significant quantification (Rib Collapse Index) of parasol rib deformity in patients with SMA
- We demonstrated that the Rib Collapse Index can be used as a predictor of pulmonary function in SMA patients

## References

- Bridwell KH, Baldus C, Iffrig TM, Lenke LG, Blanke K. Process measures and patient/parent evaluation of surgical management of spinal deformities in patients with progressive flaccid neuromuscular scoliosis (Duchenne's muscular dystrophy and spinal muscular atrophy). *SPINE*. 1999 Jul 1; 24(13): 1300-9.
- Brown JC, Zeller JL, Swank SM, Furumasu J, Warath SL. Surgical and functional results of spine fusion in spinal muscular atrophy. *SPINE*. 1989 Jul; 14(7): 763-70.
- Chng SY, Wong YQ, Hui JH, Wong HK, Ong HT, Goh DY. Pulmonary function and scoliosis in children with spinal muscular atrophy types II and III. *J Paediatr Child Health*. 2003 Dec; 39(9): 673-6.
- Campbell RM, Smith MD. Thoracic insufficiency syndrome and exotic scoliosis. *J Bone Joint Surg Am*. 2007 Feb; 89 Suppl 1: 108-22.
- Finkel R, Mercuri E, Mayer O, Simonds A, Schroth M, Graham R, Kirshner J, Iannaccone S, Crawford T, Woods S, Muntoni F, Wirth B, Montes J, Main M, Mazzone E, Vitale M, Snyder B, Quijano-Roy S, Bertini E, Davis R, Qian Y, Sejersen T for the SMA care group. Diagnosis and management of spinal muscular atrophy: Part 2: Pulmonary and acute care; medications, supplements and immunizations; other organ systems; and ethics. *Neuromuscular Disorders* 2018; 28: 197-207.
- Lenhart R, Youlo S, Schroth M, Noonan K, McCarthy J, Mann D, Hetzel S, Sund S, Halanski M. Radiographic and respiratory effects of growing rods in children with spinal muscular atrophy. *J Pediatr Orthop*. 2017; 37 (8): e500-e504.
- Livingston K, Zurakowski D, Snyder B; Growing Spine Study Group; Children's Spine Study Group. Parasol rib deformity in hypotonic neuromuscular scoliosis: a new radiographical definition and a comparison of short-term treatment outcomes with VEPTR and Growing Rods. *SPINE*. 2015 Jul 1; 40(3): E780-6.
- Mercuri E, Finkel R, Muntoni F, Wirth B, Montes J, Main M, Mazzone E, Vitale M, Snyder B, Quijano-Roy S, Bertini E, Davis R, Mayer O, Simonds A, Schroth M, Graham R, Kirshner J, Iannaccone S, Crawford T, Woods S, Qian Y, Sejersen T for the SMA care group. Diagnosis and management of spinal muscular atrophy: Part 1: Recommendation for diagnosis, rehabilitation, orthopedic and nutritional care. *Neuromuscular Disorders* 2018; 28: 103-115.
- Portney LG, Watkins MP. Reliability of measurements. In: Portney LG, Watkins MP. *Foundations of clinical research: applications to practice*. 3<sup>rd</sup> ed. Philadelphia (PA): F.A. Davis Company; 2015: 77-94.
- Shrout PE, Fleiss JL. Intraclass correlations: uses in assessing rater reliability. *Psychol Bull* 1979; 86: 420-8.