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

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Spinal Muscular Atrophy

• Degeneration of anterior horn cells
• Proximal muscle weakness
• Parasol deformity
  – Campbell 2007
• Pulmonary decline
  – Bridwell 1999, Brown 1989
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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
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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:
  - Measurement of X-rays
    • 35 spine radiographs
    • 15 measured parameters
  - Inter-reliability
    • Repeated by 2 additional raters
  - Intra-reliability
    • Repeated by all 3 raters 2 weeks later
  - Statistical Analysis
    • Reliability
    • Clinical Correlations
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
- Rib Vertical Angle Difference
- Cobb Angle
- Pelvic Obliquity
35 patients
- 22 male, 13 female
Age: 8.3 ± 3.7 years
Weight: 26.5 ± 13.3 kg
Height: 123.1 ± 21.8 cm
BMI: 17.3 ± 4.7
Cobb angle: 33.6 ± 26.4°
### 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
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 * $\frac{\text{Apical thoracic width normalized}}{\text{T6 thoracic width normalized}}$

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: $R = -.528, p = .014$
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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
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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
**SMA: Reliable Quantification & Pulmonary Function**

**References**


