The Classification for Early-Onset Scoliosis (C-EOS) & Outcomes in Early-Onset Scoliosis

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-Disclosures-

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Other: CWSDSG - BOD
POSNA - BOD

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Improving the Evidence Base in EOS

Development of a Research Infrastructure
Via four parallel efforts

Endpoints
- Development and Validation of a Disease Specific QoL Measure

Equipoise
- Evaluating clinical equipoise in the field of EOS

Classification
- Developing an EOS Subgroup Classification Schema to facilitate collaboration and communication

Consensus
- Determining inclusion criteria, treatment options and outcome measures for future research efforts

Purpose of the Classification for EOS (C-EOS)

To classify EOS patients in order to:

1) **Predict** the disease course of individual patients

2) **Prognosticate** and determine beneficiaries of differing treatment modalities

3) **Improve communication** among EOS providers and facilitate research
Key ‘Philosophical’ Aspects of the (C-EOS)

• **Comprehensive**
  Applicable to all EOS pts

• **Practical**
  Utilized in daily practice

• **Prognostic**
  Predictive of course

• **Guide**
  Informs treatment decisions

An EOS ‘One Liner’

Methods: Validation Pathway

- **Phase 1**
  Development phase
  Interviews, Literature Review and Working Session
  Classification proposal
  Pilot agreement studies

- **Phase 2**
  Reliability and accuracy in clinical setting
  Multicentre agreement study
  Reliability Testing

- **Phase 3**
  Association with patient outcome(s)
  Clinical studies
  Future Work

Iterative Survey & Group Discussion

- **Group Discussion**
  - Proposing Variables
    - POSNA – May 2011

- **Iterative Survey**
  - Assessing Variables
    - May-July 2011

- **Group Discussion**
  - Finalizing Variables
    - ICEOS – November 2011

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Results of Variable Identification Survey

<table>
<thead>
<tr>
<th>Variable</th>
<th>Not Useful</th>
<th>Useful</th>
<th>Essential</th>
<th>CVR</th>
<th>Sum of Ranks</th>
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<td>4</td>
<td>0</td>
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</tbody>
</table>
C-EOS Variables: Etiology

• Challenging variable due to heterogenous population
• Numerous iterations based on study group feedback

Etiology

- Idiopathic
- HTNM
- LM
- LTNM
- Syndromic
- Congenital

C-EOS Variables: Etiology

**Congenital/Structural**: Curves developing due to a structural abnormality/asymmetry of the spine and/or thoracic cavity (i.e. hemivertebrae, fused ribs, post-thoracotomy, or CDH)

**Neuromuscular**: Patient with neuromuscular disease (i.e. SMA, Cerebral Palsy, muscular dystrophies, etc.)

**Syndromic**: Syndromes with known or possible association with scoliosis (including spinal dysraphism)

**Idiopathic**: No clear causal agent (can include children with a significant co-morbidity that has no defined association with scoliosis)
C-EOS Variables: Cobb Angle

Cobb Angle: Measurement of major spinal curve in position of most gravity

Cobb Angle (Major Curve):
1: <20°
2: 20-49°
3: 50-89°
4: ≥90°

C-EOS Variables: Kyphosis

Maximum Total Kyphosis: Between any two levels throughout spine

Maximum Total Kyphosis:
(-) <20°
N: 20-49°
(+): ≥50°
### C-EOS Variables: Progression Modifier (Optional)

Minimum of 6 months x-ray follow-up

\[
\text{[Cobb at } t_2] - [\text{Cobb at } t_1] \times 12 \text{ months/year}
\]

[Months between \( t_1 \) and \( t_2 \)]

<table>
<thead>
<tr>
<th>Progression Modifier (Optional)</th>
<th>Description</th>
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<td>P0: &lt;10°/yr</td>
<td></td>
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<tr>
<td>P1:10-19°/yr</td>
<td></td>
</tr>
<tr>
<td>P2: ≥20°/yr</td>
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### C-EOS Finalized

<table>
<thead>
<tr>
<th>Etiology</th>
<th>Cobb Angle (Major Curve)</th>
<th>Maximum Total Kyphosis</th>
<th>Progression Modifier (optional)</th>
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<tr>
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<td>1: &lt;20°</td>
<td>(-) &lt;20°</td>
<td>P0: &lt;10°/yr</td>
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<tr>
<td></td>
<td>2: 20-49°</td>
<td>N: 21-49°</td>
<td>P1:10-19°/yr</td>
</tr>
<tr>
<td></td>
<td>3: 50-89°</td>
<td>(+): ≥50°</td>
<td>P2: ≥20°/yr</td>
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<td>4: ≥90°</td>
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<td></td>
</tr>
<tr>
<td>Idiopathic</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Etiology (In order of priority):**

- **Congenital/Structural:** Curves developing due to a structural abnormality/asymmetry of the spine and/or thoracic cavity; includes hemivertebrae, fused ribs, post-thoracotomy, or CDH.
- **Neuromuscular:** Pt with neuromuscular disease
- **Syndromic:** Syndromes with known or possible association with scoliosis (including spinal dysraphism)
- **Idiopathic:** No clear causal agent (can include children with a significant co-morbidity that has no defined association with scoliosis)

**Cobb Angle:** Measurement of major spinal curve in position of most gravity

**Maximum measurable Kyphosis:** between any 2 levels

**Annual Progression Ratio Modifier (optional):**

- Progression per year;
- min. 6 months between observation

\[
(Cobb \at \ t_2) - (Cobb \at \ t_1) \times 12 \text{ months/}[t_2-t_1]
\]
CASE 1

History:
- 19 mo old female
- 38wk, C-section
- L thoracotomy for PDA repair @ 4 mo, scoliosis noted post-op
- Acquired rib fusion b/w concave 4th-5th rib

Physical:
- Hypotonic UE and trunk, hypertonic LE
- Rigid right thoracic curve

CASE 1: 9 months later

1. Etiology
   - Acquired chest wall deformity → Congenital/structural

2. Cobb Angle
   - 42° → 2

3. Kyphosis
   - Lateral x-ray reveals 35° maximum total kyphosis → N

4. Progression Modifier (optional)
   - \([(42°-27°)/(9 mo.))] \times 12 = 20°/yr → P2

C/2/N/P2
CASE 2

History
- 8 y/o girl w Rett Syndrome
- H/o seizures & hip subluxation
- Non ambulatory

Physical
- Hypokyphotic
- L. lumbar curve when seated
- Variable tone in lower extremities

1. Etiology
   - Rett Syndrome ➔ Neuromuscular

2. Cobb Angle
   - 23° ➔ 2

3. Kyphosis
   - 8° ➔ -

4. Progression Modifier
   (optional)
   - Not Available

M/2/-
CASE 3

History
• 4 y/o girl w/ Congenital Myotonic Dystrophy
  • Mother as well

Physical
• Hyperkyphosis
• Bilateral equinus s/p percutaneous heel lengthening
  – 30° of dorsiflexion
• Full ROM at knees and hips

CASE 3

1. Etiology
• Congenital?
  • Neuromuscular?
  • Syndromic?
  → Neuromuscular by priority

2. Cobb Angle
• 50° → 2

3. Kyphosis
• 96° → +

4. Progression Modifier
   (optional)
   • Not available

M/2/+
CASE 4

History
• 4 y/o girl w/ Pena-Shokeir Syndrome
• Developmentally delayed
• Right hip dislocation
• Nonambulatory
  • Wheelchair

Physical
• Lays comfortably on table
• Stiff left thoracolumbar curvature
  • Rib cage rests on pelvis
• Multiple contractures

CASE 4

7 months later

88°

97°

26°
CASE 4

1. Etiology
   - Pena-Shokier Syndrome

2. Cobb Angle
   - 97° \( \rightarrow \) 4

3. Kyphosis
   - 26° \( \rightarrow \) N

4. Progression Modifier (optional)
   - \[ \frac{(97°-88°)}{(7 \text{ mo.})} \times 12 = 15.4°/\text{yr} \rightarrow P1 \]

S/4/N/P1

Methods: Validation Pathway

- **Phase 1**
  - Development phase
  - Interviews, Literature Review and Working Session
  - Classification proposal
  - Pilot agreement studies
  - Nominal Group Technique: Iterative Surveying and Group Discussion

- **Phase 2**
  - Reliability and accuracy in clinical setting
  - Multicentre agreement study
  - Reliability Testing

- **Phase 3**
  - Association with patient outcome(s)
  - Clinical studies

Future Work

Thank You