



C-EOS

Classification for Early-Onset Scoliosis

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Development and Initial Validation of a Novel Classification System for Early Onset Scoliosis

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-Disclosures-

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Other: CWSDSG - BOD

POSNA BOD

Improving the Evidence Base in EOS

*Development of a Research Infrastructure
Via five parallel efforts*

Endpoints

Development/Validation of a Disease-Specific QoL Measure -- EOSQ

Equipose

Identifying Clinical Equipose in the Field of EOS

Classification-EOS

Development / Validation of Classification for EOS

Complications
Classification

Standardize Way We Define and Report Complications

Clinical Trials

Proximal Anchors: Rib Vs Spine – Retrospective
(Prospective Underway)

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

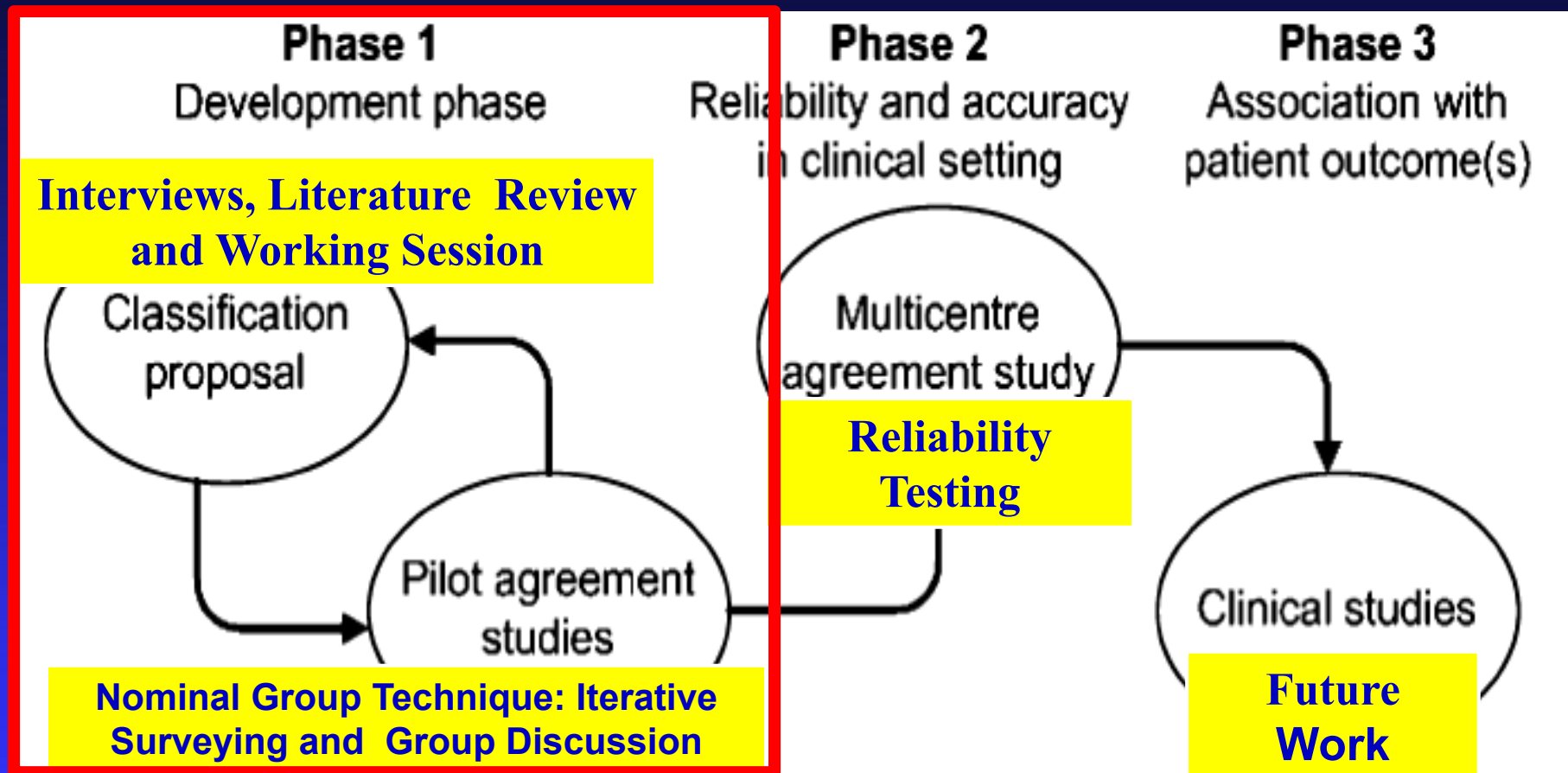
Key 'Philosophical' Aspects of the (C-EOS)

- **Comprehensive**
Applicable to all EOS pts
- **Practical**
Utilized in daily practice
- **Prognostic**
Predictive of course
- **Guide**
Informs treatment decisions



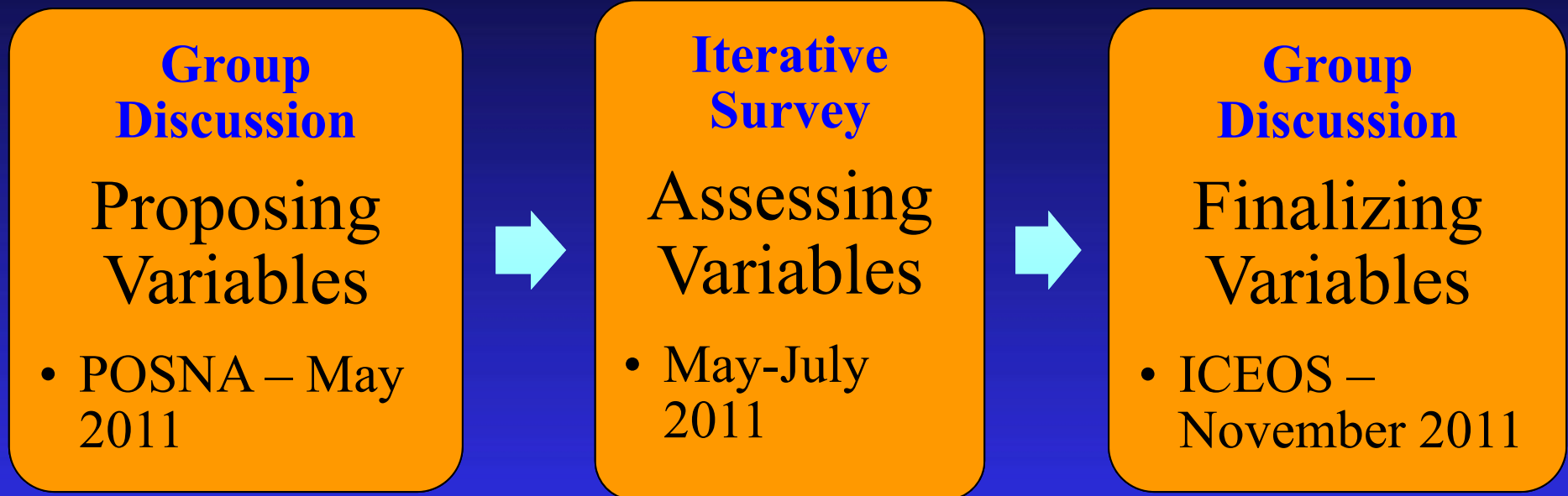
*An EOS
'One Liner'*

Methods: Validation Pathway



Audige L et al. (2005). A concept for the validation of fracture classifications. J Orthop Trauma. 19:404-409

Iterative Survey & Group Discussion



Iterative input by 24 surgeons

Results of Variable Identification Survey

	Not Useful	Useful	Essential	CVR	Sum of Ranks
COBB	0	1	14	0.87	29
ETIOLOGY	0	3	12	0.60	27
KYPHOSIS	0	4	11	0.47	26
AGE	5	0	10	0.33	20
PROGRESSION	3	5	7	-0.07	19
CHEST WALL ABNORMALITIES	2	9	4	-0.47	17
FLEXIBILITY	4	6	5	-0.33	16
OTHER CO-MORBIDITIES	3	8	4	-0.47	16
PULMONARY FUNCTION	3	9	3	-0.60	15
AMBULATORY ABILITY	2	12	1	-0.87	14
NUTRITIONAL STATUS	5	8	2	-0.73	12
MENTAL FUNCTION	10	5	0	-1.00	5
BONE QUALITY	11	4	0	-1.00	4

C-EOS Variables: Etiology

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

Etiology

```
graph TD; Etiology --> Idiopathic_pa[Idiopathic pa]; Etiology --> HTNM; Etiology --> JM; Etiology --> LTNM; Etiology --> dr; Etiology --> Syndromic; Etiology --> Congenital;
```

Idiopathic pa: HTNM JM LTNM dr Syndromic Congenital

C-EOS Variables: Etiology

Etiology

Congenital/Structural

Neuromuscular

Syndromic

Idiopathic

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 (ie. 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 (Major Curve)

1: $<20^{\circ}$

2: $21-50^{\circ}$

3: $51-90^{\circ}$

4: $>90^{\circ}$

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

C-EOS Variables: Kyphosis

Maximum Total Kyphosis

(-) $<20^{\circ}$

N: 21-50 $^{\circ}$

(+): $>50^{\circ}$

Maximum Total Kyphosis: Between any two levels throughout spine

C-EOS Variables:

Progression Modifier (Optional)

Progression Modifier (Optional)

P0: $<10^\circ/\text{yr}$

P1: $10\text{-}20^\circ/\text{yr}$

P2: $>20^\circ/\text{yr}$

Minimum of 6 months x-ray follow-up

$$\frac{[\text{Cobb at } t_2] - [\text{Cobb at } t_1] \times 12 \text{ months / year}}{[\text{Months between } t_1 \text{ and } t_2]}$$

C-EOS Finalized

Highest



Lowest
Priority

Etiology	Cobb Angle (Major Curve)	Maximum Total Kyphosis	Progression Modifier (optional)
Congenital/Structural	1: $<20^{\circ}$	(-) $<20^{\circ}$	P0: $<10^{\circ}/\text{yr}$
Neuromuscular	2: $21-50^{\circ}$	N: $21-50^{\circ}$	P1: $10-20^{\circ}/\text{yr}$
Syndromic	3: $51-90^{\circ}$	(+): $>50^{\circ}$	P2: $>20^{\circ}/\text{yr}$
Idiopathic	4: $>90^{\circ}$		

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: Pts 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 Total Kyphosis: between any 2 levels

Annual Progression Ratio Modifier (optional):

Progression per year;

min. 6 months between observation

$$\frac{(\text{Cobb @ } t_2) - (\text{Cobb @ } t_1) \times 12 \text{ months}}{[t_2 - t_1]}$$

CASE 1

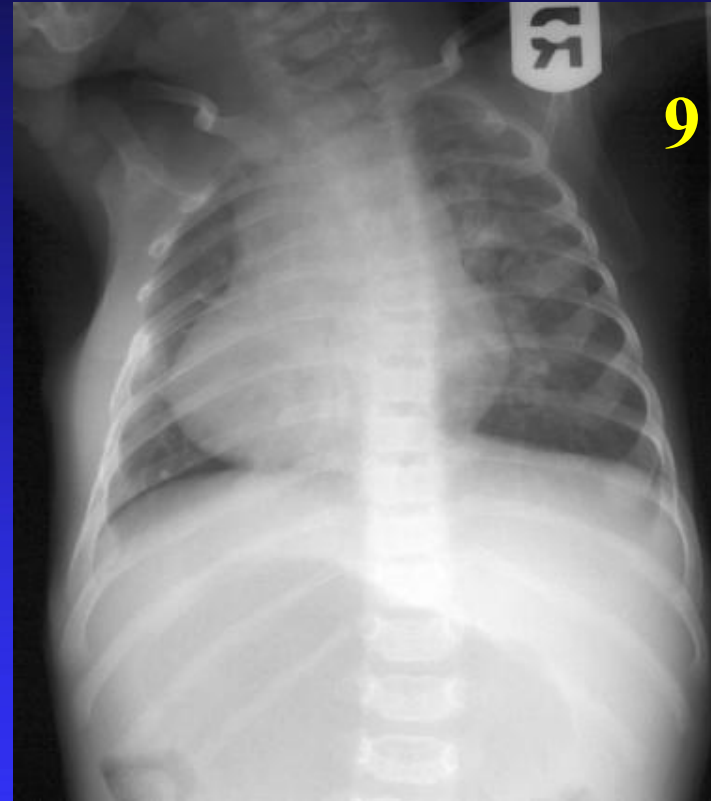
History:

- 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

C7-T6 = 24°



9 months later

Post-PDA surgery

CASE 1: 9 months later

T1-T8 = 42°

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)

- $[(42^\circ - 24^\circ) / (9 \text{ mo.})] \times 12 = 24^\circ/\text{yr} \rightarrow \text{P2}$



C/2/N/P2

Etiology	Cobb Angle (Major Curve)	Maximum Total Kyphosis	Progression Modifier (optional)
Congenital/Structural	1: <20°	(-) <20°	P0: <10°/yr
Neuromuscular	2: 21-50°	N: 21-50°	P1: 10-20°/yr
Syndromic	3: 51-90°	(+) >50°	P2: >20°/yr
Idiopathic	4: >90°		

CASE 2

History

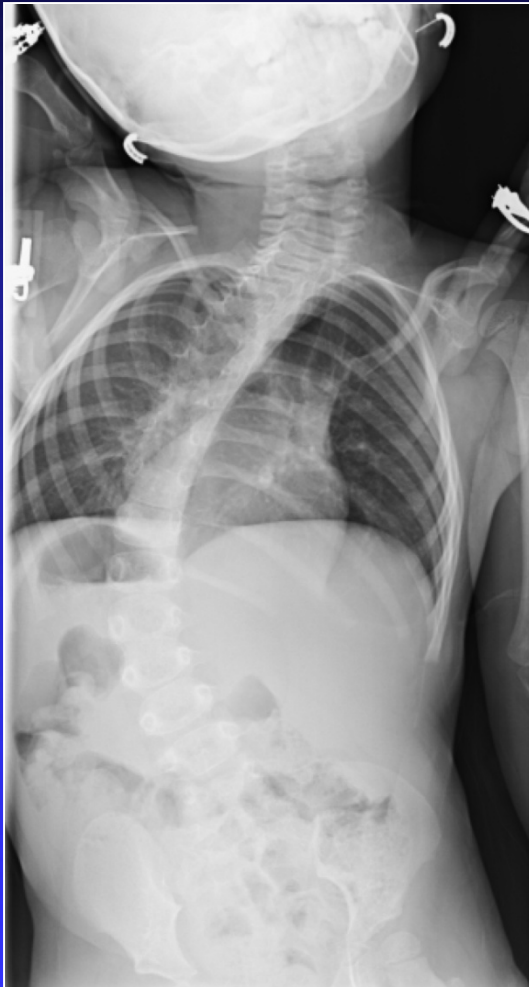
- 4 y/o girl w/ Congenital Myotonic Dystrophy
 - Mother as well

Physical

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

CASE 2

C7-L4 = 50°



1. Etiology

- Congenital? Neuromuscular? Syndromic? →

• Cobb Angle

- 50° → 2

1. Kyphosis

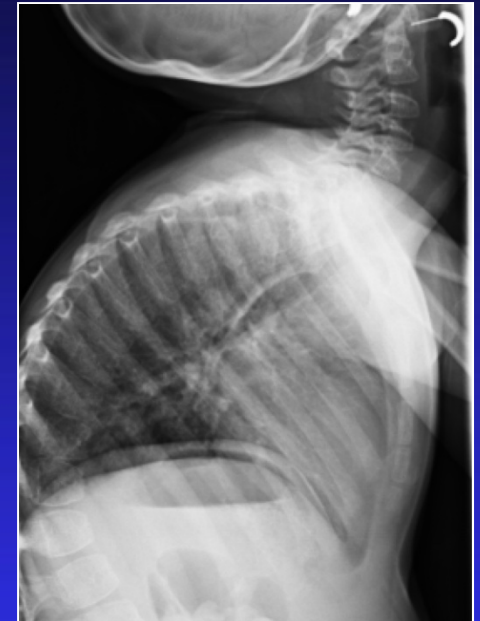
- 96° → +

2. Progression Modifier (optional)

- Not available

N/2/+

Kyphosis = 96°



Etiology	Cobb Angle (Major Curve)	Maximum Total Kyphosis	Progression Modifier (optional)
Congenital/Structural	1: <20°	(-) <20°	P0: <10°/yr
Neuromuscular	2: 21-50°	N: 21-50°	P1: 10-20°/yr
Syndromic	3: 51-90°	(+): >50°	P2: >20°/yr
Idiopathic	4: >90°		

CASE 3

History

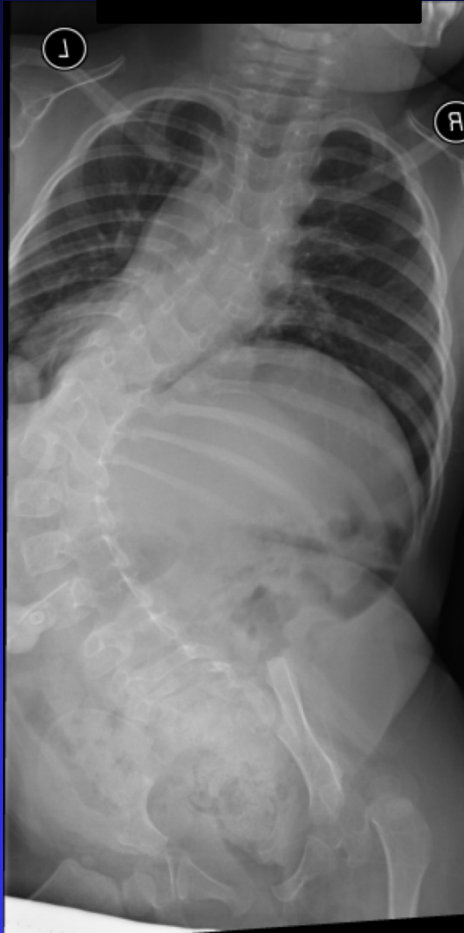
- 4 y/o girl w/ Pena-Shokeir Syndrome
- Developmentally delayed
- Right hip dislocation
- Nonambulatory
 - Wheelchair

Physical

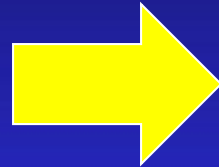
- Lays comfortably on table
- Stiff left thoracolumbar curvature
 - Rib cage rests on pelvis
- Multiple contractures

CASE 3

T9-L4= 88°



7 months later



Coronal Cobb= 97°



Kyphosis= 26°



Etiology	Cobb Angle (Major Curve)	Maximum Total Kyphosis	Progression Modifier (optional)
Congenital/Structural	1: <20°	(-) <20°	P0: <10°/yr
Neuromuscular	2: 21-50°	N: 21-50°	P1: 10-20°/yr
Syndromic	3: 51-90°	(+) >50°	P2: >20°/yr
Idiopathic	4: >90°		

CASE 4

1. Etiology

- Pena-Shokier Syndrome → **Syndromic**

2. Cobb Angle

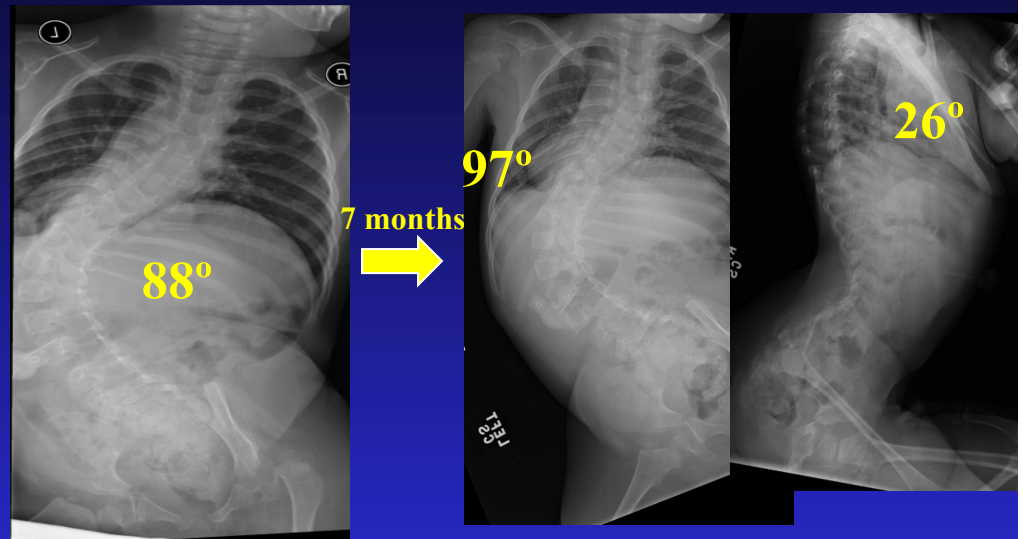
- $97^\circ \rightarrow 4$

3. Kyphosis

- $26^\circ \rightarrow N$

4. Progression Modifier (optional)

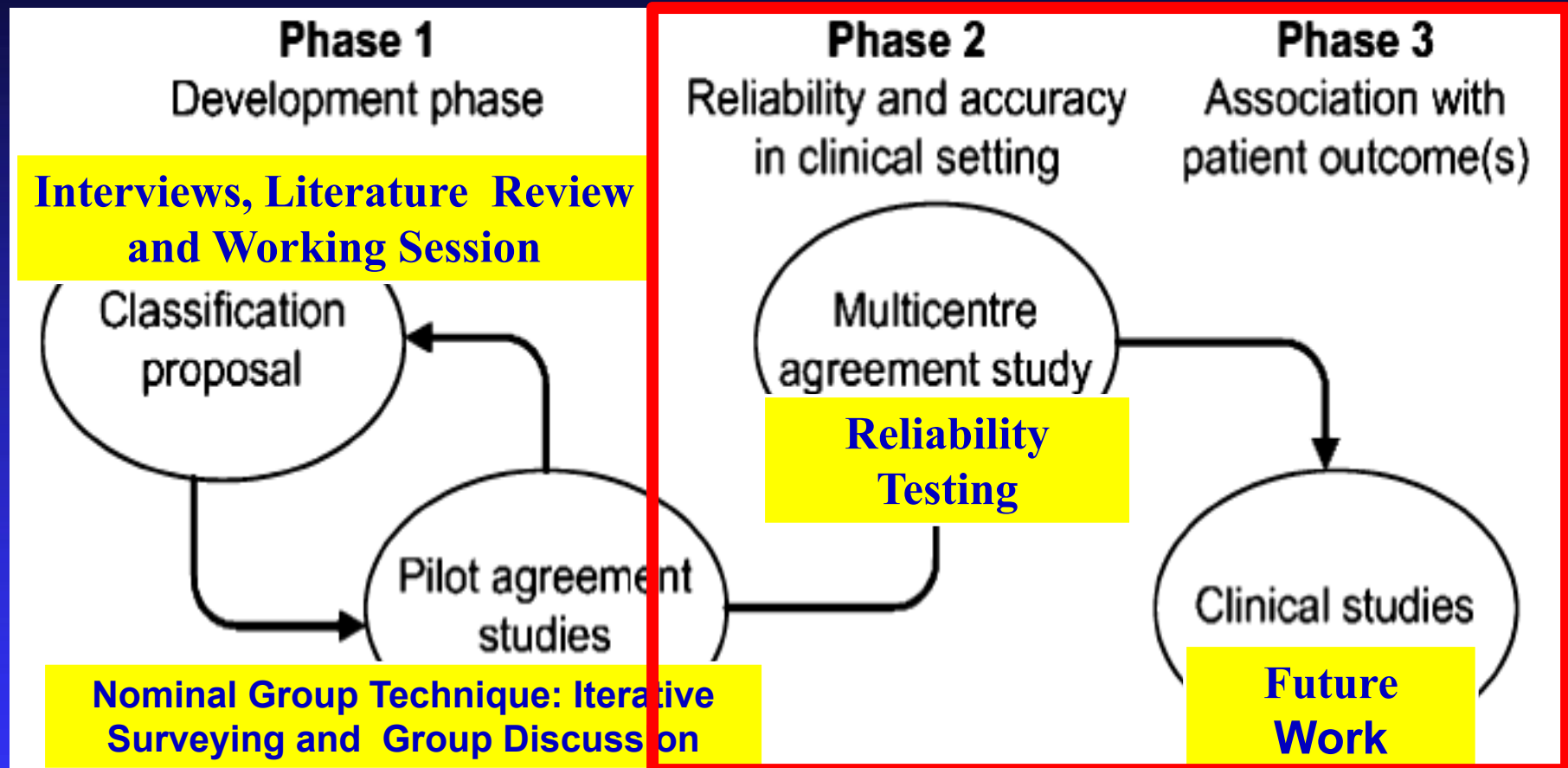
- $[(97^\circ - 88^\circ) / (7 \text{ mo.})] \times 12 = 15.4^\circ/\text{yr} \rightarrow P1$



S/4/N/P1

Etiology	Cobb Angle (Major Curve)	Maximum Total Kyphosis	Progression Modifier (optional)
Congenital/Structural	1: $<20^\circ$	(-) $<20^\circ$	P0: $<10^\circ/\text{yr}$
Neuromuscular	2: $21-50^\circ$	N: $21-50^\circ$	P1: $10-20^\circ/\text{yr}$
Syndromic	3: $51-90^\circ$	(+) $>50^\circ$	P2: $>20^\circ/\text{yr}$
Idiopathic	4: $>90^\circ$		

Methods: Validation Pathway

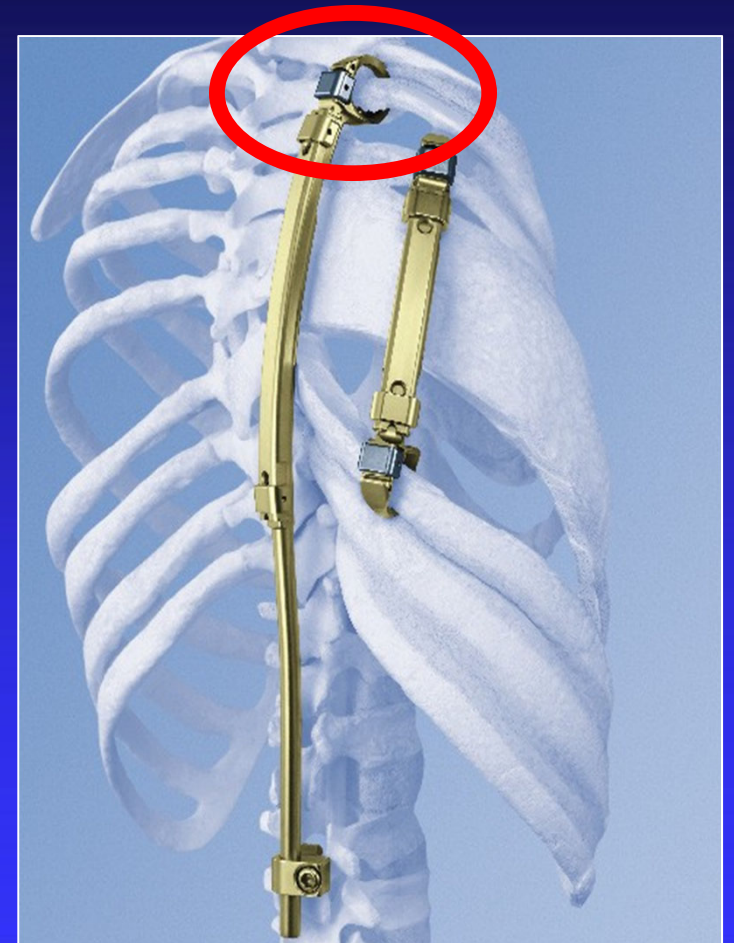


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The Classification for Early-Onset Scoliosis (C-EOS) Predicts Timing of VEPTR Anchor Failure

Purpose: To assess C-EOS' ability to prognosticate outcomes in a clinical setting

Hypothesis: Timing to VEPTR fixation failure will differ among C-EOS classes



Methods

Design:

- Retrospective review of prospectively enrolled patients
 - Sourced from a national registry, *Chest Wall Spinal Deformity Study Group (CWSDSG)*

Participants: Enrollees of the CWSDSG from 2005-2011

– Inclusion

- EOS diagnosis
- >2 yrs follow-up
- VEPTR surgery patients
- Experienced VEPTR proximal fixation failure

Methods

Endpoints:

- Time (months) to VEPTR proximal fixation failure
 - *Definition:* Radiographic diagnosis of failure by an EOS surgeon requiring operative revision of the rib cradle

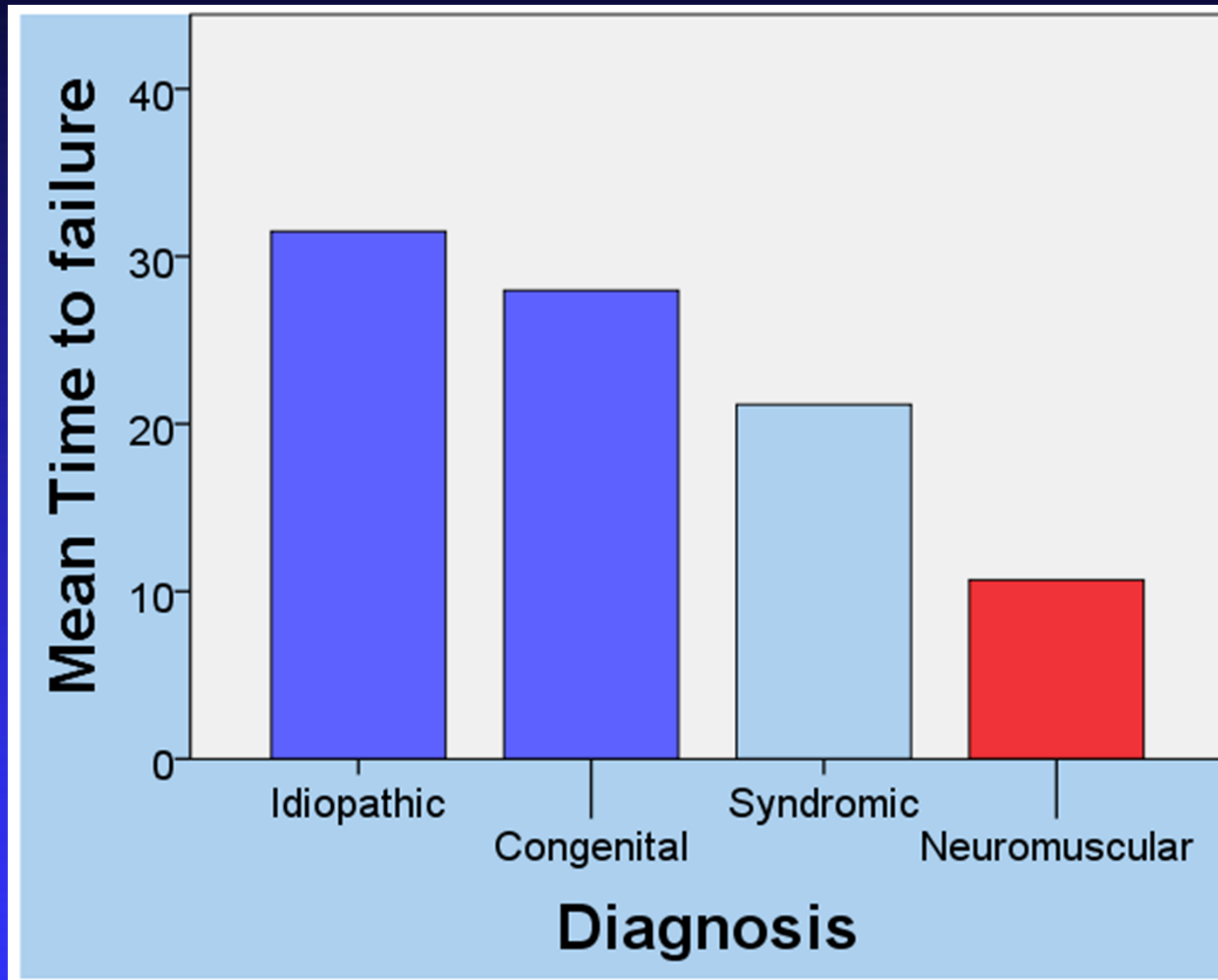
Inclusion:

- Of 446 VEPTR patients with adequate follow up,
- 105 with proximal fixation failure

Statistical Analysis:

- Analysis of Variance (ANOVA) for solitary C-EOS variables
- Kaplan-Meier Survivorship Analysis by C-EOS classes w $n > 3$

Neuromuscular Pts Exhibit Rapid Failure

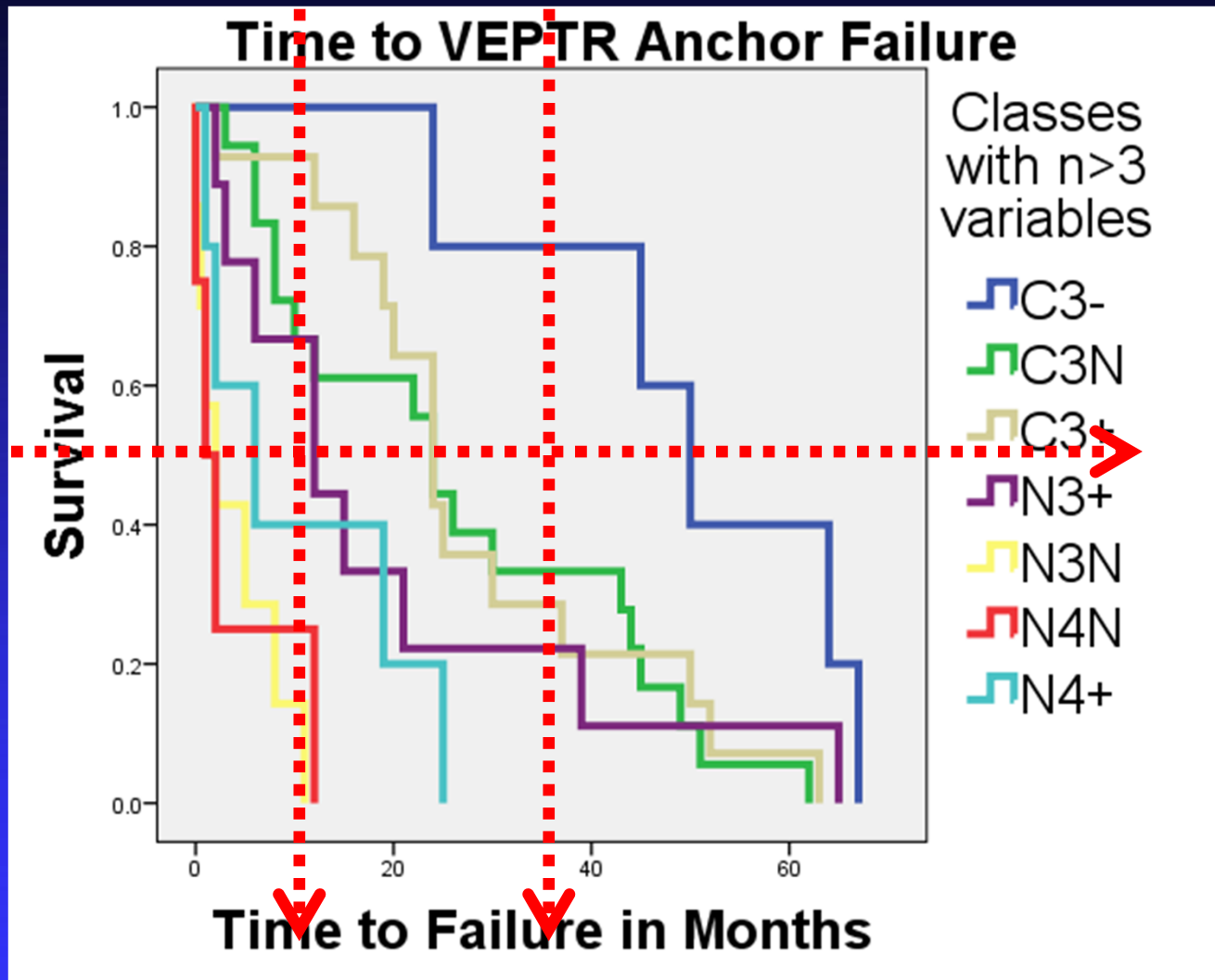


ANOVA

-NM vs. Idiopathic
($p=.026$)

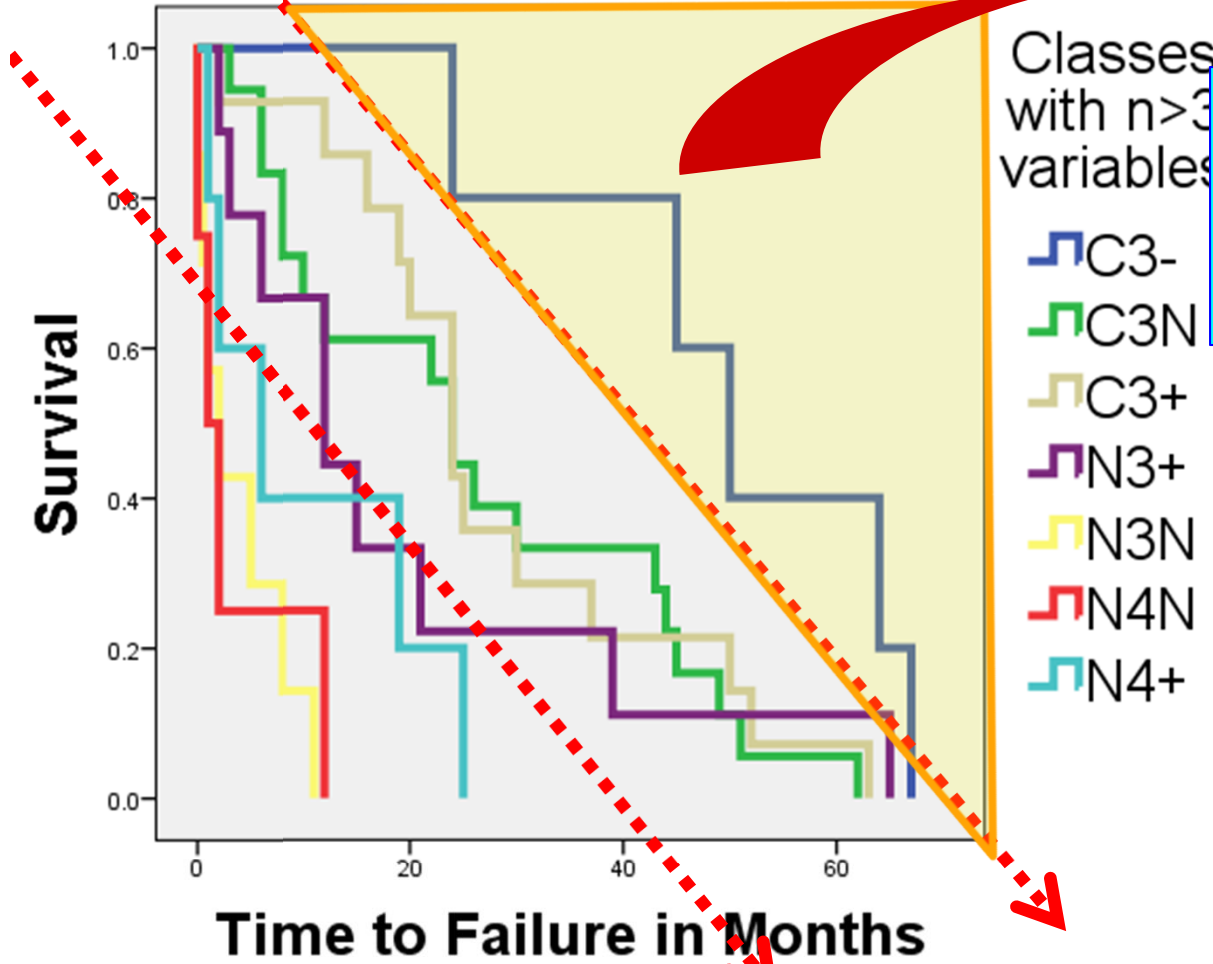
-NM vs. Congenital
($p<.001$)

C-EOS Stratifies Low, Medium, and High Risk



Low Risk of Failure by C-EOS

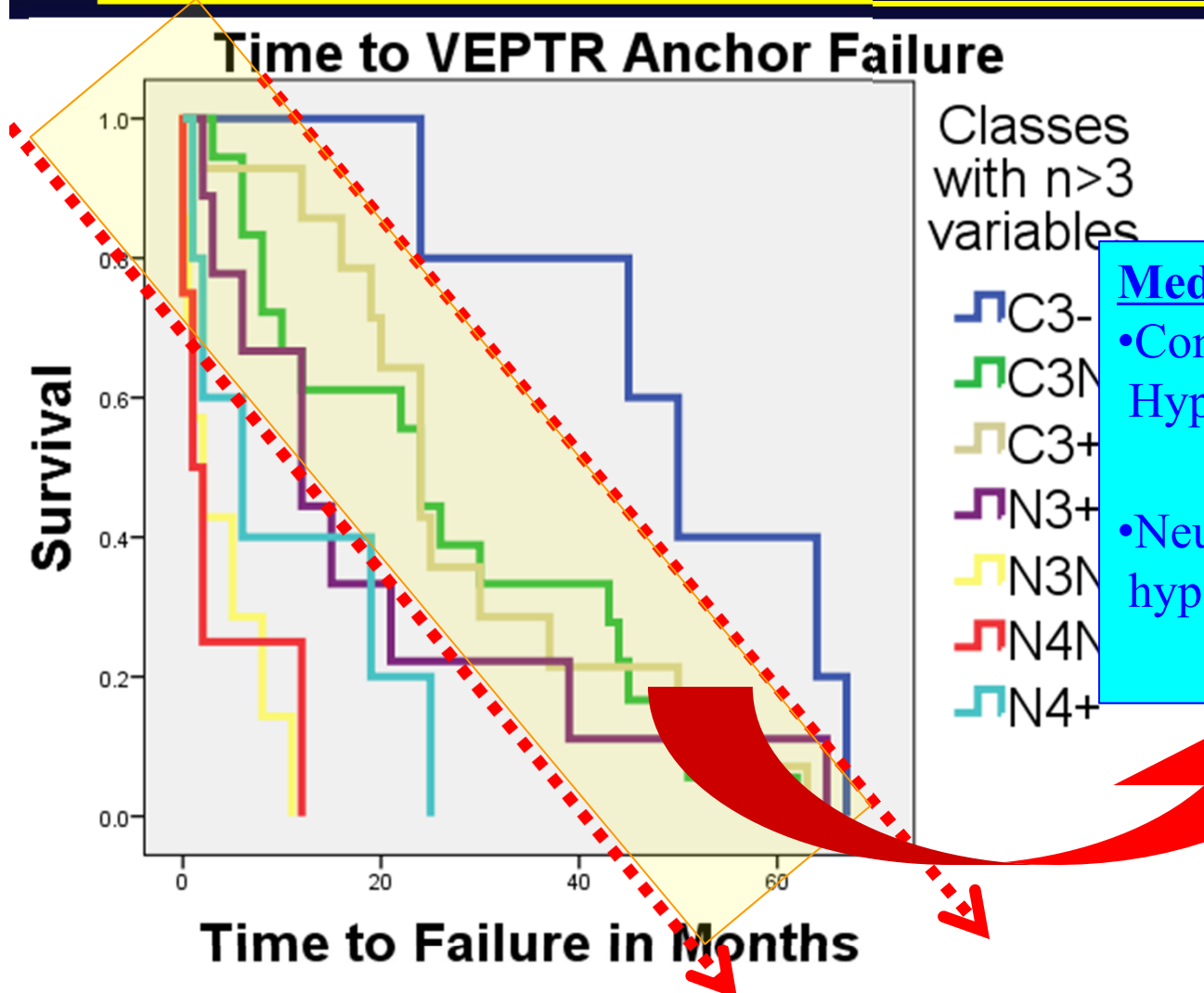
Time to VEPTR Anchor Failure



Low Risk

- Congenital / Curve: 51-90 ° / Hypokyphotic
- C3-

Medium Risk of Failure by C-EOS

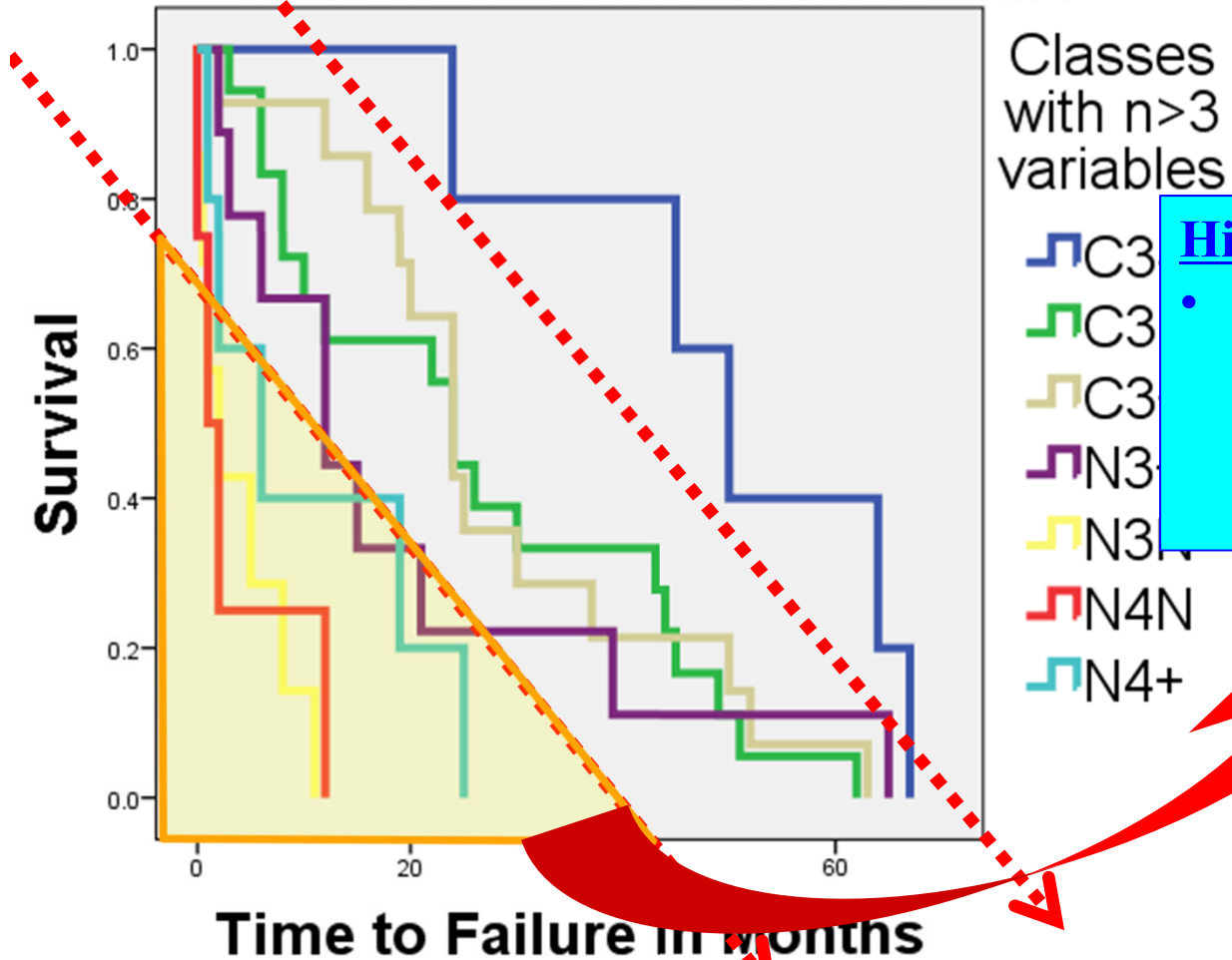


Medium Risk:

- Congenital/51-90° / Norm & Hyper-kyphosis
 - C3N, C3+
- Neuromuscular / 51-90° / hyperkyphosis
 - N3+

High Risk of Failure by C-EOS

Time to VEPTR Anchor Failure



High Risk:

- Neuromuscular / Curve 51-90° & >90° / Norm & hyperkyphosis
- N3N, N4N, N4+

Conclusions

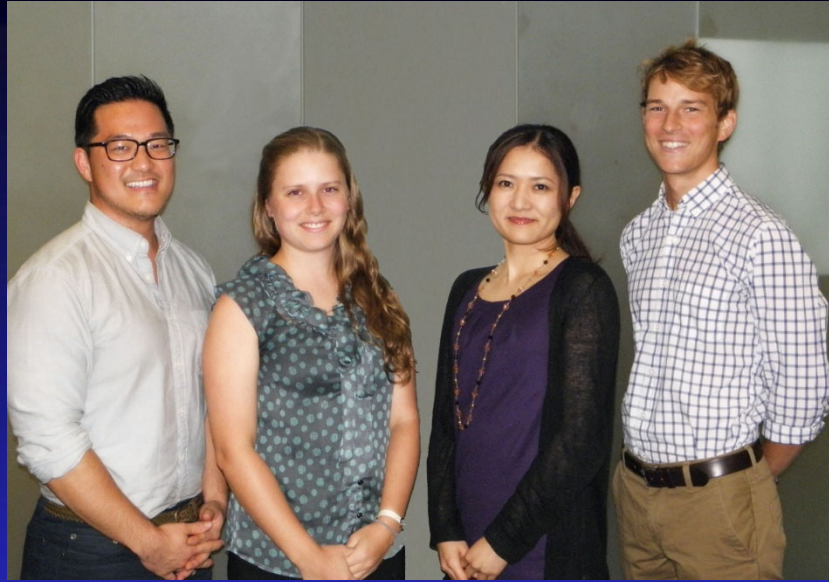
1. C-EOS able to stratify risk of rapid VEPTR anchor failure
 - Supports validity of C-EOS instrument
 - Potential for use in clinical setting
2. Neuromuscular etiology and curves $> 90^\circ$ as individual variables at high risk of rapid anchor failure
3. With further study, C-EOS may guide treatment decisions and inform providers

Work in Progress

Associate C-EOS with Patient Outcome

C-EOS applied to min. 5 Yr follow up pts:

- **Purpose:** Apply C-EOS to identify trends
- **Methods:**
 - Retrospective review of CWSDSG & GSSG database
 - Min 5 year follow-up
- **Endpoints:**
 - Complications
 - Change in coronal and sagittal curve over time
- **Status:** Pending data collection from CWSDSG and GSSG Registry



Thank You

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