## Outcome of "Early" Fusion in EOS

Charles E. Johnston, MD ICEOS Toronto 11/2010 Is short & straight better than long & crooked..... Outcome Determination

- Pulmonary avoid TIS morbidity Tidemark: FVC < 43% pred @ maturity (Pehrsson et al)
- Deformity control historically minimal correction 2° in situ fixation or ineffective constructs Tidemark: T1-12 = 18 cm (Karol et al)

### Nat'l hx EOS not benign

- Pehrsson '92
- Branthwaite '86
- Scott/Morgan '55

Unethical to withhold rx from children with progressive curves - unable to repeat nat'l hx data



#### Respiratory Death 2° Scoliosis

#### The Characteristics of Thoracic Insufficiency Syndrome Associated with Fused Ribs and Congenital Scoliosis

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#### The inability of the thorax to support normal respiration & lung growth

→ 1st attention to TIS in patients with early fusion already performed

Hypothesis : Early Fusion of the Thoracic Spine Stops growth of Th spine and thorax Impairs alveolar hypertrophy (and maybe hyperplasia if done early enough)

## Spinal Growth - Dimeglio



#### Hyperplasia + Hypertrophy



### Role of Deformity Control (keep it straight)

 Untreated large curves → ↑ early mortality (Pehrsson)



## Why control deformity?

 Prevent extrinsic chest wall deformity from producing restrictive/obstructive respiratory disease





#### Extrinsic deformity impairs normal respiration

 Narrowing/stiffening of convex chest wall as rib hump increases loss of compliance (=inability to change volume)





Spinal penetration Windswept thorax



L intercostals non functional -volume not expandable



Concave lung underperfused (Redding)

Fusion w/o deformity control or correction = double jeopardy

- Shortens T spine -> alveolar growth effects
- 2. Fails to alter chest wall dysfunction

"In situ" fusion -> not a good idea

#### Deformity Control - Karol

- Mean age 3.3, 26/28 had ASF
- 11/28 needed revision
- preop final B/W/S • Scoliosis (n=24) 63 59 9/12/3 (45-98) (35-99)
- Kyphosis (n=7) 70 72 3/3/1 (49-108) (30-109)

## **Deformity Control**

 Early surgery incl. ASF may not work (Goldberg)

	pre	іро	f/u (16+)
< age 10 (mean 4)	70	58	80
> age 10 (mean 12.9)	81	46	63

"....early surgery, even with anterior growth arrest... did not halt the deformation of scoliosis and did not reliably preserve respiratory function in this group whose scoliosis presented before age 4." Deformity Control - Winter (1982) PSF only < 5 yr. for cong. scoliosis

 9 cases f/u to Pre 47 (25-75) maturity i.p.o. 34 (17-60) fusion 3+6 Final 40 (18-66) f/u 16 yr

Sitting ht. < 3<sup>rd</sup> % in 6 pts

23 cases < mature</li>
 Fusion 3
 f/u 7 yr
 Pre 60 (22-110)
 i.p.o. 44 (18-82)
 f/u 50 (24-84)

NO f/u pulmonary function

Deformity Control - Winter (1982) PSF only < 5 yrs old for cong. kyphosis

17 cases
Age 2+3
F/u 9 yr
(2 mature)

Pre 58 (28-137) i.p.o. 38 (16-95) F/u 26 (-5-98)

Progressive correction of kyphosis due to anterior growth with solid posterior tether

- Goldberg ('03)
- Emans ('04)
- Karol ('08)
- Vitale ('08)



- Limits growth of the Th spine —> limits volume of thorax & contents
- Poor deformity control chest wall dysfunction

**Respiratory Failure** 

Well established that thoracic fusion < age 5-8 is associated with TIS

Goldberg et al Spine 2003

11 patients < age 8 yr (1.4-7.8) PFT's @ 20.5 yr. (15-30) FEV1 = 41% (14-72) FVC = 41% (12-67)

"....early surgery, even with anterior growth arrest...did not halt the deformation of scoliosis and did not reliably preserve respiratory function in this group whose scoliosis presented before age 4."

- If fusion delayed to age  $10 \rightarrow$  PFT's = 70% mean (45-100%)
- Controversy : is pulmonary function degraded by the treatment (fusion) ...OR by the deformity itself

 Historically.... Ethically.... bad curves get fusion earlier Can we do better?

#### TSRHC study (Karol et al, JBJS 6/08) Fusion age 3.3 yr, f/u 11 yr FVC 58% (27-99) FEV1 55% (23-91)



- FVC most normal in limited distal thoracic fusions
- 8/11 patients with fusions beginning at T1 or T2 have FVC's < 50% of normal</li>
- Proximal thoracic fusions correlated with  $\downarrow$ FVC (p<0.0001)



#### Role of sternum/sternal ribs in proximal thoracic fusion Canavese, Dimeglio et al, Spine '07

- T1-T6 psf in young rabbits  $\longrightarrow$
- 1. | length of segment compared to control
- 2. ↓ a/p diameter of thorax (CT) = ellipse
- 3. I length of sternum compared to control
- 4. ↓ lung growth (CT volumes)

#### T1-12 length vs. FVC



Karol L. A. et.al. J Bone Joint Surg 2008:90:1272-1281

## Th Length Fused vs. Observed

Bowen etal JPO '08

- All congenital curves, f/u 6.7 yr
- Fused < 5yr. -> T1-12 growth = .48 cm/yr
- Observed -> .97 cm/yr
- Normal (Dimeglio) -> 1.4 cm/yr (age 0-5)
- Not matched for severity, more deformity rx'd

## Conclusions?

- 1. Fusion < 4-5 not good for thoracic and pulmonary growth (>18 cm)
- Early fusion with minimal correction → chest wall deformity uncontrolled → set up T.I.S.
- 3. Early fusion must correct deformity <u>effectively</u> (one-time or modulation) while minimizing # segments

4. Early rx must correct or prevent progressive spinal deformity producing windswept thorax



#### Conclusions

- 5. ASF/PSF "in situ" or w/  $1^{st}$ generation implants tend to be <u>ineffective</u>  $\rightarrow$ ? role of in situ fixation
- 6. Await pulmonary and growth results of more effective correction +/- segmental fixation to know if outcomes improved compared to historical results



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