Outcome of "Early" Fusion in EOS

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Is short & straight better than long & crooked…..
Outcome Determination

1. Pulmonary – avoid TIS morbidity
   Tidemark: FVC < 43% pred @ maturity
   (Pehrsson et al)

2. Deformity control - historically minimal correction 2° in situ fixation or ineffective constructs
   Tidemark: T1-12 = 18 cm (Karol et al)

Respiratory Death 2° Scoliosis

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

By Robert M. Campbell Jr., MD, Melvin D. Smith, MD, Thomas C. Mayes, MD, John A. Mangos, MD, Donna B. Willey-Courand, MD, Nusret Kose, MD, Ricardo F. Pinero, MD, Marden E. Alder, DDS, Hoa L. Duong, MD, and Jennifer L. Surber, BS

Investigation performed at The Thoracic Institute, Christus Santa Rosa Children's Hospital, San Antonio, Texas

J Bone Joint Surg 3/03

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

Melvin Smith, MD

1941-2008

Hypothesis : Early Fusion of the Thoracic Spine

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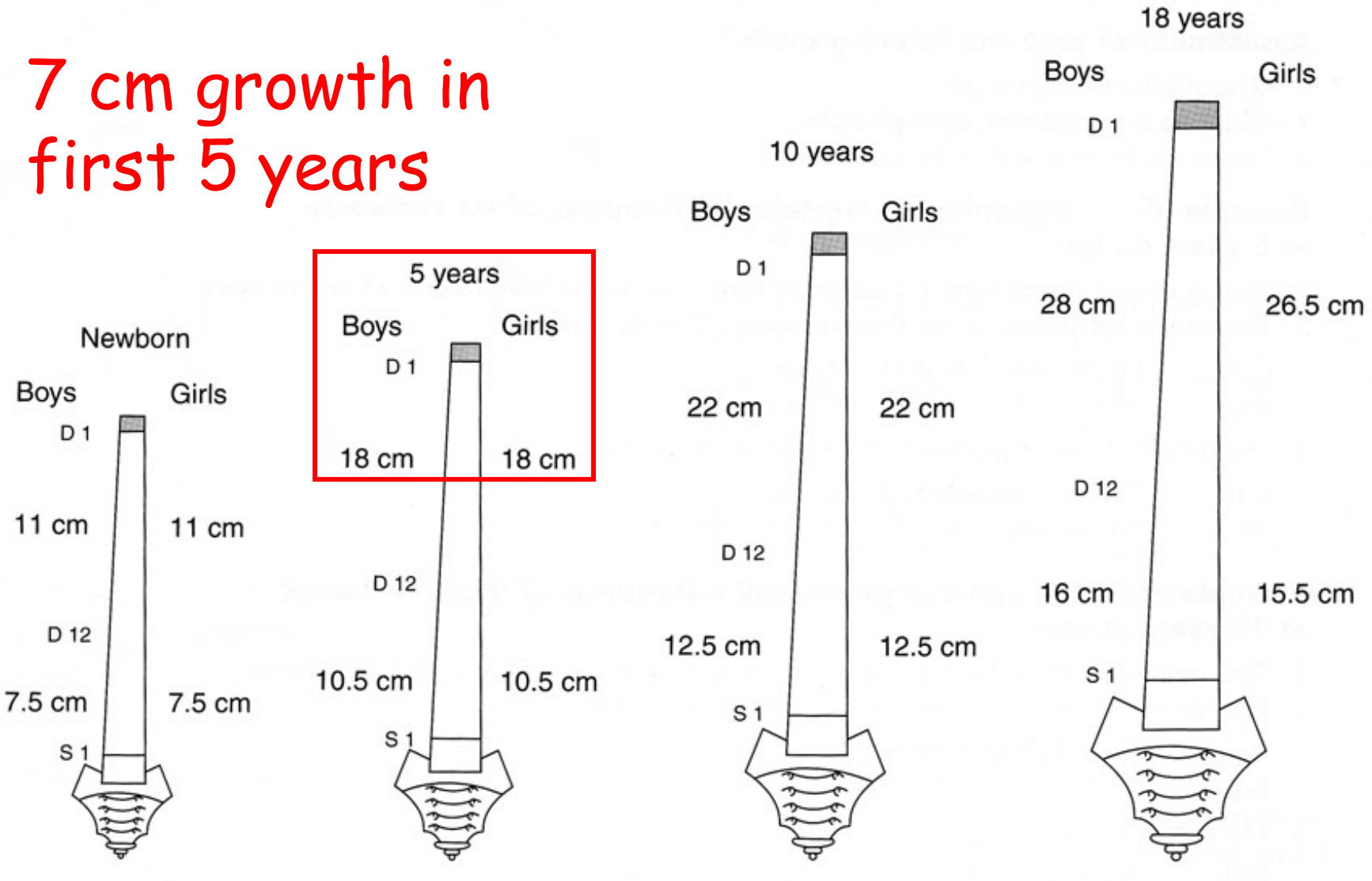
Stops growth of Th spine and thorax

↓

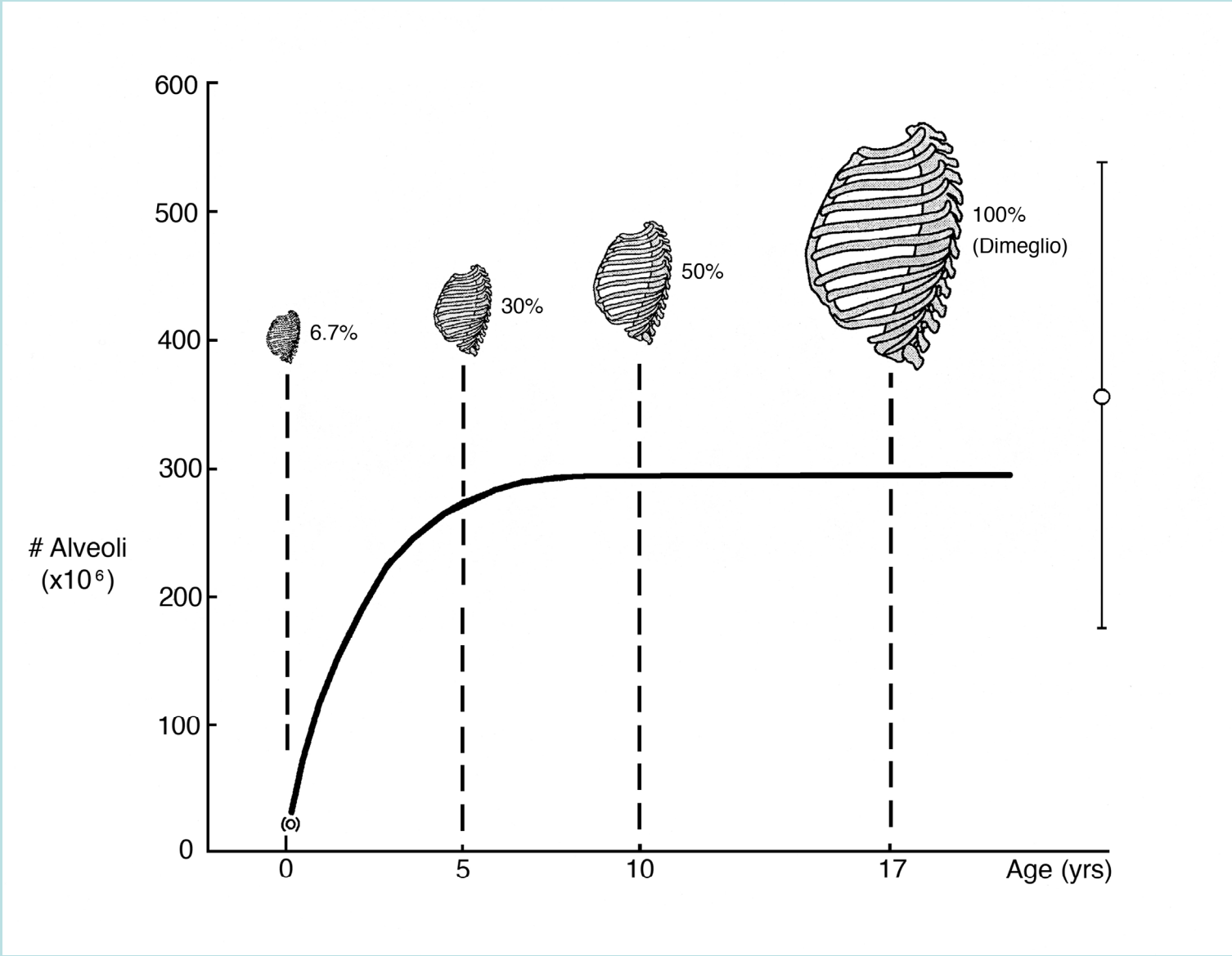
Impairs alveolar hypertrophy (and maybe hyperplasia)

Spinal Growth - Dimeglio

7 cm growth in first 5 years



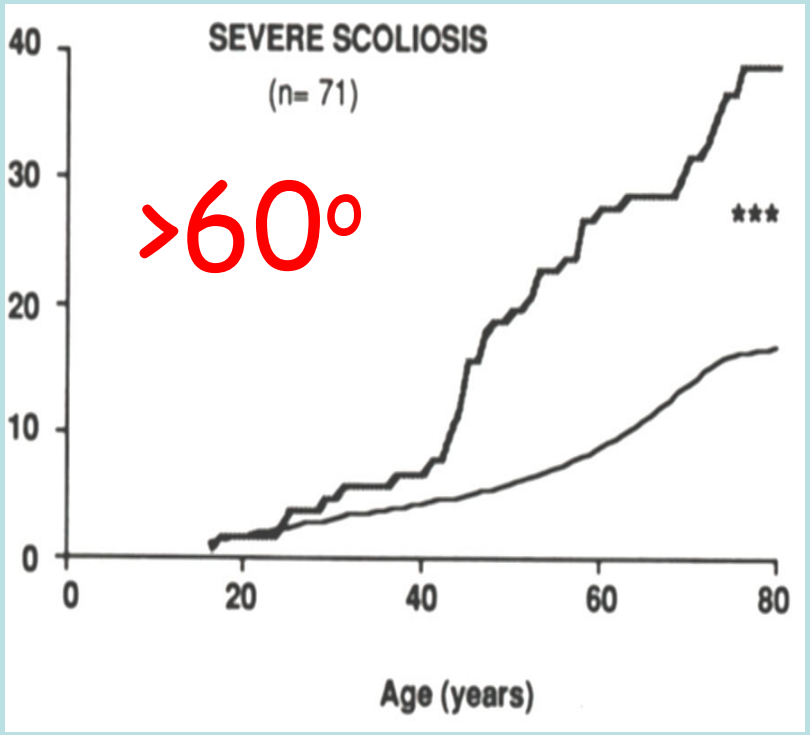
Hyperplasia + Hypertrophy

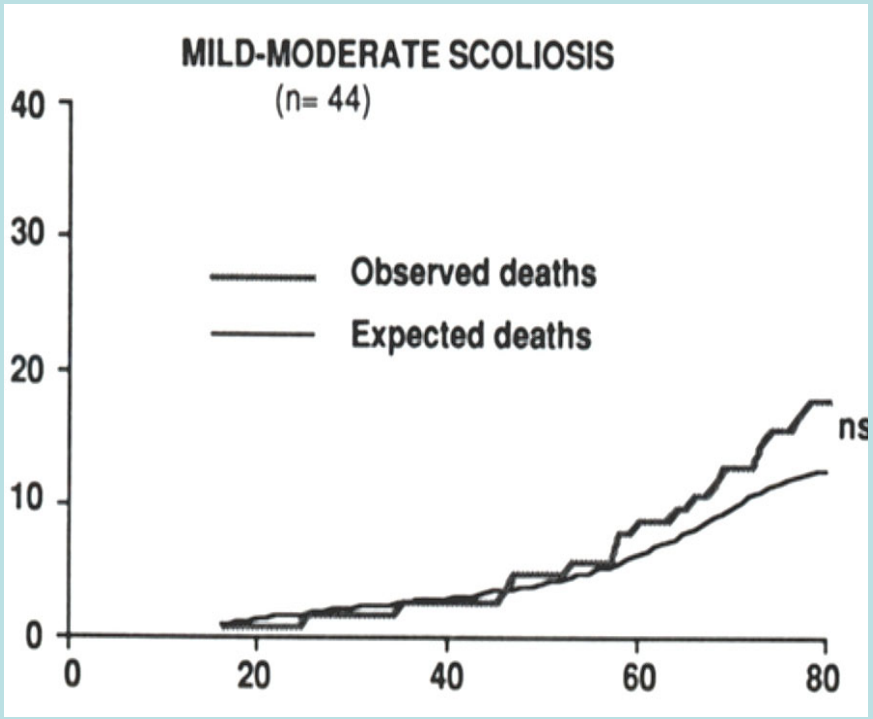


Role of Deformity Control

(keep it straight)

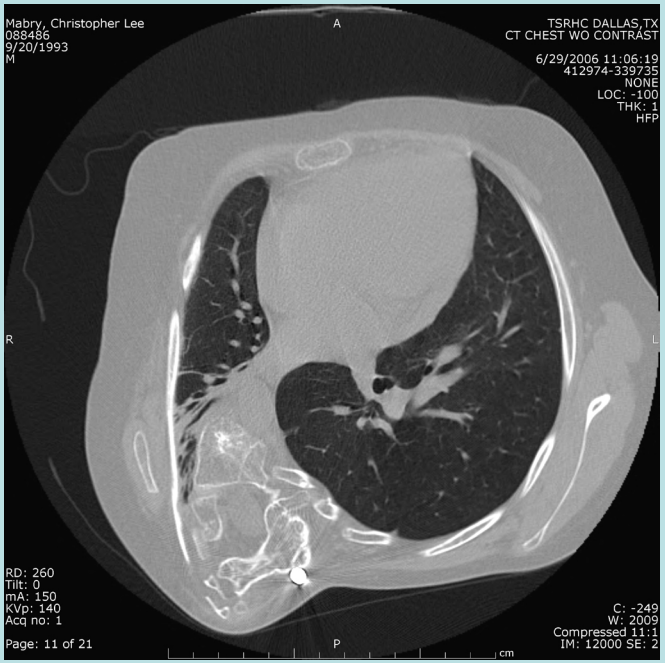
- Untreated large curves ⟶ ↑ early mortality (Pehrsson)

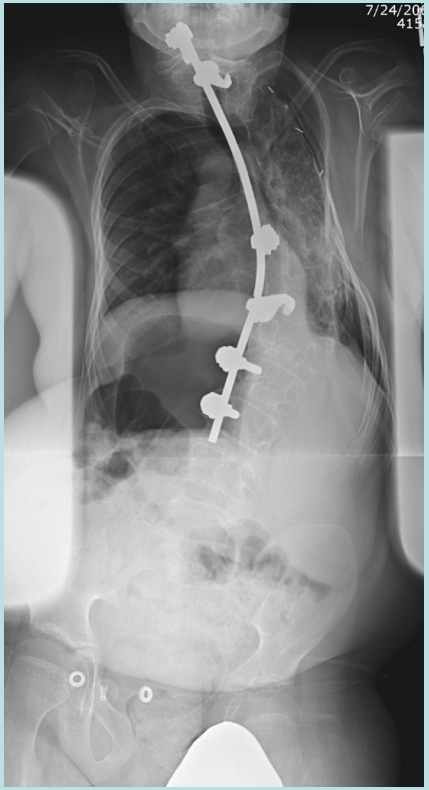




Why control deformity?

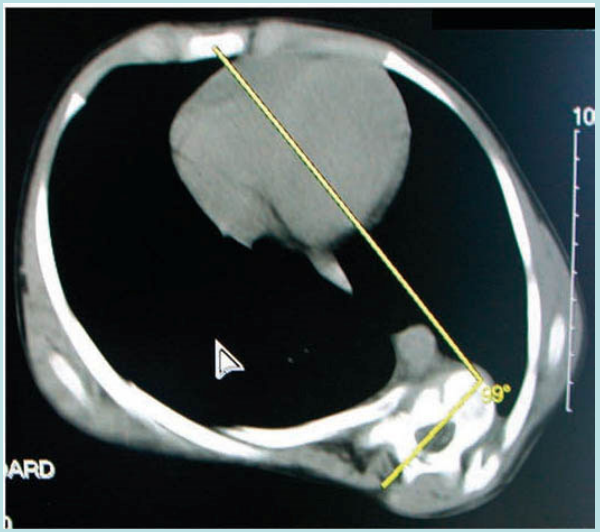
- Prevent extrinsic chest wall deformity from producing restrictive/obstructive respiratory disease





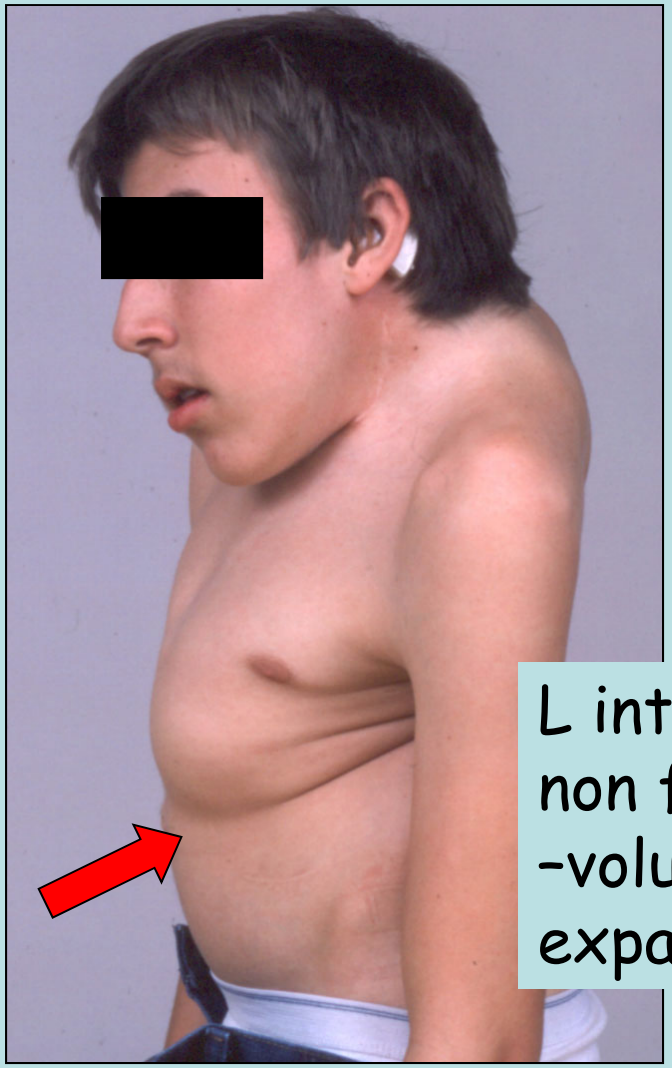
Extrinsic deformity impairs normal respiration

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

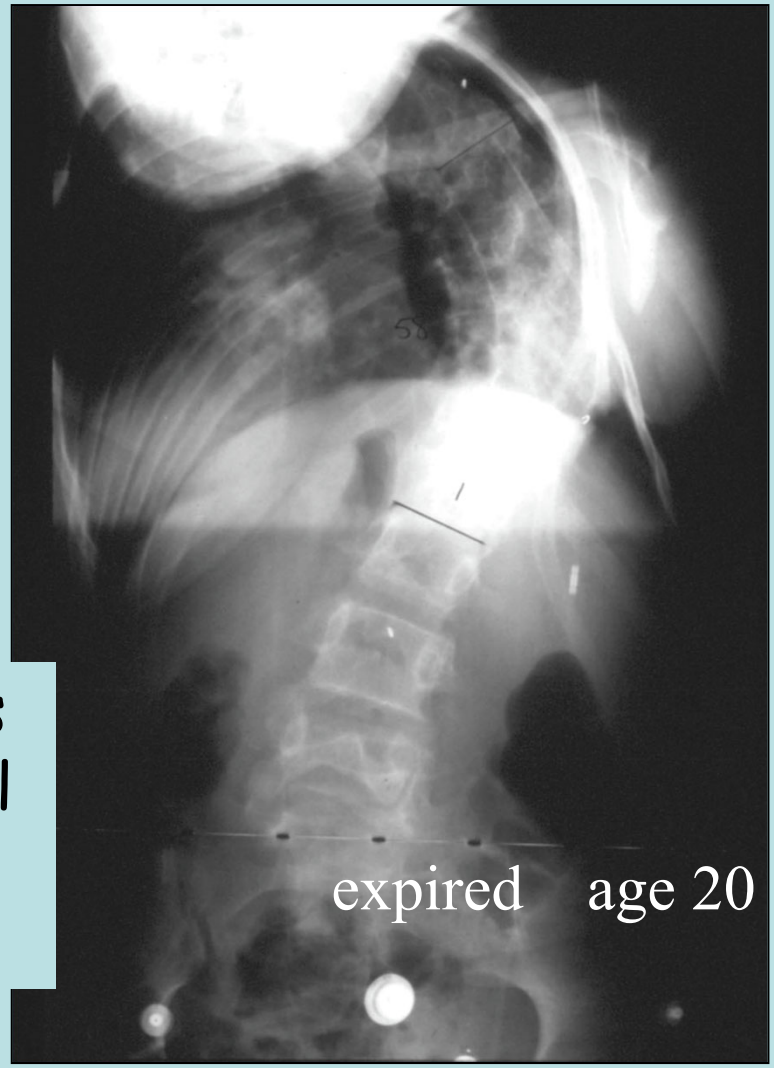


Spinal penetration

Windswept thorax



L intercostals non functional -volume not expandable



Concave lung underperfused (Redding)

- 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 ⟶
  PFT's = 70% mean (45-100%)

- Controversy : is pulmonary function degraded by the treatment (fusion) …OR by the deformity itself

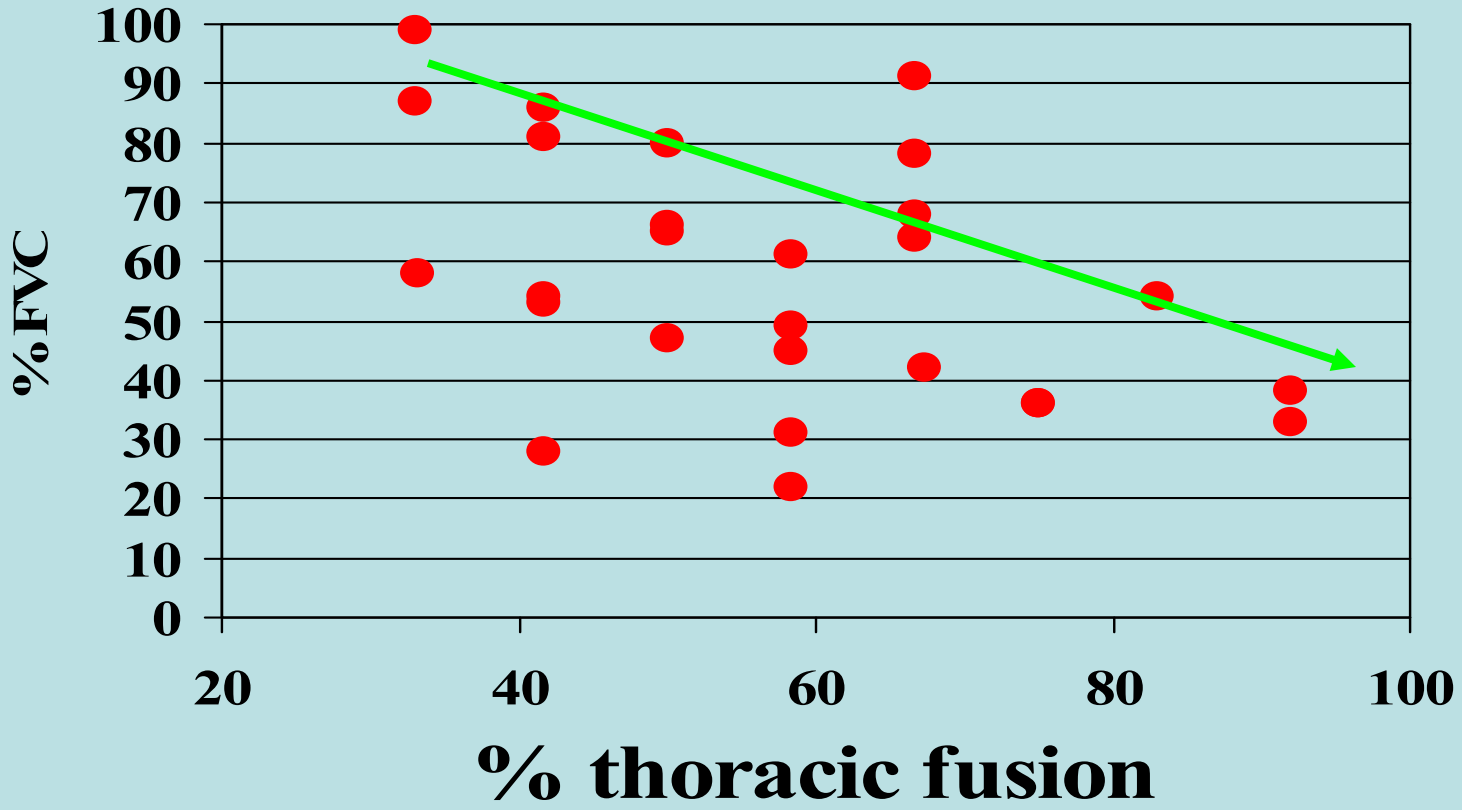
- Historically….
  bad curves get fusion earlier
  Can we do better?

TSRHC study (Karol et al, JBJS 6/08)

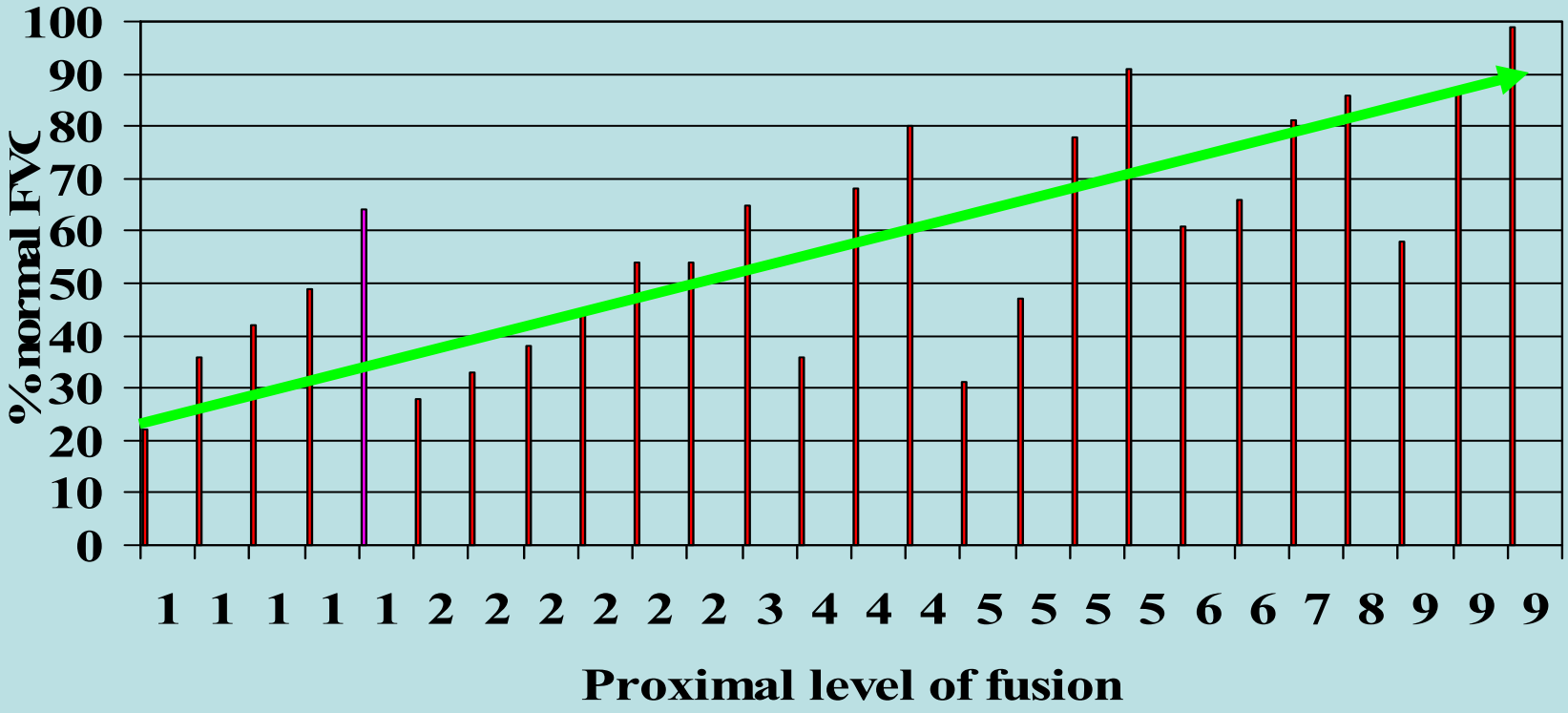
Fusion age 3.3 yr, f/u 11 yr

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
- Proximal thoracic fusions correlated with ↓FVC ($p<0.0001$)



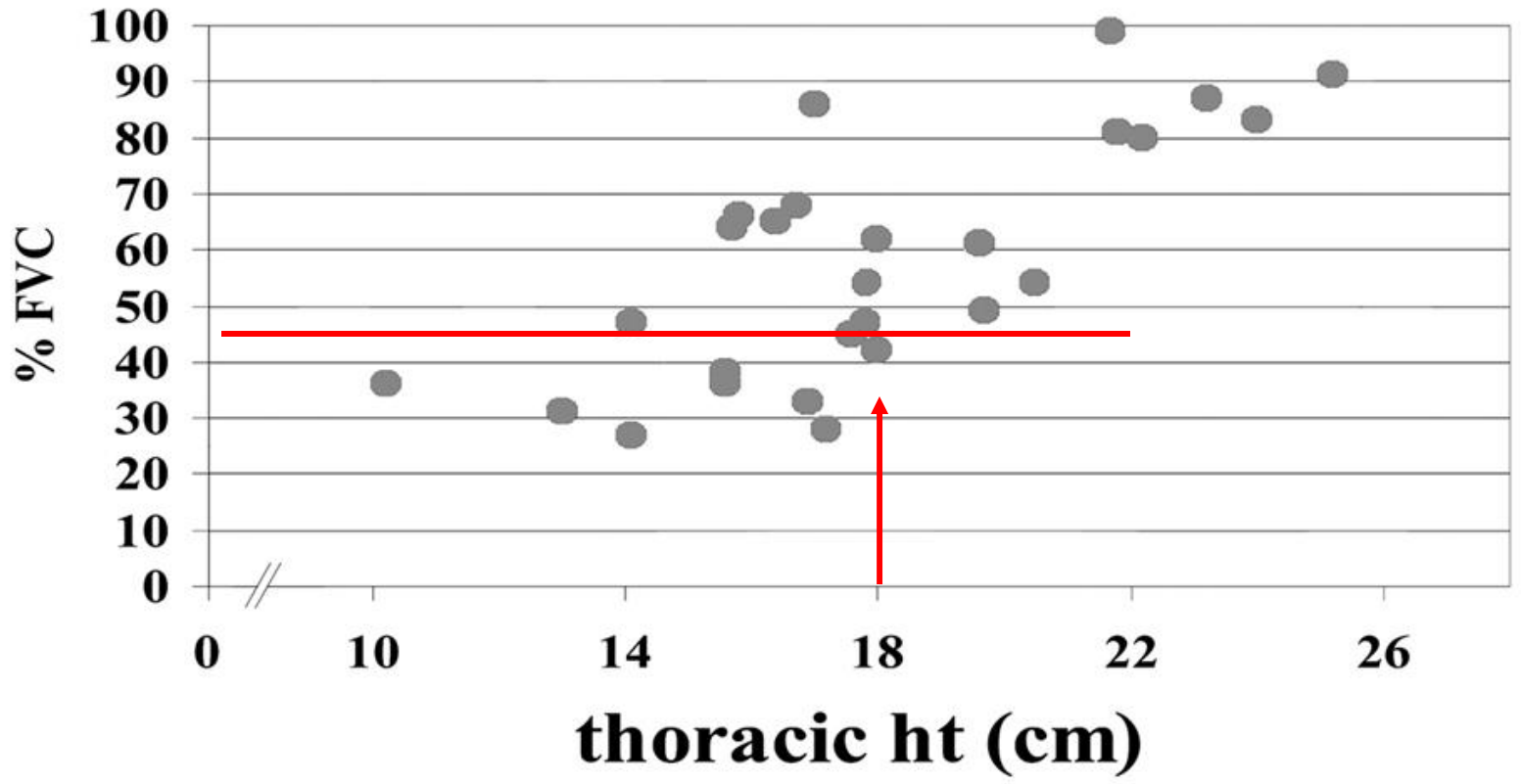
Role of sternum/sternal ribs in proximal thoracic fusion

Canavese, Dimeglio et al, *Spine* '07

T1-T6 psf in young rabbits ⟶

1. ↓ length of segment compared to control
2. ↓ a/p diameter of thorax (CT) = ellipse
3. ↓ 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

Deformity Control

- Early surgery incl. ASF may not work (Goldberg)

	pre	ipo	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 - Karol

- Mean age 3.3, 26/28 had ASF
- 11/28 needed revision

	preop	final	B/W/S
• Scoliosis (n=24)	63 (45-98)	59 (35-99)	9/12/3
• Kyphosis (n=7)	70 (49-108)	72 (30-109)	3/3/1

Deformity Control – Winter (1982)

PSF only < 5 yr. for cong. scoliosis

- 9 cases f/u to maturity
  fusion 3+6
  f/u 16 yr

Pre	47 (25-75)
i.p.o.	34 (17-60)
Final	40 (18-66)

Sitting ht. < 3rd % in 6 pts

- 23 cases < mature
  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)

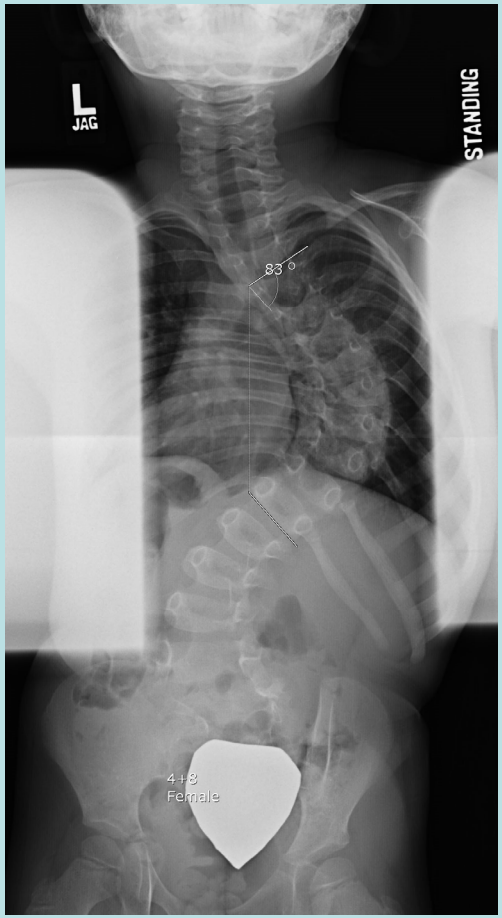
Pre 58 (28-137)
i.p.o. 38 (16-95)
F/u 26 (-5-98)

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

Conclusions ?

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

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



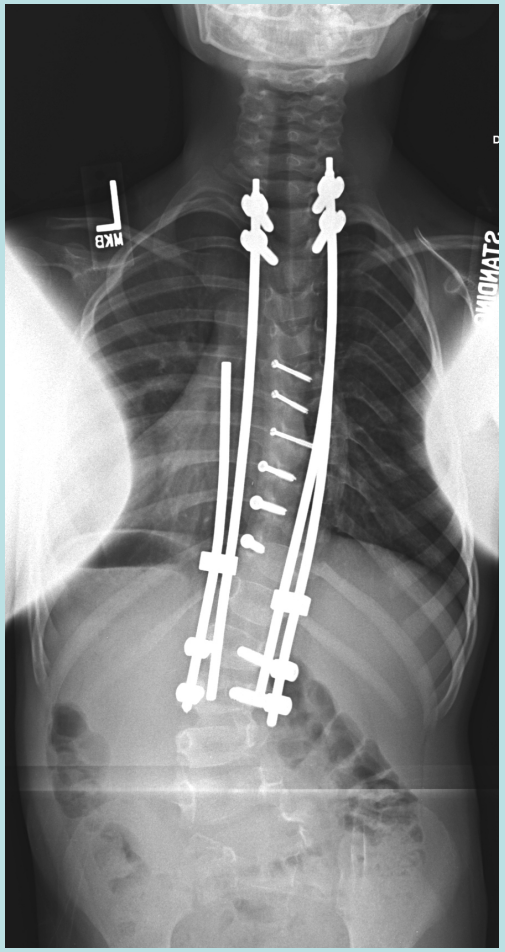
4+8

Lipo-meningocoele

NCS

arrest +

GR's



Conclusions

5. ASF/PSF "in situ" or w/ 1st generation implants tend to be ineffective → ? 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