Early Onset Scoliosis:

Why We Don't Do What We Used To!

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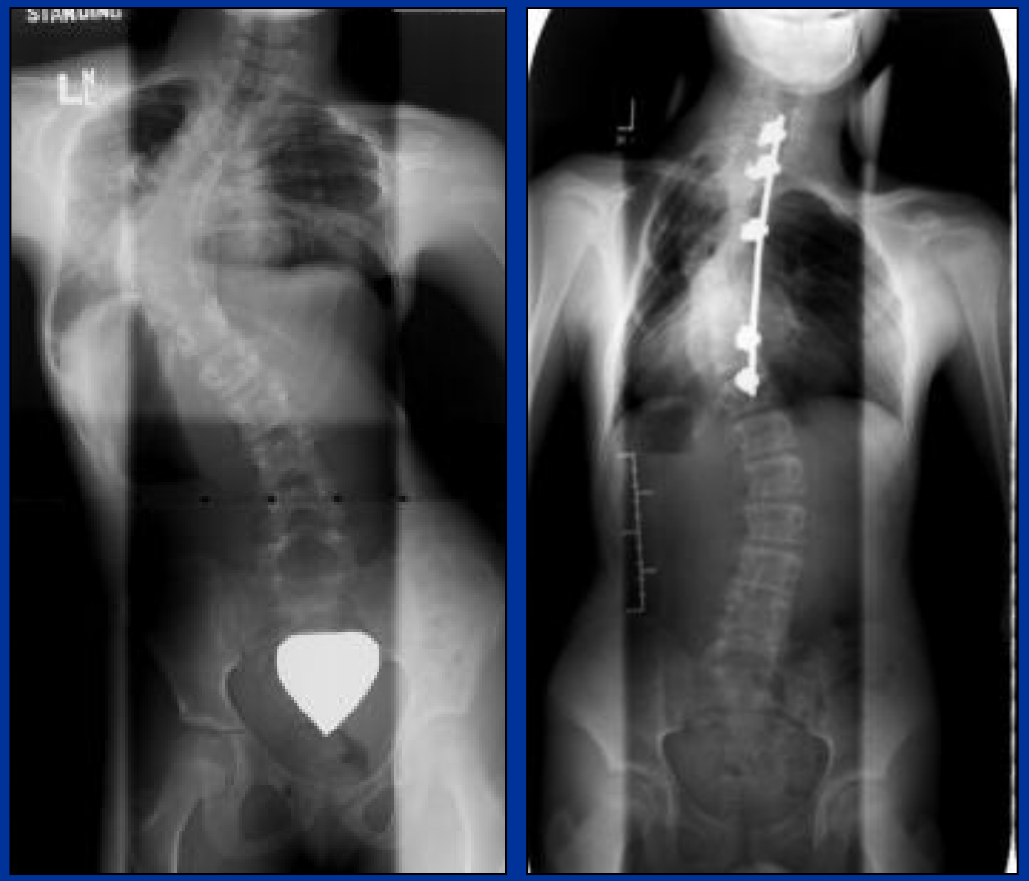
Typical Treatment 1990's

- Observation
- Bracing
- Early fusion for continued progression
  - PSF
  - ASF/PSF after Duboussset taught us about crankshaft phenomena

Congenital Scoliosis

- Felt that “short and straight” better than curve progression

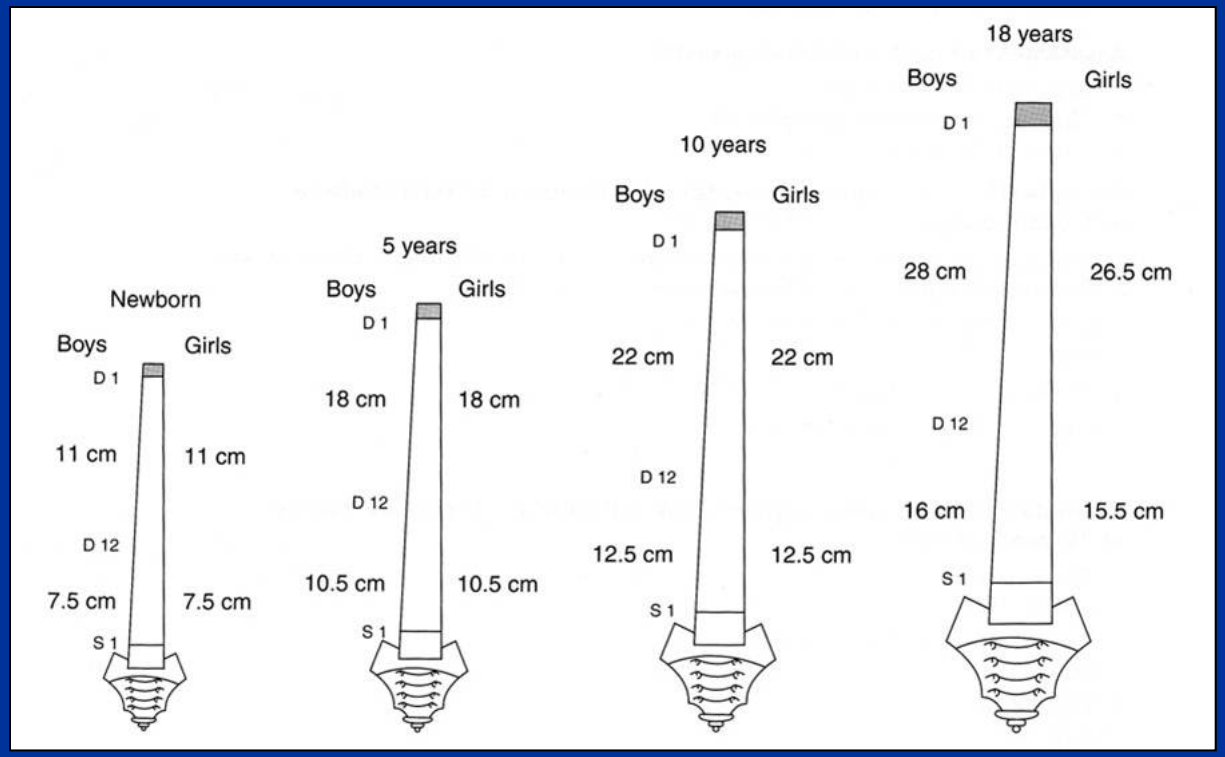


So Did It Work?

- Reoperation for Progression
  - Goldberg (2002) 37% reop for “adding on”
  - TSRH (2008) 39% reop at 11 yr f/u even after asf/psf
  - Vitale 24% congenital scoliosis reop at 7 yr f/u
  - Progression, adding on, or continued growth??

Spinal Growth (Dimeglio)

- Most rapid in first 5 yrs of life when spine increases by 50%
- Slower growth from 5-10



The Problem is…

- Spine growth limited by congenital malformations
- Early fusion prohibits continued spine and therefore pulmonary growth

Mortality in nonop EOS

- Higher than juvenile and adolescent combined
- Pehrsson described early mortality in 29 patients with IIS (ave age 54 yo)
  - Death as young as 16 yrs of age
- Scott and Morgan 4/28 patients died between 17 and 19 yrs
- Respiratory failure

PFT Studies

- Goldberg:
  - 23 infantile scoliosis patients
  - FVC average 41% (12-67%) if fused before age 10 yrs
  - FVC average 68% (48-88%) if fused later
  - Early fusion resulted in smaller lungs

WHO gets into trouble?

- We undertook our study to try to figure out which patients “got into pulmonary trouble” from early fusion, and which tolerated it.
- Who needs growth-friendly surgery?

    - Karol, Mladenov, Johnston, Schochet, Walters

INCLUSION CRITERIA

- Patients fused age ≤ 8 yrs 1983 - 1998
- Some portion of thoracic spine fused
- Minimum 5 yr f/u
- No significant thoracic comorbidities (i.e. diaphragmatic hernia), bone dysplasias which would affect growth, or neuromuscular disease

METHODS

- Records reviewed for diagnosis, comorbidities, extent of fusion, anterior vs. posterior, need for further surgeries.
- Radiographs reviewed from preop, postop, and final follow-up.
  - Coronal and sagittal deformity
  - Thoracic height T1-T12
- PFT's performed.

RESULTS

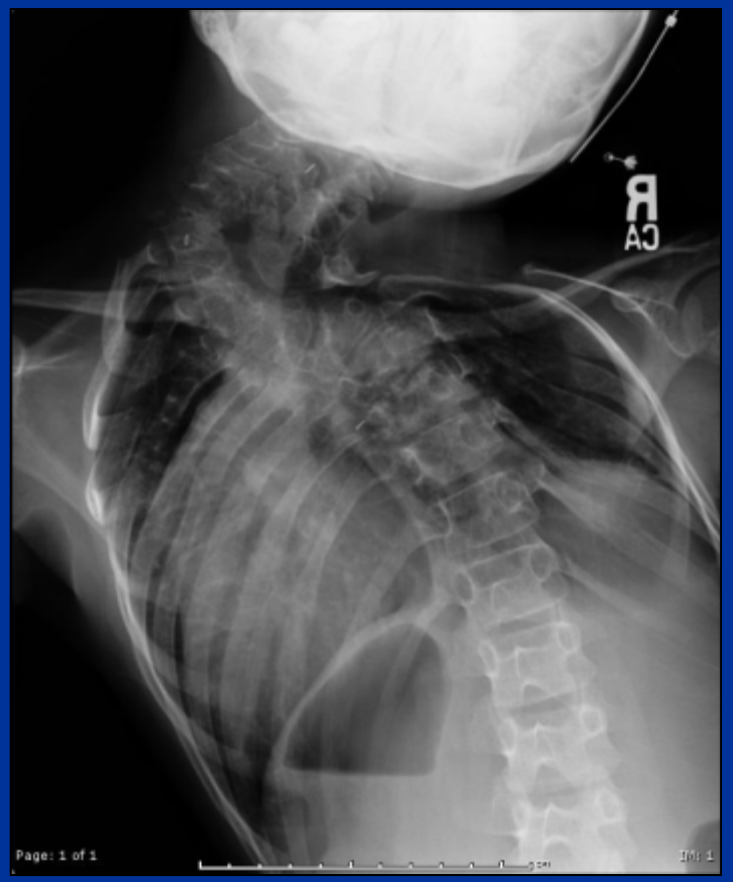
- 28 patients tested
- Age at surgery = 3.3 yrs (4 mos – 8.4 yrs)
- Age at testing =14.6 yrs (7.3 – 22.8 yrs)
- Ave f/u 11.2 years (6.4 – 20.5 yrs)
- 27/28 had anterior surgery
- 11/28 had revision surgery

RESULTS

- Diagnoses:
  - **20 congenital scoliosis**
  - 3 neurofibromatosis
  - 2 infantile scoliosis
  - 1 juvenile scoliosis
  - 1 syndromic
  - 1 kyphosis

RESULTS

- All patients alive.
- No patient ventilator dependent.
- 4/28 have thoracic insufficiency syndrome
- 1 additional patient in distress after 2008

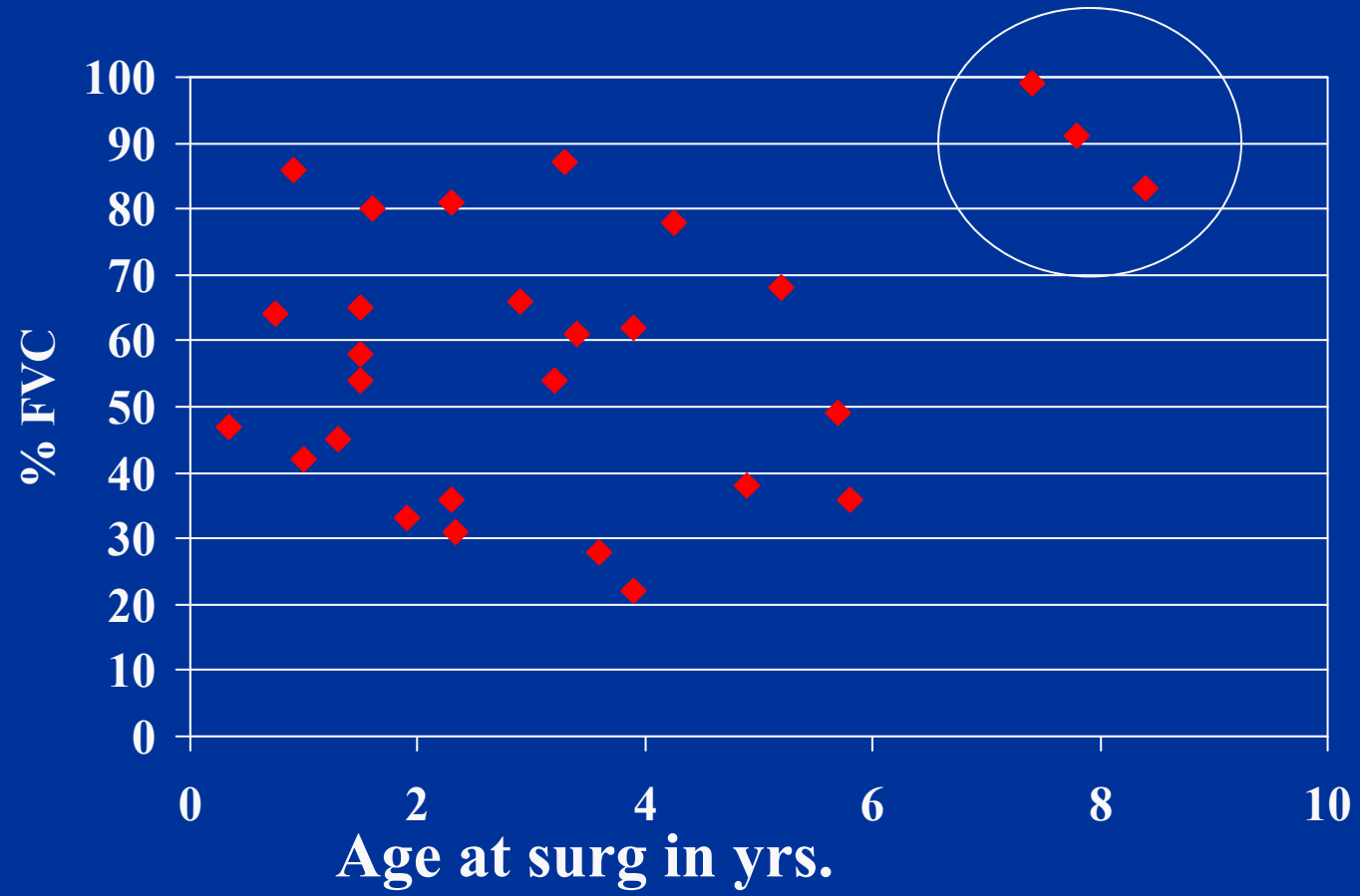


RESULTS: PFT'S

- FVC ave 57.8% normal (22-99%)
- FVC < 50%: 12/28 patients
  - *Severe RESTRICTIVE airway disease in **43%***
- Max inspiratory pressure < 80: 11/25 patients
  - *WEAKNESS of chest in **44%***
- FEV1/FVC < 85%: 10/28 patients
  - *OBSTRUCTIVE airway disease in **36%***

RESULTS: %FVC VS. AGE

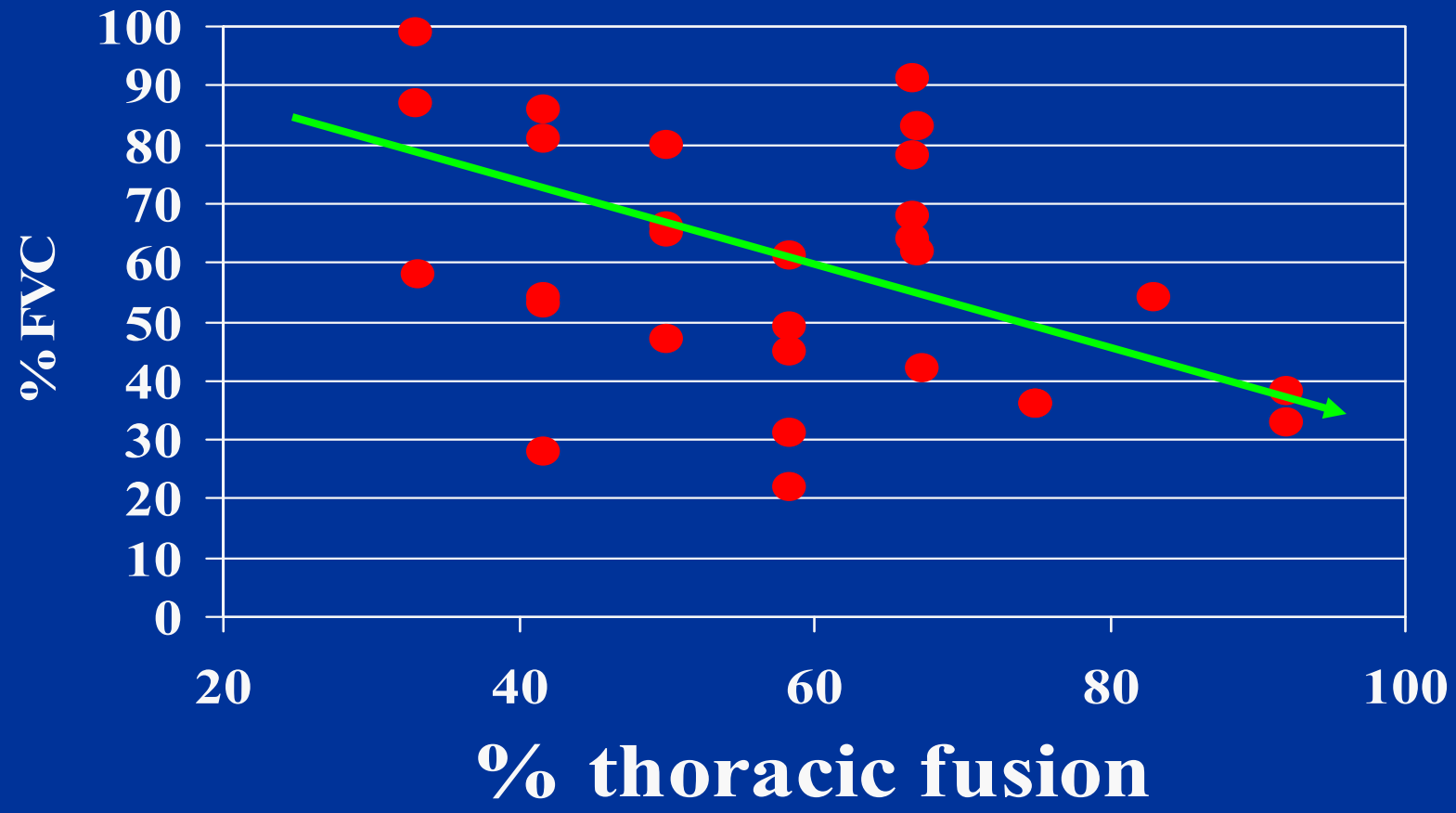
% FVC did not correlate with age at surgery (all < 8 y when operated) (r=0.28, p=0.15)



FVC vs. % THORACIC FUSION

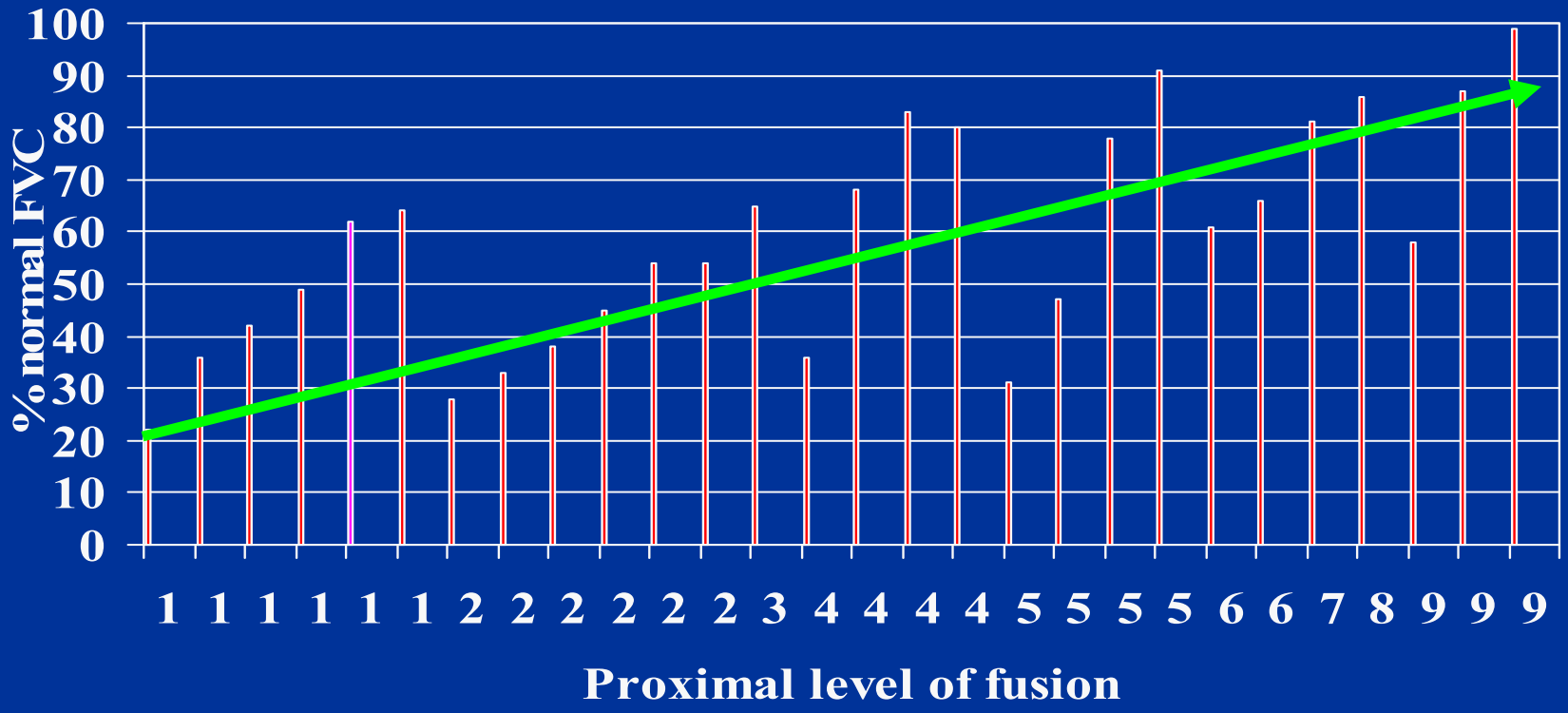
Larger % of thoracic fusion correlates with diminished FVC.

(p=0.01)

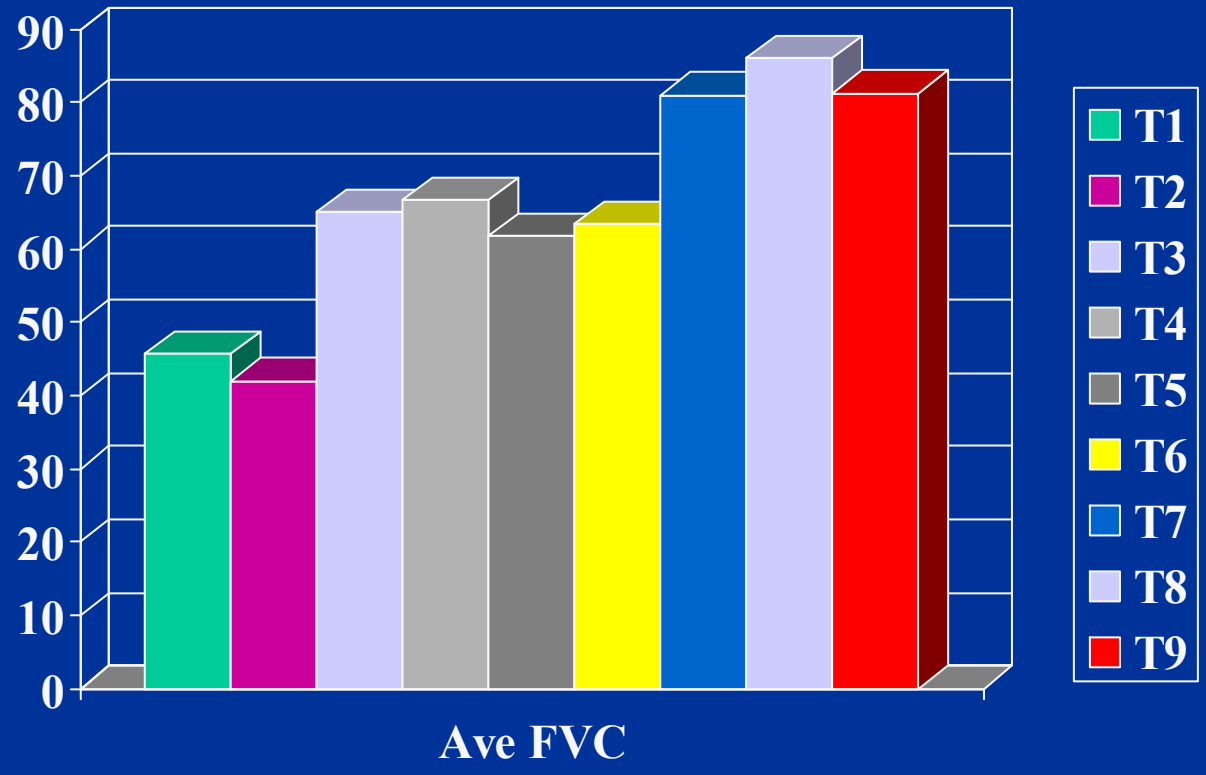


PFT'S: SPIROMETRY

- **FVC best in distal thoracic fusions**
- **Proximal thoracic fusions correlated with ↓FVC (p<0.0001)**



AVE FVC VS. PROXIMAL LEVEL OF FUSION

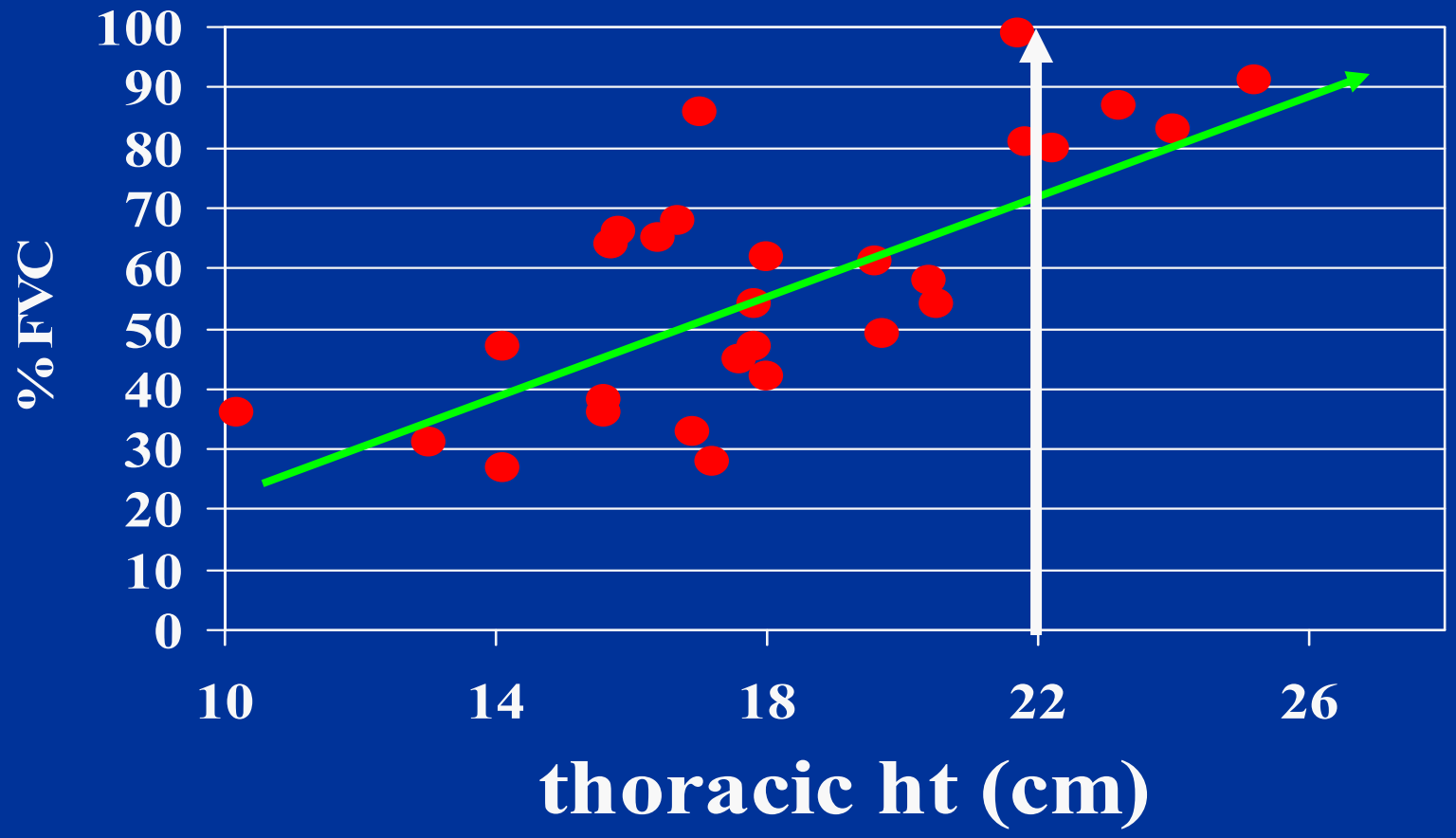


Fusions beginning at T1 - T2 lead to inhibition of pulm development, fusions T3-T6 are intermediate, and fusions beginning at T7-T9 have minimal effect

FVC < 50%

- 67% (8/12) of fusions beginning at T1 or T2
- 25% (4/16) of fusions beginning at T3-T9
- r=0.62, P=0.0004
- Stronger correlation than % fusion vs. FVC!

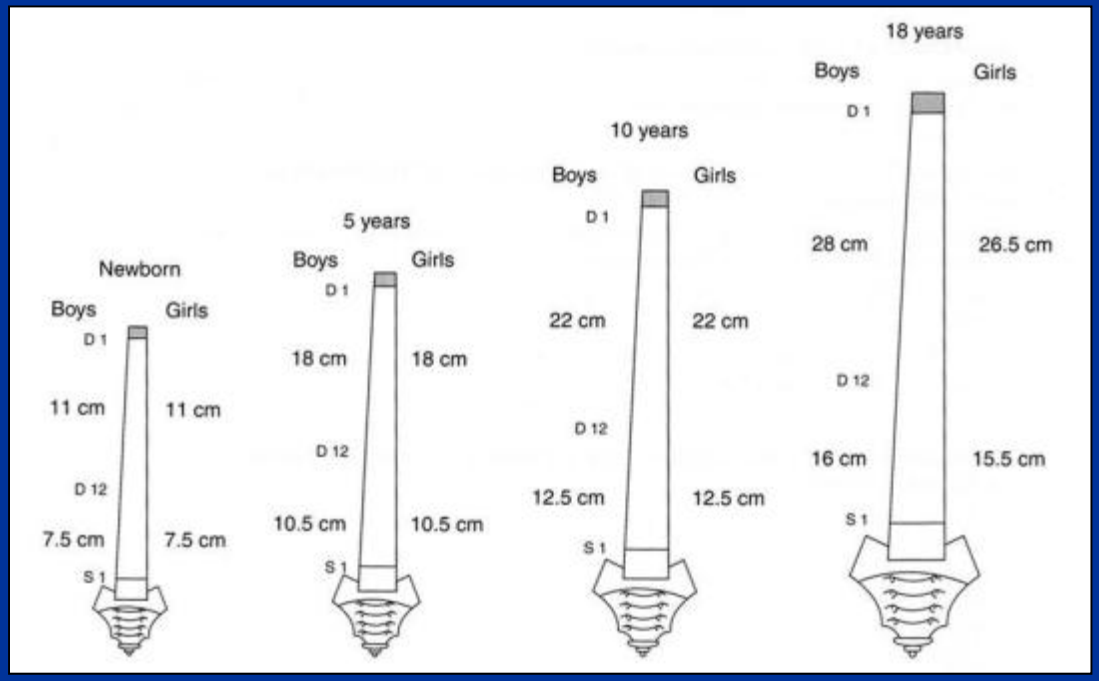
FVC VS. THORACIC HEIGHT



(r=0.73, p<0.001)

THORACIC HEIGHT

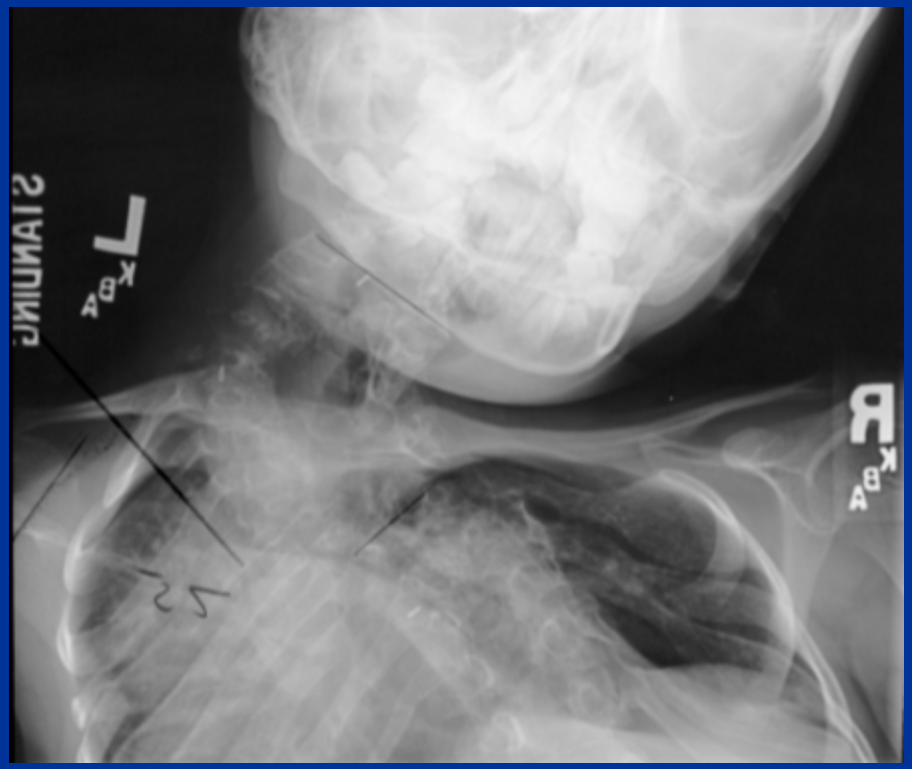
- 16/28 patients had thoracic height < 18 cm
  - Less than normal for 5 year old
  - 62.5% of these patients had FVC<50%



Case 1

- 9+2 year old female
- s/p excision of neuroenteric cyst
- ASF C7-T9/ PSF C3-T8 age 2+4 years
- Fused ribs
- 73% of thoracic spine fused

- Age 9 yrs (6.8 yrs postop)
- Hospitalized for pneumonia and required O2
- FVC 36% predicted
- FEV1 35% predicted



- Last seen 17+3
- Occasional pneumonia
- Not on BIPAP or O2

Case 2

- 10.7yo with toxoplasmosis
- ASF/PSF T2-L1 age 1.9 y
- 92% thoracic fusion
- Revision ASF/PSF T2-L4
- On BIPAP at night
- FVC 33%

- age 14+1 yrs
- On BIPAP qhs
- On O2 via vent q1hr
- Wheelchair for distance

- Now 19 + 8
- s/p VCR Jan 2011
- Thorax 17 cm long
- FVC 30-35% normal
- BIPAP nightly
- No O2 during day

Case 3

- 12 yo female
- Congenital scoliosis
- 8 y s/p asf /psf of 59% of thoracic spine
- Proximal level of fusion T1

- Age 12 years old
- FVC 27% predicted
- On BIPAP nightly

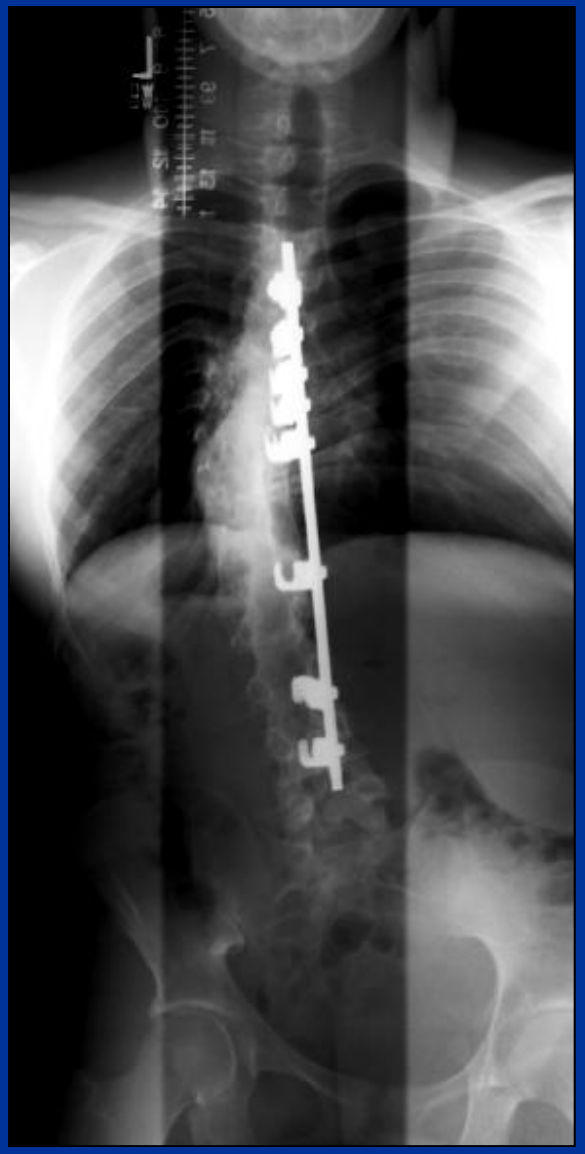
- Now 17 yrs
- On BIPAP qhs
- FVC 16% normal (WORST)
- Thoracic ht 13.7cm (<5yo)

Case 4 *** New TIS

- Asf/psf 2+4 T4-T11
- FVC 31% at time of study
- VCR 2011
- BIPAP qhs
- FVC 24%

Longest Follow-Up

- 22+10 year old male
- s/p PSF T7-T12 age 2 for congenital scoliosis
- Revised x 2
- FVC 81% predicted at 21 yr followup of 50% thoracic fusion



DISCUSSION

- Emans (SRS 2004) FVC ave. 62% compares well to our 58% predicted
- Also support Dimeglio and Canavese's work on proximal thoracic spine development and link with pulmonary growth

Other studies…

- Goldberg: fusion < age 10 for infantile scoliosis resulted in FVC of 41%
- Bowen: FVC 67% following early fusion for congenital scoliosis but short f/u (min 2 yrs, ave 6.7 yrs)
- Vitale: thoracic fusions <age 5 had FVC 64% and decreased quality of life

But It Gets Worse…

- Average age at study in our paper 14.6, Vitale study 12.6, and Bowen study 10.5
- Therefore PFT's done before the end of growth in many patients
- % predicted PFT's will worsen
- Evident in re-review of TSRH patients

NATURAL HISTORY

- PFT'S decline with age in normal adults beginning in the mid 30's
- 700cc loss in normal adult male (Kory)
- Pehrsson reported that when FVC is < 43% predicted, eventual respiratory failure is likely in adults with scoliosis

LIMITATIONS

- No nonoperated controls
- Untreated deformity will compromise respiratory function
- We’re simply saying that early fusion does not permit maintenance in lung growth!

CONCLUSIONS

36% of the patients have obstructive pulmonary disease.

- Important to test these children so they can be offered medical treatment
- Refer to pulmonologists

CONCLUSIONS

- 42% of our patients have restrictive lung disease s/p early fusion and 4/28 are symptomatic
- Low PFT's correlate with % thoracic fusion and thoracic height.
- **Children whose proximal level of fusion is T1 or T2 at highest risk!!**

Early Onset Scoliosis

- What we did then…
  - Were we always wrong?
  - Not really: older kids, distal isolated fusions
- What we do now…
  - Are we doing better?