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 Dubousset 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 $\leq$ 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)
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)
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
• 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?