Long term outcomes of EOS fusions
Average 34 years since surgery

John E Lonstein
Clinical Professor, Dept Orthopedics, University of Minnesota.
Twin cities Spine Center, Mpls.
Gillette Children’s Specialty Healthcare, St. Paul

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Disclaimers

F Royalties
  – Biomet Spine
Why don't we Fuse early?

• Effects of early fusion on PFT and growth of spine
Respiratory Function and Cosmesis at Maturity in Infantile-onset Scoliosis

- 21/32 patients
- 11 patients fused < age 10
  - Mean age at surgery 4.1 (1.4-7.8)
  - Mean age at follow-up 16.6 yrs. (12.6-23.9)
- Mean FVC 40.8% predicted (12-67%)

Goldberg CJ et al
Spine 28:2397, 2003
Pulmonary Function Following Early Thoracic Fusion in Non-Neuromuscular Scoliosis

By Lori A. Karol, MD, Charles Johnston, MD, Kiril Mladenov, MD, Peter Schochet, MD, Patricia Walters, RRT-NPS, and Richard H. Browne, PhD

- **28/54 patients**
  - Congenital scoliosis 20
  - Idiopathic 3
  - NFT 3
  - Congenital kyphosis 1
  - Syndromic 1

- Average 59% thoracic spine fused
- Average age at surgery 3.3 yrs.
- Average age at follow-up **14.6 yrs. (7.3-17.8)**
- Average follow-up **11.3 years**

JBJS 90A:1272, 2008
Results

- FVC average 57.8% predicted normal
- Inverse relationship to extent of thoracic fusion
- 16/20 had thoracic height <18cm
- Shorter the thoracic spine, smaller the FVC
- Correlation between proximal level of fusion and decreased FVC

Karol LA et al JBJS 90A:1272, 2008
FVC VS. THORACIC HEIGHT

(r=0.73, p<0.001)
A Retrospective Cohort Study of Pulmonary Function, Radiographic Measures, and Quality of Life in Children With Congenital Scoliosis

- **21/62** patients
- Mean of **5.1** levels fused
- **12** thoracic fusions
- Mean age at surgery **4.9 yrs.**
- Mean age at follow-up **12.6 yrs. (7-19)**

Vitale MG et al
Spine 33:1242, 2008
Results

F FVC mean 74% predicted
F Thoracic fusions mean of 64% predicted

Vitale MG et al
Spine 33:1242, 2008
PFT in Congenital Scoliosis

F 43 patients
  - 30 NS (No surgery)
    • Av. curve 52°
    • Age at presentation 5.6 yrs.
    • Age at f/u 10.8 yrs. (5.2 yrs. f/u)
  - 13 ES (Early surgery)
    • Av. Curve 80°
    • Age at presentation 2.9 yrs.
    • Age at f/u 9.7 yrs. (6.8 yrs. f/u)

PFT in Congenital Scoliosis

- Restrictive lung disease
  - NS – 69% FVC
  - ES – 67% FVC
- FVC correlated with
  - Decreased SAL
  - Higher thoracic apex
  - Decreased normalized thoracic width
- In ES group trend to FVC with
  - Longer follow-up
  - Longer fusion

All short follow-up
What happens in adulthood with longer follow-up?
Long term F/U study

F Inclusion criteria
- EOS scoliosis (Congenital or syndromic)
- Fusion < age 8 years
- 5+ thoracic levels fused
- Current age > 20 years
Study

- Retrospective chart and x-ray review
- Identify patients
- Trace patients
- Follow-up
  - X-ray
  - Questionnaire
  - PFT with blood gases
52 patients met criteria
23 traced
21 agreed to participate
12 complete data
Diagnoses

F 7 males
F 5 females

F 10 Congenital scoliosis
F 1 Diastrophic dysplasia
F 1 Camptomelic dwarfism
Congenital Scoliosis

F Vert. anomalies
  - 8 HV
    • 4 with an unsegmented bar
  - 2 unsegmented bar

F 8 fused ribs
  - 6 with an unsegmented bar

F Other
  - 3 Klippel-Feil
  - 2 diastematomyelia
Average Age

- At presentation 1.3 (0-4.2)
- At surgery 5.1 (1.2-7.8)
- At end of growth 17.3 (15-24)
- At follow-up 39.2 (22.7-59.8)
- Surgery to F/U 34.1 (15.6-54.4)
Scoliosis

- Presentation 52° (1.3 yrs.)
- Pre-op 54° (4.5 yrs.)
- 2 yr. Post-op 44° (6.2 yrs.)
- 5 yr. Post-op 37° (10.0)
- EOG 46° (17.6 yrs.)
- Follow-up 50° (39.2 yrs.)
Fusion Extent

Average 10 levels fused
PFT

- FVC average 58% predicted (18-131%)
  - PaO$_2$ average 87 mmHg (63-114)
  - PaCO$_2$ average 42 mmHg (35-76)

- 1 permanent O$_2$ and BiPap
  - Congenital
  - FVC 18%, PaO$_2$ 63, PaCO$_2$ 76

- 1 CPap for sleep apnea
  - FVC 49%, PaO$_2$ 69, PaCO$_2$ 40
TCSC 0 20 40 60 80 100 120 140
# Thoracic levels fused
FVC % Predicted
P=0.09

P=0.08

All
Congenitals
All

P=0.46

Congenital

P=0.90
Congenitals

![Graph showing FVC % Predicted vs. SAL with data points and trend line]
All

FVC % Predicted vs. T1T12 Length

P=0.02

Congenital

FVC % Predicted vs. T1T12 Length

P=0.06
T1-T12

Dimeglio & Bonnel, 1990

- Normal thoracic height by age

  - Newborn: 11 cm
  - 5 yo: 18 cm
  - 10 yo: 22 cm
  - Adult Female: 26.5 cm
  - Adult Male: 28 cm
T1T12 height
Congenitals in study

F Average 20.3 cm. (14.4-26.9)

F 4 Females
- Average 18.8 cm. (16.3-21.2)
- DiMeglio 26.5 cm.

F 6 males
- Average 21.4 cm. (14.4-26.9)
- DiMeglio 28 cm.
Height of Patients at the Visit Closest to Their 16th Birthday Versus 3rd, 50th, and 97th Centiles for Age

**Congenitals - 38 Boys**
- Av Age (yrs.) 16.4
- Mean height (cm) 161.89

**Normals**
- 97th centile (cm) 184.7
- 50th centile (cm) 171.5
- 3rd centile (cm) 158.3

**Congenitals - 79 Girls**
- Av Age (yrs.) 16.8
- Mean height (cm) 151.79

**Normals**
- 97th centile (cm) 173.5
- 50th centile (cm) 162.6
- 3rd centile (cm) 151.7

Goldberg CJ et al
Spine 27:1191, 2002

TCSC
Congenital T1T12 Height Female

- **Standing height 152 cm.** (Goldberg)
- **Sitting height 76 cm.** (50%)
- **T1S1 38 cm.** (50%)
- **T1T12 22.8 cm.** (60%)
  - Study (4 pts) **18.8** (16.3-21.2)
  - Normal **26.5 cm.** (Dimeglio)
Congenital T1T12 Height
Male
- Standing height 158 cm. (Goldberg)
- Sitting height 79 cm. (50%)
- T1S1 39.5 cm. (50%)
- T1T12 23.7 cm. (60%)
  - Study (6 pts) 21.4 (14.4-26.9)
  - Normal 28 cm. (Dimeglio)
Questionnaire

F Employment
- 4 Full time
- 1 Part time
- 2 each homemaker, student
- 2 Disability
Questionnaire

F Respiratory

- Dyspnea
  - 4 yes, 8 no
  - 4 with stairs, 8 no
  - 3 ADL, 9 no
Oswestry

- Average 8%
  - 7-0%
  - 2 – incomplete
  - 10, 34 & 46%
SF-36

- 4 domains low values
  - Physical component 48 (27-60)
  - Mental component 53 (34-66)
  - General health 66 (5-100)
  - Vitality 62 (20-90)
Summary

At average age of 39, 34 yr. F/U
- T1-T12 height shortened compared to congenitals
  - 2.3 cm in males
  - 4 cm in females
## Summary

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th># Patients</th>
<th>Av. Age at F/U (yrs.)</th>
<th>FVC%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Goldberg (2003)</td>
<td>Idiopathic</td>
<td>11 fused &lt;age10</td>
<td>16.6</td>
</tr>
<tr>
<td>Karol (2008)</td>
<td>Mixed</td>
<td>28 (20 cong.)</td>
<td>14.6</td>
</tr>
<tr>
<td>Vitale (2008)</td>
<td>Congenital</td>
<td>21 (12 thor.)</td>
<td>12.6</td>
</tr>
<tr>
<td>Bowen (2008)</td>
<td>Congenital (NS)</td>
<td>30</td>
<td>10.8</td>
</tr>
<tr>
<td></td>
<td>Congenital (ES)</td>
<td>13</td>
<td>9.7</td>
</tr>
<tr>
<td>Lonstein (2013)</td>
<td>Mixed</td>
<td>12 (10 cong.)</td>
<td>39.2</td>
</tr>
</tbody>
</table>
Summary

- FVC 58% predicted
  - 1 on permanent oxygen
  - 4/12 dyspnea
- FVC not correlated with
  - Extent of thoracic fusion
  - Proximal level fused
  - T1T12 length
  - SAL
- Restrictive lung disease in non fused congenital scoliosis
Summary

- Function well
  - Work
  - Back pain – Oswestry av. 8%

- SF-36
  - Wide range
  - Low physical and mental components
  - Slightly low general health and vitality scores
Problems with studies

- Small numbers
- Follow-up
  - Selection bias due to small numbers
- Mixes diagnoses
- Do not know “Normal” values for
  - IIS, JIS
  - Congenital
  - Syndromes
Thank You