Syndromes and cardiac problems: What Do We Need to Know and What are the Risks?

Glen Iannucci, MD
Assistant Professor of Pediatrics, Emory University School of Medicine
Sibley Heart Center Cardiology Aortic and Vascular Clinic
iannuccig@kidsheart.com
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## Disclosures

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<thead>
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<tbody>
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Children's Healthcare of Atlanta

Scoliosis

- Congenital Scoliosis
- Adolescent Idiopathic Scoliosis (80%)
- Neuromuscular Scoliosis
- Syndromic Scoliosis (3.5%)
- Increased risk of:
  1) Mitral valve prolapse (13-26%)
  2) Septal defects

30% Risk of CHD

Risk Depends on Underlying syndrome

? Cardiomyopathy
? Pulmonary hypertension

Risk Depends on Underlying syndrome

Increased risk of: 1) Mitral valve prolapse (13-26%) 2) Septal defects
Should all patients undergoing surgical repair have a preop echocardiogram?

13.6% had MVP

24% had valvar abnormalities

20% had abnormalities

Same institution as initial article referenced – this time excluded patients with known or suspected cardiac disease, neuromuscular disease, or suspected connective tissue disorder. Only 6% had “significant findings” (2 with ASD, 7 with aortic abnormalities).
Syndromic Scoliosis

11 yo M with mosaic Trisomy 17 – CXR
Courtesy of Hassan Sashemi (CHOA imaging scientist)
Marfan Syndrome

• Most common syndrome causing spine deformity (1:5000)
• Up to 60% have scoliosis with ~ 25-50% requiring intervention
• Some suggest higher blood loss
• Recent review suggests 5.8% risk of cardiac event during childhood
  – *Archives of Cardiovascular Disease; 2019 in press*
  – All events occurred in those with at least moderate aortic dilatation (Z score > 3)
### Marfan Syndrome – Complications during Spinal Fusion

#### Table 2

<table>
<thead>
<tr>
<th>Complications (%)</th>
<th>Marfan</th>
<th>Control</th>
<th>p Value</th>
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</thead>
<tbody>
<tr>
<td>Neurologic</td>
<td>2.4</td>
<td>0.79</td>
<td>.01*</td>
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<tr>
<td>Cervical spine-related</td>
<td>0</td>
<td>0.85</td>
<td>.11</td>
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<tr>
<td><strong>Pulmonary</strong></td>
<td>8.2</td>
<td>6.0</td>
<td>.19</td>
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<tr>
<td><strong>Cardiac</strong></td>
<td>1.7</td>
<td>2.8</td>
<td>.37</td>
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<tr>
<td>Thromboembolic</td>
<td>1.1</td>
<td>0.33</td>
<td>.11</td>
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<tr>
<td>Renal</td>
<td>0.35</td>
<td>0.53</td>
<td>.69</td>
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<tr>
<td>Infectious</td>
<td>0.69</td>
<td>1.4</td>
<td>.35</td>
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<tr>
<td>Implant-related</td>
<td>6.5</td>
<td>5.0</td>
<td>.36</td>
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<tr>
<td>Incidental durotomy</td>
<td>0.71</td>
<td>1.6</td>
<td>.25</td>
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<tr>
<td>UTI</td>
<td>1.6</td>
<td>2.8</td>
<td>.23</td>
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<tr>
<td>Any complication</td>
<td>20.0</td>
<td>19.2</td>
<td>.76</td>
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<tr>
<td>Died (%)</td>
<td>0.35</td>
<td>0</td>
<td>.3</td>
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<tr>
<td><strong>Blood transfusion (%)</strong></td>
<td>19</td>
<td>20.5</td>
<td>.62</td>
</tr>
<tr>
<td>Bone graft (%)</td>
<td>41</td>
<td>41.4</td>
<td>.9</td>
</tr>
<tr>
<td>BMP (%)</td>
<td>13.1</td>
<td>12.6</td>
<td>.8</td>
</tr>
<tr>
<td>Thoracoplasty (%)</td>
<td>1.4</td>
<td>2.0</td>
<td>.5</td>
</tr>
<tr>
<td>Osteotomy (%)</td>
<td>5.8</td>
<td>5.0</td>
<td>.61</td>
</tr>
<tr>
<td>Total charges, dollars (mean)</td>
<td>143,401</td>
<td>140,414</td>
<td>.66</td>
</tr>
<tr>
<td>Length of stay, d (mean)</td>
<td>7.2</td>
<td>6.2</td>
<td>.2</td>
</tr>
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</table>

PSM, propensity-score matching.
* Significance at p<.05.

N = 310 patients with MFS vs. controls (1:5)
Loeys-Dietz Syndrome

- Autosomal Dominant
- Classic Triad
  - Hypertelorism (90%)
  - Cleft palate or bifid uvula (90%)
  - Aortic aneurysm and arterial tortuosity
- Aggressive vascular disease (>> Marfan)
Ehlers Danlos Syndrome

• Four traditional forms
• Types I, II – Classic
  – Skin findings + hypermobility
• Type III – Hypermobile
  – Joint hypermobility dominates
• Type IV – Vascular
• Type VI - Kyphoscoliotic

Am J of Med Genet Part A 2010:152A;556-564
What condition does this patient have?

- Atrophic scar from skin tear
- Easy bruising
Cardiovascular Involvement in EDS

- Type I, II (“Classic”)
- Type III (“Hypermobile”)
- Type IV (“Vascular”)
- Type VI (“Kyphoscoliotic”) – very rare – vessel fragility reported, however no arterial rupture during spine fusion repair in recent case series (*Scoliosis* 2010;5:26)

Types I-III: ~ 6% incidence of mitral valve prolapse (adults)
~ 6% risk of aortic dilatation (adults)

General population: 3-4% incidence of mitral valve prolapse
2% incidence of aortic dilatation

**Virtually all patients with vascular EDS have a vascular event or organ rupture by age 40**
Trisomy 21 / Down Syndrome

- 1 in 700 live births in the US
- Scoliosis affects 10-55%
- High risk of scoliosis surgery complications
- 50% risk of congenital heart disease in this population
- Increased risk for pulmonary hypertension

Prader Willi

- 1 in 15,000 live births
- ~50% risk of scoliosis
- Increased risk of airway obstruction and pulmonary hypertension
Known heart disease – what is the risk?

Cardiac Risk Factors and Complications After Spinal Fusion for Idiopathic Scoliosis in Children

Christopher T. McKee, DO, David P. Martin, MD, Dmitry Tumin, PhD, and Joseph D. Tobias, MD

*Department of Anesthesiology and Pain Medicine, Nationwide Children’s Hospital, Columbus, Ohio
bDepartment of Pediatrics, Nationwide Children’s Hospital, Columbus, Ohio

Cardiac risk factors
304 / 7086 (4.3%)

Minor CHD (N = 140)
Major CHD (N = 144)
Cardiomyopathy (N = 20)

Rate of Complications
5%*
10%
40%
17 yo M with MFS s/p spinal fusion → recurrent R PTX x 2 in 3 months since spinal fusion
Infantile Marfan Syndrome – Cautionary Tale

Shortening of Growing-Rod Spinal Instrumentation Reverses Cardiac Failure in Child with Marfan Syndrome and Scoliosis

A Case Report

By David L. Skaggs, MD, Gerald Bushman, MD, Todd Grunander, MD, Pierre C. Wong, MD, Wudbhav N. Sankar, MD, and Vernon T. Tolo, MD

Investigation performed at Childrens Orthopaedic Center, Childrens Hospital Los Angeles, Los Angeles, California

14 mo old underwent growing rod placement without complication
- At time of initial lengthening (20mm) → developed acute severe ventricular dysfunction with elevated troponin and EKG changes

First report of orthopedic surgeon curing severe cardiac dysfunction

Aortic Dilatation – Beware if Hardware present

...
MAGEC rods → Magnets make MRI Artifact

MAGEC Rods: MRI conditional. Artifact extends out about 20 cm

Implications for patients who require serial surveillance imaging (ie Loeys Dietz Syndrome)
If you remember nothing else:

- High rates of echocardiographic abnormalities
  - Near 30% for all those undergoing surgery
  - Listen for murmur at apex while standing (MVP)
  - Low threshold to consider echo if considering surgery
- Red flags warranting cardiac evaluation before the OR
  - Symptoms
  - Exam findings suggestive of MFS or LDS
  - Concern for significant neuromuscular disease / OSA \(\rightarrow\) risk of pulmonary hypertension
- Cardiomyopathy = DANGER (40% risk of complication)
Thank you