Children's℠
Healthcare of Atlanta

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

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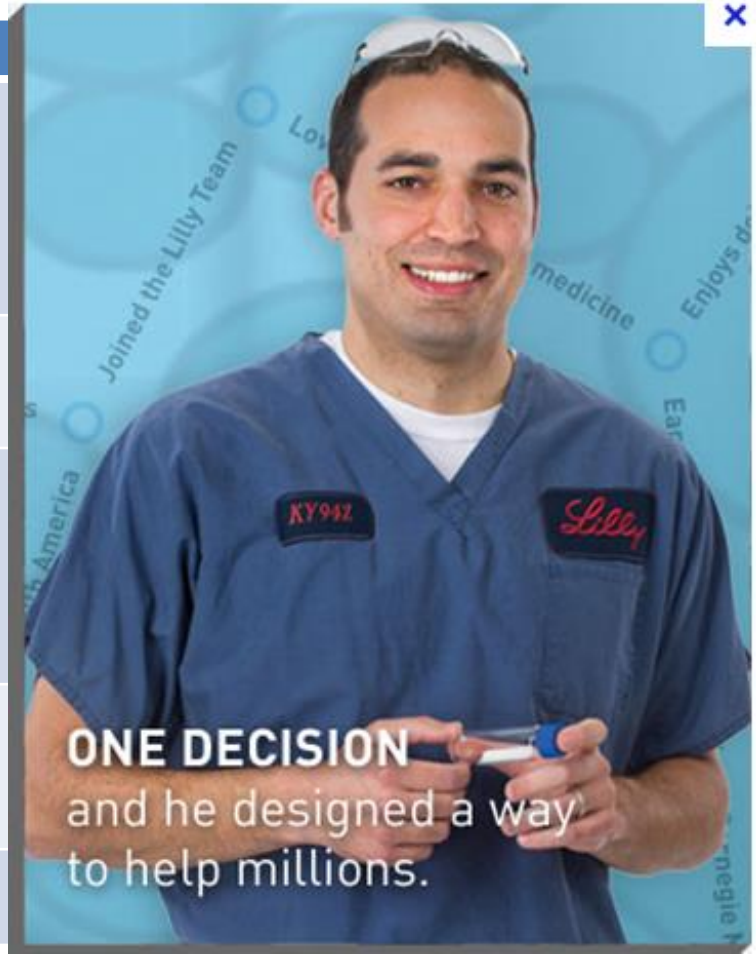


Scoliosis

Congenital Scoliosis

30% Risk of CHD

Syndromic Scoliosis (3.5%)

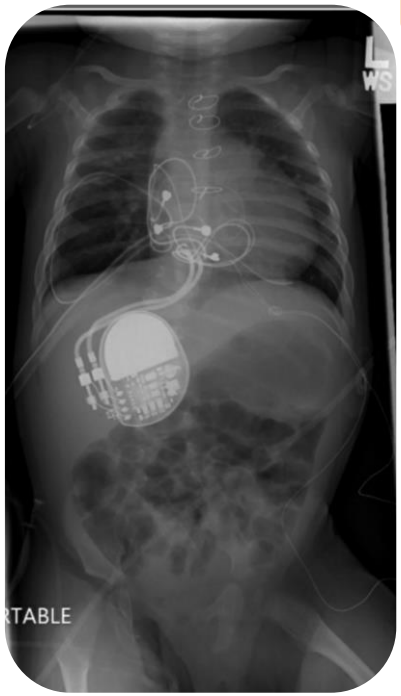
Risk Depends on Underlying syndrome

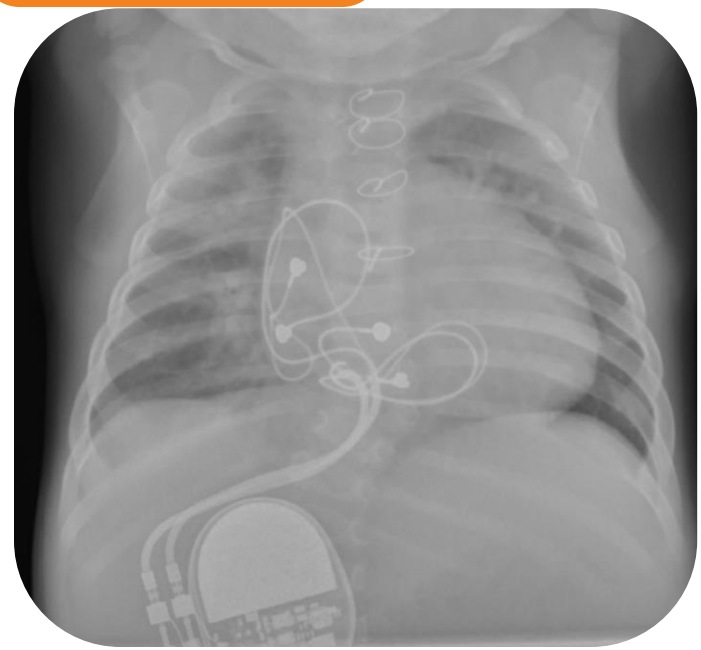
Neuromuscular Scoliosis

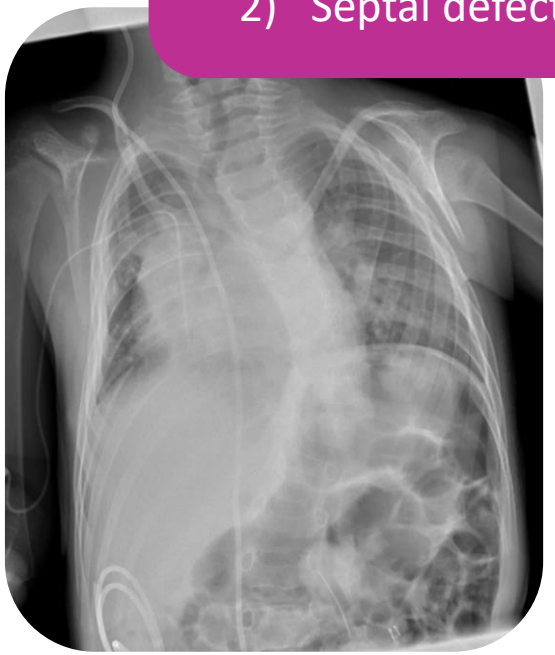
? Cardiomyopathy
? Pulmonary hypertension

Adolescent Idiopathic Scoliosis (80%)

Increased risk of:
1) Mitral valve prolapse (13-26%)
2) Septal defects ?







Should all patients undergoing surgical repair have a preop echocardiogram?

Pediatr Cardiol 18:425–428, 1997

Pediatric Cardiology

© Springer-Verlag New York Inc. 1997

Incidence and Risk Factors for Mitral Valve Prolapse in Severe Adolescent Idiopathic Scoliosis

13.6 % had MVP

Pediatr Cardiol 23:426–429, 2002
DOI: 10.1007/s00246-002-1443-2

Pediatric Cardiology

© Springer-Verlag New York Inc. 2002

Prevalence of Asymptomatic Cardiac Valve Anomalies in Idiopathic Scoliosis

24 % had valvar abnormalities

Prevalence of Cardiac Dysfunction and Abnormalities in Patients With Adolescent Idiopathic Scoliosis Requiring Surgery

20 % had abnormalities

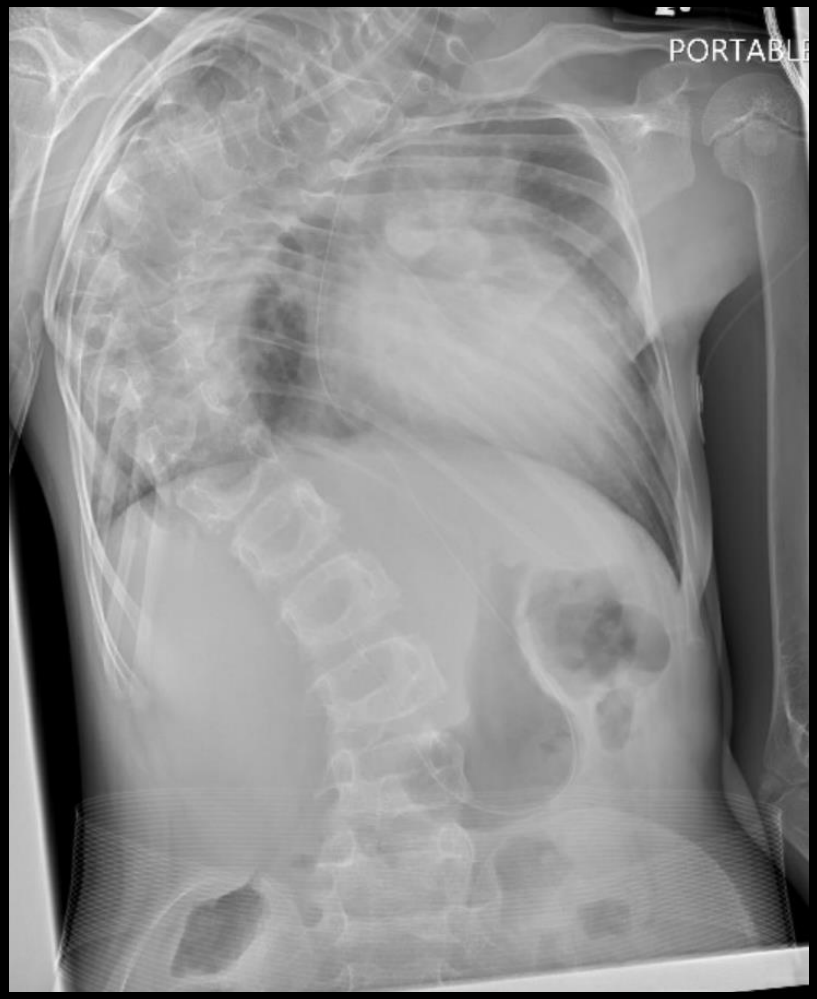
Journal of Pediatric Orthopaedics. 31(7):764–766, OCTOBER/NOVEMBER 2011
DOI: 10.1097/BPO.0b013e31822f14d6, PMID: 21926874
Issn Print: 0271-6798
Publication Date: October/November 2011

The Findings of Preoperative Adolescent Idiopathic Scolios

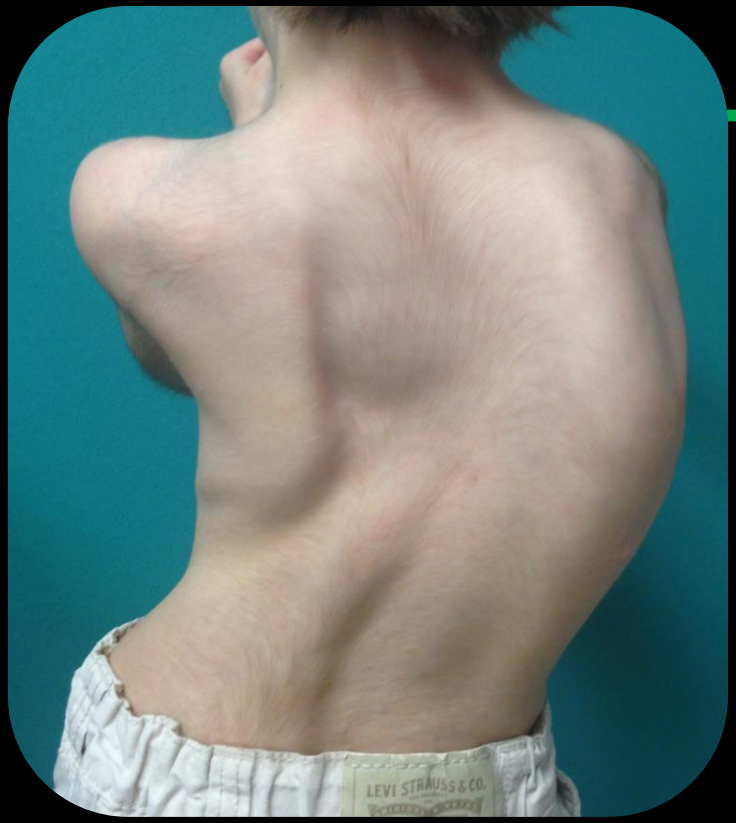
Lisa Ipp;Patrick Flynn;John Blanco;Daniel Green;Oheneba Boachie

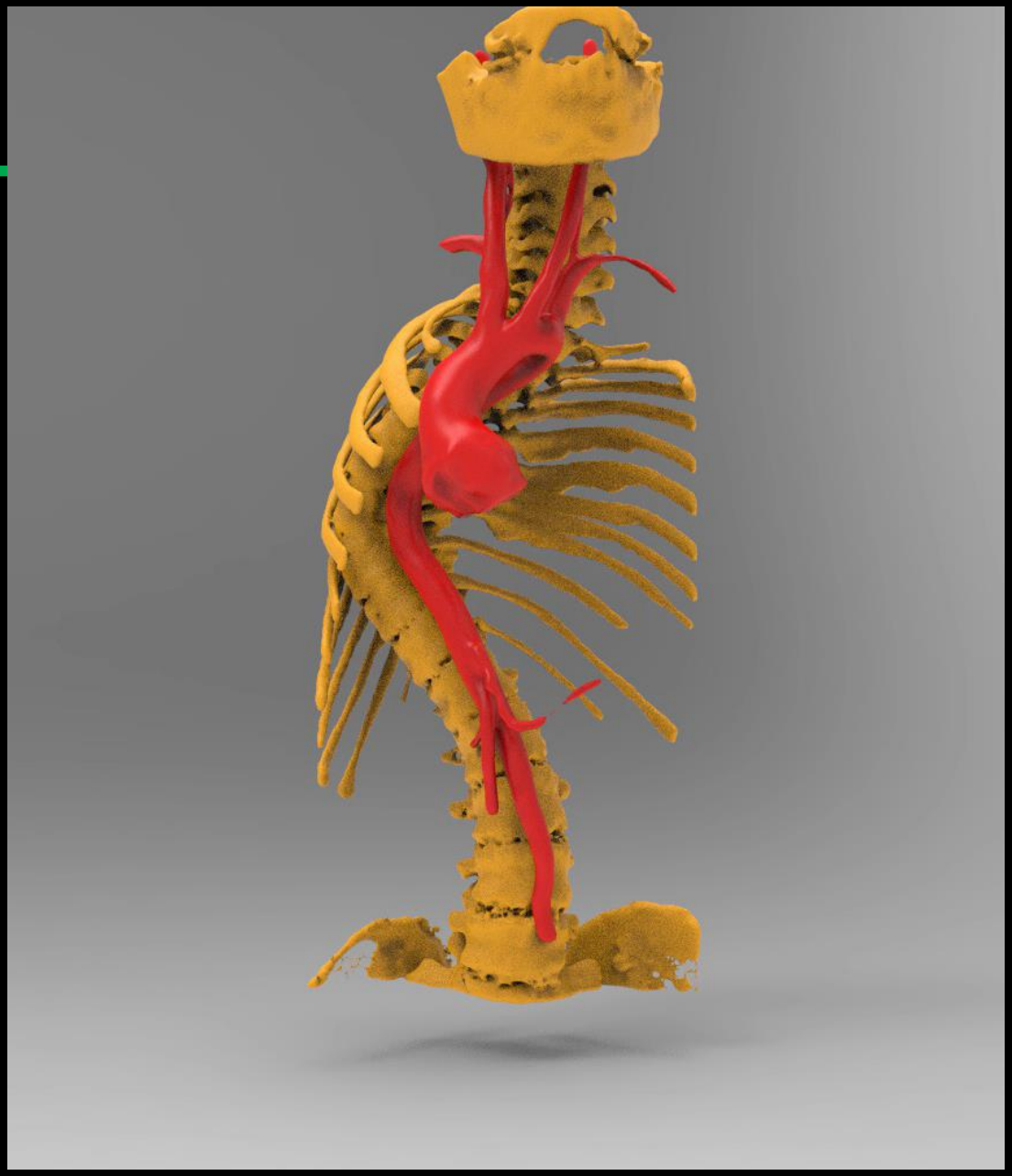
Same institution as initial article referenced – this time excluded patients with known or suspected cardiac disease, neuromuscular disease, or suspected connective tissue disorder
Only 6% had “significant findings” (2 with ASD, 7 with aortic abnormalities)

Syndromic Scoliosis



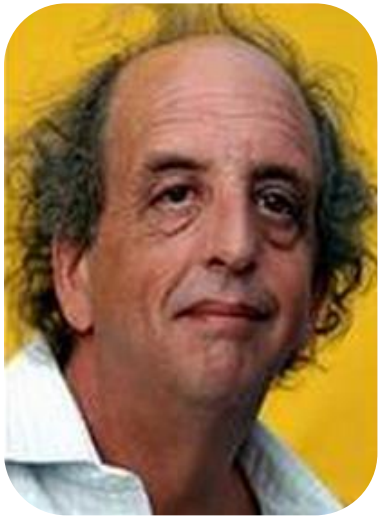
11 yo M with mosaic Trisomy 17 – CXR



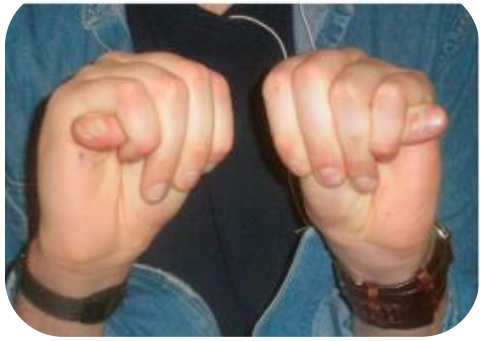


Courtesy of Hassan Sashemi (CHOA imaging scientist)

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)



PEDIATRICS Volume 132, Number 4, October 2013

Marfan Syndrome – Complications during Spinal Fusion

THE SPINE JOURNAL

Trend … rome

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Table 2
Outcomes of PSM-matched patients

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

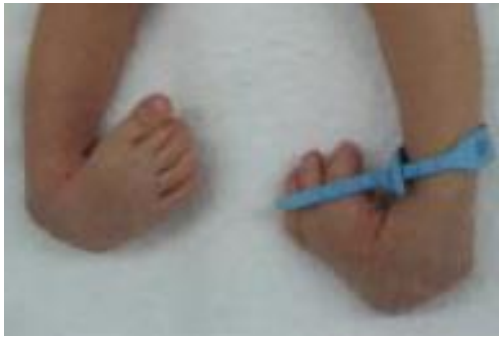
PSM, propensity-score matching.
* Significance at $p<.05$.

R… .4

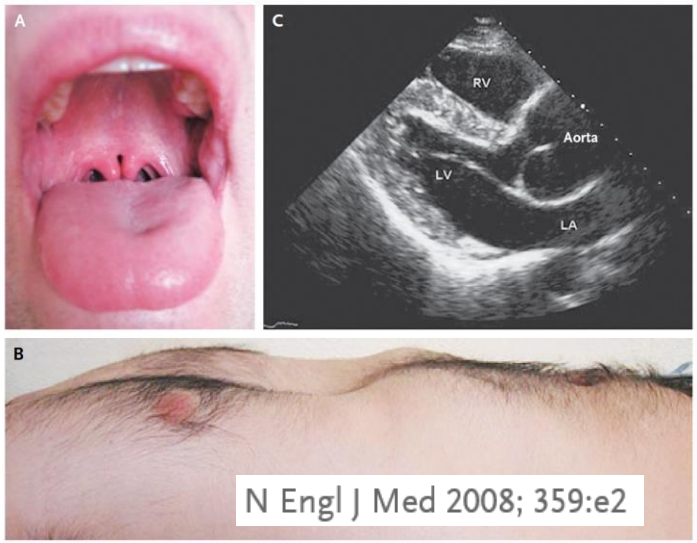
N = 310 patients with MFS vs. controls (1:5)

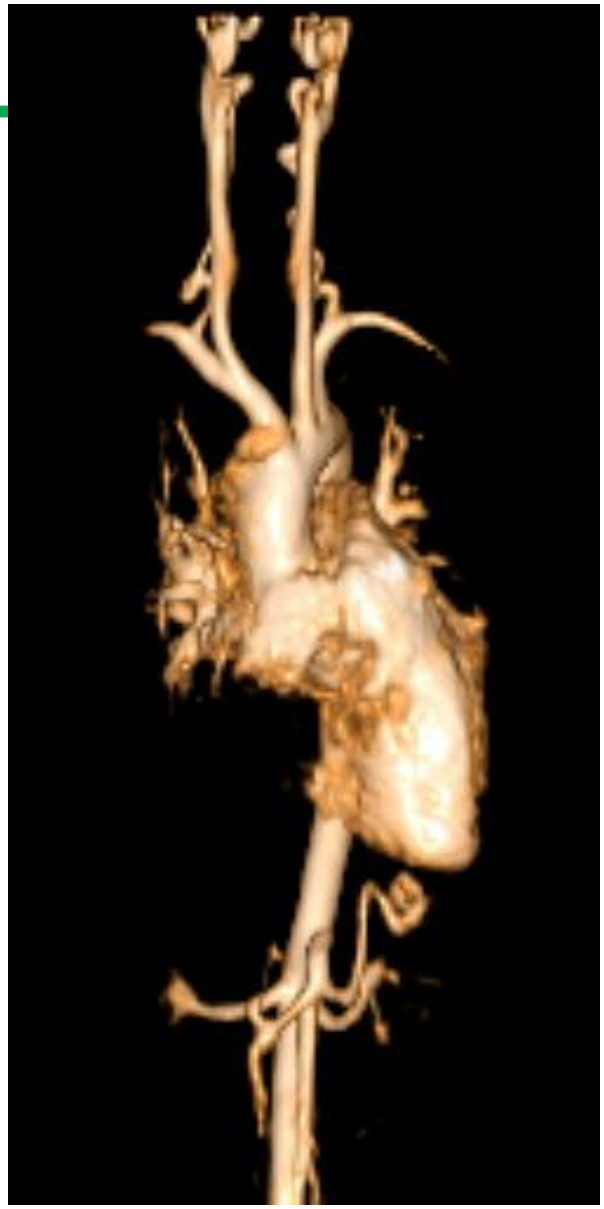
Loeys-Dietz Syndrome

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



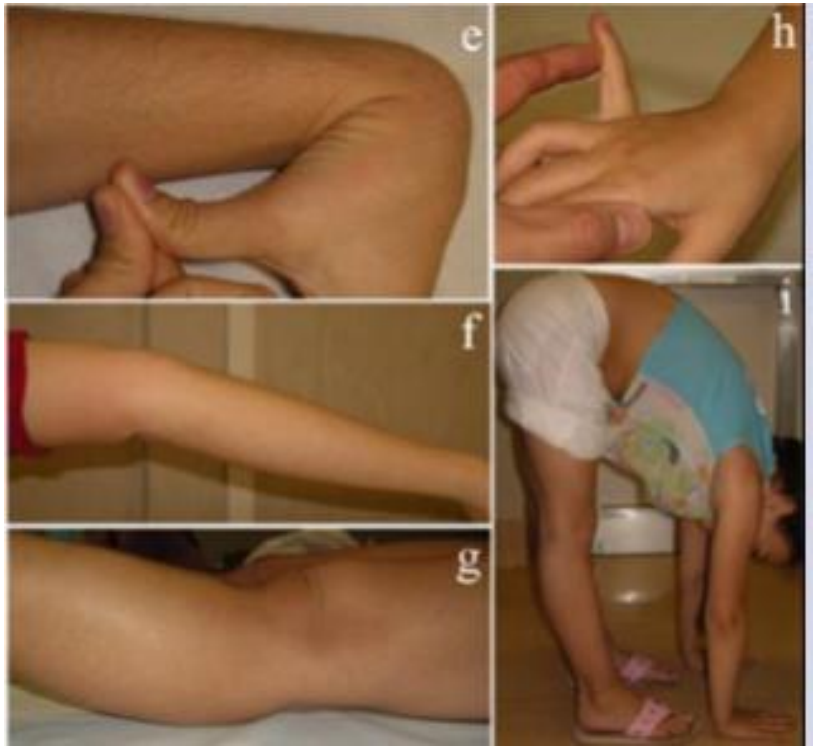
Am J Med Genet Part A 152A:417–421.



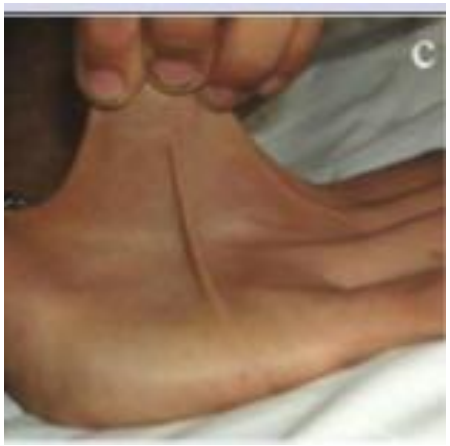


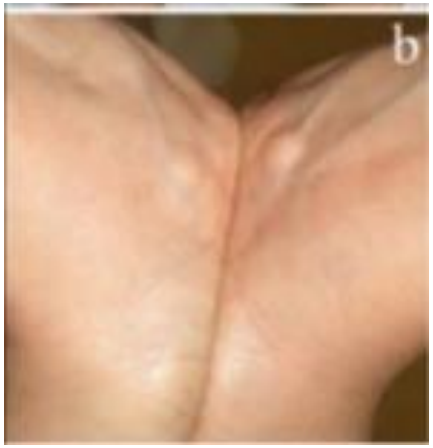
Ehlers Danlos Syndrome

- 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?

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,[a,b,] David P. Martin, MD,[a] Dmitry Tumin, PhD,[a] and Joseph D. Tobias, MD[a,b]*

[a] *Department of Anesthesiology and Pain Medicine, Nationwide Children's Hospital, Columbus, Ohio*
[b] *Department of Pediatrics, Nationwide Children's Hospital, Columbus, Ohio*

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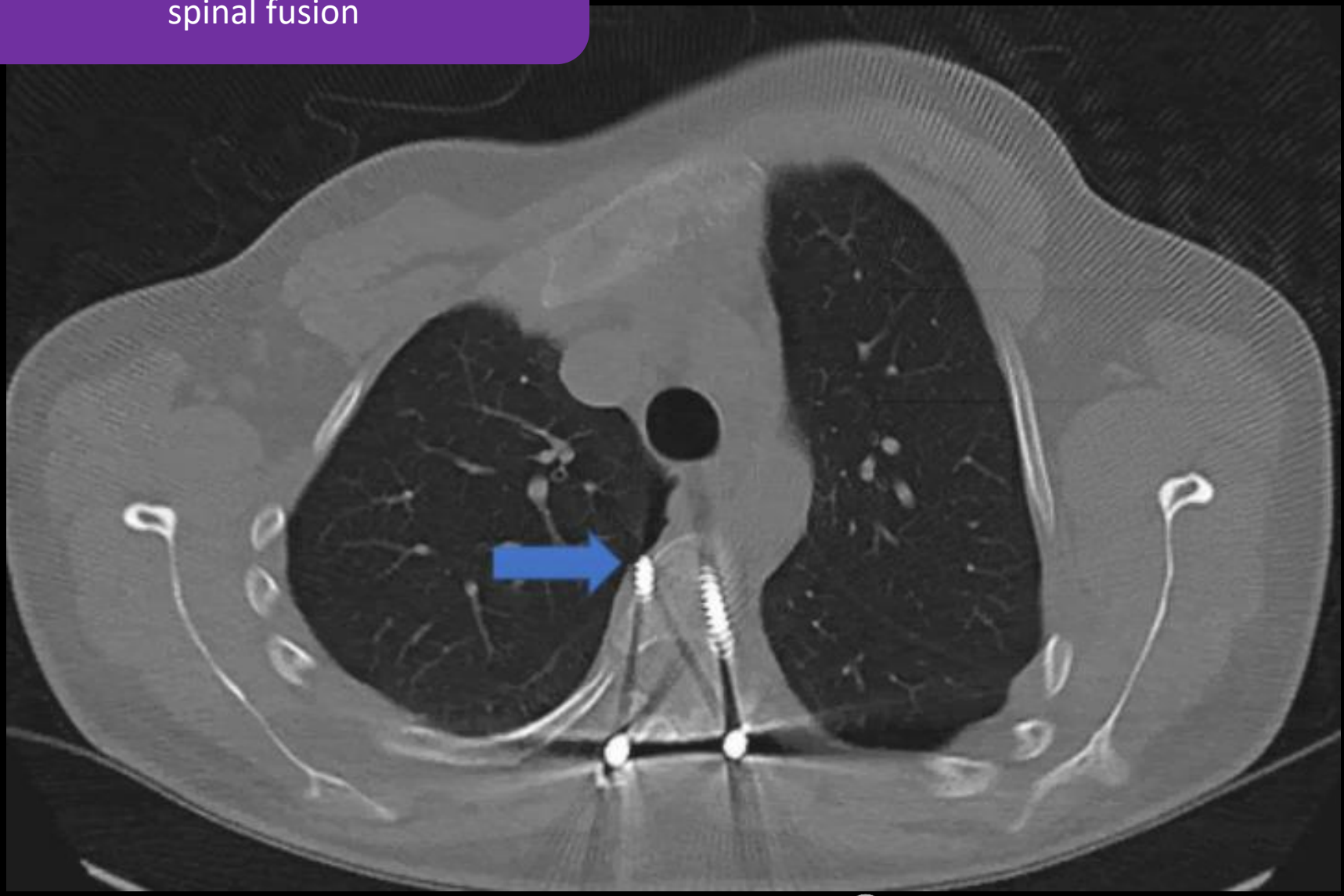
Cardiac risk factors
304 / 7086 (4.3%)

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

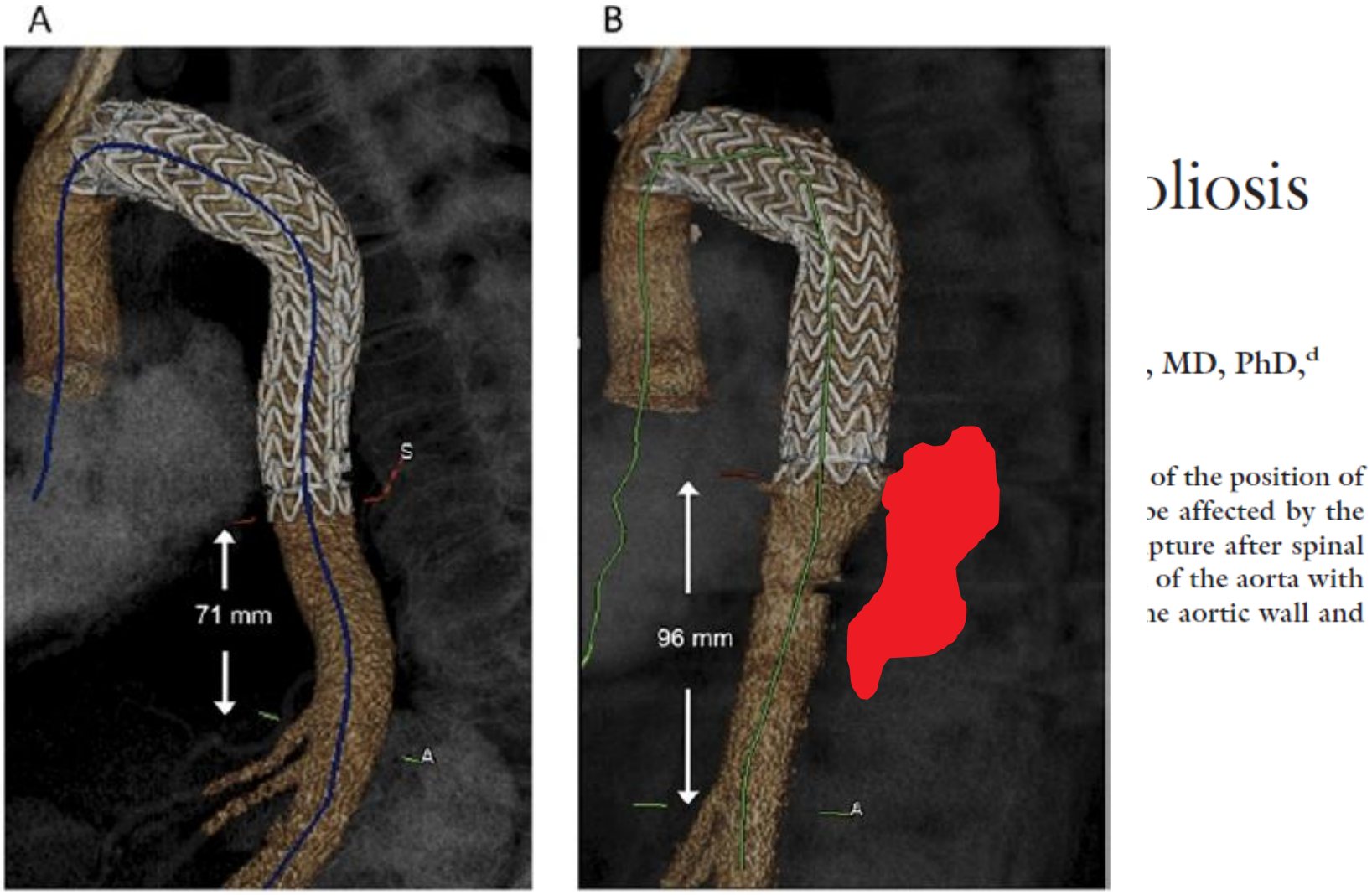
Investigation performed at Childrens Orthopaedic Center, Childrens Hospital Los Angeles, Los Angeles, California

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

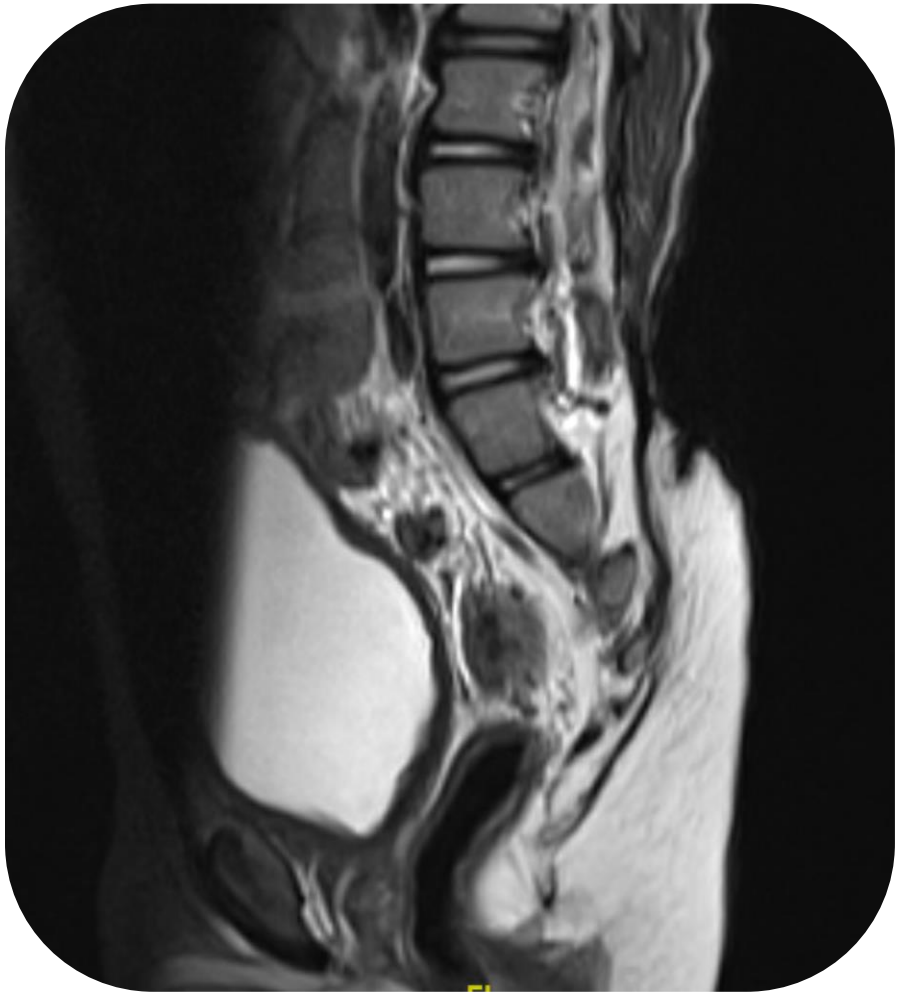
First report of orthopedic surgeon curing severe cardiac dysfunction

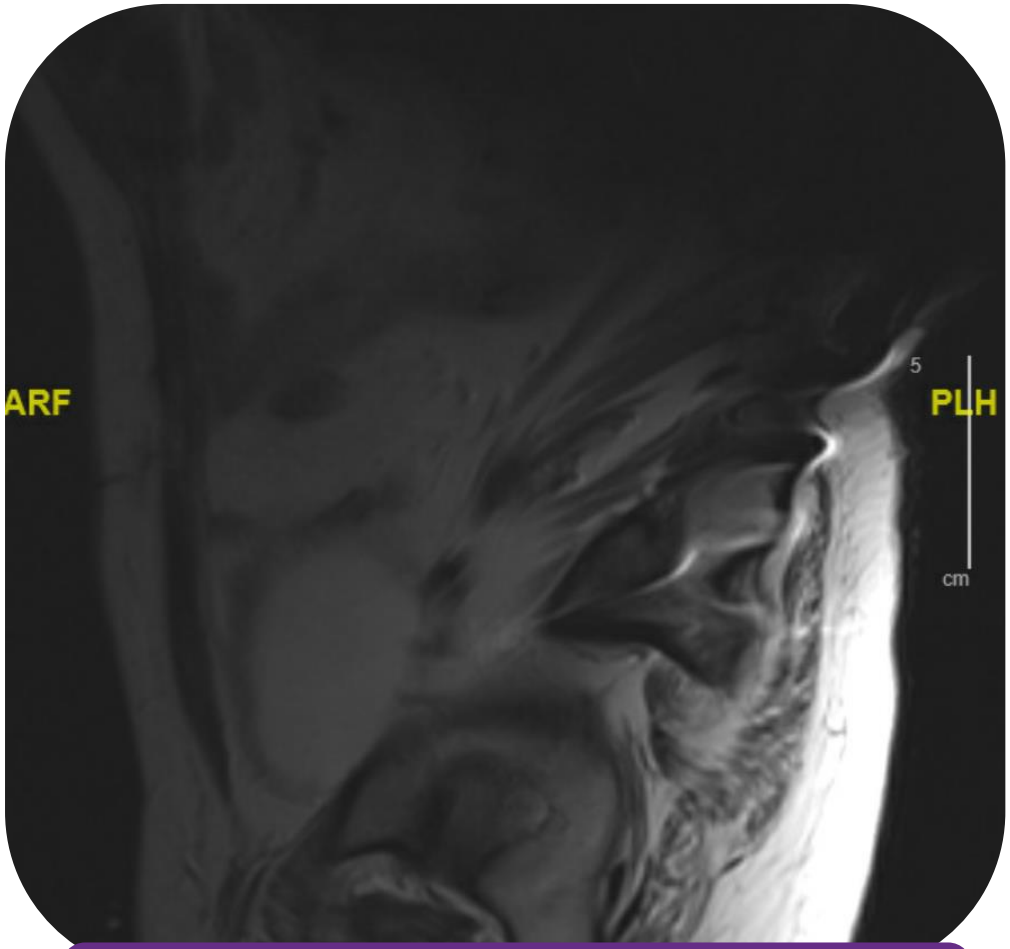
J Bone Joint Surg Am. 2008;90:2745-50

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 → risk of pulmonary hypertension
- Cardiomyopathy = DANGER (40% risk of complication)

Thank you