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Growth-friendly instrumentation for the treatment of early-onset scoliosis in Marfan Syndrome

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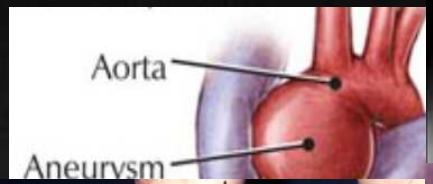
Background

- Marfan Syndrome: multisystem involvement
  - Cardiac
  - Ocular
  - Orthopaedic



Background

- ~60% of pts with Marfans (MFS) develop scoliosis
  - MOST present as teens
- Early-onset scoliosis (dx at <10yo)
  - **Progress more rapidly** (esp. 0-3yo)
  - **Respond poorly** to conservative treatments
  - **Little data** on surgical treatment

Sponseller PD, et al. The thoracolumbar spine in Marfan syndrome. J Bone Joint Surg Am. 1995;77(6):867-876.

Background

- Growth-friendly instrumentation (VEPTR, TGR, MAGEC)—multiple surgeries, **complications**
  - Fixation fail
  - Infection
  - Progress
  - Reduced

Do MFS patients experience unique outcomes and complications with these techniques?

Sponseller PD, Thompson GH, Akbarnia BA, Glait SA, Asher MA, Emans JB, Dietz HC 3rd. Growing rods for infantile scoliosis in Marfan syndrome. Spine (Phila Pa 1976). 2009 Jul 15;34(16):1711-5.

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Methods

Retrospective review: all pts with Marfans in CSSG and GSSG over 20 year span (1996 – 2016)

Inclusion Criteria:

- Clinical or genetic dx
- Initial eval for scoli at <10yo
- Growth-friendly instrumentation

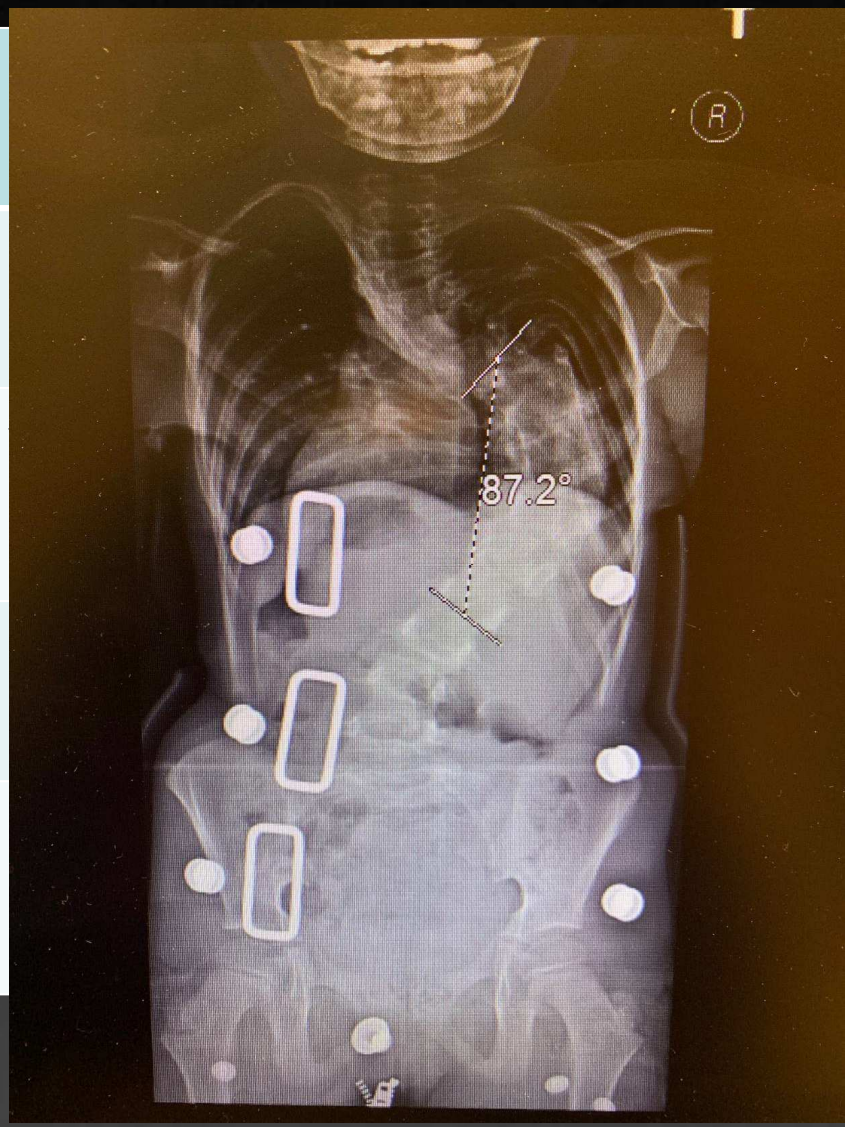
Exclusion criteria:

- Patients with less than 2 years f/u
- Treatment with initial definitive spinal fusion

→ **42 total patients met all of the above criteria.**

Results

Demographics	
% Male / Female	53 / 47
Mean age at 1st surgery	5.5 (±2.8)
Bracing attempted	23%
Ambulatory without device	77%



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Preop Radiographs

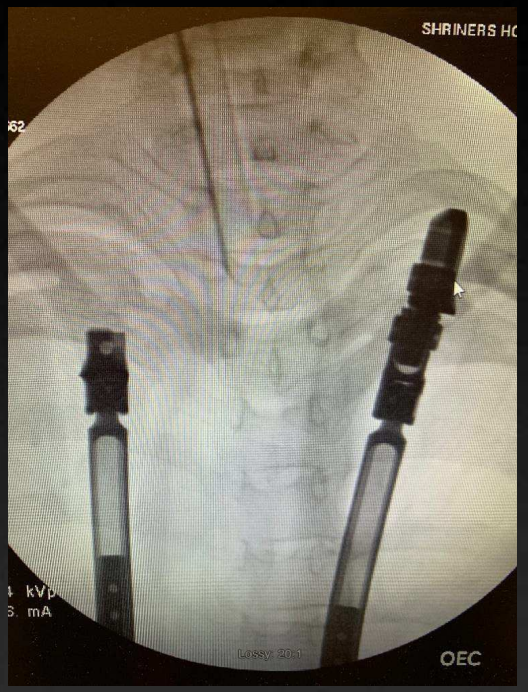
XR Measure	Cobb angle	Range
Major Coronal Cobb angle	**77.2** (±19.1)°	45 - 113°
Secondary Cobb angle	47.6 (±19.1)°	8 - 87°

- Mean thoracic height: **20.5 cm,** Smallest **10.8** cm
- Mean max kyphosis: **50.3** (±24.2)°
  - Mean T5-T12 kyphosis: 23.5 (±23.6)°
  - **Ranged -20 to 86 degrees**.

Karol LA, Johnston C, Mladenov K, Schochet P, Walters P, Browne RH. Pulmonary function following early thoracic fusion in non-neuromuscular scoliosis. J Bone Joint Surg Am. 2008;90: 1272–1281.

Surgical Data

- 28 spine-based TGRs
- 4 rib-based VEPTRs
- 8 MAGEC: 5 spine based, 3 rib based
  - 7 more pts converted to MAGEC during study period
- 2 Shilla constructs



Procedure Profile

Over the course of treatment, pts averaged:

- 5.7 lengthening surgeries
- 2.4 revision surgeries
- 7.1 total surgical procedures

Does not account for clinic MAGEC lengthenings

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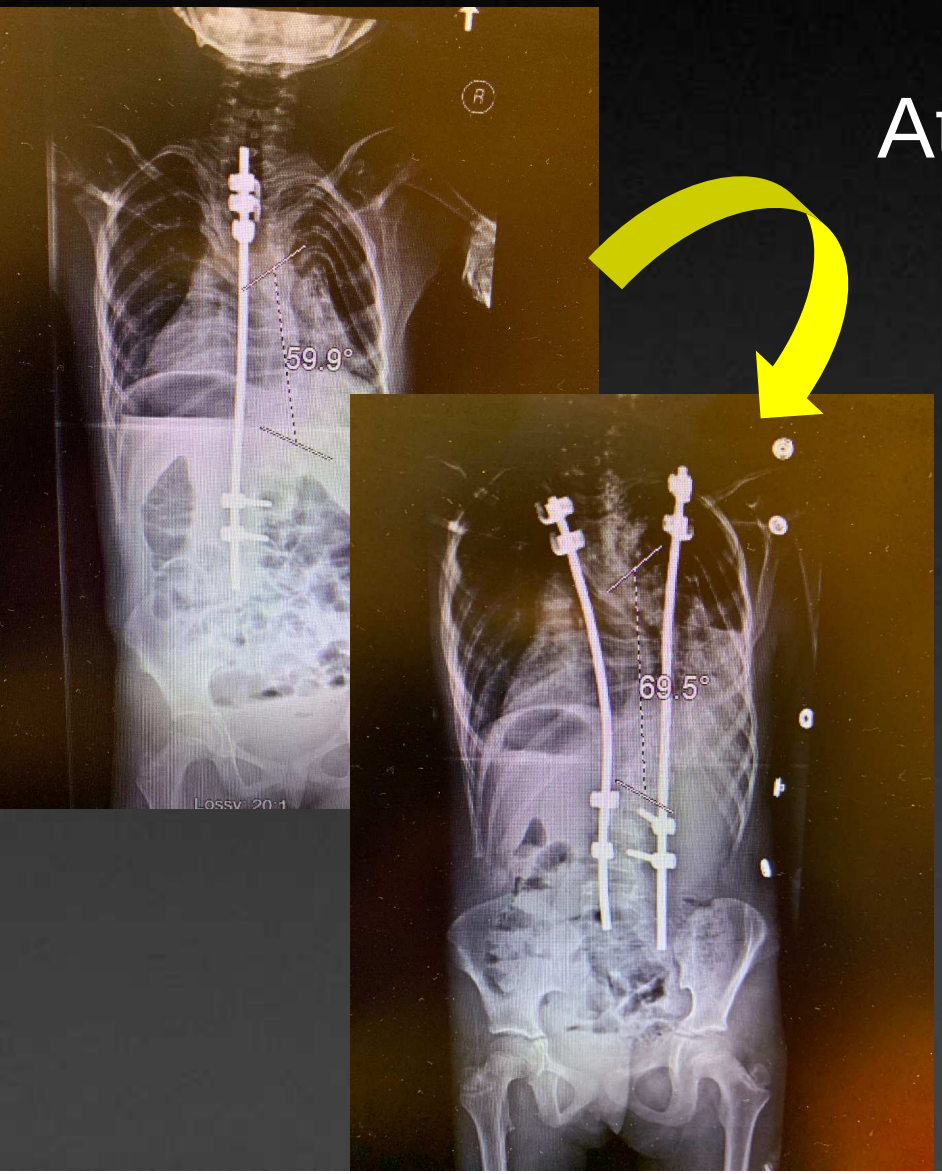
Implant Revisions



At time of final follow-up:

33% still had VEPTR/TGR

35% converted to fusion

30% MAGEC rods (18% at start of study)

1st Postop → Final XRs

Cobb:
- Spine: reduced 40 ±22°
- Rib: reduced 20 ±11°

p=0.004

Kyphosis:
- Spine: reduced 19 ±25°
- Rib: reduced 11 ±27°

p=0.038

Mean f/u: ***6.5 (±4.1) yrs***

nal Cobb 42.4 (±18.2)°

1° correction lost

Final max kyphosis: 42.3 (±20.8)°

10° correction lost from first f/u

Thoracic spine height: 23.8 (±4.2) cm

Complications

- 111 total complications
- Comp's per pt: **2.6** over study period
  - 0.40 comps per pt per year

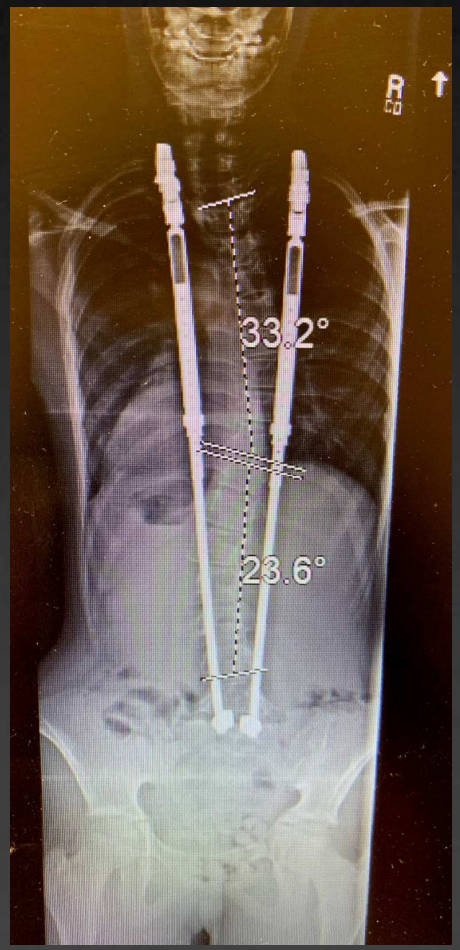
- **35% had no complications**
- 16% had 1 complication, 9% had 2….
- **14% had 6+**

Complications

- **42% implant failures/migration**
- 19% neurogenic symptoms, pain
- 14% dehiscence without infection
- 9% superficial infection
- 5% deep infection

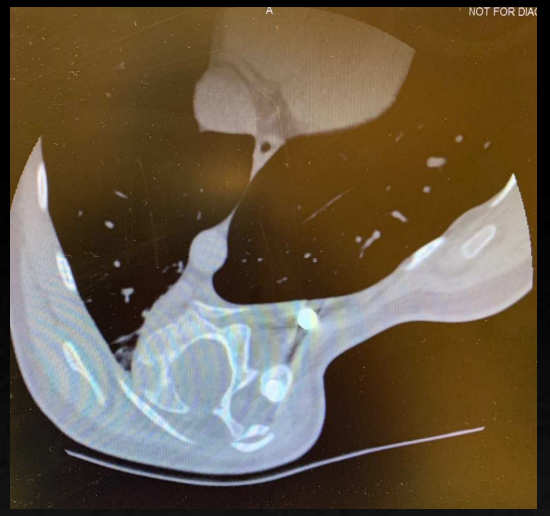
Conclusions

- Growing implants **allow for and maintain** radiographic correction in MFS
- *Bess et al, JBJS 2010*: 140 EOS pts
  - Total 897 growing rod surgeries (avg **6.4 surgeries per pt**)
  - 58% had 1+ complication, mean **2.2 complications per pt**



Bess S, Akbarnia BA, Thompson GH, et al. Complications of growing-rod treatment for early-onset scoliosis: analysis of one hundred and forty patients. J Bone Joint Surg Am. 2010;92(15):2533–2543

Conclusions



- Reoperation, complication rates similar to greater EOS
- Further collaboration needed to better study:
  - Spine vs rib based constructs
  - Other pt, surgical factors that impact correction
  - PRO’s