Scoliosis in children and adolescents with Friedreich’s Ataxia:

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Scoliosis in Friedreich’s Ataxia

1. What do we know?
2. Bracing: frequently asked questions
3. Surgery: frequently asked questions
What do we know?

- No enough!
- Why?
  - FA relatively rare disorder
  - Management of all spine deformities is rapidly evolving
  - No large series of FA scoliosis patients to provide proof that certain treatments are much better than others
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What do we know?

• Can we extrapolate from types of scoliosis we see often?
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What do we know?

• Can we extrapolate from types of scoliosis we see often?

• Probably not, because we don’t know what type of scoliosis children with FA have
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What kind of scoliosis?

Different types of scoliosis

• Idiopathic
• Neuromuscular
• Congenital
• Others
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What kind of scoliosis?

Idiopathic Scoliosis

• By far the most common type we see
• Pre-teen and teen girls > boys
• Strongly genetic—but gene(s) not known yet
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What kind of scoliosis?

Idiopathic Scoliosis

- Bracing recommended for progressive curves >25° in growing kids
- Surgery: curves greater than 45-50°
Neuromuscular Scoliosis

- Seen in children with neuromuscular disorders like
  - Cerebral palsy
  - Spina Bifida
  - Muscular Dystrophy
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What kind of scoliosis?

Neuromuscular Scoliosis

- Caused by the underlying condition
- Braces don’t prevent progression
- Different threshold and techniques for surgery
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What do we know?

2 published reviews on FA scoliosis

Montreal 1986

- 56 patients, 8-33 y/o
- Scoliosis > 60° progressed
- Scoliosis < 40° did not progress
- Scoliosis before puberty predicts progression
- No correlation between scoliosis progression, age at FA diagnosis, degree of muscle weakness, walking ability
What do we know?

2 published reviews on FA scoliosis

Dallas and Lexington Ky 2008

- 77 patients, 49 (63%) had scoliosis
- No association:
  - >10° at age <10 y/o and progression
  - Gender and progression
  - Age of scoliosis dx and progression
- Bracing
  - 10 pts braced, 8 eventually required surgery
- Surgery
  - 16/49 pts had surgery
  - Complications in 5/16 (30%)
What do we know?

- We know most will get scoliosis.
- We don’t know which kids with scoliosis will get worse.
- Braces less successful for FA scoliosis than AIS, but may delay progression.
- Surgery has a higher complication rate for FA than AIS.
Should your child with FA wear a scoliosis brace?

• Evidence is weak, but….
• There are no other options to slow progression
• Is there a downside? (walking balance, usual psycho-struggles with pre-teens and teens, etc)
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Bracing FAQs

When is the right time to start bracing?

- Follow rules of idiopathic scoliosis?
  - 25 degrees with significant growth remaining
  - Documented progression of the curve
How much should the brace be worn?

- Follow rules of idiopathic scoliosis?
  - 20 hours standard
  - Success drops considerably if brace is worn <16 hrs/day
When should the brace be stopped?

- Generally when there is “no growth remaining” (not getting taller)
- Bracing not used after skeletal maturity
- Can comfortably stop earlier if far from surgical range
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Surgery FAQs

When should we start thinking about surgery?

- Persistent progression above about 45-50°
- Especially if curve getting stiff
- Medical issues
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Surgery FAQs

What happens if we don’t do surgery?

- Progressive deformity throughout adulthood in curves bigger than 50°
- Progression is very slow
- Does not worsen life expectancy unless curves get very large (>100°?)
- Pulmonary effects of large deformity in the chest cage
What are the goals of surgery?

- Halt progression of the deformity by getting a solid fusion of the curved part of the spine
- Correct the deformity as much as is safely possible
What happens in the surgery?

- Place “anchors” in the bone (screws, hooks, wires)
- Loosen up spine by removing some of the joints (facets)
- Connect 2 rods, straightening as much as possible
- Large amounts of bone graft for fusion
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Surgery FAQs

What happens in the surgery?

[Images of X-rays showing spine before and after surgery]
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Surgery FAQs

What happens in the surgery?
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Surgery FAQs

What does NOT happen in the surgery?

- We do not use “Harrington rods” (1970’s technology)
- We do not take bone from the pelvis
- We do not use body casts after surgery
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Surgery FAQs

What is the typical in-hospital recovery (for children without FA)?

• 4-5 days inpatient stay
• Day 1: relax and recover in bed
• Day 2: sit up as start some eating
• Day 3: walk with PT, more normal diet
• Day 4: climb steps, pain medicine by mouth, x-rays and go home if ready
What is the typical in-hospital recovery?

• FA children who are walking before surgery would need MUCH more PT, perhaps even a brief period of in-hospital rehab

• Other medical considerations

• Expect a longer stay!
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Surgery FAQs

What is the typical early at home recovery (idiopathic scoliosis)?

- Can walk around, climb steps, get to bathroom, go outside
- No special bed or equipment necessary
- Very low energy level
- Each week stronger, most can return for a full school day after 4 weeks.
Thank You