The Use of Rib-Based Distraction in Dysplastic Early Onset Scoliosis associated with Neurofibromatosis Type 1

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Paper #33: The Use of Rib-Based Distraction

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Scoliosis associated with NF-1

- Develops early
- Aggressive
- Dysplastic
- Associated rib anomalies
- Poor response to bracing

Literature

- Jain et. al.: ICEOS, 2012
  - GSSG database
  - 14 pts with NF1
  - Treated with growing rods
  - High complication rate (50—70%)
  - Most common complication was failure of proximal spine anchors

Purpose

- Evaluate the effectiveness of rib-based distraction in managing dysplastic EOS in NF-1
- Evaluate complications with this technique

Methods

- Retrospective review
- CSSG database
- 12 patients with NF-1 treated with rib-based distraction
- Minimum 2 year follow (2-9 years)




Results

- Mean age at implantation: 6.3 years
- Mean Pre-op Cobb: 61 degrees
- Mean Post-op Cobb: 51 degrees
- Average lengthenings: 5.2
- No patient progressed beyond their pre-op Cobb
- Two patients have reached final fusion

Complications

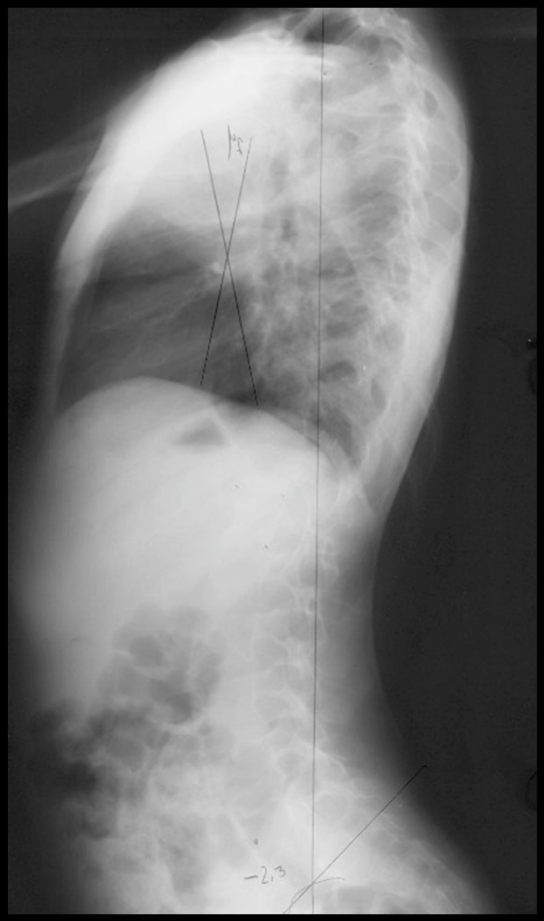
- 15 complications in 8 patients
  - Migration: 4
  - Wound Dehisence: 2
  - Rod breakage: 1
  - Medical issues: 5

Complication Grading

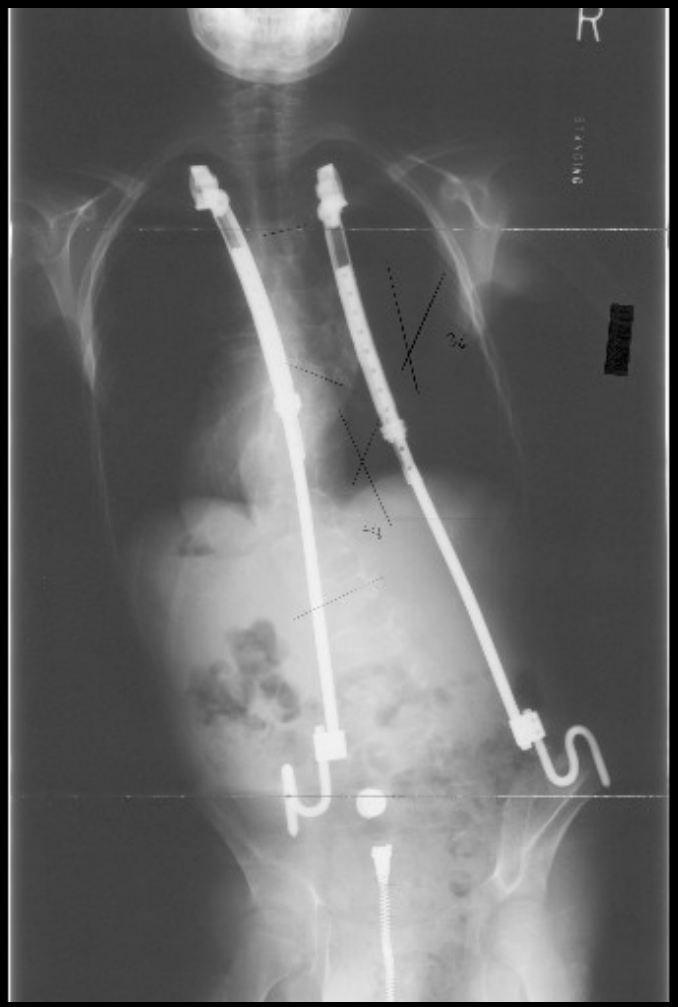
- Grade I: 8
- Grade IIA: 7
- Grade III: 0

Smith et. al. ICEOS, 2012

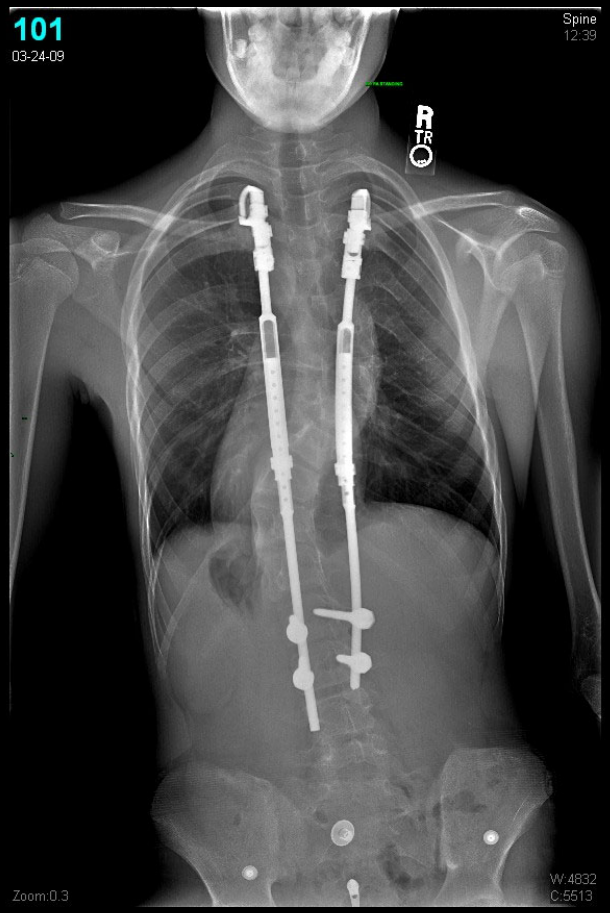
Case Example: 6 y/o male with NF1 and scoliosis



Initial VEPTR implant age 7

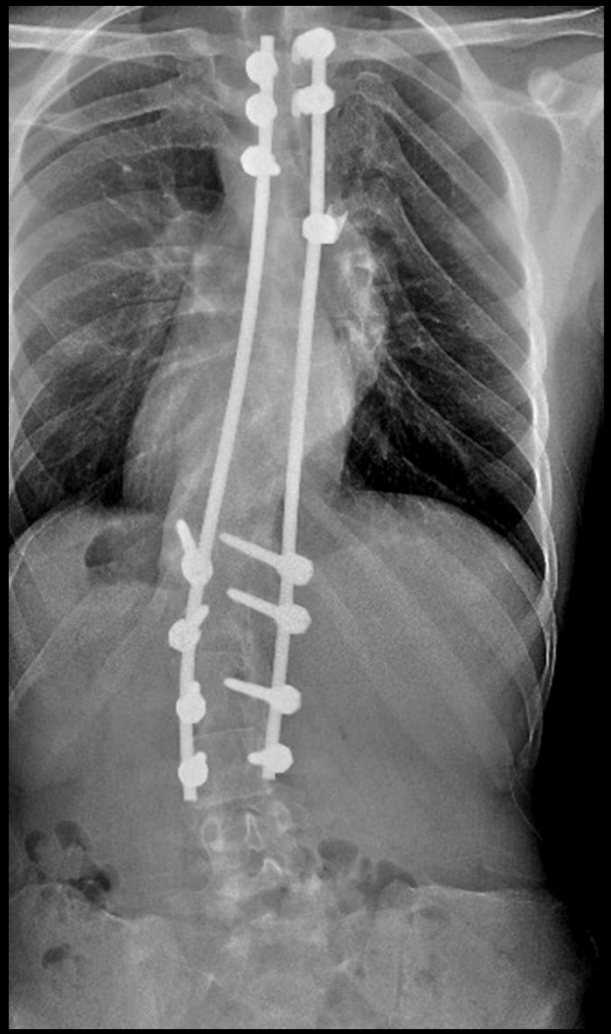


Five years s/p VEPTR



Make a wish……!

Final Fusion Age 16



Discussion

- Advantages of rib vs. spine anchors for growth-friendly systems are debatable
- NF-1 bone is often dysplastic and provides poor fixation as a *spine* anchor
- Consequences of screw anchor failures are potentially significant (neurologic injury)
- Consequences of rib failure are less significant and easily revised

Conclusions

- Rib-based distraction techniques effectively managed EOS in NF-1
- Incidence of rib migration was acceptable
- Complication rate as expected for growth-friendly treatment strategies

Thank you