

Growing Rods in Early Onset Scoliosis with Neurofibromatosis Type I (NF1)

Viral Jain, MD

Abhishek Ray, MD

Alvin H. Crawford, MD

John B. Emans, MD

Paul Sponseller, MD

Growing Spine Study Group



Disclosures

- Viral Jain None
- Abhishek Ray None
- Alvin Crawford Depuy Spine (A,B,C,E)
- John Emans Medtronic (B)
Synthes (E)
- Paul Sponseller Depuy Spine (A,B,E)
Globus Medical (E)
Oakstone Medical (E)
- GSSG GSF (A)

Introduction

- Skeletally immature patients with NF1 pose a unique challenge.
 - Dystrophic curves
 - Rapid progression
 - Brace/Cast ineffective
 - Fusion not an ideal treatment
 - Fusion stops the growth of the spine and chest
 - Crankshaft

Introduction

- Growing rods in the treatment of early onset scoliosis of various etiologies has shown to be successful

Purpose

- To evaluate the results of growing rods in patients with early onset dystrophic spinal deformities with NF1
- Compare the results with published results of growing rod procedures

Materials and Methods

- Pooled data
 - Growing Spine Study Group (GSSG)
 - Patients treated at our own institute

Materials and Methods

- Type of Study
 - Retrospective (level IV)
- Inclusion Criteria
 - Diagnosis of NF1 confirmed with genetic testing
 - Early onset spinal deformities (scoliosis $>20^\circ$ or Kyphosis $>45^\circ$ diagnosed before age 11)
 - Presence of dystrophic features
 - Minimum two years of follow-up

Materials and Methods

- **Exclusion Criteria**

- Incomplete radiographic follow-up
- Incomplete or inadequate follow-up (<2 years)

Materials and Methods

- **Complications**
- Wound complications
 - Superficial or deep infections
 - Others: painful scars.
- Implant related complications
 - Breakage, loss of foundation and prominent implants.
- Alignment complications
 - Junctional issues
- Neurological complications
 - Transient or permanent neurological deficit.
- General complications
 - Dural leaks, hematomas, and postoperative cardiopulmonary and gastrointestinal complications.

Results

- GSSG database query
 - 17 patients met inclusion criteria
 - 4 patients were excluded due to incomplete data
 - 3 were excluded due to <2 years of follow-up
- Cincinnati Children's Data
 - 4 patients met all the inclusion and exclusion criteria.
- Total of **14** patients were included in the study

Results

Total Procedures	71	5 per patient
------------------	----	---------------

General

Average follow-up	54 months	Range 24 to 95 months
Average Age	6.8 years	Range 2.7 to 9.7 years
Graduates	5/14	36%

Correction

Preop Cobb	74	52%
Post op Cobb	36	

Spine Growth (T1-S1)

After 1st surgery	3 cm
Total	6.6 cm
Rate	2cm / year

Complications of Growing-Rod Treatment for Early-Onset Scoliosis

Analysis of One Hundred and Forty Patients

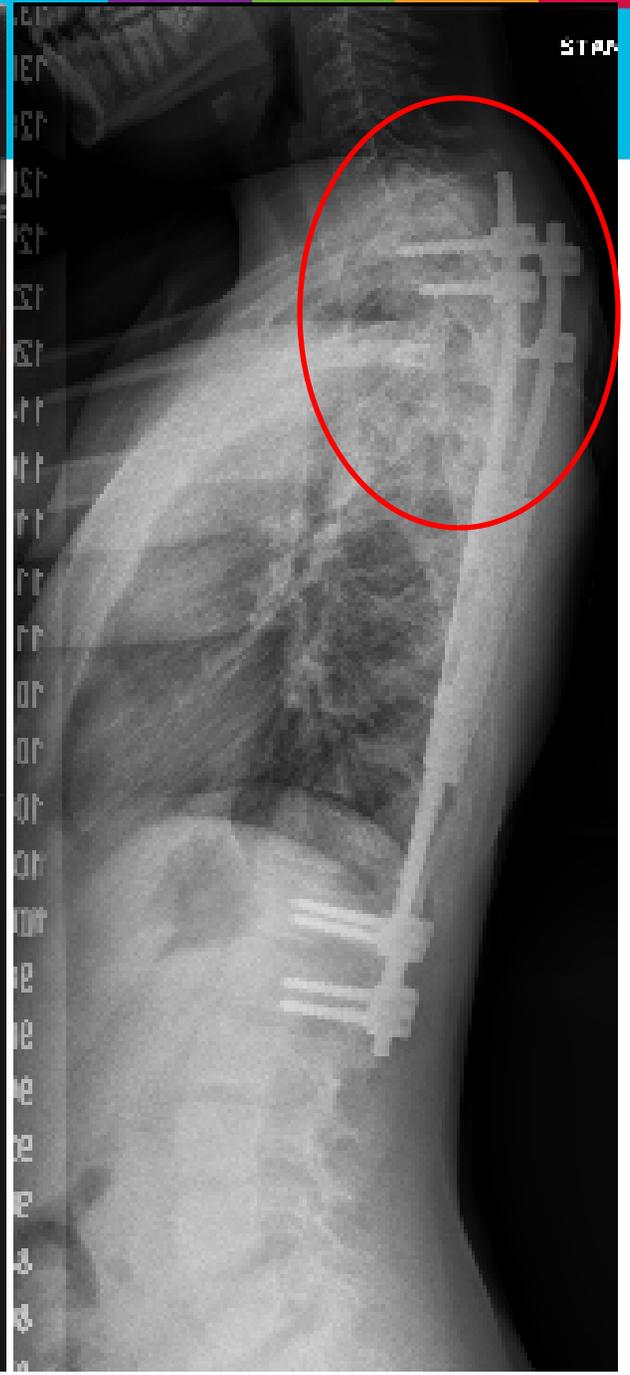
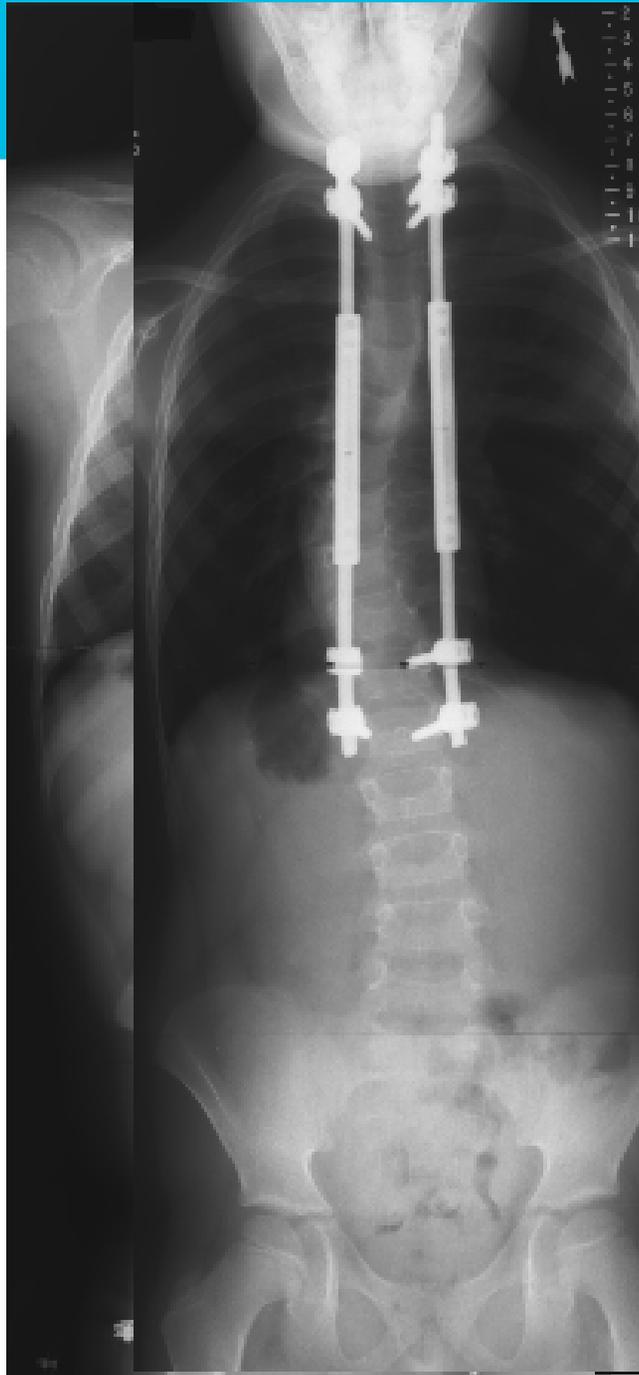
By Shay Bess, MD, Behrooz A. Akbarnia, MD, George H. Thompson, MD, Paul D. Sponseller, MD, Suken A. Shah, MD,
Hazem El Sebaie, FRCS, MD, Oheneba Boachie-Adjei, MD, Lawrence I. Karlin, MD, Sarah Canale, BS,
Connie Poe-Kochert, RN, CNP, and David L. Skaggs, MD

Results

- Complications
 - 17 complications in 14 patients (1.2 per patient)
 - 17 complications in 71 surgeries (23%).
- Bess et al JBJS 2010:
 - 1.3 complications per patient
 - 177 complications in 823 surgeries (20%)

Results: Complications

Type		Numbers	Percentage	Bess et al
	Total	9/14	64%	45%
Implant related	Failure of proximal construct	6/9	66%	
	Rod Breakage	2/9	22%	
	Prominent Implants	1/9	11%	
Alignment	Proximal Junctional Kyphosis	5/14	36%	7%
Wound	Deep Infection	2/14	14%	10%
Neurological	Sensory loss	1/14	7%	



Discussion

- Higher rates of proximal junctional kyphosis and failure of proximal construct
 - Type of anchor (screw vs hook) : Not significant
 - Pre-op Kyphosis: Not significant
 - Inherent bony dysplasia
 - Nature of the dystrophic curve
 - Rigid,
 - Unyielding
 - Sharply angulated
 - Proximity of dural ectasia

Conclusion

- Growing rod instrumentation for dystrophic spinal deformities in NF1 leads to excellent correction of deformity while allowing the growth of the spine.
- The complication rate of this technique is high but comparable to growing rod procedures for other etiologies.
- The most common complications are the failure of proximal anchors and proximal junctional kyphosis

References

1. DiSimone RE, Berman AT, Schwentker EP. The orthopedic manifestation of neurofibromatosis. A clinical experience and review of the literature. *Clin Orthop Relat Res*. May 1988(230):277-283.
2. Crawford AH, Parikh S, Schorry EK et al. The immature spine in type-1 neurofibromatosis. *J Bone Joint Surg Am*. Feb 2007;89 Suppl 1:123-142.
3. Akbarnia BA, Marks DS, Boachie-Adjei O et al. Dual growing rod technique for the treatment of progressive early-onset scoliosis: a multicenter study. *Spine (Phila Pa 1976)*. Sep 1 2005;30(17 Suppl):S46-57.
4. Durrani AA, Crawford AH, Chouhdry SN et al. Modulation of spinal deformities in patients with neurofibromatosis type 1. *Spine (Phila Pa 1976)*. Jan 2000;25(1):69-75.
5. Sirois JL, 3rd, Drennan JC. Dystrophic spinal deformity in neurofibromatosis. *J Pediatr Orthop*. Jul-Aug 1990;10(4):522-526.
6. 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*. Nov 3 2010;92(15):2533-2543.