EOS and NF
Surgical Challenges and Solutions

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NF Symposium- Solutions

- **10:40 Cervical Spine Challenges (kyphosis, spontaneous dislocation, cervicothoracic scoliosis) in NF1**

  Ilkka J. Helenius, MD, PhD

- **11:10 The Role for Anterior/Posterior vs. Posterior only Fusion for NF**

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Incidence of EOS in NF

- **NF 1:3000**
- **NF1**

  loss of neurofibromin protein

  which downregulates oncogene p21-Ras

Spine deformity in NF **8-60%**

NF in scoliosis clinic **2%**

Incidence of EOS in NF

- **BCH**
  - 102 spine operative procedures for deformity
  - 1993 to present
- **30 cases in patients under age 10**
- **Youngest 2.5**
- **# of procedure range 1- 5+**

Skeletal Manifestations of NF1

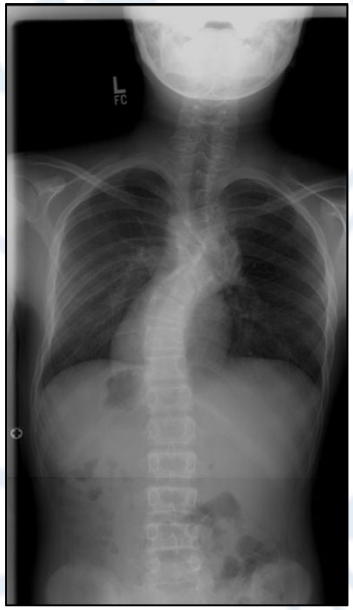
- **Generalized**
  - Low bone density -osteopenia and osteomalacia
  - Short stature
  - Macrocephaly
- **Focal skeletal conditions**
  - Spine deformity
  - Tibia and long bone osseous lesion
  - Chest wall deformity

Spine deformity In NF

- **Google Scholar "NF, Spine" 32,000**
- **Theorized**
  - Pressure erosive effect of ectasia or paraspinal neurofibroma
  - Local active biochemical interaction
- **Twin studies – discordant for dystrophy and severity**
  - Heritable factors
  - Non heritable factors

NF1 Scoliosis

- **Non dystrophic**
  - Similar to AIS



- **Dystrophic**
  - Rapid progression
  - Poor prognosis
  - May not be apparent at presentation

Durrani et al, Spine 2000

- Modulation occurred in 81% of patients scoliosis presented before 7 years and 25% after 7 years
- Rib penciling only factor influenced progression
- Progression rate: scoliosis 12˚ and kyphosis 8˚

Dystrophic Radiographic appearance

Table 2. NINE RADIOGRAPHIC CHARACTERISTICS OF DYSTROPHIC DEFORMITY IN NF1.

Characteristics	% incidence
Rib penciling	62
Vertebral rotation	51
Posterior vertebral scalloping	31
Vertebral wedging	36
Spindling of transverse processes	31
Anterior vertebral scalloping	31
Widened intervertebral foramina	29
Enlarged intervertebal foramina	25
Lateral vertebral scalloping	13

From Durrani AA, Crawford AH, Choudry SN, et al. Modulation of spinal deformities in patients with Neurofibromatosis type 1. Spine 2000:25:69-75

Reliability of Radiographic Findings

- **Ledonio et al NF study group**

- **5 expert readers**
- **122 radiographs**

Characteristic	Fleiss' kappa
Dystrophic diagnosis	0.612
Vertebral wedging	0.619 - max
Vertebral rotation	0.589
Sharp angular curve	0.602
Rib penciling	0.414
Vertebral scalloping	0.140 - min
Widened interpedicular distance	0.182
Atypical location	0.276
Spindling of transverse processes	0.424

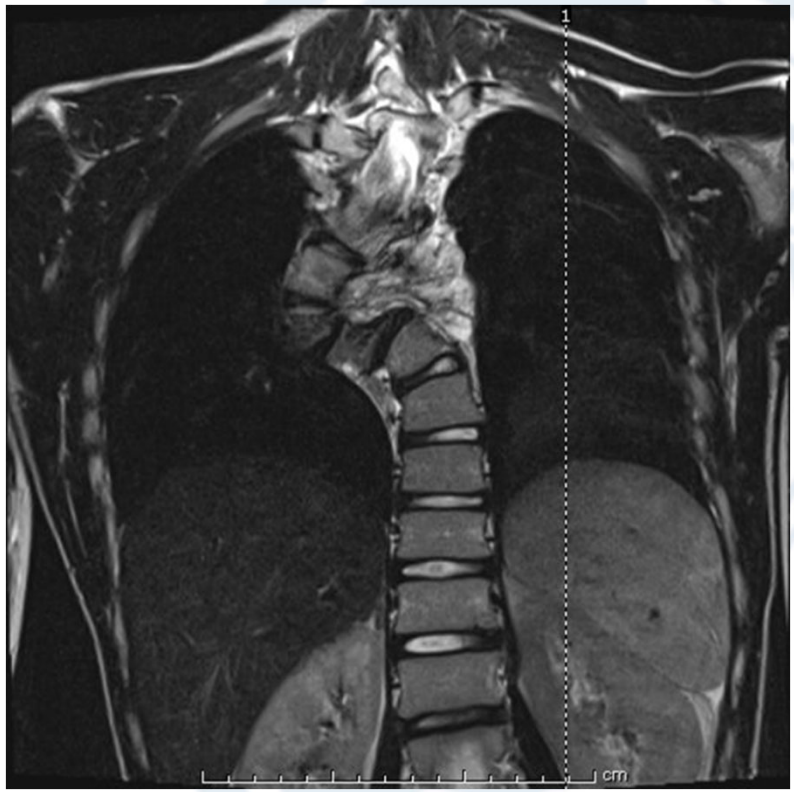
Actual Dystrophic diagnosis

Variable Name	Rate observed in all 610 readings	Rate observed in truly dystrophic (sensitivity)	Rate observed in truly non-dystrophic (1-specificity)
Vertebral wedging	61.5 %	75.9 %	30.8 %
Vertebral rotation	61.2	76.1	29.2
Sharp angular curve	52.5	65.3	25.1
Rib penciling	42.8	54.4	18.0
Vertebral scalloping	40.7	46.8	27.7
Widened interpedicular distance	36.1	43.9	19.5
Atypical location	22.3	29.6	6.7
Spindling of transverse processes	15.1	18.3	8.2

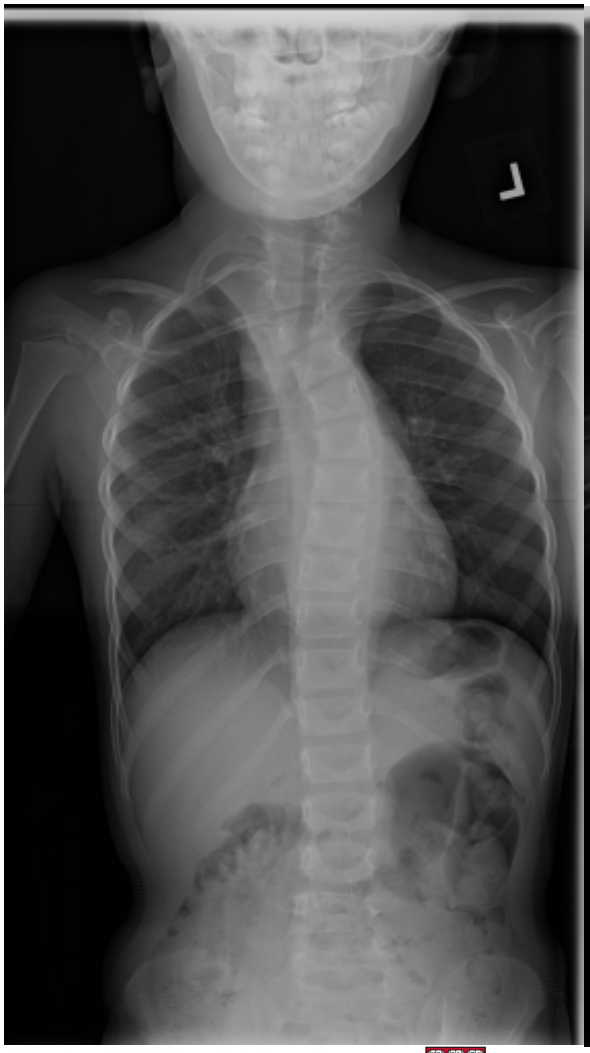
The association between each characteristic and true dystrophic diagnosis is highly significant (chi-square test, p-value < 0.0001) for seven of the eight characteristics, and slightly less significant (p-value = 0.0011) for the eighth (spind).

Non radiographic Factors

- **Age of presentation**
- **Size of curve**
- **MRI findings**
  - Dural ectasia
  - Paraspinal neurofibroma



2013 5yo



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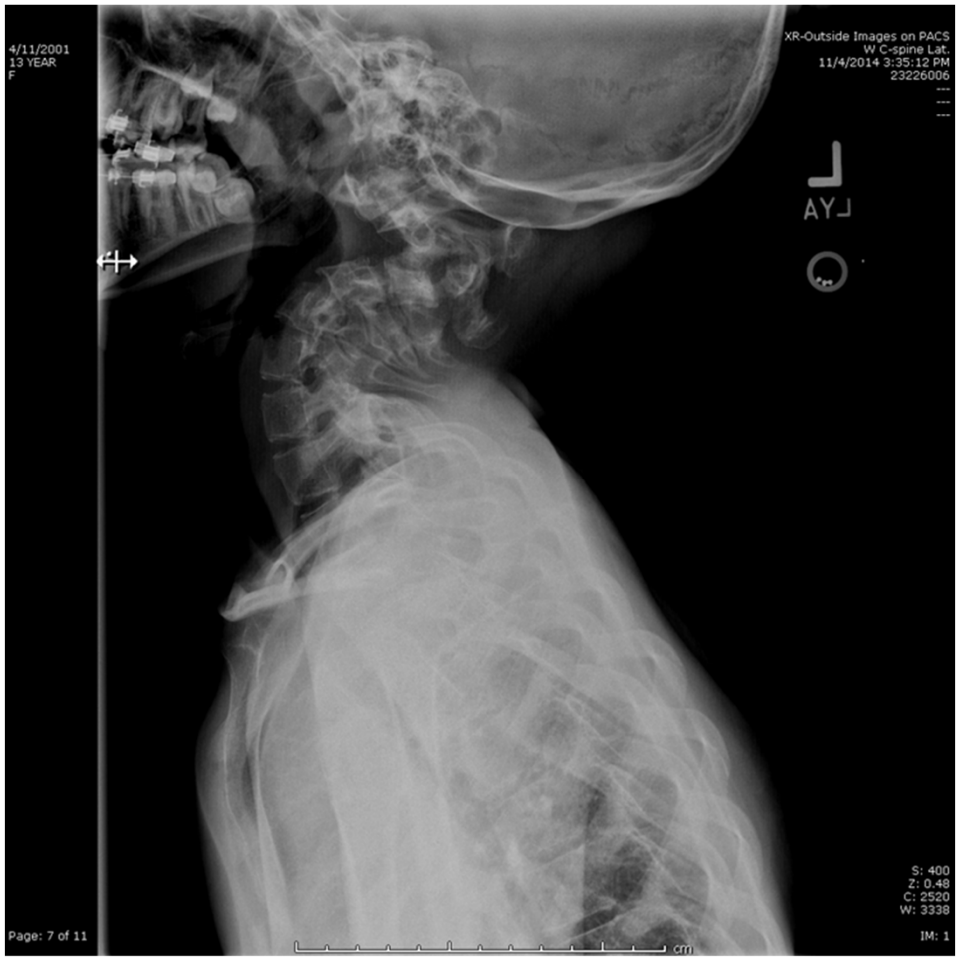
Cervical Spine



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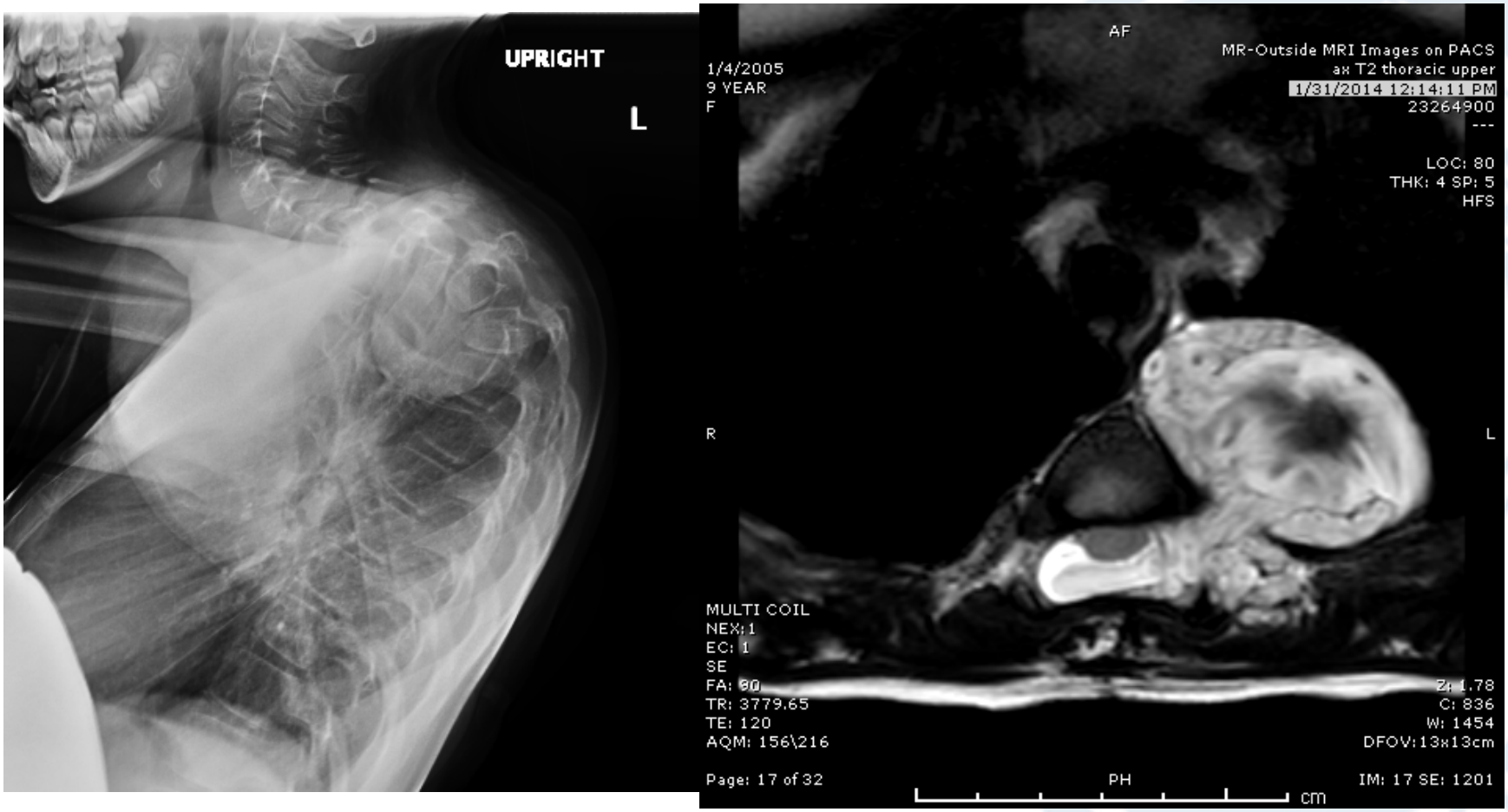
NF Post Laminectomy kyphosis



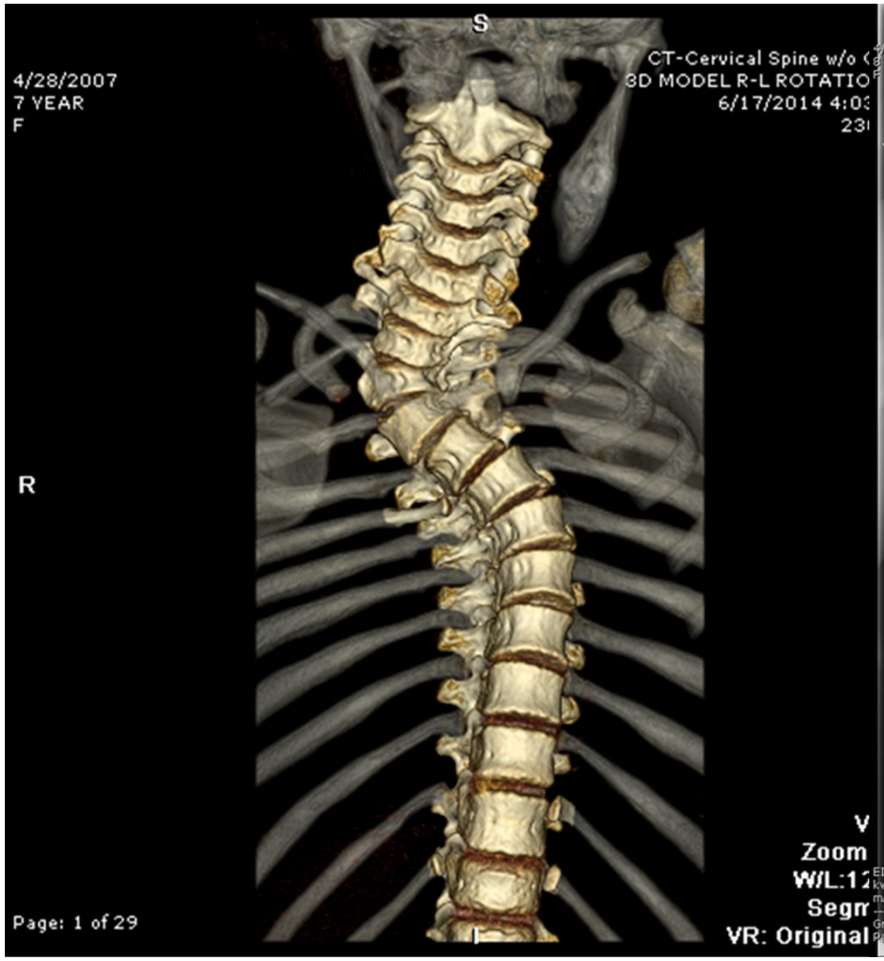
Challenges Pre Op

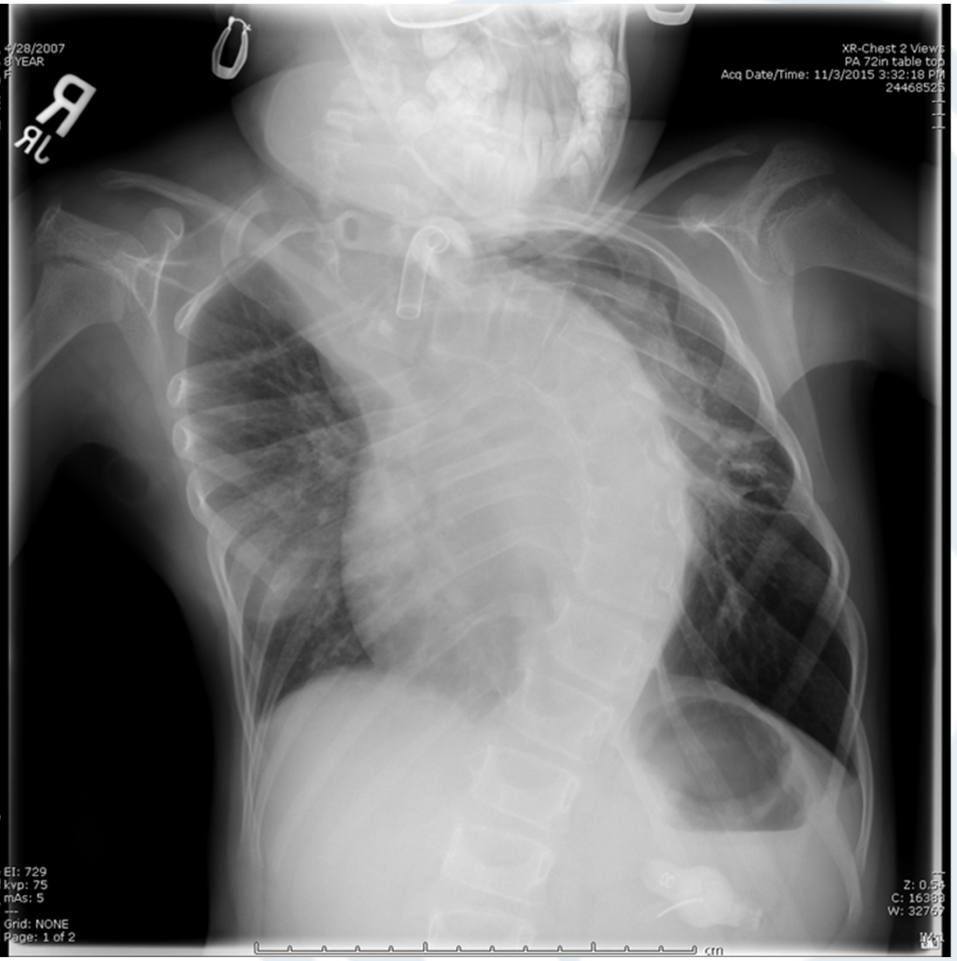
- General health
- Skin
- Need for future imaging with MRI
- Bone density
- Air way obstruction
- Neurofibroma transformation

Mediastinum/cardiac involvement

2014

2015





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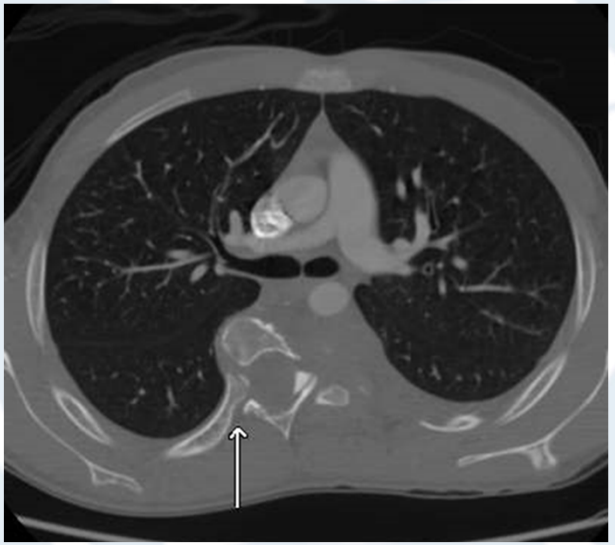
Treatment options

- **Observation**
- **Casting**
- **Bracing**
- **Traction**
- **Surgical**
  - Instrumented AP or P
  - Autogenous graft
  - ?BMP?

Traction

NF Spinal Fusion

- **High rate of non union**
  - Winter et al (1988) J Spinal Disord 1:39–49
- **Anterior posterior fusion recommended**
- **At 40 degree or more**
- **Thin /absent pedicles**
- **Dural ectasia**
- **Displace rib heads in canal**

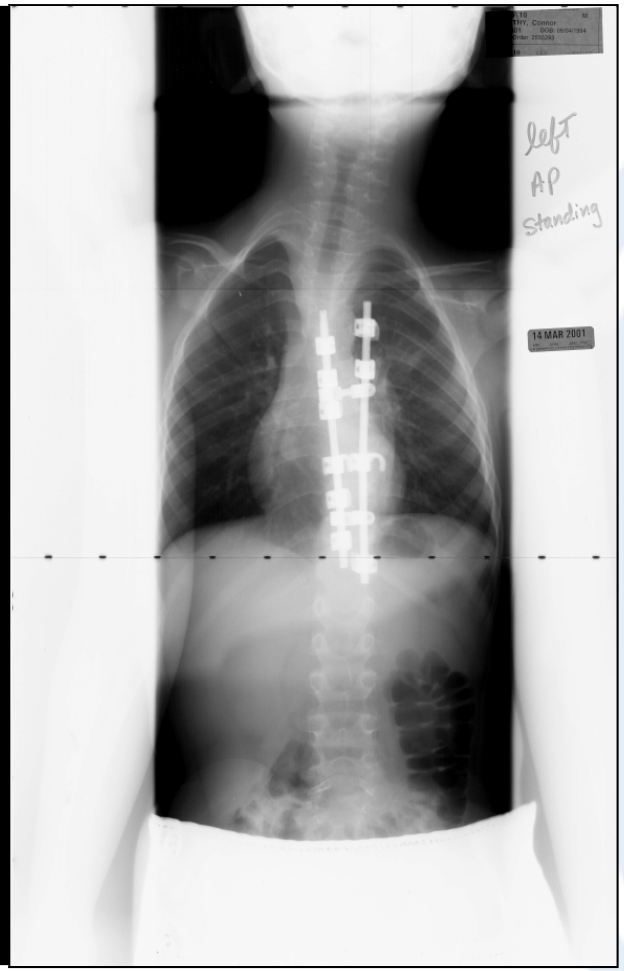


Surgical Approach

- **Scoliosis 20- 40 and Kyphosis < 50 – posterior fusion alone**
- **AP if greater**

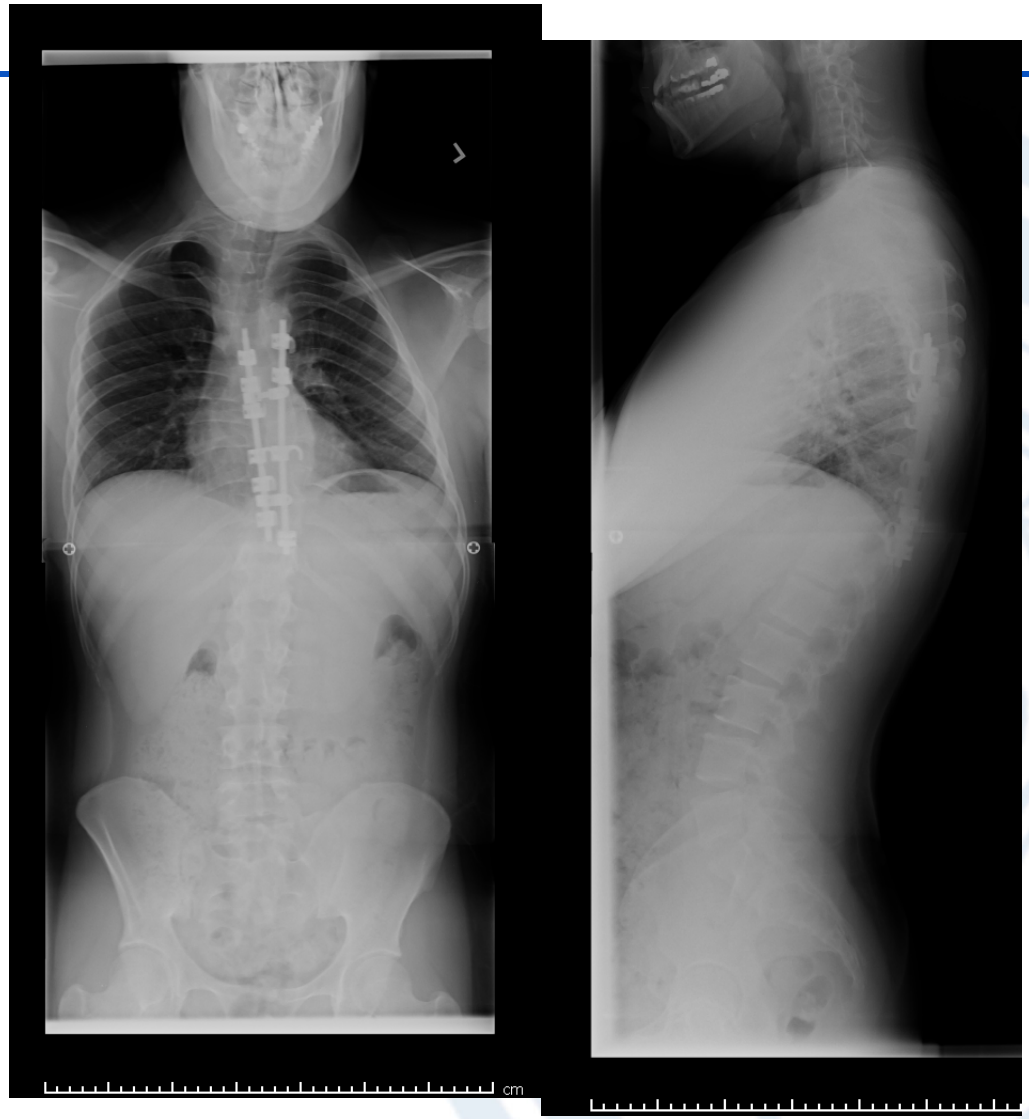
Kim HW, Weinstein SL (1997) Spine update: the management of scoliosis in neurofibromatosis. Spine 22:2770–2776

Age 6 2001 VAT+ PSF



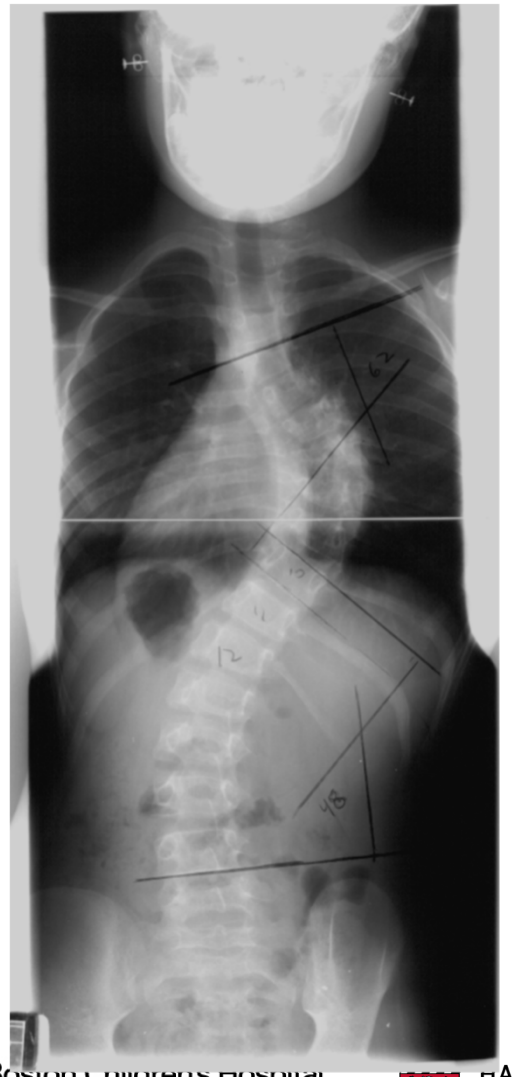
2014 age 19

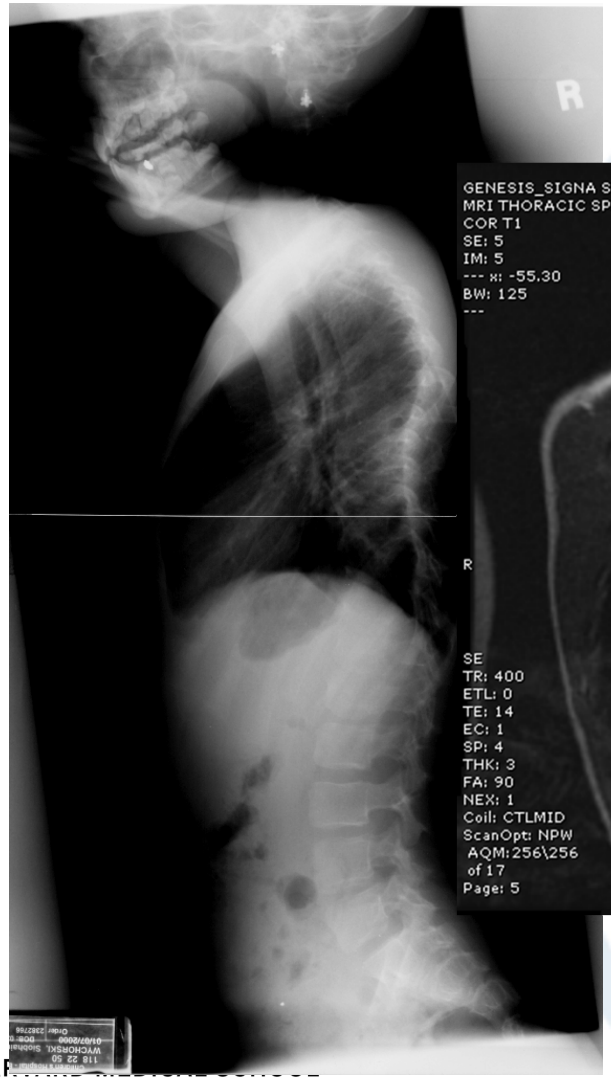
- College
- No Dyspnea
- AC pain



No further surgery!

2000 age 8





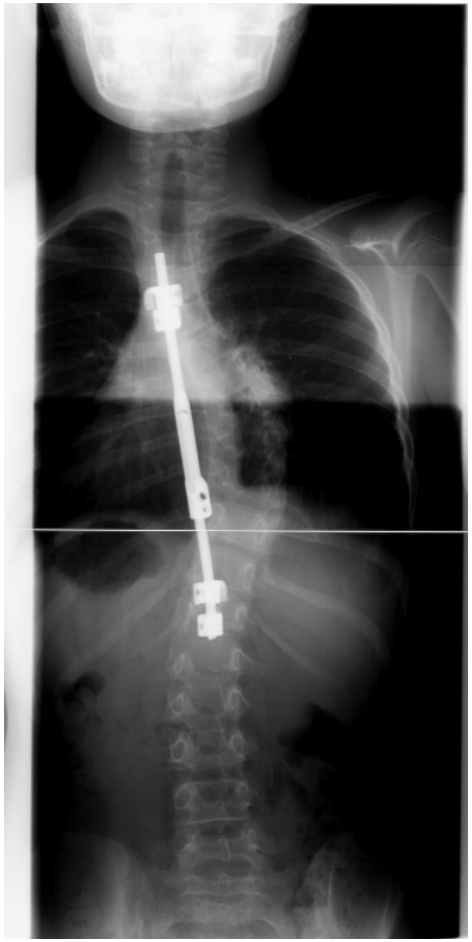

GENESIS_SIGNA SYS#MRS2OC0
MRI THORACIC SPINE WO CONTR
COR T1
SE: 5
IM: 5
--- x: -55.30
BW: 125

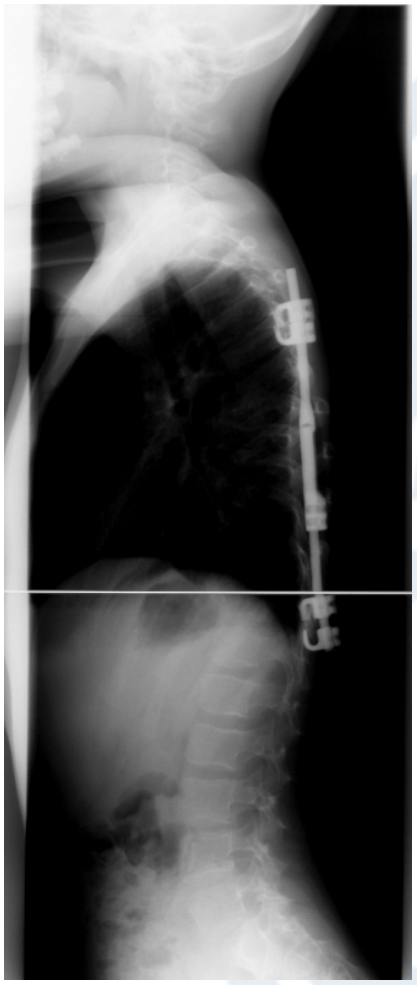
H
Acc Num: 1194075
F 7 YEAR 1182250
DOB: 2/22/1992
Acq Date/Time: 1/13/2000 8:54:24 AM

R
L
SE
TR: 400
ETL: 0
TE: 14
EC: 1
SP: 4
THK: 3
FA: 90
NEX: 1
Coil: CTLMID
ScanOpt: NPW
AQM:256\256
of 17
Page: 5

Z: 2
W: 1049 C: 469
DFOV:28x28cm
Ctr: 1200 0 0
DIM: 0 0 0
F
cm

VAT T6-9 + Growth rod T4-11

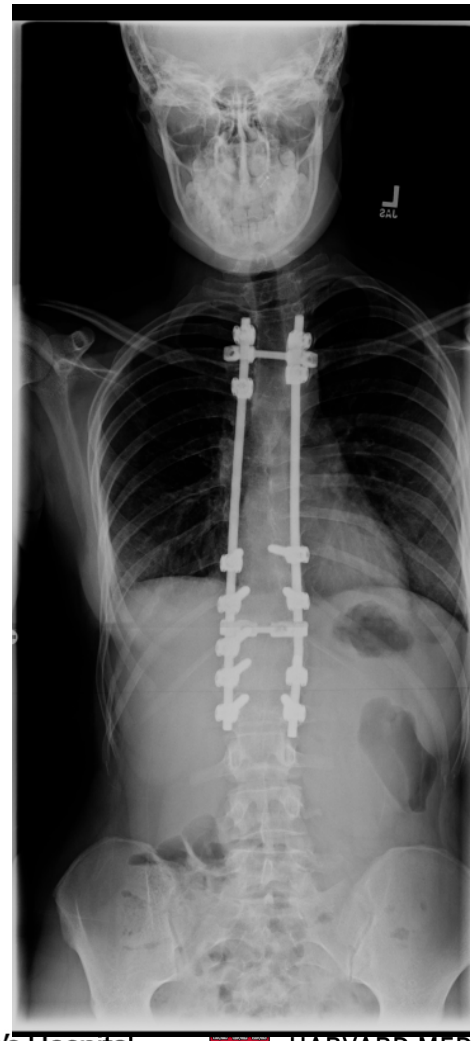




NF Scoliosis

- 2001 At 2nd lengthening – extended to L1
- 2002 Infected bursa
- 2002 Revision anterior and posterior fusion
- 2004 removal painful implants
- 2005 Revision AP fusion

2008 16 yo



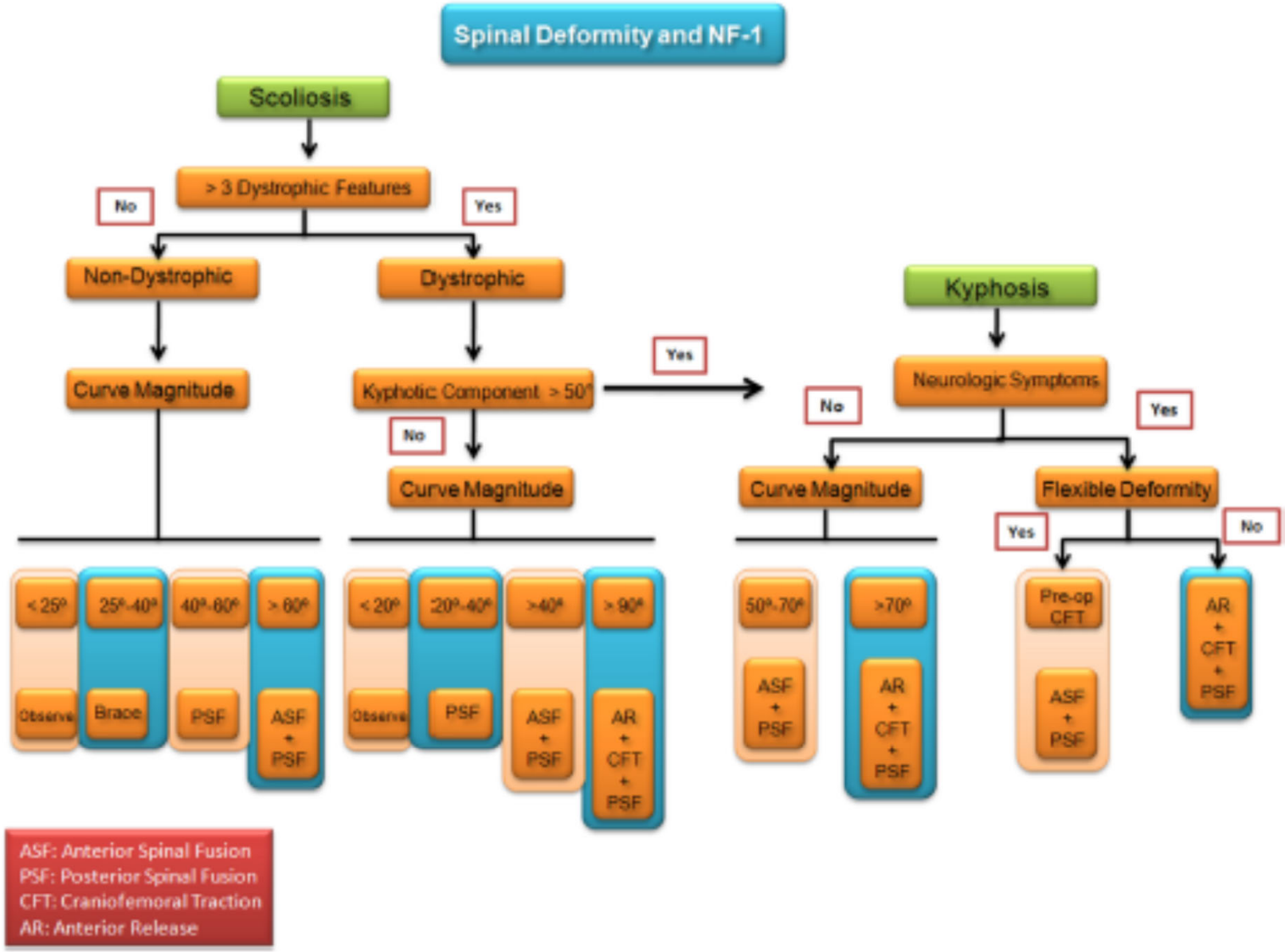


Fig. 6. Treatment algorithm in patients with neurofibromatosis 1 (NF-1) and spinal deformity.

Levy et al. Scoliosis (2015) 10:14
DOI 10.1186/s13013-015-0035-x

Scoliosis

REVIEW Open Access

Complications associated with surgical repair of syndromic scoliosis

Benjamin J Levy[1], Jacob F Schulz[1*], Eric D Fornari[1] and Adam L Wollowick[2]

- **Dural tear** **5.9%**
- **Non union** **5.7%**
- **Infection** **10.3%**
- **Implant failure** **22%**
- **Neuro compromise** **5.7%**

Use of Rib-Based Distraction in the Treatment of Early-Onset Scoliosis Associated With Neurofibromatosis Type 1 in the Young Child

John A. Heflin, MD[a,*], Andrew Cleveland, MD[b], Scott D. Ford, PA-C[a], Jessica V. Morgan, BS[a], John T. Smith, MD[a]

- **12 patients**
  - mean age 6
  - Follow 5 yr
- **Maintained or improved curve**
- **6 device migration**

- **High thoracic curve**
- **Poor pedicle anatomy**

– Spine Deformity 3(2015)

NF 1 Challenges

- Recognition of Dystrophic characteristics
- When to perform early growth friendly instrumentation or AP fusion?
- Spine or rib based?
- Use of BMP?
- Do I need to worry about ectasia?
- Many more….

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Thank You!

- **Timothy.Hresko@childrens.harvard.edu**