Neurofibromatosis type 1 and Dystrophic Scoliosis: A Multicenter Study of Accuracy of Surgeons' Radiographic Assessment

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Disclosures

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Scoliosis in Neurofibromatosis type1: Dystrophic or non-dystrophic

- Nondystrophic and dystrophic
- Most common osseous defect
- 2% of pts with scoliosis will have NF-1
- 30% of patients with NF-1 have spine disorders
- Dystrophic more severe

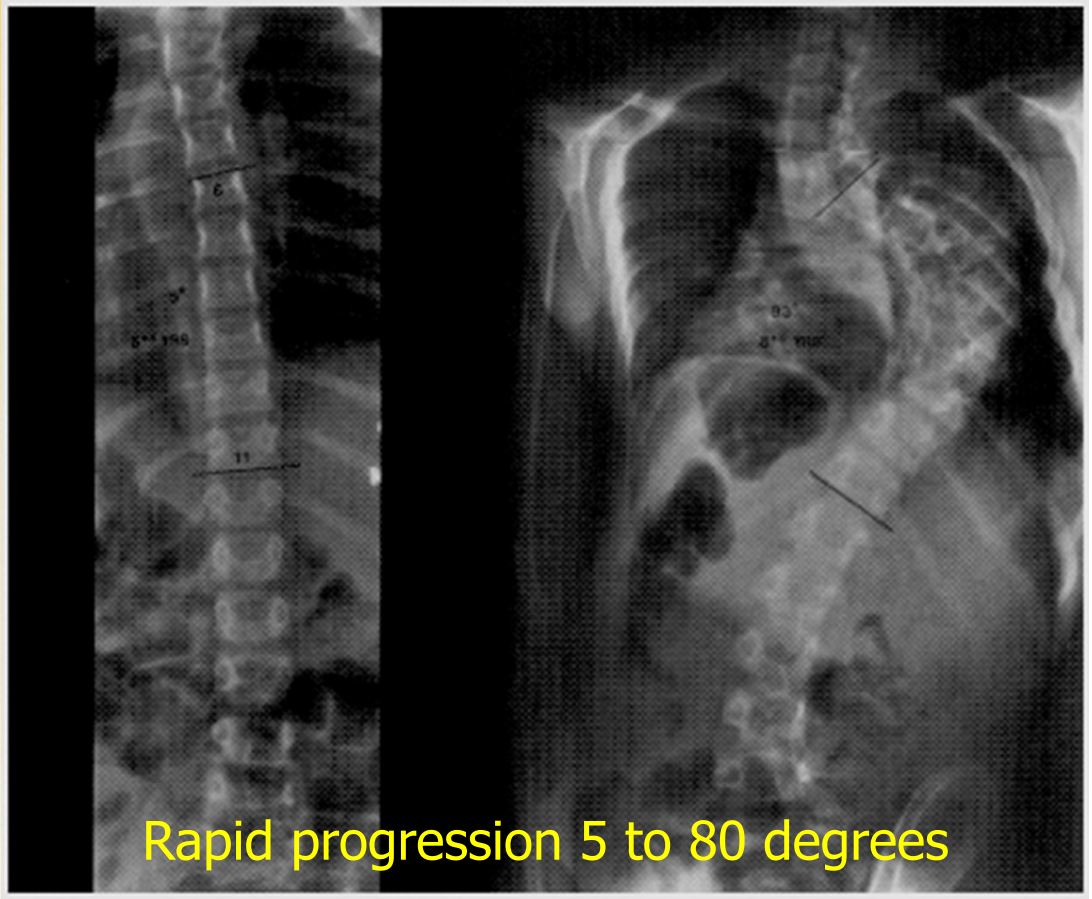
Crawford OCNA 2007

Table 1. Prevalence of scoliosis in NF1

	Scoliosis Prevalence	Dystrophic	Non-Dystrophic
McCarroll (1950)	41%		
Hunt & Pugh (1961)	14.6%	30%	5.0 %
Lewis & Pallios (1963)	39%		
Scott (1965)	12-20%		
Chaglessian (1976)	26%		
Held (1979)	36%		
Crawford (1986)	64%		
DeSimone (1988)	53%		
Sirais & Brennan (1990)	69%		
Akbarnia (1992)	10%	61%	39%

Natural History

- *Calvert et al, JBJS Br 1989*
  - Treated (n=34) and untreated (n=32) w/ NF1 scoliosis
  - 75% untreated group had kyphoscoliosis
  - Severe anterior scalloping – progressed 23˚ /yr
  - All others 7˚ /yr progression and 8˚ /yr of kyphosis
- *Wilde et al, Spine 1994*
  - Vertebral subluxation, disc wedging and peripheral skeletal dystrophy prognostic factors that predict progression after arthrodesis

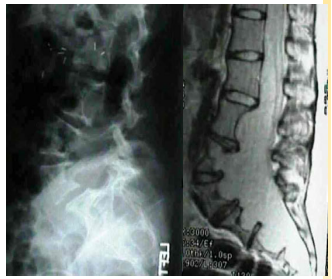


Radiographic characteristics of dystrophic scoliosis

- Certain radiographic characteristics have been reported to predict dystrophic scoliosis, but their predictive value is not well described.

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

Objective

This study aims to determine the accuracy of radiographic assessment of dystrophic modulation in NF1 patients with scoliosis.

Materials and Methods

- Multicenter contribution
- 122 sets (AP & Lat) of patient radiographs with NF1 & scoliosis assessed by 5 Spine surgeons
- 8 Radiographic characteristics dystrophic scoliosis
- Blinded to final diagnosis

- Sensitivity and specificity were calculated

- The association between each characteristic and dystrophic scoliosis was tested using chi-square and quantified as a relative risk (RR).

- **Vertebral wedging**
- **Vertebral rotation**
- **Sharp angular curve**
- **Rib penciling**
- **Vertebral scalloping**
- **Widened interpedicular distance**
- **Atypical location**
- **Spindling of transverse processes**

- ***The 'gold standard' clinical diagnosis for each x-ray, made by the patient's surgeon based on clinical data***
- ***Combination of Hx, PE, MRI and CT scans, surgical observations and results.***

Results

- The *actual* diagnosis was dystrophic for 83 of the 122 x-rays, or 68% and 39(32%) were non-dystrophic
- Readers underestimated the proportions that were dystrophic.

Reader	Frequency Non-dystrophic (percent)	Frequency Dystrophic (percent)
1	47 (39%)	75 (61%)
2	45 (37%)	77 (63%)
3	40 (33%)	82 (67%)
4	48 (40%)	74 (60%)
5	67 (55%)	55 (45%)
Total	247 (41%)	363 (59%)

Results

- Dystrophic scoliosis: Sensitivity of 75% (310/415 reads)
- Non-dystrophic: Specificity of 73%(142/195 reads).
- Positive predictive value = 85%
- Negative predictive value = 57%.

Reader	Sensitivity	Specificity
OVERALL	*74.7 %*	*72.8 %*
1	77.1	71.8
2	77.1	66.7
3	83.1	66.7
4	74.7	69.2
5	61.5	89.7

Results

- All 8 characteristics are strongly associated with dystrophic scoliosis ($p<0.002$).
- The association is strongest for atypical location (RR=4.45) and weakest, (still significant) for scalloping (RR=1.69).

Characteristic	Sensitivity	Specificity	Relative Risk* (95% CI)
Vertebral rotation	76.1 %	70.8 %	2.60 (2.08 – 3.26)
Vertebral wedging	75.9	69.2	2.47 (1.98 – 3.07)
Sharp angular curve	65.3	74.9	2.60 (2.02 – 3.34)
Rib penciling	54.4	82.0	3.03 (2.22 – 4.15)
Vertebral scalloping	46.8	72.3	1.69 (1.32 – 2.17)
Widened interpedicular distance	43.9	80.5	2.25 (1.66 – 3.05)
Atypical location	29.6	93.3	4.45 (2.58 – 7.67)
Spindling of transverse processes	18.3	91.8	2.23 (1.34 – 3.72)

*Risk of a rater seeing the indicated characteristic in dystrophic x-rays vs. in non-dystrophic x-rays.

Discussion: Dystrophic Modulation

- *Durrani et al, Spine 2000*
  - Modulation occurred 65% of patients
  - 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 modulation may explain underestimation of dystrophic diagnosis by 5 raters.

Summary

- The 8 radiographic characteristics were significantly associated with dystrophic modulation in NF1 patients with scoliosis.
- Wedging and rotation were most sensitive, atypical location and transverse processes spindling were most specific.
- On balance, atypical location and rib penciling had the strongest association with dystrophic scoliosis.

Thank you

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