

Scoliosis in Fibrous Dysplasia/McCune-Albright Syndrome

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Polyostotic Fibrous Dysplasia

- ▶ Wide range of clinical severity
- ▶ Disease proliferation during childhood
- ▶ Frequent fractures
- ▶ Bone pain
- ▶ Extremity deformity
- ▶ Associated endocrine involvement

Scoliosis

Often not appreciated if the whole patient is not examined carefully

The extremity deformities are obvious and often symptomatic

Scoliosis is not usually symptomatic in early cases.

Scoliosis

- ▶ Requires serial radiographs over time to determine progression
- ▶ Mild: 10 to 30 degrees
- ▶ Moderate: 30 to 45 degrees
- ▶ Severe: over 45 degrees

National Institutes of Health Bethesda, Maryland, USA

- ▶ Subset of ongoing clinical cohort study
- ▶ January 2018
- ▶ Journal of Bone and Mineral Research
- ▶ Allison Boyce, et al
- ▶ 138 patients

Scoliosis

- ▶ 61% had scoliosis
- ▶ 55 (65%) Mild (10 to 30 degrees)
- ▶ 11 (13%) Moderate (30 to 45 degrees)
- ▶ 18 (22%) Severe (over 45 degrees)

Scoliosis Surgery

- ▶ 12 had surgery, but only 10 had follow up
- ▶ One instrument failure
- ▶ Nine had stable fusion at follow up (average 6.1 years)
- ▶ Two Untreated patients had death as a result of restrictive lung disease.

Scoliosis Progression

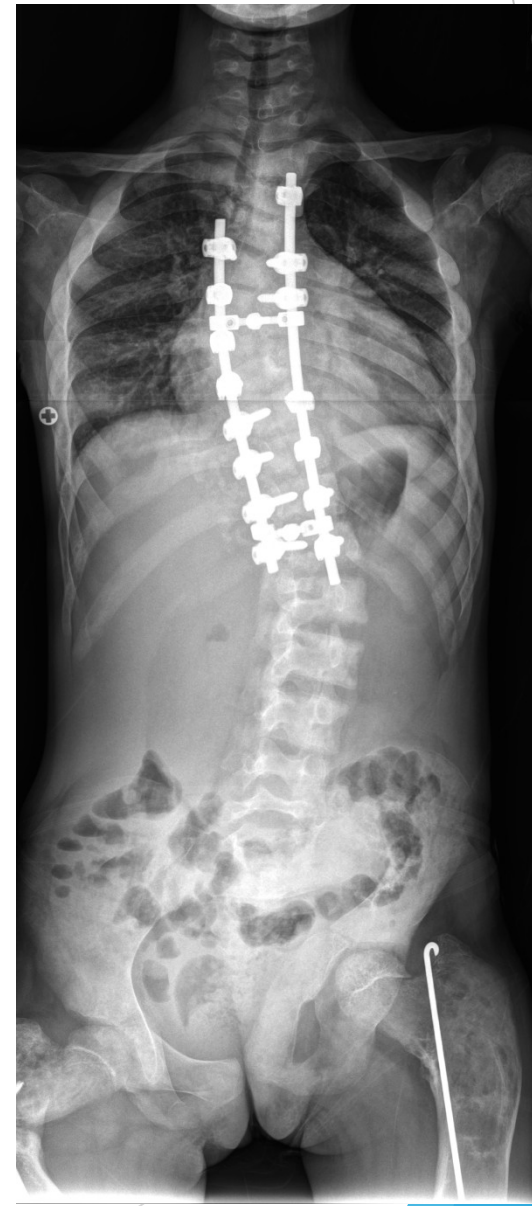
- ▶ Associated with:
 - ▶ 1. Hypothyroidism
 - ▶ 2. FGF-23 mediated Hypophosphatemia
 - ▶ 3. leg length discrepancy
 - ▶ 4. Total body disease burden

Scoliosis Progression

- ▶ Not affected by Bisphosphonate administration



October 2009 (age 9)



Scoliosis in Fibrous Dysplasia

- ▶ Look and you will find
- ▶ More common than appreciated
- ▶ X-rays are required
- ▶ Serial follow up especially in growing children
- ▶ Conventional surgery is effective in stabilizing the spine.