

# Rib head dislocation causing spinal canal stenosis in a child with neurofibromatosis, type 1

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Radiology Case. 2017 Aug; 11(8):8-15 :: DOI: 10.3941/jrcr.v11i8.3113

## ABSTRACT

A 10-year-old female with neurofibromatosis type 1 and severe dystrophic scoliosis presented with a two-month history of difficulty ambulating due to low back pain. The patient did not have any neurological symptoms. MRI of the thoracolumbar spine demonstrated subluxation of the right posterior tenth and eleventh ribs through their respective neural foramina, with mild mass effect on the thecal sac without abnormal cord signal or cord compression. Groups of neurofibromas were present along the right ribs and paravertebral soft tissues around these levels. CT evaluation was completed for greater osseous definition. Cases of rib head dislocation into the central canal in the setting of dystrophic scoliosis were documented in only a limited sample of case reports. The angulated short-segment curvature in dystrophic scoliosis causes vertebral body rotation, foraminal enlargement, spindling of transverse processes, and penciling of the apical ribs. These changes can alter the articulation of the rib along the transverse process. The enlarged foramina can also create a larger space into which a rib may displace. As a result, in most reported cases, the subluxed ribs were on the convex apex of the curve in the mid-to-lower thoracic region. The risk of cord injury from rib head dislocation makes the complete depiction of the anatomy essential for proper surgical management.

## CASE REPORT

### CASE REPORT

#### Clinical History:

A 10-year-old female with neurofibromatosis, type 1 (NF-1) and severe dystrophic scoliosis presented with progressive difficulty to ambulate due to lower back pain for two months. There were no reported neurological symptoms, such as numbness or incontinence.

#### Imaging findings:

Initial radiograph of the spine demonstrated the patient's severe dextroscoliotic curve with a Cobb angle measurement of 81 degrees measured from the superior endplate of T7 to the inferior endplate of T11 (Figure 1). Cross-sectional imaging workup began noncontrast magnetic resonance imaging (MRI) of the spine, which most notably demonstrated dislocated right posterior tenth and eleventh ribs through their corresponding neuroforamina. The protruded ribs were not causing cord compression but were indenting the

thecal sac (Figure 2). Paraspinal neurofibromas and dural ectasia were appreciated at multiple levels (Figure 3). The MRI also depicted the extent of plexiform neurofibromas along the contour of the posterior right 9<sup>th</sup> through 12<sup>th</sup> ribs (Figure 4).

Further evaluation with a noncontrast computed tomography (CT) scan better demonstrated the short-segmented, sharply angulated kyphoscoliotic curve (Figure 5). The invasive right tenth and eleventh ribs were again visualized (Figure 6). Multilevel vertebral body wedging and scalloping was found (Figure 7), as well as the widening of the intervertebral foramina (Figure 8), primarily on the convex sides of the curve. After the discovery of these findings, the decision was made to move forward with surgical management.

#### Management and Follow-up:

Complex posterior spinal fusion and segmental spinal instrumentation were planned. The surgery involved a T10-T11 thoracoplasty with resection of the 10<sup>th</sup> and 11<sup>th</sup> right rib. Posterior column wedge osteotomy, also known as Ponte osteotomy, was performed to help correct the thoracic kyphosis. A Ponte osteotomy involves resection of the inferior articular processes of the cephalad vertebra, the superior articular process of the caudal vertebra, and the ligament flavum. The purpose of the osteotomy was to allow gradual compression of the posterior elements to correct the kyphosis. The patient was noted to have excellent outcomes without neurologic symptoms and reduced pain after correction.

## DISCUSSION

#### Etiology & Demographics:

NF-1, also known as von Recklinghausen disease, is typically an autosomal dominant inherited disorder involving the mutation of the neurofibromin gene on chromosome 17; however, sporadic mutations have been reported in up to 50% of cases. NF-1 is the most common neurocutaneous disorder, which involves all three germ cell layers, and thus can affect any organ system. Classically, NF-1 presents with cutaneous café-au-lait spots, skeletal deformities (pseudoarthrosis, hypoplasia of sphenoid wing, and severe kyphoscoliosis), and cutaneous and plexiform neurofibromas. Other common associated findings include optic gliomas, Lisch nodules (pigmented iris hamartomas), and axillary/inguinal freckling. The collection of clinical criteria is used to make the diagnosis (1-4).

The most common spinal deformity is kyphoscoliosis. Hypoplasia of the pedicles, transverse processes, and spinous processes can also be seen affecting the osseous spine in NF-1. Neurofibromas can be seen along the paraspinal and presacral soft tissues, as well as the spinal cord. Dumbbell-shaped enlargement of the neural foramina is common. Lateral and anterior intrathoracic meningoceles can also be seen (1-11).

#### Clinical & Imaging findings:

A dystrophic scoliotic curve, which can be seen in NF-1, is characterized by a short-segmented and sharply angulated curve. Abnormal development of the vertebral bodies in NF-1 may result in vertebral bodies becoming wedged and rotated. Vertebral body scalloping is a result of dural ectasia from weakened meninges, which allows the transmission of normal cerebrospinal fluid pulsations. Widening of the intervertebral foramina is often seen, especially in the convex parts of the curve(2,6,7,10-12).

Limited case reports have documented rib head dislocation as a complication of dystrophic scoliosis. A literature review by Ton et. al. cited 21 reported cases (9). The affected rib heads tended to be on the convex side of the curve where the neuroforamina were large. Most patients were asymptomatic, which may underestimate incidence. Symptoms included back pain, mild sensory or motor deficits, paraplegia, and paraparesis (9). Cases were typically found in the teenage years with no gender predilection (2,9). Imaging findings can be subtle and require close attention to make the diagnosis.

No single mechanism has been linked to an increased propensity for rib head dislocation. It has been postulated that this involves entities that alter the articulation of the rib, including vertebral body rotation, neuroforaminal enlargement, spindling of the transverse processes, and penciling of the apical ribs. The enlarged foramina also create a larger space into which a rib may displace (9). In a study by Sun et. al., they cited an average Cobb angle of 76.6 degrees and average kyphosis of 58.8 degrees in their sample of six patients (8).

Spine radiography is the usual first test of choice to evaluate the scoliotic curve with Cobb angles primarily. Further imaging with CT will allow for the full three-dimensional evaluation of the osseous spine and ribs. MRI is a useful adjunct study especially for evaluating the cord, prevertebral, and paraspinal soft tissues (2,8).

#### Treatment & Prognosis:

No clear consensus has been made regarding surgical management. Thus, it is rendered on a case-by-case basis. With the review of previous cases, rib head excision has been consistently favored in the surgical management, since it was first thought that the rib head could disturb the cord in the process of correcting the kyphoscoliosis. There have been case reports of postoperative rib head dislocation in NF-1 patients who have developed paraparesis after posterior spinal fusion without prior recognition. Cord compression has been documented both before and after spinal instrumentation<sup>2</sup>. Sun et. al. suggests rib head reduction without rib head resection as a good alternative to avoid the risk of impaling the spinal cord or tearing the dural sac (8). Regardless, the prognosis appears good with proper surgical management, but further investigation is needed. Regardless, recognition of this finding is necessary to assist the surgical team in preoperative planning.

Differential Diagnosis:

Differential considerations are limited. Fracture-dislocation in the rib or thoracic spine can be considered in the setting of trauma with imaging findings of fractures, surrounding hemorrhage, and edema. Trauma could also inherently increase the risk of cord compression from rib head dislocation in patients with dystrophic scoliosis (8) (Table 2).

## TEACHING POINT

Rib head dislocation in patients with dystrophic scoliosis, especially in neurofibromatosis, type 1, is usually asymptomatic. However, it is important to recognize this entity during imaging evaluation, as it is a major factor in preoperative planning.

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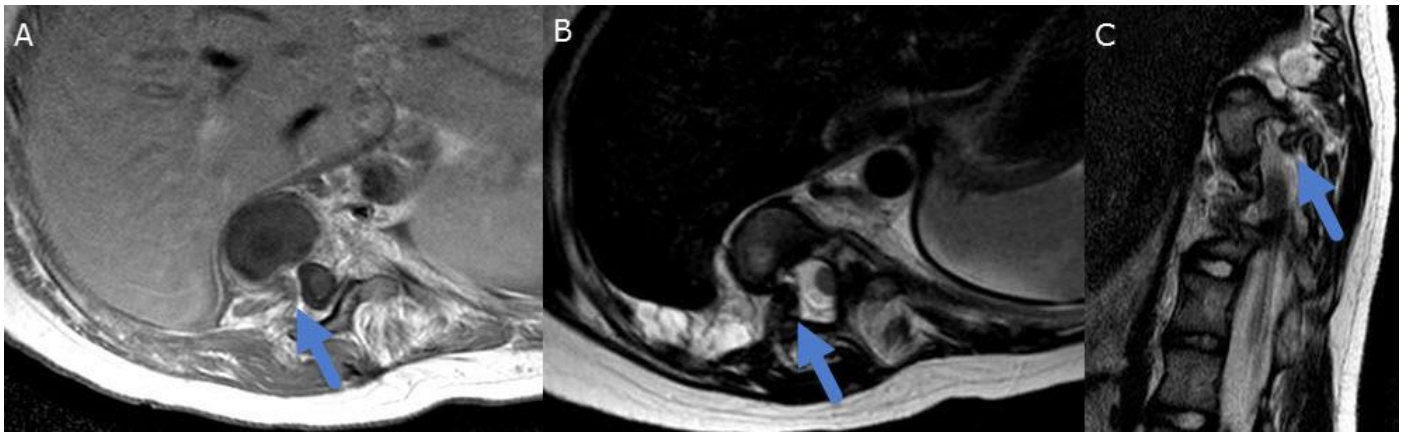
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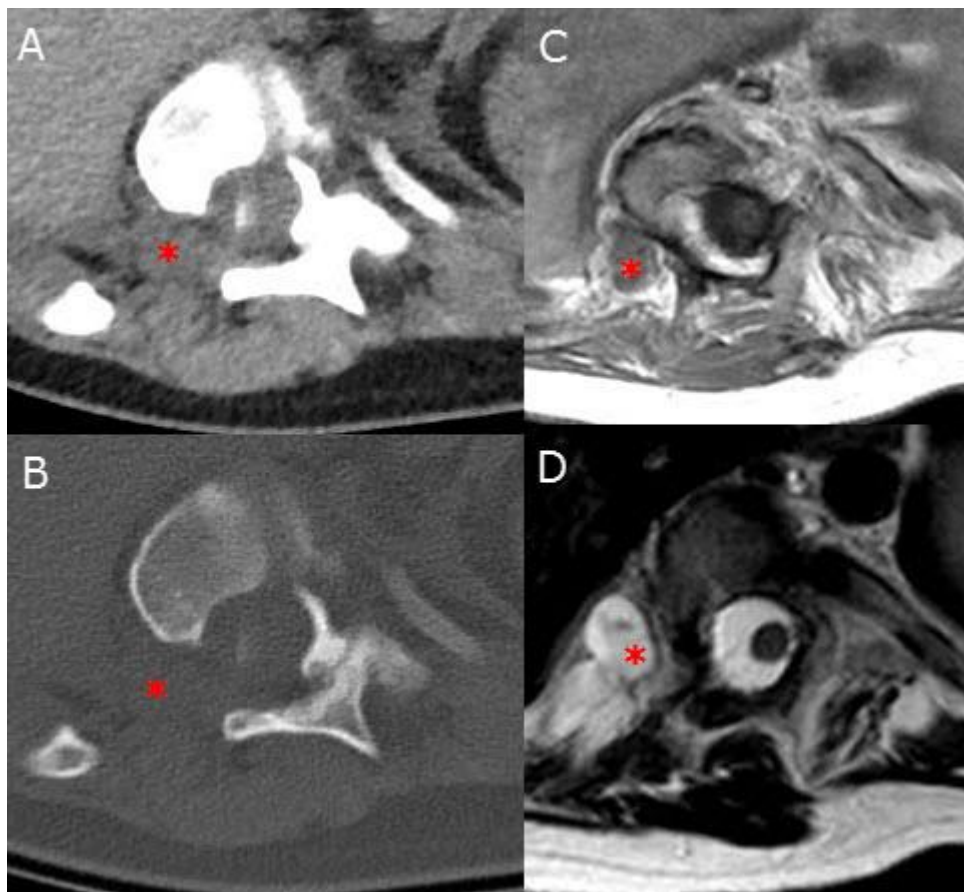
## FIGURES



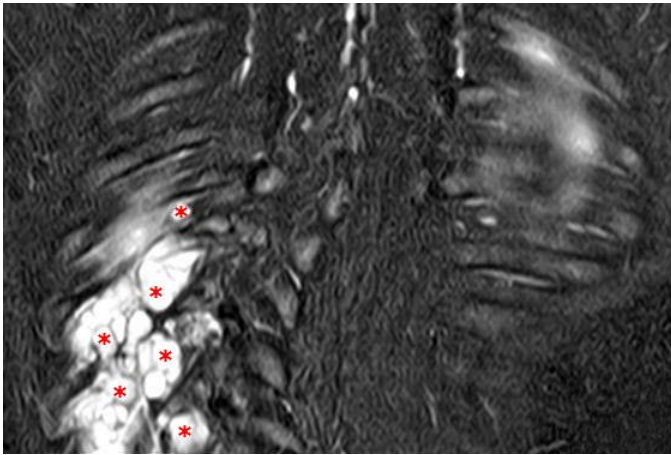
**Figure 1:** Preoperative radiographs. 10-year-old female with rib head dislocation from dystrophic scoliosis. FINDINGS: Severe dextroscoliotic curvature with a Cobb angle of 81 degrees (green lines) measured from the superior endplate of T7 to the inferior endplate of T11. TECHNIQUE: Scoliosis radiograph, AP view (70 mAs, 85 kVp)



**Figure 2:** Rib head dislocation on MRI. 10-year-old female with rib head dislocation from dystrophic scoliosis.  
**FINDINGS:** (A-C) Right tenth rib head indents the thecal sac (blue arrow). There is no abnormal cord signal or cord compression.  
**TECHNIQUE:** 1.5 T MRI. A) Postcontrast axial T1 (1.5 T, ET 3, TR 550 ms, TE 8.0 ms, Slice thickness 6.0 mm, 5 mL Multihance). B) Axial T2 (1.5 T, ET 30, TR 4200 ms, TE 120 ms, Slice thickness 7.0 mm). C) Sagittal T2 (ET: 36, TR: 3400.0, TE: 120.0, 3.0 thk/1.0 sp)



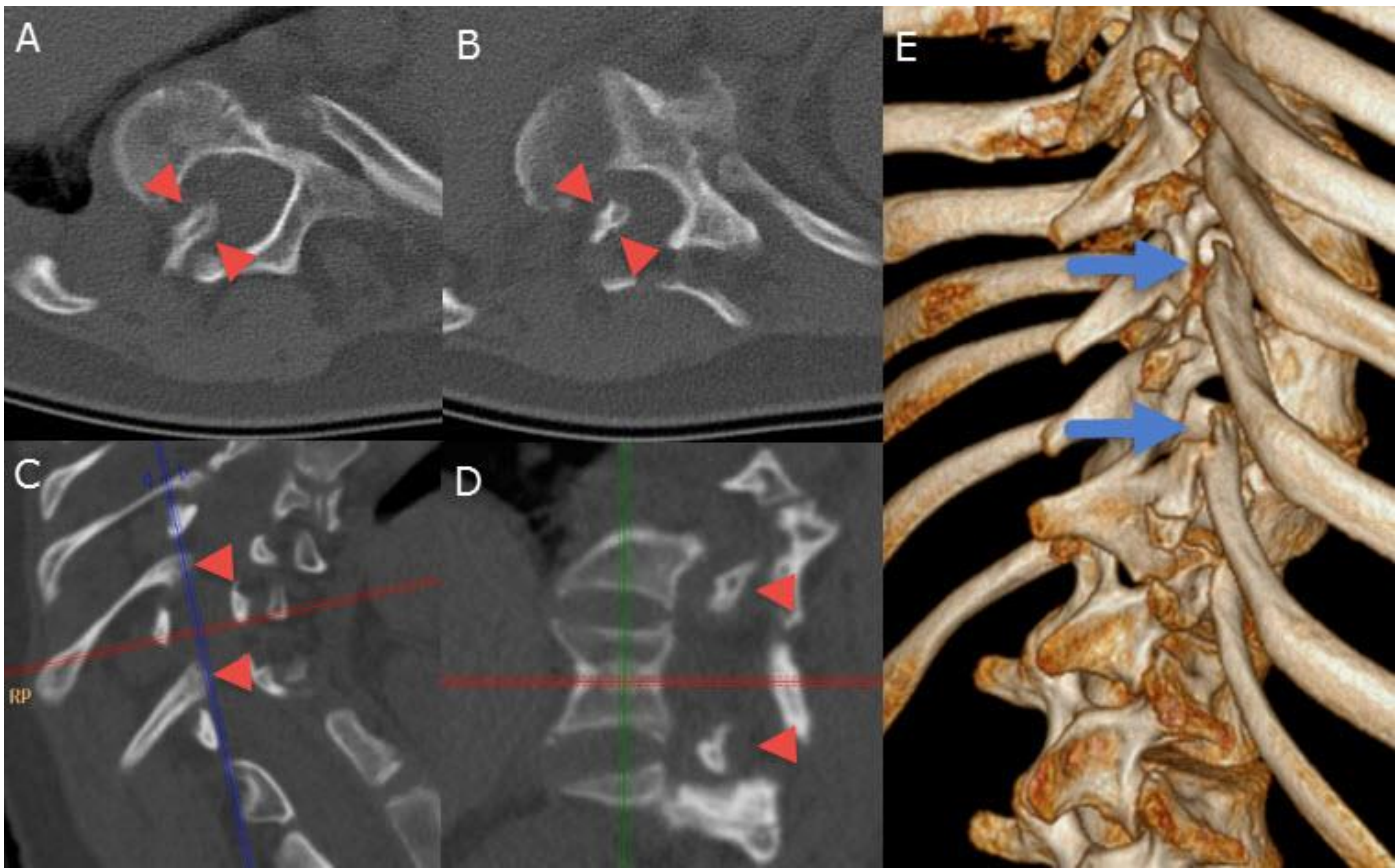
**Figure 3:** Paraspinal neurofibroma and dural ectasia. 10-year-old female with rib head dislocation from dystrophic scoliosis.  
**FINDINGS:** (A, B) Iso-attenuating paraspinal soft tissue lesion representing a neurofibroma (red star). (C, D) Paraspinal soft tissue lesion with decreased signal on T1 and increased signal on T2. Dural ectasia was also found with the cord displaced to the left due to severe scoliosis.  
**TECHNIQUE:** A) Noncontrast axial CT (100 mAs, 120 kVp, Slice thickness: 2.5 mm), bone windows. B) Noncontrast axial CT, soft tissue windows. C) Postcontrast axial T1 (1.5 T, ET 3, TR 550 ms, TE 8.0 ms, Slice thickness 6.0 mm). D) Axial T2 (1.5 T, ET 30, TR 4200 ms, TE 120 ms, Slice thickness 7.0 mm)



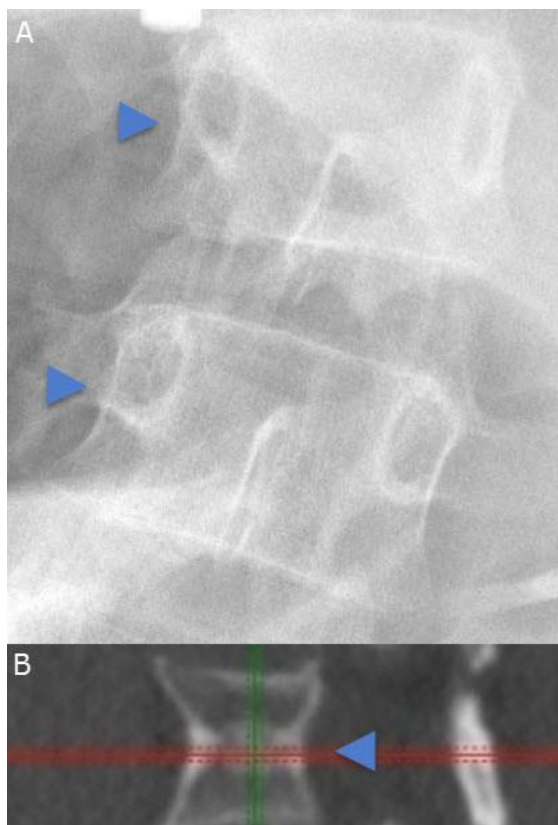
**Figure 4:** Plexiform neurofibromas. 10-year-old female with rib head dislocation from dystrophic scoliosis. FINDINGS: Multiple neurofibromas arranged along the contour of the right 9th through 12th ribs (red stars). TECHNIQUE: Coronal STIR (1.5 T, TI 180 ms, TR 2396.4 ms, TE 60 ms, Slice thickness 4.0 mm).



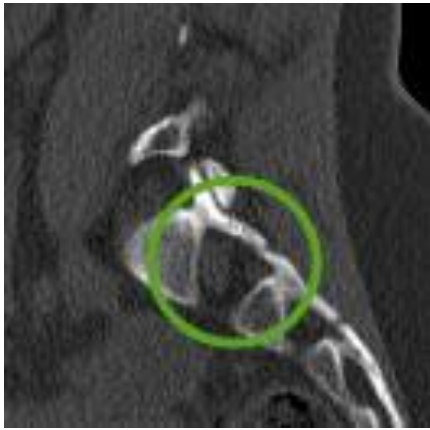
**Figure 5 (right):** Kyphoscoliosis in the lower thoracic and lumbar spine. 10-year-old female with rib head dislocation from dystrophic scoliosis. FINDINGS: 3-D reconstructed CT image of the thoracolumbar spine demonstrates a severe S-shaped scoliotic curve in the lower thoracic and lumbar spine. TECHNIQUE: Noncontrast axial CT (100 mAs, 120 kVp, Slice thickness: 2.5 mm).



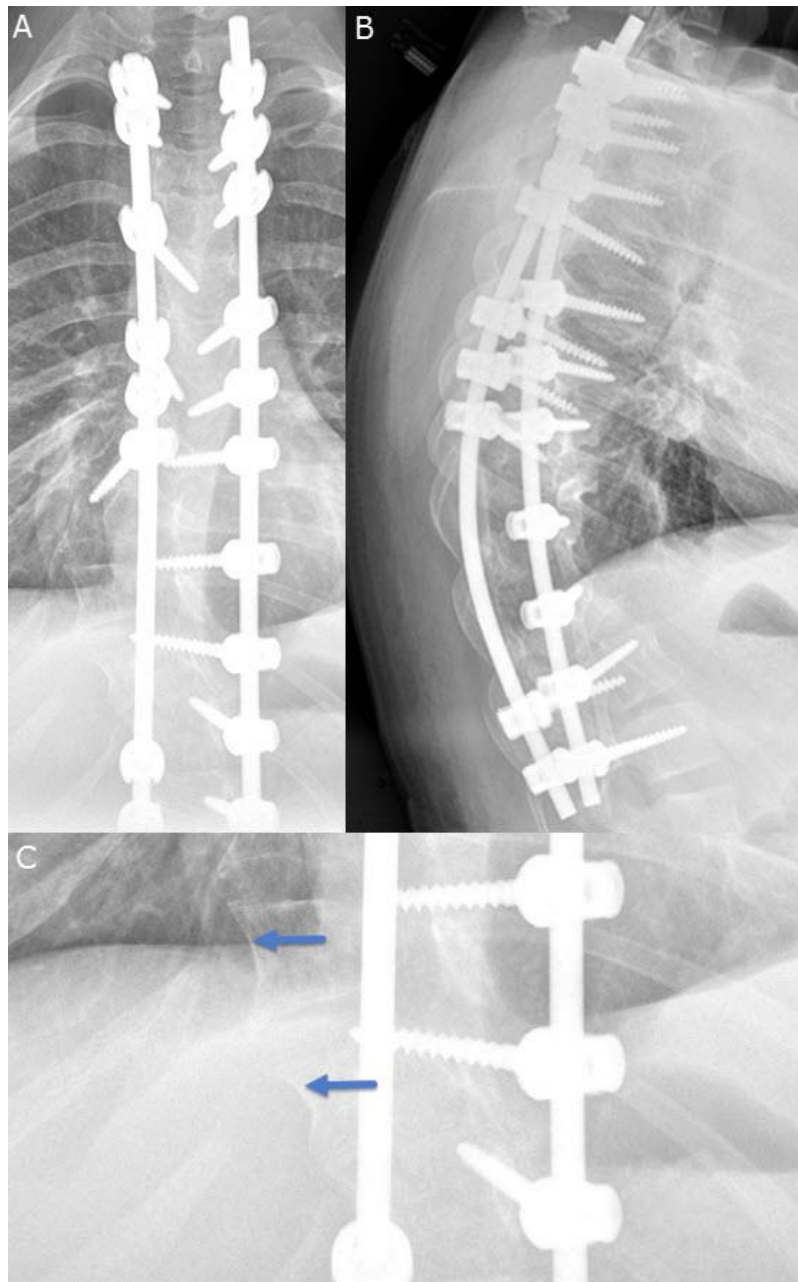
**Figure 6:** Rib head dislocation on CT. 10-year-old female with rib head dislocation from dystrophic scoliosis. **FINDINGS:** (A, B) Rib heads contact the thecal sac at the T10 and T11 level (red arrowheads) (C, D) Images show the relationship of the right rib head with the neuroforamen at each level (red arrowheads). (E) 3-D relationship of the right rib head with the neuroforamen is shown (blue arrows). **TECHNIQUE:** (A, B) Noncontrast axial CT image (100 mAs, 120 kVp, Slice thickness: 2.5 mm). (C) Coronal CT reconstruction (100 mAs, 120 kVp, Slice thickness: 2.5 mm). (D) Sagittal CT reconstruction (100 mAs, 120 kVp, Slice thickness: 2.5 mm). (E) 3D CT reconstruction (100 mAs, 120 kVp, Slice thickness: 2.5 mm).



**Figure 7 (left):** Scalping of vertebral bodies. 10-year-old female with rib head dislocation from dystrophic scoliosis. **FINDINGS:** (A) Anterolateral scalping of the vertebral bodies (blue arrowheads). (B) Posterior vertebral body scalping. **TECHNIQUE:** (A) Scoliosis radiograph, AP view, digitally magnified (68 mAs, 85 kVp) (B) Noncontrast sagittal CT image (100 mAs, 120 kVp, Slice thickness: 2.5 mm).



**Figure 8 (left):** Neuroforaminal enlargement. 10-year-old female with rib head dislocation from dystrophic scoliosis. **FINDINGS:** Neuroforaminal enlargement in the sacrum (green circle). **TECHNIQUE:** Noncontrast sagittal CT image (100 mAs, 120 kVp, Slice thickness: 2.5 mm).



**Figure 9:** Postoperative radiographs. 10-year-old female with rib head dislocation from dystrophic scoliosis. **FINDINGS:** Postsurgical scoliosis AP (A) and lateral (B) plain films. S-shaped scoliosis has improved with multilevel spinal fusion hardware in place. Magnified selected AP view (C) demonstrates the resected right 10th and 11th rib heads (blue arrows). **TECHNIQUE:** Scoliosis radiograph, AP view, digitally magnified (68 mAs, 85 kVp)

<b>Etiology</b>	Head of the rib dislocated into the neighboring neuroforamen which may or may not cause cord compression
<b>Incidence</b>	Documented in selected case reports Incidence may be underestimated since it is often asymptomatic
<b>Gender ratio</b>	No gender predilection
<b>Age predilection</b>	Teenage years
<b>Risk factors</b>	Multiple factors that alter the articulation of the rib with the spinous process 1) Vertebral body rotation 2) Neuroforaminal enlargement 3) Spindling of the transverse processes, 4) Penciling of the apical ribs
<b>Treatment</b>	No clear consensus Rib head excision common in most cases Specific surgical management, including approach to osteotomy and fusion, regarding correction of scoliosis is decided on a case-by-case basis
<b>Prognosis</b>	Proper surgical management seems to have good outcomes but needs more investigation
<b>Findings on imaging</b>	Rib heads dislocate into the neuroforamen indenting the thecal sac Commonly seen on the convex side of the thoracic curve Cord may or may not be involved Can be seen with neurofibromas along the rib

**Table 1:** Summary table for rib head dislocation in dystrophic scoliosis.

	<b>Rib head dislocation</b>	<b>Fracture-dislocation</b>
<b>Radiograph</b>	<ul style="list-style-type: none"> <li>Severe scoliotic curvature can be appreciated, but rib head dislocation can be subtle</li> </ul>	<ul style="list-style-type: none"> <li>Severe scoliotic curvature can be appreciated, but fracture lines or dislocations can be subtle</li> </ul>
<b>CT</b>	<ul style="list-style-type: none"> <li>Rib head dislocates into the neuroforamen on the convex side of the scoliotic curve</li> </ul>	<ul style="list-style-type: none"> <li>Fracture lines with dislocated fragments with surrounding hemorrhage and edema and possible neighboring organ damage</li> </ul>
<b>MR</b>	<ul style="list-style-type: none"> <li>Same as CT.</li> <li>Cord compression and/or increased cord signal on T2 less common</li> <li>T1 and DWI appearances vary.</li> </ul>	<ul style="list-style-type: none"> <li>Same as CT.</li> <li>Possible additional cord compression and/or increased cord signal on T2</li> <li>T1 and DWI appearances vary.</li> </ul>
<b>Contrast enhancement</b>	<ul style="list-style-type: none"> <li>None</li> </ul>	<ul style="list-style-type: none"> <li>May have enhancement if there is hemorrhage</li> </ul>
<b>US</b>	<ul style="list-style-type: none"> <li>Of limited value</li> </ul>	<ul style="list-style-type: none"> <li>Of limited value</li> </ul>
<b>Scintigraphy</b>	<ul style="list-style-type: none"> <li>Bone scan may present with increased uptake depending on chronicity</li> </ul>	<ul style="list-style-type: none"> <li>Bone scan may present with increased uptake depending on chronicity</li> </ul>
<b>PET</b>	<ul style="list-style-type: none"> <li>Varied appearance depending on chronicity</li> </ul>	<ul style="list-style-type: none"> <li>Varied appearance depending on chronicity</li> </ul>

**Table 2:** Differential diagnosis table for rib head dislocation in dystrophic scoliosis.

**ABBREVIATIONS**

CT = Computed tomography  
MRI = Magnetic resonance imaging  
NF-1 = Neurofibromatosis, type 1

**KEYWORDS**

Neurofibromatosis type 1; Dystrophic scoliosis; Spinal canal stenosis; Rib head dislocation; Thoracic spine

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