http://dx.doi.org/10.2106/JBJS.N.00680

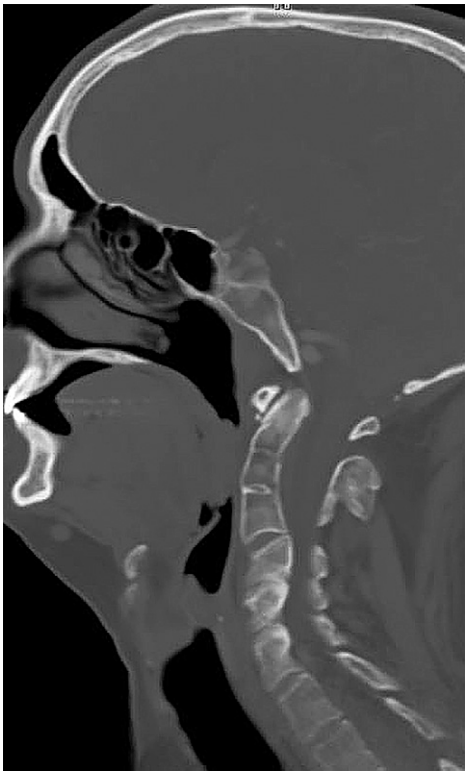

Fig. E-1

Sagittal computed tomogram of a thirty-two-year-old male patient with elongation (5.1 cm) and apex-anterior angulation of the dens.

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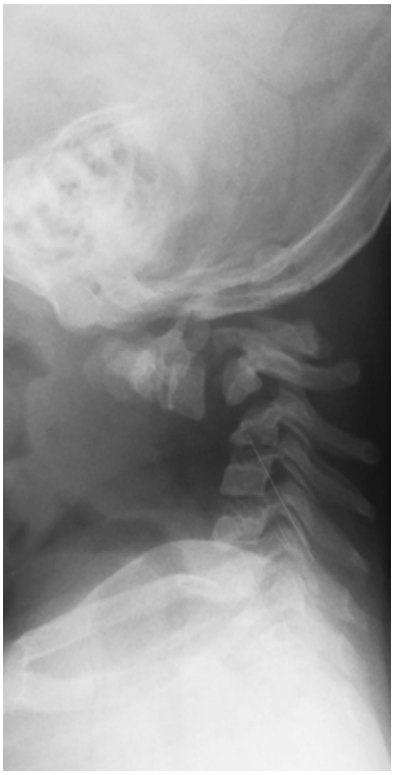
Fig. E-2A

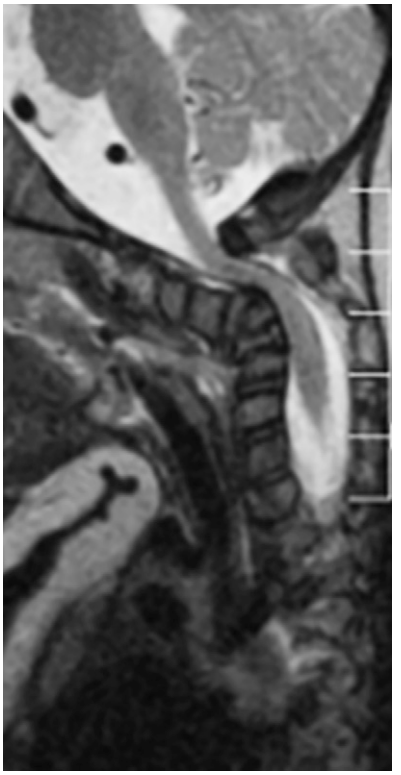
Fig. E-2B

Figs. E-2A through E-2D Images of a boy with C2 spondylolysis and C3-C4 hypoplasia, resulting in a 90° C2-C3 kyphosis and instability. The boy, who had severe contractures, bilateral clubfoot, and motor delay, was evaluated at three months of age after presentation with an apneic episode. **Figs. E-2A and E-2B** Lateral neutral radiograph and T2-weighted magnetic resonance image. The patient was managed with bracing until surgical fixation could be attempted. At seventeen months of age, he underwent C2-C4 anterior spinal fusion and occiput-to-C4 posterior spinal fusion. At twenty-three months of age, he was noted to have occipital implant loosening and pseudarthrosis of C1-C2 and C2-C3, requiring revision posterior spinal fusion from the occiput to C4. He remained in a halo vest until thirty months of age; after halo removal, he developed C4-C5 junctional kyphosis, which remained stable until the age of seven. At this time, subluxation at C4-C5 acutely worsened, requiring revision and extension of anterior implants to C7. Two months after surgery, failure of fixation was noted, and the patient underwent revision of anterior instrumentation and extension posteriorly to T2.

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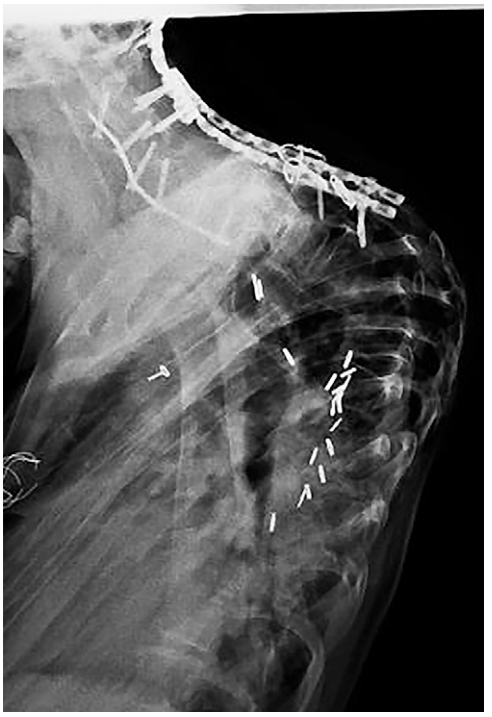

Fig. E-2C

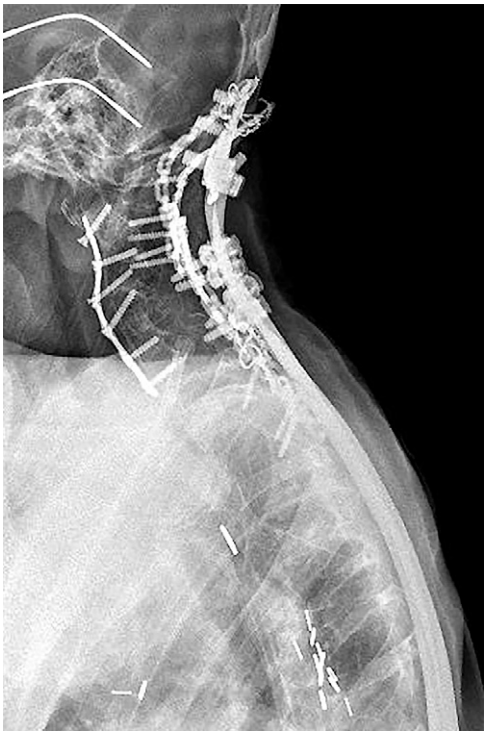

Fig. E-2D

Fig. E-2C As evidenced on the sagittal radiograph, the patient subsequently developed a T2-T3 junctional kyphosis, along with respiratory failure, and was treated with growing rods from the existing construct extending to the sacrum. **Fig. E-2D** Postoperative radiograph. At twelve years of follow-up after the initial cervical procedure, the cervical spine remained fused.

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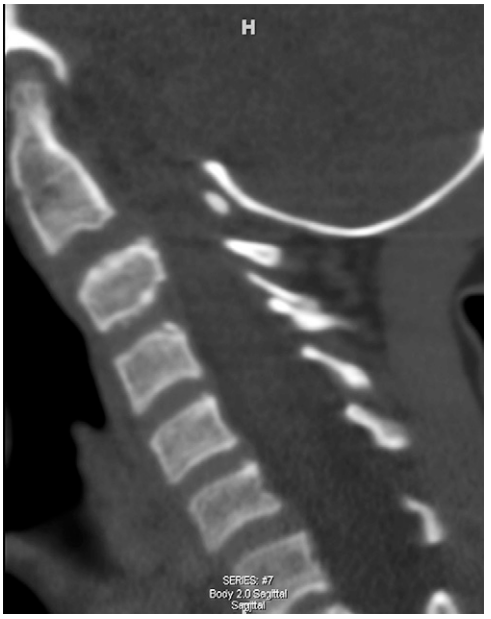

Fig. E-3A

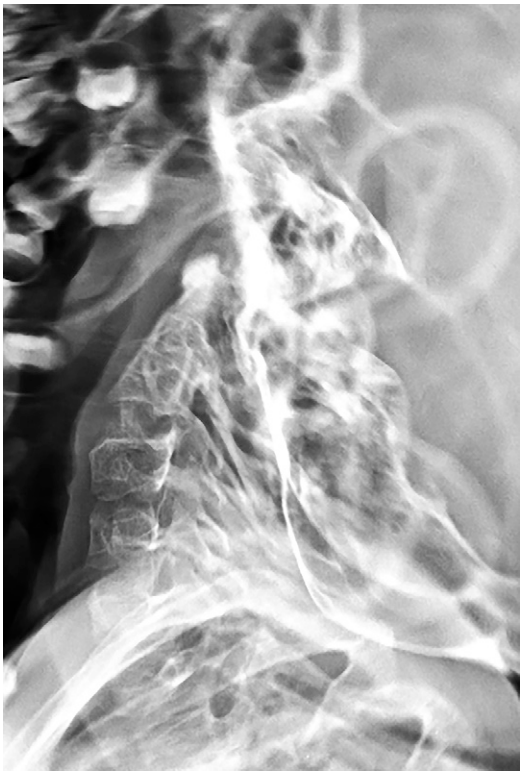
Fig. E-3B

Figs. E-3A through E-3E Images of a girl with Loeys-Dietz syndrome who, at ten years of age, began experiencing bilateral upper-extremity weakness and paresthesia. She previously had been managed with T2-to-pelvis growing rods for thoracic scoliosis. **Figs. E-3A and E-3B** Sagittal computed tomogram at six years of age and lateral extension radiograph at nine years of age showing no evidence of subluxation, which also was not evident on imaging at ten years. At twelve years of age, she presented with increased upper-extremity symptoms and difficulty emptying the bladder.

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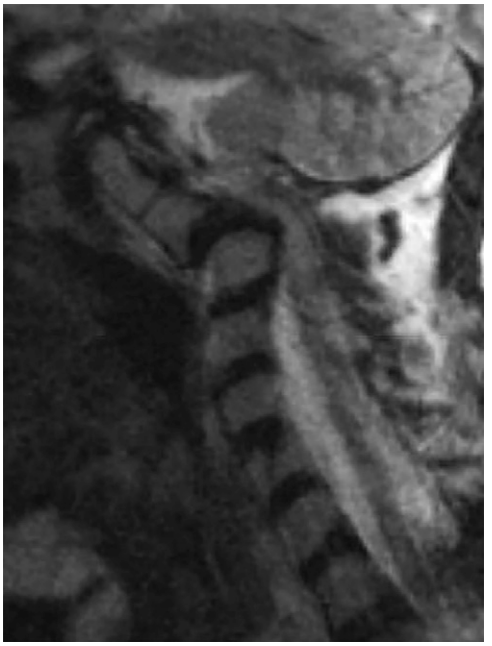
Fig. E-3C

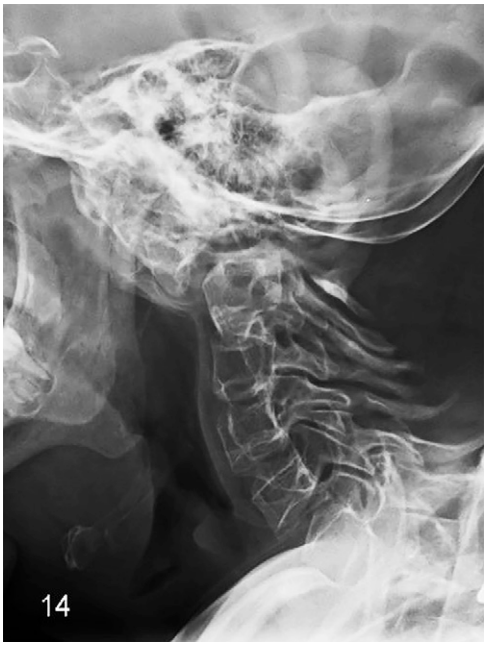

Fig. E-3D

Figs. E-3C and E-3D T2-weighted sagittal magnetic resonance image and lateral neutral radiograph showing C2-C3 subluxation and kyphosis and spinal cord compression at the apex and at the craniocervical junction. Other osseous anomalies were noted, including a C1 posterior arch defect, elongated C2 with basilar impression, hypoplasia of the C2-C3 facets, and C2 spondylolysis.

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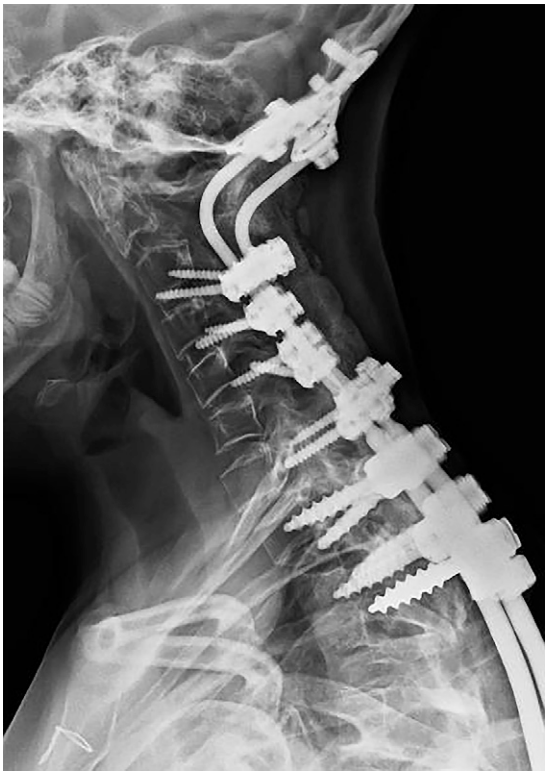

Fig. E-3E

The kyphotic deformity was reduced with traction, and the patient underwent occiput-to-T3 posterior spinal fusion (extension of the growing rod construct). At her most recent follow-up, eighteen months after the initial procedure, her upper-extremity weakness had returned to baseline, with normal strength but some preexisting interphalangeal joint contractures.