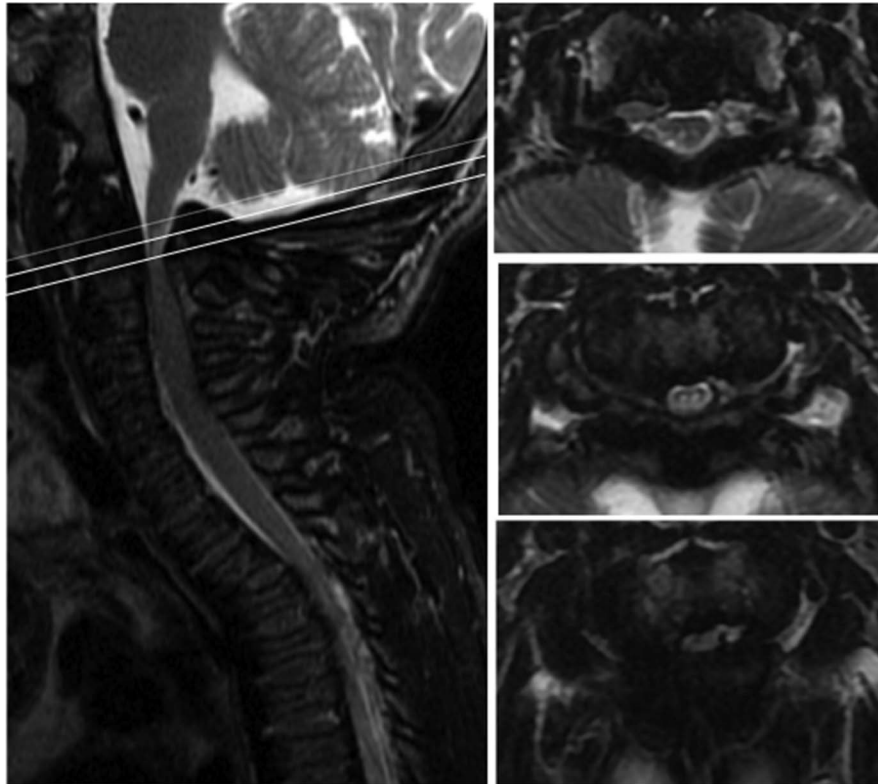


Remote spinal cord injury in mucopolysaccharidosis type IVA after cervical decompression

Figure 1 Preoperative spinal cord MRI



Sagittal T2-weighted image shows abnormal hyperintensity signal at C1 to C2 levels. The 2 upper axial images show the “snake eyes” myelomalacia appearance, secondary to ischemic lesion due to atlantoaxial subluxation; the lowest axial cut shows the maximum degree of spinal cord compression in this patient.

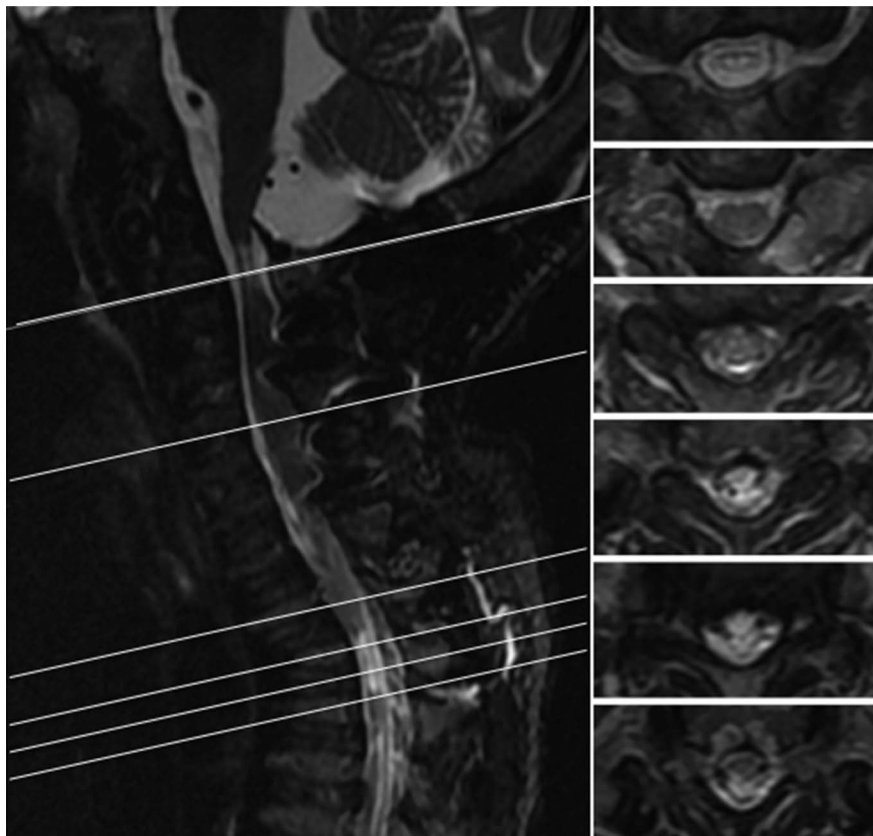
Morquio A syndrome (Online Mendelian Inheritance in Man #253000) is a lysosomal storage disease caused by deficiency of N-acetylgalactosamine-6-sulfatase encoded by the *GALNS* gene. Key clinical features are skeletal dysplasia and short stature.

A 17-year-old boy with Morquio A syndrome had slowly progressive signs of spinal cord compression at C1 to C3. His examination showed Medical Research Council grade 3 in the upper and 2 in the lower limbs, hyperactive reflexes without clonus or Babinski sign, and no sensorial impairment. After occipitocervical fixation in the prone position, he developed acute paraplegia, sensory losses below T3, and striking MRI abnormalities (figures 1 and 2).

Patients with mucopolysaccharidosis may develop remote spinal cord injuries from compression sites after general anesthesia in the prone position due to impaired cardiac output.¹ This outcome is unusual,^{1,2} but providers and patients should be alert to this possible complication.

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Sagittal and axial T2-weighted images. Persistent abnormal hyperintensity signal at C1 to C2 is seen in the sagittal and upper axial cuts; marked narrowing of cord with epidural lipomatosis and an extensive myelomalacia area from T2 to T6 is seen.

Author contributions: Dr. Borlot: study concept and design, drafting the manuscript, accepts responsibility for conduct of research, and acquisition of data. Dr. Arantes: drafting the manuscript, analysis or interpretation of data, accepts responsibility for conduct of research, and acquisition of data. Dr. Cardoso: analysis or interpretation of data, accepts responsibility for conduct of research, revising the manuscript, and final approval. Dr. Kim: interpretation of data, accepts responsibility for conduct of research, study supervision, and final approval.

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