

## Recommended assessments for diagnosing and monitoring spinal involvement in patients with Morquio A<sup>1</sup>

Assessment	At diagnosis	Frequency <sup>a</sup>
Neurological examination	Yes	6 months
Plain radiography cervical spine (AP, lateral neutral, and flexion-extension) <sup>b</sup>	Yes	2–3 years
Plain radiography spine (AP, lateral thoracolumbar)	Yes	2–3 years if evidence of kyphosis or scoliosis
MRI neutral position, whole spine <sup>c</sup>	Yes	1 year
Flexion-extension of cervical spine by MRI <sup>c, d</sup>	Yes	1–3 years
CT neutral region of interest		Preoperative planning

<sup>a</sup> Additional examinations may be necessary before surgical procedures or if there have been no recent examinations.

<sup>b</sup> If plain x-rays are technically inadequate then CT neutral and flexion/extension, usually done without anesthesia, may be considered for the evaluation of instability.

<sup>c</sup> MRI evaluation may require anesthesia which carries substantial risk for patients with Morquio A. Risk may be reduced by minimizing examination times, using continuous monitoring, and performing these examinations in experienced MPS treatment centers.

<sup>d</sup> MRI flexion-extension sequences can be obtained if flexion-extension radiographs are abnormal or inconclusive or when stenosis on MRI is present.

Adapted from Solanski, *J Inherit Metab Dis*, 2013.

Abbreviations: AP, anteroposterior; CT, computed tomography; MPS IVA, mucopolysaccharidosis IVA.

**Reference: 1.** Solanki GA, Martin KW, Theroux MC, et al. Spinal involvement in mucopolysaccharidosis IVA (Morquio-Brailsford or Morquio A syndrome): presentation, diagnosis and management.

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