

Clinical manifestations of Morquio A

Organ system	Complications
Cardiovascular ¹⁻³	Severe thickening of mitral and aortic valves is common and leads to <ul style="list-style-type: none"> • Valve regurgitation • Left ventricular hypertrophy • Valve stenosis • Death
Pulmonary ^{1,4-7}	Multifactorial etiology occurs as a result of prominent GAG accumulation throughout the airways, chest wall restriction, respiratory muscle weakness, and cervical myelopathy leading to <ul style="list-style-type: none"> • Sleep-disordered breathing (eg, obstructive sleep apnea) • Recurrent respiratory infections • Collapse of the upper airway • Respiratory failure • Death
Neurological ^{4,8}	Severe upper cervical spinal cord compression resulting from soft-tissue thickening and odontoid hypoplasia is common and leads to <ul style="list-style-type: none"> • Cervical myelopathy • Tetraplegia • Death
Musculoskeletal ⁴	Significant and well-characterized skeletal and connective tissue involvement resulting in <ul style="list-style-type: none"> • Short stature • Neck/cervical spine compression • Bone deformity • Dysostosis multiplex • Odontoid hypoplasia • Protrusion of chest • Kyphoscoliosis • Joint laxity
Rheumatologic ^{9,10}	Multiple rheumatologic manifestations including <ul style="list-style-type: none"> • Hypermobility of joints • Degradation of connective tissue • Weak grip, progressive difficulties with dressing, personal hygiene, and writing
Ophthalmologic ¹¹⁻¹⁴	GAG and lipid accumulations in keratocytes and endothelial cells leading to <ul style="list-style-type: none"> • Diffuse corneal clouding • Cataracts • Reduction in visual acuity
ENT ¹⁵	Sensorineural GAG accumulation, recurrent upper respiratory tract infections, and deformity of ossicles lead to <ul style="list-style-type: none"> • Mixed hearing loss • Severe, progressive hearing loss by second decade of life
Abdominal ¹⁰	<ul style="list-style-type: none"> • Hepatomegaly • Splenomegaly
Dental ^{4,16}	Severe dental signs and symptoms <ul style="list-style-type: none"> • Poor dentition • Dental caries • Abnormally thin enamel • Widely spaced teeth • Fracture risk

Abbreviations: ENT, ear, nose, and throat; GAG, glycosaminoglycan.

References: **1.** Harmatz P, Mengel KE, Giugliani R, et al. The Morquio A clinical assessment program: baseline results illustrating progressive, multisystemic clinical impairments in Morquio A subjects. *Mol Genet Metab.* 2013;109(1):54-61. doi:10.1016/j.ymgme.2013.01.021. **2.** Ireland MA, Rowlands DB. Mucopolysaccharidosis type IV as a cause of mitral stenosis in an adult. *Br Heart J.*

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