

Clinical manifestations of Morquio A (MPS IVA)

Organ system	Complications
Cardiovascular ^{1,7-9}	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 ^{1,6,7,10,11}	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 ^{6,12}	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 ^{1,6}	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 ^{5,13}	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 ^{1,14,15}	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
Ear, nose, and throat	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 ¹³	<ul style="list-style-type: none"> • Hepatomegaly • Splenomegaly
Dental ⁶	Severe dental signs and symptoms <ul style="list-style-type: none"> • Poor dentition • Dental caries • Abnormally thin enamel • Widely spaced teeth • Fracture risk

Abbreviations: GAG, glycosaminoglycan; MPS, mucopolysaccharidosis.