

Most frequent respiratory involvement in patients with MPS¹¹

Anatomic region	Symptoms	Pathophysiology
ENT aspect	<ul style="list-style-type: none"> • Chronic rhinorrhea • Otitis media with effusion • Hearing loss • Adenotonsillar hypertrophy 	<ul style="list-style-type: none"> • Unclear • Conductive and sensorineural • GAG deposits
Upper airway	<ul style="list-style-type: none"> • Macroglossia • Pharyngeal narrowing • Limitation of full mouth opening • Stridor, laryngomalacia 	<ul style="list-style-type: none"> • GAG deposits • Multifactorial • Decreased temporomandibular joint mobility • Deposits in epiglottis and arytenoids and decreased muscle tone
Lower airway	<ul style="list-style-type: none"> • Subglottic stenosis • Tracheomalacia/stenosis • Bronchomalacia • Bronchitis/pneumonia 	<ul style="list-style-type: none"> • GAG deposits • GAG deposits in submucosa and cartilage with instability of tracheal rings • GAG deposits in submucosa • Increased secretions, poor airway clearance
Restrictive lung disease	<ul style="list-style-type: none"> • Progressive respiratory inefficiency • Abdominal distension 	<ul style="list-style-type: none"> • Kyphosis, scoliosis, flattening of the vertebral bodies • Abnormal ribs and costovertebral angle • Hepatosplenomegaly
Sleep disturbance	<ul style="list-style-type: none"> • Mostly obstructive apnea • Central apnea and disordered sleep cycle possible 	<ul style="list-style-type: none"> • Airway obstruction • Central apnea: hydrocephalus, spinal cord compression, neuronal degeneration

Adapted from Muhlebach, *Paediatr Respir Rev*, 2011.

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