Most frequent respiratory involvement in patients with MPS[1]

Anatomic region	*Symptoms*	*Pathophysiology*
ENT aspect	• Chronic rhinorrea • Otitis media with effusion • Hearing loss • Adenotonsillar hypertrophy	• Unclear • Conductive and sensorineural • GAG deposits
Upper airway	• Macroglossia • Pharyngeal narrowing • Limitation of full mouth opening • Stridor, laryngomalacia	• GAG deposits • Multifactorial • Decreased temporomandibular joint mobility • Deposits in epiglottis and arytenoids and decreased muscle tone
Lower airway	• Subglottic stenosis • Tracheomalacia/stenosis • Bronchomalacia • Bronchitis/pneumonia	• 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	• Progressive respiratory inefficiency • Abdominal distension	• Kyphosis, scoliosis, flattening of the vertebral bodies • Abnormal ribs and costovertebral angle • Hepatosplenomegaly
Sleep disturbance	• Mostly obstructive apnea • Central apnea and disordered sleep cycle possible	• Airway obstruction • Central apnea: hydrocephalus, spinal cord compression, neuronal degeneration

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

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

Reference: 1. Muhlebach MS, Wooten W, Muenzer J. Respiratory manifestations in mucopolysaccharidoses. *Paediatr Respir Rev*. 2011;12(2):133-138. doi:10.1016/j.prrv.2010.10.005.