

ANESTHESIOLOGIST SPECIALIST GUIDE

General anesthesia can be difficult and risky due to the anatomic changes, including short neck, jaw immobility, and upper airway obstruction, seen in MPS II patients.¹ Ideally, MPS II patients should undergo general anesthesia in centers staffed with anesthesiologists experienced in MPS disorders, and local or regional anesthesia should be considered where possible.^{1,2} The possibility of combining multiple minor surgeries into one general anesthesia session should be evaluated, however, it is important to remember that extending the operation time greatly increases the risk of respiratory complications.^{1,3}



AJ, age 3

**HUNTER
SYNDROME IS A
MULTISYSTEMIC
AND PROGRESSIVE
DISEASE**

Before Surgery

Prior to surgery the patient should be assessed by a multidisciplinary team that includes a cardiologist, otorhinolaryngologist, and anesthesiologist, and a full cardiac assessment should be conducted.¹ In order to evaluate the individual anatomy of the airway, a flexible nasendoscopy and computed tomography scan, or a video recorded bronchoscopy using a fiberoptic bronchoscope can be carried out prior to surgery.^{1,3} Parents should be consulted about the possibility of an acute airway obstruction resulting in an urgent tracheotomy or cricothyrotomy during intubation and extubation.³

During Surgery

As the atlantoaxial joint is unstable in MPS II patients, careful positioning is required to avoid hyperextension of the neck.² In some centers it is common practice for an otorhinolaryngologist or pediatric pulmonologist to be available during the induction of anesthesia and intubation of the patient.³ Bronchoscopic intubation is often the most appropriate intubation technique, because of the tracheomalacia of the airway.^{1,3} In difficult patients or for brief procedures a laryngeal mask airway may provide adequate airway control.³

After Surgery

Early extubation can reduce the risk of post-obstruction pulmonary edema exacerbating upper-airway obstruction.¹ Using a helium–oxygen breathing mixture at the time of extubation may relieve the upper airway obstruction and improve outcomes because the reduced density of this air mixture decreases the work of breathing and increases linear flow rates.³ Recovery from anesthesia can be slow in MPS II patients.²

1. Scarpa M et al. *Orphanet J Rare Dis* 2011; 6: 72. 2. Neufeld EF, Muenzer J. In: Scriver CR, eds. *The Metabolic and Molecular Bases of Inherited Disease*. New York, NY: McGraw-Hill; 2001: 3421-3452. 3. Muenzer J et al. *Pediatrics* 2009; 124(6): e1228-1239.

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