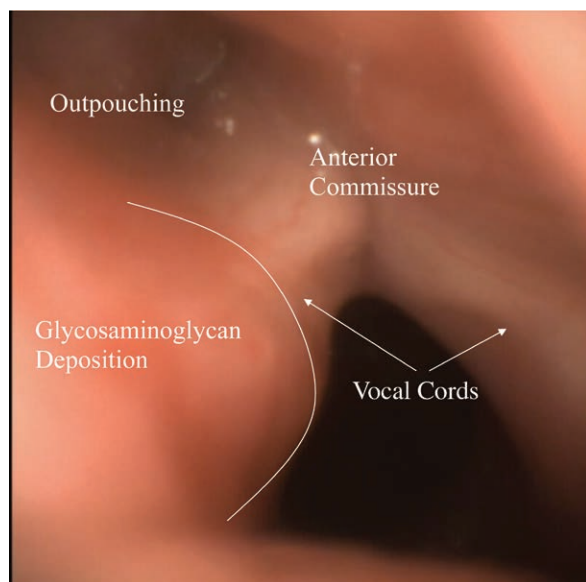


# Airway Anatomy of an Adult with Hurler's Syndrome

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**H**urler's Syndrome is a rare genetic disorder characterized by a deficiency in the enzyme  $\alpha$ -L-iduronidase, leading to an accumulation of glycosaminoglycans in the soft tissues. Without treatment, these patients experience severe multisystem dysfunction leading to airway obstruction, cardiopulmonary failure, and eventually death at 5 to 10 yr of age.<sup>1</sup> Allogeneic hematopoietic stem cell bone marrow transplantation significantly reduces disease progression, increasing survival into the third decade of life.<sup>2</sup>

These videos are from a 19-year-old patient with Hurler's Syndrome presenting for surgery whereby a "cannot intubate" scenario occurred requiring urgent supraglottic airway placement and intubation using a flexible bronchoscope through the supraglottic airway. While maintaining spontaneous ventilation and after passing the bronchoscope through the supraglottic airway, the aryepiglottic folds are engorged and appear edematous (Supplemental

Digital Content, video 1, <http://links.lww.com/ALN/C343>). After advancing the scope past the engorged tissue, the vocal cords could be seen in addition to an out-pouching cephalad to the anterior commissure. Polysaccharide deposition between the tracheal rings made discerning tracheal structures difficult (Supplemental Digital Content, video 2, <http://links.lww.com/ALN/C344>).

Patients with Hurler's Syndrome are presenting for surgery with increased frequency to nonpediatric centers. It is important for anesthesiologists to be familiar with common airway changes occurring in this disease, including the presence of a short, immobile neck, narrow nasopharyngeal passages, limited temporomandibular joint movement, macroglossia, tonsillar hypertrophy, and tissue engorgement specifically within tracheal cartilage.<sup>3</sup> Planning for difficult airway management and performing a controlled induction with a multitude of ventilation and intubation devices readily available can lead to high first-attempt success and mitigation of perioperative complications.

## Competing Interests

The authors declare no competing interests.

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Published online first on March 25, 2020. From the Departments of Anesthesiology and Critical Care Medicine (N.M.D., O.A.I., C.M.T., T.P.T.) and Otolaryngology/Head and Neck Surgery (N.M.D.), Johns Hopkins University School of Medicine, Baltimore, Maryland.

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