

Place of Tracheotomy in Management of Air Way Obstruction for a Child with Hunter Syndrome Case Report

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Abstract : *Mucopolysaccharidoses (MPS) are composed of inherited conditions coupled with errant mucopolysaccharide degradation. In connective tissues, since these substances are ubiquitous, the manifestations are large and challenging. Deficiencies in the lysosomal enzyme underlie the inadequate degradation of mucopolysaccharides and result in the accumulation in connective tissues of incompletely catabolized substances. There are 11 known deficiencies of enzymes that generate seven recognized MPS phenotypes. It has defined subtypes.11 In patients with type mucopolysaccharidosis (MPS), progressive multi-level upper airway obstruction can develop. Tracheostomy remains an effective technique to protect the airway where other procedures have failed or when the airway obstruction requires several locations, considering the particular challenges posed by these complex patients. The presence of airways is primarily responsible for the substantial anesthetic risk seen in MPS.*

Keywords: Tracheostomy Airway Mucopolysaccharidoses Hunter's syndrome enzyme deficiencies

1. Case Report

A 5-year-old Saudi male with hunter s syndrome presented in emergency room with signs and symptoms of respiratory failure that were secondary to inflammation of the upper respiratory tree and increased breathing rate. The Flaring of the Nasal. A rapid physical examination showed that the child was alert with extreme difficulty breathing. He was drooling, his breathing rate with accessory muscles utilizing .cyanosis was 34 per minute .temperature was 37.5 Oxygen saturation was 80 percent with a bilateral wheezing and the patient from mechanical ventilation was not possible according to Weaning, and a tracheostomy was required.

Otolaryngologic examination showed increased mandibular length, making it difficult to enter the anterior neck,

Tracheostomy was completed successfully and without any complications and the patient improved over time and corrected respiratory conditions fig 1

Shiely tube size 4 was the tracheostoma tube used in this operation. Fig 2

After the tracheostomy and stabilization of the patient s conditions Xray and ct chest were requested for further evaluation and follow up



Figure 1: Tracheostomy in a patient with Hunter's syndrome with sheily tube size 4

2. Discussion

Management of the airway in patients with MPS can be difficult, including quantification of the risk of endotracheal intubation and extubation while tracheostomy is a very efficient way of securing the long-term airway, due to the

significant risk of tracheostomy-related complications, it should be treated with caution. Because of the short neck, GAG deposits anterior to the trachea and a relatively small trachea, it may be difficult to perform a tracheostomy itself in MPS patients[1]. In MPS patients, accidental decannulation is relatively normal due to a short neck and

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deeper trachea [16]. By using 'maturation sutures' that secure the skin to the edges of the tracheotomy, this issue can be resolved. In the event of unintended decannulation, this not only allows quick tracheal reinsertion, but also decreases the risk of developing a 'false passage' anterior to the trachea during tracheostomy tube reinsertion. There were no major issues with the alteration of the tracheostomy tube in our patients. Jeong et al. have also used a similar surgical procedure in patients with MPS II [16]. However, in patients with thickened soft tissues and those with difficult surgical access to a deeply placed trachea, maturation sutures are not possible.

Local damage to the tracheal mucosa, resulting in inflammation, scarring and potential GAG deposition due to increased cell turnover, can be caused by tilting of the tracheostomy tube resulting from a relatively short neck, exacerbating any tracheal narrowing [2].

This inflammation may result in granulation tissue formation at the point of contact between the tracheostomy tube tip and the tracheal mucosa [3]. The resultant infratip granulation may necessitate additional rigid airway endoscopy and surgical debridement [4]

MPS II patients also have poor mucociliary clearance which results in recurrent tracheitis and obstruction of tracheostomy tubes by thickened secretions [3].

Repeated inflammation worsened by granulation tissue formation may eventually lead to tracheal In MPS II patients, tracheal stents (metallic or silicone) were used to treat tracheal narrowing and/or collapse [4]. Silicone stents, particularly with development, tend to migrate and inhibit clearance of tracheobronchial secretions[5]. In the tissue, metallic tracheal stents become implanted and do not develop with the child's normal development. Also, both types of stents can cause stenosis. Despite these risks, tracheostomy will be the most appropriate treatment in some children as their disease progresses and dependent upon response to treatment such as enzyme replacement therapy.

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3. Conclusion

In children with MPS, progressive upper airway obstruction is widespread. Local airway procedures aimed at improving airway patency, including adenotonsillectomy or microsurgical debulking of prolapsing laryngeal soft tissue, are part of the initial treatment of upper airway obstruction in patients with MPS. If local airway activities are no longer sufficient, , or when there is significant tracheobronchial involvement, non-invasive CPAP may be tolerated by some

patients during sleep. Tracheostomy is a very effective way of managing airway obstruction when less invasive The techniques are no longer sufficient, but they are not without complications. In a patient with the anatomical features and soft tissue characteristics of MPS, chronic tube blockage is prevalent due to excessive secretions and replacement of a blocked or displaced tube. Adjustable flange and more flexible tracheostomy tubes are an important addition to the equipment available for use in these patients. There are no large-scale prospective studies of the airway manifestations of MPS disorders available, with most reports representing the experience of each center. Similarly, the effect of enzyme replacement therapy (ERT) on the airway is not yet well known, and in those patients who do not respond to ERT or in those patients who live into adulthood, tracheostomies are likely to remain a significant airway adjunct. A multidisciplinary team of physicians with expertise in the treatment of these patients must be involved in the management of the airways of all patients with MPS in order to offer the highest level of care for this rare condition.

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