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Case report

Montgomery© T-tubes in the management of multilevel airway obstruction in mucopolysaccharidosis

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Introduction

The mucopolysaccharidoses (MPS) represent a heterogeneous group of inherited metabolic disorders. The underlying abnormality is a deficiency of one of the hydrolytic enzymes present in the lysosomes, which normally degrade glycosaminoglycans (GAGs) in a stepwise fashion. Deficiency of these enzymes leads to accumulation of partially degraded GAGs within the cells, as they cannot be broken down by alternative methods. The accumulated GAGs affect cell/tissue function and the clinical phenotype is dependent upon the enzyme deficiency involved and the cell type this substrate is most prevalent in [1,2]. The MPS subtypes are inherited in an autosomal recessive fashion except for MPS II, which is X-linked. Each subtype displays a wide variety of signs and symptoms as well as differing levels of severity [3].

Inherent to a number of the MPS disorders is airway compromise, particularly MPS I (Hurler/Hurler Scheie), MPS II (Hunter), MPS IVA (Morquio), MPS VI (Maroteaux–Lamy) and MPS VII (Sly) [4]. Airway obstruction may be due to adenotonsillar hypertrophy, macroglossia, reduced mouth opening, laryngopharyngeal GAG deposits and tracheobronchomalacia [4]. The airway obstruction is often multilevel and multifactorial, with the accumulation of GAG deposits within the airway leading to progressive airway obstruction [5]. Airway obstruction in MPS is considered by many to represent the most challenging paediatric airway, encompassing the attempt to ameliorate airway obstruction and to safely secure the airway during general anaesthesia [6].

We present the first case series detailing the use of tracheal T-tubes to assist in the management of airway obstruction in children and young people with MPS. Our management of progressive airway obstruction in MPS will be discussed along with the treatment algorithm developed in our tertiary referral centre.

Methods

We undertook a retrospective case note review of all MPS patients requiring a T-tube to assist in the management of airway obstruction at our tertiary Centre. The primary outcome measure for this study was the effectiveness of the T-tube insertion on airway patency and the secondary outcome measures were complications directly attributable to the T-tube and the requirement for further airway surgery.

Case 1

A seventeen year-old boy with MPS II (Hunter syndrome) developed progressive shortness of breath, initially on exertion

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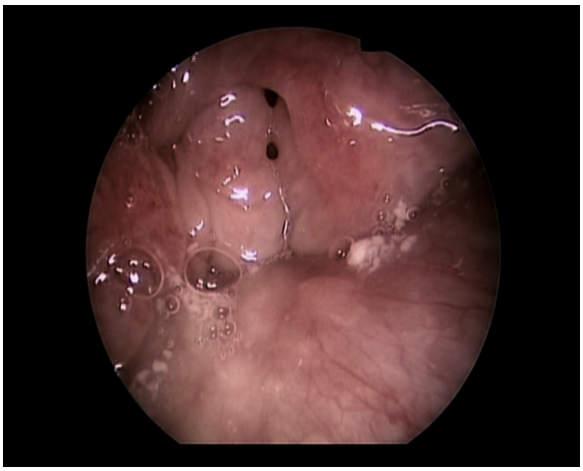

Fig. 1. Supraglottic GAG deposits with limited view of the vocal cords.

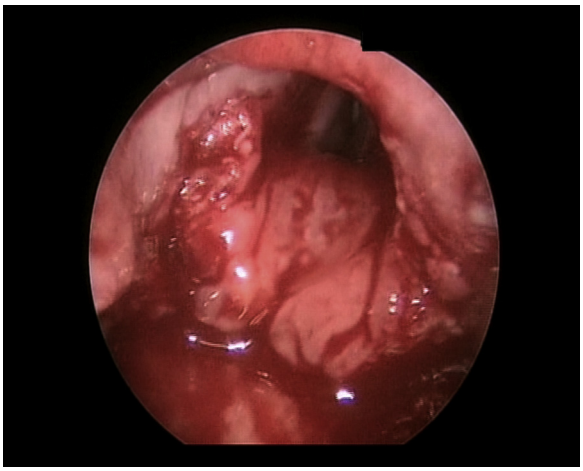

Fig. 2. Post-operative view of vocal cords following microdebrider removal of supraglottic GAG deposits.

and then at rest, accompanied by nocturnal choking and deteriorating respiratory function (FEV1 and FVC 40% of expected). Clinically he had macroglossia with no view of the soft palate and subsequent pharyngolaryngoscopy revealed a large adenoidal pad and extensive supraglottic GAG deposits with no view of his larynx.

A rigid airway endoscopy was attempted but had to be abandoned secondary to a failed attempt at establishing an anaesthetic airway. A grade 4 view of the larynx was noted (Cormack and Lehane grading system) at that time and a decision was made to support the airway conservatively. This multidisciplinary decision was made in light of the difficulty of airway access, the level of perceived risk from further attempts at airway surgery and the lack of immediately life-threatening airway disease. Six months later the adolescent was re-admitted through accident and emergency with rapidly deteriorating respiratory function and an urgent awake fibreoptic intubation with a size 5.0 microlaryngoscopy tube was performed.

At rigid airway endoscopy only the posterior larynx was visible with the 0° 4 mm Hopkins rod. A tracheostomy was fashioned and a 4.5 mm Microcuff® paediatric endotracheal tube (Kimberly-Clark) was customized and used as a temporary tracheostomy tube. This was later up sized to a size 5.5 mm customized Microcuff® tube and then replaced with a Bivona® size 5.5 uncuffed paediatric Flextend™ silicone tracheostomy tube (Smiths Medical). The pharyngeal and supraglottic GAG deposits were debulked one week later using a Medtronic Skimmer® Laryngeal microdebrider blade until true cords were visible. Tracheoscopy revealed tracheomalacia extending from the immediate subglottis to the carina. Fig. 1 (GAG deposits supraglottic), Fig. 2 (glottis following debridement). His ventilatory support and oxygen requirements were slowly weaned.

Due to the ongoing need for ventilatory support, alongside persistent upper airway disease, his tracheostomy could not be removed. A decision was made to replace this with a 9.0 mm Montgomery© T-tube, which has remained in-situ for six months. The T-tube is 'capped off' during the day and the patient has a good voice. His most recent assessment diagnostic endoscopy under anaesthetic showed a recurrence of supraglottic disease, requiring further debulking. His T-tube remains in-situ to assist with the on-going airway obstruction.

2.2. Case 2

A fifteen year-old boy with type VI (Maroteaux–Lamy) MPS developed upper and lower limb motor weakness secondary to cervical spinal cord compression. A decision was made to surgically stabilise his C-spine and prevent further neurological deterioration. He had established aortic and mitral valve incompetence and corneal clouding causing blindness. He also had severe airway compromise with significant stridor witnessed in outpatients. As seen in patients with MPS VI, he adopted an extended neck position to maximize his airway.

Prior to his spinal surgery, airway endoscopy under general anaesthetic confirmed narrowing at the level of the cricoid cartilage extending into the upper trachea, due to anterior and posterior soft-tissue swelling resulting in an 80–90% luminal reduction. A further GAG deposit was identified approximately 2 cm above the carina. A post-operative computer tomography (CT) scan was performed which confirmed that these masses were intrinsic to the trachea.

A tracheostomy was performed prior to his spinal surgery to ensure safe maintenance of his airway throughout his perioperative period. It was not possible to decannulate his tracheostomy due to progression of his airway deposits and heavy reliance on the tracheostomy tube. A year later his tracheostomy was exchanged for a size 7.0 Montgomery© T- tube, which has now remained in-situ for four years. At his last diagnostic endoscopy under anaesthetic he was found to have granulation tissue within the subglottis and 30% obstruction of his distal trachea secondary to GAG deposits.

Case 3

A six year-old boy with type II (Hunter) MPS presented acutely at the age of six years. He had suffered a nocturnal respiratory arrest but had been successfully resuscitated by his parents. Over the preceding six weeks it had been noted that he had developed increasing weakness and hypertonicity. An MRI scan demonstrated a significant narrowing of the cranio-cervical junction leading to severe spinal cord compression at this level explaining his motor symptoms. The scan also demonstrated significant upper airway narrowing and a diagnostic upper airway endoscopy confirmed GAG deposits in the supraglottis. A size 6.0 Shiley® (Covidien) tracheostomy tube was inserted. This was later changed to a softer Bivona® size 7.0 uncuffed paediatric Flextend™ silicone tracheostomy tube (Smiths Medical). He could not be decannulated over the ensuing years secondary to on-going upper airway obstruction.

At the age of seventeen, he underwent an elective mitral valve replacement. His post-operative recovery was difficult, requiring high-pressure support on intensive care unit. This was secondary to GAG deposits within the anterior and posterior distal tracheal walls,