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Transnasal adenoidectomy in mucopolysaccharidosis

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ABSTRACT

Background: Mucopolysaccharide (MPS) diseases are a heterogeneous group of inherited, metabolic disorders characterized by accumulation of partially degraded glycosaminoglycans (GAG) in multiple organ systems. Due to accumulation in the airway, patients often present with multilevel airway obstruction and obstructive sleep apnoea (OSA). Adenotonsillar surgery leads to a significant improvement in the severity of OSA in MPS patients. However, access to secure the airway and for conventional surgery can be challenging, due to limited neck extension, macroglossia and reduced mouth opening. This study was undertaken to evaluate the role of transnasal microdebridement and radiofrequent plasma ablation (Coblation) in adenoidectomy to treat OSA in patients with MPS and restricted airway access.

Methods: A retrospective case review was performed including patients with MPS undergoing adenoidectomy for OSA in the period between June 2015 and March 2017. In all cases, either a microdebrider (Gyrus Diablo) or a Coblation wand (EVAC70, Smith&Nephew) was used via a transnasal approach guided by nasendoscopy. The primary outcome was effect upon OSA, measured by sleep oximetry and parental report of benefit. The secondary outcomes were surgical complications and risk factors for persistent OSA after surgery.

Results: A total of nine patients were identified with a mean age of 9 years (range 3–14 years) at surgery. Postoperative sleep study data was available for eight patients (8/9). Six patients (6/8) had improvement in 4% oxygen desaturation index (ODI-4) with a mean of 8.11 pre-operatively (range 2.69–14.0) and 4.99 postoperatively (range 0.68–8.48). ODI-4 did not improve in two (2/8) patients. Irrespective of sleep oximetry results, improvement in OSA-related symptoms was noted by all parents postoperatively. No risk factors for persistent OSA were identified. Furthermore, no complications were noted in this cohort.

Conclusion: Transnasal Coblation and Microdebrider adenoidectomy is a safe and effective surgical treatment for OSA in patients with Mucopolysaccharidosis and adenoidal hypertrophy. As lifespan increases for patients with the Mucopolysaccharidoses, greater emphasis is being given to optimising airway management over the longer-term. This technical note describes the novel application of endoscopic techniques for the management of primary adenoidal hypertrophy when transoral access is restricted, or to debulk recurrent disease that would be challenging to remove via the standard transoral route.

1. Introduction

Mucopolysaccharidosis (MPS) is a rare lysosomal storage disorder with an incidence of around 1 in 22,000 [1]. Inheritance is in an autosomal recessive manner, except for MPS II, which is X-linked. MPS is caused by a deficiency in specific enzymes that affect the degradation of glycosaminoglycans (GAG) causing accumulation of this substance within cells. There are 11 known enzyme defects causing 7 different sub-types of MPS (Table 1). Patients are born with few symptoms but as GAG accumulates within cells, clinical disease is manifest and management of complications is required [1]. As GAG is ubiquitously expressed throughout the body, MPS is a multi-system disorder that causes a wide range of symptoms including cognitive delay, musculoskeletal dysplasia, airway problems, cardiorespiratory complications as well reduced vision and hearing [2].

Common features in otolaryngology include adenotonsillar hypertrophy, otitis media with effusion, hearing loss, chronic rhinitis and obstructive sleep apnoea (OSA) [3]. Airway issues have been shown to predominantly affect patients with MPS I, II, IV, VI and VII, with MPS III patients less severely affected [1]. A significant number of patients will also have laryngomalacia, tracheobronchomalacia, structural abnormalities of the trachea or craniofacial abnormalities including

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Table 1
Types of mucopolysaccharidosis.

Type	Alternative name	GAG	Incidence [8]	Common ENT features [9]
MPS IH	Hurler	α-L-iduronidase	1 in 100,000 [8]	Adenotonsillar hypertrophy and obstructive sleep apnoea (OSA), progressive multi-level airway obstruction, airway deposits, otitis media with effusion (OME), sensorineural hearing loss (SNHL)
1H/S	Hurler-Scheie		1 in 100,000–500,000 [8]	
1S	Scheie		1 in 500,000 [8]	
MPS II	Hunter	Iduronate-2-sulfatase	1 in 300,000 [8]	Adenotonsillar hypertrophy and obstructive sleep apnoea (OSA), progressive multi-level airway obstruction, airway deposits, otitis media with effusion (OME), sensorineural hearing loss (SNHL)
MPS IIIA	Sanfilippo A	N-sulfoglucosamine sulfohydrolase	1 in 70,000 [8]	OME, adenotonsillar hypertrophy and OSA, SNHL
IIIB	Sanfilippo B	N-alpha-acetylglucosaminodase		
IIIC	Sanfilippo C	Heparan acetyl-CoA: alpha-glucosaminide		
IIID	Sanfilippo D	Nacetyltransferase N-acetylglucosamine-6-sulfatase		
MPS IVA	Morquio syndrome A	Galactosamine-6-sulfatase b-D-galactosidase	1 in 200,000-300,000 [8]	Tracheomalacia and/or tracheal tortuosity secondary to mismatch between thoracic and tracheal dimensions, OME, adenotonsillar hypertrophy and OSA, restrictive lung disease
IVB	Morquio syndrome B	β-D-galactosidase		
MPS VI	Maroteaux-Lamy	N-acetylgalactosamine -4-sulfatase	1 in 250,000–600,000 [8]	Progressive multi-level airway obstruction, adenotonsillar hypertrophy and OSA, OME, airway deposits, SNHL
MPS VII	Sly	b-glucuronidase	1 in 250,000 [8]	Similar ENT manifestations to MPS I & VI

temporomandibular joint immobility, resulting in multilevel airway obstruction [1,4,5]. This often results in sleep disordered breathing (SDB) or OSA, diagnosed in up to 81% of patients with MPS [1].

SDB is a spectrum of disorders ranging from simple snoring through to OSA [6]. OSA can cause a wide range of adverse effects in affected children including cognitive impairment as well as severe physical effects such as failure to thrive and cor pulmonale [6]. To limit the potential adverse effects of untreated OSA in these susceptible patients with co-existing multi-system disease, they often require airway interventions [1].

First line surgical management of OSA in MPS is adenotonsillectomy, with tracheostomy reserved for the most severe cases [1]. It has been our experience that patients with MPS require revision adenoidectomy more frequently due to regrowth of adenoidal tissue. Accessing the airway for conventional adenotonsillectomy can be problematic in MPS for a variety of reasons, including limited neck movements or cervical instability, macroglossia and reduction in mouth opening. Reduced mouth opening is an emerging pathology in MPS that we will discuss in a future manuscript.

We describe our experience of endoscopic adenoidectomy in patients with MPS and restricted airway access.

2. Methods

9 patients with MPS underwent endoscopic adenoidectomy between June 2015 and March 2017, with the choice of surgical approach considered pre-operatively in clinic and at the start of surgery. As a result of limited surgical access and the extent and position of adenoidal tissue in relation to the posterior choana and posterior nasal cavity it was decided that the transnasal endoscopic approach would provide optimal results.

Case notes were retrospectively reviewed and demographic data, operative details and pre- and post-operative sleep study findings were recorded. Patients were cared for post-operatively either in a high dependency unit (HDU), for children requiring one-to-one nursing and non-invasive ventilatory support, or on our routine post-operative ward. Demographic data is represented in Table 2.

2.1. Operative technique

The procedures were performed under general anaesthesia with endotracheal intubation or laryngeal mask ventilation. Topical decongestion with xylometazoline 0.5 mg/mL was performed prior to surgery. A 2.7 mm 0° Storz endoscope and Gyrus Diablo microdebrider (500RPM) or EVAC70 Smith&Nephew coblation (settings varied between 7/3 and 9/3) were used to endoscopically remove any obstructing adenoidal tissue (see Fig. 1). Care was taken to avoid debridement of the Eustachian cushions. Haemostasis was achieved with topical 1/10,000 adrenaline soaked patties. Patients were kept in hospital for a minimum of one night with overnight oxygen saturation monitoring. Sleep oximetry studies were repeated post operatively in eight patients.

Statistical analysis was not possible due to the small number of subjects. The presence or absence of subjective improvement was recorded for all patients.

3. Results

3.1. Operation details

Three (3/9) patients underwent radiofrequency ablation (coblation) endoscopic adenoidectomy and six (6/9) underwent endoscopic adenoidectomy with a microdebrider (Table 3).

Seven (7/9) patients had previously undergone adenoidectomy and a primary endoscopic procedure was chosen for two (2/9) patients. For the two patients undergoing a primary procedure one patient was noted to have grade 1 tonsils and therefore the decision was made to leave the tonsils in situ. The other patient underwent a coblation tonsillotomy, where a complete tonsillectomy was not possible due to limited mouth opening. The reasons stated for utilising endoscopic techniques included C-spine subluxation, difficulty extending the neck and limited mouth opening.

No intra-operative complications occurred, although one patient had a prolonged operating time of 2.5 h due to difficult surgical access. No post-operative complications occurred. Four (4/9) patients had a planned admission to the High Dependency Unit (HDU) post-operatively and five (5/9) were monitored overnight on the ward.