



## CASE REPORT

# Airway management in mucopolysaccharidosis: a retrospective case series review



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### KEYWORDS

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**Abstract** Airway management can be difficult in surgeries of patients with mucopolysaccharidosis. We performed a retrospective review of 31 surgeries performed between 2015 and 2019. The mean age of the patients was 127.6 months. MPS-IV and MPS-VI were the most frequent subtypes. Orthopedic surgeries were the most common surgery type. Difficult intubation was seen in 10 procedures. All patients with difficult intubation were aged over 36 months. Video laryngoscopy was the most common intubation method. Seventeen patients were followed up in the intensive care unit. Although video laryngoscopy seems to be a safe method in these patients, one should always be prepared for alternative methods.

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## Introduction

Mucopolysaccharidosis (MPS) is an inherited and progressive lysosomal storage disorder associated with glycosaminoglycan aggregation in various tissues.<sup>1</sup> Patients with MPS often undergo surgery due to organ pathologies and extremity anomalies. Airway management is difficult in these patients due to facial deformities and mucopolysaccharide aggregation in the nasopharynx.<sup>2</sup> In this case review, we aimed to report the problems in airway management of patients with MPS undergoing various surgical procedures.

## Case description

After ethical committee approval was obtained, we performed a retrospective review of patients with MPS who underwent surgical procedures at Hacettepe University Hospital between 2015 and 2019. Demographic, clinical, and airway management data were obtained from anesthesia charts in hospital records. For patients who underwent multiple surgeries, each encounter was analyzed as a separate event. Informed consent was obtained from all patients via phone calls and email.

We identified difficulties in airway management according to the signs of difficult intubation and difficult mask ventilation on the anesthesia charts. Difficult intubation was explained by the number of intubation attempts. Multiple

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intubation attempts were defined as difficult intubation. Difficult mask ventilation was defined for patients who could not be ventilated using the double-hand technique.

Thirty-seven of 359 patients with MPS who were followed in our hospital underwent surgery for various reasons between 2015 and 2019. Anesthesia and airway management data were available for 23 of these 37 patients. The 23 patients underwent surgery 31 times over 5 years. As a result, we included 31 surgical procedures (9 females, 22 males) in our case review.

The mean age of the patients was  $127.6 \pm 84.2$  (range, 7–348) months, and the mean body weight was  $24.9 \pm 17.6$  (range, 7–85) kg (mean  $\pm$  SD [minimum-maximum]). Maroteaux-Lamy syndrome (Type 6,  $n = 9$ ) and Morquio syndrome (Type 4,  $n = 9$ ) were the most common MPS types. Orthopedic surgeries were the most common surgery type ( $n = 9$ ).

Airway management and postoperative follow-up data are given in Table 1. Difficult mask ventilation and laryngeal mask airway (LMA) use were not encountered in patient records. Airway intervention was not required in five patients (four with tracheostomy, one was already intubated). Video laryngoscopy (VL) was the most common intubation method ( $n = 20$ ). All patients with difficult intubation ( $n = 10$ ) were aged over 36 months. Half of the patients with difficult intubation were adults ( $n = 2$ ) and teenagers ( $n = 3$ ).

Eight patients with difficult intubation were intubated using VL. One patient who was scheduled for emergency tracheotomy could not be intubated and underwent surgery (FONA: front of neck access) under mask ventilation. One patient with limited neck extension due to narrowness of the foramen magnum was intubated via awake fiberoptic bronchoscopy. There was no need for laryngoscopy in five patients because three had a tracheotomy and two were

**Table 1** Airway management and postoperative follow-up of cases.

	Toddler (0-36m) $n = 3$	Preschool (37-60 m) $n = 4$	School aged (61-144 m) $n = 12$	Teenager (145-216 m) $n = 9$	Adults (216+m) $n = 3$
<b>MPS Type</b>					
Hurler's (I) ( $n = 3$ )	3	0	0	0	0
Hunter (II) ( $n = 3$ )	0	0	1	2	0
Sanfilippo (III) ( $n = 7$ )	0	0	3	3	1
Morquio (IV) ( $n = 9$ )	0	1	7	0	1
Maroteaux-Lamy (VI) ( $n = 9$ )	0	3	1	4	1
<b>Airway device</b>					
ETT ( $n = 25$ )	3	4	10	5	3
LMA ( $n = 0$ )	0	0	0	0	0
FONA ( $n = 1$ )	0	0	0	1	0
Other ( $n = 5$ )	0	0	2 <sup>a</sup>	1 <sup>a</sup> , 2 <sup>b</sup>	0
<b>Intubation technique</b>					
DL ( $n = 4$ )	0	2	1	1	0
VL ( $n = 20$ )	3	2	9	4	2
FOB ( $n = 1$ )	0	0	0	0	1
FONA ( $n = 1$ )	0	0	0	1	0
N/A ( $n = 5$ )	0	0	2 <sup>a</sup>	1 <sup>a</sup> , 2 <sup>b</sup>	0
<b>Intubation Difficulty</b>					
Easy ( $n = 16$ )	3	2	7	3	1
Difficult ( $n = 10$ )	0	2	3	3	2
N/A ( $n = 5$ )	0	0	2 <sup>a</sup>	1 <sup>a</sup> , 2 <sup>b</sup>	0
<b>Mask difficulty</b>					
Easy ( $n = 26$ )	3	4	10	6	3
Difficult ( $n = 0$ )	0	0	0	0	0
N/A ( $n = 5$ )	0	0	2 <sup>a</sup>	1 <sup>a</sup> , 2 <sup>b</sup>	0
<b>Postoperative Extubation</b>					
Extubated ( $n = 22$ )	3	4	9	3	3
Intubated ( $n = 3$ )	0	0	1	2	0
N/A ( $n = 6$ )	0	0	2 <sup>a</sup>	4 <sup>a</sup>	0
<b>Postoperative Follow-up</b>					
Service ( $n = 14$ )	2	3	6	2	1
ICU ( $n = 17$ )	1	1	6	7	2

ETT, Endotracheal tube; LMA, Laryngeal Mask Airway; DL, Direct Laryngoscopy; VL, Videolaryngoscopy; FOB, Fiberoptic Bronchoscopy; FONA, Front of neck airway; ICU, Intensive Care Unit.

<sup>a</sup> Had a tracheotomy.

<sup>b</sup> Already intubated.



**Figure 1** A 4-year-old female patient with MPS-VI who had a short neck and kyphoscoliosis. Informed consent was obtained from the patient shown in the figure.

already intubated orally during admission to the operating room.

In the postoperative period, 17 patients were followed up in the intensive care unit (ICU). Most patients ( $n = 11$ ) were admitted to the ICU as extubated. One patient undergoing Bentall surgery was reintubated using VL due to acute respiratory dysfunction during their postoperative ICU stay. Elective tracheostomy was performed for this patient whose airway pressure was high. There was no other perioperative airway complication in patient records.

## Discussion

Anesthetic management can be challenging in many aspects in patients with MPS (Fig. 1). The risk of difficult intubation must always be considered due to macroglossia, short neck, hypertrophic tonsils, and adenoids, kyphoscoliosis, immobile jaw, narrowed nasal passages, and atlantoaxial instability.<sup>3</sup>

Similar to the literature, the incidence of difficult intubation increased with age in our patients.<sup>4</sup> All difficult intubations were seen in children aged over 3 years. In addition, the number of patients with tracheotomy and/or already intubated was higher at older ages. Studies show that, unlike intubation, the use of LMA and mask ventilation does not become more difficult with older age in patients with MPS.<sup>5</sup> In our case series, it was remarkable that difficult mask ventilation and LMA use were not encountered in any patient records. This may be due to missing data or the effective double-hand mask ventilation technique. In addition, there were very few patients in whom surgery could be performed using LMA. This could be another factor. The most important limitation of our case series is its retrospective, non-randomized and uncontrolled design.

According to our case series, VL seems to be a safe intubation method in patients with MPS. However, an experienced

anesthesiology team should always be ready with alternative plans during the perioperative management of these patients regardless of the choice of equipment. In these patients, regional anesthesia is also an alternative method for anesthesia management if it can be used.

## Conflicts of interest

The authors declare no conflicts of interest.

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This article, in whole or in part, has not been published elsewhere and is not being evaluated by any other journal. The data of the study were presented for the first time as a poster presentation at the Euroanaesthesia on Nov 28–30<sup>th</sup>, 2020.

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