Airway management in Hurler's syndrome: A persistent challenge for anaesthesiologists

INTRODUCTION

The difficulty in airway management of Hurler's syndrome or mucopolysaccharidosis (MPS) type I patients has been described as the worst.[1] These patients may develop serious complications under anaesthesia including airway obstruction leading to severe hypoxaemia, inability to ventilate or intubate and post-extubation problems.[2] Despite many potential benefits of using supraglottic devices for primary airway management in these patients over endotracheal tube (ETT) intubation, their usage has been limited. We at our centre decided to insert a Proseal laryngeal mask airway (PLMA) in awake state in a patient of Hurler syndrome to circumvent the problem of the difficult airway.

CASE REPORT

A 15-year-old male child diagnosed as a case of Hurler's syndrome with bilateral corneal haze was scheduled for keratoplasty of the right eye. He exhibited characteristic features of a severe form of MPS type I including short stature, facial dysmorphism, protruded abdomen and multiple joint deformity of limbs and complaint of obstructive sleep apnoea for which he was using nasal bilevel positive airway pressure (BiPAP) device for the last 5 years. There was no history of any other significant illness or anaesthetic exposure.

On examination, the child was found to be of normal intelligence, cooperative and with stable vital parameters. His airway examination revealed reduced mouth opening of 2.5 cm, large tongue, receding chin, thyromental distance of 4.5 cm, modified Mallampati class 4 and adequate range of neck movements [Figures 1 and 2]. On systemic examination, significant findings were systolic murmur in aortic area on auscultation and hepatosplenomegaly on abdominal palpation. Routine laboratory haematological examinations were unremarkable. Chest X-ray showed cardiomegaly and on echocardiogram, thick calcified aortic valve with mild mitral regurgitation was found. The X-ray of cervical spines ruled out atlantoaxial instability.

During pre-anaesthetic visit, the patient was explained about the necessity to perform awake supraglottic device insertion or awake fibreoptic bronchoscopy (FOB) guided intubation to which he agreed. He was

also asked to bring nasal BiPAP machine to the hospital on the day of surgery.

In the morning of surgery after appropriate duration of fasting, the patient was administered 0.1 mg of intramuscular glycopyrrolate injection 45 min before surgery, and topical preparation of airway was accomplished using lignocaine preparations. In operation theatre, standard anaesthetic monitors were attached, and baseline vitals were noted. Appropriate difficult airway cart was kept prepared. Intravenous (IV) midazolam 0.5 mg, ranitidine 25 mg and metoclopramide 5 mg were administered as pre-medication.

The patient was asked to insert the PLMA number 2½ on his own in the manner of swallowing a lollipop. The patient complied and thereafter the cuff of the PLMA was inflated with 10 ml air and connected to Bains circuit. Appropriate placement of PLMA was confirmed by bag movement with respiration, capnogram and easy passage of the gastric tube through drain channel. For analgesia, fentanyl 25 μg IV was administered, and anaesthesia was induced with thiopentone sodium 75 mg given slow IV. After confirming the ease of ventilation with a gentle, positive pressure by bag, injection vecuronium bromide 2 mg was given. Anaesthesia was maintained with sevoflurane (<2%) in oxygen-nitrous mixture (33:66). Surgery was completed uneventfully in 2 h. Anaesthetic gases were discontinued before reversal of neuromuscular blockade with injection glycopyrrolate 0.2 mg and neostigmine 1.25 mg. After 10 min, the patient was fully awake and obeying commands. PLMA was then taken out by the patient himself, and he was then shifted to PACU with stable vitals.

DISCUSSION

Hurler's syndrome is a rare inherited metabolic disorder characterised by the widespread progressive accumulation of unmetabolised glycosaminoglycans (GAGs) within the cells of various organ systems. The most common and difficult task encountered by anaesthesiologists in these patients during perioperative period is the establishment of a patent airway.[3] The various anatomical changes in airway due to deposition of GAGs lead to difficulty in mask ventilation, laryngoscopy and intubation. The use of LMA in these patients has been limited as an aid to FOB intubation or rescue device after failed intubation attempts. The LMA devices are particularly useful for airway management in MPS patients because of multiple reasons. First, MPS patients are at risk of upper airway obstruction following the induction of anaesthesia. The LMA provides a patent airway from exterior to rima glottidis. Second, MPS patients can have atlantoaxial joint instability which precludes neck hyperextension during airway management.[4] LMA can be inserted in neutral position and thus avoids catastrophic neurological consequences. In addition, MPS patients are at increased risk of post-operative airway oedema due to multiple factors including multiple attempts at intubation, history of obstructive sleep apnoea (OSA). These can be avoided by elective use of LMA as it is less invasive compared to ETT. Fourth, the children with MPS often present for multiple surgical procedures; the successful usage in one setting might influence the decision to use LMA in subsequent general anaesthesia.

Despite these advantages, there is no agreement about its elective use in MPS. The main concerns while choosing LMA as primary airway device in

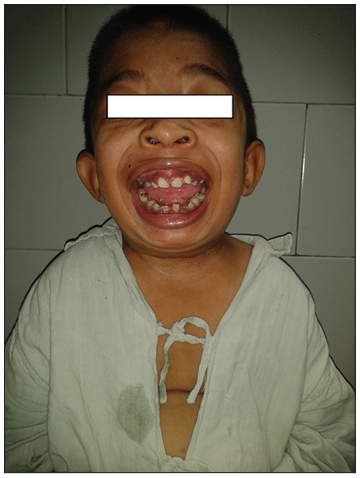

Figure 1: Patient of mucopolysaccharidosis type I showing decreased mouth opening and large tongue

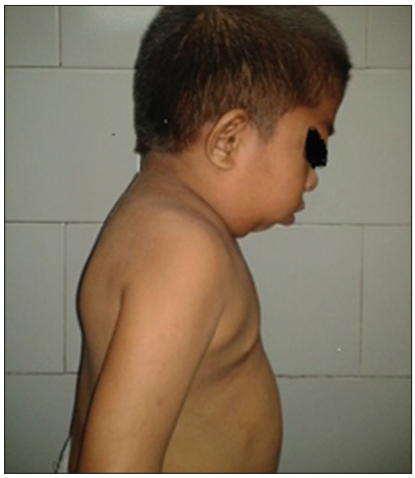

Figure 2: Lateral view picture of patient

patients of the difficult airway is failure of proper device placement and risk of unsuccessful ventilation through the device. There are no validated parameters which can predict the success of LMA insertion. Saito *et al*. found four independent risk factors for difficult ventilation via a supraglottis device: male sex, age >45 years, short thyromental distance and limited neck movement.[5] Our patient had two of these risk factors; hence, we decided to insert PLMA in awake patient after topicalisation of airway. This approach also mitigated the risk of developing airway obstruction following sedation or induction of anaesthesia. Awake insertion of other supraglottic devices has also been reported for difficult airway management.[6,7]

We used sodium thiopentone for induction of anaesthesia as the airway of the patient including posterior pharyngeal wall was adequately anaesthetised by prior topicalisation with lignocaine nebulisation and lignocaine gargle which obviated the possibility of hyperreflexia associated with the use of thiopentone.

Other options for airway management would have been securing airway with awake fibreoptic intubation. We decided to use PLMA as primary airway device as awake passage of LMA is a less noxious stimulus and requires less expertise compared to awake FOB intubation. Extubation is smooth compared to ETT insertion. In ophthalmic surgeries, there is additional desirable benefit of less increase in intraocular pressure and haemodynamic parameters[8] over ETT.

CONCLUSION

The laryngeal mask family devices are useful in handling difficult airway situations as in the present case. Since there are no validated parameters which can predict the successful placement of supraglottic devices, awake insertion would be safer compared to insertion under anaesthesia.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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