Safe intubation in Morquio-Brailsford syndrome: A challenge for the anesthesiologist

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Abstract

Morquio-Brailsford syndrome is a type of mucopolysaccharidoses. It is a rare disease with features of short stature, atlantoaxial instability with risk of cord damage, odontoid hypoplasia, pectus carinatum, spine deformities, hepatomegaly, and restrictive lung disease. Neck movements during intubation are associated with the risk of quadriparesis due to cervical instability. This, along with the distortion of the airway anatomy due to deposition of mucopolysaccharides makes airway management arduous. We present our experience in management of difficult airway in a 3-year-old girl with Morquio-Brailsford syndrome posted for magnetic resonance imaging and computerized tomography scan of a suspected unstable cervical spine. As utmost sagacity during intubation is required, the child was intubated inside operation theatre in the presence of experienced anesthesiologists and then shifted to the peripheral location. Intubation was done with an endotracheal tube railroaded over a pediatric fibreoptic bronchoscope passed through the lumen of a classic laryngeal mask airway, keeping head in neutral position.

Key words: Cervical instability, classic laryngeal mask airway, cord damage, difficult airway

Introduction

Morquio-Brailsford syndrome is a type of mucopolysaccharidoses which leads to abnormal deposition of keratin sulfate in tissues, distorting upper airways.[1,2] It is classified as a rare disease with a prevalence of about one in 2,50,000.[3] As there is a risk of atlantoaxial subluxation and quadriparesis during intubation, safe anesthesia in Morquio syndrome is a formidable challenge.[4] We report the anesthetic management of a 3-year-old child with Morquio syndrome posted for magnetic resonance imaging (MRI) and computerized tomography (CT) scan of cervical spine.

Case Report

A 3-year-old child weighing 9kg diagnosed with Morquio-Brailsford syndrome was posted for MRI and CT of cervical spine after she had developed sudden weakness of the upper and lower limbs with inability to walk. She was diagnosed as Morquio syndrome at the age of 1 year, with a delay in achieving developmental milestones. She had short neck, pectus carinatum thoracolumbar kyphoscoliosis with a suspected unstable cervical spine based on clinical deterioration and cervical spine X-ray [Figure 1]. Cardiovascular, abdominal, and hematological examinations were unremarkable. Airway assessment could not be done as the child was uncooperative. A high-risk informed consent was taken in view of difficult airway and possible cervical instability which could worsen cord compression during the procedure.

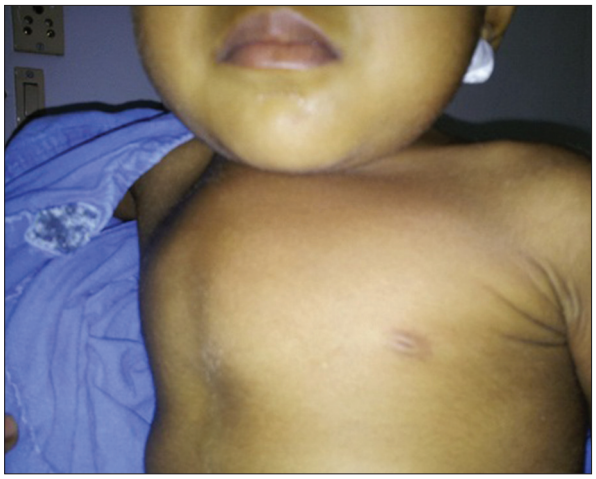

Figure 1: Child with Morquio syndrome having short neck and pigeon chest

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Intubation was done inside the operation theatre (OT) in presence of senior anesthesiologists and then shifted to the MRI suite. Different sizes of endotracheal tubes (ETTs), laryngeal mask airways (LMAs), proseal LMA, and pediatric fibreoptic bronchoscope were kept in the difficult airway cart. An intravenous (IV) access was secured and child was shifted to OT after administration of 0.1mg IV glycopyrrolate and 0.5mg IV midazolam. Baseline vitals were heart rate of 150/min, blood pressure 94/60mmHg and oxygen saturation (SpO_2) of 99%. Anesthesia was induced with titrating doses of propofol along with 3% sevoflurane in 100% oxygen; ensuring spontaneous ventilation was maintained while monitoring SpO_2 and end tidal carbon dioxide ($EtCO_2$). A total of 10μg IV fentanyl was given and after adequate depth of anesthesia, a size 2 classic LMA which was shaped like a proseal LMA using stylet was gently inserted with manual in-line stabilization (MILS) [Figure 2]. LMA position was confirmed by adequate chest expansion and presence of capnogram. A right-angle tracheal tube connector with seal was attached to the LMA [Figure 3]. This allowed fibreoptic bronchoscope to be inserted through the LMA while the child was ventilated with 100% oxygen and 2%-3% sevoflurane.

A 4.0 mm uncuffed portex ETT was railroaded over the 2.4 mm Outer Diameter (OD) pediatric bronchoscope. Then the bronchoscope was introduced through the lumen of LMA into the trachea and the ETT was advanced over the bronchoscope. On connecting the breathing circuit to the ETT, though a typical capnogram was present but visualization through the fibreoptic scope showed that the ETT tip was just above the glottis, even on introducing the ETT to the maximum depth possible in the presence of right-angle tracheal tube connector. Eventually, the right-angle tracheal tube connector was removed and reintubation was done using similar technique, now ensuring that the tip of ETT was placed about 2cm above carina by fibreoptic visualization.

The ETT with LMA was fixed as a single unit after confirming the ETT tip position by visualization and auscultation. Care

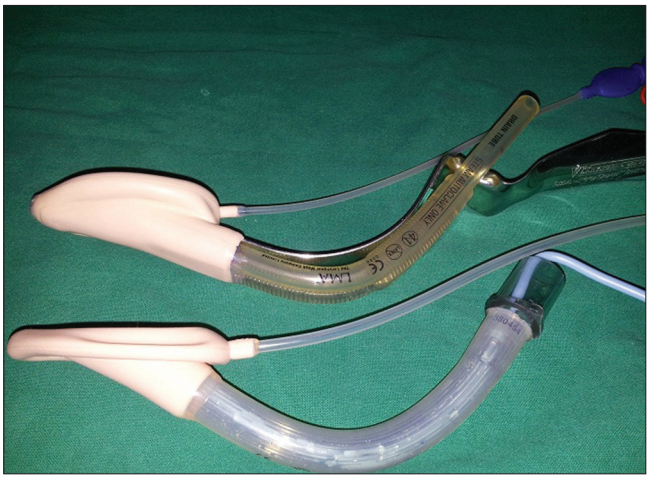

Figure 2: Shape of classic LMA changed like a PLMA for better position at laryngeal aperture after insertion in a neutral head position

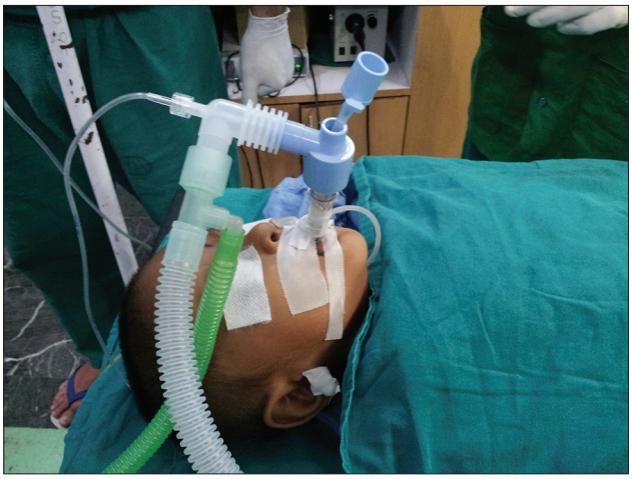

Figure 3: Classic LMA with ETT and the right-angled connector

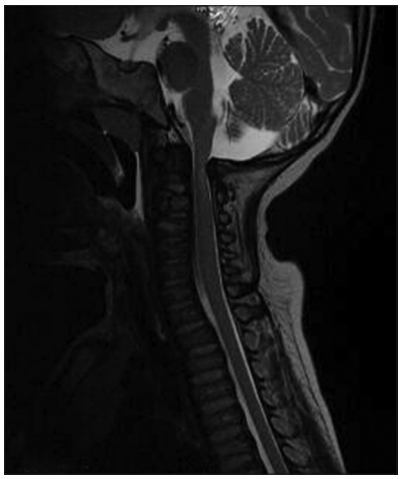

Figure 4: MRI of the child showing cervical cord compression

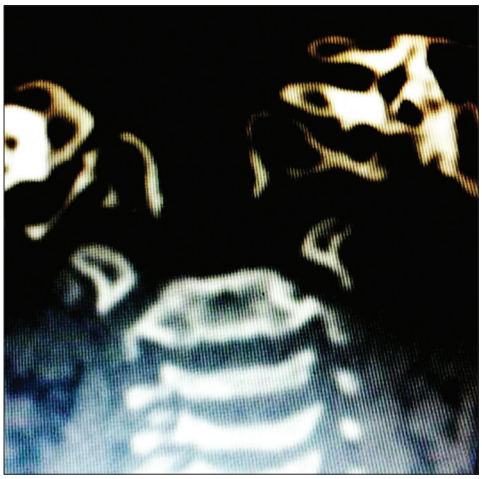

Figure 5: CT of the child with nonossification of the dens

was taken to avoid kinking of the ETT at the point of exit from LMA. Throughout the procedure, saturation was maintained between 96% and 100%. A total of 3mg IV atracurium was then given and child was shifted to MRI suite on oxygen trolley with monitors. Anesthesia during the MRI and CT was uneventful, after which child was shifted to pediatric intensive care unit and was extubated in ICU. MRI showed cord compression involving the cervicomedullary junction, hypoplastic split atlas, and posterior atlantooccipital fusion [Figure 4]. CT confirmed hypoplasia of C1 vertebra, split atlas, and nonossification of the dens [Figure 5]. Child was then put on a cervical collar and eventually discharged.

Discussion

Morquio syndrome is characterized by odontoid hypoplasia, atlantoaxial instability with risk of quadriparesis, distorted upper airway anatomy, spine deformity, cardiorespiratory, and liver abnormalities.[1-4]

Our patient was posted for MRI in a closed MRI suite which is a remote location. We chose to secure airway inside OT in the presence of experienced anesthesiologists. Intubation was done due to the possibility of displacement of LMA inside the MRI console where our accessibility to the head end is limited. Head was kept in neutral position during intubation with MILS as quadriplegia after anesthesia has been reported.[5] It has also been recommended to ensure spontaneous ventilation till intubation in these children as bleeding from abnormally deposited soft tissues in airway along with airway collapse may lead to extremely difficult intubation, so atracurium was administered postintubation.[6]

We chose LMA as a conduit for fibreoptic-guided intubation as was done by Mourao *et al.*,[6] after their initial attempt to intubate a child with Morquio syndrome using fibreoptic endoscope was futile due to mucuous membrane hypertrophy of oropharynx. LMA with ETT *in situ* was kept as a single unit, as LMA provides a backup in case of ETT dislodgement, especially in scenarios where our accessibility to head end is restricted.[7] We could not use a proseal LMA inside the MRI suite as the metal coil could produce a large black hole in the image and thus lead to image deterioration.[8] However, proseal LMA would be useful alternative in other situations.

The use of fiberscope for intubation without any airway conduit may be extremely difficult in Morquio syndrome. These patients may have a "hanging epiglottis" making the passage of fiberscope toward the glottis impossible.[3]

The LMA classic was bent by us to resemble a proseal LMA, as it would help in better positioning of LMA over the laryngeal aperture in the absence of sniffing position.[9] The introduction of stylet in a classic LMA to change its shape has been recommended.[9] However, our notion to use a right-angle tracheal tube connector with seal to allow ventilation through LMA, while fibreoptic-guided intubation was attempted had a drawback. The length of the connector allowed ETT to be maximally inserted to just above the glottis, even though on connecting the breathing circuit to the ETT in that position gave an acceptable capnogram. Thus, we want to emphasize that when LMA is used as a conduit for intubation, appearance of a typical capnogram should not assure the anesthesiologist of optimal ETT position. It has to be confirmed visually using fibreoptic bronchoscope. We chose a smaller ETT as recommended in these patients as they have mucopolysaccharide deposition in their airway along with subglottic stenosis.[2,3]

Thus, safe intubation is a challenge in Morquio syndrome and a "cannot ventilate, cannot intubate" scenario can be encountered by anesthesiologist due to the possibility of total airway collapse because of laxity of the trachea and main bronchi.[2,4,10] We must also be aware of other comorbidities in them like aortic regurgitation, restrictive lung disease, and hepatosplenomegaly.[3]

All the precautions taken by us during intubation were essential as the MRI showed cord compression in the child.

Conclusion

LMA as a conduit for fibreoptic-guided intubation is helpful in Morquio syndrome, where the movement of fibreoptic scope may be impeded by abnormal soft tissue of the airway. In situations where proseal LMA is unavailable or cannot be used, the shape of a classic LMA can be modified for optimal LMA placement in a neutral head position. Intubation has to be done by experienced anesthesiologists preferably inside OT and not in remote locations.

References

1. Walker PP, Rose E, Williams JG. Upper airway abnormalities and tracheal problems in Morquio's disease. Thorax 2003;58:458-59.
2. McLaughlin AM, Farooq M, Donnelly MB, Foley K. Anaesthetic considerations of adults with Morquio's syndrome: A case report. BMC Anaesthesiol 2010;10:2.
3. Kadic L, Driessen JJ. General anaesthesia in an adult patient with Morquio syndrome with emphasis on airway issues. Bosn J Basic Med Sci 2012;12:130-3.
4. Tobias JD. Anesthetic care for the child with Morquio syndrome: General versus regional anesthesia. J Clin Anesth 1999;11:242-6.
5. Tzanova I, Schwarz M, Jantzen JP. Securing the airway in children with the Morquio-Brailsford syndrome. Anaesthesist 1993;42:477-81.

6. Mourao J, Chow E, Tavares J. Anesthetic implications of Morquio syndrome: Impossible intubation though fiberoptic endoscopy. Eur J Anaesthesiol 2007;24:191.
7. Kuzma PJ, Calkins MD, Kline MD, Karan SM, Matson MD. The anesthetic management of patients with multiple pterygium syndrome. Anesth Analg 1996;83:430-32.
8. Dorsch JA, Dorsch SE. Equipment for the magnetic resonance imaging environment. In: Dorsch JA, Dorsch SE, editors. Understanding anesthesia equipment. 5th ed. Lippincott Williams and Wilkins; 2008. p. 871-82.
9. Dorsch JA, Dorsch SE. Supraglottic airway devices. In: Dorsch JA, Dorsch SE, editors. Understanding anesthesia equipment. 5th ed. Lippincott Williams and Wilkins; 2008. p. 461-518.
10. Kurdi MS, Deshpande SS. Mucopolysaccharidoses: An adventurous anaesthetic encounter. Indian J Anaesth 2008;52:453-8.

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