

Anaesthetic implications in a patient with Morquio A syndrome

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Abstract

In patients with Morquio A syndrome, almost all the systems of body are affected and this has a bearing on their anaesthetic management. Herein, we describe the management of child with Morquio A syndrome with atlanto-axial dislocation who underwent posterior fixation under general anaesthesia.

Key words: Anaesthesia, atlanto-axial dislocation, Morquio A syndrome, mucopolysaccharidosis

INTRODUCTION

Mucopolysaccharidosis (MPS) refer to an inherited group of diseases caused by deficiency of different lysosomal enzymes responsible for degradation of glycosaminoglycans. Intracellular accumulation of these partially metabolized glycosaminoglycans hinder normal functioning of the tissues involved. Morquio A syndrome (MPS IV-A) is an autosomal recessive MPS caused by deficiency of N-acetyl galactosamine-6-sulfatase enzyme resulting in deposition of large amount of keratan sulphate and chondroitin sulphate in various tissues of body.^[1] The manifestations are primarily due to mesenchymal abnormalities and mental intelligence is normal.

Nearly every system of body may be involved affecting anaesthetic preparation and management. Cervico-vertebral junction anomaly, deposition of mucopolysaccharides in various airway structures and

involvement of temporo-mandibular joints result in a difficult airway state.

We hereby report a case of MPS IV-A with atlanto-axial dislocation (AAD) scheduled for transoral odontoidectomy and posterior fixation.

CASE REPORT

A 6-year-old female child known case of MPS IV-A, weighing 14 kg was admitted with history of gradually increasing weakness of all limbs for last 5 years and increased frequency of micturition for last 7 months. Patient had a complete cessation of growth for last 2 years and at present, height was 120 cm. She had normal developmental milestones and intellect with no history of obstructive sleep apnoea.

On examination, the child had a short stature with dolichocephaly, short neck, kyphosis at thoraco-lumbar region, pectum excavatum with prominent costal margins, genu valgum and equino valgum [Figure 1]. Central nervous system examination revealed a power of 4/5 in all limbs with spasticity and brisk reflexes. Computed tomography (CT) scan of craniovertebral junction showed AAD with obliteration of normal cervical lordosis with occipitalisation of C1 posterior arch. Patient was planned for occipitocervical fusion with sublaminar wiring.

Access this article online	
Quick Response Code:	Website: www.jnaccjournal.org
	DOI: 10.4103/2348-0548.154238

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Preanaesthesia assessment revealed a systolic murmur at apical area on auscultation which was confirmed by echocardiographic finding of myxomatous degeneration of heart valves with moderate mitral regurgitation with normal left ventricular function. Airway examination revealed adequate mouth opening with Mallampati grade II with large tongue and short neck. A cervical collar was placed around neck. X-ray neck revealed enlarged soft tissue shadow in prevertebral area with apparently normal laryngeal and tracheal contour [Figure 2]. Possibility of difficulty in securing airway was anticipated in view of presence of congenital AAD, large tongue, short neck, soft tissue deposit and cervical collar in situ. Preoperative complete haemogram, renal function test, lung fields on chest X-ray were normal. Bedside pulmonary function tests (PFT) of the patient were equivocal as the child could not cooperate to maximum ability. We did not administer infective endocarditis (IE) prophylaxis, as there is no recommendation regarding IE prophylaxis in a patient with congenital mitral regurgitation, according to the guidelines.^[2]

Patient was premedicated with intravenous glycopyrrolate 0.1 mg, 10 min before induction of anaesthesia. Difficult airway cart was prepared in view of anticipated difficult intubation. It comprised oral airways, stylets and bougie, laryngoscopes with Miller and Macintosh blades, different sizes of endotracheal tubes, laryngeal mask airway (LMA), paediatric fiberoptic bronchoscope, glidescope, equipment for cricothyrotomy and surgical tracheostomy. The neurosurgeon was present in the operating room and was asked to be prepared for performing tracheostomy, in case need arises. Anaesthesia was induced with sevoflurane following which a peripheral venous access was secured. Intravenous (iv) fentanyl 2 µg/kg was administered and after ensuring adequate mask ventilation, rocuronium 10 mg iv was given. Patient was intubated successfully with 5.5 mm cuffed, oral, flexometallic tracheal tube with the help of paediatric

fiberoptic bronchoscope. Maintenance of anaesthesia was done with sevoflurane in air/oxygen mixture and intermittent boluses of rocuronium.

Monitoring included electrocardiography, pulse oximetry, invasive arterial blood pressure, urinary output and endtidal CO₂ and anaesthetic gas measurement. Heart rate was kept between 90 and 100/min to decrease the regurgitant flow across mitral valve. Patient was carefully positioned prone for surgery. Surgery lasted for an uneventful period of 4 hours.

At end of posterior fixation, patient was turned supine and a check laryngoscopy was performed to assess the laryngeal view. A complete glottic view was visualized [Figure 3] and subsequently, neuromuscular blockade was reversed.^[15] When child became fully conscious with resumption of normal breathing, trachea was extubated without any complication. She was then transferred to the neuro-intensive care unit (NICU), where she recovered uneventfully and was monitored overnight. Duration of intensive care unit (ICU) stay and hospital stay was 1 and 5 days, respectively. At the time of discharge, spasticity had decreased with power 4/5 power in all limbs and glasgow outcome scale (GOS) was 4.

DISCUSSION

The multisystemic involvement^[3-7] in a patient with MPS IV-A makes thorough preoperative assessment imperative. Based on the findings [Table 1], adequate preparation should be done to avoid any sudden crisis.

Several authors have highlighted airway abnormalities in these patients. Not only it is difficult to intubate trachea but also structural changes may lead to laryngeal stenosis and distorted lower airway resulting in ventilation problems. History of obstructive sleep apnoea (OSA) has been found to be associated with



Figure 1: Image of the patient showing pectum excavatum with prominent costal margins and short neck



Figure 2: X-ray neck showing soft tissue mass in prevertebral area with normal laryngeal and tracheal contour

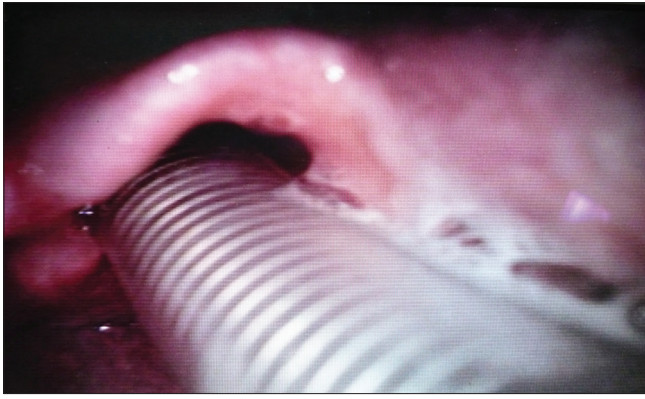


Figure 3: Glidescopic view clearly showing full view of glottis

Table 1: Multisystemic involvement in Morquio A syndrome

System
Cardiac
Valvular regurgitant or stenotic lesions, reduced myocardial compliance, hypertension, coronary disease, arrhythmias, cardiac failure
Respiratory
Macroglossia, dental abnormalities, short neck, subglottic narrowing, hanging epiglottis, oro-pharyngeal deposits, large adenoids/ tonsils, restricted opening of temporomandibular joints, restrictive and even obstructive respiratory pattern
Vertebral
Cranio-vertebral junction anomaly, odontoid hypoplasia, cervical cord compression
Skeletal
Short stature, short diaphysis, curved metaphysis, poorly developed epiphysis, wide acetabula, hypoplastic femoral heads, pigeon chest, genu valgum, equino valgum
Others
Prognathism, hepatomegaly, corneal clouding, glaucoma, degenerative retinal lesions, hearing loss

difficult airway.^[8] Such children are prone to respiratory obstruction during induction of anaesthesia and post extubation. The incidence of difficult intubation in patients with Morquio syndrome varies from 0–50%.^[9–11] In a retrospective review of 17 patients of MPS, overall incidence of difficult intubation was 25%; highest in MPS VI (86.7%) and 0% in MPS IV.^[9] However, small number of MPS IV patients in study population may be responsible for this finding.

The decision regarding airway management can be based on radiology findings. In our patient, the preoperative X-ray neck did not show any evidence of laryngeal or tracheal stenosis. Another modality like preoperative multidetector computed tomography film can also

delineate airway anatomy and assist in decision-making regarding intubation and extubation.^[12] Recently, use of ultrasound for assessment of airway anatomy has also been highlighted.^[12] This simple method not only allays the harmful effects of radiation but can also give a real time picture.^[13]

Rigid instruments like glidescope, lighted stylet etc., are preferred for intubation in these patients as they can displace soft tissue more easily than flexible fibrescope.^[10] We opted for flexible bronchoscope as the first choice as it causes no movement at atlanto-axial joint. Several authors recommend awake fibreoptic intubation and avoiding muscle relaxants for this purpose. As our patient was a child, we did not resort to awake intubation. Though the intellect is normal in these patients, behavioural abnormalities like anxiety and depression are known to occur.^[14] We were successful in intubating the patient with fibreoptic bronchoscope under anaesthesia in the first attempt. Literature search reveal reports of augmented difficult intubation once the patient is paralysed. Loss of muscular tone and floppy soft tissues after administration of muscle relaxants, makes fibreoptic intubation after induction of anaesthesia even more difficult.^[7,15] Supraglottic airway device such as I-gel has also been used successfully used as a channel for fibreoptic-guided tracheal intubation in a patient with Hunter syndrome.^[16] The ability to mask ventilate and back-up of glidescope, cricothyrotomy and tracheostomy made us go ahead with muscle relaxation. The choice of muscle relaxant is debatable and many will straightway negate the use of rocuronium in such cases and will prefer to use succinylcholine if at all. But we want to emphasize that succinylcholine is contraindicated in such cases of upper motor neuron paresis and should not be used. Life-threatening hyperkalaemia may complicate the anaesthetic management and even cardiac arrest may ensue. However, another safer alternative could have been fibreoptic intubation under sevoflurane induction along with ‘spray as you go technique’ omitting the administration of muscle relaxant. The inhalational induction also allows rapid awakening in case intubation fails. Fibreoptic intubation under dexmedetomidine sedation with preservation of spontaneous breathing is another alternative. Fibreoptic intubation under influence of muscle relaxants requires expertise and should only be attempted if one is proficient and has back-up help. In a difficult airway situation, we should use the technique we are comfortable with.

Posterior fixation renders the airway more difficult. So at the end of surgery, we did a check laryngoscopy with glidescope to assess the laryngeal view [Figure 3]. An indirect glottis visualization with help of videolaryngoscope can help in assessing the airway at

time of extubation in cases where posterior fixation of cervical spine has been done. If reintubation is required in cases where extension of cervical spine is absent, at least we can be sure of being able to visualize glottis with videolaryngoscopes. With additional help of stylet/bougie, we expect to be able to reintubate 100% of patients.

Absence of OSA, uneventful intubation at start and visibility of full glottis on check video laryngoscopy gave an assurance that we can go ahead with extubation of trachea. Patients may require continuous positive airway pressure in the postoperative period and should be closely watched for any desaturation episodes.

CONCLUSION

MPS IV-A affects multiple systems and patient should be assessed in total. Then, depending on the expertise and resource availability, an individual anaesthetic plan can be chalked out for these patients. Decision to extubate the patient can be reinforced by a check laryngoscopy with help of video laryngoscope.

REFERENCES

1. Singh J, Di Ferrante N, Niebes P, Tavella D. N-acetylgalactosamine-6-sulphate sulphatase in man. Absence of the enzyme in morquio disease. *J Clin Invest* 1976;57:1036-40.
2. Wilson W, Taubert KA, Gewitz M, Lockhart PB, Baddour LM, Levison M, *et al.* American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee; American Heart Association Council on Cardiovascular Disease in the Young; American Heart Association Council on Clinical Cardiology; American Heart Association Council on Cardiovascular Surgery and Anesthesia; Quality of Care and Outcomes Research Interdisciplinary Working Group. Prevention of infective endocarditis: Guidelines from the American Heart Association: A guideline from the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee, Council on Cardiovascular Disease in the Young, and the Council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, and the Quality of Care and Outcomes Research Interdisciplinary Working Group. *Circulation* 2007;116:1736-54.
3. Braunlin EA, Harmatz PR, Scarpa M, Furlanetto B, Kampmann C, Loehr JP, *et al.* Cardiac disease in patients with mucopolysaccharidosis: Presentation, diagnosis and management. *J Inherit Metab Dis* 2011;34:1183-97.
4. Algahim MF, Almassi GH. Current and emerging management options for patients with morquio A syndrome. *Ther Clin Risk Manag* 2013;9:45-53.
5. Walker PP, Rose E, Williams JG. Upper airway abnormalities and tracheal problems in morquio's disease. *Thorax* 2003;58:458-9.
6. Muenzer J. Overview of the mucopolysaccharidosis. *Rheumatology (Oxford)* 2011;50 Suppl 5:v4-12.
7. Walker R, Belani KG, Braunlin EA, Bruce IA, Henrik H, Harmatz PR, *et al.* Anaesthesia and airway management in mucopolysaccharidosis. *J Inherit Metab Dis* 2013;36:211-9.
8. Apfelbaum JL, Haqberg CA, Caplan RA, Blitt CD, Connis RT, Nickinovich DG, *et al.* American society of anesthesiologists task force on management of the difficult airway. Practice guidelines for management of the difficult airway: An updated report by the American society of anesthesiologists task force on management of the difficult airway. *Anesthesiology* 2013;118:251-70.
9. Frawley G, Fuenzalida D, Donath S, Yaplito-Lee J, Peters H. A retrospective audit of anesthetic techniques and complications in children with mucopolysaccharidoses. *Paediatr Anaesth* 2012;22:737-44.
10. Theroux MC, Nerker T, Ditro C, Mackenzie WG. Anesthetic care and perioperative complications of children with Morquio syndrome. *Pediatr Anesth* 2012;22:901-7.
11. Walker RW, Darowski M, Morris P, Wraith JE. Anaesthesia and mucopolysaccharidoses. A review of airway problems in children. *Anaesthesia* 1994;49:1078-84.
12. Ingelmo PM, Parini R, Grimaldi M, Mauri F, Romagnoli M, Taqliabue G, *et al.* Multidetector computed tomography (MDCT) for preoperative airway assessment in children with mucopolysaccharidosis. *Minerva Anestesiol* 2011;77:774-80.
13. Marciniak B, Fayoux P, Hébrard A, Krivosic-Horber R, Engelhardt T, Bissonnette B. Airway management in children: Ultrasonography assessment of tracheal intubation in real time? *Anesth Analg* 2009;108:461-5.
14. Davison JE, Kearney S, Horton J, Foster K, Peet AC, Hendriksz CJ. Intellectual and neurological functioning in Morquio syndrome (MPS IVa). *J Inherit Metab Dis* 2013;36:323-8.
15. 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.
16. Michalek P, Hodgkinson P, Donaldson W. Fiberoptic intubation through an I-Gel supraglottic airway in two patients with predicted difficult airway and intellectual disability. *Anaesth Analg* 2008;106:1501-4.

How to cite this article: Mahajan C, Adarsha KN, Jena BR, Gupta D, Srivastava S. Anaesthetic implications in a patient with Morquio A syndrome. *J Neuroanaesthesiol Crit Care* 2015;2:130-3.

Source of Support: Nil, **Conflict of Interest:** None declared.