International Journal of Medical Anesthesiology

E-ISSN: 2664-3774
P-ISSN: 2664-3766
www.anesthesiologypaper.com
IJMA 2023; 6(1): 16-19
Received: 09-10-2022
Accepted: 15-11-2022

Dr. Roly Mishra
FIACTA Fellow, Department of Cardiac Anaesthesia, Fortis Escorts Heart Institute Delhi Ex-DNB student, Department of Anaesthesia and Pain Management, Sir H.N Reliance Foundation Hospital and Research Centre, Mumbai, Maharashtra, India

Dr. Vidhya N Deshmukh
Senior Consultant, Anaesthesiologist, Department of Anaesthesia and Pain Management, Sir H.N. Reliance Foundation Hospital and Research Centre, Mumbai, Maharashtra, India

Dr. Rashmi D Gujaran
Consultant Anaesthesiologist, Department of Anaesthesia and Pain Management, Sir H. N. Reliance Foundation Hospital and Research Centre, Mumbai, Maharashtra, India

Corresponding Author:
Dr. Roly Mishra
FIACTA Fellow, Department of Cardiac Anaesthesia, Fortis Escorts Heart Institute Delhi Ex-DNB student, Department of Anaesthesia and Pain Management, Sir H.N Reliance Foundation Hospital and Research Centre, Mumbai, Maharashtra, India

Anaesthetic challenges and perioperative management of a child with morquio syndrome posted for atlanto axial fusion and decompression: A case report

Dr. Roly Mishra, Dr. Vidhya N Deshmukh and Dr. Rashmi D Gujaran

DOI: https://doi.org/10.33545/26643766.2023.v6.i1a.367

Abstract
Morquio syndrome, also described as Mucopolysaccharidoses (MPS) type IV A in literature, are rare progressive and autosomal recessive lysosomal storage diseases characterised by deficiency of enzymes N- acetyl-galactosamine-6-sulphate sulphatase and beta-galactosidase that decreases the catabolism of the glycosaminoglycans (GAGs) that cause accumulation of glycosaminoglycans in soft tissue, bone and cartilage giving rise to severe skeletal dysplasias including critical pectus carinatum, kyphoscoliosis, odontoid hypoplasia resulting in notable atlanto-axial instability, cervical stenosis, other joint deformities and dwarfism. It also poses major airway challenges as there is limited mouth opening, deformation of the oropharyngeal and laryngeal tissues, hypertrophied tonsils adenoids resulting in marked sleep apnea and macroglossia. Keratan sulphate (predominant GAG) accumulates in the hyaline cartilage of the trachea giving rise to tracheal stenosis and tracheomalacia. Pulmonary, cardiac and other vital organs dysfunction also adds to the risk of anesthesia related complications. In this article, authors will discuss the case of a 9 years old female child with Morquio syndrome and the successful management of airway and general anaesthesia challenges they faced.

Keywords: Morquio syndrome, Mucopolysaccharidoses (MPS) GAG

Introduction
Patients with MPS have multiple comorbidities, many of which require surgical interventions. Because MPS is associated with specific phenotypic facial and airway characteristics, substantial challenges for perioperative airway management may be expected. Besides various airway issues, patients with MPS have other comorbidities that may have an impact on ventilation. Specifically, they frequently have restrictive or obstructive lung disease, recurrent lung infections, and obstructive sleep apnea. Propensities for bronchospasm and oxyhemoglobin desaturation may complicate airway management in MPS patients. Furthermore, skeletal dysplasia such as atlantoaxial instability, spinal cord compression, limited neck mobility, pectus carinatum, and scoliosis is common [1].

The MPS group involves a huge range of clinical manifestations with differences in life span and quality of living. All types are autosomal recessive inherited except type II which is X-linked; all forms are characterized by distinct somatic manifestations but not subjects with type III (Sanfilippo), progressive mental retardation is invariably present in severe forms of types I, II (Hurler, Hunter), and VIII (Sly). In Type III, due to the massive central nervous system (CNS) involvement, can be affected by severe mental retardation as well as aggressive behavior, hyperactivity, sleep problems, and loss of the capacity to perceive dangers [2].

Despite of the severity of their somatic presentation types, IV and VI do not have mental impairment. It is observed that patients with severe somatic affections may show a higher scale of neurological defects than they actually have, due to the language disturbance because of decreased hearing, enlarged tongue, poor vision, reduced manual skills, results of joint stiffness, and respiratory insufficiency [3].

Table 1: Classification of mucopolysaccharidoses with the deficient enzymes and the Accumulated Glycosaminoglycans

Mucopolysaccharidoses Disease	Deficient Enzyme	Accumulated Glycosaminoglycans
Mucopolysaccharidoses I: IH as Hurler syndrome, IH/S as Hurler-Scheie syndrome, and IS as Scheie syndrome	a-L-iduronidase	Dermatan sulfate, Heparan sulfate
Mucopolysaccharidoses II: Hunter syndrome	Iduronate sulfate sulfatase	Dermatan sulfate, Heparan sulfate
Mucopolysaccharidoses III A-D: Sanfilippo syndrome	A: Heparan-S-sulfaminidase, B: N-Acetyl-a-d-glucosaminidase, C: Acetyl-Co-A glucosaminidase, D: N-Acetylglucosidase N-acyltransferase	Heparan sulfate
Mucopolysaccharidoses IV A, B: Morquio syndrome	A: Galactosamine-6-sulfate sulfatase, B: Galactosamine-6-sulfate sulfatase	A: Keratan sulfate, Chondroitin 6-sulfate; B: Keratan sulfate, Chondroitin 6-sulfate
Mucopolysaccharidoses VI: Maroteaux Lamy syndrome	N-acetyl-galactosamine a-4-sulfate sulfatase	Dermatan sulfate
Mucopolysaccharidoses VII: Sly syndrome	P-glucuronidase	Dermatan sulfate, Heparan sulfate, Chondroitin sulfate
Mucopolysaccharidoses IX	Hyaluronidase 1	Hyaluronan

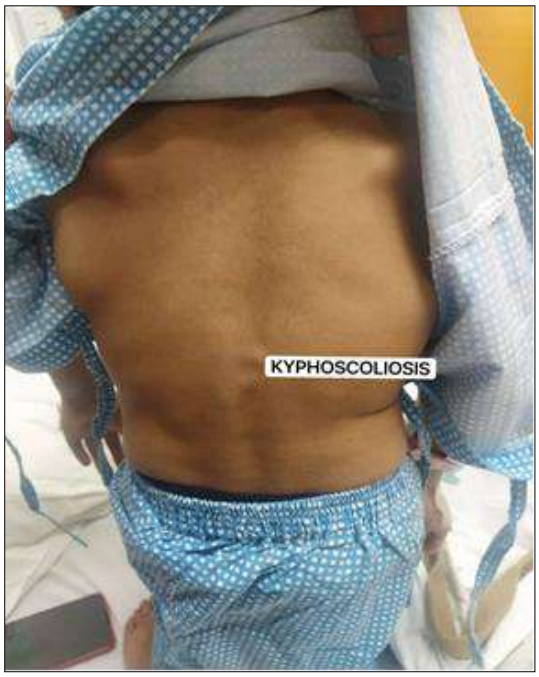

Fig 1: Kyphoscoliotic spine

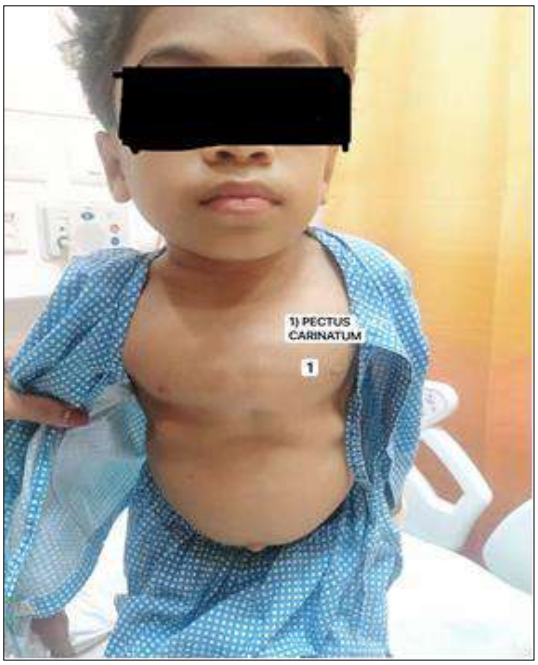

Fig 3: Pectus Carinatum

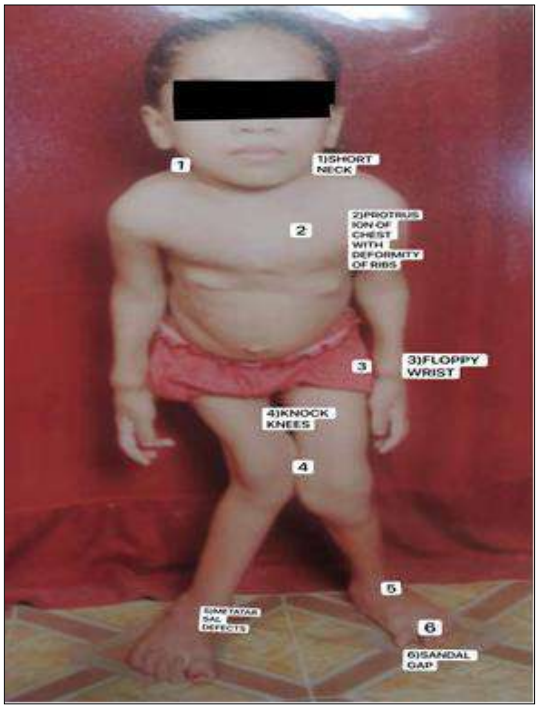

Fig 2: Skeletal Defects in Morquio syndrome

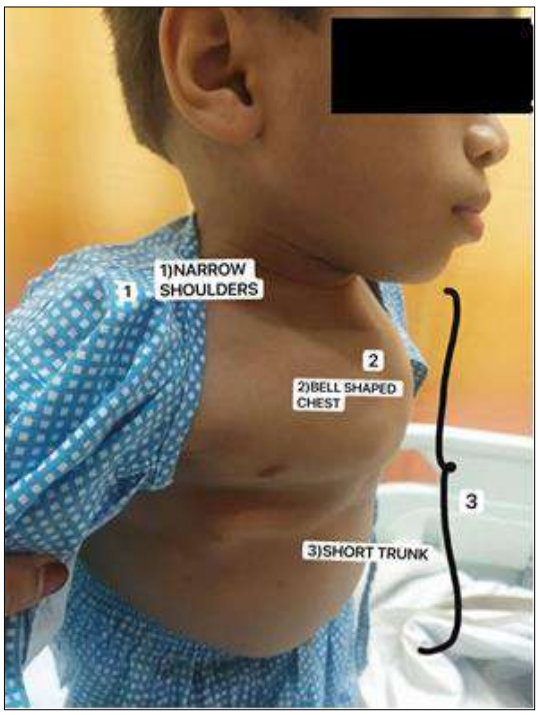

Fig 4: Thoracic Deformities

MPS subjects are often subjected to surgical and diagnostic anaesthetic procedures. The common surgeries being herniorrhaphy, adenotonsillectomy, and median nerve decompression. The nonroutine procedures are cardiac valve replacement, spinal cord decompression, ventricular peritoneum shunt and various orthopedic surgeries to rectify skeletal defects [3].

Standard preoperative preparations done for an MPS patient is often insufficient and mostly ineffective. A full overview of every single case and stream-lined multidisciplinary approach must be taken for a successful execution of the anaesthetic procedure. The mortality related to anaesthesia in such patients is way higher versus the general population. The anesthesiologist must deal with three major concerns in such cases:

a) Difficult intubation
b) Chronic pulmonary disease,
c) Dangerous neck manipulation for cervical instability

Our main aim is to review the current literature on this topic in a procedural algorithm of anesthetic management for these high-risk patients, needing surgical procedures [3].

Case Report

Our patient was a 9 year old child born of a 3rd degree consanguineous marriage, A known case of type 4 MPS (Morquio Syndrome). Patient got admitted for atlantoaxial decompression and fusion. Parents first noticed that the child wasn't gaining weight at 1.5 years and child had anterior broadening of chest wall along with widening of ends of long bones in all four limbs for which child was taken to paediatrician. With on-going evaluation at the age of four diagnosis of metabolic disorder Type 4 MPS and multiple skeletal dysplasias was made.

Patient's general and physical examination in brief:

FUll term normal delivered, cried immediately after birth, no ICU stay, birth weight-3 kgs Immunisation-Complete as per NIS

Development

1. Gross motor-Child can walk but cannot run. Can clim stairs with one step at a time.
2. Fine motor-Can hold spoon, can draw.
3. Social skills-Can participate in conversation.
4. Language-Can make stories with 10-12 meaningful sentences.
5. Diet-Mixed type of diet with no protein or calorie deficit. Family history- No significant family history.

On examination: Afebrile with vitals stable

Anthropometric examination

1. Weight-12.10 kgs Below 3rd percentile
2. Height- 82 cms below 3rd percentile 3. US:LS- 0.9
3. Chest diameter AP-12 cms
4. Chest transverse diameter- 18 cms

Physical examination

Short stature Genu valgus

Systemic Examination: Central Nervous System Neuro power grade 4+ Reflexes +
Plantars up going GCS-15/15

Respiratory system

AEBE clear, reduced lung capacity, breath holding time 15 seconds Gastrointestinal and genitourinary system- NAD

Airway examination

MOA MPC-ll No loose teeth Short neck limited extension

Blood Investigations

CBC-14/5600/3.43 Lacs
Creatinine-0.32 Aptt-T-35.8 PT/INR-12.8/1.19
HHH-Non Reactive Na/K-143/4.3

Radiological investigation

MRI-Severe C1-C2 stenosis. Myelomalacia with chronic compressive myelopathy CT Angiography-Shows no vertebral artery anomaly.

Xray chest and spine: Atlantoaxial instability, Dysplastic vertebrae, thoracolumbar kyphosis 2D Echo- Normal ventricular function, Myxomatous mitral and tricuspid valves, No MR, Trivial TR, Mild thickened Aortic valve with No AS, Tortuous descending aorta after complete anaesthesia check-up, Multidisciplinary team was formed involving paediatric cardiologist, intensitivist, orthopaedician and pediatri anaesthetist. High risk consent explaining prolonged ICU stay and ventilator support was taken and Occipitocervical fusion with C1 laminectomy was planned. Adequate blood and blood products were also arranged from the blood bank for the surgery.

Patient was premedicated with injection Fentanyl 10 micrograms I.V., injection Glycopyrrolate 40 micrograms I.V. injection Midazolam 0.5 milligrams I.V. An antisialogogue is an essential component of preanaesthetic medication in view of the copious upper airway secretions, and antibiotic prophylaxis is advised in view of the high incidence of cardiac valvular lesions.

After adequate preoxygenation, anaesthesia induction was done with injection Propofol 40 mg I.V. till the patient lost consciousness maintaining spontaneous respiration. We gave titrated doses under strict respiratory monitoring. We took care of the cervical spine by limiting the neck movements and stabilizing it during induction and intubation throughout the procedure. Check fibreoptic bronchoscopy was done to see the upper airway anatomy. Epiglottis and vocal cords were visualised and any extreme anatomical variation was ruled out. After confirming ability to mask ventilate, muscle relaxation was achieved with injection Atracurium 10 mg I.V. using cmac videolaryngocope, trachea was intubated using cuffed 5 mm endotracheal tube and position was confirmed and fixed. Radial artery was cannulated for invasive blood pressure monitoring. A peripheral IV cannula of 22 G was also inserted. After adequate eyes and pressure points padding patient was made prone. Neuromonitoring was done intraoperatively. For maintenance of anasethesia, Inj. Propofol (100 mcg/kg/minute) and Inj. Dexmedetomidine (0.4 mcg/kg/minute) intravenous infusions were used (dosages titrated as per effect) and inhalational anaesthesia and muscle relaxants were avoided. BIS was monitored and maintained between 40-60 throughout the surgery. Total intraoperative blood loss was 310 ml.For fluid maintenance 650ml of 1% dextrose RL was given. Intraoperative urine output was 75ml. Post-surgery arterial blood gas was within normal limits with Hb being 10.2 gram percent. Extubation was done once patient became conscious with good

respiratory efforts. Extubation was planned to be done when patient was full conscious and awake, considering the possibility of respiratory depression and difficult intubation if reintubation was required. Postoperatively intensive monitoring for airway patency and adequacy of respiration and hemodynamic stability was planned. Paracetamol and diclofenac suppositories were given per rectal for postop analgesia. Patient was shifted to ICU for postoperative care with injection Dexmedetomidine infusion at 0.25 mcg/kg/min. Recovery was uneventful and the patient was shifted out of ICU the next day and discharged on postoperative day four.

Discussion

At birth, a patient with Morquio syndrome may appear healthy; however, as the child grows into adulthood, various manifestations of this syndrome begin to emerge.Cardiac involvement is usual. Valve involvement may cause incompetent or stenotic lesions especially of the aortic valve whilst myocardial deposits cause reduced compliance. Systemic and pulmonary hypertension may occur and death is often due to cardiac or respiratory failure [4, 5, 6]. In our patient, we noticed coarse facial features like frontal slanting, narrowed nasal opening, macroglossia, reduced oropharyngeal space, dental caries, restricted neck movements and the skeletal abnormalities like kyphoscoliosis contributing to difficult ventilation and intubation. The most important problems concerning the airway management are based on intracellular accumulation of mucopolysaccharides resulting in macroglossia, limited mouth opening due to involvement of temporo-mandibular joints, short neck and subglottic narrowing. These features of the patent can lead to “cannot intubate -cannot ventilate situation”. Oral fiberoptic technique might be difficult due to large, heavy, anteriorly placed, “hanging epiglotis” (sometimes resting on the tong base) making the passage of the fibrescope towards the glotic opening impossible. Nasal fiberoptic technique is really helpful in such times, but might be difficult because of mucopolisaccharoide deposition in the nasopharynx, with higher chances of bleeding. Careful preoperative airway examination should be conducted to predict and evaluate the possible airway difficulties. When difficult airway is predicted, loco-regional technique should be applied. However, performing spinal anesthesia can be difficult due to progressive scoliosis. Also the safe dose of local anesthetic agent is difficult to calculate due to extreme rowth retardation in most of these patients. Regardless of the technique we choose, all necessary equipments to manage difficult airway should be readily available in case of complications or failure to intubate. There are also controversies about the appropriate ways of induction. Some anaesthesiologists prefer the use of intravenous agents and others of inhalational agents (especially in children), but most of the authors advise to avoid the use of muscle-relaxants especially the long acting ones. The relaxation of the supraglottic tissue may preclude effective bag-mask ventilation. Endo-tracheal intubation using fiberoptic technique, being minimally invasive and allowing neutral head position is strongly recommended in these patients. Fiberoptic bronchoscope may not be available at most centres and it also requires special skills [7].

Conclusion

As we discussed MPS children are extremely high risk patients because of airway narrowing, bone dystrophy, cardiac illness, and neurological impairment. They require the cooperation of counselors with different professional specialties such as otorhinolaryngologists (ORLs), anesthesiologists, and surgeons. We can achieve that with the creation of a communicative multidisciplinary team, available to cooperate and draw a perfect case management plan with other professionals as well. The administration of anesthesia should be performed only in specialized centers by experienced anesthesiologists and trained personnels. Indication for surgery should be carried out only after consulting the anesthesiologist, who has the duty to discuss risks and benefits with the parents and relatives [3].

Conflict of Interest
None declared

Financial Support
Not applicable

References

1. Clark BM, Sprung J, Weingarten TN, Warner ME. Anesthesia for patients with mucopolysaccharidoses: Comprehensive review of the literature with emphasis on airway management. Bosn J of Basic Med Sci [Internet]. 2018 Feb. 20 [cited 2022Aug.5];18(1):1-7. Available from: https://www.bjbms.org/ojs/index.php/bjbms/article/view/220.
2. Mohammed YM, Khan S. Advanced Techniques for Securing Airway in Mucopolysaccharidoses and the Impact of New Therapeutic Approaches. Cureus. September 22, 2020;12(9):e10582. doi:10.7759/cureus.10582
3. Concetta Maria Spinello, Lorena Maria Novello, Sofia Pitino, Claudia Raiti, Paolo Murabito, Fortunato Stimoli, Marinella Astuto. Anesthetic Management in Mucopolysaccharidoses, International Scholarly Research Notices; c2013, Article ID 791983, 10 pages, 2013. https://doi.org/10.1155/2013/791983.
4. Renteria VG, Ferrans VJ, Roberts WC. The heart in the Hurler syndrome. Am J Cardiol. 1976;38(4):487-501. [Medline]
5. Ireland MA, Rowlands DB. Mucopolysaccharidosis type 4 as a cause of mitral stenosis in an adult. Br Heart J. 1981;46(1):113-15. [Medline] [Free PMC Article]
6. King DH, Jones RM, Barnett MB. Anaesthetic considerations in the Mucopolysaccharidoses. Anaesthesia. 1984;39(2):126-31. [Medline]
7. Thorat, Wasmatkar, Gore. Anesthetic considerations in Morquio syndrome: A case report. Anaesthesia,Pain and Intensive Care. 2013;17(1):75-78. http://www.apicareonline.com/wordpress/wp-content/uploads/2013/05/18-Anesthetic-co nsiderations-in-Morquio.pdf

How to Cite This Article
Mishra R, Deshmukh VN, Gujaran RD. Anaesthetic challenges and perioperative management of a child with morquio syndrome posted for atlanto axial fusion and decompression: A case report. International Journal of Medical Anesthesiology. 2023;6(1):16-19.