Case Report

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Mucopolysaccharidosis and Anesthetic Challenges

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Abstract

Rare diseases and syndromes are of special interest to pediatric anesthesiologits, as each of them has very specific anesthesia concerns. Knowledge of the pathophysiology, symptomatology, treatment options of these diseases and tailored anesthesia management forms the basis of provision of safe anesthesia care to these groups of children. Mucopolysaccharidosis are a group of lysosomal storage disorders. They are caused by the total or partial deficiency of one of the eleven enzymes involved in the metabolism of glycosaminoglycans. This deficiency leads to gradual accumulation of glycosaminoglycans in the lysosomes leading to permanent, progressive cellular damage which affects appearance, physical abilities, organ and system functioning and mental development. From the anesthesiologist point of view, these patients have problems with airway management and positioning. Few anesthesiologists get to routinely care for these patients. But individual patients undergo multiple surgical procedures for improvement in quality of life. We present a case of an 11-year-old girl with MPS posted for herniotomy.

Keywords: Mucopolysaccharidosis; Difficult airway; Anesthesia

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Introduction

The mucopolysaccharidoses are a group of inherited, chronic progressive, metabolic diseases and include seven eponymously named syndromes caused by deficiency of 11 different lysosomal enzymes that are required for the catabolism of glycosaminoglycans (heparain sulfate, dermatan sulfate, chondroitin sulfate).[1] The incidence of MPS is 1:25,000. There is accumulation of glycosaminoglycans (GAG) in several body tissues leading to the involvement of multiple organ systems including airway, cardiac, respiratory and skeletal systems. systemic involvement and severity of the disease progress with time. MPS I (Hurler's syndrome) and MPS II (Hurler's syndrome) manifest tracheobronchial complications, cardiac disease (mitral valve involvement, aortic valve anomaly, left ventricular hypertrophy) and hepatosplenomegaly. Restrictive lung disease due to skeletal involvement is seen in MPS IV (Morquio's syndrome) and MPS VI (Maroteaux–lamy syndrome).

Airway obstruction occurs due to macroglossia adenotonsillectomy hypertrophy and deposition of GAG in the pharyngeal wall and larynx.[2] GAG infiltrate the connective tissues of the oropharynx and airways causing airway obstruction, obstructive sleep apnea, difficult mask ventilation and intubation. Submucosal GAG deposits in the upper airway (tongue, floor of mouth, epiglottis,

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ary-epiglottic folds) and tracheal wall impart a rigid anatomy. They also can have a short, immobile neck with limited mobility of the cervical spine and temporomandibular joint further contributing to difficult airway. Odonoid dysplasia and radiographic subluxations of C1 on C2 is common in MPS 1, IV, and VI and may cause anterior dislocation of the atlas and spinal cord compression. They can also have abnormal laryngeal and tracheal cartilage and copious secretions compounding the situation. Older age is associated with increased risk of difficult intubation, due to effect of alteration of MPS.

GAG accumulation in the cardiac tissues, causing valvular abnormalities and insufficiency (myocardial deposits). They also accumulate in the bones, joints, ligaments leading to atlantoaxial instability. Developmental delay and progressive neurological decline are also seen in these patients.

The incidence of anesthesia-related complications is high in MPS patients. Incidence of difficult intubation is 25–80% failed intubation is 2–10% and that of perioperative cardiorespiratory complications range between 5–25%.

In a retrospective review of anesthesia records of 19 children with MPS over a 9-year period, the incidence of respiratory and cardiovascular complications was 24% and 4% respectively.[3] The respiratory complications were airway obstruction at induction, difficult mask ventilation, difficult intubation and failed intubation. Airway obstruction during emergence occurred after 13 anesthetics. The cardiac complications were hypotension, bradycardia and perioperative circulatory arrest.

In a recently published retrospective analysis of 54 patients with MPS in an Italian tertiary referral center, 16 patients had at least 1 anesthetic complication during their clinical course.[4] Hypoxia, airway obstruction, hypoventilation and laryngospasm was observed in 11, 4, 4 and 2 procedures, respectively. 19 (29%) of intubation attempts were difficult and of these 3 were achieved with fiberoptic technique, 6 by video-assisted laryngoscopy and the remaining by repeated direct laryngoscopies. During 3 (1.8%) of these cases, face mask ventilation was inadequate to provide oxygenation, emergency intubation failed and patients were salvaged with the LMA. However, there is also a case report of failure of the LMA to secure the airway in a patient with MPS type II (Hunter's syndrome), where a subsequent rigid bronchoscopy revealed a pedunculated polyp just above the epiglottis, with diffuse infiltration of the pharyngeal and laryngeal mucosa and a smaller trachea.[5]

Postobstructive pulmonary edema during anesthesia in 5 patients with severe form of MPS has also been reported.[6] Chronic myelopathy can lead to central hypoventilation, which together with respiratory muscle weakness can lead to difficulty in extubation.

Case Report

An 11-year-old girl was posted for inguinal hernia repair. She was diagnosed as a case of MPS at one year of age and is under regular management with physiotherapy. At the time of diagnosis she was found to have a left inguinal hernia with bowel loops, left ovary and uterus as contents. She also had a small VSD. Because of these reasons, the hernia repair was deferred to a later age. At present, she was short for her age with delayed milestones. She was alert, very vocal and her intellect seemed to be normal. There was mild scoliosis and deformities of fingers. Facial features were coarse with flat nasal bridge, small mouth with crowded teeth and retrognathia. She had a history of snoring and slept comfortably only in lateral decubitus position. Her skin was dry with poorly visible veins. She had a skin dimple in the sacral region. Port-wine stains were present in the neck and chest. Lab investigations were within normal limits. Echocardiogram showed no evidence of residual VSD. Airway assessment showed a small mouth with crowding of teeth. Mallampatti was grade 2. MRI neck did not show any evidence of spinal cord compression.

An intravenous access was secured in the preoperative ward. Because of anticipated airway shifting on to the table, she was hooked on to monitors. Sevoflurane stepwise increase from 2% to 6% was used to induce the patient. Once induced, trial laryngoscopy was done. Vocal cords were not visualized with Cormack lehane grade 4. A size 2.5 LMA was inserted successfully and anesthesia maintained with O_2/N_2O 33%/66% and titrated Sevoflurane through a Jackson Rees circuit. Analgesia provided with 2 mics/Kg Fentanyl and hernia block with 10 mL 0.25% Bupivacaine. Intraoperative period was uneventful. Sevoflurane was cut at the last skin suture and LMA was removed after full recovery. Oxygen supplementation was given for 6 hours in the postanesthesia care unit and she was discharged after 36 hours.

Discussion

MPS are rare conditions, incidence varying from 1 in 24,000–5, 00,000 population.[7]

There are seven types of MPS. Hurler's syndrome being the prototype.

MPS cases are notorious for airway difficulties and are a challenge in anesthesia. They are rare and few anesthesiologists get to serve these patients. In contrast, these patients are in frequent need of medical attention. MPS causes accumulation of unmetabolized molecules in the connective tissue leading to swollen tongue and parapharyngeal tissues.[8] This can lead cause airway obstruction in a ball valve fashion in anesthetized and paralyzed patients. In addition, in some types of MPS there is a specific risk for compression of the cervical spinal cord (Metabolic myelopathy). Hurler's (1H), Morquio (IV), Maroteaux-Lamy (VI) and Sly (VII) syndromes are known for cervical canal narrowing.[8,10] This is due to thickening of duramater and hypoplasia of dens axis. These changes make direct laryngoscopic visualization of vocal cords both difficult and risky. Failure to control the airway is the largest single cause of perioperative mortality. Supraglottic airway devices like the laryngeal mask airway have been found to be useful in maintaining airway in this population. The advantage of LMA is the ease of positioning without excessive neck extension and without muscle relaxant.[2,9] The use of volatile induction and maintenance anesthesia (VIMA) helps in easy titration of depth thereby giving greater control over airway during induction. The deposits of mucopolysaccharides in lower airways results in diffusion defects — hypoxia and hypercarbia. This may lead to secondary pulmonary hypertension.[7,10] Supplemental analgesia with nerve blocks decreases requirement of opoids enabling earlier discharge from post Anesthesia Care Unit.

Conclusion

Children with congenital syndromes with multiple anomalies need a multidisciplinary approach to their care. It is advisable to actively look for specific anomalies associated with the particular syndrome. Systems that may be affected include cardiovascular, respiratory, airway, spine, coagulation, metabolic and endocrine systems. Specific websites and books are also available as references. An individualized tailored approach to anesthesia care is important to avoid anesthetic complications in these special groups of children.

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