Hurler's Syndrome: Anaesthetic Challenges and Management

Dr.Arun Kr. Gupta, Dr. Vinod Kharde, Dr. Varun Gawer, Dr.R.V.Sidhaye, Dr.D.S.Divekar.

Department. Of Anaesthesiology, Rural Medical College, Loni

Corresponding Author: Dr.Arun Kumar Gupta

Assistant Professor

Dept. of Anaesthesiology & Critical Care

Rural Medical College, Loni

Maharashtra, India, 413736

guptaarun71@yahoo.com

ABSTRACT

Mucopolysaccharidosis (MPS) are a group of inherited disorder of connective tissue metabolism. These disorders are uncommon and few anesthetists care for these patients on a regular basis although individual patients often undergo multiple anesthetics for procedures intended to improve their quality of life. Herein we describe the anaesthetic considerations and management of an 8 year old child with Hurler syndrome, who presented for an inguinal hernia repair.

Key Words: Mucopolysaccharidosis; Hurler syndrome; Anesthesia

INTRODUCTION

Mucopolysaccharidoses (MPS) are a group of progressive hereditary disorders of connective tissue metabolism in which lysosomal enzyme deficiency leads to intralysosomal deposition of mucopolysaccharides or glycosaminoglycans in the airway, cornea, brain, heart, liver, spleen, bones, ligaments, blood vessels, skin producing various symptoms.[1,2] Seven different Types of mucopolysaccharidosis have been described and categorized into syndromes according to both the clinical features and demonstrated enzyme deficiencies.

Anaesthetic management of patients with Hurlers syndrome - a rare metabolic disorder is very challenging & needs to be planned well. Our experience with an 8 year old patient who presented for herniorraphy is described below. Breathtaking advances like stem cell transplant, bone marrow transplant & enzyme replacement therapy will increase life expectancy of such patients exposing them to a variety of problems & coming generation of anesthesiologists are more likely to see greater number of such patients in the days to come.

CASE REPORT

Our patient diagnosed to have MPS at five years of age & on regular management by physiotherapy, presented to us for inguinal hernia repair at the age of 8 years. He was short in stature (96cm) weighed 35kg and had an apathetic face with coarse features. He had attention deficit with mild mental retardation & hyperkinetic behavior. His developmental milestones were delayed. He used to snore during sleep and preferred to sleep in prone position. His respiratory system and cardiovascular systems were within normal limits. His routine investigations including ECG and chest X-ray were normal. Echocardiography ruled out any cardiac abnormality. MRI Brain showed Patchy T_2 prolongation in the deep

cerebral white matter bilaterally. His airway assessment revealed normal mouth opening, a thick and large tongue with a modified Mallampati Grade IV view. He had a short neck with limited extension (<30◦). X-ray for soft tissue of neck showed narrow air column.

In preanaesthesia room during IV access, his skin was found to be dry, thick and rough. ECG, NIBP and SpO2 monitors were attached. Following premedication with glycopyrrolate and preoxygenation inhalational Anesthesia was induced by progressively increasing concentration of Halothane upto 3.5%- 4.5% and laryngoscopy was done in deeper plane with the patient breathing spontaneously. Laryngeal structures were not visible even after posterior displacement of laryngeal cartilages. Repeated attempts were made ensuring oxygen saturation & at the final attempt tip of the epiglottis was seen markedly shifted to left side. During next attempt by a straight blade laryngoscope introduced through left angle of mouth, cricoid pressure by the assistant improved the laryngeal view with the posterior extremity of glottis barely visible. A thin smooth plastic bougie was introduced along the posterior surface of epiglottis. A 6mm ID cuffed ETT failed to pass over the bougie & even smaller sized tubes of 5.5 and 5mm ID were tried with no success. Finally a 4.5mm uncuffed tube could be passed over the bougie into the larynx. The bougie was removed and the ETT was connected to Jackson-Rees circuit and manual ventilation started. There was very minimal air leak even after using a smaller uncuffed ETT. Anesthesia was then maintained with oxygen in nitrous oxide and halothane. Inj. fentanyl 1µg/kg was given to maintain intraoperative analgesia while muscle relaxation was maintained with intermittent doses of atracurium. At the end of the surgery residual neuromuscular blockade was reversed with neostigmine (0.05mg/kg) and glycopyrollate (0.01mg/kg). His trachea was extubated when he started responding to verbal commands and had

adequate respiratory effort. He was shifted to recovery room and oxygen supplementation was continued for 24hrs and was discharged from hospital 48 hours after surgery.

DISCUSSION

Hurler syndrome or MPS 1 H is the prototype of MPS and is the most severe form of it[3]. In Hurler's syndrome airway problem has been described as the worst in pediatric anesthesia[4]. Perioperative mortality rates averaging 20% have been reported for patients with this disease, with failure to control the airway as the largest single cause of mortality[5].

MPS are rare conditions, incidence varying from 1 in 24,000-5, 00,000 population[2, 6].

The anesthesiologist may face diverse problems while anaesthetizing these patients so knowledge of the anesthetic implication of the disease is essential to prevent any catastrophe[3]. The most important and life threatening problem is difficulty in maintaining airway while anaesthetizing such patients because of the anatomical changes in upper airway due to deposition of mucopolysaccharides in tongue, tonsils, adenoid, epiglottis, glottis and trachea[2,6-8]. These patients also have excessive tracheobronchial secretion with frequent upper respiratory infection[2]. Chest deformity along with deposits in lower respiratory tract and lung interstitium may cause obstructive lung disease and diffusion defects leading to hypercapnia, hypoxia and elevated airway pressures[4, 9]. They are also likely to develop myocardial hypertrophy, ventricular dysfunction, cardiomyopathy and heart failure following pulmonary hypertension[2, 6]. Nerve and tendon entrapment is common in them. Due to deposition of mucopolysaccharides in brain cells they may develop progressive mental retardation of varying degree. Thickening of meninges may

lead to hydrocephalus and hypertrophic cervical pachymeningitis that may result in myelopathy associated with nerve root compression[6].

During preanesthetic check up the type of MPS syndrome should be confirmed as it will have a great implication on severity of difficulty to be anticipated during airway management. Child's intelligence and behavior are important. In Children with MPS behavior may vary from uncooperative belligerent to placid, cooperative and lovable. Child's favorable sleeping position should be inquired, since this may be the position in which airway is held open[3]. History of snoring and sleep apnea should be inquired. Preoperative investigations should include arterial blood gas analysis, hemoglobin, serum electrolytes, chest and cervical spine radiographs, electrocardiography, echocardiography and pulmonary function tests if possible. In our case the child was thoroughly investigated for blood chemistry, ECG, echocardiography and skeletal radiography. SBE prophylaxis with antibiotics is recommended for all MPS patients with valvular cardiac lesion[2]. In all cases of MPS excessive deposition of mucopolysaccharides continues throughout the life with a special predilection for tracheal cartilages, worsening clinical features in all of them as age advances[1, 2].

In all cases inhalation induction with maintenance of spontaneous ventilation is preferred but in mentally retarded and uncooperative patient intravenous induction is more satisfactory. In all these patients spontaneous ventilation should be maintained until adequate airway control is achieved, as after administration of muscle relaxant muscle tone is lost and the thickened supraglottic tissue and large tongue obstruct the airway and act as a ball valve during manual ventilation leading to upper airway obstruction[1, 3, 7]. Therefore as rule of thumb spontaneous ventilation should be maintained until the airway is secured.

In our case, the child was premedicated with glycopyrrolate and general endotracheal anaesthesia with inhalational induction technique was adopted in view of difficult airway and abdominal surgery.

Recovery after GA in patients with MPS is often slow and accompanied by periods of breath holding, apnea, bronchospasm, cyanosis and respiratory arrest[8.] Therefore anesthetic sequences which ensure early return of consciousness and airway reflexes are strongly recommended[10]. As an awakened child with MPS struggles to breathe against the high airway resistance, pulmonary hypertension is exacerbated and negative pressure pulmonary edema may develop[2, 11].

Regional anesthesia offers a valuable safe alternative to children with MPS undergoing lower abdominal, perineal, upper and lower extremitysurgery[2] and should be preferred over GA if cooperation of child is ensured.

CONCLUSIONS

In children with MPS, both anesthesiologist and surgeons should be aware of the expected complications. The benefit of the surgical procedure should be balanced against the risk of exposing the child to GA. The parents of the patient should properly be informed of the risk involved. Anesthesia should ideally be given by the anesthesiologists who are experts in handling pediatric airway problems and resuscitation, in a center in which pediatric intensive care facilities are available.

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