Anaesthetic Challenge in a Child with Hurler’s Syndrome: Encouraging Experience with Neuraxial Blockade

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Abstract

Mucopolysaccharidoses are uncommon genetic diseases with multisystem involvement. The potential for airway difficulties and varying success rate of neuraxial blockade demands considerable expertise in the anesthetic management in these patients. Airway management has evolved significantly in such syndromic patients with the increasing use of video laryngoscopes and Supraglottic airway devices. However, airway catastrophes are not uncommon. This calls for better utilization of neuraxial and peripheral nerve blocks along with the use of newer agents for sedation having a favorable pharmacological profile. Here, we present a case of child having Hurler disease with an inguinal hernia successfully operated under subarachnoid blockade.

Keywords: airway management, dexmedetomidine, Hurler disease, pediatric anesthesia, regional anesthesia.

Introduction

Mucopolysaccharidoses are lysosomal storage diseases with multi organ involvement leading to decreased life span. 1,2 The need for surgeries in pediatric age group and multi organ involvement of the disease pose challenges in the anesthetic management in these patients. Although case reports of general anesthesia using multiple airway devices are available, the use of central neuraxial blockade seems to be less popular. The possible reasons could be the
difficulty associated with cognitive dysfunction in these patients and due to chances of failed blockade due to varying levels of spinal fusion and deposition of Mucopolysaccharides.

Case report
A 1-and-a-half-year-old, 13 kg male child was posted for bilateral inguinal hernia repair. The child had been diagnosed as compound heterozygous variant of reported phenotype of Hurler syndrome (MPS1H). In the pre-anesthetic checkup, the mother gave a history of noisy breathing, snoring, and abrupt awakening from sleep suggestive of possible obstructive sleep apnea. There was also a history of thick and copious secretions and frequent respiratory infections in the child. A positive family history with an elder male sibling who died of the same disease at the age of 5 years was also revealed. On examination, the child had a large head, coarse facies and scalp hair, thick eyebrows, prominent alae nasi, macroglossia, and a short neck (Figure 1). Chest X-ray showed cardiomegaly (Figure 2). X-ray of the Dorso-lumbar spine showed scoliosis. Presence of prominent subarachnoid space over bilateral frontal cortex was present in the CT head during workup (Figure 3). However, there were no signs and symptoms suggestive of raised intracranial tension. The echocardiogram showed trivial mitral regurgitation. The rest of the systemic examination and laboratory parameters were within the normal range.

In view of the anticipated difficult airway, our plan of anesthesia was subarachnoid blockade under dexmedetomidine sedation. On the day of surgery, one hour prior to the procedure, eutectic mixture of local anesthesia was applied over the area of third to fifth lumbar space. Difficult airway cart along with ENT surgeon for surgical airway were kept standby for any possibility of airway problems was encountered. The case was conducted by trained pediatric anesthesiologist. Child was sedated with dexmedetomidine 1 µg kg-1 given as a slow intravenous infusion over 15mins under strict monitoring before initiating SAB. Supplemental moist oxygen was given with a Hudson mask. The child was positioned in the left lateral position with neck in neutral position. Using a 26 G Quincke needle, 5 mg of Bupivacaine heavy was injected in the subarachnoid space at L3/L4 level. Surgery lasted for 30 minutes and was uneventful and the child was shifted out of OR (Operating Room) 15 minutes post-surgery after being fully awake. Further, the child was monitored for 2 hours in the postoperative room before shifted to the ward.

Discussion
The primary concern in the anesthetic management in a case of Hurler disease is attributed to extensive anatomical and physiological abnormalities rendering airway management difficult.

The difficult airway encountered in Hurler’s syndrome is attributed to the deposition of mucopolysaccharides in tongue, tonsil, adenoid, epiglottis, glottis and trachea, macroglossia, narrow nasal passages, short neck with or without cervical instability, Obstructive sleep apnoea makes bag mask, laryngoscopy and intubation difficult.
Increased thick secretions, and frequent upper and lower respiratory tract infections increase the risk of bronchospasm, cyanosis, and negative pressure oedema following extubation.

Cardiac system abnormalities including ventricular hypertrophy, valvular heart disease, myocardial dysfunction, cardiomyopathy, heart failure, arrhythmia, and sudden cardiac death.

Added to this, vertebral anomalies and cognitive impairment could make regional anesthesia also challenging.

This complexity has made it difficult to lay down a common management protocol in the anesthetic management for such patients. There is hardly any literature available detailing the use of neuraxial blockade in Hurler patients of this age group\textsuperscript{4,5} although, neuraxial blockade has been used with reasonable success rates in Mucopolysaccharidoses in elder age groups.\textsuperscript{6,7}
The primary challenge in administering neuraxial blockade was getting the patient adequately sedated while keeping in view the potential problems of airway obstruction post-sedation. Studies using propofol and ketamine as sedating agents in these patients were noted. Use of Sevoflurane before spinal anesthesia has also been reported as it has an effect on analgesia and sedation and partial skeletal muscular relaxation. Narcotic premedication should be avoided because of airway problems. If a benzodiazepine is administered as premedication, a reversal agent should be available.

We opted for Dexmedetomidine as a sedating agent. Dexmedetomidine is a relative selective alpha 2A agonist, with analgesic effect due to spinal, supra spinal, direct analgesic and or vasoconstrictive effect. A single iv bolus over 15min followed by spinal blockade causes anxiolysis with quick arousal, minimal respiratory depression, preserves spontaneous respiration without secretions and maintains hemodynamic well. In children after spinal anaesthesia, a maintenance infusion might result in greater degree of hypotension and bradycardia, requiring resuscitation.

There have been instances of failed neuraxial blockade hypothesized by researchers due to the possible deposition of mucopolysaccharides in and around the nerve sheath. However Regional can be a successful anaesthetic technique as the amount of mucopolysaccharides in younger children could be small.

Even with the patient being operated under central neuraxial blockade, preparation for anticipated difficulties in securing the airway also needs to be addressed given the high incidence of airway difficulties in these patients. With the introduction of different types of intubating LMAs, the approach generally has been to administer anesthesia using these LMAs or intubating through these using fibreoptic guidance. Besides airway difficulties, general anesthesia might create other complications in these children due to the presence of cervical instability and alterations associated with chronic pulmonary disease. As a result, regional anesthesia appears to be a feasible choice wherever possible, along with well-defined backup plans and the appropriate expertise to deal with any unexpected difficulties.

Conclusion

Anesthetic management of patients with congenital syndromes is challenging and to formulate or follow any common guideline is difficult. Neuraxial blockade which can avoid airway manipulations can be considered even in the pediatric age group with necessary back up plans.

References