Pediatric difficult airway management and anesthetic in children with mucopolysaccharidoses undergoing ventriculoperitoneal shunt surgery: A case report

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Abstract

The mucopolysaccharidoses has high prevalence of airway obstruction and restrictive pulmonary disease in combination with cardiovascular manifestations poses a high anaesthetic risk to these patients. A 11 year old boy, weighing 27 kg, a known case of mucopolysaccharidoses was admitted with hydrocephalus. Patients are planned for elective ventriculoperitoneal shunt surgery. He has problems include airway obstruction, intubation difficulties or failure, possible emergency tracheostomy. We describe the successful airway and anesthetic management of pediatric mucopolysaccharidoses who underwent ventriculoperitoneal shunt surgery for acute hydrocephalus.

Keywords: Mucopolysaccharidoses; Difficult airway; Ventriculoperitoneal shunt; Acute hydrocephalus

Introduction

The mucopolysaccharidoses are genetic diseases characterized by storage of incompletely degraded glycosaminoglycans and caused by mutations in lysosomal hydrolase enzymes. The high prevalence of airway obstruction and restrictive pulmonary disease in combination with cardiovascular manifestations poses a high anaesthetic risk to these patients. Typical anaesthetic problems include airway obstruction after induction or extubation, intubation difficulties or failure [can't intubate, can't ventilate (CICV)], possible emergency tracheostomy and cardiovascular and cervical spine issues. Because of the high anaesthetic risk, the benefits of a procedure in patients with MPS should always be balanced against the associated risks. Therefore, careful evaluation of anaesthetic risk factors should be made before the procedure, involving evaluation of airways and cardiorespiratory and cervical spine problems. We present the case of a patient with pediatric diagnoses of hydrocephalus, scaphocephaly and mucopolysaccharidoses. We decided to perform ventriculoperitoneal shunt surgery to avoid increasing intracranial pressure to become heavier and considering a progression towards difficult intubation in mucopolysaccharidoses get older.

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Case description

A 11 year old boy, weighing 27 kg, a known case of mucopolysaccharidoses was admitted with complaints of left head establishes the bathroom floor, the patient looks nervous, do not want to eat and speak, sleep snoring and difficult to breathe. A CT-scan of head revealed communicating hydrocephalus with dilatation the right and left lateral ventricle, macrochephaly with scapocephaly picture, crouzon syndrome with brachycephaly. Patients are planned for elective ventriculoperitoneal shunt surgery. The child had a history of delayed developmental, coarse facies, dolicocephalic head, large tongue, at nasal bridge with wide intercanthal distance, short nose bone and multiple skeletal deformities.

The patient was premedicated with atropine 0.25 mg and dexamethasone 5 mg with existing central venous access. Before intubation, patient was administered topical anesthesia to the patient with lidocaine aerosol at 10% into the oral cavity and the pharynx. We performed bilateral superior laryngeal nerve blocks using the external approach. The patient is placed in the supine position and need a degree of neck extension to facilitate identification of the hyoid bone. Once identified, the hyoid bone is gently displaced to the side where the block is to be performed and a 25 gauge needle is inserted from the lateral side of the neck, aiming toward the greater cornu using 2 mL of local anesthetic (ropivacaine 0.75%) would anesthetize the airway. Patients with hypopharyngeal disease are more dependent on awake muscle tone to maintain airway patency, spontaneous ventilation was maintained along.

Tracheal intubation was performed using Macintosh laryngoscope blade size 2. We look a laryngoscopic view of CL grade IV after doing external laryngeal manipulation with the help of an assistant who provided jaw lift. Intubation was difficult by direct laryngoscope and glidescope video laryngoscope. We decided to perform surgical tracheostomy procedure in the operating theatre with otorhinolaryngology (ENT) surgeon. During surgical tracheostomy procedure, we look collapse of tracheal

cartilage and failed to perform tracheostomy tube insertion size 4.5 cuffed. Because difficult tracheostomy tube insertion, the bougie should be inserted via the side of the tracheostomy and then used as a guide over which the endotracheal tube size 4.5 can be railroaded and replacement ETT success. Correct position of the ETT was confirmed by bilateral chest auscultation. Anesthetic management underwent ventriculoperitoneal shunt surgery success and required in PICU after surgery to monitoring and periodic suctioning is necessary to avaoid secretion retention, infection, and mucus plugging of the ETT (**Figure 1**).

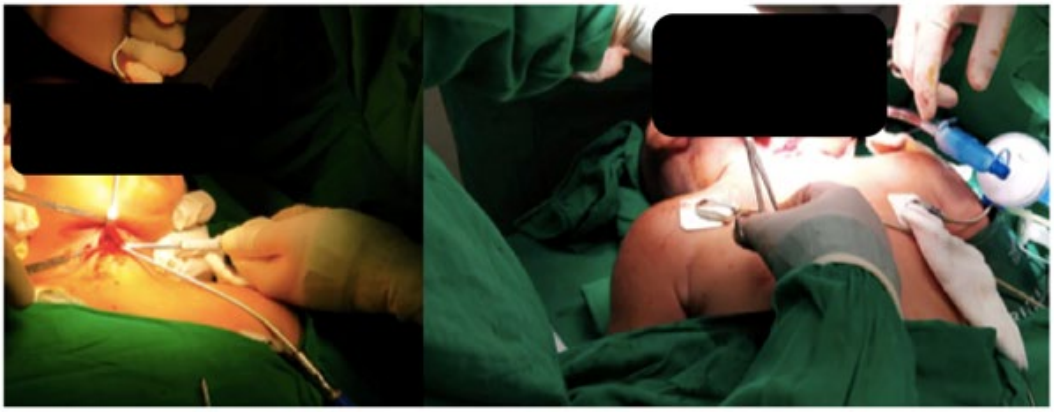

Figure 1: *Endotracheal tube inserted through the tracheostomy (soon after insertion)*

Discussion

Mucopolysaccharidoses are uncommon genetic diseases related to the metabolism of connective tissue.1 MPS is characterized by progressive craniofacial, articular, and skeletal deformities, cardiac involvement, and early death due to pulmonary infections or heart failure, often before adulthood. 2,3 We report case mucopolysaccharidoses with 2 main concerns in such situations difficult ventilate, difficult intubation, and unable for neck manipulation. In this case, we maintain a spontaneous breath to ensure the airway remains safe during the airway management. During airway management, we maintain the principles of neuroanesthesia with preventing secondary brain injury because pain during intubation or tracheostomy surgery. Anesthetic considerations involve attention to the possibility of increased intracranial pressure.4 A difficult intubation equitment should always be ready in operating room before the administration of anesthesia. Sedative premedication should be avoided. Therefore, ensuring spontaneous ventilation until the airway has been secured.5 Nevertheless, the focus of management airway includes avoidance of a situation is called "can't ventilate, can't intubate".

In our case, airway and anesthesia management in patient with mucopolysaccharidoses with hydrocephalus with presenting increasing intracranial pressure will be challenging and requires a balance between neuroanesthetic principles and difficult airway management techniques.

Conclusion

For the children with mucopolysaccharidoses, who are at higher risk at times of ventriculoperitoneal shunt surgery for acute hydrocephalus. By the use of anesthetic management with a successful airway management creates an opportunity for a successful surgery.

Acknowledgment

None.

Conflict of Interest

The authors declare that there are no conflicts of interest.

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