

ANESTHESIA MANAGEMENT IN A PATIENT WITH MAROTEAUX-LAMY SYNDROME

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ABSTRACT

Mucopolysaccharidoses are uncommon hereditary disorders leading to organ dysfunction and anatomic abnormalities. They are accompanied by increased risk of perioperative and postoperative complications. Mucopolysaccharidoses (MPS) are a group of syndromes in which there is an inherited lack of a lysosomal enzyme. Widespread, progressive mucopolysaccharide tissue deposition may cause both organ dysfunction and anatomical abnormalities. The Maroteaux-Lamy syndrome (MPS VI) is a very rare

mucopolysaccharidosis with variable severity of somatic and possible cervical spine involvement.

These children have difficulties during the preoperative, perioperative, and postoperative periods like other children with MPS. We present here two cases with Maroteaux-Lamy syndrome (Type VI) and their problems with the establishment and maintenance of anesthesia.

Key Words: Mucopolysaccharidoses, anesthesia, Maroteaux-Lamy Syndrome *Nobel Med 2012; 8(1): 114-116*

MAROTEAUX LAMY SENDROMLU HASTADA ANESTEZİ YÖNETİMİ

ÖZET

Mukopolisakkaridozlar lizozomlar içinde spesifik enzim eksikliğine bağlı olarak glikozaminoglikan birikimi ile karakterize, kronik ilerleyici ve multi system tutulumu yaparak kalıcı anatomik anormalilik ve organ disfonksiyonuna neden olan nadir görülen hastalık grubudur. Bu hasta grubunda komplikasyon riski hem peroperatif hem de postoperatif dönemde yüksektir. Maroteaux-Lamy sendromu değişik formlarda

somatik ve vertebra defekti olabilen nadir görülen bir mukopolisakkaridoz tipidir. Bu çocuklar diğer mukopolisakkaridozlar gibi preoperatif, peroperatif ve postoperatif dönemde anestezi açısından bazı zorluklar taşırlar.

Burada Maroteaux-Lamy sendromu tanımlanmış iki hastada anestezi yönetimi ve karşılaşılan sorunlar sunulmuştur.

Anahtar Kelimeler: Mukopolisakkaridozlar, anestezi, Maroteaux-Lamy Sendromu *Nobel Med 2012; 8(1): 114-116*

INTRODUCTION

Mucopolysaccharidosis is a hereditary disease in which there is a deficiency of lysosomal enzymes.¹ The systemic deposition of mucopolysaccharides causes disorders of various organs, such as central nervous system and cardiovascular system.² Airway complications are due to possible difficult intubation and upper airway obstruction in this group of patients. These conditions require attention during anesthetic management. Depending on the specific type of lysosomal enzyme deficiency, mucopolysaccharidosis can be classified into six subtypes, with mucopolysaccharidosis type VI (Maroteaux-Lamy syndrome) being extremely rare.

We present two cases, who were scheduled for repair of an umbilical hernia, providing anesthesia to Maroteaux-Lamy syndrome (Type VI) patients and their problems with the establishment and maintenance of anesthesia.

CASE I

The 9-year-old boy (15 kg, 100 cm), diagnosed with Maroteaux-Lamy syndrome (MPS VI), was scheduled for umbilical hernia operation. He had normal mental status, dwarfism with a large head, a very short neck with limited mobility, a prominent breast bone and kyphosis. The patient was evaluated for intubation; Mallampati III airway, macroglossia, and small interincisor distance were observed. Routine preoperative laboratory values were unremarkable, chest examination revealed pectus excavatum and moderate kyphoscoliosis at the level of T12-L4 was observed. Echocardiogram revealed mild mitral valve prolapse and mitral insufficiency. Induction of anesthesia was performed with 30 mg propofol and sevoflurane 2% with O₂, and an air mixture was used for maintenance. Airway topicalization was accomplished with 2% xylocaine. At first, a laryngoscopy attempt was made for the purpose of evaluation of the oropharynx. Subsequently, the soft stilet was inserted to find and feel the tracheal rings and the endotracheal tube was successfully slid over the stilet. Atracurium besylate was administered after successful intubation. Mechanical ventilation continued with sevoflurane 2% with 50% air and 50% O₂. The epidural analgesia was planned for the perioperative and postoperative periods. The epidural insertion was tried at the levels of L3-4, L3-L2 and, L2-L1. The successful insertion was performed at the level of T12-L1 by using the loss of resistance technique. Levobupivacaine 0.75%, 30 mg, 1 ml kg⁻¹ volume and 0.4 mg morphine were given. The epidural block was confirmed intraoperatively with hemodynamic response after 30 minutes. The surgery took one

hour, the maintenance of anesthesia was uneventful, extubation was performed while the patient was completely awake, and there was no desaturation or spasm after the extubation. The patient's visual analog scale (VAS) was 3 in the postoperative period.

CASE II

The 5-year-old boy (11 kg, 70 cm) with MPS was scheduled for repair of an umbilical hernia. His physical examination revealed dwarfism, short neck, scaphocephaly, macroglossia, and stiff joints in the extremities. His routine laboratory values and chest x-ray were normal. Echocardiogram revealed mild mitral valve prolapse and mitral insufficiency. General anesthesia was induced with 20 mg propofol. Sevoflurane 2% and remifentanyl (0.1-0.05 mcg kg⁻¹ min⁻¹) was used for anesthesia maintenance. After the adequate tidal volume was obtained with mask ventilation, intubation was performed without problem. After the intubation, atracurium besylate was used for neuromuscular blockage. The surgery took nearly one hour and it was completed uneventfully. The extubation was performed after the neuromuscular blockage was reversed and adequate tidal volume was observed. However, after the extubation, rapid desaturation and laryngospasm occurred. The patient was sedated with propofol and mask ventilation was continued manually with 100% O₂ and dexamethasone was administered intravenously. Excessive secretion was aspirated and he was observed in the operation room until adequate ventilation was obtained.

DISCUSSION

The incidence of Maroteaux-Lamy syndrome is 1/100 000. The arylsulfatase β enzyme deficiency causes accumulation of dermatan sulfate in the skin, brain, heart, bone, liver, spleen, blood vessels, cornea, and tracheobronchial airways.³ The physical symptoms of the syndrome include macrocephaly, coarse face, macroglossia, tongue protrusion, mouth breathing, excessive tracheobronchial secretions, and frequent upper respiratory infection. MPS deposition in the soft tissues of the upper airway may cause acquired laryngotracheobronchomalacia and obstructing tracheal mucopolysaccharide deposits. The temporomandibular joints are stiff, the larynx shifts anteriorly and cephalad with age and further MPS deposition occurs in the upper airway.³ Another factor for difficult airway management is neck instability, which is demonstrated as a radiological subluxation of C1 on C2: odontoid dysplasia. Odontoid dysplasia may dislocate to atlas anteriorly and cause spinal cord compression. →

The establishment and maintenance of the airway is the major problem for these patients. The 9-year-old patient was Mallampati III and his intubation was more difficult than the 5-year-old patient. Khan et al. emphasized that increased age and weight correlates with increased airway difficulty.⁴ Meanwhile, other reports showed that there was no clear data about higher incidence of airway problems related to age.² These problems may worsen due to the progressive nature of somatic involvement. Other problems for these groups of patients are upper airway obstruction and laryngospasm after extubation.^{1,4} MPS infiltration of the lips, tongue, epiglottis, tonsils, adenoids, and lower airway may predispose the complications. We observed laryngospasm with the 5-year-old patient after extubation because of the excessive amount of secretions, big tongue, and the deposition of mucopolysaccharidosis around the upper airway and trachea. We sedated the patients with propofol 1 mg kg⁻¹ and administered 100% O₂ while the patient was in a seated position. Preoperative atropine administration, perioperative hydration, and postoperative oxygen with humidity also minimized the problem.

The patients with MPS may have some spinal deformities that cause epidural block failure. Vas et al. reported regional block failure even though the correct epidural catheter placement was used.⁵ It was speculated that the deposition of MPS in the epidural space or in the sheaths of the nerve fibers prevented the local anesthetic effect. Our epidural experience was problematic because of kyphoscoliosis at the level of L4-T12. The epidural insertion was successfully

performed at the level of T12-L1, but the blockage started later than expected. The MPS deposition of nerve or epidural space may affect the quality, beginning, and duration of the blockage.⁵ Spinal deformities may affect the distribution of local anesthetic drugs and may change the level of the blockage.⁶ In the second patient, we preferred the intravenous analgesia method for both the perioperative and postoperative periods to compare to the regional methods. The patient was stable with remifentanyl infusion during the perioperative period, and efficient analgesia was established with tramadol 1 mg/kg after the operation. The vertebrae and thoracic skeletal deformities, which cause obstructive and restrictive ventilatory defects, impact the postoperative airway complications in MPS patients with copious airway secretions, big tongue, and soft tissue deposition. Our patient with spinal deformities had pneumonia 48 hours after the operation and he was treated with antibiotics and physiotherapy for two weeks.

Children with mucopolysaccharidosis are a high risk group, with an estimated 20% perioperative mortality rate.¹ The major problems are with intubation and postoperative airway difficulties.⁷ The respiration problem may continue during the postoperative period, especially in patients with severe spinal deformities. The regional block practice may be problematic and less successful than expected. It is important to evaluate the patient and to take proper precautions to decrease complications. The management of these patients also requires the attention of an experienced anesthesiologist to greatly enhance the probability of success.

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✓	DELIVERING DATE: 26 / 01 / 2011 • ACCEPTED DATE: 16 / 05 / 2011

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