Case Report

Airway management of mucopolysaccharidosis with cervical spine involvement

Altan Sahin, MD, Didem Dal, MD, Turgay Ocal, MD, Ulku Aypar, MD.

ABSTRACT

Mucopolysaccharidoses are a group of inherited disorders occasionally accompanied by cervical spine involvement complicating tracheal intubation. In this study, we review and discuss 5 cases of mucopolysaccharidosis with cervical spinal involvement.

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Mucopolysaccharidoses (MPS) are a group of inherited disorders caused by incomplete degradation and storage of acid mucopolysaccharides. Abnormal accumulation of mucopolysaccharides in the tissues characterizes these disorders. Syndromes may be associated with skeletal abnormalities that may require surgical correction. Particularly, the cervical spine involvement may influence the anesthetic management. Atlantoaxial subluxation or translocation with spinal cord compression occurs in Morquio-Brailsford syndrome (MPS IV) because of underdevelopment of the odontoid process. Maroteaux-Lamy syndrome (MPS VI) is also a rare disorder that might be associated with cervical spine abnormalities. Airway management in these patients deserves special attention as careless intubation attempts may complicate the neurological status.

In this report, we review and discuss the airway management and associated problems of 5 cases of MPS with cervical spinal involvement.

Case Reports. ***Patient One.*** A 10-year-old male patient with the diagnosis of MPS IV was scheduled to have left scrotal hernia and right inguinal hernia repair. This 15 kg and 90 cm patient was hypotonic and had a short neck, pectus excavatum and extremity deformities. Anesthesia was induced with thiopentone sodium and succinylcholine. Direct orotracheal intubation was performed cautiously with MacIntosh 2 laryngoscope blade as he had a short neck and possible craniovertebral junction anomaly. After the 45-minute operation, he was easily extubated and transferred to the pediatric surgery ward devoid of any complications.

Patient 2. A 20-year-old, 28 kg, 115 cm male patient with the diagnosis of MPS IV was scheduled for diaphragmatic hernia repair. He had diastasis recti, pectus carinatum and a short neck. Thiopentone sodium and succinylcholine were used in anesthetic induction. As he had a craniovertebral junction anomaly that might complicate orotracheal intubation, he was intubated with MacIntosh 3 laryngoscope blade avoiding extreme neck movement. After the 95-minute operation, he was easily extubated and transferred to the pediatric surgery ward without any complications.

Patient 3. A 14-year-old male patient with the diagnosis of Morquio-Brailsford syndrome was scheduled for surgery for atlantoaxial subluxation and spinal stenosis. This 18 kilogram and 95 cm high patient had micrognathia, and his mouth could not be opened more than 2 centimeters. His neck

From the Department of Anesthesiology, Faculty of Medicine, Hacettepe University, Ankara, *Turkey*.

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Address correspondence and reprint request to: Dr. Altan Sahin, Assistant Professor of Anesthesiology, Department of Anesthesiology, Faculty of Medicine, Hacettepe University, Ankara, *Turkey*. Tel. +90 (312) 3051255. Fax. +90 (312) 3109600. E-mail: asahin@hacettepe.edu.tr

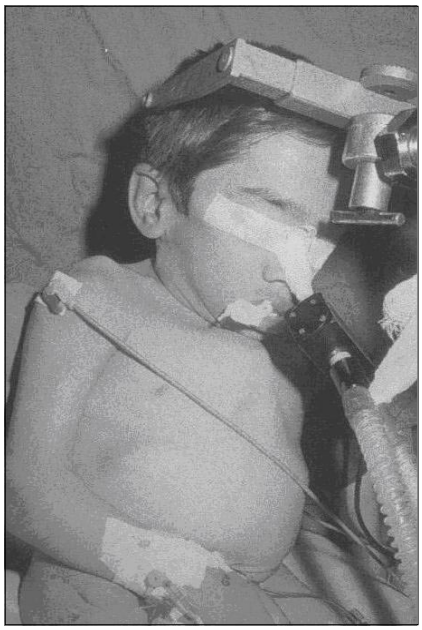

Figure 1 - Nasally intubated patient with Morquio syndrome.

movement was limited, and he had a pigeon chest, prominent thoracal lordosis and lumbar kyphosis. He had a decrease in muscle tone and a mild motor deficit prominent in the proximal muscles. He rejected awake intubation; thus, a fiberoptic intubation under deep sedation was planned. After premedication with atropine and diazepam, routine monitoring and intravenous access; sedation was initiated with intravenous (IV) propofol 2 $mg/kg^{-1}/hr^{-1}$. Topical lidocaine 10% was sprayed on the tongue and pharynx and a nasal decongestant was applied to both nostrils immediately. Pediatric fiberoptic bronchoscope (Storz, Germany) was inserted through the right nostril and larynx was visualized easily. Two milligrams of vecuronium bromide were administered, and the patient was intubated without difficulty (**Figure 1**). After the end of the surgery, he was extubated in the operating theatre without any additional neurological deficits. He spent an uneventful night in the neurosurgical intensive care unit, where he was transferred to the pediatric neurosurgery ward the next day and discharged from the hospital on the 7th postoperative day.

Patient 4. A 17-year-old, 40 kg, 120 cm male patient was scheduled for posterior decompression surgery. He and his older brother had the diagnosis of MPS VI (Maroteaux-Lamy disease) and his cervical and lumbar MRI showed compression of the dural sac but not the spinal cord. Movement of his neck was limited, and he gave a history of cancellation of a hip arthroplasty operation because of intubation difficulty in another center. After premedication with atropine and midazolam and routine monitoring; anesthesia was induced with sevoflurane. Topical lidocaine 10% was sprayed to the tongue and pharynx and a nasal decongestant was applied to both nostrils immediately. Fiberoptic bronchoscope was inserted through the right nostril, and the larynx was visualized easily. Four milligrams of vecuronium bromide were administered, and the patient was intubated without any difficulty. After the operation he was extubated in the operating theatre without any additional neurological deficits. He spent the night in neurosurgical intensive care unit and was transferred to the pediatric neurosurgery ward the next day without any problems.

Patient 5. A 24-year-old, 25 kg, 100 cm high male patient admitted to our hospital with the complaint of syncope with flexion of his neck. He had the diagnosis of Morquio-Brailsford syndrome at the age of 18 months. He was scheduled for decompression and posterior fusion. After monitoring and intravenous access, a nasal decongestant was applied to both nostrils. The patient was easily ventilated with the face-mask after induction with propofol 100 milligrams. Fiberoptic bronchoscope was inserted through the right nostril. Tonsils and the larynx could not be visualized easily. The perilaryngeal tissue was infiltrated with mucopolysaccharides, and the adult fiberscope could not be advanced into the larynx in 2 attempts. As the pediatric bronchoscope was not available at the time, a senior anesthesiologist intubated the spontaneously breathing patient using a Macintosh 2 blade with a 5.5 mm internal diameter endotracheal tube. After the operation the patient was transferred to the intensive care unit intubated. Attempts to extubate the patient were unsuccessful and a tracheotomy was performed. He was discharged from hospital on the 80th postoperative day because of respiratory problems. He was admitted to the emergency services 45 days after his discharge with cardiopulmonary arrest. He responded to cardiopulmonary resuscitation, but died the next day in the intensive care unit.

Discussion. Five cases of MPS with cervical involvement were operated under general anesthesia for various surgeries. Direct orotracheal or fiberoptic intubation techniques were used. According to our observations, presence of experienced staff in both techniques and careful manipulation of the neck and the airway are essential to avoid complications. The most common anesthesia related problem encountered in patients with MPS is the establishment and maintenance of an adequate airway in the perioperative period.[1] The potential airway problems are usually in the upper airway because of mucopolysaccharide infiltration of the lips, tongue, epiglottis, tonsils and adenoids. Additionally, obstructive and restrictive

ventilatory diseases result from abnormal laryngeal and tracheal cartilage, airway secretions and skeletal deformities involving vertebrae and thorax.[2,3] The complications we met in patient 5 might be due to mucopolysaccharide deposition in the upper and lower airway. Abnormal pharyngeal structure made the fiberoptic intubation impossible, and postoperative respiratory complications were possibly because of tracheomalacia induced by abnormal tracheal cartilage structure.

Craniofacial abnormalities, stiffening of the temporomandibular joints, micrognathia and anteriorly positioned larynx may complicate laryngoscopy and intubation.[4] Our third patient had micrognathia and temporomandibular joint involvement, which made direct laryngoscopy impossible. Fortunately, an easy and smooth fiberoptic intubation could be performed in this patient without complication under premedication and deep sedation.

Involvement of the cervical spine may lead to odontoid hypoplasia and potential atlantoaxial subluxation, which may necessitate careful intubation with minimal positioning of the head and neck.[5] A fatal outcome has been reported in a child with Morquio syndrome developing total cervical cord transection related to endotracheal intubation.[6] Different techniques of establishing the airway have been introduced including awake or sedated intubation, laryngeal mask airway, retrograde intubation, fiberoptic intubation and tracheostomy.[7-9] The critical point in consideration is to secure the airway while preventing further spinal injury. Fiberoptic intubation enables intubation without movement of the cervical spine preventing neurological injury.

The first 2 patients in this report were the cases before the fiberoptic intubation was possible in our institution. After these 2 patients, our first choice technique in these patients was awake fiberoptic intubation. However, this method is not accepted by most of our patients. Deep sedation without respiratory depression or general anesthesia induction are performed as alternatives to the awake technique. Both techniques have the risk of airway obstruction by the collapsing of the large tongue and supraglottic tissue. A gentle jaw thrust maneuver, or pulling the tongue out of the mouth smoothly may help to open the airway and better visualize the larynx in these patients.[10] The key point is to prevent neurological injury due to extreme flexion or extension of the neck.

Our patients were of various ages, and the most difficult intubation was encountered in the oldest. This was probably due to more mucopolysaccharide accumulation in the upper airways due to his age. The trachea was difficult to discriminate in this patient, and the internal diameter 6 mm tube could not be advanced into the trachea. The poor visualization with the fiberoptic bronchoscope was due to the mucopolysaccharide infiltrated tissue. Airway management of MPS with cervical spine involvement needs special attention. Fiberoptic tracheal intubation seems to be the method of choice to prevent neurological injury where available, but it might be problematic in patients with extreme mucopolysaccharide deposition of the upper airway.

In conclusion, fiberoptic intubation is advisable in patients of MPS with cervical involvement. However, when this technique is not available or for excessive mucopolysaccharide deposition in the airway, orotracheal intubation may be cautiously performed. During orotracheal intubation, avoiding extreme neck movement, applying inline neck stabilization and an experienced anesthetist are necessary. Also after the operation, mechanical ventilation in the intensive care unit might be required.

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