



Case Report

Deep Sedation in Difficult Airway Patient: Case Report About Dental Treatment of Hurler Scheie Syndrome (MPS IH/IS) Patient

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Abstract

There are a wide range of special care patients in dentistry anesthesia. As a consequence of this condition, rare syndromes are encountered. Hurler-Scheie syndrome is an inherited disorder of mucopolysaccharide metabolism, which is caused by a defect in genetically controlled pathways of lysosomal degradation. As a result, mucopolysaccharide accumulation occurs in various tissues and organs. Structural and functional deficits resulting from Hurler-Scheie Syndrome can lead to difficulties in providing safe airway, especially during anesthetic applications. There is limited data on the literature about the deep sedation practice in patients with Hurler-Scheie syndrome. We aimed to describe our experience of deep sedation for dental treatment in a child with Hurler-Scheie syndrome.

Keywords: Deep sedation, dental treatment, difficult airway, hurler scheie syndrome

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Mucopolysaccharidoses (MPS) are rare seen hereditary metabolic diseases caused by genetic disorders in lysosomal enzyme production.^[1] MPSs are classified into seven types (I, II, III, IV, VI, VII and IX) according to the enzyme affected. Mucopolysaccharidosis I is an autosomal recessive disease caused by the deficiency or absence of the lysosomal hydrolase alpha-L-iduronidase enzyme.^[2] The worldwide incidence of Hurler's syndrome is 1: 100000.^[3] The cases have large head structures due to hydrocephalus. The syndrome has distinctive features such as dwarfism, mental retardation, skeletal anomalies, hernia, hypertension, cardiac diseases, hepatosplenomegaly, corneal opacities, mucopolysaccharide accumulation changes in the soft tissues of the pharynx and larynx, short thick neck, kyphosis, decreased joint mobility and flexion contractures.^[4, 5] Structural and functional deficits resulting from Hurler-Scheie Syndrome can lead to difficulties in providing safe

airway, especially during anesthetic applications. There is limited data on the literature about the deep sedation practice in patients with Hurler-Scheie syndrome. We aimed to describe our experience of deep sedation for dental treatment in a child with Hurler-Scheie syndrome.

Case Report

A 10-year-old male patient who was planned to undergo dental treatment by anesthesia was seen in preoperative examination. Rough face and short stature were the first to be noticed as physical properties of patient. His mallampati score was evaluated as class IV (Scoring system based on the visibility of pharynx structures while the patient is sitting, mouth open, tongue is out and patient is not emit sound. Class I: soft palate and uvula visible; Class II: soft palate and one portion of uvula visible; Class III: soft palate vis-

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ible; Class IV: soft palate not visible). In the continuation of the examination, a large and big tongue, cataract-like eyes and plump and swollen lips were noted. During the preoperative examination, the patient was found to be mentally depressed and the family confirmed this. The patient has previous scoliosis, adenoid, tonsil, inguinal and umbilical hernia operations. General pediatric consultation was requested for the patient who was diagnosed with Hurler-Scheie syndrome (MPS IH/IS) and second degree of mitral insufficiency according to the history of the patient's family.

For the patient who evaluated by general pediatrics; pediatric metabolism, pediatric cardiology, pediatric neurology and ear, nose and throat polyclinic consultations were requested. The joint decision of these consultations was directed at the treatment of dental treatment with local anesthesia, if possible, in the context of infective endocarditis prophylaxis.

The patient was brought to the surgical service by the family. Infective endocarditis prophylaxis was applied to the patient as suggested and the patient was reevaluated by the pedodontist and anesthesiologist. The pedodontist commented that the patient's dental treatment could not be done with local anesthesia because of patient would not allow operations and that the dental treatment was dangerous in this patient in terms of the instruments used. As an anesthesiologist we made a decision that the procedure could be carried out with under deep sedation with the condition of difficult airway by talking with the pedodontist who would make dental treatments.

Operation was initiated with propofol at a dose of 2 mg/kg after standard monitoring. Nasal mask was applied to the patient being oxygenated by the mask. Patient who was ventilated with nasal mask was followed up with pressure mode of ventilator. Additional intermittent propofol was used to achieve the appropriate sedation depth at which the dental treatment and ventilation could be performed comfortably. The depth of sedation that the pedodontist can comfortably work with is determined by the Ramsey sedation scores and the score is kept between 4-5 (Table 1).^[6] No anesthetic complications (eg. decreasing blood ox-

xygen saturation, arrhythmia, hypotension, aspiration etc.) was encountered during the procedure. Also, as a result of using mouth prob apparatus for dental treatment and giving slight extension position to the head of the patient, the back of the tongue is prevented from falling back and the ventilation continued smoothly. During the procedure 3 teeth extraction and 2 teeth filling were performed to the patient and it took about 30 minutes.

At the end of the operation nasal CPAP mask removed and maneuver of chin lift and jaw thrust made smoothly. Recovery time, the primary outcome, was evaluated by a modified Steward score; a score of ≥ 6 means that the patient is awake or responds to verbal stimuli, has purposeful motor activity, and coughs on command. The time to reach a modified Steward score of ≥ 6 was 25 minutes after deep sedation.

Discussion

The pedodontic patient population to be treated with dental treatment under general anesthesia is composed of patients who are young enough to be unable to cooperate or syndromic patients accompanied by mental retardation. This is why, in dentistry anesthesia, there are quite a wide spectrum of syndromic patients. Especially for patients with mental retardation, the risk of odontogenic infection is higher than that of healthy individuals due to poor oral hygiene and suppression of the immune system.^[7]

Mucopolysaccharidoses (MPS) are a group of metabolic diseases with inherited, chronic, progressive and multi-system involvement that go through glycosaminoglycan accumulation due to a specific enzyme deficiency in lysosomes. Hurler-Scheie syndrome, type of MPS in our case, occurs as a result of alpha-L-iduronidase deficiency. Clinical findings begin around 2 years of age. The most important clinical findings are progressive joint stiffness and widespread degenerative bone disease.^[8] Intralysosomal GAG accumulation can lead to cellular, vascular, tissue and organ dysfunction leading to different degrees of mental and physical retardation and, in severe cases, premature death. Normally, mucopolysaccharides are the component of connective tissue, but in MPSs it is stored much in skin, respiratory tract, cornea, central nervous system, heart, liver, spleen, bone and ligaments. As a result; cardiovascular functions, airway, joint movements, visual acuity, hearing may be affected.^[9] In our case, the influence of the heart, mental, cornea, bone and organ such as liver was the subject. Progressive involvement in many organs and tissues often causes these children to be exposed to many surgical procedures. The most commonly performed surgery was reported as ear nose throat procedure.^[10] In our case the

Table 1. Ramsey sedation score

1. Nervous, agitated and/or restless
2. Cooperative, orientated, quiet patient
3. Only obeying orders
4. Sleeping, hitting the glabella and responding to high voice suddenly
5. Sleeping, hitting the glabella and responding to high voice slowly
6. No response to any of these stimulations

Source: (1).

patient has previous scoliosis, adenoid, tonsil, inguinal and umbilical hernia operations.

MPS patients should be carefully assessed for anesthesia risk factors before anesthesia, and if necessary, should be considered with departments such as pediatrics, cardiology and chest diseases. Our patient was also institutionalized with department of pediatric metabolism, pediatric cardiology, and with department of ear nose throat. Typical anesthesia problems include airway obstructions after induction or extubation, intubation difficulties, intubation failures, urgent tracheostomy needs, cardiovascular and cervical vertebrae problems.^[11] In our case, adenoid and tonsil hypertrophy due to GAG accumulation in soft tissues and associated obstructive sleep apnea (OSAS) was present, and there was a previous history of adenoidectomy and tonsillectomy depending on them. In a previous study; it has been suggested that the possibility of confrontation with difficult airway during anesthesia is high in patients with severe OSAS history.^[12]

Because of the anatomical problems and excessive secretions in MPS patients, face mask and sedation are not recommended.^[13] The medical story of the patient we were presented had a history of OSAS-related adenoidectomy and tonsillectomy and also, in our patient, the procedure was performed using the mouth probe and mouth tampon, and the physician assistant who assisted the dentist applied continuous aspiration. Therefore, no problems were encountered in ventilation with nasal mask under deep sedation. Since the device we used is a nasal mask, the anesthesia and pedodontic procedure has progressed without problems.

The most common cardiac finding seen in patients with MPS is valve disease. Progressive valve thickening secondary to GAG storage is caused by valve regurgitation and/or stenosis. Routine valvular disease severity should be considered in MPS patients because severe aortic and mitral stenosis, as in non-MPS patients, also increases the risk of mortality during operation in MPS patients.^[14, 15] The most common associated valve problems in patients with MPS are mitral and aortic regurgitation.^[16] Our case was also followed by second degree mitral regurgitation and mitral valve prolapse and was using digoxin, lasix and enapril.

Uncooperative, mental retarded or very young children patients may require general anesthesia for dental treatment. But; for patients with additional medical problems or considered difficult intubations, sedation should be the preferred option in condition of difficult airway preparation. In our experience with MPS, sedation was considered suitable for dental treatment. We performed noninvasive mechanical ventilation in pressure control mode to deep

sedated patient and the procedure was completed without any problems.

Disclosures

Informed Consent: Written informed consent was obtained from the parents of patient for the publication of the case report.

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