

Sedoanalgesia During Dental Treatment of a Patient with Sanfilippo Syndrome: A Case Report

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Abstract

Sanfilippo syndrome, a type of mucopolysaccharidosis (MPS), is an autosomal recessive lysosomal storage disease. The disorder is characterized by bone abnormalities (i.e. abnormally long arms and legs when compared with the trunk length), as well as abnormal bone shapes and skin thickening. In general, patients with Sanfilippo syndrome have sharp and coarse facial features, thickening of the lips and enlargement of the mouth and tongue. Hypoplasia of hard dental tissue is also present. The parents of a 5-year-old female patient were referred to Konya Oral and Dental Health Hospital because of tooth pain. A full dental examination could not be conducted because the patient was not cooperative. Thus, we decided to perform the patient's treatment under general anaesthesia. As the patient had a history of difficult intubation and the planned dental procedures could be completed in a short time, under sedoanalgesia.

Keywords: Sanfilippo; Mucopolysaccharidosis; Sedoanalgesia; Dental Treatment

Introduction

Sanfilippo syndrome, a type of mucopolysaccharidosis (MPS), is an autosomal recessive lysosomal storage disease. The disorder is caused by a lack of lysosomal alpha N- acetylglucosaminidase [1]. Due to the deficiency of N-acetylglucosaminidase, heparan sulphate accumulates in various tissues of the body instead of breaking down. There are four types of Sanfilippo syndrome (A, B, C and D), which are classified according to mutations in different areas of the gene responsible for the production of specific enzymes [2]. The global frequency of Sanfilippo syndrome is estimated at 1 in 70,000 live births [3].

Although the symptoms of MPS differ among patients, there are general similarities in the clinical course of the disorder. Patients have a normal appearance at birth, and the diagnosis is not made until the infant is aged between 6 and 24 months. The diagnosis of MPS is based on the level of heparan sulphate in urine. A blood test to determine enzyme levels or a skin biopsy is required for a definitive diagnosis. Enzyme activity in leukocytes or fibroblasts confirms the diagnosis and allows differentiation between the different subtypes of Sanfilippo syndrome [4-6].

Clinical symptoms appear around the age of 2 years. Physically, patients present with bone abnormalities (i.e. abnormally long arms and legs when compared with the trunk length), as well as abnormal bone shapes and skin thickening [7]. In general, patients have sharp and coarse facial features, thickening in the lips, enlargement of the mouth and tongue, thick and prominent eyebrows, coarse hair and hirsutism. Hypoplasia of hard dental tissue is also observed [8]. Additional clinical symptoms are hepatosplenomegaly, heart involvement, pulmonary hypertension, joint and skeletal deformities and inguinal and umbilical hernias [9]. Central nervous system involvement is one of the most obvious characteristics of the syndrome. In older patients, psychomotor retardation and delayed speaking skills are evident, in addition to aggressive behaviour, mental retardation, uncontrolled hyperactivity and sleep disorders. Mobility disappears in the last stage of the disease, with no response to environmental stimuli. A nasal drip and pain can accompany the symptoms [8,10,11].

Thus far, there is no efficient treatment for Sanfilippo syndrome. Therapies to address the symptoms of the syndrome include enzyme replacement treatment, hematopoietic cell transplantation, gene therapy, and substrate deprivation therapy and enzyme development studies [2]. We report the case of a 5-year-old female with Sanfilippo syndrome whose dental treatment was performed under sedoanalgesia.

Case Report

The parents of a 5-year-old female were referred to Konya Oral and Dental Health Hospital because of tooth pain. During the anamnesis obtained from the patient's parents, they confirmed that the child had sanflippo syndrome, mucopolysaccharidosis tip III b. The clinical examination showed short arms and legs, coarse facial features, thickening of the lips, short stature, thickening of the skin, mitral regurgitation grade 3 (managed by enapril in tablet form), hepatosplenomegaly, diabetes mellitus (regulated with diet), hyperactivity, spasticity and mental motor retardation.

The patient's extra-oral examination showed coarse facial features, thick lips and a large mouth. The intra-oral examination showed macroglossia. A dental examination could not be completed because the patient was not cooperative. For this reason, we decided to perform the patient's treatment under general anaesthesia. The patient had a history of an adenotonsillectomy with tracheal intubation when she was 18 months old. The case records revealed that intubation was difficult during the procedure due to a laryngeal obstruction. The patient was subsequently diagnosed with Sanfilippo syndrome. Due to the patient's history of difficult intubation, we decided to perform the dental treatment under sedoanalgesia.

Prior to the procedure, haemogram, glucose, liver function and kidney function tests were performed, and the results of all these tests were normal. Echocardiography showed symptoms of enlargement in the left cardiac cavity and increased wall thickness, with mitral regurgitation grade 3. The patient had a Mallampati score of 4. Anaesthesia was planned according to the guidelines of the American Society of Anesthesiologists for patients with ASA 3.

Oral midazolam (0.2 mg/kg) was administered to the patient as premedication prior to the procedure. Intravenous (IV) route was administered with 24 G intracath. The patient was monitored by ECG (electrocardiogram) pulse pressure; oxygen saturation and arterial pressure were monitored non-invasively. Cefazolin (40 mg/kg) was administered intravenously as infective endocarditis prophylaxis. Thirty minutes later, the patient's Ramsey sedation scale was 3, and the patient was taken to the operation theatre and monitored. A balanced electrolyte IV solution was started, and 0.8 mg/kg of ketamine and 0.1 mg of atropine were administered intravenously. Nasal oxygen (3 L/min) was started. After the induction of infiltrative and/or regional local anaesthesia (80 mg of articaine hydrochloride plus 0.2 mg of epinephrine), depending on the dental procedure to be performed, 0.04 mg/kg of midazolam was added via the IV route. The patient's respiration became superficial following the administration of midazolam, and a jaw thrust manoeuvre intervention was made. The procedure commenced after the patient's respiration recovered and she became immobile. Ketamine (10 mg) was added to the anaesthesia protocol 10 min later.

The intra-oral examination was undertaken when the patient was immobilized. The examination revealed poor oral hygiene and intense plaque accumulation, especially in the lower anterior teeth. Extreme crown damage was seen in teeth 84, 85 and 74 according to the tooth numbering system of the World Dental Federation and crown decay was seen in teeth 64, 54 and 75. Teeth 84, 85 and 74 were extracted. Teeth 64, 54 and 75 were treated with class I amalgam restorations.

The procedure took 15 min. At the end of the procedure, flumazenil (0.04 mg/kg) was administered intravenously and the patient was taken to the recovery room. Thirty minutes later, the patients had an Aldrete score of 10. In total, the treatment time was 150 min (i.e., from the time of anaesthesia induction to discharge).

Discussion

The incidence of difficult intubation is very high in patients with mucopolysaccharidosis [12]. Airway management difficulties in these patients during anesthesia are due to various factors, including a coarse face, macroglossia, a short neck, cervical thickening due to mucopolysaccharide accumulation, odontoid dysplasia, thickening in the otorhinolaryngologic area and tracheal stenosis. Thus, the patient assessment should include a detailed physical examination, and necessary consultations should be asked for pathologies in other systems. In selecting the most suitable type of anaesthesia for the patient, all necessary precautions should be taken. In patients with a history of difficult intubation, the use of intubation devices, such as laryngeal masks and fiberoptic intubation, should be avoided [13]. In addition, in such cases, an otorhinolaryngologist should be present in the operation theatre in case an emergency tracheostomy is required [13].

After the dental surgery, care should be taken while extubating the patient. During extubation, a range of factors, such as subglottic oedema and post-obstructive pulmonary oedema, should be taken into consideration. If necessary, the patient should be transferred to the intensive care unit after the procedure [8].

A previous study reported that sedoanalgesia (inhalation or IV) can cause severe hypoxemia in MPS patients by causing airway obstruction [12]. The same study reported that midazolam and fentanyl could be safely used because they acted as antagonists of undesirable effects (i.e. hypoxemia) of sedation. Ketamine is also safe to use, especially during fiberoptic intubation, as it does not cause airway obstruction. Other agents, such as atropine or glycopyrrolate, can be useful to decrease secretions and not to block oral vision [12]. In the present study, we added atropine to the sedoanalgesia protocol to prevent ketamine-induced increases in the *secretion of*.

Various methods of airway intubation have been used in MPS patients. Such patients require a multi-disciplinary approach. As

shown by the literature, a full history of the pre-operative status of the patient, together with knowledge of MPS disorders, can assist intra-operative and post-operative approaches. In 141 anaesthesia procedures involving 30 MPS patients, the possibility of seeing vocal cords in direct laryngoscopy was inversely proportional to the age of the patient, and intubation was easier in patients weighing less than 12 kg and more than 15 kg [14]. In two sisters with MPS aged 5 months old and 13 years, respectively, in whom anaesthesia was administered prior to hernia surgery, direct laryngoscopy was successfully used for airway intubation in the younger sister and the patient was extubated without any problems after the procedure. However, in the 13-year-old sister, direct laryngoscopy failed three times, and an emergency tracheostomy was required to provide an airway. At the end of the operation, extubation was unsuccessful and the patient was transferred to the intensive care unit. Subsequently, the patient developed pulmonary oedema and respiratory acidosis and she died on post-operative day 8 [15]. In a 3-year-old child with MPS type 1 who was referred to the emergency services for foreign matter aspiration, bronchoscopy was implemented successfully using midazolam, sevoflurane, remifentanyl and rocuronium [16]. However, after the foreign matter was removed, a laryngeal and facial mask used for maintaining the patient's airway failed. Direct laryngoscopy was successful in airway intubation on the third try. In a 20-year-old female diagnosed with MPS type 4 who underwent foramen magnum decompression, airway intubation was successful using fiberoptic intubation, and extubation was conducted without any problems [17]. In a 4-year-old patient with MPS type 3 who underwent circumcision, a laryngeal mask airway was successful in airway management [13].

Conclusion

In our patient, pre-operative hemodynamic findings were stable. Due to the difficult intubation history of the patient as revealed by the anamnesis and the fact that the planned dental procedures could be completed in a short time, the patient's procedure was performed under sedoanalgesia. In continuation anaesthesia, ketamine was safe and effective.

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