Tooth Extraction Under Sedation in a Patient Diagnosed with I-Cell: Case Report

I-Cell Tanılı Hastada Sedasyon Altında Diş Çekimi

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ABSTRACT I-cell disease (Mucolipidosis Type II) is a lysosomal storage disorder, caused by a deficiency of the enzyme, GlcNAc-phosphotransferase. Due to the possible occurrence of complications resulting in preoperative mortality, careful assessment, experience and preparation are necessary for patients diagnosed with I-cell disease during anesthesia procedures of these patients. A difficult airway is a common clinical problem and one of the most significant causes of major anesthesia-related morbidity. During dental treatment for patients with mental and psychiatric disorders, sedation methods combined with local anesthesia is a safe alternative to general anesthesia. This case report presents the anesthesia method used during the tooth extraction of a 3- year-old, mentally retarded patient who was diagnosed with I-cell disease.

Key Words: Deep sedation; tooth extraction

ÖZET I-cell (Mucolipidozis Tip II) hastalığı; otozomal resesif kalıtımlı GIcNAc fosfotransferaz enziminin eksikliğinden kaynaklanan lizozomal depo hastalığıdır. I-cell tanılı hastalarda perioperatif mortaliteyle sonuçlanabilecek ciddi komplikasyonların ortaya çıkabilmesi nedeniyle, hastaların anestezi uygulamalarında dikkatli değerlendirme, deneyim ve hazırlık gereklidir. Zor hava yolu anestezide morbidite ile sonlanabilen en önemli klinik problemdir. Mental retarde hastalarda diş tedavisinde lokal anestezi ile kombine edilen sedasyon yöntemi genel anesteziye güvenli bir alternatiftir. Bu olgu sunumunda, 3 yaşında, I-cell hastalığı tanılı mental retarde hastada diş çekimi sırasında uyguladığımız anestezi yönetimimiz tartışılmıştır.

Anahtar Kelimeler: Derin sedasyon; diş çekimi

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I-cell disease (Mucolipidosis Type II) is an autosomal recessive disorder of lysosomal storage caused by a deficiency of the enzyme GlcNAc-phosphotransferase.[1] This disease was defined by Leroy et al. for the first time in 1969.[2] Clinical and radiological findings are similar to those of Mucolipidosis Type I (Hurler syndrome); however, Mucolipidosis Type II can be differentiated by early symptoms and the lack of excreted mucopolysaccharides in the urine.

Babies diagnosed with I-cell disease have low birth weight, possess hypotonic muscle weight, and appear to have course facial features. In addition to mental and physical retardation, in I-cell patients, macroglossia,

hyperplasia in the gums, adenoids, tonsils and nasal tissues, a narrowed trachea, vocal cord thickening, a short neck, restricted cervical motion, and mucoid secretions leading to airway obstruction may cause difficulties in ventilation and intubation. As children with I-cell disease mature, their airways become increasingly limited due to easily traumatized narrowed passages.[2]

In these patients, intubation and maintenance of the airway are the most important problems for the anesthesiologists.[3] Due to the fact that serious complications, including the potential for preoperative mortality, may occur in patients diagnosed with I-cell disease, careful assessment, experience and preparation are warranted during the anesthesia procedures.

In patients with mental and psychiatric disorders, sedation methods combined with local anesthesia are a safe alternative to general anesthesia, during dental treatment.[4] An ideal sedation method is expected to reduce anxiety, be non-invasive and reliable, have an amnesic effect, and provide immobility and analgesia. While providing these effects, the sedation method must not depress the patient's respiration or cardiovascular reflexes. The administered drugs must take affect quickly but the lasting effects of the drugs must be short and the dose must be adjustable according to the patient's response.[5] Although many drugs, such as midazolam, ketamine, opioids, and propofol, are commonly used for sedation, anesthesiologists hold no consensus on the most appropriate approach.[5,6] Two other alternative sedation applications are nitrous oxide (N_20) and sevoflurane inhalation, which can be used for deep sedation applications.[7-10]

There are numerous cases in the literature in which anesthesia applications are reported to carry high risks in lysosomal storage disorders. In these cases, airway safety was maintained by methods of either intubation with fiberoptic bronchoscopy, LMA, or emergency tracheostomy.[11-13]

This case report aims to present a patient diagnosed with I-cell in whom tooth extraction under sedation was performed without intubation, unlike the cases reported in the literature.

CASE REPORT

This case involved a 3-year-old, mentally retarded female patient, with a weight of 11 kg and height of 85 cm, who was diagnosed with I-cell disease. After consent was received from the patient's relatives, anesthesia under sedation was planned in order to maintain immobility during tooth extraction at the Oral and Jaw Surgery Clinic, Faculty of Dentistry, XXX University (Figure 1). During the preoperative assessment, it was discovered that this patient, who had consanguineous marriage in her family history, was born spontaneously and vaginally. At birth, she measured 50 cm in length and weighted 3400 grams. She had a course facial appearance, her hands and feet were short-blunt, and her voice was hoarse. When she was 18 months old, she contracted bronchitis and, upon examination, she was diagnosed with I-cell disease.

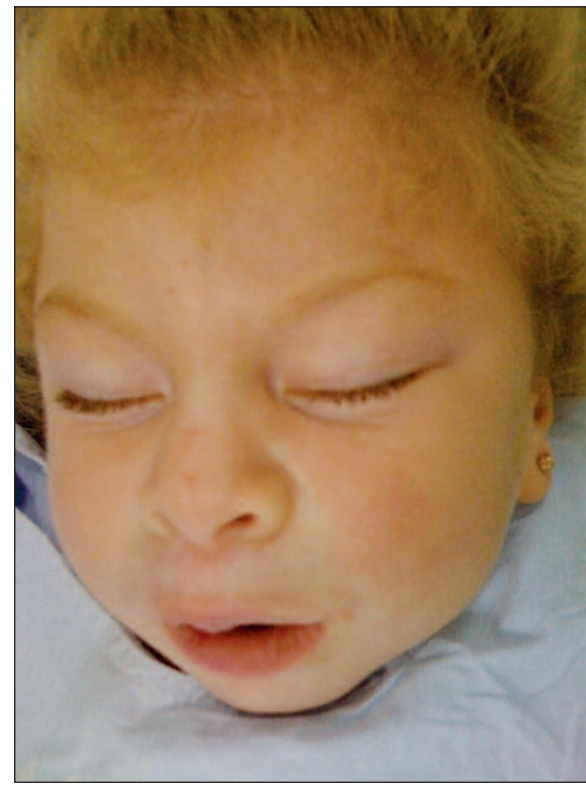

FIGURE 1: Patient diagnosed with I-cell.
(See for colored form http://anestezi.turkiyeklinikleri.com/)

During the preoperative head and neck examination, the patient was determined to have a thick, course facial structure, an enlarged tongue and lips, limited movement of the head, neck, and jaw joints and a mouth opening of 2.5 cm. Furthermore, she had a Mallampati score of IV. Examination of this patient's eyes revealed bilateral corneal opacity. Findings from the examinations of other organ systems were normal.

The results from the patient's laboratory tests were as follows: hemoglobin: 9.6 g dL^{-1}; hematocrit 29.2%; WBC: 8.4 x103 mm^{-3}, and platelet: 451 x103 mm^{-3}. Urine metabolic screening and metabolic screening using tandem mass spectrometry yielded normal results. The patient had a normal abdominal ultrasonograph. An echocardiograph showed that the patient had a 2-3 mm secundum type atrial septal defect. The patient, who had cranial magnetic resonance imaging consistent with neurometabolic disease, was determined to have a spinous formation indicating a mucolipidose deposit in the anterior of the thoracic-lumbar vertebrae. Based upon the results from the extremity radiograph, the patient was found to have shortening and thickening of the metacarpals and phalanges. A chest radiograph revealed that she also had flattening and thickening of the ribs. In addition to the routine anesthesia preparations, and taking into consideration possible intubation difficulties, and the possible need for fiberoptic laryngoscopy, emergency cricotiroidotomia, and tracheostomy, drugs with inotropic, diuretic, and bronchodilator effects were prepared in the preparation room.

The patient was admitted to the operation room without applying premedication after a 6-hour fasting period.

The patient was taken to the operation room and followed using a monitor (Datex-Ohmeda, Helsinhi, Finland), which measures noninvasive systolic and diastolic arterial pressure (SAP, DAP), heart rate (HR), peripheral oxygen saturation (SpO_2), a capnograph ($ETCO_2$), and a precordial stethoscope. Respiratory depression (respiration rate below 8≤ min, or apnea lasting more than 15 seconds, or SpO_2 <90), hypotension (initial value dropping by 20%), hypertension (initial value rising by 20%), bradycardia (HR<45/min), tachycardia (initial value rising by 20%) were assessed. The measured values were as follows: HR: 95/min, SAP/DAP: 95/65 mmHg, and SpO_2: 97%.

Sedation induction was applied with a face-mask, at a constant speed, maintaining 0.5-1% sevoflurane spontaneous respiration in a 60% O_2 + 40% N_2O mixture. Liquid infusion, at a rate of 50 mL h^{-1}, was started by establishing a 24 G peripheral intravenous access at the back of the patient's hand. Next, 10 μg kg^{-1} atropine was intravenously administered. Five minutes after anesthesia induction, once the patient reached level 3, as determined by the Ramsey Sedation Scale (Table 1), the dentist infiltratively administered 1 mL local anesthetic (Ultracain DS,HOECHST) on the primary incisors in the upper front area of the patient's mouth (51,61).[14] The reaction shown to local anesthesia and the child's reactions to the tooth extraction were assessed by the scale defined by Houpt et al. (Table 2).[15] The face mask was removed when scale values ≥ 8 were obtained, which meant that the patient was considered to be ready for the procedure.

Tooth extraction started five minutes after local anesthetic infiltration. After two teeth were extracted and sutures were placed, a face mask containing 0.5-1 sevoflurane in a 60% O_2 + 40% N_2O was applied for about two minutes (until the patient reached level 3 as determined by the Ramsey Sedation Scale) and anesthesia was deepened. During the 10-minute surgical

TABLE 1: Ramsay sedation scale.

Score	Response
1	Anxious or restless or both
2	Cooperative, orientated and tranquil
3	Responding to commands
4	Brisk response to stimulus
5	Sluggish response to stimulus
6	No response to stimulus

TABLE 2: Rating scale for movement, crying, and overall behavior.

Score	Movement	Crying	Behavior
1	Continuous treatment difficult	Hysterical	Impossible-procedure cancelled
2	No interference with treatment	Treatment difficult	Poor-treatment difficult
3	None-quiet patient	Mild, no interference	Good-limited difficulty
4		None---quiet patient	Very good-no problems

procedure, two teeth were extracted. In order to maintain the patient's airway safety during the procedure, in-mouth aspiration was applied to avoid aspiration during tooth extraction. Additionally, sutures were placed in both gums after extraction. Throughout the procedure, oxygen support was provided at a rate of 5 L/min using a nasal cannula. No significant change occurred in the patient's blood pressure and pulse rate during the operation (Table 3). Bronchospasm, laryngospasm, and respiratory depression were not observed. Upon completion of the surgery, the patient was awakened with 100% O_2. The patient was monitored in the recovery room for six hours, and then discharged without any complications.

TABLE 3: Hemodynamic parameters of the patients before and after sedation.

	BASELINE	5 min	10 min	15 min
Oxygen saturation (%)	97	96	95	96
Systolic pressure (mmHg)	95	82	102	90
Diasstolic pressure (mmHg)	65	55	60	60
Heart rate (beats/minute)	95	102	105	87
Ramsey sedation scala	6	3	5	3

DISCUSSION

I-cell disease is an autosomal recessive disorder caused by a deficiency in the transfer of many lysosomal hydrolase enzymes. In these patients, intubation and maintenance of the airway are the most important problems for anesthesiologists.[3] Due to the fact that serious complications, potentially resulting in preoperative mortality, may occur in patients diagnosed with I-cell disease, careful assessment, experience, and preparation are warranted during the anesthesia procedures.

As a result of our literature search, we did not find any research focusing on anesthesia methods used in I-cell disease cases in Turkey. Despite the small number of publications presenting data on the anesthesia methods used in I-cell disease, in the literature, clinical similarities with mucopolysaccaridosis provided guidance in the planning and the implementation of the anesthetic method. In mucopolysaccaridosis patients, airway difficulty is the problem most frequently encountered by anesthesiologists.[3] The difficult airway is a common clinical problem and one of the most important causes of major anesthesia-related morbidity.[16] In patients who are expected to have difficult intubations, sedation implementations can provide a safe method of treatment without necessitating general anesthesia and, in recent years, various sedation methods have been developed as an alternative to general anesthesia.[5]

Sedoanalgesia is a procedure applied to provide additional anesthesia and amnesia in patients who receive local anesthesia during their operations. Combinations of ketamine and midazolam have been stated to be among the best and safest techniques for sedoanalgesia. However, the use of midazolam may be associated with paradoxical reactions (delayed recovery, anxiety, behavioral changes, and agitation). Furthermore, irregular breathing patterns, skipped heartbeats, respiratory failure, and unusual or involuntary muscle movements have been observed, rarely, in some children.[17] An infusion of propofol, an ultra-short-acting intravenous anesthetic, has been used successfully for sedation in outpatient oral surgery.[18] However, propofol does cause dose-dependent depression of the cardiovascular and respiratory systems.[19,20] Administration of N_2O/O_2 is widely used to induce both analgesia and

sedation and to improve patient cooperation during dental treatment. Although N_2O inhalation sedation (IHS) enjoys a high 83-96% treatment completion success rate, this does indicate that N_2O HIS is not successful in all cases. The use of N_2O + Sevoflurane was an efficacious and safe HIS method when administered by an anaesthetist.[7]

Our patient did not allow us to conduct tooth extraction under local anesthesia due to mental retardation and expected difficult intubation, based on pre-anesthetic examination. Therefore, we performed the tooth extraction under deep sedation, eliminating intubation-related risks.

Sevoflurane is a non-irritant inhalation agent whose effect starts and ends quickly due to its low solubility. When it is used at sub-anesthetic concentrations, these properties allow the drug to be used in controlled deep anesthesia cases.[21]

In recent years, surprising number of publications have reported on the successful use of 40% N_2O added to low concentrations (0.1-0.3%) of sevoflurane inhalation in the sedation of pediatric cases without any side effects.[8] Lahaud et al. reported that during the dental treatment of cases involving 3-10-year-old pediatric patients, low dose sevoflurane inhalation added to N_2O and O_2 was more effective compared to the application of only O_2 and N_2O.[8]

We administered the 0.5-1% sevoflurane in a 60% O_2+ 40% N_2O mixture to the patient using a face mask. Additionally, the literature emphasized the importance of maintaining breathing, through the use of a mask in patients with difficult airways. As a result, we planned the treatment so that spontaneous respiration would not be lost with the inhalation of the sevoflurane.[22]

Since we used a non-invasive administration for the N_2O-sevoflurane mixture and we could easily change or terminate the sedation level, we believe this combination can safely be used in dental surgical procedures. With regard to successful treatment completion, a study by Soldani et al suggested that inhalation sedation with O_2 and N_2O + sevo IHS was as successful as inhalation sedation with O_2 and N_2O IHS alone. Moreover, no adverse events related to either sedation method were found that required termination of treatment or administration of any emergency medications.[7] Hall et al performed odontotherapy in 22 patients under sedation with 30-70% N_2O and 0.2-0.8% Sevoflurane.[23] No statistically significant changes in SAP, DAP, and SpO_2 were observed at any dosage of either gas compared with the combined control sessions and with each subject acting as his/her own control. Since we also gave the patient 0_2 through nasal cannula during the operation, no respiratory depression was observe in our monitoring.

In mucopolysaccharidosis cases, many researchers recommend the use of anticholinergics in order to insure control of intra-oral secretions.[22] Due to the clinical similarities between mucopolysaccharidosis and I-cell disease, after intravenous cannulation, we administered atropine to our patient in order to provide an anticholingeric effect.

In these patients, hemodynamic changes present another expected set of problems. Toda et al.report encountering complete heart block during the anesthesia applications in 4-year-old patients diagnosed with Type VII MPS.[24] Despite the fact that the echocardiograph findings for our patient led to a diagnosis of a 2-3 mm secundum type atrial septal defect, we did not encounter any problems during the hemodynamic monitoring. The absence of hemodynamic problems may be associated with the fact that the volatile agent we selected was easily titratable.[25]

Since Sjogren et al reported on a number of deaths in cases of patients diagnosed with mucopolysaccaridosis who had received opioid and bezodiazepine premedication, we avoided the use of these agents during sedation.[26] We preferred sevoflurane inhalation, which is an alternative method used for deep sedation.

CONCLUSION

In conclusion, it should be kept in mind that establishing general anesthesia and a safe airway in patients with I-cell disease is always a big risk;

however, as long as the administration route provides sufficient immobility and analgesia for the surgical procedure, a combination sedation method should be taken into consideration as a good alternative in cases where intubation is expected to be difficult. During sedation, in order to provide a safe airway, all equipment, including those pertaining to emergency tracheostomy, should be kept ready, taking into consideration possible respiratory depression.

REFERENCES

1. Dierks T, Schlotawa L, Frese MA, Radhakrishnan K, von Figura K, Schmidt B. Molecular basis of multiple sulfatase deficiency, mucolipidosis II/III and Niemann-Pick C1 disease - Lysosomal storage disorders caused by defects of non-lysosomal proteins. Biochim Biophys Acta 2009;1793(4):710-25.
2. Baines DB, Street N, Overton JH. Anaesthetic implications of mucolipidosis. Paediatr Anaesth 1993;3(5):303-6.
3. Benumof JL. Anesthesia&Uncommon Diseases. 4th ed. Philadelphia: WB Saunders Company; 1998. p.533.
4. Ryder W, Wright PA. Dental sedation. A review. Br Dent J 1988;165(6):207-16.
5. Haas DA. Oral and inhalation conscious sedation. Dent Clin North Am 1999;43(2):341-59.
6. Clarkson K, Power CK, O'Connell F, Pathmakanthan S, Burke CM. A comparative evaluation of propofol and midazolam as sedative agents in fiberoptic bronchoscopy. Chest 1993;104(4):1029-31.
7. Soldani F, Manton S, Stirrups DR, Cumming C, Foley J. A comparison of inhalation sedation agents in the management of children receiving dental treatment: a randomized, controlled, cross-over pilot trial. Int J Paediatr Dent 2010;20(1):65-75.
8. Lahoud GY, Averley PA. Comparison of sevoflurane and nitrous oxide mixture with nitrous oxide alone for inhalation conscious sedation in children having dental treatment: a randomised controlled trial. Anaesthesia 2002;57(5):446-50.
9. Haraguchi N, Furusawa H, Takezaki R, Oi K. Inhalation sedation with sevoflurane: a comparative study with nitrous oxide. J Oral Maxillofac Surg 1995;53(1):24-6.
10. Wang CY, Chiu CL, Har KO, Chan C, Rahman ZA. A comparative study of sevoflurane sedation with nitrous oxide sedation for dental surgery. Int J Oral Maxillofac Surg 2002;31 (5): 506-10.
11. Walker RW. Management of the difficult airway in children. J R Soc Med 2001;94(7):341-4.
12. Kempthorne PM, Brown TCK. Anaesthesia and the mucopolysaccharidoses: A survey of techniques and problems. Anaesth Intensive Care 1983;11(3):203-7.
13. M Mahfouz AK, George G, Al-Bahlani SS, Al Nabhani MZ. Difficult intubation management in a child with I-cell disease. Saudi J Anaesth 2010;4(2):105-7.
14. Ramsay MA, Savege TM, Simpson BR, Goodwin R. Controlled sedation with alphaxalone-alphadolone. Br Med J 1974;2(5920):656-9.
15. Houpt MI, Weiss NJ, Koenigsberg SR, Desjardins PJ. Comparison of chloral hydrate with and without promethazine in the sedation of young children. Pediatr Dent 1985; 7(1):41-6.
16. Uzun Ş, Şahin A, Köse A, Aypar Ü. [Fiberoptic intubation via laryngeal mask in a patient with unexpected difficult airway: case report]. Turkiye Klinikleri J Anest Reanim 2008;6(1): 28-32.
17. Bozkurt P. Premedication of the pediatric patient - anesthesia for the uncooperative child. Curr Opin Anaesthesiol 2007;20(3):211-5.
18. Valtonen M, Salonen M, Forssell H, Scheinin M, Viinamäki O. Propofol infusion for sedation in outpatient oral surgery. A comparison with diazepam. Anaesthesia 1989;44(9):730-4.
19. Lippmann M, Mok MS. Propofol causes cardiovascular depression III. Anesthesiology 1990;72(2):395.
20. Taylor MB, Grounds RM, Mulrooney PD, Morgan M. Ventilatory effects of propofol during induction of anaesthesia. Comparison with thiopentone. Anaesthesia 1986;41(8): 816-20.
21. Averley PA, Lane I, Sykes J, Girdler NM, Steen N, Bond S. An RCT pilot study to test the effects of intravenous midazolam as a conscious sedation technique for anxious children requiring dental treatment--an alternative to general anaesthesia. Br Dent J 2004; 197(9): 553-8.
22. Ard JL Jr, Bekker A, Frempong-Boadu AK. Anesthesia for an adult with mucopolysaccharidosis I. J Clin Anesth 2005;17(8):624 6.
23. Hall DL, Weaver J, Ganzberg S, Rashid R, Wilson S. Bispectral EEG index monitoring of high-dose nitrous oxide and low-dose sevoflurane sedation. Anesth Prog 2002;49(2):56-62.
24. Toda Y, Takeuchi M, Morita K, Iwasaki T, Oe K, Yokoyama M, et al. Complete heart block during anesthetic management in a patient with mucopolysaccharidosis type VII. Anesthesiology 2001;95(4):1035-7.
25. Gupta A, Stierer T, Zuckerman R, Sakima N, Parker SD, Fleisher LA. Comparison of recovery profile after ambulatory anesthesia with propofol, isoflurane, sevoflurane and desflurane: a systematic review. Anesth Analg 2004;98(3):632-41.
26. Sjøgren P, Pedersen T, Steinmetz H. Mucopolysaccharidoses and anaesthetic risks. Acta Anaesthesiol Scand 1987;31(3):214-8.