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Sedation for dental treatment of child with angelman syndrome: Case report

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Abstract

Angelman syndrome (AS) is a genetic disorder characterized by intellectual disability, movement or balance disorders, specific abnormal behaviors, and severe speech and language limitations. Many patients with AS may have difficulty with medical evaluations and may require general anesthetics for seemingly innocuous procedures including the dental care. At the same time AS poses significant challenges for the anaesthetist due to sensitivity to anesthetic agents and the craniofacial abnormalities. This paper describes the use of deep sedation for dental treatment provided for a 7-year-old patient with AS.

Keywords: Sedation, angelman syndrome (AS), dental treatment, complications

Introduction

Patients with genetic disorders present unique challenges to the health care provider responsible for administering sedation and anesthesia during surgical or technical procedures. It is important for health care providers to recognize risk factors and potential complications before sedation or anesthesia [1]. Angelman syndrome (AS) is a neurogenetic disorder. It was first reported in 1965 by the English pediatrician Dr. Harry Angelman who first described three children with developmental delay [2]. Because of the apparent demeanor of the children, they were first known as 'happy puppet children' [3, 4]. The name of this syndrome changed into AS in 1987 [5]. AS is resulting from deficient expression or function of the maternally inherited allele of UBE3A gene on chromosome 15, which plays an important role in the cellular ubiquitin-proteasome pathway and synaptic development [6]. AS is mainly characterized by intellectual disability, movement or balance disorders, specific abnormal behaviors, and severe speech and language limitations [7].

Because of their intellectual disability and behavioral abnormalities (anxiety etc.), patients with AS may have difficulty with medical evaluations and may require general anesthetics for seemingly innocuous procedures such as dental care [8]. Other characteristics like hyperactive tendon reflexes, frequent drooling, sucking/swallowing disorders, increased sensitivity to heat, sleep disturbances, and fascination with water and reflective surfaces may be helpful or disruptive during dental treatment [9, 10]. During the general anesthesia, the dentist offers all dental treatment that the child needs. If the behavioral management, instead, offers the possibility to treat child with AS without general anesthesia, and/or dental procedures are not too invasive, the dentist has to use the correct techniques and psychological approach, to establish communication, and to obtain compliance even if in more visits [11].

On the other hand, there are several features of the syndrome that have theoretical implications for anesthetic management. For example, GABA (A) receptor abnormalities could affect responses to commonly used intravenous anesthetics. Both decreased and increased sensitivity to anesthetic agents have been reported [12, 13]. Patients with AS also may have high vagal tone. There are reports of malignant bradycardias triggered by laughing episodes [14, 15], and profound bradycardias have occurred during anesthetics [12, 16]. These patients may have craniofacial abnormalities and excessive drooling that could complicate airway management [12, 17]. Lastly, these patients have a "happy" demeanor, which can confound pain severity assessment [18].

This case report aimed to share our experience with anesthetic management of a patient with AS who underwent dental treatment under sedation.

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Case Report

In the faculty of dentistry, Bulent Ecevit University a 7 year-old female patient need teeth extraction was referred from the department of pedodontics to the department of oral and maxillofacial surgery due to the lack of cooperation.

The patient's medical history showed that the patient has AS, the diagnosis was made 9 months after birth. The patient was admitted to the hospital because of strabismus, and during patient assessment a genetic study was performed and the test confirmed the AS.

The medical history showed that the patient had her first seizure after 1 year of birth, and it was treated by 50 mg Sodyum valproat (Convilex ®) syrup per day, as a result the seizures stopped for a year, then 3 seizures happened again during the next second year, then the treatment was changed to 1 mg Clonazepam (Rivotril®) per day and for the last five years no seizure had happened.

In the physical examination; there were signs of speech impairment, gait ataxia, tremulous limb movement, frequent smiling, laughter, easily excitable personality, peripheral spasticity, muscular atrophy, hyperactivity, and a short attention span. The mouth was normal with frequent salivation. Thyromental distance was sufficient. Difficult intubation and/or difficult airway findings were not present. Listening lung sounds were natural and bilateral. There was no rale and rhonchi. Heart sounds were rhythmic, no additional sounds and murmurs were heard. ECG was in sinus rhythm. Laboratory values were within normal limits. In light of this information, it was planned to perform deep sedation with taking into consideration that no cooperation could be established with the patient.

The extraction of teeth was done under deep sedation with the support of local anesthesia in the operation room where monitoring of vital signs was performed. The presedation measurements revealed: blood pressure 100/56 mm Hg, Heart rate 95 beats per minute, and oxygen saturation 99%. An intravenous route was secured, and midazolam 2 mg IV was administered, but sufficient deep sedation did not obtain. Inhalation sedation was performed with 8 % Sevoflurane, oxygen and nitrous oxide. Then, sedation was maintained by nasal cannula with 4 % sevoflurane, nitrous oxide, and 4 L/min O₂ flow. All the time of work the spontaneous breathing was monitored. At the time of work the heart rate was between 85- 95 beats per minutes, and oxygen saturation 99-100 %. When the dental treatment completed, sevofluran and nitrous oxide were closed and the patient was awakened by giving only oxygen. After 2 hours of follow-up in post-anesthesia care unit the Aldrete recovery score was 10, so the patient referred to the ward.

Discussion

AS is a rare genetic neurological disorder. The exact incidence of the syndrome is unknown but the reported incidence of the disease varies from one in 10,000 to one in 40,000, with speculation that the disease is underdiagnosed due to its various phenotypes. Also there is no reported male or female predominance^[9, 19]. Our patient in this case report was a girl and there is no evidence that this syndrome is more common in girls.

Some associated clinical features of children with AS are of extreme importance to dentists. Abnormal function of the tongue, lips and cheeks which may adversely affect the mouth's natural cleaning patterns and alter the natural guidance of teeth into proper alignment. Sucking and swallowing difficulties may cause food to linger in the mouth,

increasing the risk of tooth decay^[9, 20]. In our case, excessive crown damage occurred in the permanent teeth of the patient due to many reasons, especially inadequate oral hygiene. As there was no indication for restorative treatment, these teeth were extracted under deep sedation with the support of local anesthesia.

AS patients with micro deletions and mutations leading to functional absence of UBE3A may have abnormal GABA receptors and thus may be more sensitive to anesthetic agents that stimulate these receptors^[21]. Altered regulation of $\beta 3$ subunit of the GABAA receptors may modify receptor kinetics and response to anesthetic agents^[13]. Many drugs, such as anxiolytics, sedative hypnotics, general anaesthetics, and anticonvulsants, become effective by interacting with these receptors. Midazolam, propofol, and potent inhalation agents all activate the GABA receptors. Therefore, the frequency of unexpected complications due to abnormal responses of these receptors increases in the anesthesia management of these patients^[5, 22]. Bujok and Knapik 2004 reported a prolonged recovery time after a general anesthesia for dental treatment in 12-year old boy with AS^[12].

In our case, 2 mg midazolam followed by sevoflurane inhalation and mask anesthesia were applied considering the patient's anxiety in both procedures. Studies have reported that bradycardia may occur during anesthesia. Some authors even recommend prophylactic atropine administration before anesthesia^[8, 16, 18]. In our case, no bradycardia was observed during the procedure.

In literature reviews, there is no recommendation regarding the duration of postoperative PACU care. However, Landsman *et al.* reported a mean PACU follow-up of 81.6 minutes in a retrospective study^[5]. In our case, although there was no complication in the peroperative period, the patient was referred to the ward after approximately 2 hours of PACU follow-up.

These patients typically appear to be happy and frequently laugh and smile, even under inappropriate circumstances. These characteristics could complicate assessment of pain postoperatively^[18]. In our case, postoperative pain palliation was not performed because local anesthesia was applied to the surgical field.

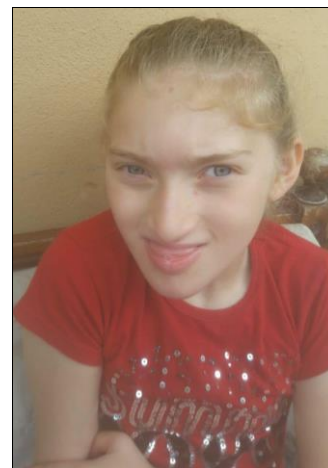


Fig 1: Patients with Angelman Syndrome are known to have a Good-Natured Disposition with Frequent Smiling, Laughing, and salivation.

Conclusion

Due to some features of the AS, dental treatments are frequently performed. It is very important that the patient remains stationary during the treatment. Also the degree of

cognitive impairment and limited communication skills limits their ability to cooperate during medical procedures. Therefore, they often require anesthetic even when undergoing minor procedures.

On the other hand, many features of the AS have theoretical implications for anesthetic management, and both decreased and increased sensitivity to anesthetic agents have been reported. Also successful anesthesia has been reported, although adverse events have occurred. Therefore, care should be taken against complications that may occur during anesthesia procedure.

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