OTJHS
Online Turkish Journal of Health Sciences

e-ISSN: 2459-1467

OTSBD
Online Türk Sağlık Bilimleri Dergisi

Online Turkish Journal of Health Sciences 2023;8(3):410-412 *Online Türk Sağlık Bilimleri Dergisi 2023;8(3):410-412*

Anesthesia Management in A Case with Prader-Willi Syndrome

Prader Willi Sendromlu Bir Olguda Anestezi Yönetimi

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ABSTRACT
Prader Willi (PWS) is the most common and rare genetic cause of obesity. Airway problems associated with obesity and hypotonia, increased risk of aspiration due to gastrointestinal (GI) motility and hyperphagia, obstructive sleep apnea syndrome (OSAS), difficult airway management, and postoperative respiratory failure risk due to narrow airway are the factors that complicate anesthesia management. It should be noted that there are many anatomical and physiological factors that may adversely affect perioperative anesthesia management in patients with PWS. For this reason, preoperative anesthesia evaluation and all preparations should be performed completely.
Keywords: Anesthesia management, difficult airway, prader-willi syndrome

ÖZ
Prader Willi (PWS), obezitenin en yaygın ve nadir görülen genetik nedenidir. Obezite ve hipotoni ile ilişkili hava yolu sorunları, gastrointestinal (GI) motilite ve hiperfaji nedeniyle artan aspirasyon riski, obstrüktif uyku apne sendromu (OUAS), zor hava yolu yönetimi, dar hava yoluna bağlı postoperatif solunum yetmezliği riski anestezi yönetimini zorlaştıran faktörlerdir. PWS'li hastalarda perioperatif anestezi yönetimini olumsuz etkileyebilecek birçok anatomik ve fizyolojik faktör olduğu unutulmamalıdır. Bu nedenle ameliyat öncesi anestezi değerlendirmesi ve tüm hazırlıklar eksiksiz yapılmalıdır.
Anahtar Kelimeler: Anestezi yönetimi, prader-willi sendromu, zor havayolu

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Yayın Bilgisi / Article Info:
Gönderi Tarihi/ Received: 08/08/2022
Kabul Tarihi/ Accepted: 19/07/2023
Online Yayın Tarihi/ Published: 06/09/2023

Atıf / Cited: Tunç Y and et al. Anesthesia Management in A Case with Prader-Willi Syndrome. *Online Türk Sağlık Bilimleri Dergisi* 2023;8(3):410-412. doi: 10.26453/otjhs.1159374

INTRODUCTION

Prader-Willi syndrome (PWS) was first described in 1956 and is a genetic disease caused by loss of gene expression (15q 11-13) occurring in the proximal long arm of paternal chromosome 15. Its incidence is approximately 1/15000. PWS is the most common and rare genetic cause of obesity.[1,2]

In this syndrome, craniofacial anomalies (dolichocephaly, narrow bifrontal diameter, strabismus, thin upper lip, small upturned nose and downturned corners of the mouth, enamel hypoplasia), mental retardation, infantile hypotonia, growth and other hormone deficiencies, hypogonadism/ hypogenitalism, behavioural problems in early childhood play a role in clinical manifestations. It is a neurodevelopmental disorder with hyperphagia that leads to obesity if not controlled.[1]

Airway problems associated with obesity and hypotonia, increased risk of aspiration due to gastrointestinal (GI) motility and hyperphagia, obstructive sleep apnea syndrome (OSAS), risk of postoperative respiratory failure due to narrow airway, and difficulty in providing intravenous access are factors that complicate anesthesia management. It should be noted that many anatomical and physiological factors may adversely affect perioperative anesthesia management in patients with PWS. For this reason, preoperative anesthesia evaluation and all preparati-

ons should be performed completely.
In this case report, we aimed to present the anesthesia management of a 9-year-old male patient who presented to the emergency department of our hospital due to foreign body obstruction in the esophagus.

CASE REPORT

A 9-year-old male patient diagnosed with PWS by genetic analysis and followed-up in an outer center, with a height of 131 cm, weight of 55 kilograms, and body mass index of 32 kg/m2, was consulted by our pediatric surgeon for emergency esophagoscopy under general anesthesia due to foreign body obstruction (coin) in the esophagus. When the patient was evaluated preoperatively, it was observed that he had mental retardation, hyperphagia, and OSAS findings. It was learned that he had growth retardation and thyroid hormone deficiency and had used 1 mg/day recombinant human growth hormone and 12.5 mcg/day levothyroxine for treatment. In the physical examination, craniofacial anomalies such as a thin upper lip, downward curved corners of the mouth, and high palate were detected. The patient had adequate mouth opening and a Mallampati score of III. The patient was clinically euthyroid, and there was no anomaly in other laboratory parameters. The preoperative fasting period of the patient was 8 hours. Considering that there may be a risk of postoperative respiratory failure and reintubation, pharyngeal hypotonia and stenosis in the patient with OSAS, preparations were made with the necessary equipment (ETT and LMA in different diameters and sizes, Videolaryngoscope etc.). The patient had peripheral vascular access, which was controlled. The patient was taken to the operating table without any premedication, and standard monitoring was performed with electrocardiography (ECG), peripheral oxygen saturation (SpO_2) and non-invasive blood pressure (NIBP) measurement. Case baseline measurement values were heart rate: 87/min, NIBP: 121/75 mmHg, and SpO2: 97%. Preoxygenation was provided with 5 L/min 100% O_2 in spontaneous respiration. In the induction of anesthesia, 1 mg/kg IV lidocaine, 2 mg/kg IV propofol and 3 mcg/kg IV remifentanil were administered. Neuromuscular blocker administration was avoided in the patient. Before esophagoscopy, the laryngeal and esophageal entrance part of the patient was visualised with the help of a C-MAC videosyngoscope, and it was observed that the coin was inserted into the first stenosis of the esophagus, and the foreign body was successfully removed by the pediatric surgeon using magell forceps. Esophagoscopy was performed to control the esophagus, and no other foreign body was found. The patient, whose hemodynamic data were within normal limits and no additional medication was administered during the procedure, was ventilated with a mask until spontaneous breathing returned. In the case that lasted 20 minutes in total, no complications developed, and the patient's spontaneous breathing returned to normal. He obeyed verbal commands, and after the patient woke up, he was interned in the postanesthetic care unit (PACU). The patient, who was kept under observation for 30 minutes in PACU, was transferred to the pediatric surgery service without any problem, with a Modified Aldreate Score of 10.

DISCUSSION AND CONCLUSION

PWS is a genetic disease with a clinical course consisting of 2 stages. The hypotonic infantile stage is the first stage, which is characterised by difficulty in sucking, muscle hypotonia, weakness in the cough reflex, and growth retardation. The hyperphagic obese stage is the second stage that follows the first stage and starts with the recovery of muscle activity between the ages of 2-4 and causes the development of mental retardation, behavioral disorders, hyperp-

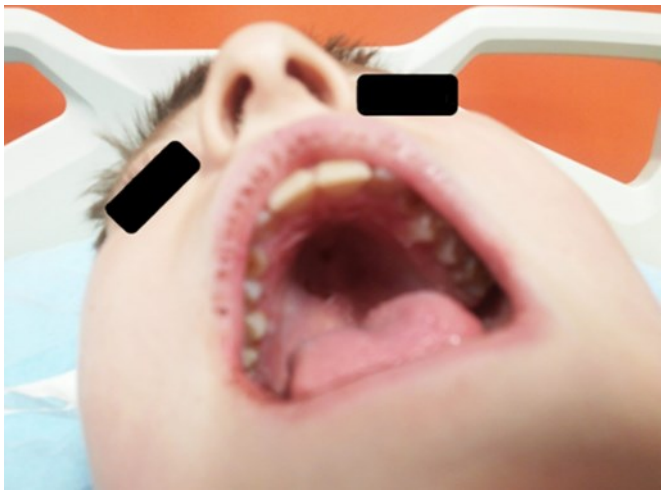

Figure 1. Craniofacial anomalies such as a thin upper lip, downward curved corners of the mouth, and high palate.

hagia and obesity.[3]

In studies conducted with PWS patients, it has been reported that morbid obesity, central hypotonia and OSAS are the most common and important factors of perioperative complications. In these patients, general and regional anesthesia may be more risky than normal due to the perioperative risks arising from hypotonia and morbid obesity, especially in general anesthesia.[4]

If access to food and weight gain are not adequately controlled in PWS, diabetes mellitus, right heart failure, hypoventilation, obstructive sleep apnea (OSAS), and narrow airway, GI dysmotility, esophageal dysmotility, delayed gastric emptying may accompany the clinical picture associated with hyperphagia and obesity. The presence of conditions such as water intoxication may make anesthesia management more difficult.[1] In these patients, the difficulty in maintaining hunger control before the procedure due to hyperphagia should be considered, and the last food intake period should be followed closely. It is stated that the risk of aspiration is high due to preoperatively uncontrolled food intake. In patients with OSAS, close follow-up should be applied after the procedure because of the risk of postoperative respiratory failure. (4) In addition, venous access difficulties, thermoregulation disorders and metabolic problems are also common.[2]

Tseng et al. presented a 5-year-old, 50 kg male PWS patient who was admitted with OSAS and underwent general anesthesia with IV propofol and face mask and sevoflurane induction for bilateral tonsillectomy and uvulopalatopharyngoplasty. The patient was intubated endotracheally without the use of muscle relaxants. It has been reported that the patient, who was extubated when fully awakened, was reintubated and sent to the pediatric intensive care unit when the O2 saturation (SpO2) decreased to 70%, and the respiratory rate increased to 70/min within a few minutes after the follow-up in the postanaesthesia care unit (PACU).[5]

In our case, the patient with OSAS, growth retardation and thyroid hormone deficiency, hyperphagia, mental retardation and behavioral disorder; to eliminate the foreign body obstruction developed in the esophagus as a result of coin ingestion, the procedure was performed without any problem with the help of a videoryngoscope under general anesthesia by avoiding the use of neuromuscular blocking agents. The patient was followed closely in the PACU in the postoperative period. The patient, who did not develop any complications during the follow-up, was transferred to the pediatric surgery service.

In conclusion, it should not be forgotten that many anatomical and physiological factors may adversely affect the perioperative process in patients with PWS. Therefore, the preoperative anesthesia evaluation should be detailed, and all preparations should be made completely. Necessary equipment (laryngeal mask, videoryngoscope, flexible bronchoscopy, tracheostomy materials, etc.) that may be needed should be readily available and easily accessible in the operating room, taking into account possible difficult airway in the patient, and respiratory complications that may arise intraoperatively and postoperatively. Using long-acting muscle relaxants should be avoided, especially in cases with severe OSAS. In this patient group, individualised and well -planned anesthesia management before the procedure can provide uneventful anesthesia.

Ethics Committee Approval: Ethics committee approval was not required for the case report. The patient /relatives have signed an informed consent/ consent form, and the study was conducted in accordance with international declarations, guidelines, etc.

Conflict of Interest: No conflict of interest was declared by the authors.

Author Contributions: Concept – EK, YT; Supervision – EK, YT; Materials – YT; Data Collection and/ or Processing – EK, ASÖ; Analysis and/ or Interpretation – EK, YT, ASÖ; Writing –EK, YT.

Peer-review: Externally peer-reviewed.

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