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CLINICAL REPORT

Anesthesia for a 16-month-old patient with Prader–Willi syndrome

Basak C. Meco · Zekeriyya Alanoglu · Ozgur Sinan Cengiz · Neslihan Alkis

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Abstract Prader–Willi syndrome (PWS) is a rare disorder of chromosome abnormalities in which the paternal genes in chromosome 15 are lacking. The clinical course is characterized by hypotonia, hyperphagia, and morbid obesity. Both general and regional anesthesia in these patients is difficult due to morbid obesity and hypotonia. We report our anesthetic management in a patient with PWS with a body mass index (BMI) of 29.43 kg/m^2 who underwent orchiopexy and hypospadias repair. The clinical course of the patient was uneventful during the procedure and postoperative period. However, arrangements with a pediatric intensive care setting for the postoperative period are recommended for patients with PWS undergoing surgery.

Keywords Prader–Willi syndrome · Anesthesia · Hypotonia

Introduction

Prader–Willi syndrome (PWS) is a rare (1:15,000) and multisystem genetic disorder caused by the lack of expression of paternal genes in chromosome 15q11–13 [1, 2]. In early childhood, PWS is characterized by hypotonia, hyperphagia, and morbid obesity. After childhood, learning disabilities, mental retardation, behavioral problems, and seizure may occur [3]. Anesthesia management of these patients is challenging due to airway problems related to hypotonia and obesity, postoperative respiratory failure, temperature control, and intravenous line placement difficulty. We present our anesthetic experience with a 16-month-old patient with genetically proven PWS undergoing orchiopexy and hypospadias repair.

B. C. Meco (✉) · Z. Alanoglu · O. S. Cengiz · N. Alkis
Department of Anesthesiology and ICM, Faculty of Medicine, Ankara University, Mesa Koza sitesi, 2.blok no:15, GOP, 06700 Ankara, Turkey
e-mail: basakceyda@hotmail.com

Case report

A 16-month-old boy with PWS and a body weight of 17 kg and height of 76 cm [body mass index (BMI) 29.43 kg/m^2] was scheduled for a bilateral orchiopexy and hypospadias repair. His medical history revealed no surgeries or anesthesia. In the preanesthesia visit, physical investigation revealed an anticipated difficult airway (obesity, known pharyngeal hypotonia and narrowness), and adequate preparations were made for airway management prior to anesthesia. Standards for basic anesthesia monitoring consisting of electrocardiography, pulse oximetry, and noninvasive blood pressure in the operating room. Body temperature was monitored with a nasopharyngeal probe. Difficulty in intravenous line placement was predicted, and therefore, the line was initiated in the ward prior to the procedure. General anesthesia was induced with sevoflurane 8% in 100% oxygen via facemask and was deepened with intravenously administered ketamine 0.5 mg/kg, propofol 3 mg/kg, and remifentanil 0.5 μg/kg boluses. The trachea was intubated uneventfully at the first attempt with a 3.5-mm endotracheal cuffed tube without the use of a muscle relaxant. Methylprednisolone 2 mg/kg was given intravenously to prevent laryngeal edema and bronchospasm, and fentanyl 1 μg/kg was given intravenously for postoperative pain management. Anesthesia was maintained with sevoflurane in oxygen and air mixture. The patient's vital signs and body temperature remained stable

during the surgery, which was uneventful. The endotracheal tube was removed once the patient regained spontaneous respiration and was fully awake. After extubation, the patient was arousable (crying) with painful stimulation, and oxygen saturation remained 100%. The patient was then transferred to the pediatric intensive care unit (ICU) for close monitoring. He was discharged to the ward the day after, with no respiratory events.

Discussion

The combination of hypotonia, developmental delay, hypogonadism, and obesity was first described by Prader, Labhart, and Willi in 1956 as Prader–Willi syndrome [4]. It is known that PWS results from the absence of paternal genes on chromosome 15q11–13. The classic presentation is biphasic, with an infantile hypotonic phase followed by a childhood obese phase. According to the literature related to anesthesia management of patients with PWS, both general and regional anesthesia is challenging. Difficult airway is frequently accompanied with general anesthesia. Also, difficult intravenous line placement, disturbance in thermoregulation, and metabolic problems are common.

Few cases related to anesthesia management for patients with PWS are reported in the literature. Tseng et al. reported two cases of PWS and noted the importance of preoperative patient evaluation. They emphasized the importance of obstructive sleep apnea (OSA) as a complication and recommended postoperative intensive care for such patients [5]. Milliken and Weintraub [6] reported a case of 10-year-old boy who developed severe cardiac arrhythmias during a bilateral orchiopexy procedure. In our case, cardiac rhythm and hemodynamic parameters were stable throughout the surgery and during postoperative care. The patient remained in the pediatric ICU for close monitoring to avoid OSA periods after surgery and was discharged uneventfully. Dearlove et al. [7] reviewed a case series of PWS patients and reported that body weight was the determinant and sole factor for anesthesia-related difficulties. In the case presented by Mayhew and Taylor [8], difficult intubation and malignant hyperthermia developed during and orchiopexy procedure and the operation was delayed. The surgery was then performed without any problems on the second attempt. Tseng et al. [5] reported intermittent bronchospasm, stridor, desaturation, and reintubation of a patient during a tonsillectomy procedure. In our case, intraoperative and postoperative airway management was uneventful, and the patient was extubated in the operating room without delay. Respiratory problems are vital during and after the operation. Respiratory arrest secondary to hypotonia and hypoventilation is a major cause of mortality in PWS patients. To prevent this complication, special care must be taken during the preoperative evaluation related to the respiratory system. Regional anesthesia techniques may be an alternative to general anesthesia in order to prevent respiratory complications. However, difficulty defining landmarks for regional anesthesia due to obesity is another concern.

Several manifestations of PWS in different organ systems have been defined. Of primary concern is obesity leading to difficult airway management and respiratory failure. Rarely cardiac involvement is present. Thermoregulation imbalance, metabolic disorders, and mental retardation should be considered during the perioperative and early postoperative periods. Therefore, detailed preoperative evaluation, adequate preparation, and special postoperative follow up are recommended for PWS patients undergoing surgery.

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