Anesthesia management of a pediatric patient with Wolf-Hirschhorn Syndrome: A case report

Wolf-Hirschhorn Sendromlu pediatrik bir hastanın anestezi yönetimi: Olgu sunumu

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Abstract

Wolf-Hirschhorn syndrome (WHS) is a rare chromosomal anomaly caused by the deletion of the short arm of chromosome 4. The leading causes of death are aspiration pneumonia, epilepsy, and infection. In this study, we aimed to present our anesthesia experience in a patient with WHS who was scheduled for cleft palate surgery and in whom we performed total intravenous anesthesia (TIVA).

In the clinical examination of our 8-year-old patient with WHS, weighing 13 kg, who was planned to undergo cleft palate surgery. Growth retardation, cleft palate, micrognathia, epicanthus and hyptonia were present. Anesthesia induction was performed with 1 mg/kg i.v lidocaine, 2 mg/kg i.v propofol and 1 mg/kg i.v fentanyl, 0.5 mg/kg i.v rocuronium bromide. TIVA (0.5 μg/kg/min remifentanil and 75 μg/kg/min propofol i.v) was used for maintenance of anesthesia. In the perioperative period, the pulse rate was 170 beats/min for a short time. Extubation was achieved 13 minutes after the end of surgery.

We think that TIVA (Propofol, remifentanil) and C-MAC videolaryngoscope as intubation device is a reliable method in the anesthetic management of children with WHS syndrome.

Keywords: Airway management; anesthesia; laryngoscope; Wolf-Hirschhorn syndrome

Öz

Wolf-Hirschhorn sendromu (WHS) 4. kromozomun kısa kolunun silinmesiyle ortaya çıkan nadir görülen bir kromozomal anomalidir. Önde gelen ölüm nedenleri aspirasyon pnomonisi, epilepsi ve enfeksiyondur. Bu çalışmamızda WHS' lu yarık damak cerrahisi planlanan Total intravenöz anestezi (TİVA) uyguladığımız bir olguda anestezi deneyimimizi sunmayı amaçladık. WHS tanısıyla takip edilen, yarık damak cerrahisi planlanan 8 yaşında, 13 kg olan olgumuzun klinik muayenesinde, gelişme geriliği, yarık damak, mikrognati, epikantus ve hiptoni mevcuttu. Anestezi indüksiyonunda 1mg/kg i.v lidokain, 2mg/kg i.v propofol ve 1mcg/kg i.v fentanil, 0.5 mg/kg i.v rocuronium bromide uygulandı. Anestezi idamesinde TİVA (0.5 μg/kg/dk remifentanil ve 75 μg/kg/dk propofol i.v) kullanıldı. Perioperatif dönemde nabız kısa süreli 170/atım/dk oldu. Cerrahi bitiminden 13 dk sonra ekstübasyon sağlandı.

WHS sendromlu çocukların anestezi yönetiminde TİVA (Propofol ,remifentanil)' nın ve entübasyon cihazı olarak C-MAC videolaringoskopun güvenilir bir yöntem olduğunu düşünüyoruz.

Anahtar Sözcükler: Anestezi; havayolu yönetimi; laringoskop; Wolf-Hirschhorn sendromu

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INTRODUCTION

Wolf-Hirschhorn syndrome (WHS) is a rare chromosomal abnormality characterized by deletion of the short arm of the 4th chromosome especially in the 4p16.3 region (1).

Clinical findings are usually observed in the early period of life. Short upper lip, wide mouth, craniofacial anomalies (hypertelorism, protruding glabella, depressed nasal root, and micrognathia), severe mental and developmental retardation, cleft palate and lip among midline defects, congenital heart defects (atrial septal defect, patent ductus arteriosus, and pulmonary stenosis), skeletal and ocular anomalies, hypotonia and epilepsy are the most common anomalies (2,3).

The incidence of WHS is 1:50,000-20,000. The female/male ratio is 2:1. The mortality rate in the first two years is 17-30%. The leading causes of death are aspiration pneumonia, epilepsy, and infection (4).

Many surgical interventions are required because of congenital anomalies accompanying WHS cases. Increased incidence of malignant hyperthermia (MH), difficult airway, and difficulty in intubation due to abnormal oral structure and craniofacial anomalies make anesthesia management specific (3).

In this study, we aimed to present our anesthesia experience in a patient with WHS in whom we performed TIVA for cleft palate surgery.

CASE REPORT

Our 8-year-old, 13 kg patient, who was followed up with the diagnosis of WHS and planned for cleft palate surgery, had a normal vaginal birth as the 6th child of a 34-year-old mother, weighing 1600 g delivery. He was operated due to intracranial hemorrhage on the 2nd postpartum day and stayed in the intensive care unit for 40 days. Clinical examination revealed growth retardation, cleft palate, micrognathia, epicanthus, hypertelorism, pesechinovarus, and hyptonia. His history included mental retardation, epilepsy, secundum Atriyal Septal Defekt (ASD) and frequent pneumonia attacks. When the laboratory values of the patient were examined, urine, biochemical and hemogram values were found to be within normal limits.

After explaining the procedure and possible risks to the patient's relatives, written consent was obtained for anesthesia.

The patient was placed on the operating table and standard monitoring was performed with electrocardiography (ECG), heart rate (HR), peripheral oxygen saturation (SpO_2) and noninvasive blood pressure (NIBP). The patient was preoxygenated with 4 L/min 100% O_2 via nasal cannula. Initial vital signs were within normal limits (heart rate 115/min, NIBP: 92/55 mmHg and SpO_2: 95%). Temperature monitoring was also performed. Initial body temperature was 36.7°C.

For induction of anesthesia, 1 mg/kg IV lidocaine, 2 mg/kg IV propofol and 1 mcg/kg IV fentanyl were administered. Upon no difficulty in mask ventilation, IV rocuronium bromide was administered at 0.6 mg/kg as a muscle relaxant. Mallampati score was III in preoperative evaluation. There was no restriction in neck movements. Intubation conditions were optimized by using a shoulder roll. After adequate muscle relaxation was achieved, intubation was achieved on the first attempt with a C-Mac Videolaryngoscope (number 2 bleyd) and an endotracheal tube with a number 5.5 cuff and a spiral stylet. After intubation, ventilation was provided with tidal volume (6-8 ml/kg) and $EtCO_2$ value of 35-40 mmHg in 50% O_2-air mixture.

Surgery lasted a total of 3.5 hours. Vital signs were stable for the first 60 minutes. HR suddenly increased up to 170/min/beat. As a result of volume support and deepening of anesthesia, HR (90-110 min/beat) returned to normal limits. $EtCO_2$ was 35-48 mmHg, SpO_2 was 95-97, and temperature was around 34.7-36.7 Co. At the end of surgery, neostigmine 0.04 mg/kg and atropine 0.02 mg/kg were administered i.v. to reverse neuromuscular blockade. Extubation was achieved 13 minutes after the end of surgery successfully. She was followed up in PACU for 1 hour. Postoperative analgesia was provided with 15 mg/kg i.v paracetamol in patients with (VAS) >5.

When the modified Aldrete score was >9, the patient was transferred to the pediatric intensive care unit (5). The patient was visited in the relevant service at 12-24 hours. In the follow-up, it was observed that vital signs were stable and there was no respiratory distress.

DISCUSSION AND CONCLUSION

WHS is a rare genetic disorder characterized by deletion of the distal portion of the short arm of chromosome 4 (4p-) and is associated with many anomalies. Anesthetic management in WHS patients has many difficulties. Cleft lip/palate, gastroesophageal reflux, muscular hypotonia, skeletal anomalies (scoliosis), malignant hyperthermia, perioperative respiratory complications, micrognathia, oral dental anomalies, congenital heart defects are the main causes (2).

There are some case reports in the anesthesia literature indicating an association between malignant hyperthermia and WHS. Ginsburg R et al. used halothane and succinylcholine as anesthetics in a 21-month-old child who underwent surgery for cleft palate (6). The rectal temperature increased to 38.5 C° at the 7th minute following induction and to 42.2 C° in the following period. This was evaluated as malignant hyperthermia. The temperature rise was taken under control with necessary interventions (6). In a study by Jae Ho Cho et al. in a 33-month-old male pediatric patient scheduled for tympanoplasty and myringotomy, the entry body temperature was measured as 36.2 C°. At the end of the surgical operation, the body temperature was 37.9 C° in the recovery room and 39 C° at the 7th hour postoperatively. Body temperature was controlled with antipyretics and phenobarbital. In the initial evaluation, persistent temperature increase was evaluated as delayed malignant hyperthermia. On detailed examination, it was evaluated as upper respiratory tract infection and the diagnosis of malignant hyperthermia was excluded (7).

Despite the case examples of malignant hyperthermia in the literature. There are also WHS cases in which inhalation anesthetics and succinylcholine were used safely (8). The relationship between WHS and malignant hyperthermia has not been fully proven. In anesthesia practice, propofol, barbiturates, benzodiazepines, narcotics and non-depolarizing muscle relaxants are known as safe anesthetic agents in terms of malignant hyperthermia (9).

In our case, we did not use inhalation anesthetics and succinylcholine to avoid the risk of malignant hyperthermia. In addition, our patient had a history of epilepsy. Therefore, we preferred the use of TIVA (propofol and remifentanil) as an anesthetic because of the safer epileptogenic potential of propofol compared to sevoflurane anesthesia and the antiemetic properties of propofol.

In WHS, cleft palate, cleft lip, micrognathia, defects in oral dentition and skeletal anomalies cause difficult airway management and intubation difficulties (2,10,11).

Therefore, necessary preliminary preparations should be made for difficult airway management in patients with WHS. In our case, the presence of cleft palate was hypothesized to cause intubation difficulties. Care should be taken to administer muscle relaxants in patients who cannot be ventilated after induction of anesthesia (7). In our patient, there was no difficulty in ventilation after induction and we proceeded to the intubation stage. We prepared for difficult airway (flexibil bronchoscopy and C-MAC videolaryngoscope) before induction of anesthesia. We performed intubation on the first attempt with the C-MAC videolaryngoscope without difficulty.

The incidence of hypotonia in WHS cases is >75%. In the presence of hypotonia, care should be taken in the dose of muscle relaxant and the choice of the agent to be used. It is known that the use of succinylcholine is a risk factor for malignant hyperthermia (12). Non-depolarizing muscle relaxants are used more frequently. However, in the presence of hypotonia, the dose of NMBAs should be reduced as it may cause prolonged extubation and recovery time. (13, 7, 14).

In our case, rocuronium from NMBAs was safely used as a neuromuscular agent. Alternatively, there are cases in the literature in which intubation was performed without the use of muscle relaxant (8).

Muscle hypotonia is one of the reasons provoking gastroesophageal reflux in WHS patients. Because of the risk of aspiration during induction and in the postoperative period, it is important to prepare the aspirator and pay attention to the fasting period. In some publications in the literature, rapid sequence intubation is recommended against the risk of aspiration (2).

Congenital heart defects are observed in 50% of WHS cases. The most common form is atrial septal defects (27%) (3). Our patient had a secundum ASD. During the operation, HR: 170 beats/min during operation. Therefore, the patient should be closely moni-

tored to ensure hemodynamic stability and the patient should be prepared for possible deterioration in hemodynamics and intervened in a timely manner.

In conclusion; we think that TIVA (Propofol, remifentanil) and C-MAC videolaryngoscopy as an intubation device is a reliable method in the anesthetic management of children with WHS syndrome.

Conflict-of-interest and financial disclosure

The author declares that he has no conflict of interest to disclose. The author also declares that he did not receive any financial support for the study.

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