



Anesthetic Management of a Patient With Jarcho-Levin Syndrome

Jarcho Levin Sendromlu Olguda Anestezi Yönetimi

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Abstract

Jarcho-Levin syndrome (JLS) is a rare disease involving costovertebral anomalies and is accompanied by hydrocephalus, neural tube defect, and cardiac, renal and gastrointestinal problems. Due to respiratory system pathologies, there is a high mortality rate at young ages. Due to its rarity and little information in the literature related to anesthetic practice for this syndrome, we present our anesthetic experience of inguinal hernia surgery in a patient with JLS.

Keywords: Jarcho-Levin syndrome, laryngeal mask, anesthesia management

Öz

Jarcho Levin sendromu (JLS) kostavertabral anomalilerin ve bunlara hidrosefali, nöral tüp defekti, kardiyak, renal ve gastrointestinal problemlerin eşlik ettiği nadir görülen bir hastalıktır. Solunum sistemi patolojileri nedeniyle erken yaşta mortalite oranı yüksektir. Nadir karşılaşılması ve literatürde JLS'ye dair anestezi pratiğinin az olması nedeniyle inguinal herni ameliyatı yapılan hastadaki anestezi deneyimimiz sunulmuştur.

Anahtar Sözcükler: Jarcho Levin sendromu, laringeal maske, anestezi yönetimi

Introduction

Jarcho-Levin syndrome (JLS) is a congenital genetic disease characterized by vertebral and rib anomalies. It is inherited as an autosomal dominant and recessive trait (1). It was first described in 1938 by Jarcho and Levin (2). The prevalence is determined as 0.25/10.000 (3). In 1978, Solomon et al. (4) have classified the syndrome into two different phenotypes as spondylothoracic dysplasia and spondylocostal dysplasia according to prognosis, inheritance and distribution of skeletal anomalies (5). The syndrome is accompanied by multiple anomalies. Hydrocephalus, hydroureteronephrosis, meningomyelocele, atrial septal defects, ventricular septal defects, renal agenesis, hypoplasia, polycystic kidneys, anal atresia, diaphragmatic hernia, esophageal fistula, Meckel's diverticulum, and bifid uvula are among the anomalies that have been reported (6). This report shares

our anesthetic experience of a JLS patient operated on for inguinal hernia.

Case

Family consent was given for this case report. The 19-month, 8 kg male infant with planned operation for left inguinal hernia was the second pregnancy of a 27-year-old mother and was born by cesarean at 36 weeks with birth weight of 2.700 g. The case's history included parents who were second-degree relatives, the mother attended regular prenatal check-ups and apart from iron deficiency anemia, had no diseases, did not use cigarettes or alcohol, had no teratogeny, was not exposed to radiation, first child had glucose six-phosphaate dehydrogenase deficiency, and there were no other children with JLS. Pregnancy scans identified costovertebral anomalies in the fetus with additional hydrocephalus in the prenatal 34th week and

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postnatal lumbar meningocele. Operated one week after birth for meningocele and hydrocephalus, the patient had a ventriculoperitoneal shunt inserted, remained in the intensive care unit for one month and was hospitalized twice for pneumonia. Physical examination noted hydrocephalus, pectus carinatus, pes equinovarus, accessory nipple on the right and disfiguration of the thorax (Figure 1). Preoperative examination requested ear nose and throat and pediatric consultation and found all other systems to be normal, other than paraplegia. Mallampati was not assessed. Routine preoperative laboratory tests were normal. Examination of current x-rays of the case showed secondary deformed appearance due to fusion anomalies at the level of the costovertebral joints on the right ribs, reduction in the number of ribs, and fenestration (crab appearance) between the ribs. Due to S-shaped scoliosis of the thoracolumbar spinal column, there were vertebra segmentation anomalies at multiple levels and in the lumbar region, vertebra had appearance in accordance with posterior arc defect (closure defect) (Figure 2). Cranial tomography showed the shunt catheter entering the right parieto-occipital bone and ending at the posterior horn of the left lateral ventricle and hydrocephalic dilatation in ventricles at the supratentorial level; the corpus callosum and interventricular septum were not observed (partial-complete callosal agenesis?). Renal ultrasound revealed left renal agenesis.



Figure 1. The patient's physical appearance

The case had standard monitoring (electrocardiography, peripheral oxygen saturation, non-invasive blood pressure measurement, temperature) applied. Relevant preparations were made for possible difficult airway before general anesthesia. For induction, 2.5 mg/kg propofol and 1 mg/kg lidocaine were administered. The case did not encounter problems with mask ventilation and had a size 1.5 laryngeal mask airway (LMA) (Supreme® LMA) inserted. Anesthesia maintenance was provided by 2.5-3% sevoflurane and 50/50% oxygen/nitrous oxide. After the surgery, intravenous 10 mg/kg paracetamol was administered for analgesia. The case was transferred to the pediatric surgery clinic after 45 minutes in the postoperative care unit. The case was discharged on postoperative day one.

Discussion

JLS is a rare congenital disease with costovertebral anomalies (hemivertebra, vertebral agenesis, butterfly, wedge and hypoplastic vertebra and costovertebral fusion defects) and accompanied by hydrocephalus, neural tube defects, and cardiac, renal and gastrointestinal problems (1,6). Patients experience severe respiratory problems causing early death due to pneumonia or restrictive-type pulmonary disease. Additionally, pulmonary hypertension and congestive heart failure negatively affect prognosis in these cases (3,7). To date, the longest surviving reported JLS case has been monitored for 33 years (6). It was determined that the intelligence level of all reported cases was normal (1,3).

Schulman et al. (7) did not observe distal tracheal rings in two JLS cases with airway anomalies, but stated that they had wider carina and narrowing of both bronchial lumen



Figure 2. The patient's chest X-Ray

reducing airflow. They emphasized that these anomalies of the rings may cause primary, or secondary to repetitive aspiration, dysplastic changes. Additionally, anesthesia for any operation to be performed on JLS patients may be complicated by insufficient pulmonary development, narrower chest cage, lower respiratory infections and respiratory pathway problems. Due to being an inherited disease and observed with different clinical forms, it is necessary that families be informed of abnormal airway and risks. In our case, the family was informed of possible difficult airway, required consent was obtained and ear, nose and throat and pediatric consultations were requested. For our case, all necessary airway devices for possible difficult ventilation and intubation were prepared preoperatively.

Due to the short duration of the operation, the procedure was completed with intravenous induction of general anesthesia with LMA to protect spontaneous respiration and reduce complications related to intubation and muscle relaxants in the postoperative period. Developed in 1980 for the first time, LMA is widely used in anesthetic practice and provides successful results especially for difficult airway and intubation (8,9). Geze et al. (9) emphasized that use of LMA was more reliable for a JLS case. With surgery for inguinal hernia, we chose the Supreme® LMA with general anesthesia and for anesthetic safety, chose short-effect anesthetic agents. Our case did not experience any postoperative respiratory problems.

Yilmaz et al. (10) described accessory nipple in JLS for the first time. Our case had accessory nipple on the right, hydrocephalus, meningomyelocele, pectus carinatus, and renal agenesis.

Şen et al. (1) reported cleft palate in a patient with JLS diagnosis in the newborn period and stated the necessity of detailed investigation of accompanying systems in situations where this syndrome is suspected.

Conclusion

Under medical care conditions developed in recent times with effective treatment and prevention of recurrence of respiratory infections, the life expectancy of JLS patients will lengthen and surgeries planned for a variety of reasons will increase; we believe that LMA is a reliable anesthetic method for JLS patients.

Ethics

Informed Consent: It was taken.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: Gamze Küçükosman, Aslı Yalçın. Concept: Gamze Küçükosman, Bengü Gülhan Aydın. Design: Gamze Küçükosman, Bengü Gülhan Aydın, Hilal Peri Ayoğlu. Data Collection or Processing: Gamze Küçükosman, Bengü Gülhan Aydın, Aslı Yalçın. Analysis or Interpretation: Gamze Küçükosman, Bengü Gülhan Aydın, Hilal Peri Ayoğlu. Literature Search: Gamze Küçükosman, Bengü Gülhan Aydın, Aslı Yalçın. Writing: Gamze Küçükosman, Bengü Gülhan Aydın, Hilal Peri Ayoğlu.

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