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Case Report / Olgu Sunumu

Spinal Anaesthesia in a Neonate with Hypoplastic Left Heart Syndrome and Duodenal Atresia

Hipoplastik Sol Kalp Sendromu ve Duodenal Atrezili Yeni Doğanda Spinal Anestezi

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Abstract / Özet

Hypoplastic left heart syndrome (HLHS) is a lethal, congenital cardiac defect of neonates. It has been reported to occur in approximately 0.016 to 0.036 per 1000 live births and surgical palliation or cardiac transplantation is needed to treat this defect. This syndrome is more common in males and is associated with other congenital extra cardiac abnormalities, and these patients may need surgery for non-cardiac reasons. Here we report a neonate with HLHS and duodenal atresia who required emergency surgery due to intestinal obstruction with spinal anaesthesia. This case had experienced various challenges due to cardiac and extra-cardiac abnormalities. Surgery was performed under spinal anaesthesia.

Key Words: Hypoplastic left heart syndrome, anaesthesia, spinal, neonate

Hipoplastik sol kalp sendromu (HSKS) yeni doğanda görülen ölümcül, kalıtsal bir kalp kusurudur. Sıklığı her bin canlı doğumda 0,016 ile 0,036 arasında bildirilmiştir ve kusurun düzeltilmesi için cerrahi girişim veya kalp transplantasyonu gerekmektedir. Erkeklerde daha sık görülen bu sendrom, kalp dışı diğer kalıtsal anormalliklerle beraber olabilmekte ve bu nedenlerle cerrahiye ihtiyaç duyulabilmektedir. Bu sunuda duodenum obstrüksiyonu nedeniyle acil cerrahi gerektiren ve spinal anestezi uygulanan hipoplastik sol kalp sendromlu bir hasta sunulacaktır. Kalp ve kalp dışı nedenlerle bazı zorluklar içeren bu vakada spinal anestezi ile uygulanmıştır.

Anahtar Kelimeler: Hipoplastik sol kalp sendromu, anestezi, spinal, yeni doğan

Introduction

Hypoplastic left heart syndrome (HLHS) is a congenital disorder with a defect in the left-sided cardiac structures. The syndrome includes underdevelopment of the left ventricle, aorta and mitral valve. Hypoplasia of the left ventricle, aortic valve stenosis or atresia, mitral valve stenosis or atresia and hypoplasia of the ascending aorta with discrete coarctation of the aorta with varied levels of severity may be seen (1, 2). Although HLHS was once invariably a fatal cardiac anomaly, advances in surgery and neonatal intensive care have improved outcomes for many infants. Coexisting non-cardiac anomalies may increase mortality and complicate treatment. The 30-day mortality rate of patients with major cardiac anomalies following non-cardiac surgery was reported to be 11.3% and similarly, the mortality rate of children with HLHS who underwent non-cardiac surgery was found to be around 19% (3). To avoid haemodynamic and respiratory disturbance during the pre and postoperative period, spinal anaesthesia had been suggested as an alternative to general anaesthesia, especially in high-risk infants with congenital cardiac disease. This provides efficient motor blockage and pain relief, and blunts neurohumoral stress (1).

In this case report we describe a newborn with HLHS suffering from duodenal atresia that required emergency surgery under spinal anaesthesia.

Case Report

A five-day-old newborn male, weighing 2100 g, with a prenatal diagnosis of duodenal atresia, was transferred to the paediatric surgery department after delivery by elective caesarean section. The neonate was born in the 36th gestation week with a complex heart malformation. Echocardiography showed severe mitral valve hypoplasia (tricuspid annulus 9 mm, mitral annulus 5 mm), a hypoplastic left ventricle, a double outlet right ventricle, pulmonary hypertension, an atrial septal defect (5 mm), and a ventricular septal defect (large outlet), with the aorta and pulmonary artery originating from the right ventricle, and a patent ductus arteriosus. The consultant paediatrician initiated prostaglandin E1 infusion and digoxin therapy to stabilize cardiac function in the intensive care unit.

After initial management of the patient, urgent surgery was requested for duodenal atresia. The parents were informed about the procedure and their written consent was received. The infant had moderate cyanosis and was breathing and crying actively in the operating room. Laboratory findings were within normal limits at the preoperative evaluation, with the exception of moderate physiological hyperbilirubinaemia. Electrocardiography, invasive blood pressure, body temperature and pulse oximetry were monitored and forced air warming was used to keep the infant warm. On admission to the operating room, heart rate, mean arterial pressure and oxygen saturation were

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134 beats min^{-1}, 55 mmHg and 85-87% respectively. A mixture of Lactated Ringer's and 5% dextrose was infused through an IV line at a rate of 4-6 mL kg^{-1} h^{-1}. Arterial blood gas analysis (ABG) was performed before spinal anaesthesia (ABG analysis: pH 7.32, PaO_2 68 mmHg, PCO_2 63 mmHg, SpO_2 89%, BE 4.1, HCO_3 28.8 mmol L^{-1}). EMLA™ was applied one hour before the procedure. Spinal block was performed in the left lateral decubitus position, using a median approach with injection of 0.6 mL isobaric bupivacaine 0.5% using a 25-gauge spinal needle in the L4-L5 interspace. After subarachnoid injection, the neonate was placed in the supine position and was unable to move his feet and legs after 5 minutes. Surgical incision was started after the onset of complete motor blockade. At the initiation of surgery, the infant's rate and depth of breathing was adequate. During surgery, no additional medication was necessary for sedation or analgesia. Mean arterial pressure decreased to 30-35 mmHg, heart rate was stable, at 120-140 beats/min, but SpO_2 levels started to decrease to 75-70% (ABG analysis: pH 7.27, PaO_2 48 mmHg, PCO_2 60 mmHg, BE 0.7, HCO_3 24 mmol L^{-1}). Spontaneous respiratory efforts of the infant were observed to be becoming weaker after 10 minutes of surgery. Breathing was supported by applying manual ventilation with an O_2-air mixture (40-60%) for 20 minutes via a mask. The SpO_2 was 70-85%, and the $ETCO_2$ was 35-47 mmHg during mask ventilation. The same gas mixture was continued with a facemask after spontaneous breathing efforts recovered. The SpO_2 values were maintained at 85-90% with his spontaneous breathing effort. The haemodynamic status was stable and there was no significant change in either mean arterial pressure or heart rate during surgery.

A duodenojejunostomy was performed; the procedure lasted for 120 minutes without complications. The systolic and diastolic arterial pressures and heart rate of the patient were 60-65 mmHg, 30-40 mmHg and 140-155 beats min^{-1} respectively in the last 15 minutes of surgery. An ABG analysis was obtained at the end of surgery. Values were as follows: pH 7.31, PaO_2 78 mmHg, $PaCO_2$ 57 mmHg, SpO_2 92%, BE 2.7, HCO_3 26 mmol L^{-1}. Around 20 mL of crystalloid mixture and 10 mL of fresh frozen plasma were given during surgery. The patient was crying, pulling up and flexing his legs at the end of the operation, and he was admitted to the paediatric surgery intensive care unit with efficient spontaneous breathing.

Discussion

Hypoplastic left heart syndrome is a complex and fatal congenital heart disease that reduces left ventricular blood flow, causing obligatory left-to-right shunting at the atrial level with an atrial septal defect or foramen ovale and right-to-left shunting from the ductus arteriosus. The right ventricle works as a single ventricle and pumps blood through the pulmonary valve (2, 4). The prognosis of a neonate depends on the patent ductus arteriosus, which balances the pressure in the aorta and the pulmonary artery and maintains systemic blood flow (2).

Patients with HLHS may suffer from coexisting congenital anomalies including midline anomalies such as duodenal atresia and neural tube defects, and may need non-cardiac surgery prior to or after cardiac palliation. Anaesthetic management of these patients must lead to minimal haemodynamic changes without disturbing the systemic and pulmonary circulation, balancing both systems to ensure optimum oxygen delivery to the tissues. Spinal anaesthesia has been suggested as a safe technique for high-risk neonates and infants (1, 5). General anaesthesia affects vascular resistance and cardiac function, causing haemodynamic changes. Spinal anaesthesia induces fewer haemodynamic and respiratory disturbances in infants than in adults (6). Katznelson et al. (7) reported that spinal anaesthesia did not cause any significant changes in haemodynamic or respiratory parameters between the preprocedural and intraprocedural period during cardiac catheterization. It was also found that oxygen desaturation, bradycardia and central apnea were more frequent during normal sleep after general anaesthesia compared with spinal anaesthesia (8).

Spinal anaesthesia also causes a lower surgical stress response with minimum haemodynamic changes (9). Anaesthesia management of a patient with HLHS needs to balance pulmonary and systemic blood flow. Perioperative care of these patients is a challenge due to systemic hypoperfusion and pulmonary overcirculation. We preferred not to intubate the patient to avoid the haemodynamic response of laryngoscopy and negative effects of inhalation anaesthetics and mechanical ventilation on cardiac performance and pulmonary vascular resistance. The main goal of spinal anaesthesia was to maintain vital signs within the physiological limits during spontaneous breathing. It was anticipated that pulmonary and systemic circulation would not tolerate the alterations in systemic and pulmonary vascular resistance encountered during general anaesthesia and mechanical ventilation. At the start of spinal anaesthesia the neonate was haemodynamically stable but his breathing effort later weakened. We considered that accessory breathing muscles had been affected by the spinal block, and his effort improved with regression of the block at the end of surgery.

Infants tolerate spinal block with minimal cardiovascular changes due to dominance of the parasympathetic nervous system rather than the sympathetic system (6). Spinal anaesthesia prevented the potential risks of general anaesthesia including haemodynamic disturbances in the perioperative period in this infant. The prematurity of the patient and his low birth weight seemed to be important factors for spinal block level in the perioperative period. Other factors, which contributed to the failure of the breathing muscles, might have been abdominal surgery and severe cardiac abnormalities. Caudal anaesthesia could have been considered as another option in the management of this patient. However, we preferred spinal anaesthesia due to its faster onset, profound sensory blockage and flaccid paralysis with a smaller dose of local anaesthetic, and because confirmation of needle placement was easier by cerebrospinal fluid flow.

Kachko et al. (6) performed spinal anaesthesia in 505 neonates and infants with a mean gestational age of 30.8±3.3 weeks and a mean body weight of 1565.66±639.3 g using a mean dose of bupivacaine of 0.68±0.16 mg kg^{-1}. Haemodynamic disturbances were not observed other than bradycardia without hypotension in nine patients. High spinal block was defined in three premature infants with low birth weight and these patients also experienced perioperative apnea. Four out of 505 infants had postoperative apnea and two of them needed mechanical ventilation in the postoperative period; these patients were premature and had bronchopulmonary disease. The authors suggested that spinal anaesthesia was a safe alternative for surgery in infants. Most studies suggested that spinal anaesthesia was applicable in infants presented for outpatient surgery and complications were rare (1, 10, 11). Williams et al. (12) presented 1554 infants with a body weight ranging from 540 to 7800 g, who underwent general, urologic, orthopaedic, neurosurgical and thoracic procedures under spinal anaesthesia. They

observed that 5% of patients required supplemental oxygen and five patients were intubated, four of them undergoing abdominal surgery and one surgery for inguinal hernia. They recommended spinal anaesthesia even in small premature infants undergoing major surgical procedures. Shenkman et al. (13) reported that the rate of minor complications was 38% in 62 infants with coexisting diseases who underwent inguinal hernia and pyloromyotomy. They speculated that two of the patients suffered hypoxaemia due to diuretic therapy and metabolic status. There are several reports about general anaesthesia in HLHS patients during non-cardiac surgical procedures, but these patients had cardiac palliation prior to surgery and thus might have been more tolerant to the general anaesthetics and their deleterious haemodynamic and respiratory effect in the peri and postoperative period (3, 5).

Conclusion

Anaesthesia care of high-risk premature infants with congenital cardiac abnormalities must include maintenance of a steady state in haemodynamic and ventilatory parameters within the physiological limits of carbon dioxide and oxygen levels. Spinal anaesthesia has been recommended as an alternative method for premature infants with a history of cardiac abnormalities and respiratory problems. Although the use of general anaesthesia is more prevalent in many institutions, the safety and efficacy of spinal anaesthesia has been reported in large groups of paediatric patients (6, 11). The rate of complications of anaesthesia increases with additional risk factors including coexisting disease, type of surgery and the metabolic status of the patient. We recommend spinal anaesthesia as an alternative method for high risk and premature patients having suitable operations.

***Conflict of Interest* / Çıkar Çatışması**
No conflict of interest was declared by the authors.
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***Author Contributions* / Yazar Katkıları**
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