

Caudal Anesthesia in a Neonate with Dandy-Walker Syndrome

Keywords: Dandy-walker syndrome; Caudal block-neonatal

Abstract

Dandy-Walker Syndrome (DWS) is an intrauterine developmental anomaly, which consists of 4-12% of all infants having hydrocephalus its prevalence is 1/30000 in all live births [1,2]. Cranio-facial anomalies such as cleft lip and palate, micrognathia and hypertelorism, together with cardiac and renal anomalies, limb anomalies (syndactyly, polydactyly, etc.) and vertebral anomalies can all be detected in patients with DWS. They may have to undergo a single operation or a series of operations during their childhood period, related to these anomalies. Additionally, these patients may present with features of a difficult airway, together with neurological disorders such as increased intracranial pressure and brain stem dysfunction. All of these situations create numerous serious challenges to the anesthesiologist [3,4]. We were unable to meet any report published in the literature, related to the anesthetic management of patients with DWS until now. We aimed to present our anesthetic management, in a newborn male patient with Dandy-Walker Syndrome, in whom we used caudal block for umbilical cord hernia repair.

Introduction

Dandy-Walker Syndrome (DWS) is characterized by cerebellar hypoplasia, together with cysts communicating with the fourth ventricle of the posterior cranial fossa, and hydrocephalus. Additionally, cleft lip and palate, facial retrognathia, high-arched palate, and abnormal dentition are observed with increased frequencies in patients with DWS [5]. Major concerns of the anesthesiologist should be the difficult airway and neurological problems such as increased intracranial pressure (ICP) and brain stem dysfunction in patients with DWS [3]. The anesthesiologists might encounter difficulties in their preferences of either general or regional anesthesia in patients with such characteristics. Our presented case has shown that regional anesthesia was preferable, when compared to general anesthesia in patients with DWS.

Case Report

The patient, who had male gender, was born at 32 weeks of gestational age with Apgar scores of 6 and 7 and he was diagnosed with prematurity and DWS. He had an antenatal medical history of polyhydramnios and early membrane rupture. He was followed in neonatal intensive care unit.

At his first physical examination, his weight was 1010 gr (low birth weight), he had a syndromic facial appearance, micrognathia, standing right metatarsus adductus, bilateral ulnar deviation, and umbilical cord hernia. His head circumference was 33 cm (>97 percentile) and was interpreted as macrocephaly.

His echocardiographic assessment revealed a pulmonary artery pressure of 45 mm Hg, presence of a bicuspid aortic valve, small perimembranous ventricular septal defect, secundum type atrial septal defect, and thin patent ductus arteriosus.

His cranial ultrasonography findings were mild dilation of both



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ventricles and hydrocephalus. His renal ultrasonography showed a parapelvic cyst, 7x6 cm in size and located at the middle pole of the right kidney.

He was consulted with outpatient clinic of our anesthesia department, prior to his planned umbilical cord hernia repair. His preoperative hemogram values were as follows: Hb: 20.1 g/dl, Htc: 54.3%, platelet 107000/mcl.

The patient was admitted to the operating room. Pulse oxymetry, heart rate and blood pressure were monitored. For prevention of infective endocarditis, ampicillin and amikacin were administered. Following administration of ketamine 2 mg iv, caudal block was performed with 1.25 ml/kg of 0.25% bupivacaine and 0001% epinephrine. Throughout the surgery, his SpO₂ remained between 92 and 94. His heart rate was between 130-140/min, and remained stable for 40 minutes. At 40th minute, the heart rate fell down to 98/min following intravenous administration of atropine 0.02 mg/kg, it returned to normal values. Forty five minutes following the initiation of anesthesia, operation was completed, and the patient was transferred to the neonatal intensive care unit uneventfully in a transport incubator, without being intubated.

On the second postoperative day, the patient deteriorated. Sepsis was diagnosed and due to his respiratory problems, endotracheal intubation was required. However, his intubation by conventional laryngoscopy was not possible. The patient died on his 7th postoperative day, related to sepsis.

Discussion

In patients having DWS, particularly in whom maxillofacial deformities are present, difficult airway management and intubation are frequently observed. Difficult or failed endotracheal intubation is a condition feared by all anesthesiologists for prediction of difficult intubation, numerous attempts have been made until now. Since the use of predictive tests as a part of routine clinical practice is limited in pediatric patients, difficult intubation is especially important in this age group. A clue which might help in predicting the potentially

difficult intubation is the presence of associated congenital anomalies [4].

Most of the conducted studies have reported that approximately 20% to 30% of healthy former preterm infants, undergoing inguinal hernia repair under general anesthesia, manifest at least one apnea during the postoperative period. Regional anesthesia reduces the need for postoperative ventilatory support in very low birth weight infants undergoing hernia repair [6]. We avoided general anesthesia, due to this knowledge. However, Craven et al. have claimed that there was no reliable evidence in the reviewed trials concerning the effect of spinal anesthesia on the incidence of postoperative apnea, bradycardia, or oxygen desaturation in ex-preterm preterm infants undergoing hernia repair, when compared to general anesthesia [7] this issue is controversial.

It is well known that the sympathetic nervous system is sufficiently developed in newborns. Therefore, signs such as systemic hypertension, indicative of hemodynamic instability are very rare during caudal block in newborns and children [8]. Increased intracranial pressure is the most frequent finding in patients with Dandy-Walker Syndrome (90%) and is caused by obstructive hydrocephalus. It was observed in our patient, also. Although there is an ongoing debate on performing caudal epidural analgesia/anesthesia in patients with hydrocephalus and ventricular shunt devices, it has been reported that uneventful ICP changes were observed due to epidural local anesthetic injection [3]. We preferred caudal block for anesthesia for our patient taking support from this report.

Conclusion

Caudal anesthesia can be recommended as an effective

technique for avoiding intraoperative and postoperative anesthetic complications in neonates with DWS.

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