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Anesthetic management of a neonate with Dandy-Walker syndrome

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Introduction

Dandy-Walker syndrome is a congenital disease characterized by absence of cerebellar vermis and cyst development in the posterior fossa which is connected with the fourth ventricle; it results in enlargement of the posterior fossa and hydrocephalus [1].

We reported the anesthetic management of a neonate with Dandy-Walker syndrome who underwent neurosurgery twice and had prolonged convulsive episodes.

Case report

The patient, a male neonate, was born at the 39th gestational week with a birth weight of 3.3 kg. Apgar scores were 9 at 1 min and 10 at 5 min. Anomalies were not found by the examination at birth. There was no particular family history. He was noted to have poor muscle tonus, a poor sucking reflex, and tachypnea during the first few days. On the 6th day, he was transferred to the NICU of our hospital. Computed tomography (CT) showed a cyst in the posterior fossa, and he was diagnosed as having Dandy-Walker syndrome.

Drainage operation through an occipital burr hole for the Dandy-Walker cyst was done on the day of admission. Atropine 0.1 mg i.m. was given as premedication. After administration of lidocaine 4.0 mg i.v., nitrous oxide—oxygen—sevoflurane mask induction was started. Assisted and controlled ventilation by mask was easy, and endotracheal intubation was undertaken with vecuronium 0.3 mg as a muscle relaxant. The trachea was easy to intubate with the standard laryngoscopic technique, and anesthesia was maintained with nitrous oxide—oxygen—sevoflurane and controlled hyperventilation. Vital signs were stable during the operation. At the end of the operation, the return to spontaneous breathing was smooth, and the endotracheal tube was extubated.

At midnight on the day of the operation, he had convulsive episodes. Endotracheal intubation was done in the NICU, and convulsion was controlled with intravenous infusion of thiopental 3 mg/kg per h for 72 h and then phenobarbital 5 mg/kg per day p.o. During the next few days, his head circumference increased, and he was noted to have a tense anterior fontanelle. Lateral ventricle-cyst and cyst-peritoneal shunt operation was done on the 15th day postpartum. He was still intubated. Premedication was atropine 0.1 mg i.m., and anesthesia was maintained with nitrous oxide—oxygen—sevoflurane and controlled hyperventilation with vecuronium. The operation was uneventful. Controlled ventilation was continued after the operation in the NICU. After the second operation, he had convulsive episodes again despite phenobarbital, and rectal administration of diazepam 1.2 mg/kg per day was added for the treatment of convulsions. Weaning from the ventilator succeeded on the 68th day postpartum.

Discussion

Common clinical signs of Dandy-Walker syndrome are the characteristic enlargement of the occiput, macrocrania, mental retardation, cerebellar ataxia, and those of increased intracranial pressure (ICP) [2,3]. About one-fourth of such patients exhibit these signs in the neonate period. Associated congenital anomalies are craniofacial, cardiac, renal, and skeletal abnormali-

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ties [2]. Cerebral anomalies may lead to respiratory failure, such as apneustic breathing [4]. Diagnosis of Dandy-Walker syndrome is usually easy with clinical signs, CT, or magnetic resonance imaging (MRI).

Most patients require shunting surgery to reduce increased intracranial pressure (ICP). A ventriculoperitoneal (VP) shunt operation is usually enough, but some patients may need a double-shunt in the ventricles and the cyst. Generally speaking, intubation should be performed under general anesthesia to avoid increasing the ICP during induction [5]. However, tracheal intubation or airway management is often difficult due to craniofacial malformations, such as micrognathia and cleft palate. Awake intubation is required despite the increased ICP if difficulties in airway management are anticipated [5]. Careful mask induction with inhalational anesthetics to determine whether mask ventilation and laryngoscopy are possible, as done in the present case, is thought to be a safe and useful method in some cases. Intravenous administration of lidocaine may help to attenuate changes in ICP and blood pressure in the induction period. Maintenance of anesthesia should include controlled hyperventilation with muscle relaxation [5]. Close attention to circulatory responses by monitoring changes in ICP is also important. Respiratory failure, or convulsion may occur postoperatively, so observation in the ICU is essential [5]. Patients who had episodes of apnea, convulsion, or aspiration pneumonia due to cerebral or pharyngeal abnormalities often need mechanical ventilation in the ICU, and weaning from the ventilator may be difficult for those patients [5].

In conclusion, careful evaluation of ICP, airway, and associated anomalies, and postoperative intensive care is important in the anesthetic management of Dandy-Walker syndrome.

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