Letter to the Editor

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Anesthetic management of an adolescent with Dandy-Walker syndrome

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Dandy-Walker syndrome is classically described as a neuropathological triad consisting of hypoplasia of the cerebellar vermis, cystic dilatation of the fourth ventricle, and hydrocephalus. Clinical manifestations of the syndrome usually appear in the first year of life, but can occur during the neonatal period. Asai et al. [1] reported that as many as 89% of patients are diagnosed before 1 year of age. However, in the absence of prenatal diagnosis, the symptoms usually become apparent in early infancy and include slow motor development, bulging anterior fontanel, and progressive enlargement of the skull. We report a rare case of anesthetic experience for an adolescent with Dandy-Walker syndrome.

A 13-year-old, 140 cm, 29 kg, male adolescent who had been born prematurely at 28 weeks, weighing 850 g visited our urology for surgical treatment of scrotal hydrocele. The patient had mental retardation and general muscular dystrophy. Six months prior to admission, he had an episode of seizure with vomiting, and brain magnetic resonance imaging was performed. Magnetic resonance imaging showed a cyst in the posterior fossa, hypoplastic cerebellar hemispheres, and elevation of the hypoplastic cerebellar vermis due to the cyst (Fig. 1). He was diagnosed with Dandy-Walker syndrome. In the preoperative evaluation, he was found to have macrocephalus and micrognathia, which could render intubation difficult. He was also afflicted with general muscular dystrophy. During surgery, his vitals were monitored by electrocardiogram, pulse oximetry, noninvasive blood pressure (BP) monitoring, end-tidal CO_2, bispectral index (Aspect Medical Systems, Newton,

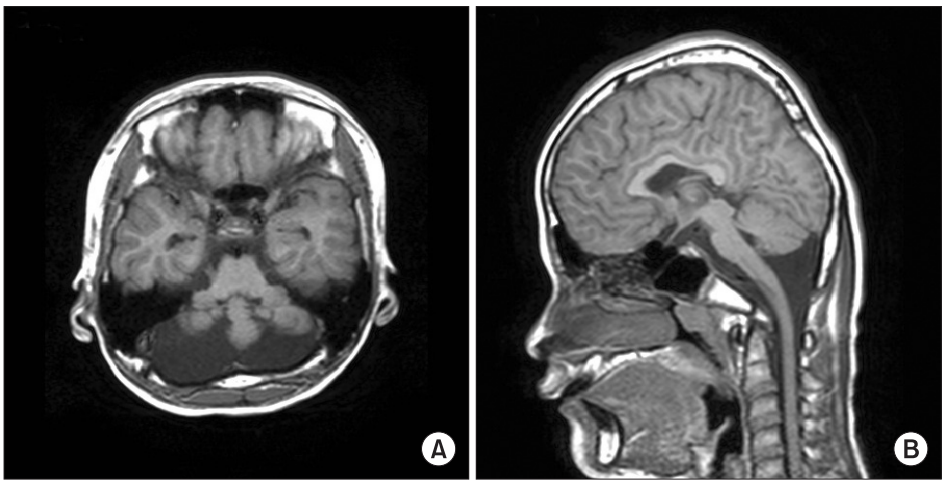

Fig. 1. Brain magnetic resonance imaging shows (A) a cyst in the posterior fossa and hypoplasia of both cerebellar hemispheres, and (B) upward elevation of the hypoplastic cerebellar vermis due to the cyst.

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MA, USA), and a train-of-four monitor (Organon Ireland Ltd., Dublin, Ireland). A 24-gauge catheter was inserted into the left radial artery for continuous BP measurement. After sufficient muscle relaxation, the patient was successfully intubated with a 5.5-mm inner diameter tube, using the standard laryngoscope technique. During the operation, the BP and pulse rate remained stable at 110−120/70−80 mmHg and 60−80 beats/min, respectively. The operation took 2 h and 5 min, and was uneventful.

In older children with Dandy-Walker syndrome, symptoms of increased intracranial pressure (ICP), including irritability and vomiting, and signs of cerebellar dysfunction, including gait disturbance and lack of muscle coordination, may occur [2]. In the present case, the patient had general muscular dystrophy and macrocephalus as well as craniofacial abnormalities such as micrognathia and cleft palate, which can make airway management challenging. Ewart and Oh [3] reported difficult awake intubation of 2-week-old Dandy-Walker patient due to micrognathia and an anteriorly placed larynx. When difficulties in airway management are anticipated, awake intubation is required despite the increased ICP. In our case, fortunately, intubation was less difficult than suspected based on the appearance of a difficult airway.

In this patient, another important anesthetic concern was ICP management. As the patient had shown symptoms of increased ICP, including seizures and vomiting, 6 months before surgery, it was important to prevent a further increase in ICP. Induction and maintenance of anesthesia were attained by continuous infusion of propofol, which reduced ICP. Additionally, continuous infusion of remifentanil was used for pain and BP control. For continuous BP monitoring, an arterial line was inserted into the left radial artery, and the end tidal CO_2 pressure was maintained between 30 and 35 mmHg for ICP control [4]. Finally, owing to possible respiratory difficulty as a result of the muscular dystrophy, train-of-four monitoring was conducted. As convulsions and respiratory failure are possible postoperatively [3], the patient was closely observed in the intensive care unit.

In conclusion, we reported a rare case of anesthetic management of an adolescent with Dandy-Walker syndrome. Careful evaluation of airway anatomy, appropriate airway management, ICP and BP monitoring and control, cautious muscle relaxation, and postoperative intensive care are required for anesthetized patients afflicted with Dandy-Walker syndrome.

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