EDİTÖRE MEKTUP/LETTER TO THE EDITOR
J Turgut Ozal Med Cent 2015;22(4):280-1

Journal of Turgut Ozal Medical Center
www.jtomc.org

General Anesthesia Management of a Pediatric Patient with Dandy-Walker Syndrome

Dandy-Walker Sendromlu Çocuk Hastada Genel Anestezi Yönetimi

Ahmet Selim Özkan, Muharrem Uçar, Hacer Gülerce Uçar, Mahmut Durmuş

İnönü University, Faculty of Medicine, Department of Anaesthesiology and Reanimation, Malatya, Turkey

Dear Editor,

Characterised by the presence of a neuropathological triad of partial or complete agenesis of the cerebellar vermis, cystic dilatation of the fourth ventricle, and enlarged posterior fossa and accompanied by ataxia, increased intracranial pressure, and hydrocephalus, Dandy-Walker syndrome (DWS) is a congenital anomaly with a prevalence rate of 1/30000. Clinical findings especially surface in the neonatal period (25-30%) and the first years of life (75-80%). In addition to craniofacial congenital abnormalities like clefts in the lip and palate, micrognathia, macrocrania, and hypertelorism, several limb and vertebral anomalies like cardiac and renal anomalies, syndactyly, and polydactyly may accompany DWS; therefore, anaesthetic management of these patients is important (2). In this letter, we aim to present the anaesthesia management of a child patient with DWS in the light of the current literature.

A seven-year-old female Dandy-Walker Syndrome patient was scheduled for surgery due to tooth extractions under general anesthesia. The medical history of the patient revealed that she was the child of first-degree relatives and was born with cesarean section in the 37th gestational week from a 35-year-old mother. She was followed in the newborn intensive care unit after the birth. Having fourth ventricle cystic dilatation and vermin hypoplasia, the patient had formerly undergone surgery for ventriculoperitoneal shunt in the second and sixth months after the birth and for cleft palate at the age of one. On physical examination, we noticed an atypically large head and face, large and low-set ears, a high arched palate, microphthalmia, and micrognathia (Figure 1).

We administered standard anaesthesia monitoring and the measured values were normal. Keeping the possibility of potential airway management difficulties in mind, we prepared several different airway equipments, masks, endotracheal tubes, and laryngeal masks in various sizes along with the necessary materials for fiberoptic intubation. After a five-minute pre oxygenation, we applied the induction. As the ventilation was difficult, we provided ventilation by placing airway in the mouth. Seeing that the mask ventilation properly functioned, we administered the muscle relaxants. Even though the orotracheal intubation was first tried by an experienced anaesthetist by using number 4,5 and 4 cuffed transparent tubes, this attempt failed. We therefore assumed that there was narrowing in the vocal cords. We eventually achieved intubation with the help of a stylet and a number 3 tube. The larynx was reported to be a Cormack-Lehane Grade II case. Anaesthesia was maintained with 50/50% nitrous oxide/oxygen along with 2% sevoflurane. Vital signs remained stable during the surgery. During the operation, 12 teeth were taken out. After a two-hour operation and obtaining the adequate spontaneous respiratory movements, the patient was successfully extubated and transferred to the clinic services.

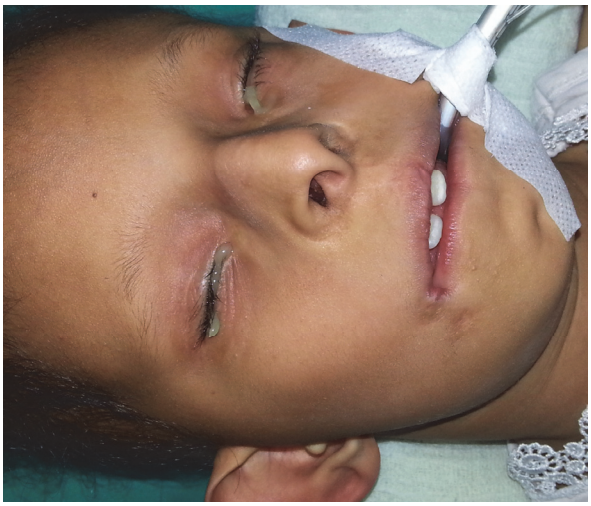

Figure 1. The appearance of the patient with Dandy-Walker syndrome.

Although its etiology is not entirely known, Dandy-Walker syndrome, which is thought to be a combination of environmental and genetic factors, is a rare central nervous system malformation (3). The development mechanism of the syndrome is usually associated with the

hindbrain development process in the 7th-8th intrauterine weeks (4). Clinical findings like large head, hydrocephalus, increased intracranial pressure, and mental retardation widely occur (70-85%) in the early years of life (5).

It has been reported that the risk of difficult intubation is high in children with maxillofacial abnormalities (6). Required preparations should be made and potential reasons that might cause difficult intubation should be identified in advance to ensure intubation safety in these patients (7). In the absence of a previous difficult intubation in patient's history, intubation can be carried out under general anaesthesia. In case of a challenging intubation situation in patients with head and neck anomalies, different approaches such as oral or nasal intubation with fiberoptic bronchoscopy, blind or nasal intubation, supraglottic airway intubation tools, or intubation with video-laryngoscope can be preferred. If required, practitioners are also recommended to use intubation while patient is awake according to the difficult airway algorithm in these patients. Ödeş et al. have reported the successful intubation case of a 2,5-month-old patient with DWS in which they did not use muscle relaxants and applied uncuffed intubation in the first try (8). Demircioğlu et al. have reported a similar case of a DWS patient who was successfully administered endotracheal intubation in the second attempt without the use of muscle relaxants with direct laryngoscopy (9). Jang et al. have reported the case of a 13-year-old DWS patient with micrognathia, macrocephaly, and muscular dystrophy who was intubated in the first go after administering muscle relaxants (10). In addition to the atypical head structure, the presence of micrognathia and cleft palate in our patient strengthened the possibility of a difficult intubation. Despite the easily provided mask ventilation, we encountered a difficult intubation. We did not favour supraglottic airway tools at first since the patient was undergoing surgery in the oral region. Following the induction, we could achieve ventilation by placing an airway to the mouth and this is why we had to use muscle relaxants. Seeing that our case would not be a difficult intubation, we preferred intubation under general anesthesia, and made the necessary preparations. Failing in the first two attempts, we thought that we were facing stenosis in the vocal cords and tried a smaller tube and therefore achieved successful intubation.

Intubation success is directly proportional to the experience of the practitioner. Skills and experience in difficult intubations reduce mortality and morbidity. Therefore, it is important that an experienced anaesthesiologist be present in the operation theatre for such patients. It is needless to say that our case was intubated by an experienced anaesthesiologist.

Consequently, it should be kept in mind that obstructed airway is one of the possible pictures due to concomitant abnormalities in DWS patients and requires preparation. It should also be remembered that intubation must be performed by an experienced anaesthesiologist.

Presented at the Turkish Anaesthesiology and Reanimation Association Congress, 25-29 October 2014, Ankara, Turkey.

REFERENCES

1. Alexiou GA, Sfakianos G, Prodromou N. Dandy-Walker malformation:analysis of 19 cases. J Child Neurol 2010;25:188-91.
2. Wakeling EL, Jolly M, Fisk NM, et al. X-linked inheritance of Dandy Walker variant. Clin Dysmorphol 2002;11:15-8.
3. Çakmak A, Zeyrek D, Çekin A, Karazeybek H. Dandy Walker Syndrome together with occipital encephalocele. Minerva Pediatr 2008;60:465-8.
4. Ten Donkelaar HJ, Lammens M, Wesseling P, Thijssen HOM, Renier WO. Development and developmental disorders of the human serebellum. J Neurol 2003;250:1025-36.
5. Jha VC, Kumar R, Srivastav AK, Mehrotra A, Sahu RN. A case series of 12 patient with incidental asymptomatic Dandy-Walker syndrome and management. Childs Nerv Syst 2012;28:861-7.
6. İsada T, Miwa T, Hiroki K, Fukuda S. The management of the difficult pediatric airway. Masui 2005;54:490-5.
7. Çelik F, Tüfek A, Temel V, Akdemir S, Yıldırım ZB, Kavak GÖ. Dandy-Walker sendromlu çocuk hastada anestezi yönetimi. J Clin Exp Invest 2011;2:327-9.
8. Ödeş R, Özer AB, Gedik F, Erhan ÖL. Dandy-Walker Malformasyonlu hastada genel anestezi uygulaması. Fırat Üniversitesi Sağlık Bilimleri Derg 2006;20:447-8.
9. Demircioğlu Rİ, Özanbarancı A, Karabayırlı S, Usta B, Andıran F. Dandy Walker Sendromlu Hastada Genel Anestezi Uygulaması. Anestezi Dergisi 2012;20:245-8.
10. Jang JS, Lee JJ, Park WJ, Kim EY, Lim SY. Anesthetic management of an adolescent with Dandy-Walker syndrome. Korean J Anesthesiol 2013;64:180-1.

Received/Başvuru: 17.11.2014, Accepted/Kabul: 03.02.2015

Correspondence/İletişim

Ahmet Selim ÖZKAN
İnönü Üniversitesi Tıp Fakültesi, Anesteziyoloji ve Reanimasyon Anabilim Dalı, MALATYA, TÜRKİYE
E-mail: asozkan61@yahoo.com

For citing/Atıf için

Ozkan AS, Ucar M, Ucar HG, Durmuş M. General anesthesia management in a pediatric patient with dandy-walker syndrome. J Turgut Ozal Med Cent 2015;22:280-1 DOI: 10.7247/jtomc.2014.2513