Using the Sugammadex and Gum Intubation Catheter for Difficult Airway Management of a Newborn with Dandy-Walker Syndrome

Letter to Editor

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Abbreviations: DWS: Dandy-Walker Syndrome; NMB: Neuromuscular Blockades; BP: Blood Pressure; ICP: Intracranial Pressure; LMA: Laryngeal Mask Airway

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Dandy-Walker syndrome (DWS), with an incidence of approximately 1 in 30,000 individuals, is a congenital brain malformation characterized by an enlarged posterior fossa, hypoplasia of the cerebellar vermis, and a high position of the tentorium [1]. The clinical expression of DWS occurs 25-30% of the time during the neonatal period, and its clinical manifestations include gait ataxia and obstructive hydrocephalus [2]. Congenital and acquired upper airway obstruction and failed or difficult tracheal intubation in infants can be a challenge or a disaster to the anesthetist, because it is commonly manifested as acute and remains an important cause of mortality and morbidity during anesthesia in pediatric patients due to anatomical variations [3]. The sugammadex sodium injection removes neuromuscular blockades (NMB) in a quick and safe manner, and it plays a growing role in the management of difficult airways. However, its use in difficult pediatric airways is less well documented, although numerous manuscripts have been published about sugammadex in the literature. Here, we report a case of anesthesia for a newborn with Dandy-Walker syndrome using a bridion and gum intubation catheter for difficult tracheal intubation. A 40day-old, 50 cm, 3300 g, male adolescent, who had been born at 36 weeks, visited our pediatric surgery department for the surgical treatment of an inguinal hernia. The patient had a typical facial appearance, PDA, atrophic cerebellar vermis, larger than normal cisterna manga, and hypoplastic bilateral hemispheres; therefore, the diagnosis of Dandy-Walker syndrome was made. In the preoperative evaluation, the patient was found to have microcephalus, as well as craniofacial abnormalities, such as micrognathia, which could render intubation difficult. During surgery, his vitals were monitored via electrocardiogram, pulse oximetry, noninvasive blood pressure (BP) monitoring, and end-tidal CO2. Suspecting that intubation would be difficult in our patient, and to avoid an increase in intracranial pressure (ICP), we used low dose propofol (2 mg/kg) with spontaneous respiration for anesthesia, and attempted to place a laryngeal mask airway (LMA). The patient could not be aerated, since the LMA was not exactly in place, and intubation was initiated. The intubation turned out to be as difficult as we expected. Although the glottis was not visible after the introduction of a laryngoscope (Cormack-Lehane grade 4), utilizing the presited Cook frova gum intubation catheter (Frova, Cook, USA) as a guide, with a 2.5 mm inner diameter tube and tracheal ETT positioning, was confirmed via end-tidal capnography. Rocuronium (Esmeron, Organon, Hollande) was used as a muscular relaxant and for anesthesia maintenance, and pain control was maintained with a propofol and remifentanil infusion, which reduced the ICP. During the operation, the BP and pulse rate remained stable at 60-70/30-35 mmHg and 120-130 beats/min, respectively. The operation took 35 min, and was uneventful. At the end of the operation, for extubation, 2 mg/kg of Sugammadex (Bridion, MSD, Greece) was used, and the patient was extubated without problems and transferred to the intensive care unit. Congenital anomalies, such as a cleft palate, micrognathia and hypertelorism, cardiac, renal, and skeletal malformations, and limb and vertebral abnormalities, are present in 48% of individuals with Dandy-Walker syndrome [4]. Agenesis of the corpus callosum and pontine lesions interfere with the control of respiration, leading to respiratory failure, respiratory spasms, or apneustic breathing [5]. Intubation should be performed under general anesthesia and the maintenance of anesthesia can include muscle relaxation. This is because conscious intubation can have a deleterious effect on the ICP, since Ukwuoma et al. [1] suggested that increased mortality associated with DWS is directly related in DWS patients who did not undergo drainage surgery for hydrocephalus. The other issue which should be kept in mind is that are turn to spontaneous breathing may require more time, because of the structural abnormalities in respiratory control [6]. The gum elastic bougie has been an important tool in the management of difficult intubation in patients with congenital anomalies, such as a cleft palate and micrognathia, since 1949 [7]. When in the trachea, it is easily advanced, and the operator might feel "clicks" from the tracheal rings. The ETT is then moved out over the guide to the trachea via the Seldinger method [8]; however, the definitive and objective confirmation can only be performed by end-tidal capnography. Post-operative residual curarization in postoperative patients can cause delayed recovery, hypoxia, or much worse [9]. The neuromuscular junction in infants is not sufficiently mature, and a child's diaphragm is more vulnerable to NMB. Therefore, the muscles can be easily depolarized, but cannot be easily reversed with cholinesterase inhibitors. In such cases, sugammadex provides extubation with a shorter recovery time and more safely in pediatric patients [10]. The possibility of

reversing a neuromuscular block with sugammadex to awaken the patient adds to the safety of an otherwise difficult airway algorithm in children. The return of spontaneous breathing, in which the patient cannot intubate and cannot ventilate, with sugammadex is almost immediate [11]. In the present patient, since there were structural problems during the awakening stage, it was thought that a mask would be difficult, and since it was also known that intubation was difficult, sugammadex was used.

Conclusion

In conclusion, a gum elastic bougie and sugammadex are useful and essential requirements in difficult airway management in pediatric patients, just as in adults (Figure 1).

Figure 1: The patient with Dandy Walker Syndrome.

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