Case report

Navigating Anaesthetic Challenges in Paediatric Cataract Surgery: A Case Report of Dandy-Walker Malformation

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Abstract

Dandy-Walker malformation (DWM) is a complex disorder which involves malformation in the posterior fossa and cerebellum. It involves various systems like skeletal, cardiac, renal and facial malformation. This case reports highlights the anaesthetic implications and management of complications that can be encountered in a child with such malformation.

Keywords

Dandy-Walker malformation, pediatric cataract surgery, general anesthesia, Sevoflurane, anesthetic management, developmental delay

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Introduction

Dandy-Walker malformation (DWM) is a rare congenital cerebellar malformation that presents with developmental delays, neurological impairments, and often associated with other systemic abnormalities. This case report describes the anaesthetic management of a 14-year-old boy with DWM and multiple comorbidities undergoing cataract surgery under general anaesthesia.

Case Report

A 14-year-old, 21 kg male, with a history of DWM, global developmental delay, non-verbal communication, excessive drooling, clenching of the jaw, churning of teeth, and sleep disturbances, was scheduled for cataract surgery on each eye on different days, with a 7-day interval between the procedures. He was also diagnosed with kyphoscoliosis, developmental cataract, limb hypotonia, crowding of teeth and had a history of recurrent respiratory infections, Figure 1 and 2.

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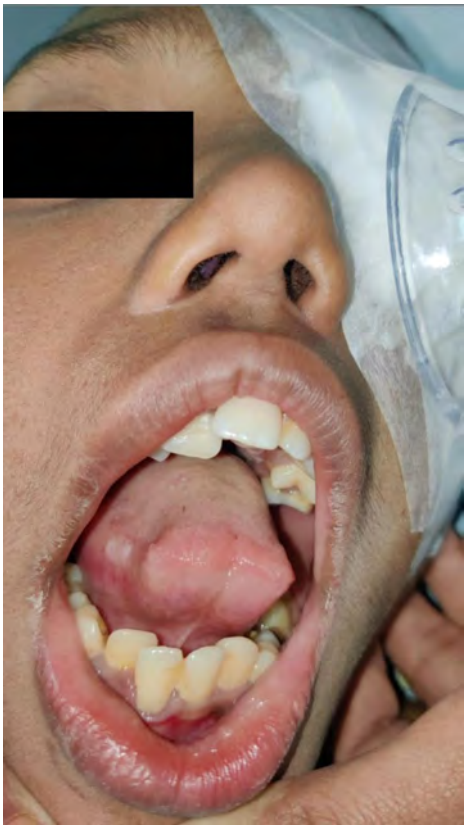

Figure 1- Crowding of teeth

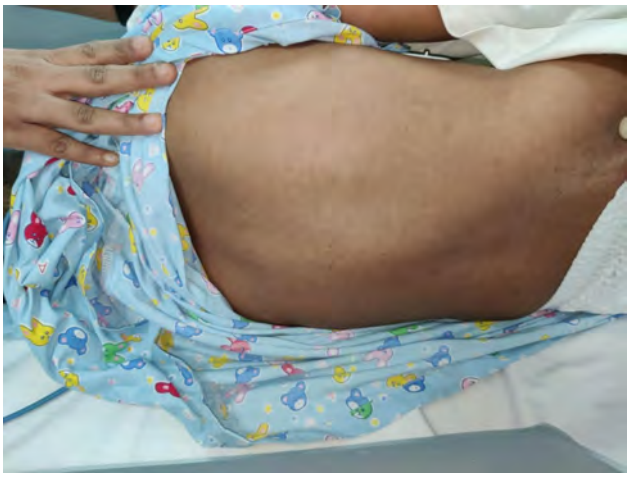

Figure 2- Gross kyphoscoliosis seen on the patient

The patient was born to consanguineous parents with a birth weight of 2.5 kg. He had a history of gastroesophageal reflux disease (GERD), which contributed to feeding difficulties and subsequent developmental delays, along with growth retardation and short stature. There were no cardiovascular and uro-genital abnormalities in the patient. Preoperative assessment included consultation with a paediatric neurologist.

The patient's haemoglobin was 13.2 g/dL, renal function and electrolytes were within normal limits. Given the patient's complex medical history and multisystem involvement, meticulous anaesthetic planning was essential. High risk consent was taken. For the postoperative care, arrangement of ambulance and backup bed in paediatric hospital was done.

On the day of surgery, the patient was taken to the operating room, where an intravenous cannula was secured. We utilized standard ASA monitoring, which includes SpO_2, five-lead electrocardiogram, non-invasive blood pressure measurement, and end-tidal carbon dioxide monitoring. Preoperative medications, including Injection Glycopyrrolate (0.01 mg/kg) to reduce secretions and Injection Ondansetron (0.1 mg/kg) to prevent nausea and vomiting, were administered. General anaesthesia was induced with 30 mg of propofol.

In the first surgery, the patient's airway was secured with a size 2.5 Ambu AuraGain laryngeal mask airway (LMA), and the cuff was inflated. Ventilation was supported with a synchronized intermittent mandatory ventilation (SIMV) mode, with respiratory rate of 18/min and sevoflurane at 1.5 MAC. Intraoperative haemodynamic parameters were stable, with a pulse rate of 98/min, blood pressure of 108/65 mmHg, and SpO_2 of 100%. Surgery lasted for about 20 minutes, the end-tidal CO_2 was monitored throughout the procedure, which remained stable at 28 mmHg. IV paracetamol was administered for analgesia. The LMA was successfully removed without any complications.

In the second surgery, after administering 30 mg propofol, loading dose of Injection Atracurium 0.25 mg/kg IV was given. The patient was intubated with size 5 cuffed endotracheal tube. The ventilatory settings were kept in Volume Control (VC) mode, with tidal volume 160ml, respiratory rate of 16/min, $EtCO_2$ around 30-28 mmHg, maintaining with sevoflurane 1 MAC. Intraoperative vitals were stable, with a pulse rate around 80- 86/min, blood pressure of 98/56 mmHg, and oxygen saturation of 100%. The surgery lasted for about 20 minutes. Reversal was achieved using Injection Myopyrrolate (neostigmine and glycopyrrolate). The patient was extubated successfully, exhibited no signs of delayed recovery, and remained active with stable postoperative vitals.

Post operatively in both surgeries, we observed the patient in the recovery room for 2 hours while administering oxygen via Hudson mask at 5 litres/min. The patient was shifted to step down unit when the modified Aldrete score of 10 was achieved.

Discussion

DWM is a complex disorder which involves the posterior fossa and cerebellum. It is a rare condition with an estimated incidence of 1 in 10,000 to 30,000 births. DWM commonly includes craniofacial abnormalities such as microcephaly, cleft palate, micrognathia, hypertelorism, and cardiac, renal and skeletal malformations such as limb and vertebral anomalies. Cerebral anomalies include agenesis of the corpus callosum leading to poor intellectual development.

Our patient had microcephaly, global developmental delay, absent speech, behavioural issues, stunted growth, kyphoscoliosis, abnormal dentition, hypotonia. Ultrasound abdomen and echocardiogram ruled out renal and cardiac anomalies respectively. While there are numerous reports on DWS in other medical fields, publications related to anesthesia are limited. This case highlights the importance of a thorough preoperative assessment and individualized anesthetic management in pediatric patients with DWM, particularly when associated with multiple comorbidities The anaesthetic management plan focused on securing the airway with an LMA, maintaining stable haemodynamics, and closely monitoring respiratory status. DWS has been linked to various chromosomal abnormalities and presents challenges in airway management. Managing the airway of these patients require careful planning and the use of alternative techniques to ensure airway security. In view of history of GERD, during the first surgery we have used the second-generation LMA that features a gastric port, which helps reduce the risk of aspiration by allowing the drainage of stomach contents and preventing regurgitation into the airway. This design improvement enhances safety, particularly in patients at higher risk of gastric reflux or full stomach during general anesthesia. Selim et al. successfully used an LMA for airway management in a 5-month-old child with DWS.[3] Similarly Buget et al could not visualize the vocal cords in their patient with DWS, and to avoid airway irritation, they utilised LMA.[4] In our patient, during the first surgery, we avoided using a muscle relaxant as the child was hypotonic, which could have increased the risk of delayed recovery.

Considering the child's hypotonia, atracurium was selected for second surgery which undergoes quick elimination through Hoffmann degradation. To minimize the risk of delayed recovery, a lower dose (0.25 mg/kg) was administered, as the cataract surgery was brief in duration. Additionally, the placement of an endotracheal tube eliminated the risk of aspiration. Given the patient's complex medical history, including kyphoscoliosis, developmental delay, and history of respiratory infections, intraoperative and postoperative monitoring was crucial for ensuring a safe outcome. The use of Sevoflurane, which is commonly used in pediatric anesthesia due to its smooth induction and recovery profiles, helped maintain a stable anaesthetic depth.

Additionally, the appropriate use of preoperative medications such as Glycopyrrolate and Ondansetron contributed to the reduction of perioperative risks like excessive secretions and nausea.

Conclusion

Anaesthetic management of paediatric patient with Dandy-Walker malformation requires careful planning and monitoring due to the complexity of the condition and associated comorbidities. This case demonstrates that with appropriate preoperative preparation, intraoperative management, and postoperative care, paediatric cataract surgery can be successfully performed in such patients. Collaboration between the anaesthesiology, neurology, and surgical teams is critical for optimizing outcomes in this unique patient population.

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Conflicts of interest

There are no conflicts of interest.

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