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LETTER TO EDITOR

Anesthesia Management of a Patient with Dandy–Walker Syndrome for Pseudocyst Excision and a Short Literature Review

Sir,

Dandy–Walker syndrome (DWS) is a rare congenital malformation with multiple anomalies. We report a patient with DWS with multiple anomalies who underwent abdominal pseudocyst excision.

A 10-year-old male patient (height: 90 cm and weight: 23.5 kg) with DWS with a ventriculoperitoneal shunt *in situ* was taken for excision of a cerebral spinal fluid abdominal pseudocyst [Figure 1]. He had already undergone postpalatoplasty surgery and multiple lower limb corrective surgeries and had a history of multiple failed intubations during previous surgeries. Airway examination revealed macrocephaly, micrognathia, retrognathia, Class IV Mallampati airway, and arthrogryposis [Figure 2]. The patient's serum creatinine was measured at 0.3 mg/dl, and blood urea was 16 mg/dl. A transthoracic echocardiography revealed a left ventricular ejection fraction of 60%. Intubation with direct laryngoscopy with a stylet was planned in anticipation of the difficult airway. In addition, the difficult airway carts were kept ready. In the operating room, standardized monitors were placed and preoxygenated with 100% oxygen along with 100 microgram of fentanyl and 0.2 mg of glycopyrrolate intravenously (IV). The patient was induced with 100 mg of IV propofol. Once bag and mask ventilation were achieved, an injection of 20 mg of atracurium IV was given. Laryngoscopy revealed a Cormack–Lehane Grade IV, and a 5.5 mm internal diameter-cuffed endotracheal tube was inserted into the glottis using a stylet, resulting in unintentional esophageal intubation. We proceeded with fiberoptic intubation. Given that a muscle relaxant had already been administered, a fiberoptic bronchoscope was used. On the first attempt, a 5.5 mm internal diameter tube was successfully placed inside the trachea. The tube was railroaded over the bronchoscope into the trachea. Correct tube placement was confirmed using end-tidal carbon dioxide monitoring and the auscultation method, verifying proper ventilation and placement. Anesthesia was maintained with O2:N2O (40:60 ratio) and 1%–1.5% sevoflurane. The surgery lasted for 4 h, and extubation of the trachea was uneventful. Postoperative course was uneventful.

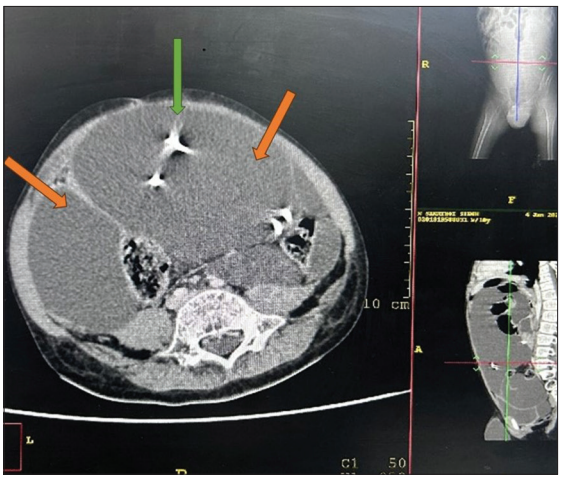

Figure 1: Computed tomography scan of the abdomen of the patient showing a large fluid collection (orange arrow) along with the ventriculoperitoneal shunt tip (green arrow)

DWS affects 1 in every 30,000 live births, resulting in hydrocephalus and elevated intracranial pressure (ICP) in 2%–4% of patients within the 1st year.[1] Increased ICP may produce vomiting, necessitating fluid resuscitation before surgery. A study on craniotomy demonstrated that a 10° head-up position can lead to a reduction in ICP without significantly affecting cerebral perfusion pressure (CPP).[2] We avoided excess sympathetic stimulation using judicious opioids to maintain ICP and CPP. We maintained normocarbia to preserve the vasoconstrictive and vasodilatory cascade that helps regulate cerebral blood flow and the ICP. We chose sevoflurane as the optimal anesthetic agent for maintaining the CPP and ICP. We avoided neuraxial block since 90% of DWS patients have elevated ICP with hydrocephalus.[1]

Cardiac abnormalities are observed in 18% of DWS patients.[3] Renal and cardiac anomalies must be ruled out during the preanesthetic checkup.[3] Cerebral agenesis interferes with the medullary control of respiration, resulting in respiratory failure. Commonly associated anomalies with DWS create difficulties in airway management.[4] Anesthesia

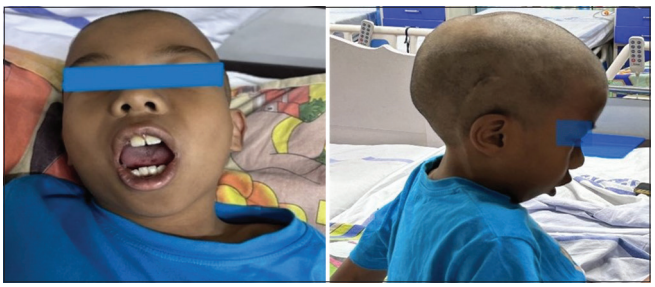

Figure 2: Airway imaging showing macrocephaly, micrognathia, retrognathia, and Class IV Mallampati airway

management necessitates a thorough airway evaluation, planned difficult intubation and extubation, and postoperative care to ensure close observation and monitoring for respiratory problems. Our patient had a history of multiple failed intubations during previous surgeries due to a difficult airway and large head size. In light of this, we took special precautions by ensuring that alternate airway access methods were readily available, including a fiberoptic bronchoscope and appropriate airway adjuncts. We followed the Difficult Airway Society guidelines for pediatric airways. Other options for intubation in our patient included fiberoptic intubation with airway block.

In conclusion, patients with DWS should be anticipated to have difficult airway management. A viable alternative method for airway control, ICP, and CPP, together with a careful preoperative examination, should be considered.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Data availability statement

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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