Case Report

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Dandy Walker Syndrome – AlwaysA Challenge

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Abstract

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. Obstructive hydrocephalus with dilatation of the third and lateral ventricles occurs frequently.' The syndrome was first described in 1914 by Dandy and Blackfan who postulated the pathogenetic theory of atresia of the fourth ventricle's foramina of Luschka and Magendie; but today most authors consider the cerebellar hypoplasia to be a malformation independent of this atresia. DWS is also associated with abnormalities in the skeletal, cardiac, and genitourinary systems. In this report, we aimed to present a case of a patient with DWS who was scheduled for VP shunt surgery and was managed successfully without any perioperative complications.

Keywords: Dandy-Walker Syndrome, Hydrocephalus, VP Shunt.

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Introduction

Dandy–Walker Syndrome (DWS) is a rare congenital brain anomaly affecting the cerebellum and the fourth ventricle. It has a reported incidence of 1: 25 000–1: 30 000 with a slight female predominance. The chief components of the syndrome include cystic dilatation of the fourth ventricle and agenesis or hypoplasia of the cerebellar vermis. These abnormalities are typically associated with hydrocephalus. Patients often present in infancy for cerebrospinal fluid shunt procedures. Anaesthetic management of patients with DWS may prove to be challenging due to the frequent association of abnormalities in other systems. Airway management also proves to be difficult as patient have a large head size and may also be associated with craniofacial abnormality. We describe a case of four year old child with Dandy-Walker syndrome and malfunctioned VP shunt in-situ for revision of VP shunt. Written consent was taken from mother to publish a case report.[1-5]

Case Report

A four year old male child known case of Dandy-walker syndrome previously operated for VP shunt in view of hydrocephalous and raised intracranial pressure (ICP) was posted in emergency operation theatre for revision of VP shunt procedure as patient had signs of raised ICP and drowsiness since 10 days.

Patient had history of seizure disorder for which he was on antiepileptic medications. No history of feeding difficulties was reported and he had well-coordinated swallowing with no fatigue on feeding. There was no history of apneas or breathing abnormalities neither there was a history of aspiration or recurrent aspiration. On examination the child weighed 18kg and height of 122 cm with a head circumference of 59cm. there was no abnormalities of eye movements and showed no signs of cranial nerve abnormalities.

A difficult intubation was anticipated in view of large head size and craniofacial abnormalities, so difficult airway cart was kept ready.

Patient's preoperative haemoglobin was 11.2 g/dl. Renal and liver functions were in normal range.

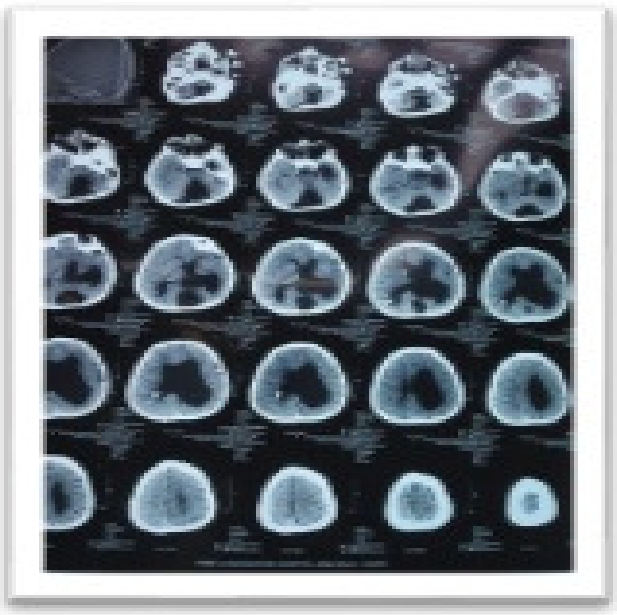
After all standard monitors were placed on patient, induction of anaesthesia was achieved with sevoflurane and oxygen. Intravenous line was secured with 22G cannula on dorsum of left hand. Patient's position was optimized with the use of rolls of towels to elevate body to prevent hyperflexion of neck. Analgesia was given with fentanyl (2mcg/kg). When end-tidal sevoflurane was 2.8% and patient was induced properly, endotracheal intubation done and airway secured with size 4.5(ID), and ETT was fixed at 12cm marking and left side angle of mouth after conforming bilateral air entry equal and adequate. After securing airway non-depolarizing muscle relaxant (Atracurium,0.5 mg/kg) was given to facilitate mechanical ventilation. Mechanical ventilation was adjusted to prevent hypercarbia. After induction left radial artery was cannulated with 24g arterial catheter for continuous hemodynamic monitoring. Forced air warmer was used to prevent hypothermia.

Local anaesthetic infilteration was done with bupivacaine by surgeon.

The patient was haemodynamically stable throughout the

surgery. Good respiratory efforts were seen at the end of surgery and muscle relaxation was reversed with glycopyrrolate and neostigmine.

After an uneventful stay in recovery room for one hour, patient was shifted to general ward. Patient did not experience any apnoea or respiratory complication.



Discussion

The term Dandy Walker Syndrome was first used by Benda to describe the group of congenital posterior fossa abnormality. Whilst DWS has been discussed extensively in paediatric and radiological literature, relatively little has been published about it in anaesthesia-related literature. This probably relates to its rare incidence. Despite this, anaesthetists should be aware of the syndrome and its associations since up to 80% of cases will develop hydrocephalus and will require anaesthesia and surgery. The aetiology and pathogenesis of this condition have not yet been determined, but the presence of associated abnormalities is suggestive of an early embryonal developmental disturbance affecting more than just the posterior fossa structures.

In children with Dandy-Walker syndrome, symptoms of increased (ICP), which includes irritability and vomiting, and signs of cerebellar dysfunction, which includes gait disturbance and lack of muscle coordination, may occur.[6,7]

When difficulties in airway management are anticipated as in this case, awake intubation may be required despite the increased ICP. In this case, fortunately, intubation was not too difficult despite large head size and craniofacial abnormality. Another important anesthetic concern was ICP management. As the patient had shown symptoms of raised ICP, i.e seizures and drowsiness, it was important to prevent a further increase in ICP. Induction and maintainence of anesthesia were attained by sevoflurane at higher inhalational levels to keep patient deeply anaesthesized, thereby reducing ICP.

Additionally Paracetmol infusion 10 mg/kg infusion 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 CO2 pressure was maintained between 30 and 35 mmHg for ICP control. As convulsions and respiratory failure are possible postoperatively, the patient was closely observed in the intensive care unit.

Conclusion

Wereported a rare case of anesthetic management of a child 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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