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ANAESTHETIC MANAGEMENT OF A CHILD WITH DANDY WALKER SYNDROME FOR OPHTHALMIC SURGERY

Anaesthesiology

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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.Asai et al. [1] reported that as many as 89% of patients are diagnosed before 1 year of age. However, in the absence of prenatal diagnosis, the symptoms usually become apparent in early infancy and include slow motor development, bulging anterior fontanel, and progressive enlargement of the skull. Dandy-Walker syndrome (DWS) is a very rare congenital malformation. Its characteristic features are cystic dilatation of the fourth ventricle, enlarged posterior cranial fossa with upward displacement of the tentorium, and lateral sinuses with agenesis or hypoplasia of the cerebellar vermis. DWS is also associated with abnormalities in the skeletal, cardiac, and genitourinary systems.

KEYWORDS

Anaesthetic Management, Dws, Cataract Surgery

INTRODUCTION:

The etiopathology of Dandy–Walker malformation (DWM) consists of the developmental failure of the roof of the fourth ventricle during embryogenesis that results in the cystic expansion of the fourth ventricle in the posterior fossa. It presents early in life, 80–90% within 1st year with features of raised intracranial pressure (ICP). It is well-established that Dandy-Walker syndrome (DWS) is frequently associated with other systemic anomalies including cardiovascular malformation.

Anesthetic management of DWS patients can pose serious threats because of multisystemic association of craniofacial abnormalities, hydrocephalus, renal, and cardiac pathologies [2]. Difficulties in intubation are frequently encountered .

Course in the hospital :

- A 38 weeks born female baby, at present 6 months weighing 7.4 kgs, was diagnosed with DWS with infantile spasms. Birth weight 2.3 kgs and had signs of global developmental delay.
- All haematological investigations were unremarkable.
- EEG revealed severe epileptiform dysfunction with modified hypsarrythmia. Baby was started on T. Vigabatrin 500 mg ¼ BD
  T. Clonazepam 0.25 mg BD
  Neurology clearence obtained

This baby was referred from a neighbouring country to our institution and was scheduled for Lens aspiration and intra ocular lens placement. An informed high risk consent was obtained and pre-op neurology clearence was also obtained. Perioperatively all anti epileptic medications were continued After following universal fasting guidelines, and premedication with glycopyrolate, the baby was induced with oxygen, nitrous oxide and sevoflurane. An I.V. access was obtained using 24 G cannula and 0.45% DNS was started. Intra op monitoring was done with ECG, NIBP, SaO2, EtCO2 and body temperature. Baby was kept warm using hot air blanket.

After ascertaining the correct depth of anaesthesia, the trachea was intubated with 4 mm plain ET tube and connected to ventilator. Fentanyl 1 MCG/ kg was used for analgesia. Anaesthesia was maintained with oxygen, nitrous oxide and sevoflurane. Vitals remained stable for the duration of surgery which lasted for about one and half hours. A naso gastric tube was passed to deflate the stomach. Baby was extubated completely awake. Baby was observed in PICU for 3 hours and was shifted to ward after feeding was established.

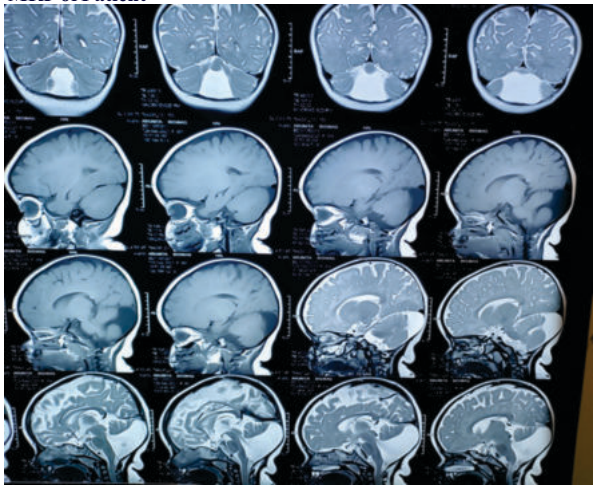
DISCUSSION :

The term 'Dandy–Walker Syndrome' was first used by Benda in 1954 to describe this group of congenital posterior fossa abnormalities.
The aetiology of DWS is heterogeneous and it may result from chromosomal disorders or environmental factors. In addition a single gene error has been implicated in cases where it occurs as part of other syndromes like Warburg Syndrome or Meckel–Gruber Syndrome.[3] Dandy–Walker malformations may present as part of PHACES syndrome (posterior fossa abnormalities of the brain, haemangiomas of the face and scalp, arterial abnormalities, cardiac defects, eye anomalies and sternal abnormalities). There is also evidence that heterozygous deletions of ZIC1 and ZIC4 genes may play a role in the development of DWS[4] Dandy–Walker syndrome is characterized by various neuro pathological features like, cystic dilatation of fourth ventricle, hypoplasia of cerebellar vermis, hydrocephalus, elevation of tentorium and or transverse sinus, enlargement of posterior fossa, lack of patency of foramen of luschka and/or magendie[2]. The incidence of DWM is 1:25,000–30,000 of live births and signs and symptoms of hydrocephalus and raised ICP appear within the 1st year of life in 2–4% of patients [3] In addition, associated cerebral anomalies like agenesis of corpus callosum, pontine lesions, and interference with medullary control of respiration can precipitate frequent episodes of apnea, apneustic breathing, and respiratory failure[5,6,7] Anesthetic management of DWS patients can pose serious threats because of multisystemic association of craniofacial abnormalities, hydrocephalus, renal, and cardiac pathologies. Difficulties in intubation are frequently encountered .

Thus, a multidisciplinary approach is helpful in safe conduct of anaesthesia.

Financial Interest : nil
Conflict of Interest : nil
(An oral permission was obtained from parents for publication without revealing the patient's identity.)

MRI of Patient



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