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Dandy Walker Syndrome: A case report

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Abstract

Background: Dandy-Walker malformation is a rare congenital malformation which was first described by Dandy and Blackfan in 1914. It is defined as the characteristic triad of complete or partial agenesis of the vermis, cystic dilatation of the fourth ventricle and an enlarged posterior fossa with upward displacement of lateral sinuses, tentorium, and torcularherophili. It is frequently associated with genetic anomalies and systemic malformations.

Case summary: A 4 months old infant weighing 4 kg born full term normal vaginal delivery presented with abnormal head size since birth. As our case was anticipated to have difficult airway due to large size of head, incomplete neck extension, cleft palate, micrognathia and incompletely formed tongue, airway management was a challenge. Hence it was important to properly evaluate and manage the airway. Difficult airway cart was kept ready. Patient was brought to the edge of the operating table with an assistant lifting shoulders and supporting neck to facilitate intubation. Measures to control ICP like controlled hyperventilation, normocapnia, normotension, patient positioning, euglycemia, anticonvulsants and temperature monitoring were taken.

Conclusion: Major anesthetic concerns are control of intracranial pressure and evaluation of accompanying organ anomalies. We report successful anesthetic management with special mention of difficult airway in an infant with Dandy-Walker malformation for ventriculo-peritoneal shunt procedure.

Keywords: Dandy-Walker malformation, hydrocephalus, ventriculo – peritoneal shunt

Introduction

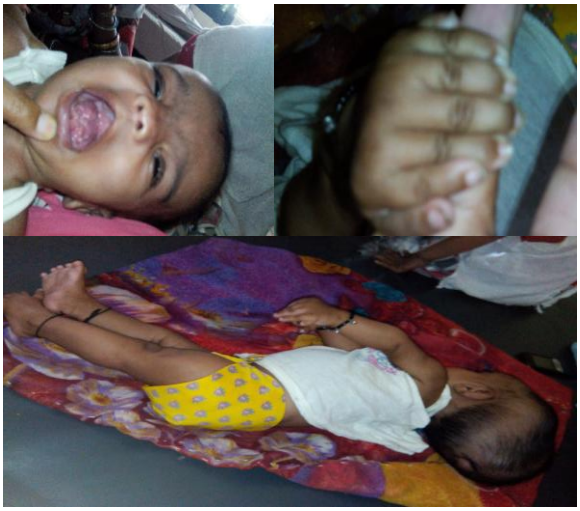
DANDY-WALKER syndrome (DWS) is a nonfamilial syndrome characterized by cystic dilatation of the fourth ventricle and aplasia or partial or total atrophy of the cerebellar vermis. It has a slight female predominance and an incidence of around 1 in 25000-30000 live births. It is typically found in association with supratentorial hydrocephalus, clinical presentation is most often heralded by symptoms and signs of hydrocephalus with focal neurologic findings. Associated congenital anomalies are said to be present in 48% of cases. These commonly include craniofacial abnormalities such as cleft palate, micrognathia and hypertelorism, cardiac, renal and skeletal malformations including syndactyly, polydactyly, limb and vertebral abnormalities. Cerebral anomalies include

agenesis of the corpus callosum with poor intellectual development, irregular respiration and apneic spells.[1,2]

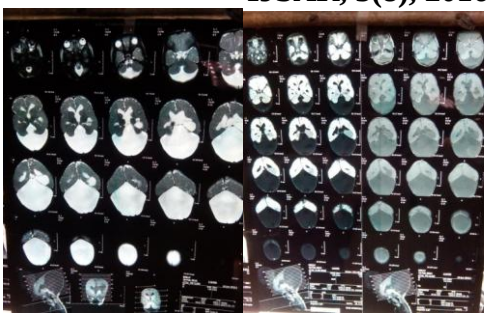
We report a rare anesthetic experience of a 4 months old infant diagnosed with dandy walker syndrome posted for V-P shunt surgery

Case report

A 4 months old infant weighing 4 kg born full term normal vaginal delivery presented with abnormal head size since birth. Head circumference was 52 cm, lax anterior fontanelle, occipital bone elongated and bulging posteriorly. Cleft lip, cleft palate, incompletely formed tongue, micrognathia and polydactyly were present. Milestones were delayed, social smile was present but head holding was absent. Crying to pain and consolable .Sucking was normal. There was no other associated complaints. Lab reports were within normal limits.

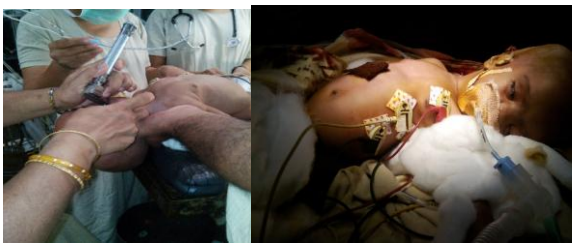


MRI brain showed cystic lesion in posterior fossa communicating with 4th ventricle with cerebellar atrophy and vermis hypoplasia, corpus callosum agenesis with left temporal arachnoid cyst.



Preoperative preparation was specifically done keeping in mind an anticipated difficult airway. Difficult intubation cart was kept ready.

Appropriate positioning was given by bringing the patient to edge of the table with assistant supporting the neck and shoulders to allow for head extension and to accommodate the large sized head. This was done to facilitate intubation. As it was anticipated difficult airway, child was sedated with sevoflurane 5% and was kept on spontaneous ventilation. First attempt of laryngoscopy was unsuccessful. Bag mask ventilation was done. Laryngoscopy was done again, and child was intubated with 2.5 mm ID uncuffed ET Tube with the help of a stylet.



Intraoperatively, vitals were monitored with ECG, pulse oxymetry, NIBP, ETCO2 and temperature probe. Patient was maintained on sevoflurane with oxygen and nitrous oxide with intermittent dose of inj atracurium 0.4 mg. Inj. fentanyl 8 mcg was given to deepen the plane of anesthesia. During the course of the surgery child remained hemodynamically stable .Procedure lasted for an hour. During extubation “at risk” algorithm was followed.

When the child started having good regular respiratory efforts and was fully awake, reversal consisting of inj neostigmine 0.2 mg and inj glycopyrolate 0.04 mg was given intravenously.

We extubated the patient as saturation was maintained on room air at 99-100% and was moving all four limbs. Extubation was uneventful.

Post operatively patient was shifted to ICU.Post operative pain management was done with paracetamol (25 mg) suppository.

Discussion

Clinical manifestations of the syndrome usually appear in the first year of life, but can occur during the neonatal period. Asai et al reported that as many as 89% of patients are diagnosed before 1 year of age.[3]

However, in the absence of prenatal diagnosis, the symptoms usually become apparent in early infancy and include slow motor development, poor head control, irritability, setting sun sign, bulging anterior fontanelle, progressive enlargement of the skull, cranial nerve palsies, irregular respiration and apneic spells.

In older children with Dandy-Walker syndrome, symptoms of increased intracranial pressure (ICP), including irritability and vomiting, and signs of cerebellar dysfunction, including gait disturbance and lack of muscle coordination, may occur.[4]Most patients with DWS develop hydrocephalus and require a ventriculoperitoneal shunt to reduce ICP.

Children undergoing anesthesia for neurosurgical procedures present unique challenges for the anesthesiologists.

The method of induction is decided by the circumstances of the case, goal is to avoid rise in ICP and avoid hypotension. Intravenous induction allows for rapid control of airway if patient is not fasted.

Inhalational induction should be avoided as they dilate cerebral blood vessels in a dose dependent manner causing raised ICP. However Isoflurane and Sevoflurane in less than 1.0 MAC concentration does not affect the operating conditions significantly.[5] Proper positioning using a pad or bolster under shoulders or using lateral position may help in difficult intubation. Endotracheal intubation should be very smooth. If tracheal intubation or airway maintenance is difficult, like in our case due to hydrocephalus, micrognathia , cleft palate and incompletely formed tongue the advantages of awake intubation must be weighed against the deleterious effect on ICP. Maintenance of general anesthesia should include controlled ventilation with muscle relaxation. General measures like maintaining normotension, normocapnia, euglycemia, normal acid base balance, control of seizures with anticonvulsants and specific measures like cerebrospinal fluid drainage, controlled hyperventilation, use of barbiturates, hypothermia helps in reducing ICP as in the case of hydrocephalus.[6] A period of postoperative ventilation in ICU may be needed in these patients as they may suffer from recurrent apnea.[7] But in our case post op respiratory efforts were good and patient did not require mechanical ventilation in post-operative period.

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