Contents lists available at www.innovativejournal.in

INNOVATIVE JOURNAL OF MEDICAL AND HEALTH SCIENCE

Journal homepage: http://www.innovativejournal.in/index.php/ijmhs

ISSN No. 2277-4939

DANDY WALKER SYNDROME: CASE REPORT

Dr Shweta Mane[1] Dr Shruti Rao[2] Dr S.D Ladi [3] Dr S.S Aphale[4]

1 Lecturer, Department of Anaesthesiology, Bharati vidyapeeth deemed university, Pune, Maharashtra, India.
2 Junior Resident II, Department of Anaesthesiology, Bharati vidyapeeth deemed university, Pune, Maharashtra , India.
3 Professor, Department of Anaesthesiology, Bharati vidyapeeth deemed university, Pune, Maharashtra , India.
4 Professor and Head of Department, Department of Anaesthesiology, Bharati vidyapeeth deemed university, Pune, Maharashtra , India.

ARTICLE INFO

Corresponding Author:

Dr Shruti Rao

Junior Resident II, Department of Anaesthesiology, Bharati vidyapeeth deemed university, Pune, Maharashtra , India.

Keywords: Dandy Walker Syndrome, shunt surgery, hydrocephalus

ABSTRACT

Dandy Walker Syndrome is rare congenital malformation of the brain occurring i 30,000 births.It is characterized by a classical neuropathological triad consisting the cerebellar vermis, cystic dilatation of the fourth ventricle and hydrocephalus. year female child diagnosed as Dandy Walker Syndrome at four months of age, pr excessive crying, irritability, delayed milestones and increase in the size of the he operated at 6 months of age for VP shunt surgery, as the symptoms recurred; she revision of VP shunt surgery in our institution.As the case was anticipated as a dif due to the large size of the head and inadequate neck extension which made airw management challenging in this patient.Hence, careful evaluation of airway anato appropriate positioning and airway management intra operatively were essential anaesthetize a patient afflicted with Dandy Walker Syndrome.A rare syndrome p considerable airway challenge, managed successfully.

©2014 IJMHS, All Right Reserved

INTRODUCTION

Dandy walker syndrome is characterized by a nueropatholigical triad comprising of hypoplasia of the cerebellar vermis , cystic dilatation of the fourth ventricle and hydrocephalus.

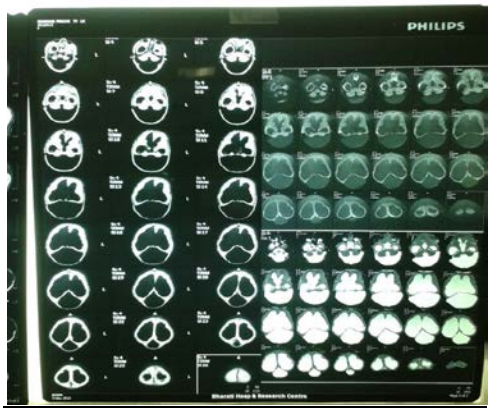
Obstructive hydrocephalus with dilatation of the third and lateral ventricles occurs frequently. This syndrome was first described by Dandy and Blackfan in 1916. Associated congenital anomalies are said to be present in 48 % of cases. These commonly include craniofacial anomalies such as cleft palate, micrognathia, hypertelorism, cardiac, renal and skeletal malformations. Cerebral anomalies include agenesis of corpus callosum with poor intelligence and interference with medullary control of respiration often resulting in respiratory failure. Signs and symptoms of this disease become evident in the first year of life with slow motor development, bulging anterior fontanel and progressive enlargement in the size of skull. We report a rare anesthetic experience of a seven year old child diagnosed with dandy walker syndrome posted for revision of VP shunt surgery.

CASE REPORT:

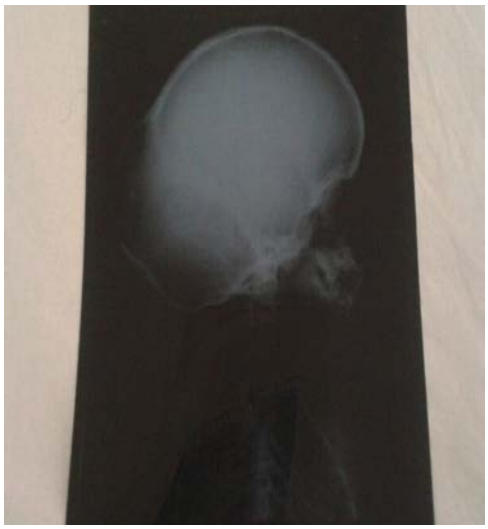
The patient was a seven year old child weighing 35 kgs. She was born full term caesarian section in view of large size of head. She was diagnosed at 6 months of age with Dandy Walker Syndrome. She presented with irritability, excessive crying and increase in size of head. She was operated thereafter for VP shunt surgery. Her symptoms correspondingly reduced till about 6 months back. She presented with symptoms of frequent episodes of severe headache intermittently accompanied with irritability. On preanaesthetic examination, she had a macrocephalus measuring 74 cms (head circumference) and micrognathia. Her mouth opening was adequate (MP 1) and neck

movements were restricted due to the size of her head. Her mental development was normal with delayed milestones. The rest of her systemic examination was normal.

Her MRI brain revealed a large cyst in posterior fossa causing enlargement of the fossa and agenesis of cerebellar vermis, severe stenosis of aqueduct of sylvius with severe enlargement of 3rd and lateral ventricles. (Figure 1)

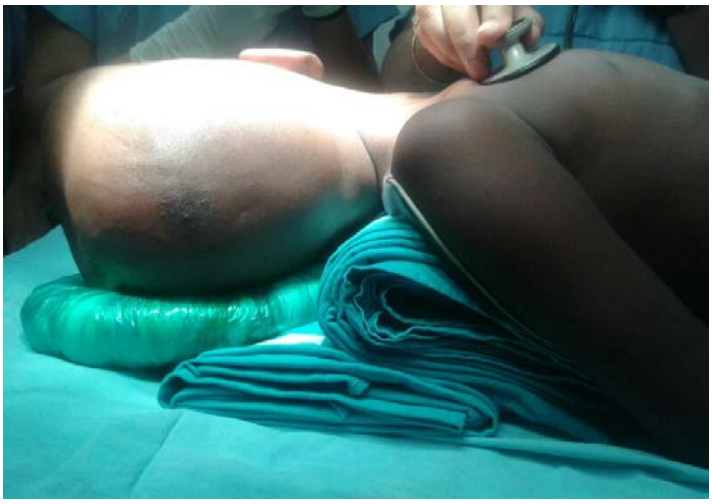


Her X-ray skull showed bulging posterior fontanelle, doliocephalic with enlarged occipital segment. VP shunt in situ. (Figure 2)



Preoperative preparation was specifically done keeping in mind an anticipated difficult airway due to the patient's macrocephalus and micrognathia. Difficult intubation cart kept ready. Adequate bolsters and head rings to facilitate intubation.Intraoperatively her vitals were monitored by an ECG, pulse oxymetry, NIBP, ETCO2. An intravenous access secured inside the operation theatre with a 22 G intracath. She was given adequate positioning prior to induction of anesthesia.

This positioning comprised of placing adequately sized bolsters underneath her shoulders providing neck extension and a head ring underneath her head to accommodate the large size. This position gave an appropriate position for intubation. She was induced on injection thiopentone sodium with a dose of 5mg/kg and injection succinycholine 2mg/kg was given to facilitate endotracheal intubation. She was successfully intubated with a 5.5 number cuffed flexometalic tube using the standard laryngoscopic technique.



During the course of the surgery she remained haemodynamically stable with her pulse rate being maintained between 70-90/minute and blood pressure between 110-120 /70mm of hg. She was being maintained on oxygen with nitrous oxide and sevoflurane with injection vecuronium given intermittently bolus doses for muscle relaxation. The procedure took one hour and went uneventful. Prior to extubation patient was given same position as given to intubation. Patient showed smooth regular respiratory attempts prior to extubation which supported the decision to extubate the patient. She maintained saturation on room air between 99- 100 % so we proceeded with extubation with injection neostigmine0.04mg/kg and injection glycopyrollate 0.01mg/kg. It was uneventful.post operatively patient was shifted to pediatric intensive care unit for further monitoring.

DISCUSSION:

DANDY WALKER SYNDROME requires neurosurgery. Neither marsupialisation nor excision of the membranes of the cystic fourth ventricle has shown definitive improvement. A VP Shunt operation frequently suffices.

An Anesthetist may encounter the DWS in a few situations, first to anaesthetize or sedate these patients for CT or MRI investigations. And secondly when they are posted to surgery. If the ICP is raised it will be exacerbated by any depression of ventilation so general anesthesia with controlled ventilation is preferred. Return to spontaneous breathing may be delayed because of inherent abnormalities in respiratory control and may need a period of post operative ventilation [1].

In older children with Dandy-Walker syndrome, symptoms of increased ICP, including irritability and vomiting, and signs of cerebellar dysfunction, including gait disturbance and lack of muscle coordination, may occur [2]. In the present case, the patient had macrocephalus as well as craniofacial abnormalities such as micrognathia which can make airway management challenging. Ewart and Oh reported difficult awake intubation of 2-week-old Dandy-Walker patient due to micrognathia and an anteriorly placed larynx. When difficulties in airway management are anticipated, awake intubation is required despite the increased ICP. In our case, fortunately, intubation was less difficult than suspected based on the appearance of a difficult airway. In this patient, another important anesthetic concern was ICP management. As the patient had shown symptoms of increased ICP, including seizures and vomiting, 6 months before surgery, it was important to prevent a further increase in ICP. Therefore the patient was maintained on sevoflurane and regular doses of muscle relaxants.Hydrocephalus may cause raised ICP where anesthetic induction should be performed without increasing cerebral blood flow and cerebral metabolic rate. General measures like maintaining normocapnia, normotension and euglycemia helps in reducing ICP. Maintenance of anesthesia should include controlled ventilation and muscle relaxants.[3] Endotracheal intubation should be very smooth as difficulties are encountered due to large size of head in hydrocephalus by placement of shoulder bag.

A period of postoperative ventilation may be required for such patients, as they suffer from recurrent episodes of apnea. Our patient returned back to spontaneous ventilation soon after the surgery and did not need ventilatory support as respiration was good.

CONCLUSION:

Thorough evaluation of the patient's condition, measures taken to control ICP and postoperative care lead to successful management of a patient afflicted with dandy walker syndrome.

ABBREVIATIONS:

DWS: Dandy Walker Syndrome

MRI: Magnetic resonance imaging

MP: Mallampatti grading

ICP: Intracranial pressure

VP: Ventriculoperitonial

REFERENCES:

1. Ewart MC,Oh TE. The Dandy Walker syndrome. Relevance to anesthesia and intensive care.Anesthesia 1990;45:646-8.
2. Ji Su Lang, Jae Lee. Anesthetic management of an adolescent with Dandy Walker Syndrome. Korean journal of anesthesia 2013 february64(2): 180-181
3. Kim SS, Kwak S, Kim YK. An Anesthetic Experience In a Patient with Dandy Walker Malformation: Case Report. Anesth Pain Med 2008 Apr;3(2):139-144.