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Case Report on: Dandy Walker Malformation Syndrome

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Introduction: Dandy Walker Syndrome is a congenital brain malformation that affects the cerebellum and the fluid filled areas that accompany it. The cerebellum is a part of the brain that deals with movement coordination as well as cognition and behavior.
Case Presentation: The male patient 8 year old who was apparently admitted in A.V.B.R.H on 18/8/2019 is diagnosed with a known case of dandy walker malformation and came with the complaints of headache since 4 days, fever since 3 days, vomiting since 5 days and 3 episodes of seizures. All the necessary blood investigations has been done. The CT scan as well as MRI reports has shown that large posterior fossa cyst with open communication with fourth ventricle with hypoplastic left cerebellar hemisphere and no visualized cerebellar vermis. Before taking this case, information was given to the patients and their relatives and Informed consent was obtained from patient as well as relatives.

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Therapeutic Intervention: The patient was treated with NSAIDS, antibiotics, anticonvulsant, antiemetic and antacid and physical therapy to help keep muscle strength and flexibility The patient underwent VP shunt head circumference assessed regularly, vital signs hourly, provided vitamins enriched diet as per dietician's order assisted dressing for drainage and physical therapy. **Conclusion:** The patient was admitted in A.V.B.R.H in a critical condition with the diagnosis of dandy walker syndrome, after providing the required treatment by the health care team members of hospital the patient condition was improved and satisfactory.

Keywords: Dandy walker malformation; ventricles; syndrome; cerebellar vermin hydrocephalus; shunting; cystic enlargement.

1. INTRODUCTION

The malformation of DandyWalker is a congenital (present at birth) disorder that affects the cerebellum, the back section of the brain that regulates speech, actions and cognitive abilities. Dandy Walker may cause blockage of normal cerebrospinal fluid drainage (CSF), resulting in CSF buildup and hydrocephalus. Today researchers do not fully understand the causes and forms of the classic DandyWalker malformation. Chromosomal defects which affect the development of fetal brain some mother's viral infections that transfer to the developing unborn infant's susceptibility to other toxins. Dandy Walker malformation signs usually turn up by age 1year [1].

Those symptoms could include: Developmental motor and language skills delays, such as sitting, Low muscle tone, flexibility and posture Problems with eye movement, particularly jerky eye motion hearing loss, hydrocephalus complicates the majority of Dandy-Walker malformations. Symptoms of hydrocephalus include: Abnormally rapid head growth, Irritability, Vomiting, sleepiness. The cause of this malformation is based on the finding that the development of the fetal rhombencephalon cap is defective, along with a substantial or complete lack of patentability Magendie's foramen [2].

DandyWalker malformation has complex impact on mental development. Many children have normal comprehension, and some never reach proper mental growth, even though excess CSF is removed early [3].

Incidence: Dandy Walker malformation is a by birth deformity of the posterior cranial fossa and the high occurrence of Dandy–Walker syndrome (DWS) is 1:25,000–1:35,000 live births. The incidence of DWS was 1.0 per 100,000 live births per year. The syndrome can appear dramatically or develop unnoticed [4].

1.1 Objective

- To determine the general idea regarding disease condition.
- To develop knowledge regarding medical, surgical and nursing management.

2. PATIENT INFORMATION

Patient present history: The male patient 8 year old who was apparently admitted in A.V.B.R.H on 18/8/2019 is diagnosed with a known case of dandy walker malformation and came with the complaints of headache since 4 days, fever since 3 days, vomiting since 5 days and 3 episodes of seizures. After undergoing investigation he was diagnosed as Dandy walker malformation.

Past history: At the time of admission the history was taken patients was apparently alright 6 days back then he started high grade fever, and was intermittent relieved on taking medication. Patient also started complaining of vomiting and episodes of seizures for which he has been taking ayurvedic medicines also. Patients were taken to private practitioner where he was given some treatment but there was no any relief.

Etiology: DWM is the product of abnormalities in early embryonic cerebellum and associated structures. A few patients suffer chromosome abnormalities including chromosome deletion, or chromosome replication. This may be attributed to other more nuanced genetic and environmental factors such as teratogens, such as the chance of recurrence in siblings below 5%. The risk of recurrence is higher in these families, up to 25 per cent [5].

Clinical Findings: The patient demonstrates on physical examination the signs and symptoms of conditions that frequently arise in early childhood, including sluggish motor growth and

gradual skull enlargement.In older children there may be symptoms of increased intracranial press ure such as irritability and vomiting, as well as sig ns of cerebellar dysfunction such as instability, la ck of muscle coordination, or jerky eye movemen ts. Many signs include raised head size, bumping at the back of the brain, eyecontrol nerve issues, facial and neck issues, and erratic breathing habi ts [6].

Diagnostic Assessment: Different findings are discovered on medical tests and patient records and other examinations and the diagnosis varies based on the degree of abnormalities. Although unspecific, the early signs of DW couldbe observed in the newborns including macrocephaly, skull enlargement in the occipital area ultrasound in the brain, CT scan or brain MRI is used to diagnose complex Dandy-Walker [7].

MRI And CT SCAN Reports: Big posterior fossa cyst with clear contact with the fourth ventricle, with hypoplastic left cerebellar hemisphere and nonvisualized cerebellar vermis s / o dandy walk er mild to severe hydrocephalus malformation.

Medical Management:

- Antibiotics: culture based, and resistance t o infection shunting For eg, septicemia, ve ntriculitis, meningitis prescribed as prophyl axis
- Anticonvalsants**:** Helps to control the impulse transmission of cerebral cortex and prevent seizures.
- Diuretics**:** Acetazolamide (ACZ) and furosemide (FUR) treat neonates with posthemorrhagic hydrocephalus; all are diuretics that often tend to decrease CSF secretion at the choroid plexus level.[8]
- Oral steroids, Antacids and local anesthetic etc.
- Physical therapies to improve muscle tone and coordination [9]

Table 1. Etiological factors

S. no	Book Picture	Patient picture
1.	**Genetic factors-** There is strong evidence for a genetic predisposition.	**Yes**
2.	**chromosome abnormalities**	**Yes**
3.	**Environmental factors** • Teratogens are affected like Warfarin use during pregnancy have developmental anomalies including eye dysgenesis, microcephalus, corpus callosum agenesis, structural abnormalities and heart defects. It was also linked • viral infection, and maternal diabetes: maternal virus infections (rubella, toxoplasma, and cytomegalovirus) and alcohol consumption.	**No**

Table 2. Patient blood investigation report

S. no	Investigation	Normal Value	Patient value	Remark
Blood Investigation				
1.	Hemoglobin	11-15(Fe) 12-16(male) gm/dl	14.2 gm/dl	Normal
2.	Neutrophils	40-80%	700%	Normal
3.	Lymphocytes	20-80%	55%	Normal
4.	Monocytes	2-10%	1%	Decrease
5.	Eosinophils	1-6%	02%	Normal
6.	Basophils	<1-2%	1%	Normal
7.	WBC	4,500 to 11,000 per microliter	10800/cu.mm	Normal
8.	TLC	4.5-11.0 x 10^9 /L	4700/l	Normal
9.	Platelets	1,50,000-4,00,000 cells/cumm	2.85lacs/cumm	Normal
10.	HCT	45% to 52%	35.5gm%,	Decrease
11.	MCV	76-100 fl.	80 cub micron	Normal
12.	MCH	27.5–33.2 pg	28.6 pico gm	Normal
13.	MCHC	32-35 %	32.6%	Normal
14.	CSF	**CSF** total protein: 15 to 60 mg/100 mL	300mg/dl	Increased

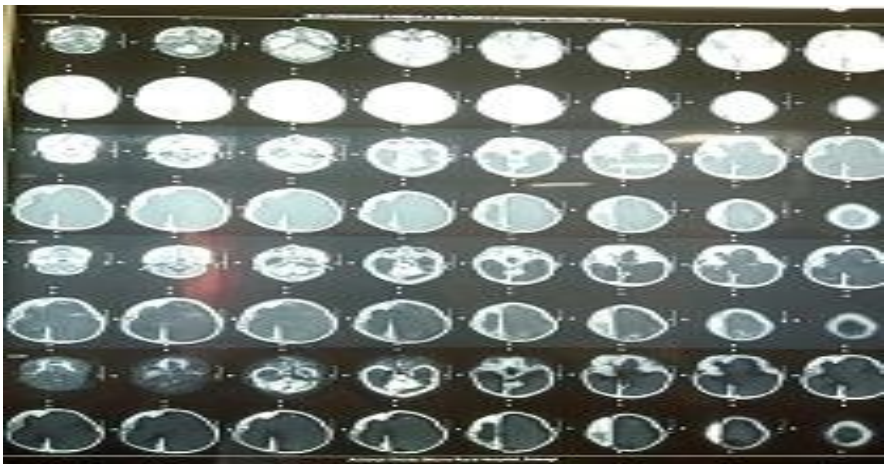

Fig. 1 A. Big posterior fossa cyst with clear contact with the fourth ventricle, with hypoplastic left cerebellar hemisphere and nonvisualized cerebellar vermis s

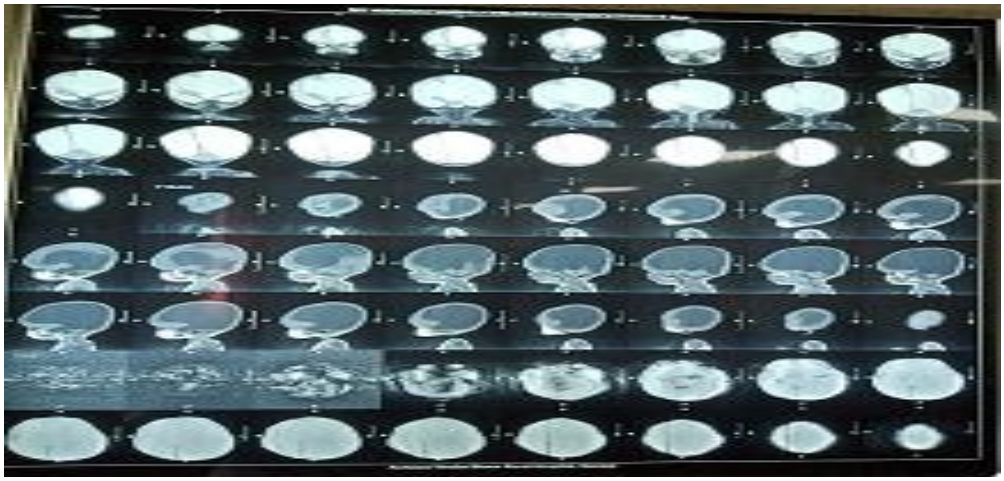

Fig. 1B. O dandy walker mild to severe hydrocephalus malformation

Table 3. Complication of patients compare with book pictures

S. no	Book picture	Patient Picture
1.	Hydrocephalus	Present
2.	Spastic paraplagia	Absent
3.	Occipital encephalocele	Absent
4.	Ataxia	Present
5.	Visual impairment	Absent

3. THERAPEUTIC INTERVENTION

Nursing Management:

- Measurement of the head of the newborn is essential. • Vital signs and neurological signs. Precise critical and neurological signs are to be collected before and after surgery. •Check fontanels. When the fontanels are not locked, then check them closely for any signs of bulging.
- **Preventing injury.**
- Promoting safe skin
- To avoid infection. **.**
- To foster growth and development. The inf ant wants social contact and the potential f or

Movement wants to be spoken to, interacted with, and offered

- Reducing anxiety within the family. Explain to the family in terms they can understand the state and anatomy of the surgical proc edure
- Physical therapy: Improve muscle tone and coordination
- Providing instruction within the home**.** Dem onstrate shunt treatment to family member s and help them conduct a return demonstr ation; provide them with a list of signs and effects that need to be identified, and addr ess the child's reasonable growth and deve lopment goals, and emphasize practical tar gets [10-12].

Complication: The following are the complications may present in the child with dandy walker malformation.

Follow up and outcomes: Inspite of all the care the patients progress was satisfactory. Parents was advised about the drug therapy and personal hygiene and also physical therapy for improving muscle tone and flexibility. The parents instructed about drug regimen and advised to come for follow up after 10 days for routine assessment to see the disease outcome.

4. DISCUSSION

The prenatal diagnosis of DandyWalker syndrom e, usually achieved after 18 weeks of gestation, i s particularly significant.The life and wellbeing pr ognosis relies on the occurrence of concomitant genetic defects, chromosomal abnormalities and medical timeliness. The present study had big posterior fossa cystwith clear contact with the fourth ventricle, with hypoplastic left cerebella r hemisphere and nonvisualized cerebellar vermi s. In a similar study by Paladini and Volpe mentioned in their study in 2006 that the degree of vermian hypoplasia is substantially related to the prevalence and severity of mental impairment. Thus, it appears that the more aberrant the vermis, the worse the prognosis, and our patient showed just moderate hypoplasia of the cerebellar vermis, indicating a milder presentation. Today, a variety of neuroimaging methods are utilized to identify and confirm diagnoses in fetuses, newborns, and anybody else who may be suspected of having Dandy Walker Malformation.

5. CONCLUSION

Patient was admitted to hospital with the chief complaint headache since 4 days, fever since 3 days, vomiting since 5 days and 3 episodes of seizures immediate treatment was started by health team member now the patient condition was satisfactory. Neurologists and neurosurgeons have considerable expertise in diagnosing and treating malformations caused by Dandy Walker. Members of the health team shou ld develop plans which ensure continuity of care f rom the early stages of pregnancy, after birth, an d throughout childhood.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author.

CONSENT

As per international standard, parental written consent has been collected and preserved by the author(s).

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

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