Case Report

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Case Report On: Dandy Walker Malformation as Congenital Defect

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Abstract

Introduction: The Dandy Walker Malformation is a congenital defect affecting the cerebellum, the back part of the brain that controls movement, behaviour and cognitive ability. The central part of the cerebellum (the vermis) is absent or very small and may be abnormally positioned. The incidence rate is approximately 1 per 25,000 – 35,000 live births and the male and female ratio is 1:3.

Patient history: Master Rupesh aged 8 years was admitted in A.V.B.R.H. on 18/08/2019 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 for which he has been taking ayurvedic medication.

Clinical finding: The patient has undergone various kind of blood test, CSF examination, CT scan, EEG record and MRI brain. The CBC reports were found as Hb% - 14.2 gm%, RBC – 5.23 M/cu. mm, WBC- 10800/cu.mm, platelets - 2.85 lacs/cu.mm; whereas the CSF reports were found with increased protein-CSF,i.e. 300mg/dl and the CT scan as well as MRI reports has shown that a large posterior fossa cyst with open communication with fourth ventricle with hypoplastic left cerebellar hemisphere and non visualized cerebellar vermis S/O dandy walker malformation and mild to moderate hydrocephalus.

Pharmacology: The patient was treated with NSAIDs, anti-biotics, anticonvulsant, antiemetic and antacid.

Surgical Management: The patient underwent V.P. shunt.

Nursing management: Checked for head circumference regularly, vital signs hourly, provided vitamins enriched diet as per dietician's order and assisted dressing for drainage.

Conclusion: The patient was admitted in A.V.B.R.H. in a critical condition with the chief complaints of headache since 4 days, fever since 3 days, vomiting since 5 days, 3 episodes of seizures, poor muscle tone and ataxia due to developmental delay, but after providing the required treatment by the health care team members of A.V.B.R.H., the patient's condition was improved and satisfactory.

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INTRODUCTION

Dandy Walker syndrome is the rare congenital brain malformation involving the cerebellum

(an area in the back of the brain which controls the movement) and the fluid filled spaces around it. The key feature of this syndrome is an enlargement of fourth ventricle (a small channel allow the fluid to flow between upper and lower areas of brain and spinal cord). A partial or complete absence of cerebellar vermis (the area between the two cerebellar hemisphere), a cyst formation near the internal base of skull. An increase in the size of fluid spaces surrounding the brain as well as increase the pressure may also be present. The syndrome appear dramatically or develop unnoticed.

Incidence

The incidence rate of Dandy Walker Syndrome was 1.0 per 100,000 live births per year. The incidence by sex per 100,000 live births per year was 1.24 for males and 0.78 for females. Dandy Walker Syndromeformed 3.5% of our cases of infantile hydrocephalus.

OBJECTIVE

1. To know general idea regarding disease condition.
2. To explore knowledge regarding pharmacology, medical and nursing management.

Patient Information

Patient history: Master Rupesh, aged 8 years was admitted in A.V.B.R.H. on 18/08/2019 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 for which he has been taking ayurvedic medication. The patient has undergone various kind of blood test, CSF examination, CT scan, EEG record and MRI brain. The CBC reports were found as Hb% - 14.2gm%, RBC – 5.23 M/ cu.mm, WBC - 10800/cu.mm, platelets - 2.85 lacs/ cu.mm; whereas the CSF reports were found with increased protein-CSF, i.e. 300mg/dl and the CT scan as well as MRI reports has shown that a large posterior fossa cyst with open communication with fourth ventricle with hypo plastic left cerebellar hemisphere and non visualized cerebellar vermis- S/O dandy walker malformation and mild to moderate hydrocephalus.

Past history: He was diagnosed with dandy walker malformation five years back.

Causes

In most cases, the exact cause of Dandy-Walker complex is unknown. It is thought that most cases are caused by a combination of genetic and environmental factors that affect early development before birth. In some cases, exposures that occur during pregnancy, such as a pregnant woman having diabetes or an infection such as rubella, are thought to cause an increased risk for the developing baby to have Dandy-Walker complex. In some cases, Dandy-Walker complex is caused by an underlying genetic change. For example, some people with Dandy-Walker complex have extra or missing pieces of chromosomes in each cell of the body (chromosome abnormalities). Dandy-Walker complex can also occur as a symptom of another genetic syndrome. For example, people with Dandy-Walker complex may have a change in a gene that causes them to develop Dandy-Walker complex as well as other health problems.

Classification of complete heart block

- Dandy walker malformation (cystic dilation of the 4th ventricle, partial or complete agenesis of cerebellar vermis and an enlarge posterior fosse.
- Dandy walker variant (cystic posterior fossa mass with variable hypoplasia of cerebellar vermis and no enlargement of posterior fossa).
- Mega cistern magna (enlarge cistena magna with normal cerebellar vermis).

Clinical Finding

- Developmental delays in motor and language skills such as sitting up, walking, and talking.
- Poor muscle tone, balance, and coordination.
- Problems with eye movement, mainly jerky eye movement.
- Vision and hearing impairment.
- Seizures.

Diagnosis evaluation

1. **History collection**: Done (history of Hydrocephalus and V.P. Shunt).
2. **Physical examination:** Done (poor muscle tone and ataxia due to developmental delay).
3. **CBC test:** Done
4. **ECG:** Done

5. **CSF Study:** Report were found with increased protein
6. **CT scan:** Done
7. **MRI scan:** Its hows that a large posterior fossa cyst with open communication with fourth ventricle with hypoplastic left cerebellar hemisphere and non visualized cerebellar vermis

Blood Investigation Report

Investigation	Normal Value	Patient Value	Justification
Biochemistry			
Kidney function test			
Urea serum	25 mg%	18-40 mg%	Normal
Creatine serum	0.96 mg%	0.7-1.5 mg%	Normal
Sodium serum	142 meg/l	142-136 meg/l	Normal
RBS	172 mg%	70-150 mg%	Increase
Potassium	4.8 meg/g	3-5 meg/l	Normal
Pathology complete blood count			
HB%	14.2 gm%	13-15.5 gm%	Normal
MCV	77 cub.micro	80-90 cub	Decrease
MCH	22.3 pico.gm	26.5 - 33.5 picogm	Decrease
MCHC	33.8%	30.36. 5%	Normal
RBS count	4.13 millioncumm	4.5-6 million cumm	Normal
RDW	12.3%	10-15%	Normal
HCT	36.5%	40.50%	Increase
WBC count	5000-10000/cumm	6000-10000 cumm	Normal
Total platelet count	150000-450000/micro liter	420000/micro liter	Normal
Monocytes	02%	06%	Increase
Granulocytes	60%	40-60%	Normal
Lymphocytes	36%	17-48%	Normal
Esosinophils	0.2%	0.5%	Increase

Treatment

Pharmacologic Therapy

- NSAIDs
- Antibiotic
- Anticonvulsant
- Antiemetic
- Antacid

Medical Management: As follow by:

- Tablet - Atorin 10mg
- Tablet - Alprax 0.25mg
- Tablet - Telma 20mg
- Tablet - Chymoral forte 1mg
- Injection - Ceftrazone 1gm
- Injection - Amikacin 500mg-2ml

General Measures for the management of Dandy Walker Malformation

- Explanation of nature of disease, treatment and self help strategies
- Good general nutrition
- Speech therapy to help with speech and language
- Physical therapy to improve the muscle tone and coordination
- Surgical insertion of ventriculoperitonial shunt in case of severe or worsening hydrocephalus
- Occupation al therapy to help build the self car and mobility skills such as eating, getting dressed and walking
- Special education as necessary for cognitive and learning problems

Surgical Management

- Ventriculoperitonial shunt
- Cystoperitonial shunt

Nursing Management: The nurse is responsible for administering the medications and for assessing their beneficial and detrimental effects to the patient. It is the balance of these effects that determines the type and dosage of pharmacologic therapy. Nursing actions to evaluate therapeutic effectiveness include the following:

- Keeping an intake and output record to identify a negative balance (more output than input).
- Weighing the patient daily at the same time and on the same scale.
- Physical examination to check muscle tone, mobility, head circumference *etc*.
- Monitor head circumference and vital signs.
- Provide vitamins rich diet as per dietician.
- Assisted dressing for drainage.

Monitoring and Managing Potential Complications: Profuse and repeated diuresis can lead to hypokalaemia (ie, potassium depletion). Signs are weak pulse, faint heart sounds, hypotension, muscle flabbiness, diminished deep tendon reflexes, and generalized weakness. Hypokalaemia poses new problems for the patient with HF because it markedly weakens cardiac contractions. In patients receiving digoxin, hypokalemia can lead to digitalis toxicity. Digitalis toxicity and hypokalemia increase the likelihood of dangerous dysrhythmias (see Chart 30-3). Low levels of potassium may also indicate a low level of magnesium, which can add to the risk for dysrhythmias. Hyperkalaemia may also occur, especially with the use of ACE-Is or ARBs and spironolactone.

Nursing Diagnoses: Based on the assessment data, major nursing diagnoses for the patient with HF may include the following:

- Activity intolerance (or risk for activity intolerance) related to imbalance between oxygen supply and demand because of decreased CO
- Excess fluid volume related to excess fluid or sodium intake and retention of fluid because of HF and its medical therapy
- Anxiety related to breathlessness and restlessness from inadequate oxygenation
- Powerlessness related to inability to perform role responsibilities because of chronic illness and hospitalizations.
- Non compliance related to lack of knowledge.[5]

Collaborative Problems/ Potential Complications Based on the assessment data, potential complications that may develop include the following:

- Cardiogenic shock
- Dysrhythmias
- Thromboembolism
- Pericardial effusion and cardiac tamponade

Continuing Care: Depending on the patient's physical status and the availability of family assistance, a home care referral may be indicated for a patient who has been hospitalized. Elderly patients and those who have long-standing heart disease with compromised physical stamina often require assistance with the transition to home after hospitalization for an acute episode of HF. It is important for the home care nurse to assess the physical environment of the home. Suggestions for adapting the home environment to meet the patient's activity limitations are important. If stairs are the concern, the patient can plan the day's activities so that of stairs climbing is minimized; for some patients, a temporary bedroom may be set up on the main level of the home. The home care nurse collaborates with the patient and family to maximize the benefits of these changes. The home care nurse also reinforces and clarifies information about dietary changes and fluid restrictions, the need to monitor symptoms and daily body weights, and the importance of obtaining follow-up health care. Assistance may be given in scheduling and keeping appointments as well. The patient is encouraged to gradually increase his or her self-care and responsibility for accomplishing the therapeutic regimen.[5]

Evaluation

Expected Patient Outcomes

Expected patient outcomes may include:

1. **Demonstrates tolerance for increased activity**
   a. Describes adaptive methods for usual activities.
   b. Stops any activity that causes symptoms of intolerance.

c. Maintains vital signs (pulse, blood pressure, respiratory rate, and pulse oximetry) within the targeted range

d. Identifies factors that contribute to activity intolerance and takes actions to avoid them

e. Establishes priorities for activities

f. Schedules activities to conserve energy and to reduce fatigue and dyspnea

2. Maintains fluid balance

a. Exhibits decreased peripheral and sacral oedema

b. Demonstrates methods for preventing oedema

3. Is less anxious

a. Avoids situations that produce stress

b. Sleeps comfortably at night

c. Reports decreased stress and anxiety

4. Makes decisions regarding care and treatment

a. States ability to influence outcomes

5. Adheres to self-care regimen

a. Performs and records daily weights

b. Ensures dietary intake includes no more than 2 to 3 g of sodium per day.[5]

DISCUSSION

CHB occurs when auricular and ventricular contractions are not communicated to each other beating at their own paces to result in a negative effect in cardiac function. CHB may occur in AV node, intra-Hisian, or infra-Hisian sites. Intranodal or intra-Hisian blocks almost always feature escape rhythms with narrow QRS complexes in the mean while infra-Hisian block often presents with wide QRS complex escapes.

Strength: Patient was 71 year male patient tolerate all the medication and well response within seven days to the therapeutic treatment of the hospital which was given as a treatment.

Informed Consent

Before taking this case, information was given to the patients and their relatives and Informed consent was obtained from patient as well as relatives.

CONCLUSION

Treatment of asymptomatic CHB with narrow complex escape rhythm is challenging. Those patients are often very young, and implanting a permanent PM is not always an easy decision. The likelihood of renewing multiple generators, potential of developing infections, and vascular complications sometimes outweigh the benefits of early intervention, and the ideal time for implanting a PM in those patients still remains a subject for further investigation. Nevertheless, the select group will benefit from close follow-ups, annual echocardiography, and rhythm monitoring by a loop recorder when they opt for a conservative approach without PM therapy.

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