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REVIEW ARTICLE
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"HYDROCEPHALUS: CAUSES, PATHOPHYSIOLOGY, DIAGNOSIS, AND TREATMENT APPROACHES-A REVIEW"

Muskan Parween[1], Sageer Ahmad[2], Rajesh Kumari Patil[3*], Hanumanthrao C Patil[4]

[1] PharmD (Student), Adesh Institute of Pharmacy and Biomedical Sciences, Adesh University, Bathinda
[2] Assistant Professor, Adesh Institute of Pharmacy and Biomedical Sciences, Adesh University, Bathinda
[3] *Professor & HoD, Department of Pharmacy Practice, Adesh Institute of Pharmacy and Biomedical Sciences, Adesh University, Bathinda
[4] Professor & HoD, Department of Pharmacology, Akal College of Pharmacy, Mastuana, Sangrur

[*]**Corresponding Author:** Rajesh Kumari Patil
*Professor & HoD, Department of Pharmacy Practice, Adesh Institute of Pharmacy and Biomedical sciences, Adesh University, Bathinda[5] Email: rkpatil3014@gmail.com

Abstract :-
Hydrocephalus, commonly known as "water on the brain," is a neurological disorder characterized by the abnormal accumulation of cerebrospinal fluid (CSF), leading to brain enlargement. This condition can result from various factors, including age, genetic predisposition, infections, and other underlying causes. Hydrocephalus is classified into two main types: communicating hydrocephalus, where CSF flow is blocked after leaving the ventricles, and non-communicating hydrocephalus, where the obstruction occurs within the ventricular system. Diagnosis typically involves imaging techniques such as MRI, ultrasound, and CT scans. Treatment options include medications like diuretics to manage fluid buildup and surgical interventions such as shunt placement or endoscopic procedures to restore normal CSF flow.

Keywords: - Hydrocephalus, CSF, ICP, Communicating and Non- Communicating, Brain enlargement, shunting.

Introduction :-

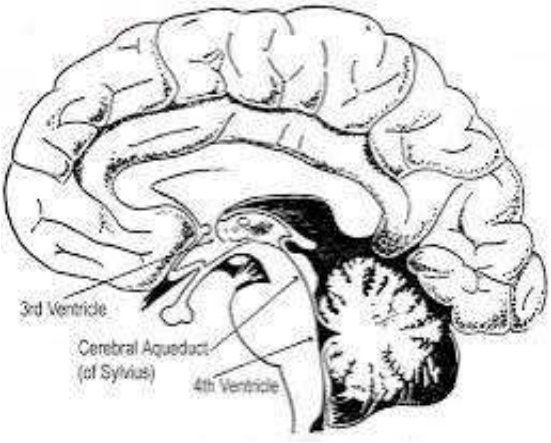
It is a chronic neurological disorder also called water in the Brain it can be fatal if left untreated also brought about blindness, spasticity, affect child growth and development, mental retardation and death. it is characterized by brain enlargement or increased intracranial pressure [ICP] due to access accumulation of [CSF] cerebral spinal fluid inside the cerebral ventricles. CSF is watery fluid which fills the space between the arachnoids membrane and the pia matter the normal functioning of CSF is assists brain in protecting, nourishing, and removal of wastage. Hydrocephalus is mainly occurs due to overproduction of CSF fluid in the brain premature children typically exhibit progressive macrocephaly, while older children typically display signs and symptoms of cerebral hypertension if we see epidemiology then most of the cases registered due to neonatal infection like 1 in every 500 babies in US born with hydrocephalus called congenital hydrocephalus a complicated interplay between genetic and environmental variables results in hydrocephalus. Prevalent cause is constriction of aqueduct. Another common cause is chiari malformations [in combination with spinia Bifida] or

blockage of the fourth ventricle exit [Dandly walker syndrome] congenital hydrocephalus present with

In Infants: high pitch cry, Big head, poor feeding, excessive sleepiness, vomiting, difficulty in eye movement, fontanelles.

In Older child: Lethargy, severe headache, Irritability, problems with balance, trouble walking or standing, bladder control loss, extreme fatigue, problems with coordination, seizures, trouble to remember, trouble in focus, unexpected shifts in temperament etc.

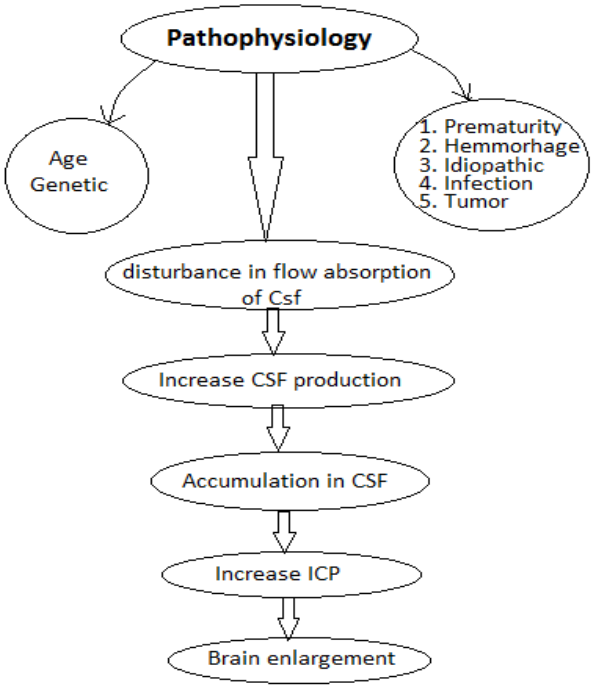
PATHOPHYSIOLOGY:-



Numerous developmental problems or shocks can result in congenital or neonatal hydrocephalus; the most common causes include neural tube anomalies, infections, intraventricular hemmorrhage, trauma, and tumours. This disorder is particularly harmful to children because the growing ventricles and rising CSF pressure force the flexible skull to grow, which in turn compresses and stretches the surrounding brain tissue.

OR

- Age , Genetic
- Idiopathic, tumor, infection, prematurity , hemorrhage
- Can cause disturbances in flow or absorption of CSF leads o increase CSF production CSF get accumulated then increase intra cranial pressure results enlargement of brain



Aetiology:- Congenital hydrocephalus has a highly diverse aetiology. Most are secondary types caused by wounds or trauma, teratogens, cerebral hemorrhages ,tumors and intrauterine infections ,Neural tube abnormalities can also cause congenital hydrocephalus, which is frequently connected with other abnormalities of central nervous system. There are 0.3–0.9 cases of 1° congenital hydrocephalus for every thousands live births . Of patients with congenital hydrocephalus, roughly 35% have a potential genetic cause. This covers multifactorial traits, monogenically or complicated hereditary ailments, including abnormalities results from cytogenetics. The cause of 1° congenital hydrocephalus is contemporary unknown in though.

Classification :

It can be classified into two main categories

1. Non- communicating [Obstructive]

a. Acquired hydrocephalus

b. congenital hydrocephalus

2. Communicating [non- obstructive]

a. Hydrocephalus ex vacuo

b. Normal Pressure Hydrocephalus.

- so, [Birth defects] congenital hydrocephalus comes in non- communicating hydrocephalus so let's discuss it.

Obstructive hydrocephalus, or non-communicating hydrocephalus, is a condition in which 1 or more of the small channels that connect the ventricles are impeded, preventing the flow of CSF. Aqueductal stenosis, or narrowing of aqueduct of Sylvius, tiny channel connecting brain's 3rd and 4th ventricles, is one of the most frequent causes.

Sign and symptoms

The signs and symptoms of hydrocephalus can fluctuate significantly between age groups and individuals. Increased [ICP] intracranial pressure can cause symptoms such as vomiting in infants and early children, and in adults, it can lead to loss of normal functions such as walking or thinking, problem with coordination, hypersomnia, seizures, severe cephalagia, sweating , loss of bladder control, trouble remembering and focusing, irritability and head symptoms include rapid head growth.

Risk factors :-

1. One of the most common side effects of pediatric brain tumors is hydrocephalus, which can remain in 10–40% of cases following surgical resection and is present in roughly 50% of cases at the time of tumor diagnosis. This is a retrospective study conducted at a single institution to identify the factors that may indicate whether pediatric patients with brain tumors may require therapy for persistent hydrocephalus. Techniques: Case note retrospectively.
2. Tina Noergaard Munch 2013 Future studies should focus on the increased incidence of single CHC in first term infants as well as behavioural problems and comorbidities related to using antidepressant by mothers. It is yet unclear how antidepressants could cause hydrocephalus through potential biochemical routes
3. .Melese Shenkut Abebe 2022 Congenital hydrocephalus has been associated with the development of several risk factors for the child and the mother. Pregnancy-related factors increase the risk of hydrocephalus include maternal diabetes, oligohydramnious, gestational hypertension, consanguinity, lack of parental care, drug use during pregnancy, and multiple pregnancy.

4.T.T Kitova 2020 The risk of Congenital hydrocephalus and related hydrocephalus is increased when the mother's age is above 40 years and consanguinity is present..

5.Girma seyoum 2022 Multivariate logistic analysis showed a significant association between congenital hydrocephalus and alcohol consumption and iron and folic acid supplementation during pregnancy..

6. Stephanie Walsh 2017 Lack of prenatal care, multiparous gestation, gestational diabetes, prolonged hypertension in mothers, hypertension in mothers throughout pregnancy, and alcohol consumption in mothers were amidst the risk variables found.
7. Based on the limited information that is currently available, it is possible that modifiable risk factors such as maternal obesity, not using prenatal multivitamin supplements, and high HDL cholesterol levels in adults are associated with both congenital and acquired hydrocephalus.
8. Bin Chen, MD 2021 In children diagnosed with bacterial meningitis, the most important indicators of hydrocephalus are severe clinical presentation and a significant laboratory index at admission.
9. Among the risk variables for hydrocephalus in neonatal purulent meningitis, female sex, CSF glucose <2 mmol/L, punctate white matter lesions and congenital hydrocephalus affected 2.68 per 1,000 pregnancies. found that pyogenic intraventricular empyema has significant consequences. When planning a pregnancy, these factors must be taken into account to avoid developing of congenital hydrocephalus in fetus.
10. TORCH [congenital toxoplasmosis] infection in the fetus, living near a highway, unregistered marriage, chemical exposure to both sexes, paternal pregnancy before conception, smoking, and military service are all risk factors for congenital hydrocephalus. A higher level of parental education and the use of folic acid in the first trimester of pregnancy are probably protective factors against congenital hydrocephalus in the offspring. When planning a pregnancy, these factors must be taken into account to avoid the development of congenital hydrocephalus in fetus.

Diagnosis of Hydrocephalus

1 . The diagnosis of hydrocephalus is primarily as per clinical presentations like :-

a. Enlargement of brain
b. Severe Headache
c. Loss of coordination
d. Impaired vision
e. Nausea and vomiting etc.

2. Neurological examination

Age of patient determines neurological test that is most appropriate. Assessing a patient's muscular health, mobility, general health, and sensory function is as easy as asking a few questions and doing a few tests.

3.Brain imaging

a. MRI

When it comes to identifying hydrocephalus in all its forms and manifestations, MRI is gold standard. However, axial plane CT imaging is enough for elucidating the explanation for the finding. Interstitial edema, also known as transspendymal CSF flow and hyperdynamic CSF flow in shunt-responsive normal pressure hydrocephalus (NPH), may be better detected using MRI than CT scan. MRI scans may detect severe white matter ischemia, that might play a role in the idiopathic kind of NPH.

b. Ultrasound

Due to its simplicity and lack of danger, this test can be given to infants as their first screening. A sensitive area on baby's skull is used to position ultrasound machine. During standard prenatal checkups, ultrasound may identify hydrocephalus.

C. CT Scan

A cross-section of the brain may be obtained using this specialized X-ray method. Scanning takes very little time and has little side effects. A little sedative is often administered to the youngster before this test since it also needs the child to remain calm. An MRI scan reveals more information than a CT scan. Plus, there is a little quantity of radiation that CT scans expose patients to. Emergency investigations are the only ones that often employ CT scans for hydrocephalus.

1. A lumbar puncture is a technique that involves inserting a needle into lower back to draw cerebrospinal fluid. In the next step, pressure in cerebrospinal fluid sample is assessed.

2. Lumbar infusion test

Lumbar infusion test involves gradually injecting fluid in lower back while monitoring pressure.

3. Cranial ultrasonography

Cranial ultrasound remains an important tool in the evaluation of hydrocephalus, periventricular leukomalacia (PVL, also known as hypoxic ischemic encephalopathy of prematurity), and intracranial hemorrhage.The most common reason for cranial ultrasound is to look for hemorrhage. Screening for prematurity, germinal hemorrhage, intracranial hemorrhage.

X- ray skull Radiography

Treatment of Hydrocephalus

Treatment Type	Description	Indications	Considerations
LCR Shunting (VP Shunt)	A surgical procedure to drain excess CSF from the brain to the abdominal cavity, helping to relieve increased ICP.	Standard treatment for hydrocephalus	Complications include infection, shunt malfunction, and need for revisions.
Endoscopic Third Ventriculostomy (ETV)	Triflingly hostile process to create a pathway for CSF flow, used primarily in non-communicating hydrocephalus.	Alternative for certain cases	Not suitable for all patients; requires specific anatomy.
Drug Therapy	Medications to manage CSF dynamics temporarily; includes:	Temporary management	Not effective for long-term treatment; used before surgery.
	- **Acetazolamide**: Reduces CSF production.		Can cause side effects.
	- **Isosorbide/Furosemide**: Increases CSF resorption.		Less common; used for specific cases.
	- **Osmotic Diuretics**: Increase water secretion to lower ICP.		Used as temporary measures.
Anti-inflammatory Agents	Used experimentally to prevent hydrocephalus post-infection or hemorrhage.	After meningitis or hemorrhage	Mixed results on effectiveness; may reduce other complications.
Calcium Channel Blockers (e.g., Nimodipine)	Experimental use to reduce cognitive decline associated with hydrocephalus.	Cognitive decline	No definitive evidence for effectiveness in hydrocephalus.
Medications for Neuroprotection	Agents like bifemelan and bromocriptine have shown benefits in specific cases of hydrocephalus.	Certain cases of hydrocephalus	Limited evidence; requires further research.
Observation	Monitoring the condition without immediate intervention for mild cases.	Asymptomatic or mild cases	Regular follow-up needed; potential for progression.

Surgical Treatment Procedure (Shunting)	
Procedure	Details
Shunt Surgery	Involves implantation of a shunt for draining additional CSF to abdomen.
Components	A valve body and catheter; drains CSF from the brain to the abdominal cavity for absorption.
Risks	Infection, shunt failure, need for revisions; high rate of complications associated with VP shunts.
Outcomes	Generally improves neurological outcomes; requires lifelong management and monitoring.

In detail :-

Both surgical and non- surgical treatment can do in hydrocephalus but only surgical treatment can be efficient in treating hydrocephalus.

1. Treatment via medication

- While LCR shunts are the gold standard for hydrocephalus therapy, other medical interventions might be considered as an alternative or used in conjunction with shunts. The etiology of hydrocephalus dictates the course of treatment. To postpone surgical operations for hydrocephalus, pharmacological therapy is used. Although medical therapy is not helpful in treating chronic hydrocephalus in the long run, it may be maintained for the time being to balance the dynamics of

cerebrospinal fluid production and absorption. Some medications may decrease the production of CSF by the choroidal structures (acetazolamide), enhance the resorption of CSF, or reduce intracranial pressure by boosting the secretion of water.

- Carbonic anhydrases are metalloenzymes that facilitate the reversible hydration of bicarbonate and carbon dioxide. They are present in several organs and tissues, including the brain, stomach mucosa, pancreas, liver, lung, and renal cortex. This means that the concentrations of CO2, H, and HCO3 both within and outside of cells may be controlled. Additionally, the brain's secretory glia and choroid plexus contain these enzymes. In comparison to the choroid plexus halo, the enzyme concentration here is much greater. CSF production drops by 50% when choroid plexus carbonic anhydrase is completely inhibited. Restraining carbonic anhydrase reduces, according to several research.

- To balance nor epinephrine levels in the striatum and cortex, bifelan is utilized as a mild antidepressant and brain metabolic activator. It's a monoamine oxidase inhibitor. Following bypass, a patient with NPH was given 20 mg of methylphenidate, it inhibits the dopamine and nor epinephrine transporters. This helped with cognitive function and decreased apathy. Hydrocephalus and kinetic mutism were successfully treated with bromocriptine and ephedrine in yet another report. Taking trazodone helped a patient suffering severe hydrocephalus who was uncontrollable and hurting themselves.

- Inflammation of the CSF fluid is clearly associated with the progression of hydrocephalus. Experimental testing of anti-inflammatory drugs for prevention of hydrocephalus following meningitis and hemorrhage has been conducted. While the effects of corticosteroid treatment on hydrocephalus are poorly understood, many studies have shown that it considerably decreases hearing loss and narcoleptic squeal following acute bacterial meningitis. While some research indicates steroids may raise risk of hydrocephalus in children, other studies demonstrate no effect.

- As a calcium channel blocker, nimodipine is often used to manage hypertension. Research has demonstrated that nimodipine may mitigate the cognitive and motor impairments associated with hydrocephalus. Although nimodipine has been demonstrated to be safe in clinical tests, there is currently no conclusive proof that it effectively treats hydrocephalus. Additionally, magnesium, a calcium antagonist, has a less effective protective effect.

2. Treatment via surgery

- Hydrocephalus patients who do not respond to endoscopic third ventricle transfer may be considered for a further course of therapy that involves CSF shunting from ventricular system to the peritoneum. Surgical consequences are a major medical and social burden, yet they are essential for treating hydrocephalus. P atient may also need further surgeries and shunt revisions throughout the course of their lives due to shunt issues. A considerable amount of the morbidity linked to ventriculoperitoneal (VP) shunt surgery is caused by shunt problems, repairs, or failures, which may arise from either proximal or distal sources. S hunt revision or replacement is necessary for a very large number of individuals with hydrocephalus, even though neurosurgical shunting is remarkably successful owing to technique and therapy. Research has shown that VP shunt surgery is a viable option for people suffering from hydrocephalus, leading to better neurological results.

- **Procedure :-**

During shunt surgery, a thin tube called a shunt is implanted into the brain. Excess cerebrospinal fluid (CSF) in the brain is drained through a shunt to another part of the body, usually the abdomen. From there, it is absorbed into the blood .The shunt has an internal valve that controls the flow of cerebrospinal fluid so that it does not drain too quickly. Patient will feel the flap on the scalp like a lump under the skin. A cerebral shunt is a device that is permanently implanted in the head and body to remove excess fluid from the brain. They are often used to treat hydrocephalus, or swelling of the brain, caused by excessive accumulation of cerebrospinal fluid (CSF). If left unchecked, excess CSF

can cause increased intracranial pressure (ICP), which can lead to intracranial hematoma, brain edema, ruptured brain tissue, or herniation. The drainage provided by a shunt can alleviate or prevent these problems in patients with hydrocephalus or related conditions. Shunts come in various forms, but most consist of a valve body connected to a catheter, the lower end of which is usually placed inside. The abdominal cavity. The main differences between shunts are usually the materials they are made of, the type of valves used (if any) and whether or not the valve is programmable.

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