Hydrocephalus

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Abstract: Hydrocephalus is one of the commonest neurosurgical pathologies, and a neurosurgeon has to deal with it daily. It has a number of etiologies at any age. The congenital type of hydrocephalus is more common at pediatric age. The commonly applied surgical treatments for this condition are ventriculo-peritoneal (VP) shunts and endoscopic third ventriculostomy (ETV). Etiology, classification, clinical features, radiological findings, and surgical options are discussed in a focused form. A short description of the surgical procedures for hydrocephalus, including the installment of a ventriculo-peritoneal shunt, endoscopic third-ventriculostomy, and external ventricular drainage (EVD), is provided along with possible complications and strategies for complication management. In the later part of this chapter, normal-pressure (NPH) hydrocephalus is discussed.

Abbreviations

CSF	cerebrospinal fluid	CT	computed tomography
ETV	endoscopic third ventriculostomy	EVD	external ventricular drain
FH	frontal horn	HCP	Hydrocephalus
MRI	magnetic resonance imaging	NPH	normal pressure hydrocephalus
OFC	occipito-frontal circumference	TH	temporal horn

1. Introduction

Excessive collection of cerebrospinal fluid (CSF) within the ventricular system is called hydrocephalus (HCP). We know that the brain has a ventricular system, which contains CSF that supports the brain. Due to CSF, the weight of the brain is significantly reduced in the body in individuals with HCP.

Hydrocephalus can strike anyone at any age, but it is more frequent in infants and people over the age of sixty. Most of these instances are recognized before birth, at the time of parturition, or in early infancy (Figure 1) (Greenberg 2010).

Figure 1. A premature malnourished baby with hydrocephalus. Source: Photo by authors.

2. Etiology

CSF accumulates in the brain either through its excess production, an absorption defect pertaining to it, or its blockage in its pathway. We know CSF is secreted by the lateral ventricular choroid plexus and passes from one ventricle to another lateral ventricle through the foramen of Monro. From the lateral ventricle, CSF passes to the third ventricle and subsequently the fourth ventricle via the aqueduct of Sylvius. Hydrocephalus can be passed down through the offspring, linked to congenital anomalies like spinal dysraphism or encephalocele, or caused by cerebral tumors, head injuries, hematoma, or infections (meningitis). Hydrocephalus is classified into six categories depending on the onset, existence of structural problems, or high vs. normal CSF pressures.

3. Categories of HCP

1. Acquired HCP;
2. Congenital HCP;
3. Communicating HCP;
4. Non communicating HCP (obstructive);

5. Normal-pressure HCP, which is more common in the elderly and marked by enlarged ventricles with normal CSF pressure within the spine.
6. Hydrocephalus ex vacuo, which is common in adults and can occur when the brain is damaged by a degenerative condition, such as Alzheimer's disease, stroke, or head trauma, and in which the brain tissue shrinks.

4. Specific Etiologies of HCP in Pediatric Population

4.1. Congenital

(a) Chiari-2 malformation +/− myelomeningocele;
(b) Chiari-1 malformation;
(c) Primary cerebral aqueductal stenosis;
(d) Secondary cerebral aqueductal gliosis;
(e) Dandy–Walker malformation: Foramina of Luschka and Magendie atresia;
(f) X-linked inherited disorder: rare.

4.2. Acquired

(a) Infectious
- Post-meningitis (Figure 2);
- Cysticercosis.

(b) Post-
- Post-SAH;
- Post-intraventricular hemorrhage (IVH).

(c) Due to mass effect
- Non-neoplastic: such as arteriovenous malformation;
- Neoplastic lesions: e.g., medulloblastoma, colloid cyst of the third ventricle, pituitary tumor, and apoplexy with suprasellar extension (Kalangu et al. 2009).

(d) Postoperative;
(e) Neurosarcoidosis;
(f) Tuberculosis (Figure 3).

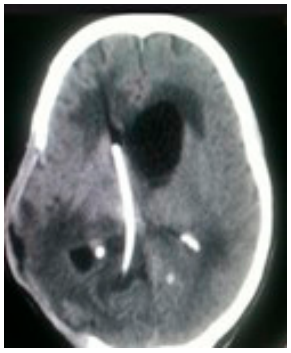

Figure 2. CT scan showing decompressive craniectomy with evidence of a VP shunt. Source: Figure by authors.

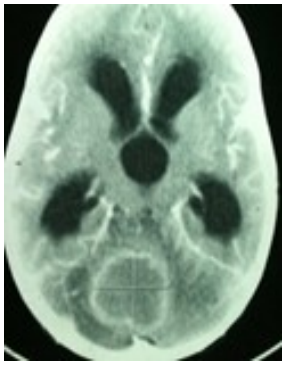

Figure 3. Posterior fossa tuberculoma with triventriculomegally. Source: Figure by authors.

Common etiologies of hydrocephalus are shown in Table 1.

Table 1. Common etiologies of hydrocephalus in pediatric patients.

Etiology
Hemorrhage within ventricle
Myelomeningocele
Neoplasm
Cerebral aqueductal stenosis
Meningitis
Cranial trauma
Others
Idiopathic
Two or more etiologies

Source: Authors' compilation based on data from Kirollos et al. (2019).

5. Classifications of HCP

Hydrocephalus can be classified into various types, which are listed below.

5.1. Noncommunicating Hydrocephalus (Obstructive)

Potential results that can be obtained from lesions that obstruct the CSF pathway from the lateral ventricle to the fourth ventricle and its outlet are noted below:

a. Dilated lateral and third ventricles and a normal-size fourth ventricle;
b. Dilation of the lateral ventricle due to an obstruction at the Forman of Monro, commonly by a colloid cyst;
c. A posterior Fossa mass lesion (tumor, cyst, hematoma), an intraventricular mass lesion (tumor, IVH, cyst), and/or aqueductal stenosis.
d. Lumber puncture is contraindicated in this type of hydrocephalus.

5.2. Communicating Hydrocephalus

This type of hydrocephalus refers to situations in which the intracerebral CSF pathways are active and patent but there is an excessive collection of CSF, commonly from abnormalities in CSF absorption. Here, all four ventricles are dilated. This condition occurs because of meningitis, SAH, adult IVH, and IVH in premature babies. To treat communicating hydrocephalus, a lumber drain can be inserted to decrease intracranial pressure.

6. Clinical Features of HCP

HCP's clinical features vary according to the age limit of the patients. Newborn babies present with enlarge heads and are often born via Caesarean section.

In infancy, the head will grow much larger, with a prominent scalp vein with widened fontanelles. Commonly, the anterior fontanelle closes at 5–15 months' time, and the posterior fontanelle closes at around 2–3 months.

Babies are commonly irritable and cry because of hydrocephalus, and their brains become compressed. Their brain tissue (cortical mantle) becomes thin and varies from a few millimeters to a few centimeters. Normally, the cerebral mantle is 5–6 cm long.

At birth, the head circumference of a baby is 35 cm at 6 months, 39 cm at 1 year, 45 cm at 1.5 years, and 49 cm at 2 years.

Commonly, the head enlarges at a rate of 1–2 cm per month.

6.1. The Various Symptoms of Hydrocephalus

6.1.1. Infants

- Unusually large head size;
- Fastly growing head circumference;
- Bulged and tense fontanelles;
- Prominent and engorged scalp veins;
- Downward deviation of eyes or sunset sign;
- Nausea and vomiting;

- Sleepiness;
- Irritability;
- Seizures.

6.1.2. Children and Adolescents

- Nausea as well as vomiting;
- Papilledema;
- Blurred vision or diplopia;
- Imbalance and gait disturbances;
- Delays in or lack of milestones of development;
- Personality changes;
- Poor concentration;
- Seizures;
- Loss of appetite;
- Urinary incontinence.

6.1.3. Adults

- Headache;
- Nausea as well as vomiting;
- Difficulty walking and gait disturbances;
- Imbalance or incoordination;
- Lethargy;
- Urinary incontinence;
- Visual disturbances;
- Impaired cognitive function skills;
- Loss of memory;
- Mild dementia.

7. Radiological Features

Hydrocephalus is best seen via CT or MRI
Based on experience, a clinician can identify HCP by its characteristics on a CT scan or MRI.

7.1. Specific Imaging Criteria for HCP

1. The width of both temporal horns (THs) of the lateral ventricle is $\geq$2 mm.
2. Both THs are less than 2 mm wide, and the FH/ID ratio is greater than 0:5. (where FH is the greatest width/breath, and ID is the internal diameter from inner-table to inner-table at this level) (Greenberg 2010).

7.2. Other Features Alluding to Hydrocephalus

1. "Mickey Mouse" ventricles (ballooning of the FH of the lateral ventricles) and/or third-ventricular ballooning (the third ventricle is usually slit-like).
2. Periventricular lower density as seen in a CT scan or periventricular higher signal intensity on T2WI as shown via MRI.
3. The FH/ID ratio: <40% is normal, 40–50% is considered borderline, and >50% is hydrocephalus (Omidi-Varmezani 2015).
4. An Evans ratio, i.e., the ratio of FH to BPD (height biparietal diameter calculated in the same CT slice), of >0.3 alludes to hydrocephalus
5. Sagittal view shows thinning and/or superior bending of the corpus callosum (Greenberg 2010).

7.3. Radiological Features in 'Chronic HCP'

1. Beaten copper/silver cranium plain skull X-ray;
2. The third ventricle is herniating into the sellae (as seen via CT scan or MRI);
3. Erosion of sellae turcica;
4. Macrocrania: OFC greater than the 98th percentile;
5. Atrophy of the corpus callosum (Figure 4A);
6. In infantile patients:

(a) Sutural gap (diastasis);
(b) Delay in closure of the fontanelles;
(c) Failure to thrive;
(d) Developmental diverticulum (Figure 4B).

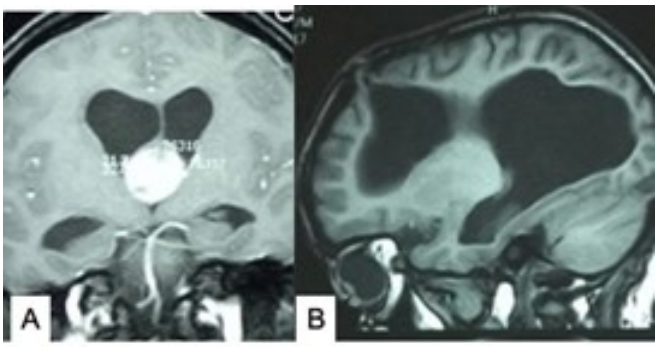

Figure 4. (**A**) MRI shows chronic HCP with diverticulum. (**B**) MRI shows colloid cyst of the 3rd ventricle with biventricular HCP. Source: Figure by authors.

8. Management of HCP

Hydrocephalus needs to be treated to allow normal development of the brain. Otherwise, brain activity such as cognition, motor activity, vision, and urinary control will all gradually decline. HCP strongly affects babies' developmental milestones. Commonly, a baby will raise its head at 3–4 months, crawl at 6–7 months, stand with support at 10–11 months, and be able to walk by year one.

8.1. Endoscopic 3RD Ventriculostomy (ETV)

ETV is an excellent and alternative approach to treating hydrocephalus. For the last 20 years, ETV has worked excellently for most patients. In the case of congenital hydrocephalus occurring when a baby is less than six months old, the surgeons generally do not prefer ETV as it has a more than 50% chance of failure. ETV works better in cases of adult-onset aqueduct stenosis and tumor-related obstructive hydrocephalus. The aim is to create a pathway between the basal cistern and the ventricular system.

8.1.1. Surgical Procedure of ETV

The head is positioned at 30–45-degrees, elevated, and kept straight. There is no need to rotate the head to avoid disorientation. Kocher's point is the commonest point to choose for ETV.

A "U"-shaped skin flap is chosen instead of a linear paramedian wound to allow better closure of the wound.

The optimal burr hole is positioned 3–4 cm laterally from the midline and 1 cm in front of the coronal suture (Oka et al. 1999).

Following the creation of a 10 mm burr hole, the dura is cut in a cruciate fashion. The underlying brain is cauterized, and an incision is made.

After making all preparations, such as employing a camera with a cover, a telescope, a sheath with an obturator, and Fogarty catheters, saline and then the brain canula are introduced through the burr hole.

Clear CSF normally comes out after applying 5–6 cm or less.

At this stage, an ETV telescope is introduced.

The telescope will show the location of the landing, allowing for navigation through the foramen of Monro by revealing the choroid plexus, fornix, and thalamostriate vein so that one can reach the floor of the third ventricle.

The floor of the third ventricle can be identified by the two mammillary bodies in the posterior and infundibular recess in the anterior and the thin or thick membrane (tuberous cinereum) beneath the tuber cinereum Basilar artery pulsation can be observed if the membrane is thin.

The next step is making a puncture in the membrane either with ventriculostomy forceps or by using unipolar or bipolar tips.

Following fenestration, the membrane needs to be dilated at least 4-6 mm, which can be performed either by using a 3F Fogarty catheter or via opening the space using ventriculostomy forceps.

Pulsation of the membrane should now be evident.

Sometimes another membrane is there: it is known as the Liliequist membrane, and it also needs to be punctured to allow the free flow of CSF into the prepontine space.

Hemostasis must be ensured before the withdrawal of the endoscope.

Commonly, hemostasis is achieved via compression using a Fogarty catheter or via direct cauterization.

Profuse irrigation is applied before removing the telescope.

A gel foam is placed in the port, and the wounds in all the layers are closed after the removal of the telescope.

Complications of ETV

The commonest complication of ETV is bleeding; hence, proper hemostasis is mandatory before the closure of an ETV scalp wound. Hemostasis is commonly achieved via continuous irrigation. Another way of achieving hemostasis is the application of either a unipolar or bipolar diathermy. It is recommended to ensure hemostasis in this case and EVD should be employed for safety (Nader et al. 2014).

Success Rate of ETV

ETV success rate is a matter of debate (Quiñones-Hinojosa 2012; Chowdhury et al. 2017) and shown in Table 2.

Favorable clinical as well as radiographic characteristics for ETVare shown in Table 3.

8.2. Choroid Plexus Cauterization

ETV plus choroid plexus cauterization increases the efficacy of ETV. Flexible endoscopic support is required to cauterize the entire length of the choroid plexus on both the right and left sides.

8.3. EVD

EVD is a temporary CSF diversion procedure commonly performed at Kocher's point; however, there are other points at which to perform EVD, such as via Frazier's, Dandy's, and Keene's points. Commonly, EVD is performed using an at least 5 cm subcuticular tunnel; in one study, it was shown that a long subcutaneous tunnel is associated with a reduced chance of brain infection. However, EVD placement after more than two weeks is complicated by meningitis.

Table 2. Third Ventriculostomy success rates according to HCP cause.

Higher Success Rates ($\geq$75%)
Acquired cerebral aqueductal stenosis neoplasms obstructing ventricular CSF outflow
Tectal tumour
Pineal tumour
Thalamic tumour
Intraventricular tumour
Intermediate-Level Success Rates (50–70%):
Myelomeningoceles (shunted earlier, in older patients)
Congenital cerebral aqueductal stenosis
Cystic lesion obstructing CSF flow
Arachnoid cysts
Dandy–Walker malformation
Earlier-shunted patients with difficulties
Slit ventricle syndrome
Intractable or recurrent shunt infections
Intractable or recurrent shunt malfunction
Low Success Rates (<50%)
Myelomeningoceles (not shunted earlier, in neonatal patients)
Posthemorrhagic HCP
Postinfectious HCP

Source: Authors' compilation based on data from Quiñones-Hinojosa (2012); Chowdhury et al. (2017).

Table 3. Favorable clinical as well as radiographic characteristics for ETV.

Clinical
HCP etiology in a higher or intermediate success group
Age of more than six months at time of HCP diagnosis
Age of more than six months at time of intervention
No history of prior radiation therapies
No history of prior meningitis or hemorrhage
Patient was shunted earlier
Radiographic
Definite proof of ventricular noncommunication
Obstructive type of HCP
Anatomical obstruction of aqueduct
Absence of aqueductal CSF flow void in T2-weighted MRI images
Suitable 3rd ventricular anatomy
Width of foramen of Monro is enough to accommodate endoscope
Rigid endoscope > 7 mm
Flexible endoscope > 4 mm
Third ventricular floor is thin
Downward bulging floor of 3rd ventricle draped over clivus
Basilar artery is posterior to mammillary bodies
Absence of anatomical anomalies related to surgery
AVM or neoplasm obscuring floor of third ventricle
Large massa intermedia
Inadequate space between mammillary bodies, the basilar artery, and the clivus Ectasia of basilar artery

Source: Authors' compilation based on data from Chakraborty et al. (2012); Quiñones-Hinojosa (2012); Chowdhury et al. (2017).

8.3.1. EVD Points

(i) Paine's point: "The junction at 90° angles of the lines measured 2.5 cm above from the floor of the anterior cerebral fossa (lateral orbital roof) and 2.5 cm front to the Sylvian fissure," according to Paine's point definition. The frontal horn base is reached via a ventriculostomy through this site.
(ii) Kocher's point: The entry site is 2–3 cm from the midline, which is roughly the mid-pupillary line from the front of the skull, and 1 cm from the front of the coronal suture, which is nearly 11 cm above from the nasion (this positioning is intended to protect the motor strip).
(iii) Keene's point: The entry point is 2.5–3 cm behind and 2.5–3 cm above the pinna; placement is at the trigone.
(iv) Frazier's point: The entry point is 3–4 cm from the midline laterally and 6–7 cm above the inion.
(v) Dandy's point: The entry point is 2 cm from the midline laterally and 3 cm above the inion (using this point may render the patient more prone to an optic radiation injury than they would be if Frazier's point were used).

EVD is commonly carried out in the case of acute hydrocephalus or obstructive hydrocephalus just before or at the time of primary surgery.

8.3.2. Complications of EVD

1. Bleeding along the tract.
2. EVD migration: The EVD tool can come out through the tract if not secured well.
3. The withdrawal of too much CSF leads to subdural hygroma or hematoma.

8.4. Shunting

Shunting is the most common treatment for hydrocephalus for both the congenital and acquired varieties. Among all shunting procedures, a VP shunt is the commonest, followed by ventriculo-atrial (VA) or ventriculo-pleural shunts.

8.4.1. Ventriculo-Peritoneal Shunt

About 80% of all shunting procedures are VP shunts. This is the only surgery wherein long exposure of the wound can take place, so there is an about 20% chance of shunt infection.

Exposure from the head to the abdomen through the neck is required.

Here, a burr hole is commonly made at Keene's, Frazier's, or Kocher's points and traced up, leading to a subcutaneous Daniel lot. The end of the shunt is introduced into the peritoneal cavity, commonly in the paramedian plane.

Complications of VP Shunt

1. Common complication:
   (i) *Shunt infection:* Shunt infection rates range from 8-15%, with 10% being generally accepted. The greatest risk of shunt infection is within 6 months of its original implementation. Shunt infection is often treated with antibiotics and shunt externalizations.
   (ii) *Shunt obstruction:* Shunt failure occurs when a shunt becomes blocked at either the cranial or caudal ends. Excess protein in the CSF might block the shunt valve at the proximal end. The additional protein will deposit at the drainage point and clog the valve over time. If the shunt is dragged out of the abdominal cavity (as with VP shunts), or if similar protein buildup occurs, the shunt can clog at the end.
   (iii) *Shunt over drainage:* The possible complications of over shunting include the following:
      (a) Slit ventricles;
      (b) Intracranial hypotension;
      (c) Subdural hematoma;
      (d) Craniosynostosis and microcephaly;
      (e) Stenosis or occlusion of sylvian aqueduct;
   (iv) Disconnection or breakage at any point;
   (v) Shunt leaking;
   (vi) Hardware erosion through skin;
   (vii) Seizure;
   (viii) Malposition of catheter tip (Figure 5);
   (ix) Silicon allergy.

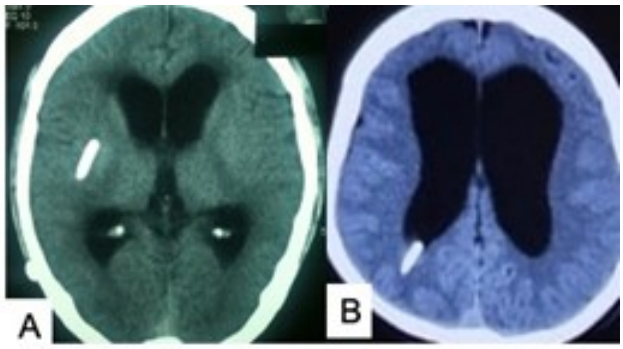

Figure 5. (**A**) CT scan showing that the cranial end of the VP shunt is not in the ventricle; (**B**) CT scan showing that the cranial end of the VP shunt has migrated out of the ventricle. Source: Figure by authors.

2. Others
   (i) Inguinal hernias;
   (ii) The need to lengthen the catheter with growth;
   (iii) Obstructions of peritoneal catheter with growth:
      (a) Omentum;
      (b) Peritoneal cysts;
      (c) Peritoneal adhesion.
3. Complication specific for V-P shunts:
   (i) Peritonitis;
   (ii) Hydrocele;

(iii) CSF ascites;
(iv) The migration of the tip of the shunt or the whole thing into the ventricular system (Figure 6);
(v) Intestinal obstruction;
(vi) Volvulus;
(vii) Intestinal strangulation;
(viii) Over-shunting: Over shunting can cause other complications; such as slit ventricles, intracranial hypotension, subdural hematoma, craniosynostosis, microcephaly, and stenosis or occlusion of sylvian aqueduct.

Shunt Infection

1. Risk factors for shunt infection:
   - Young age of the patient: Waiting until a child is two weeks old before administering antibiotics to treat a myelomeningocele (MM) may reduce the infection rate dramatically.
   - Long operations.
   - Open neural-tube defect.
2. Pathogens:
   - Early infection:
     - Staphylococcus epidermidis
     - Staphylococcus aureus
     - Gram-negative bacilli
     - E. coli and Streptococcus Hemolyticus in neonates.
   - Late infection:
     - Staphylococcus epidermidis
3. Medical treatment/Antibiotics for VP shunt infection:
   - Commonly, the surgeons choose injectable antibiotics, such as Meropenem, Vancomycin, Clindamycin, Linezolid, and Rifampicin.
4. Surgical treatment of shunt infection

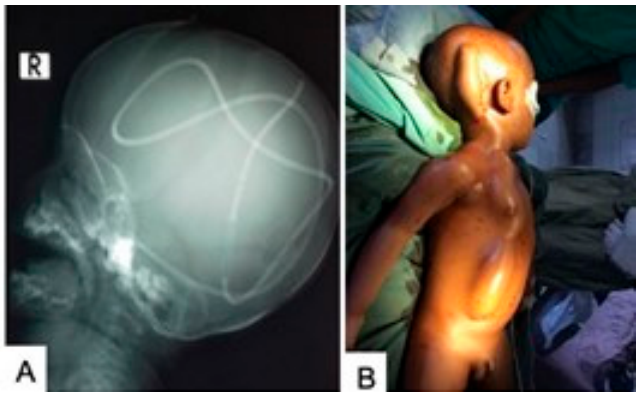

Figure 6. (**A**) X-Ray shows total migration of shunt tube within the brain. (**B**) Picture shows accommodation of CSF within the subcutaneous tunnel because of lower end block. Source: Figure by authors.

Management of a CSF shunt infection usually requires the removal of the shunt as well as the insertion of a temporary ventricular reservoir (Ommaya reservoir) until the infection is over. There are four main methods of managing VP shunt infection:

(i) Appropriate antibiotics;
(ii) Removal of the infected shunt with simultaneous replacement;
(iii) Exteriorization of the shunt with subsequent replacement;
(iv) External ventricular drain (EVD) insertion with removal of the shunt with subsequent shunt re-insertion (this is the best technique, with an over 95% success rate) (Greenberg 2010).

5. VP shunt Replacement
   - VP Shunt externalization:
     Indications:

- VP shunt infection;
- Abdominal pseudocysts;
- Peritonitis.

Contraindications:

- Proximal tube obstruction;
- Valve malfunction;
- Pus in the valve.

- Surgical technique: A small incision is made over the neck along the subcutaneous tube.

Blunt dissection is carried out until the white, shiny VP shunt tube can be seen. Once the tube is observed, the lower end of the VP shunt tube should be gently pulled. All distal tubing removed from the abdomen is connected by a connector system to a closed external drainage system. The wound is closed in layers using an occlusive dressing.

8.4.2. Ventriculo-Atrial (VA) Shunt

The VA shunt is an alternative to the VP shunt. Here, the common facial vein is selected as the area where the lower end of the catheter will be placed. From the common facial vein, the shunt goes to the internal jugular vein and then the right atrium of the heart.

The placement of the lower end by the right atrium can be confirmed via a chest X-ray, where the tip will be located at the D2 level, and at the time of the insertion of the shunt tube, there may be a rhythm change of the heart.

Complications of VA shunt:

- The shunt may be blocked by a blood clot.
- It may be displaced.

8.4.3. Ventriculo-Pleural Shunt

Another alternative shunting technique is the V pleural shunt, where the lower end is placed in the lower chest in the interpleural space, commonly in the fifth or sixth intercostal space and in the mid or posterior axillary line.

Here, the big pleural surface acts as an absorber of CSF (Nader et al. 2014).

8.4.4. Lumbo-Peritoneal Shunt

This is a shunting procedure for the IIH or pseudo-tumor cerebri or communication variety of HCP. It is performed by placing an LP shunt where a lumbar puncture has been made using a 14-bore Touhy needle. The lower end of the shunt is introduced into the peritoneal cavity via a mini laparotomy incision through a subcutaneous tunnel in the flank (Greenberg 2010).

8.4.5. Cysto-Peritoneal Shunt

This is a bypass procedure for encysted cysts such as posterior fossa arachnoid cysts and Sylvia fissure cysts, which compress the loco-regional brain structure.

9. Normal-Pressure Hydrocephalus (NPH)

This condition is typically recognized by noting the following triad of symptoms: dementia, ataxia, and incontinence.

9.1. Epidemiology

According to recent research on probable NPH symptoms, at least 21.2% of nursing home residents have gait impairment, 9.4% have dementia, and 14.7% have incontinence (Shprecher et al. 2008).

9.2. Pathology

NPH is a condition characterized by a decrease in CSF absorption rather than an increase in CSF production. The arachnoid granulations fail to maintain their baseline clearance of CSF, whether due to a known or unknown reason. This is frequently due to fibrosis and scarring, which conceal absorptive surfaces.

Surgical therapy for patients with a severe form of dementia is generally discouraged, even in the presence of gait difficulty and incontinence, independent of radiographic results (Shprecher et al. 2008).

9.3. Radiological Features

The image below show communicating HCP observed via CT (Figure 7A) scan or MRI.

Figure 7B shows that MRI can detect periventricular white matter alterations better than CT scans. These contiguous T2/FLAIR (fluid-attenuated inversion recovery) hyperintensities are assumed to be trans-ependymal edema due to high CSF pressure, but they may seem similar to those seen in small-vessel ischemic illnesses (Shprecher et al. 2008).

9.4. Morphological Changes in MRI

9.4.1. Ventriculomegaly

- Evans' index is increased >0.3;
- The THs of the lateral ventricles are widened by >6 mm;
- Acute callosal angle;
- Superior bowing of the corpus callosum.

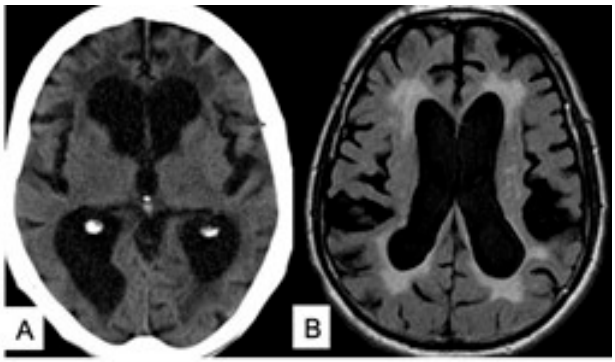

Figure 7. (**A**) CT scan showing ventriculomegaly with periventricular hypodensity suggestive of hydrostatic edema. (**B**) MRI scan shows ventriculomegaly with periventricular high-intensity signal suggestive of hydrostatic edema. Source: Figure by authors.

9.4.2. Disproportionate Alterations in Subarachnoid Spaces

- Expanded Sylvian fissures;
- Compressed high convexity (subarachnoid spaces and sulci are narrow at the vertex and medial/parafalcine region)
- Cingulate sulcus sign: the posterior portion of cingulate sulcus is slenderer than the anterior portion.
- Focal/isolated expansion of sulci on the medial surface or convexity area (sometimes known as transport sulci)

Significantly expanded subarachnoid space hydrocephalus is characterized by ventriculomegaly, Sylvian fissure expansion, and crowding at the vertex.

9.4.3. Cine Phase-Contrast MRI

Phase-contrast CSF flow in films is measured using MRI in terms of stroke volume, which is defined as the average volume of CSF traveling through the cerebral aqueduct in both systole and diastole (Shprecher et al. 2008). A stroke volume of more than 42 micro-liters may indicate that it is probable there will be an improvement after a VP shunt is placed (Scollato et al. 2008).

9.5. Differential Diagnosis of NPH

The possible neuro-imaging differential include the following:

- Normal senile brain;
- Alzheimer's disease with dementia: may demonstrate greater expansion of perihippocampal fissures;
- Obstructive HCP—due to neoplasm (e.g., pineal region, tectal plate, midbrain)

- Lewy body dementia—visual hallucinations as well as delusions are more marked;
- Parkinson's disease—it is important to differentiate one-sided symptoms;
- AIDS–dementia complex—positive HIV immunological test.

Asymmetric rest tremor, lead pipe rigidity, and visual hallucinations may indicate dementia with Lewy bodies (DLB), which induces cognitive abnormalities similar to Alzheimer's disease. Pseudodementia is also a possibility in the differential diagnosis. Aphasia, apraxia, or agnosia might arouse suspicion of dementia with cortical dysfunction, such as Alzheimer's disease (AD), multi-infarct dementia, or frontotemporal dementia, if these symptom appear early (Shprecher et al. 2008). Regardless of ventriculomegaly, a cause other than NPH should be investigated for patients with progressing dementia who do not have gait problems.

9.5.1. Tests for Differentials

Although a high-volume (>40 mL) spinal tap (also known as lumbar tap test) was the first method for diagnosing NPH and predicting shunt response, external lumbar drainage (ELD) is becoming more widely accepted as a more superior predictor for patients who do not respond to a tap test. CSF is drained at a rate of 10–15 cm^3 per hour for 72 h using a lumbar spinal catheter (Shprecher et al. 2008).

9.6. Treatment of NPH

In about 60% of NPH patients, CSF shunting techniques such as ventriculoperitoneal, ventriculopleural, and ventriculoatrial shunting can result in considerable clinical improvement in NPH symptoms (Shprecher et al. 2008).

10. Pineal Cyst and Posterior Fossa Cyst with HCP

Affected patients commonly present with a visual discrepancy and headache (Figure 8).

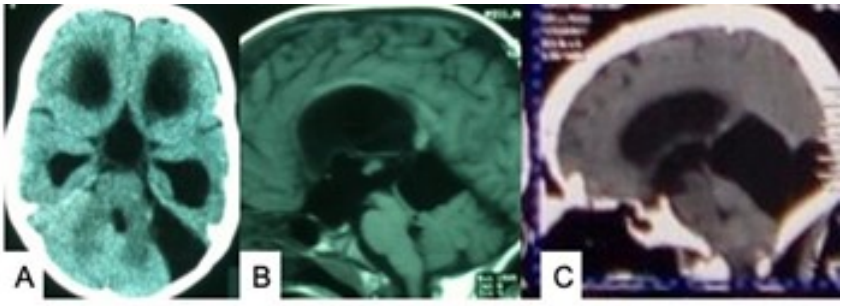

Figure 8. (**A**) CT shows CP angle arachnoid cyst with HCP. (**B**) MRI shows pineal region arachnoid cyst with HCP. (**C**) MRI shows huge pineal region arachnoid cyst with HCP. Source: Figure by authors.

The best treatment is either a endoscopic or microscopic excision of the cyst wall.

11. Trapped Fourth Ventricle

An isolated fourth ventricle can be caused by failure to communicate with either the aqueduct of the Sylvius or the fourth ventricular outlet—the foramen Luschka and Magendie (Figure 9).

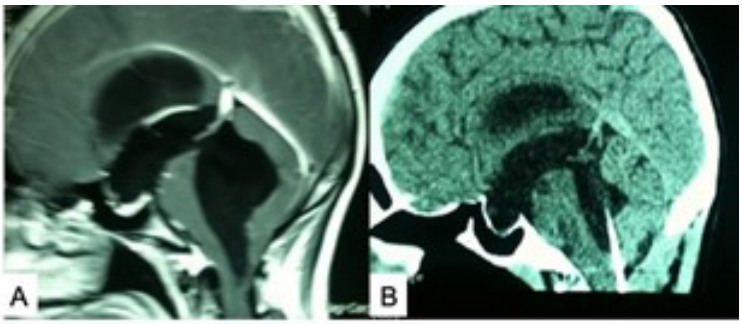

Figure 9. (**A**) MRI shows entrapped and dilated 4th ventricle. (**B**) MRI shows previous entrapped and dilated 4th ventricle treated using a 4th ventricular shunt. Source: Figure by authors.

11.1. Etiology

- Post-infective hydrocephalus;
- Chronic shunting of lateral ventricle.

11.2. Clinical Presentation

- Headache;
- Recurrent vomiting;
- Decreased consciousness level;
- Ataxia;
- Dysphagia.

11.3. Surgical Technique

- A fourth ventricular shunt plus an ETV/VP shunt;
- Opening of fourth ventricular outflow plus an aqueductoplasty +/− stent.

12. Double-Compartment Hydrocephalus

The figure below shows supratentorial and infratentorial hydrocephalus in clinical sequence and separately (Figure 10).

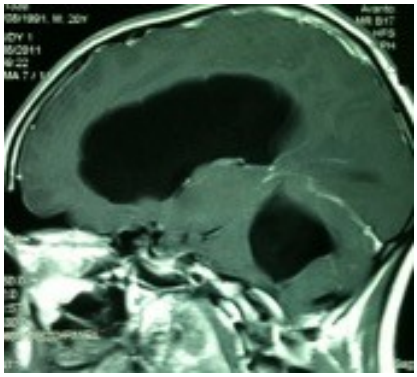

Figure 10. MRI shows double compartmental HCP. Source: Figure by authors.

13. HCP in Dandy–Walker Malformation

Overall, 80% of Dandy–Walker cysts are diagnosed by 1 year of age. The development of hydrocephalus may be explained by a hemorrhage at the time of delivery and subsequent inflammatory blockage of the foramen Luschka. Alternatively, a patient may have congenital aqueductal stenosis. Young children present with delayed motor development and poor head control. Older children present with truncal ataxia and nystagmus.

13.1. Radiological Diagnosis

- Cranial ultrasound and a CT scan of head are the initial diagnostic investigations performed. CT has revealed that the cerebellar hemisphere is typically hypoplastic. The tentorium, torcula, and venous sinuses are in higher positions.
- Massively dilated fourth ventricle.
- Concomitant hydrocephalus.

13.2. Treatment Options

1. The placement of a proximal bi-compartmental shunt at a single valve using a Y connector and distal tubing.
2. The application of a completely separate VP shunt.
3. Addressing only hydrocephalus using either a VP shunt or via ETV
4. Only ETV of the lateral ventricle, keeping posterior fossa cyst untouched (Nader et al. 2014).

14. Long Standing Overt Ventriculomegaly

LOVA—long-standing overt ventriculomegaly in adults—is a type of chronic hydrocephalus of infantile-onset severe third and lateral ventriculomegaly with a mild clinical presentation in adults. It may be a reactivated form of arrested hydrocephalus wherein compensational factors have failed. Oi et al. described it for the first time in 2000, considering the following criteria: (1) adult-onset hydrocephalus symptoms (headache, cognitive decline, imbalance, gait disturbances, and visual deterioration/diplopia); (2) macrocephaly; (3) overt triventriculomegaly observed via neuroimaging with cortical sulcal effacement and/or destruction of the sella turcica; and (4) no secondary causes of aqueductal stenosis (Oi et al. 2000). It should be differentiated from other types of hydrocephalus without macrocephaly. The clinical features are non-specific low-grade headache, early-morning nausea and occasional vomiting, sometimes insidious visual failure, and normal-pressure-hydrocephalus-like features. Magnetic resonance imaging (MRI) is the gold standard in LOVA diagnostics, combined with clinical and neuropsychological examination.

Treatments includes the following:

1. The conservative approach with regular follow-ups;
2. Endoscopic third ventriculostomy (ETV);
3. The installation of a programmable ventriculo-peritoneal shunt.

Many authors have recommended that endoscopic third ventriculostomy (ETV) is the gold standard for the treatment of LOVA, as it is a triventricular form of hydrocephalus. If ETV fails, a CSF shunt should be used instead (Tuniz et al. 2021).

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