INTRODUCTION

Anatomy of the brain

Structure

The adult human brain weighs on average about 1.5 kg [1] with a volume of around 1130 cubic centimeters (cm^3) in women and 1260 cm^3 in men, although there is substantial individual variation.[2] Men with the same body height and body surface area as women have on average 100 grams heavier brains,[3] although these differences do not correlate in any simple way with IQ or other measures of cognitive performance.[4]

The human brain is composed of neurons, glial cells, and blood vessels. The number of neurons was estimated in 1988 as 100 billion (10^{11}), interconnected by their 100 trillion (10^{14}) synapses.[5] In 2009, the estimate was lowered to 86 billion neurons, of which 16.3 billion are in the cerebral cortex, and 69 billion in the cerebellum.[6, 7]

The Cerebral Lobes

1-The frontal lobe

Extends from the frontal pole of the brain to the central sulcus. Since it lies mostly in the anterior cranial fossa, its lower surface is shallow and concave to fit the orbital roof. The lateral sulcus separates it from the temporal lobe. [7]

2-The parietal lobe

Lies between the frontal, temporal, and occipital lobes. It is separated from the frontal lobe by the central sulcus. Similarly, it is largely separated from the temporal lobe by the lateral sulcus, but at the end of the sulcus, parietal, temporal, and occipital lobes are confluent on the lateral surface of the hemisphere. [7]

3-The temporal lobe

Is the part of the hemisphere that lies below the lateral sulcus. Posterioly it is continuous with the occipital lobe. [7]

4-The occipital lobe

Is located at the posterior aspect of the brain, figures (1-4). [7]

The Brain Stem

The brain stem is composed of the hindbrain and midbrain. The hindbrain contains structures including medulla and the pons, figure (5).

The Hindbrain

The hindbrain is the structure that connects the spinal cord to the brain.

- The medulla is continuous below, through the foramen magnum, with the spinal cord and above with the pons; posteriorly, it is connected with the cerebellum by the inferior cerebellar peduncles.
- The pons lies between the medulla and the midbrain and is connected to the cerebellum by the middle cerebellar peduncles. Its ventral surface presents a shallow median groove and numerous transverse ridges, which are continuous laterally with the middle cerebellar peduncle. The dorsal surface of the pons forms the upper part of the floor of the fourth ventricle.(7)

The Midbrain

The midbrain or mesencephalon is that part of the neuroaxis located between the pons inferiorly and the thalamus superiorly. It is a short, constricted segment of the brainstem that measures only about 2cm. By definition each half of the midbrain is called the cerebral peduncle, and this is further divided into a ventral part called the crus cerebri and a dorsal part called the midbrain tegmentum. (7)

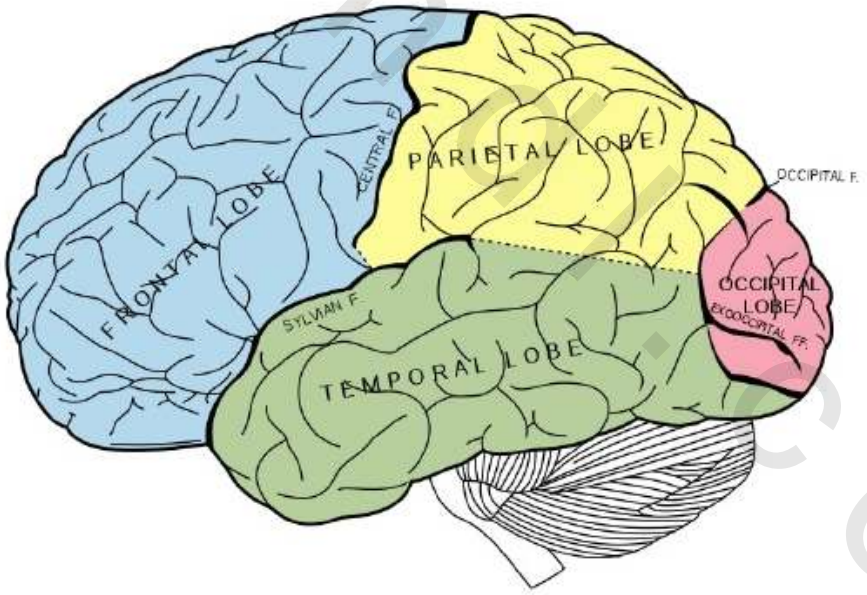

Figure (1): Showing the four cerebral hemispheres. (7)

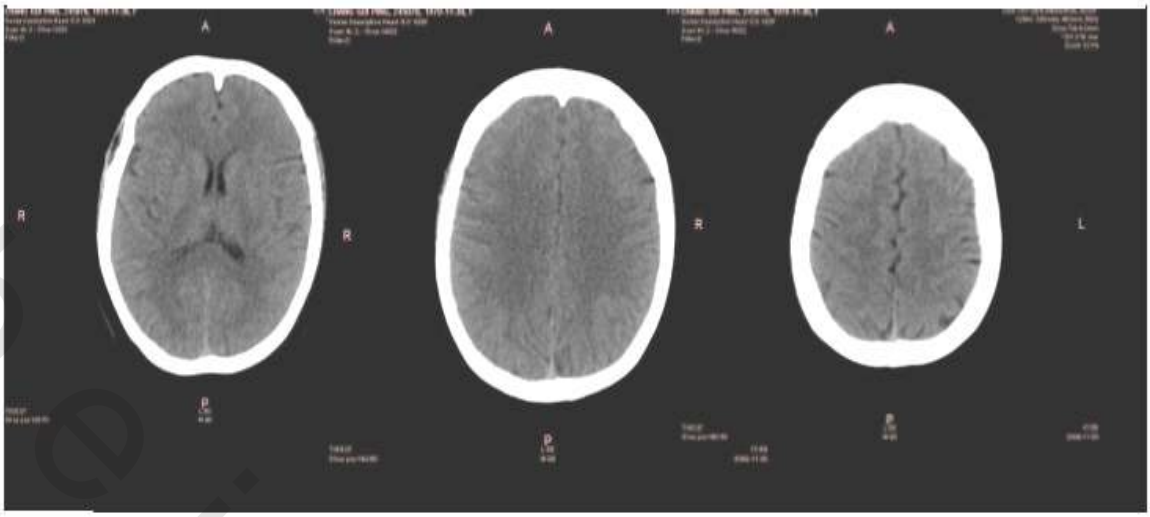

Figure (2): CT imaging of normal cerebral lobes. [7]

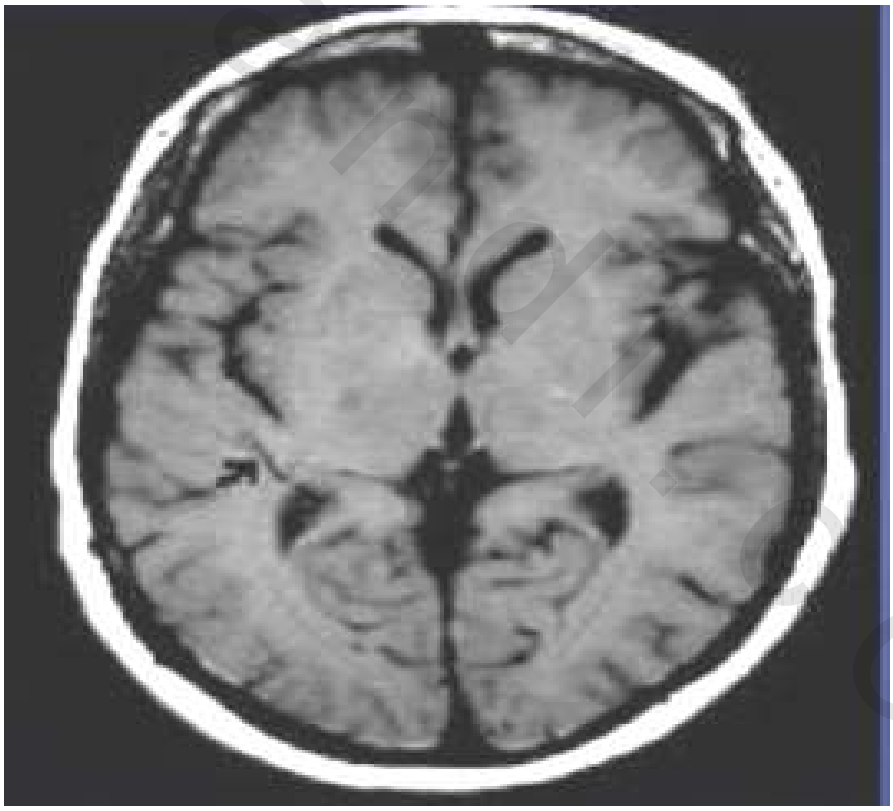

Figure (3): MRI T1WI of the normal brain. [7]

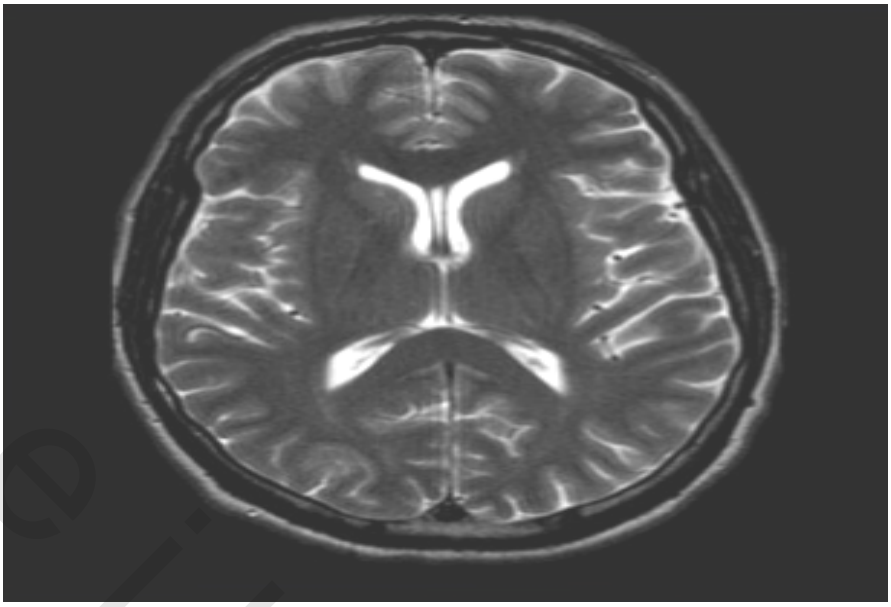

Figure (4): MRI T2WI of the normal brain. [7]

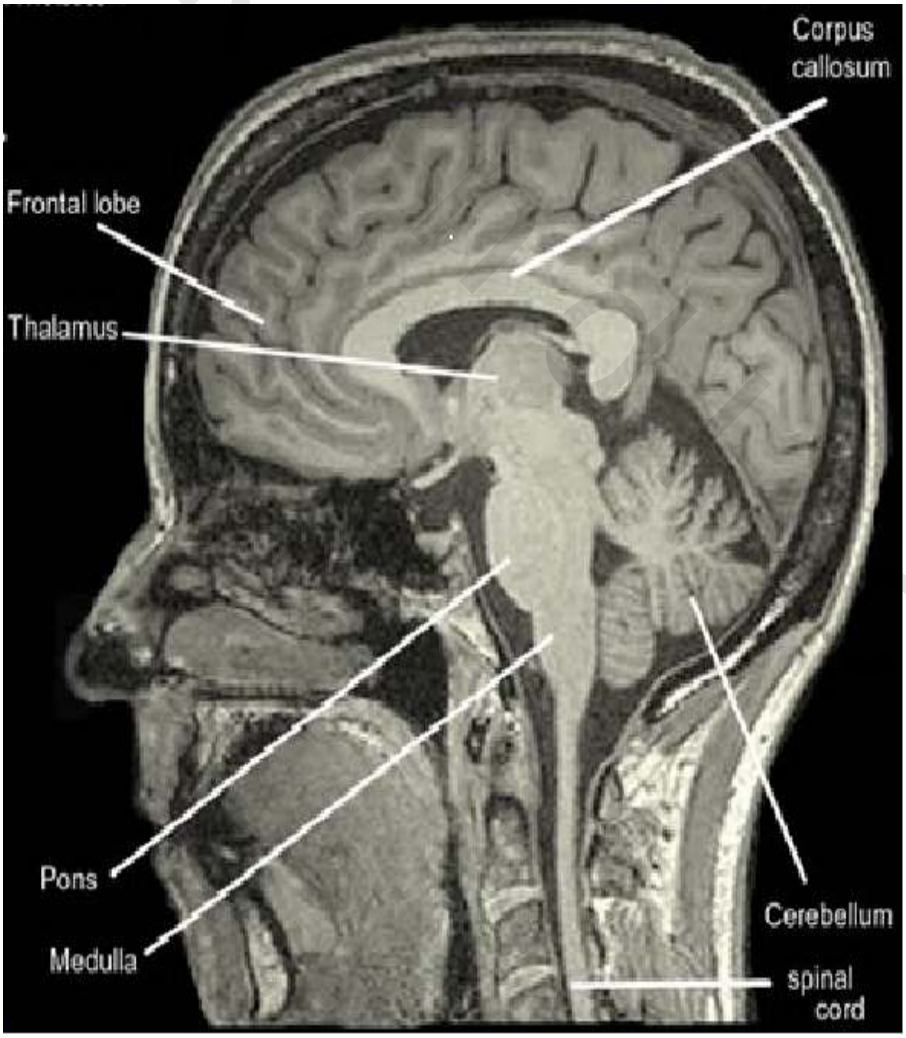

Figure (5): Sagittal T1WI MRI showing anatomy of the brain stem& cerebellum. [7]

Deep structures

Hypothalamus

The hypothalamus forms the floor of the 3rd ventricle. It includes from anterior to posterior the following: the optic chiasma, the tuber cinereum, the infundibular stalk (leading down to the posterior lobe of the pituitary), the mammillary bodies and the posterior perforated substance. [8]

Pituitary gland

The pituitary lies in the cavity of the pituitary fossa covered over by the diaphragma sellae, which is a fold of dura mater. This fold has a central aperture through which passes the infundibulum. Inferior to it is the body of the sphenoid, laterally lies the cavernous sinus and its contents separated by dura mater, with intercavernous sinuses communicating infront, behind and below. The optic chiasma lies superiorly, immediately anterior to the infundibulum. [8]

Pineal gland

The pineal gland is reddish-gray and about the size of a grain of rice (5-8 mm) in humans, located just rostro-dorsal to the superior colliculus and postroinferior to the stria medullaris, between the laterally positioned thalamic bodies. It is part of the epithalamus. It is a midline structure shaped like a pin con, and is often seen in plain skull X-rays, as it is often calcified. [8]

Thalamus

The thalamus is an oval mass of grey matter which forms the lateral wall of the 3rd ventricle; it extends from the interventricular foramen rostrally to the midbrain caudally. Laterally, it is related to the internal capsule (and through it to the basal ganglia), and dorsally to the floor of lateral ventricle. Medially, it is frequently connected with its fellow of the opposite side through the massa intermedia. [8]

Basal ganglia

The basal ganglia are located deep within the cerebral hemispheres in the telencephalon region of the brain. A component of the corpus striatum, it consists of the subthalamic nucleus and the substantia nigra. [7]

The Cerebellum

The cerebellum is the largest part of the hind-brain and occupies most of the posterior cranial fossa. It is made up of two lateral cerebellar hemispheres and a median vermis. Inferiorly, the vermis is clearly separated from the two hemispheres and lies at the bottom of a deep cleft, the vallecula; superiorly, it is only marked off from the hemispheres as a low median elevation. Small ventral portion of the hemisphere lying on the middle cerebellar peduncle is almost completely separated from the rest of the cerebellum as the flocculus. [7]

Ventricles and cerebrospinal fluid

The ventricular system is embryologically derived from the neural canal, forming early in the development of the neural tube. The three brain vesicles (prosencephalon or forebrain, mesencephalon or midbrain, and rhombencephalon or hindbrain) form around the end of the first gestational month. The neural canal dilates within the prosencephalon, leading to the formation of the lateral ventricles and third ventricle. The cavity of the mesencephalon forms the cerebral aqueduct. The dilation of the neural canal within the rhombencephalon forms the fourth ventricle, figure (6). [8]

The lateral ventricles communicate with the third ventricle through interventricular foramens, and the third ventricle communicates with the fourth ventricle through the cerebral aqueduct. [8]

During early development, the septum pellucidum is formed by the thinned walls of the two cerebral hemispheres and contains a fluid-filled cavity, named the cavum, which may persist. [8]

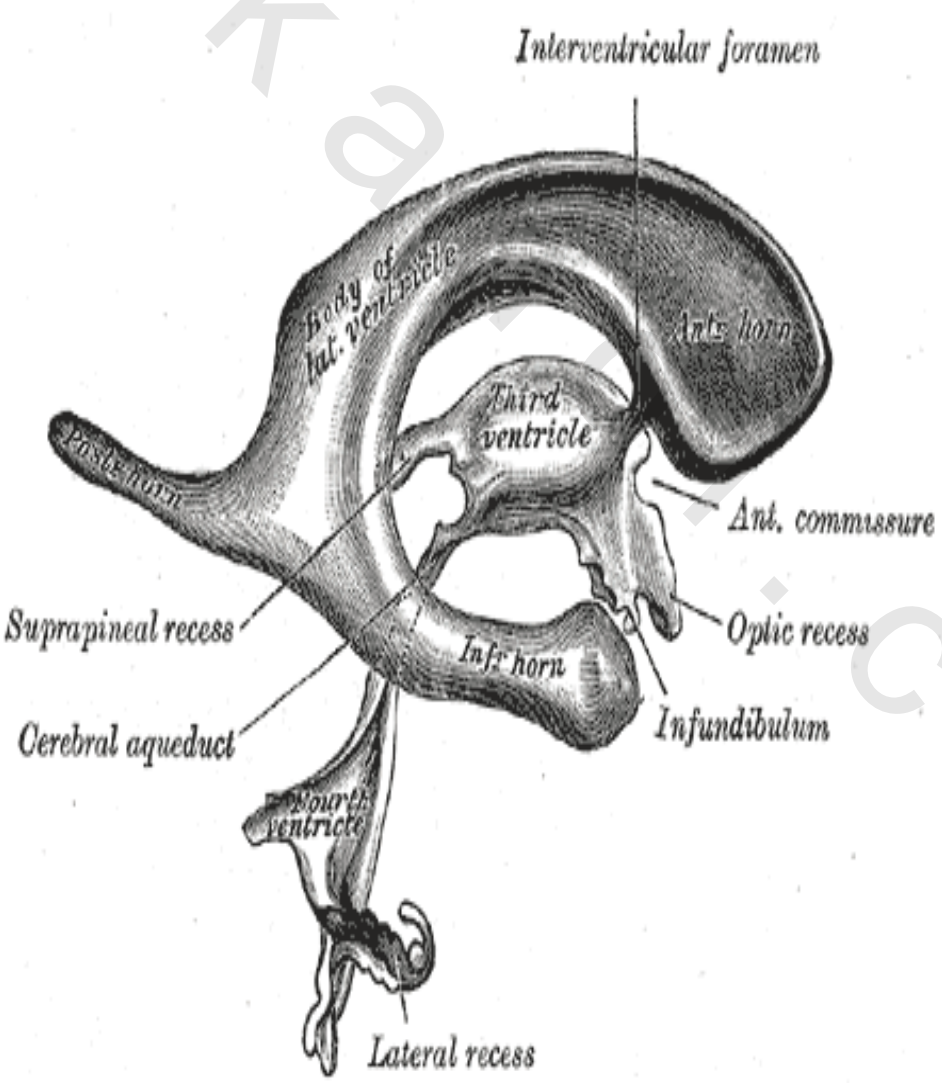

Figure (6): Anatomy of ventricular system. [8]

Tufts of capillaries invaginate the roofs of prosencephalon and rhombencephalon, forming the choroid plexuses of the ventricles. Cerebrospinal fluid (CSF) is secreted by the choroid plexuses, filling the ventricular system. CSF flows out of the fourth ventricle through the three apertures formed at the roof of the fourth ventricle by week 12 of gestation. [8]

Lateral ventricles

The largest cavities of the ventricular system are the lateral ventricles. Each lateral ventricle is divided into a central portion, formed by the body and atrium (or trigon), and three lateral extensions or horns of the ventricles. The central portion or the body of the ventricle is located within the parietal lobe. The roof is formed by the corpus callosum, and the posterior portion of the septum pellucidum lies medially. The anterior part of the body of the fornix, the choroid plexus, lateral dorsal surface of the thalamus, stria terminalis, and caudate nucleus, form the floor of the lateral ventricle. [9]

The interventricular foramen is located between the thalamus and anterior pillar of the fornix, at the anterior margin of the body. The two interventricular foramens (or foramina of Monro) connect the lateral ventricles with the third ventricle. The body of the lateral ventricle is connected with the occipital and temporal horns by a wide area named the atrium. [8, 9]

The anterior or frontal horn is located anterior to the interventricular foramen. The floor and the lateral wall are formed by the head of the caudate nucleus, the corpus callosum constitutes the roof and anterior border while the septum pellucidum delineates the medial wall. [8, 9]

The posterior or occipital horn is located within the occipital lobe. The fibers of the corpus callosum and the splenium form the roof. The forceps major is located on the medial side and forms the bulb of the occipital horn as seen in figure (7). [8, 9]

The inferior or temporal horn is located within the temporal lobe. The roof is formed by the fibers of the temporal lobe; the medial border contains the stria terminalis and tail of the caudate. The medial wall and the floor are formed by the hippocampus and its associated structures. The amygdaloid complex is located at the anterior end of the inferior horn. [9]

Third ventricle

The third ventricle is the narrow vertical cavity of the diencephalon. A thin tela choroidea supplied by the medial posterior choroidal arteries (branch of posterior cerebral artery) is formed in the roof of the third ventricle. The fornix and the corpus callosum are located superiorly. The lateral walls are formed by the medial thalamus and hypothalamus. The anterior commissure, the lamina terminalis, and the optic chiasm delineate the anterior wall. [8, 9]

The floor of the third ventricle is formed by the infundibulum, which attaches the hypophysis, the tuber cinereum, the mammillary bodies, and the upper end of the midbrain. The posterior wall is formed by the pineal gland and habenular commissure. The

interthalamic adhesions are bands of gray matter with unknown functional significance, which cross the cavity of the ventricle and attach to the external walls. [8, 9]

Fourth ventricle

The fourth ventricle is connected to the third ventricle by a narrow cerebral aqueduct. The fourth ventricle is a diamond-shaped cavity located posterior to the pons and upper medulla oblongata and anterior-inferior to the cerebellum. The superior cerebellar peduncles and the anterior and posterior medullary vela form the roof of the fourth ventricle. The apex or fastigium is the extension of the ventricle up into the cerebellum. The floor of the fourth ventricle is named the rhomboid fossa. The lateral recess is an extension of the ventricle on the dorsal inferior cerebellar peduncle. [9]

Inferiorly, it extends into the central canal of medulla. The fourth ventricle communicates with the subarachnoid space through the lateral foramen of Luschka, located near the flocculus of the cerebellum, and through the median foramen of Magendie, located in the roof of the ventricle. Most of the CSF outflow passes through the medial foramen. The cerebral aqueduct contains no choroid plexus. The tela- choroidea of the fourth ventricle, which is supplied by branches of the posterior inferior cerebellar arteries, is located in the posterior medullary velum as shown in figure (7).[8, 9]

Cerebrospinal fluid

CSF is a clear, watery fluid that fills the ventricles of the brain and the subarachnoid space around the brain and spinal cord. CSF is produced primarily by the choroid plexus of the ventricles (up to 70% of the volume), most of it being formed by the choroid plexus of the lateral ventricles. The rest of the CSF production is the result of transependymal flow from the brain to the ventricles.[10]

CSF flows from the lateral ventricles, through the interventricular foramens, and into the third ventricle, cerebral aqueduct, and the fourth ventricle. Only a very small amount enters the central canal of the spinal cord. [11]

CSF flow is the result of a combination of factors, which include the hydrostatic pressure generated during CSF production (known as bulk flow), arterial pulsations of the large arteries, and directional beating of the ependymal cilia. Hydrostatic pressure has a predominant role in the CSF flow within the larger ventricles, whereas cilia favor the movement of the CSF in the narrow regions of the ventricular system, such as the cerebral aqueduct. Immotile cilia syndrome is a rare cause of hydrocephalus in children. [11]

The ventricles constitute the internal part of a communicating system containing CSF. The external part of the system is formed by the subarachnoid space and cisterns. The communication between the two parts occurs at the level of fourth ventricle through the median foramen of Magendie (into the cistern magna) and the two lateral foramina of Luschka (into the spaces around the brainstem cerebellopontine angles and prepontine cisterns). The CSF is absorbed from the subarachnoid space into the venous blood (of the sinuses or veins) by the small arachnoid villi, which are clusters of cells projecting from subarachnoid space into a venous sinus, and the larger arachnoid granulations.[11]

The total CSF volume contained within the communicating system in adults is approximately 150 ml, with approximately 25% filling the ventricular system. CSF is produced at a rate of approximately 20 ml/h, and an estimated 400-500 ml of CSF is produced and absorbed daily. [11]

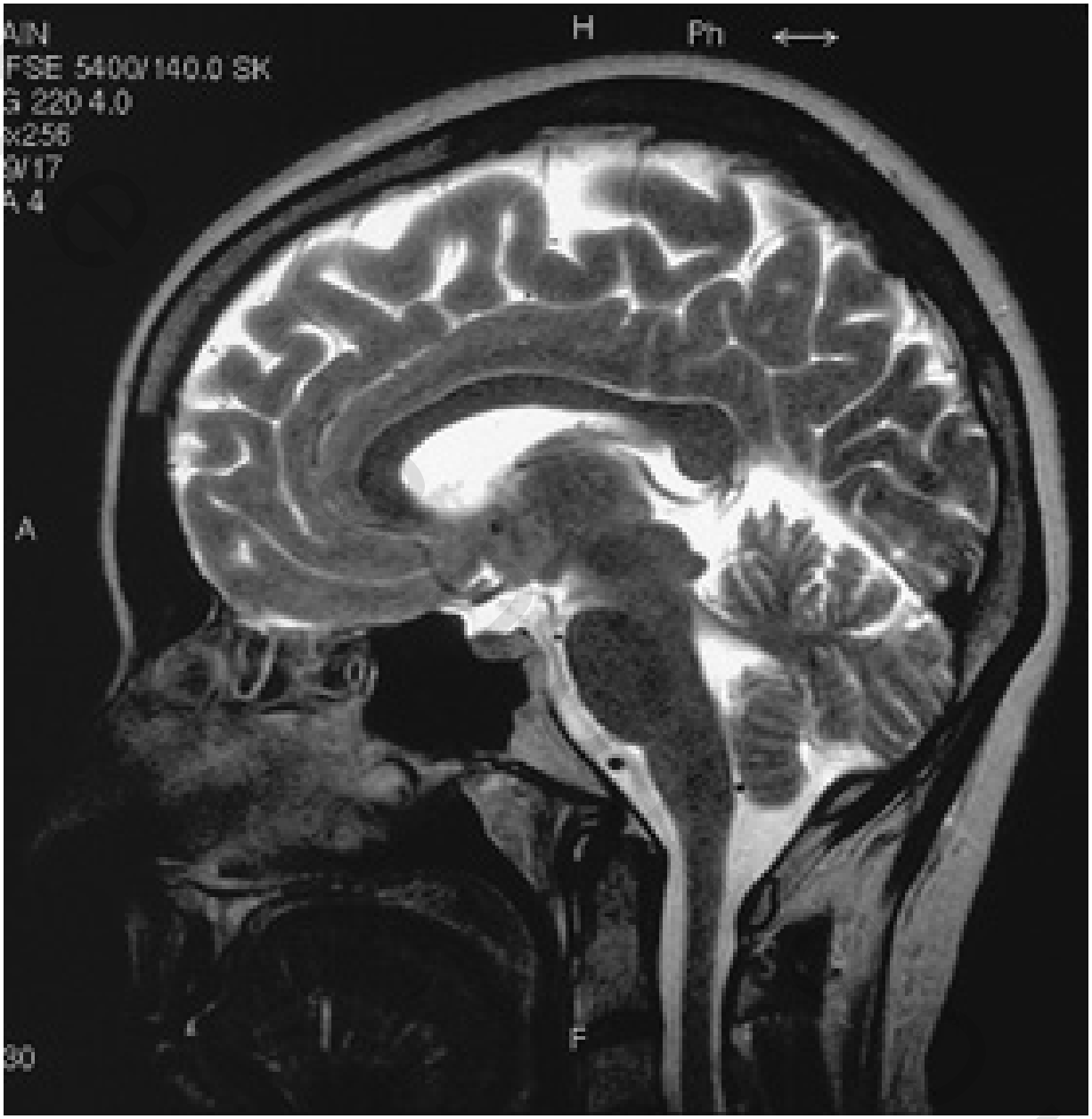

Figure (7): T2WI MRI of the ventricular system. [9]

CSF absorption capacity is normally approximately 2-4 times the rate of production. The normal CSF pressure is between 5-15 mm Hg (65-195 mm H_2 O) in adults. In children younger than 6 years, normal CSF pressure ranges between 10-100 mm H_2 O. [11]

CSF plays an important role in supporting the brain growth during evolution, protecting against external trauma, removal of metabolites produced by neuronal and glial cell activity, and transport of biologically active substances (like hormones and neuropeptides) throughout the brain, figure (8).[11]

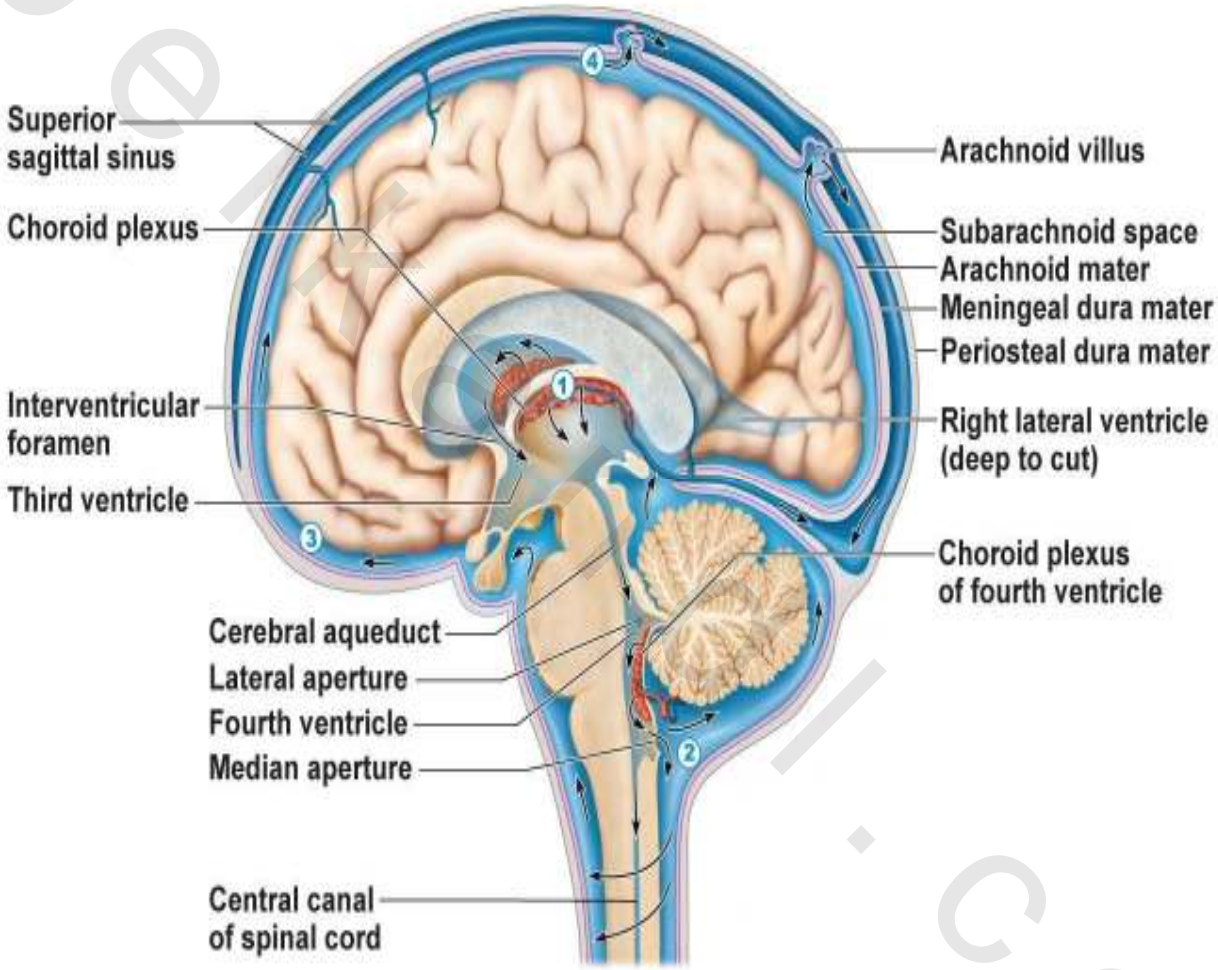

Figure (8): CSF circulation. [11]

Hydrocephalus

Definition

Hydrocephalus is defined as a neurological disorder that is caused by a disruption in the balance between the formation, flow, or absorption of CSF in the brain, resulting in an increase in the volume that the CSF occupies in the CNS, figure (9). [11]

Incidence

Hydrocephalus is one of the most common birth defects. In the U.S., the incidence of congenital hydrocephalus is believed to be approximately 200 cases per 100,000 live births. In addition, approximately 6,000 children annually develop acquired hydrocephalus during the first two years of life. Overall incidence of acquired hydrocephalus is not known.[12]

There are two peaks associated with the relationship between incidence and age of onset of hydrocephalus. First peak occurs in infancy and is associated with a number of congenital malformations. Second peak occurs in adulthood and is mostly associated with normal pressure hydrocephalus. Approximately 60% of the total cases of hydrocephalus are congenital or acquired in childhood. Hydrocephalus is a common and significant pediatric problem. [12]

In general, there is no difference in incidence between males and females. One exception is Bickers-Adams syndrome, which is an X-linked recessive hydrocephalus that is manifested only in males and affects approximately 1/30,000 males at birth. Normal pressure hydrocephalus is slightly more common in males. [12]

Pathogenesis

Hydrocephalus is usually due to blockage of cerebrospinal fluid (CSF) outflow in the ventricles or in the subarachnoid space over the brain. Normally CSF continuously circulates through the brain, its ventricles and the spinal cord and is continuously drained away into the circulatory system. Alternatively, the condition may result from an overproduction of the CSF, from a congenital malformation blocking normal drainage of the fluid, or from complications of head injuries or infections.[13]

Compression of the brain by the accumulating fluid eventually may cause neurological symptoms such as convulsions, mental retardation and epileptic seizures. These signs occur sooner in adults, whose skulls are no longer able to expand to accommodate the increasing fluid volume within. Fetuses, infants, and young children with hydrocephalus typically have an abnormally large head, excluding the face, because the pressure of the fluid causes the individual skull bones which have yet to fuse to bulge outward at their junction points.[13]

The elevated intracranial pressure may cause compression of the brain, leading to brain damage and other complications. [14]

If the foramina of the fourth ventricle or the cerebral aqueduct are blocked, cerebrospinal fluid (CSF) can accumulate within the ventricles and it results in increased

CSF pressure. The production of CSF continues, even when the passages that normally allow it to exit the brain are blocked. Consequently, fluid builds inside the brain, causing pressure that dilates the ventricles and compresses the nervous tissue. Compression of the nervous tissue usually results in irreversible brain damage. If the skull bones are not completely ossified when the hydrocephalus occurs, the pressure may also severely enlarge the head. The cerebral aqueduct may be blocked at the time of birth or may become blocked later in life because of a tumor growing in the brainstem. (14)

Classification and etiology

Hydrocephalus can be classified into congenital or acquired. Congenital causes include genetic causes like aqueductal abnormalities, chiari malformation (types I and II), congenital infections (e.g., cytomegalovirus, toxoplasmosis, rubella), Dandy-Walker syndrome, spina bifida and neural tube defects (NTDs), cerebral hemorrhage and neurofibromatosis. An unusually large head is the main sign of congenital hydrocephalus.(15)

Acquired hydrocephalus can occur at any age. Causes include, head injuries, strokes, infections, tumors, and brain hemorrhage. (15)

Also can be classified into communicating hydrocephalus occurs when full communication occurs between the ventricles and subarachnoid space. It is caused by defective absorption of CSF, rarely by overproduction of CSF, or occasionally by venous drainage insufficiency. (15)

Non- communicating hydrocephalus occurs when CSF flow is obstructed within the ventricular system or in its outlets to the arachnoid space resulting in impairment of the CSF from the ventricle to the subarachnoid space. The most common form of non-communicating hydrocephalus is obstructive and is caused by intraventricular or extra ventricular mass-occupying lesions that disrupt the ventricular anatomy. (15)

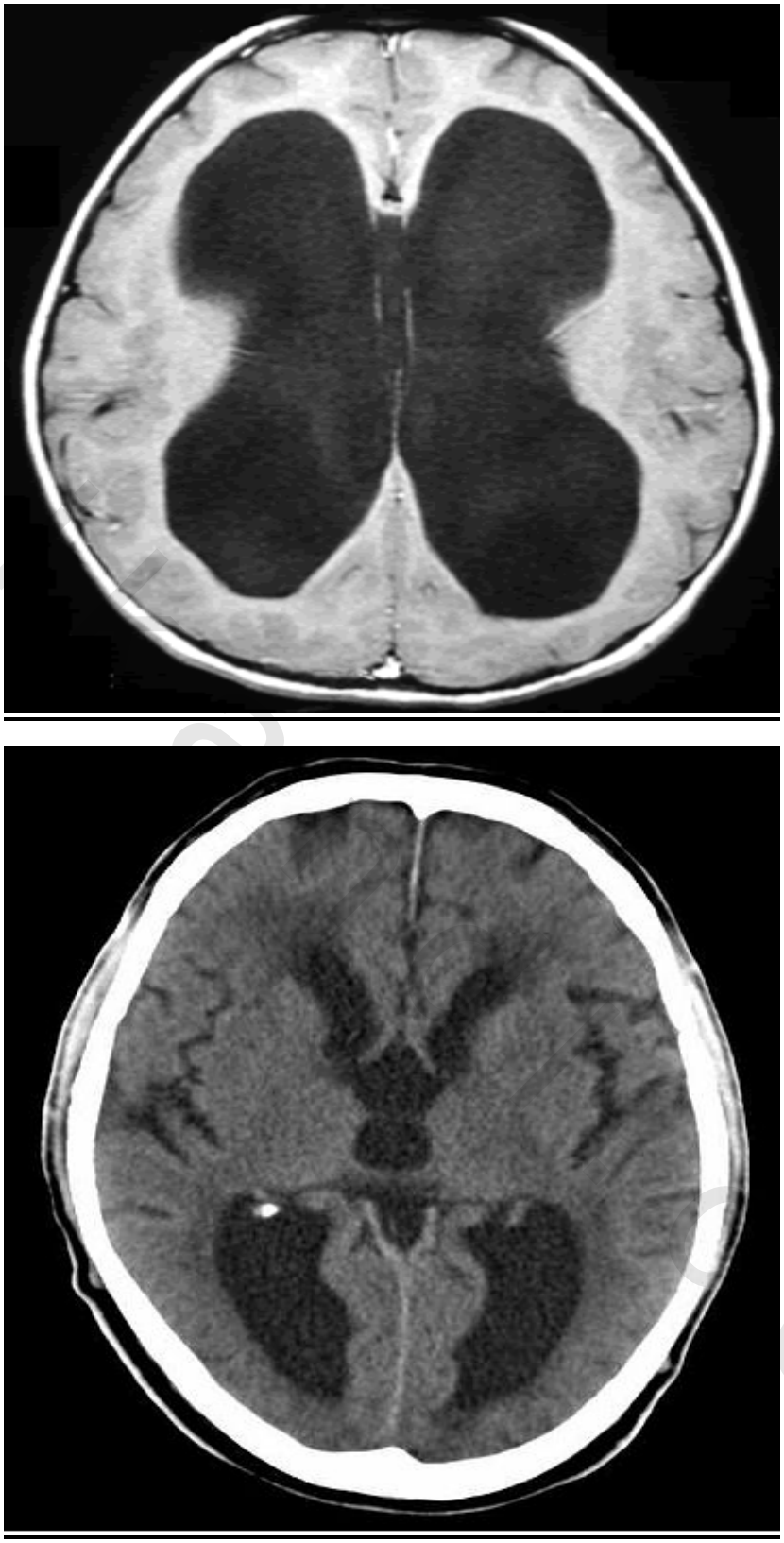

Figure (9): MRI &CT axial images demonstrating dilated ventricles. [11]

Clinical presentation

The clinical presentation of hydrocephalus varies with chronicity. The symptoms depend on the cause of the blockage, the person's age, and extent of brain damage. Acute dilatation of the ventricular system is more likely to manifest by the nonspecific signs and symptoms of increased intracranial pressure. By contrast chronic dilatation (especially in the elderly population) may have a more insidious onset presenting, for instance, with Hakim's triad (Adams triad). [15]

Symptoms of increased intracranial pressure may include headaches, vomiting, nausea, papilledema, sleepiness or coma. Elevated intracranial pressure may result in uncal and/or cerebellar tonsill herniation, with resulting life-threatening brain stem compression.[15]

Hakim's triad of gait instability, urinary incontinence and dementia is a relatively typical manifestation of the distinct entity named normal pressure hydrocephalus (NPH). Focal neurological deficits may also occur, such as abducent nerve palsy and vertical gaze palsy (Parinaud syndrome due to compression of the quadrigeminal plate, where the neural centers coordinating the conjugated vertical eye movement are located). [15]

In infants with hydrocephalus, CSF builds up in the central nervous system, causing the fontanels to bulge and the head to be larger than expected. [15]

Early symptoms may also include, eyes that appear to gaze downward, irritability, seizures, separated sutures, sleepiness &vomiting. [15]

Symptoms that may occur in older children can include high-pitched cry**,** changes in personality, changes in facial appearance and eye spacing**,** crossed eyes or uncontrolled eye movements**,** difficult feeding**,** excessive sleepiness**,** headache**,** irritability, poor temper control**,** urinary incontinence**,** loss of coordination and trouble walking**,** muscle spasm**,** slow growth (child 0–5 years), slow or restricted movement& vomiting. [15]

Management

Diagnosis

Hydrocephalus is diagnosed through clinical neurological evaluation and by using cranial imaging techniques such as ultrasonography, computed tomography (CT), magnetic resonance imaging (MRI), or pressure-monitoring techniques. [16]

A physician selects the appropriate diagnostic tool based on an individual's age, clinical presentation, and the presence of known or suspected abnormalities of the brain or spinal cord. [16]

Role of imaging

Brain imaging techniques can show enlargement of the ventricles caused by excess cerebrospinal fluid. They may also be used to identify underlying causes of hydrocephalus or other conditions contributing to the symptoms. Imaging techniques may include: [16]

Ultrasound

It uses high-frequency sound waves to produce images, is often used for an initial assessment for infants because it's a relatively simple, low-risk procedure. In the newborn infant with an open fontanel, sonography at the bedside can demonstrate the ventricular size and large subdural collections. Insonation through the mastoid can image the posterior fossa and rule out 4th ventricular masses. (16)

Computerized tomography (CT)

(CT) produces cross-sectional views of the brain. This is the imaging modality of choice for screening for hydrocephalus. It is relatively inexpensive and gives sufficient details to rule out most tumours which might obstruct the ventricular system. (16)

Magnetic resonance imaging (MRI)

MRI is good modality to image anatomical changes and can also add support for the diagnosis with CSF flowmetry studies and MR spectroscopy, findings include:

- Ventriculomegaly
  - Frontal and temporal horns of the lateral ventricles most affected
  - Upward bowing of corpus callosum
- Crowding of the gyri at the vertex(with small sulci)
- Sylvian fissures out of proportion to sulcal enlargement (which is minimal)and hippocampus and mesial temporal lobe volumes(which are near normal)
- Aqueductal flow void, best seen on T2spin echo sequences
- Periventricular high signal on T2 weighted sequence (periventricular permeation).
- MR spectroscopy – lactate peak in lateral ventricles
- CSF flow studies
  - Increased aqueductal stroke volume
  - Increased aqueductal peak velocity
  - Various publications have set various normal and abnormal ranges
    - Flow rate of>24.5ml/min 95% specific for NPH
    - Stroke volume of 42 microl predicts good response from shunting. (16)

Treatment

Hydrocephalus treatment is surgical, generally utilizing various types of cerebral shunts. It involves the placement of a ventricular catheter (a tube made of silastic), into the cerebral ventricles to bypass the flow obstruction/malfunctioning arachnoidal granulations and drain the excess fluid into other body cavities, from where it can be resorbed.[17]

Ventriculo peritoneal shunt (mechanism of action)

A shunt is a narrow, soft and pliable piece of tubing (approximately 0.25 cm. in diameter) which is surgically implanted into the ventricle through a small hole made in the skull. All shunts have a valve system which regulates the pressure of the cerebrospinal fluid and prevents backward flow of fluid into the ventricles. It opens automatically when the pressure exceeds a certain level (usually called the 'opening pressure' of the shunt) and allows CSF to drain. The valve closes again when the pressure returns to the normal level.[17]

Shunt has three basic components:

1. A catheter (or tube) which is inserted into the brain ventricles
2. A valve which regulates the flow of spinal fluid.
3. A long catheter which carries the CSF from the head to wherever the CSF is being diverted (the peritoneal or chest cavity, the jugular vein, etc.), figure (10).[18]

Most shunts drain the fluid into the peritoneal cavity (ventriculo-peritoneal shunt), but alternative sites include the right atrium (ventriculo-atrial shunt), pleural cavity (ventriculo-pleural shunt), and gallbladder. A shunt system can also be placed in the lumbar space of the spine and have the CSF redirected to the peritoneal cavity (Lumbar-peritoneal shunt). [17]

There are many different types of shunts, each with different pressure specifications, flow characteristics, and options for external adjustment. These characteristics can be matched to best solve each case. [18]

To allow for the possibility to test the shunt, a tapping or flushing chamber can be incorporated in it. This allows a neurosurgeon to test some aspects of the shunts workings.[18]

Complications of VP shunts

Include shunt malfunction, shunt failure, and shunt infection which is the most common reason for shunt failure. Although a shunt generally works well, it may stop working if it disconnects, becomes blocked (clogged) or infected. If this happens the cerebrospinal fluid will begin to accumulate again and a number of physical symptoms will develop (headaches, nausea, vomiting, photophobia, some extremely serious, like seizures. The shunt failure rate is also relatively high (of the 40,000 surgeries performed annually to

treat hydrocephalus, only 30% are a patient's first surgery), and it is not uncommon for patients to have multiple shunt revisions within their life time. [19]

Other shunt complication may include the shunt system draining fluid at the wrong rate. Over drainage of the ventricles can cause the ventricle to decrease in size to the point where the brain and its meninges pull away from the skull or the ventricles become slit like. If blood from sheared vessels in the meninges becomes trapped between the brain and the skull, resulting in a subdural hematoma, further surgery is required. This is most common in older adults with normal pressure hydrocephalus (NPH). [19]

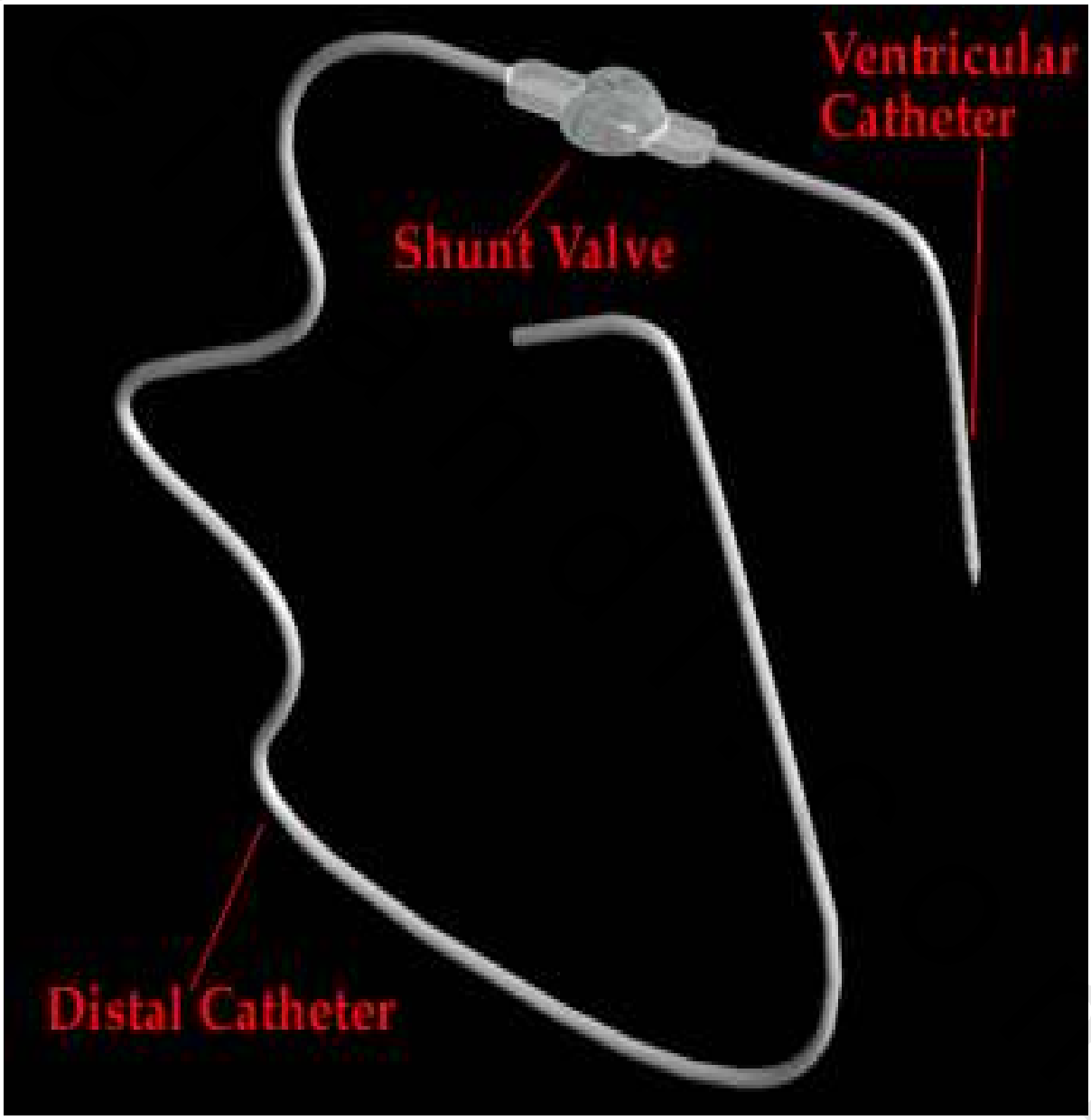

Figure (10): Parts of VP shunt. [18]

Endoscopic third ventriculostomy (ETV)

(ETV) is considered as a treatment of choice for obstructive hydrocephalus. [20]

History of ETV

Ventriculoscopy was introduced in the early 1900s. Walter E. Dandy used a primitive endoscope to perform choroid plexectomy in communicating hydrocephalus. He later introduced the sub-frontal approach for an open third ventriculostomy. [20]

An advent of valve-regulated shunt systems and the simplicity of the shunt technique resulted in minimal advances in third ventriculostomies for next 30 years. In 1947, H. F. McNickle introduced a percutaneous method of performing third ventriculostomy that decreased the complication rate and improved the success rate. [20]

We now have small neuroendoscopes with deflectable tips, working ports, and good optic resolution, in addition to the rigid endoscopes with their excellent optic resolution. High definition camera has further improved visualizing and recording. [20]

An improvement in the success of third ventriculostomy in recent time could be due to better patient selection; improvements in endoscope, better imaging, advanced surgical technique and instruments. [20]

Indications of ETV

It is indicated in:

- Hydrocephalus secondary to congenital aqueductal stenosis

Aqueductal stenosis is a common cause of obstructive (non-communicating) hydrocephalus. It could be due to congenital stenosis, acquired idiopathic, post-infectious or post-hemorrhagic and stenosis secondary to tumor. In congenital stenosis, aqueduct could be narrow or completely obstructed. Complete or near complete obstruction usually present in an early age while if obstruction is partial, the patients may be asymptomatic or present at a later age. Endoscopic third ventriculostomy instead of a shunt placement is considered better option for non-communicating hydrocephalus secondary to congenital aqueductal stenosis. Results of ETV are better in congenital hydrocephalus due to aqueductal stenosis as compared to post-hemorrhagic or post-infective hydrocephalus, figure (11). [20]

- Dandy-Walker malformation

The Dandy-Walker syndrome (DWS) is associated with the posterior fossa enlargement, partial or complete agenesis of the cerebellar vermis, and cystic dilation of the fourth ventricle. This often involves hydrocephalus and complications due to associated genetic conditions, such as spina bifida. Hydrocephalus can be treated by shunt surgery, but the ETV was found to be a preferential operative procedure. Endoscopic third ventriculostomy alone, ETV with aqueductal stent placement and ETV with fenestration of the occluding membrane could be performed effectively and safely depending on existing pathology in DWS. [21]

- Vein of Galen aneurysm

Aneurysms of the vein of Galen are uncommon vascular malformations and are frequently seen in infants and children, leading to heart failure and hydrocephalus. An obstruction of the cerebral aqueduct by an aneurysm is one of the causes of hydrocephalus. Such hydrocephalus has been treated mainly with cerebrospinal shunt procedures. ETV has been found to be successful in huge hydrocephalus in vein of Galen aneurism. [22]

- Syringomyelia with or without Chiari malformation type I

Chiari malformations refer to a spectrum of congenital hindbrain abnormalities affecting the cerebellum, brainstem, the upper cervical cord and the bony cranial base. Hydrocephalus occurs in 7% - 10% of patients with Chiari I malformation (CIM). Such patients can develop syringomyelia. Hydrocephalus and syringomyelia are commonly managed by shunt surgery. Syringomyelia and hydrocephalus with or without (C1M) can be successfully treated by ETV. [23, 24]

- Shunt malfunction

Shunt malfunction is quite common, and some of them could have repeated malfunctions. Such patients with repeated block could be better managed by ETV. Formerly shunted patients have less favorable results, somewhat greater risk of serious complications; therefore, more experience is essential when offering them an ETV. Carefully selected patients with obstructive hydrocephalus could benefit from ETV after shunt malfunction, and about 70% become shunt free. [26]

Other indications include: posterior third ventricle tumor, cerebellar infarct, intraventricular hematoma, post infective, normal pressure hydrocephalus, myelomeningocele, multiloculated hydrocephalus, posterior fossa tumor and craniosynostosis. [20]

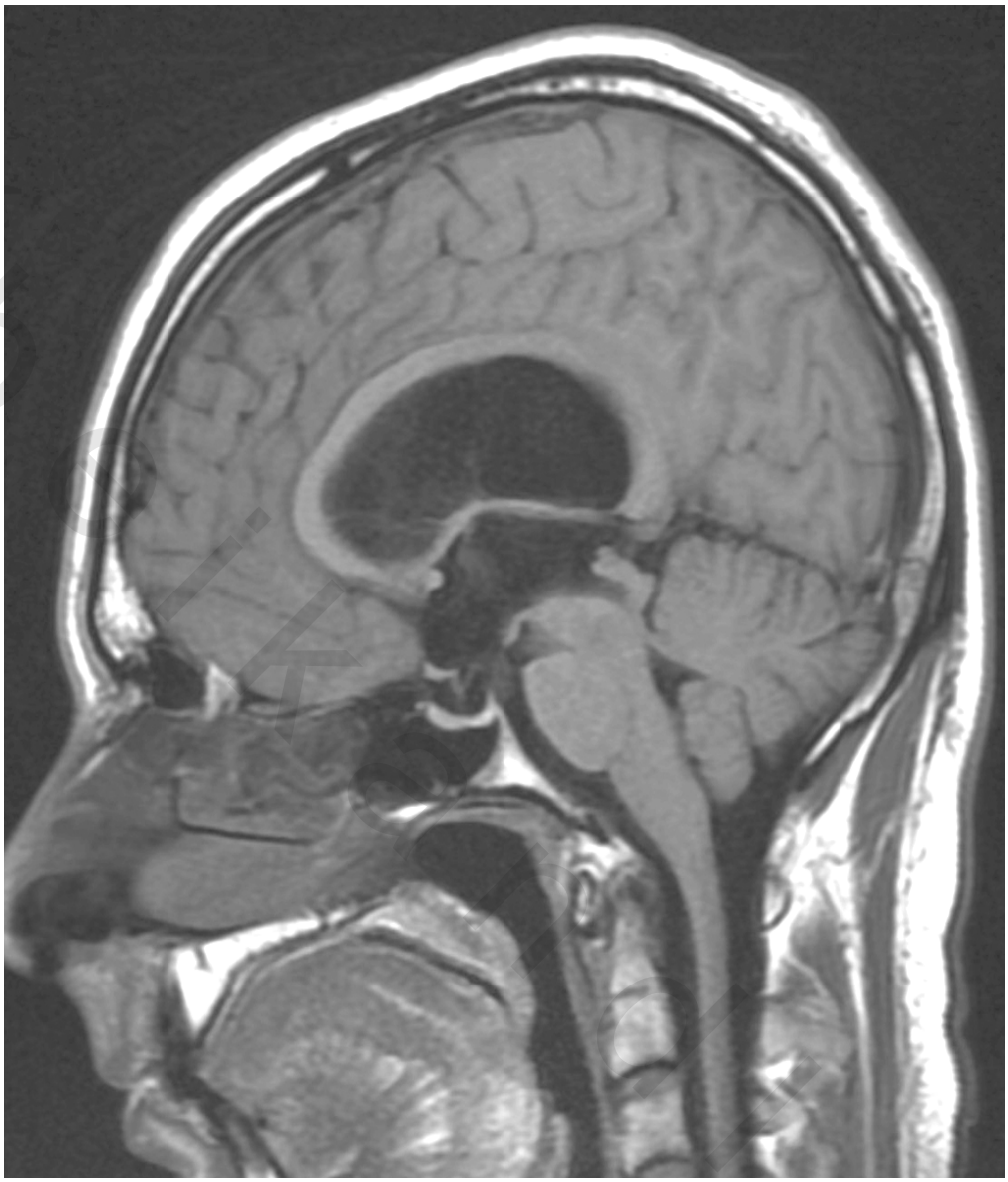

Figure (11): MRI T1W image showing aqueduct stenosis. [20]

Surgical Technique

ETV is performed in supine position with head flexed so that the burr hole site is at the highest point. This also avoids over drainage of CSF and an entry of air in the ventricles and subdural space, especially in large ventriculomegaly. Loss of CSF may also be a risk factor for post-operative subdural hematoma. Some authors even prefer semi sitting position. It is important to note that the size of the lateral ventricles, foramen of Monro and third ventricle should be sufficiently large to allow an introduction of the endoscope and to navigate it into the third ventricle. Generally, the width of the third ventricle and foramen of Monro should be approximately 7 mm or greater. If a patient has slit ventricles, caused by over shunting of CSF, it will first be necessary to externalize the shunt to control drainage until an adequate working diameter of the ventricles is achieved. Stereotactic guidance can be used as a surgical adjunct to access ventricle. [27]

The optimal trajectory into the third ventricle through foramen of Monro and into the interpeduncular cistern is usually achieved with burr hole, placed at or just anterior to the coronal suture and about 2.5 cm – 3 cm lateral to the midline. Usually, right side burr hole is performed. An exact site of burr hole can be determined by a line, extending from interpeduncular cistern and foramen of Monro on to the skull. [27]

Foramen of Monro can be identified by confluence of thalamo-striate vein, septal vein and choroid plexuses. Perforation in the third ventricle floor is made after negotiating endoscope through the foramen of Monro. Fenestration in the third ventricle floor should be in between mammillary bodies and infundibular recess, at the most transparent site. Location of basilar artery should be identified to avoid an injury and bleeding during procedure, and the fenestration should be made anterior to the artery complex. Microvascular Doppler probe inserted through the endoscope could be useful to locate artery if basilar artery is not seen. Position of dorsum sellae can be identified by gentle probing by the blunt instrument, such as bipolar forceps if the facility of doppler is not available. Fenestration should be made just posterior to dorsum sellae. Water jet dissection technique can be used to prevent an injury to vessel and bleeding if the third ventricle floor is thick and opaque. [27]

If any hemorrhaging is encountered during the procedure, copious warm fluid irrigation should be used until all bleeding is visibly stopped and the ventricular CSF is clear. [27]

We use careful intermittent closure of outflow channel to create tamponed effect. This helps in better visualization during bleeding. An external ventricular drain is kept if there was any oozing of blood. Some authors use reservoir routinely, which facilitates management and diagnosis if patients are suspected to have persistent elevation of ICP or block stoma. Ommaya reservoir in certain high-risk patients may be a useful option for achieving quick ventricular access by medical and non-medical personnel in case of deterioration after ETV. Any bleeding, from cortical surface if seen while the endoscope is removed, should be cauterized. [27]

Post-operative Imaging

A decrease in the peri ventricular haze is detected soon after successful endoscopic third ventriculostomy is associated with a satisfactory clinical outcome. This response continues during the first few months after surgery. The reduction in ventricular size is more prominent in acute forms of hydrocephalus. Reduction of the size of third ventricle width was more than the reduction in lateral ventricle size after successful ETV. This easily can be detected by doing CT scan after procedure. [28]

Success Rate of ETV and Age

There are controversies regarding the success of ETV in infants. Some authors found that the ETV success do not depend on an age of the patient. Others reported poor results, especially in neonates and in infants, younger than 2 months. [29]

It was reported that there is relative higher risk of initial failure in ETV, than shunt in children. The relative risk becomes progressively lower for ETV after about 3 months. Patient could experience a long-term treatment survival advantage after an early high-risk period of ETV failure as compared to shunt. They observed that it might take several years, however, to realize this benefit. [29]

Complications

Over all complication rate after ETV is about 2% - 15%, including. [30]

Intra-operative Complications

Intra-operative neural injury, such as thalamic, forniceal, hypothalamic and midbrain injuries are also observed. Intra-operative bradycardia and hemorrhages including fatal hemorrhage due to basilar artery rupture are also reported. Attempts to perforate the ventricular floor can lead to bleeding, especially in hydrocephalus following an infection and hemorrhage. [30]

Complication Avoidance

Forniceal injury and other neural injuries could be avoided by proper planning of burr hole, avoiding significant side movements and by selecting proper cases with significantly enlarged foramen of Monro and third ventricle. Per-operative bleeding should be avoided by using water jet dissection in thick and opaque third ventricle, avoiding significant stretching of structure, especially during perforation of tough third ventricle floor. Significant side movement should also be avoided to prevent bleeding due to an injury to structures, like fornix and veins at foramen of Monro. Rarely, blood might trickle from burr hole site into the ventricle; proper hemostasis must be achieved before entering the ventricle. Proper inspection is must before making perforation in the third ventricle floor; fenestration on the vessel must be avoided. Bradycardia due to raised ICP could be avoided by keeping outflow patent. Bradycardia due to stretching on the brain stem should be avoided, especially during perforation of tough third ventricle floor. [30]

Early Complications

Central nervous system infections, fever, stoma block, CSF leak and post-operative intracranial hematomas were also seen. Post-operative mortality is also reported. Diabetes insipidus, weight gain, precocious puberty and abnormal prolactin levels were found after ETV. However, abnormal prolactin levels were not clinically significant in the patients. It remained inconclusive whether ETV contributes to the abnormalities of prolactin levels or to other endocrine parameters in pediatric patients. They suggested the longitudinal studies to delineate the effect of ETV on endocrine regulation. [31, 32]

Chronic subdural hematoma or subdural hygroma can occur after ETV, especially after Ommaya reservoir. Rare complications like post-operative hyperkalemia, severe Parkinsonism, acute respiratory alkalosis and tachypnea can occur after endoscopic third ventriculostomy. [33]

Complication Avoidance

Post-operative CSF leak could be avoided by plugging cortical and dural opening by gel foam, direct dural closure, especially in large ventriculomegaly in infants, or by using artificial dural substitute and tissue sealant in at risk patients. Post-operative CSF leak can also be reduced by galeal-pericranial flap. Subdural hematoma could be prevented by making burr hole at highest point by flexing the neck, proper hemostasis at burr hole site and by preventing brain collapse using constant fluid irrigation. [34, 35]

Delayed Complications

Most of the complications in the ETV patients occur within 4 weeks. However, delayed complications including stoma block can occur, and therefore, a longer follow-up is desirable. Delayed stoma block though very rare, can be fatal. Patients who undergo ETV for an infective hydrocephalus and Dandy-Walker malformation should receive long-term follow-up because late closure of the stoma may occur in these patients. Intra-operative observation of thickened arachnoid membranes at the level of the interpeduncular cisterns at the time of ETV and a progressive decrease in CSF flow through the stoma on cine MR imaging should be considered a significant risk of deterioration.[36]

The total hospital stay is two days for the majority of patients. A postoperative computerized tomography scan is obtained prior to discharge. The patient should be free of headache and ambulatory at discharge. In infants, the fontanel should be soft and sunken while the patient is in an upright position. The ventricular drain is weaned in patients in whom shunts have previously been placed to verify shunt independence. The drain is then removed and the patient is observed to ensure patency of the third ventriculostomy. [37]

Although endoscopic third ventriculostomy can ideally lead to the much-desired result of a shunt-free life, doctors caution that this procedure is not appropriate for everyone. Still, for those who meet the criteria, endoscopic third ventriculostomy offers the possibility of freedom from shunt dependency. [38]

Post-operative Care

ETV results are good in obstructive hydrocephalus. Post-operative failures usually occur early; regular clinical and radiological assessment must be performed, especially in the first years after the neuroendoscopic procedure. [39]

Measurement of intracranial pressure during the immediate post-operative period after endoscopic third ventriculostomy could be required in patients who continue to have clinical features of raised ICP or failed to show an improvement after ETV. [40, 41]

Post-operative continuous ICP monitoring could be very useful in the ETV after shunt failure, it could provide an accurate assessment of the success of the ETV and patency of the stoma in the early post-operative days using CT ventriculography. [42, 43]

Some patients fail to show an improvement despite of patent stoma after ETV. This could be due to complex hydrocephalus (combination of obstructive and communicating). Lumbar puncture helps by increasing the compliance and the buffering capacities of the spinal subarachnoid spaces, it probably decreases the cerebrospinal fluid outflow resistance from the ventricular system, facilitate the decrease in the ventricular volume and allowing faster permeation of CSF in the intracranial subarachnoid spaces. A cycle of 1 to 3 lumbar punctures should always be performed in patients who remain symptomatic after ETV, before ETV is assumed to have failed and an extra cranial cerebrospinal fluid shunt is implanted. Patients with temporary defect in CSF hydrodynamics would show clinical improvement after lumbar puncture while those with permanent defect in CSF absorption or permeation will need some form of shunt. [44, 45]

Endoscopic third ventriculostomy versus cerebrospinal fluid shunt in the treatment of hydrocephalus

Advantages of Endoscopic third ventriculostomy

- No foreign object (shunt tubing and valve) implanted in the body, lowering the risk of infection.
- Fewer incisions mean slightly less discomfort.
- A lower long term complication rate compared to a shunt. [46]