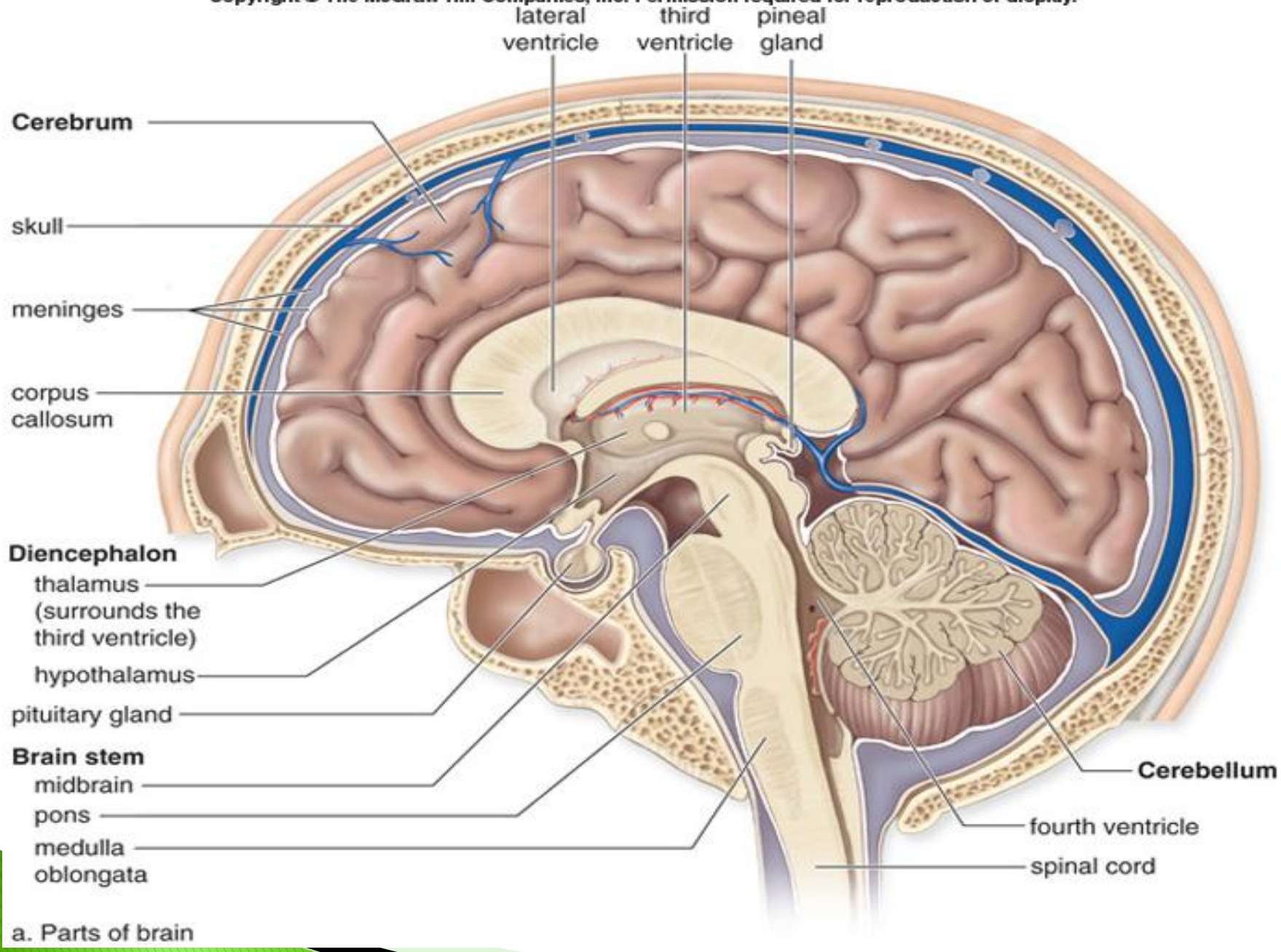
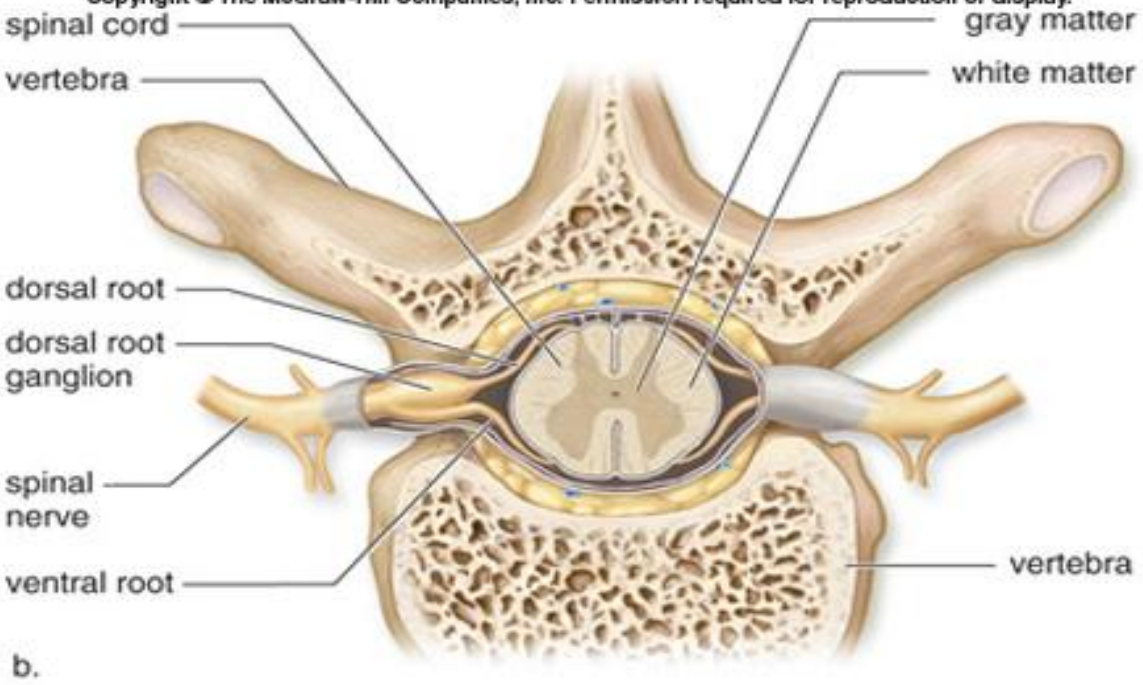
DISEASES OF THE CENTRAL NERVOUS SYSTEM
(B: Surgical diseases)



a. Parts of brain



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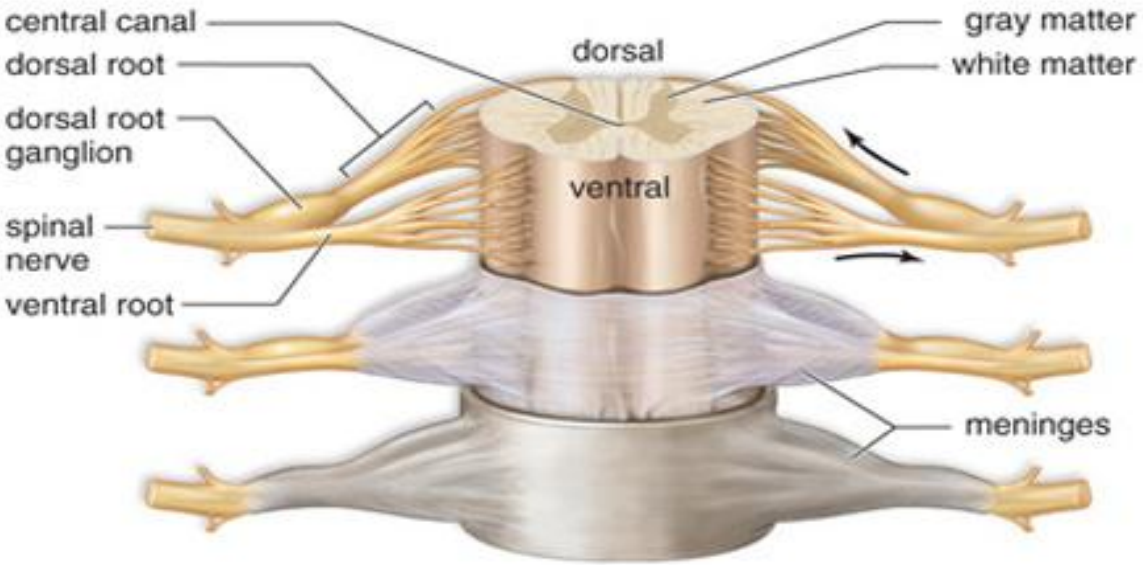

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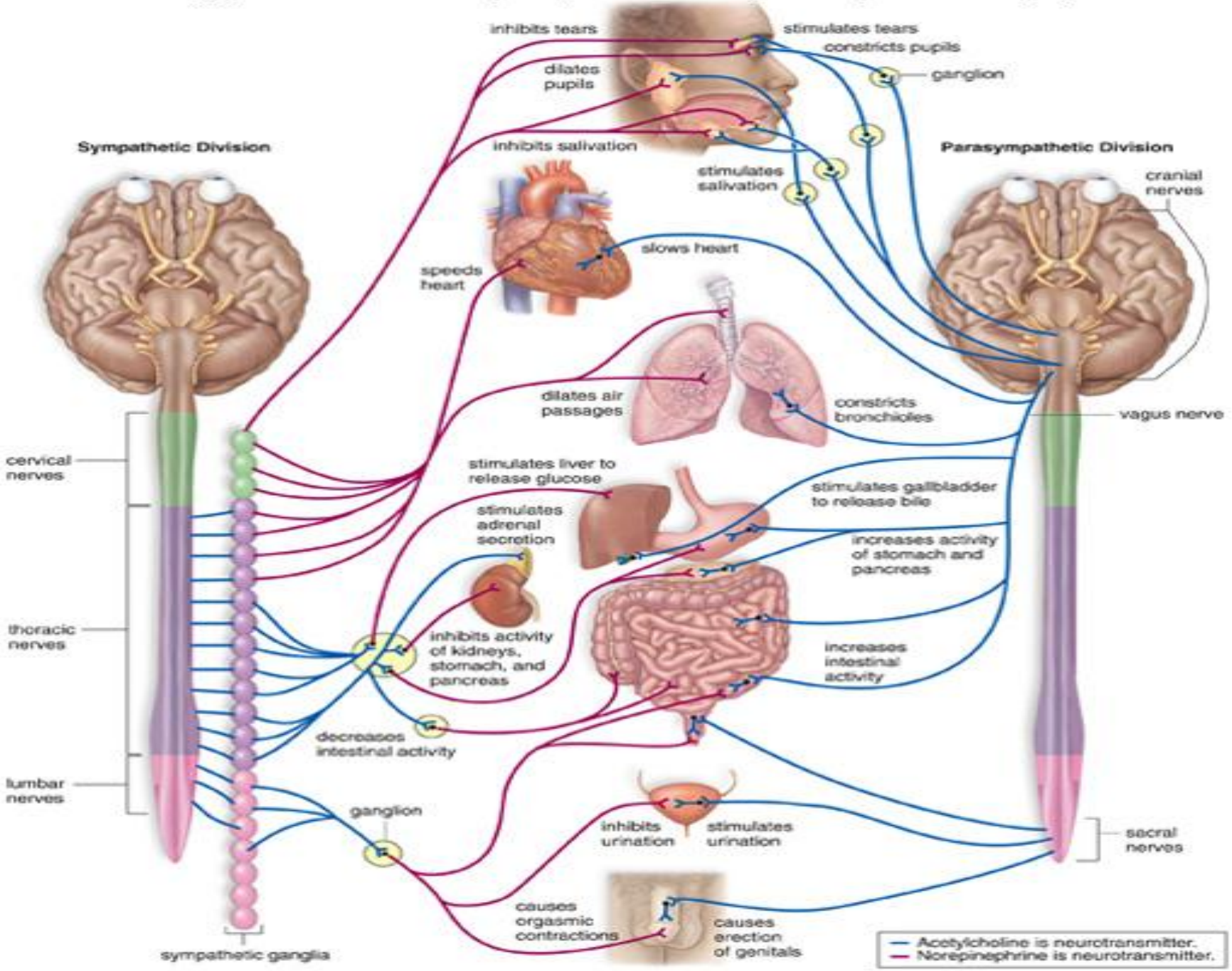


VASCULAR DISEASES OF NERVOUS SYSTEM

- The term denotes any brain abnormality caused by pathological process involving the blood vessels (thrombosis, embolism, or hemorrhage).
- **Stroke**: Abrupt onset of global neurological symptoms due to ischemia or hemorrhage with symptoms more than 24 hours.
- If symptoms resolve in less than 24 hours, the term transient ischemic attack (TIA) is applied.

Cerebral infarction

- **Causes:**
  1. Arterial occlusion mainly due to atherosclerosis with thrombosis of internal carotid, cerebral, vertebral and basilar arteries.
  2. Embolism from left side of the heart.
  3. Venous thrombosis associated with pregnancy and oral contraceptive pills.

- The commonest sites of thrombosis are: basal ganglia, internal capsule and cerebral cortex.
- Cerebral infarction is an example of liquefactive necrosis, it is changed into a fluid filled cavity surrounded by glial tissue.

INTRACRANIAL HEMORRHAGE

A. **Cerebral hemorrhage:**

- Mostly spontaneous (non-traumatic) hemorrhage.
- Accounts for 10-20% of all strokes.
- More in males.

- **Location:** basal ganglion and thalamus (60%), cerebral (20%), cerebellum (13%) and pones (7%).

- **Causes**:
  1. **Hypertension**: is the most common cause of non-traumatic hemorrhage due to:
- Hyaline arteriolar sclerosis makes the arteriolar wall weaker and more vulnerable to rupture.
- The development of minute aneurysms in the cerebral vessels (***Charcot Bouchard micro- aneurysms***).

2. **Other causes**:
    a. Rupture of cerebral aneurysm: Berry (saccular) aneurysms.
    b. Neoplasms: bleeding into a degenerated tumor.
    c. Hemorrhagic blood diseases: blood dyscrasias e.g., sickle cell anemia.
    d. Vasculitis.
    e. Trauma.

- **Morphology**:
  1. **Peticheal**: multiple small hemorrhages in the brain substance follows septicemia, hemorrhagic blood diseases, and vitamin B or C deficiency.
  2. **Massive**: extensive area of hemorrhages with destruction of the brain tissue.

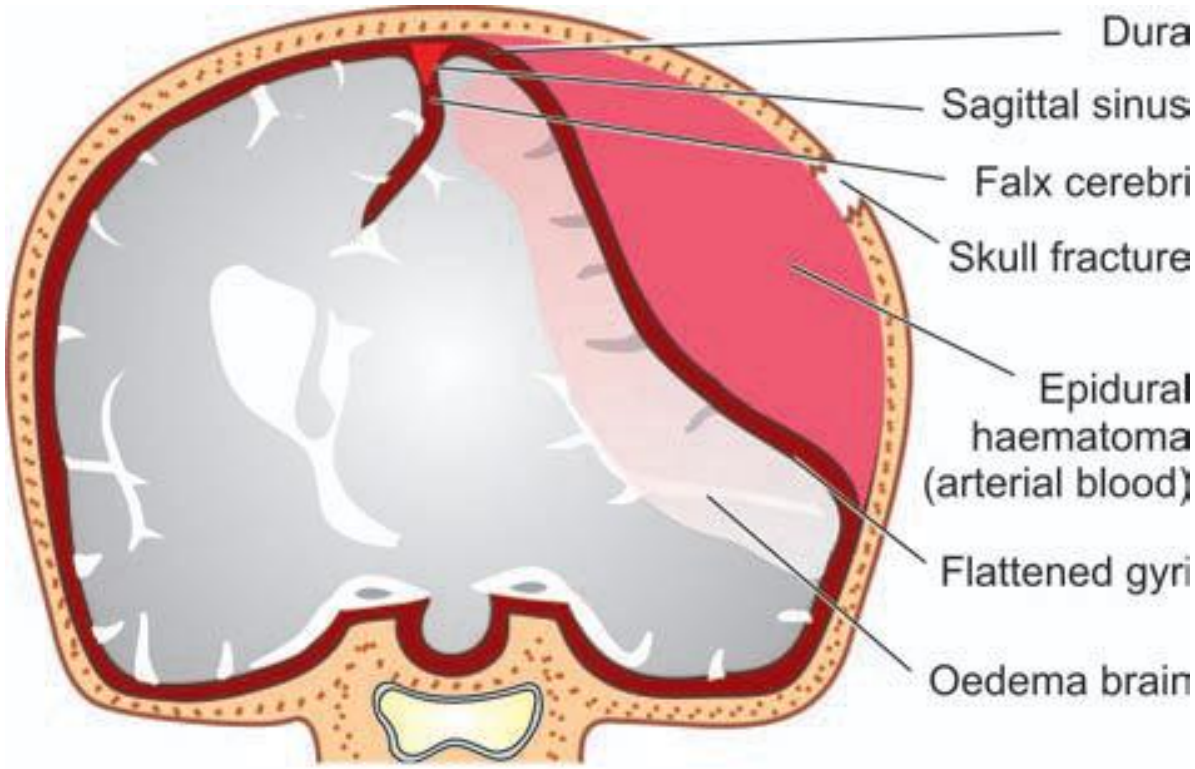
- **Site**: in the internal capsule from a branch of the middle cerebral artery.

- **Effects:**
  1. Hemiplegia.
  2. Increased intracranial tension.
  3. Compression of the brain stem and death due to tentorial herniation.

B. **Meningeal hemorrhage:**

- Mostly due to traumatic vascular injury:
  1. **Extradural or epidural hemorrhage:** Arterial blood between dura and skull bones.
- It is due to trauma to the middle meningeal artery.

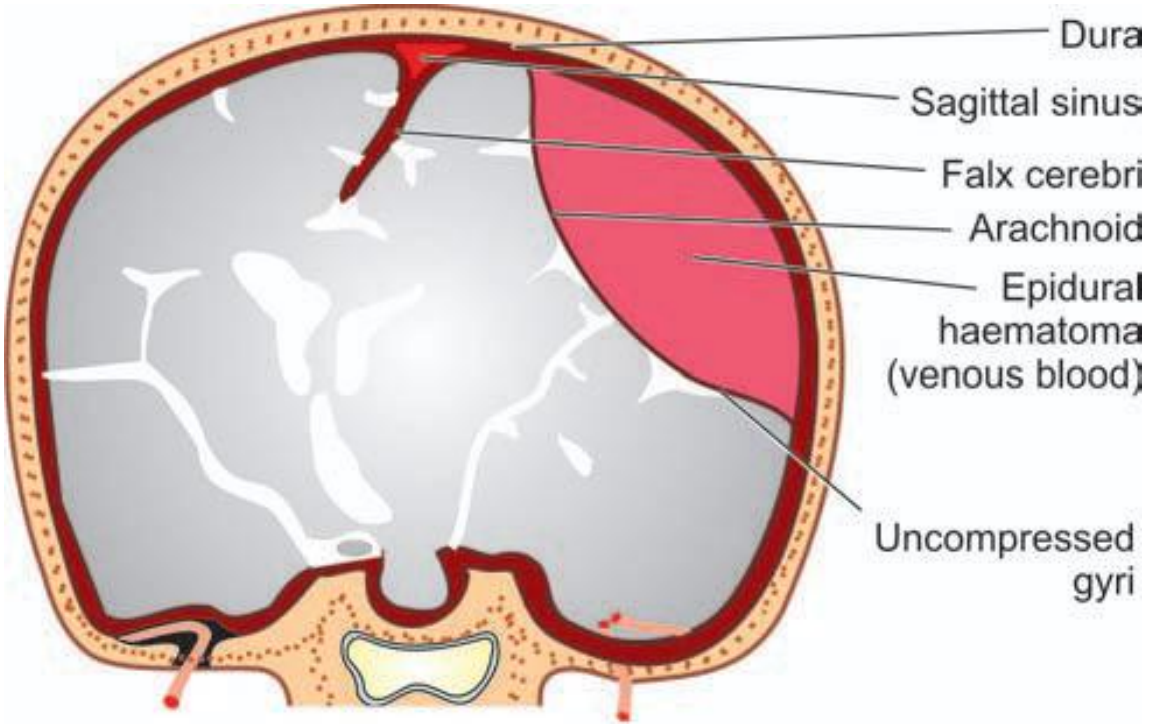
Epidural hematoma often results from rupture of artery following skull fracture resulting in accumulation of arterial blood between the skull and the dura.



2. **Subdural:** Venous blood in the subdural space between dura and arachinoid.

- It is due to traumatic rupture of cerebral veins (cortical veins) crossing the subdural space.
- The accumulated blood may form hematoma.

Subdural hematoma often results from rupture of veins crossing the cerebral convexities and is characterized by accumulation of venous blood between the dura and the arachnoid.

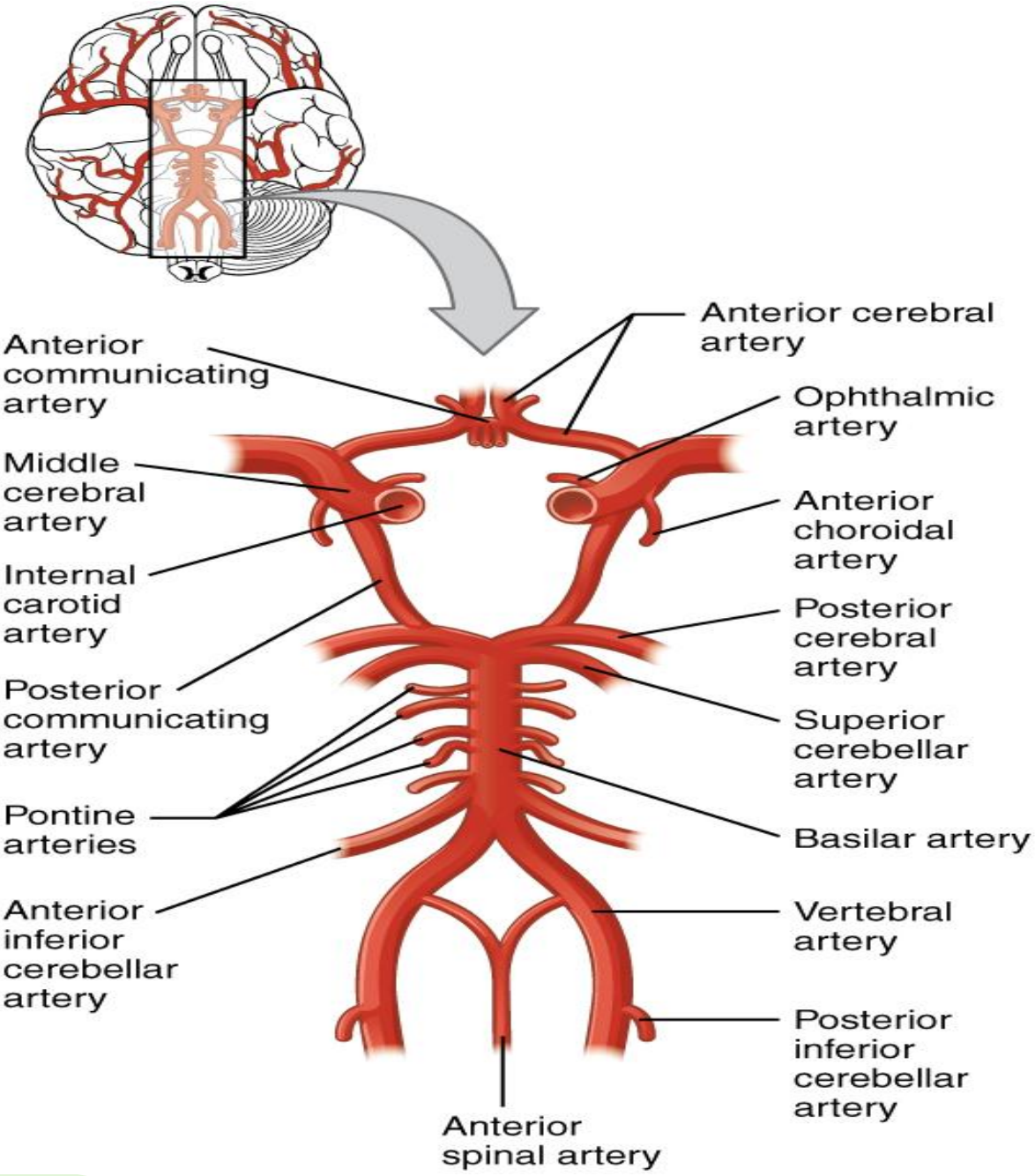


3. **Subarachnoid**: in the subarachnoids space.

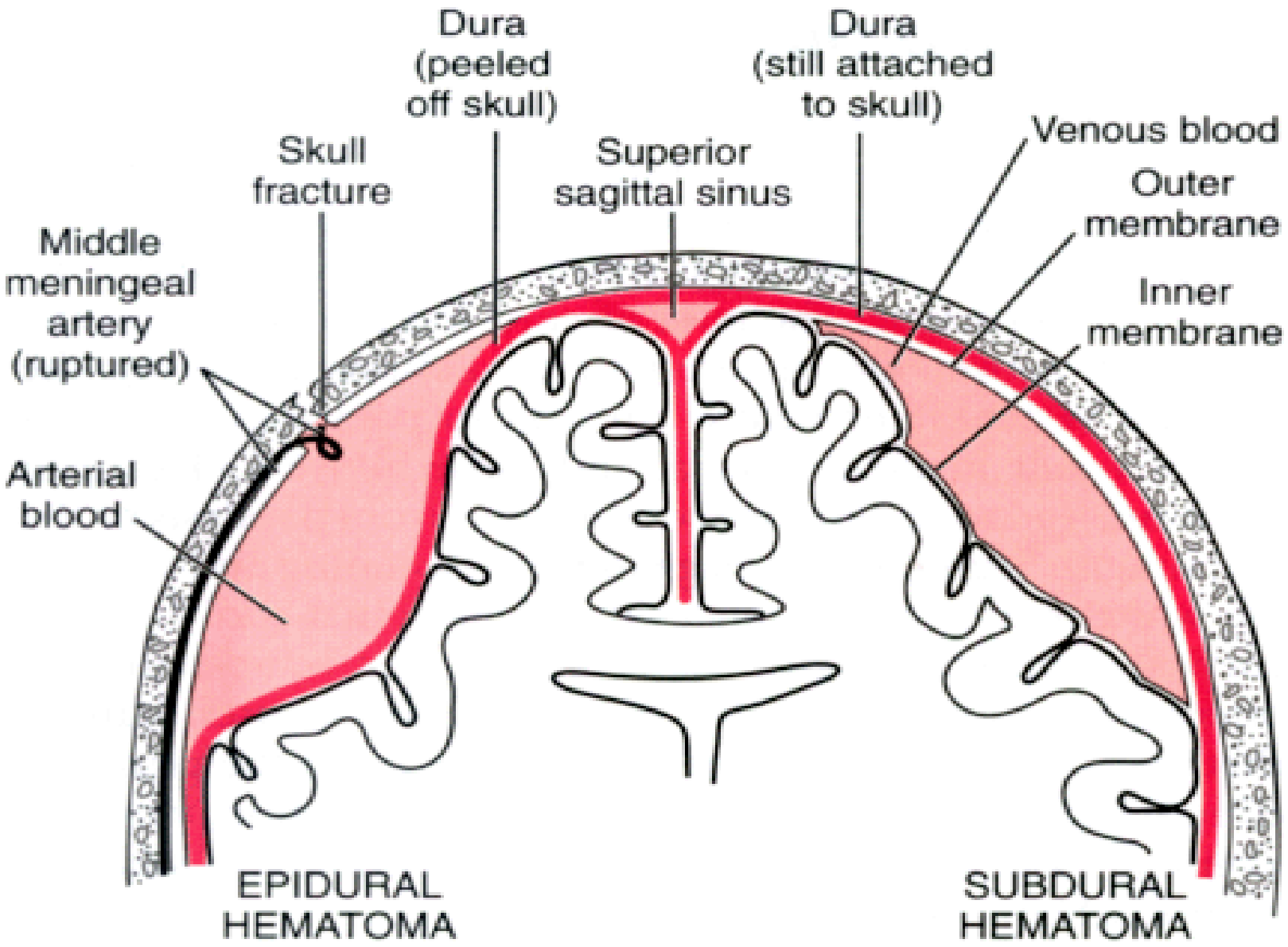
- **Caused by:**

A. Traumatic lacerations of the brain.
B. Spontaneous rupture of cerebral aneurysm.
C. Hemorrhagic blood diseases.

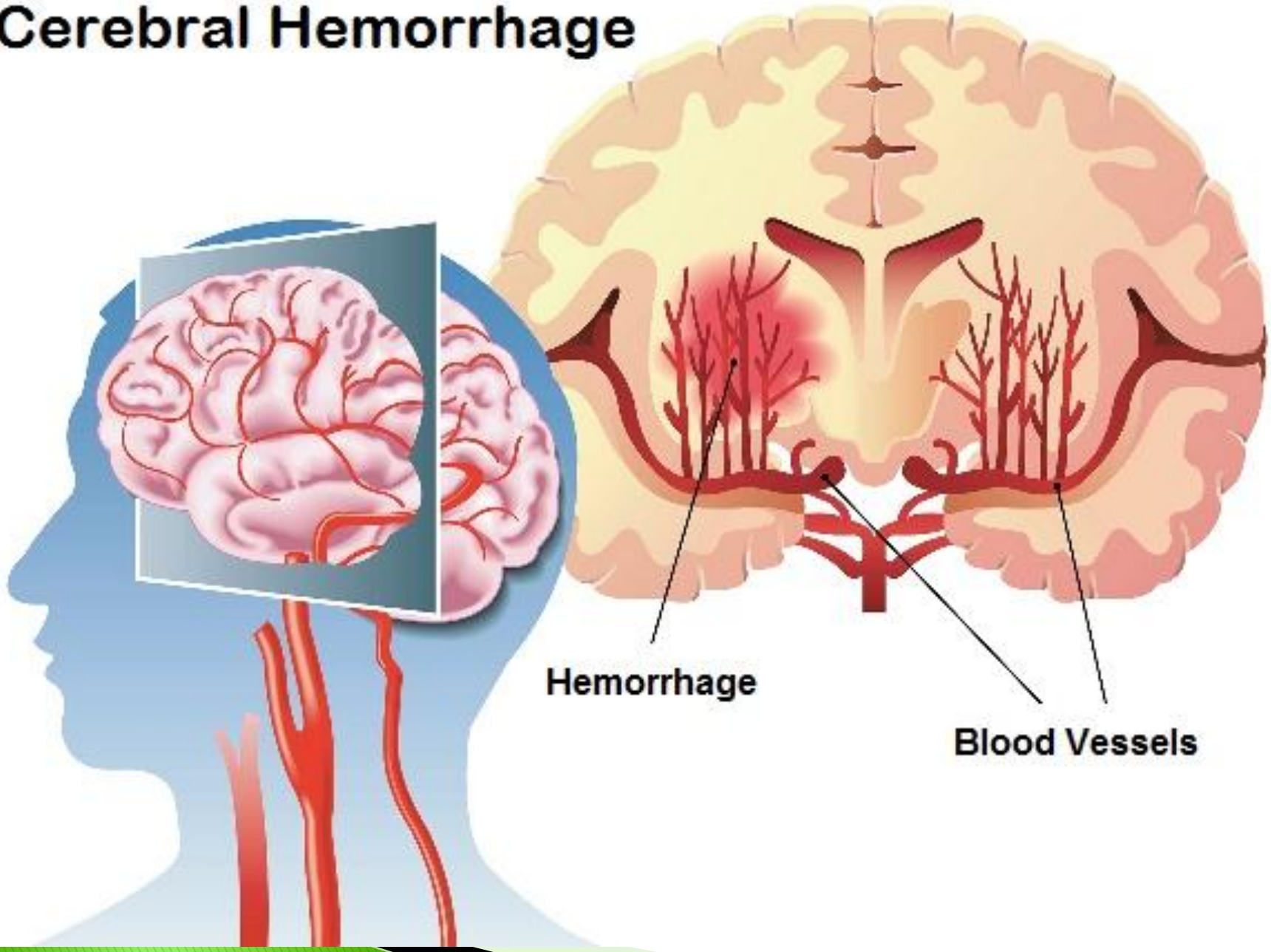
Circle of Willis



Intracranial hemorrhage



Cerebral Hemorrhage



Cerebral edema

- **Cerebral edema**: It is an excess accumulation of fluid in the intracellular or extracellular spaces of the brain, or
- It is an accumulation of excess fluid within the brain parenchyma.

- **Clinically:**
- The brain becomes soft and smooth and overfills the cranial vault, gyri (ridges) become flattened, sulci (grooves) become narrowed, and ventricular cavities become compressed.
- Symptoms include nausea, vomiting, blurred vision, faintness, and in severe cases, seizures and coma.
- If brain herniation occurs, respiratory symptoms or respiratory arrest can also occur due to compression of the respiratory centers in the pons and medulla oblongata.

Causes:

- Cerebral edema can result from **trauma** to the brain or from **nontraumatic** causes such as ischemic stroke, tumors, or inflammation due to meningitis or encephalitis.
- Vasogenic edema caused by amyloid-modifying treatments.
- The blood–brain barrier (BBB) or the blood–cerebrospinal fluid (CSF) barrier may break down, allowing fluid to accumulate in the brain's extracellular space.

- Altered metabolism may cause brain cells to retain water, and dilution of the plasma may cause excess water to move into the brain cells.
- Fast travel to high altitude without proper acclimatization can cause high-altitude cerebral edema.

Types:

- **Four types of cerebral edema have been identified:**

A. **Vasogenic edema:**

- Edema occurs when the integrity of the normal blood-brain barrier; BBB is disturbed due to breakdown of the tight endothelial junctions that make up the barrier.
- This allows subsequent increase in the vascular permeability with escape of intravascular proteins and fluid into the parenchymal extracellular space.

- Once plasma constituents and water cross the BBB → enter the white matter → move extracellularly → edema spreads and can affect the gray matter.
- This may be quite rapid and extensive.
- Mechanisms contributing to BBB dysfunction include physical disruption by hypertension, cerebral ischemia, trauma, and tumor-facilitated release of vasoactive and endothelial destructive compounds (e.g. arachidonic acid, excitatory neurotransmitters, eicosanoids, bradykinin, histamine, and free radicals), and focal inflammation.
- Edema can be either localized adjacent to inflammation or tumors or generalized.

- *Subtypes of vasogenic edema include:*

1. **Hydrostatic cerebral edema:**

- This form of cerebral edema is seen in cases of malignant hypertension.
- It results from direct transmission of pressure to the cerebral capillaries with transudation of fluid from the capillaries into the extravascular compartment.

2. **Cerebral edema from brain tumors:**

- Glioma can increase secretion of VEGF, which weakens the junctions of the BBB.
- Dexamethasone can be of benefit in reducing VEGF secretion.

3. ## High altitude cerebral edema (HACE):

- It is severe and sometimes fatal form of altitude sickness that results from capillary fluid leakage due to the effects of hypoxia on the mitochondria-rich endothelial cells of the BBB.
- Symptoms include headache, loss of coordination (ataxia), weakness, disorientation, memory loss, psychotic symptoms (hallucinations and delusions), coma and death.

B. Cytotoxic edema:

- It is an ↑ in the intra-cellular fluid secondary to neural, glial, or endothelial cell membrane injury e.g., generalized, hypoxic/ischemic insult, or exposure to some toxins.
- Cytotoxic edema is seen with various toxins, Reye's syndrome, severe hypothermia, early ischemia, encephalopathy, early stroke or hypoxia, cardiac arrest, and pseudotumor cerebri.
- Cytotoxic edema is dominant immediately following an injury or infarct, but gives way to vasogenic edema that can persist for several days or longer.

- In most instances, cytotoxic and vasogenic edema occur together.
- In cytotoxic edema, the BBB remains intact but disruption in cellular metabolism impairs function of sodium and potassium pump in glial cell membrane → cellular sodium and water retention.
- During an ischemic stroke, lack of oxygen and glucose → breakdown of the sodium-calcium pumps on the cell membranes → massive buildup of sodium and calcium intracellularly.
- This causes rapid uptake of water and subsequent swelling of the cells.

- Swollen astrocytes occur in the gray and white matter.
- Swelling of the individual cells of the brain is the main distinguishing characteristic of cytotoxic edema, as opposed to vasogenic edema, wherein the influx of fluid is typically seen in the interstitial space rather than within the cells themselves.
- While not all patients who have stroke will develop severe edema, those who do, have a very poor prognosis.

C. Osmotic edema:

- Normally, the osmolality of CSF and extracellular fluid in the brain is slightly lower than that of plasma.
- Plasma can be diluted by several mechanisms, including excessive water intake (or hyponatremia), syndrome of inappropriate antidiuretic hormone secretion, hemodialysis, or rapid reduction of blood glucose in hyperosmolar hyperglycemic state, formerly known as hyperosmolar non-ketotic acidosis.
- Plasma dilution decreases serum osmolality, resulting in a higher osmolality in the brain compared to the serum.

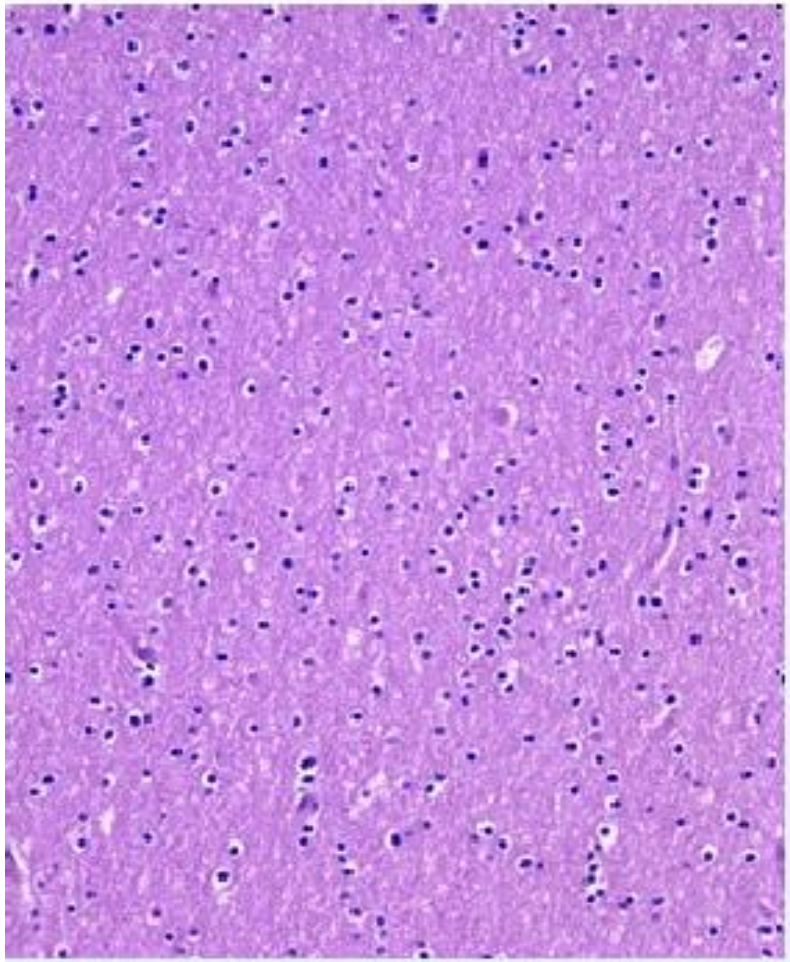
- This creates an abnormal pressure gradient and movement of water into the brain, which can cause progressive cerebral edema, resulting in a spectrum of signs and symptoms from headache and ataxia to seizures and coma.

D. Interstitial edema:

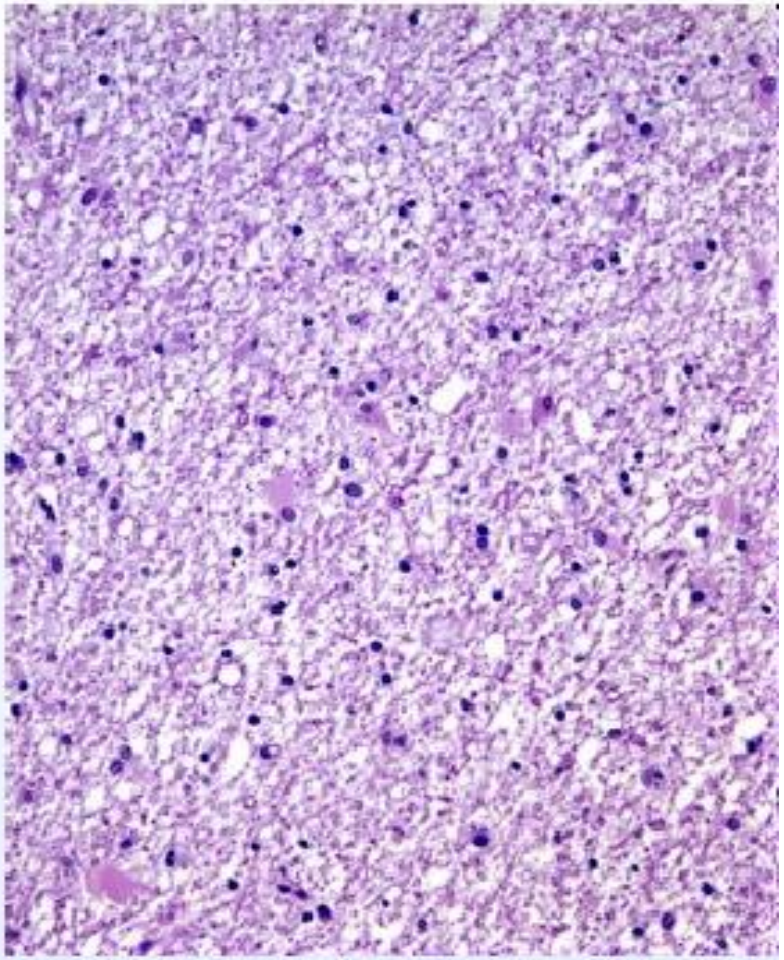
- Interstitial edema occurs in obstructive hydrocephalus due to a rupture of the CSF-brain barrier.
- This results in trans-ependymal flow of CSF, causing CSF to penetrate the brain and spread to the extracellular spaces and the white matter.
- Interstitial cerebral edema differs from vasogenic edema as CSF contains almost no protein.

- **Gross picture:**
- The edematous brain is softer, and appears to overfill the cranial vault.
- The gyri are flattened as a result of compression of the expanding brain by the dura matter and the inner surface of the skull.
- The intervening sulci are narrowed.
- The ventricular cavities are compressed.

Cerebral Edema:



Normal



Edema

CEREBRAL ANEURYSMS

- Types:

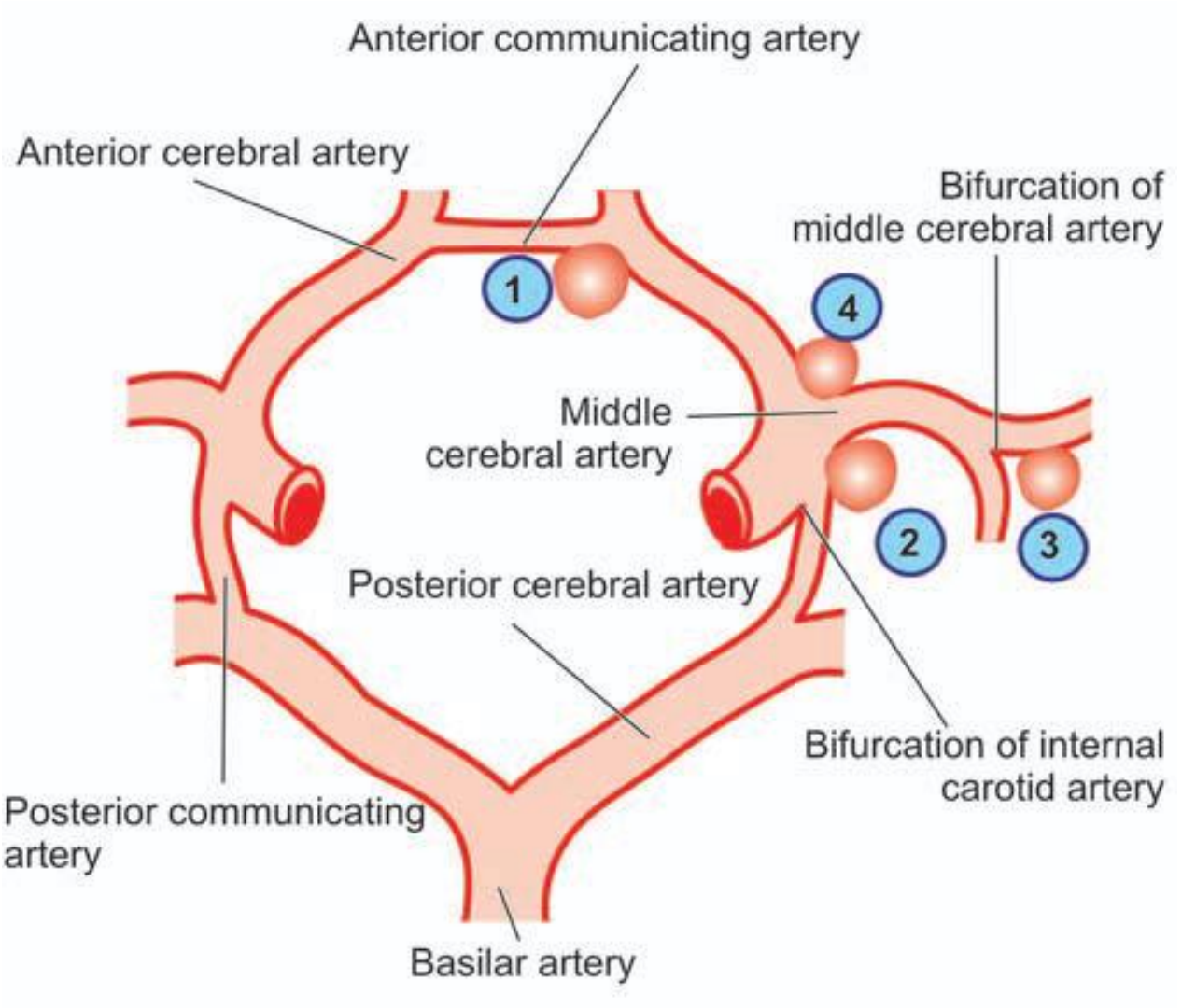
1. ***Congenital aneurysms (Berry aneurysms):***

- Single or multiple small aneurysms (pin head-2 cm size).
- Usually multiple saccular aneurysms and if they rupture, they cause subarachinoid hemorrhage.

- Found at the bifurcation of the cerebral arteries as a result of congenital defects in the media.
- Although referred to as congenital, they are not present at birth and develop overtime.
- Associated conditions: polycystic kidney disease (autosomal dominant).

- **Etiology:**
  1. Congenital defects of the media at the arterial bifurcations.
  2. Focal destruction of the internal elastic lamina due to hemodynamic alterations.
  3. Abnormalities of specific collagen subsets.

The circle of Willis showing principal sites of berry (saccular) aneurysms. The serial numbers indicate the frequency of involvement.



2. ***Mycotic aneurysms:***

- Develop in association with subacute bacterial endocarditis or polyarteritis nodosa due to inflammatory weakness in the vascular wall.
- Occurs when the wall of an artery is weakened by mild infections.
- The commonest site is the branches of middle cerebral artery.

3. ***Atherosclerotic aneurysm:***

- Fusiform in shape.
- Develops in association with hypertension or cerebral atherosclerosis.
- **Sites**:
- Internal carotid and basilar arteries.

- **Effects of cerebral aneurysms:**
  1. Increased intracranial tension.
  2. Rupture giving subarachinoid hemorrhage.
  3. Hydrocephalus.
  4. Thrombosis and calcifications.

Vascular malformations

1. **Arteriovenous malformation (AVM):**

- Admixture of arteries, veins and intermediate sized vessels.
- Accounts for 1-4% of all brain masses.
- Common in males between 10-30 years.
- It is the most common vascular malformation associated with subarachinoid hemorrhage.
- Middle cerebral artery distribution is the most common site.

2. **Cavernous malformation (cavernous angioma):**

- Large sinusoidal type vessels.
- Slight male predominance, most in young adults.

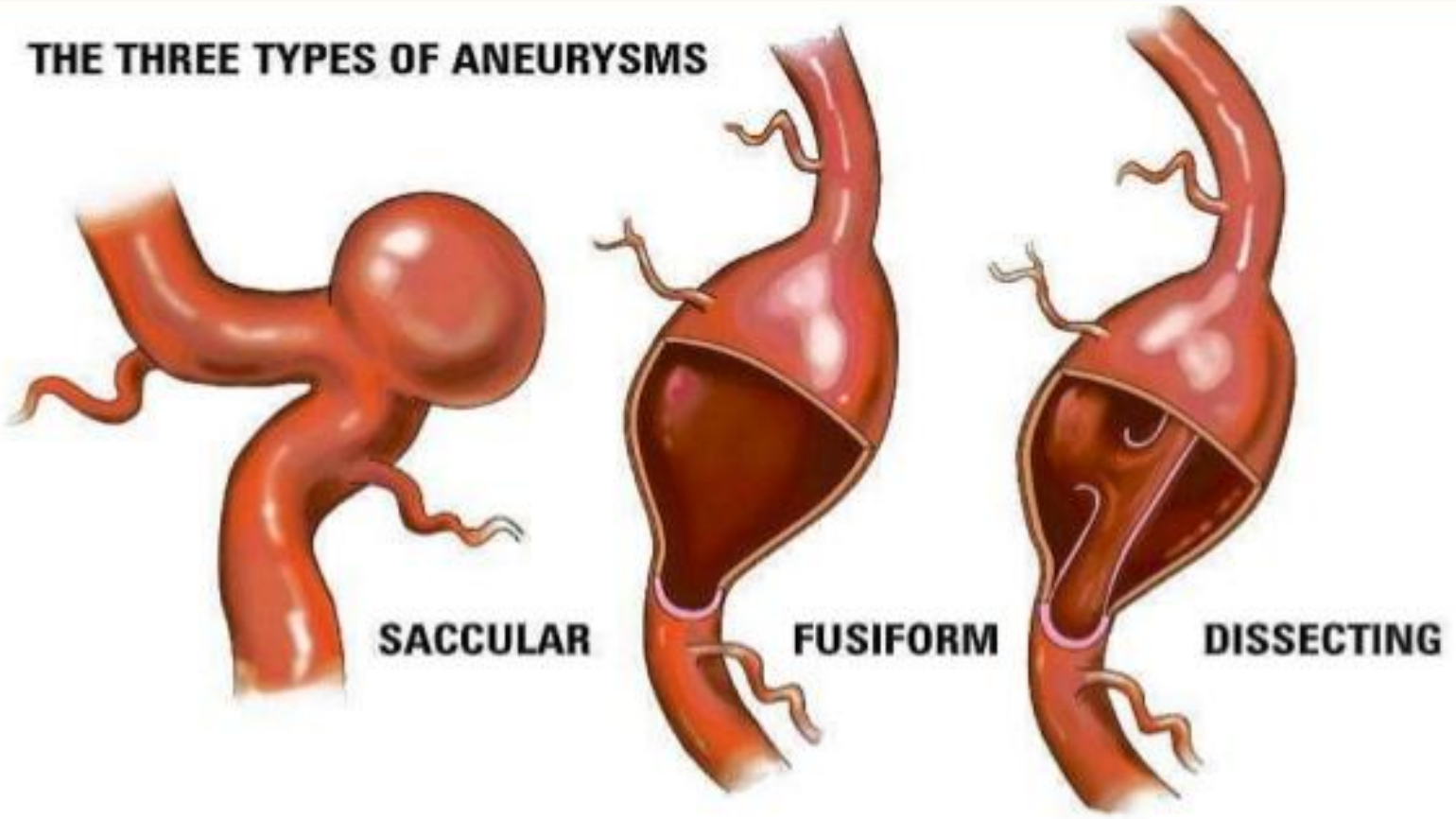
3. **Capillary telangiectasis:**

- Capillary sized vessels separated by normal neural parenchyma.

4. ***Arteriovenous aneurysm:***

- Communication between the internal carotid artery and the cavernous sinus caused by fracture of the base of the skull.
- **Effects**:
  1. Rupture leading to subarachnoid hemorrhage.
  2. Pressure effect.
  3. Thrombosis and calcification.

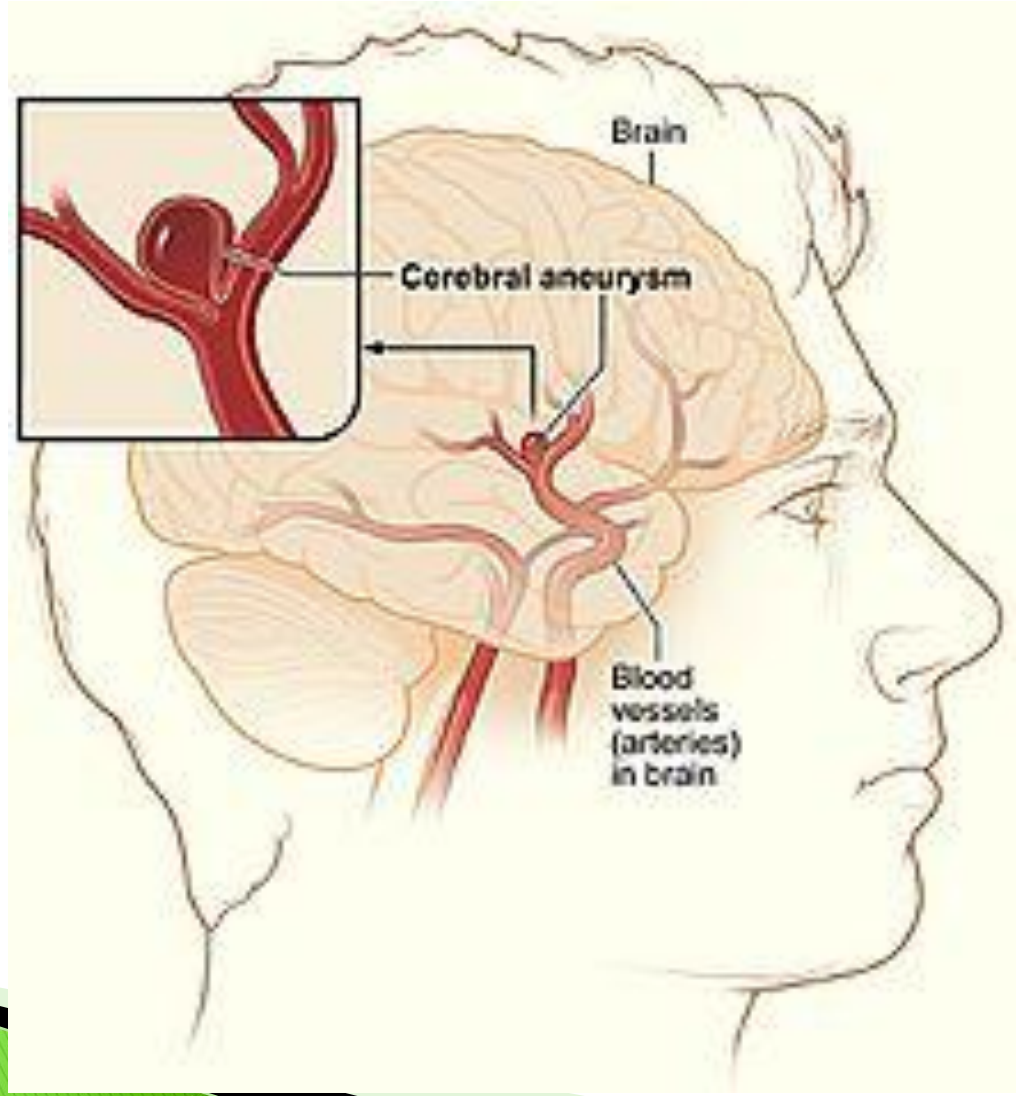
Types of Cerebral aneurysm:



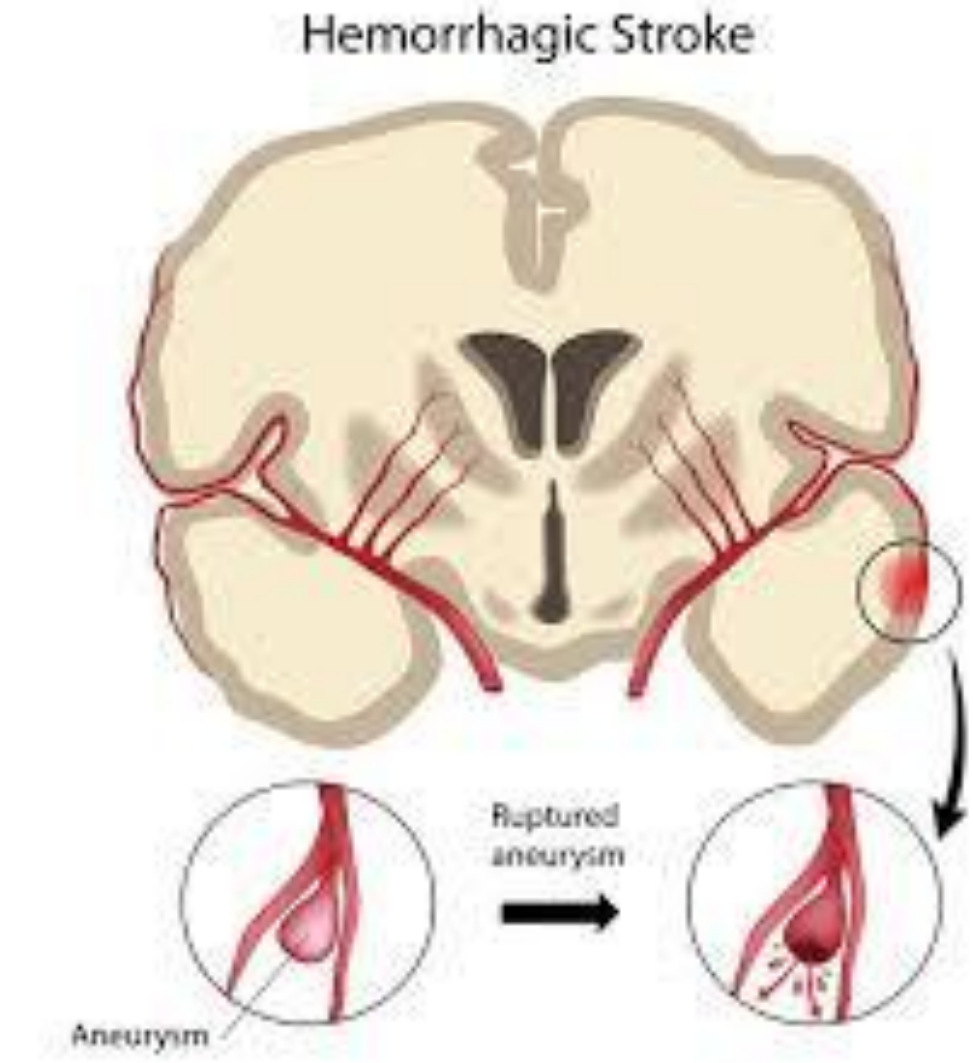
Source: New York-Presbyterian Hospital; University of Maryland Medical Center; M. Headworth; Mayfield Clinic

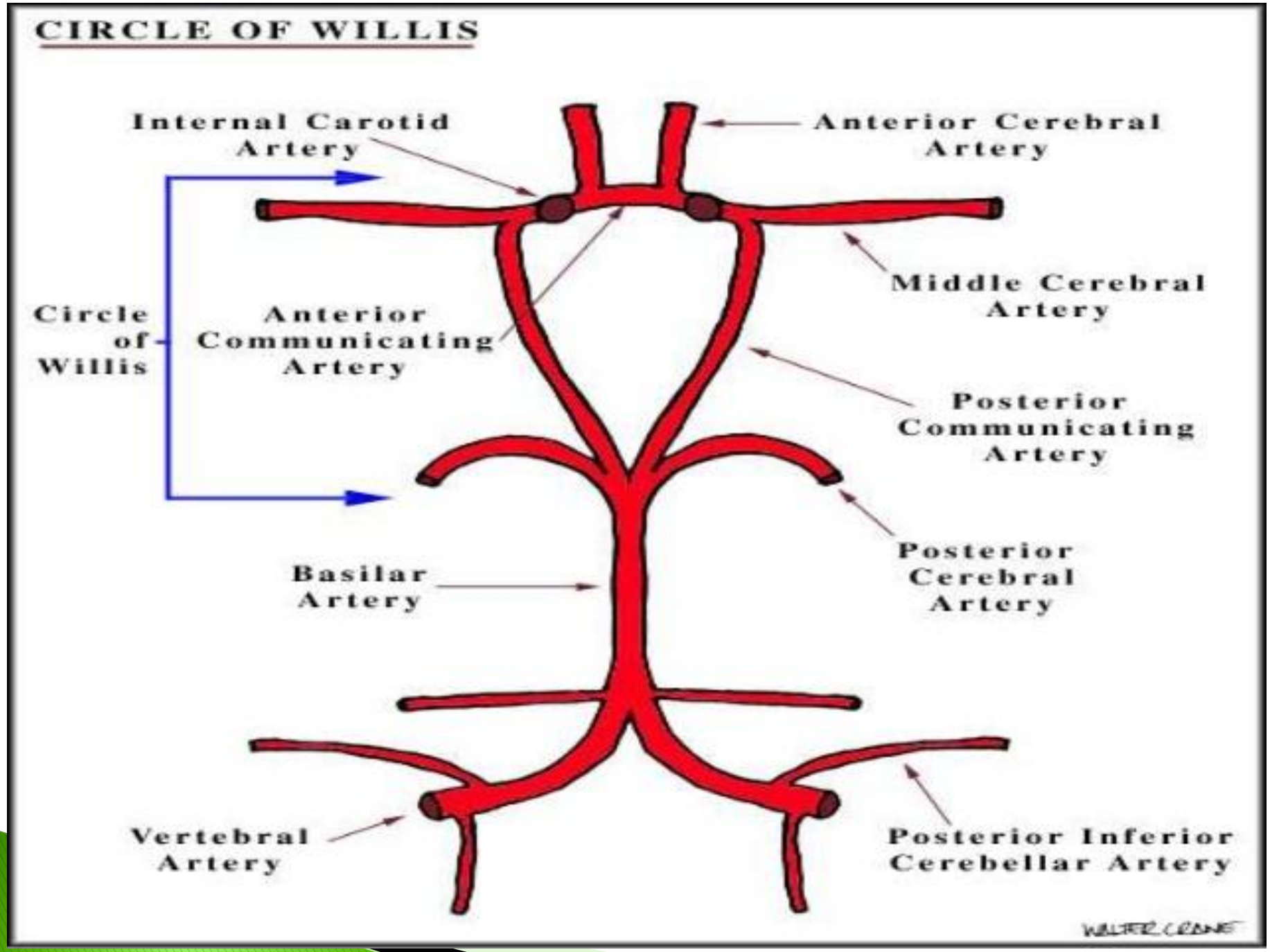
CHARLOTTE THIBAULT / Monitor staff

CEREBRAL ANEURYSMS



CEREBRAL ANEURYSMS



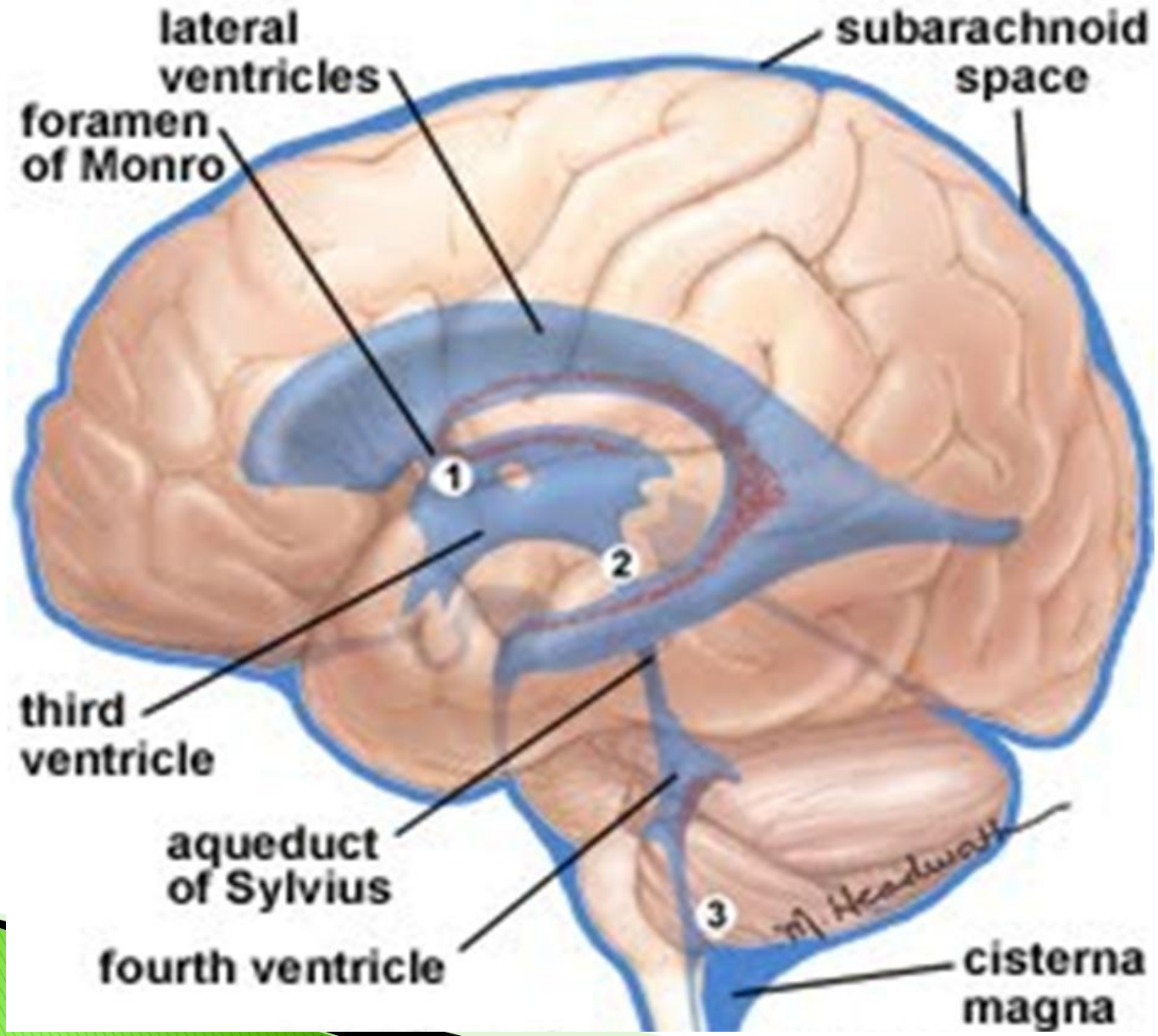

CIRCLE OF WILLIS
Internal Carotid Artery
Anterior Cerebral Artery
Middle Cerebral Artery
Circle of Willis
Anterior Communicating Artery
Posterior Communicating Artery
Posterior Cerebral Artery
Basilar Artery
Vertebral Artery
Posterior Inferior Cerebellar Artery

Normal cerebrospinal fluid (CSF)

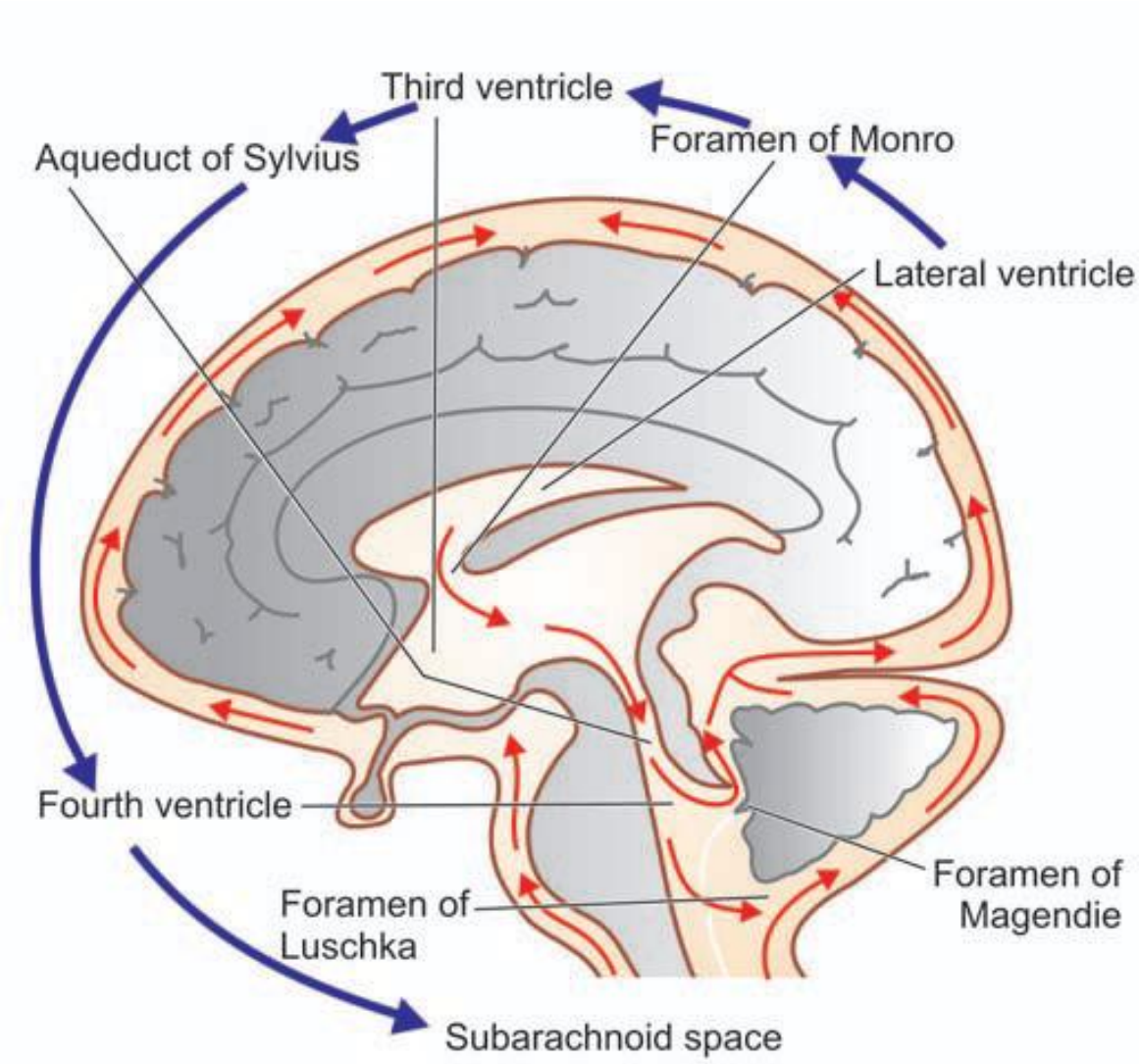
- **<u>Production and circulation:</u>**
- Produced by choroid plexus in the lateral ventricles.
- Pass from the lateral to the third ventricle through foramina of ***Monro*** (inter-ventricular foramen).
- Pass from the third to the fourth ventricle through aqueduct of ***Sylvius***.

- Passes from the fourth ventricle to the sub- arachnoids space through foramina of ***Magendie*** and ***Luschka.***
- CSF then drains to the **superior sagittal sinus** through the arachnoid villi or granulations.

Normal CSF


lateral ventricles
subarachnoid space
foramen of Monro
1
2
third ventricle
aqueduct of Sylvius
3
fourth ventricle
cisterna magna

Normal circulation of CSF



HYDROCEPHALUS

- **Definition:**
- Abnormal dilatation of the ventricular system by excess CSF with atrophy of the brain tissue.

- **Causes:**

I. ***Increased CSF production:***

   A. Coroid plexuses tumor as papilloma.

II. ***Obstrution of CSF flow:***

   A. ***Congenital causes:***

      1. Stenosis of the aqueduct cerebri; foramina of ***Monro*** and ***Sylvius***.
      2. Stenosis of the foramina of fourth ventricle; foramina of ***Magendie*** and ***Luschka***.
      3. Arnold-Chiari malformation.

B. ***Acquired causes:***

1. Meningitis → fibrous adhesions obstructing the foramina of the fourth ventricle or the subarachnoid space.
2. Brain abscess may obstruct the pathway of CSF.
3. Brain tumor may also obstruct the pathway of CSF.

III. Decreased CSF drainage or absorption:

A. Congenital causes:
   1. Aplasia of arachinoid villi.

B. Aquired causes:
   1. Damage of arachnoid villi by inflammatory exudate or fibrosis.
   2. Obliteration of the subarachnoid space by subtentorial space occupying lesion → pushes the brain up against the skull bones.
   3. Thrombosis of the superior sagittal sinus.

- Types:

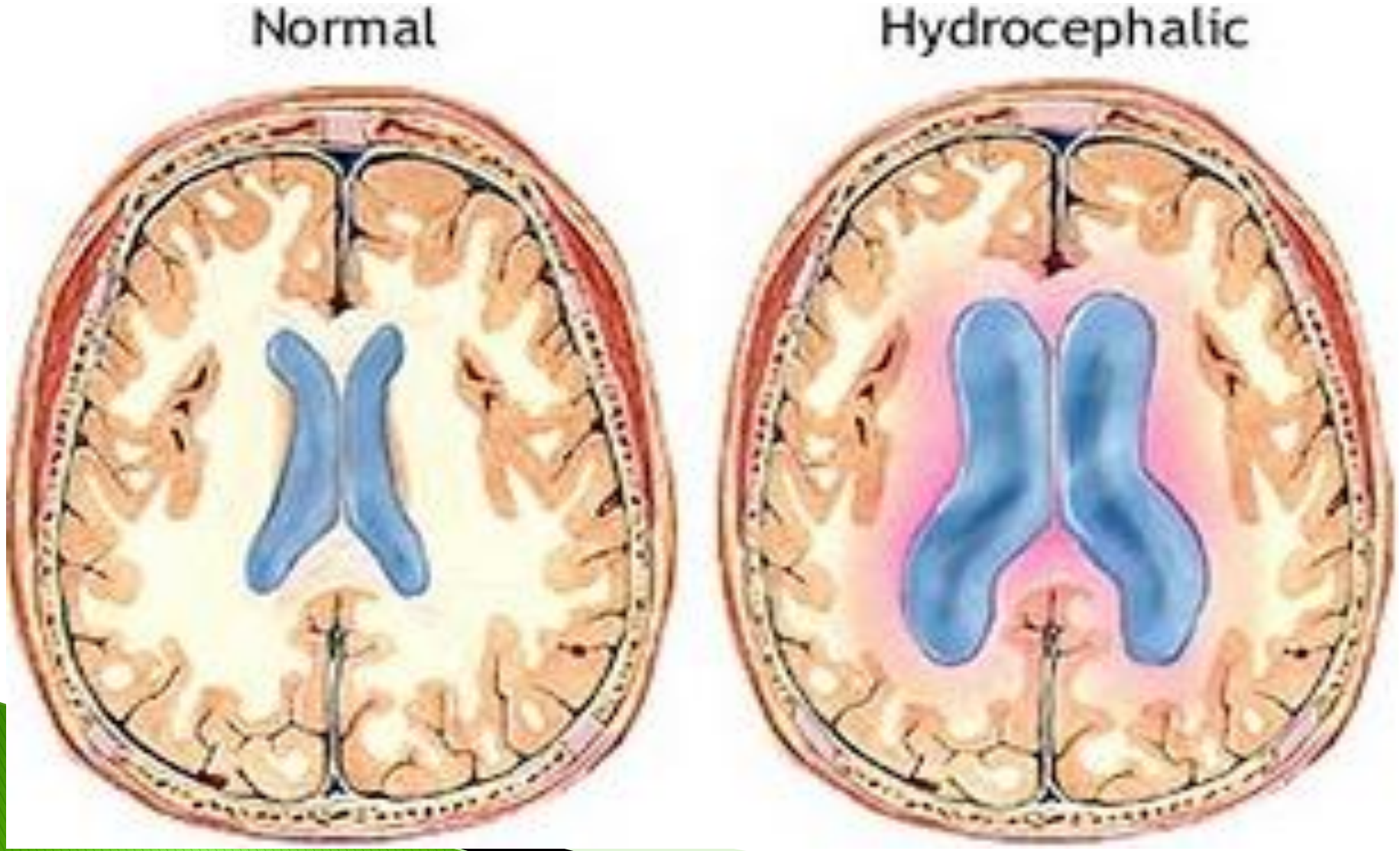
1. ***Communicating hydrocephalus:***

- The obstruction is in the subarachnoid space around the mid-brain.
- CSF can pass from the ventricular system to the subarachenoid space.

2. ***Non-communicating hydrocephalus:***

- The obstruction is within the ventricular system or in the roof of the fourth ventricle.
- CSF can not pass from the ventricular system to the subarachenoid space.

HYDROCEPHALUS



Clinical features of hydrocephalus

- **Features of ↑ ICT:**
  1. Nausea
  2. Vomiting
  3. Headache
  4. Altered consciousness
- **Other symptoms include:**
  1. Dysphagia, laryngeal dysfunction
  2. Visual disturbances
  3. Hearing impairments
  4. Weakness or numbness of the face.
  5. Difficulties with balance and walking.

Hydrocephalus

Increased intracranial tension (↑ICT)

- **Etiology**
  1. Intracranial tumors
  2. Intracranial inflammation:
     A. Acute:
        - Suppurative: septic meningitis
        - Non suppurative: viral meningitis

     B. Chronic:
        - Non specific: chronic abscess
        - Specific: TB and Gumma of syphilis

3. Intracranial vascular disorders
    a) Aneurysms
    b) Hemorrhages.
4. Hydrocephalus

- **Symptoms**:

- Persistent headache
- Vomiting
- Blurred vision

- **Effects**
  1. Flattened brain convolution on the same side of lesion.
  2. Shift of midline structures to the opposite side.
  3. Papillaedema: due to compression of retinal veins in the sub-arachenoid space.
  4. Thinning of skull bone over the lesion in chronic cases.
  5. Intracranial herniation:
     a) Tentorial herniation.
     b) Tonsillar herniation.

A. **Tentorial herniation:**

- Herniation of the cerebrum through tentorium cerebelli.
- Occurs when the space occupying lesion is located above tentorium cerebelli.

- **Effects**:
  a. Compression of midbrain and aquiduct of Sylvius → hydrocephalus.
  b. Compression of 4^{th} and 6^{th} cranial nerves → distorted eye movement.
  c. Compression of posterior cerebral artery → posterior cerebral infarction.

B. **Tonsillar herniation:**

- Herniation of the brain stem through foramen magnum.

- **Effects**:
  - Compression of medulla oblongata → sudden death.

Thank You