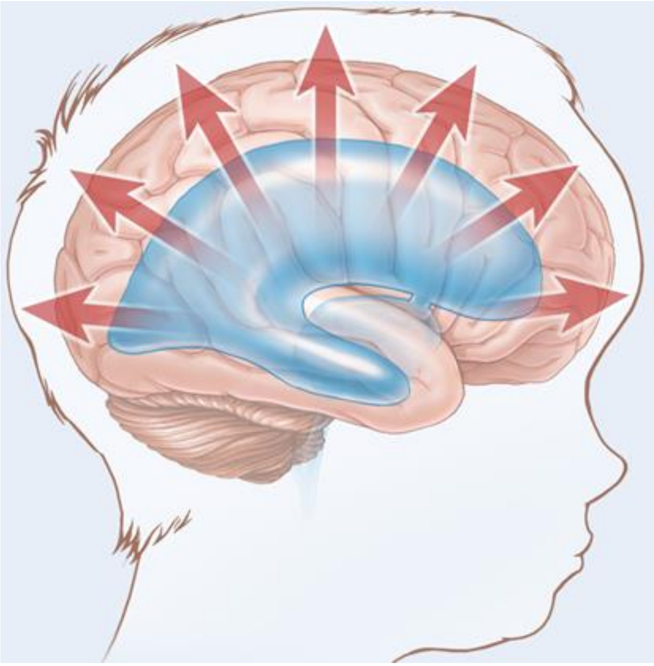
Department of Surgery
Neurosurgery Teaching Unit

SEMINAR: INTRACRANIAL HYPERTENSION AND HYDROCEPHALUS

34484 Pathology of the nervous system

Neurosurgery



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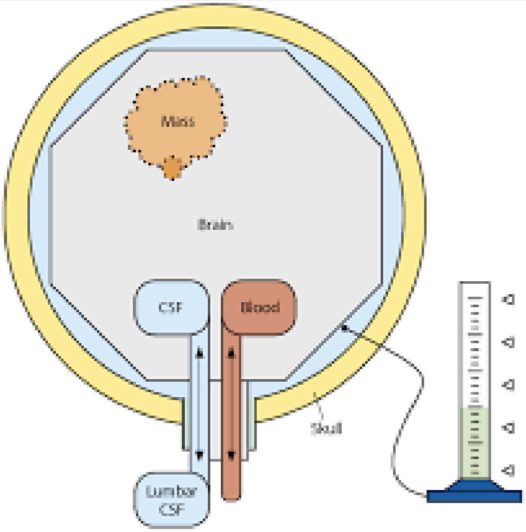
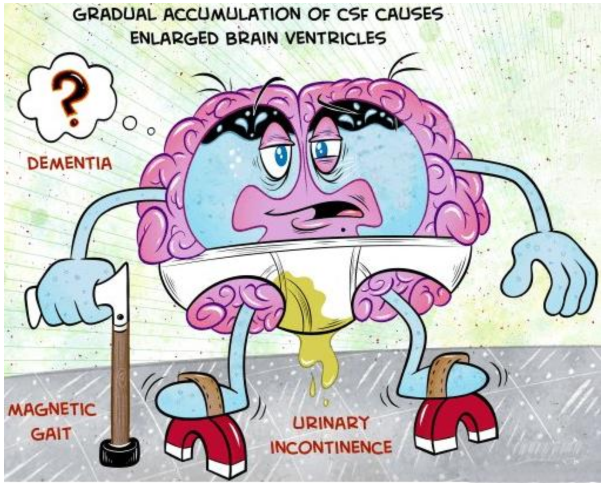
Key points to study

- Intracranial pressure and intracranial hypertension
- Benign intracranial hypertension
- Normal pressure hydrocephalus
- CSF drainage systems

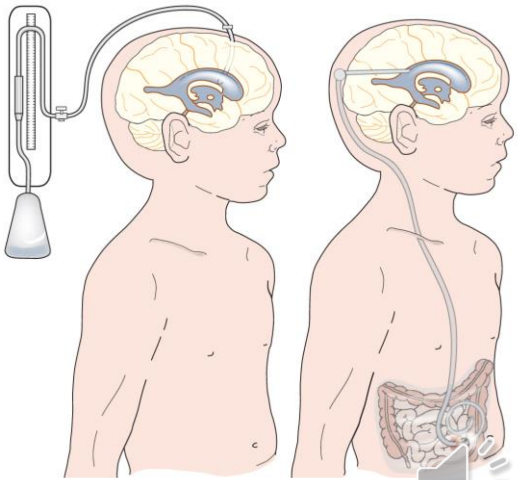


Intracranial pressure

Benign intracranial hypertension



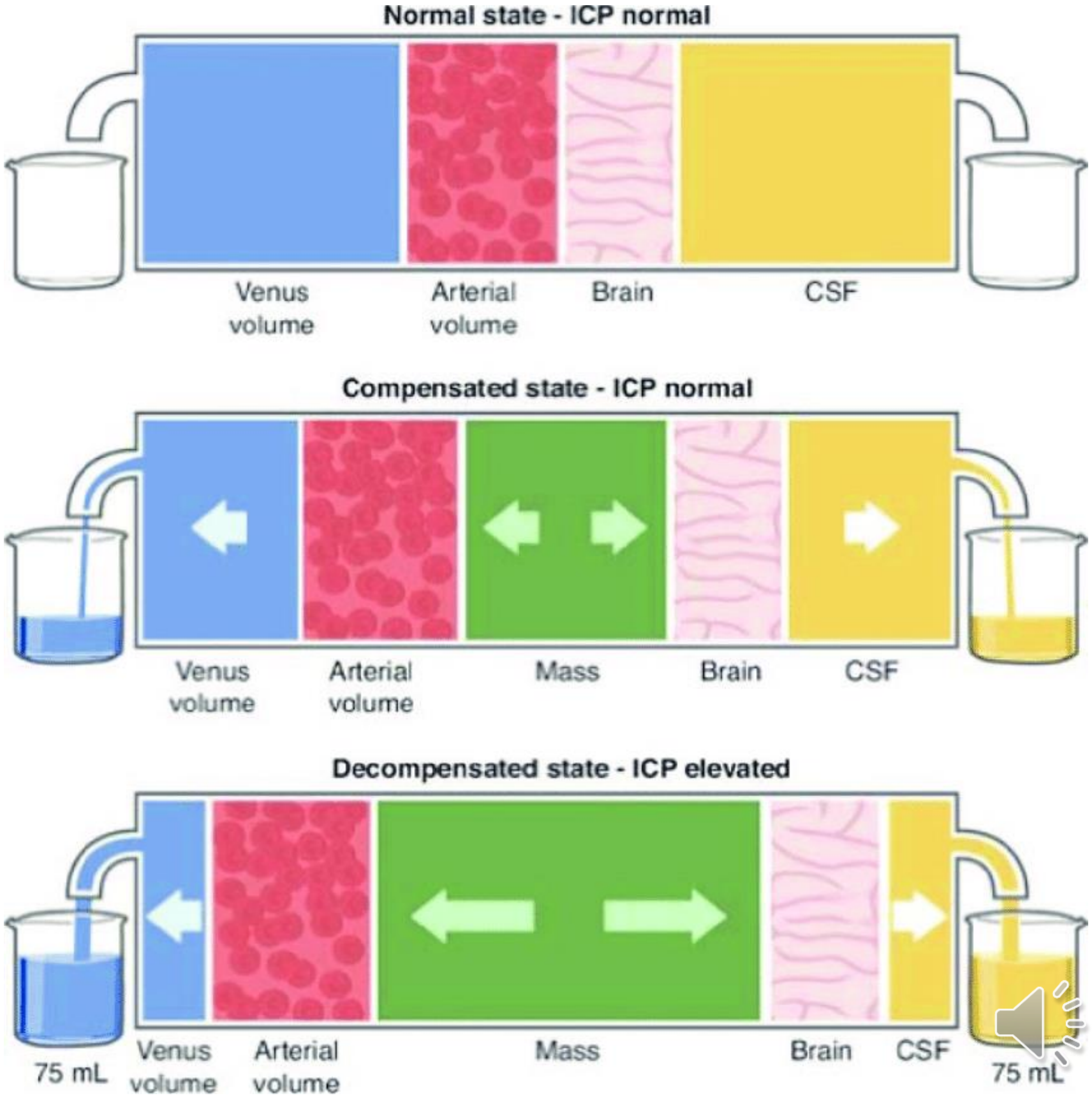
Chronic adult hydrocephalus



CSF drainage systems

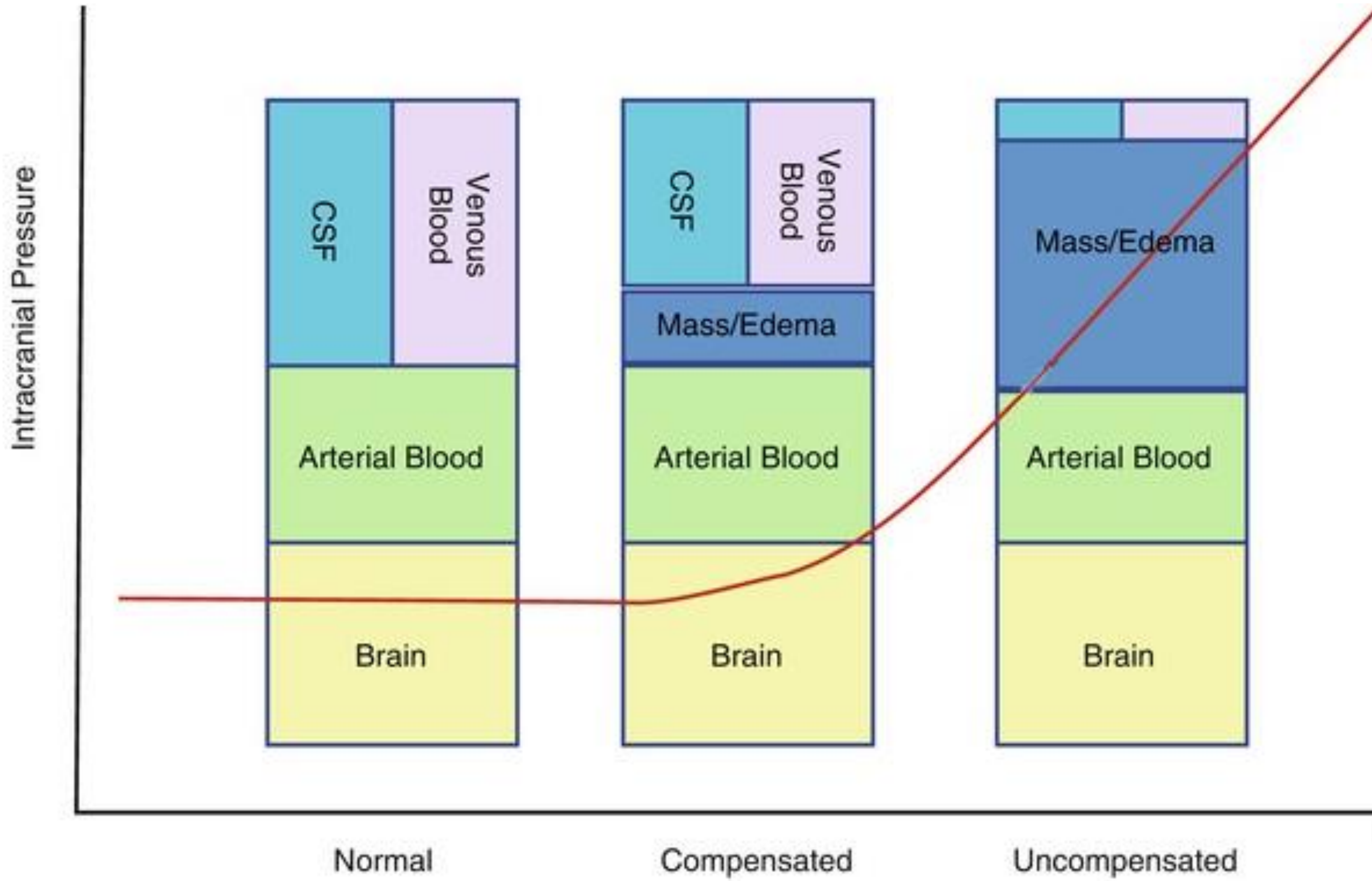
Monro-Kellie doctrine: intracranial content evolution if ↑volume

- 1st ↓ CSF
- 2nd ↓ venous blood
- 3rd ↓ arterial blood
- 4th decompensation- brain, herniations- brain, ischemi cerebral vascularization collapse - brain death



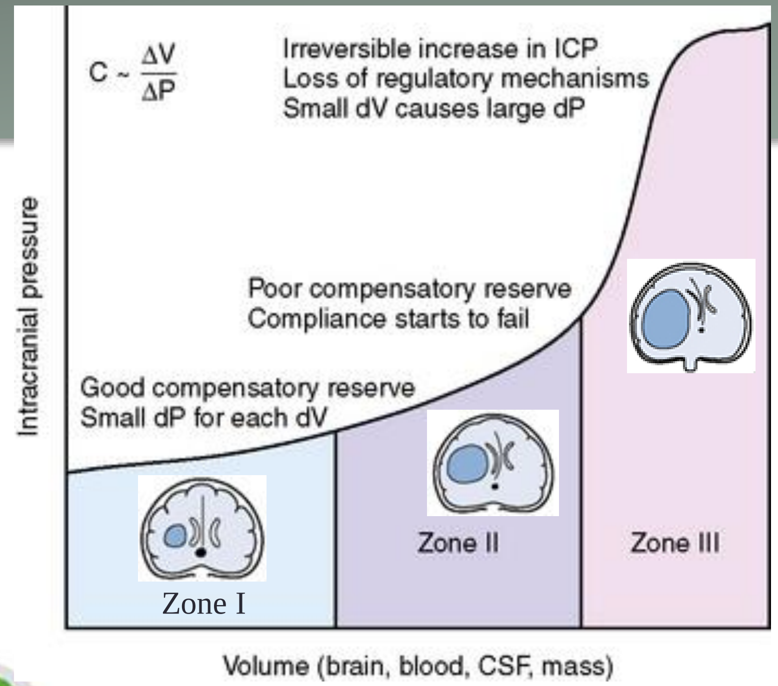
Evolution of intracranial space contents & intracranial pressure

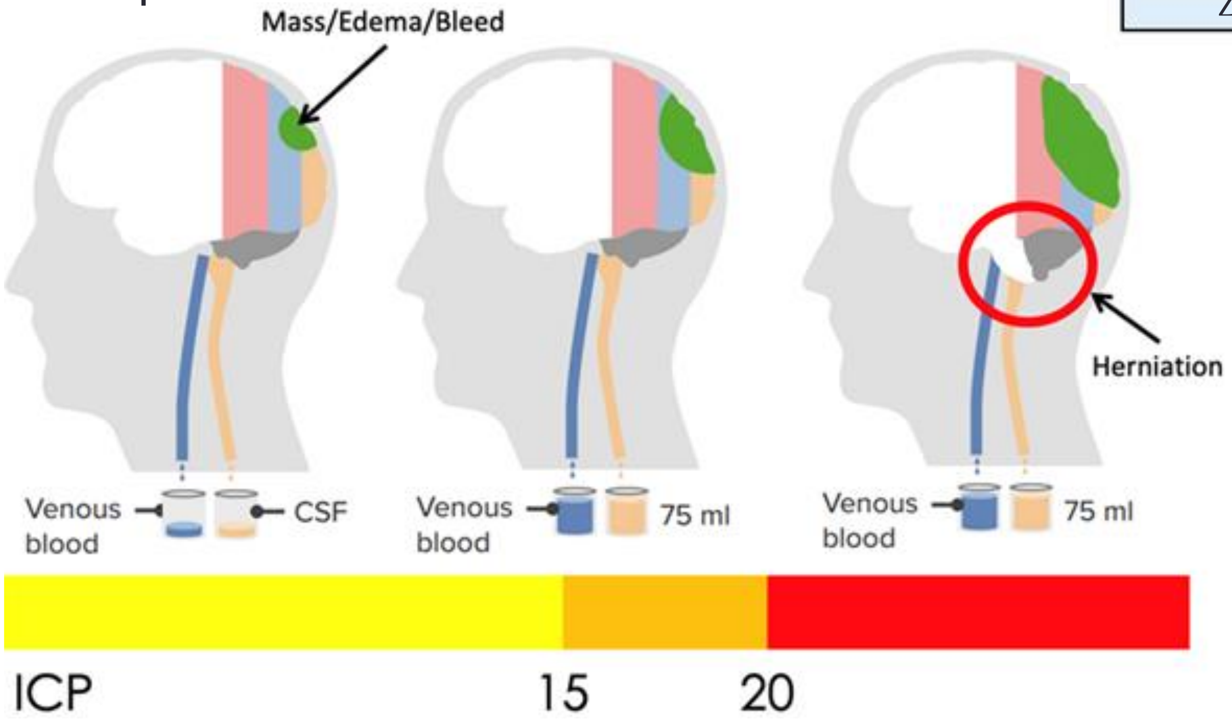
- CSF-venous blood-arterial blood-brain herniations



Steps ICP increase

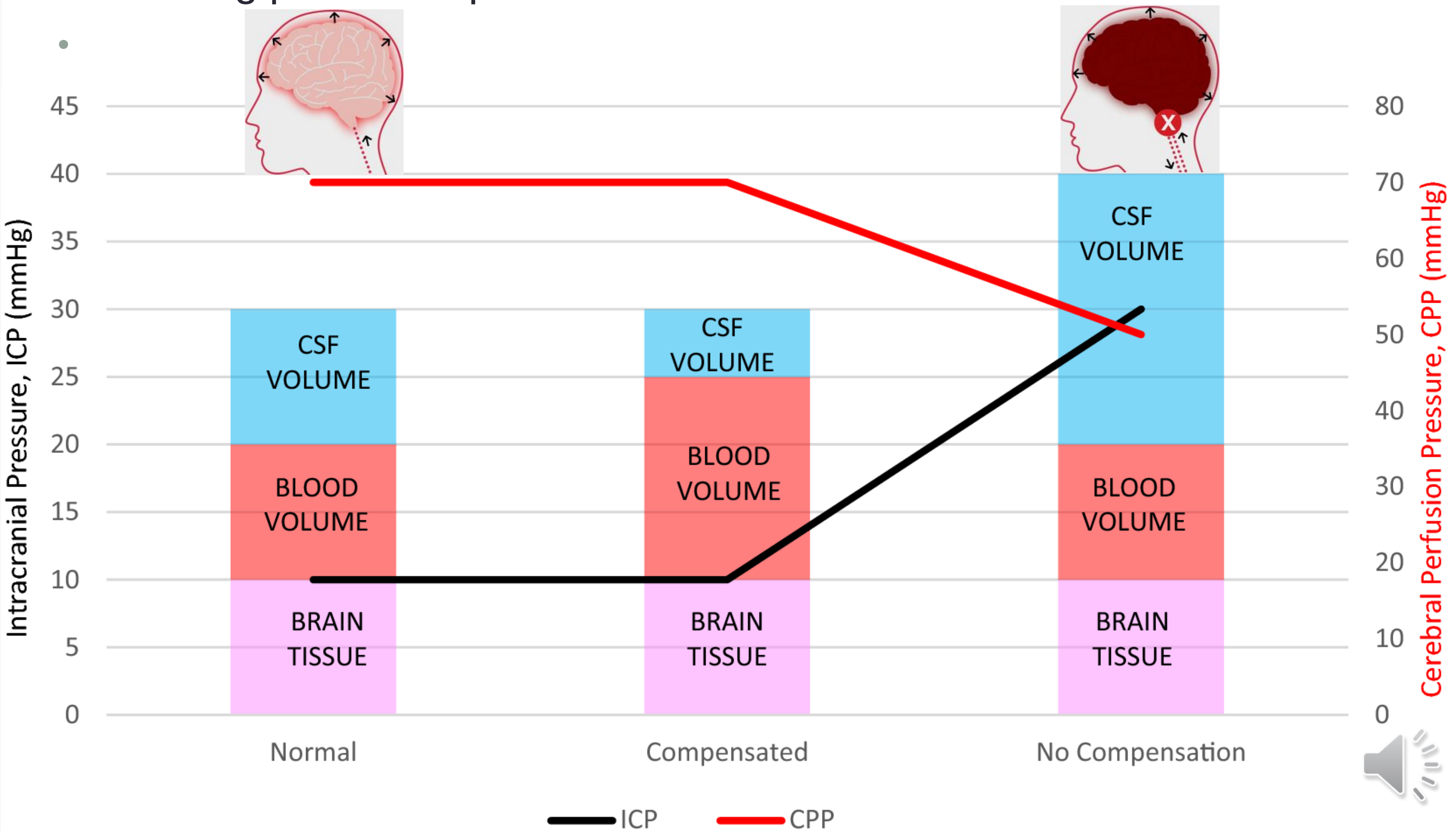
- 1st ↑Volume = little or no ↑ICP
- 2nd ↑Volume = ↑ICP
- 3rd ↑Volume = ↑↑ICP
- 4th ↑Volume = ↑↑↑ ICP-brain herniations-cerebral ischemia cerebral vascularization collapse-brain death





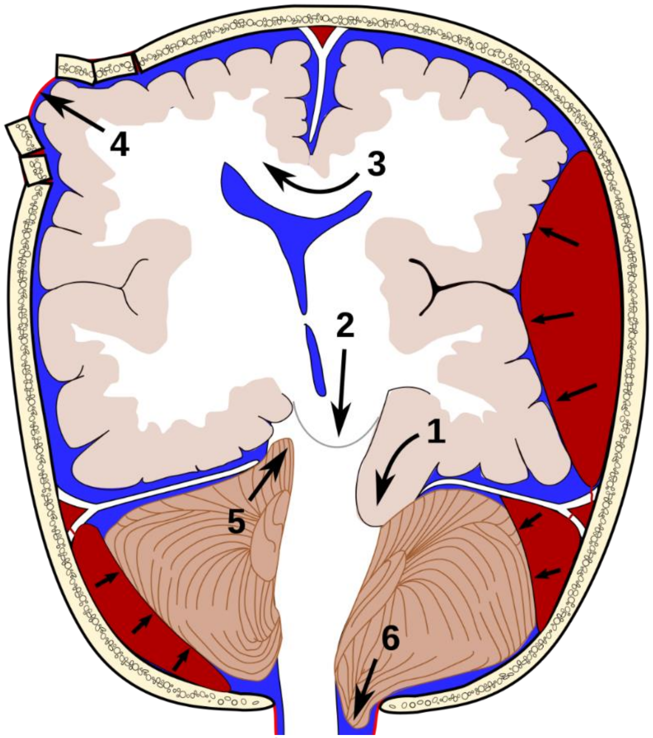
ICP/cerebral blood flow relationship

- Cerebral perfusion pressure (CPP)↓ as ICP↑
  - Crossing point = stop cerebral blood circulation = brain death

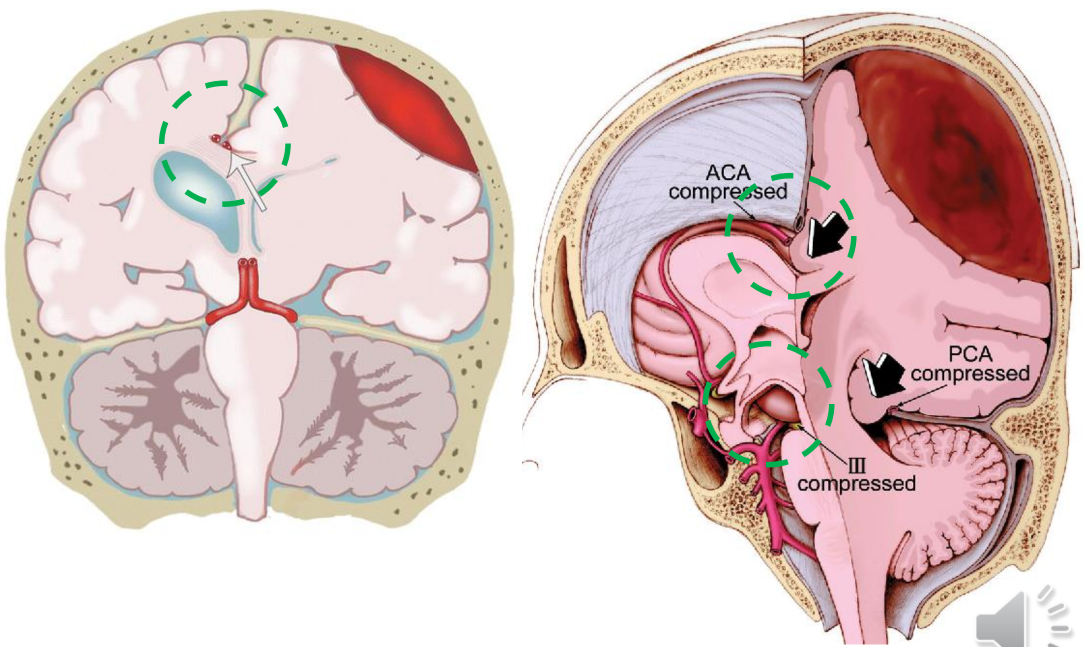


Brain herniations

- Intracranial space = partition = pressure increases in one compartment = displacement of nerve tissue = brain herniations
  - Compression of nerve tissue + vessels against falx and tentorium = added cerebral infarctions



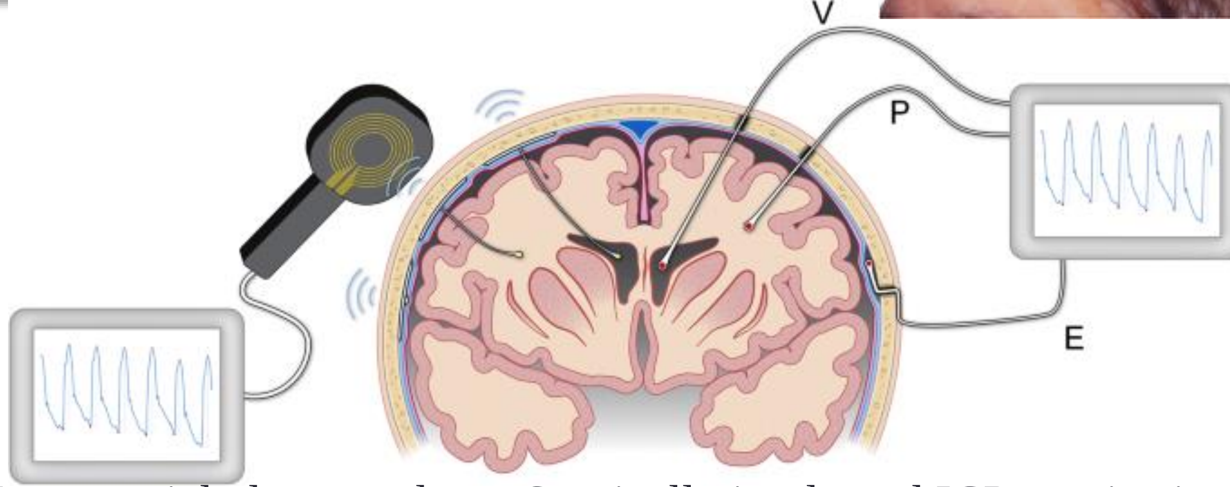
Types of brain herniations



Compression of cerebral arteries = brain infarctions

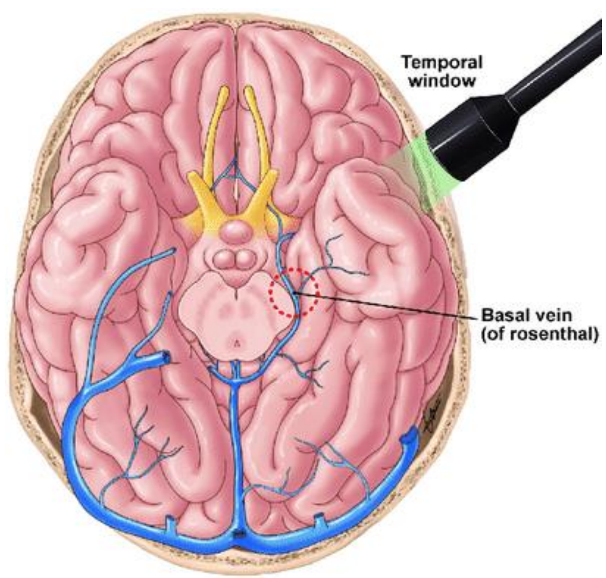
Intracranial pressure monitoring techniques

- Surgically implanted
  - Epidural (E)
  - Subdural
  - Intraparenchymal (P)
  - Intraventricular (V)
    - Allows CSF drainage → ICP reduction
- Percutaneous (ultrasound)
  - Transcranial
  - Optic nerve

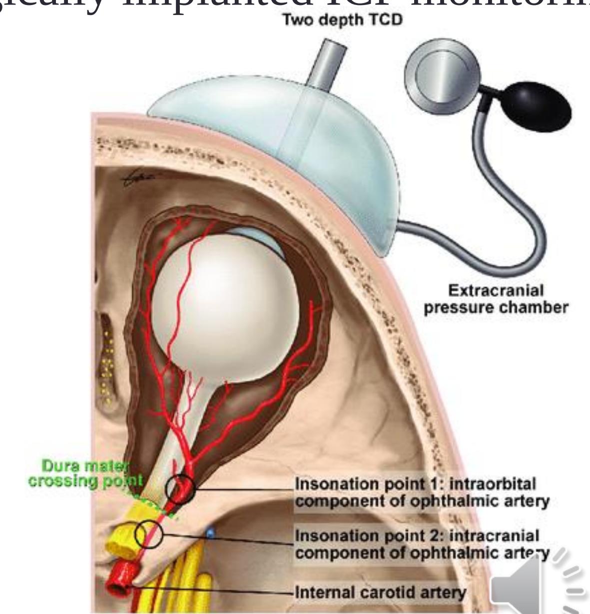


Transcranial ultrasound

Surgically implanted ICP monitoring



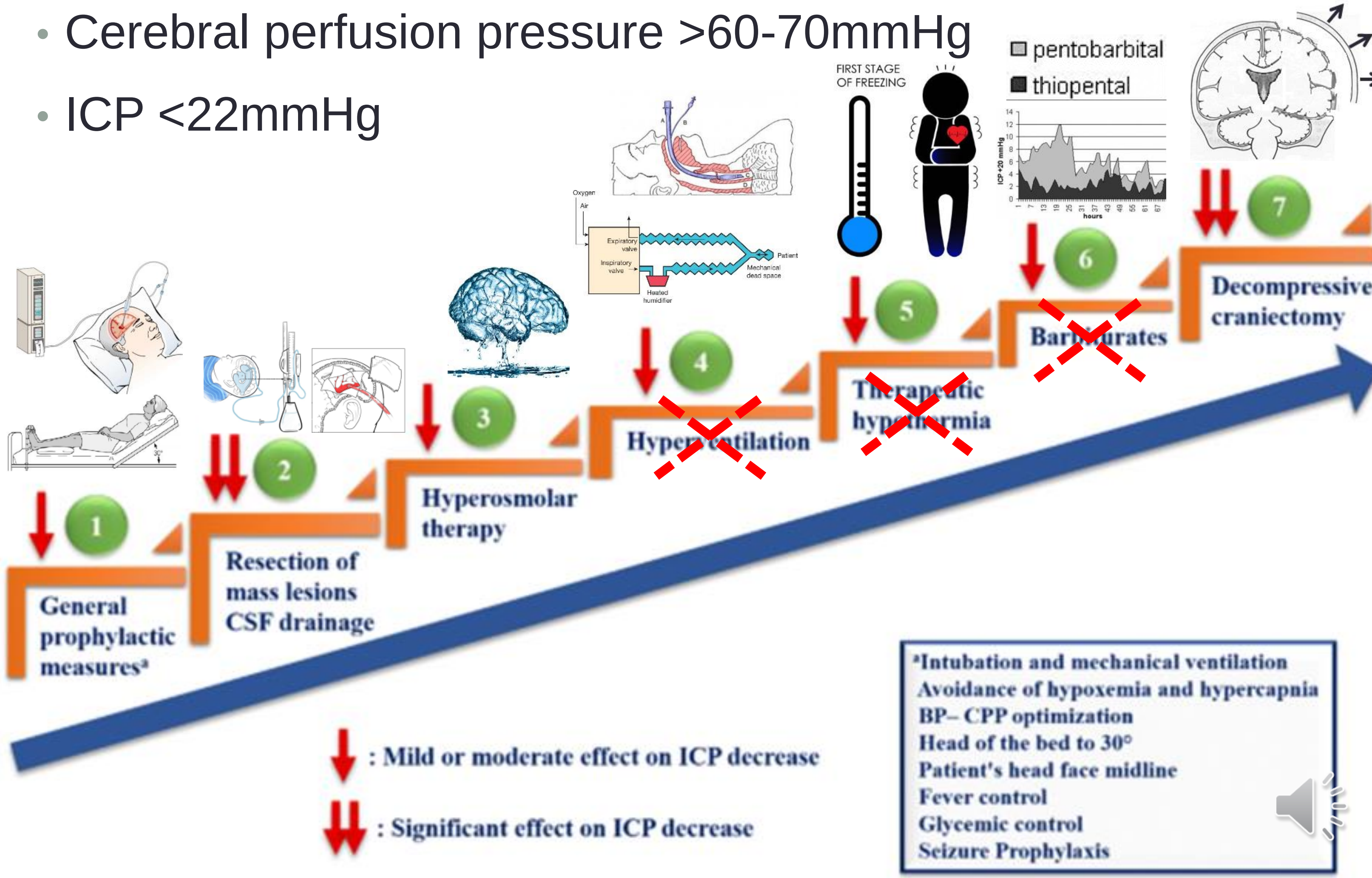
Transcranial ultrasound



Optic nerve ultrasound

Goal in treatment ↑ICP

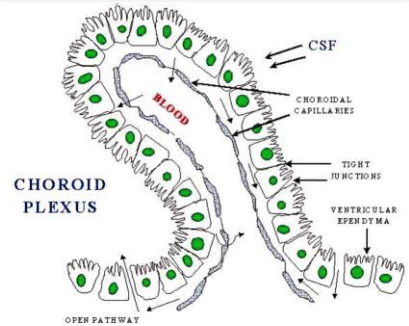
- Cerebral perfusion pressure >60-70mmHg
- ICP <22mmHg

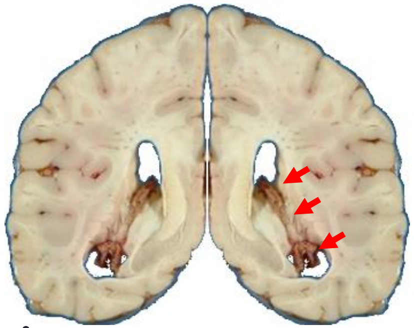


Cerebrospinal fluid (CSF)

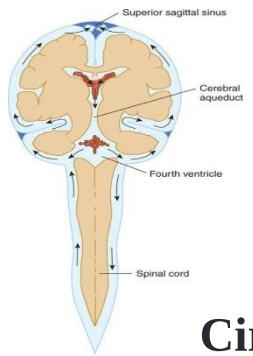
- **Production** ~ 200-500ml/day
  - Child 8 ml/h
  - Adult 20 ml/h
- **Total volume**
  - Newborn 20-50 ml
  - Baby 40-60 ml
  - Child 60-120 ml
  - Adult 120-200 ml
- **Functions**
  - Protection against blows to brain & spinal cord
  - Transport of waste products
  - Give space to avoid ⇑ICP
- **Circulation**
  - Ventricles ⇨ subarachnoid space ⇨ arachnoid villi ⇨ cerebral venous sinuses

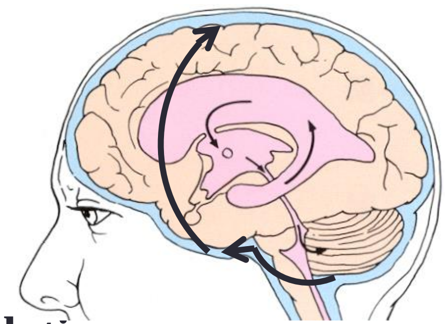
REMEMBER?



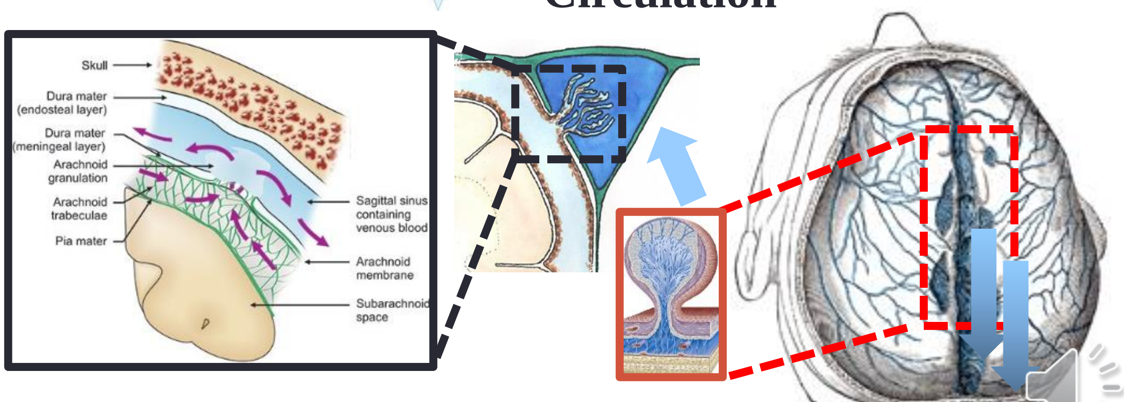


Production





Circulation

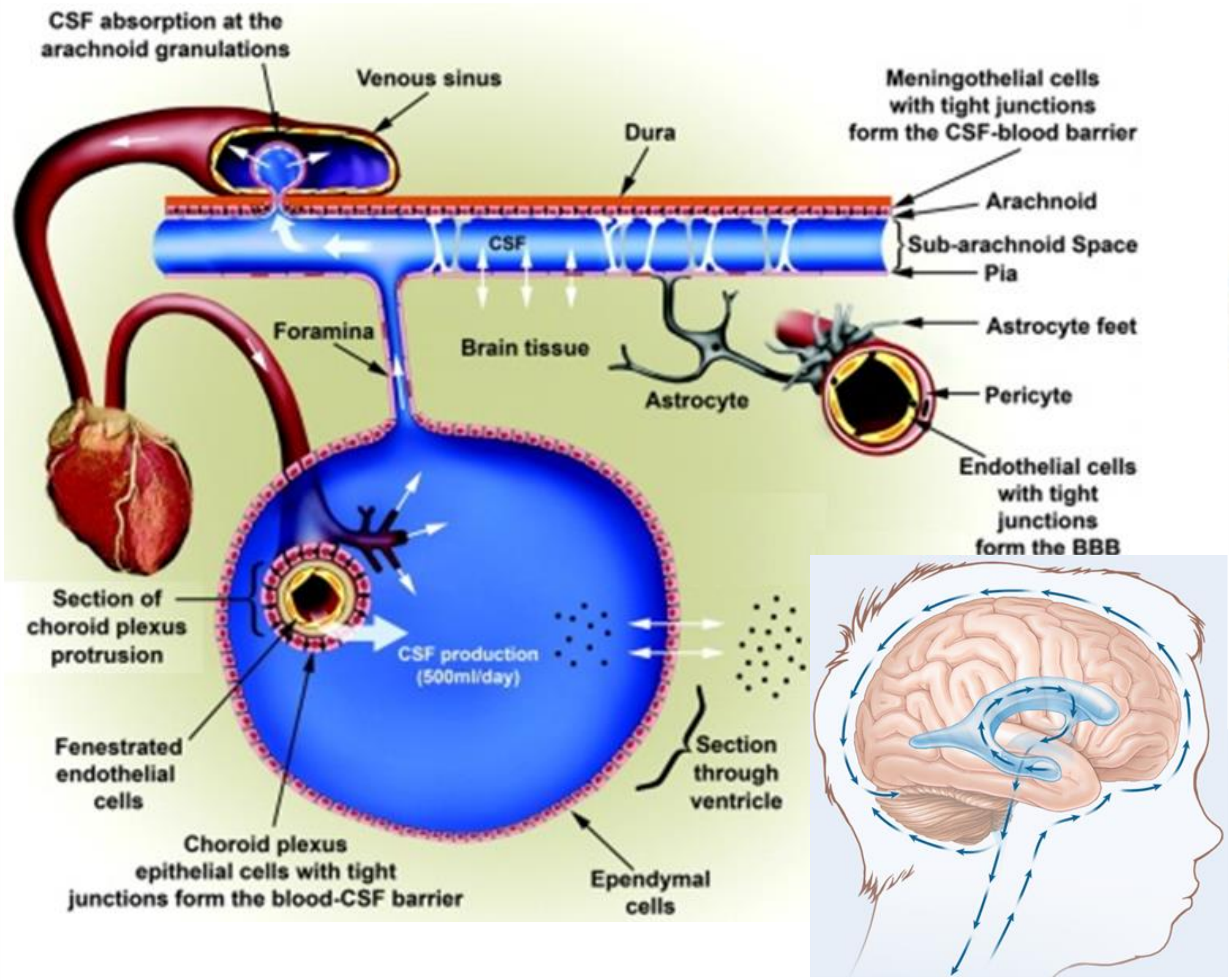


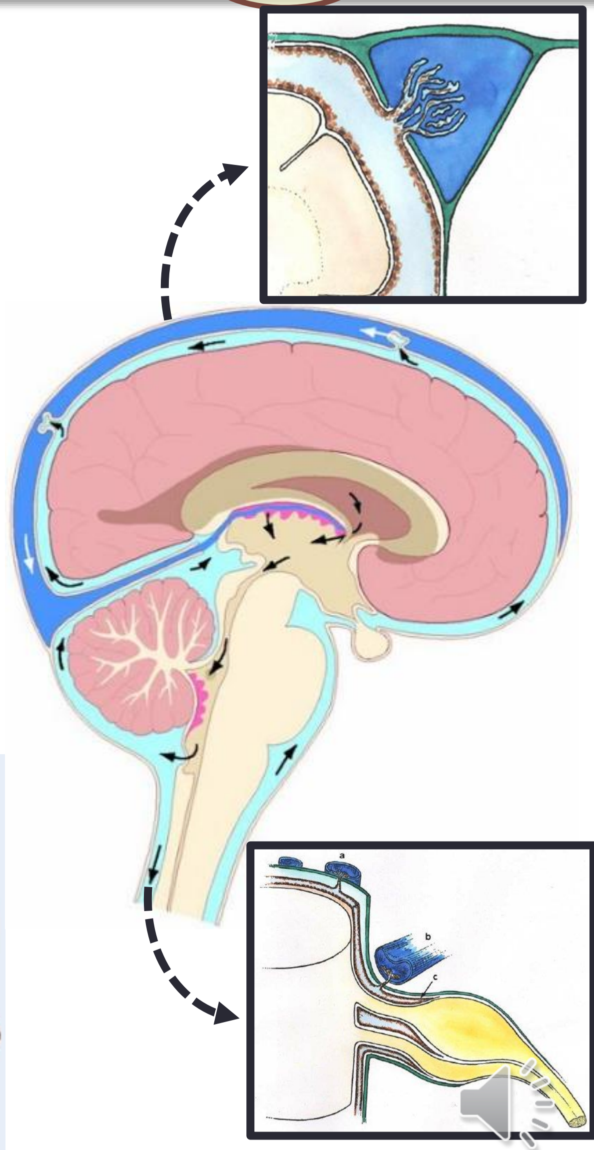
Reabsorption

CSF circulation

REMEMBER?

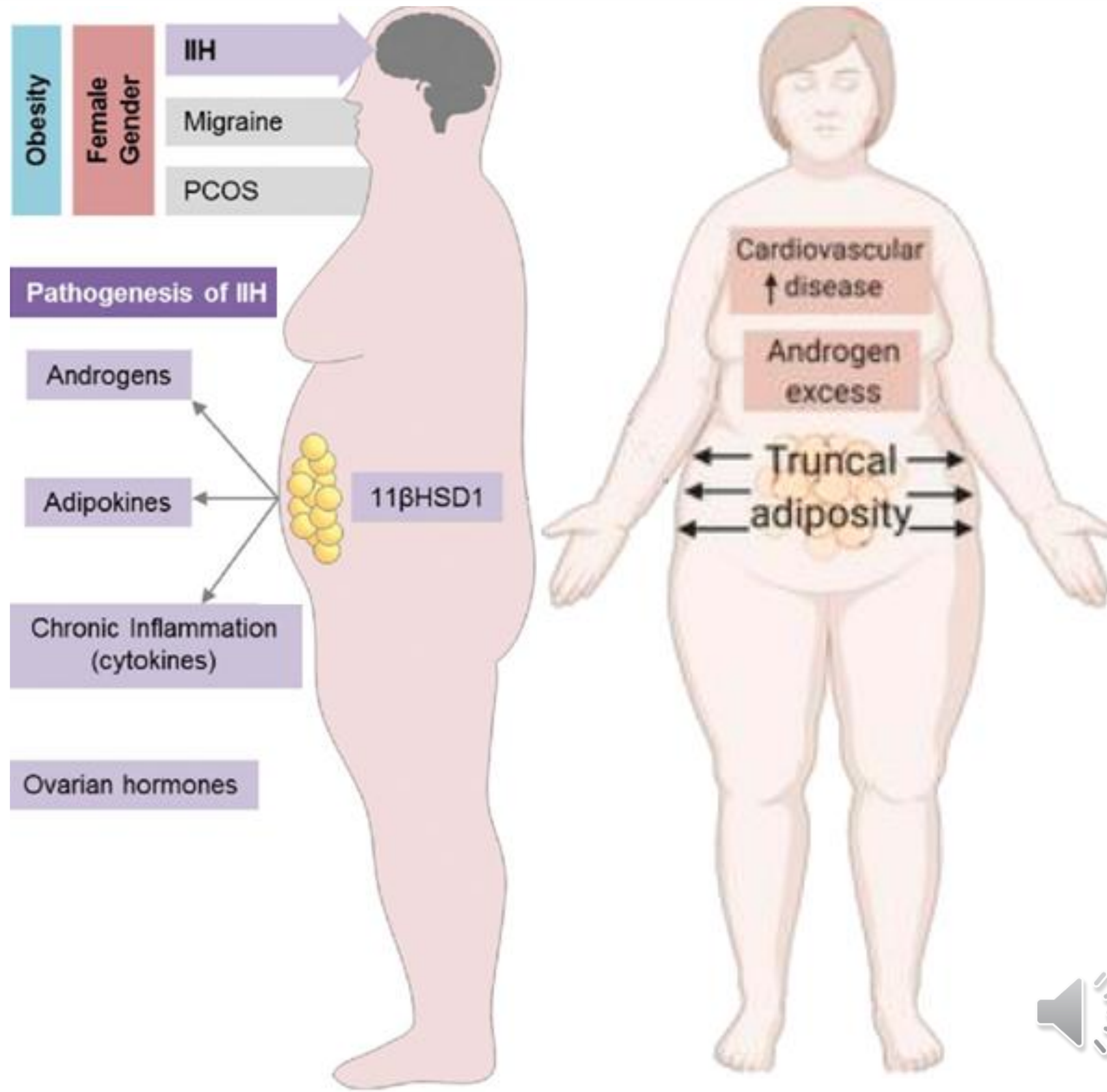
- Circulation through subarachnoid space
- Reabsorption in arachnoid villi





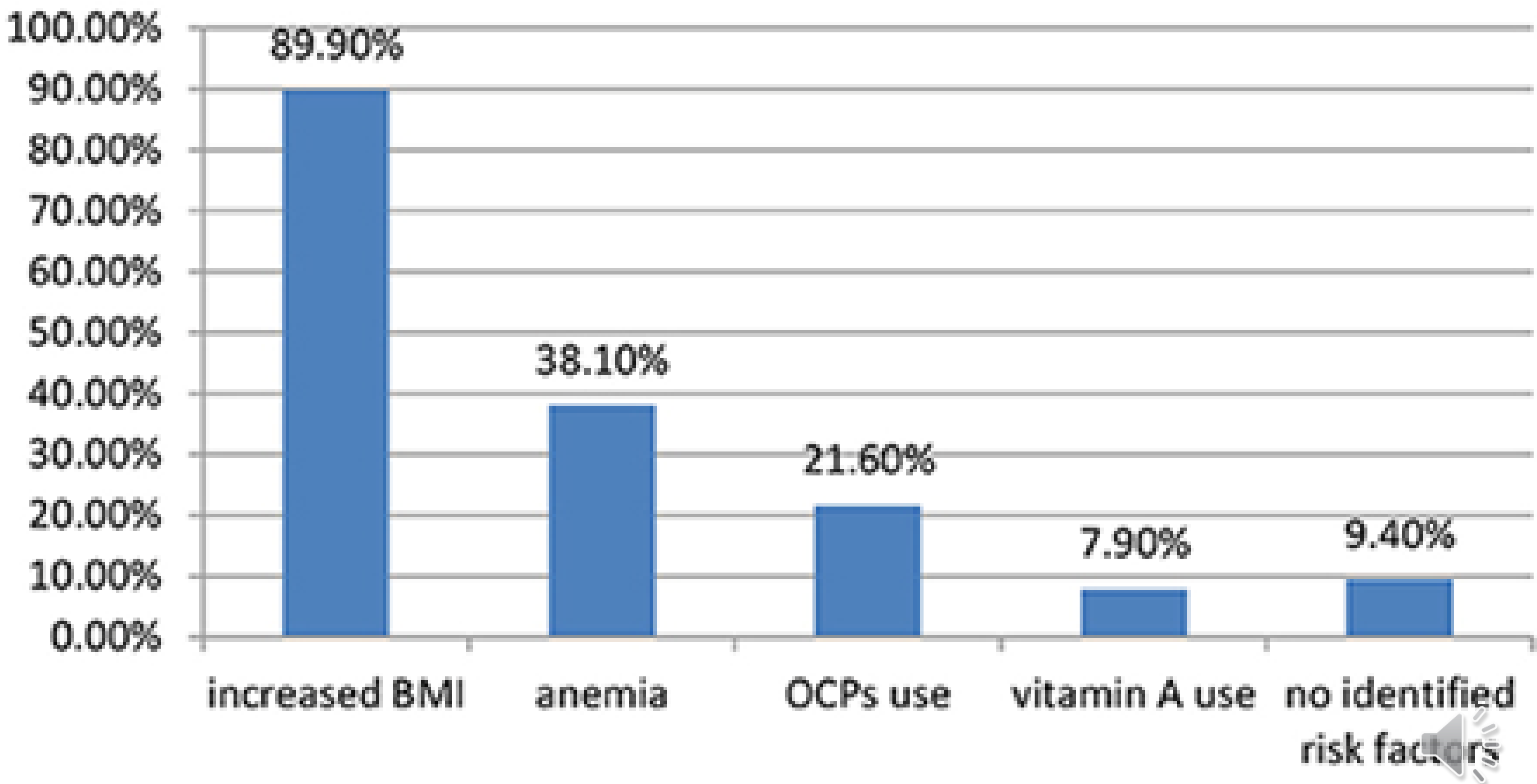
Benign intracranial hypertension

- ↑ICP of cause not always known
- Not as benign = possibility of vision loss
- Physiopathology
  - ♀
  - Obesity
  - Contraceptives
  - Other drugs



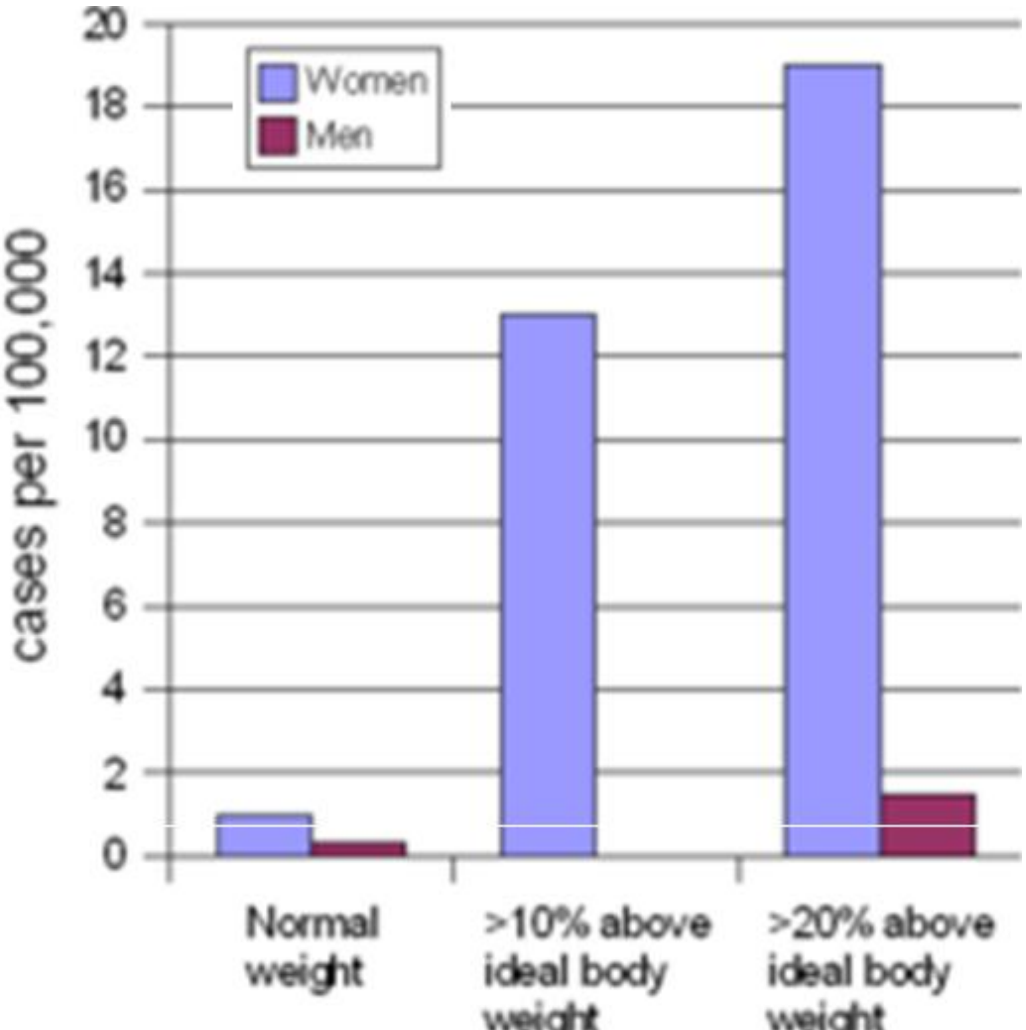
Benign intracranial hypertension: risk factors

- Obesity and contraceptive medication (OCP) the most important thing

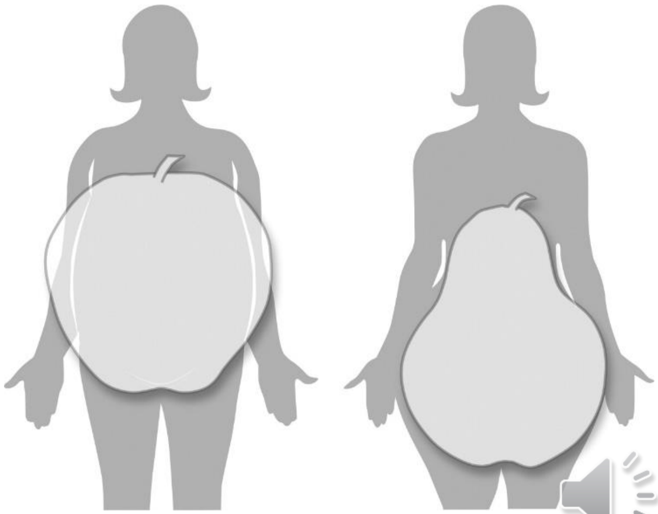


Obesity and benign intracranial hypertension (BIH) relationship

- Clearer in ♀ than in ♂



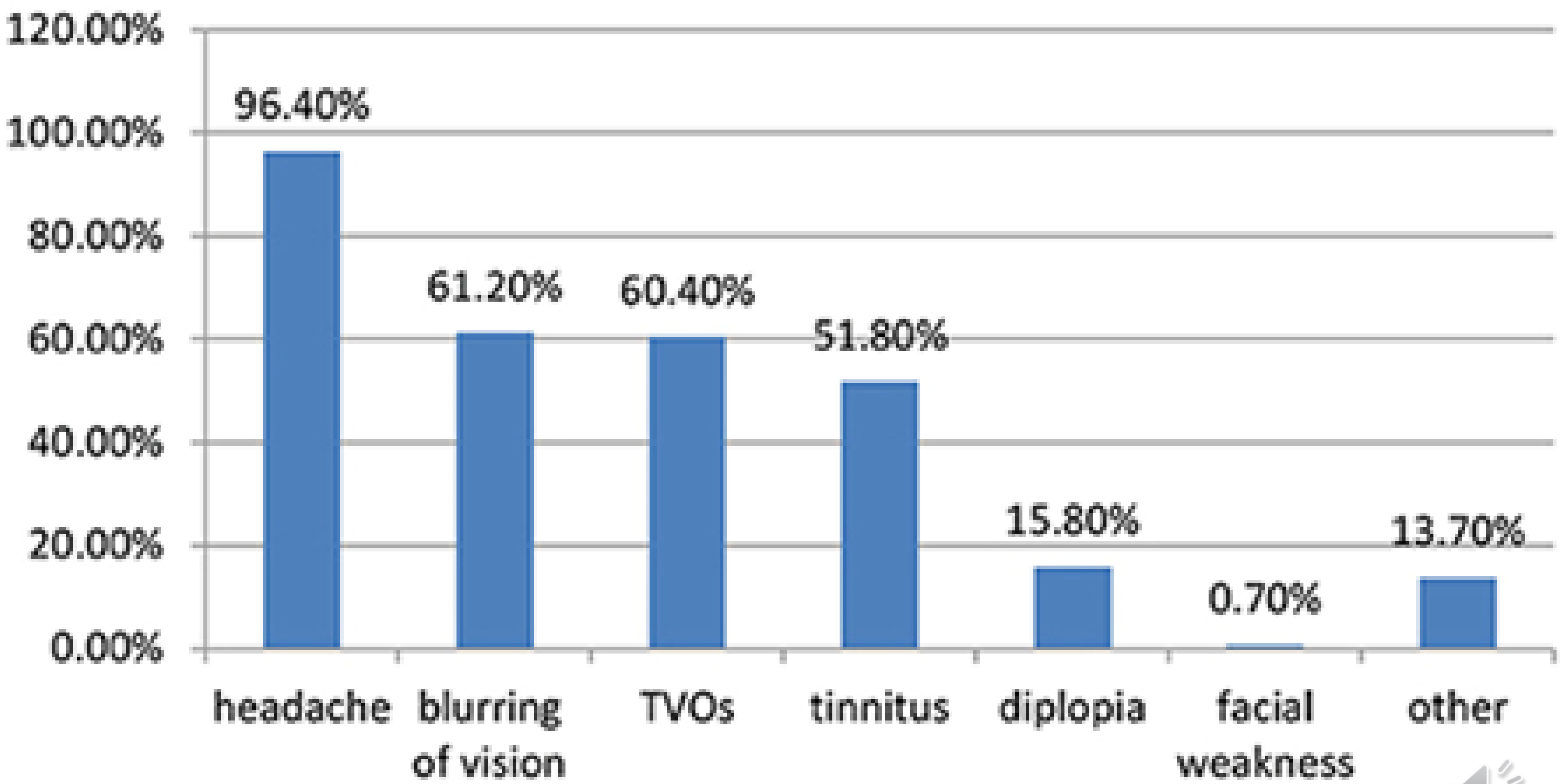
Obesity type & BIH



↑incidence

Benign intracranial hypertension: clinical features

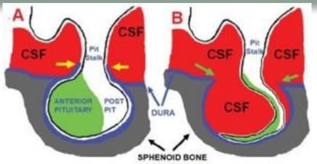
- Not as benign = visual acuity loss in 61%



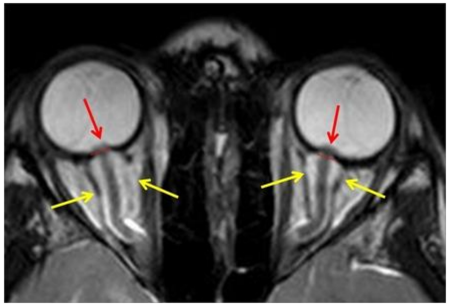
TVO = transient visual obscuration

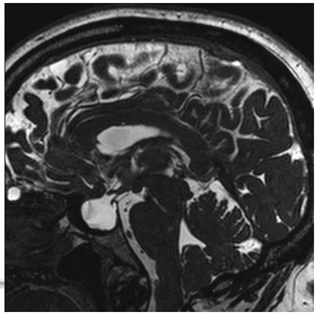
Benign intracranial hypertension: MRI

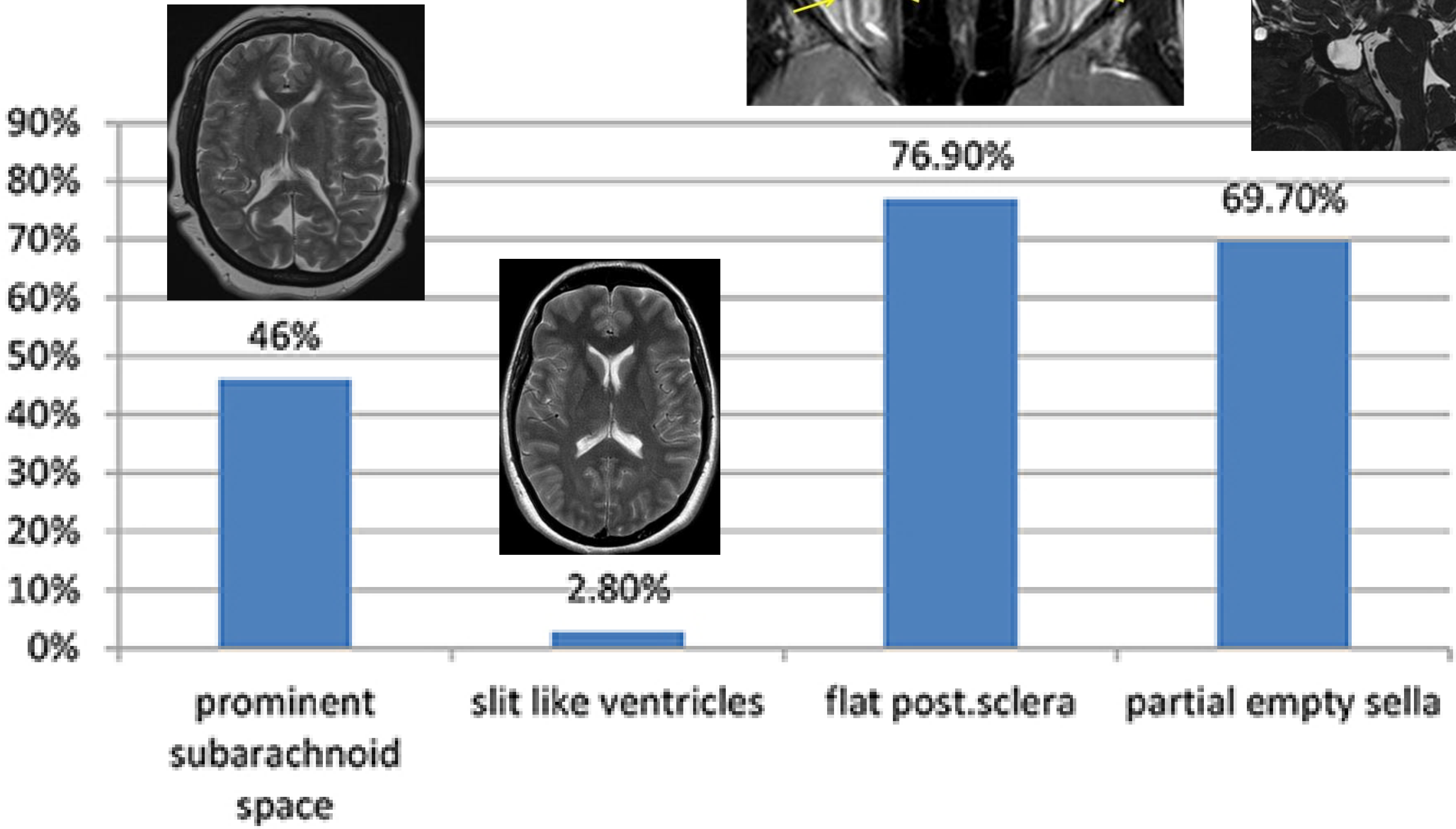
- Normally only brain MRI is asked = eye changes go undiagnosed



Normal Empty sella

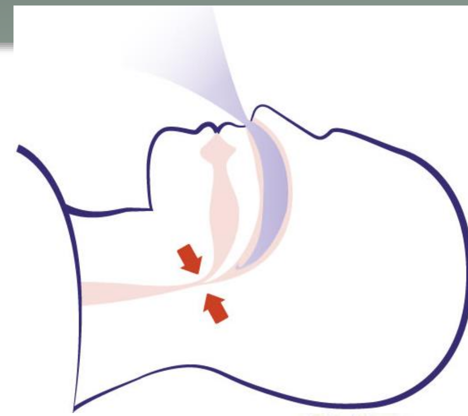


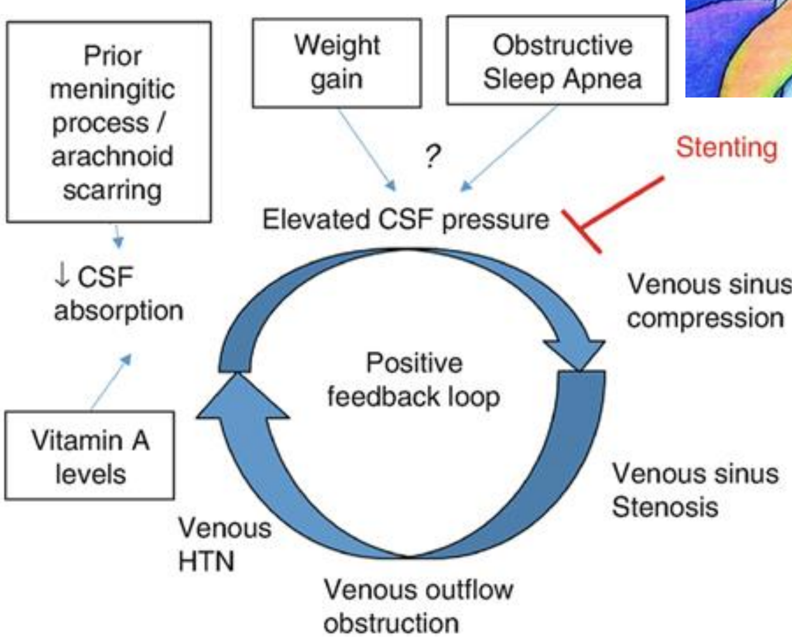


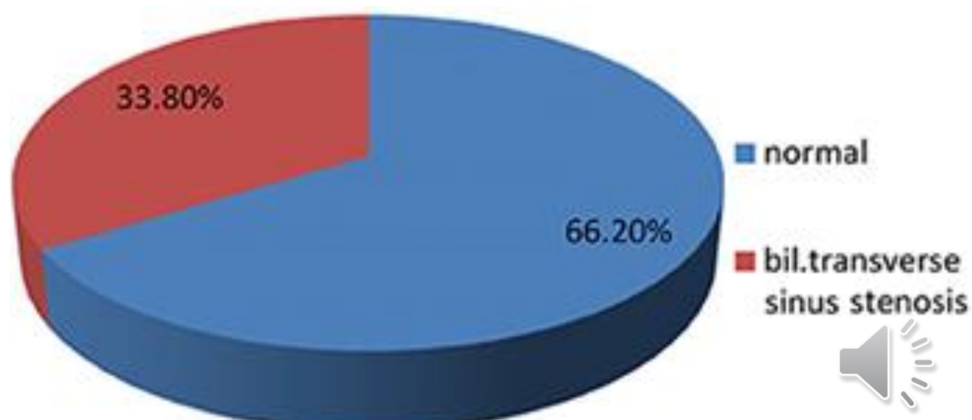


Benign intracranial hypertension: causes

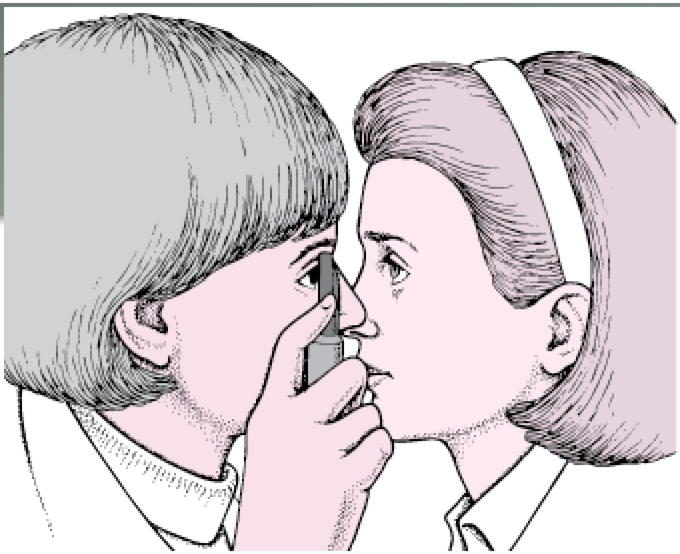
- Sleep apnea
- Stenosis intracranial venous sinuses







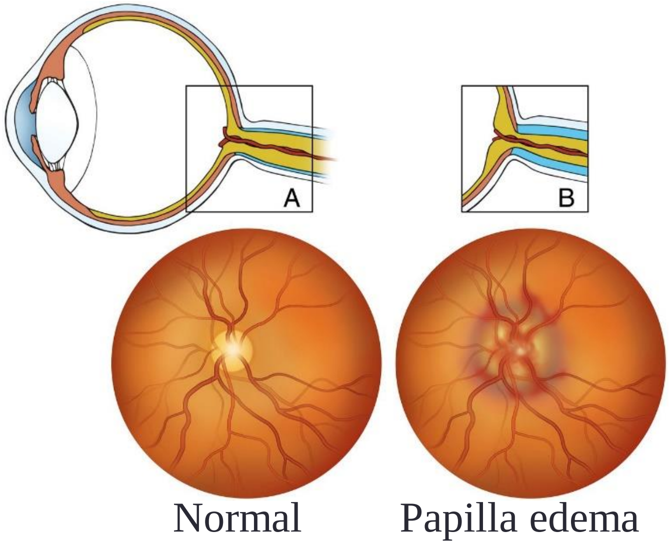
Benign intracranial hypertension: eye fundus

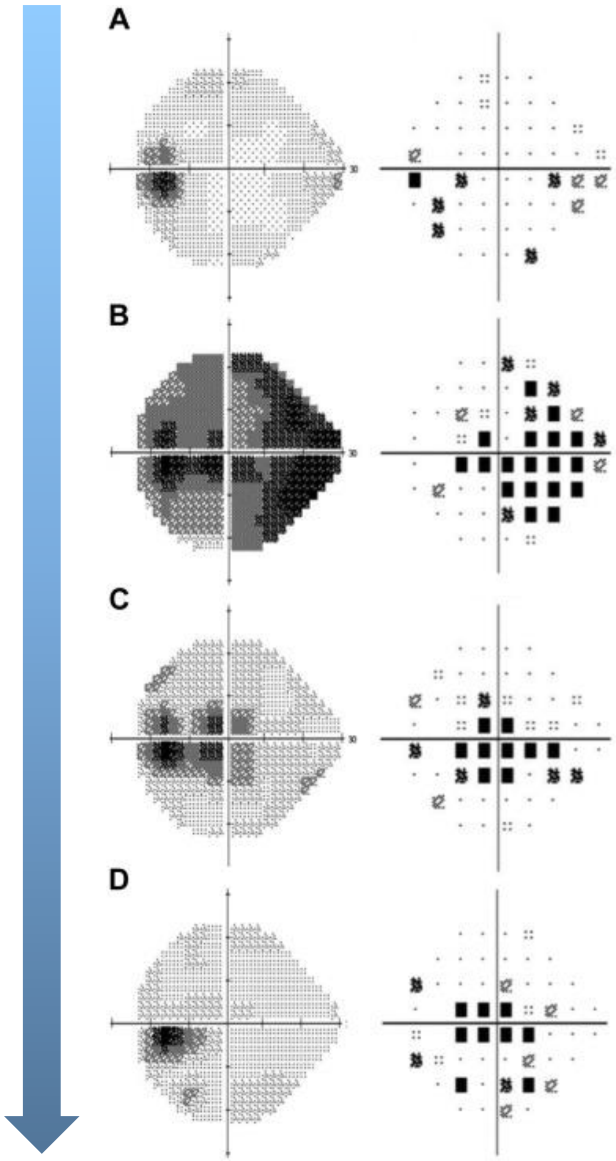


- Not usually done in out-patient consultations = risk not diagnosing the condition

Benign intracranial hypertension: loss of visual acuity

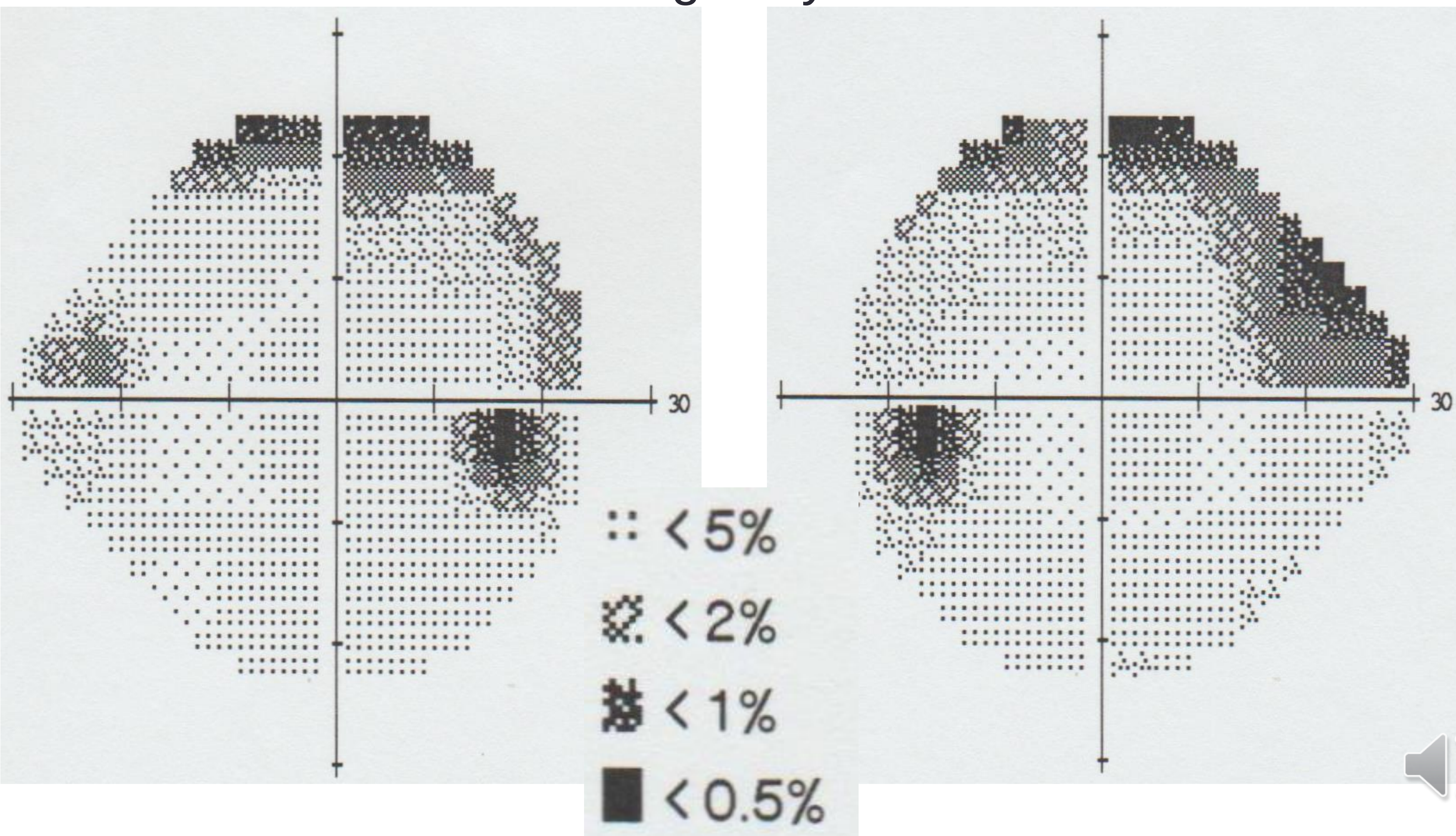
- Affects peripheral visual fields
- It can go unnoticed if serial campimetry exams are not done
- This disease is not so benign





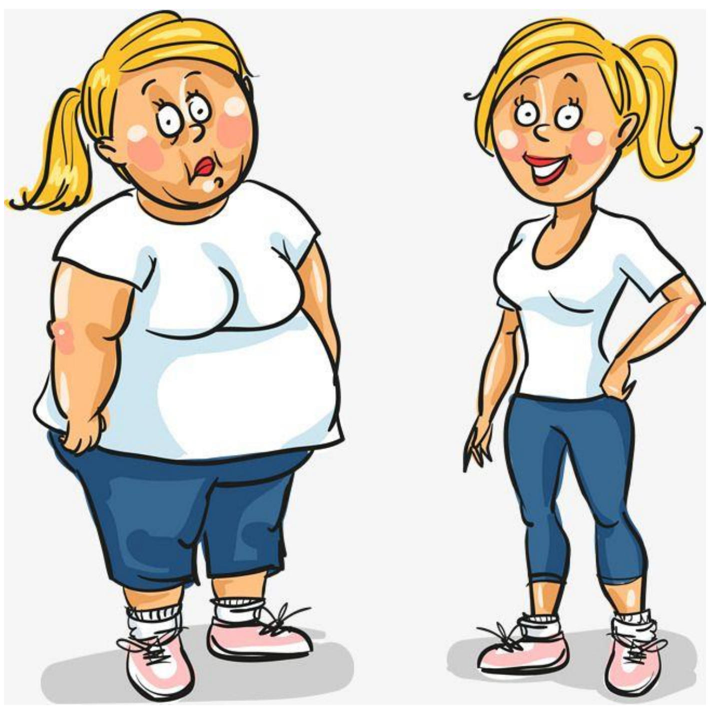
Benign intracranial hypertension: visual acuity loss pattern

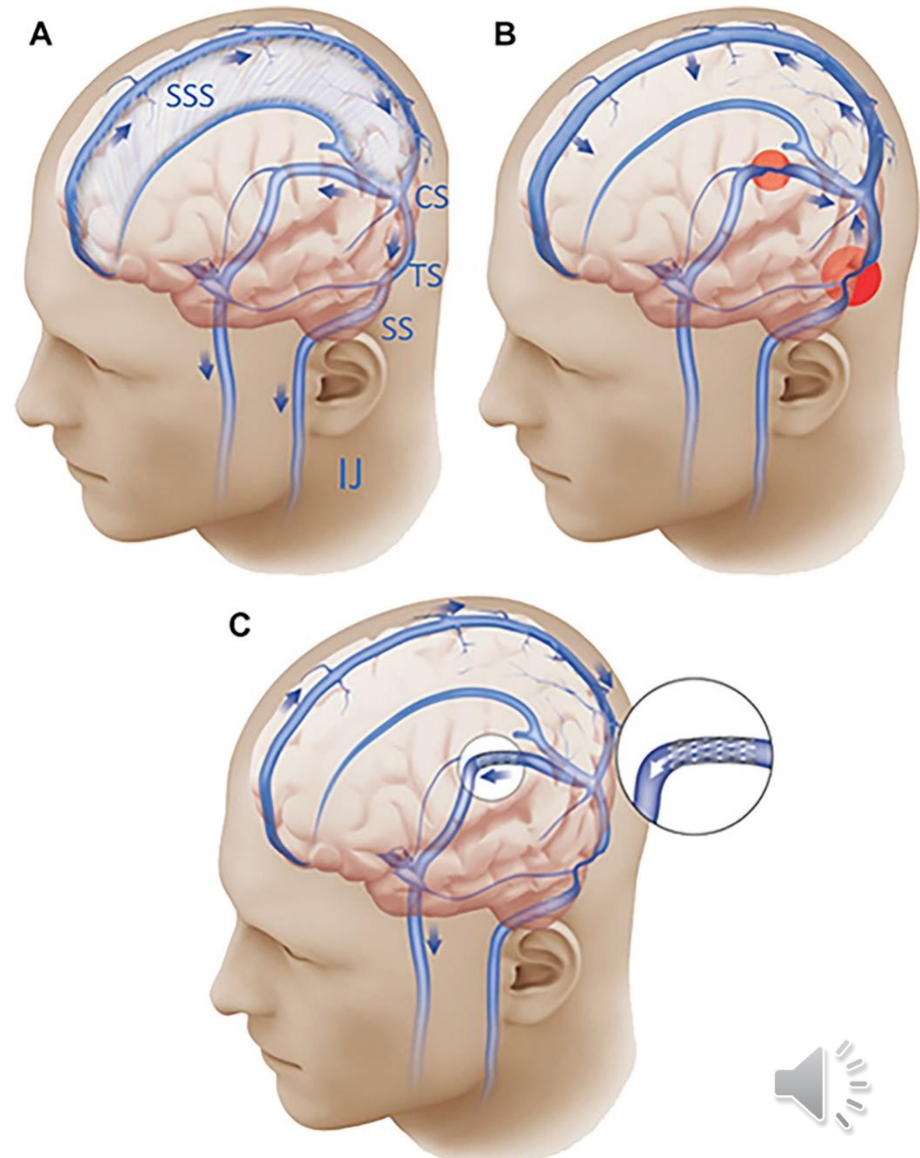
- Changes will go initially unnoticed unless visual field examinations are done regularly



Benign intracranial hypertension: treatment

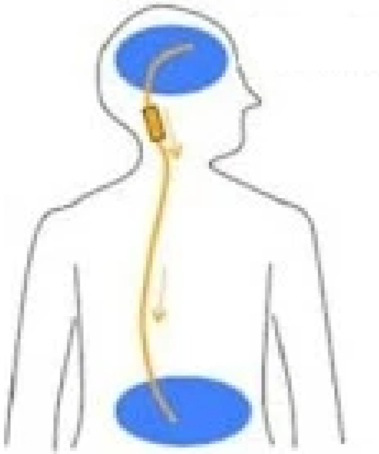
- Withdraw any suspicious drug
- Lose weight
- Stenting stenosis intracranial venous sinuses



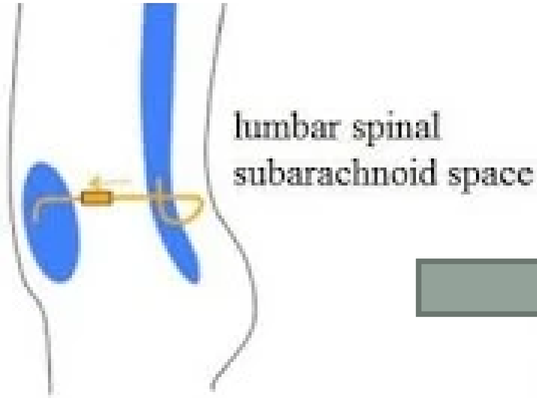


Benign intracranial hypertension: CSF shunt

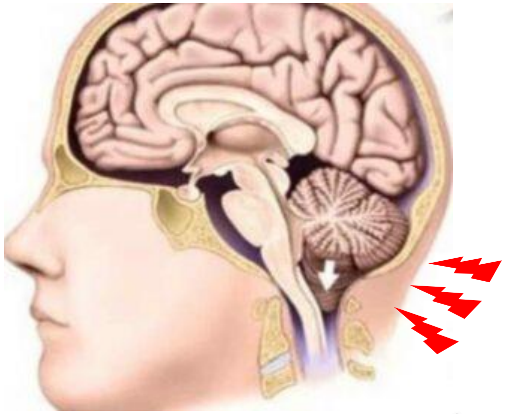
- The ideal is the ventricle-peritoneal
  - Small ventricles = difficult to cannulate
- Lumbo-peritoneal technically easier
  - Induces Chiari type 1 malformation with nuchal headache



Ventriculo-peritoneal shunt



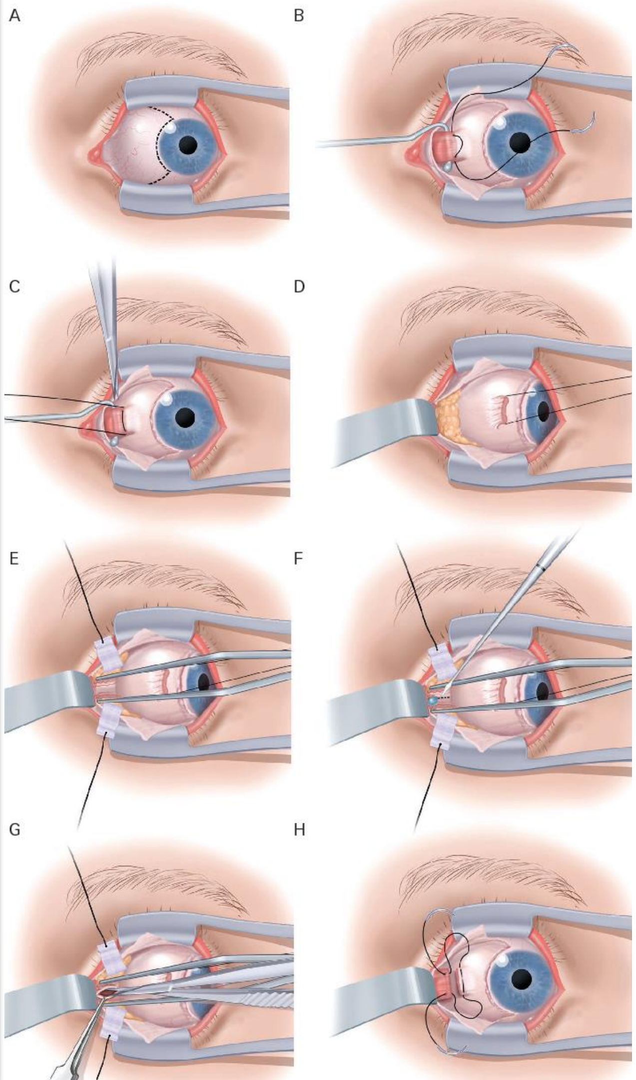
Lumbo-peritoneal shunt

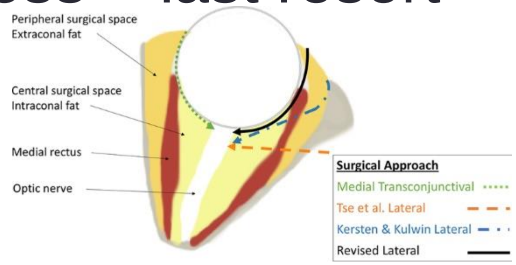


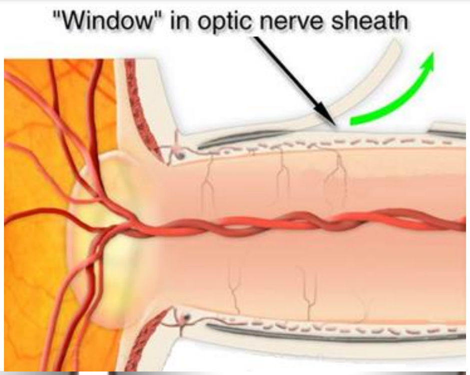
Chiari type 1 malformation

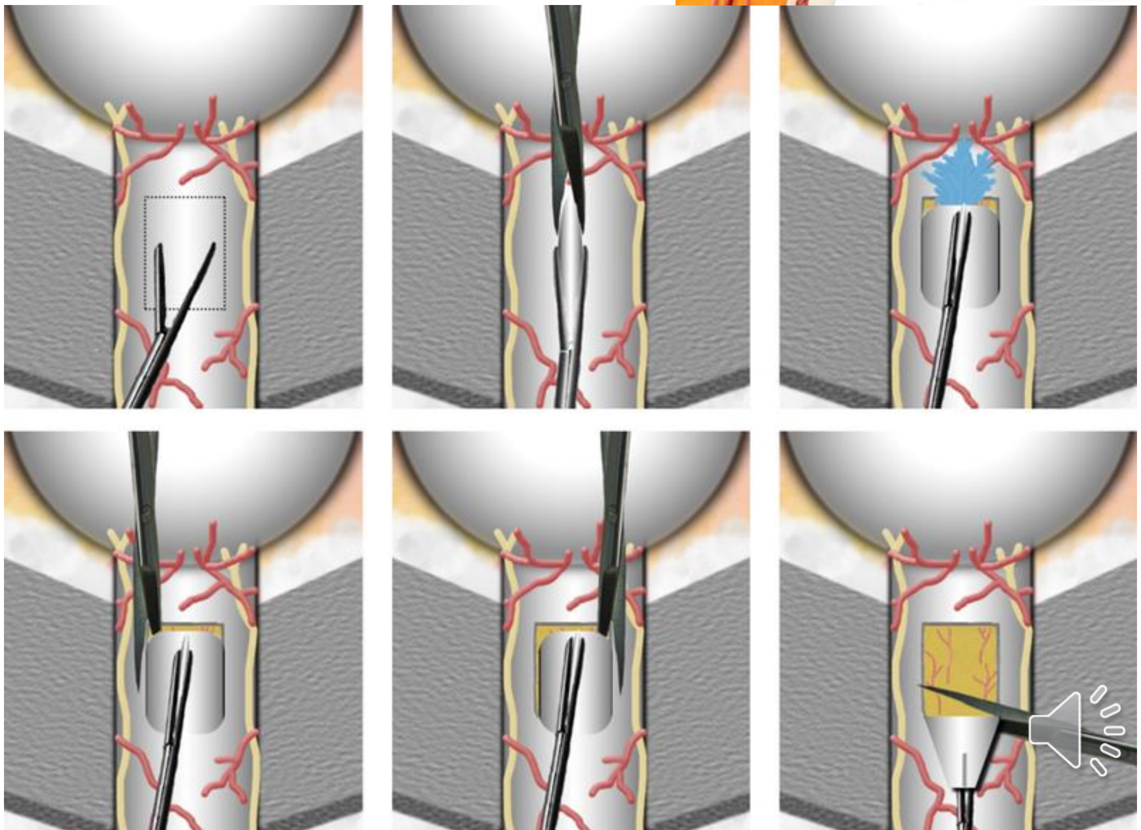
Benign intracranial hypertension: optic nerve sheath decompression

- Progressive vision loss = last resort





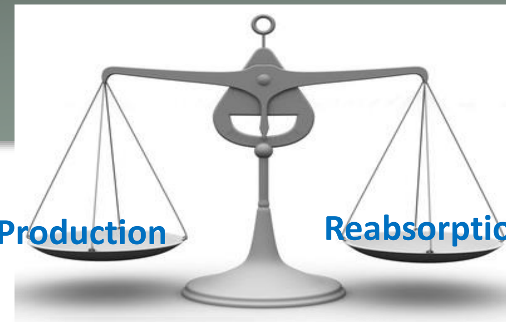


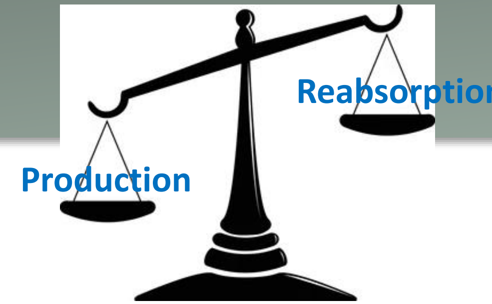


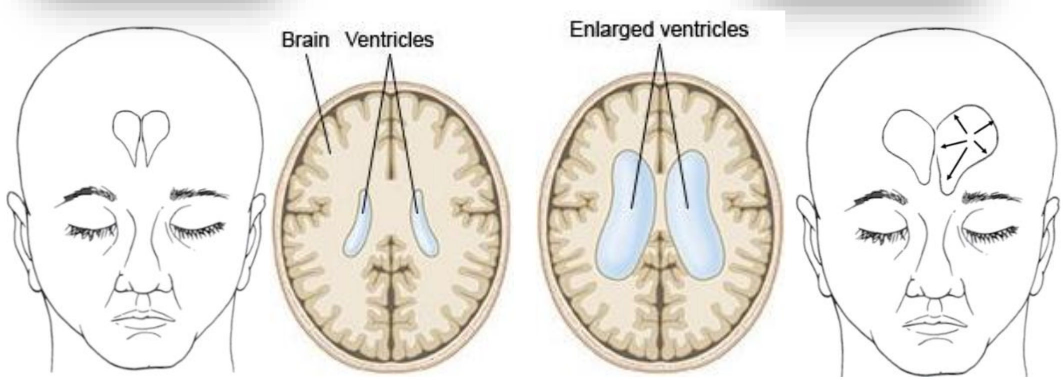
HYDROCEPHALUS

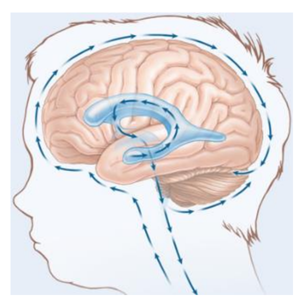
- Hydrocephalus = ⇑ CSF at intracranial level
  - Hydros = water
  - Kefalos = head
- **Causes**
  - ⇑ production (very rare)
  - Circulation block
    - Most frequent situation
  - ⇓ drainage (reabsorption)

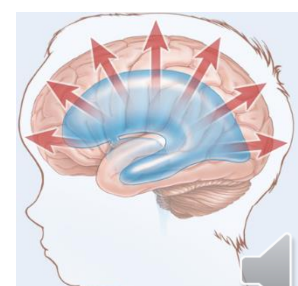
REMEMBER?





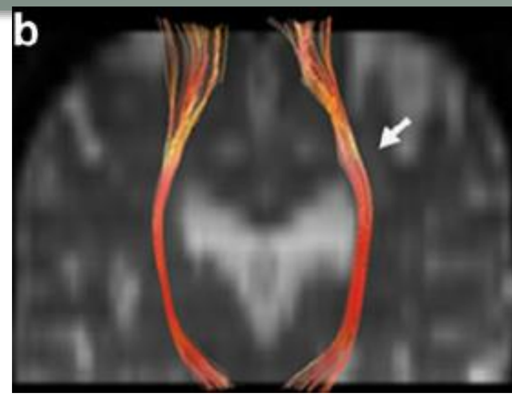



Normal

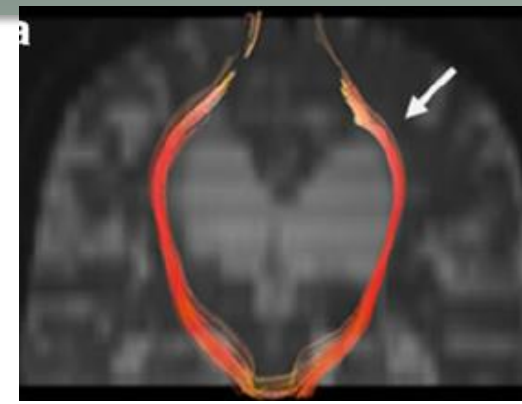

Hydrocephalus

Hydrocephalus and nerve tracts

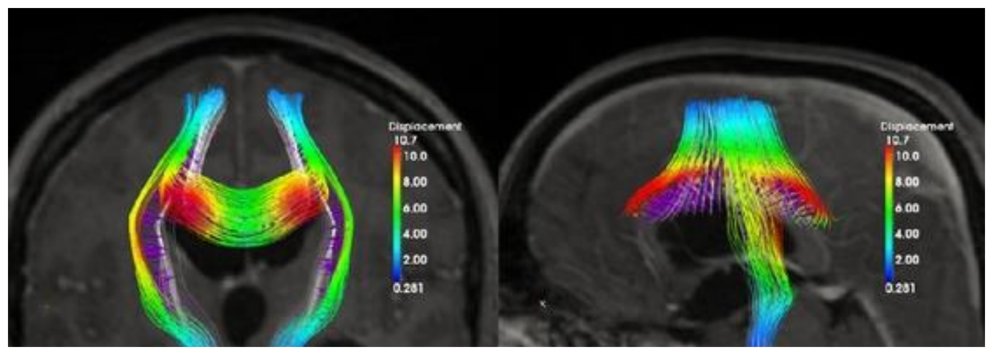
- Lateral ventricles dilation = periventricular nerve fiber stretching
  - Affects lower limb and frontal-basal fibers (gait impairment, urinary incontinence + temporo-spatial disorientation)



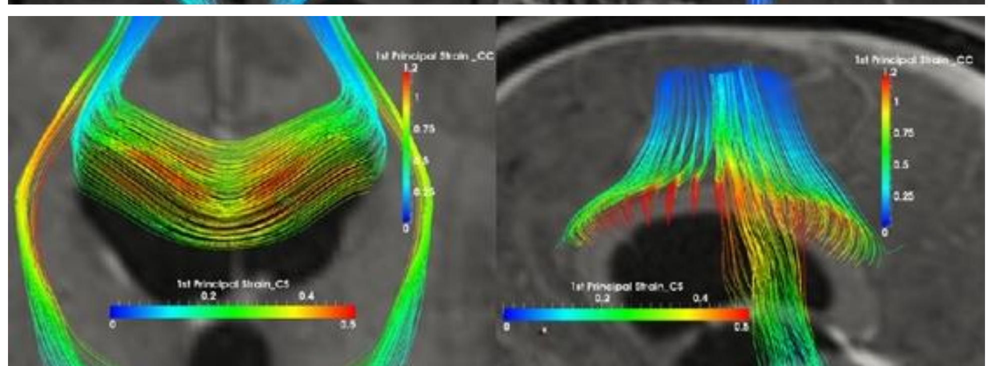
Normal (coronal cut MRI)



Hydrocephalus



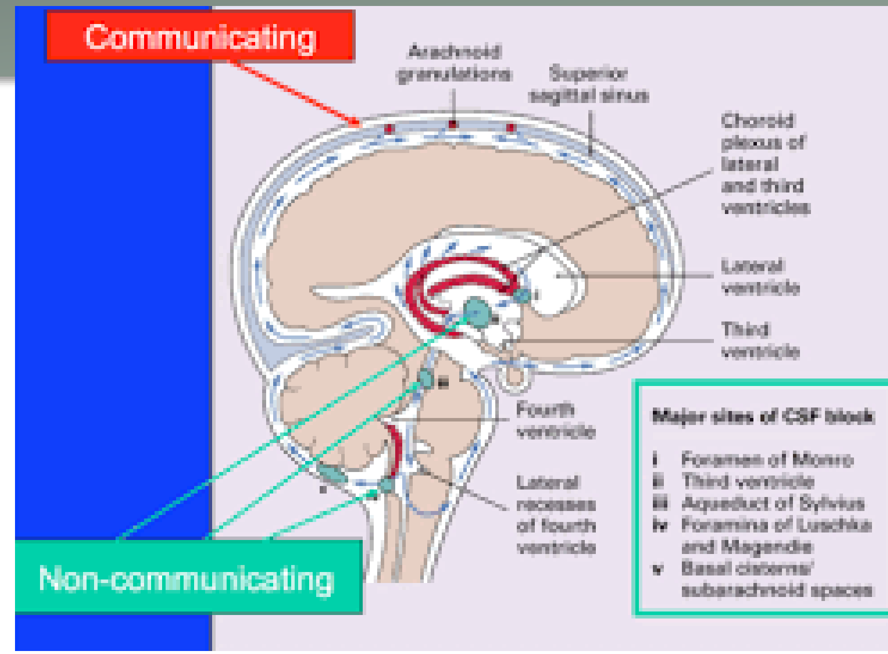
Hydrocephalus



Hydrocephalus

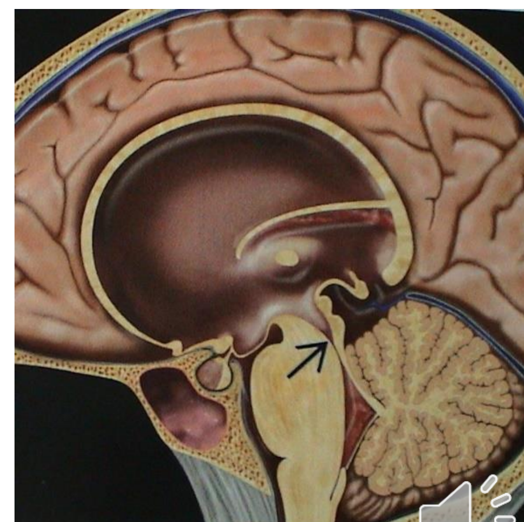
Types of hydrocephalus

- According to type
  - **Communicating**
    - CSF block in subarachnoid space
  - **Non-communicating**
    - CSF flow block inside ventricles
      - Foramen of Monro
      - Third ventricle
      - Aqueduct of Silvius
      - Fourth ventricle



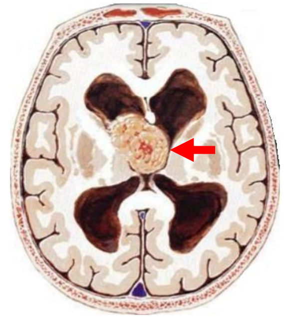
- According to presentation time
  - **Congenital**
    - Causes
      - Intraventricular haemorrhage (prematurity)
      - Stenosis aqueduct of Silvius
  - **Acquired**

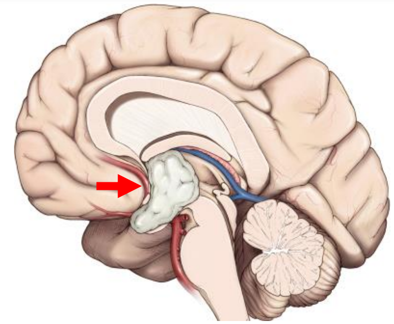

Prematurity

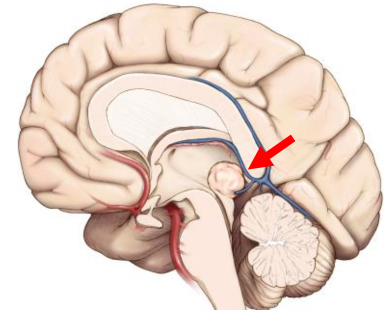

Silvio aqueduct stenosis

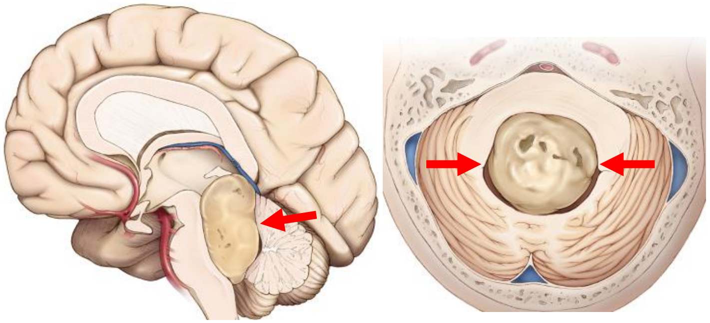
Non-communicating hydrocephalus: obstruction to CSF circulation inside ventricles

- **Tumors**/parasitic cysts
  - Lateral ventricles
  - Foramen of Monro
  - Third ventricle
  - Pineal region
  - Fourth ventricle
- **Malformations**
  - Stenosis aqueduct of Silvius
  - Atresia foramina Luschka and Magendie
  - Dandy-Walker malformation

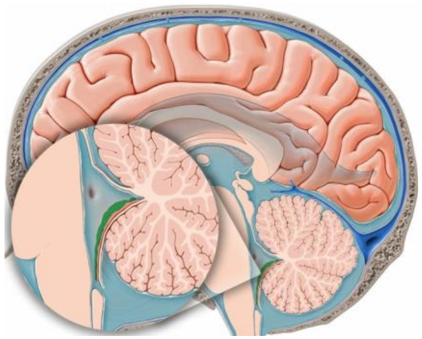

Monro foramen tumor

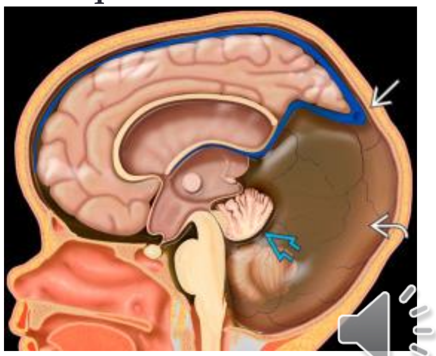

Tumor third ventricle


Pineal region tumor


Fourth ventricle tumor

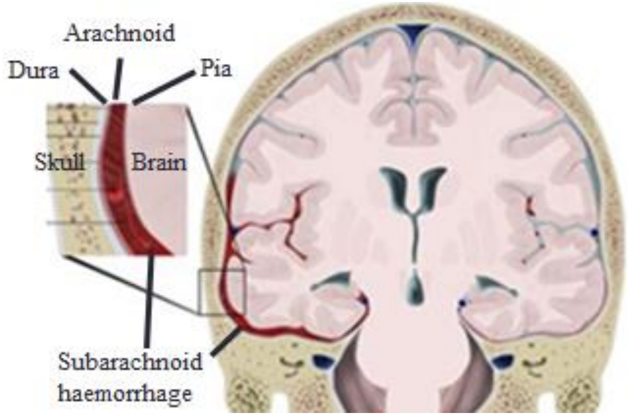

Silvio aqueduct stenosis


Atresia Luschka and Magendie

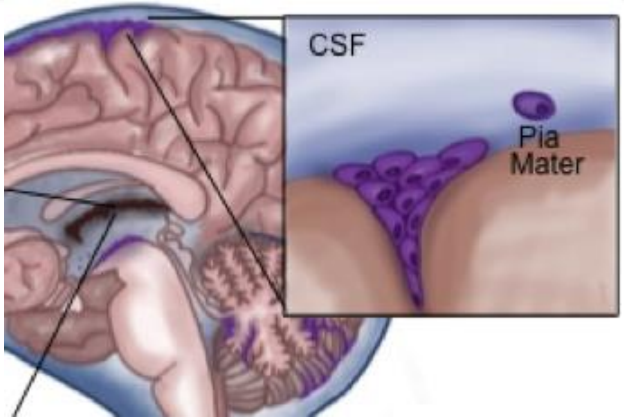

Dandy-Walker

Communicating hydrocephalus = CSF circulation obstruction in subarachnoid space or reabsorption impairment

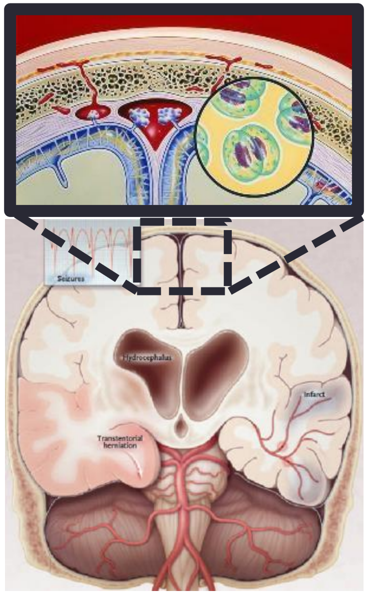
- Subarachnoid haemorrhage
- Meningitis
- Meningeal carcinomatosis
- Meningeal lymphomatosis
- Intracranial venous sinus thrombosis
- Dural arterio-venous fistulas



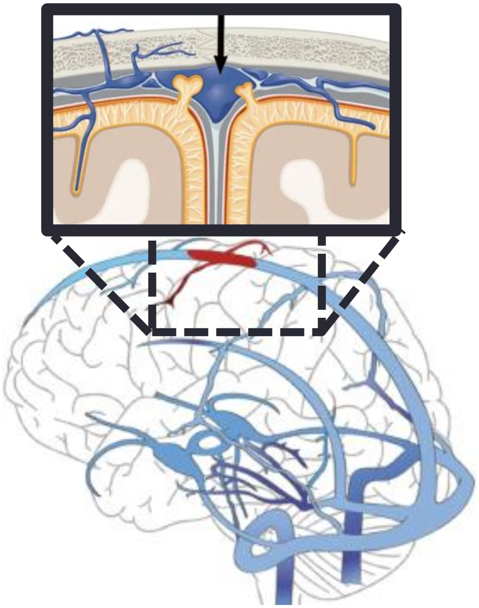
Subarachnoid haemorrhage



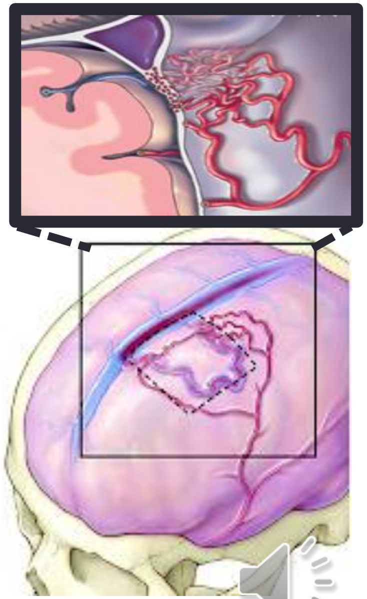
Meningeal carcinomatosis



Meningitis



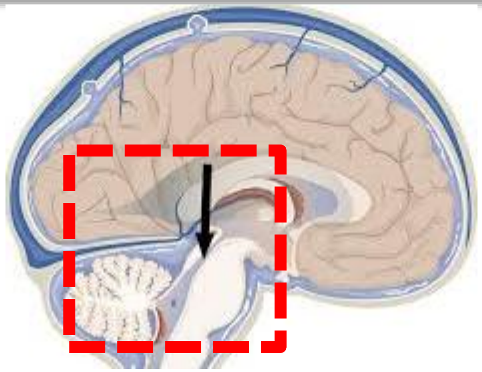
Venous sinus thrombosis

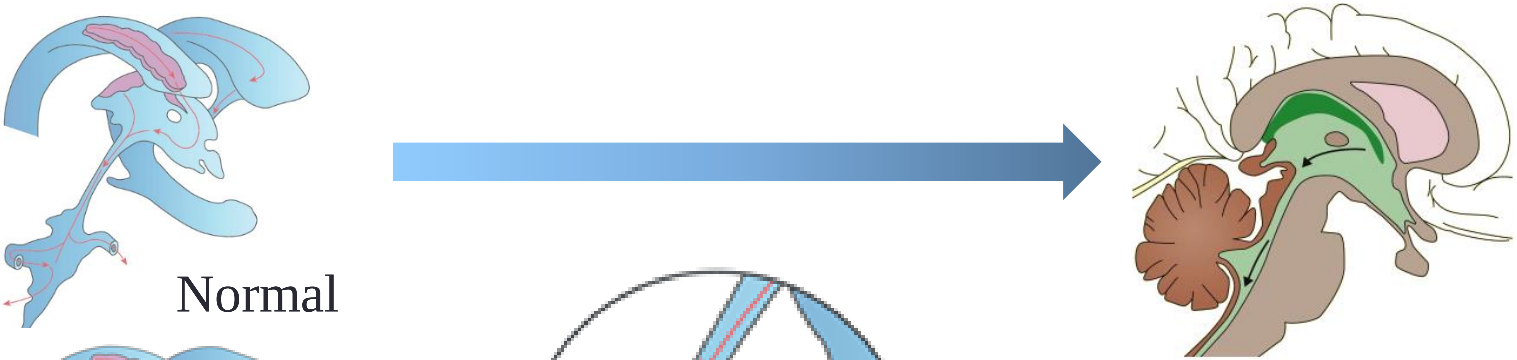


Dural AV fistula

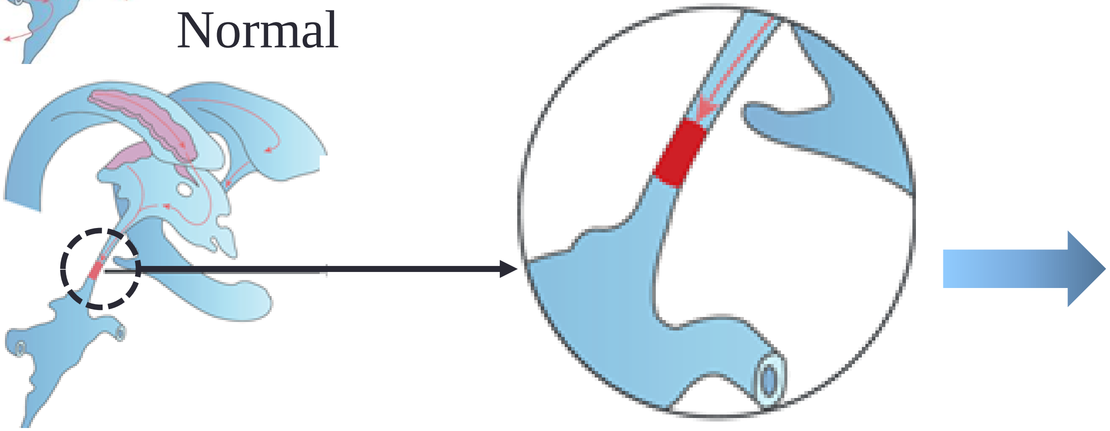
Stenosis of the aqueduct of Sylvius

- Congenital malformation
- Triventricular hydrocephalus = abnormal skull development
- Lack of nerve tissue development = slight psychomotor retardation
- Clinical symptoms often present after 40 years of age



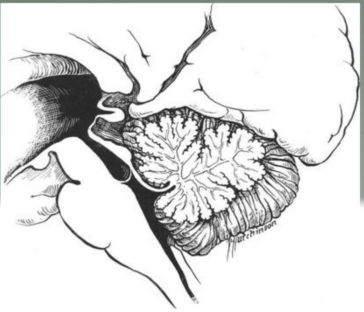


Normal

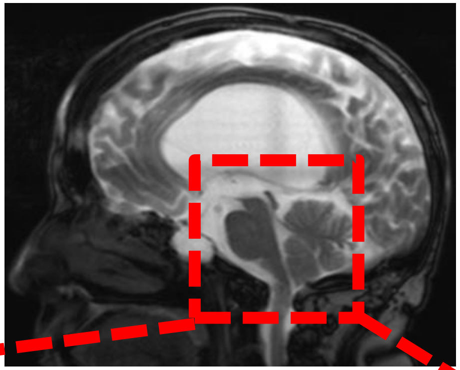


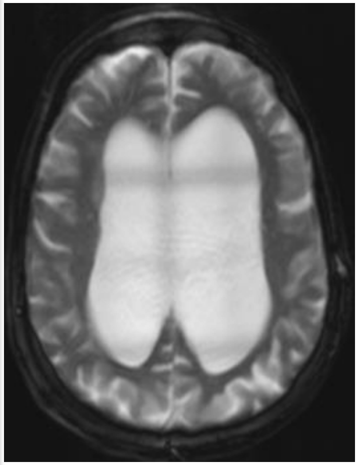
Stenosis of the aqueduct of Sylvius

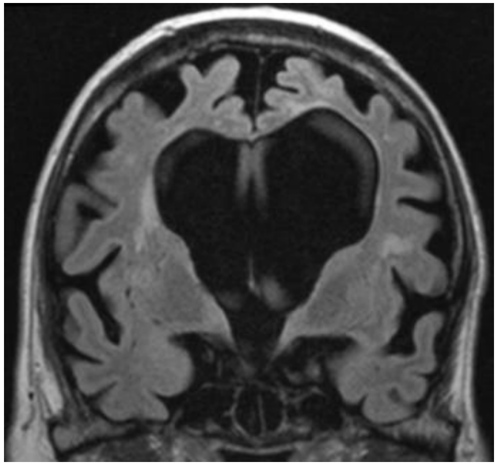
Silvius aqueduct stenosis



- Supratentorial macrocrania + small posterior fossa

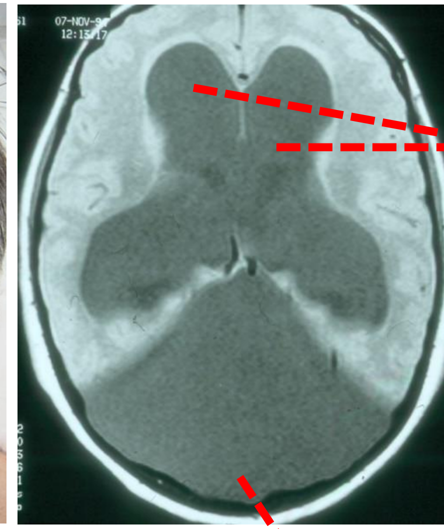


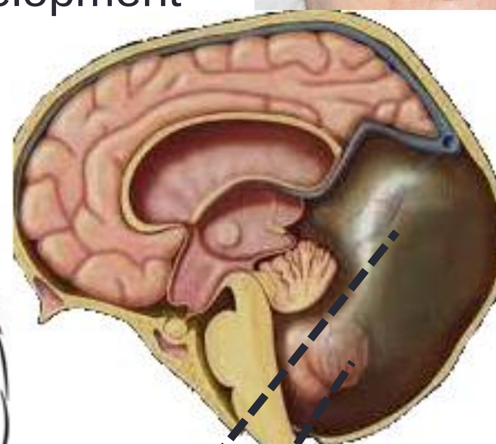


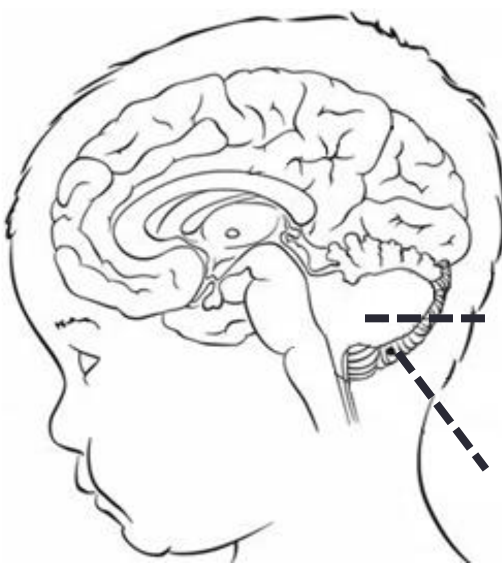


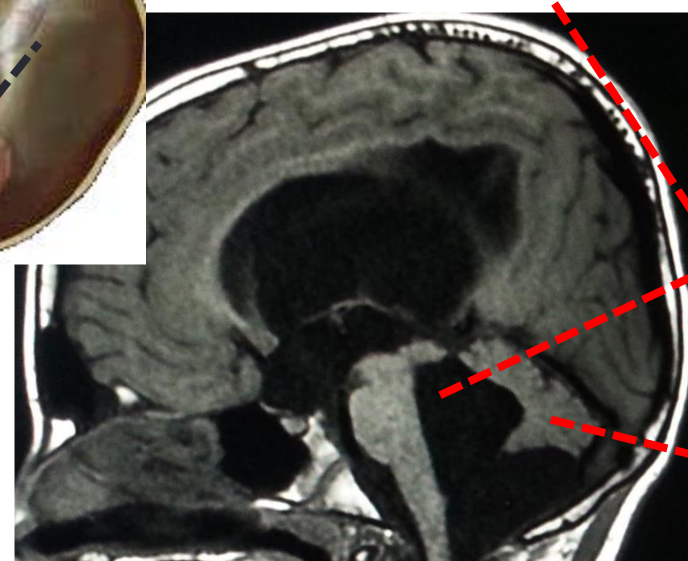
Dandy-Walker syndrome

- Very low incidence
- Partial cerebellum atrophy with atresia foramina Luschka & Magendie = cystic dilation of fourth ventricle
- Normal cognitive development



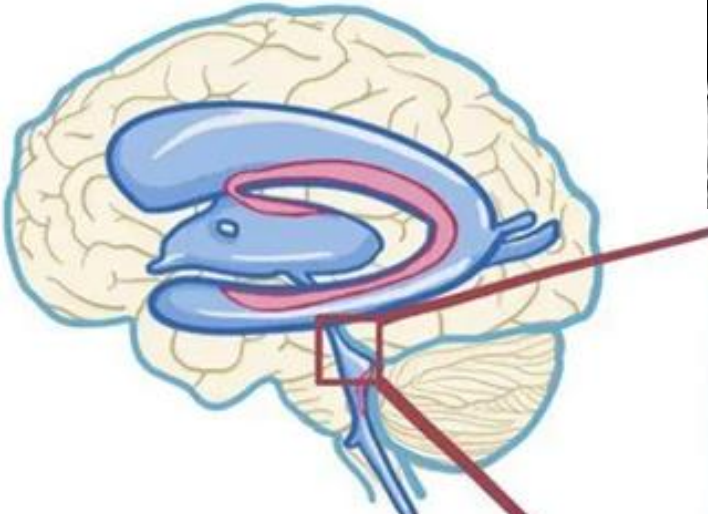


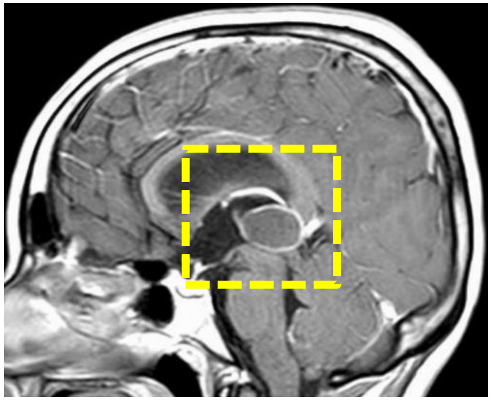


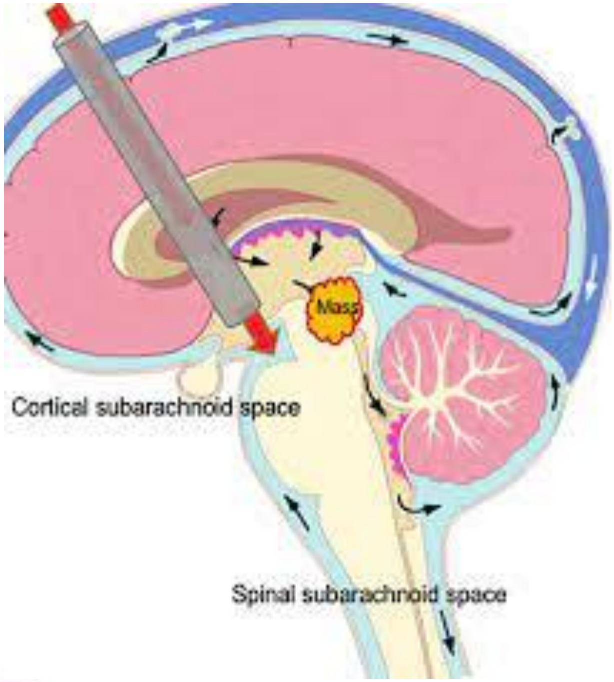


Hydrocephalus: pineal region tumors

- Uncommon
- Great histological variety
- Treatment hydrocephalus: endoscopic ventriculostomy



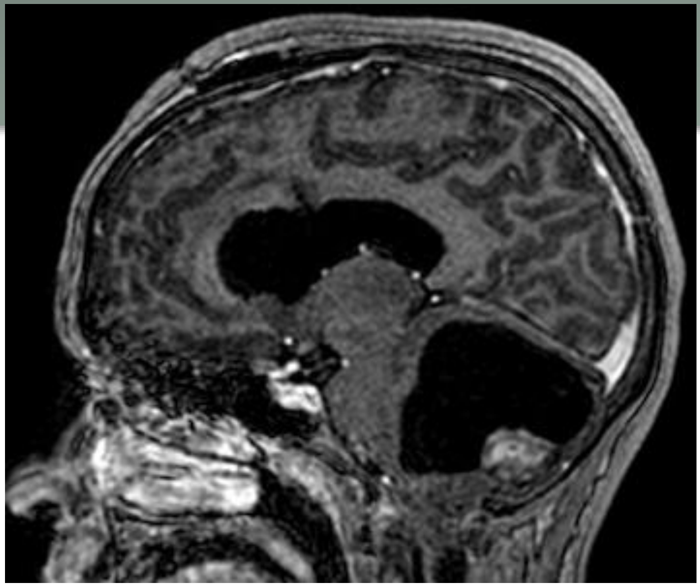


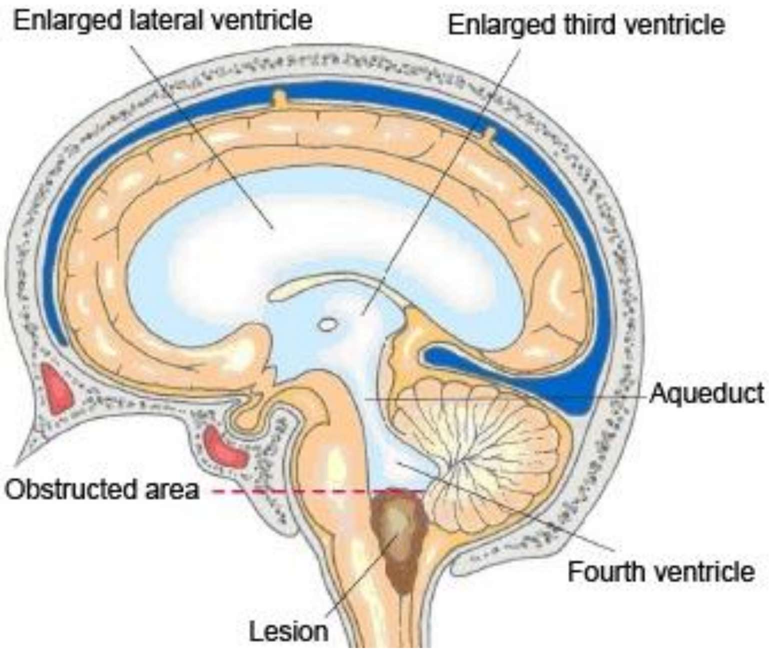


Endoscopic ventriculostomy

Fourth ventricle tumors

- ↑in children than in adults
  - Medulloblastoma
  - Ependymoma
- Obstructive hydrocephalus
- Possibility of tumor spread by CSF pathways
  - Carcinomatous meningitis
  - Meningeal carcinomatosis

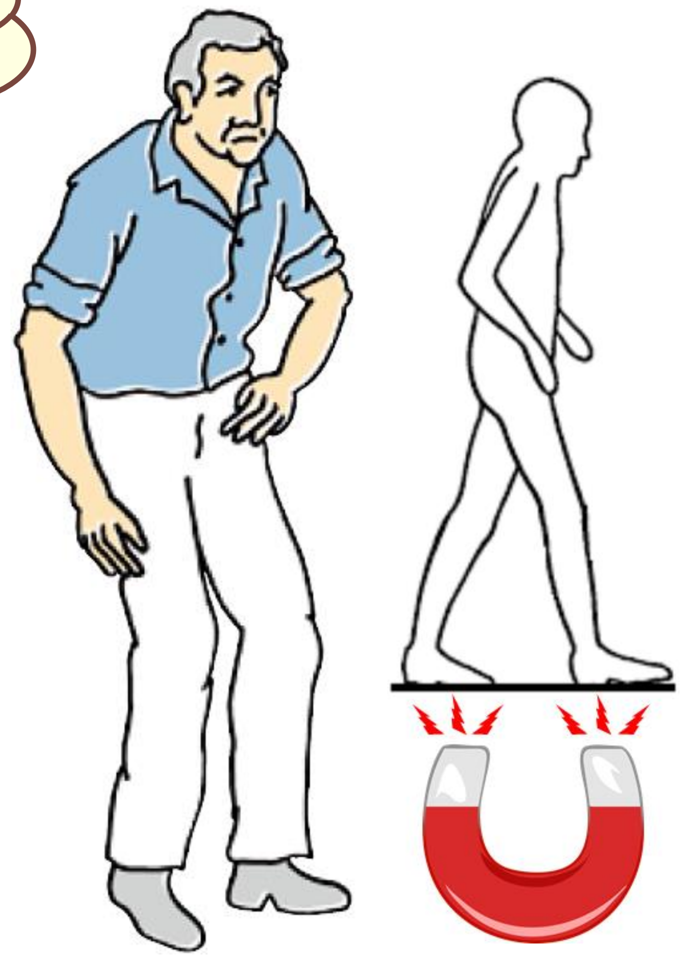




Carcinomatous meningitis

Normal pressure hydrocephalus: clinical features

Dementia

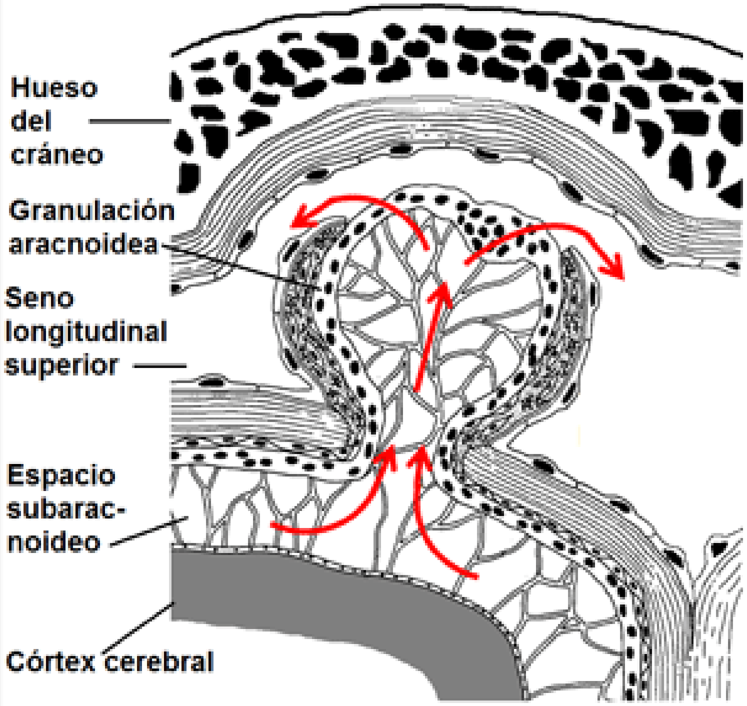


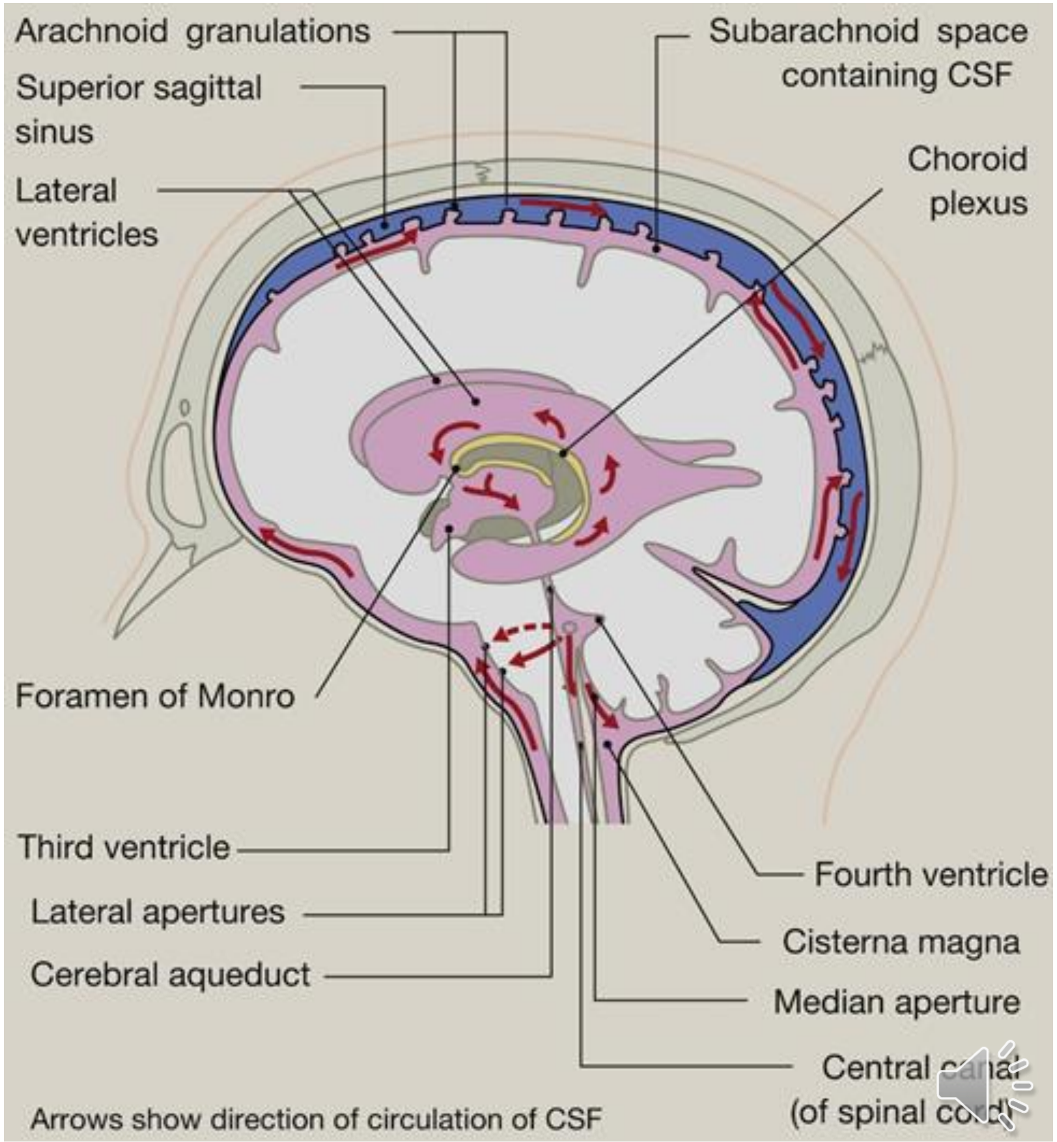
Gait disorder

Incontinence

Normal pressure hydrocephalus: communicating, CSF resorption impairment

- CSF blockage in subarachnoid space and/or reabsorption impairment at arachnoid villi





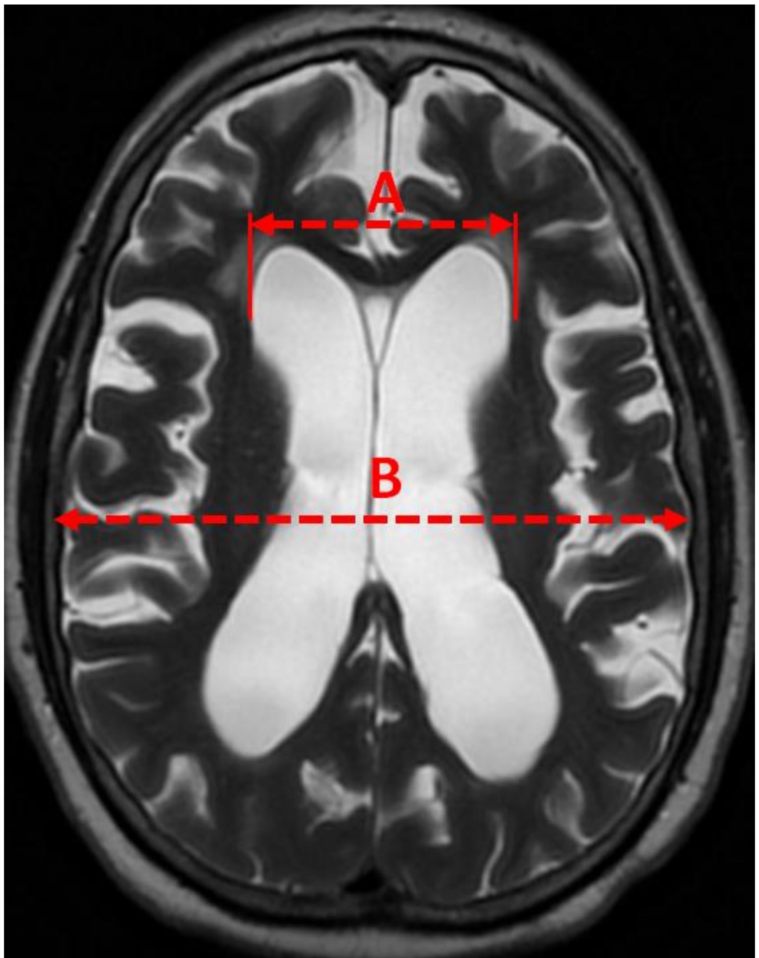
Normal pressure hydrocephalus: differential diagnosis

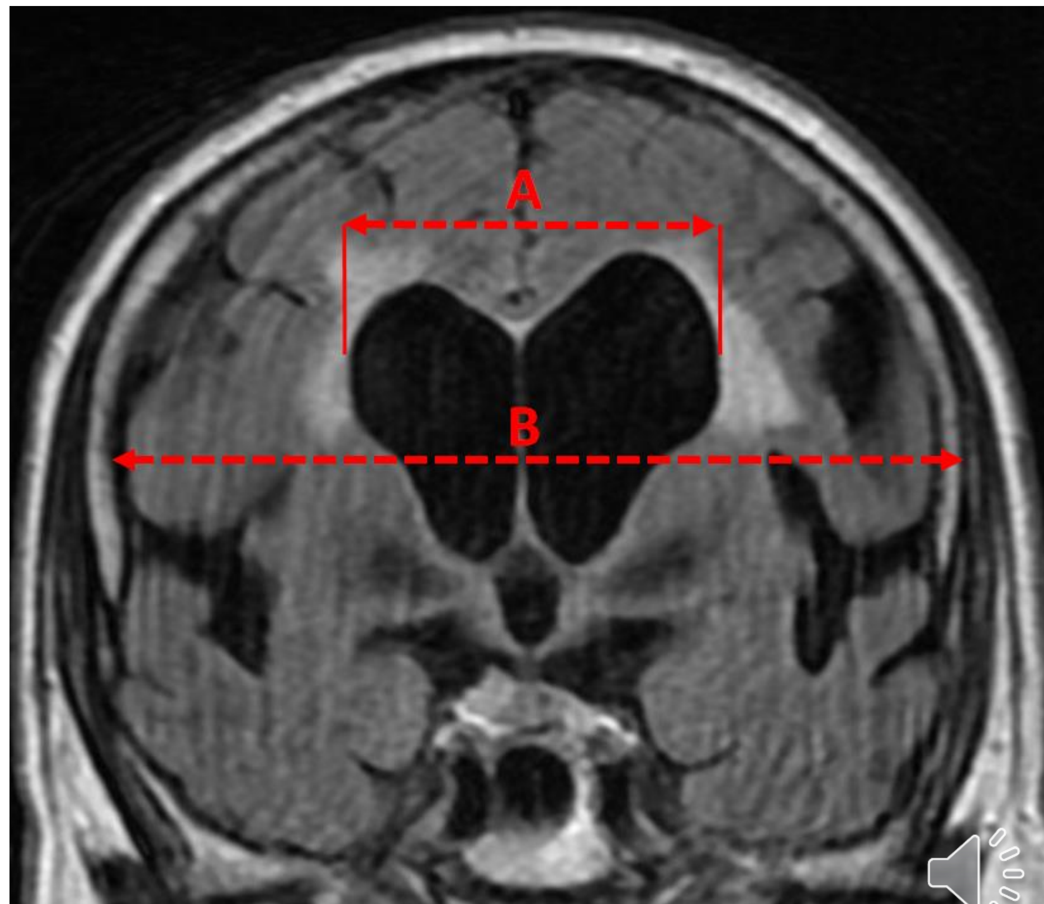
- Confusion with many other pathologies
- Possible coincidence hydrocephalus & Alzheimer's disease = poor results with surgical treatment

Common	Uncommon
Alzheimer disease	Lewy body dementia
Parkinson disease	Behavioral variant frontotemporal degeneration
Vascular dementia (Binswanger disease)	Progressive supranuclear palsy
Depression	Vestibular disorder
Urologic bladder outflow obstruction	Peripheral neuropathy
Neurodegenerative disorder	Lumbar stenosis
	Cerebral tumor
	Thyrotoxicosis

Normal pressure hydrocephalus: radiological diagnosis

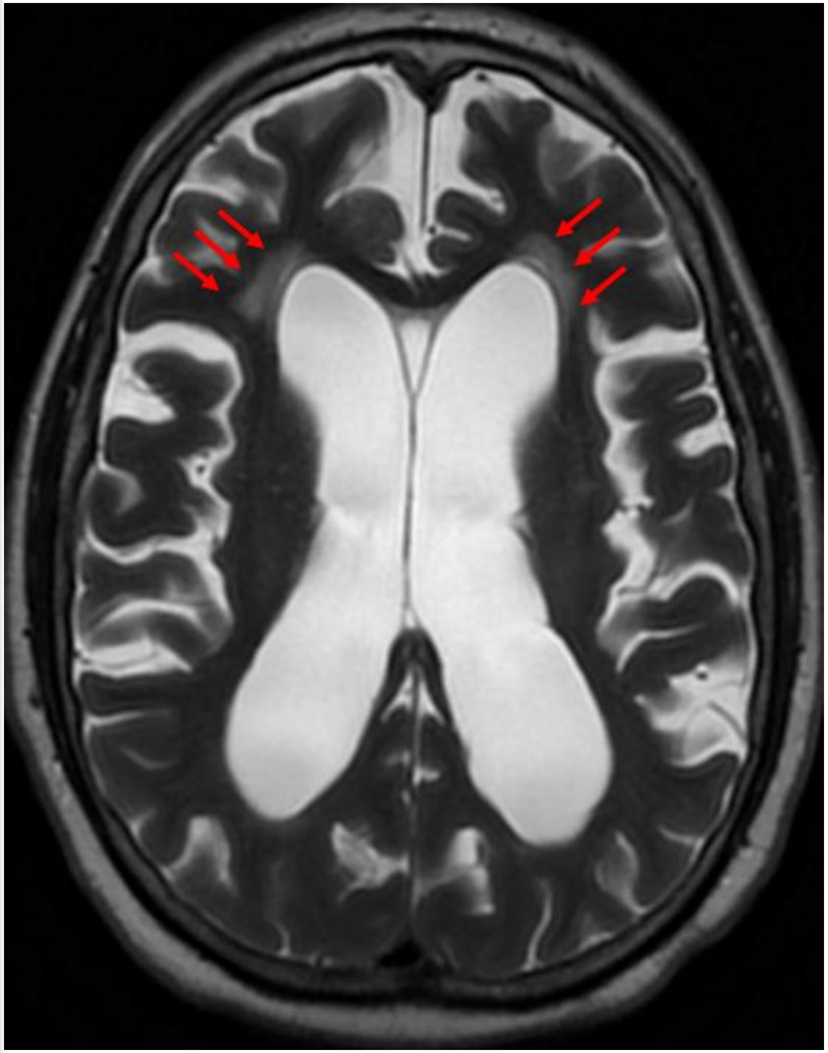
- Evans index (A/B) normal <30

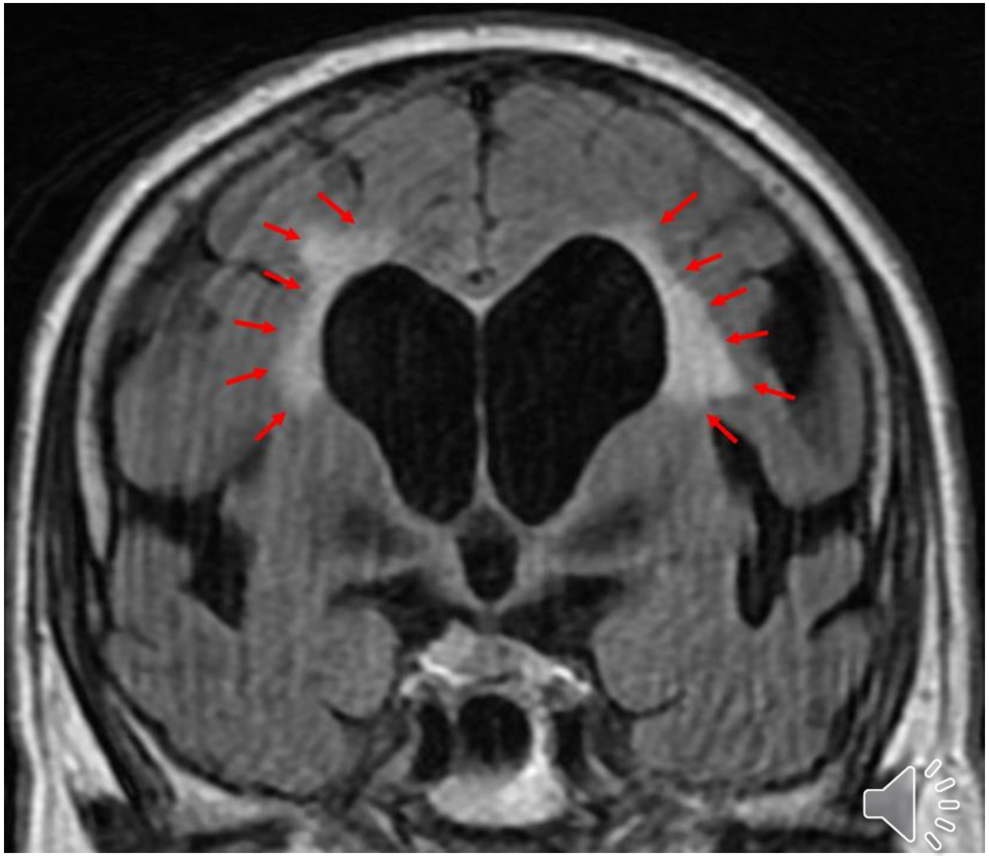




Normal pressure hydrocephalus: periependymal edema

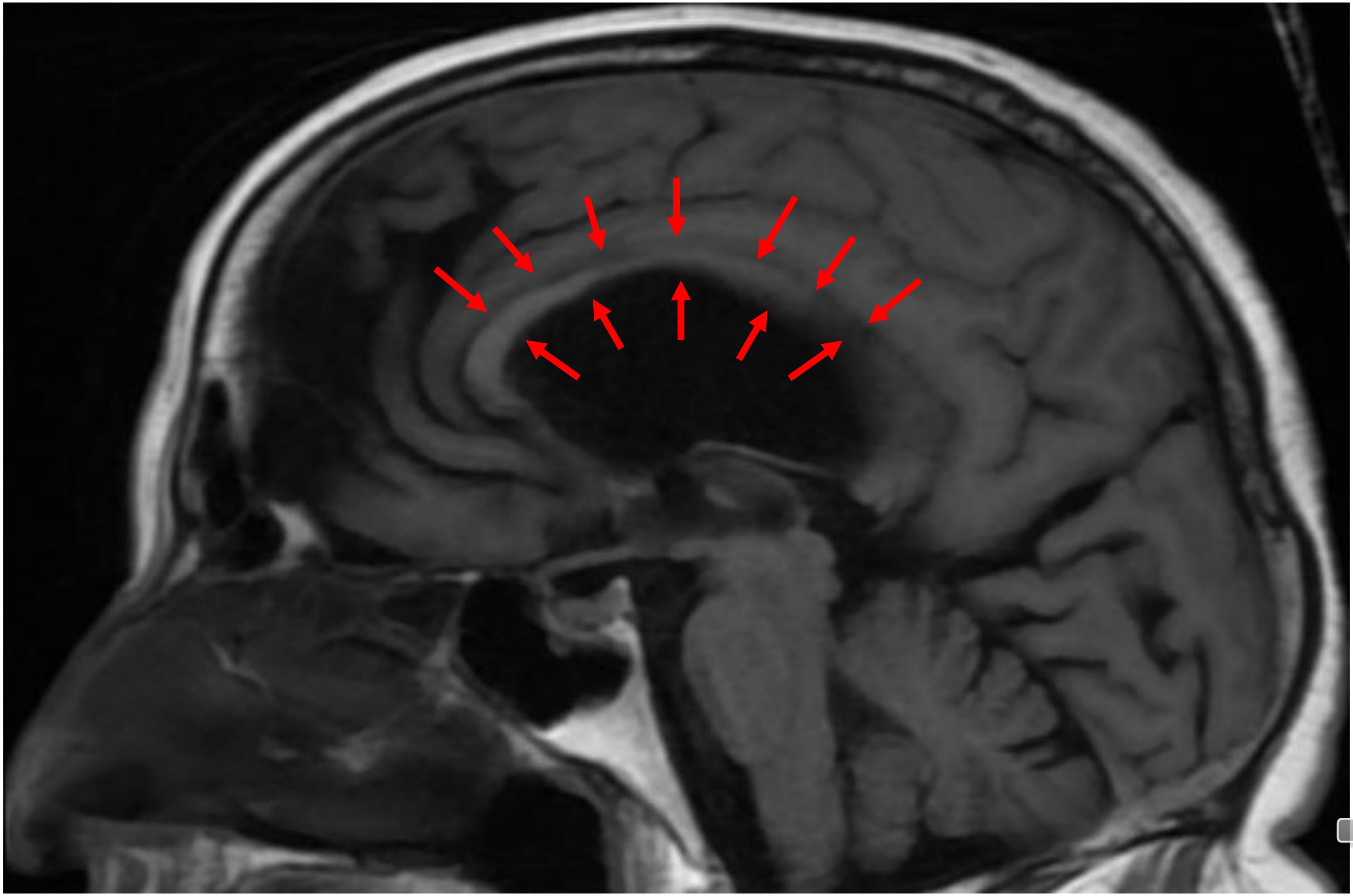
- Also occurs in other pathologies





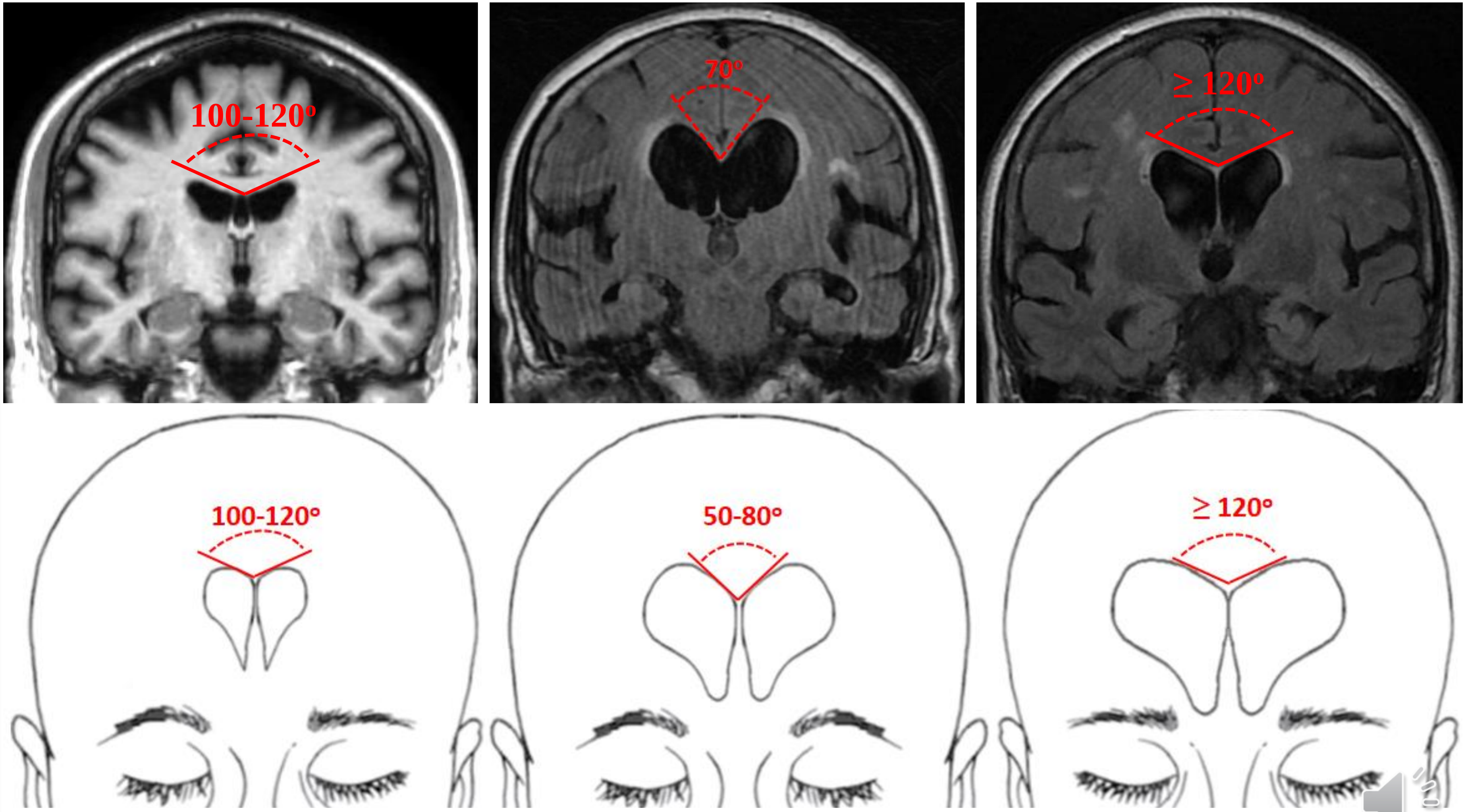
Normal pressure hydrocephalus: corpus callosum thinning

- Also occurs in brain atrophy



Normal pressure hydrocephalus: corpus callosum angle in coronal MRI images

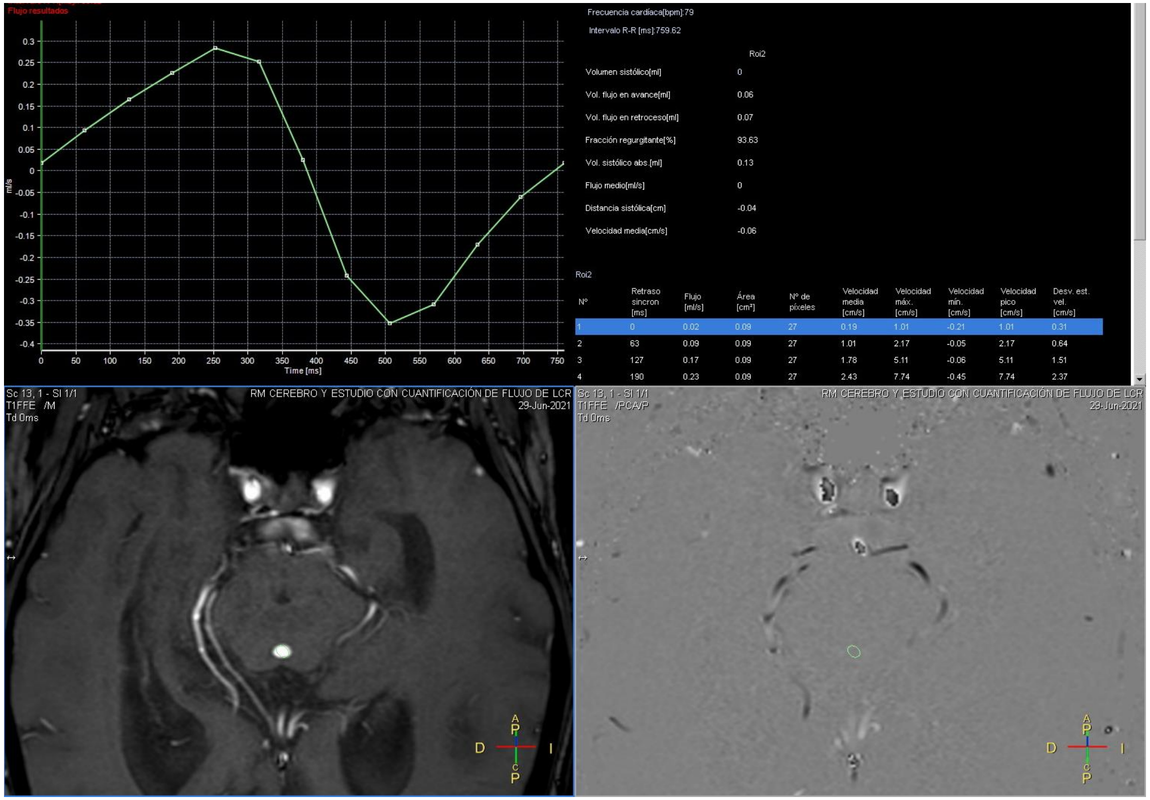
- Angle forming the upper surface of the lateral ventricles



Normal Hydrocephalus Brain atrophy

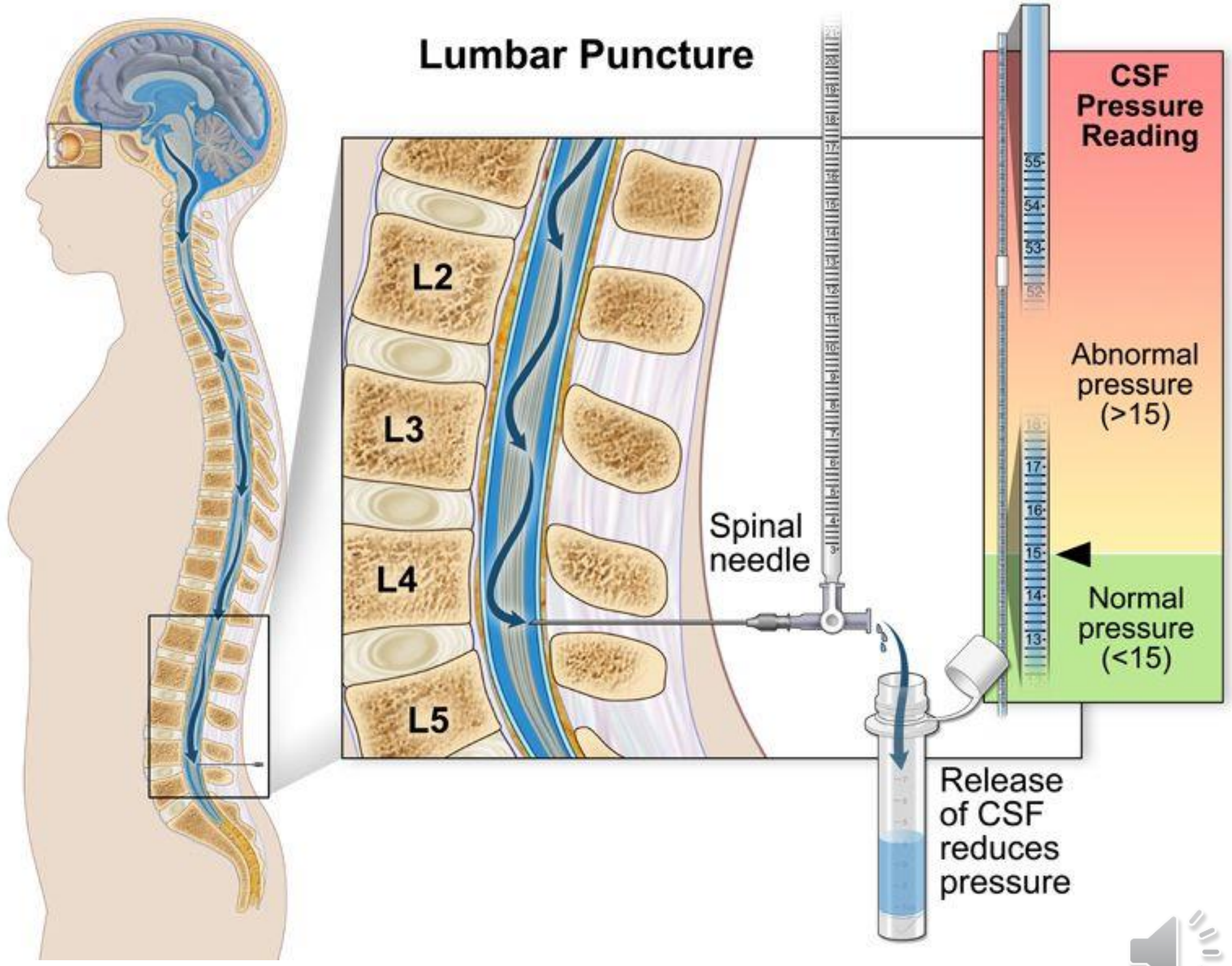
Normal pressure hydrocephalus: CSF flow measurement by magnetic resonance imaging

- Measures CSF throbbing motion during each heartbeat
  - Non-invasive and rapid CSF flow quantification
  - Measured at third ventricle, Silvius aqueduct & prepontine cistern
- Values (ml/min)
  - 18 to 27 normal pressure hydrocephalus
  - > 46 unreliable



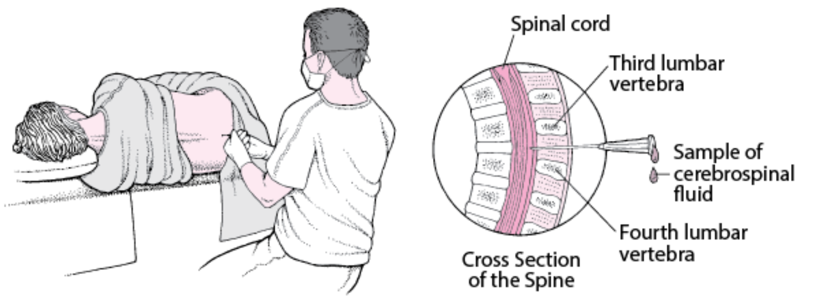
Normal pressure hydrocephalus: lumbar puncture

- CSF pressure is usually normal
  - Reason: lumbar puncture done during day and not at nighttime during the sleep REM phase
  - CSF pressure has NO diagnostic / prognostic value

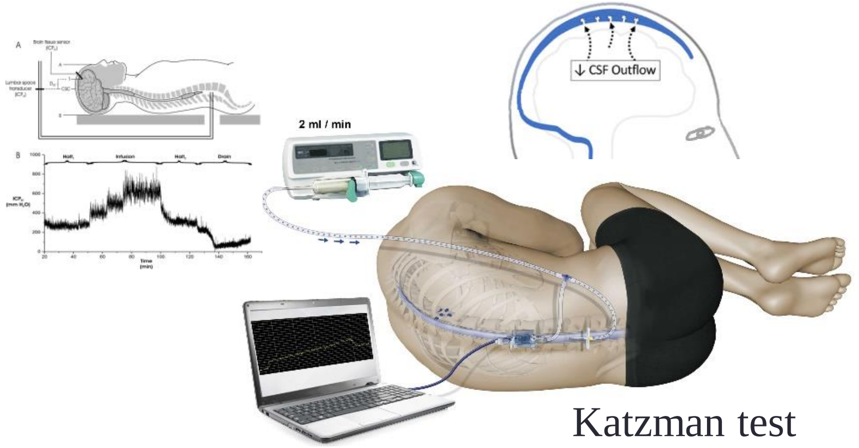


Normal pressure hydrocephalus: methods of diagnostic confirmation (1)

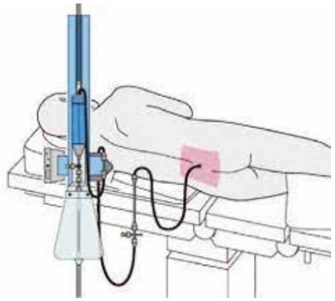
- **Lumbar puncture with CSF drainage**
  - Opening pressure measurement
  - Drainage ~ 50 ml CSF
  - Monitor symptom response
  - Practical but unreliable
- **Katzman infusion test**
  - 1st lumbar puncture
  - 2nd lumbar infusion 20 ml Ringer's lactate at constant pressure and volume
  - 3rd ICP measurement at the lumbar level
  - 4th calculation of drainage resistance (ROF)
  - 5th drainage ~ 50 ml CSF
  - Frequent false positives and negatives
- **Lumbar drainage**
  - 1st lumbar puncture
  - 2nd insertion of lumbar CSF drainage catheter
  - 3rd CSF drainage for 1-3 days
  - More reliable but uncomfortable for patients



Lumbar puncture with CSF drainage



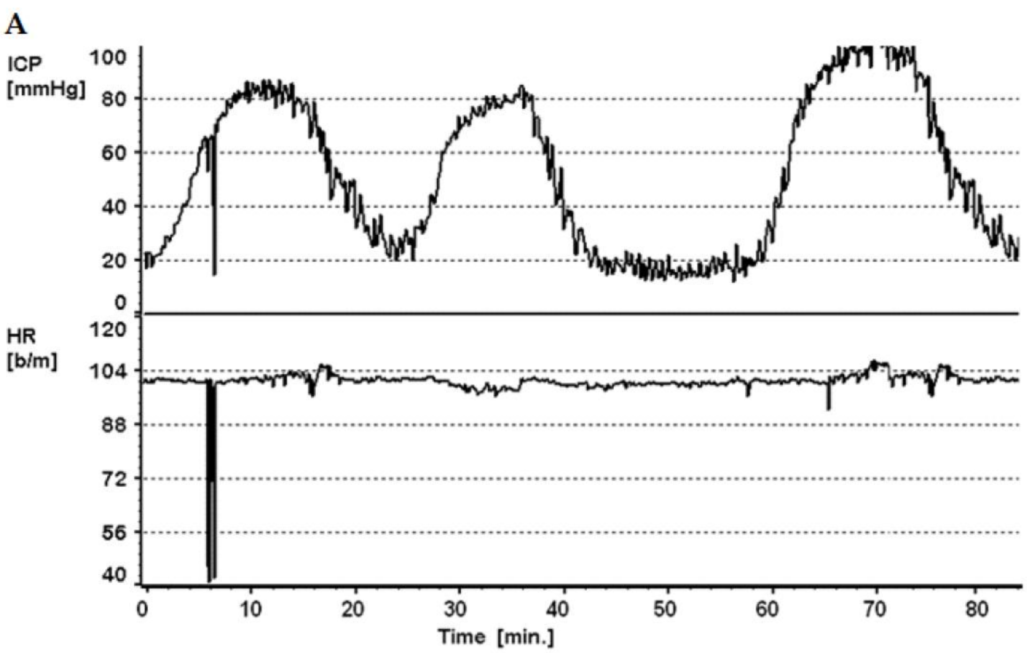
Katzman test



Lumbar drain

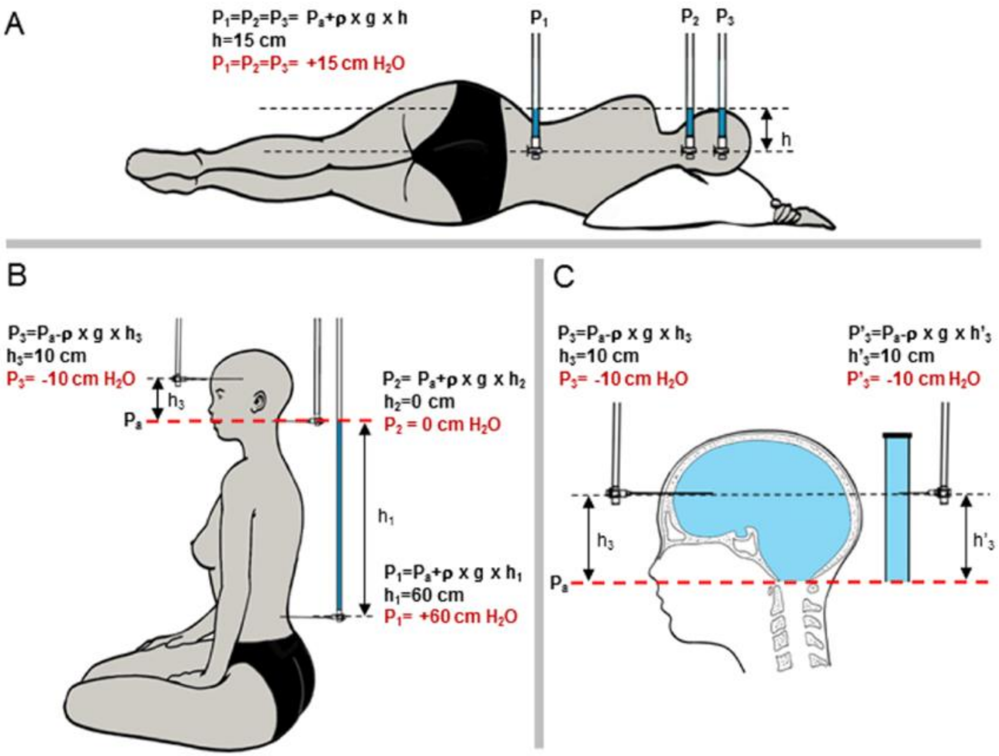
Normal pressure hydrocephalus: methods of diagnostic confirmation (2)

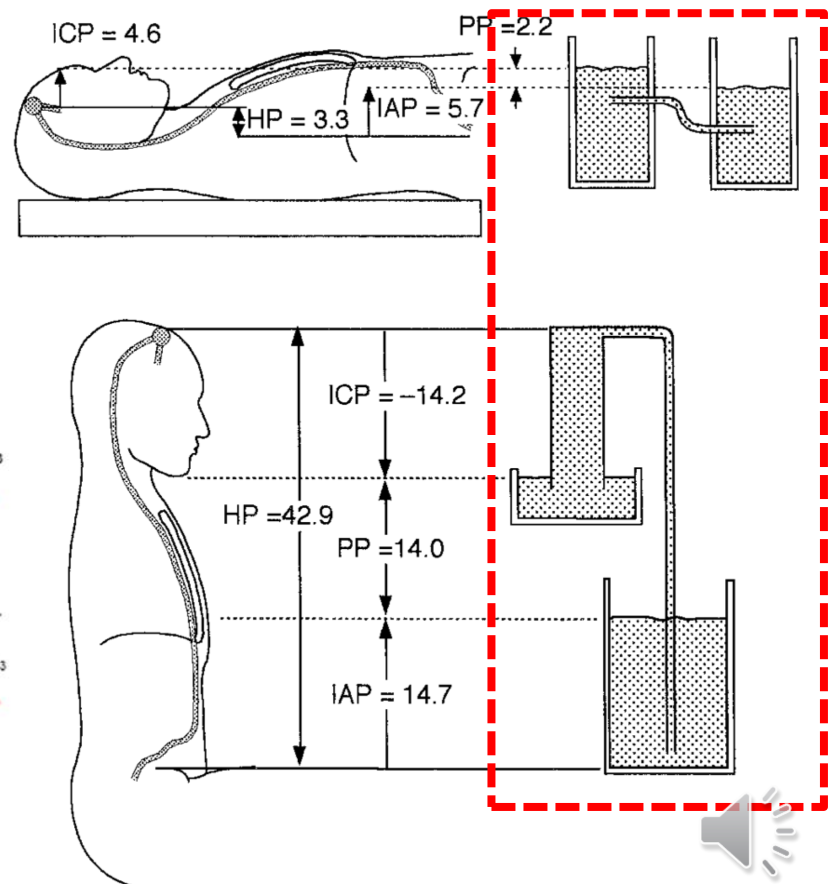
- Intracranial pressure monitoring
  - Requirements
    - Minor surgical procedure
    - 5-day hospital admission
  - The most specific and reliable diagnostic technique



HYDROCEPHALUS: TREATMENT

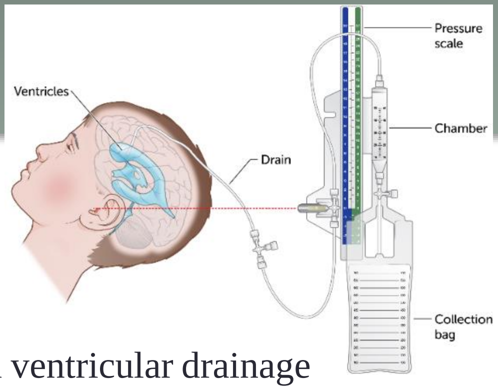
- **Goal**: ICP between -5 and +15 cm H_2O
- Position of the patient modifies
  - ICP
  - Volume of CSF drained



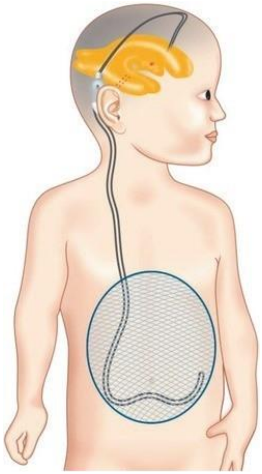


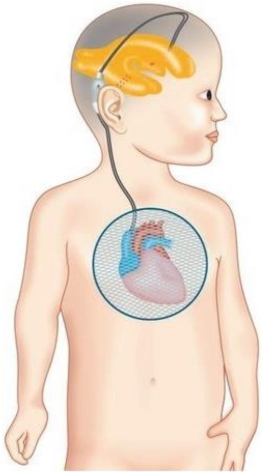
CSF drainage options

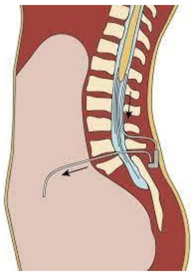
- **External ventricular drainage**
  - Temporary measure
  - If intraventricular haemorrhage (prematurity) or infection
- **Permanent CSF shunt** ~ 80% cases
  - Ventricleperitoneal
  - Ventricleatrial
  - Lumboperitoneal
  - Ventriclepleural
- **Ventriculostomy** ~ 15% cases
  - Opening floor third ventricle = CSF comes out directly into the subarachnoid space
  - CSF circumvents obstruction in third ventricle, Silvio's aqueduct or posterior fossa

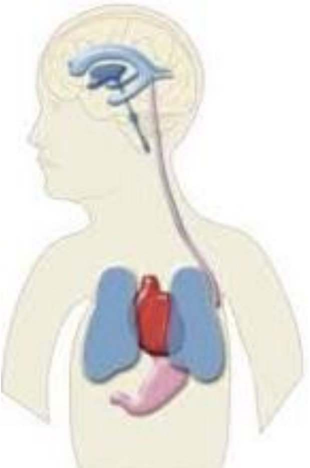


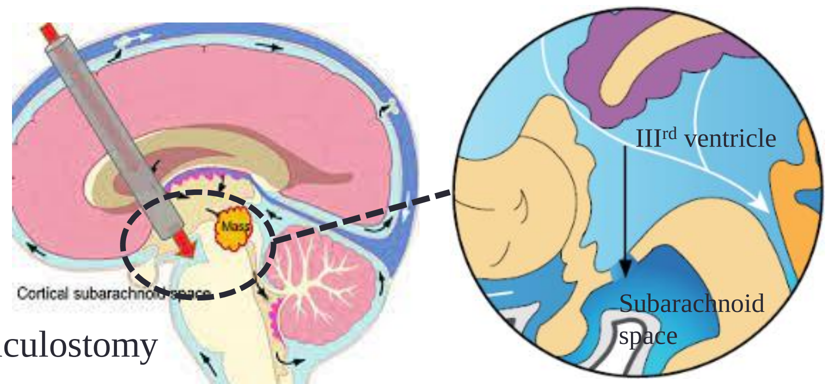
External ventricular drainage


Ventricle peritoneal

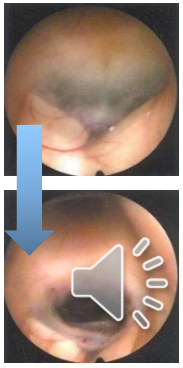

Ventricle atrial


Lumbo peritoneal


Ventricle pleural

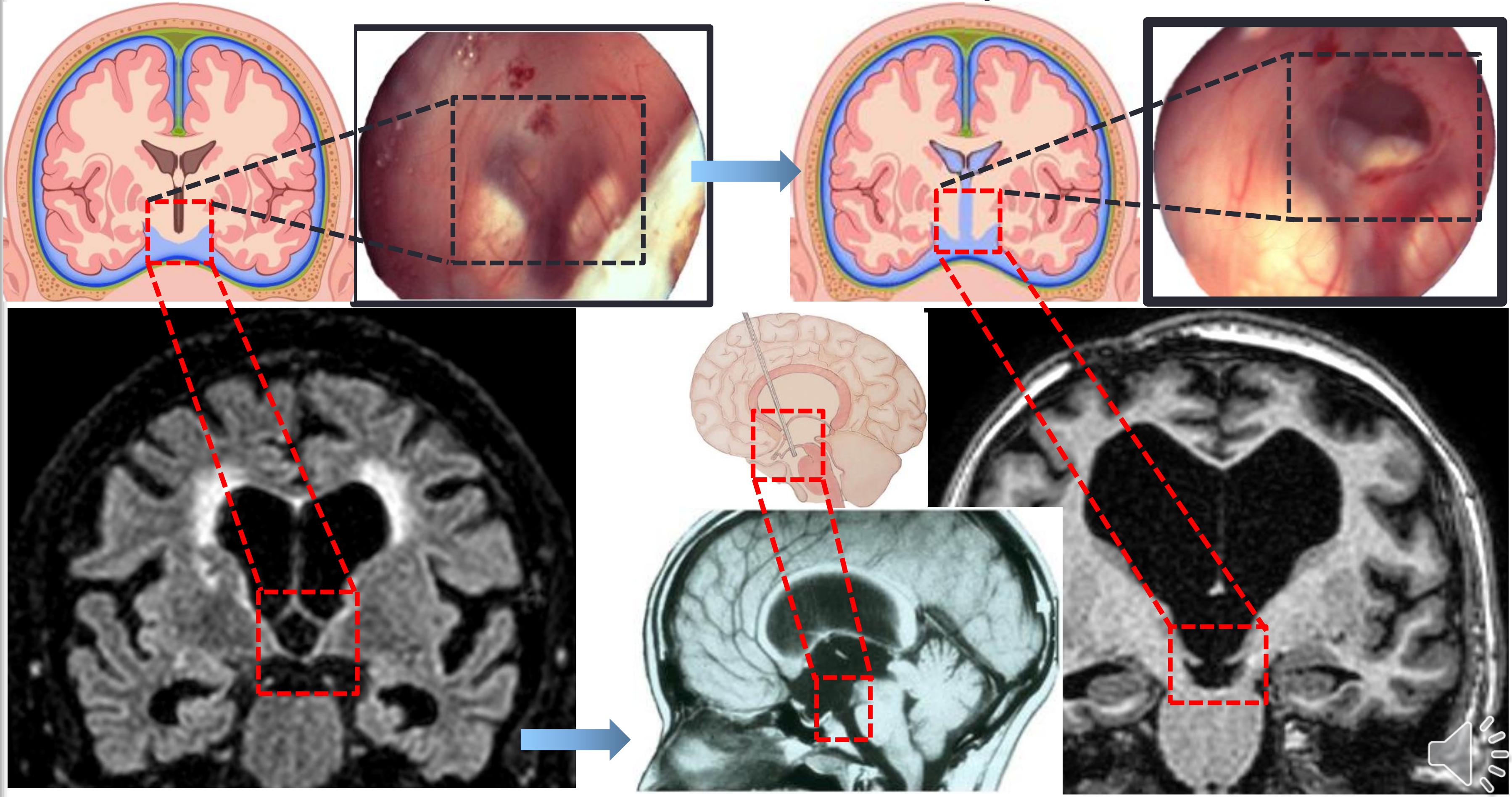


Ventriculostomy



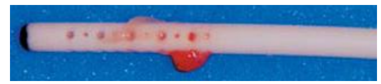
Ventriculostomy floor third ventricle

- Few indications
- Ideal: NO need for CSF shunt = no reoperations

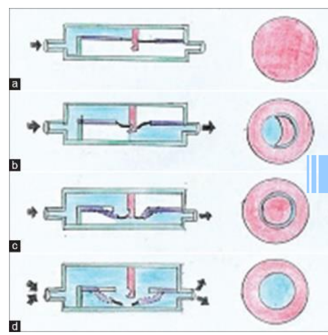


Components of the CSF shunt systems

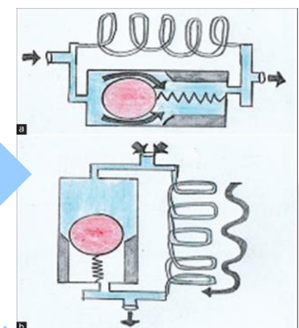
- **Ventricular catheter**
  - With time it is obstructed by the choroid plexus
- **Valve**
  - Regulates CSF drainage
  - Various mechanisms
  - Must adapt to patient's postural changes
- **Distal catheter**
  - Drainage depending on pressure cavity where it is directed
    - Abdomen pressure changes with recumbent-standing position
    - Atrium constant pressure
    - Pleura negative pressure



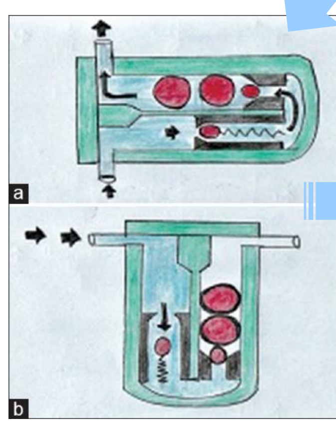
Ventricular catheter blockage caused by choroid plexus



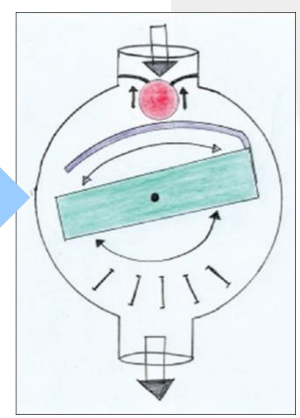
Membrane valve



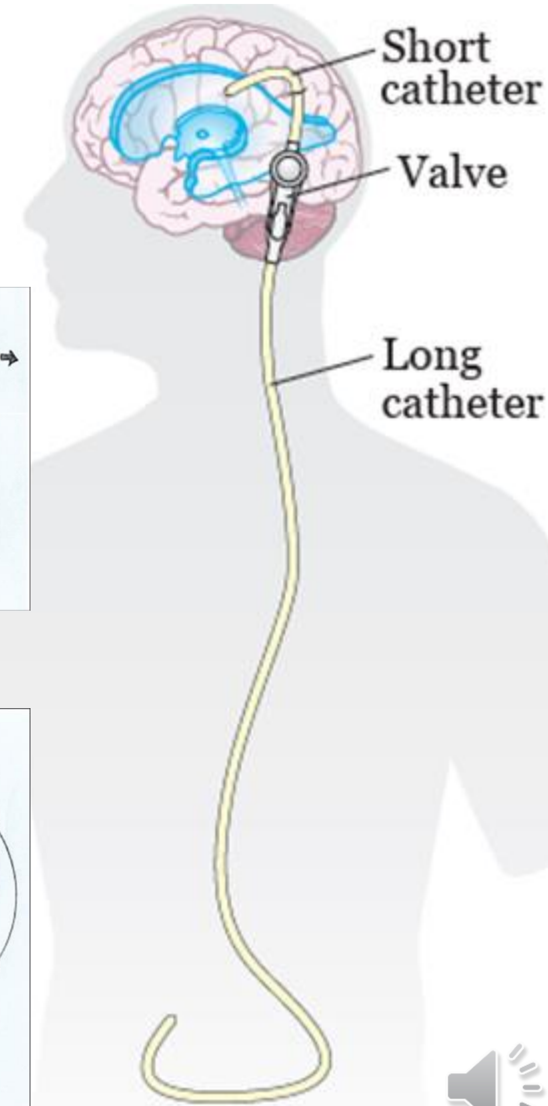
Spring valve



Ball valve

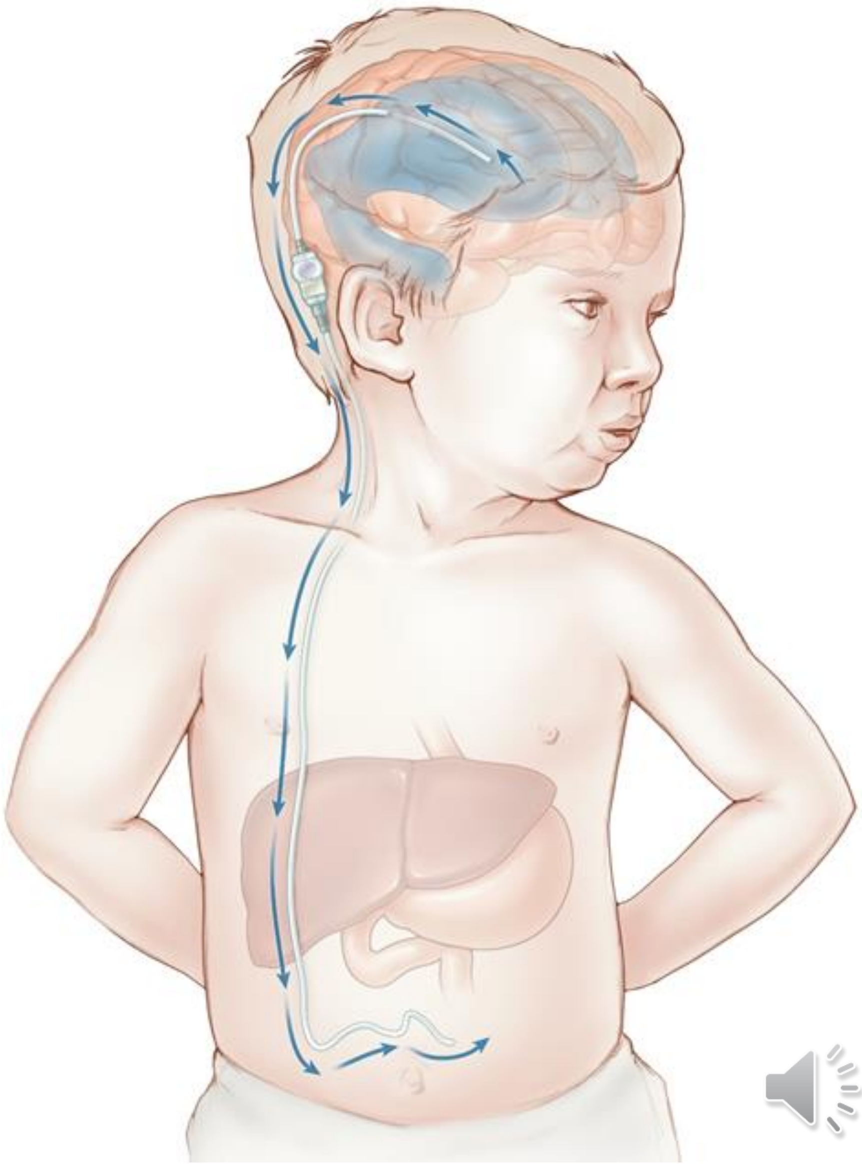


Rotor valve



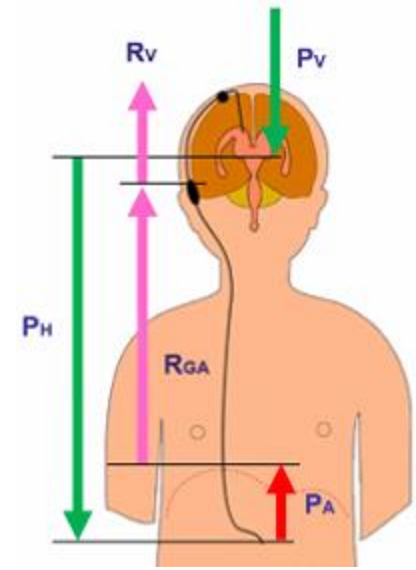
Operation of CSF shunt systems

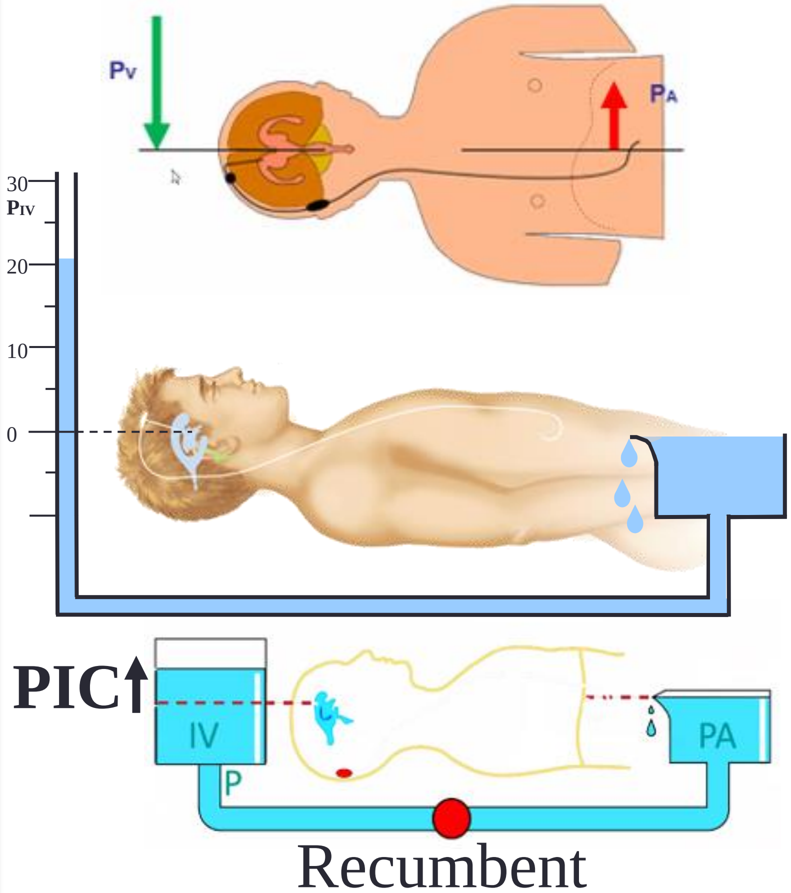
- CSF produced by choroid plexuses (in the ventricles)
- Ventricular catheter drains CSF from the ventricles
- Valve regulates CSF outlet pressure
- Distal catheter drains CSF into the chosen cavity
  - CSF drainage depending on pressure of chosen cavity
    - Atrium: constant pressure
    - Peritoneum: variable with the patient position
    - Pleural cavity: negative pressure with suction

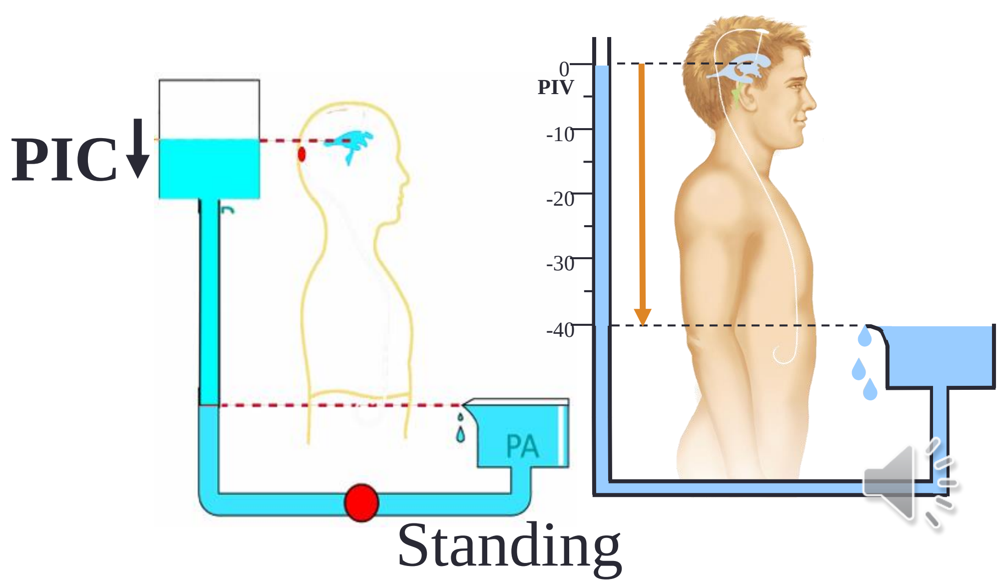


Pressures regulating CSF drainage through ventricleperitoneal shunt

- Intraventricular pressure (P_{IV})
- Hydrostatic pressure (P_H)
- Abdominal pressure (P_A)

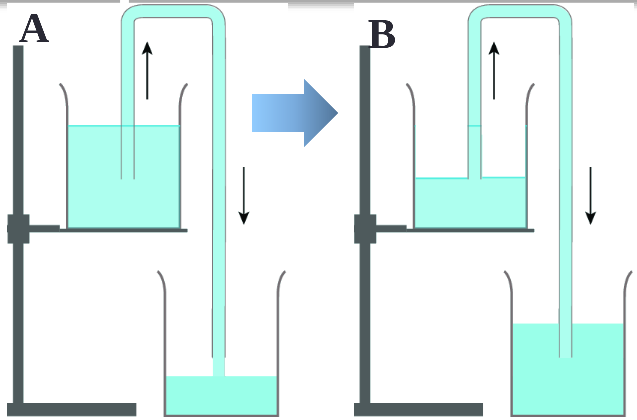




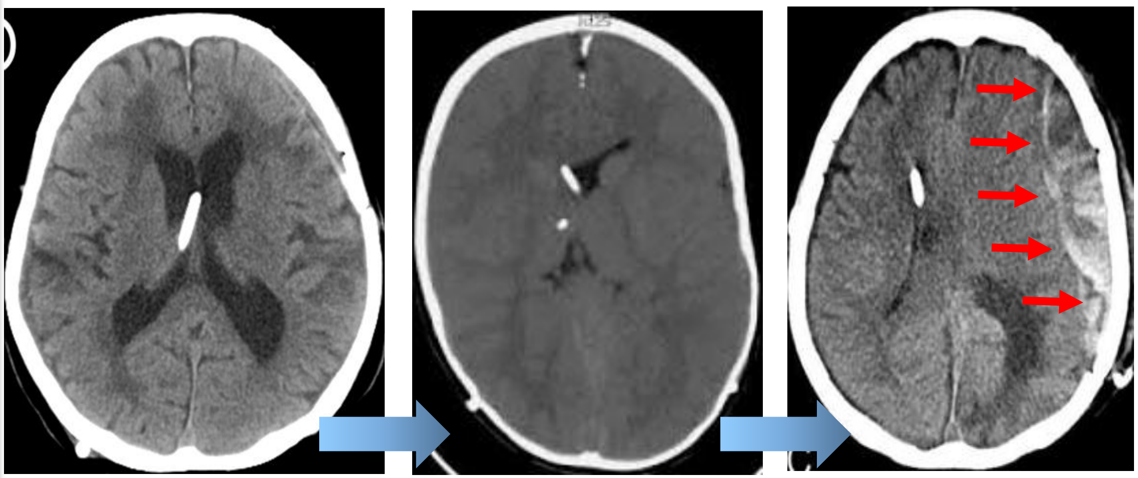


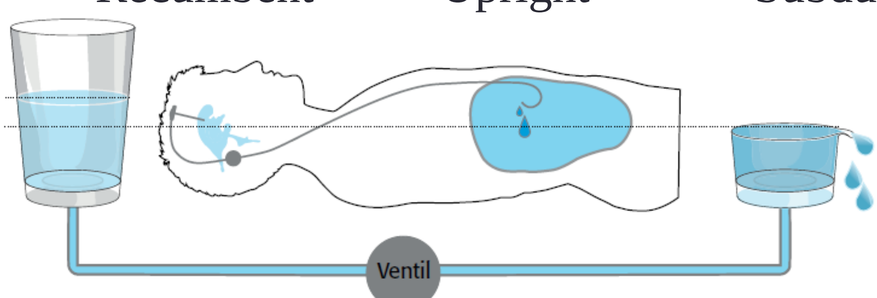
CSF drainage in standing position: ventricularperitoneal shunt siphon effect

- In standing position = suction effect of the water column of the distal catheter = excess CSF drainage = ↓PIC = headache → ventricular collapse → possibility of subdural hematoma

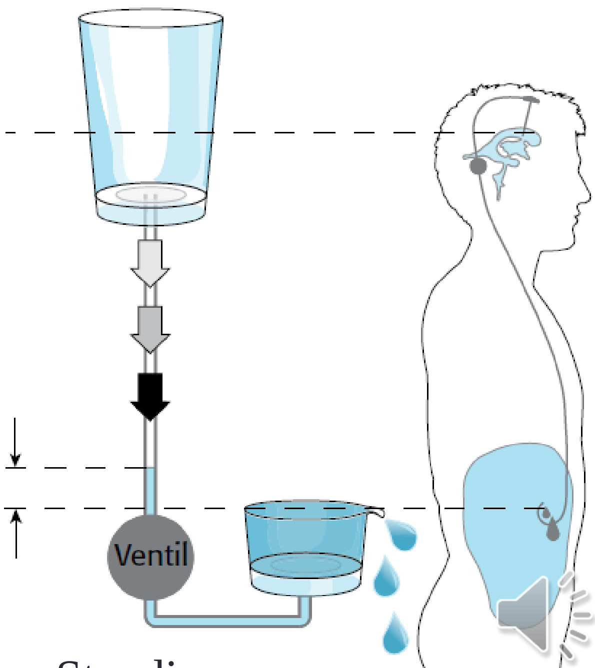


Siphon effect in standing position


Recumbent Upright Subdural hematoma



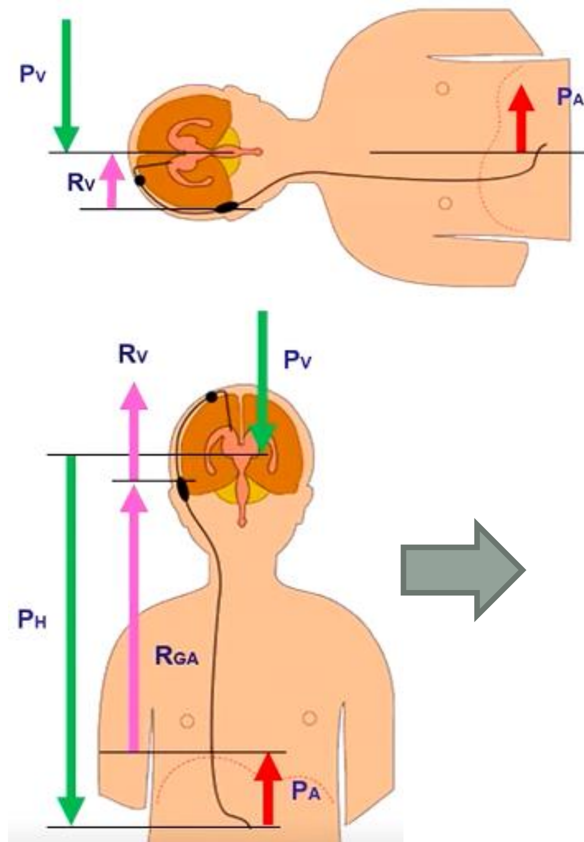
Recumbent

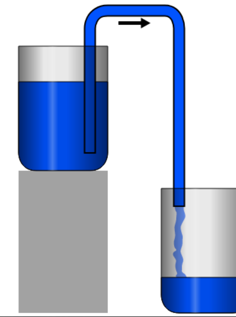


Standing

Siphon effect

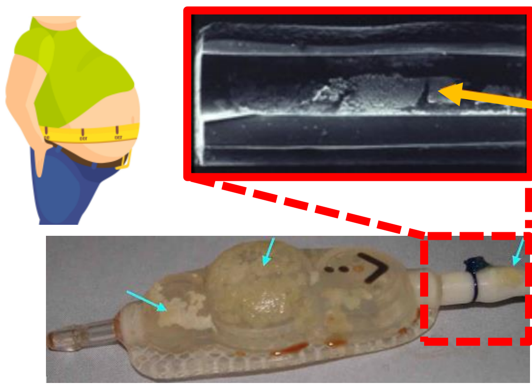
- CSF flow → suction effect = drains more CSF than desirable
  - Slit ventricle syndrome
  - Chronic hygroma/subdural hematoma
  - Orthostatic headache

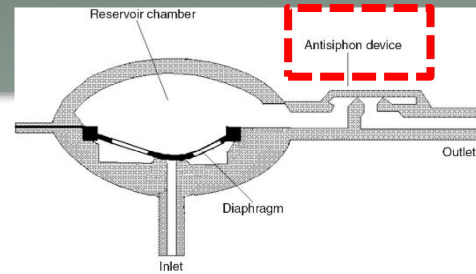


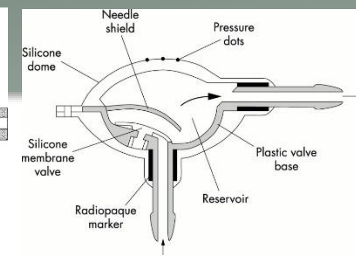


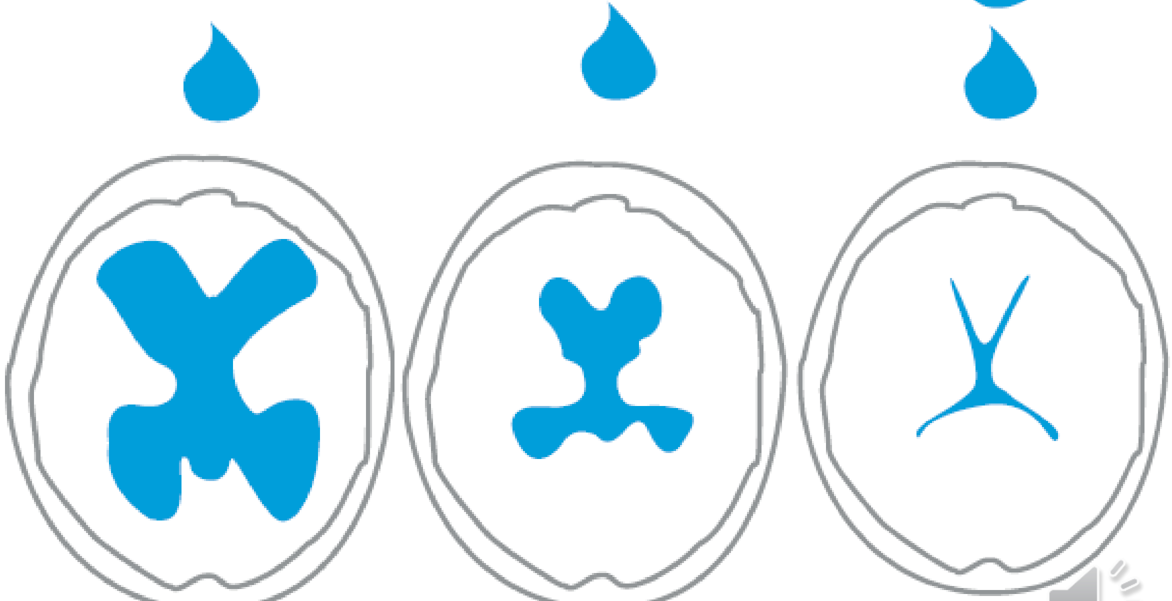
Possibilities of CSF shunt systems

- **Inadequate drainage**
  - Excessive abdominal pressure
  - Shunt colonisation by bacteria
- **Adequate drainage**
- **Excessive drainage**
  - Shunt without anti-siphon mechanism





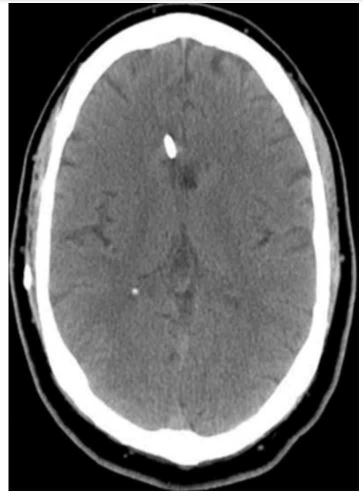




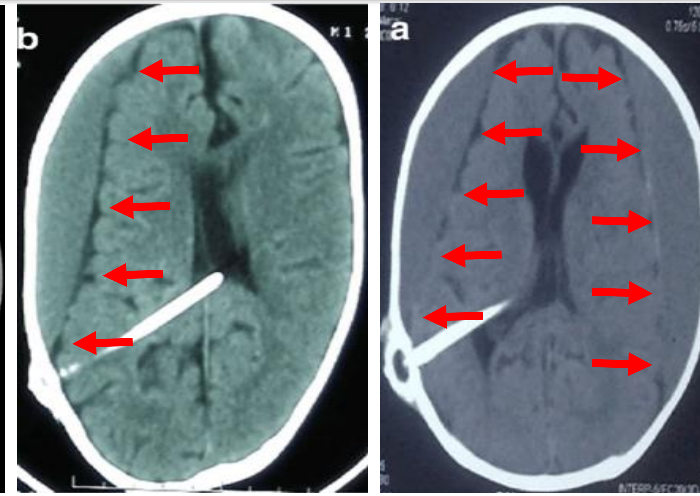
Inadequate drainage Adequate drainage Excessive drainage

Excess CSF drainage: consequences

- Ventricular collapse
  - Lower tolerance ↑ICP
- Chronic subdural hematoma
- Secondary craniosynostosis

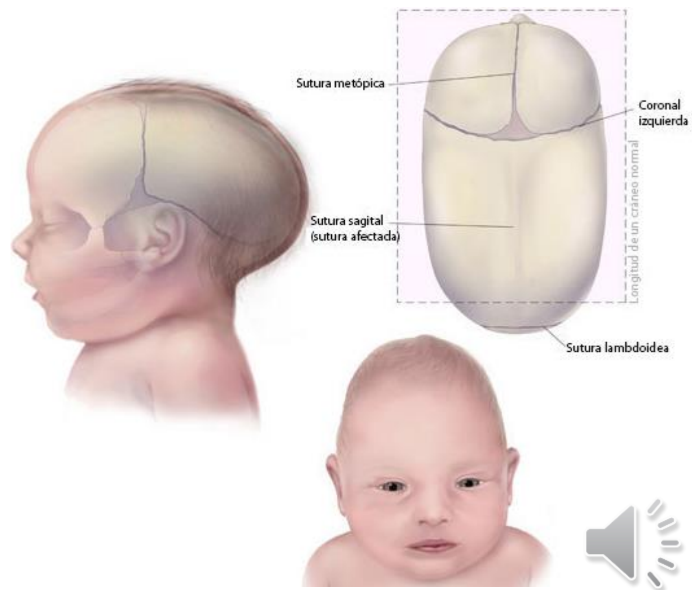


Ventricular collapse



Chronic uni- or bilateral subdural hematoma

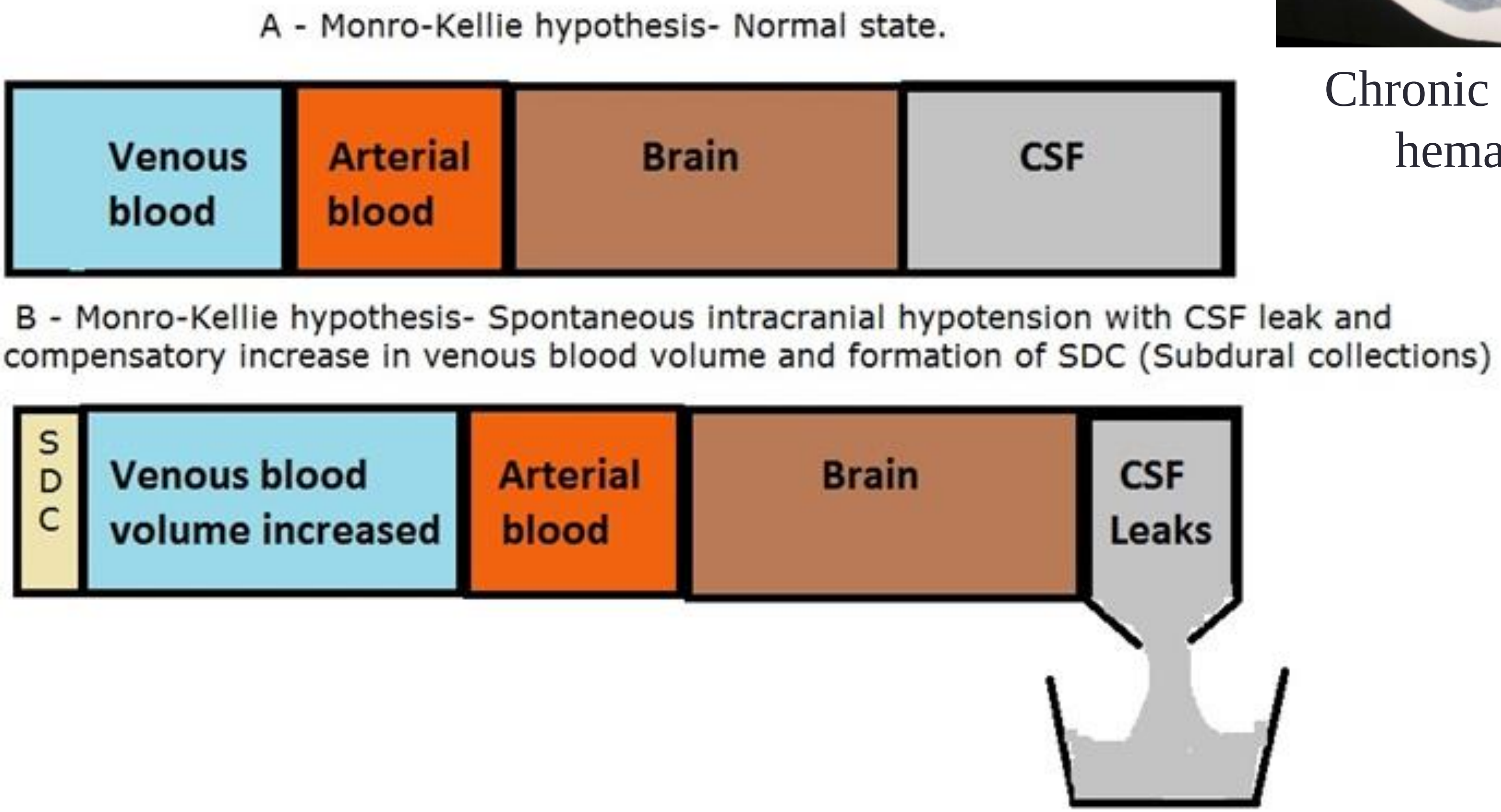
Secondary craniosynostosis



Excessive CSF drainage and chronic subdural hematoma

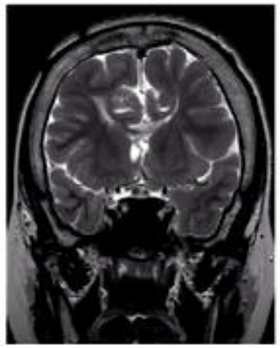
- Excess CSF drainage = possibility of chronic subdural hematoma

Chronic subdural hematoma

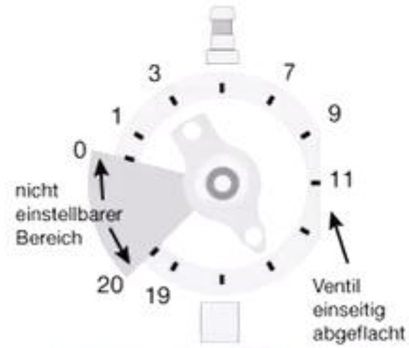


Anti-siphon mechanisms

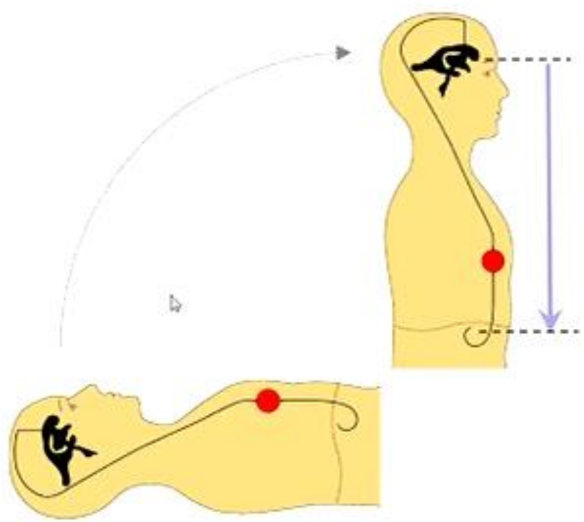
- Control excess drainage while sitting and standing but drainage not always adequate in recumbent position



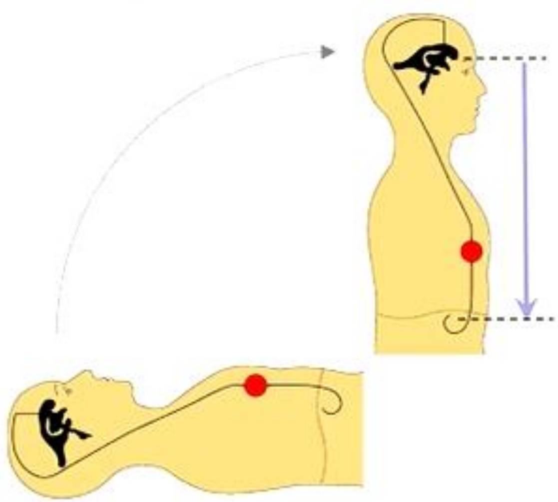
over-/ underdrainage



adjustable DP unit



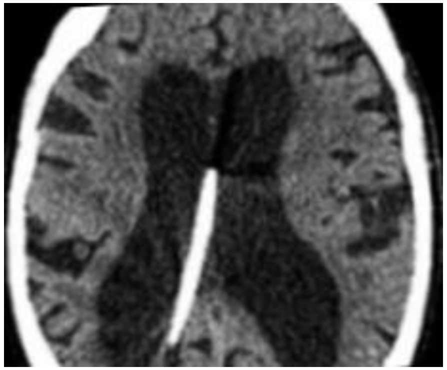
hydrostatic-effect

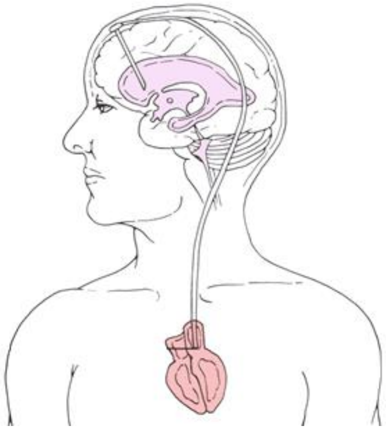


hydrostatic-effect

CSF drainage: obesity & sleep apnea

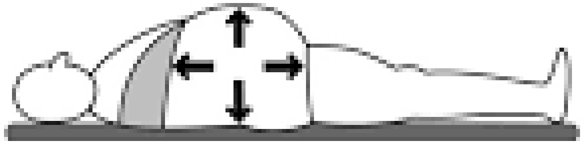
- ↑ abdominal pressure = malfunction shunt recumbent
- Sleep apnea = abdominal & chest pressure ↑ during apneas = shunt malfunction during sleep
- Option: ventriculoatrial shunt

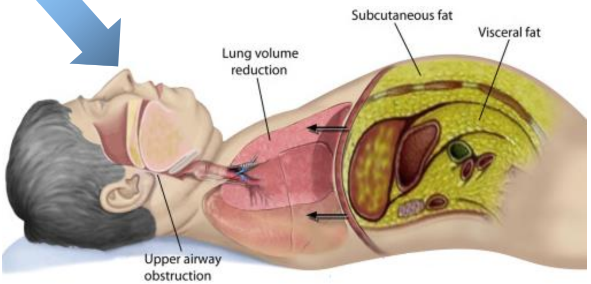




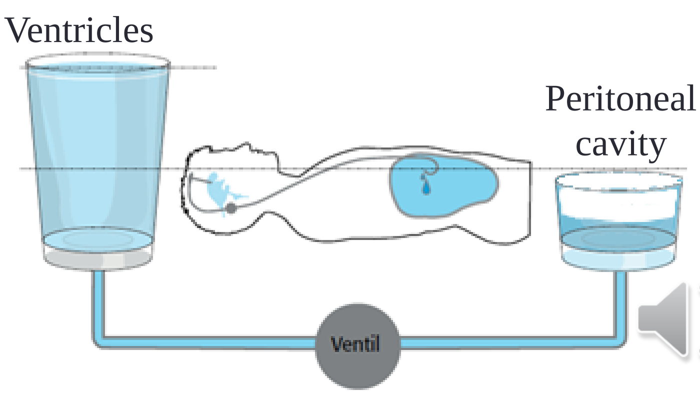
Ventriculoatrial shunt

Pharynx block in sleep apnea



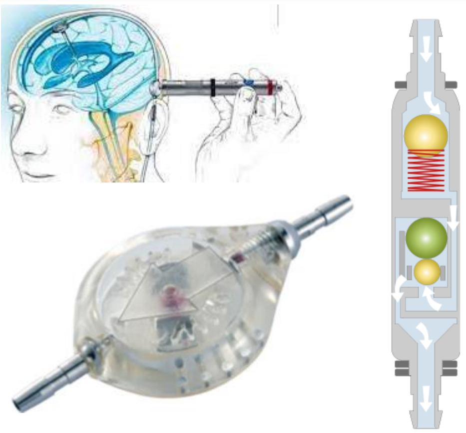


Apnea sleep

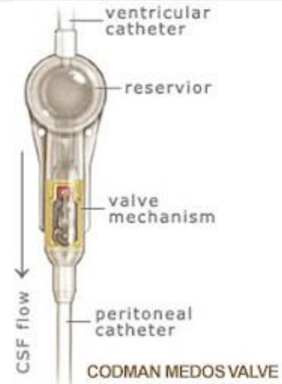


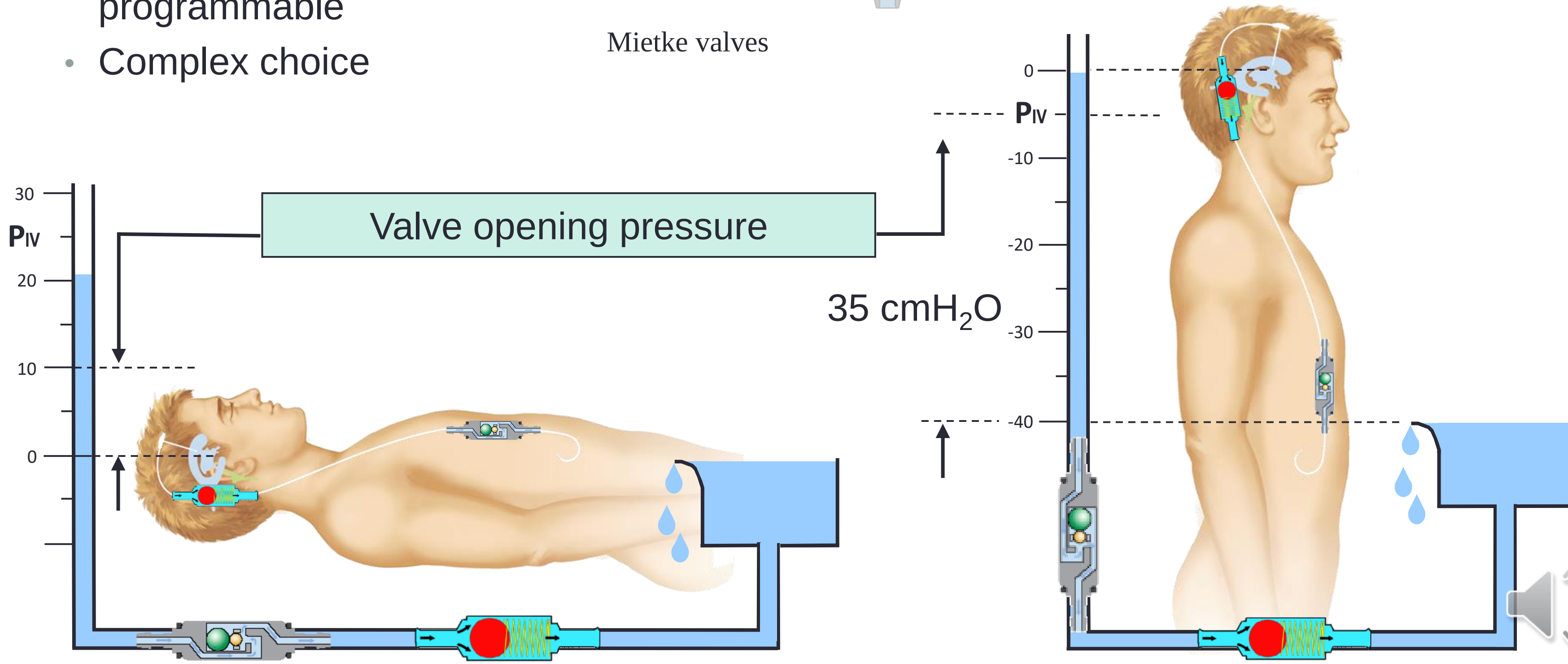
Wide variety of CSF shunt valve types

- New designs correcting problems
  - Goal: that drainage does not depend on posture or abdominal pressure
  - Percutaneously programmable
  - Complex choice



Mietke valves



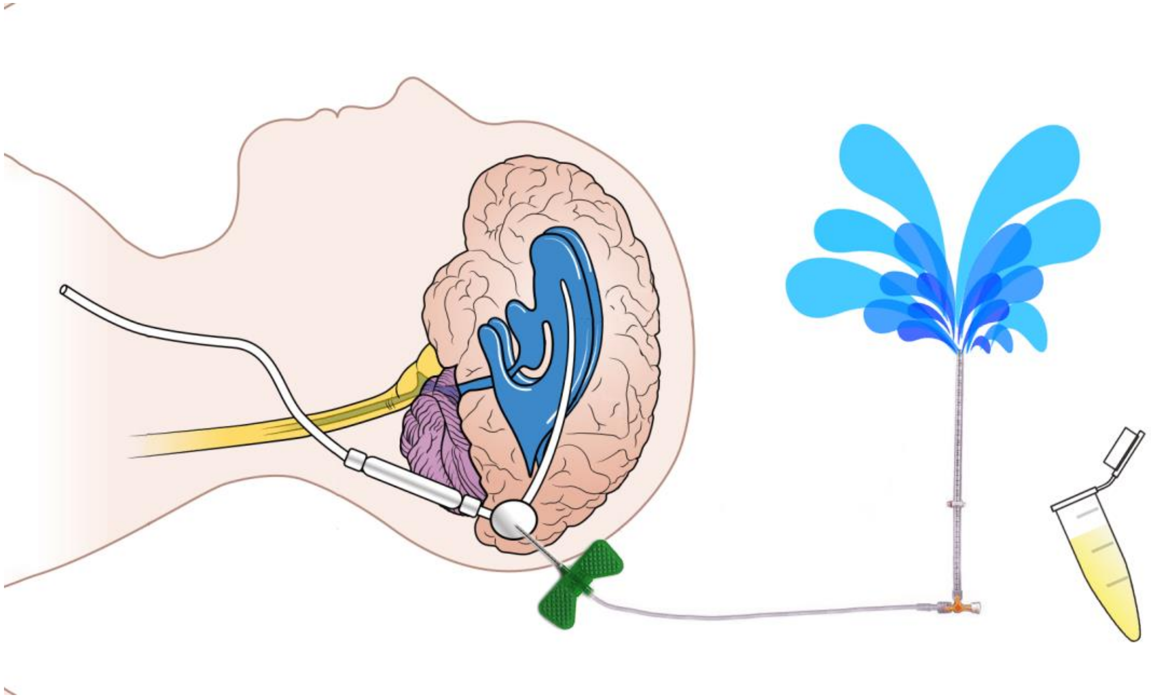


Current valves = programmable percutaneously

- Both ventricleperitoneal and lumboperitoneal
  - Allow pressure to be adjusted to patient needs
- Drawback: intense magnetic fields can affect valve parameters
  - High field magnetic resonance imaging (3 Tesla)
  - High voltage electric power lines
  - Theft detectors in shopping centers
  - Not mobile telephones

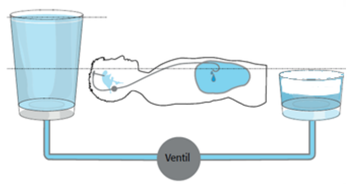
Valve reservoir puncture in emergency room

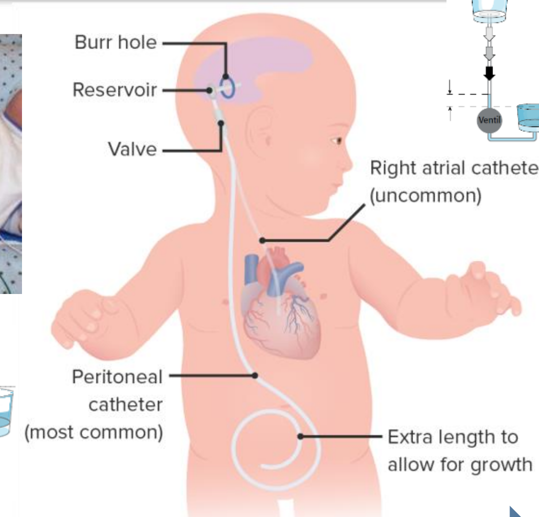
- Usefulness
  - Check ventricular catheter patency
  - Rule out CSF infection
  - Temporary measure to drain CSF?
- Risk of inducing shunt infection

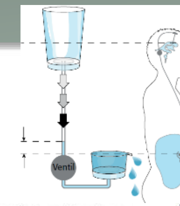


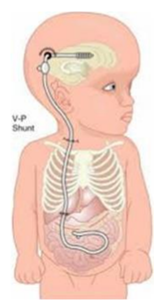
Infantile hydrocephalus problems

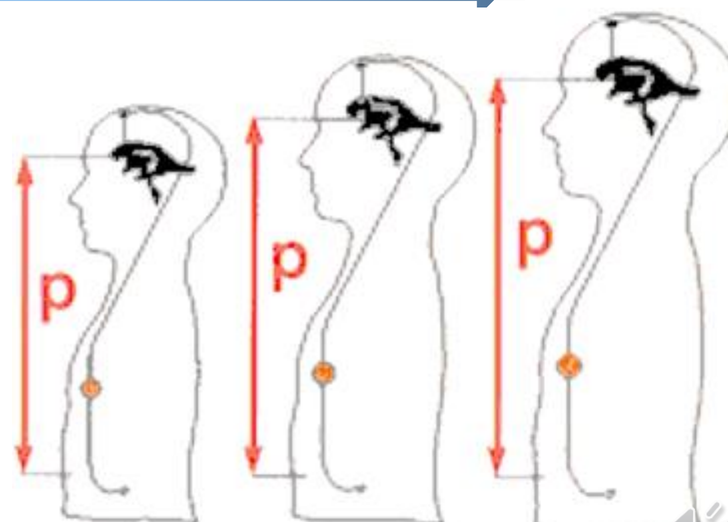
- **Patient growth**
  - Sufficiently long peritoneal catheter
  - Impossible in ventricleatrial shunt
  - Hydrostatic pressure changes as patient height increases
- **Intracranial and abdominal pressure changes**
  - Baby = recumbent, child standing
    - Siphoning effect?
  - Valve that works in infant not suitable in child / adolescent / adult
- **Result** = frequent CSF shunt surgical replacement in children





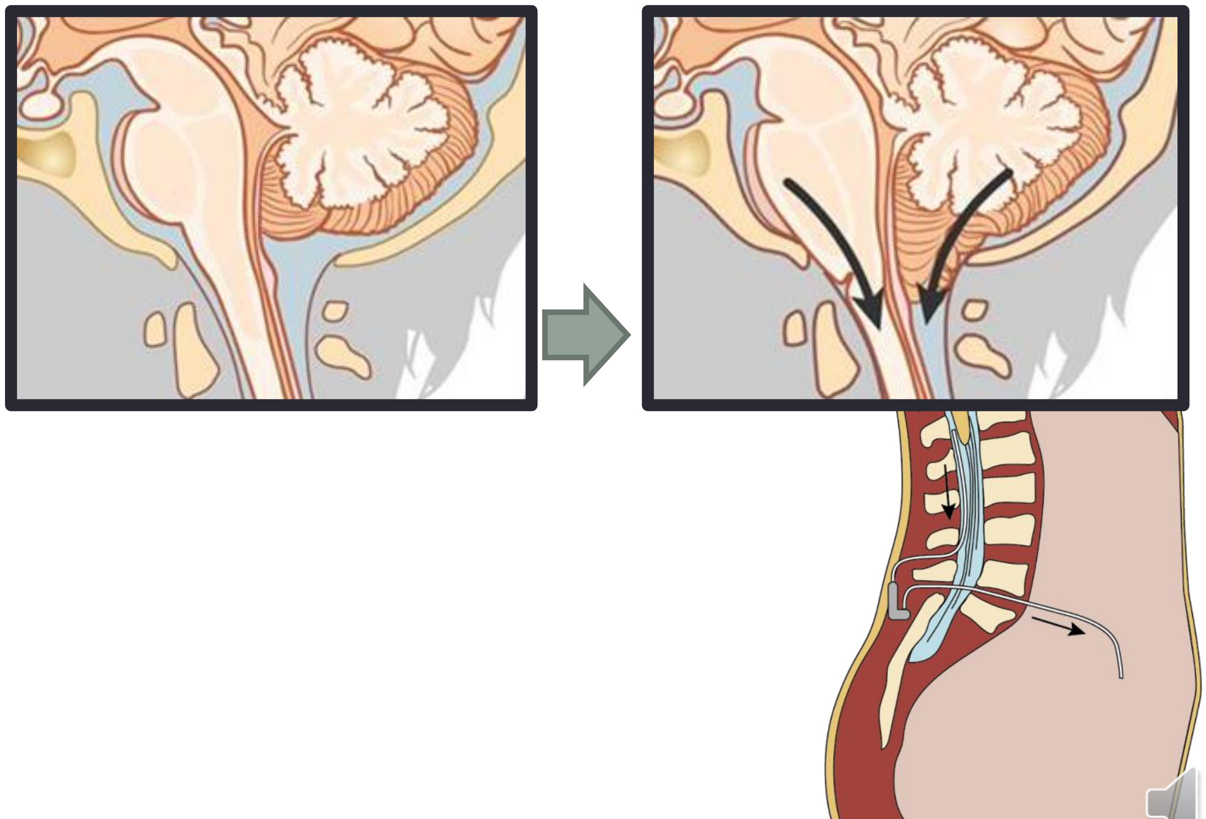






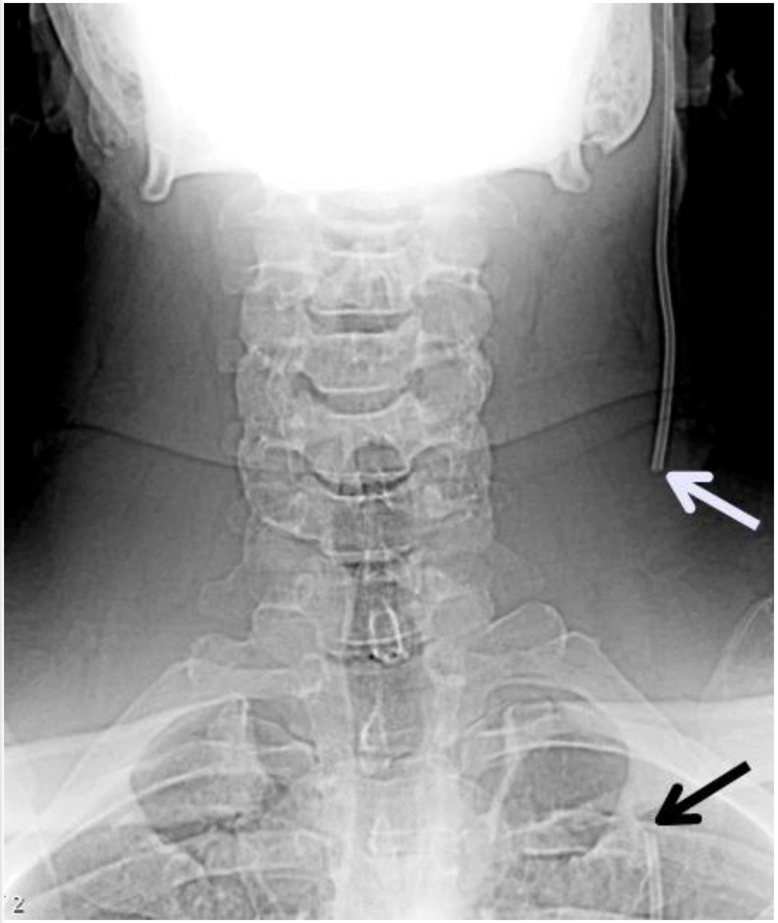
Lumbo-peritoneal shunt

- Advantage: easy surgical implantation
- Drawback: induction Chiari type 1 malformation
  - Occipital headache



Complications CSF shunts: catheter rupture

- Common in children as they grow
- ↑ at the collarbone level

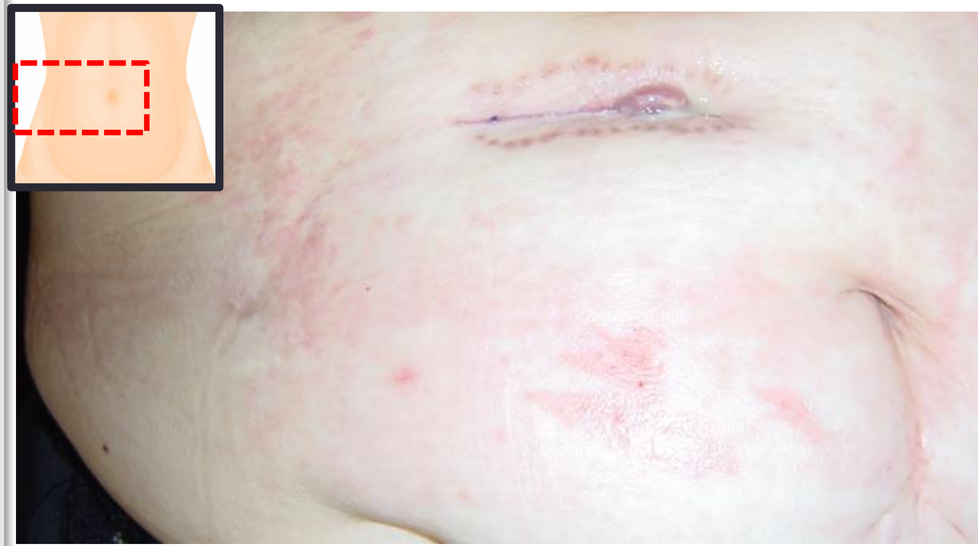


Complications CSF shunts: ventricular catheter obstruction

- The choroid plexus grows obstructing the ventricular catheter = frequent intraventricular haemorrhage when removed

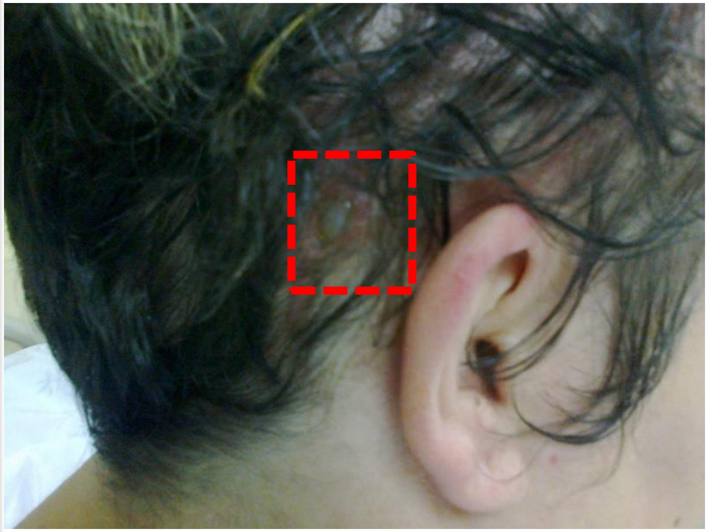
Complications CSF shunts: infection shunt system (1)

- Shunt = foreign body = ease of infection



Complications CSF shunts: infection shunt system (2)

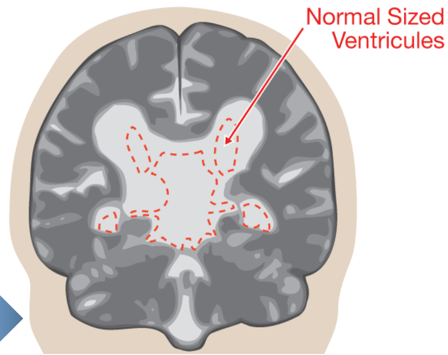
- Need shunt surgical replacement

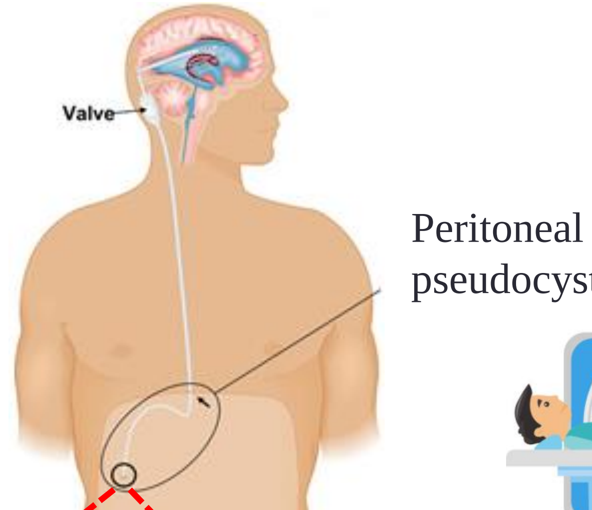


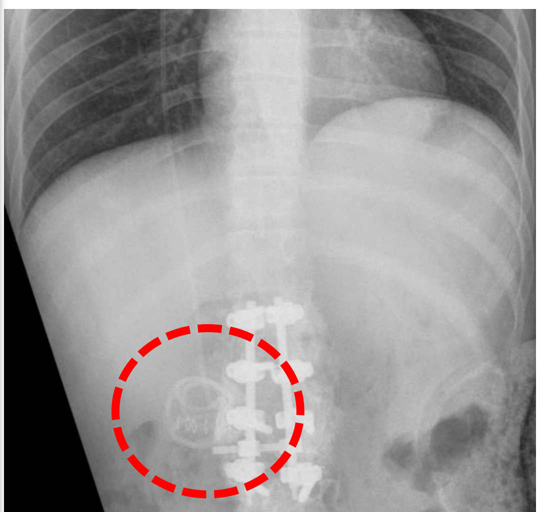
Complications CSF shunts: peritoneal pseudocyst

- Indicates shunt infection = surgical removal + antibiotics + new shunt

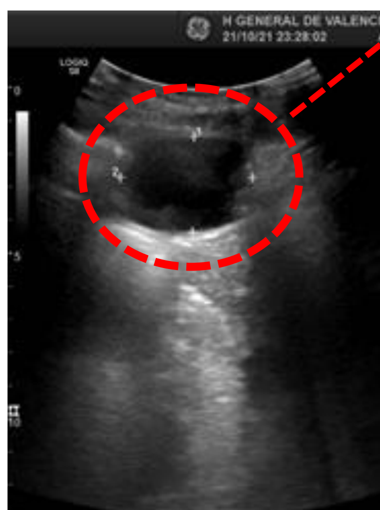
Shunt malfunction



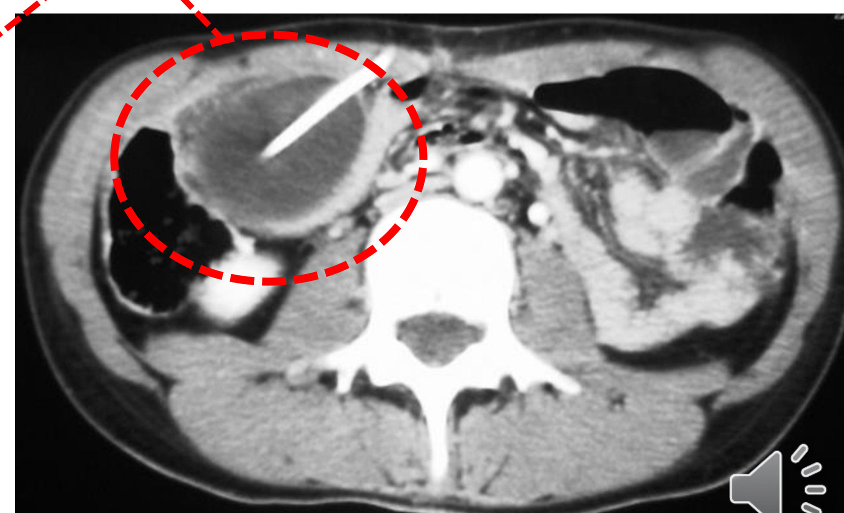




Plain x-ray



Abdominal ultrasound

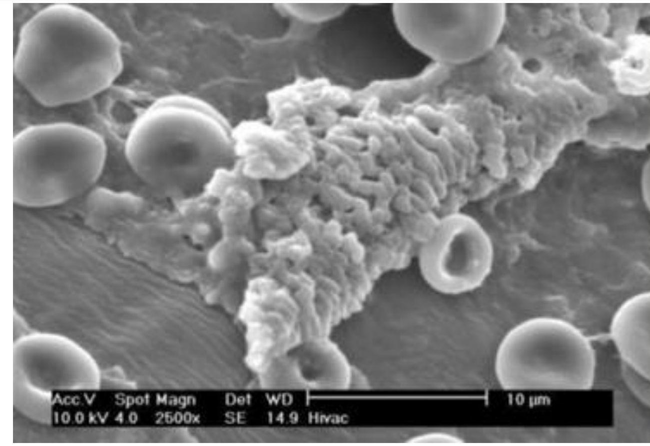


Abdominal CT

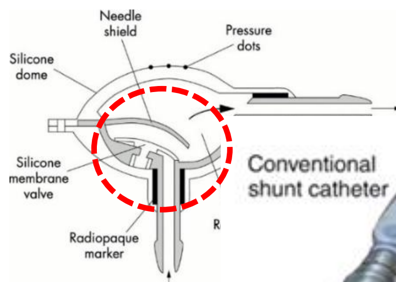
Complications CSF shunts: subclinical infection cause of CSF shunt malfunction (1)

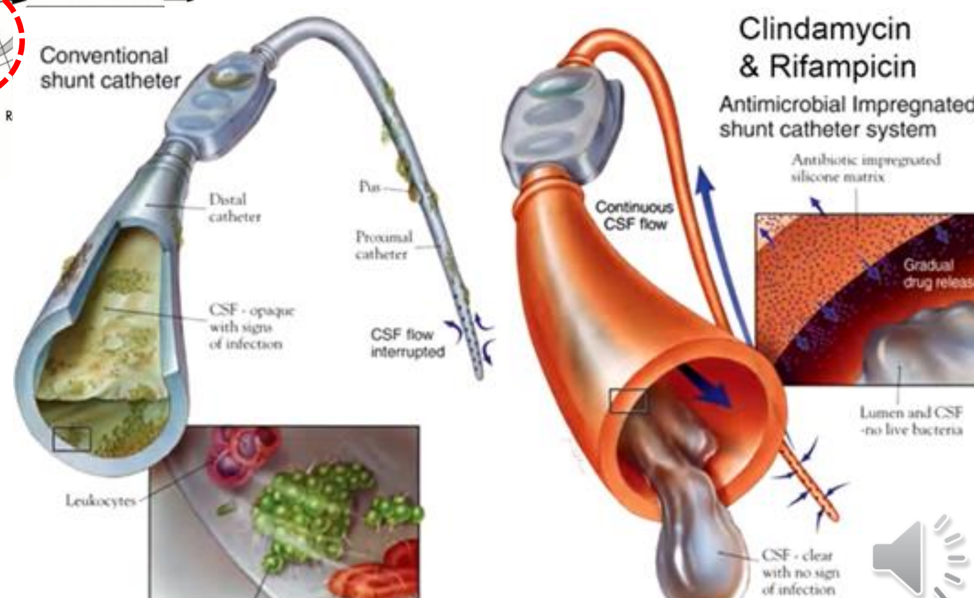
- ↑ Cause CSF shunt malfunction
- **Cause**: bacteria secreting 'slime'
  - No infection symptoms
  - Mucoprotein = valve blockage
- **Prophylaxis**: asepsis + implanting a shunt embedded with antibiotics
- **Treatment**: shunt removal, external ventricular drainage implantation, antibiotic treatment, new shunt implantation

Gram-Positive Organisms	Gram-Negative Organisms
Staphylococcus epidermidis	*Pseudomonas aeruginosa*
Staphylococcus aureus	*Serratia marcescens*
Corynebacterium spp	*Stenotrophomonas* spp
Streptococcus spp	*Enterobacter* spp
Enterococcus spp	*Escherichia coli*
Propionibacterium spp	*Klebsiella* spp



Bayston et al. 2006

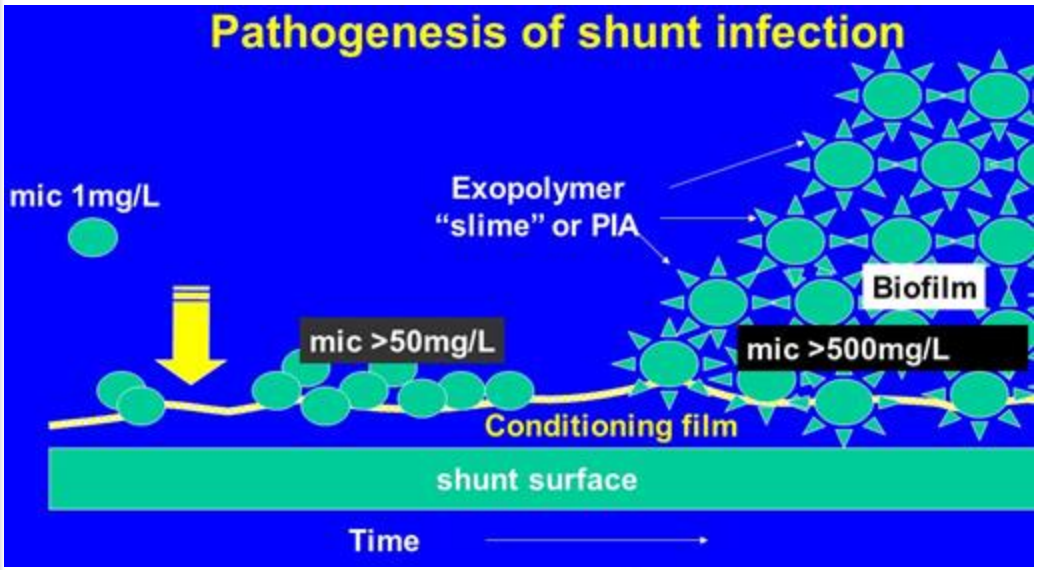


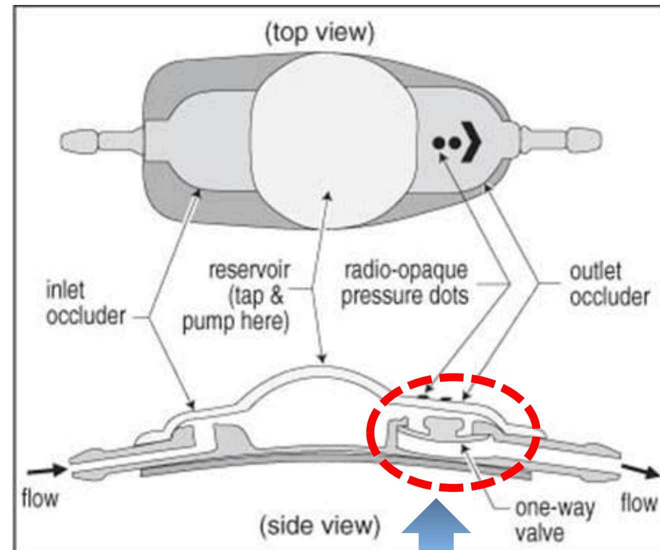


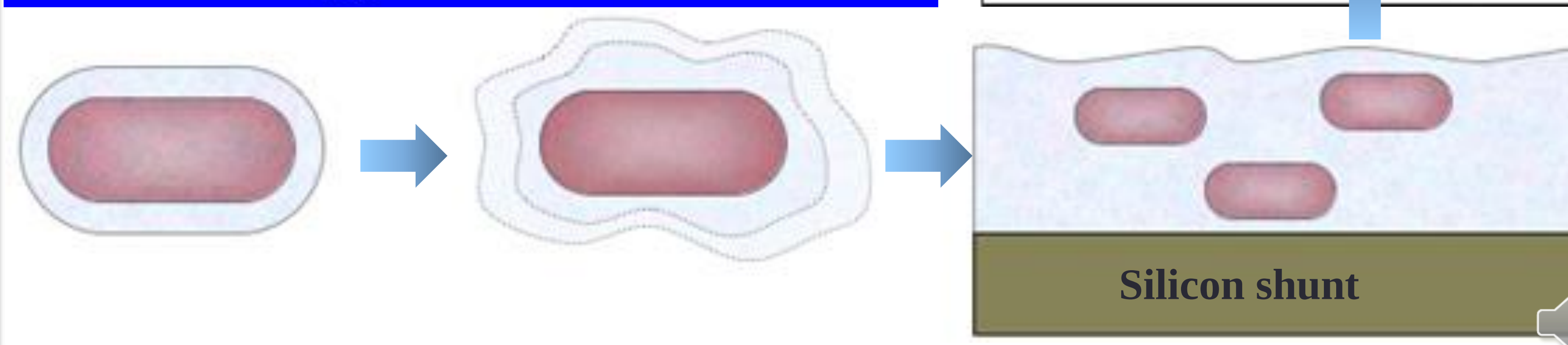
Sciubba et al., JNS 2005

Complications CSF shunts: subclinical infection cause of valve malfunction (2)

- Mucoprotein-secreting bacteria = immune system isolation = persistence low-grade infection







Bacterium Mucoprotein Biofilm

KEY CONCEPTS SEMINAR INTRACRANIAL HYPERTENSION AND HYDROCEPHALUS

- **Intracranial pressure**
  - Intracranial space volume = constant
  - Any increase in volume of one component = decrease of the other two or ↑ICP
- **Benign intracranial hypertension**
  - Mostly affects obese women of childbearing age taking contraceptive drugs
  - Treatment to correct overweight, withdraw contraceptives, and shunt CSF
- **Hydrocephalus**
  - Intracranial CSF volume increase
  - Dangers: atrophy cerebral parenchyma, psychomotor retardation, & blindness
- **Normal pressure hydrocephalus**
  - Better response for gait disorder and urine incontinence than for neurocognitive impairment
  - Vital differential diagnosis with other types of dementia
- **CSF shunt systems**
  - Valve selection appropriate for patient needs
  - Frequent complications due to inappropriate or excessive drainage or infections

ANY QUESTIONS?

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