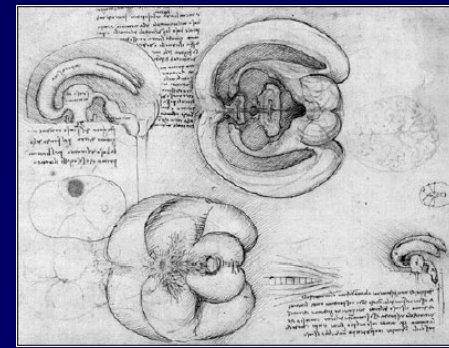


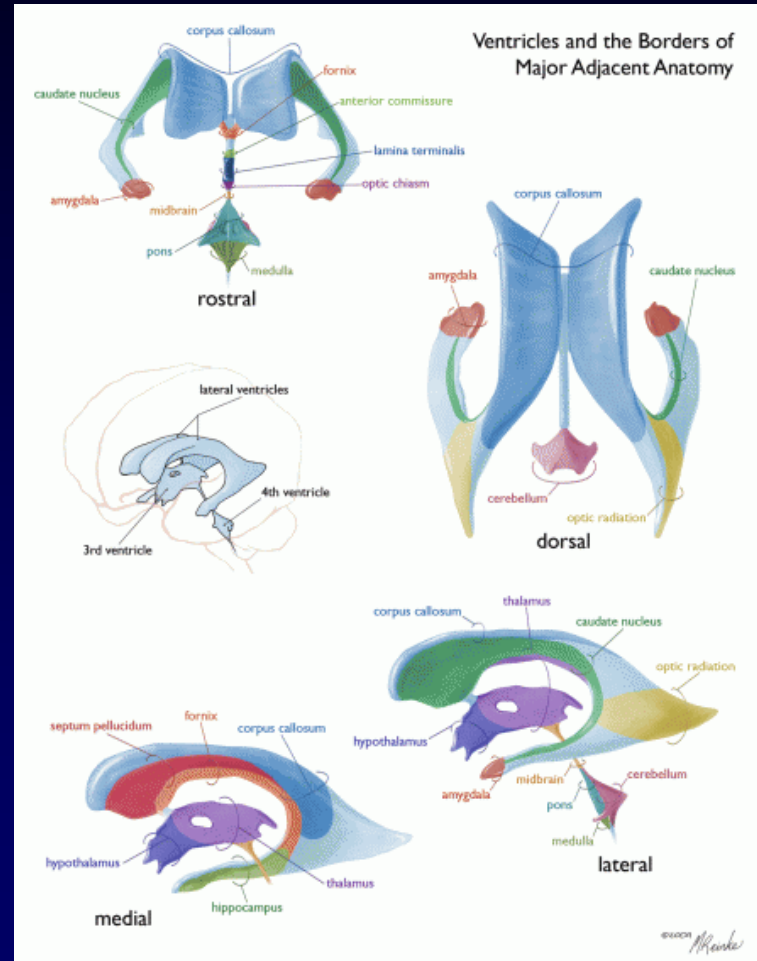
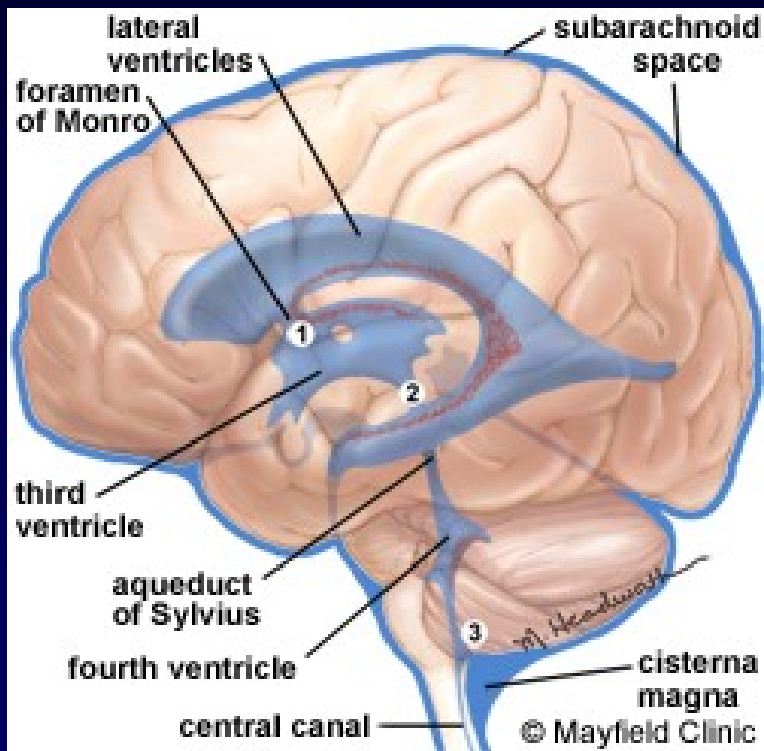
Hydrocephalus

Definition

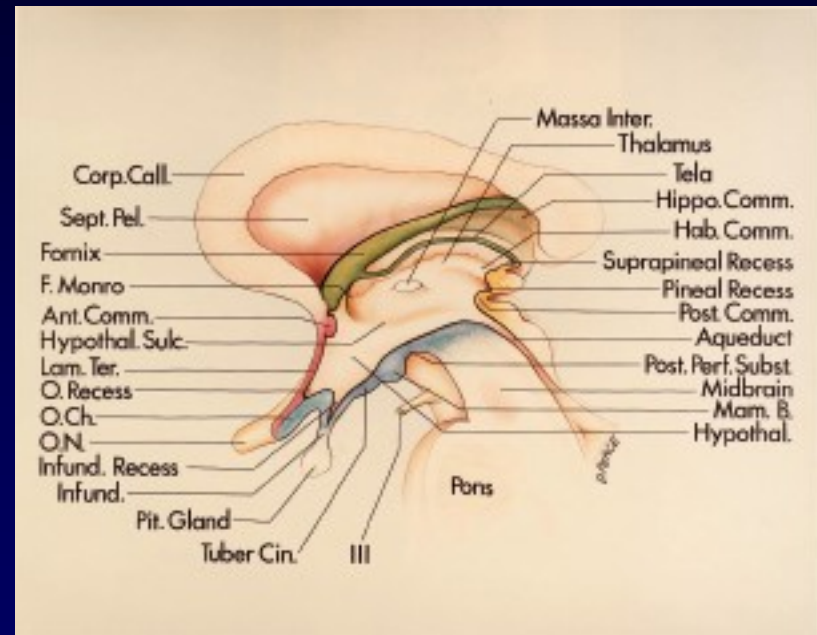
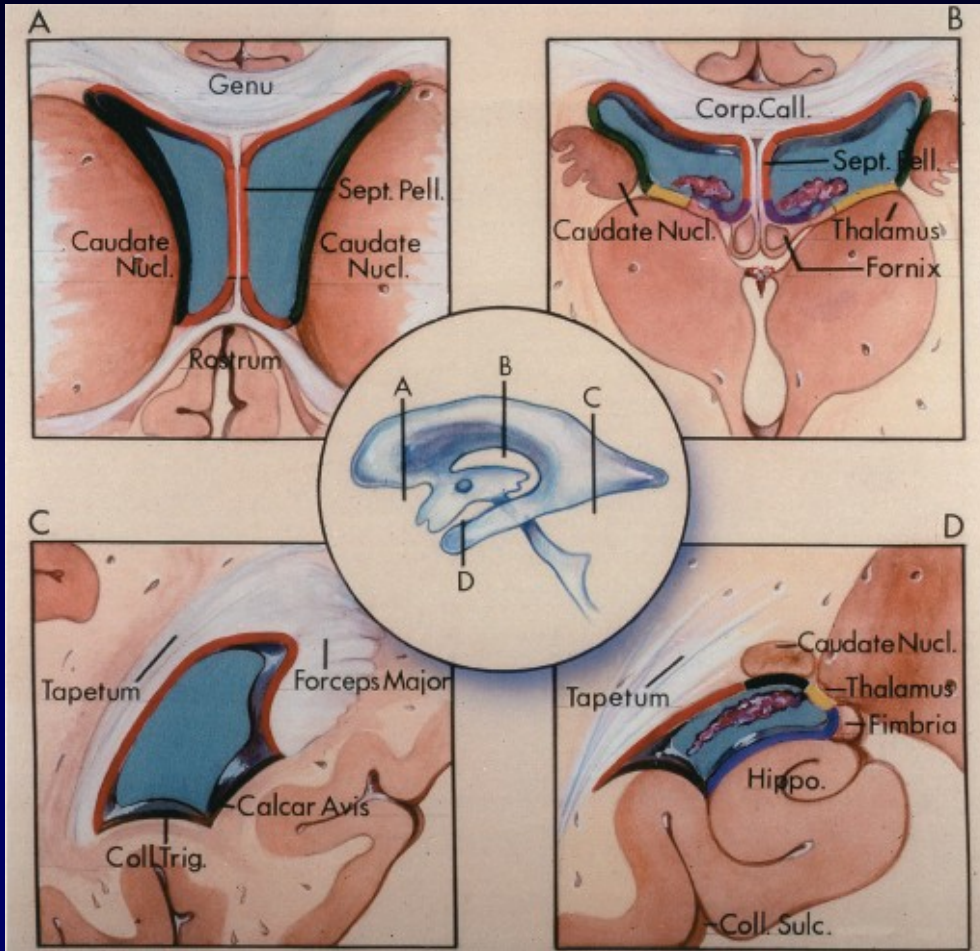
- **Hydrocephalus** – from the Greek words „hydro“ meaning water and „cephalus“ meaning head.
- **Hydrocephalus** can be defined broadly as a disturbance of formation, flow, or absorption of cerebrospinal fluid (CSF) that leads to an increase in volume occupied by this fluid in the CNS.
- This condition also could be termed a hydrodynamic disorder of CSF.
- Cerebral atrophy and focal destructive lesions - are not the result of a hydrodynamic disorder.



Anatomy

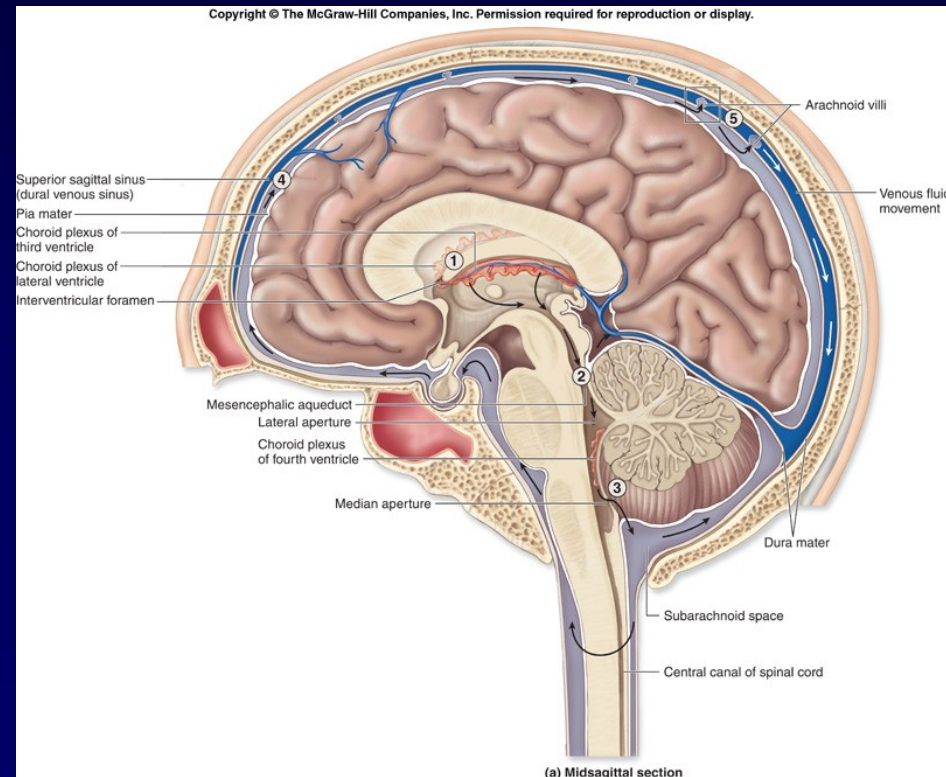


Anatomy



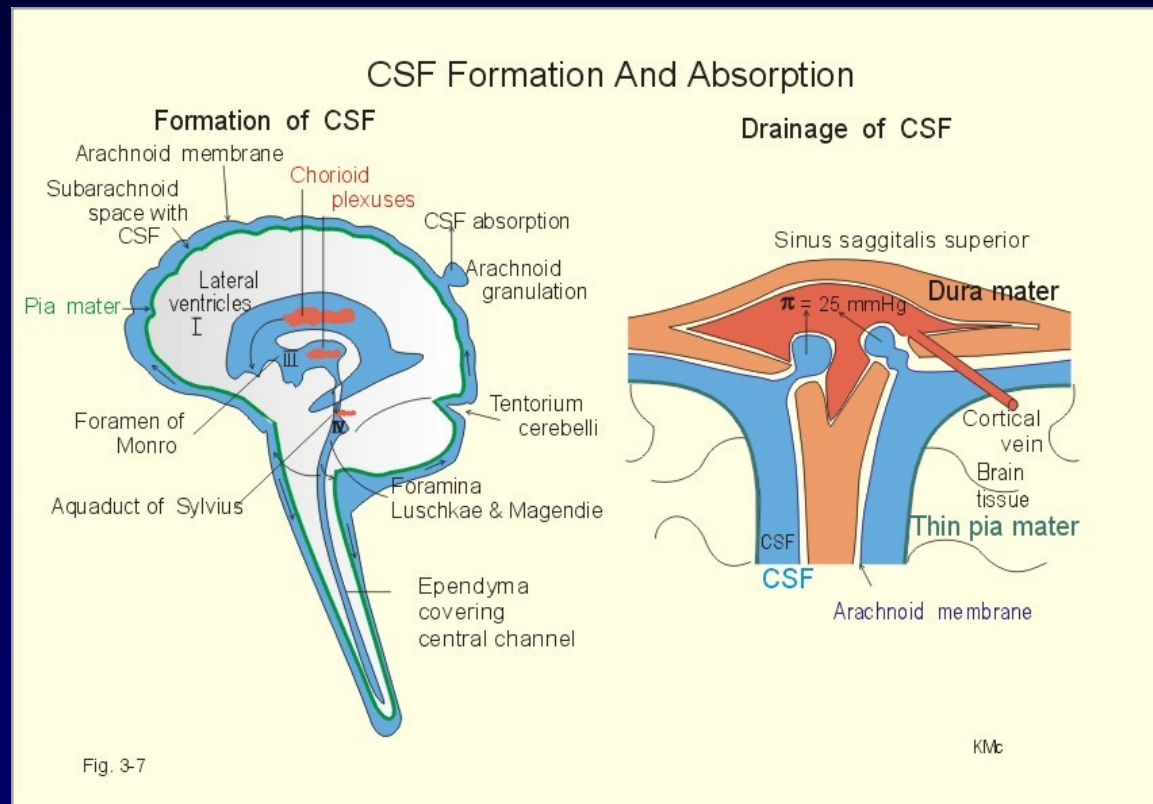
Physiology

- CSF is formed by the choroid plexus (~80%), the parenchyma (~20%) and the ependyma (negligible).
- CSF 120 ml (80-150).
- 500 ml /24 hours.
- CSF production is pressure independent under normal physiological conditions.



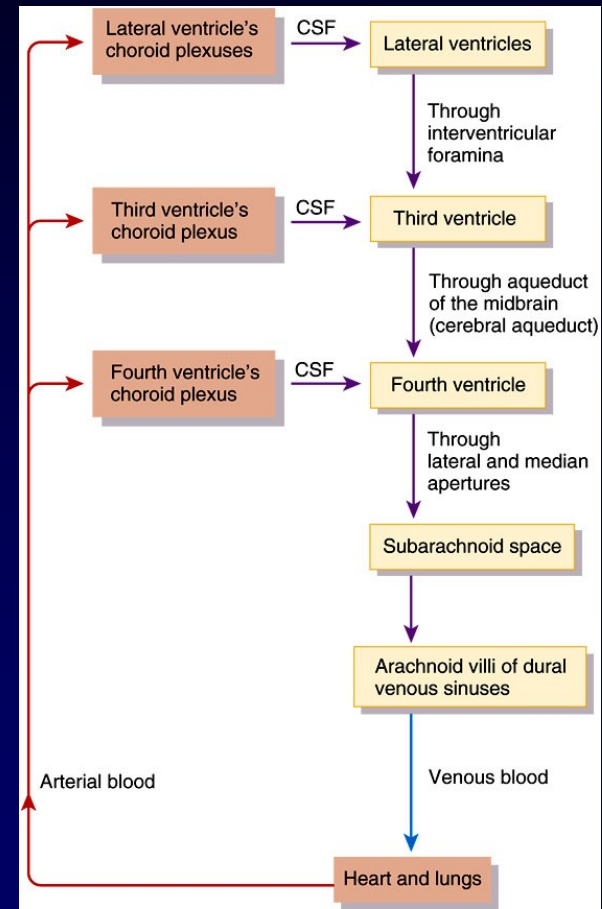
Physiology

- The only proven force responsible for bulk CSF absorption is that of a hydrostatic gradient.
- The arachnoid villi drain CSF.
- Resorption is dependent on ICP.
- Accessory routes: the mucosa of the paranasal sinuses, nasal mucosa, cranial nerve root sheaths, and cervical lymph nodes.



Physiology

Circulation of CSF

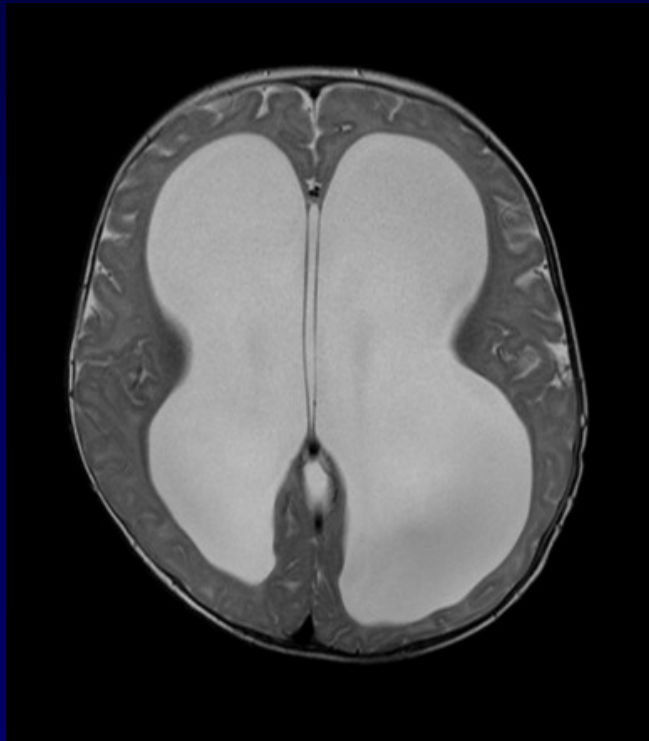


(d) Summary of the formation, circulation, and absorption of cerebrospinal fluid (CSF)

Epidemiology

Estimated prevalence: 1-1,5%.

Incidence of congenital hydrocephalus 0,5-2,5/1000 births.



Epidemiology

Some common causes of **pediatric hydrocephalus**:

- **myelomeningocele (20%)**
- **post-intraventricular hemorrhage of prematurity (15%)**
- **congenital aqueductal stenosis (12%)**
- **communicating hydrocephalus of unknown origin (10%)**
- **neoplasm (8%)**
- **post-infectious (7%)**
- **post-traumatic (3%)**
- **arachnoid cyst (5%)**
- **dural sinus thrombosis**
- **syndrome-related (Dandy-Walker malformation etc.)**

Epidemiology

Causes of hydrocephalus in **adults**:

- **subarachnoid hemorrhage (SAH)** (one third)
- **idiopathic hydrocephalus** (one third)
- **head injury**
- **tumors**
- **prior posterior fossa surgery** may cause hydrocephalus by blocking normal pathways of CSF flow.
- **congenital aqueductal stenosis** causes hydrocephalus but may not be symptomatic until adulthood.
- **meningitis, especially bacterial.**

Symptoms

Influenced by:

- patient's age
- cause
- location of obstruction
- duration and rapidity of onset
- vary greatly from person to person

Symptoms - infants

- abnormal enlargement of the head
- soft spot (fontanel) is tense and bulging
- scalp can appear thin
- bones separated in baby's head
- prominent scalp veins
- vomiting
- drowsiness
- irritability
- downward deviation of baby's eyes
- seizures
- poor appetite.



Symptoms – toddlers/children

- abnormal enlargement of baby's head
- headache
- nausea, vomiting
- blurred or double vision
- unstable balance
- irritability
- sleepiness
- delayed progress in walking or talking
- poor coordination
- change in personality
- inability to concentrate
- loss of sensory motor functions
- seizures
- poor appetite
- Older children may experience difficulty in remaining awake or waking up.



Symptoms – adults

- headache
- difficulty in remaining awake or waking up
- loss of coordination or balance
- bladder control problems
- impaired vision and cognitive skills that may affect job performance and personal skills.



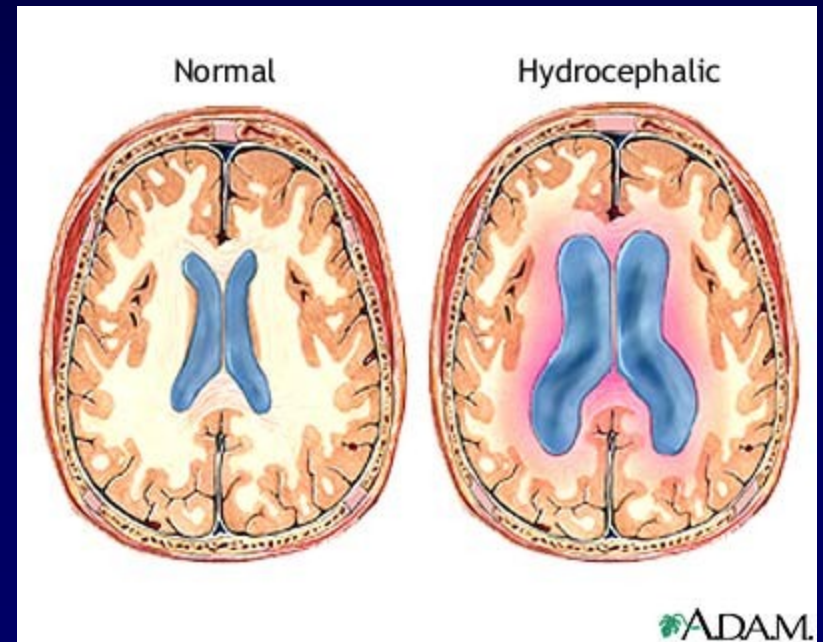
Symptoms – older adults

- loss of coordination or balance
- shuffling gait
- memory loss
- headache
- bladder control problems

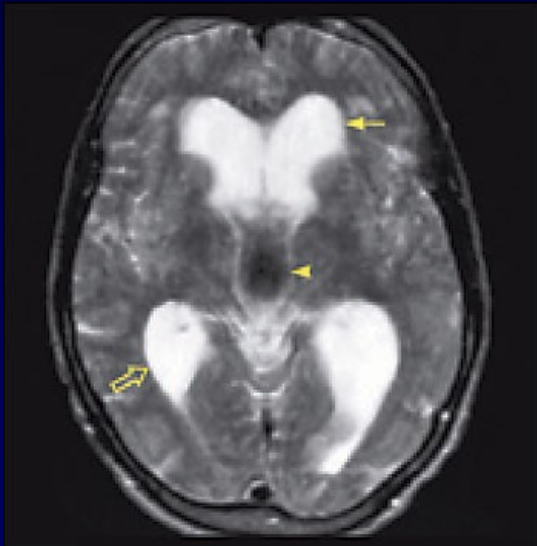


Imaging studies

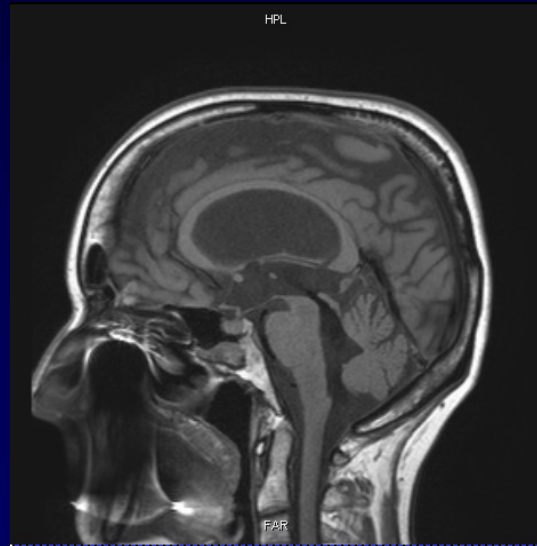
- MRI
- CT
- X-ray (plain, shunt-o-gram)
- SPECT
- PET
- Ultrasonography (infants)
- (Radionuclide cisternography)



Imaging studies - MR



T2W - axial



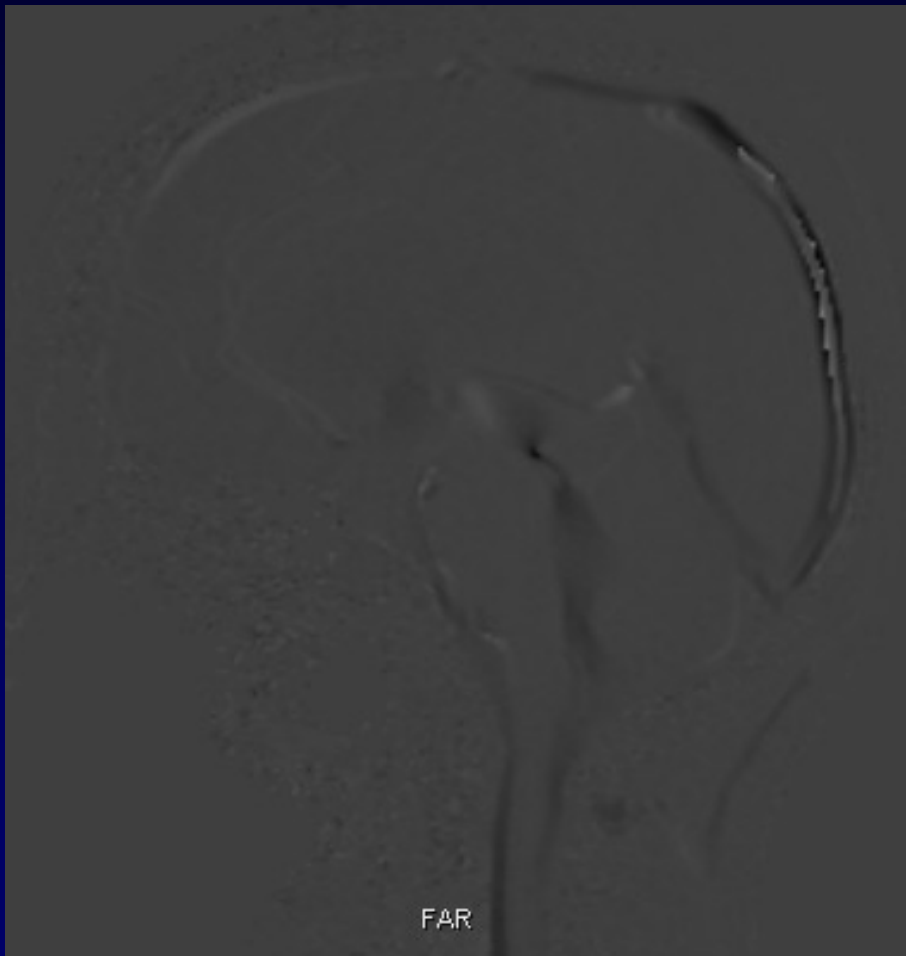
T1W - sagittal



T2W - sagittal

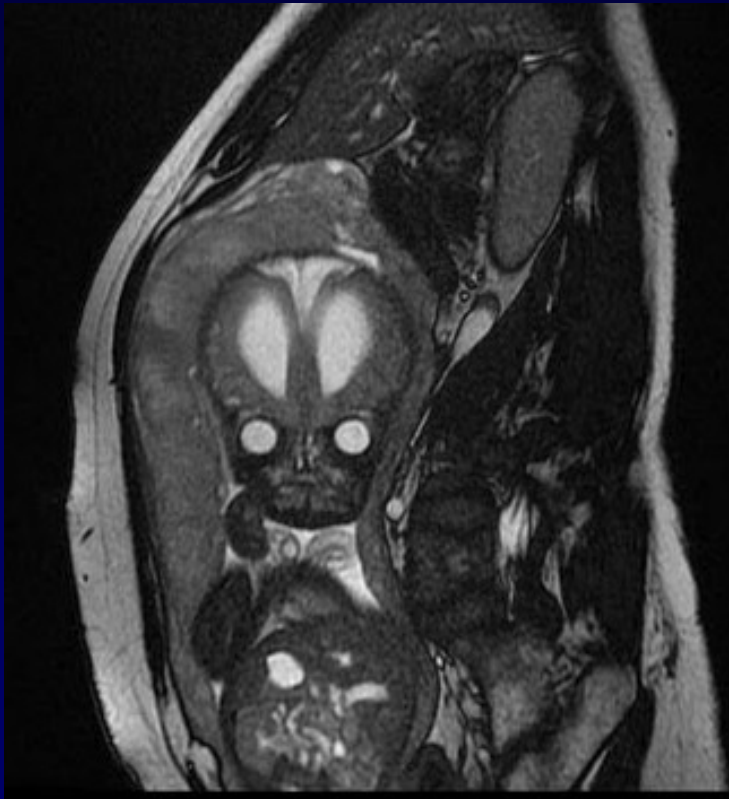
Imaging studies - MR

Cine MR

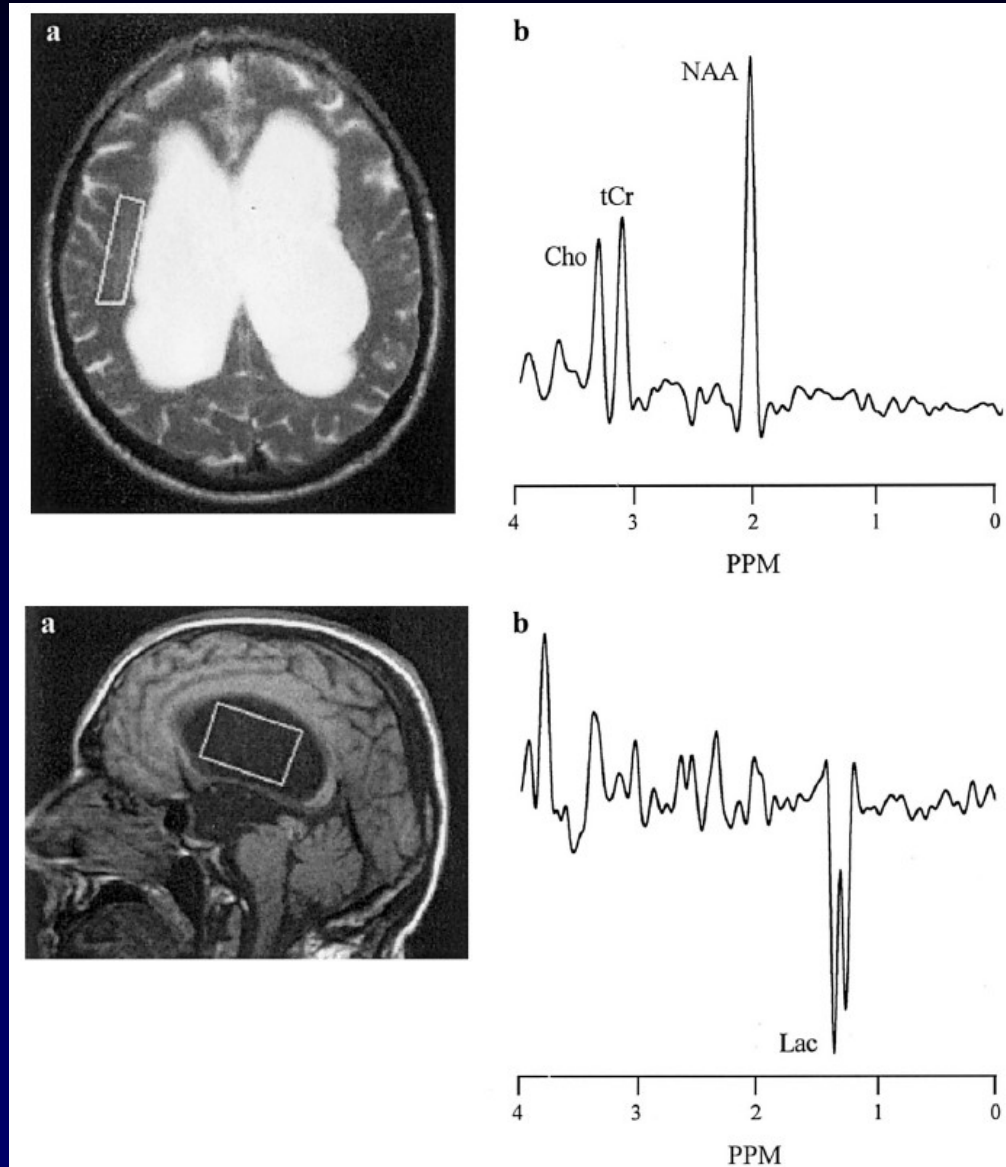


Imaging studies - MR

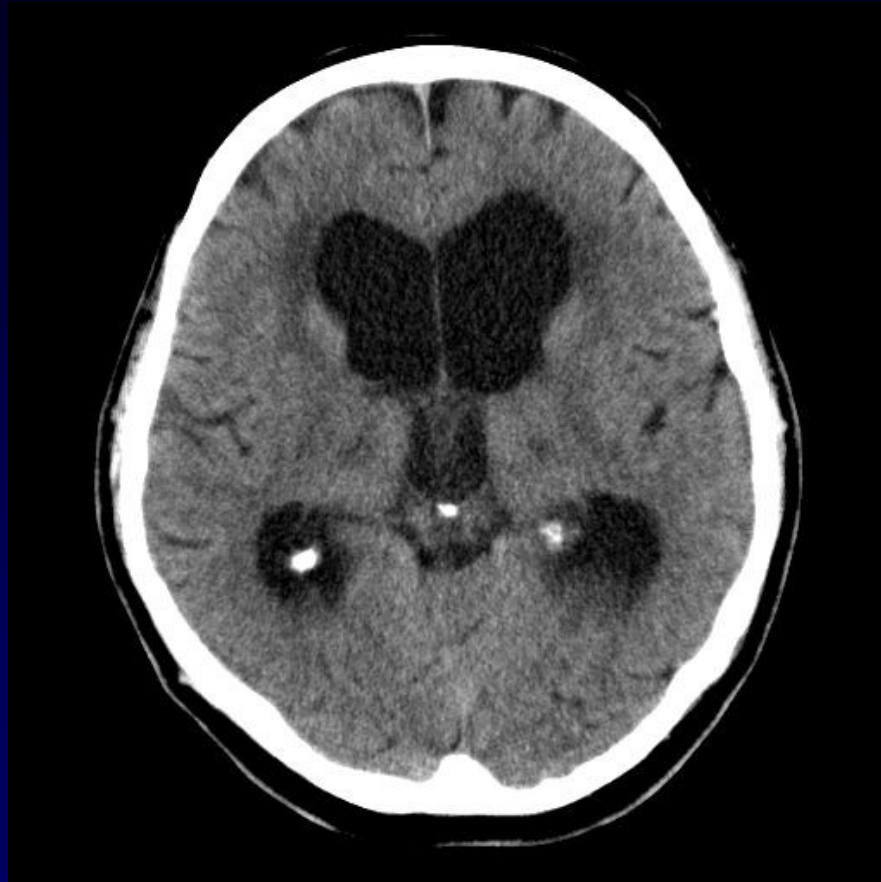
Prenatal MR



Imaging studies – MR spectroscopy

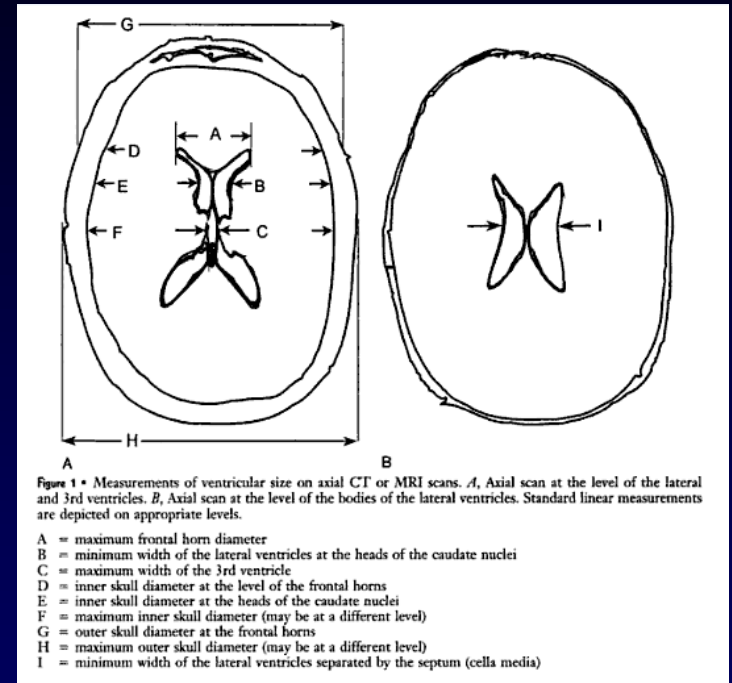
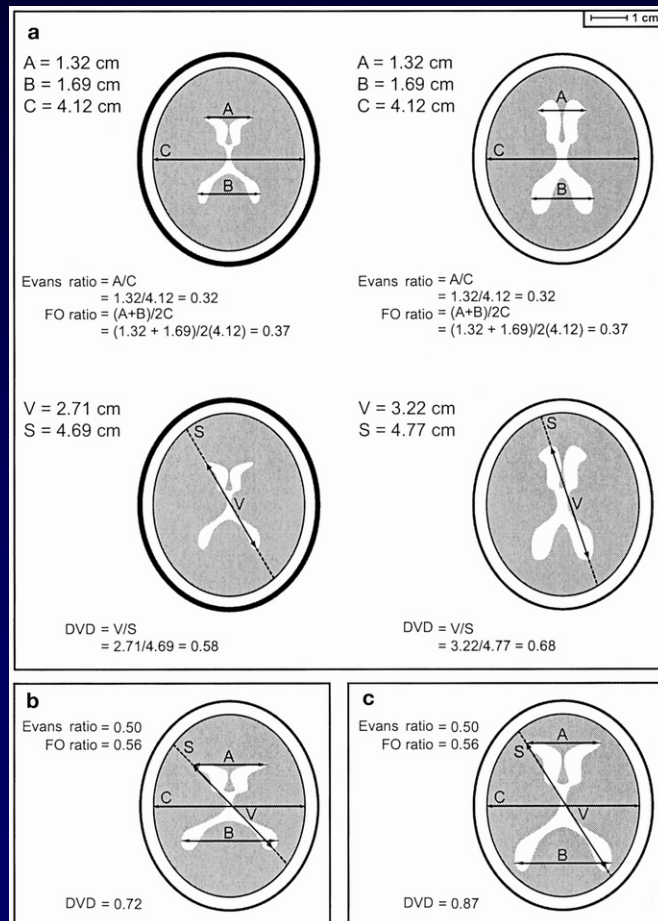


Imaging studies - CT



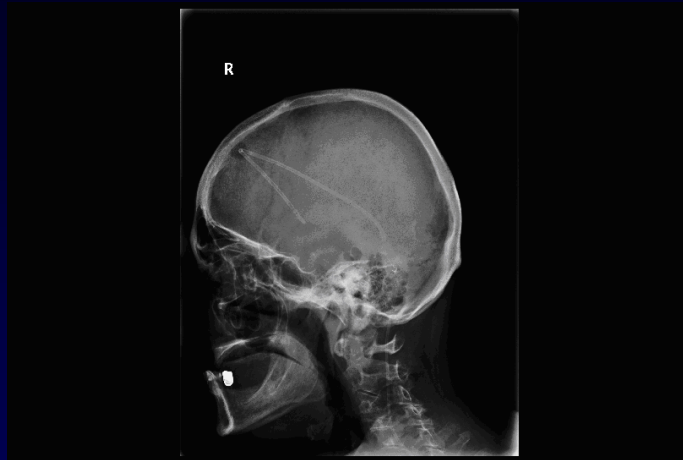
Ventricular indexes

Evan's (less than 0,3)

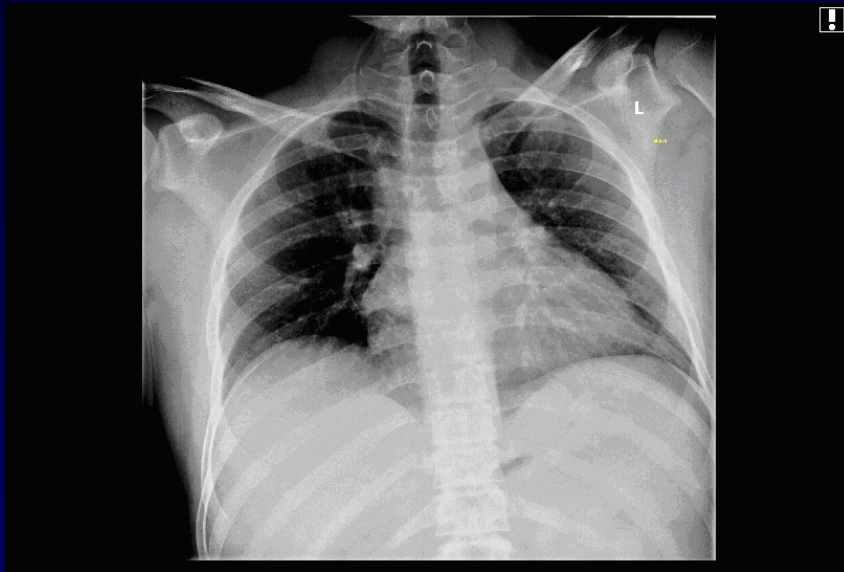


Evan's ratio (ER) = A/F
 Bifrontal index (BFI) = A/D
 Bicaudate index (BCI) = B/E
 Cella media index (CMI) = H/I
 Frontal horn index (FHI) = G/A
 Ventricular index (VI) = B/A
 Huckman number (HN) = $A + B$

Imaging studies – X-ray



Imaging studies – X-ray

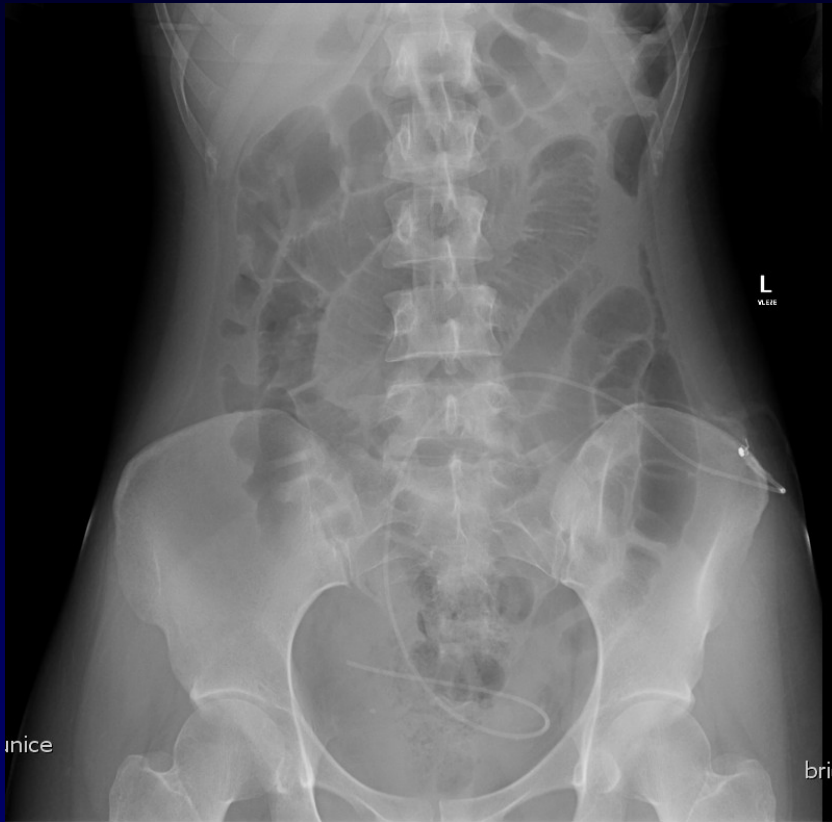


V-A shunt



V-P shunt

Imaging studies – X-ray



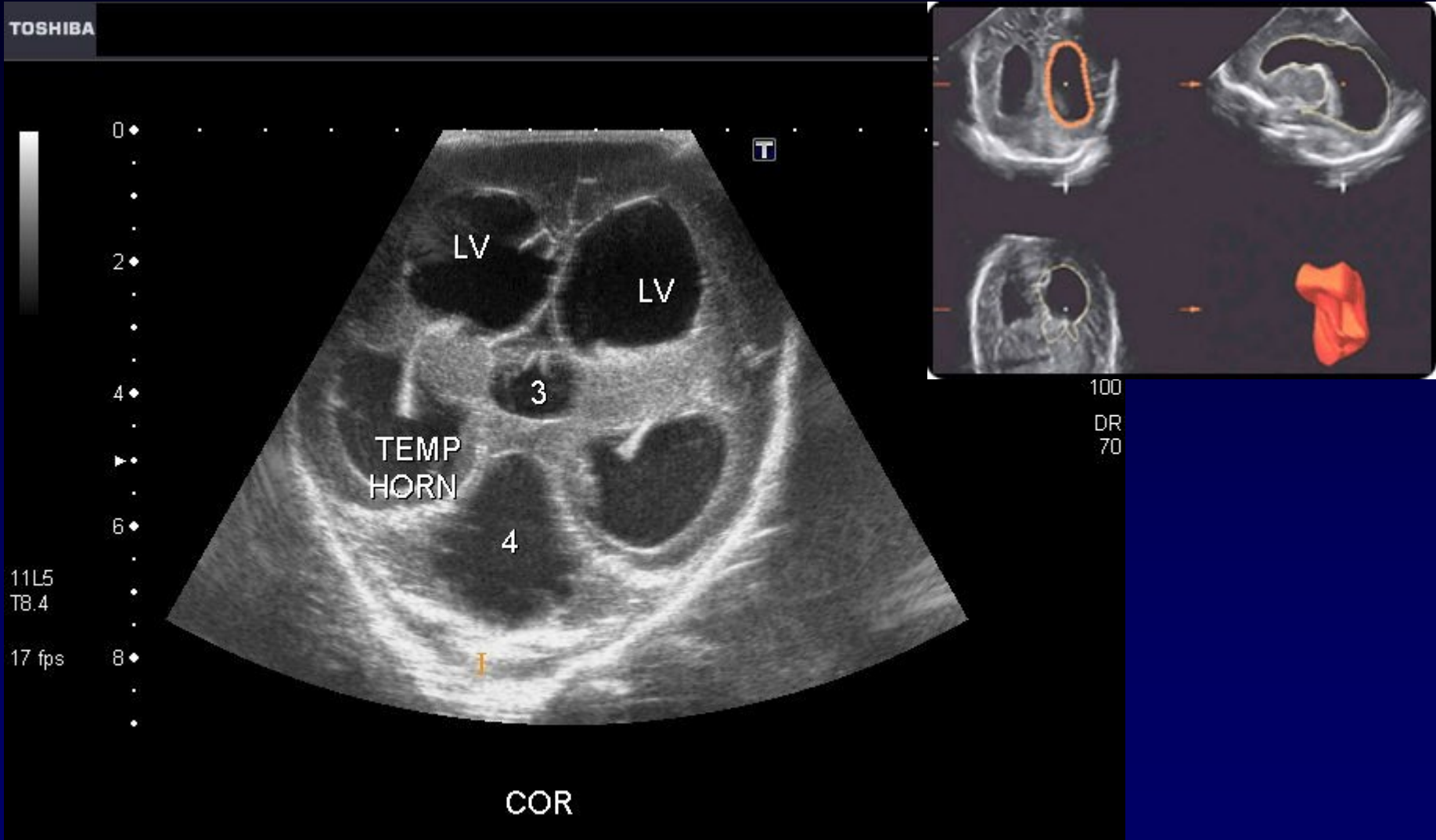
LP shunt

Imaging studies – X-ray



Shunt-o-gram

Imaging studies - ultrasonography

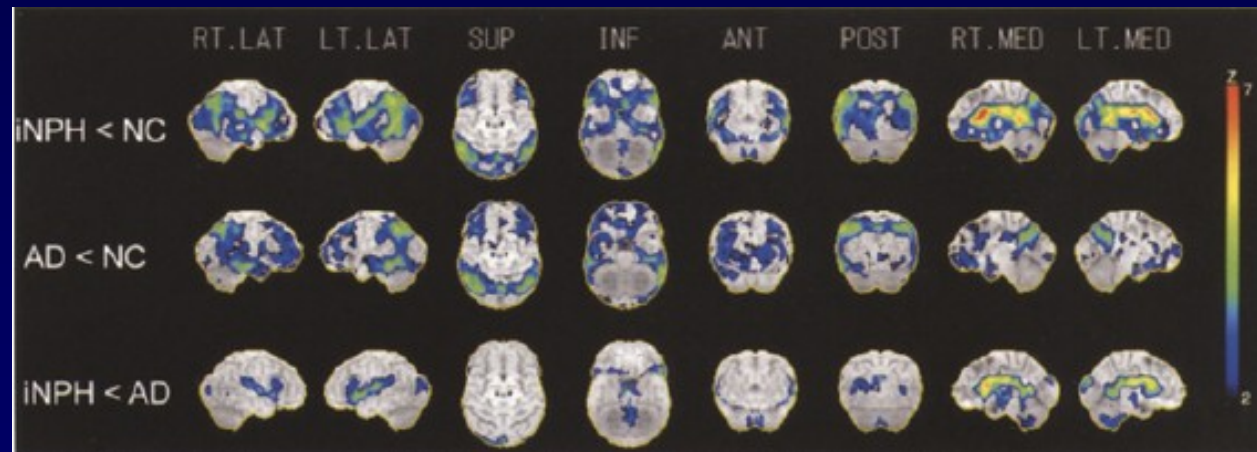
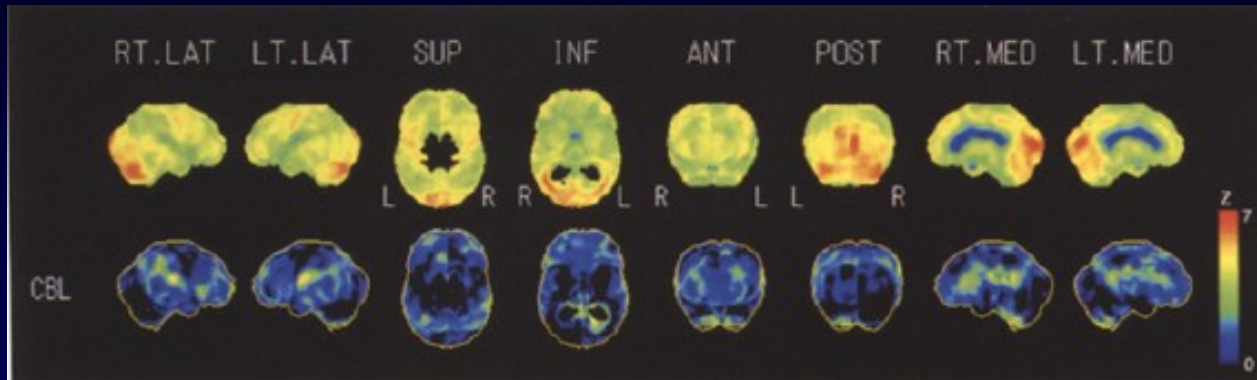


Imaging studies - ultrasonography

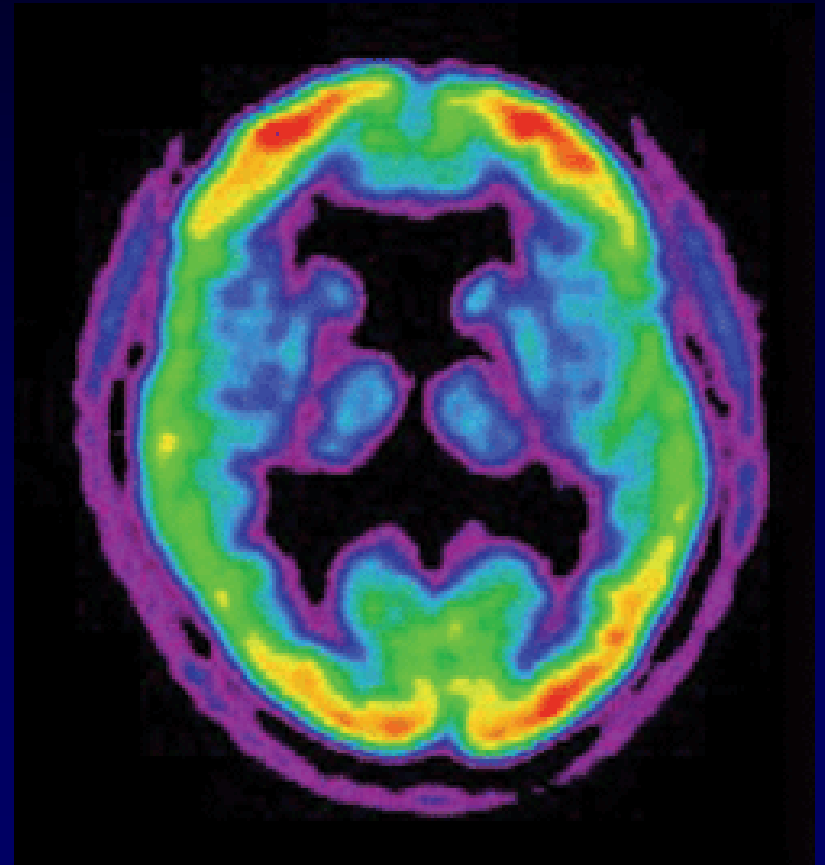
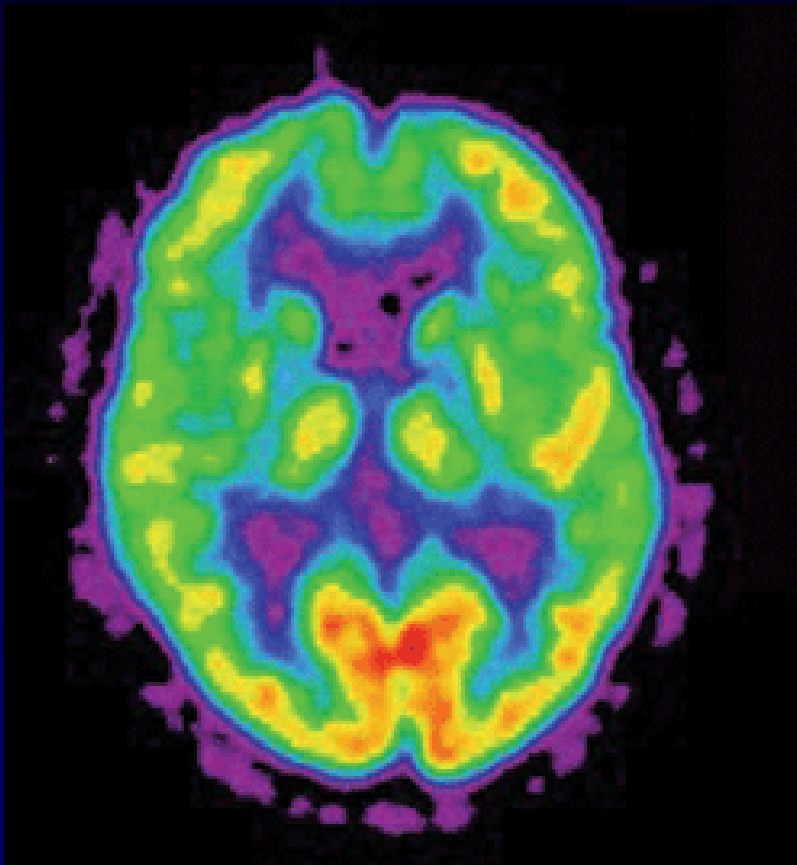


Fetus ultrasonography, 3D reconstruction

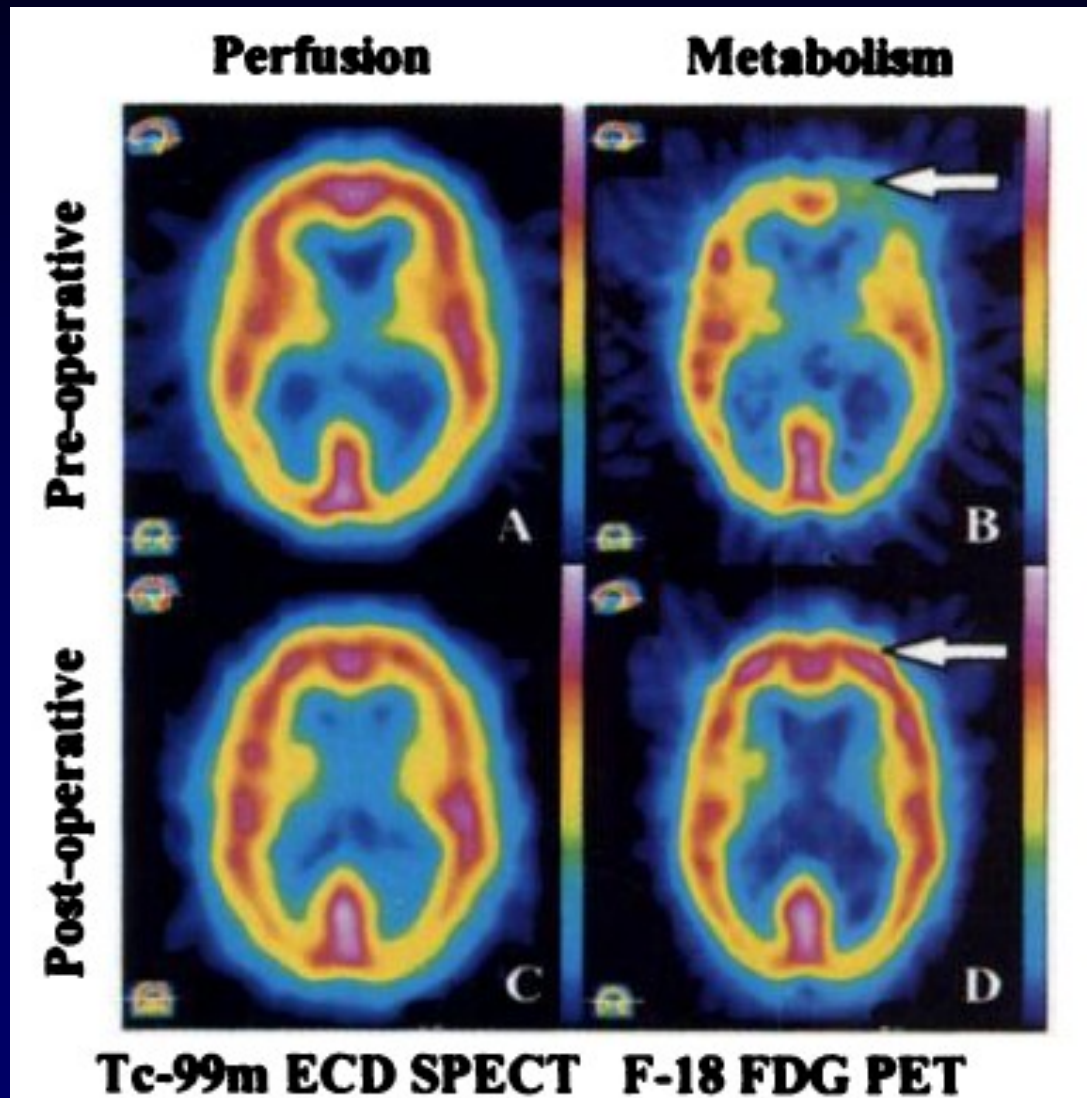
Imaging studies – SPECT



Imaging studies – PET



Imaging studies – SPECT, PET



Classification

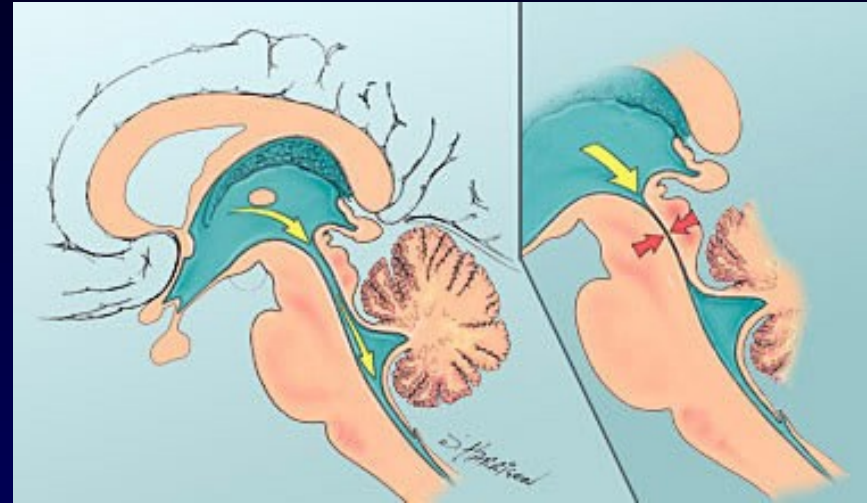
- **Communicating**
- **Non-communicating (obstructive)**

- **Congenital**
- **Acquire**

- **Pediatric**
- **Adult**

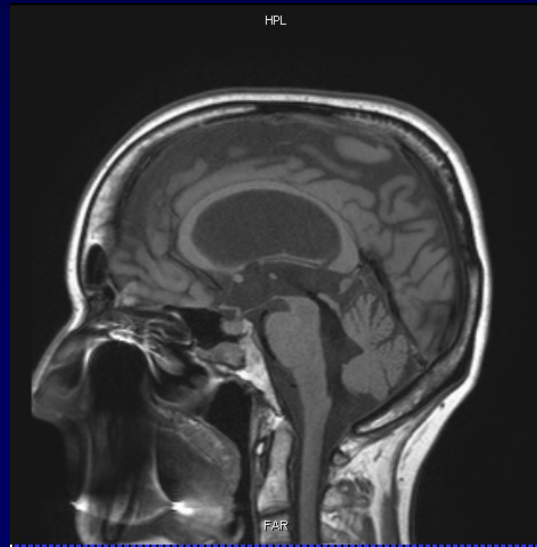
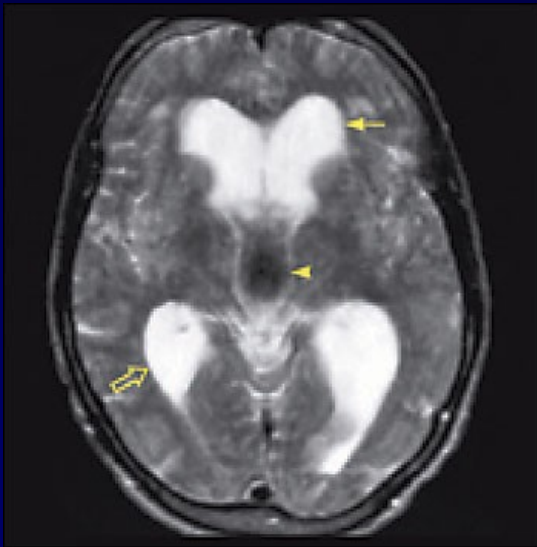
- **Special types of HCP**
 - Normal pressure hydrocephalus
 - Hydrocephalus ex vacuo
 - Others

- **Acute hydrocephalus occurs over days,**
- **subacute hydrocephalus occurs over weeks,**
- **chronic hydrocephalus occurs over months or years.**



Communicating hydrocephalus

Communicating hydrocephalus occurs when the flow of CSF is blocked after it exits the ventricles. This form is called communicating because the CSF can still flow between the ventricles, which remain open.

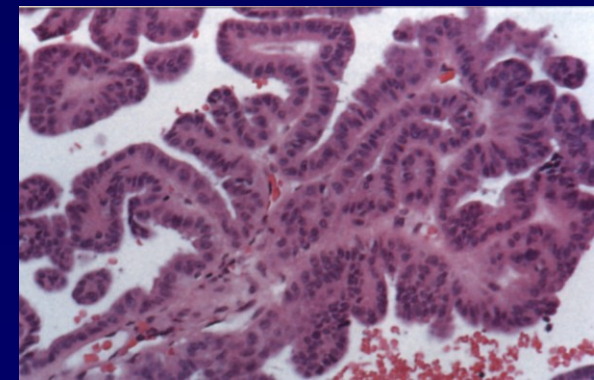
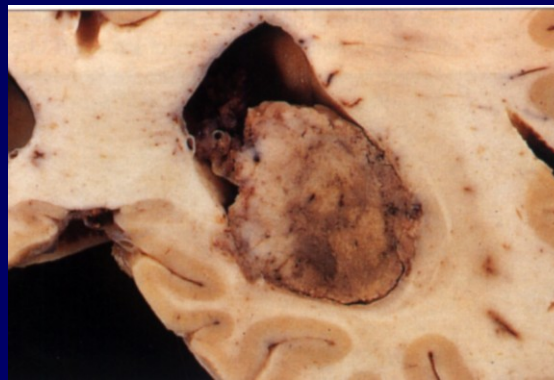
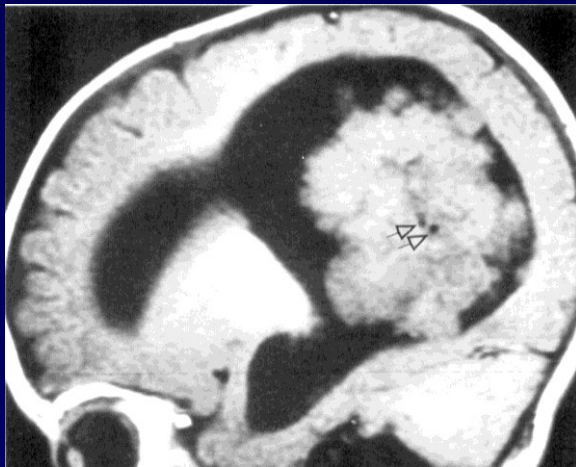
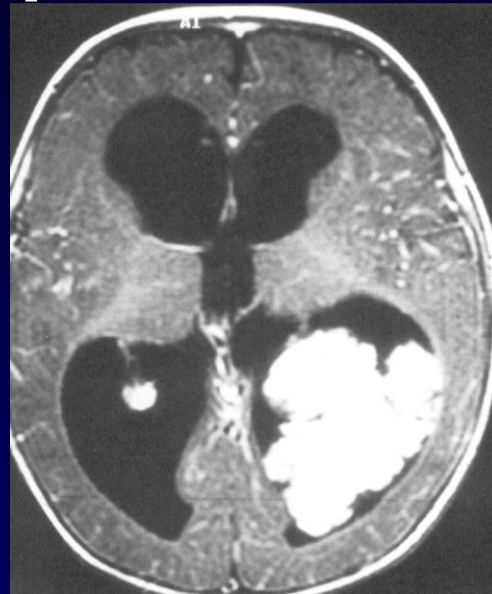


Communicating hydrocephalus

- **Increased CSF production (rarely)**
- **Decreased CSF resorption (most often)**
 - **posthemorrhagic (SAH, ICH)**
 - **postinfectious**
 - **tumors**
 - **dural sinus thrombosis**
 - **idiopathic**

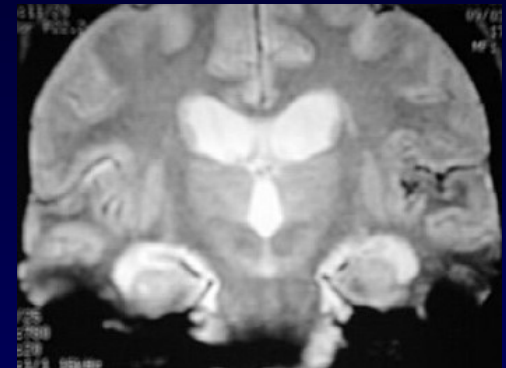
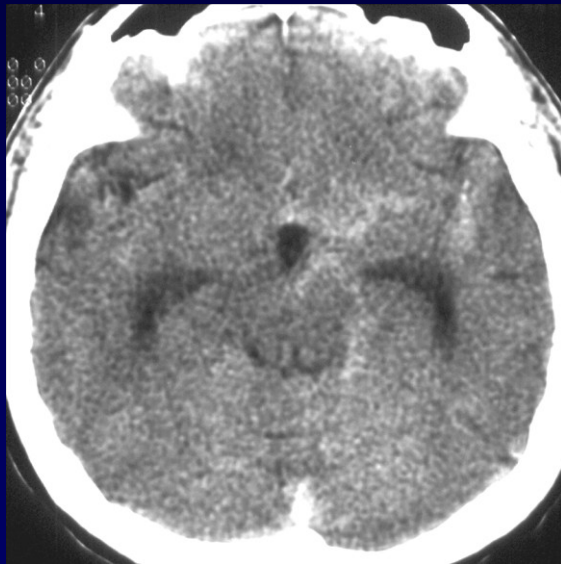
Communicating hydrocephalus

Increased CSF production
(plexus papiloma)



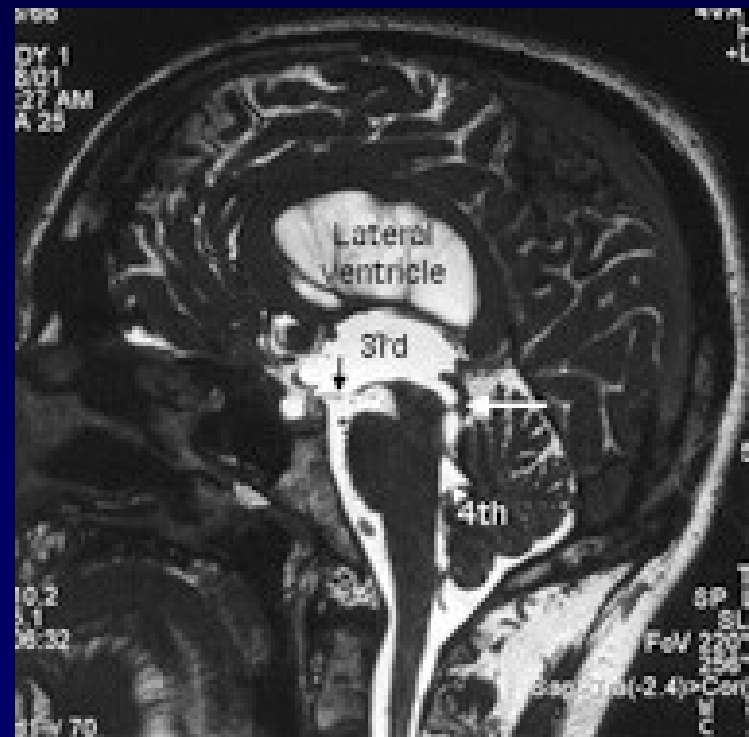
Communicating hydrocephalus

Decreased CSF
resorption (SAH)



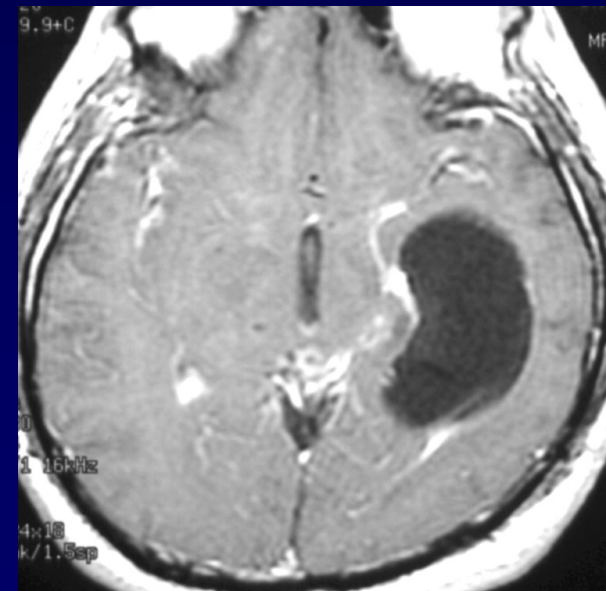
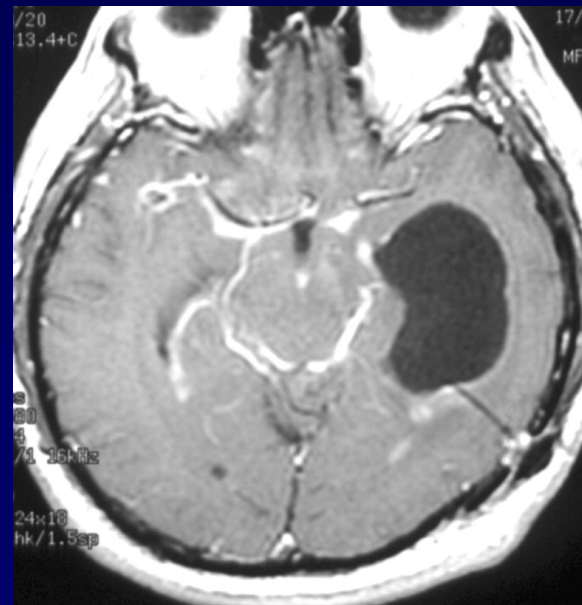
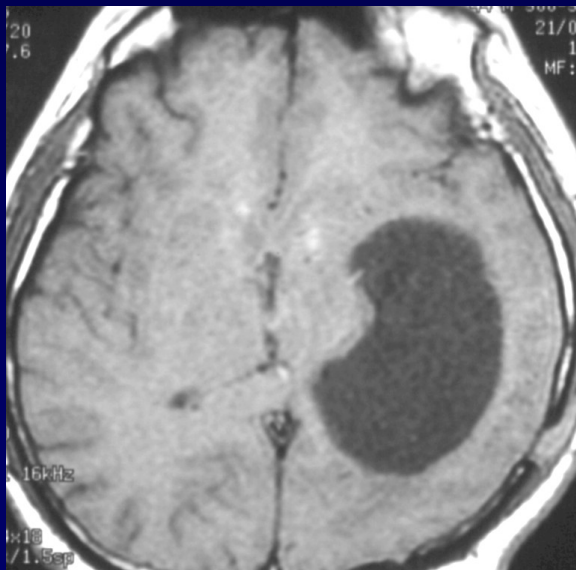
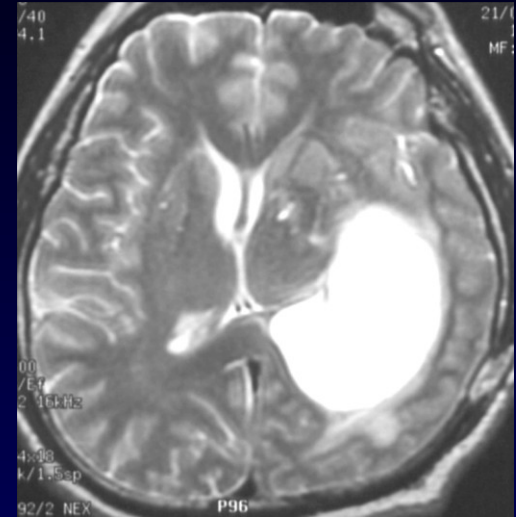
Obstructive hydrocephalus

Noncommunicating hydrocephalus, also called obstructive hydrocephalus is caused by blockage in the ventricular pathways in the brain through which cerebrospinal fluid flows. Aqueductal stenosis (most common)



Obstructive hydrocephalus

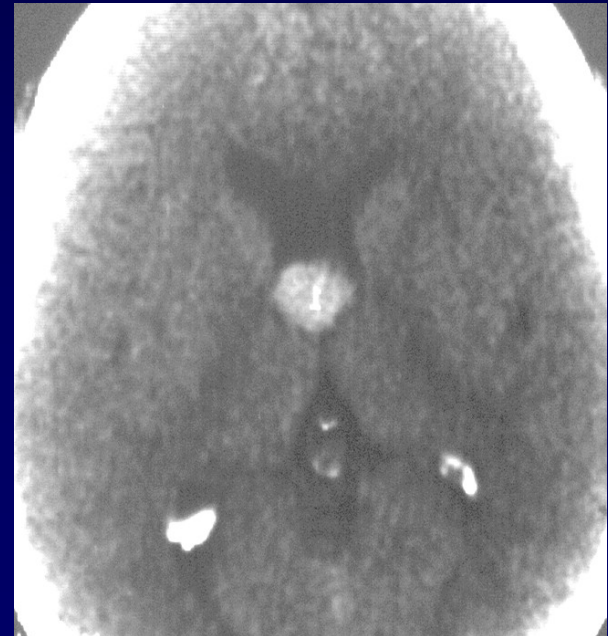
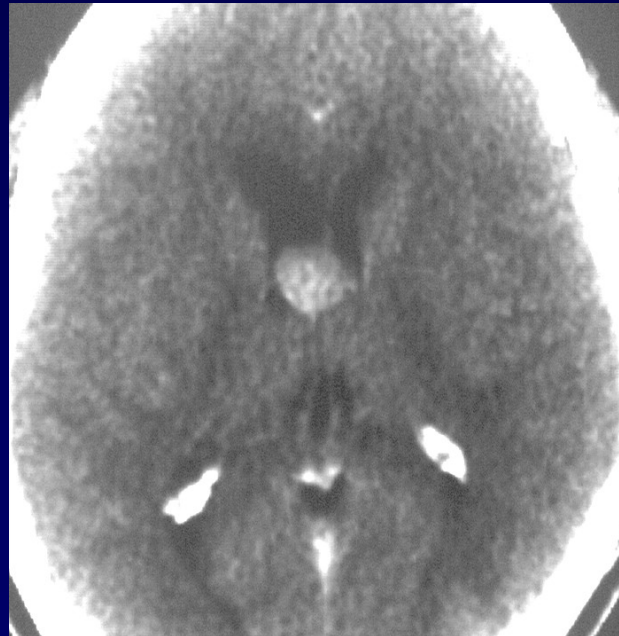
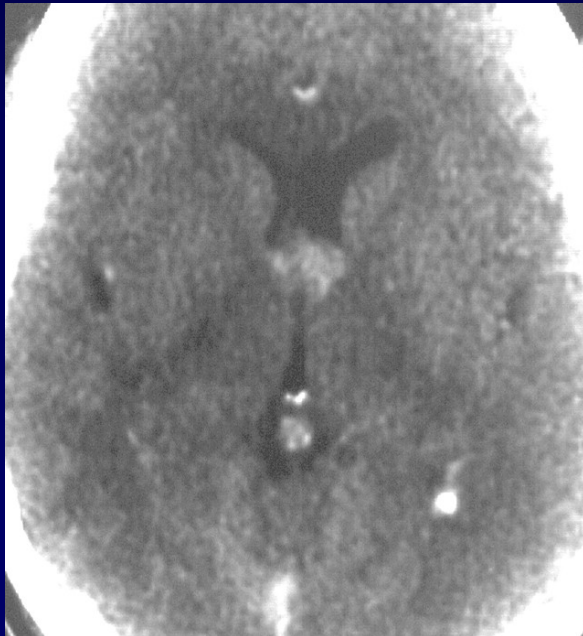
Obstructed CSF circulation
– within the ventricular system
(lateral ventricle)
postinfectious



Obstructive hydrocephalus

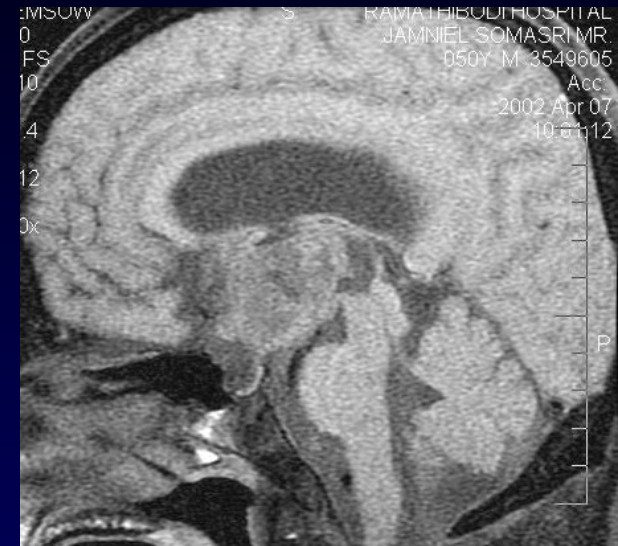
Obstructed CSF circulation

- within the ventricular system
(foramen Monroi)
colloid cyst



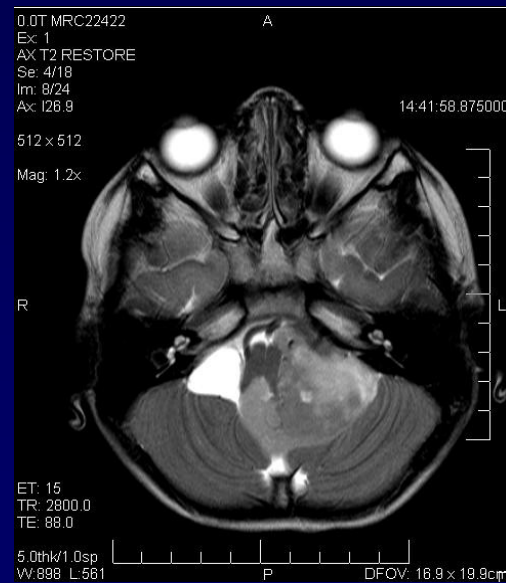
Obstructive hydrocephalus

Obstructed CSF circulation
– within the ventricular system
(3rd ventricle)
suprasellar germinoma



Obstructive hydrocephalus

Obstructed CSF circulation
– within the ventricular system
(4th ventricle)
intracerebellar hemorrhage
ependymoma



Pediatric hydrocephalus

Congenital hydrocephalus: incidence 0,5-2,5/1000 birth.

- aqueductal stenosis
- Dandy-Walker malformation
- Chiari malformation
- postinfectious
- NTD (myeloceles, meningomyeloceles)
- vein of Galen malformation
- holoprosencephaly
- neurofibromatosis
- idiopathic
- x-linked hydrocephalus (due to aqueductal stenosis)
- autosomal recessive hydrocephalus (rare)

Dandy-Walker malformation

Rare congenital malformation.

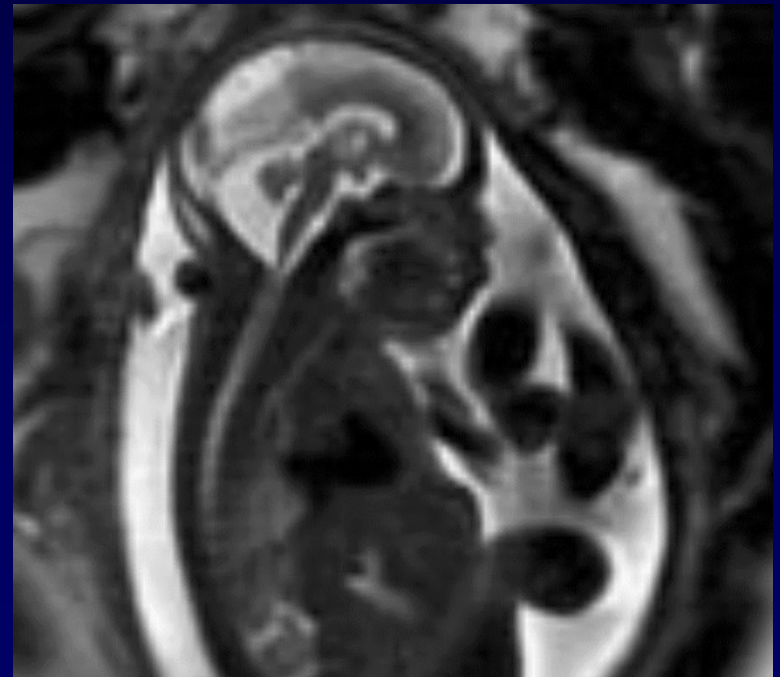
Agensis or hypoplasia of the cerebellar vermis + cystic dilatation of the fourth ventricle + enlargement of the posterior fossa.



Dandy-Walker malformation

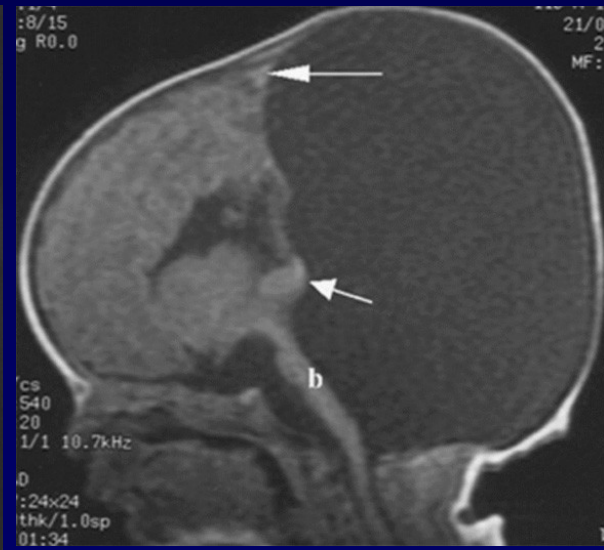
Posterior fossa cystic malformations have been divided into:

- Dandy-Walker complex
 - Dandy-Walker malformation
 - Dandy-Walker variant
 - mega cisterna magna
- Posterior fossa arachnoid cyst.



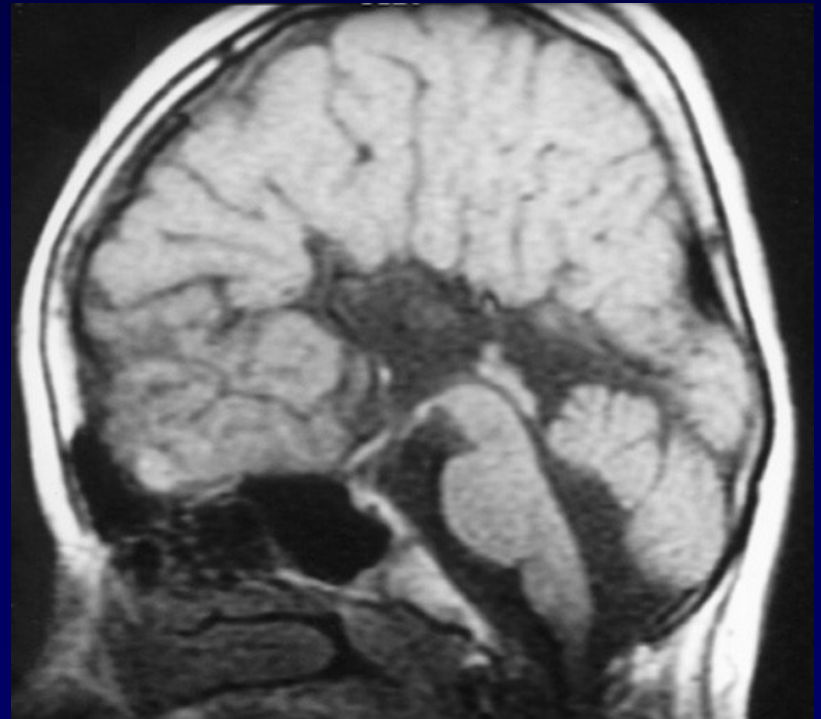
Dandy-Walker malformation

Agensis or hypoplasia of the cerebellar vermis + cystic dilatation of the fourth ventricle + enlargement of the posterior fossa.



Dandy-Walker variant

Dandy-Walker variant consists of vermian hypoplasia and cystic dilatation of the fourth ventricle, without enlargement of the posterior fossa.



Mega cisterna magna

Mega cisterna magna consists of an enlarged posterior fossa secondary to an enlarged cisterna magna, with a normal cerebellar vermis and fourth ventricle.

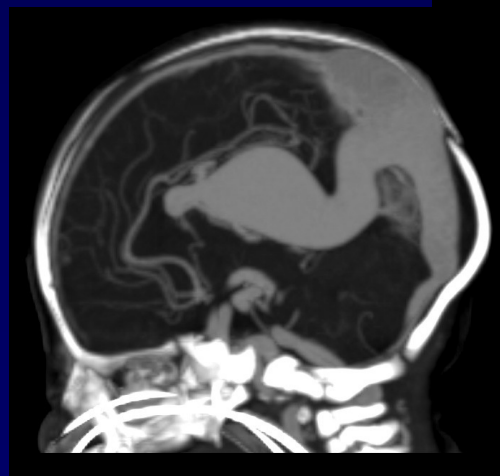
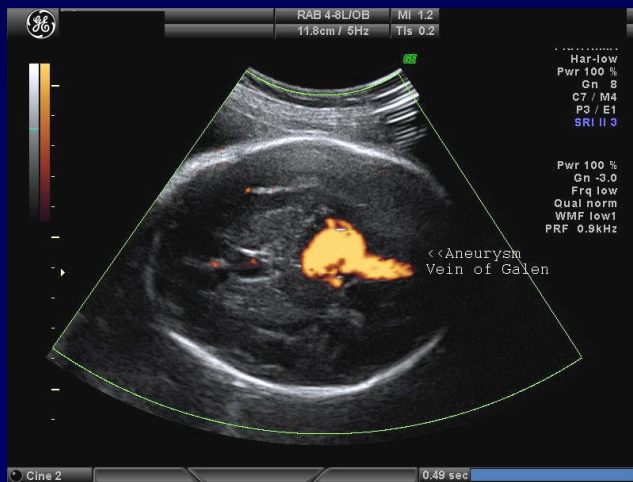
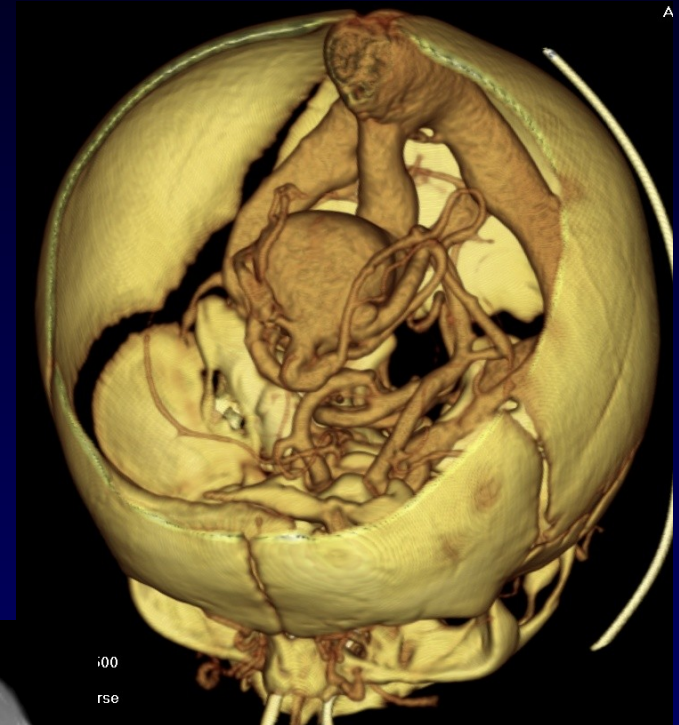
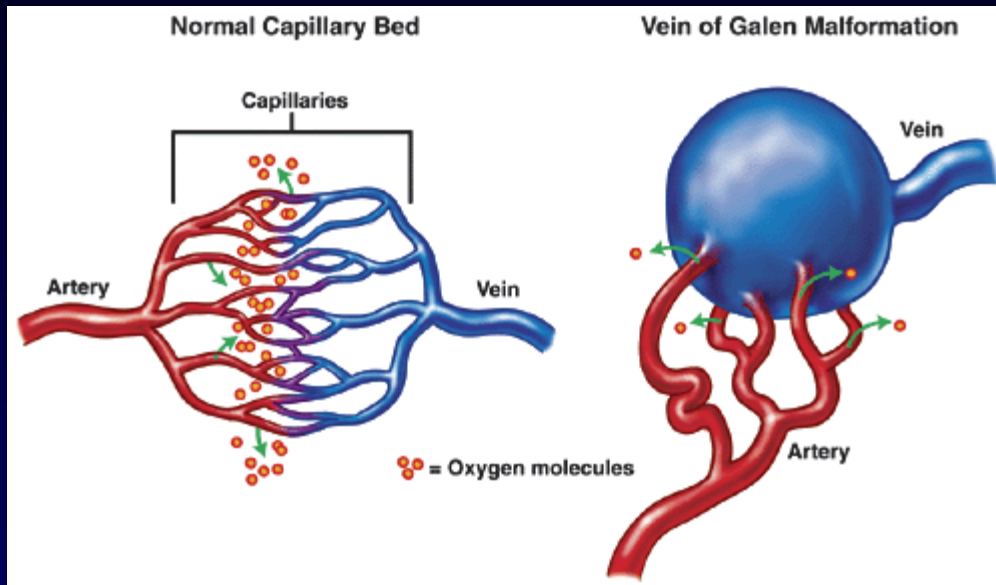


DW – arachnoid cyst

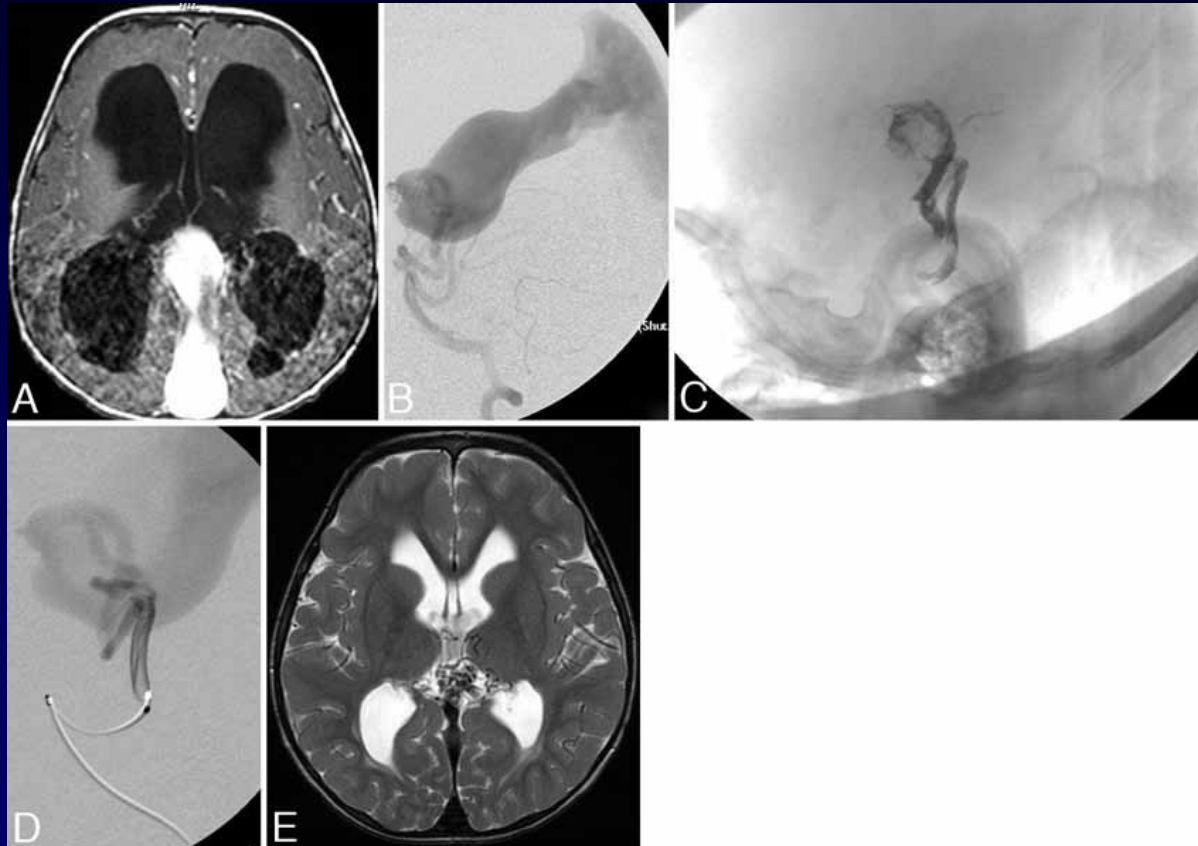
- Retrocerebellar arachnoid cysts of developmental origin are uncommon but clinically important.
- True retrocerebellar arachnoid cysts displace the fourth ventricle and cerebellum anteriorly and show significant mass effect.
- Because there are different surgical therapy approaches for posterior fossa arachnoid cyst and Dandy-Walker malformation, it is essential to differentiate between the 2 entities.



Aneurysm of the vein of Galen



Aneurysm of the vein of Galen



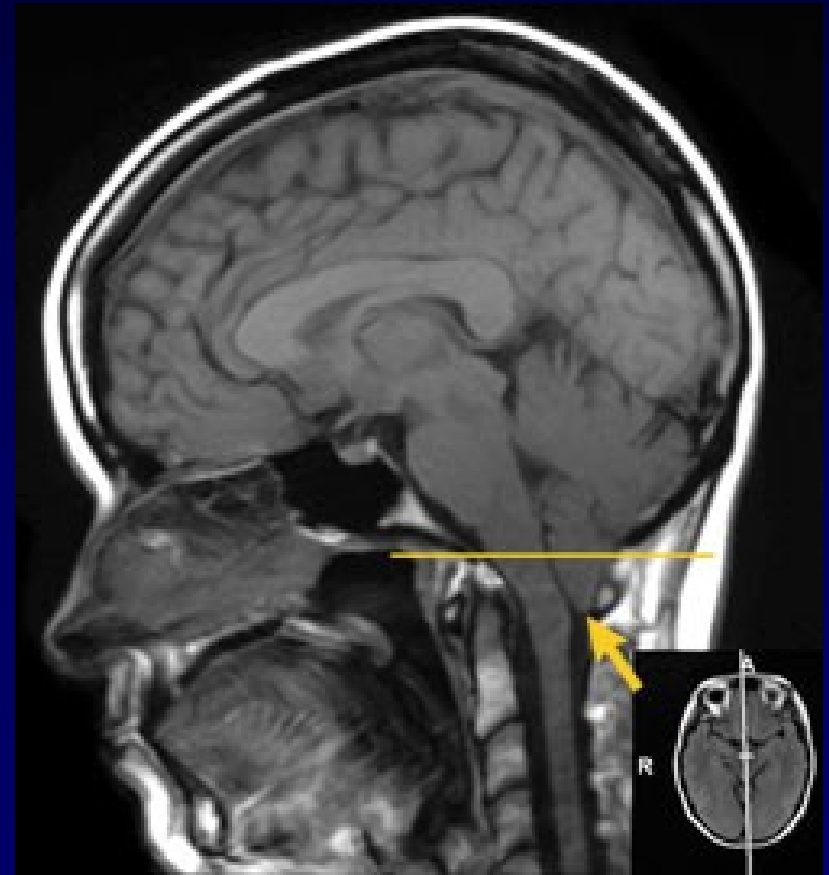
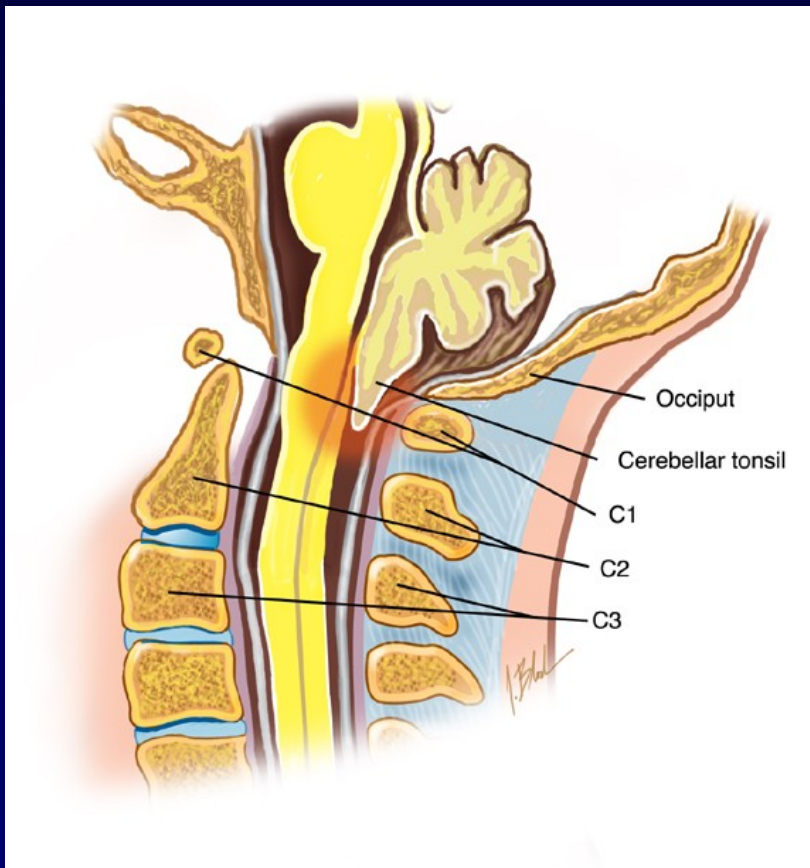
Chiari malformation

Herniation of rhombencephalonic derivatives.

Malformation	
Type	Description
Type I	Elongation of the tonsils and the medial parts of the inferior lobes of the cerebellum into cone-shaped projections, which accompany the medulla oblongata into the spinal canal
Type II	Displacement of the parts of the inferior vermis, pons, and medulla oblongata together with elongation of the fourth ventricle (most cases are associated with spina bifida)
Type III	The entire cerebellum herniates into the cervical canal
Type IV	Cerebellar hypoplasia

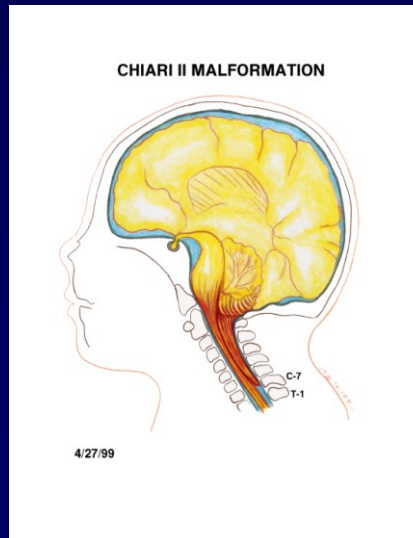
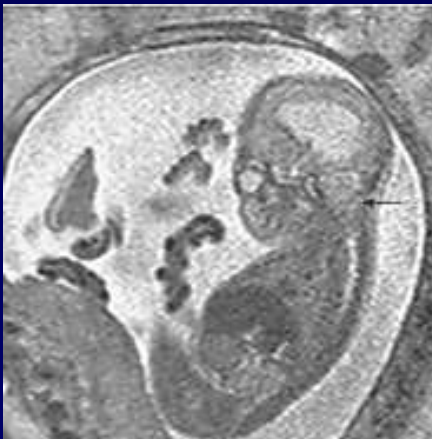
Chiari I

Caudal migration of the cerebellar tonsils through the foramen magnum, usually greater than 5 mm. Manifestation in adults. Syringomyelia (A=yes, B=no).



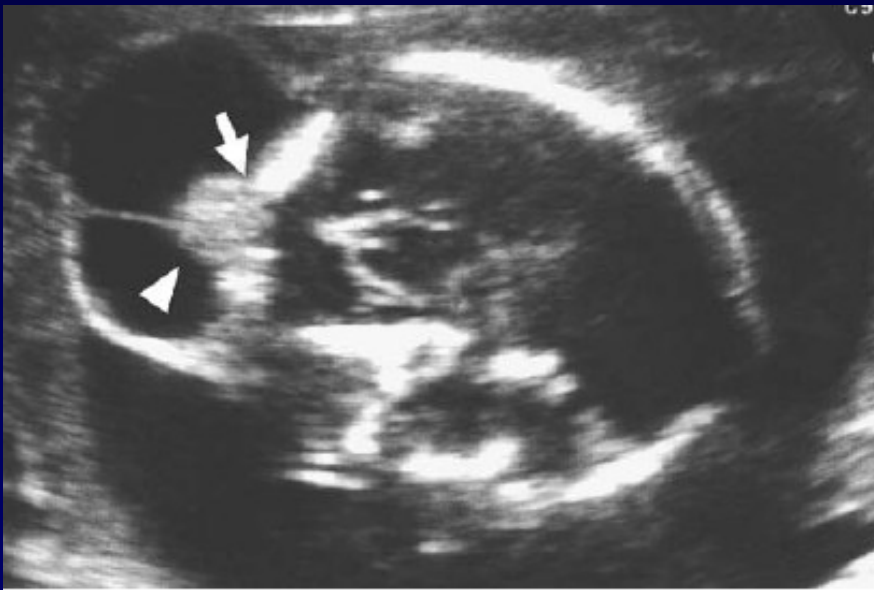
Chiari II

- Chiari type II malformation is less common and more severe.
- Symptomatic in infancy or early childhood.
- Almost invariably associated with myelomeningocele and hydrocephalus.
- Its hallmark is caudal displacement of lower brainstem (medulla, pons, 4th ventricle) through the foramen magnum.
- Symptoms arise from dysfunction of brainstem and lower cranial nerves.



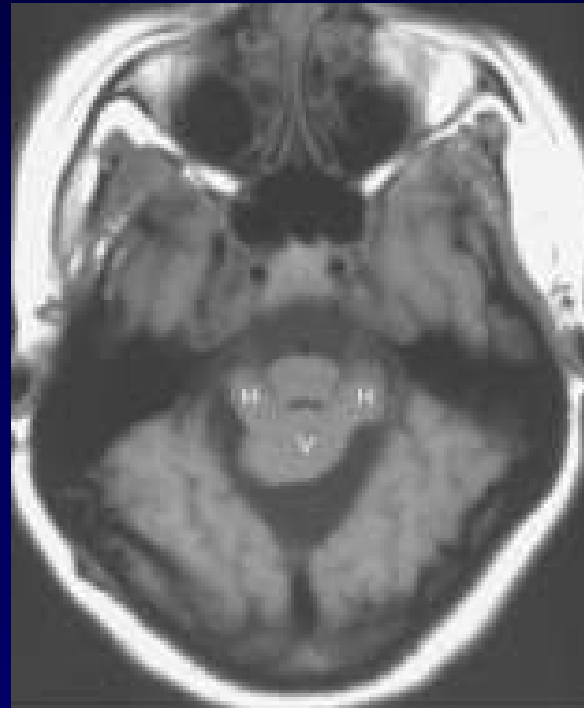
Chiari III

Type III malformation refers to herniation of cerebellum into a high cervical myelomeningocele. Exceedingly rare and incompatible with life.



Chiari IV

Cerebellar hypoplasia



Meningomyelocele (NTD)

- Protrusion of the membranes that cover the spine but some of the spinal cord itself through a defect in the bony encasement of the vertebral column.
- Infants with MMC are at risk for bacterial meningitis due to the spinal defect.
- Leak of cerebrospinal fluid (CSF) leak is commonly observed.
- The major indication for early operative repair (within 48h of delivery) is prevention of infection.
- Hydrophalus in 60%-90%.



Posthemorrhagic HCP in preterm infants

Occasionally occurs antenatally

1st day 50%, 2nd day 25%,
3rd day 15%, > 4th day 10%.

Germinal matrix – subependymal,
source of neural precursors, capillars –
vascular end zone of arterial supply.

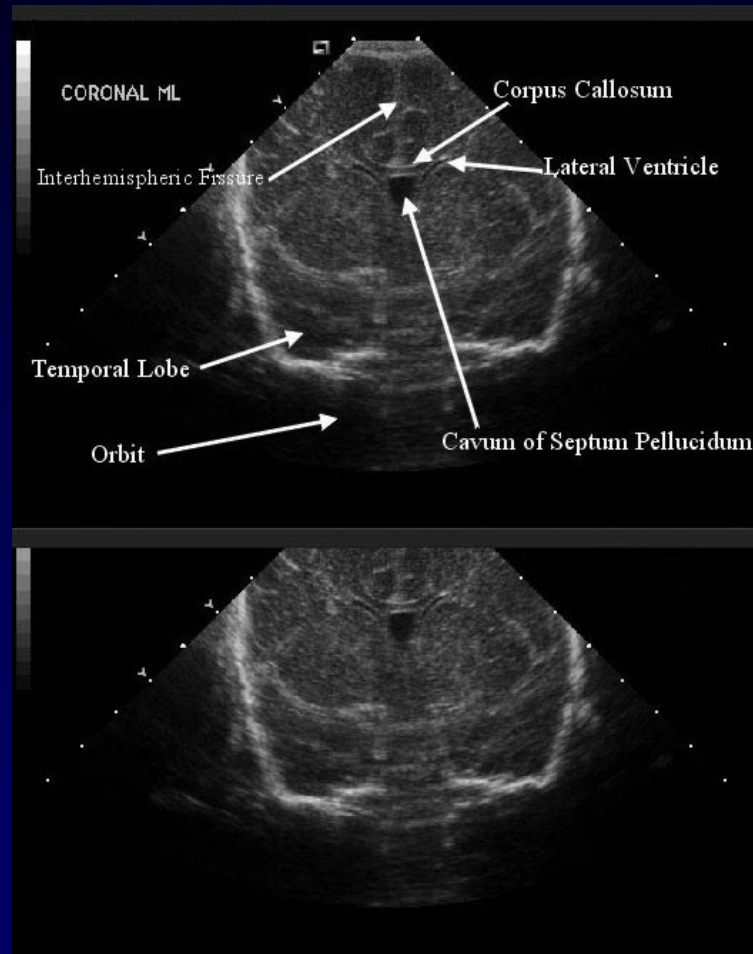
Hematoma – rupture of fragile
capillaries.

Posthemorrhagic HCP in preterm infants

Papile classification:

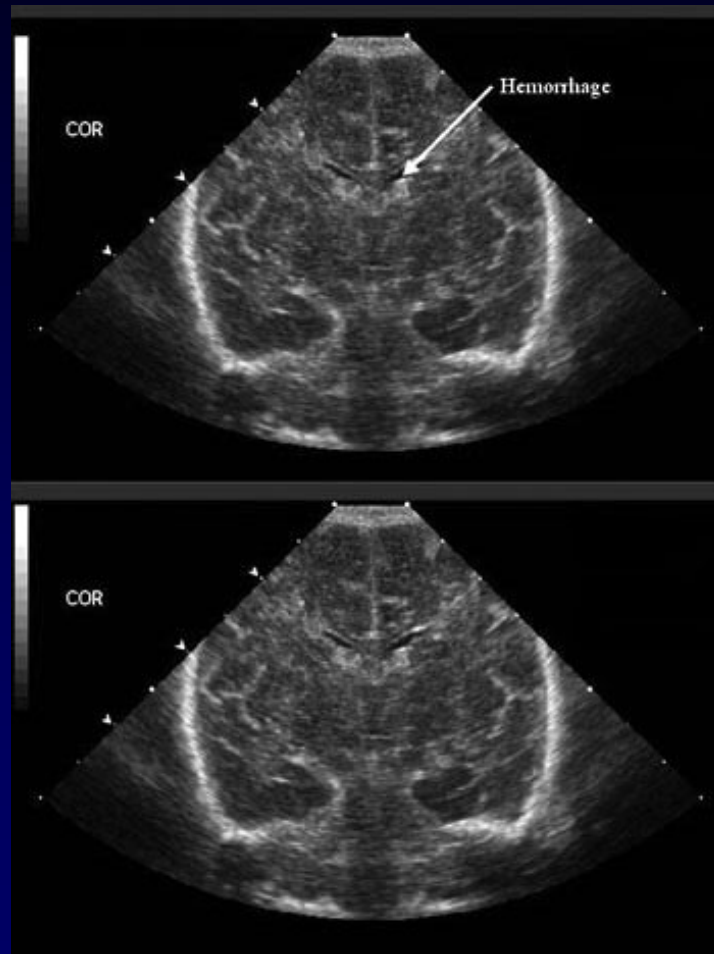
- Grade I – subependymal hemorrhage**
- Grade II – intraventricular hemorrhage without ventricular dilatation**
- grade III – intraventricular hemorrhage with ventricular dilatation**
- grade IV – intraventricular hemorrhage with parenchymal hemorrhage**

Posthemorrhagic HCP in preterm infants



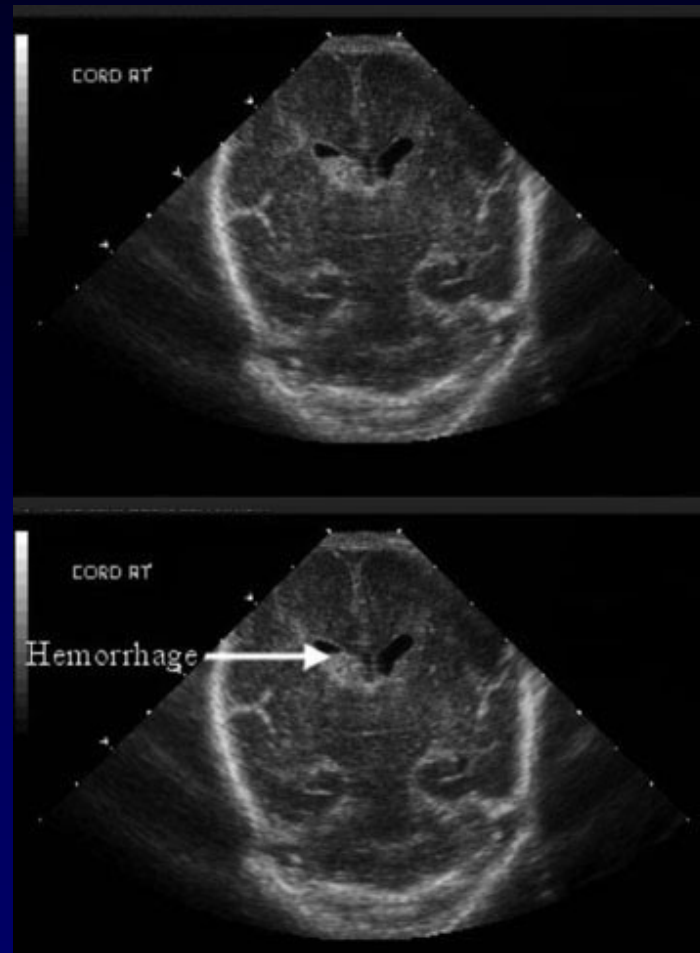
No hemorrhage

Posthemorrhagic HCP in preterm infants



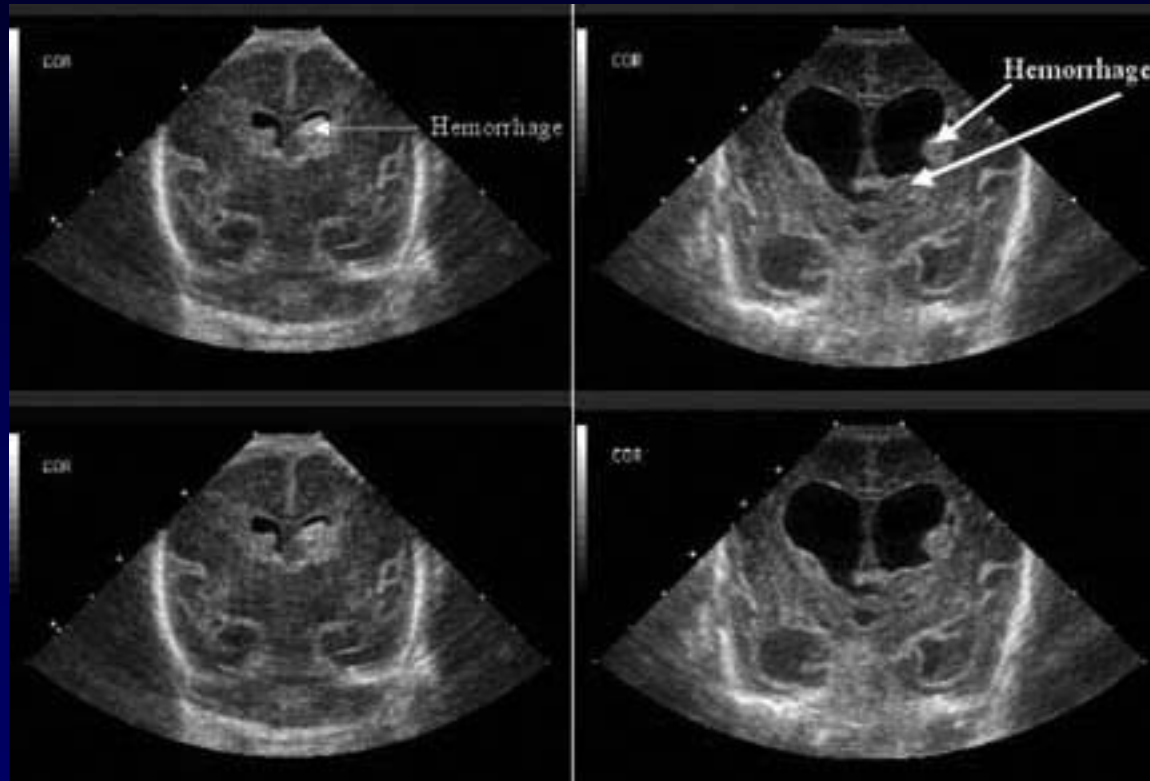
Grade I

Posthemorrhagic HCP in preterm infants



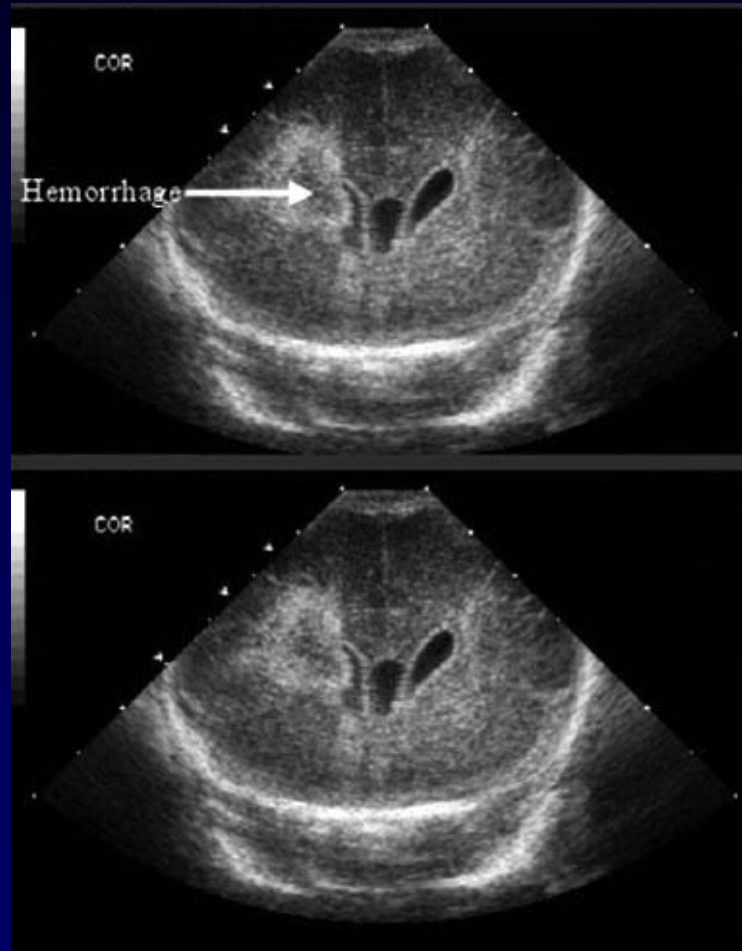
Grade II

Posthemorrhagic HCP in preterm infants



Grade III

Posthemorrhagic HCP in preterm infants



Grade IV

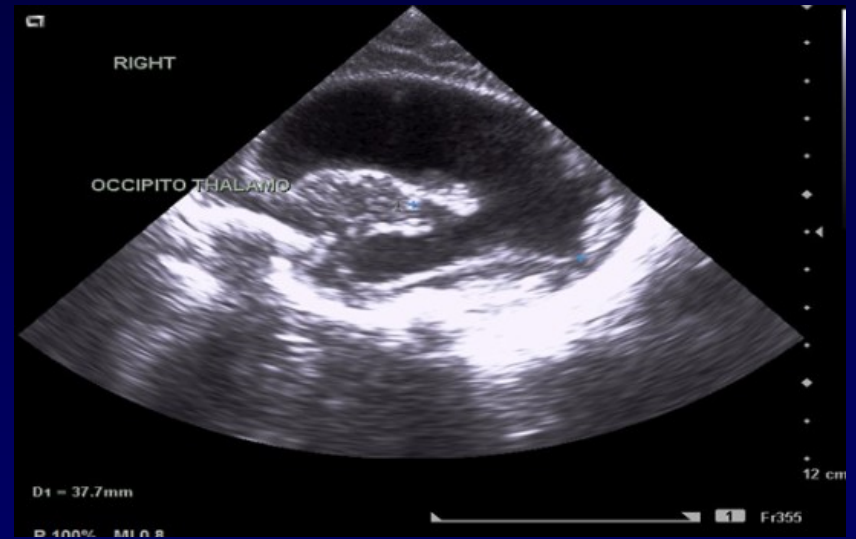
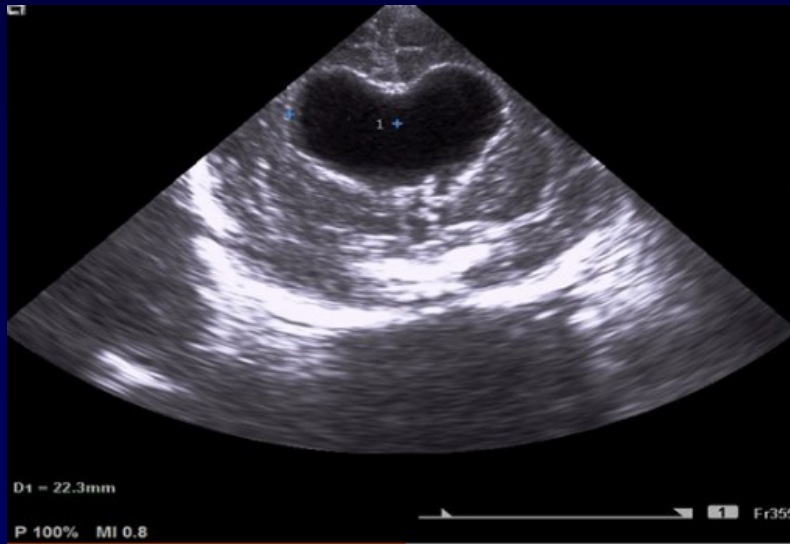
Posthemorrhagic HCP in preterm infants

Neuropathological consequences:

- Germinal matrix destruction**
- Periventricular hemorrhagic infarct 15%**
- Posthemorrhagic hydrocephalus 30%**

Posthemorrhagic HCP in preterm infants

Posthemorrhagic hydrocephalus

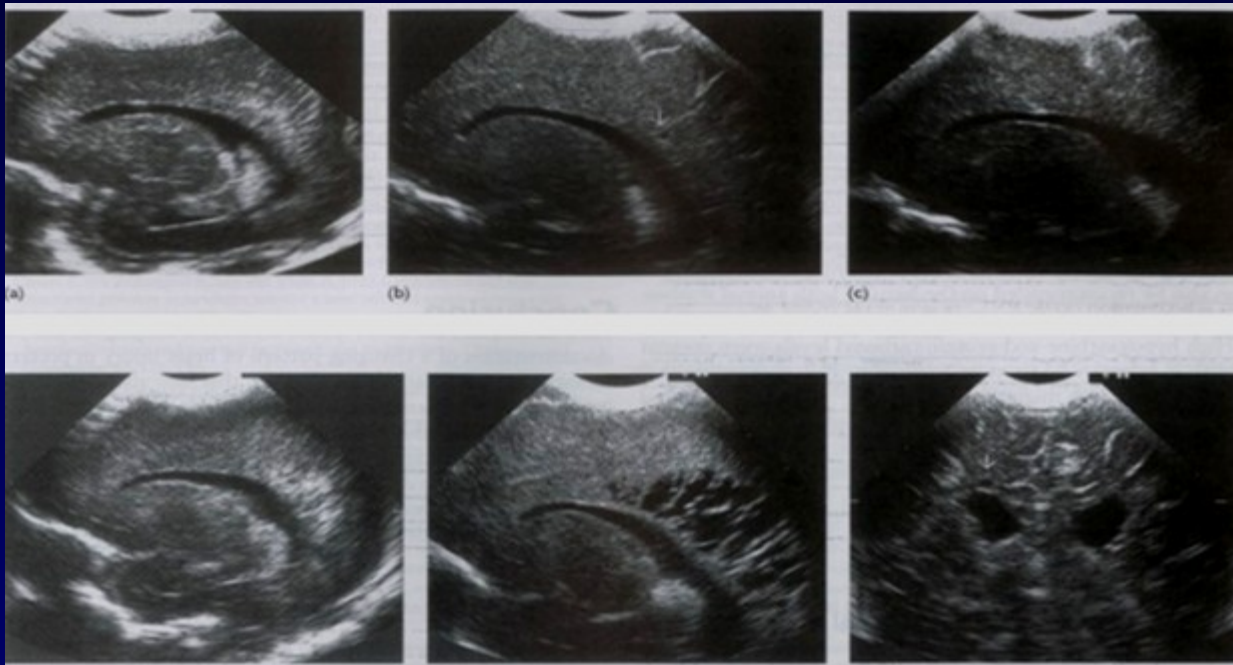


Posthemorrhagic HCP in preterm infants

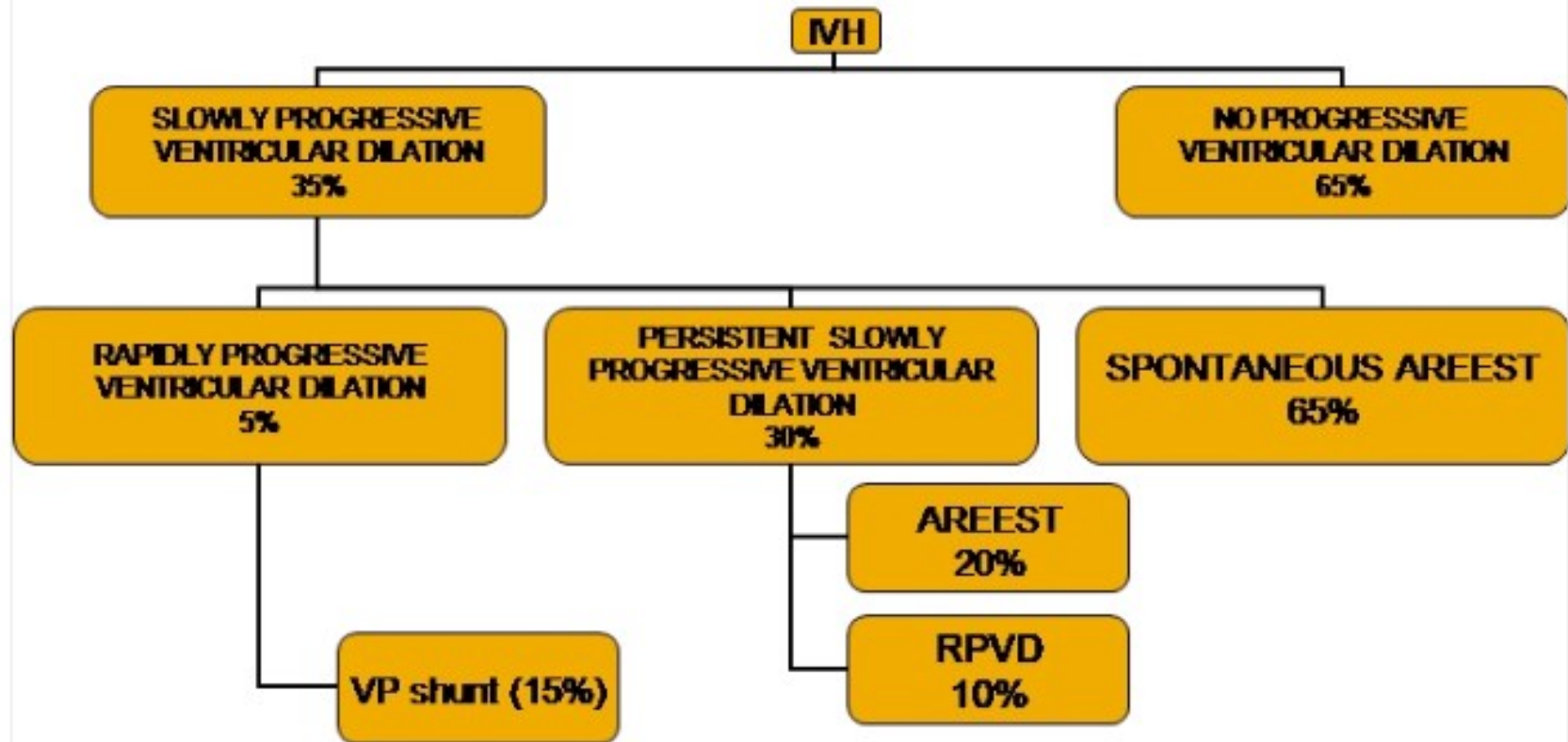
- **Neuropathological leukomalacia – bilateral, non-hemorrhagic ischemic white matter injury.**
- **Pontine neuronal necrosis – 46% to 71% of infants with IVH exhibited pontine neuronal necrosis.**

Posthemorrhagic HCP in preterm infants

Periventricular leukomalacia

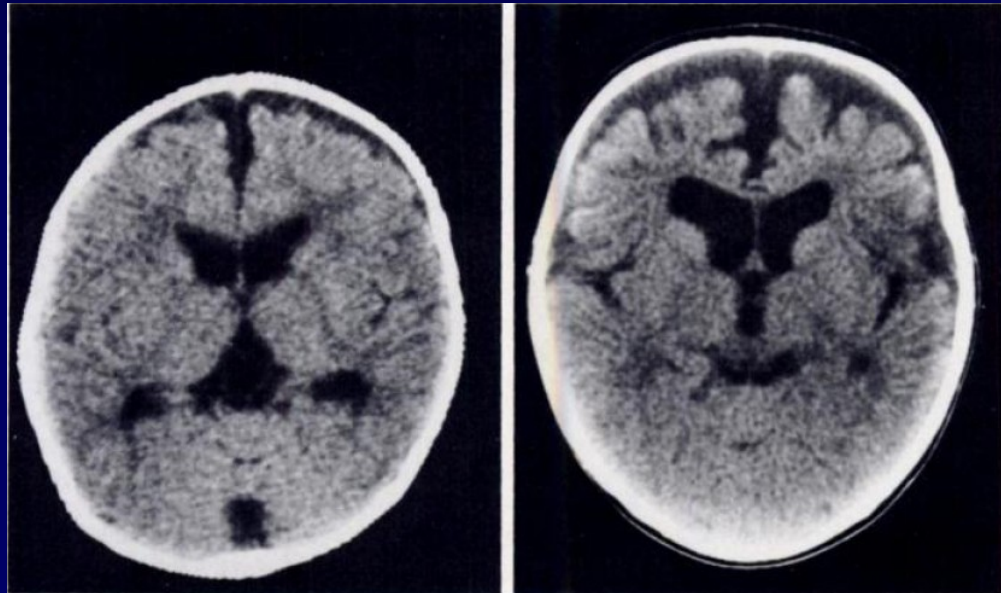


Posthemorrhagic HCP in preterm infants



Benign external hydrocephalus

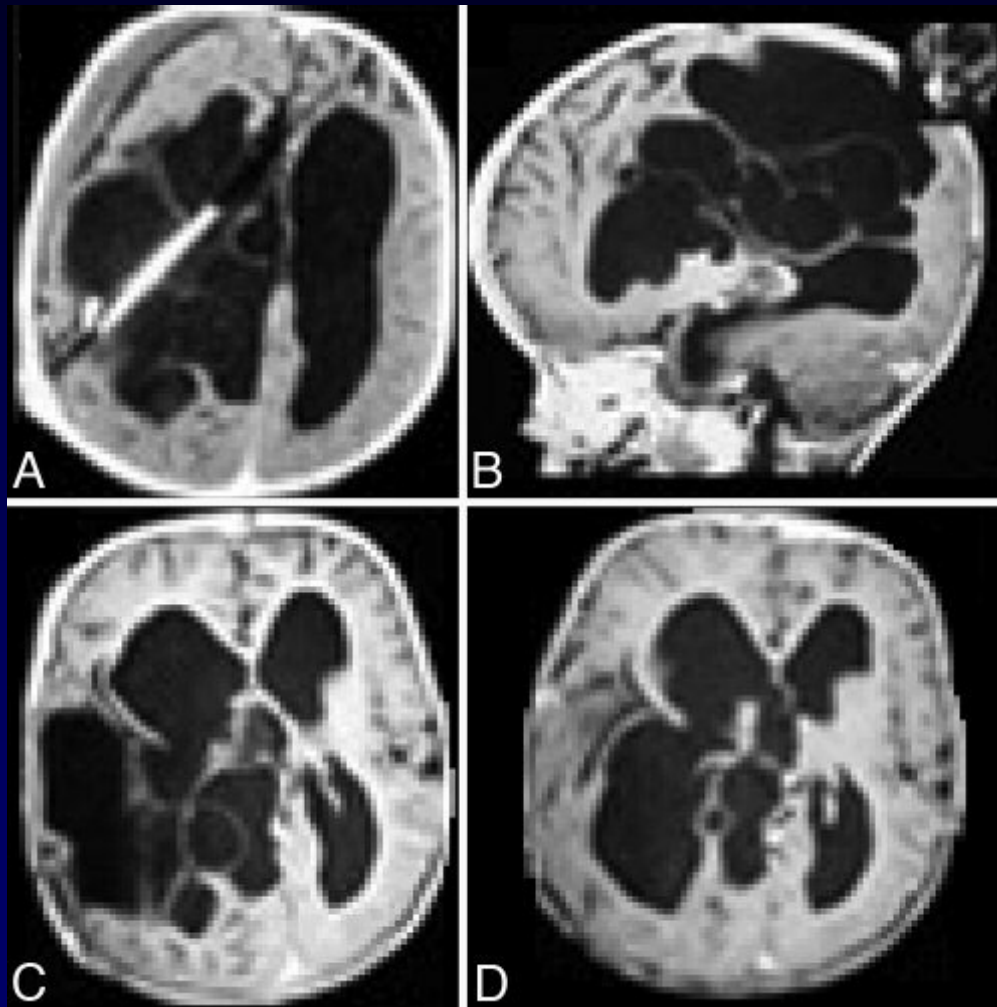
- Infants with rapidly enlarging heads.
- CT scan – widening of the subarachnoid space with mild or no ventricular dilatation
- Age-related self-limited condition occurring in infants with open cranial sutures.
- Usually resolves without intervention by 2-3 years of age.



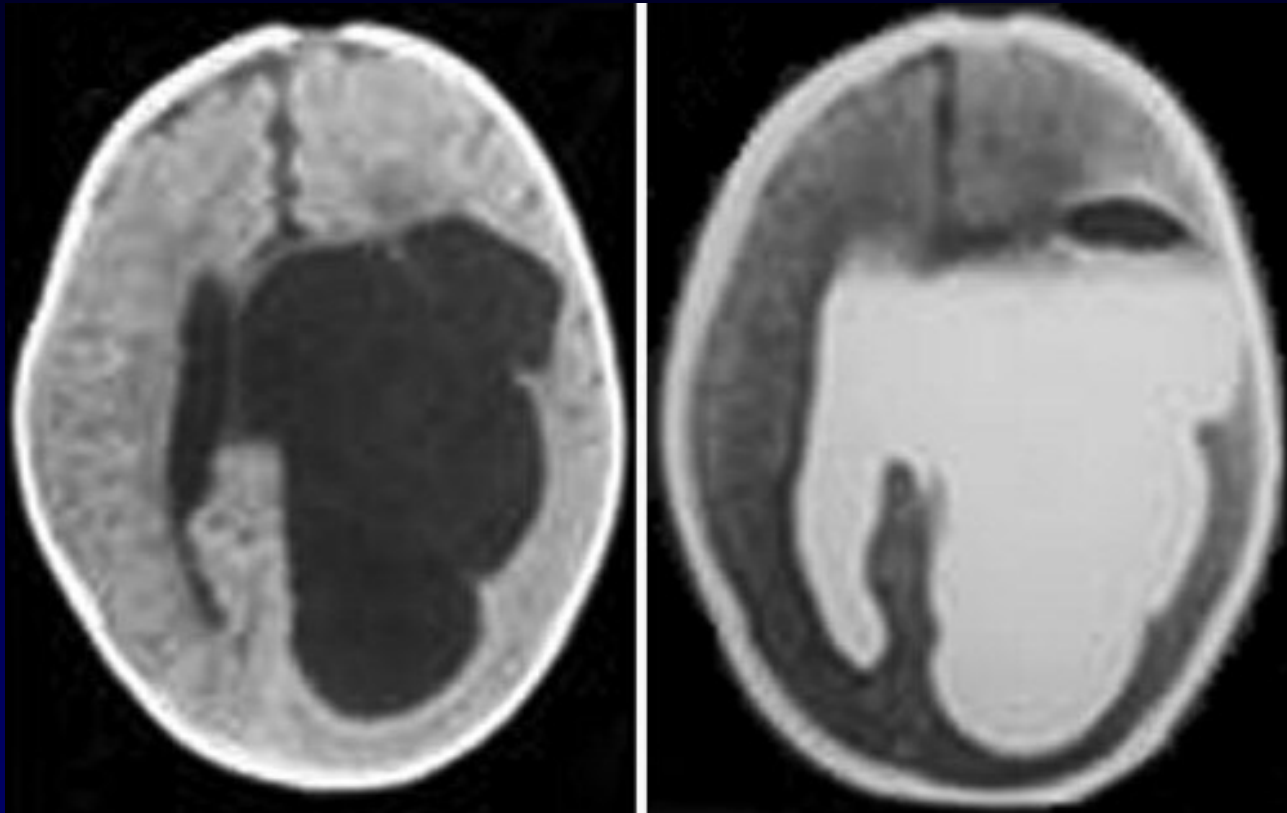
Multilocular hydrocephalus

- Hydrocephalus arising from intraventricular septations.
- Presence of multiple cysts inside the ventricles.
- Complication of neonatal hydrocephalus.
- Causes: preterm infant with intracranial hemorrhage grade II-III or central nervous system infection.
- Requires a specific therapeutic approach.

Multilocular hydrocephalus



Multilocular hydrocephalus



Postoperative exam – all parts communicate

Adult hydrocephalus

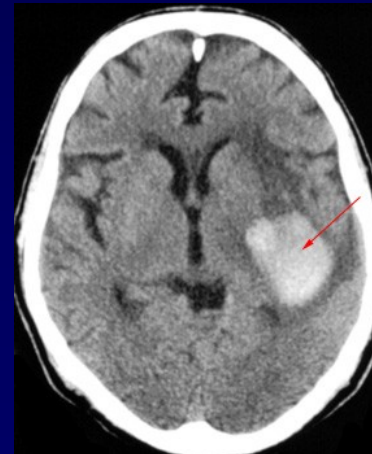
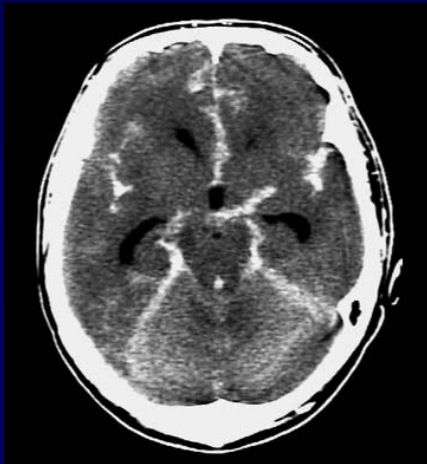
Hydrocephalus which occurs in an adult patients.

- aquired, congenital or idiopathic
- posthemorrhagic (SAH)
- aqueductal stenosis
- trauma
- tumor
- postinfectious
- special forms
 - normal pressure hydrocephalus
 - others

Posthemorrhagic hydrocephalus

Adults

- Subarachnoid hemorrhage (SAH)
- Intraventricular hemorrhage (IVH)
- Intraparenchymal hemorrhage (IPH)



Posthemorrhagic hydrocephalus

- After SAH – 2.3 to 63.4%
- STICH – 42% with IPH had IVH
 - 55% of IVH hydrocephalus
 - HCP in 23,1 % of all IPH
- Hemorrhage-related hydrocephalus is transient and patients lose their shunt dependency

LOWER INCIDENCE OF REOPERATION WITH LONGER SHUNT SURVIVAL WITH ADULT VENTRICULOPERITONEAL SHUNTS PLACED FOR HEMORRHAGE-RELATED HYDROCEPHALUS

Brian L. Hoh, M.D.

Department of Neurological Surgery,
University of Florida College of Medicine,
Gainesville, Florida

Shih-Shan Lang, M.D.

Department of Neurological Surgery,
University of Pennsylvania
School of Medicine,
Philadelphia, Pennsylvania

Michael V. Ortiz, B.S.

University of Florida College of Medicine,
Gainesville, Florida

Yueh-Yun Chi, Ph.D.

University of Florida College of Medicine,
Gainesville, Florida

Stephen B. Lewis, M.D.

Department of Neurological Surgery,
University of Florida College of Medicine,
Gainesville, Florida

David W. Pincus, M.D., Ph.D.

Department of Neurological Surgery,
University of Florida College of Medicine,
Gainesville, Florida

OBJECTIVE: The incidence of reoperation for ventriculoperitoneal shunts (VPS) in adults, although lower than in pediatric patients, is not insignificant. We hypothesize that adult VPS placed for hemorrhage-related hydrocephalus have a lower incidence of reoperation than those placed for other types of hydrocephalus.

METHODS: We retrospectively reviewed all adult (≥ 20 yr) VPS initially placed from February 2001 to August 2006 at the University of Florida. We determined the incidence and time interval to reoperation. Follow-up was conducted by telephone interview and review of medical records.

RESULTS: A total of 286 adult VPS were initially placed: 96 (34%) hemorrhage and 190 (66%) nonhemorrhage. A total of 15 (16%) hemorrhage patients underwent 22 shunt reoperations, compared with 50 (27%) nonhemorrhage patients who underwent 82 shunt reoperations ($P = 0.0316$). A Poisson regression analysis of the number of reoperations, factoring hemorrhage, age, and sex, demonstrated a significantly lower incidence of reoperation in hemorrhage patients ($P = 0.0900$). A Cox proportional hazards model analysis of time to first reoperation, factoring hemorrhage, age, and sex, demonstrated a significantly longer shunt survival in hemorrhage patients ($P = 0.0404$).

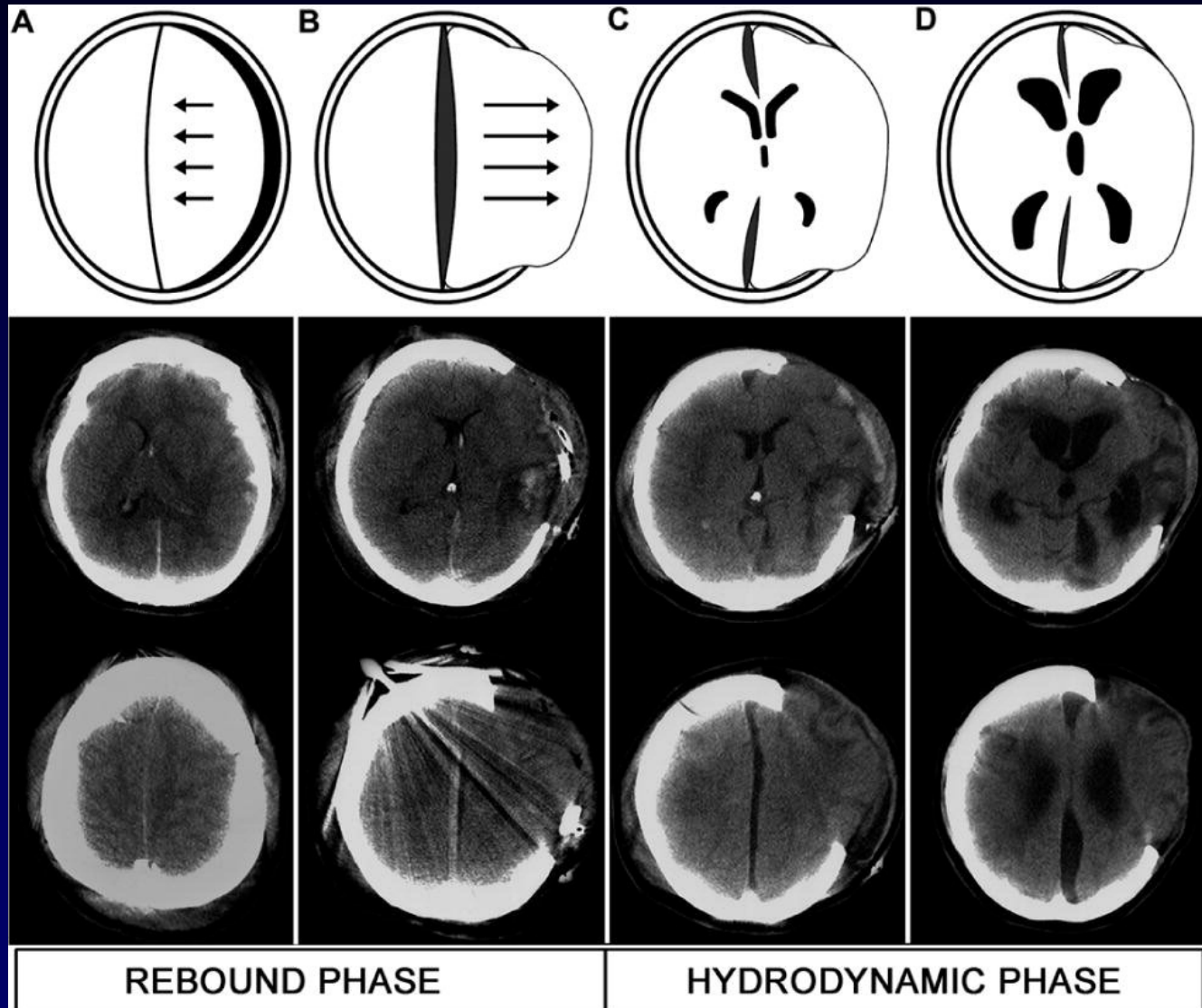
CONCLUSION: Adult VPS placed for hemorrhage-related hydrocephalus have a significantly lower incidence of reoperation and significantly longer shunt survival. This result may be related to an incidence of transient shunt dependency in patients with hemorrhage-related hydrocephalus. However, the precise mechanism remains unclear.

KEY WORDS: Adult, Hemorrhage, Hydrocephalus, Malfunction, Reoperation, Ventriculoperitoneal shunt

Posttraumatic hydrocephalus

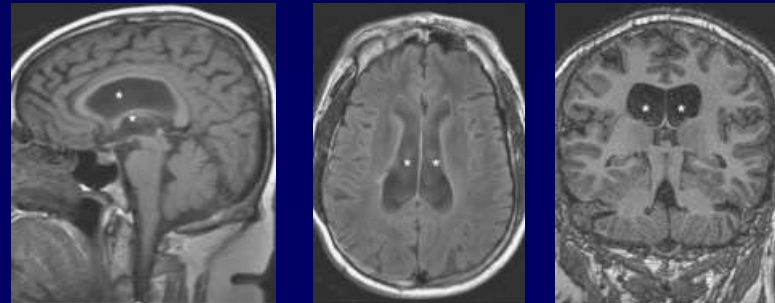
- **Posttraumatic hydrocephalus is an active and progressive process of excessive cerebrospinal fluid (CSF) accumulation due to liquorodynamic disturbances following craniocerebral injury.**
- **The incidence of PTH ranges from 0.7-86% (differences in diagnostic criteria and classification).**
- **Risk of developing PTH: coma, increased age, decompressive craniectomy and subarachnoid hemorrhage.**
- **Cave – brain atrophy (hydrocephalus ex-vacuo).**

Posttraumatic hydrocephalus



Adult hydrocephalus - SHYMA

- **SHYMA - the syndrome of hydrocephalus in young and middle-aged adults.**
- **Acquired communicating adult-onset hydrocephalus with mild ventricular enlargement and otherwise normal cerebral cortex and white matter.**
- **Symptoms: headache, subtle gait disturbance, urinary frequency, visual disturbances and some level of impaired cognitive skills that can noticeably affect job performance and personal relationships.**
- **Discrepancy between the prominence of symptoms and the subtlety of clinical signs.**
- **The degree of symptoms and their resultant effect varies widely among patients.**
- **Often remain undiagnosed and untreated.**



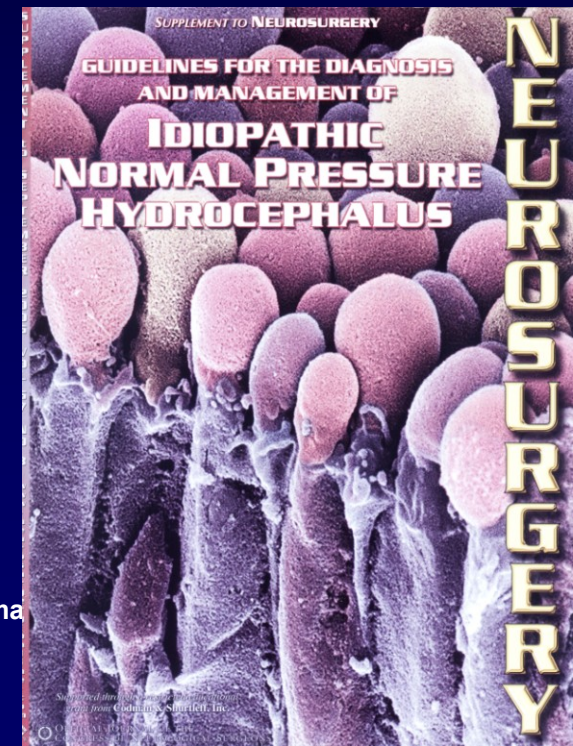
Normal pressure hydrocephalus

- Hakim S., Adams R.D. 1964, 1965*
- Normal pressure hydrocephalus - Adam's trias (gait and/or balance impairments, disturbances in cognition a control of urination) with ventricular enlargement and normal ICP (60-240 mm H₂O).
- **Idiopathic (primary) – unknown cause (INPH)**
- **Secondary (SNPH) – known cause (trauma, subarachnoid hemorrhage, stroke etc.)**

* Adams RD, Fisher CM, Hakim S, Ojemann RG, Sweet WH: Symptomatic occult hydrocephalus with “normal” cerebrospinal-fluid pressure: A treatable syndrome. N Engl J Med 273:117–126, 1965.

*Hakim S: Some observations on CSF pressure: Hydrocephalic syndrome in adults with “normal” CSF pressure. Bogata, Javeriana University School of Medicine, 1964 (Thesis No. 957).

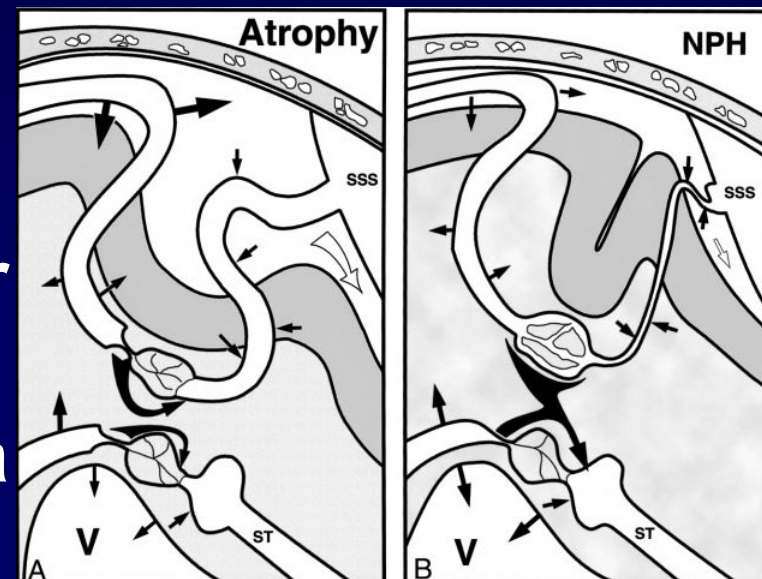
*Hakim S, Adams RD: The special clinical problem of symptomatic hydrocephalus with normal cerebrospinal fluid pressure observations on cerebrospinal fluid hydrodynamics. J Neurol Sci 2:307–327, 1965.



Normal pressure hydrocephalus

Pathophysiology: aberrations in CSF flow dynamics

- transmantle pressure gradient
- a water-hammer effect that enlarges the ventricles
- abnormalities of ventricular wall compliance may contribute to ventricular dilatation
- periventricular ischemia

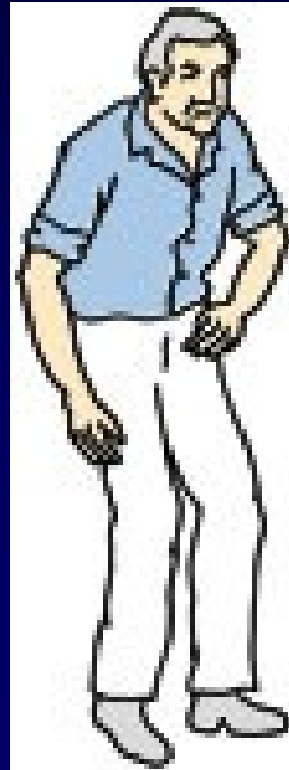


Normal pressure hydrocephalus

Patient and family member

Adam's trias

- Gait disturbances
- Cognitive impairment
- Urinary inkontinence



Normal pressure hydrocephalus

- „10 m walk test“
 - standard – time < 9 sekund, number of steps < 18
- **Videoanalysis**
 - Decreased step height
 - Decreased step length
 - Decreased cadence (speed of walking)
 - Increased trunk sway during walking
 - Widened standing base
 - Toes turned outward on walking
 - Retropulsion (spontaneous or provoked)
 - En bloc turning (turning requiring three or more steps for 180 degrees)
 - Impaired walking balance, as evidenced by two or more corrections out of eight steps on tandem gait testing
- **EMG/MEP:**
 - Hyperactivity of antagonists and antigravity muscles
 - No pyramidal tract involvement – extrapyramidal subcortical dysfunction
 - Pyramidal tracts signs : clinically unfavourable signs

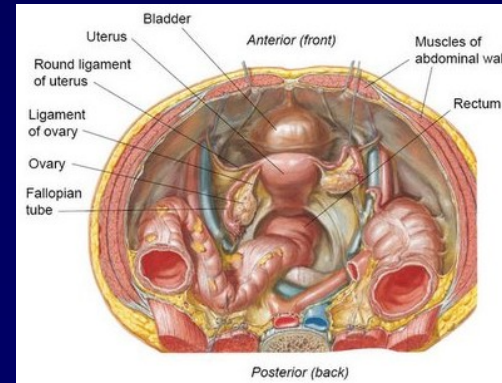
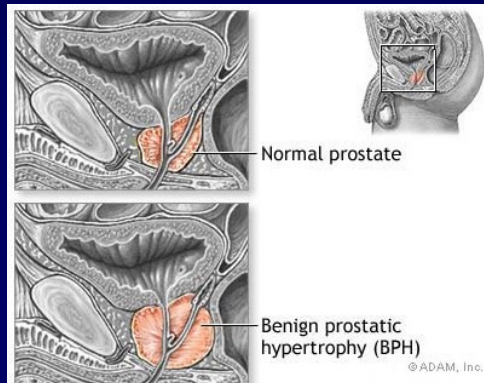
Normal pressure hydrocephalus

Impairment of cognition

- psychomotor slowing /increased response latency/
- decreased fine motor speed
- decreased fine motor accuracy
- difficulty diving or maintaining attention
- impaired recall, especially for recent events
- executive dysfunction
- behavioral or personality changes

Normal pressure hydrocephalus

- **Urinary incontinence**
 - **Urological examination**
 - Episodic or persistent urinary incontinence not attributable to primary urological disorders; persistent urinary incontinence; urinary and fecal incontinence
 - Urinary urgency as defined by frequent perception of a pressing need to void; urinary frequency as defined by more than six voiding episodes in an average 12-hour period despite normal fluid intake; nocturia as defined by the need to urinate more than two times in an average night



Normal pressure hydrocephalus

Imaging studies

- **ventricular enlargement (Evan's index $> 0,3$)**
- **no macroscopic obstruction to CSF flow**
- **at least one of the following supportive features**
 - **enlargement of the temporal horns of the lateral ventricles**
 - **callosal angle of 40 degrees or more**
 - **periventricular signal changes (no ischemic changes or demyelination)**
 - **An aqueductal or fourth ventricular flow void on MRI**
- **Other brain imaging findings**
 - **A brain imaging study performed before onset of symptoms showing smaller ventricular size or without evidence of HCP**
 - **Cine MRI study or other technique showing increased ventricular flow rate**
 - **A SPECT-acetazolamide challenge showing decreased periventricular perfusion that is not altered by acetazolamide**

Normal pressure hydrocephalus

Differential diagnosis

– Neurodegenerative disorders

- Alzheimer's disease
- Parkinson's disease
- Huntingtonova choroba
- Lewy body disease
- Frontotemporal demetia
- Corticobasal degeneration
- Progressive supranuclear palsy
- Multisystem atrophy
- Spongiform encephalopathy

– Vascular dementia

- Cerebrovascular disease
- Stroke
- Multi-infarct state
- Binswanger's disease
- Cerebral autosomal dominant arteriopathy, subcorical infarcts, and leukoencephalopathy
- Vertebrobasilar insufficiency

– Infectious disease

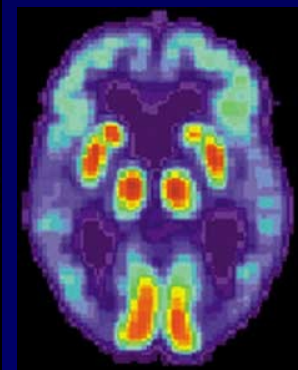
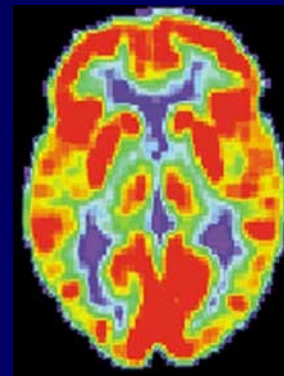
- Lyme
- HIV
- syphilis

– Urological disorders

- Urinary tract infection
- Bladder or prostate cancer
- Benign prostatic enlargement

– Miscellaneous

- B12 deficiency
- Collagen vascular disorders
- Epilepsy
- Depression
- Traumatic brain injuries
- Spinal stenosis
- Chiari malformation
- Wernicke's encephalopathy
- Carcinomatous meningitis
- Spinal cord tumor



Normal pressure hydrocephalus

- Supplemental prognostic tests
- „shunt-responsive“ versus „shunt-nonresponsive“
- „tap test“ (withdraw 40 to 50 ml, lesser volumes (25 ml or less) have low sensitivity, good sensitivity, low specificity)
- CSF outflow resistance via an infusion test (PPV of 75% to 92%)
- External lumbar drainage (Haan, Thomeer 1988, 10 ml per hour for 3 days) (PPV of 80 to 100%)

TABLE 3.2. Predictive value of cerebrospinal fluid tap test for shunt-responsive idiopathic normal-pressure hydrocephalus^a

Series (ref. no.)	INPH/SNPH (n)	Sensitivity	Specificity	PPV	NPV	Accuracy
Malm et al., 1995 (15)	35/0	62% (16/26)	33% (3/9)	73% (16/22)	23% (3/13)	54% (19/35)
Walchenbach et al., 2002 (23)	43/6	26% (9/35)	100% (12/12)	100% (9/9)	32% (12/38)	45% (21/47)
Haan and Thomeer, 1988 (10)	32/0	43% (9/21)	100% (5/5)	100% (9/9)	42% (5/17)	54% (14/26)

^a INPH, idiopathic normal-pressure hydrocephalus; SNPH, secondary NPH; PPV, positive predictive value; NPV, negative predictive value.

TABLE 3.5. Predictive value of cerebrospinal fluid outflow resistance in shunt-responsive idiopathic normal-pressure hydrocephalus^a

Series (ref. no.)	INPH/SNPH (n)	Ro threshold	Sensitivity	Specificity	PPV	NPV	Accuracy
Malm et al., 1995 (15)	35/0		58% (15/26)	44% (4/9)	75% (15/20)	27% (4/15)	54% (19/35)
Takekuchi et al., 2000 (20)	25/0		100% (12/12)	92% (12/13)	92% (12/13)	92% (12/13)	96% (24/25)

^a INPH, idiopathic normal-pressure hydrocephalus; SNPH, secondary NPH; Ro, outflow resistance; PPV, positive predictive value; NPV, negative predictive value.

TABLE 3.3. Predictive value of external lumbar drainage for shunt-responsive normal-pressure hydrocephalus (primary and secondary)^a

Series (ref. no.)	INPH/SNPH (n)	Sensitivity	Specificity	PPV	NPV	Accuracy
Haan and Thomeer, 1988 (10)	17/0	100% (12/12)	100% (5/5)	100% (12/12)	100% (5/5)	100% (17/17)
Williams et al., 1998 (25)	86 (mixed)	97% (31/32)	60% (9/15)	84% (31/37)	90% (9/10)	75% (40/47)
Walchenbach et al., 2002 (23)	43/6	50% (14/28)	80% (8/10)	80% (14/16)	36% (14/22)	58% (22/38)

^a INPH, idiopathic normal-pressure hydrocephalus; SNPH, secondary NPH; PPV, positive predictive value; NPV, negative predictive value.



Supplemental prognostic tests

- ICP monitoring

- Increased frequency in B waves is indicative of lowered compliance and/or may play an important role in the pathophysiology of the ventriculomegaly nad neuronal dysfunction
- static ICP values versus dynamic (pulsatile) ICP values

- (Radionuclide cisternography)

Diagnostic Intracranial Pressure Monitoring and Surgical Management in Idiopathic Normal Pressure Hydrocephalus: A 6-Year Review of 214 Patients

Per Kristian Eide, MD, PhD
Division of Clinical Neuroscience
Department of Neurosurgery,
Rikshospitalet University Hospital,
Oslo, Norway

Wilhelm Sorteberg, MD, PhD
Division of Clinical Neuroscience
Department of Neurosurgery,
Rikshospitalet University Hospital,
Oslo, Norway

Reprint requests:
Per Kristian Eide, MD, PhD,
Division of Clinical Neuroscience,
Department of Neurosurgery,
Rikshospitalet University Hospital,
0027 Oslo, Norway.
E-mail: perkristian.eide@
riks.hospitalet.no

Received, October 11, 2008.
Accepted, September 25, 2009.

Copyright © 2010 by the
Congress of Neurological Surgeons

OBJECTIVE: To review our experience of managing idiopathic normal pressure hydrocephalus (iNPH) during the 6-year period from 2002 to 2007, when intracranial pressure (ICP) monitoring was part of the diagnostic workup.

METHODS: The review includes all iNPH patients undergoing diagnostic ICP monitoring during the years 2002 to 2007. Clinical grading was done prospectively using a normal pressure hydrocephalus (NPH) grading scale (scores from 3 to 15). The selection of patients for surgery was based on clinical symptoms, enlarged cerebral ventricles, and findings on ICP monitoring. The median follow-up time was 2 years (range, 0.3-6 years). Both static ICP and pulsatile ICP were analyzed.

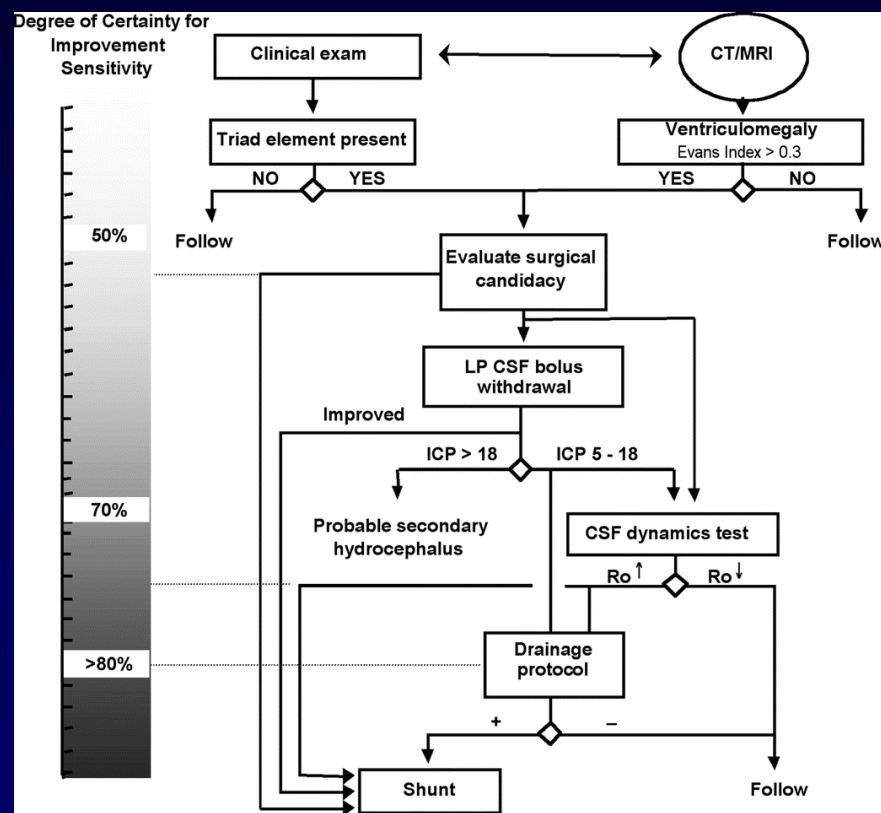
RESULTS: A total of 214 patients underwent the diagnostic workup, of whom 131 went on to surgery. Although 1 patient died shortly after treatment, 103 of the 130 patients (79%) improved clinically. This improvement lasted throughout the observation period. The static ICP observed during ICP monitoring was a poor predictor of the response to surgery. In contrast, among 109 of 130 patients with increased ICP pulsatility (ie, ICP wave amplitude >4 mm Hg on average and >5 mm Hg in >10% of recording time), 101 (93%) were responders (ie, increase in the NPH score of >2). Correspondingly, only 2 of 21 (10%) without increased ICP pulsatility were responders. Superficial wound infection was the only complication of ICP monitoring and occurred in 4 (2%) patients.

CONCLUSION: Surgical results in iNPH were good with almost 80% of patients improving after treatment. The data indicate that improvement after surgery can be anticipated in 9 of 10 iNPH patients with abnormal ICP pulsatility, but in only 1 of 10 with normal ICP pulsatility. Diagnostic ICP monitoring had a low complication rate.

KEY WORDS: Intracranial pressure monitoring, Normal pressure hydrocephalus, Shunt response

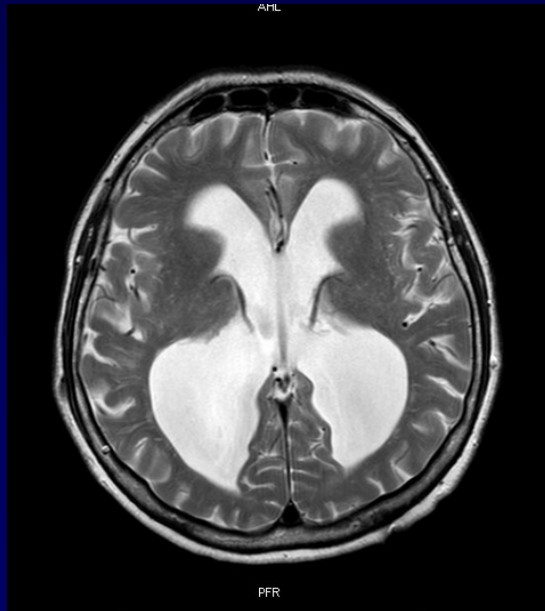
Identifying „shunt-responsive“ patients

- Correlation between clinical symptoms and CT/MRI exams.
- Resistance testing (can be repeated and compared with testing before)
- ELD
- Tap test (single application, repeated)



Aqueductal stenosis

- Symptoms are similar to normal pressure hydrocephalus.
- But treatment different – 3rd ventriculostomy.

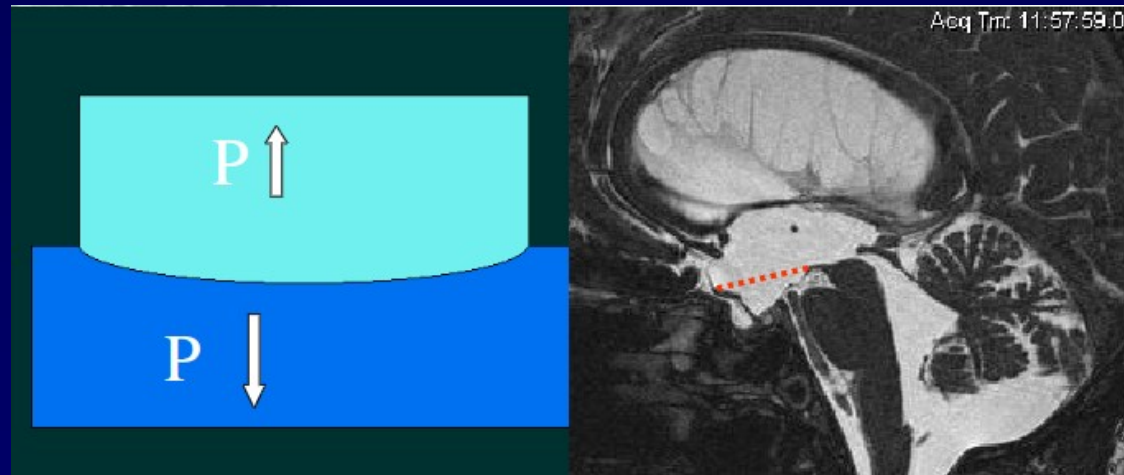


LOVA

- **LOVA = longstanding overt ventriculomegaly in adult**
- **subtype of chronic hydrocephalus**
- **develops during childhood, manifests symptoms during adulthood**
- **diagnostic criteria:**
 - **severe ventriculomegaly**
 - **macrocephaly measuring more than two standard deviations in head circumference**
 - **and/or neuroradiological evidence of a significantly expanded or destroyed sella turcica.**
- **symptoms: headache and/or Adam's trias**
- **caused by aqueductal stenosis**
- **treatment is difficult in terms of their sensitive compliance of brain parenchyma**
- **surgery: ETV, shunts (cave - over-drainage).**

InfinOH

- **InfinOH = infratentorial intracisternal obstructive hydrocephalus**
- Subtype of communicating hydrocephalus with free communication between the ventricles and subarachnoid space but with infratentorial intracisternal obstruction
- MRI – a downward bulged floor of the third ventricle and a discrepancy of sizes between the great and the prepontine cistern.
- Treatment : ETV, shunt.



Arrested (compensated) hydrocephalus

- **The condition in which the neurological status of the patient is stable in the presence of stable ventriculomegaly.**
- **CSF formation and absorption are in balance and no CSF accumulates.**
- **Spontaneous or surgical termination of a hydrocephalic condition with subsequent return to normal of the pressure across the cerebral mantle.**
- **Cave – subtle deterioration.**
- **?? Probably results from improvement of CSF circulation during growth.**

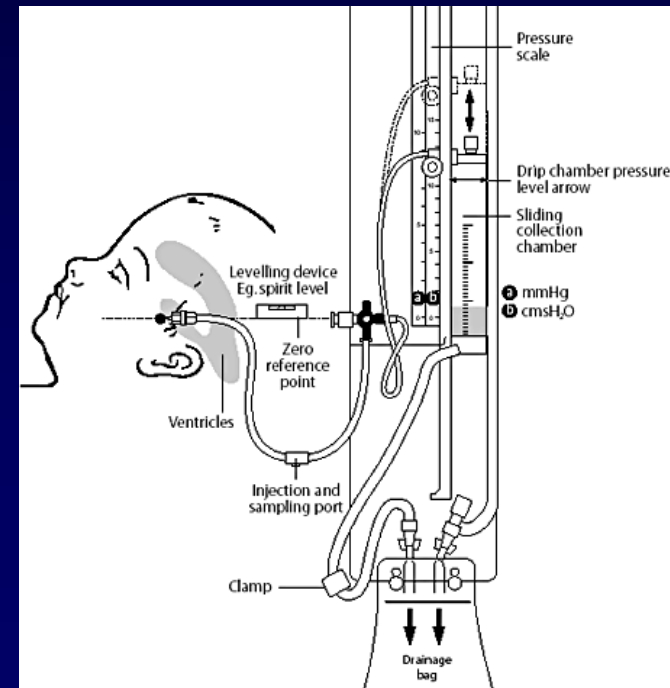
Hydrocephalus treatment

- **Observation (asymptomatic)**
- **Conservative treatment (acetazolamide, furosemide, osmotic agents)**
- **Surgery**
 - **Temporary (acute HCP)**
 - External ventricular drainage
 - External lumbar drainage
 - **Permanent (chronic HCP)**
 - Shunt
 - Neuroendoscopy
 - Others (Torkildsen drainage etc.)
- **Pediatric hydrocephalus**
- **Adult hydrocephalus**



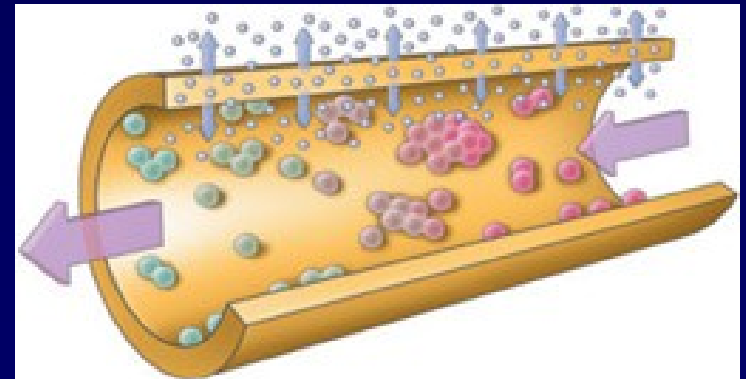
External ventricular drainage

- allows the temporary drainage of CSF
 - to relieve raised intracranial pressure
 - to divert infected CSF
 - to divert bloodstained CSF following neurosurgery/haemorrhage
 - to divert the flow of CSF
- indications:
 - to relieve raised intracranial pressure
 - to divert infected CSF
 - to divert bloodstained CSF following neurosurgery/haemorrhage
 - to divert the flow of CSF
- catheter is placed into the lateral ventricle through a burr hole made in the skull, tunnelled under the skin and connects to an external drainage system



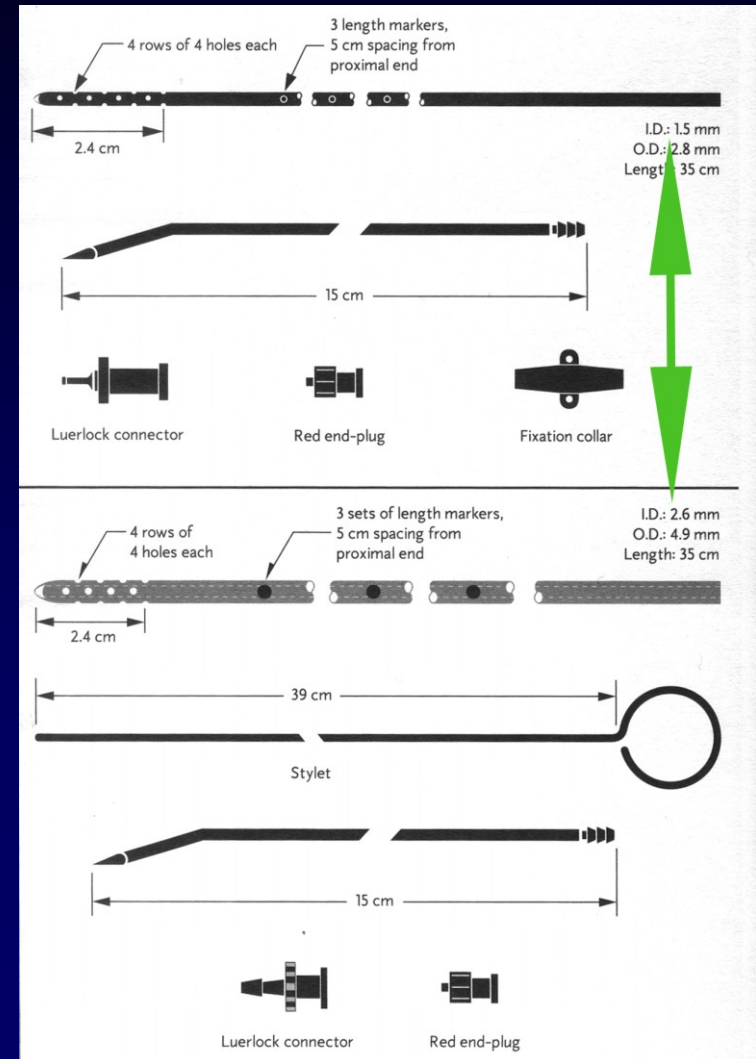
External ventricular drainage

- Spiegelberg Silverline
 - Silver ions – strong antiseptic effects with a broad spectrum.
 - Continual release of silver within 30 days in antiseptic concentration.
- Bactiseal catheter (antibiotic impregnated)



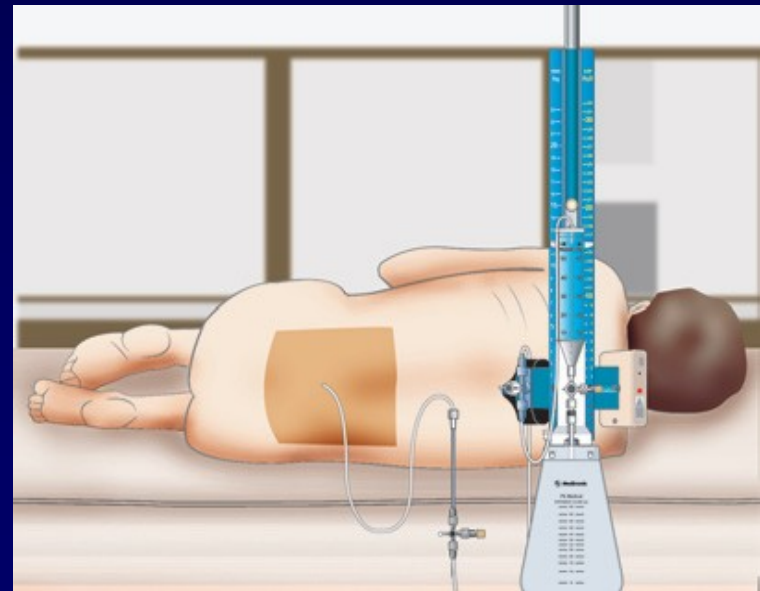
External ventricular drainage

- Special thick ELD
- Indication:
 - hemocephalus



External lumbar drainage

- **Indications: similar to external lumbar drainage, but not in case of obstructive hydrocephalus.**
- **SAH, wound complications**

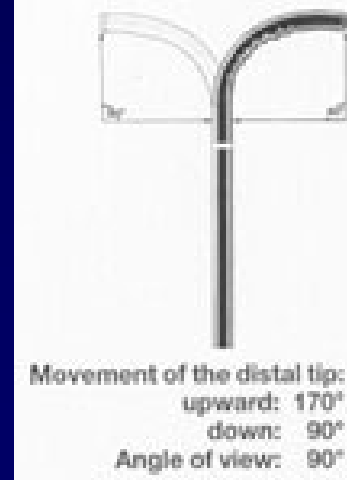
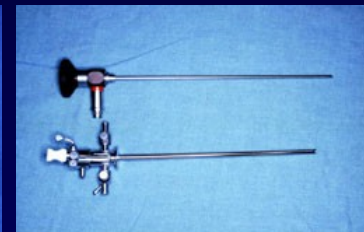
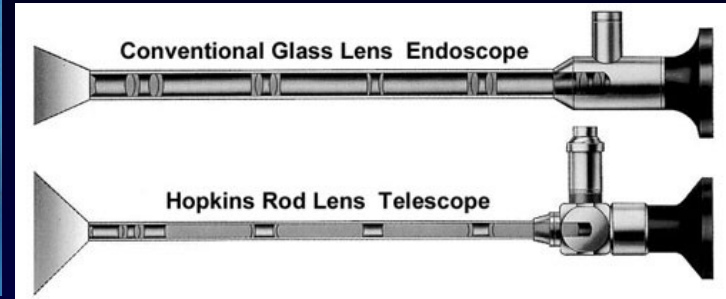


Neuroendoscopy

Endoscopes

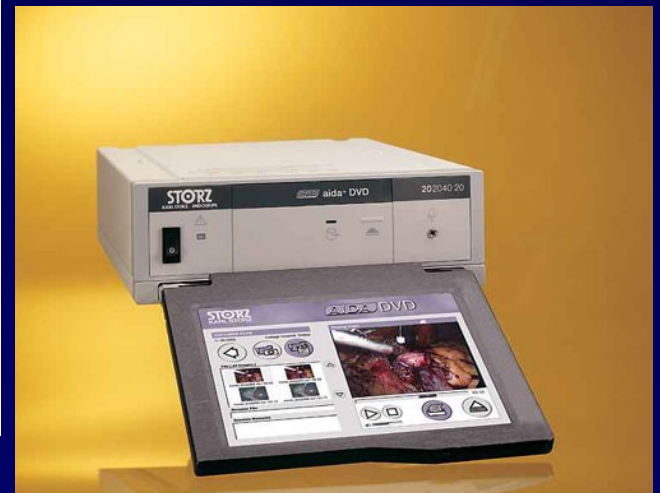
– Rigid

– Flexible



Neuroendoscopy

Light sources, cameras (full-HD),
recording devices



Neuroendoscopy

Instruments



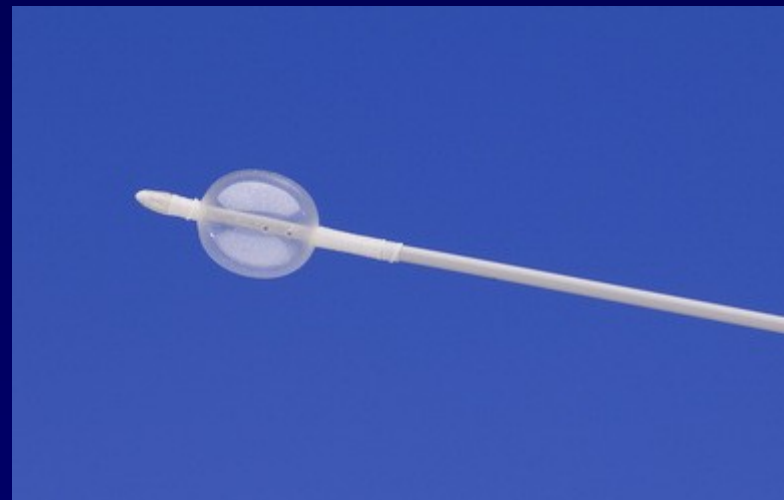
Neuroendoscopy

HOLDERS



Neuroendoscopy

- shuntscope
- balloon catheter
- introducer



Endoscopic treatment of HCP

- Endoscopic third ventriculostomy
- Aqueductoplasty
- Septum pellucidum fenestration
- Fenestration of multiloculated ventricles
- Foraminoplasty
- Lamina terminalis fenestration

Endoscopic third ventriculostomy

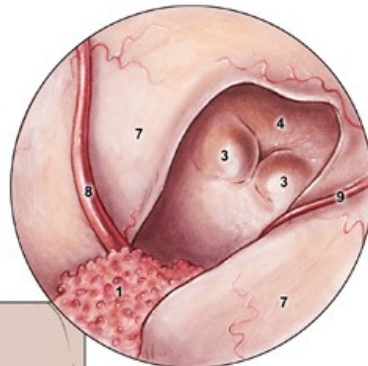
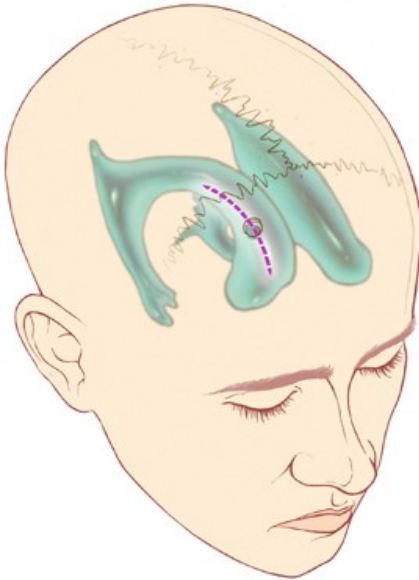
- **Indication – non-communicating HCP**
 - **LOVA: Long-standing Overt Ventriculomegaly in Adults (Oi, 2000)**
 - **InfinOH: Infratentorial Intracisternal Obstructive Hydrocephalus (Kehler)**
 - **unusual/controversial indications:**
 - **Empty sella syndrom**
 - **Posthaemorrhagic hydrocephalus**
 - **Postinfectious hydrocephalus**
 - **Chiari malformatin**
 - **Dandy-Walker malformation**
 - **Previous V-P shunt implantation**

Endoscopic third ventriculostomy

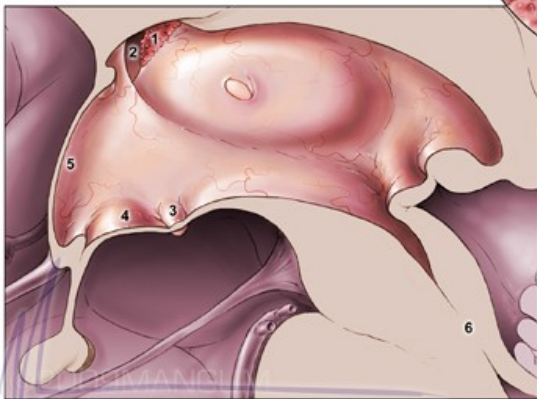
Endoscopic Third Ventriculostomy

Obstructive Hydrocephalus blocks the passage of cerebrospinal fluid (CSF) from the third ventricle to the fourth. This causes an excess of pressure in the lateral and third ventricles.

Surgical access is obtained by incising the scalp, and creating a burr-hole through the cranium. Neuroendoscopic entry is made through the lateral ventricle, then passing through the interventricular foramen.



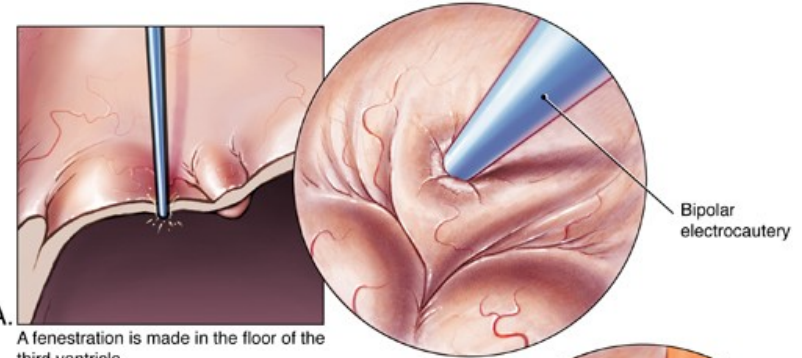
Neuroendoscopic View



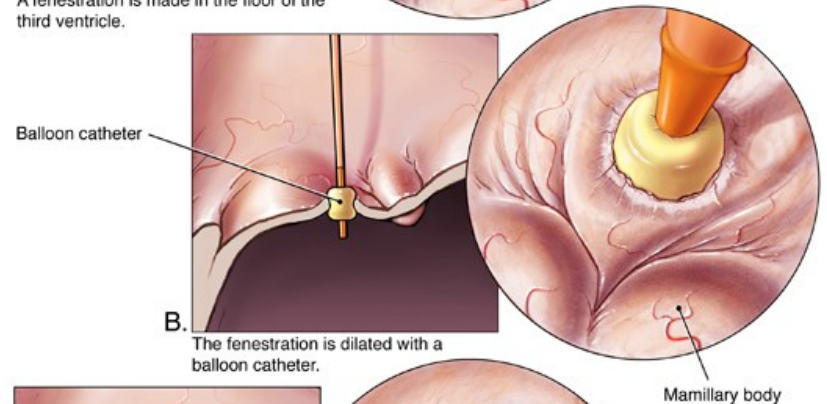
Midsagittal View

Relevant Anatomy

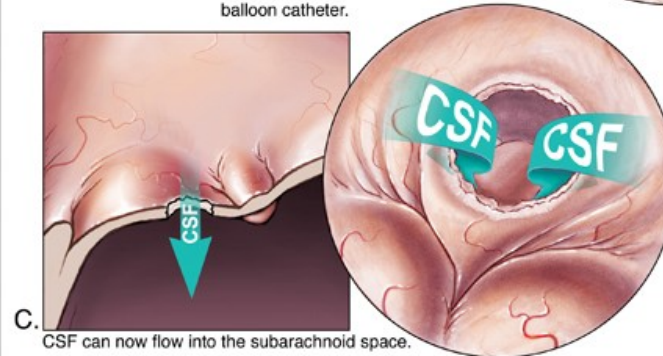
1. Choroid Plexus
2. Interventricular Foramen
3. Mamillary Body
4. Floor of Third Ventricle
5. Lamina Terminalis
6. Non-patent Cerebral Aqueduct
7. Walls of Lateral Ventricle
8. Thalamostriate Vein
9. Septal Vein



A. A fenestration is made in the floor of the third ventricle.

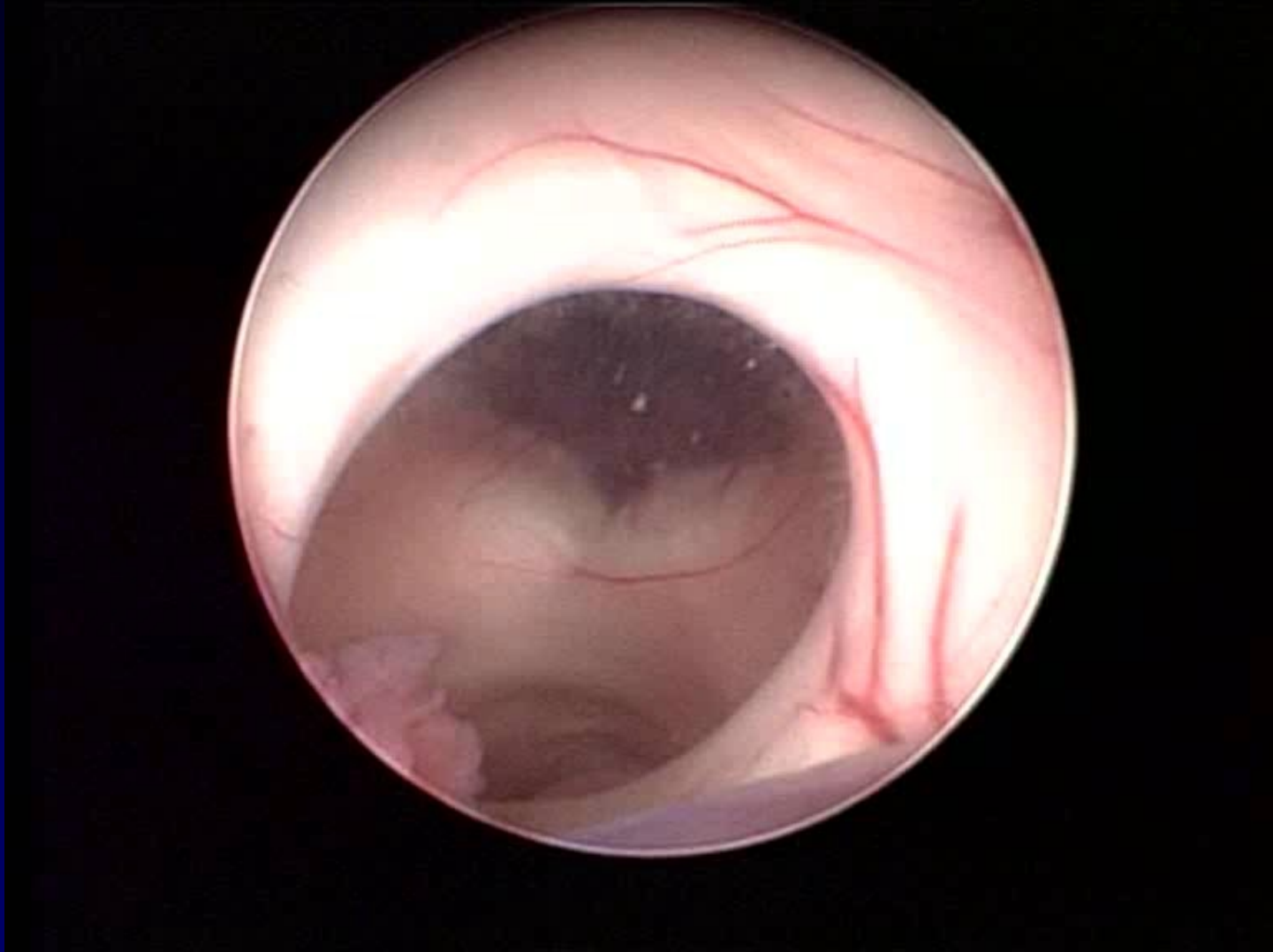


B. The fenestration is dilated with a balloon catheter.



C. CSF can now flow into the subarachnoid space.

Endoscopic third ventriculostomy



Endoscopic third ventriculostomy

60-90% succes rate

Outcome depends on etiology

– the worst results:

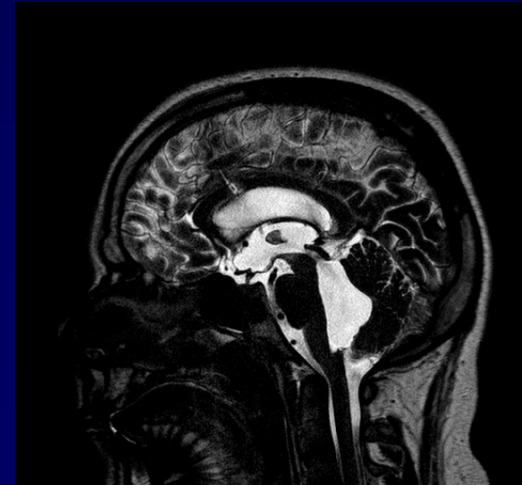
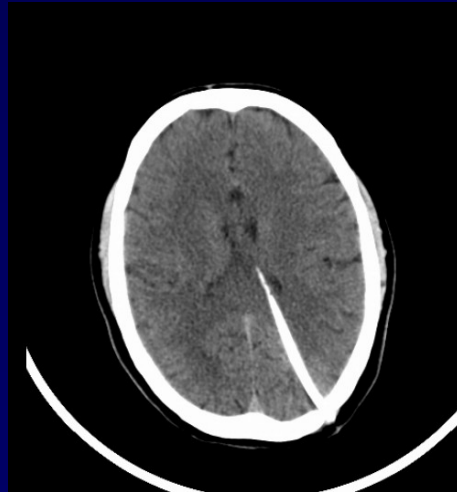
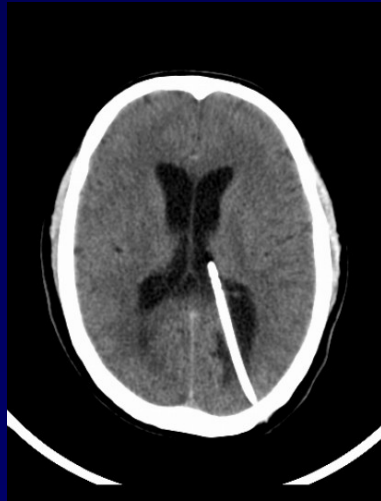
- **postinfectious hydrocephalus**
- **posthaemorrhagic hydrocephalus**
- **childern under 6 month**

Endoscopic third ventriculostomy

Patients with shunt dysfunction

- always check the possibility of endoscopic treatment

Shunt failure – MRI – if obstruction – ETV, if not – shunt revision

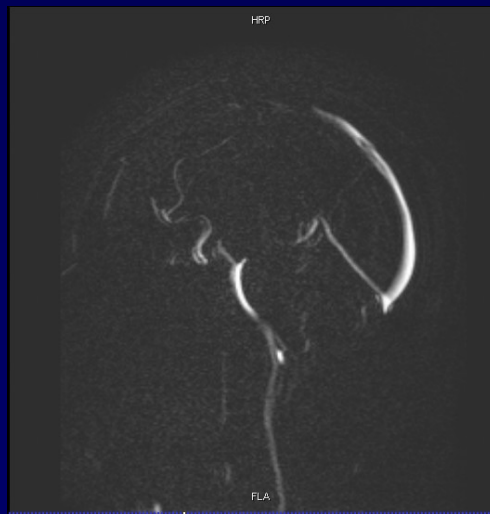


Endoscopic third ventriculostomy

Patency of the stoma on the floor of the third ventricle – MRI – phase contrast

Present in 94% of successful cases,
absent in 75% of the failures.

Cave – late failure of the stoma.



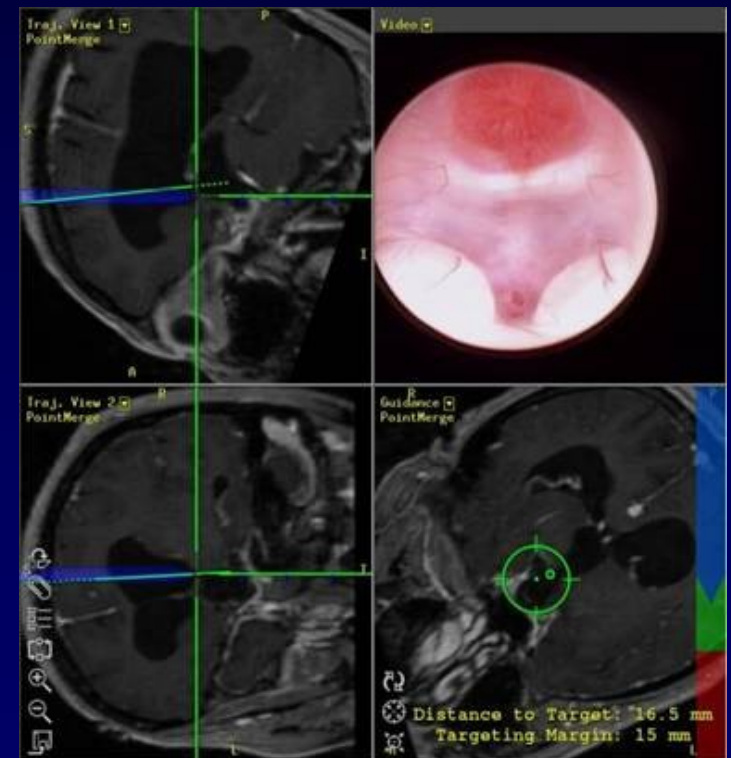
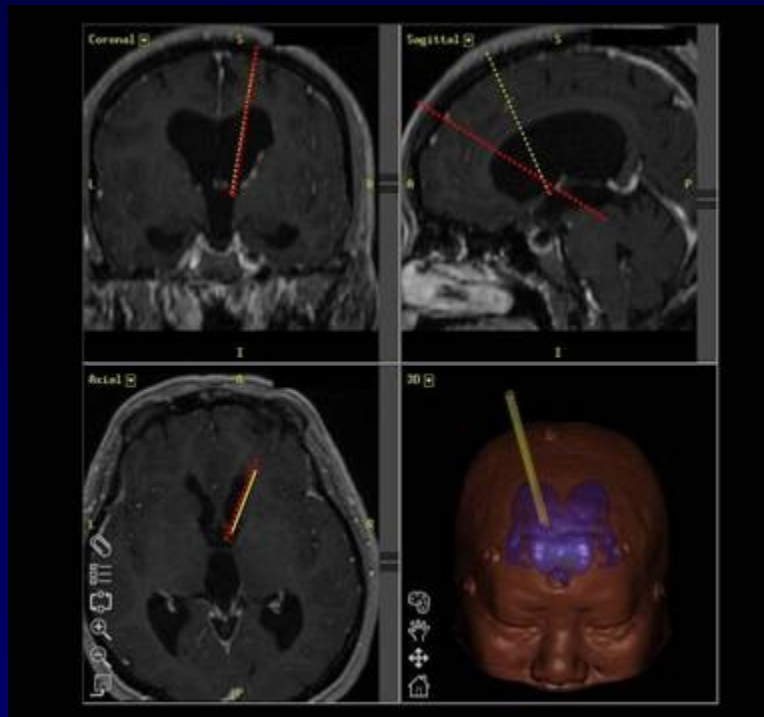
Endoscopic third ventriculostomy

Indication of repeated ETV:

- Clinical improvement after primary ETV
- Functional stoma proved on MRI
- Clinical deterioration
- Non-functional stoma on MRI in time of deterioration

Endoscopic third ventriculostomy

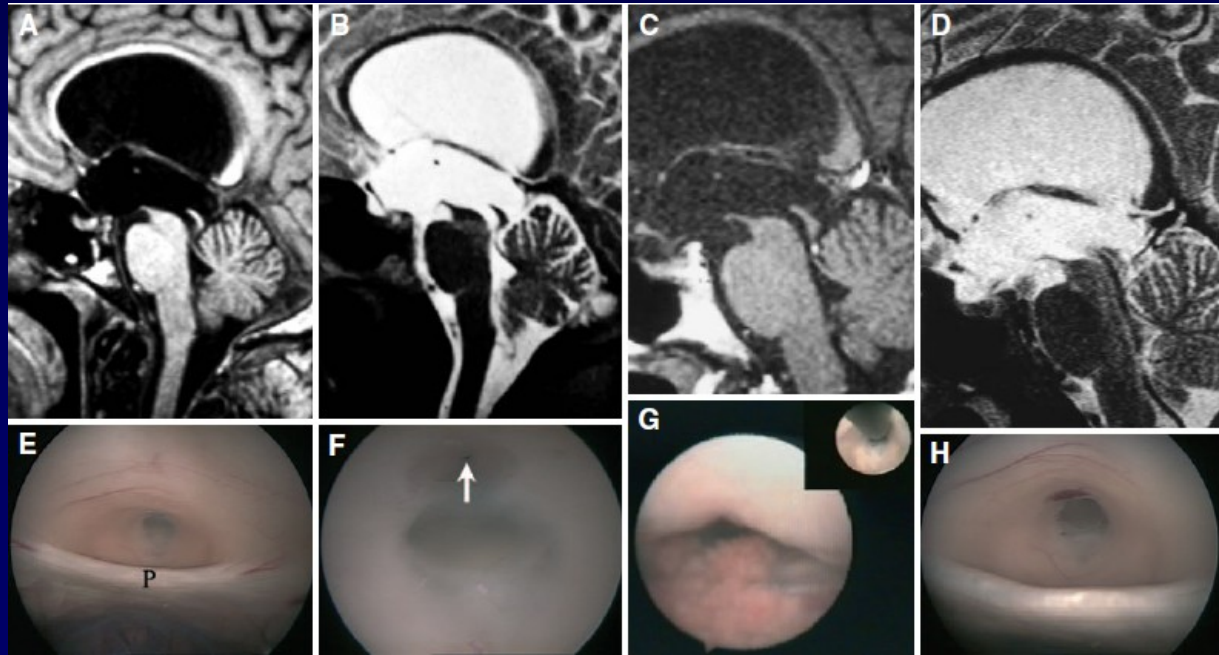
Free-hand technique x holder
Frame-less navigation



Aqueductoplasty

Indication:

- Isolated 4th ventricle
- Membrane in aqueduct

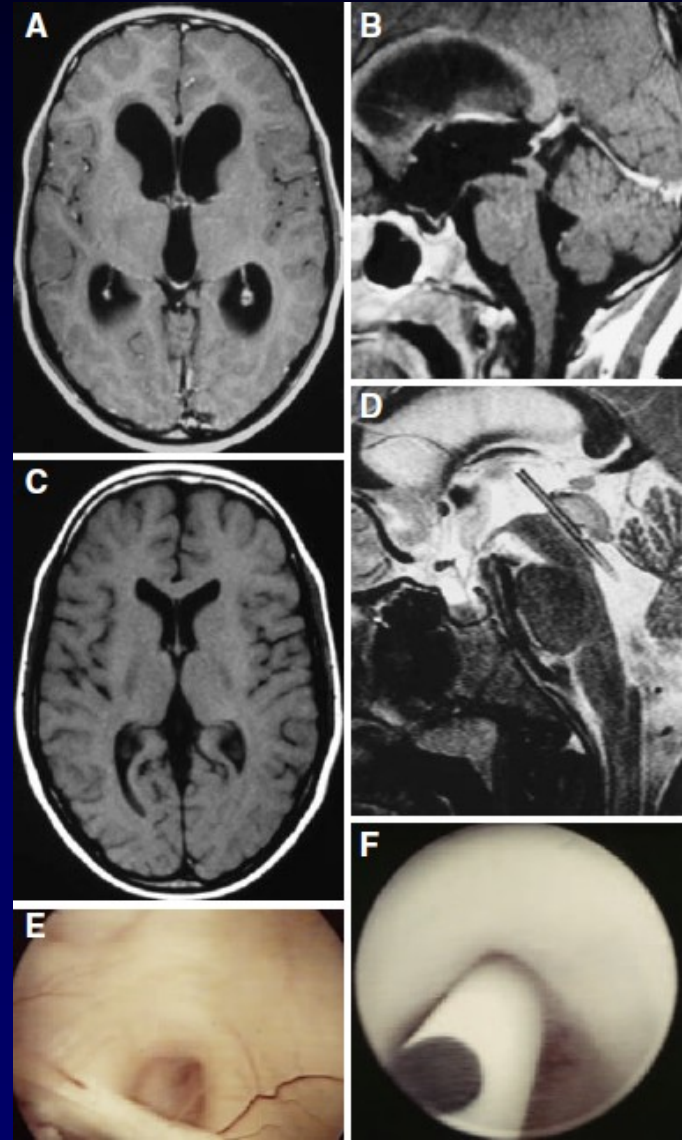


Aqueductoplasty

Aqueductoplasty with stenting

Indication:

- High risk of restenosis

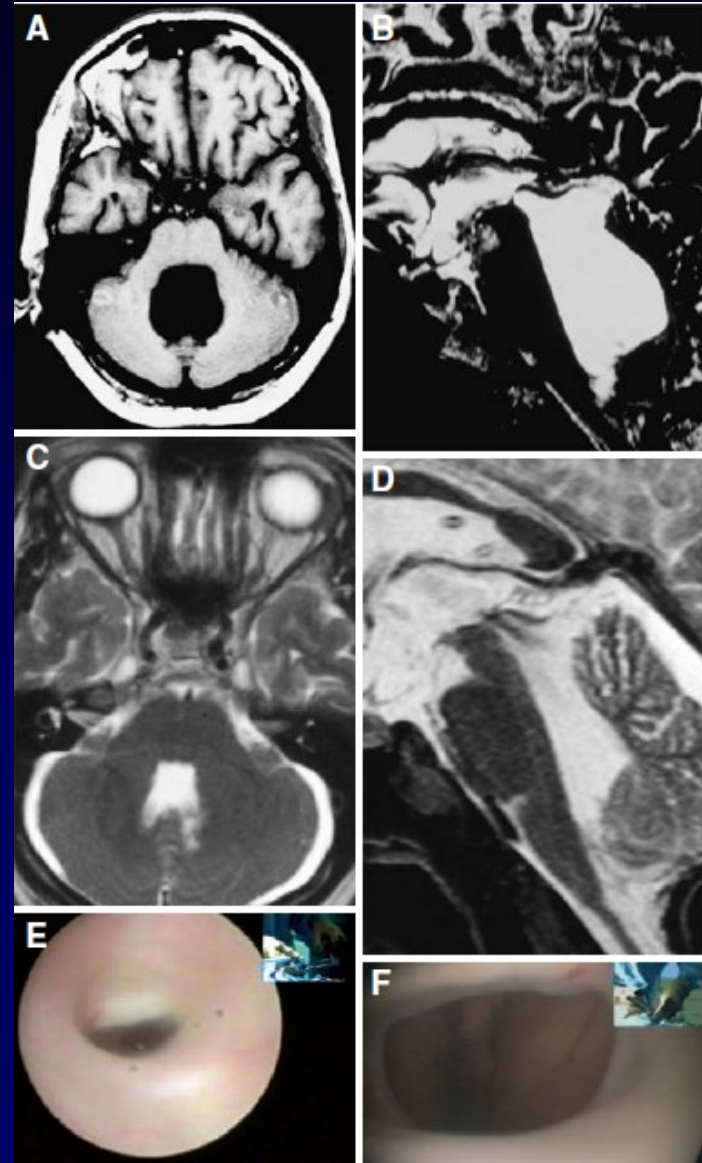


Aqueductoplasty

Aqueductoplasty retrograde

Indication:

- Isolated 4th ventricle
with slit ventricle syndrom



Endoscopic treatment of HCP

Fenestration of multiloculated ventricles

Indication

- postinfectious or poshaemorrhagic separation inside the ventricles

Frame-less navigation

Ultrasound perioperative control

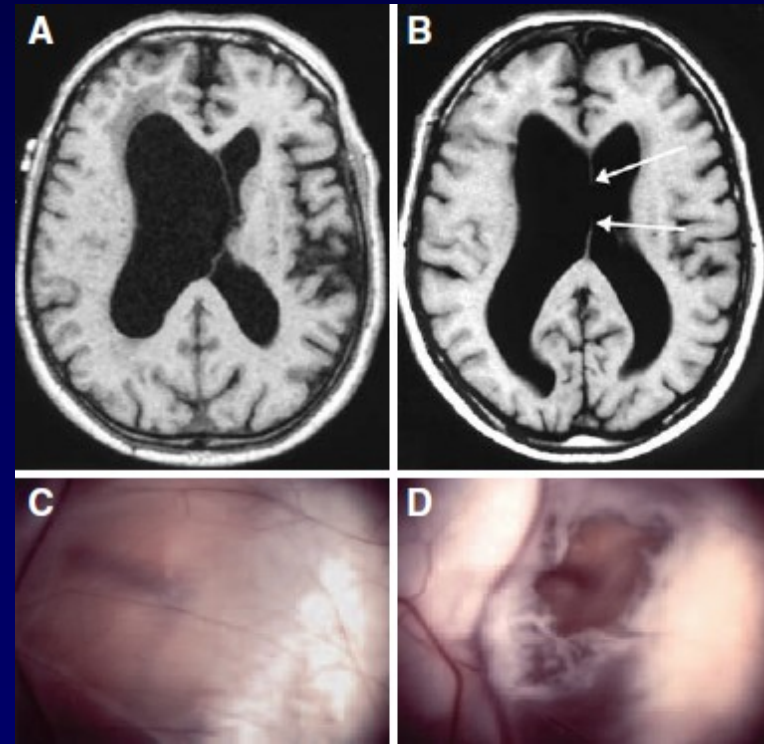


Endoscopic treatment of HCP

Septum pellucidum fenestration

Indication:

- Unilateral hydrocephalus
- „secure procedure“
in cases of unilateral
ventricular lesions

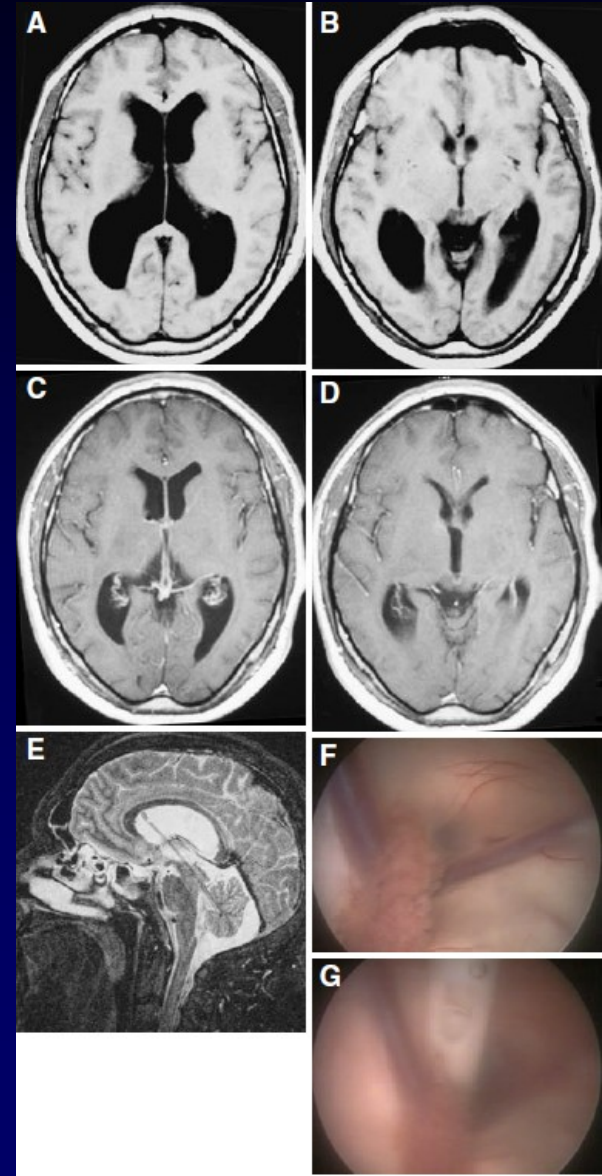


Endoscopic treatment of HCP

Foraminoplasty

– Indication:

- Obstruction of foramen Monro

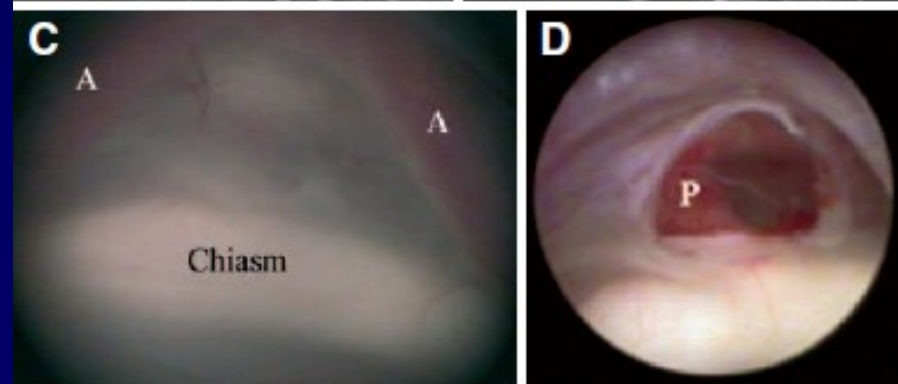
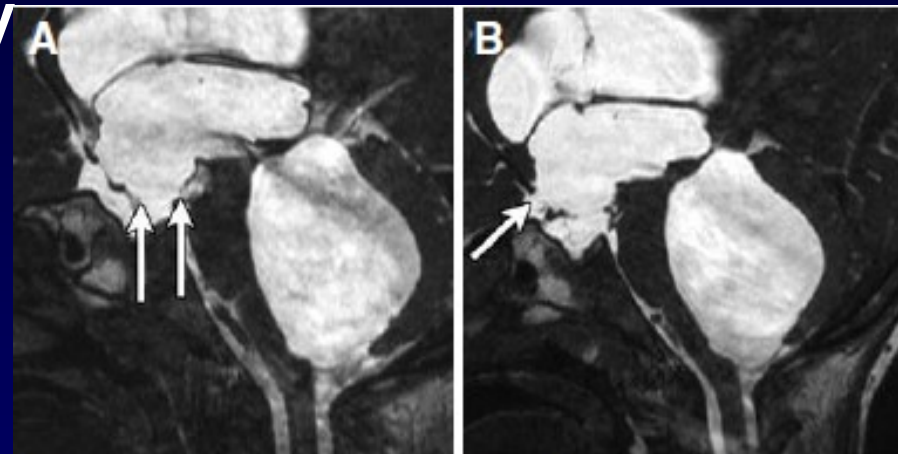


Endoscopic treatment of HCP

Lamina terminalis fenestration:

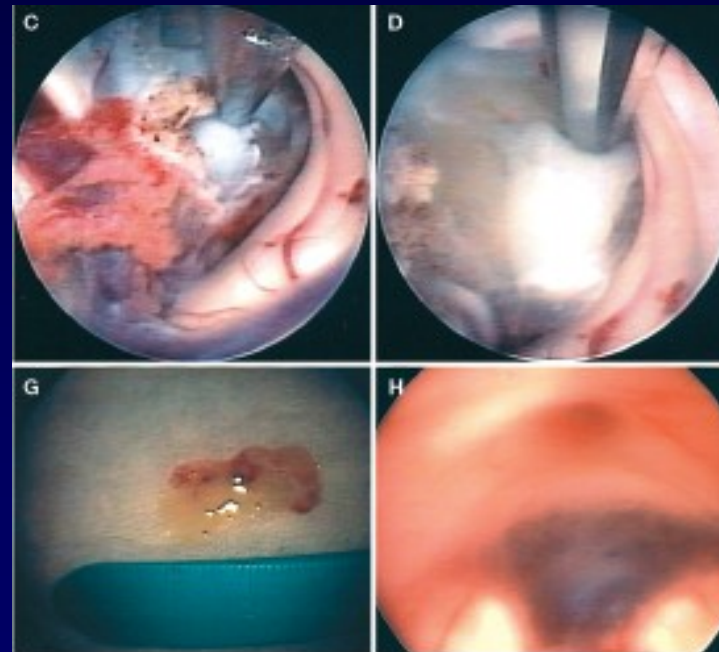
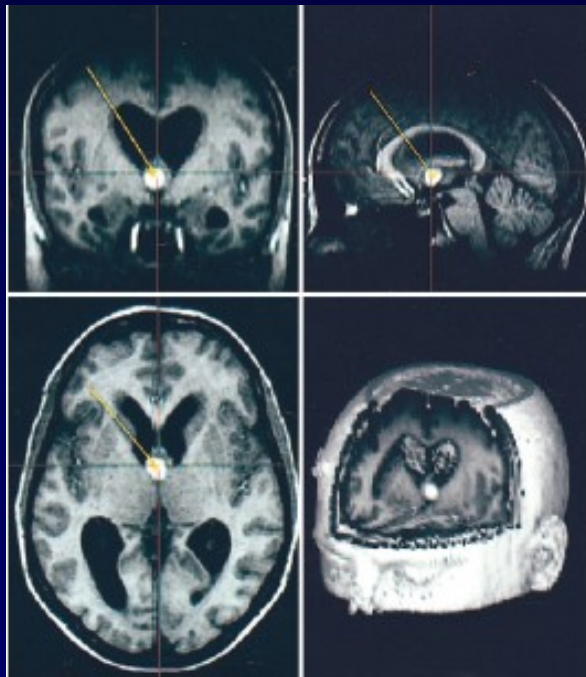
– Indication:

- If 3rd ventriculostomy cannot be performed



Endoscopic treatment of HCP

Colloid cyst – intermittent non-communicating hydrocephalus



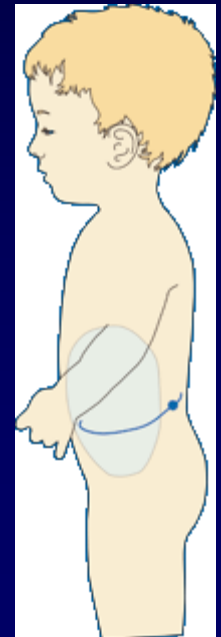
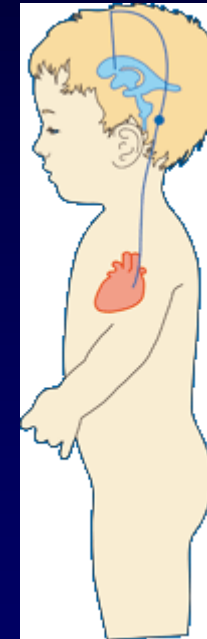
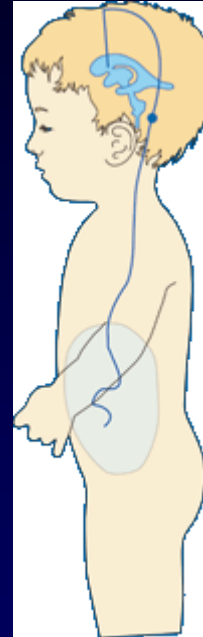
Endoscopic treatment of HCP

Complications:

- Bleeding (minor, major – can cause neurological deficit)
- CSF leak (CSF fistula, pseudomeningocele)
- Infection (contaminated endoscope, consequence of CSF leak)
- Ventricular collapse (subdural hematoma)
- Injury to brain structures (neurological deficit, death)
- Bradykarida, hypertension – elevation of ICP (to rapid irrigation etc.)

Shunts

- **Ventriculo-peritoneal**
- **Ventriculo-atrial**
- **Lumbo-peritoneal**
- **others :**
 - Ventriculo-subgaleal
 - Superior sagittal sinus
 - Ventriculo-pleural
 - Renal pelvis
 - Ventriculo-urethral
 - Ventriculo-vesical
 - Ventriculo-gastric
 - Bile ducts
 - Small intestine
 - Oviduct
 - Ventriculomastoidostomy
 - Bone marrow of the vertebra
 - Spinal epidural space



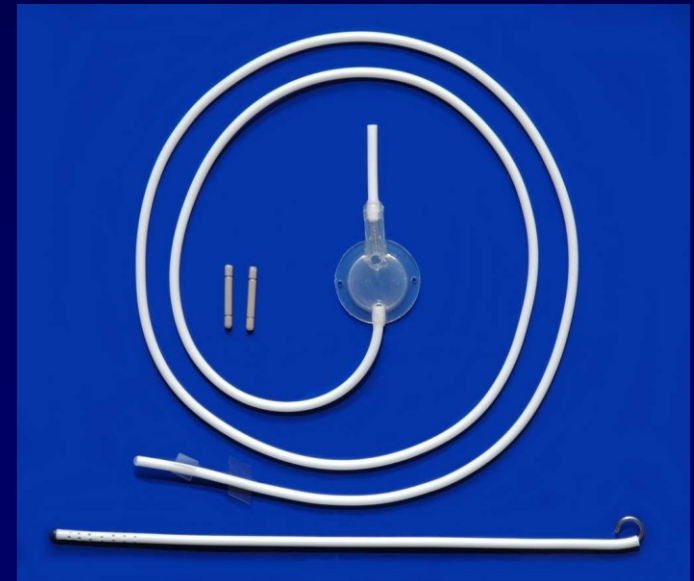
Valves

- **Differential-pressure valves**
 - static (monopressure)
 - adjustable
- **Hydrostatic valves (function of the valve depends on the change of physical parameters)**
 - Flow regulating valves
 - Valves with ASD (anti-siphon device)
 - Gravity assisted valves

Valves

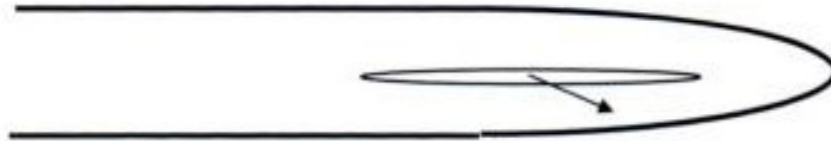
- ~ 20 manufacturers
- ~ 130 different valves
- ~ 450 different pressure setting

Chhabra shunt, low cost ~ 25 €, India

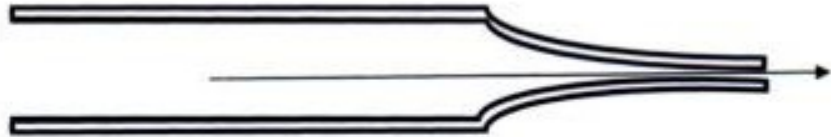


Valves

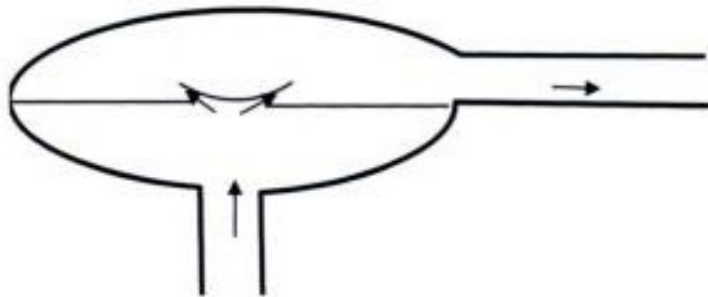
Slit valve



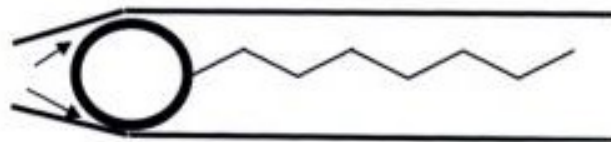
Mitre Valve



Diaphragm Valve

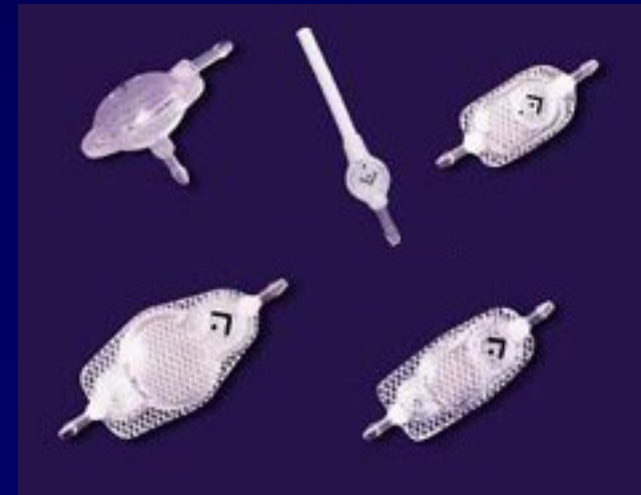
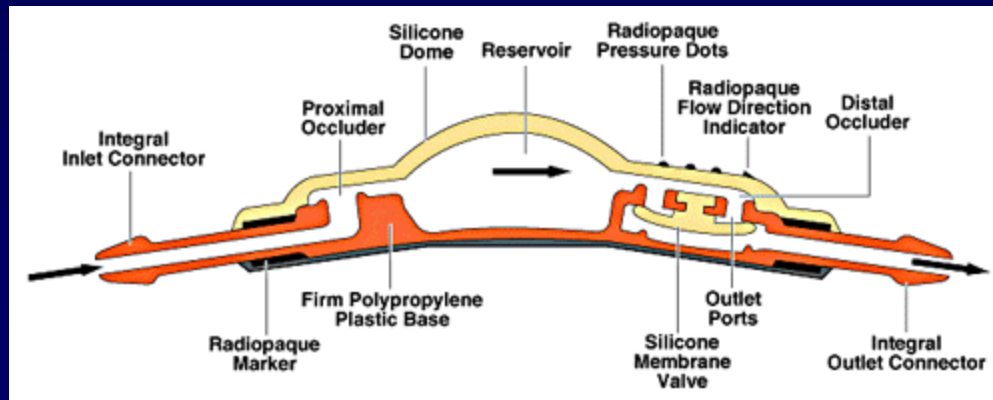


Ball in cone with spring valve

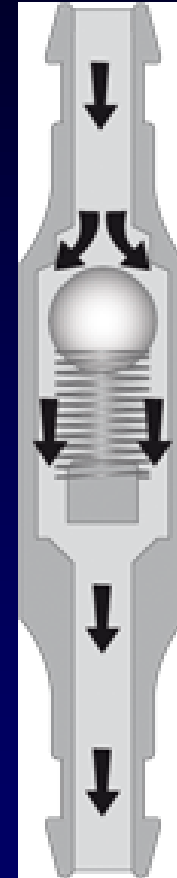
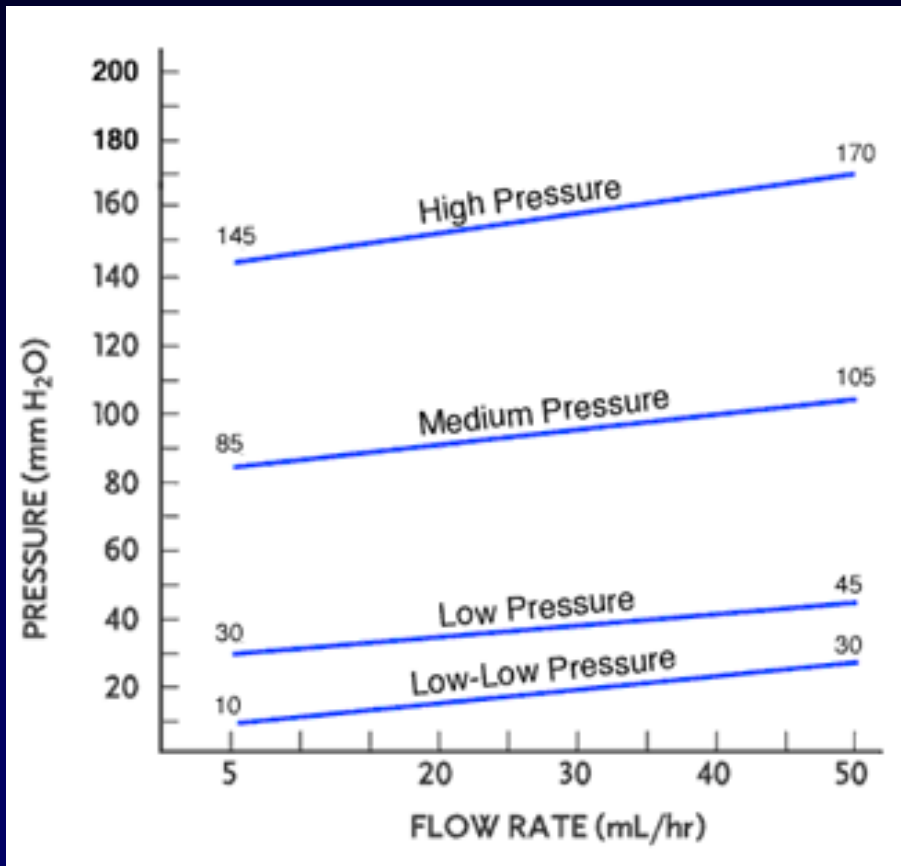


Valves

- **Differential-pressure valves**
 - static (monopressure)
 - The most common pressure ratings are:
 - Extra-low-pressure: 0-10 mm H₂O
 - Low-pressure: 10-50 mm H₂O
 - Medium-pressure: 51-100 mm H₂O
 - High-pressure: 101-200 mm H₂O

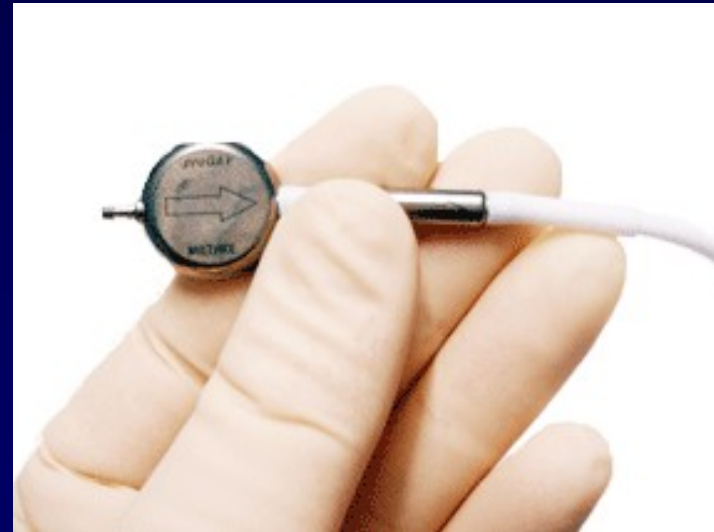
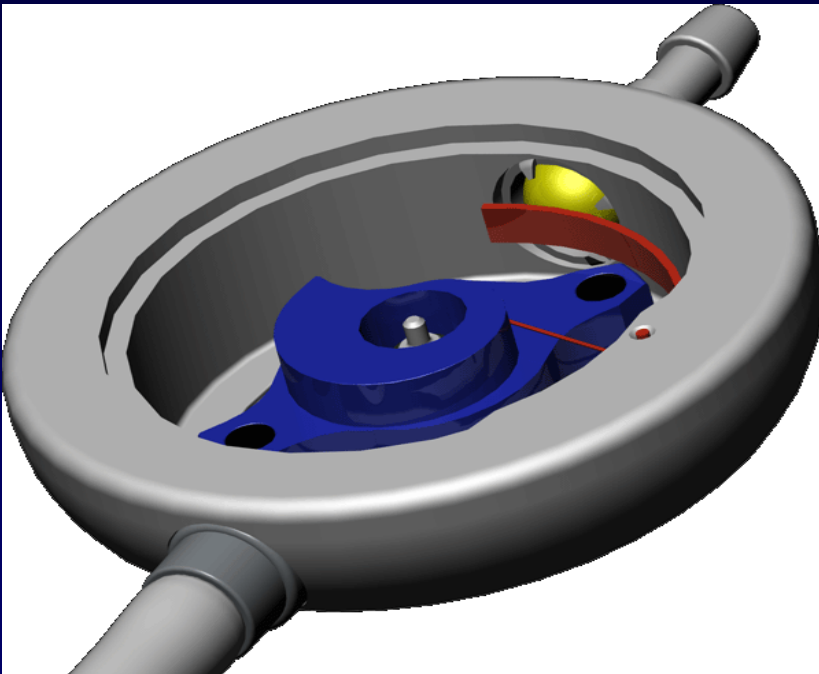


Valves



Valves

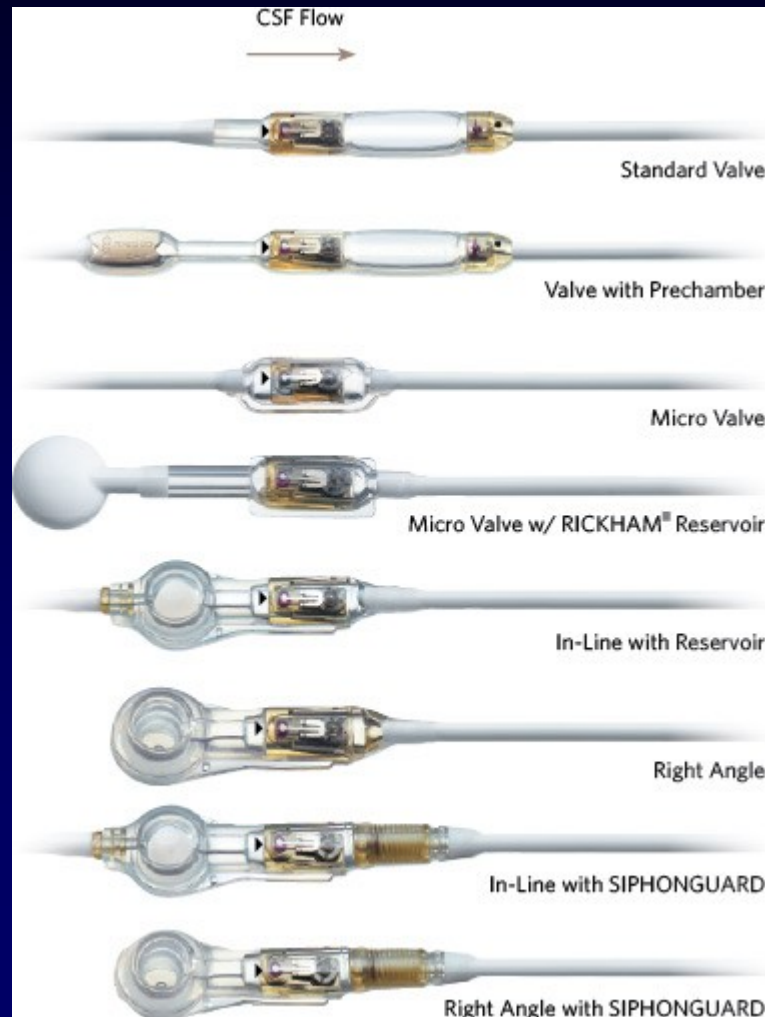
- **Differential-pressure valves**
 - adjustable



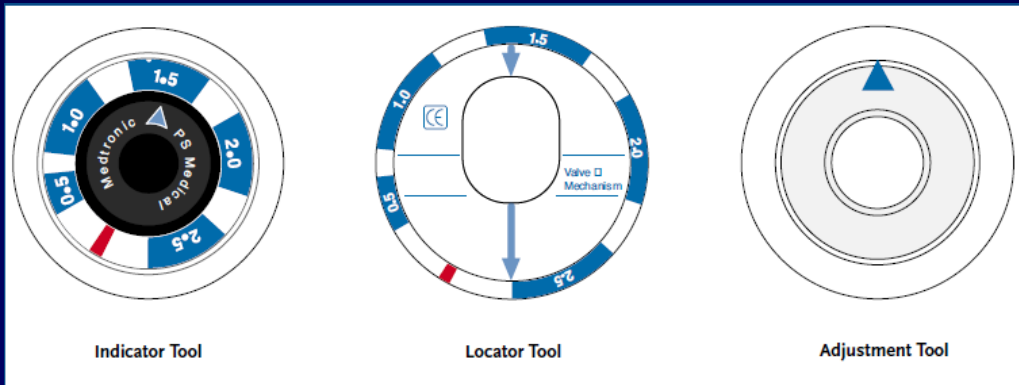
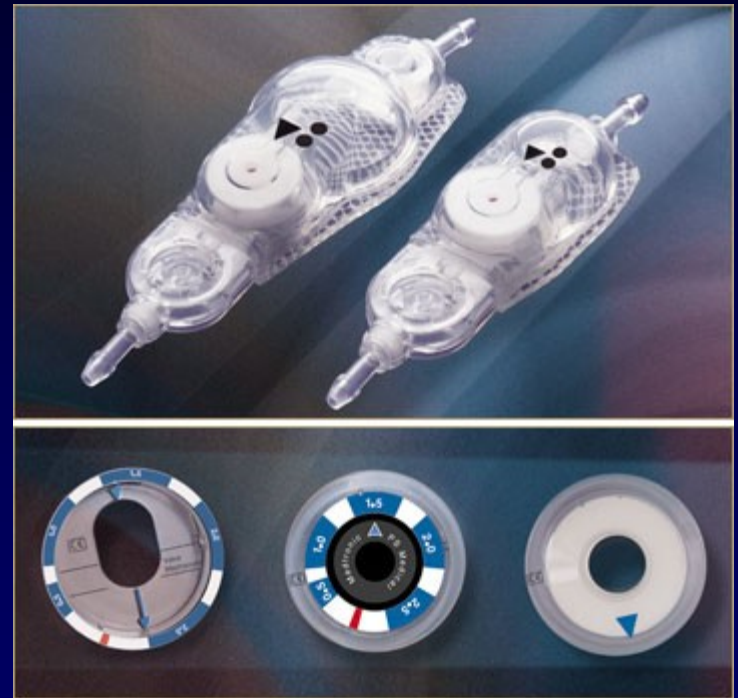
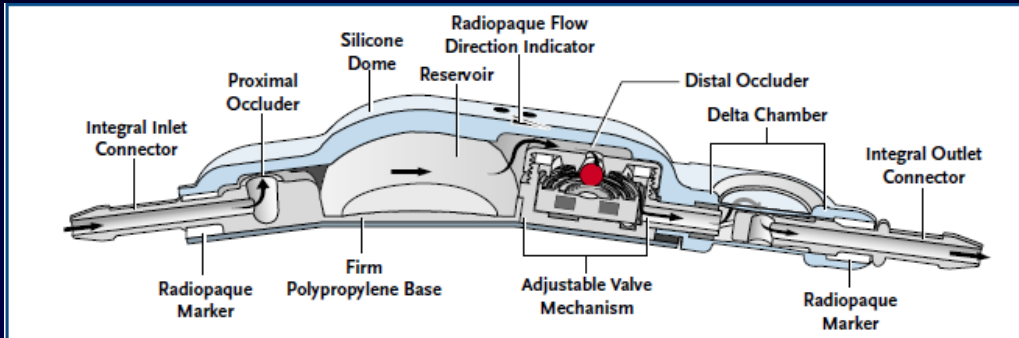
Valves



Valves



Valves



Valves

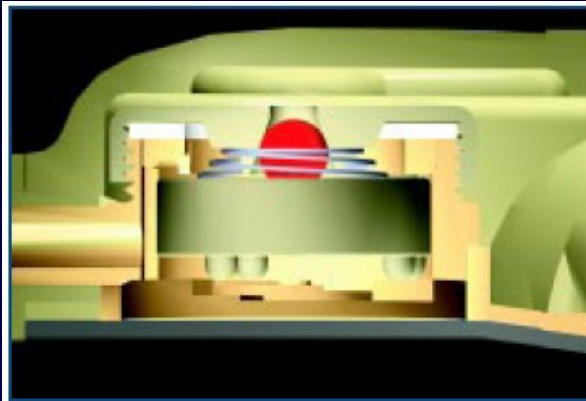


Fig. 8 Adjustable Valve Mechanism

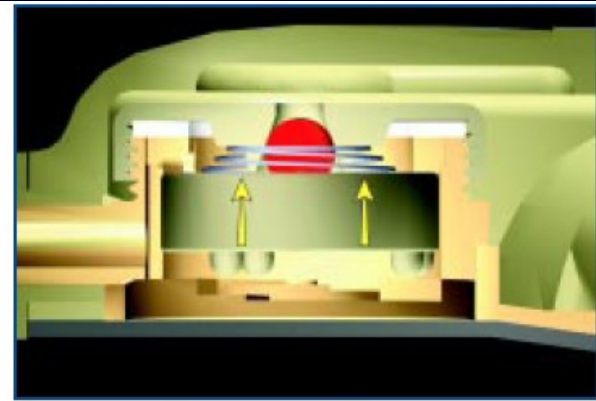


Fig. 9 Adjustable Valve Mechanism – Raised

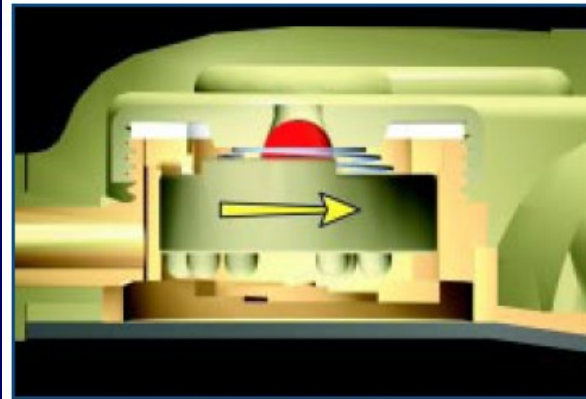


Fig. 10 Adjustable Valve Mechanism – Rotating

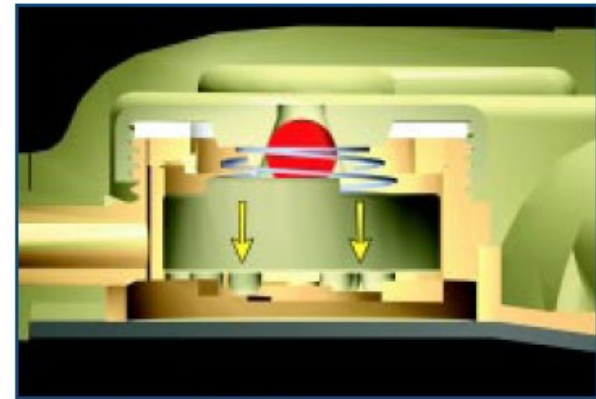
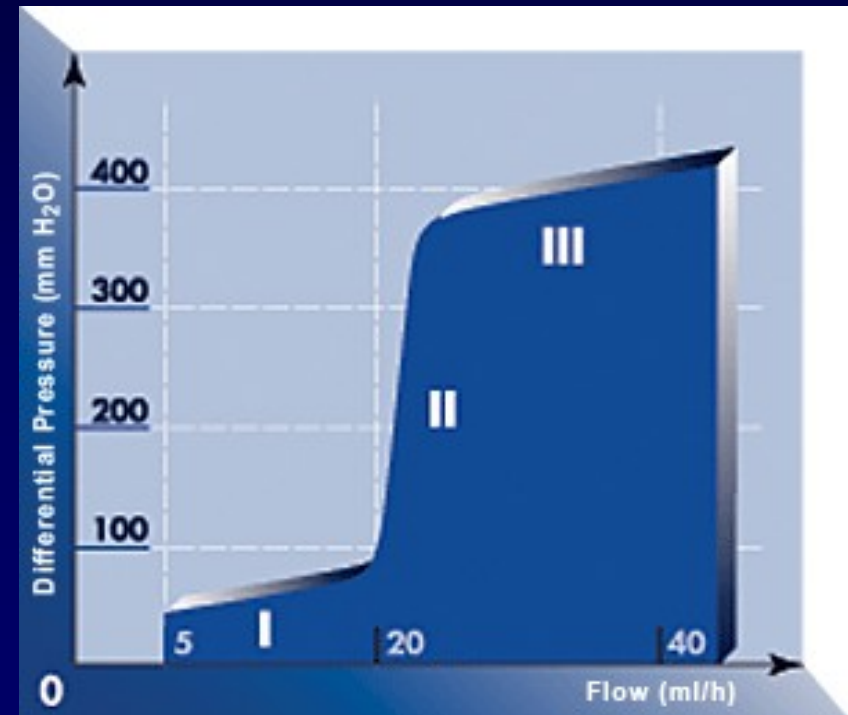


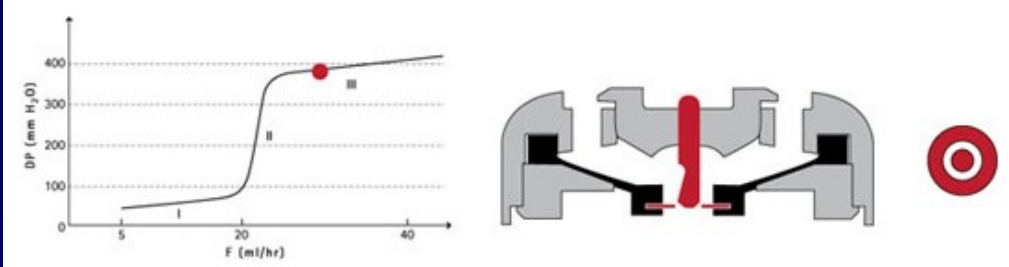
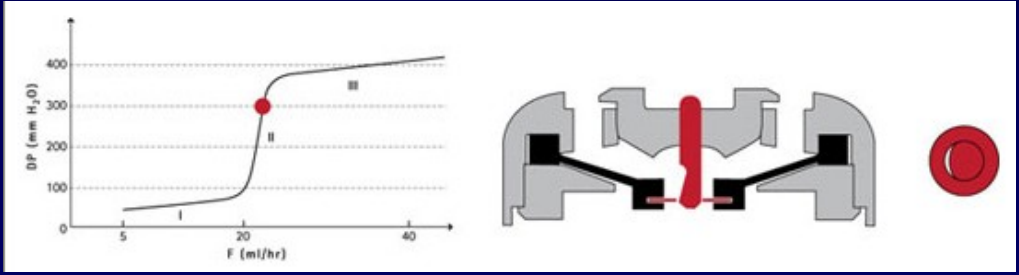
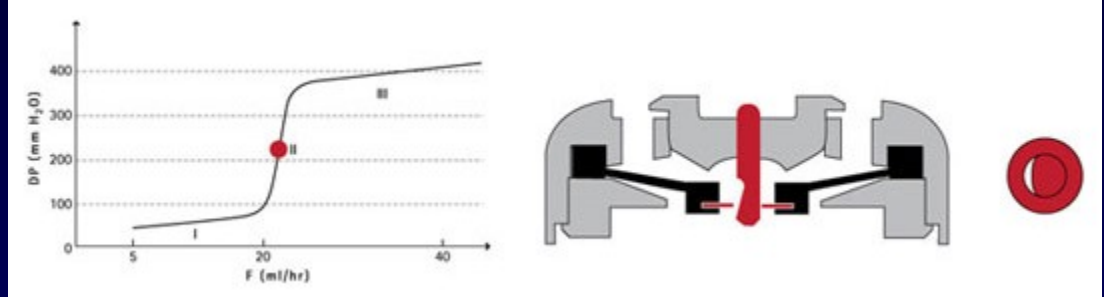
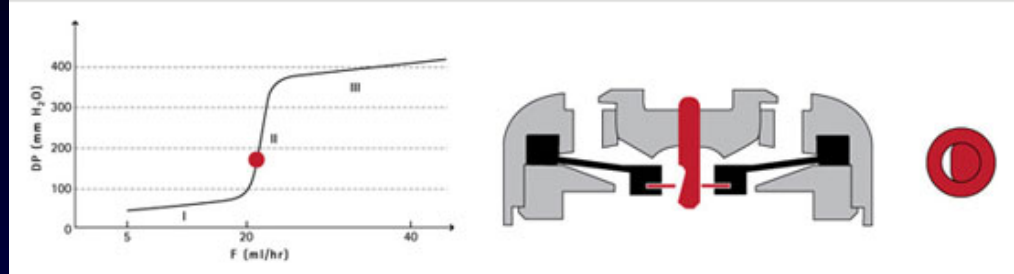
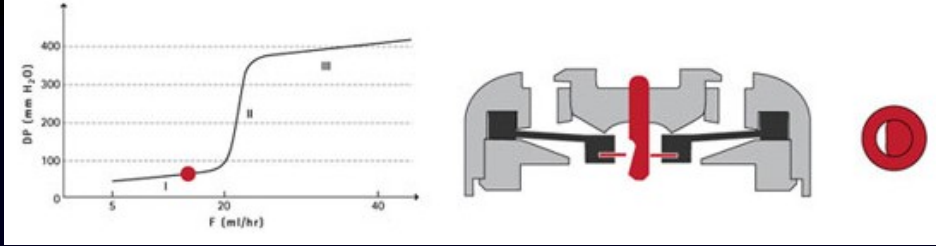
Fig. 11 Adjustable Valve Mechanism –
New Performance Level Setting

Valves

Hydrostatic valves (function of the valve depends on the change of physical parameters)

- Flow regulating valves

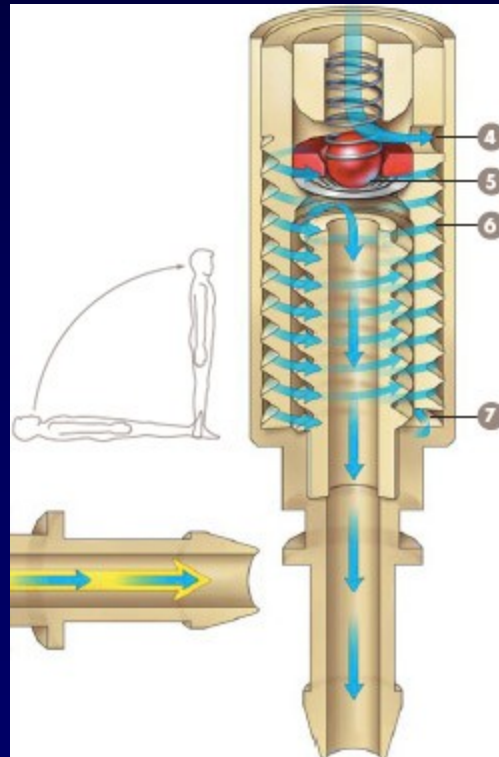




Valves

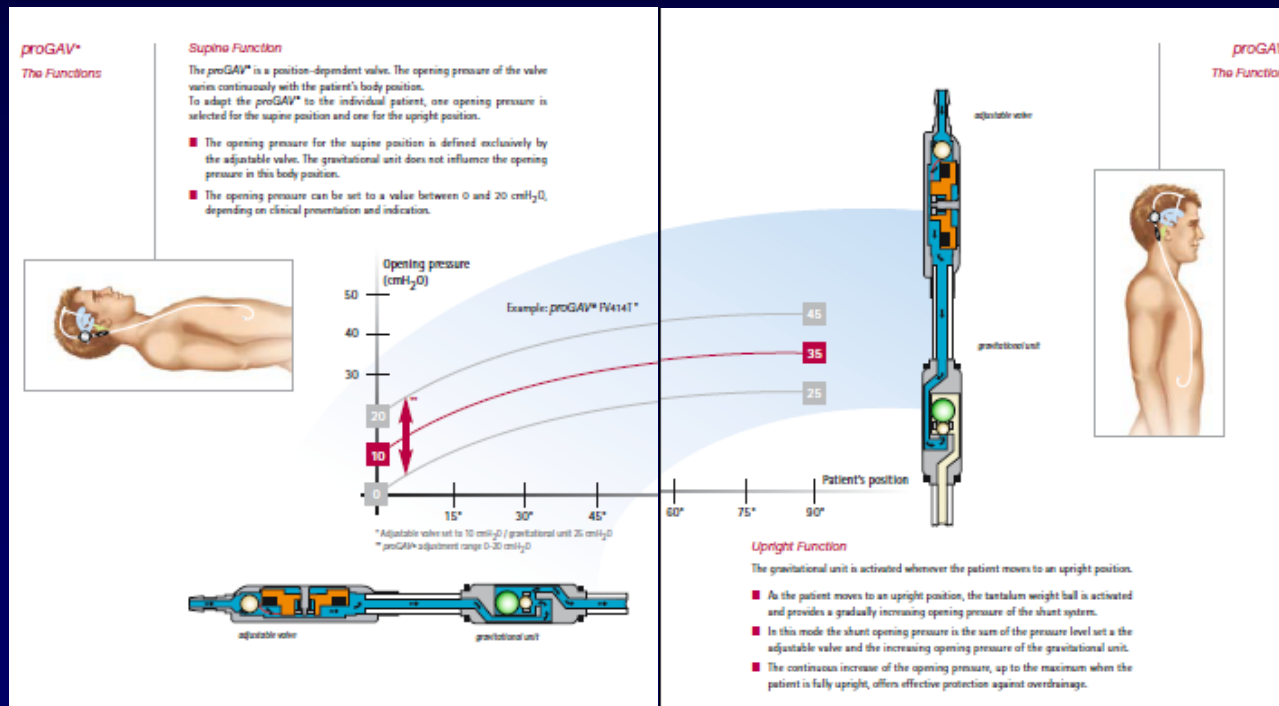
Hydrostatic valves

- valves with ASD (anti-siphon device) – on/off



Valves

Hydrostatic valves – gravity assisted valves



Introduction of a shunt

- **Careful planning**
- **Patient characteristics : CSF-free of blood, infection; skin-healthy, free of infection**
- **Appropriate ventricular access-larger ventricle, shortest path, catheter away from choroid plexus, guidance (ultrasound, image guidance available if needed).**

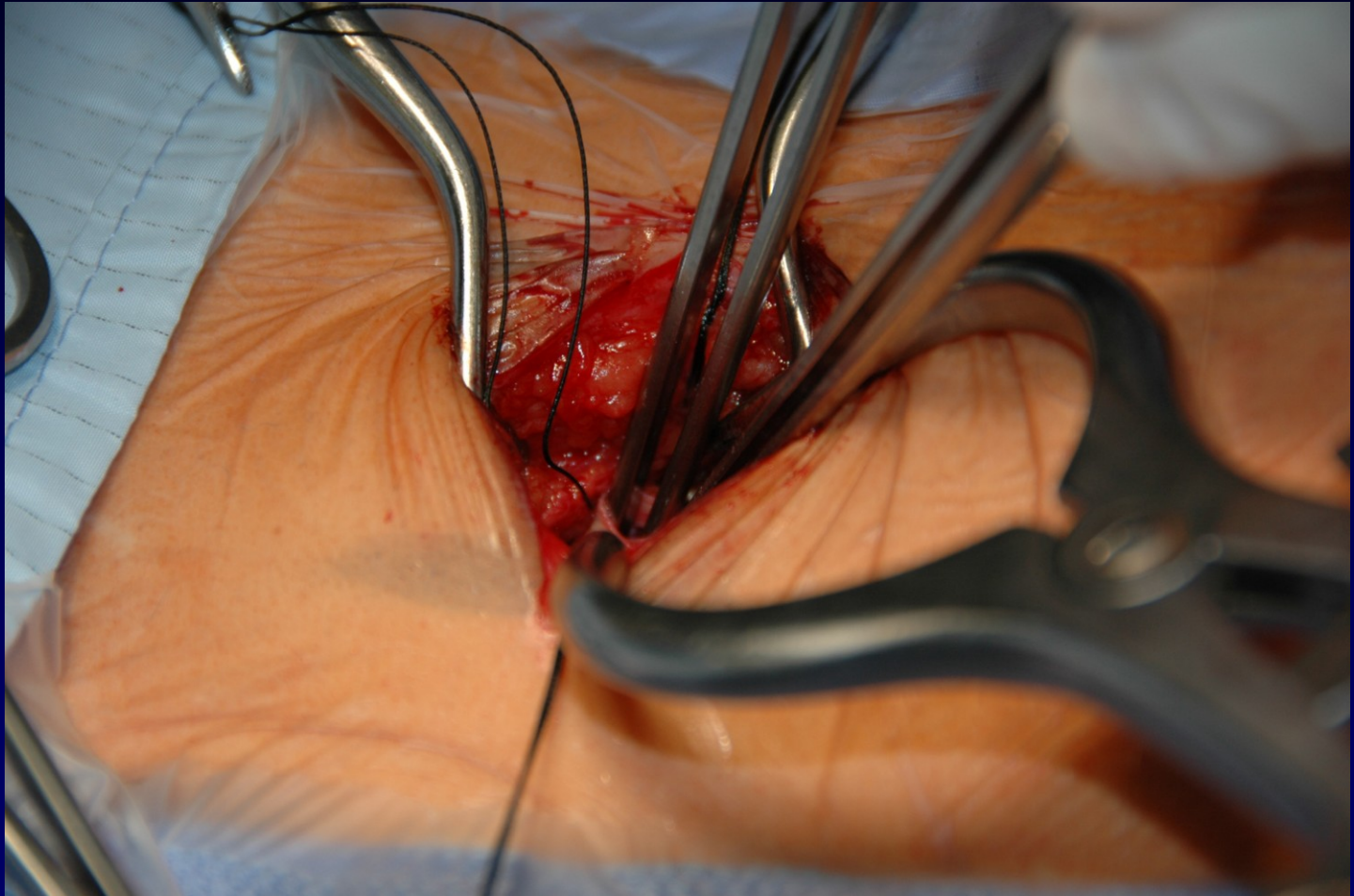
Introduction of a shunt

- **Maximum vigilance should be dedicated to prevent shunt infection: procedure carried out early in the morning at the beginning of the surgical schedule; surgical team reduced in the operating room;**
- **Shunt passer trajectory straight or intervening incision, awareness of skull defects (i.e., post fossa craniotomy), away from central lines, tracheostomy, etc.**
- **Good surgical technique is the best way to decrease shunt complications.**

VP shunt surgery



VP shunt surgery



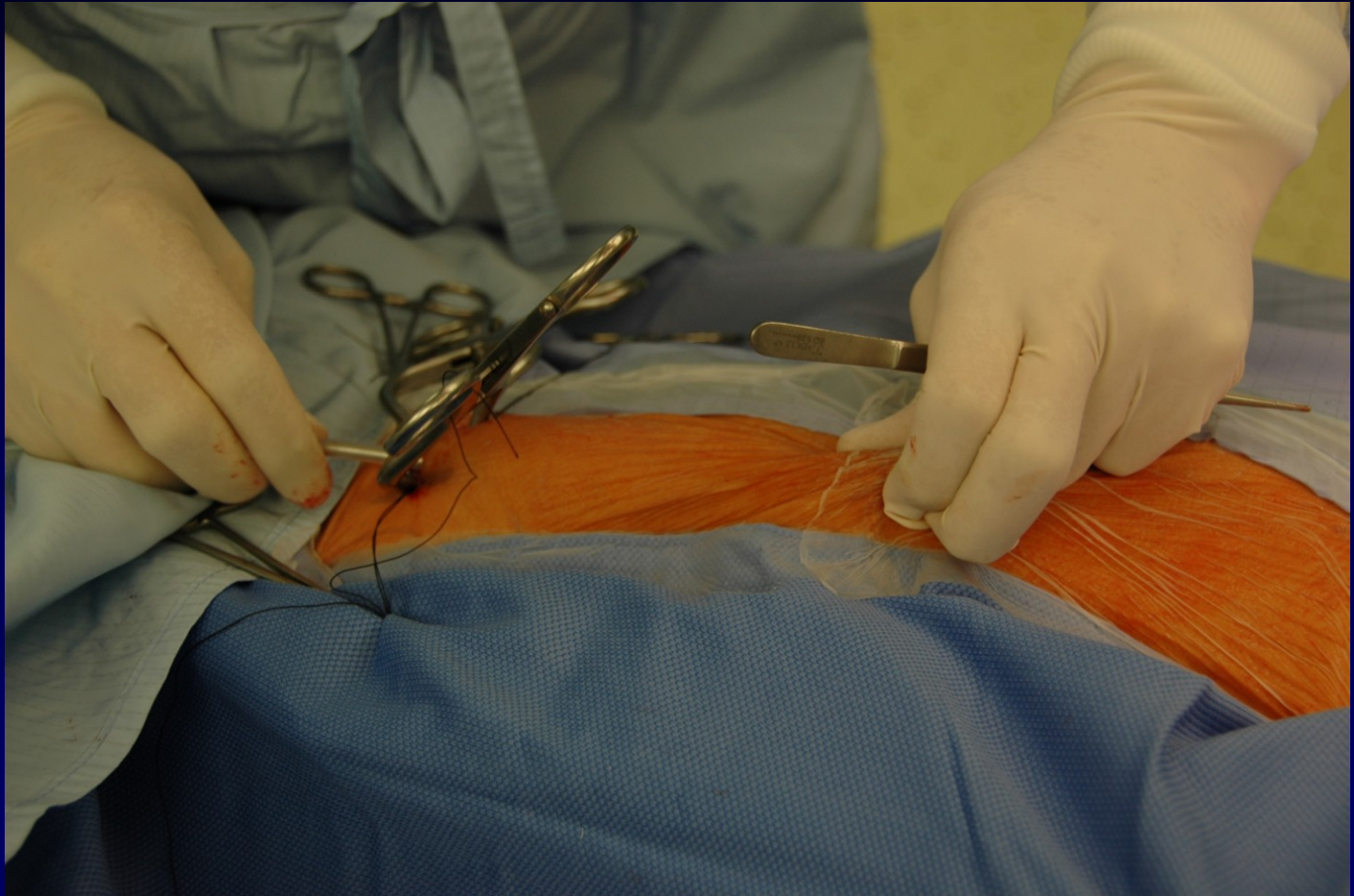
VP shunt surgery



VP shunt surgery



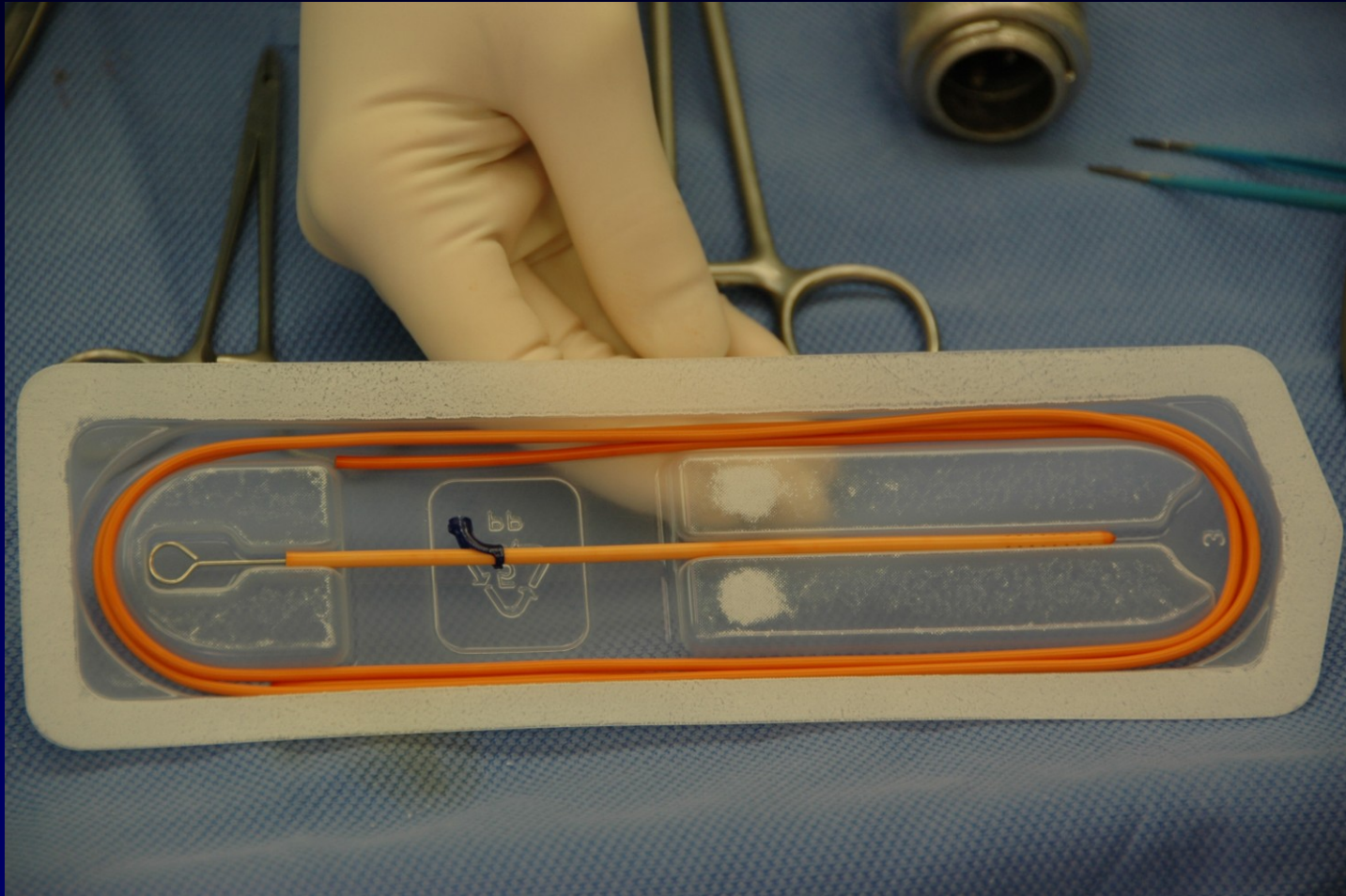
VP shunt surgery



VP shunt surgery



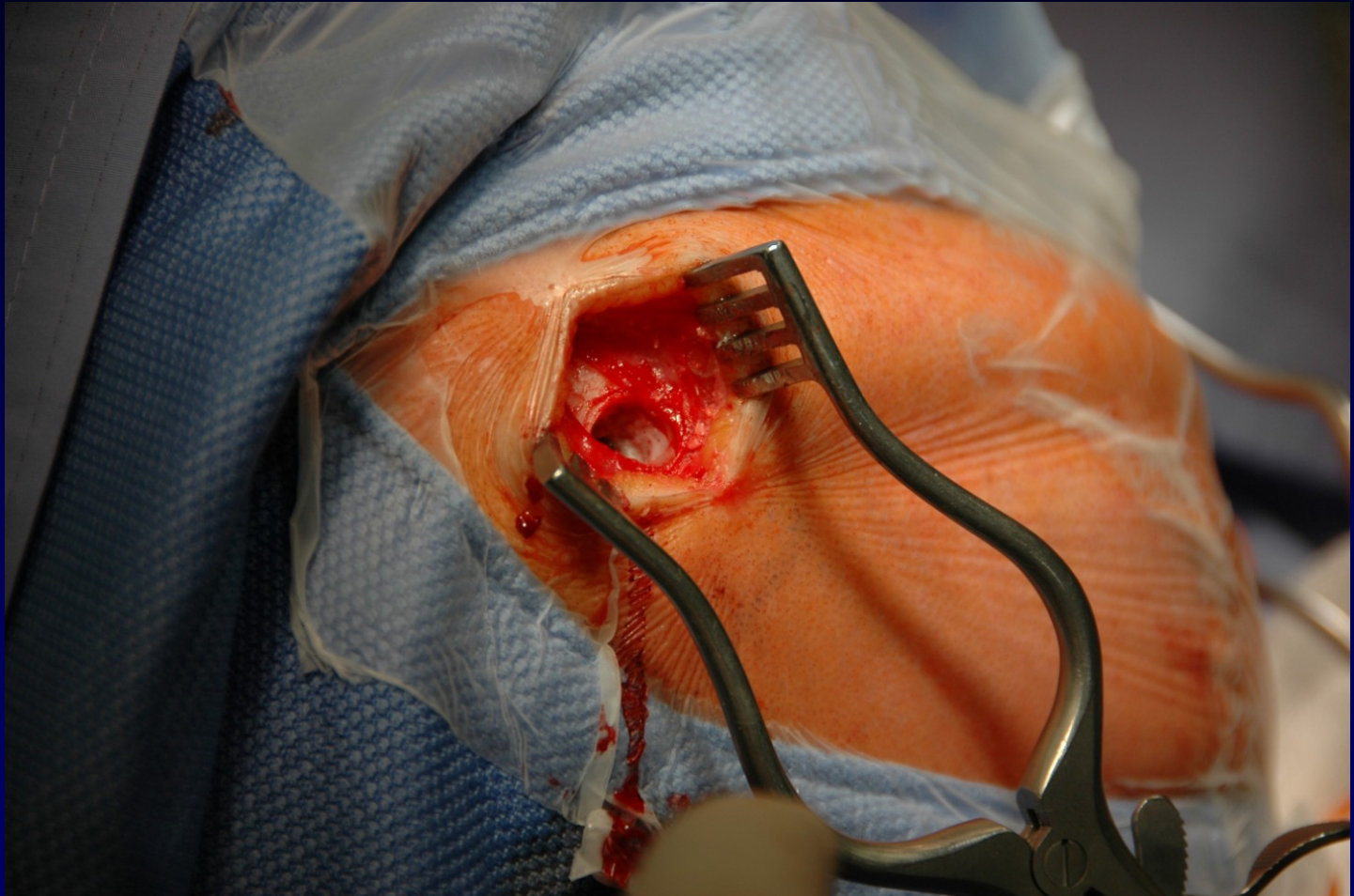
VP shunt surgery



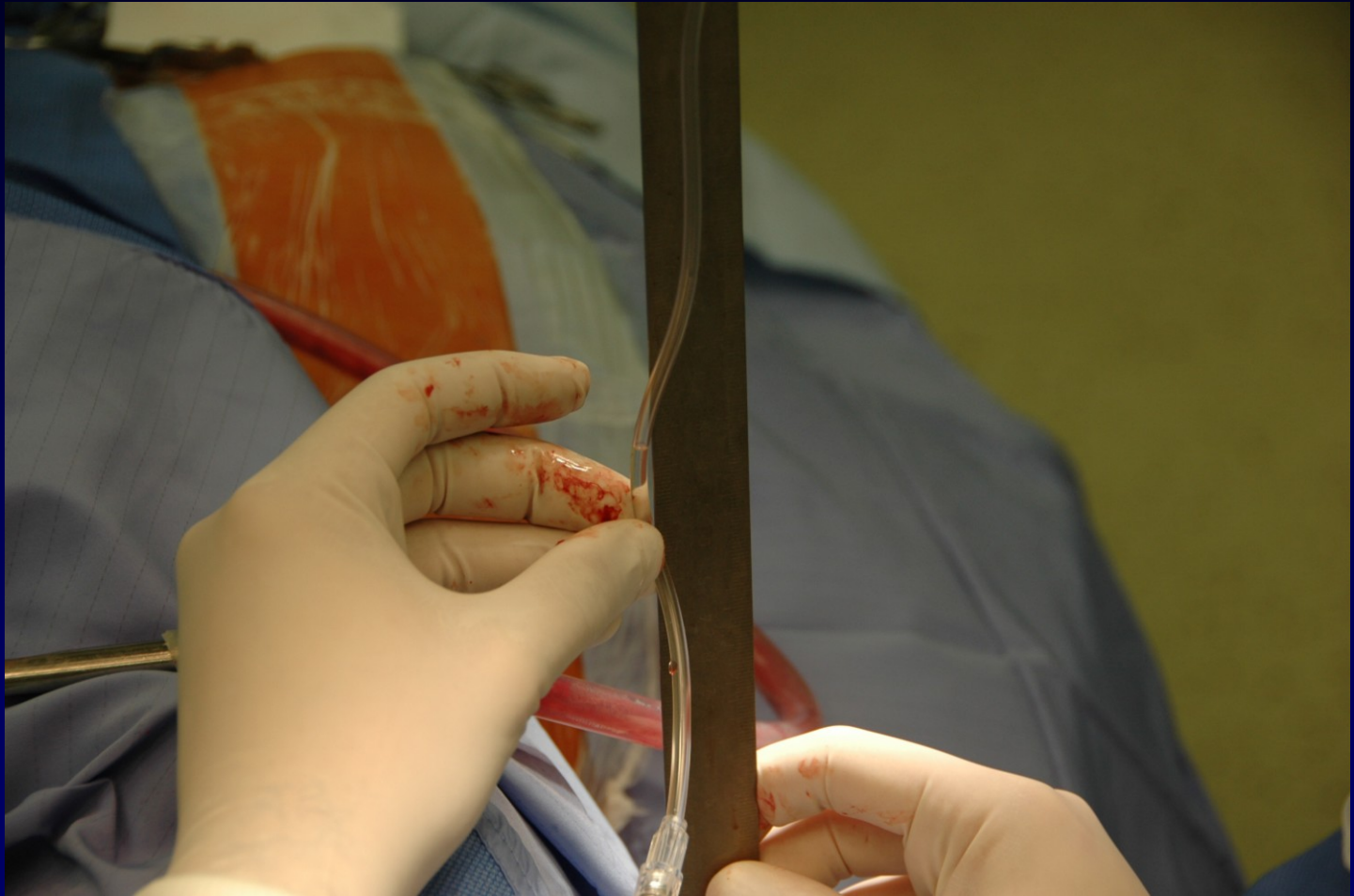
VP shunt surgery



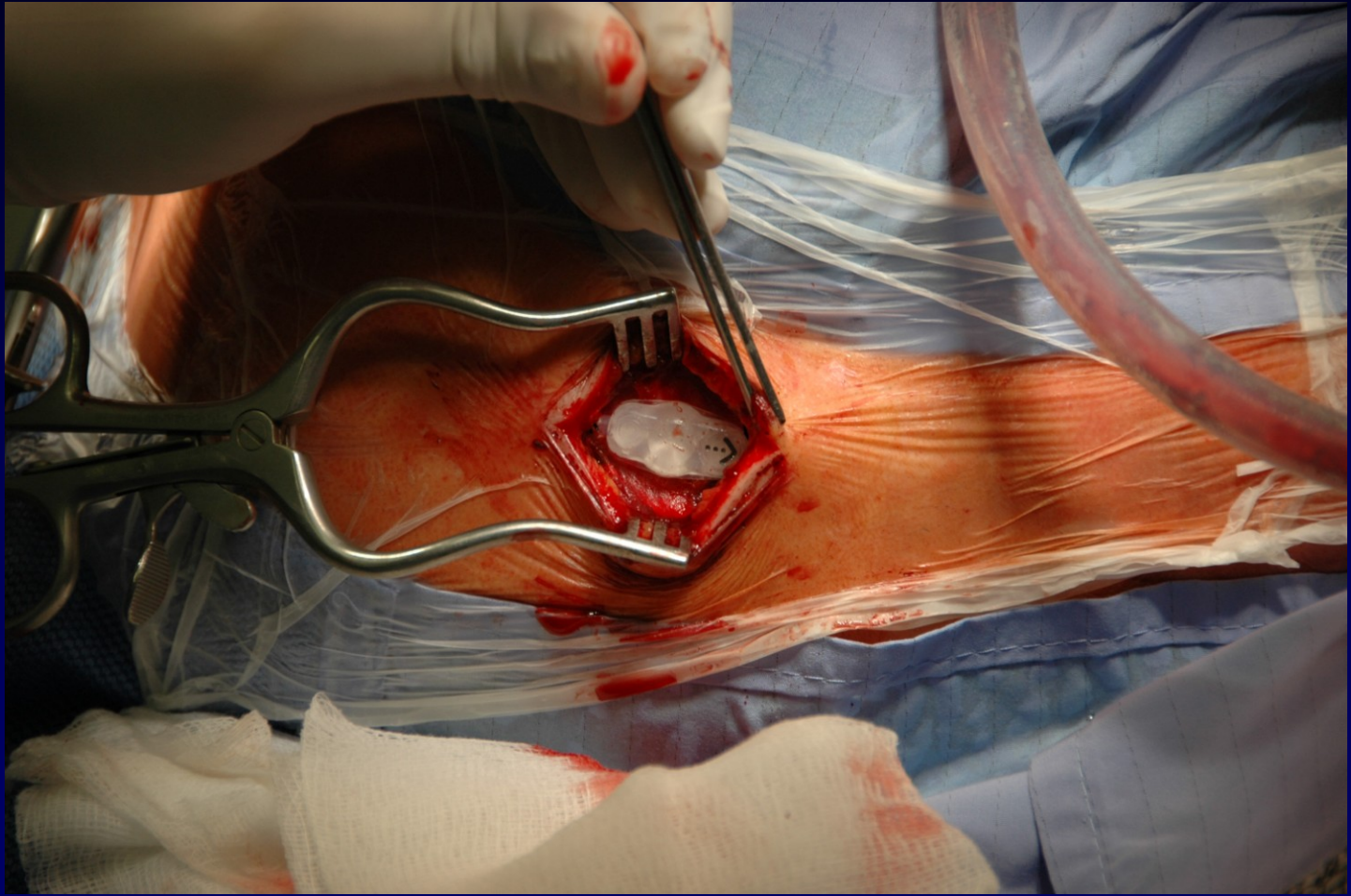
VP shunt surgery



VP shunt surgery



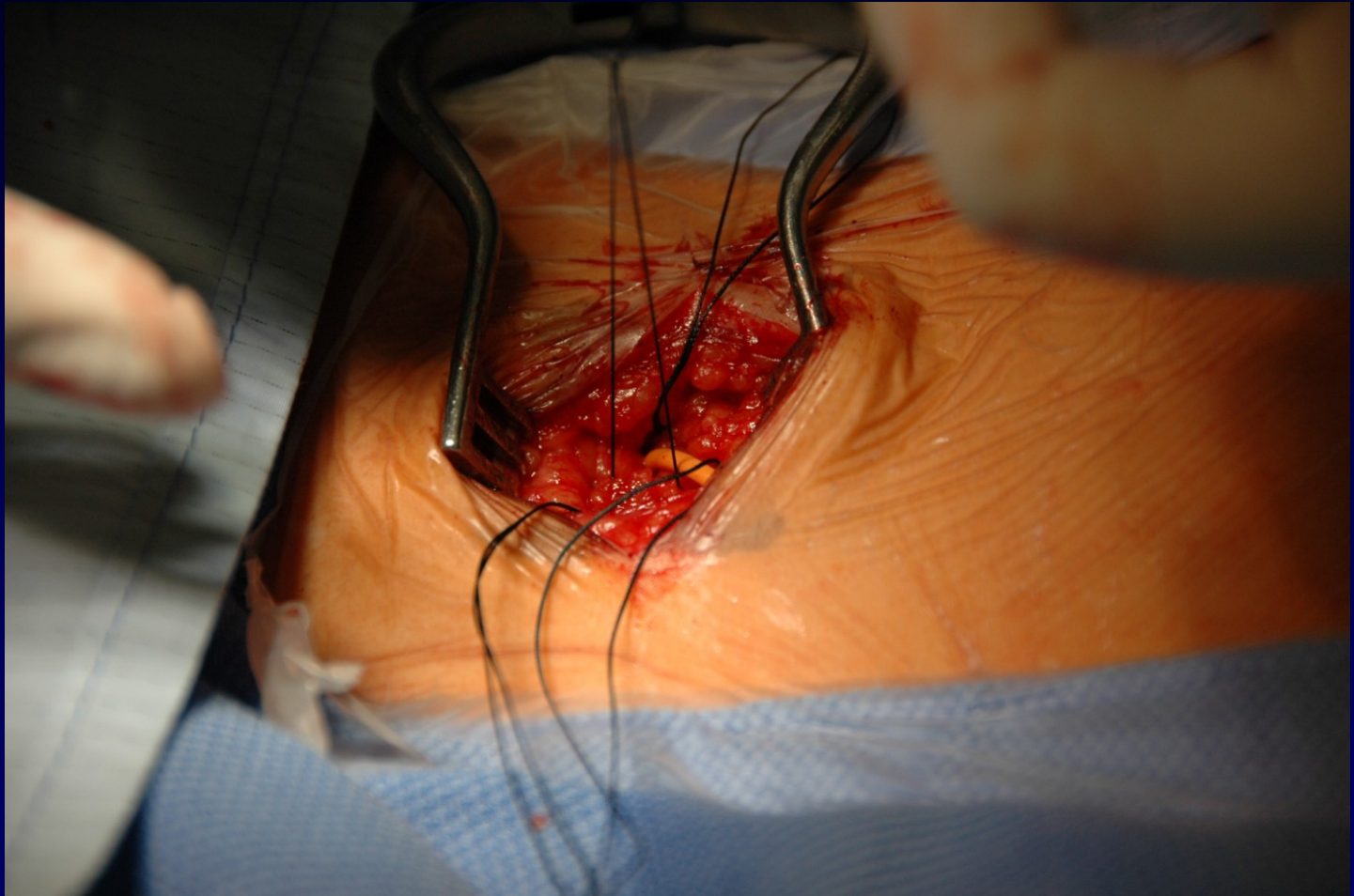
VP shunt surgery



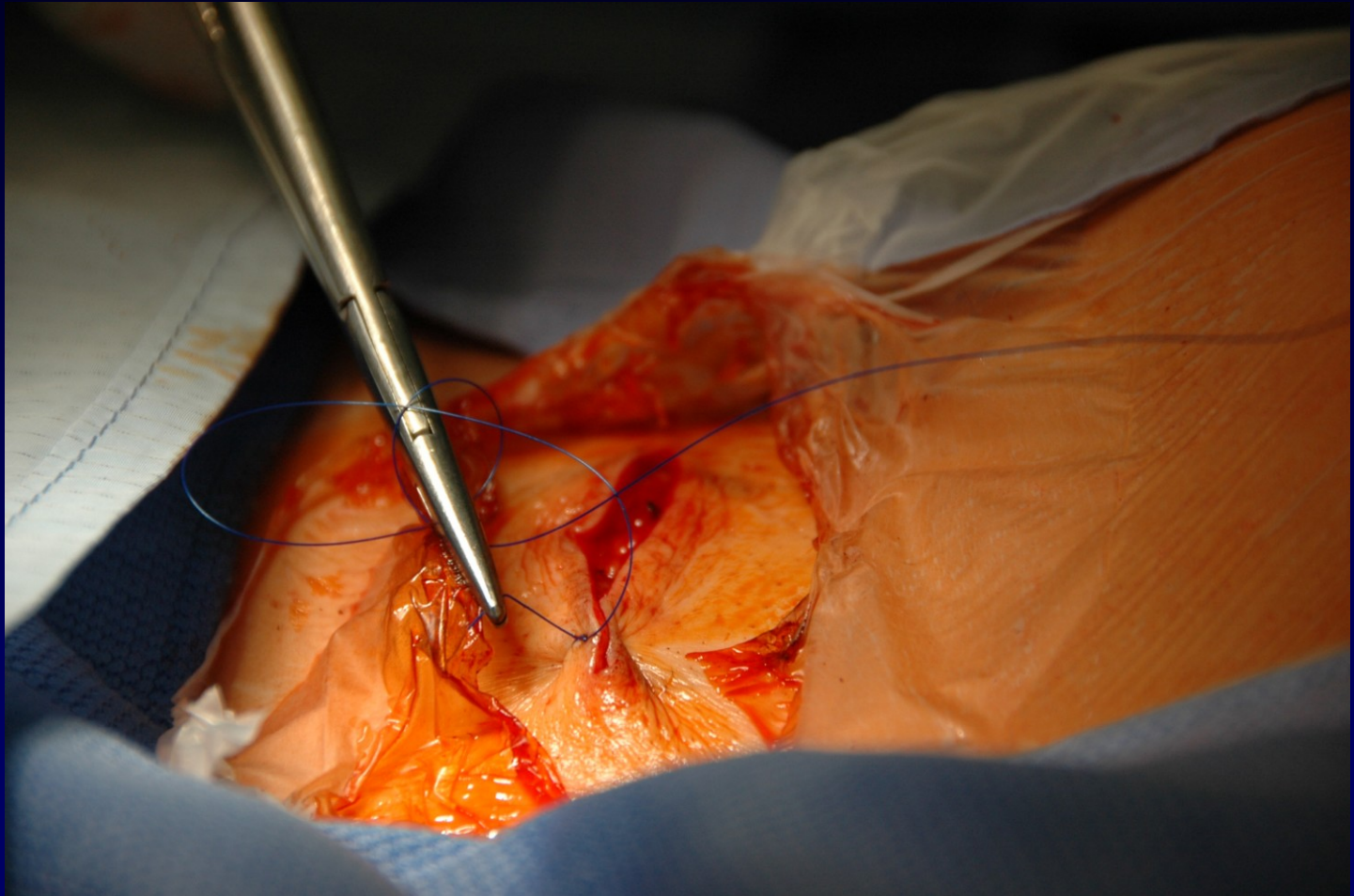
VP shunt surgery



VP shunt surgery



VP shunt surgery



VA shunt surgery

Distal catheter is placed in the atrium of the heart.

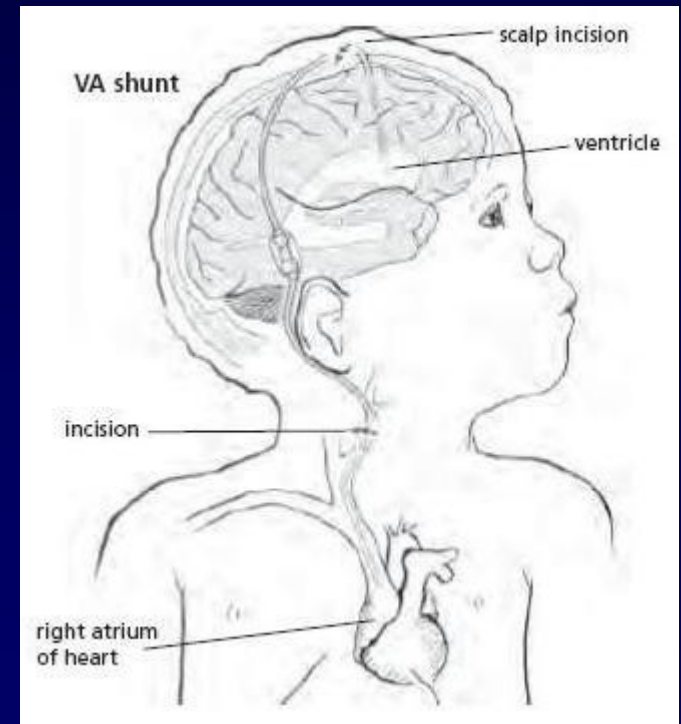
Possibilities:

- facial vein
- puncture of the VJl
- sonography guided puncture

Correct placement:

ECG changes – pulmonale wave

Possible in local anesthesia



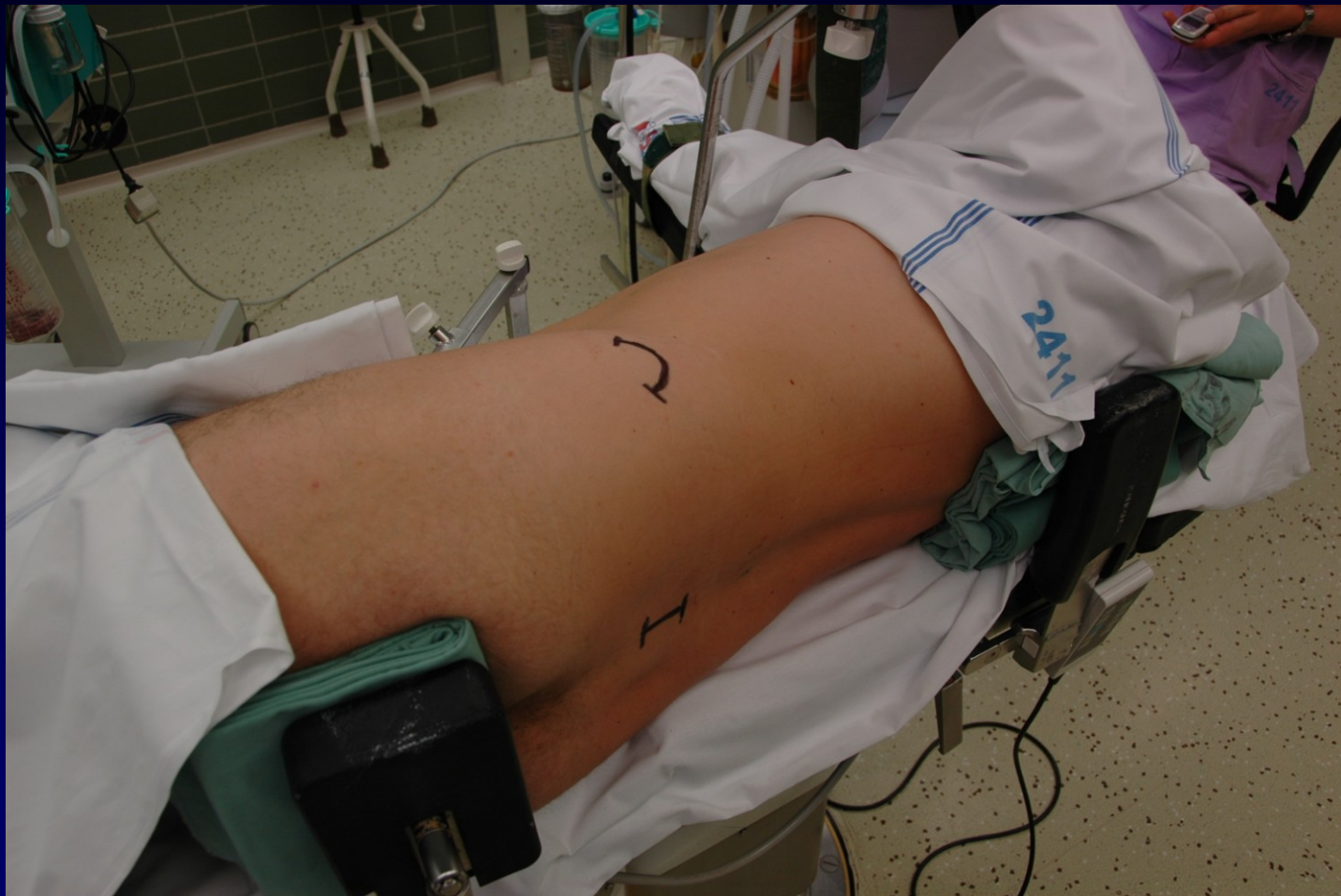
LP shunt surgery

- **Only in case of communicating hydrocephalus**
- **Preferred indications:**
 - **idiopathic or secondary intracranial hypertension (pseudotumor cerebri)**
 - **slit ventricle syndrome**

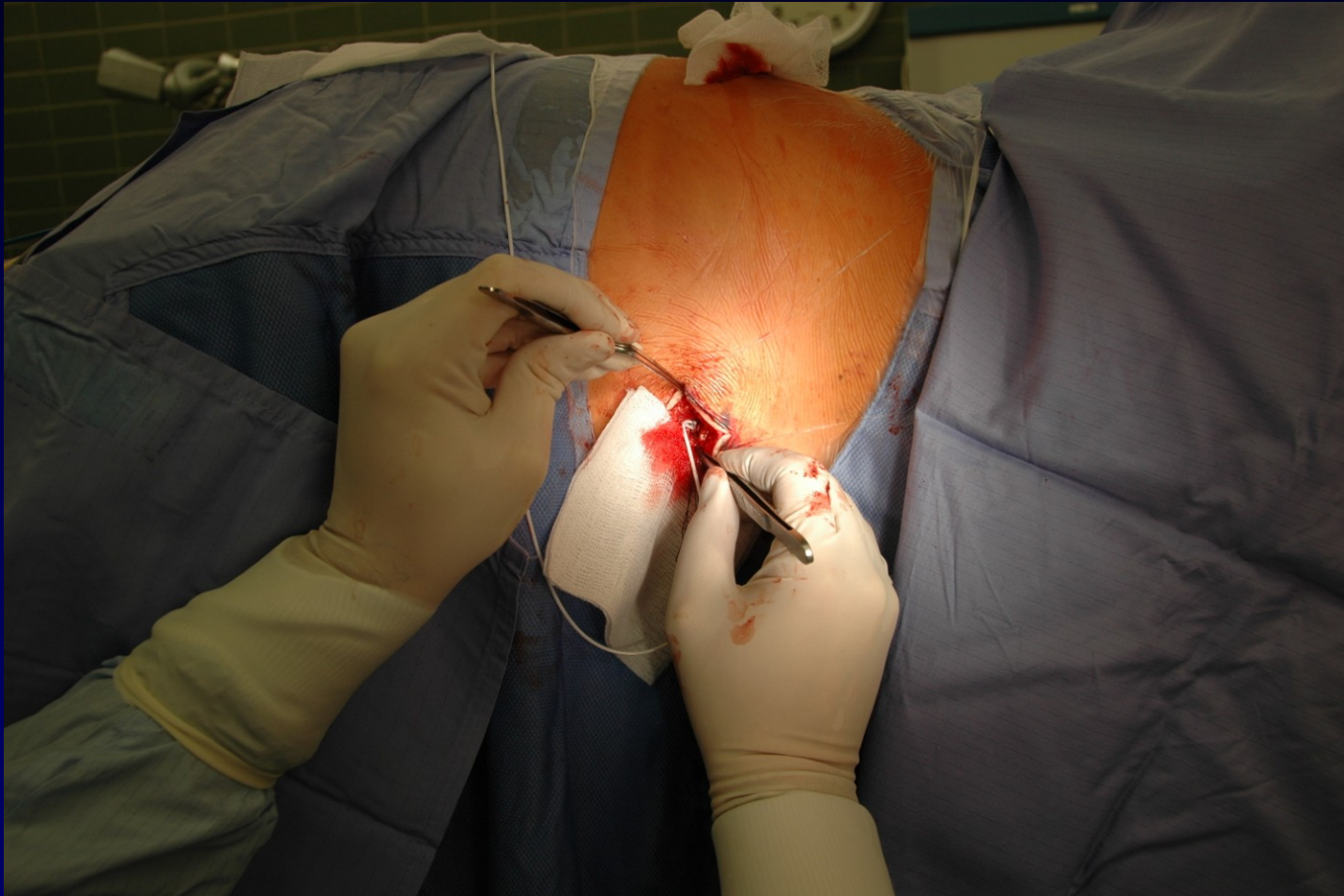
LP shunt surgery



LP shunt surgery



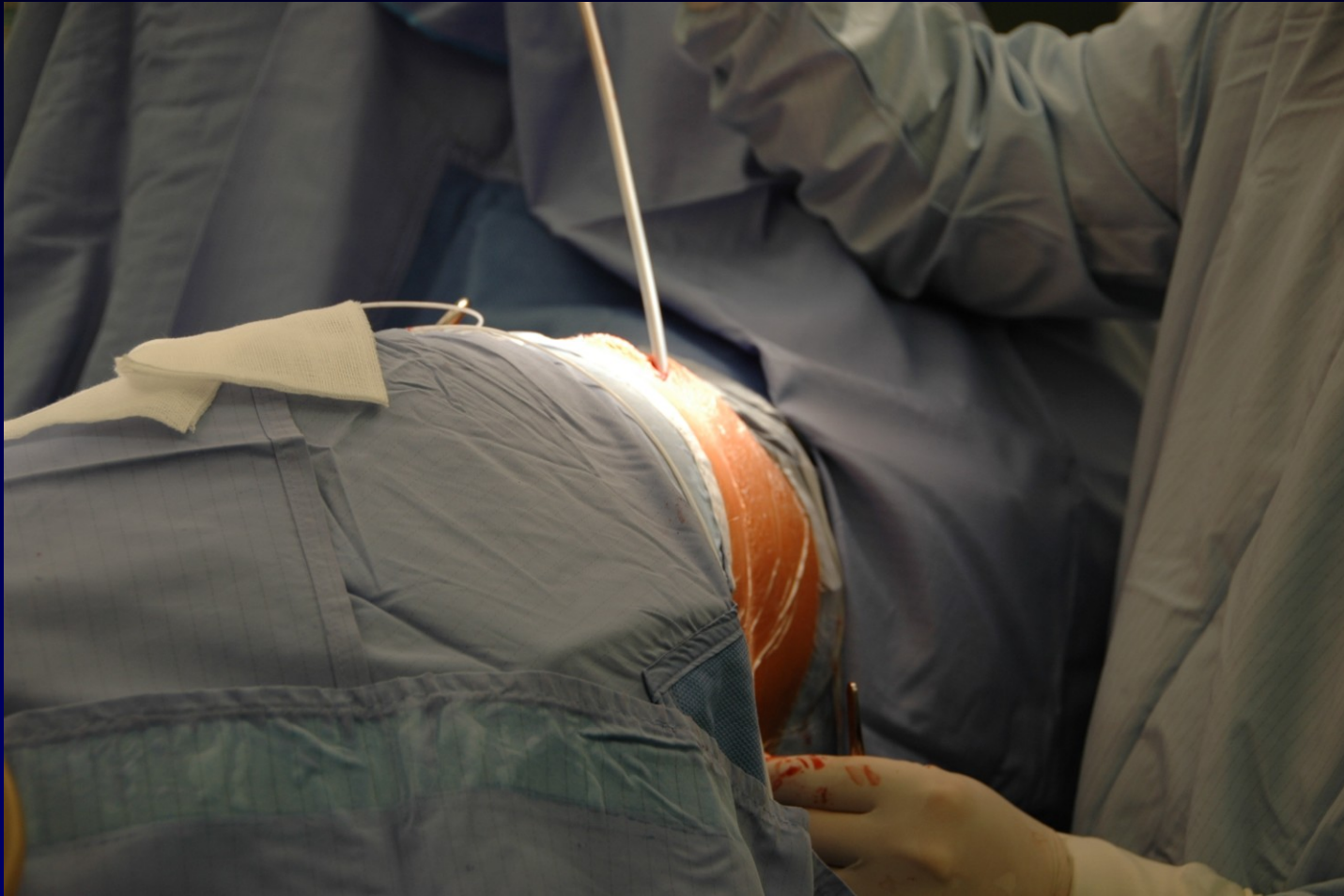
LP shunt surgery



LP shunt surgery



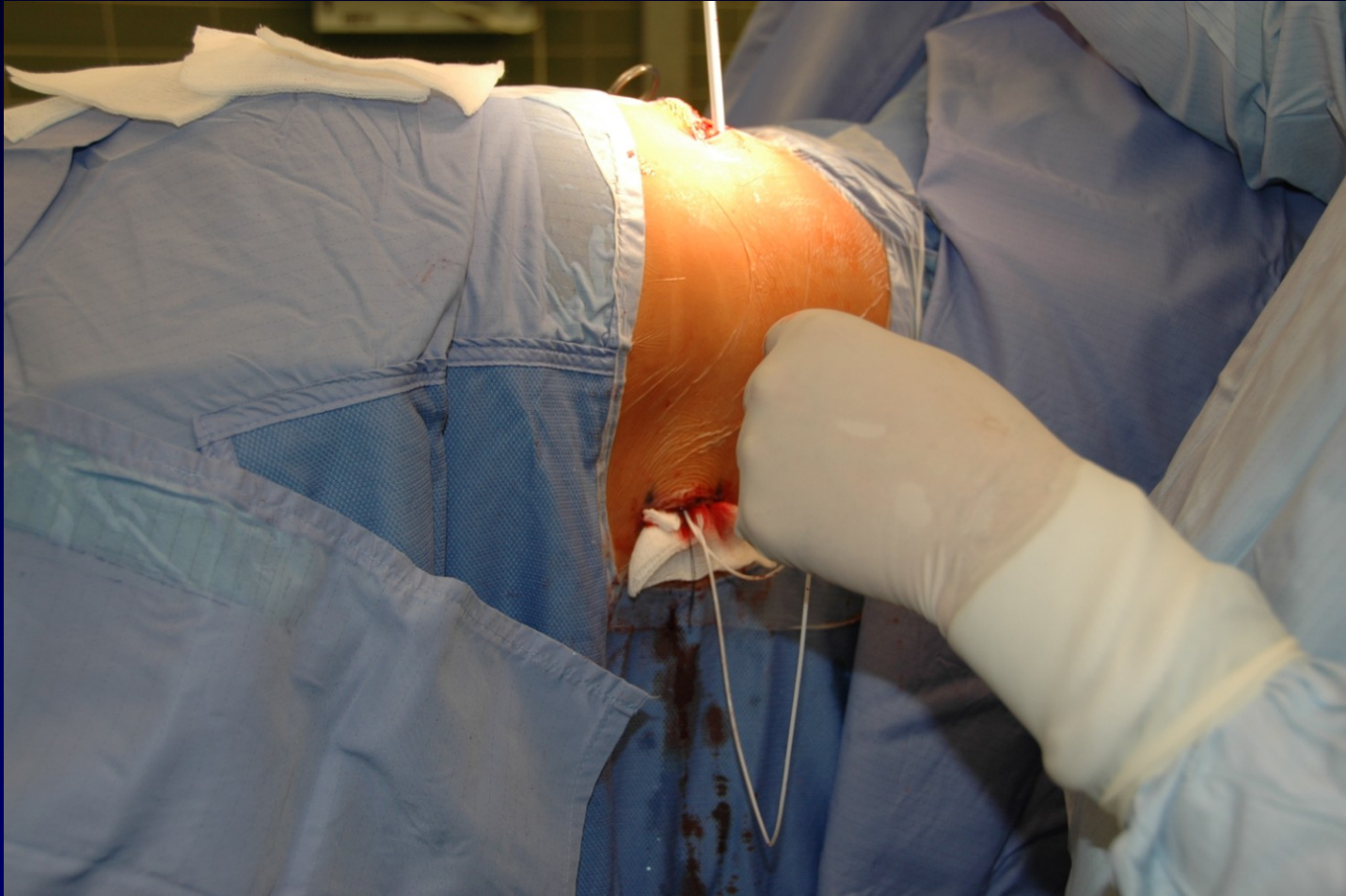
LP shunt surgery



LP shunt surgery



LP shunt surgery



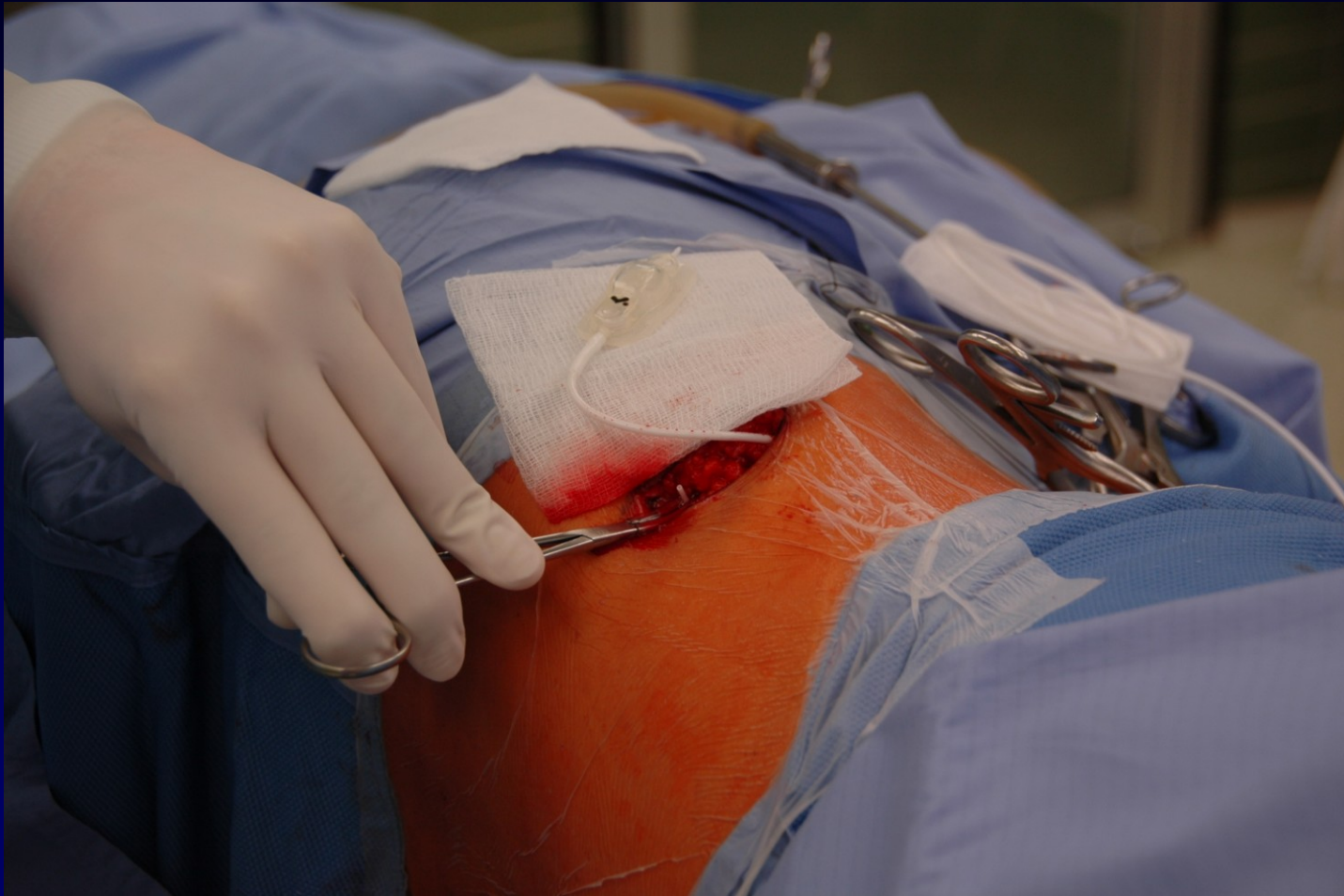
LP shunt surgery



LP shunt surgery



LP shunt surgery



LP shunt surgery



LP shunt surgery



Laparoscopy

- lesser trauma to the abdominal wall and peritoneum
- possibility of performing adhesiolysis and exquisite visualization of the peritoneal cavity, with in situ testing of catheter function
- lower risk of intraabdominal adhesions than laparotomy
- diagnosis of abdominal pain
- revision surgery
- primary placement

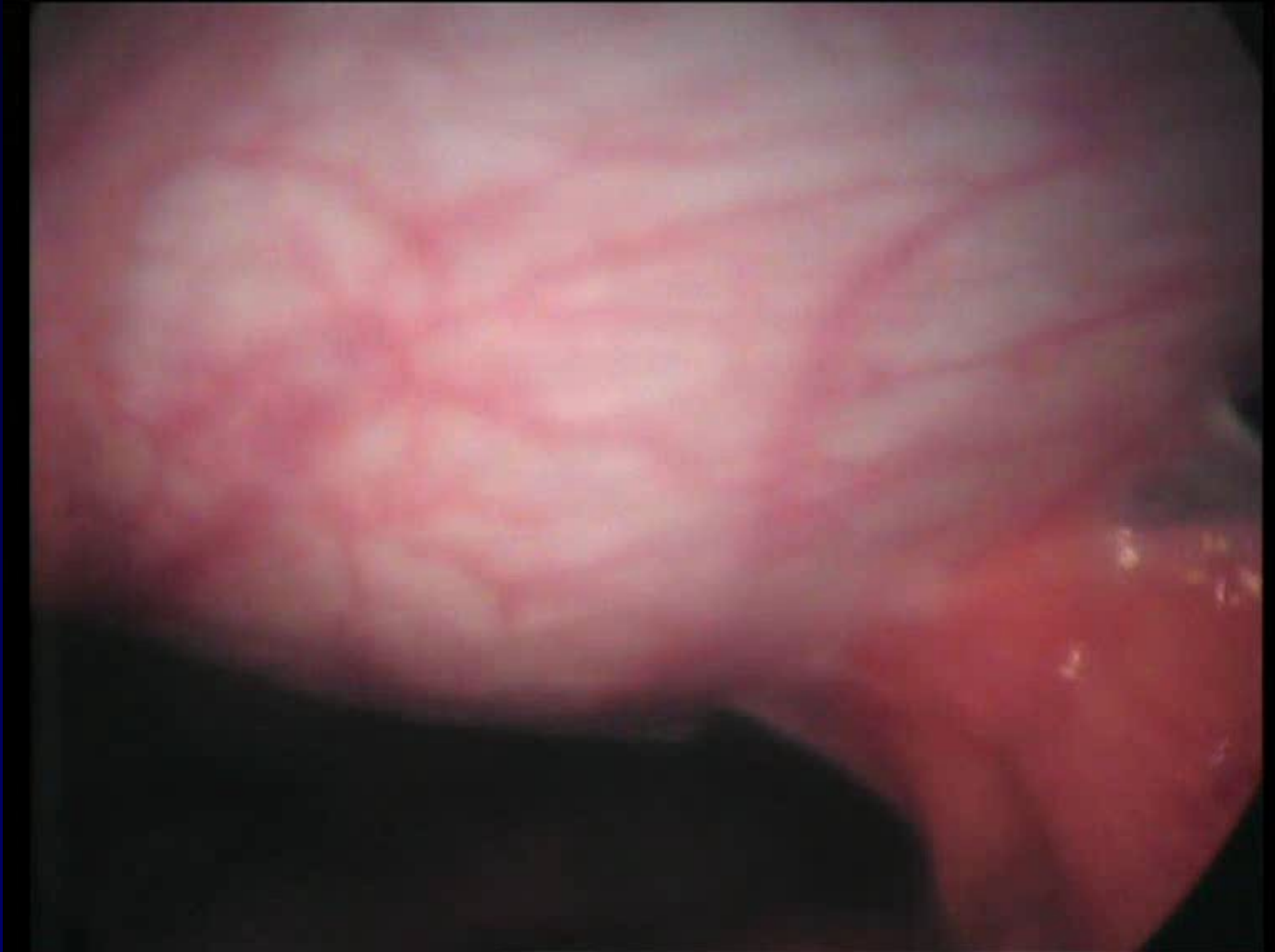


Laparoscopy

- Laparoscopy is safe even without VP catheter clamping and with only routine anesthetic monitoring (Al-Mufarrej et al, 2005).
- Risk of retrograde failfure minimal even with intraabdominal pressure as high as 80 mm Hg (Al-Mufarrej et al, 2005).



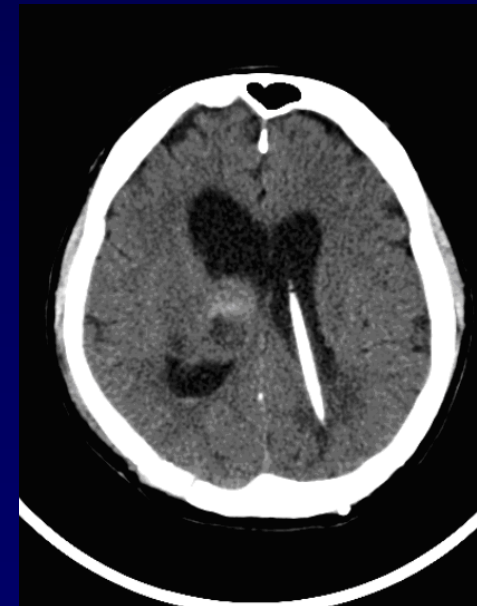
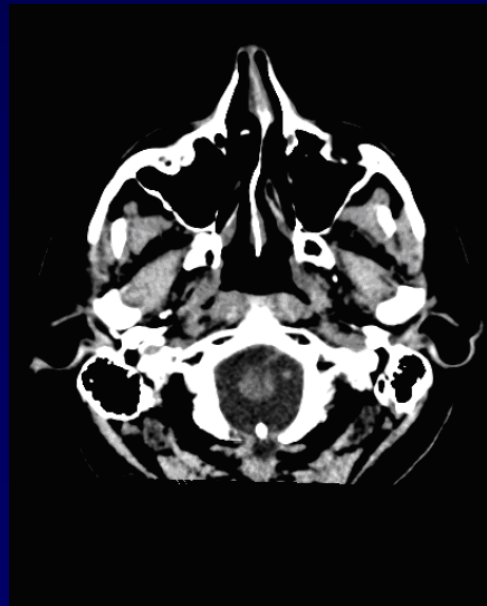
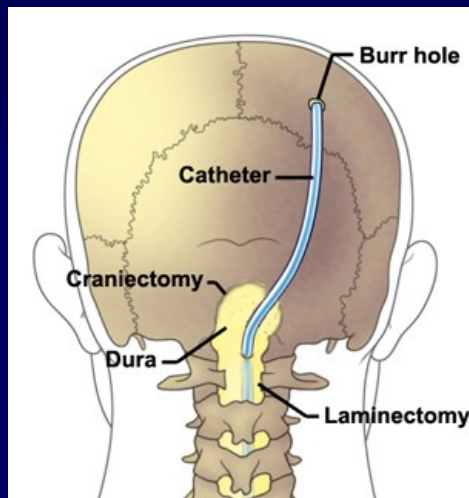
Laparoscopy



Torkildsen shunt

A ventriculocisternal shunt that diverts the cerebrospinal fluid flow from one of the lateral ventricles to the cisterna magna.

Best indication is noncommunicating hydrocephalus with a lesion in and around the third and fourth ventricles that precludes standard endoscopic approaches.



Shunt complications

- **Undershunting**
- **Overshunting**
 - **Slit ventricles**
 - **Intracranial hypotension**
 - **Subdural hematomas**
 - **Stenosis or occlusion of sylvian aqueduct**
 - **Craniosynostosis and microcephaly (controversial)**
- **Infection**
- **Seizures**
- **Obstruction**
- **Disconnection**
- **Others**

Shunt complications

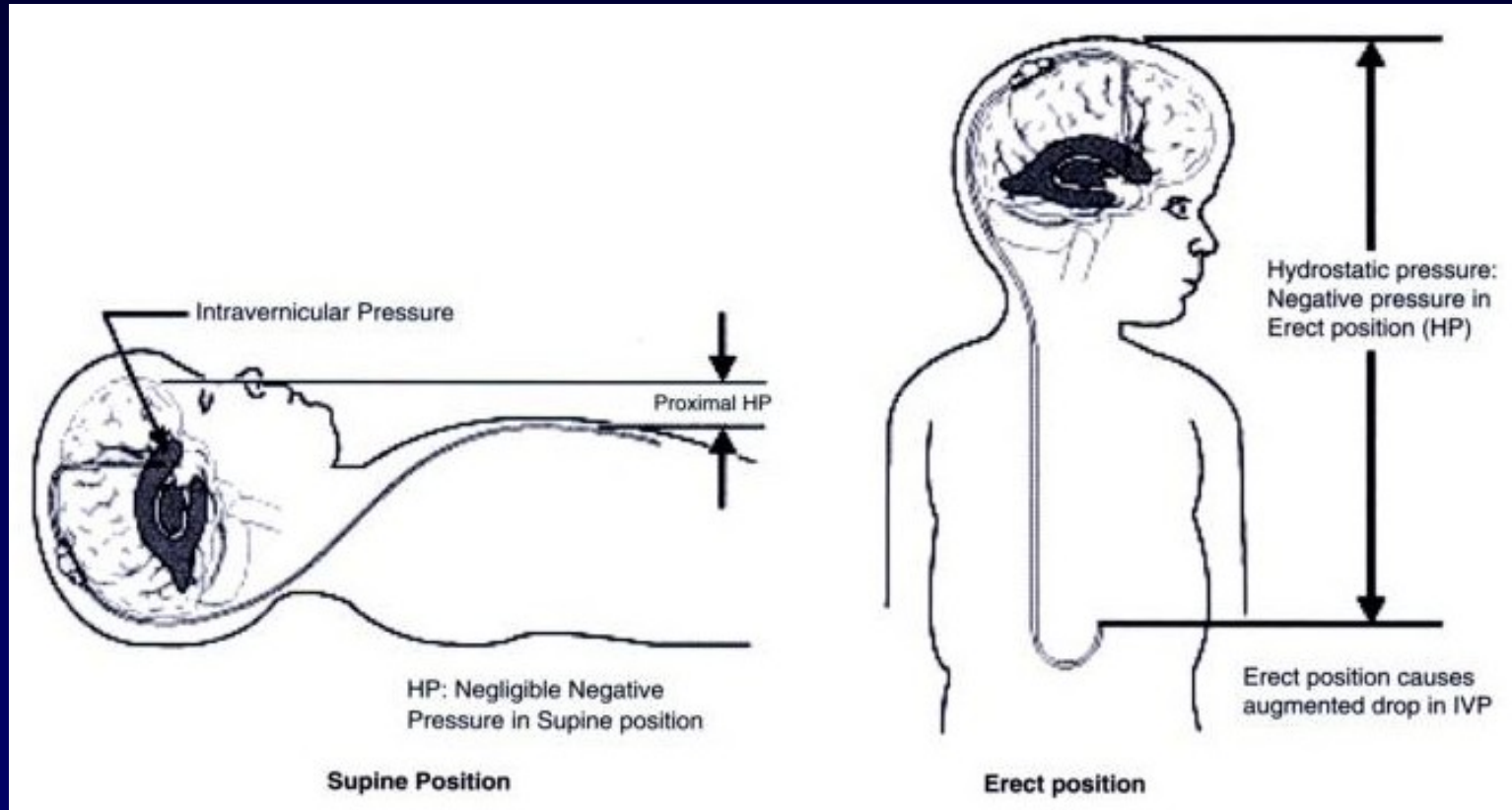
Undershunting

- Verify the function of a shunt
- Adjust the valve (lower pressure)
- NPH patients



Shunt complications

Overshunting



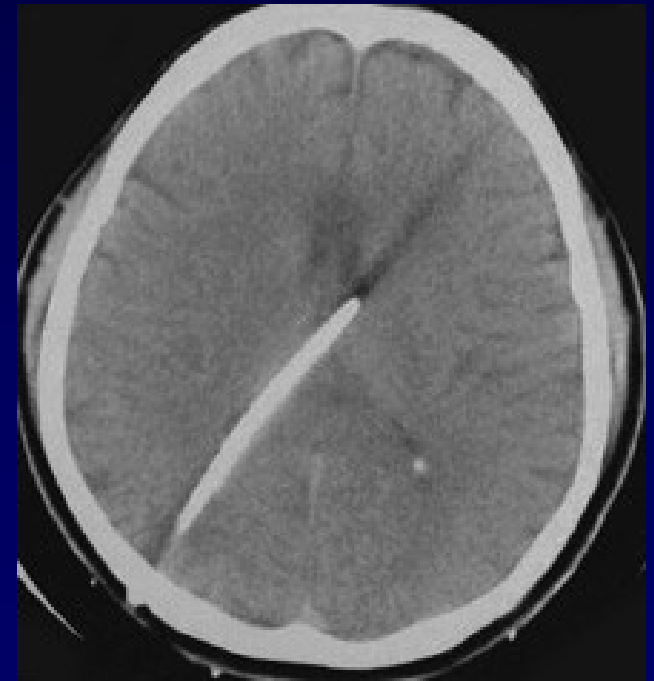
Siphoning effect

Shunt complications

Overshunting

– Slit ventricles

- Most are asymptomatic
- Slit ventricle syndrome – intermittent or permanent shunt occlusion (ventricles cannot expand because of subependymal occlusion)



Shunt complications

Overshunting

- Intracranial hypotension
 - Relieved by recumbency
 - Therapy – adjust the valve (higher pressure), change the valve, ASD etc.

Shunt complications

Overshunting

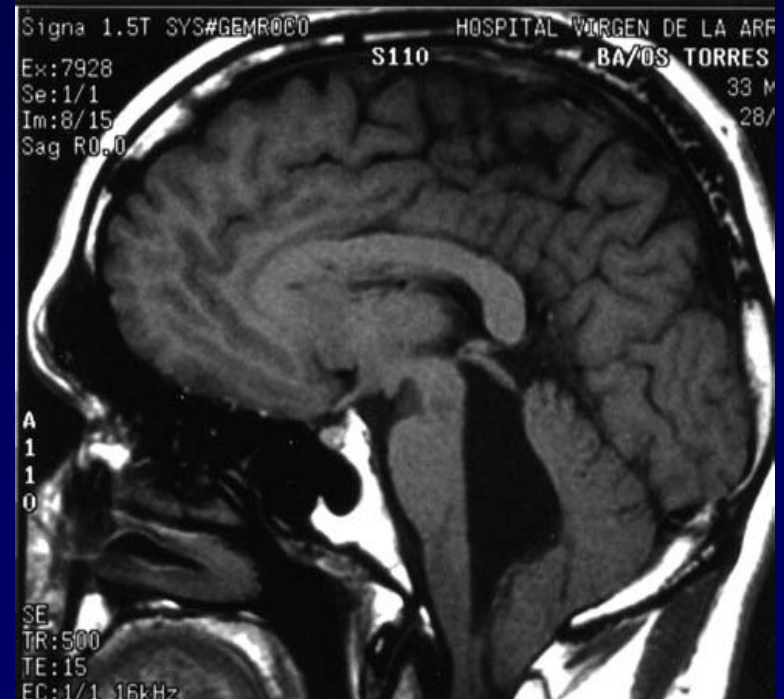
- Subdural hematomas



Shunt complications

Overshunting

- Stenosis or occlusion of sylvian aqueduct, trapped fourth ventricle



Shunt complications

- **Complications that may occur with any shunt:**
 - Obstruction (proximal, valve, distal)
 - Disconnection, break at any point
 - Hardware erosion
 - Infection, seizures etc.
- **VP shunts (abdominal complications – peritonitis, ascites, hydrocele, tip migration, intestinal obstruction volvulus, intestinal strangulation etc.)**
- **VA shunts (septicemia, shunt embolus, pulmonary hypertension, shunt nephritis etc.)**
- **LP shunts (lumbar nerve root irritation, adhesions etc.)**

