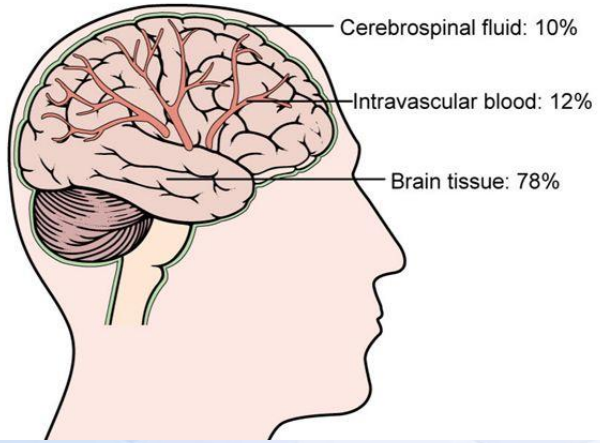
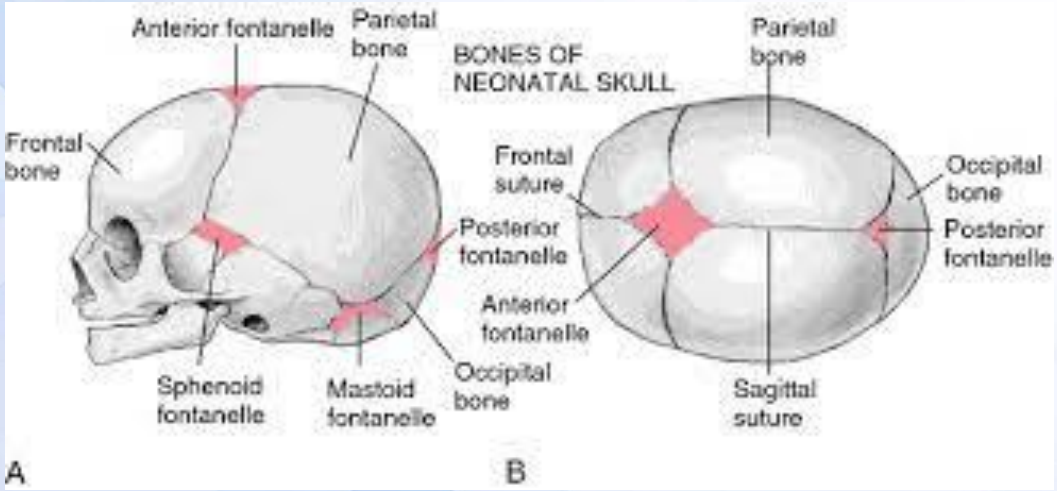
Neurosurgical approaches to size and shape of skull

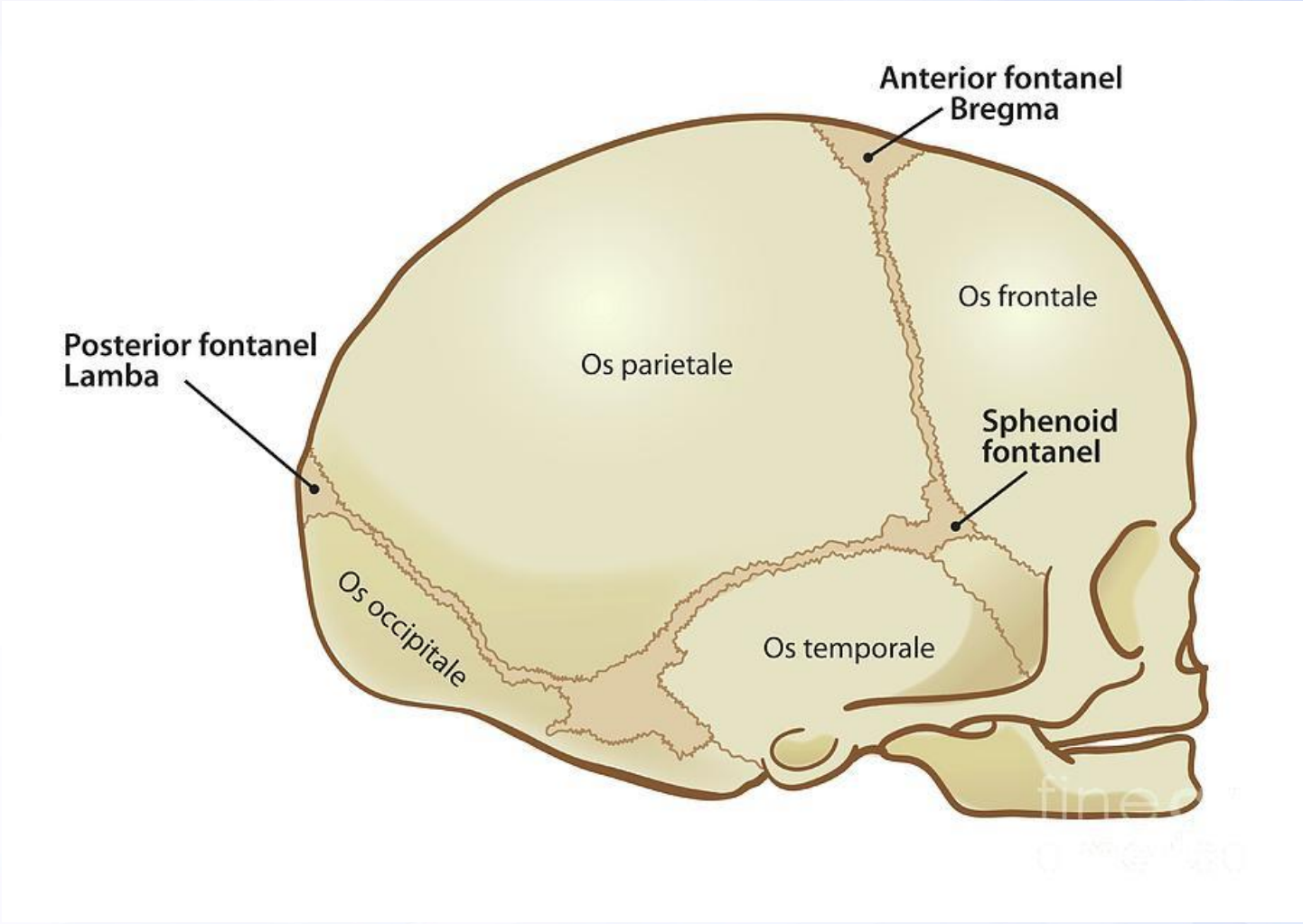
Fars Pediatric Association

Intracranial Pressure (ICP)



Skull anatomy





Timing of Closure of Sutures and Fontanelles

Type of suture/fontanelle	*Time to closure*
Metopic suture	Nine months to two years (may persist into adulthood)
Coronal, sagittal, lambdoid sutures	40 years
Anterior fontanelle	Nine to 18 months
Posterior fontanelle	Three to six months
Anterolateral fontanelle	Three months
Posterolateral fontanelle	Two years

Adapted with permission from Aviv Ri, Rodger E Hall CM. Craniosynostosis. Clin Radiol 2002;57:94.

Mature suture closure occurs by 12 years of age, but completion continues into the third decade of life and beyond.

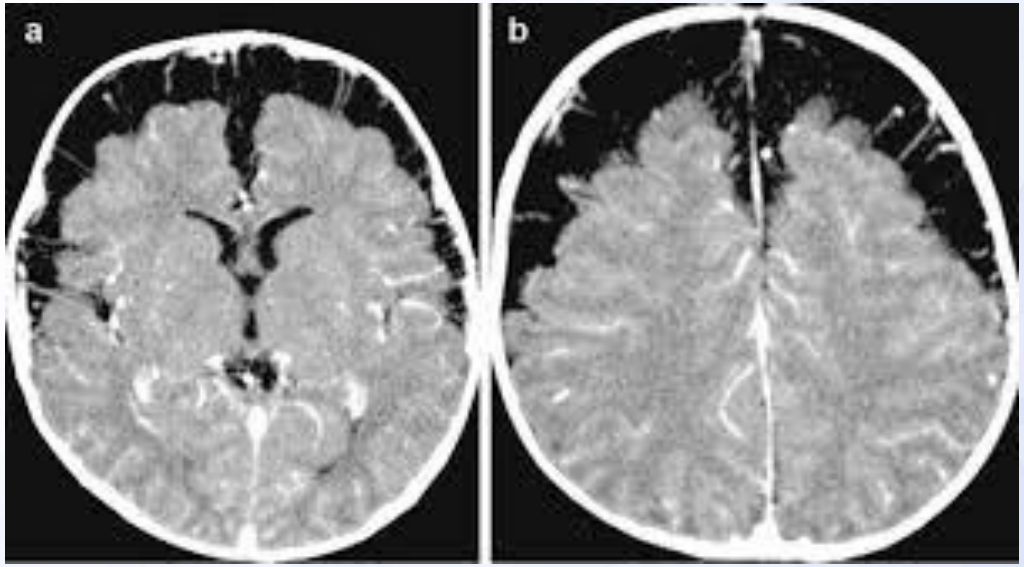
Disorders of Cranial Volume and Shape

The brain, cerebrospinal fluid (CSF), and blood are the three intracranial compartments that determine the size of the skull during infancy

Less important factors contributing to head size are the thickness of the skull bones and the rate of their fusion

The intracranial content, the fusion of the sutures, and external forces on the skull determine its shape.

Infants left supine all the time tend to develop flat occiputs. Premature infants resting on one side of the head all the time develop heads with large occipitofrontal diameter (dolichocephaly).

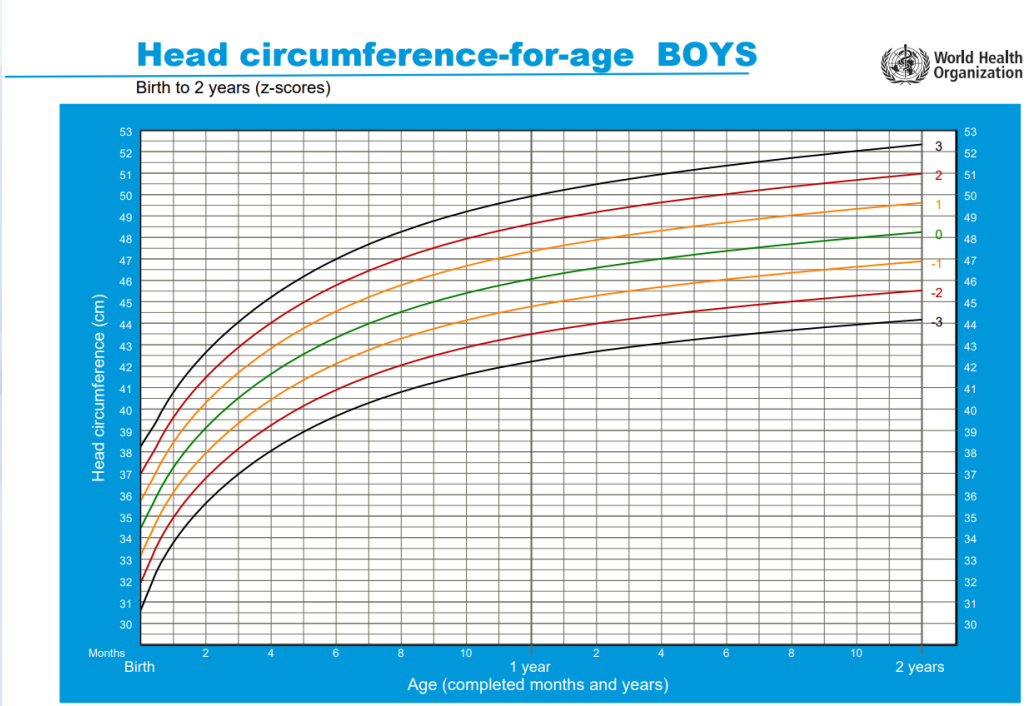


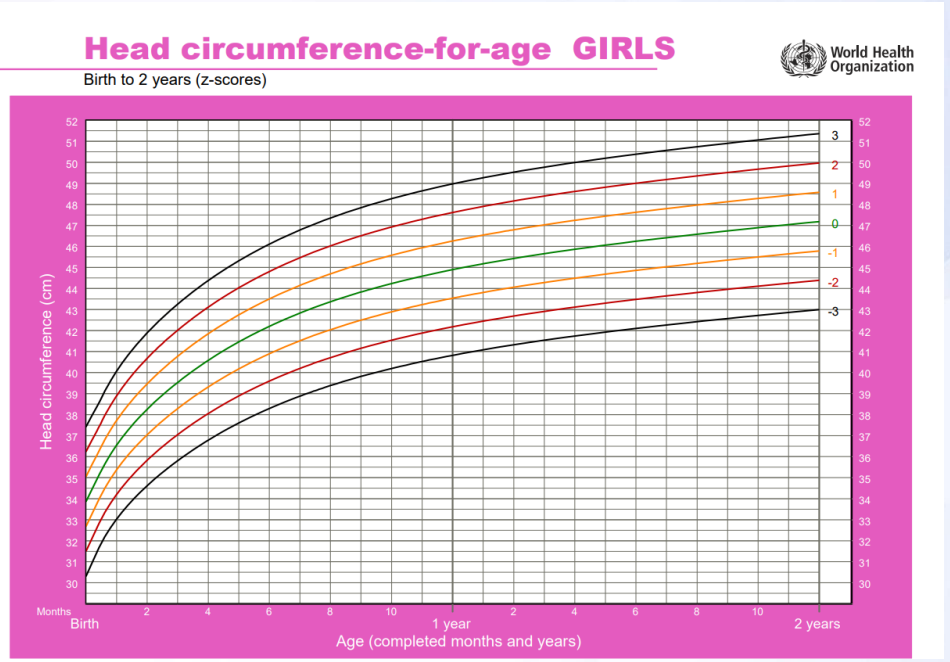
MEASURING HEAD SIZE

measuring the greatest occipitofrontal circumference

- A round head has a larger intracranial volume than an oval head of equal circumference
- A head with a relatively large occipitofrontal diameter has a larger volume than a head with a relatively large biparietal diameter

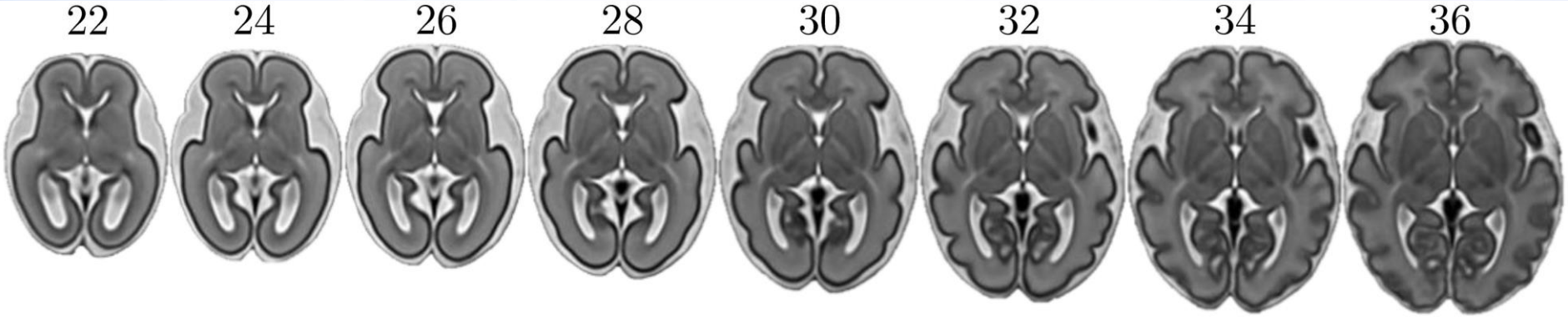
Head Circumference

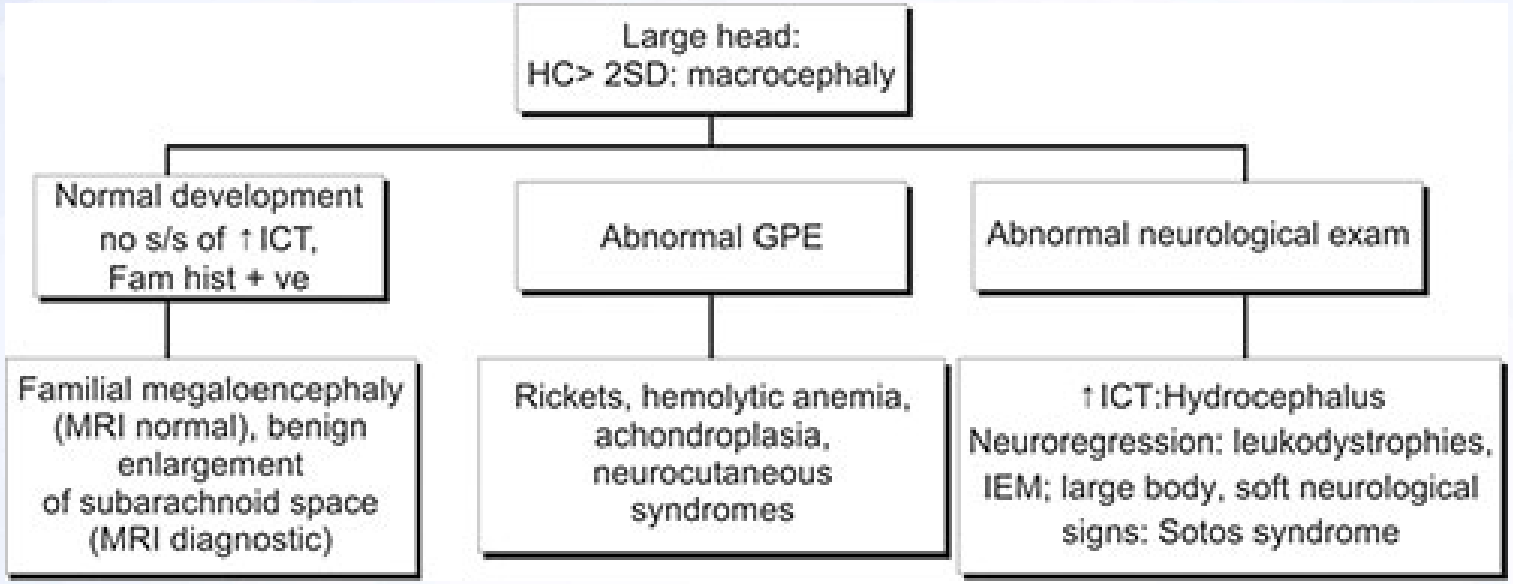




- Head circumference measurements are most informative when plotted over time (head growth).

The rate of head growth in premature infants is considerably faster than in full-term newborns
For this reason





Totally normal child

Disease	Clinical Clues	Tests
Familial megaloencephaly	Family history of large heads. Head circumference normal at birth, progressively increases to reach 98th centile. Normal development and physical examination	MRI normal
Benign enlargement of subarachnoid space/external hydrocephalus	Familial, autosomal dominant, head circumference 90th centile at birth and increases to follow 98th centile, wide open anterior fontanelle, normal development and examination. Due to development delay in reabsorbtion of CSF in arachnoid villi. Predisposed to subdural bleeds with minor trauma.	CT or MRI brain: enlarged subarachnoid space in frontal region > 5.7 mm, in sylvian fissure >7.6 mm

Disease	Management
Familial megaloencephaly	Reassurance
Benign enlargement of subarachnoid space	Reassurance
Aqueductal stenosis	Ventriculoperitoneal shunt, third ventriculostomy
Dandy-Walker cyst	Cystoperitoneal or ventriculoperitoneal or dual shunt
Communicating hydrocephalus in TBM	Ventriculoperitoneal shunt
Posthemorrhagic hydrocephalus in preterms	VP shunt when weight > 1500 gm and CSF RBCs < 1000/cc, CSF protein <500 mg/cc
Chronic subdural effusion	Surgical drainage
Glutaric aciduria I	Lysine restricted diet, supplement with riboflavin and carnitine. Valproate and baclofen may have a role.

MACROCEPHALY

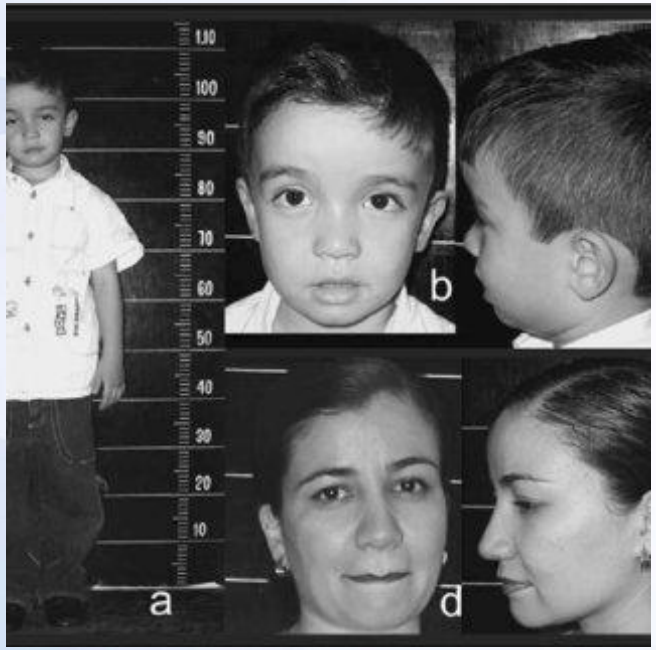
Macrocephaly means a large head, larger than two standard deviations from the normal distribution.

Thus, 2% of the “normal” population has macrocephaly

The causes of a large head include hydrocephalus (an excessive volume of CSF intracranially), megalencephaly (enlargement of the brain), thickening of the skull, and hemorrhage into the subdural or epidural spaces

Hydrocephalus is the main cause of macrocephaly at birth in which intracranial pressure is increased

The causes of megalencephaly are anatomical and metabolic



The anatomical disorders are primary megalencephaly and neurocutaneous disorders

Children with anatomical megalencephaly are often macrocephalic at birth but have normal intracranial pressure

Children with metabolic megalencephaly are usually normocephalic at birth and develop megalencephaly from cerebral edema during the neonatal period

Increased thickness of the skull bones does not cause macrocephaly at birth or in the newborn period

Macrocephaly develops during infancy

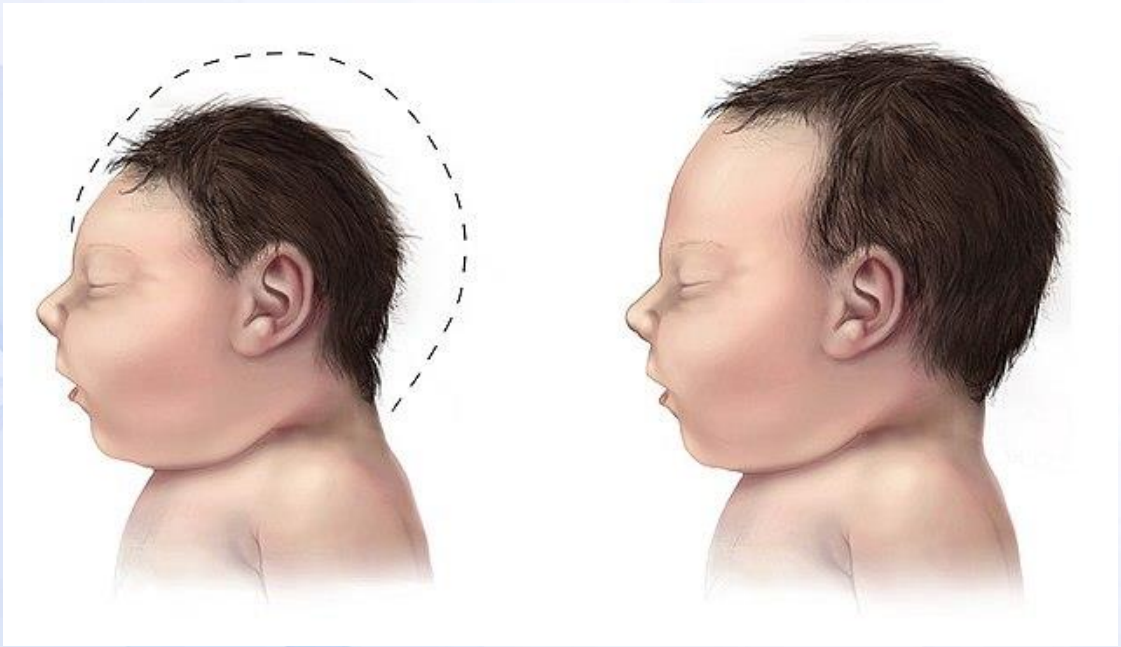
Microcephaly

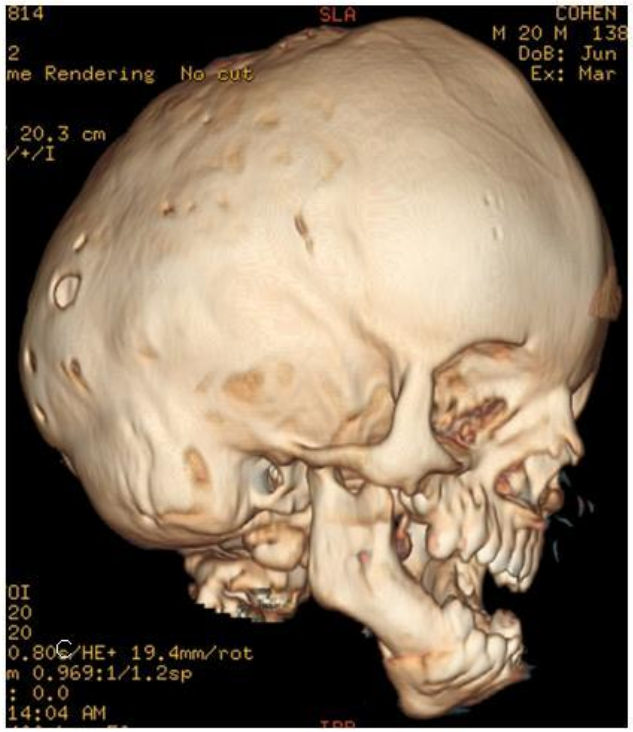
including >2 SD or >3 SD below mean HC

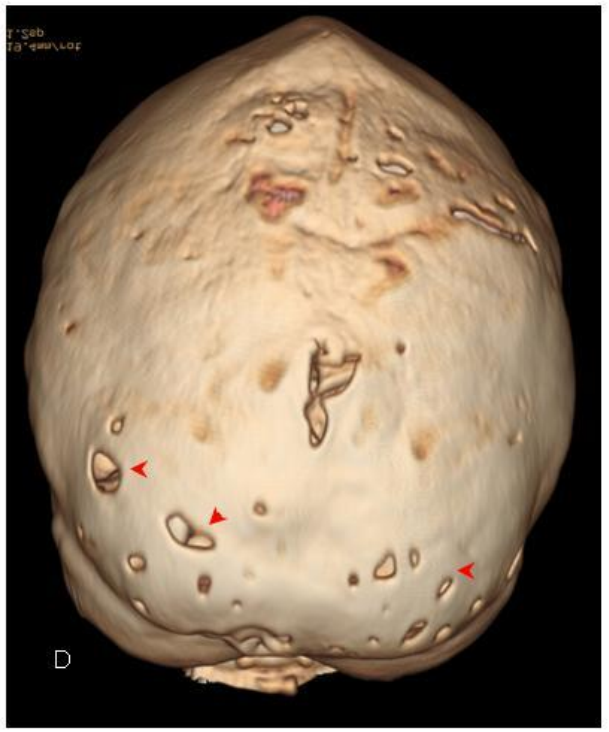
disproportionately small head circumference (HC) for gestational age

absolute (asymmetrical growth retardation—where the HC is reduced to a greater extent than length and weight)

relative (symmetrical growth retardation—where the HC, the length and weight are reduced to a similar degree)

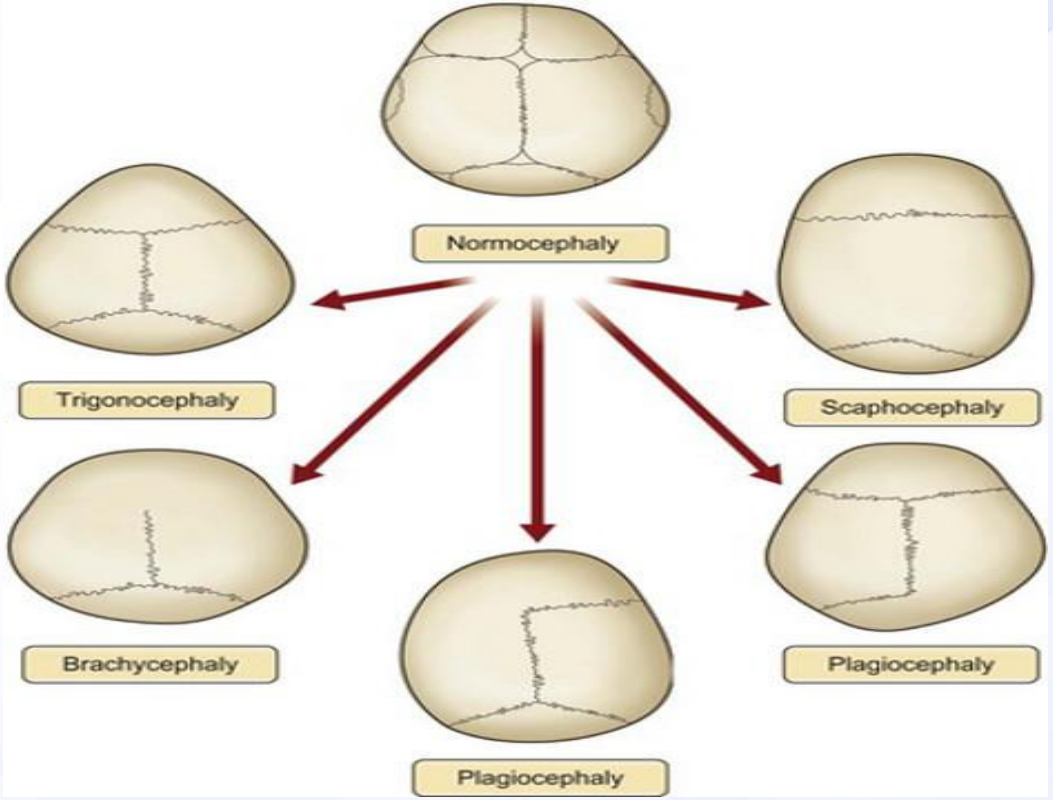




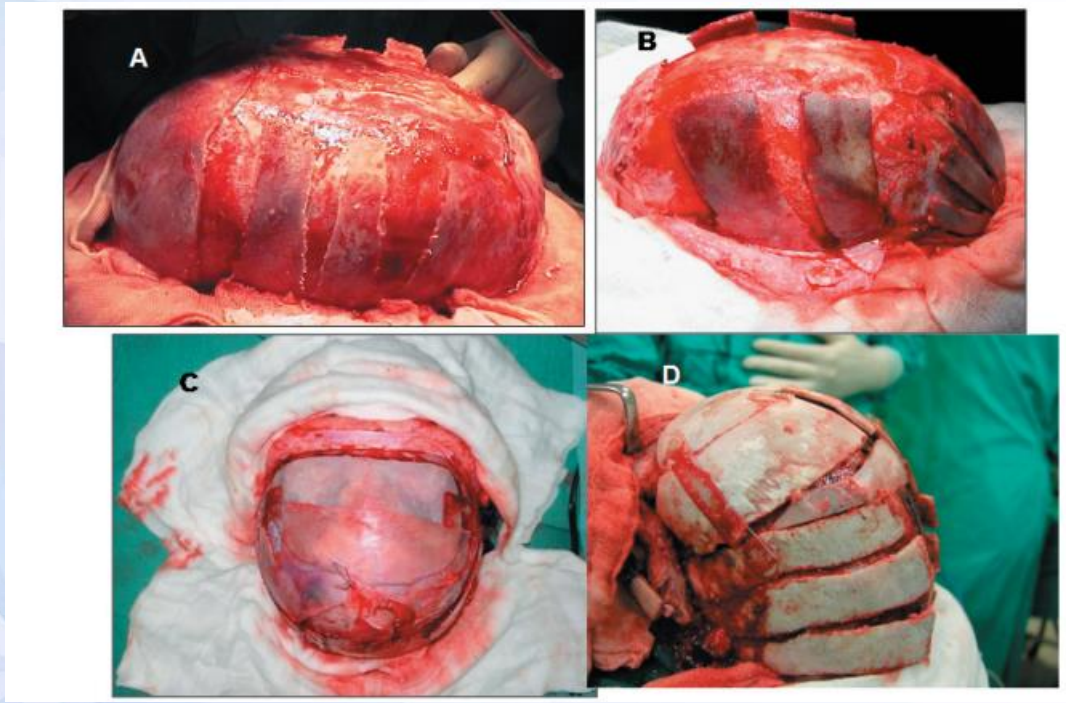


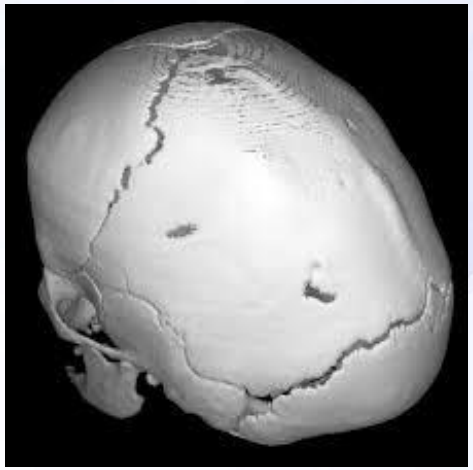
pansynostosis

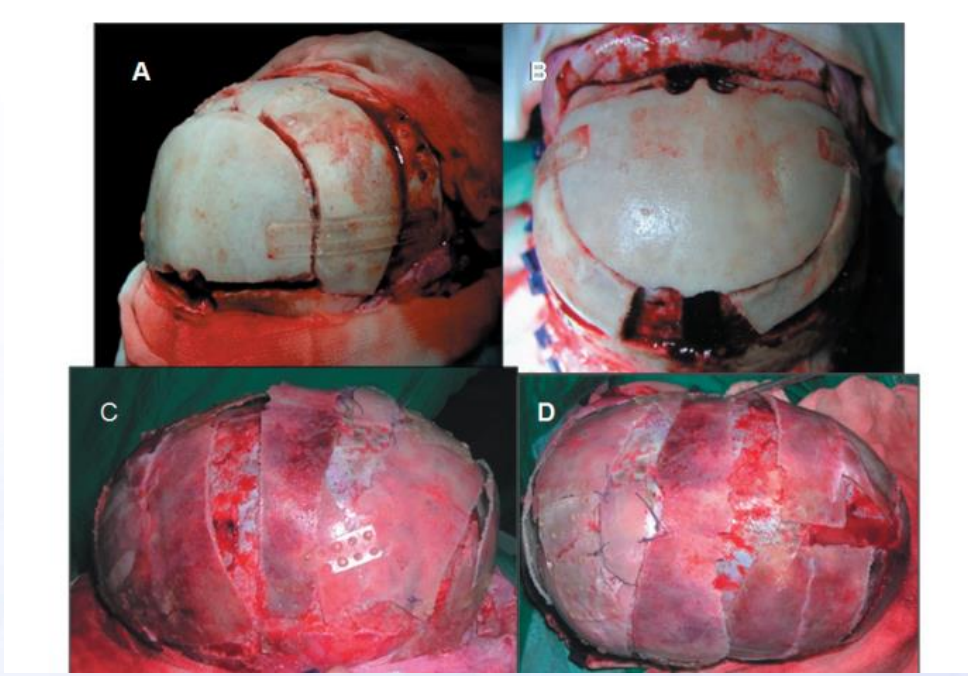
Craniosynostosis

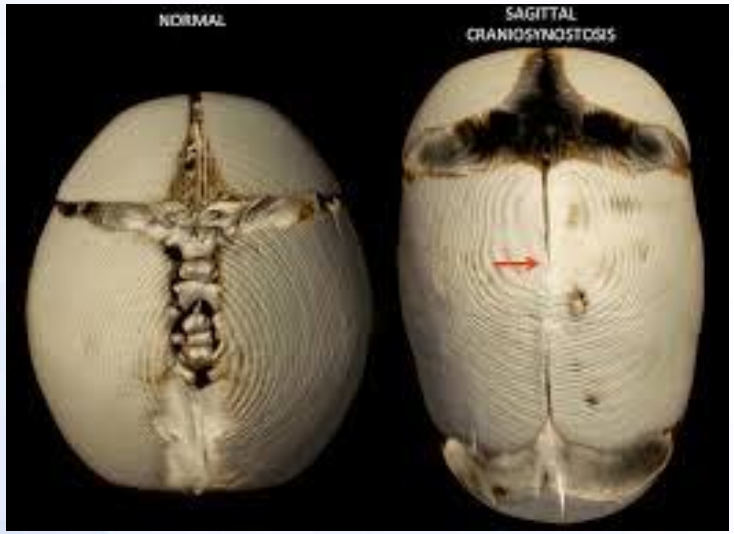


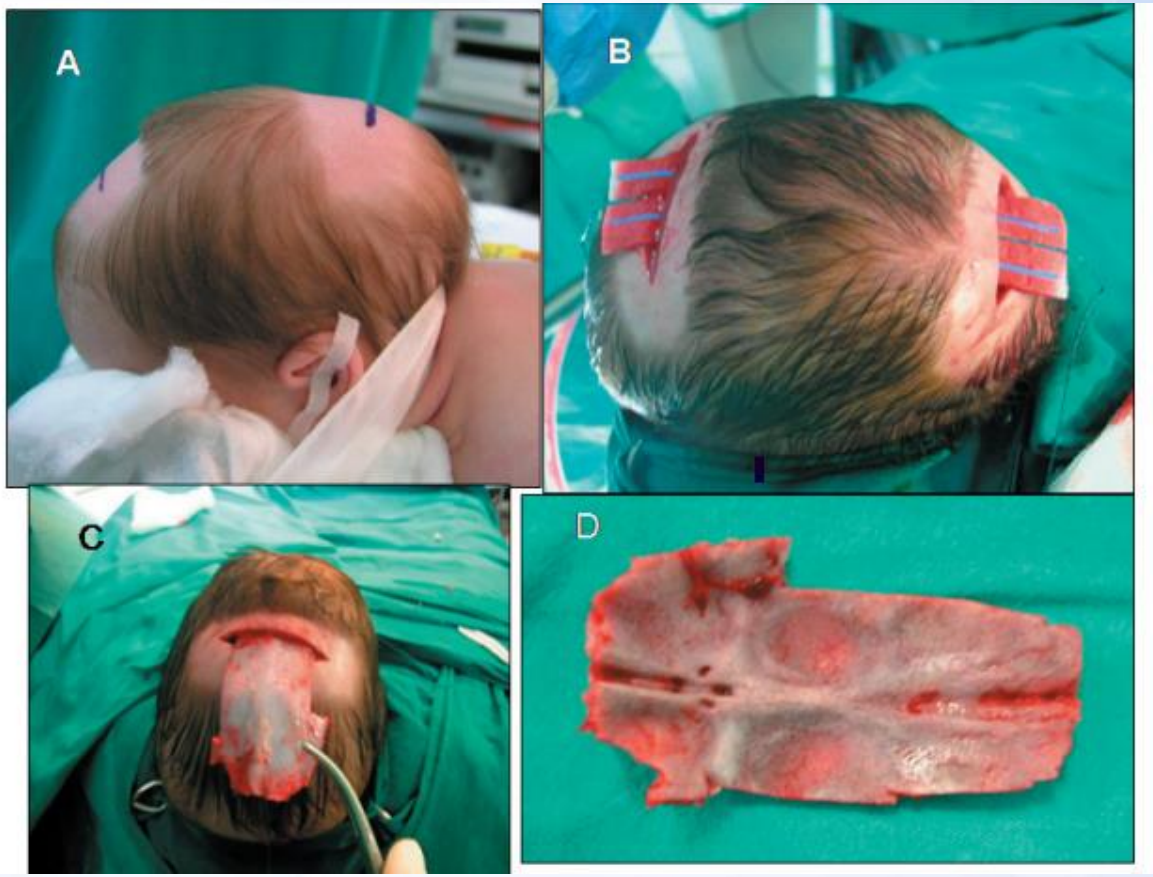
Scaphocephaly (Sagittal Craniosynostosis)



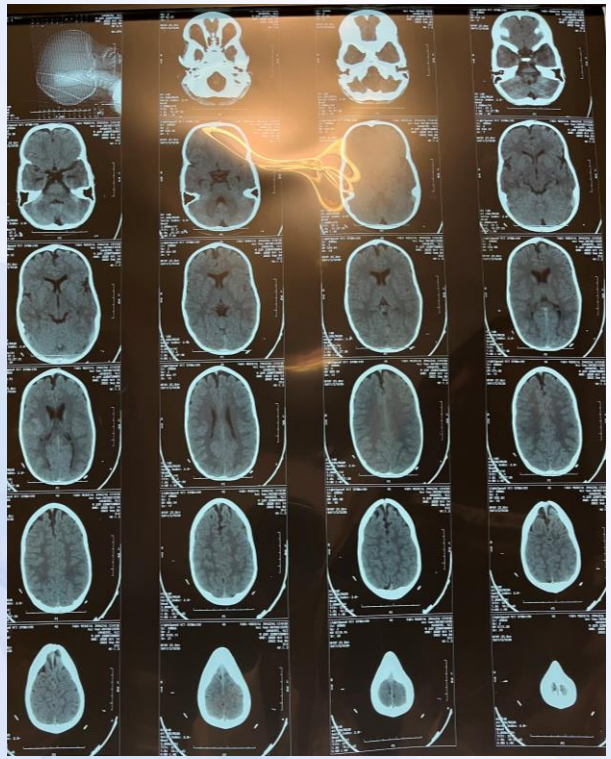


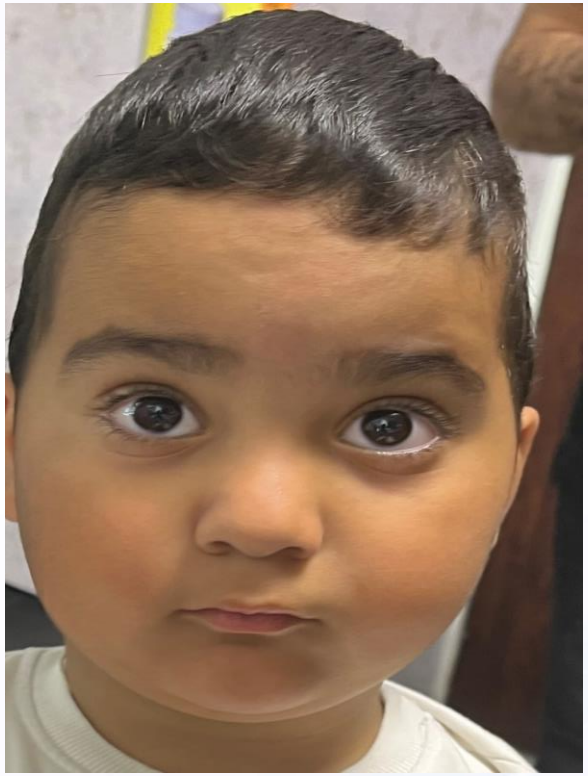



NORMAL
SAGITTAL
CRANIOSYNOSTOSIS

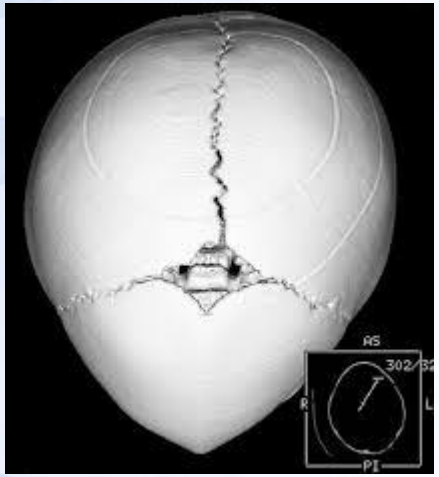

A
B
C
D

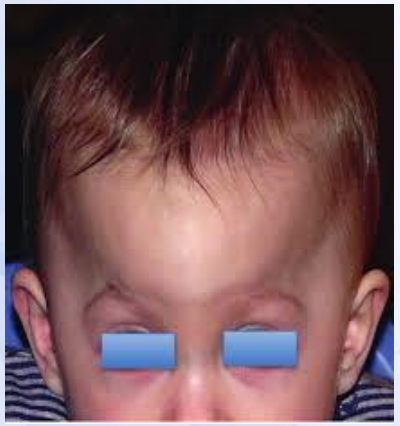
Missed Scaphocephaly

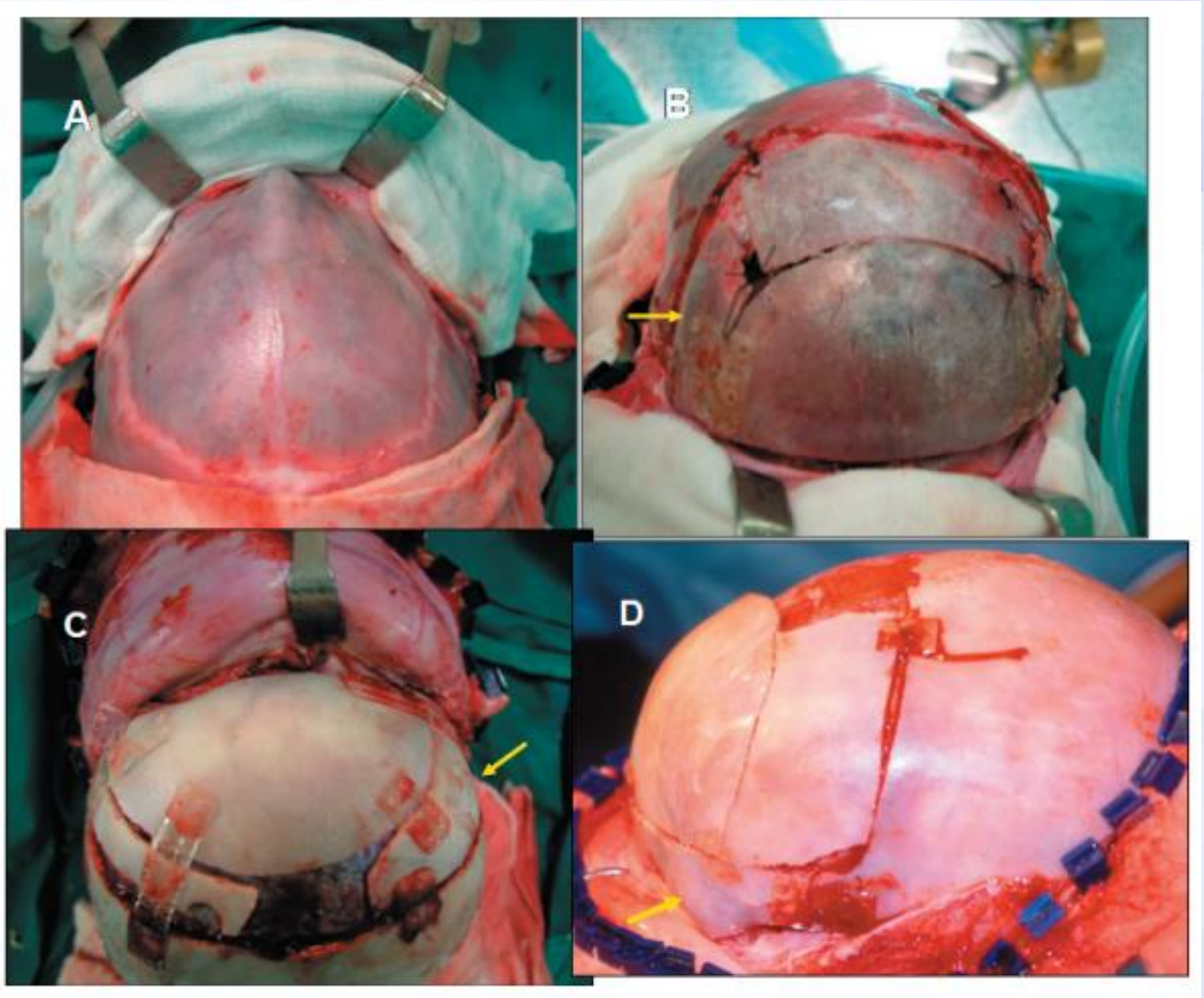




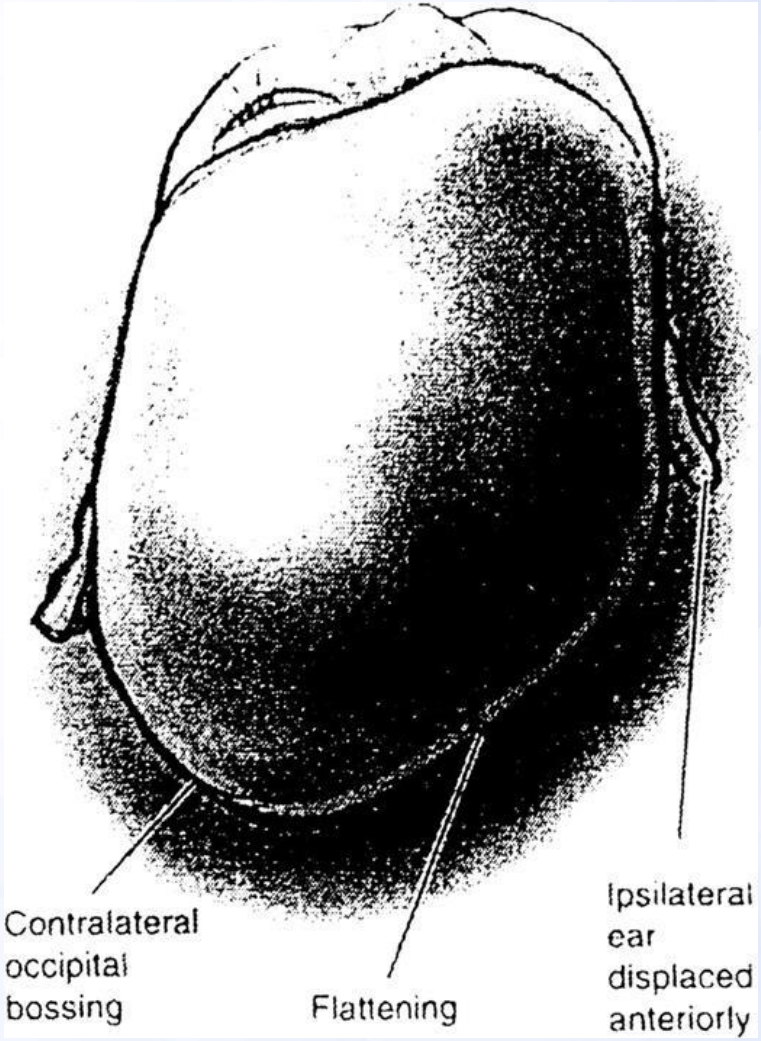
Trigonocephaly





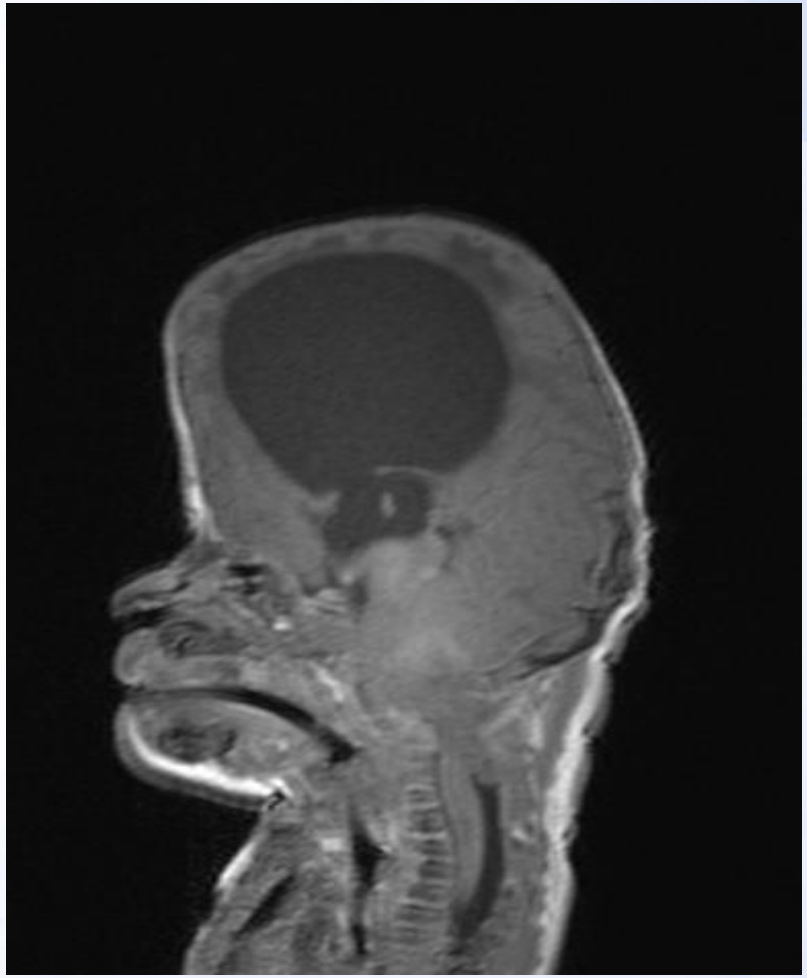


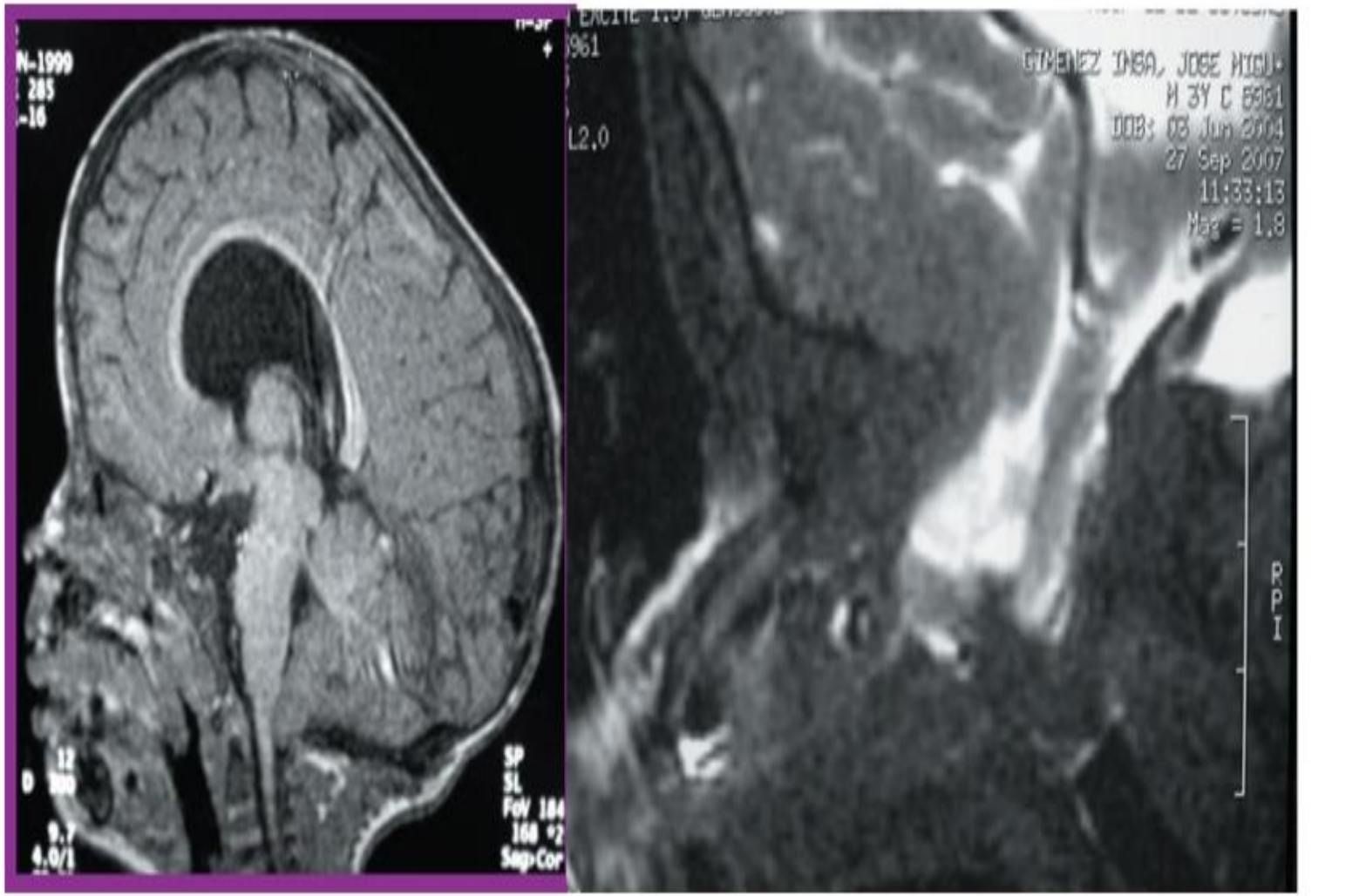
Plagiocephaly

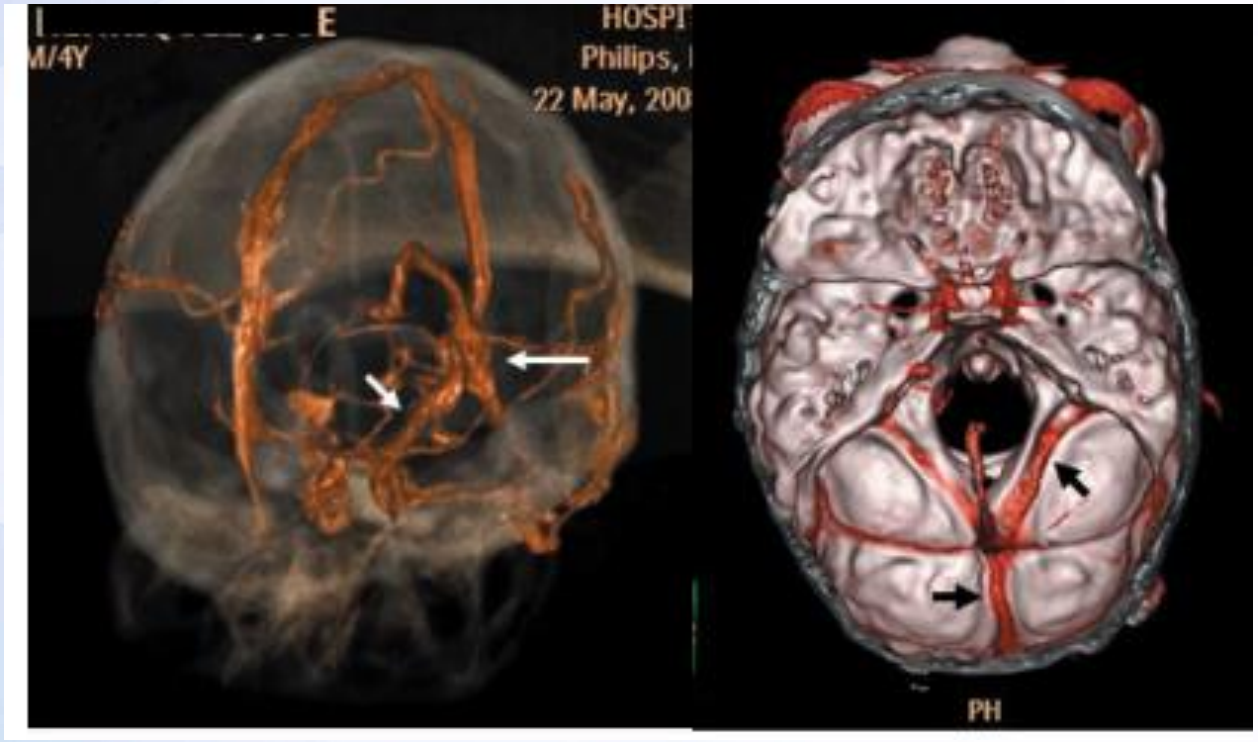


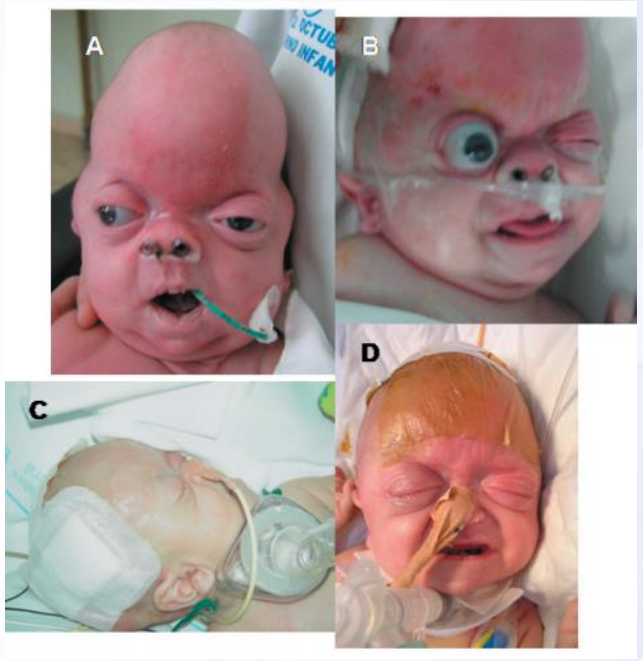
Oxycephaly (also known as **turricephaly**) is the most severe of the craniosynostoses and results from the premature closure of all sutures.
Characterized by a **tower-like skull** which may be associated with:

- 8th cranial nerve lesion
- optic nerve compression
- mental deficiency
- **syndactyly**

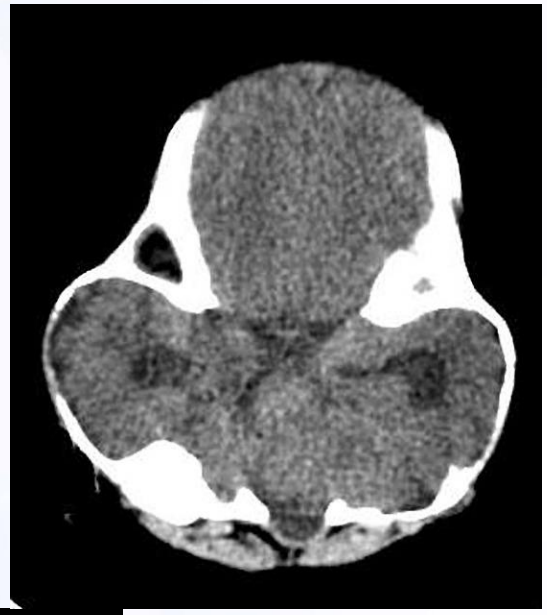


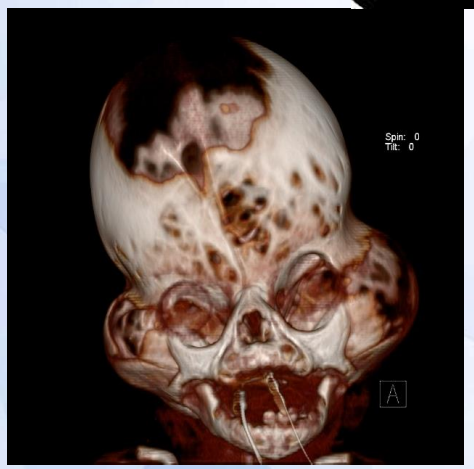

GIMENEZ INSA, JOSE MIGU
H 3Y C 8961
DOB: 03 Jun 2004
27 Sep 2007
11:33:13
L2.0
RPI

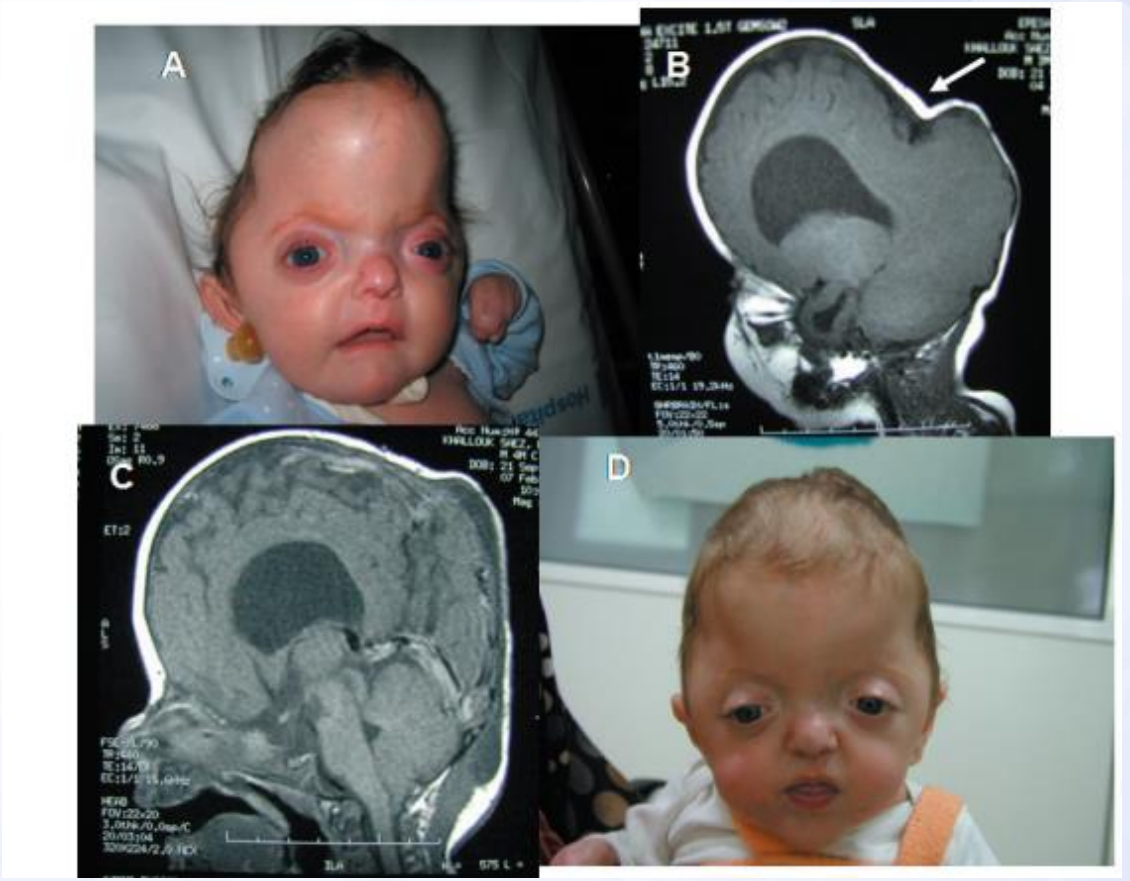

M/4Y
Philips,
22 May, 200
PH


A
B
C
D

Cloverleaf skull

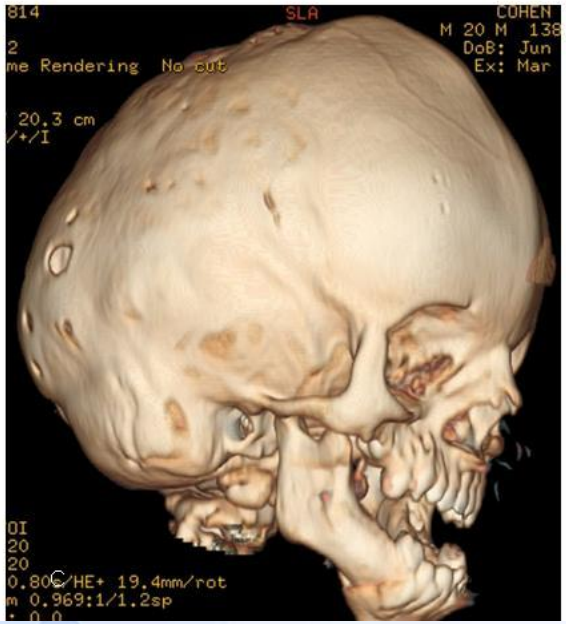


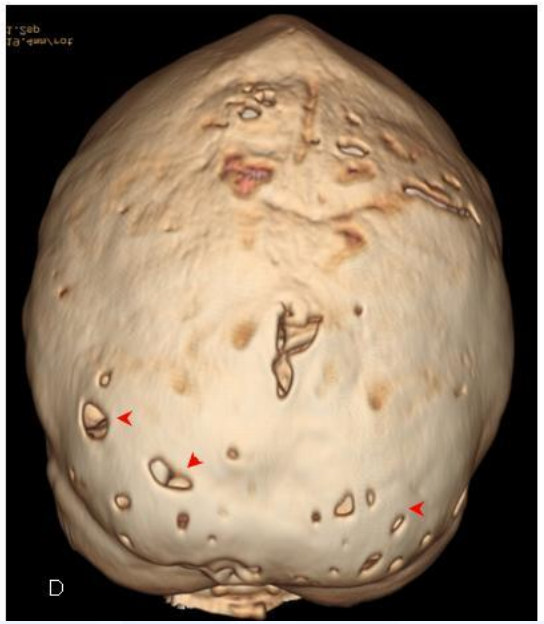




pansynostosis

- can present in several ways.
- The appearance can be the same as that seen with primary microcephaly: a markedly small head, but with normal proportions.
- The most severe form of pansynostosis is kleeblattschädel (cloverleaf skull), which presents with bulging of the different bones of the cranial vault





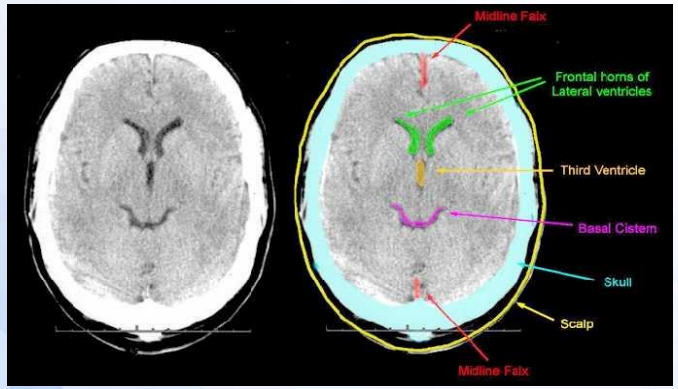
Hydrocephalus

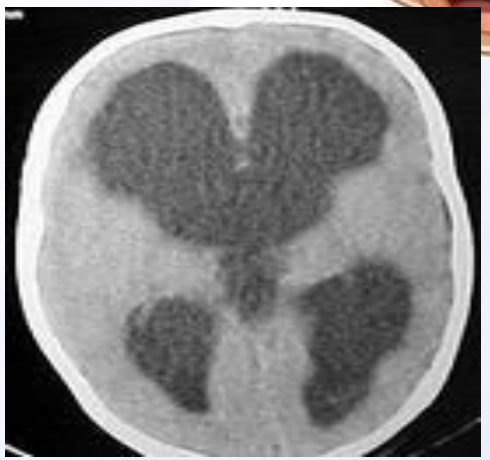
— established with neuroimaging

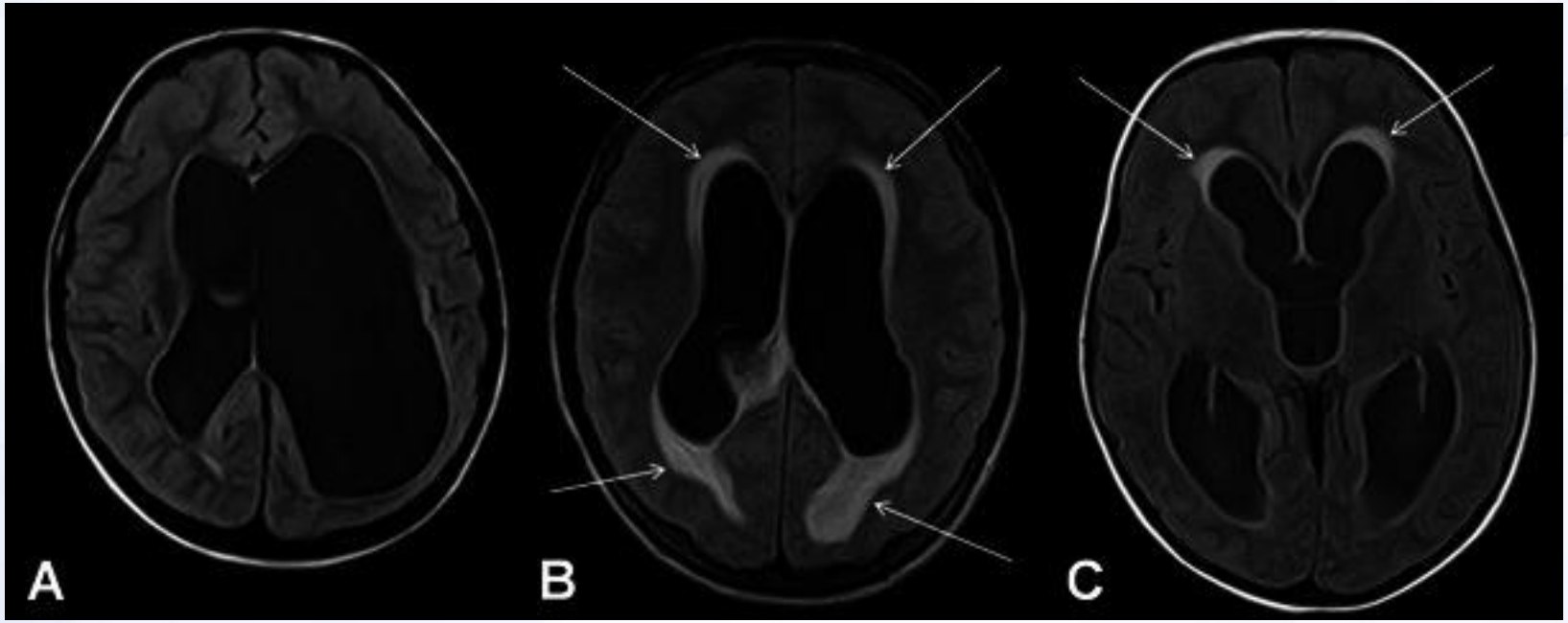
Hydrocephalus is characterized by ventriculomegaly and evidence of increased intracranial pressure (ICP)

Radiographically, findings that suggest increased pressure include:

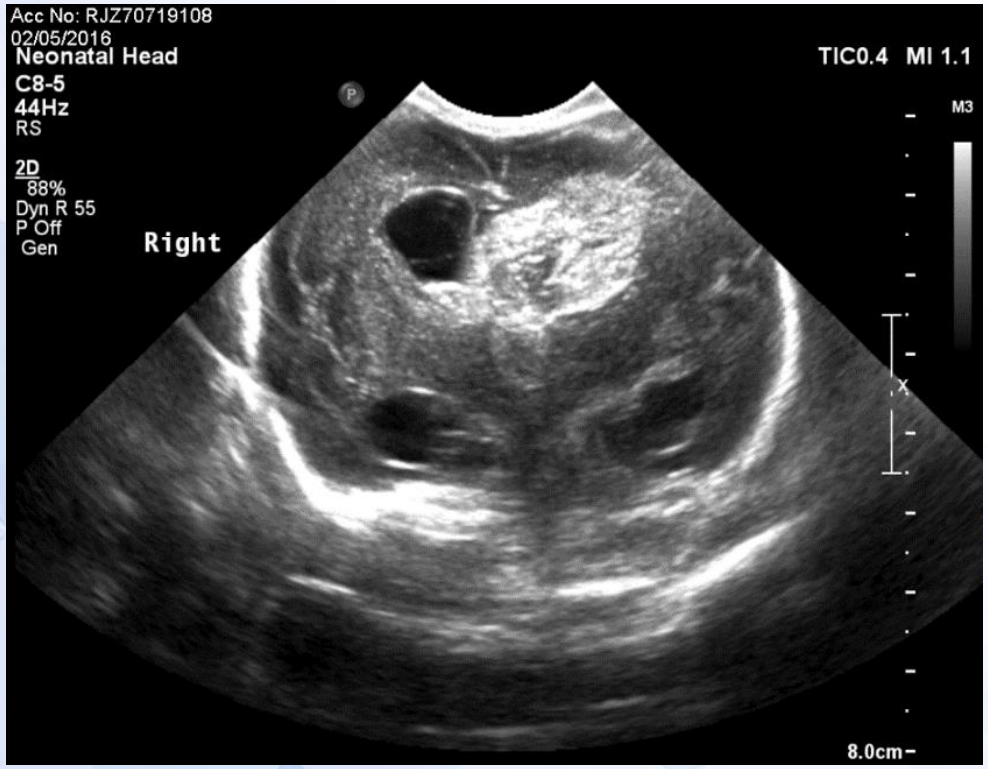
- Enlargement of the recesses of the third ventricle.
- Dilation of the temporal horns of the lateral ventricle.
- Interstitial edema of the periventricular tissues (seen on T2-weighted or FLAIR [fluid-attenuated inversion recovery] magnetic resonance imaging (MRI) sequences).
- Effacement of the cortical sulci.

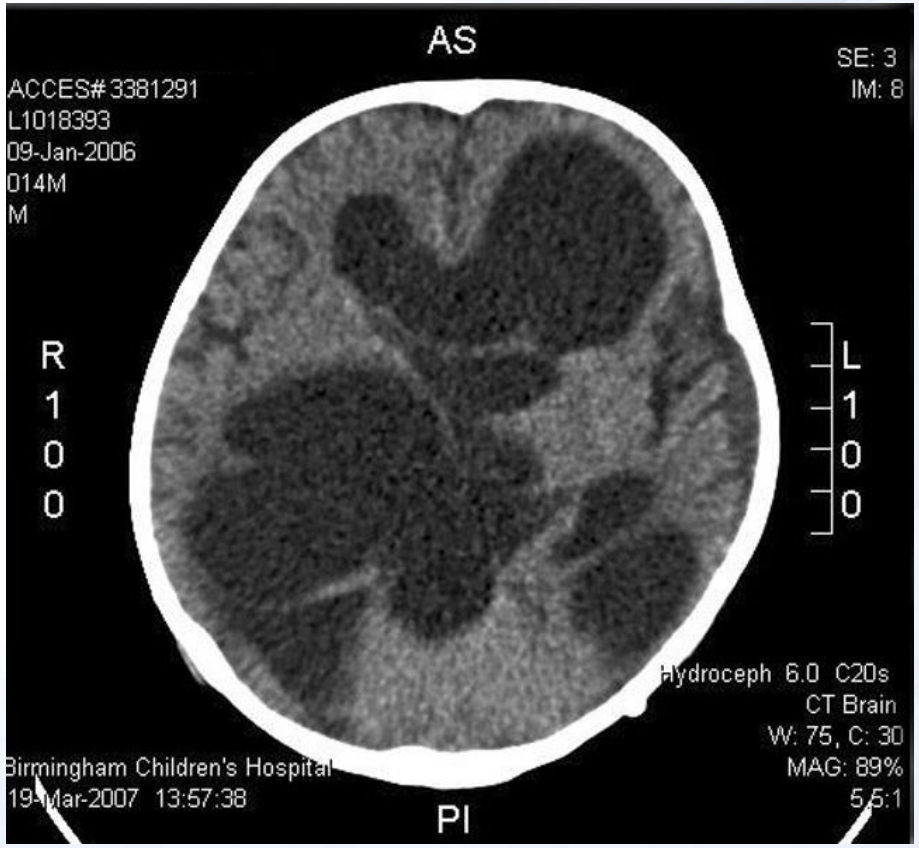





A
B
C

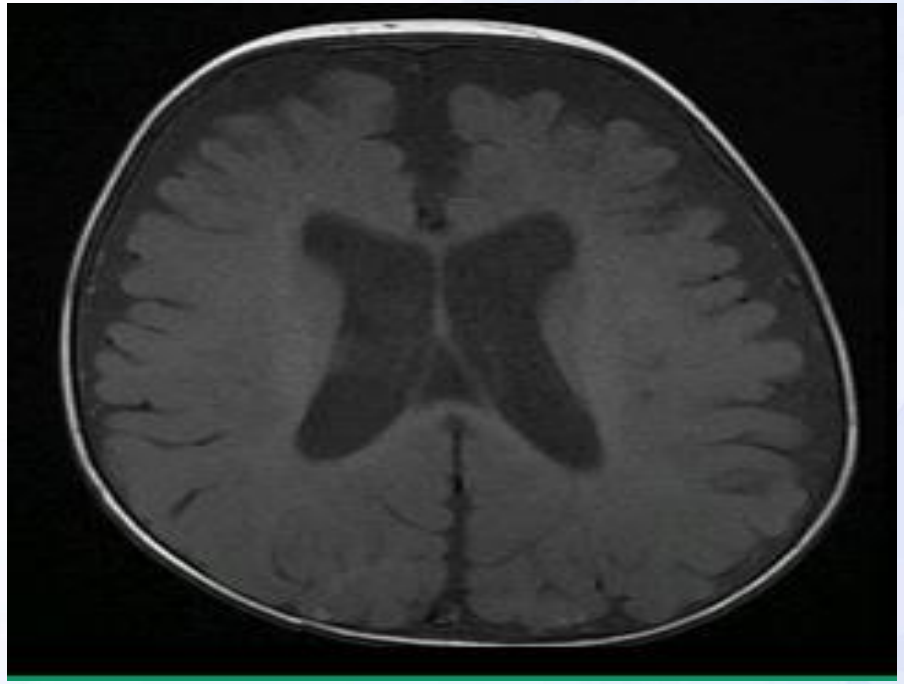
IVH of prematurity



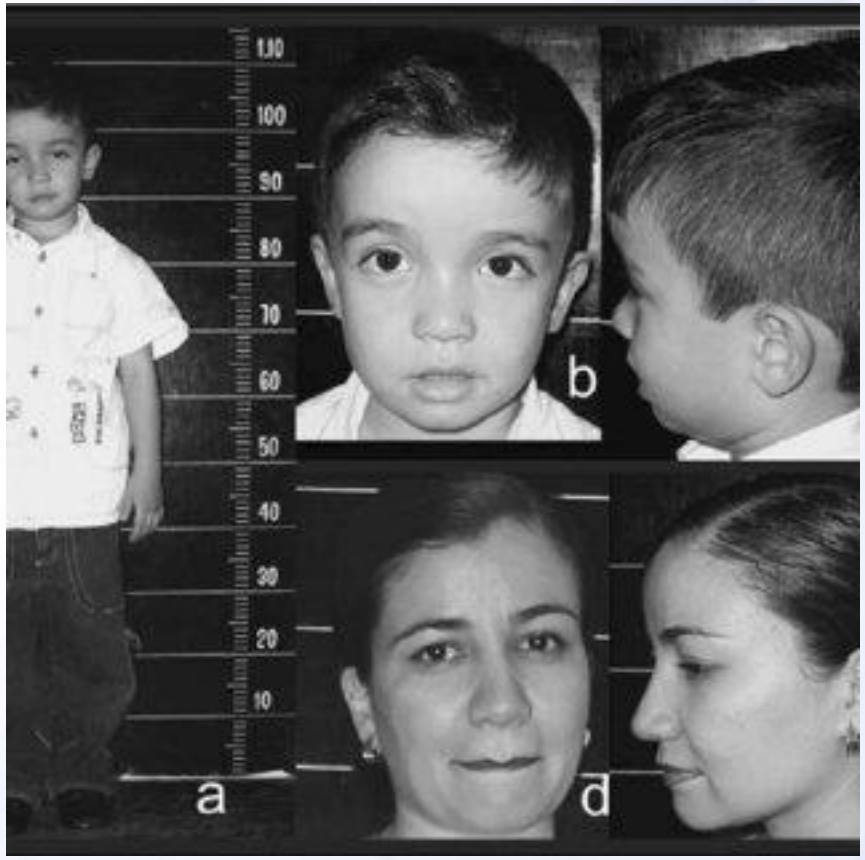


Benign external hydrocephalus"

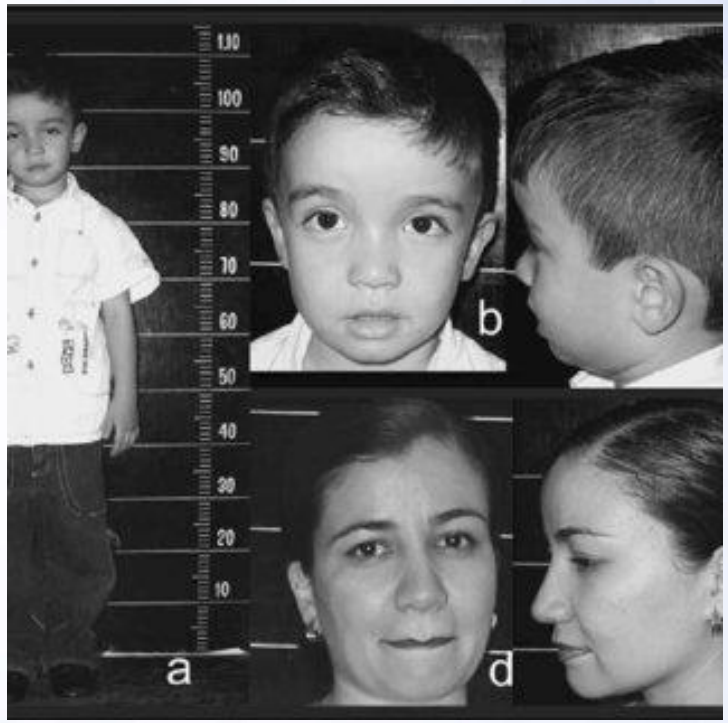
- – (also called "**benign enlargement of the subarachnoid space**" or "benign extra-axial fluid of infancy")
- is a relatively common cause of macrocephaly in infancy and frequently occurs in other family members
- As the name implies, the condition is self-limited and affected infants usually do not require any intervention



Normal children with large heads--benign familial megalencephaly



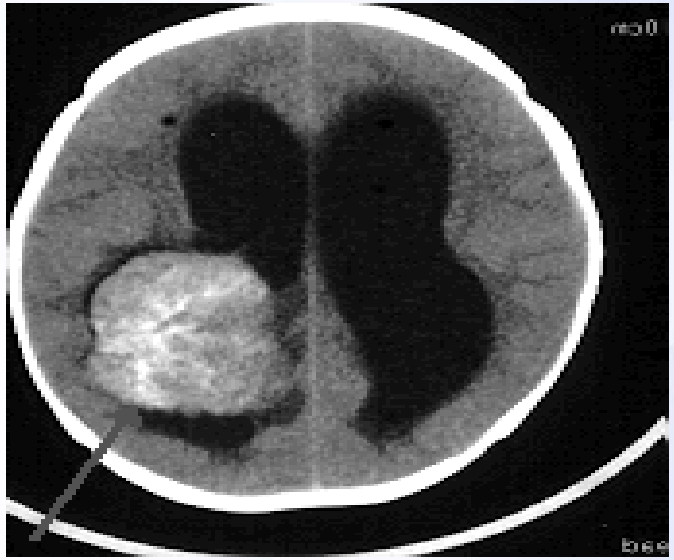
Normal children with large heads--benign familial megalencephaly

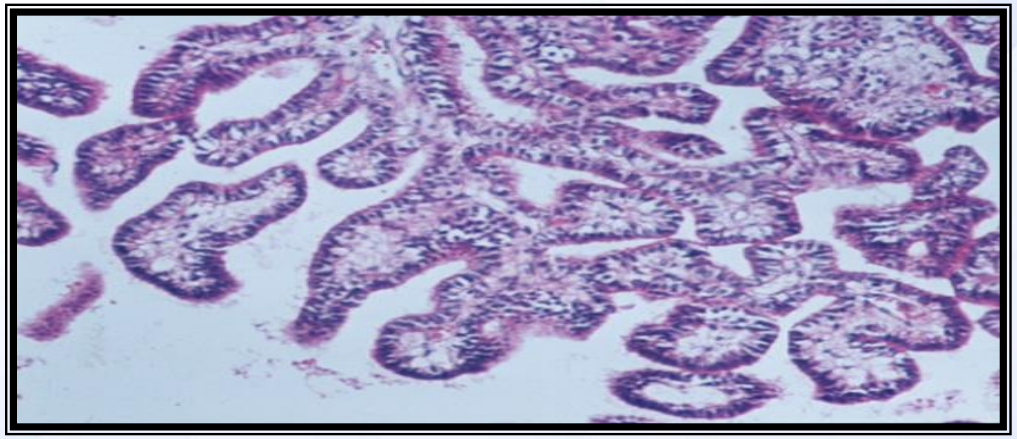


Overproduction of CSF

Excessive secretion of CSF by the choroid plexus as in cases of choroid plexus papilloma or carcinoma

This is a rare cause.

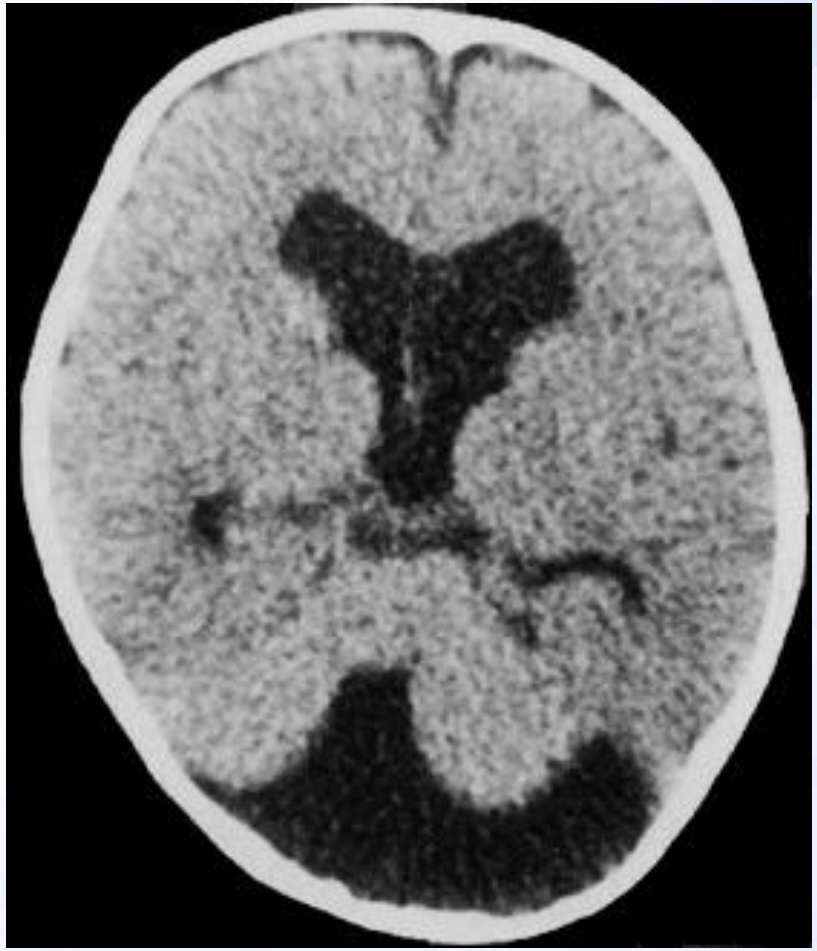




Dandy Walker Syndrome

A common cause of obstructive hydrocephalus is Dandy Walker Syndrome where there is blockage of foramina of the 4th ventricle

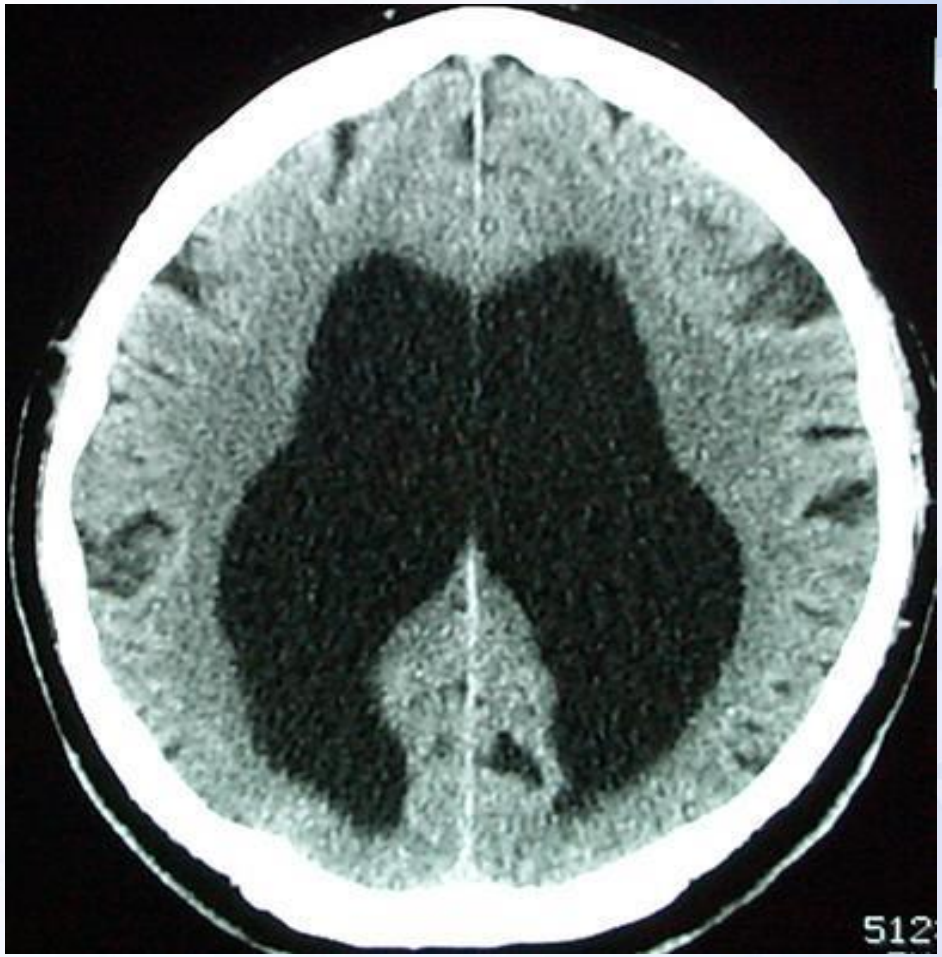
This is a congenital condition associated with agenesis of the cerebellar vermis



Normal pressure hydrocephalus

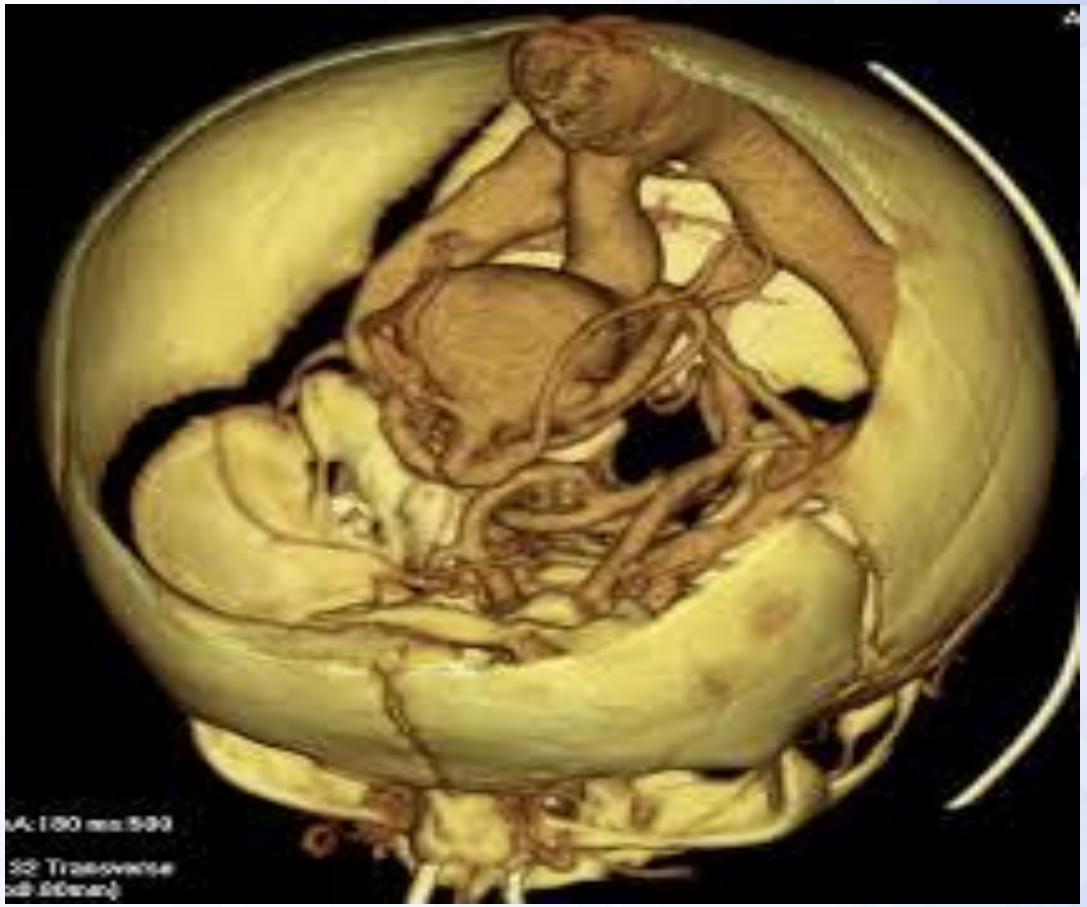
NPH is usually due to a gradual blockage of the CSF drainage pathways in the brain.

Triad:dementia.ataxia,urinary incontinency

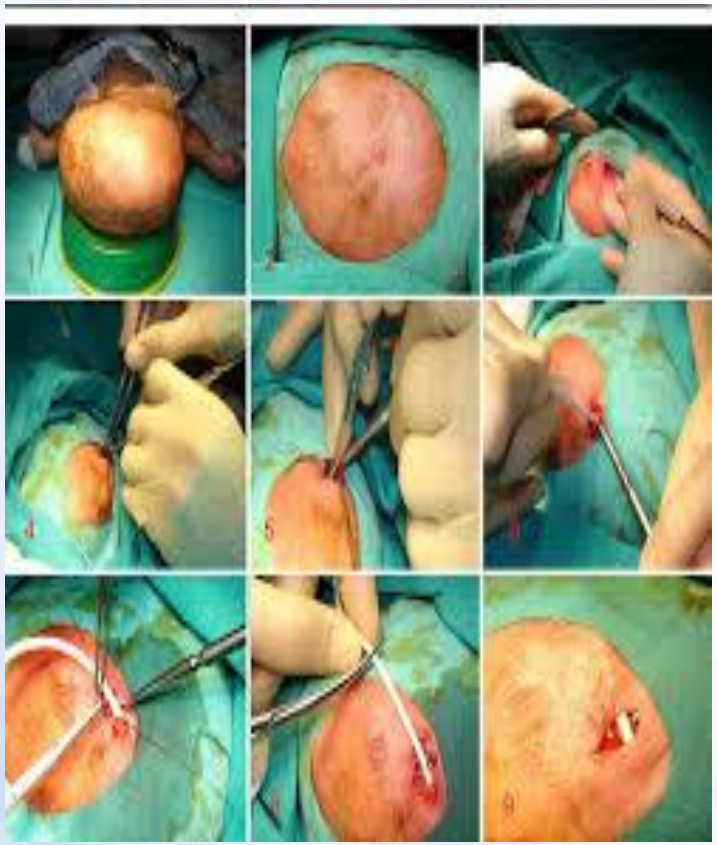


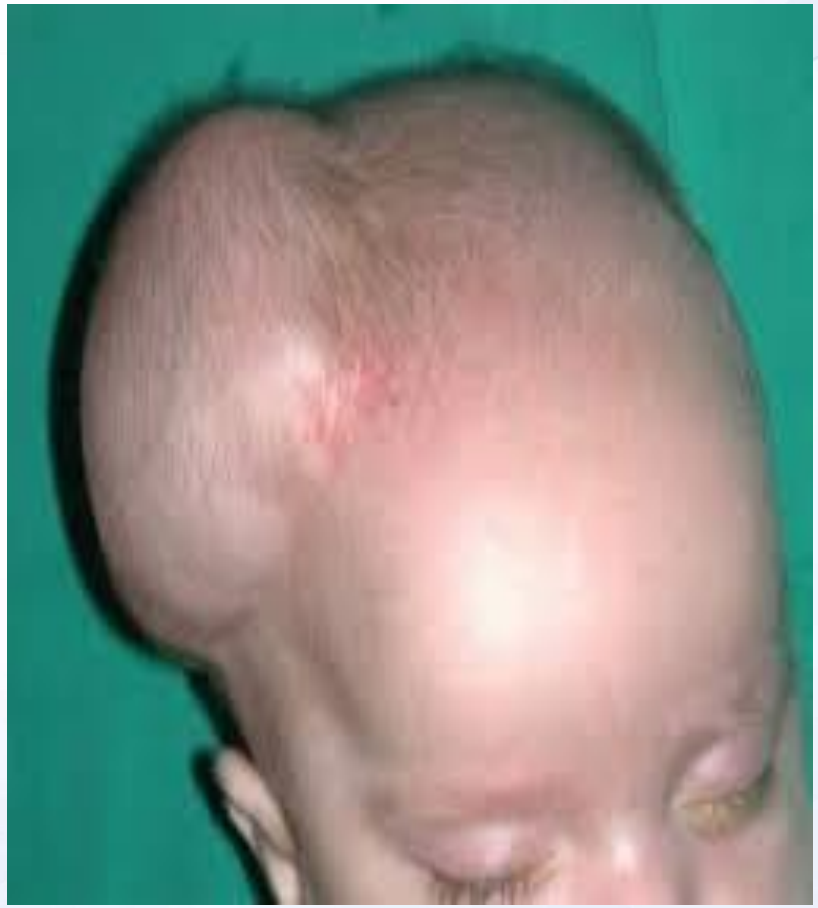
Hydrocephalus due to venous HTN

Vein of Galen anurysmal dilatation

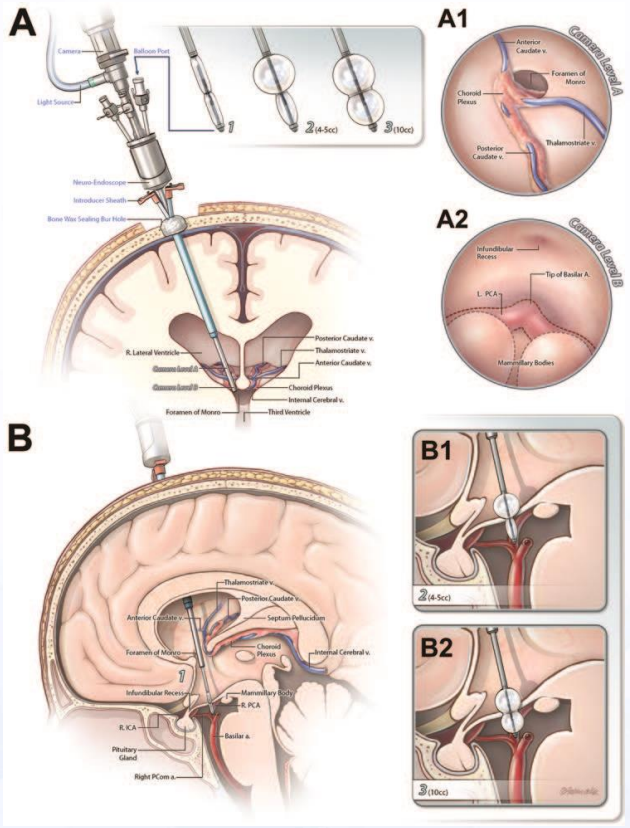


Shunt in LBW neonate

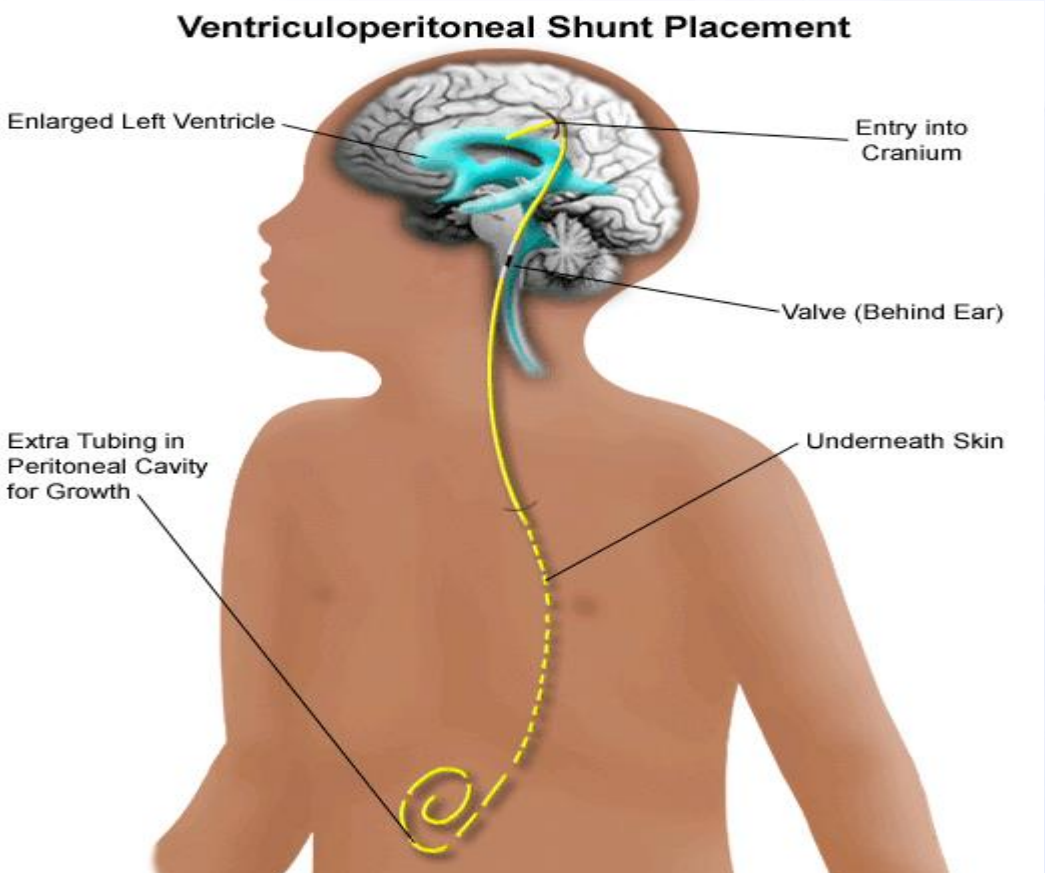




Endoscopic third ventriculostomy(ETV)



Ventriculoperitoneal shunt




جراحی مغز و اعصاب کودکان
Pediatric Neurosurgery