

The special case of infantile hydrocephalus

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- In children, hydrocephalus most prevalent in first year
- Late-recognized congenital hydrocephalus not uncommon
- Brain in infant protected by skull expandability
- Development-related vulnerability: on-going cellular proliferation, connection/synaptogenesis, myelination etc.
- Some etiologies particular to this age have specific brain morbidities

Infantile hydrocephalus (159 cases, malformations excluded)

• IVH (prematures)	34	21%	} complex
• Infection	22	13%	
• Vascular (eg: v of G)	6	4%	
• Midline cysts	15	9%	} simple
• Tumors	17	11%	
• Chiari I	9	6%	
• Parenchymal bleed	9	6%	} unknown
• Aqueduct (not tumor)	12	8%	
• 4V-cisternal	19	12%	
• Congenital	14	9%	

Prematures are not fetuses

- pericerebral spaces wide in fetuses, not in premies
- amniotic pressure in fetuses, expandable skull in premies
- CBF low in fetus/newborn, steep increase < two weeks irrespective of GA

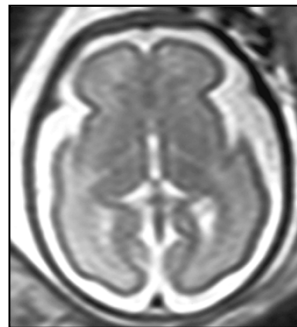
Kehrer et al. Ultrasound Med Biol 2004, 30, 283-7

Fetuses

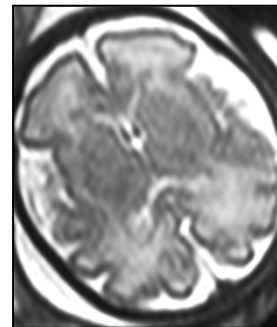
24w



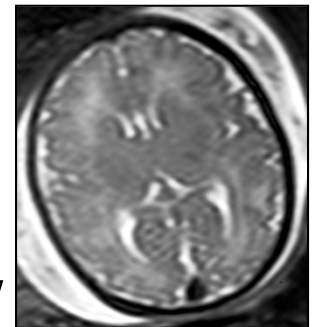
28w



33w

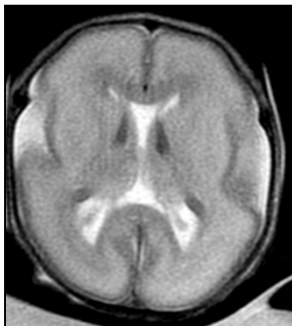


37w

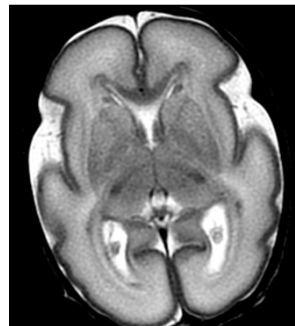


Premies

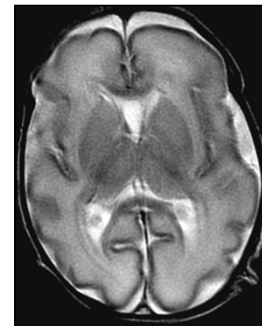
25w



28w



33w



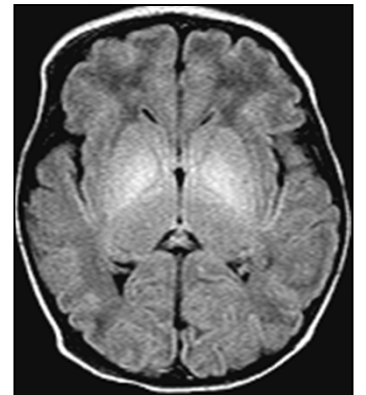
Specificities of the immature brain

1. Brain water and CSF
2. Cellularity
 1. germinal matrices
 2. oligodendroglia
 3. ependymal lining
3. Connectivity and synaptogenesis
4. Vessels: developing, immature, venous pattern incomplete

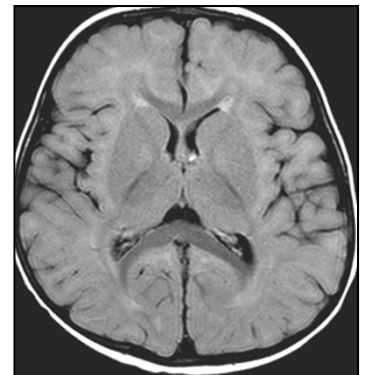
1 - Brain water and CSF

- Large extracellular spaces in infants
 - water in WM: 90% fetus/neonate, 75% mature
- Different CSF dynamics
 - no flow void in aqueduct and expandable skull
 - allows for external hydrocephalus
- Assumedly no arachnoid granulations in neonates

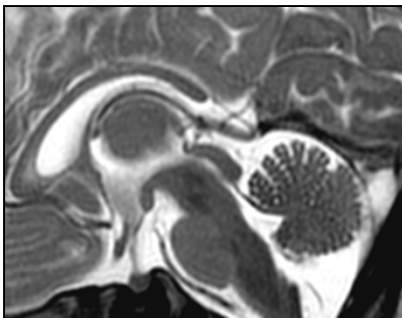
newborn



1-month

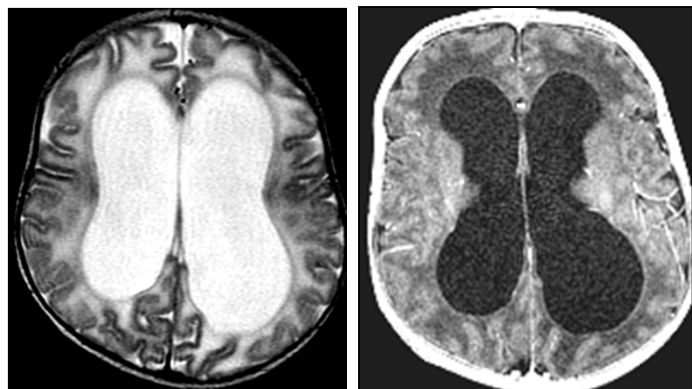


newborn

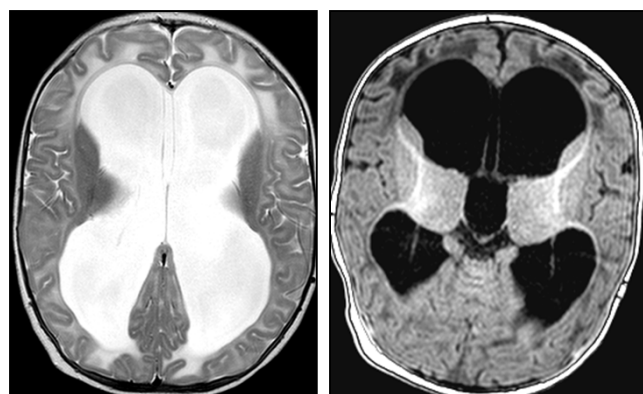


mature

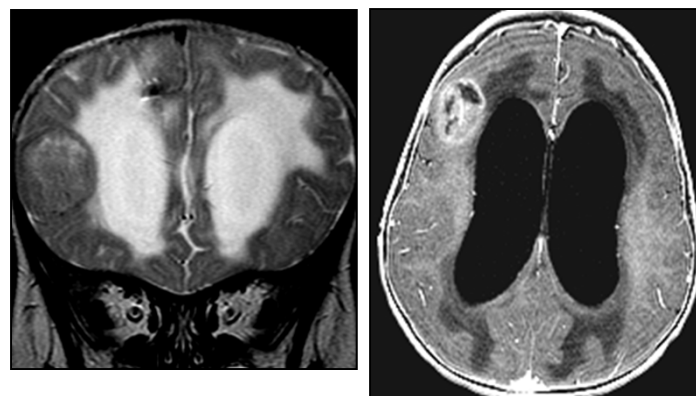




2 months

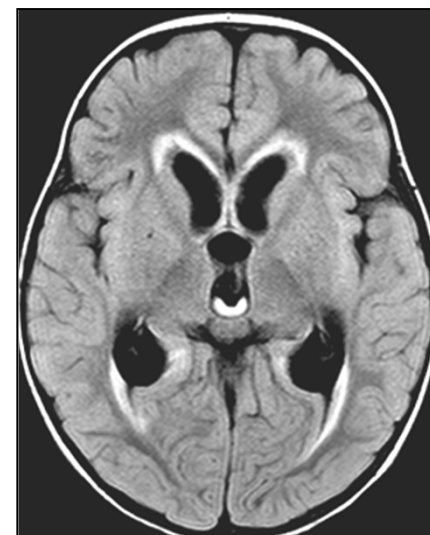


3 months

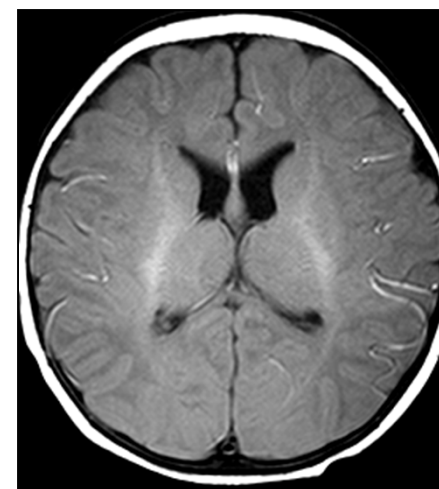


11 months

2 years

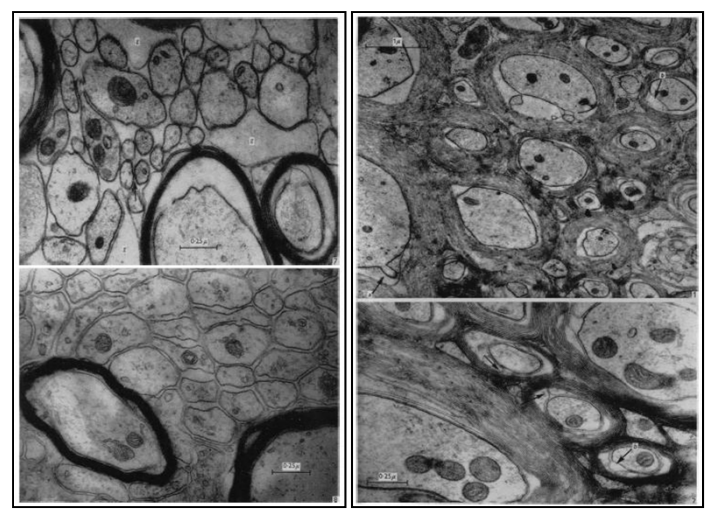
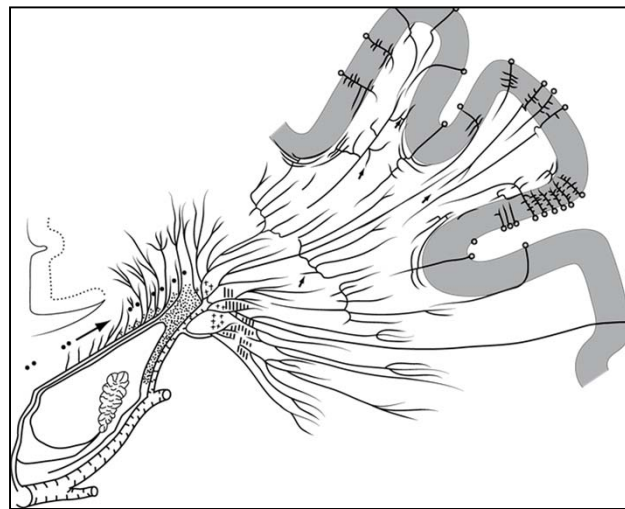


normal T1
1 month



Interstitial edema more extensive in infants

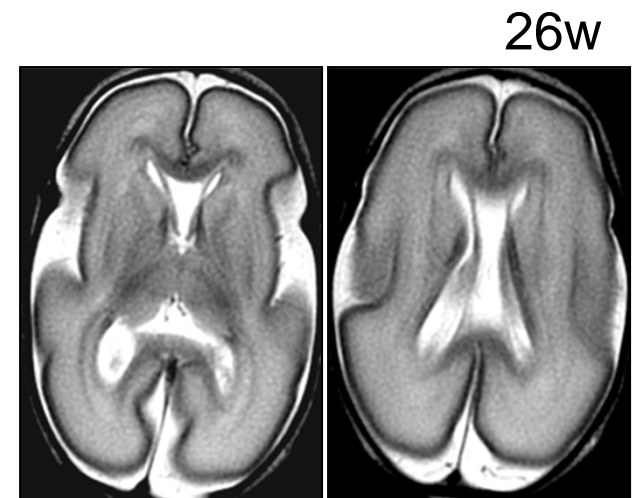
- Interstitial edema reaches the cortex in infants
 - venous drainage: subcortical incomplete, subependymal exposed
 - diffusion easier in unmyelinated brain
 - diffusion distance shorter from ependyma to cortex



2 - Cellularity of immature brain

- Pool of ependymocytes: +/- complete by midgestation
- Germinal matrix of mantle: disappears < 27w
 - late neuronal migration to superficial cortical layers
- Germinal matrix of ganglionic eminence: thickest at 25w, disappears < 36w
 - late GABAergic interneurons and thalamic neurons
- Oligodendroglia
 - intense proliferation until birth
 - myelination from 36w to well after birth

Del Bigio M, Brain 2011, 134:1344-61



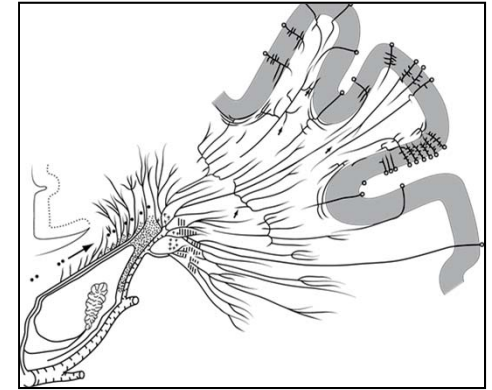
3 - Development of connectivity

- Until midgestation, all axons connected to transient targets
 - subplate, internal capsule, brainstem
- From 22 to 47w, collaterals multiply and reach cortex in sequential manner
 - layer 4: thalamocortical 22-27w
 - layers 2-3: intracortical and long association - commissural 27-32w
 - layers 2-3: short association (U-fibers) 32-47w



28w

4 - Early brain vascularity



- Germinal matrices from 8 to 36w
- Cortex, stepwise from depth to surface, from 22w to 47w
- Arterial perforators transcerebral from surface to ventricle
- Venous drainage, three components
 - transcerebral from ventricle to surface (5w to 36w), only few persist
 - subependymal to vein of Galen (9w to mature)
 - transcortical to surface (22w to mature)

In summary

- Specific causes expose parenchyma to specific damages
 - IVH: destruction of GM
 - decreased cortical, thalamic cellularity
 - Impaired venous drainage
 - IVH, infection: ependymal, choroidal fibrosis \
 - altered blood-CSF-parenchymal exchanges
 - infectious toxins: parenchymal inflammatory cascade
 - arterio-venous shunts: perfusion steal, high venous pressure, dual hydrocephalus

In summary

- Hydrocephalus exposed to time-specific developmental complications
 - perfusion impact of parenchymal distension on
 - developing parenchyma: 23-32w
 - germinal matrices: <36w
 - oligodendroglial proliferation, myelination: 28w-years
 - synaptogenesis and GM selective vulnerability: around term
 - impact of perfusion changes, interstitial edema, fiber-stretching on
 - axonal path-finding: 22w-years
 - period of possible reversibility (limits?)

Conclusion

- Hydrocephalus among children is most prevalent in infants
- Brain somewhat protected by expandable skull
- High vulnerability to developmental impairment
 - mechanically from hydrocephalus itself
 - parenchymal perfusion, ependymal tears, fiber stretching, interstitial edema
 - germinal, oligodendroglia, organizing gray matter, connectivity
 - as well as from some of its causes
 - hemorrhage, inflammation, A-V fistula
 - evolutivity (slow-fast), severity, duration