Hydrocephalus Notes

- **Sx** = triad of headache, vomiting and papilledema, ventricular enlargement Sx (ataxia/urine incontinence/dementia). In children: increased head circumference, ape face, sunset eyes, etc.
- **Acute Sx =** 'sunset eyes'

- **Overproduction**: papilloma tumor of the choroid plexus.

- **Obstruction:**
  - **a-** Congenital = aqueduct of Sylvius (stenosis due to gliosis or forking), 4th ventricle (atresia of the foramina 'Dandy-Walker), Chiari malformations.
  - **b-** Non-congenital = 3rd ventricle (colloid cyst), post. fossa (medulloblastoma)
    - Arachnoiditis secondary to either meningitis or subarachnoid hemorrhage can occlude the basal foramina and cause obstructive hydrocephalus.

- **Diminished absorption:** meningitis or SAH

- **Communicating** hydrocephalus is usually treated by shunts whereas **non-communicating** hydrocephalus is usually treated by endoscopic 3rd ventriculocisternostomy and shunts.

- Dandy-Walker cysts are developmental abnormalities characterized by a large cyst in the fourth ventricle, hypoplasia of the cerebellar vermis, and atrophy of the cerebellar hemispheres.

- Bypass Tx of Hydrocephalus:
  - **1- 3rd ventriculocisternostomy**: surgical creation of a communication between the 3rd ventricle and the interpeduncular cistern, for drainage of CSF.
    Cx: injury to the basilar artery or the mammary bodies during the operation
    Used to treat non-communicating hydrocephalus.

  - **2- Shunts**: ventriculoperitoneal shunt; most common. Drowsiness, Headache, Loss of consciousness and coma are signs of shunt malfunction

- **Normal Pressure Hydrocephalus**
  - Normal pressure (ICP) and dilated ventricles on CT scan.
  - **Hakim -Adams triad:** magnet gait (ataxia), urinary incontinence, dementia
  - **DDx**: Dementia (Alzheimer's), chronic subdural hematoma
  - **Tx**: LP

- **Hydrocephalus ex-vacuo**
  - When brain substance is lost or becomes gliosed after infarcts or aging atrophy.

Site	Causes		Notes	Management
Lateral ventricles	Choroid plexus papillomas		Endoscopic coagulation of the choroid plexus or surgical removal	
Foramina of Monro	Occur secondary to a congenital membrane, atresia, or gliosis after intraventricular hemorrhage or ventriculitis.		Unilateral ventriculomegaly	
Third ventricle	**Cysts**	**Colloid cysts**	Congenital	Stereotactic aspiration of the cyst, resection via craniotomy, or endoscopic resection
		Ependymal and arachnoid	Bobble-head doll syndrome	Endoscopic fenestration
	Neoplasms	**Craniopharyngiomas**		Surgical resection
		Chiasmal-hypothalamic gliomas		Shunt placement
Sylvian aqueduct	**Congenital aqueductal stenosis**		- Classified as true stenosis, forking, septum, or subependymal gliosis - Can be as a result of Bickers-Adams-Edwards syndrome - Secondary to in utero infections (toxoplasmosis), IVH, or mumps encephalitis.	
	- Pineal region neoplasms (germinomas) - AV malformations - Periaqueductal neoplasms (Low-grade astrocytomas)			
Fourth ventricle	- In infants, the fourth ventricle is the location for obstruction secondary to Dandy-Walker cysts or obliteration of the basal foramina. - In older children = medulloblastomas, astrocytomas, ependymomas, brainstem gliomas.			
Arachnoid granulations	- After meningitis, subarachnoid hemorrhage, or trauma. - Meningeal carcinomatosis		- Pan ventricular dilatation-communicating hydrocephalus	

Age	Causes

Premature infants	- Intraventricular hemorrhage - Cranial ultrasonography is useful
Full-term infants	- Aqueductal stenosis - Chiari II malformation - Dandy-Walker syndrome - Cerebral malformations - Vein of Galen malformations.
Older children	Secondary to trauma, meningitis or neoplasms.
Adults	- Idiopathic. - Subarachnoid hemorrhage (51%), head injury (16%), brain tumor (9%), intracranial surgery (7%), aqueduct stenosis (5%), and meningitis (5%).

Shunt complications

	Uncommon complications			
Common complications	Cranial	Subcutaneous	Peritoneal	Atrial
Infection	Subdural hygroma	Shunt migration	Peritonitis	Endocarditis
Obstruction	Subdural hematoma	Shunt disconnection	Pseudocysts	Nephritis
Inadequate flow or overdrainage	Hemiparesis Hematoma	Shunt fracture	Perforation Hernias	

SIGNS AND SYMPTOMS

Premature infants	Infants	Toddlers and older
Apnea	Irritability	Headache
Bradycardia	Vomiting	Vomiting
Tense fontanelle	Drowsiness	Lethargy
Distended scalp veins	Macrocephaly	Diplopia
Globoid head shape	Distended scalp veins	Papilledema
Rapid head growth	Frontal bossing Macewen's sign Poor head control Lateral rectus palsy "Setting-sun" sign	Lateral rectus palsy Hyperreflexia/clonus

<u>Spina Bifida Notes</u>

- **Risk factors of spina bifida**: genetics, folate deficiency, anti-epileptics (valproate).
- **Spina bifida occulta**: Swelling – Tuft of hair- Redness- Dimple
- **Spina bifida aperta**:

  1- **Lipomeningocele**: lipoma located over the lumbosacral spine associated with bowel or bladder dysfunction.

  2- **Meningocele**

  3- **Myelomeningocele**
  - **Sx =** Pain, weakness, paralysis / bowel and bladder problems / Skin ulceration
  - Highly associated with hydrocephalus (Chiari 2 and congenital aqueduct stenosis)
  - Patient may have PCKD and cardiac anomalies
  - **Cx =** Club foot (m.c) / Scoliosis or kyphosis / Osteoporosis / Hydrocephalus

  4- **Spina bifida ventralis** is less common than other, the protrusion is into retroperitoneal space and affect retroperitoneal organs (duodenum, kidneys, adrenal, aorta, IVC)

- **Tethered cord syndrome** is a rare neurological condition in which the spinal cord is attached (tethered) to the surrounding tissues of the spine. This prevents the spinal cord from moving to keep up with the lengthening of the spine as it grows. The result may be nerve damage and severe pain.
  Associated with a short, thickened filum terminale, or with an intradural lipoma.
- **Prenatal detection of NTD**: serum alpha-fetoprotein (AFP), ultrasound and Amniocentesis

Finding	Chiari type 1 (*see below*)	Chiari type 2 (*see page 238*)
caudal dislocation of medulla	unusual	yes
caudal dislocation into cervical canal	tonsils	inferior vermis, medulla, 4th ventricle
spina bifida (myelomeningocele)	may be present	rarely absent
hydrocephalus	may be absent	rarely absent
medullary "kink"	absent	present in 55%
course of upper cervical nerves	usually normal	usually cephalad
usual age of presentation	young adult	infancy
usual presentation	cervical pain, suboccipital H/A	progressive hydrocephalus, respiratory distress