IMET 2000 PAL

International Medical Education Trust - Palestine

Neuromuscular and cerebral disorders

Hydrocephalus

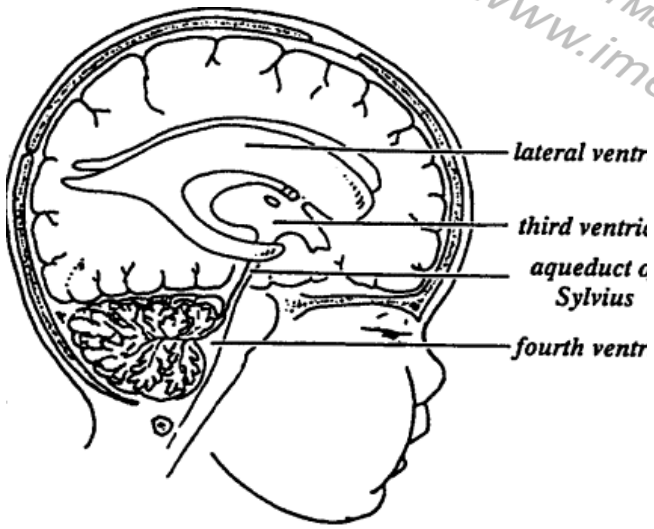
Definition of Hydrocephalus

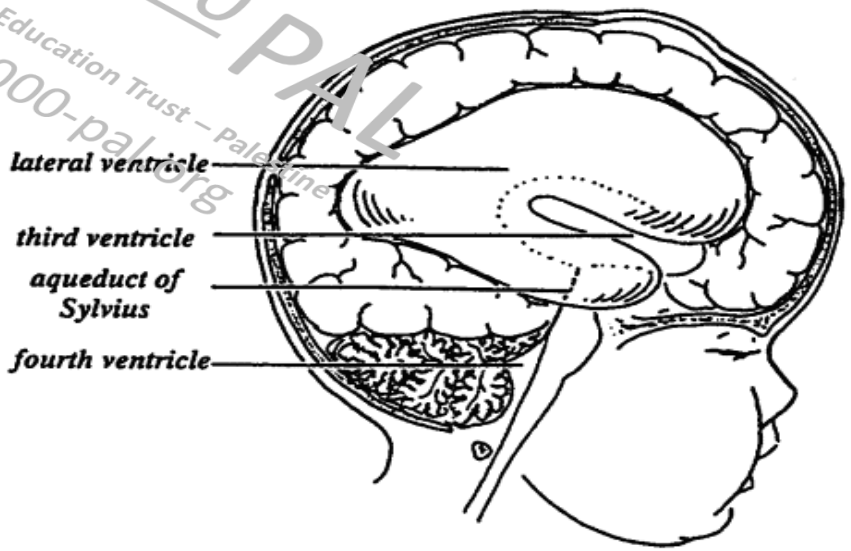
- is defined as an abnormal collection of cerebrospinal fluid with an associated dilatation of the cerebral ventricular system, a condition caused by an imbalance in the production or the absorption of the CSF through the ventricular system. An associate increase in head circumference follows.

- It could be congenital, that occurs in 3 /1000 live births
- or it could be acquired as in cases of hemorrhage, infection or neoplasm. , and if left untreated it may cause atrophy of the brain's white matter and severe neurological dysfunction

- Any imbalance in secretion or in absorption causes increase in the accumulation of CSF in the ventricles which become dilated and compress the brain substances against the surrounding rigid bony cranium.
- When this occurs before fusion of cranial sutures, it provides enlargement of skull and dilation of ventricles, but if sutures were closed, it may result in diastatic or opened sutures especially the sagital.

Enlarged ventricles with bulging fontanels





Pathophysiology

- **CSF** circulates through out the ventricular system, CSF flows from the lateral ventricle, to the third ventricle ----then flows through (aqua duct of Sylvie's)----------into the fourth ventricle, where more fluids formed,---------then through foramen of luschka and the midline foramen of magendie----------into the cisterna magna.
- Then CSF flows to the subarachnoid spaces where it is absorbed.

The pathway for CSF circulation is:

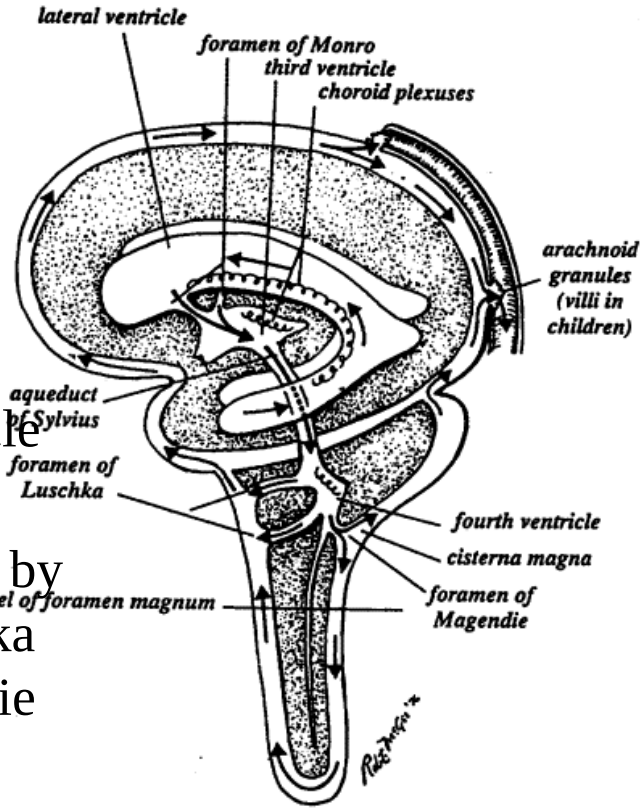
Lateral ventricles one in each hemisphere

Third ventricle

Aqueduct of Sylvius

Fourth Ventricle

To Subarachnoid Space by
(Paired Foramina of Luschka
Foramen of Magendie



Types and Causes:

- **1. Non Communicating hydrocephaly**: Obstruction to the flow of CSF through the ventricular system may result from:
- Congenital abnormalities.
- Structural defects, malformations, cyst, stenosis, tumors, and myelomeningocele.
- Prenatal maternal infections. (Toxoplasmosis or cytomegalovirus).
- neonatal infections. – (Neonatal meningo encephalitis bacterial or viral)
- Perinatal hemorrhage (anoxic or traumatic).

2. Communicating hydrocephaly

- Impaired absorption of CSF within the subarachnoid space may result from
- infection.
- subarachnoid bleeds.
- trauma.
- meningeal growth, neoplasm or tumors.
- Arachnoid cysts.

Hydrocephaly is associated with myelomeningocele.

- **Arnold –chiari**
- is One type of hydrocephaly resulted from the malformation that is
- characterized by herniation of small cerebellum, medulla pons and fourth ventricle into the cervical spinal canal through the enlarged foramen magnum results in obstruction in the flow of CSF causing hydrocephaly.

Clinical Manifestations

- C/M various according to the onset, and the associated malformations

Infancy:

- Head grows at an abnormal rate bulging fontanels + non pulsated anterior fontanel
- Dilated scalp veins palpable suture with cracked sound with percussion.
- Frontal protrusion with depressed eye + rotated down word. (Setting sun-sign).
- Irritable – lethargic infant – feed poorly.
- Changes in levels of consciousness
- Opisthotonos after extreme.
- Lower extremity sapasticity.

Infancy:

- Early infantile reflexes may persist.
- Infants with ACM – may exhibit behaviors that reflect cranial nerve dysfunction as a result of brainstem compression, including swallowing difficulties, stridor, apnea, aspiration, respiratory difficulties and arm weakness.
- If hydrocephalus is not treated it will disturb the development of the brainstem as manifested by poor sucking and feeding, high pitched cry – skull enlarged and cortex destroyed.
- If hydrophilic progressed infant may display emesis, seizures and cardio pulmonary distress.

Child hood

- **S + S** caused by high intracranial pressure mostly caused from posterior fossa neoplasm and aquduct stenosis.
- - Headache that relived after vomiting, or up right posture, papilledema strabismus, apathy, lethargic, and **Ataxia.**

Diagnostic evaluation:

- - U/S antenatal
- - Routine daily, measurement of HC in infancy and assessment for the presence of neurological signs.
- - CT + MRI are the primary diagnostic tools.
- -Echo en cephalography – to compare the ratio of lateral ventrice to cortex.
- - Sometimes isotope ventriculography. -Used to assess the flow and patency of existing shunts and to check the size of the ventricles.

Therapeutic management

- **Medical therapy is not effective**; it can be used only in slowly progressed cases as to give furosemide to decrease production of CSF, or medications to lower the intra cranial pressure if surgeries are contraindicated.

Surgical therapy

- 1- Direct removal of an obstruction such as resection of neoplasm, cyst, and hemotona, or in rare instances, fluid over production deceased by choroids plexus extirpation plexectomy or electric coagulation.
- 2-Shunt: Most children require a shunt that provides drainage of CSF from the ventricles to extra cranial compartment usually the peritoneum.

The preferred procedure is:
1- V.P shunts ventriculo peritoneal shunt

- A shunt drains CSF from the lateral ventricles to the peritoneum (VP shunt) are used most commonly because the infection implication are moderately less severe and because the VP shunt allows the child more space to grow. Children with shunts will need numerous shunt revisions while growing, and there is a greater allowance for excess tubing, which minimizes the number of Revisions needed as the child grows.

2- Ventriculo a trial shunt - V.A shunt

- A shunt drains CSF from the lateral ventricle to right atrium, is used for older children or for those with abdominal pathology but it is contra indicated in children with cardio pulmonary disease or elevated CSF protein.

3- Ventricular bypass

- Maybe used in older children to relieve the stenosis or masses but not used infant because spaces are poorly developed.

- 4- **Ventriculo pleural shunts** - are used sometimes for children over 5 years

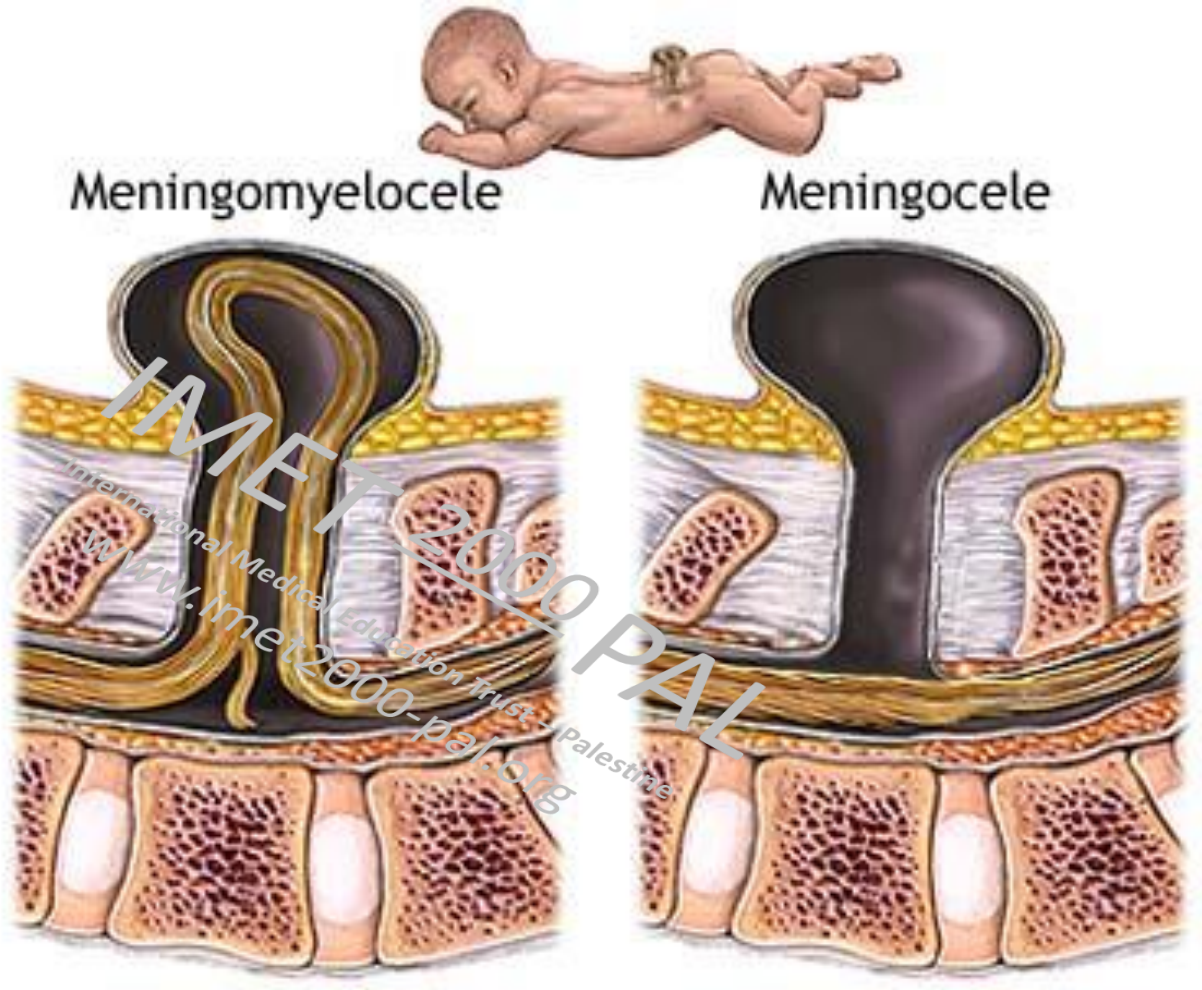
Complications associated with shunt

- - The most common complication fro shunts is **infection**. This can be life threading as it involves many vital organs such as the kidneys as well as the brain.
- - All shunts are subject to mechanical difficulties such as **kinking, plugging, separation, migration** of the tubing.
- - Mal function caused by mechanical obstruction either within the ventricles from particular matter tissue or exudates, or from thrombosis or displacement.
- -The **biggest indicator** for shunt malfunction is the **conscious level**.
- - The child will exhibit signs and symptoms of **intracranial pressure** when the shunt is obstructed and should be treated as an emergency.

Nursing considerations

- The goals of nursing care of the child with hydrocephalus include:
- - Prevent complications of hydrocephalus.
- -observe for signs of ICP which indicates obstruction of the shunt.
- - Measure head daily
- - Provide education and emotional supports to the family.
- -Neurological assessment and evaluation of pupil dilation.
- - Maintaining adequate nutrition
- -Observe signs of infection in the post operative period such as elevated temperature, poor sucking, or vomiting.

Spina Bifida



Definition:

- Malformation of the neural tube leading to herniation through an abnormality in the canal of the vertebral column.
- When the spinal cord itself is not fully formed however, the nerves do not develop as they should and the baby will have **Myelomeningocele**

Incidence

- In some cases it has occurred in the family before, but this is relatively uncommon, In the United States, spina bifida is the second most common birth defect and affects about one out of every one thousand pregnancies. also there is a high incidence of spina bifida in Palestine.

Prenatal Detection:

- There are several tests available to pregnant women which can be used to detect spina bifida before the baby is born, **maternal serum alpha fetoprotein** (MSAFP) The blood test which is done on the mother's blood around the 16th week of pregnancy. This test is not specific for spina bifida, however. An error in the dates of the pregnancy, multiple babies, as well as other birth defects can all cause an abnormal reading. If the alpha fetoprotein level is abnormal, additional testing is recommended.

Causes of spina bifida

- Causes are unknown, but in a small number of women it appears to be caused by certain medications most often anti seizure drugs. In the majority of cases the cause of the spina bifida is never determined. However, it is now known that taking folic acid a B vitamin, before a woman becomes pregnant will reduce the chances that the baby will have spina bifida and related conditions of the brain and spinal cord.

Pathophysiology

- This problem occurs very early in pregnancy, Normally at the end of the fourth week of pregnancy, the neural tube formation is complete, if there is failure for the fusion of the lamina of the vertebra or splitting closed tube will result in spina bifida that can adversely affect many body systems including the nervous system, the bones and muscles as well as the kidneys and bladder. The point along the spinal cord where the undeveloped area occurs is called the "level" of the spina bifida

pathophysilogy

- The higher up the spinal column the "level" occurs, the greater the effect on normal nerve functions. Some individuals with low levels of spina bifida can walk with little or no assistance whereas those with higher levels will require braces and in cases of very high levels, wheelchairs, to get around.

Types of spina bifida

- **1. Spina bifida occulta:** In this type the posterior vertebral arch fails to fuse, that only the bones of the spinal column will be incompletely developed, but the nerves beneath will be normal, the spinal cord and meninges does not herniated and the abnormality is not visualized externally. Does not cause neurologic problems such as paralysis or weakness and is not medically significant.

2. Spina bifida cystica

- the most severe form of spina bifida, the opening is larger, and the abnormality is shown as sac protruding in the external surface of the back and it is two types:

- **A. Meningocele:** a sac like cyst of meninges, filled with CSF, Protrude through defects in the bones of the spine
- **B. meningomyelocele:** a herniation allows for meninges, CSF, and part of spinal cord with nerves, through the bony defect.

Clinical manifestations

- a sac covered by tiny membrane likely to tear and to leak, can be located any where along spinal cord mostly lumber or lumbosacral region, and can be found at the cervical region.

Clinical manifestations

- **If the defect is below the second lumber vertebra:**
- Incontinence which means bladder nerve supply is affected. *
- *Partial paralysis of the lower limbs.
- *Sensory disturbances.
- *Poor anal sphincter control.
- **If the defect is below the third sacral vertebra**
- No motor involvement.

Treatments Available for Children and Adults with Spina Bifida

- Because spina bifida affects so many body systems it is important that professionals from many areas be consulted to provide up-to-date, comprehensive medical, psychological and social evaluation, support and treatment.

What is the initial management of patients with myelomenigocele

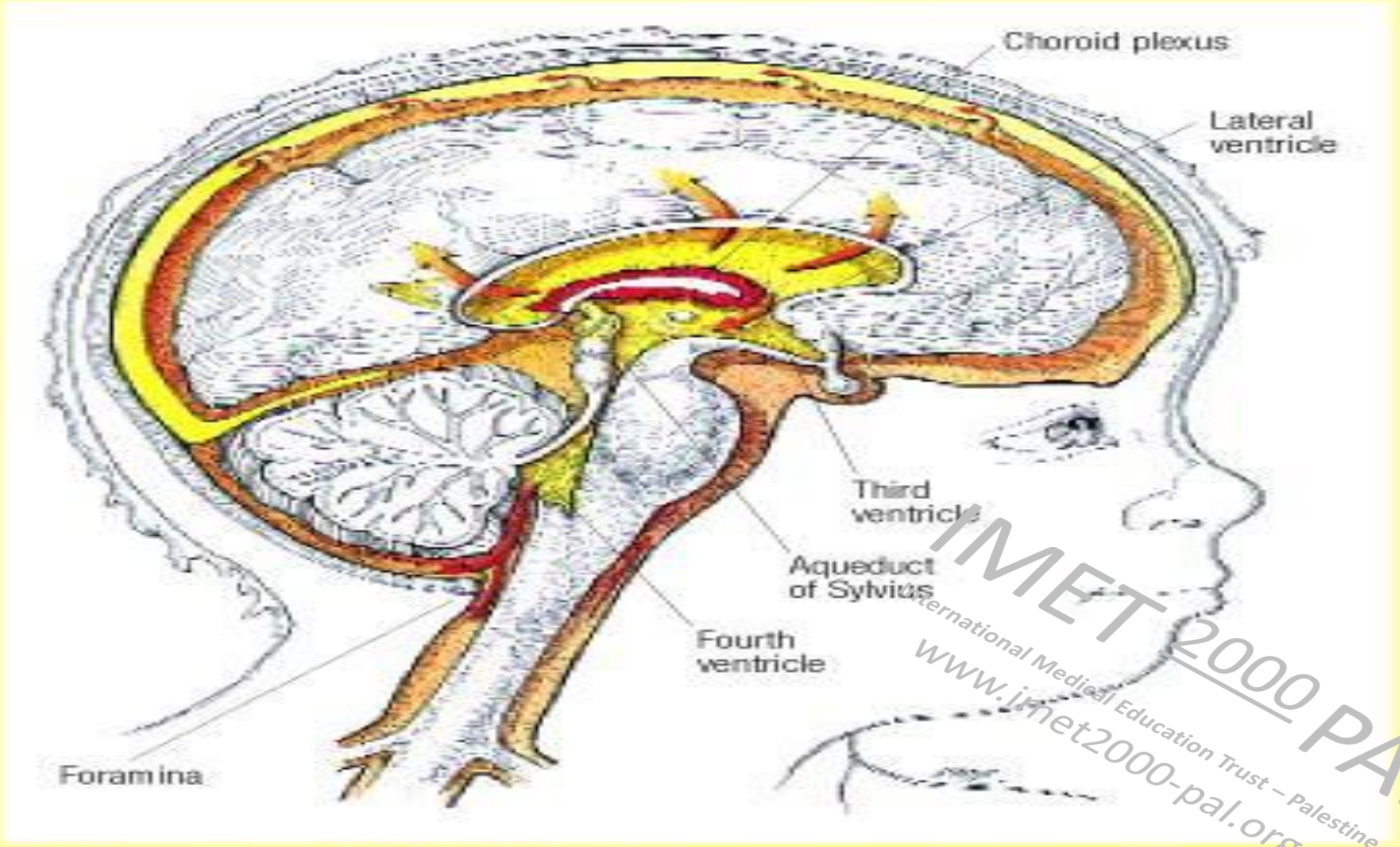
- On admission the size of the defect need to be measured.
- The lesion needs to be covered with moist dressing, and needs to be kept continuously moist.
- Surgical closure needs to be performed within 24 hours.
- Neurological assessment:
- Watch for spontaneous movement of the lower extremity (good spontaneous movement correlates with better later functional outcome)
- The lowest level of function needs to be assessed by checking the response of the lower extremities to painful stimulation.
- Hydrocephalus needs to be watched for.

What are the surgical considerations?

- Closure of the defect is not associated with improvement of neurological function, but is associated with a lower infection rate. Myelomeningoceles need to be closed with 24 hours whether or not the membranes are intact. During the operative procedure extreme care needs to be taken in protecting the exposed neural tissue. This tissue needs to be dissected and placed back into as normal of a location as possible

- After the overlying layers need to be closed in a watertight fashion. The assistance of a plastic surgeon may be necessary to close large defects.

FIGURE 4



Normal cerebrospinal fluid pathway. Produced in the choroid plexus of the lateral ventricle, CSF moves through the third ventricle, the Aqueduct of Sylvius, and on into the fourth ventricle, then out through the foramina. Any obstruction around the aqueduct or fourth ventricle leads to obstructive hydrocephalus and signs of elevated intracranial pressure.

Adapted from Edwards M, Derechin M: *About Hydrocephalus—A Book for Parents*. San Francisco, CA, The Hydrocephalus Association, 1986