MALFORMATIONS OF CNS

(1). Cranial deformities (Craniosynostosis)

Craniosynostosis is a congenital deformity of the infant skull that occurs when the fibrous joints between the bones of the skull (called cranial sutures) close prematurely. Due to this closure, the baby develops an abnormally shaped skull because the bones do not expand normally with the growth of the brain.

→ Causes & types

There are two types:

a) Nonsyndromic craniosynostosis
b) Syndromic craniosynostosis

Often the cause of craniosynostosis is not known, but sometimes it's related to genetic disorders.

- Nonsyndromic craniosynostosis is the most common type & its cause is unknown, although its thought to be a combination of genes and environmental factors.
- Syndromic craniosynostosis is caused by certain genetic syndromes, such as Apert syndrome, Pfeiffer syndrome or Crouzon syndrome, which can affect baby's skull development.

→ Complications

- Permanent head & facial deformity
- Poor self esteem & social isolation
- Seizures

→ Signs & symptoms.

- Short & wide appearance of head.
- Increased intracranial pressure.

- Developmental delays
- Cognitive impairment
- Eye movement disorders
- Seizures
- A full or bulging fontanelle
- Very noticeable scalp veins
- Increased head circumference

→ Diagnosis

- History collection
- Physical examination
- CT scan

→ Management

The key to treating craniosynostosis is early detection and treatment. Surgery is the typically the recommended treatment.

For seizures anticonvulsant drugs are prescribed such as Phenytoin, Primidone.

Surgical management

- Calvarial Vault Remodeling

In this procedure, the surgeon makes an incision in the infant's scalp & corrects the shape of the head by moving the area of the skull that is abnormally or prematurely fused, & then reshapes the skull so it can take more of a round contour. Surgery can last upto six hours.

- Endoscopic Craniosynostosis

This procedure involves the use of an endoscope, a small tube that the surgeon can look through & see immediately inside and outside the skull through very small incisions in the scalp. The surgeon opens the prematurely fused suture to enable the baby's brain to grow normally.

Nursing Management

- Child will be monitored closely.
- Tube feeding may be preferred for few days.
- The head may be raised to prevent swelling of face
- Care of tubes and lines must be taken.
- Restraints may be used to prevent touching of face or head.
- Hygiene & grooming should be encouraged.

(2) Spina Bifida

It is the congenital defect that involves the incomplete development of spinal cord or its covering. It occurs at the end of first month of pregnancy when the two sides of the embryo's spine fails to join together leaving an open area.

- Causes
  - Maternal diabetes
  - Family history
  - Obesity
  - Medications such as some anticonvulsants
  - Pregnant women taking valproic acid
  - Genetic basis
  - Folic acid deficiency
- Types

  There types

  a) Spina bifida occulta

  b) Spina bifida cystica with meningocele

  c) Spina bifida cystica with myelomeningocele

  - In Spina bifida occulta, the outer part of some of the vertebrae is not completely closed

- In meningocele, the vertebrae develop normally but meninges are forced into gaps between the vertebrae
- In myelomeningocele, the unfused portion of the spinal cord allows the spinal cord to protrude through an opening.

- Clinical Features
  - Orthopedic abnormalities (ie, hip dislocation)
  - Bladder & bowel control problems.
  - Pressure sores & skin irritations.
  - Abnormal eye movement
  - Paralysis.
  - Allergy to latex
  - Scoliosis
  - Back pain.
  - Partial or complete lack of sensation

- Complications
  - Attention deficit hyperactive disorder, Meningitis
  - Arnold Chiari II malformation (Back portion of the brain is displaced from the back of the skull down into the upper neck)

- Diagnosis
  - History collection
  - Physical examination
  - Pregnancy screening

(Testing the mother's blood (AFP screening) or a detailed fetal USG).

Increased levels of maternal serum alphafetoprotein (MSAFP) should be followed up by two tests— an USG of the fetal spine & amniocentesis of the mothers amniotic fluid (to test for alpha-fetoprotein and acetylcholinesterase).

Management

- Medical management

There is no cure for spina bifida. Damaged nerve tissues cannot be repaired nor its function be restored. Treatment depends on how severe the defect is. Medication involves treating the complications and signs of spina bifida.

- Surgical management

The surgery involves putting the meninges back in place & closing the opening in vertebral. The surgery is done within 24-48 hours after birth.

- Nursing management

Pre operative management

- Position the child in prone with legs abducted. This reduces tension & risk of trauma.
- Put the child in an incubator or warmer area without clothes.
- Apply dressing to avoid drying of the area due to heat in the incubator.
- Change dressing two - four hourly, to avoid drying
- Use Normal Saline or silver nitrate in dressing.
- Change the child's position every two hours, to promote circulation & prevent development of decubitus sore.
- Prepare the mother psychologically.
- Apply gentle pressure to suprapubic area to facilitate urine emptying.

Post-operative management

- Position the child in prone to avoid pressure on suture, or side lying position alternatively.
- monitor the child's vital signs every 30 minutes until stable.
- Use all measures to avoid any infection eg handwashing
- Encourage the mother to continue breastfeeding.
- Resume the feeding after the effects of anesthesia.
- Remove the dressing after 48 hrs to check any signs of bleeding & bulging.
- Observe for leakage.

(3) Hydrocephalus

Hydrocephalus is the abnormal accumulation of cerebrospinal fluid in the intracranial spaces.

- Causes

a) Congenital
- Intrauterine infections (eg rubella, toxoplasmosis)
- Congenital brain tumor
- Intracranial hemorrhage
- Congenital malformations like Arnold-chiari malformation
- Malformations of arachnoid villi

B) Acquired
- Meningitis, encephalitis
- Birth injury, head injury
- Degenerative atrophy of brain
- Subdural hematoma, abscess

- Types

a) Communicating hydrocephalus.
It is a condition that results when the arachnoid villi are unable to adequately reabsorb CSF.

B) Non-communicating hydrocephalus
It is a condition that results when the ventricular system does not communicate with the arachnoid villi due to some obstruction in the normal pathways of CSF flow.

- Clinical Features
- An unusually large head.
- Rapid increase in the size of head.
- A bulging or tense soft spot on the top of head
- Macewen's sign (Typical cracked-pot sound of the skull bone)

shiny scalp with prominent scalp veins
- Poor feeding
- Seizures
- Muscle tone deficits
- Vomiting
- Sleepiness
- Irritability

- Diagnosis

- History collection
- Physical examination
- Macewan's sign
- MRI
- CT Scan
- Cranial USG
- X-ray Skull

- Management

Medical management

The medical management is done to reduce increased ICP by Carbonic anhydrase inhibitor, Acetazolamide (Diamox) 50 mg/kg/day to reduce CSF production in slow progressive hydrocephalus. Oral glycerol & isosorbide can also be used.

Surgical management

- Ventriculostomy
- Choroid plexectomy
- Ventriculoperitoneal Shunt

Nursing management

- Providing adequate nutrition by exclusive breastfeeding to the neonates (the infants upto 6 months of age

For older children, offering small frequent feeding & placing the infant or child in semisitting position.
- Maintaining skin integrity. Firm or soft pillow under the child's head, frequent change of position & keeping the area clean & dry.
- Reducing parental anxiety by explanation, reassurance & encouraging to express feeling.
- Preventing infections by asptic technique.

- Complications

- Herniation of brain
- Visual problems
- Persistent increased ICP.
- Developmental delay.
- Infections.