Neural Tube Defects

Spina Bifida | Anencephaly | Encephalocele

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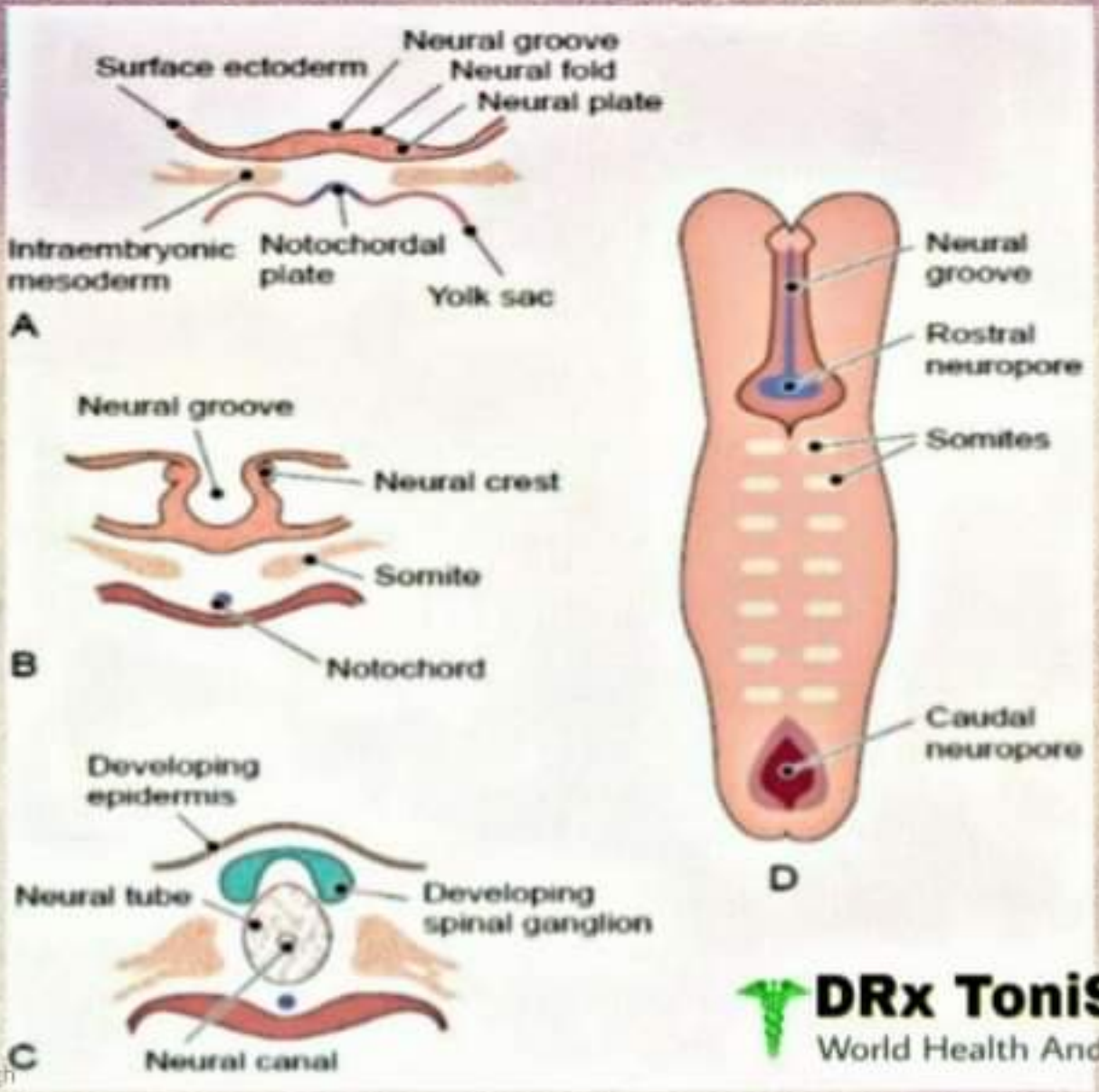
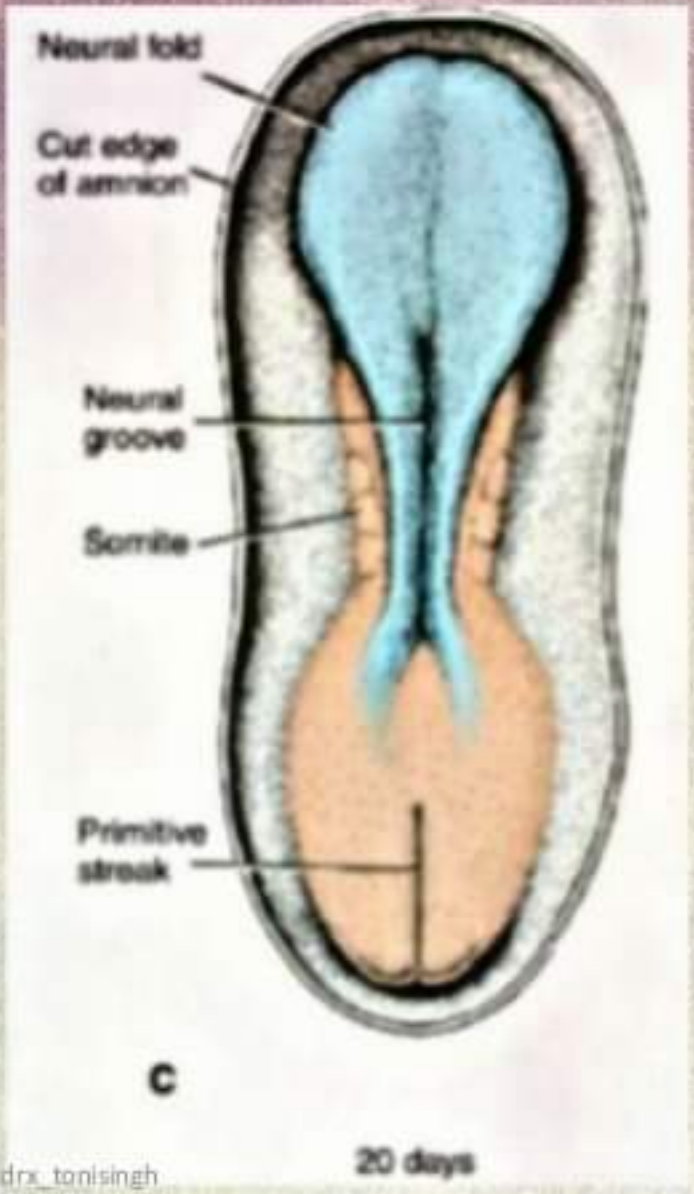
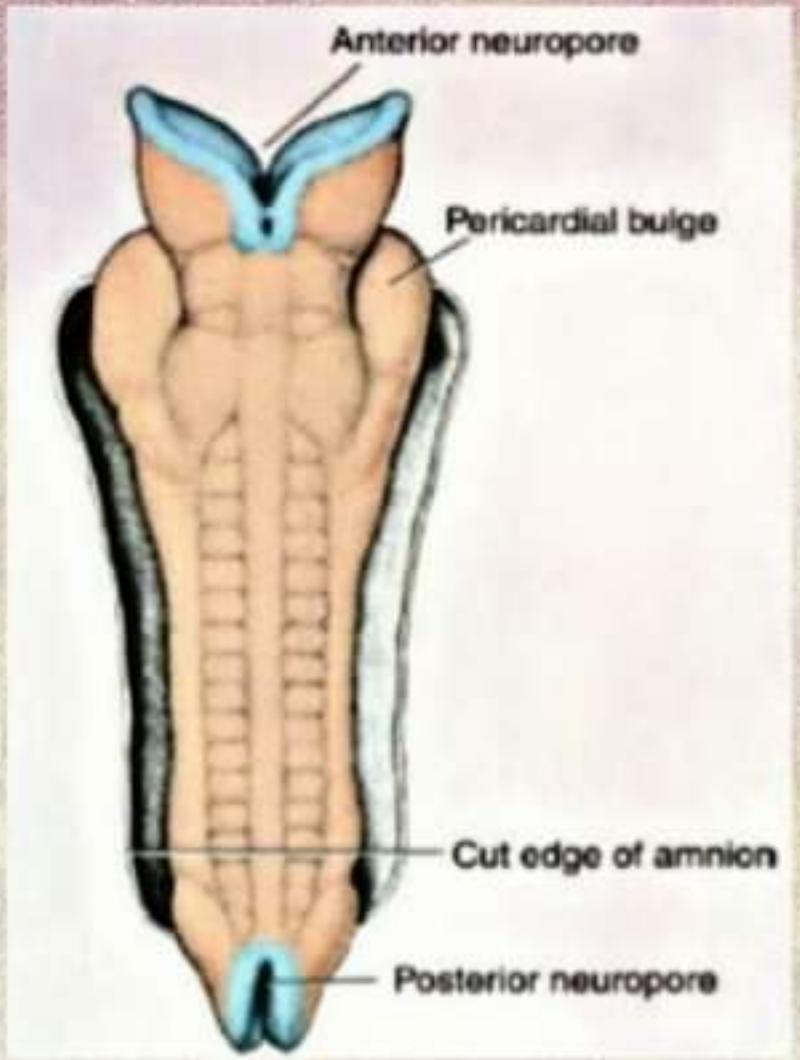
Introduction

- The human nervous system originates from the primitive ectoderm that also develops into the epidermis.
- The ectoderm, endoderm, and mesoderm form the three primary germ layers that are developed by the 3rd wk.
- The endoderm, particularly the notochordal plate and the intraembryonic mesoderm, induces the overlying ectoderm to develop the neural plate in the 3rd wk of development

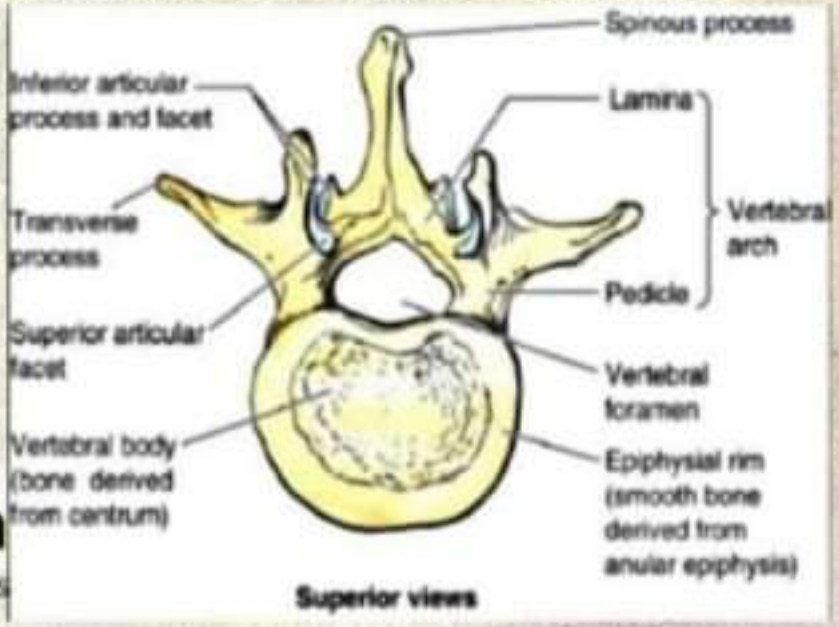
Failure of normal induction is responsible for most of the NTDs, as well as disorders of prosencephalic development.







- Neural tube defects (NTDs) account for the largest proportion of congenital anomalies of the CNS and result from failure of the neural tube to close spontaneously between the 3rd and 4th wk of in utero development.
- Although the precise cause of NTDs remains unknown,



Etiology

- The exact causes of neural tube defects are not known.
- Genetic
- Nutritional Factors
- Environmental factors.
- Poor intake of folic -acid (also known as folate)
- Obese Women
- Intake of anti epileptic medications during pregnancy

Pathophysiology

- THE NEURAL PLATE APPEARS ON THE 17TH DAY OF GESTATION AS A THICKENING OF THE EMBRYONIC ECTODERM

- ON DAY 18, THE **NEURAL PLATE** FOLDS ALONG THE MIDLINE

- BY THE END OF THE THIRD GESTATIONAL WEEK, THE NEURAL FOLDS FUSE TO FORM **NEURAL TUBE.**

- FUSION BEGINS AT THE HINDBRAIN-CERVICAL JUNCTION FIRST PROCEEDS ROSTRALLY AND THEN CAUDALLY

- THEN, THE ANTERIOR AND POSTERIOR ENDS (NEUROPORES) CLOSES TO FORM SPINAL CORD

- DEPENDING ON THE POINT OF INTERUPPTION IN NEURAL TUBE FORMATION DEVELOPS NTDS MAY AFFECT THE BRAIN (ANENCEPHALY)OR SPINAL CORD (SPINA BIFIDA).

Classification

- NTDs are classified as:

1. Open: often involve the entire CNS with neural tissue is exposed and CSF leaking
2. Closed: localised to the spine; brain rarely affected neural tissue not exposed although the skin covering the defect may be dysplastic

1. Cranial NTDs

- Anencephaly
- Encephalocele

  - meningocele

  - meningomyelocele
- Congenital dermal sinus

2. Spinal NTDs

- Spina bifida cystica
- Spina bifida occulta
- Myelomeningocele
- Meningocele
- Congenital dermal sinus
- Caudal agenesis

Risk Factors

- Family history of NTDs
- Certain syndromes and chromosomal disorders.
- Low dietary low folic acid
- Administration of sodium valproate and folic acid antagonists, e.g. some anti-epileptics, trimethoprim
- hyperthermia
- malnutrition
- low red cell folate levels
- chemicals, Radiation
- maternal obesity or diabetes
- genetic determinants (mutations in folate-responsive or folate-dependent enzyme pathways)

Sign And Symptoms

- Loss of bladder or bowel control
- Partial or complete lack of sensation
- Partial or complete paralysis of the legs
- Weakness of the hips, legs, or feet of a newborn
- Other symptoms may include:
- Abnormal feet or legs, such as clubfoot
- Build up of fluid inside the skull (hydrocephalus)
- Hair present at sacral region
- Dimpling of the sacral area

Investigations

- MRI: best for imaging neural tissue & identifying contents of the defect
- CT: direct visualisation of the bony defect and anatomy
- Ultrasound: for prenatal screening
- X-ray
- Maternal serum Alpha feto protein at 16-20 weeks.
- Aminiotic Alpha feto protein
- Aminiotic acetyl cholineterase

Prenatal Screening

- Failure of closure of the neural tube allows excretion of fetal substances (α-fetoprotein [AFP], acetylcholinesterase) into the amniotic fluid, serving as biochemical markers for a NTD.
- Prenatal screening of maternal serum for AFP in the 16th-18th wk of gestation is an effective method for identifying pregnancies at risk for fetuses with NTDs in utero

Management

Medical Care

The patient should be positioned in the prone position to prevent pressure on the defect.

- The newborn with an open NTD should be kept warm and the defect covered with a sterile wet saline dressing.
- Intravenous antibiotic should be initiated.

Surgical Management

- Neurosurgical repair of the defect is considered the mainstay of treatment for open spina bifida.
- Closed spina bifida does not usually warrant any immediate surgery.
- The cele closure is typically performed within 1 to 3 days of delivery.
- Neonates born with severe hydrocephalus should have ventriculoperitoneal shunt placed concurrently.

Nursing Management

ASSESSMENT

- Depends on the spinal involvement.
- Visible spinal defect
- Flaccid paralysis of legs.
- Altered bowel and bladder pattern.
- Perform neurological assessment

Nursing Interventions

- Evaluate the sac and measure lesion .
- Monitor for increased ICP
- Measure head circumfrence.
- Protect sac with non adherent moist dressing
- Place child in prone position
- Use aseptic techniques
- Monitor for early signs of infection
- Administer Antibiotic.
- Prepare Family for surgery

Prevention of NTDs

- Experts recommend that all women of childbearing age take a daily supplement of 400 micrograms (mcg) of folic acid.
- Educate mothers regarding intake of folic acid especially in periconception period and in first trimester as well.
- Women already had first pregnancy with NTD should take a daily 4mg tablet of folic acid for at least one month before conception and then throughout the first 12 weeks of pregnancy.
- Genetic Counseling or screening.