Congenital abnormalities in the spinal cord

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- Is due to failure of closure of neural tube between 3rd and 4th week of embryonic development

Aeitiology

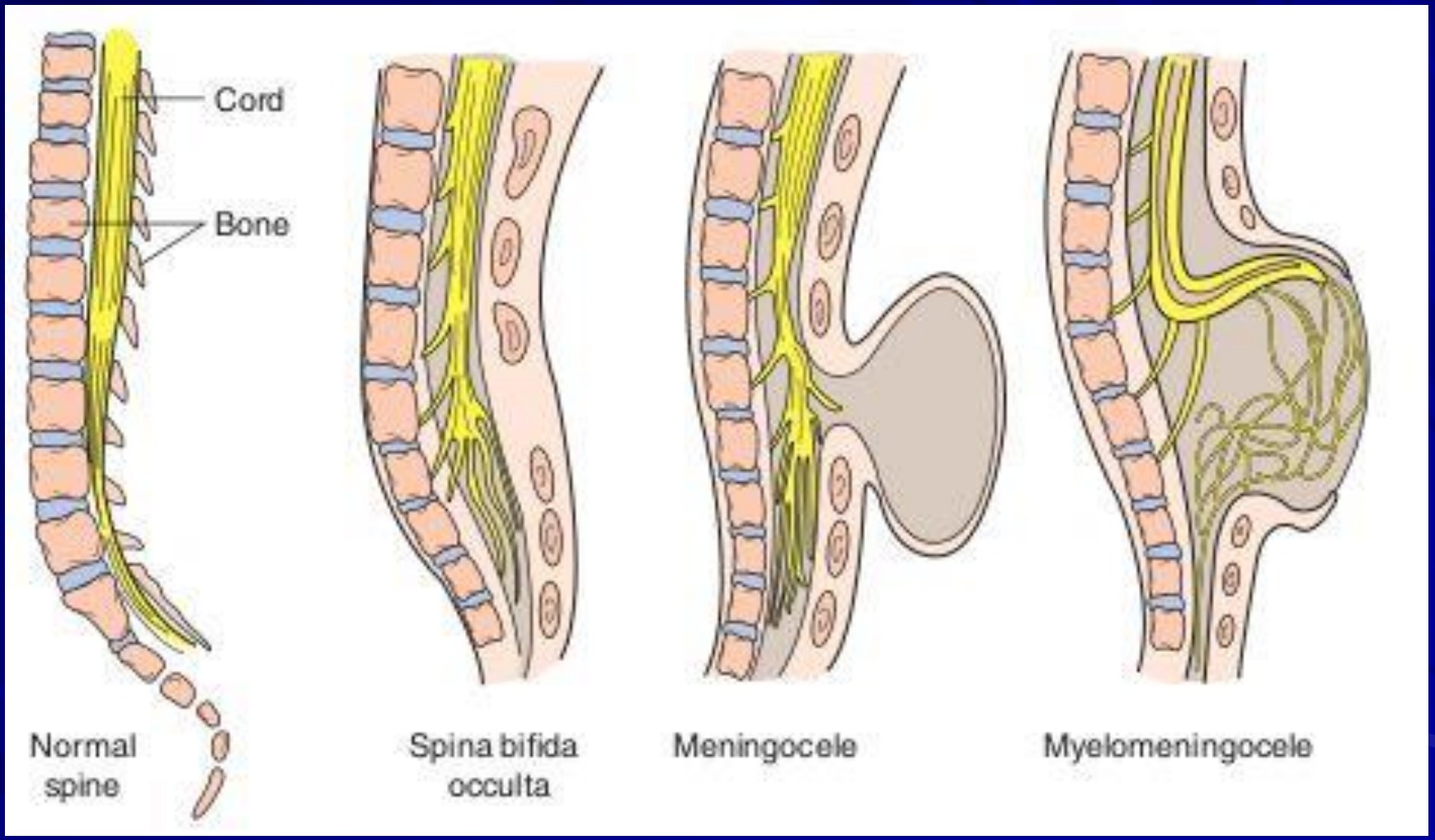
- Multifactorial
  - Genetic determinants
  - Environmental
    - Folate deficiency in pregnancy
    - Drugs (eg: valproate)
    - Hyperthermia
    - Maternal obesity/ diabetes

- Spina bifida occulta
- Meningocele
- Myelomeningocele

Spina bifida occulta

Meningocele

Myelomeningocele



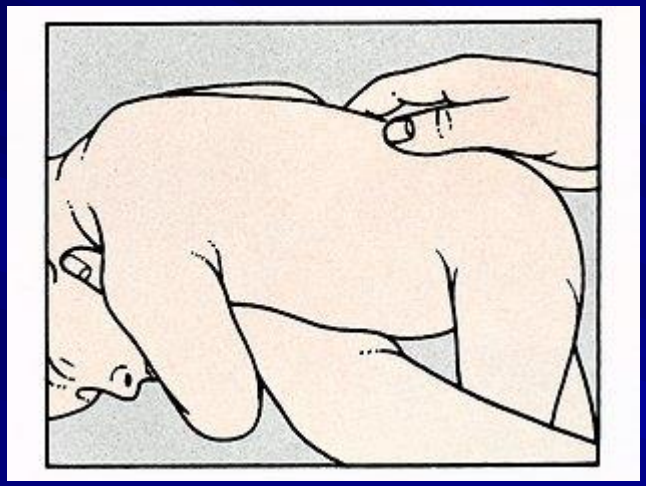
Spina bifida occulta

Meningocele

Myelomeningocele

Presentation of spinal cord abnormalities

- Usually detected during newborn examination



Spina bifida occulta

- Effects
  - Usually asymptomatic
- Cutaneous manifestation
  - Pit
  - Lump
  - Dermal sinus
  - Hemangioma
  - Discolouration
  - Hairy patch

Spina bifida occulta (cont.)

- Diagnosis
  - X ray - defect in closure of the posterior vertebral arches and laminae, typically involving L5 and S1

Myelomeningocele

- Incidence 1:4000

- Clinical features
  - Anywhere along neuraxis – 75% lumbosacral
  - Cystic structure covered by a thin layer of partially epithelialized tissue
  - Effects depend on the level of the lesion

Myelomeningocele

- Associated lesions
  - Lower limb flaccid paralysis
  - Sensory deficits
  - Bladder incontinence / retention
  - Bowel dysfunction – incontinence/ faecal impaction
  - Skeletal deformities – club foot, ankle/knee contractures, subluxation of hip
  - Hydrocephalus - type II Chiari malformation

Myelomeningocele

- Management
  - Multidisciplinary team
  - Surgery-
    - Surgery of myelomeningocele -within days
    - Operative procedures for club foot/ hips
  - Neurogenic bladder
    - CISC – to minimize residual volume
    - Prevention of UTI – prophylactic antibiotics
    - Regular assessment of renal function
    - Artificial urinary sphincter

Myelomeningocele

- Management – Neonatal
  - Lool for CSF leakage
  - Prone/lateral positioning
  - Prevent infection-Sterile gloves , cover with sterile dressing
  - Prevent heat loss-cover with plastic bag
  - Prophylactic antibiotics
  - Refer to neuro surgeons-lesion should be closed within 72 hours

Myelomeningocele

- Management
  - Bowel dysfunction
    - bowel-training with a regimen of timed enemas or suppositories
  - Physiotherapy
    - Aim – functional ambulation
  - Parental counselling
    - facts, prognostic information, management strategies and timelines
    - discussions with other parents of children with NTDs

Myelomeningocele

- Prognosis
  - Mortality rate 10%-15%
  - Majority of survivors have normal intelligence
  - Renal dysfunction is an important determinant of mortality

Meningocele

- Meninges herniate through a defect in posterior vertibral arches
- Spinal cord is usually normal
- Presentation
  - Fluctuant midline mass with translumination but well covered by skin

Meningocele (cont.)

- Some are neurologically normal

  Others….

- Investigations
  - X-ray
  - USS
  - MRI - to determine the extent of neural tissue involvement (if any) and associated anomalies

Meningocele (cont.)

- Treatment
  - Surgery –not urgent

Antenatal diagnosis

- Maternal serum alpha-foeto proteins
- Foetal ultrasound scan

Prevention of NTB

- Maternal periconceptional use of folic acid supplementation
  - Reduces the incidence of NTDs in pregnancies at risk by at least 50%.
  - should be initiated before conception and continued until at least the 12th week of gestation