Spina Bifida
(Spinal Dysraphism)
(Neural Tube Defects)

Amr M. Tayel
Lecturer of neurosurgery
South Valley university

Objectives

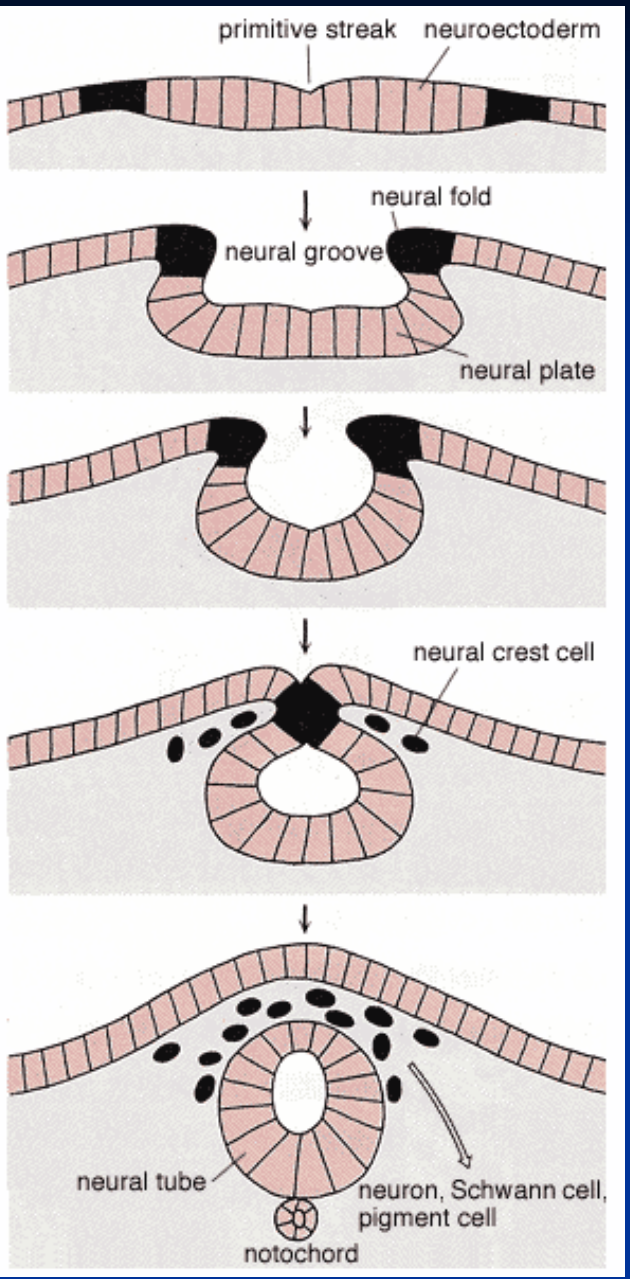
- Definition
- Pathology and types
- Etiology
- Associated pathology
- Diagnosis
- Management

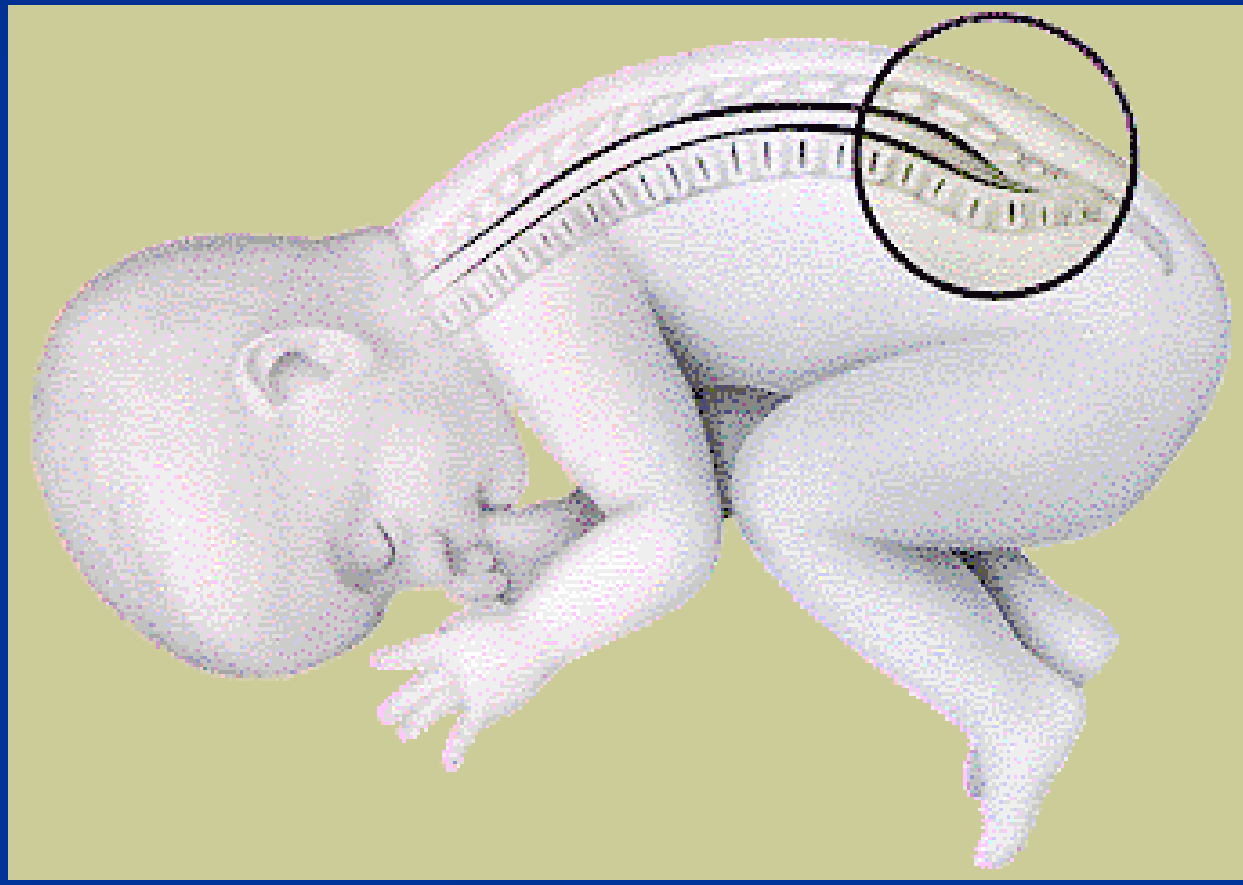
Spina Bifida definition

- Spina Bifida Latin "*split spine*"
- Failure in closure of the neural tube during early embryologic development leading to midline defect.

Pathology

- Neural tube formation days 18-27
- Neural folds fuse in midline days 22-23
- Disjunction of superficial ectoderm from neural tube
- Failure of any step → dysraphia → maldevelopment of the neural structures and adjacent mesodermal structures


primitive streak
neuroectoderm
neural fold
neural groove
neural plate
neural crest cell
neural tube
neuron, Schwann cell,
pigment cell
notochord



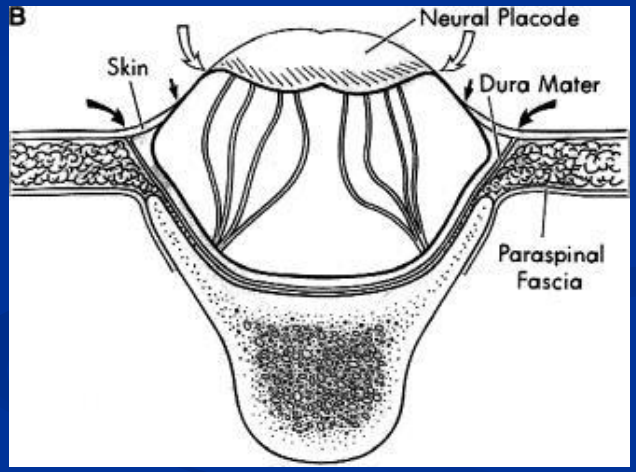
Types of spina bifida

- spina bifida cystica (manifesta):
  - ✓ Menigeocel
  - ✓ Meningeomyelocele
  - ✓ Lipomeningocele
  - ✓ Lipomyelomeningeocele
  - ✓ Myelocele = neural tissue

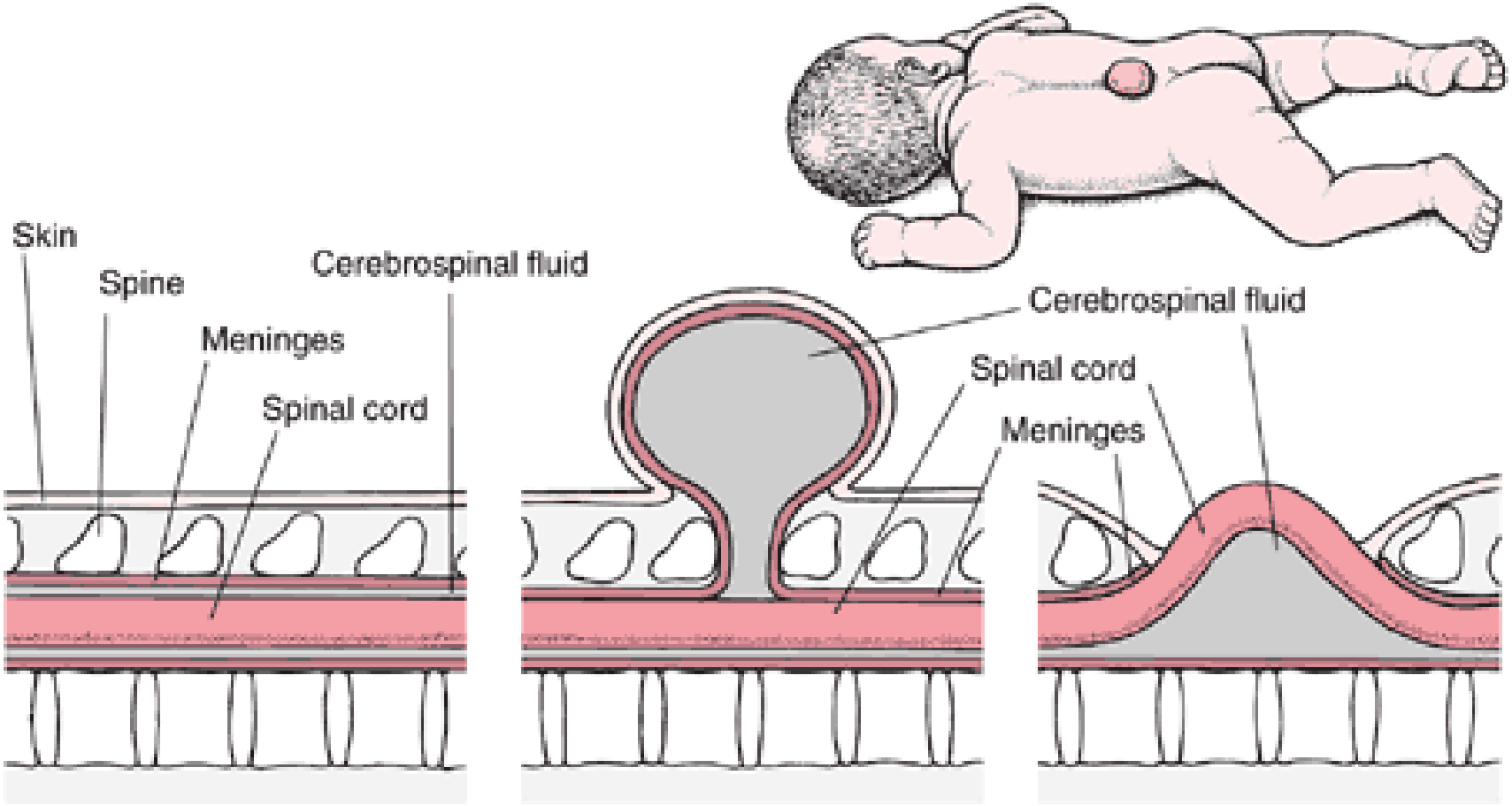
- spina bifida occulta

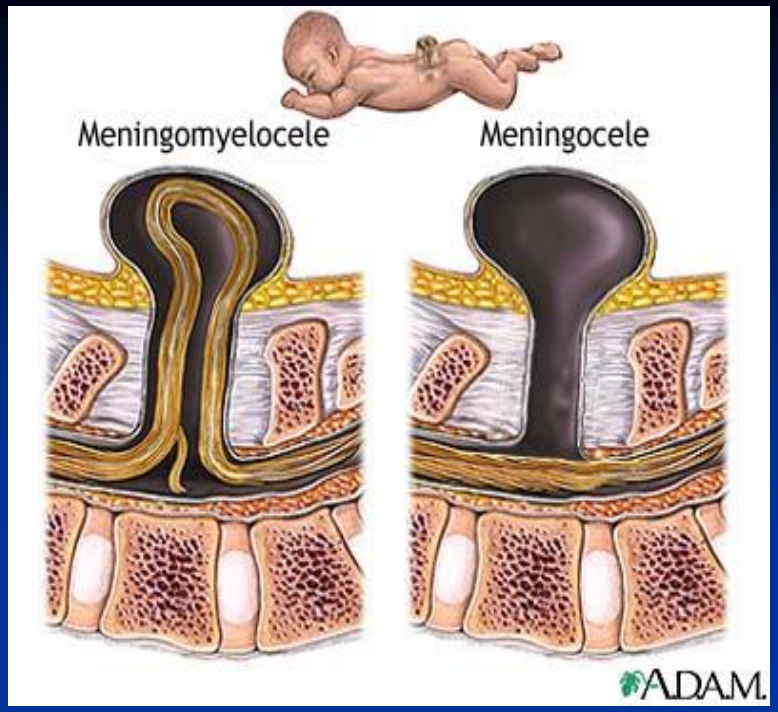
- Meningocele = meninges + CSF
- Meningomyelocele = meninges + neural tissue + CSF

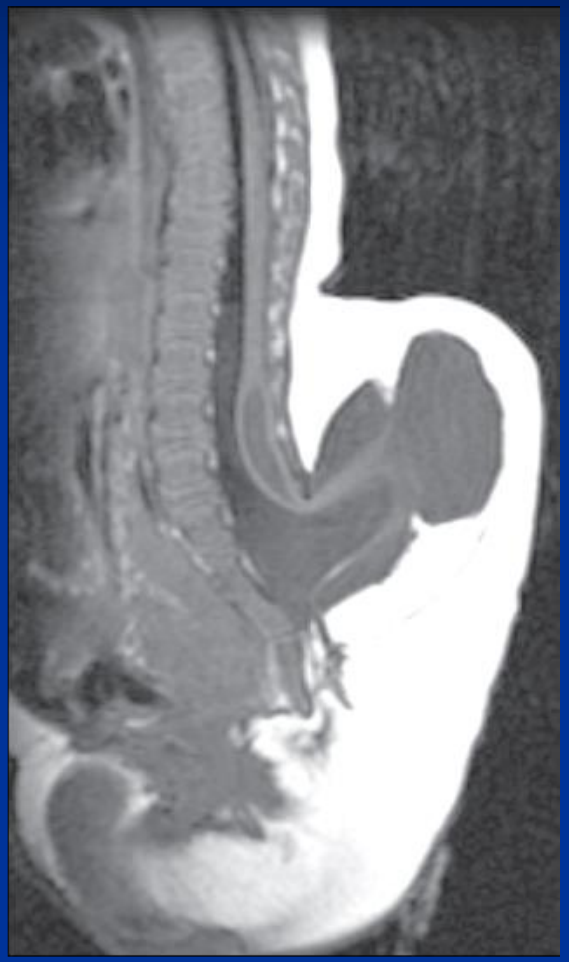
(It is the most clinically seen)

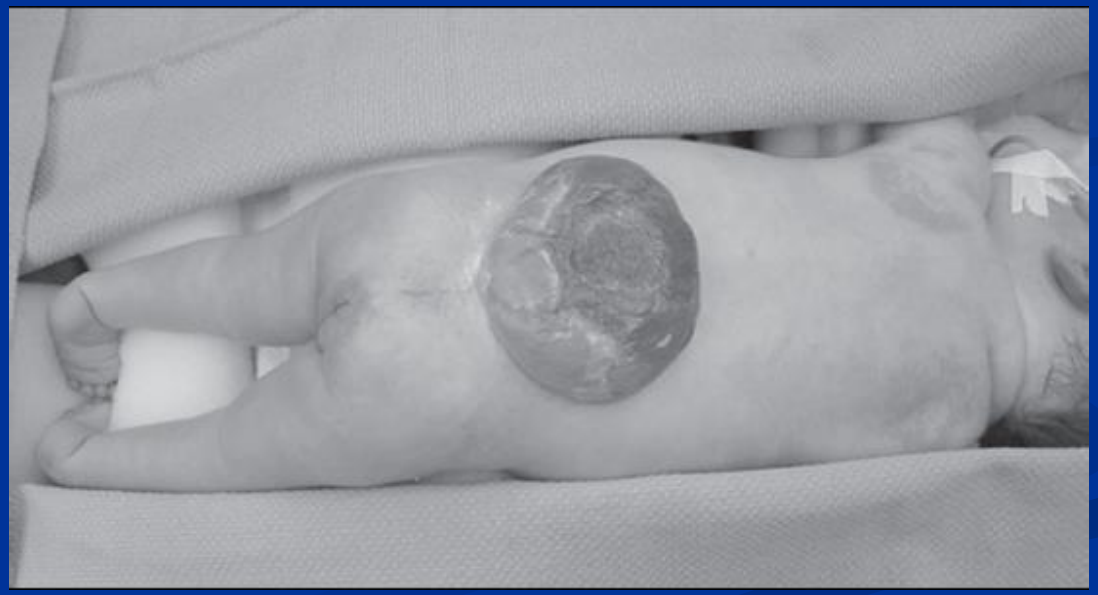


- Lipomeningocele
- lipomyelomeningeocele
- Myelocele = neural tissue


Skin
Spine
Meninges
Spinal cord
Cerebrospinal fluid
Cerebrospinal fluid
Spinal cord
Meninges
Normal Anatomy
Meningocele
Myelocele

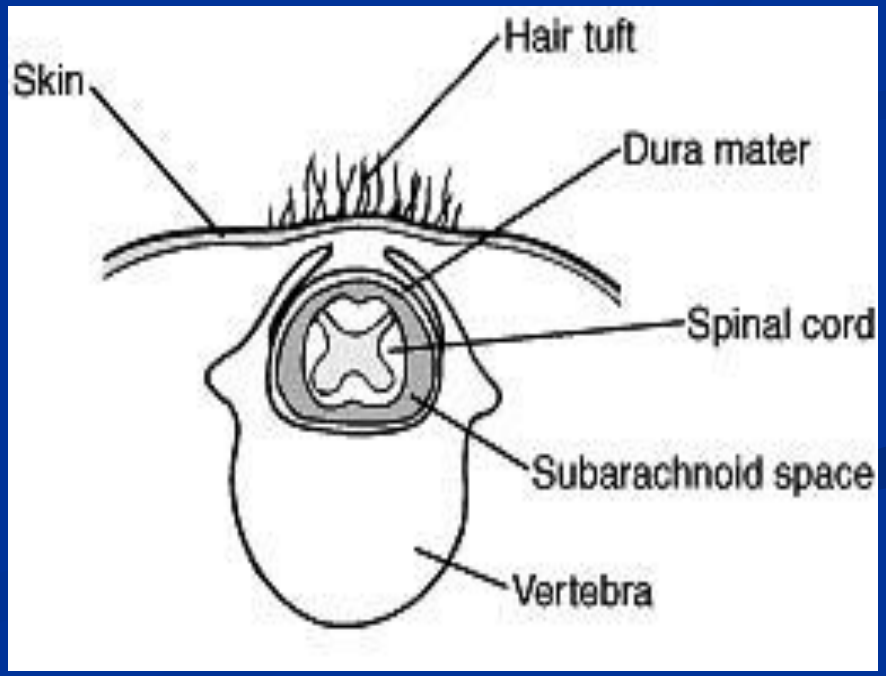

Meningomyelocele
Meningocele
A.D.A.M.





- Spina bifida occulta:

condition involving nonfusion of the halves of the vertebral arches without disturbance of the underlying neural tissue



Etiology
??????

- Nutritional deficiency / Metabolic defect
  - Folic acid deficiency
- Teratogens
  - Valproic acid, carbamezapine, cytochalasins, calcium channel blockers
- Genetics
  - Lower risk in blacks, increased risk caucasian
  - Increased risk if previous child with NTD
  - Point mutations in folate receptor genes C677G, A127T

Associated pathology

(mostly with meningeomyelocele)

- Arnold-Chiari Malformation Type II
  - ~80% incidence with MMC
  - ~20% develop symptoms of brainstem dysfunction:
    - Dysphagia
    - Poor feeding
    - Recurrent Aspiration
    - Vocal cord paralysis
    - Stridor
    - Apnea
    - Abnormal responses to hypoxia, hypercapnea
  - Hydrocephalus.

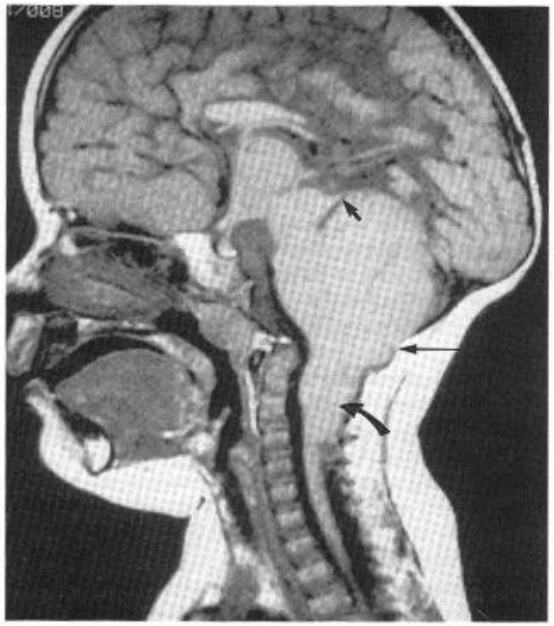
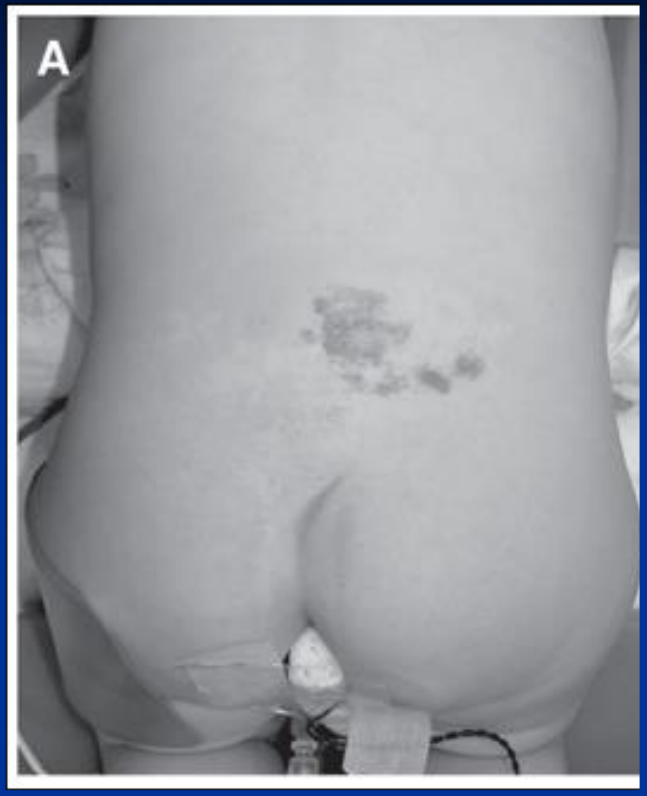
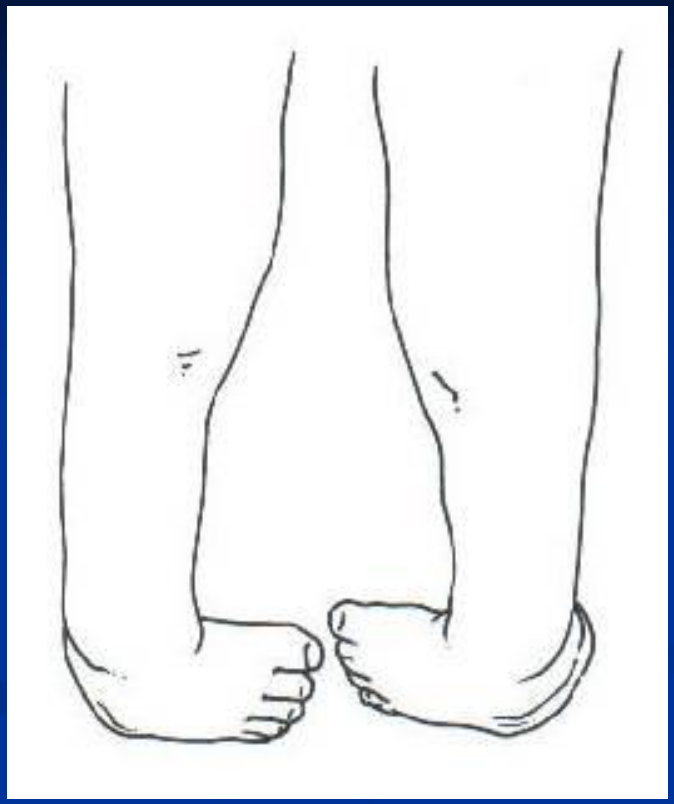


Figure 22–14. Magnetic resonance image of a patient with a Chiari II malformation. The small posterior fossa results in both upward and downward herniation of brain tissue. The short arrow indicates a "beaked" tectum showing upward herniation while the curved arrow indicates downward herniation of the cerebellar tonsils through the foramen magnum (thin arrow).

- Pulmonary: Sequellae of recurrent aspiration
- CV: Congenital cardiac defects: ASD, VSD, ToF, fallot.
- GI : Neurogenic bowel dysfunction / **incontinence, loss of anal ton**
- **GU: Urinary incontinence/obstruction, Recurrent UTIs/pyelonephritis.**
- **MSK:** motor paralysis, Kyphoscoliosis, Clubfoot, Hip dislocation.
- **Spinal cord:** tethard cord, diastematomyelia.
- **IMMUNE:** High risk population for latex allergy (up to 50%).
- **Skin:** tuft of hair or pigmentation, midline dimple, hemangioma.


A



The most associated anomalies with meningeomyelocele

1. Lower limb paralysis and bone deformities (club foot).
2. Bladder and bowel functions.
3. Association with hydrocephalus (Arnold_Chiari malformation)
4. Abnormalities of the vertebral column.

Diagnosis

⇒ **Ultrasonography**

For locating back lesion vs. cranial signs

⇒ Amniocentesis for AFP

Primary Closure of MMC

- **If ruptured:** Emergency neonatal surgery

- **If intact sac:** Goal closure within 24-48 hr of delivery before colonization of bacteria

MMC Other Surgical Procedures

- **VP Shunting**
  - Hydrocephalus 15% at primary closure
  - 80-90% post-closure
- Brain stem (Posterior fossa) decompression
- **Genitourinary procedures**
  - High incidence of bladder extrophy
  - Vesicoureteral reflux 2° neurogenic bladder
  - Urethral dilation
  - Vesicostomy
- **Orthopedic Revisions**
  - Club foot
  - Hip dislocation
- **Spine revision**: for deformities

Best wishes