Spinal Congenital Anomalies Spinal Dysraphism)

Done by :

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*3rd week of gestation

The nervous system is ectodermal in origin (develops from neuro – ectoderm) except the microglia and dura matter which are mesodermal in origin

Spinal cord development :

1. Gastrulation (conversion of the embryonic disk from a bilaminar disk to a trilaminar disk composed of ectoderm , mesoderm and endoderm)

gastrulation
*bilaminar
↓
trilaminar.

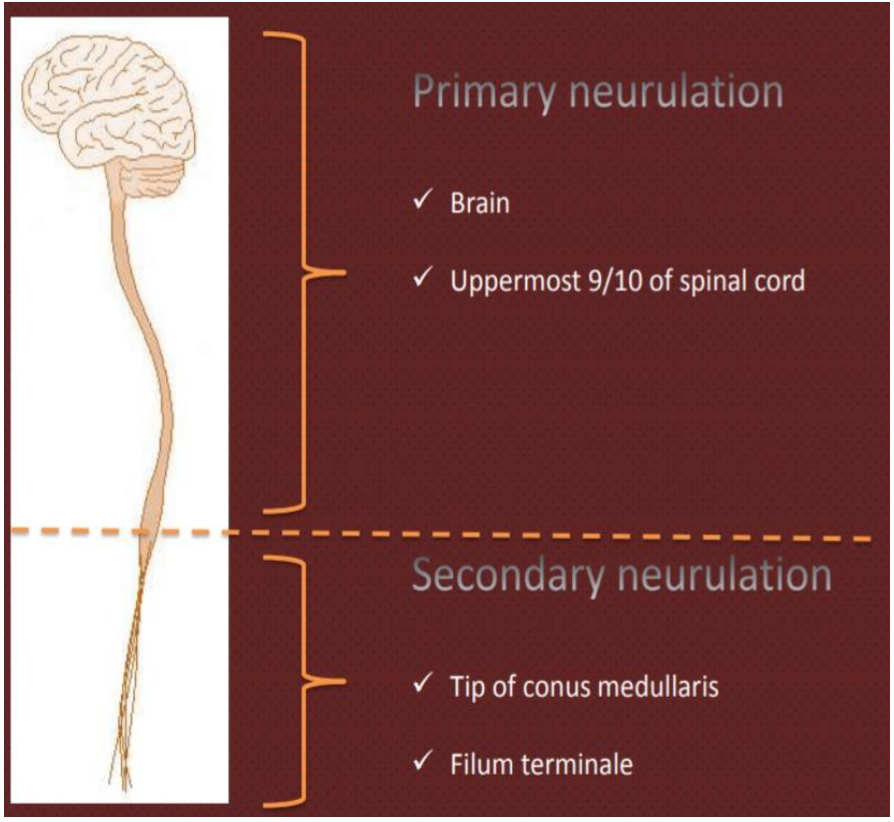
1ry neurulation

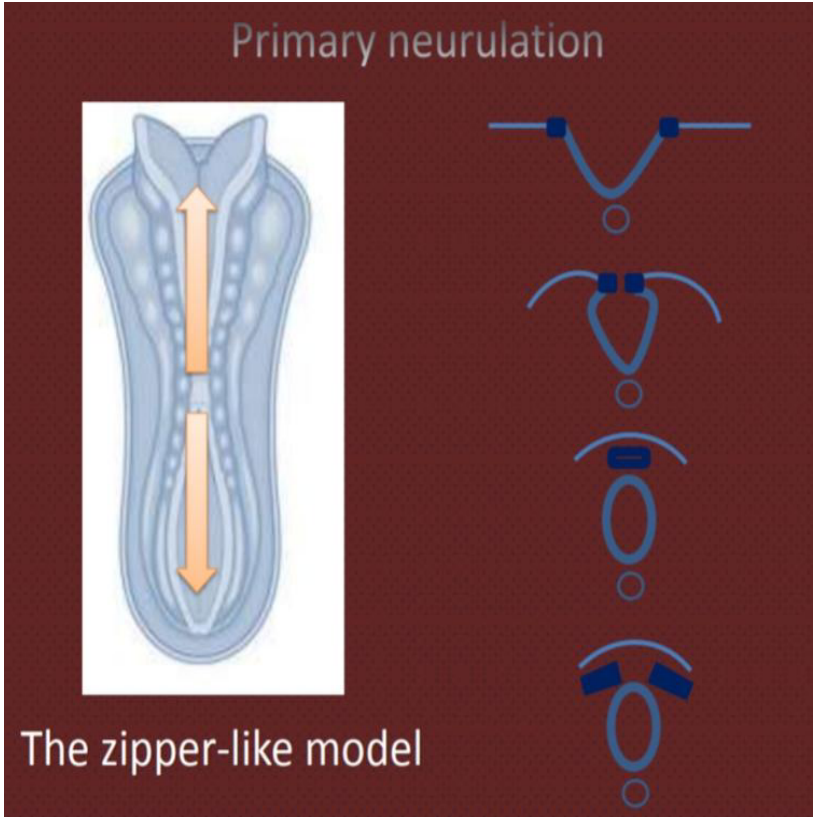
2ry neurulation

2. Primary neurulation (notochord and overlying ectoderm interact to form the neural plate)
Neural plate bends and folds to form the neural tube, which then closes bidirectionally in a zipperlike manner

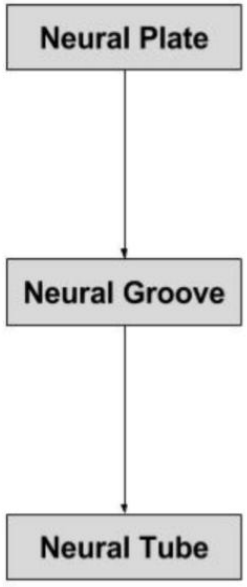
3. Secondary neurulation (neural tube is formed by the caudal cell mass)

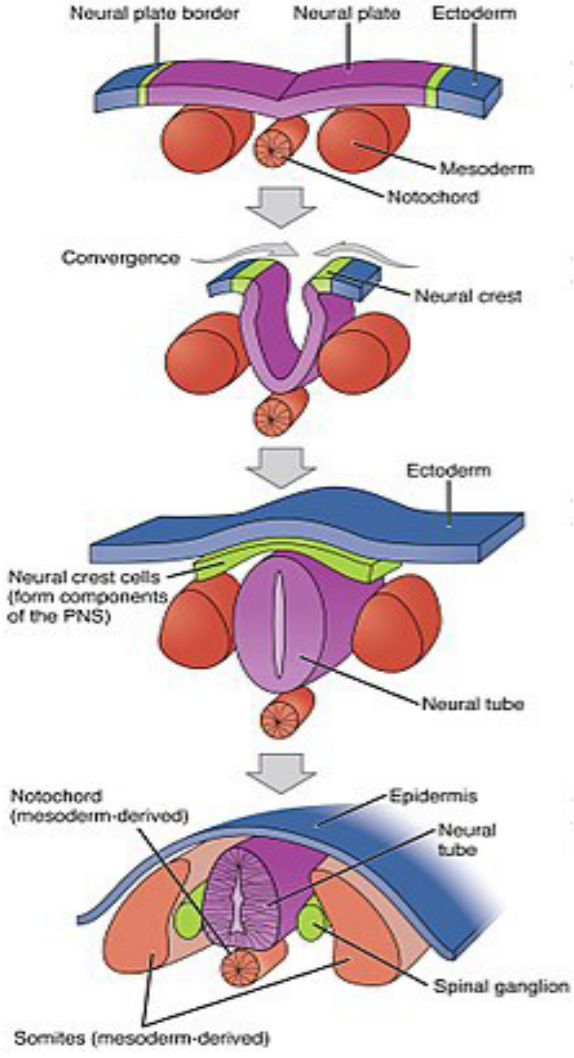
Abnormalities in any of these steps can lead to spine or spinal cord malformations.


Primary neurulation
✓ Brain
✓ Uppermost 9/10 of spinal cord
Secondary neurulation
✓ Tip of conus medullaris
✓ Filum terminale

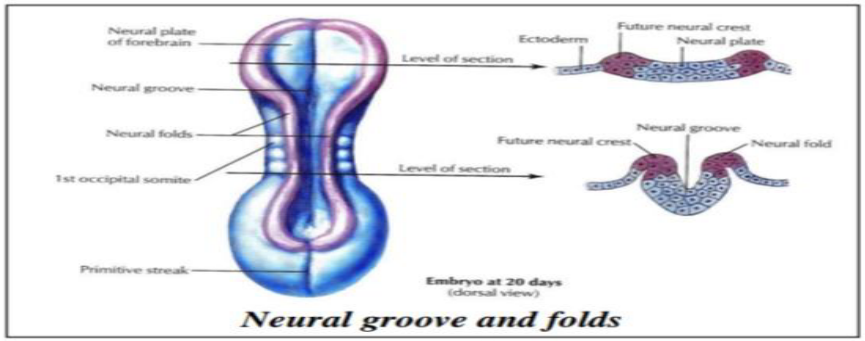

Primary neurulation
The zipper-like model

1. Formation of neural plate
2. Shaping of the neural plate
3. Bending of the neural plate
4. Fusion

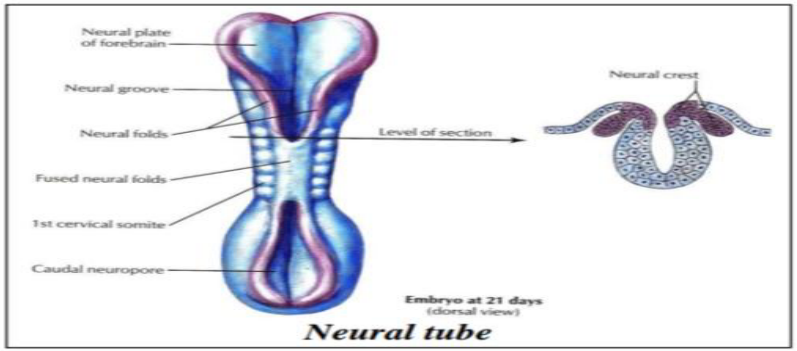




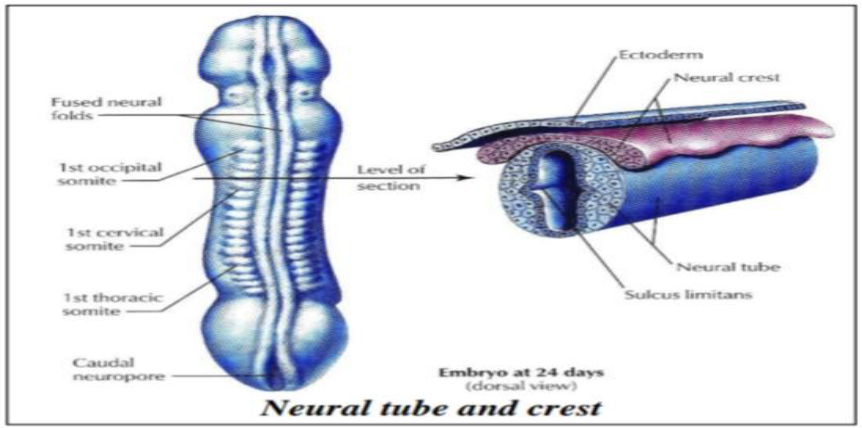
*Closure of the neural tube begins from the midline:



Neural groove and folds



Neural tube



Neural tube and crest

Fusion

Spinal Dysraphism :

Heterogeneous group of anomalies resulting from incomplete midline closure of osseous, mesenchymal and nervous tissue (Failure of normal fusion of the neural plate to form neural tube during the first 28 days following conception)

Prevalence :
Neural tube defects (NTDs) are one of the most common birth defects, occurring in approximately one in 1000 live births in the United States. common.

Neural tube defects (NTDs) cause infant mortality (death) and serious disability

Categorization of Spinal Dysraphisms (neural tube defects) :

open spinal dysraphism

closed spinal dysraphism

no skin → dysplastic tissue

1. Open spinal dysraphism
there is a defect in the overlying skin, and the neural tissue is exposed to the environment (caused by defective closure of the primary neural tube and are characterized clinically by exposure of the neural placode through a midline skin defect on the back)
. All OSDs are anomalies of 1ry neurulation

2. Closed spinal dysraphism
neural tissue is covered by skin (may be abnormal skin) , but with or without associated subcutaneous mass

Spina bifida
Spine split

occulta

cystica

Due to failure of fusion of the dorsal parts of vertebra " absence of vertebral arch or part of them "

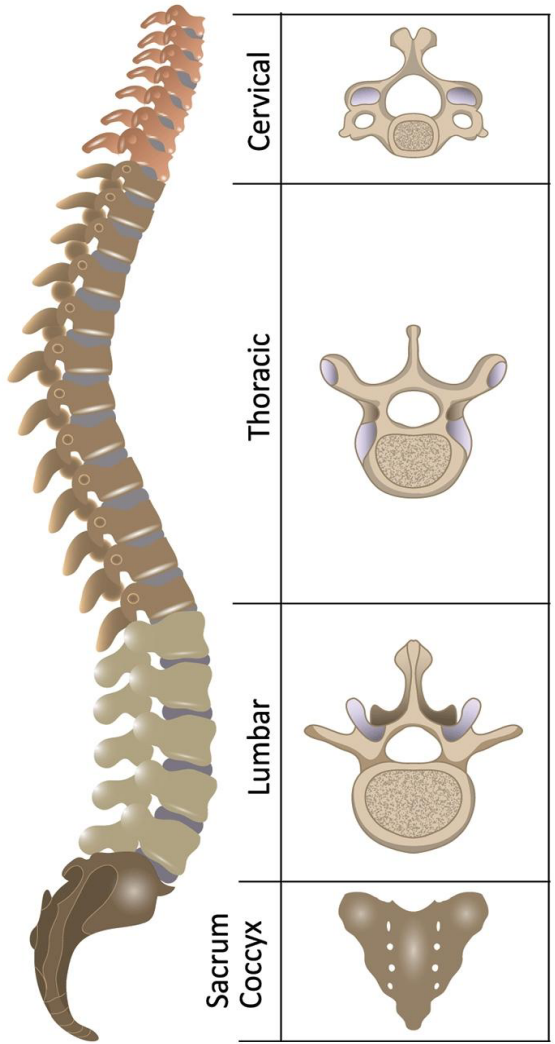
→ no posterior arch

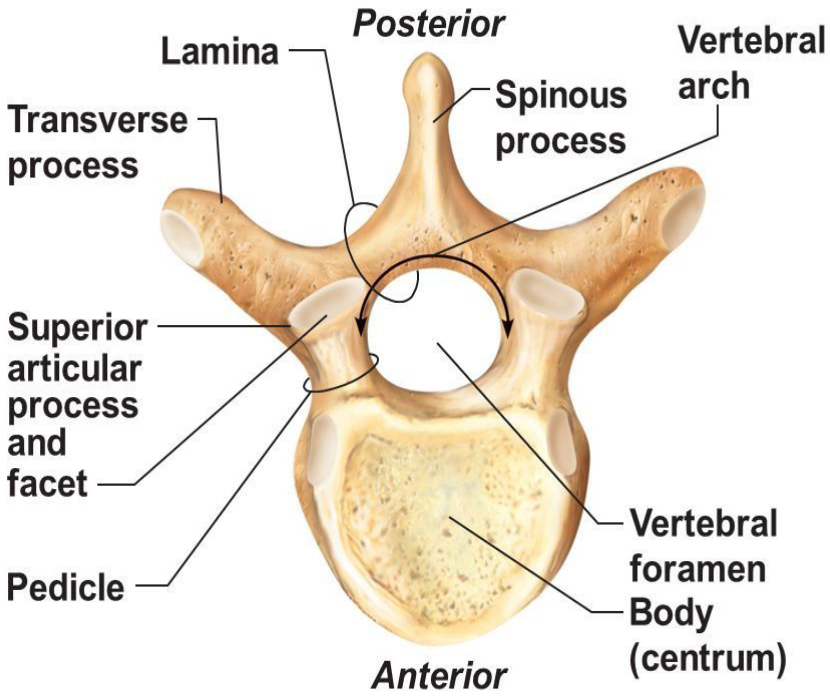
1. **Occulta**

. Most common and mildest form

. Mostly / typically affects the lumbosacral area

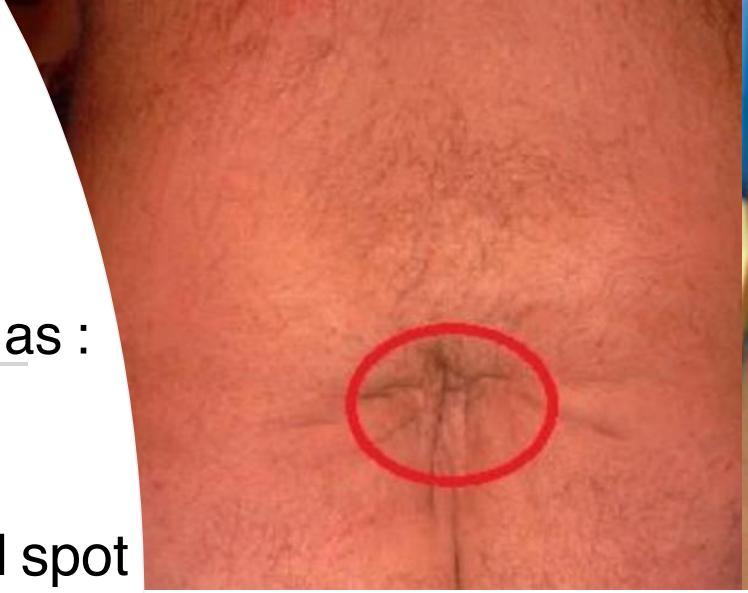
. May associated with **overlying skin lesions** as : Skin dimples , Heamangioma or red spot , Abnormal tuft of hair , sinus tract , fatty mass or lipomas

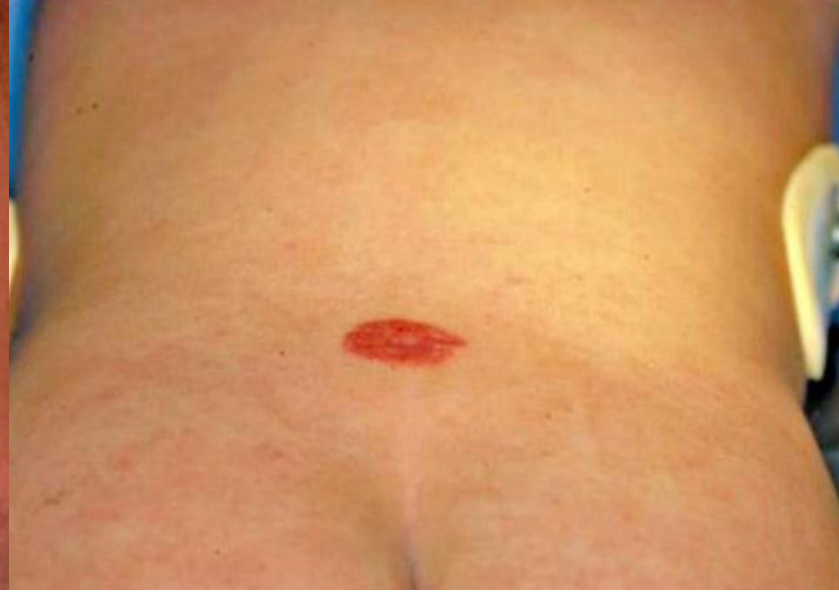

Cervical
Thoracic
Lumbar
Sacrum
Coccyx

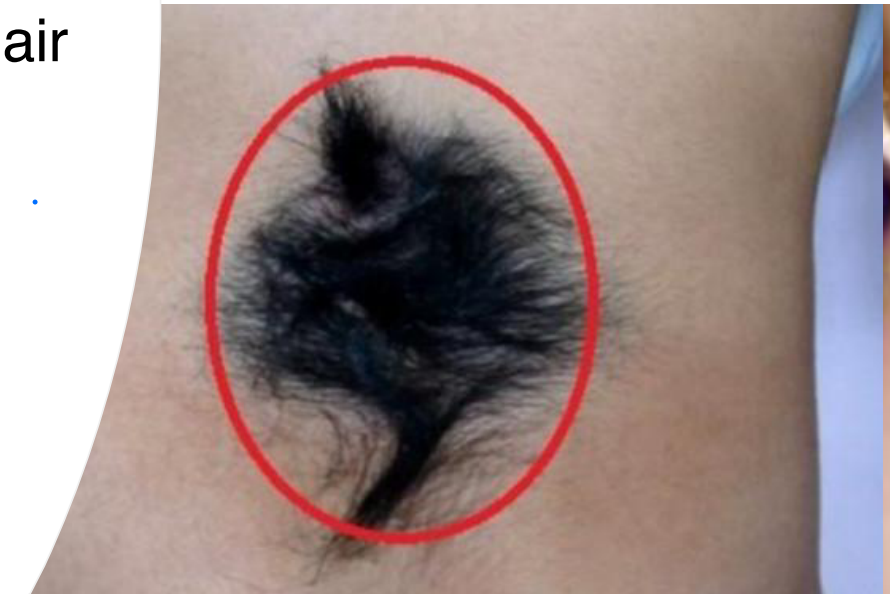

Posterior
Lamina
Vertebral arch
Spinous process
Transverse process
Superior articular process and facet
Pedicle
Vertebral foramen
Body (centrum)
Anterior

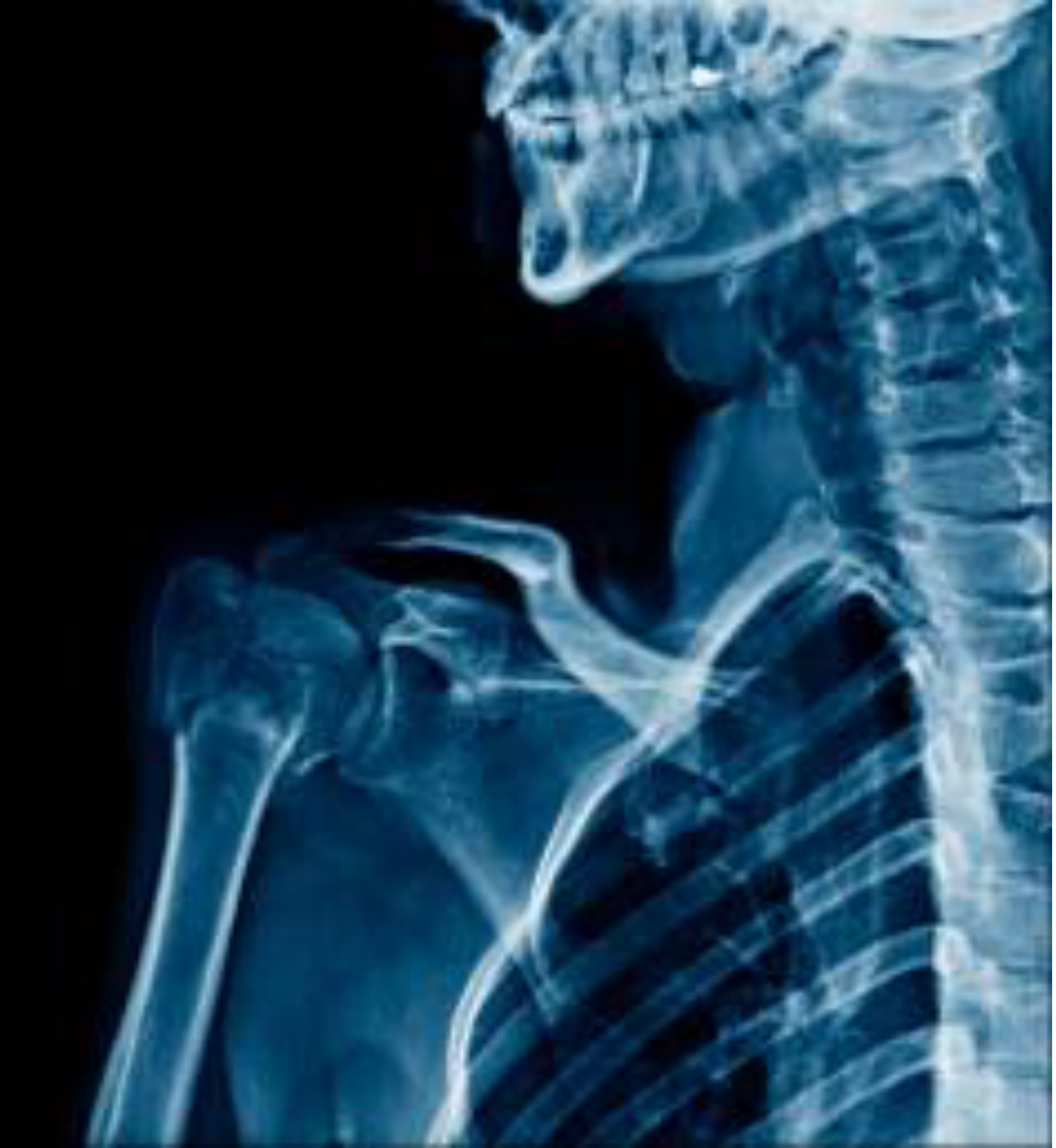
Overlying skin lesions as :

1. Skin dimples
2. Heamangioma /red spot
3. Abnormal tuft of hair
4. Sinus tract
5. Lipomas









2. **Cystica**

1 → **Meningocele** (meningeal cyst)

. The meninges herniates through the spina bifida and forming subcutaneous sac / cyst filled with CSF

2 → **Myelomeningocele** more common than meningiocele

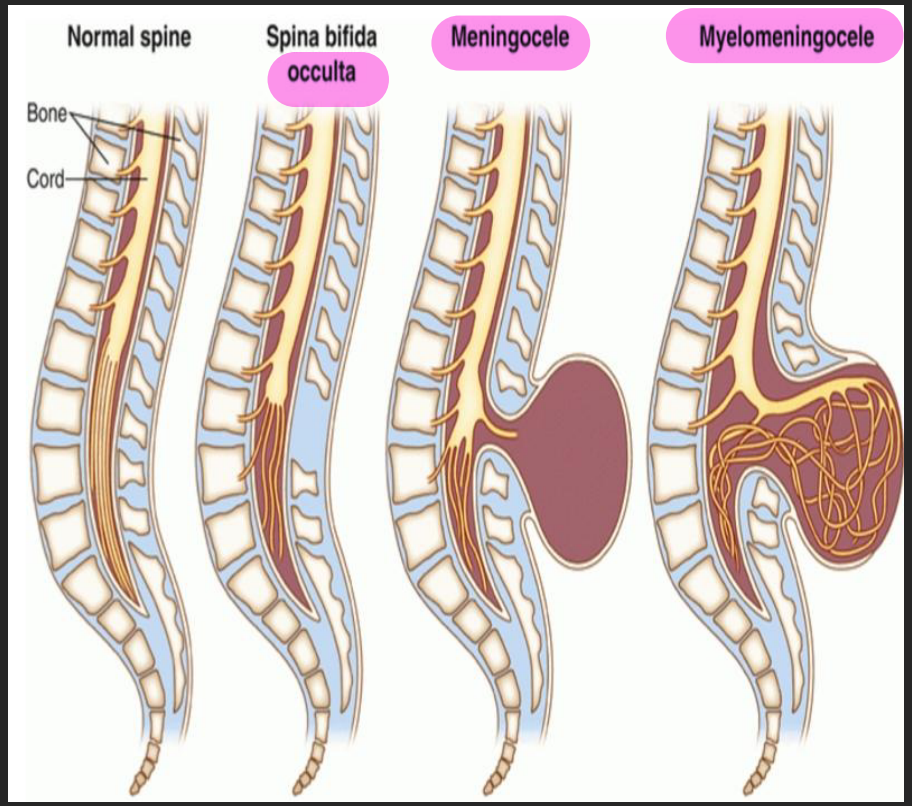
. The spinal cord herniates through the Meningocele

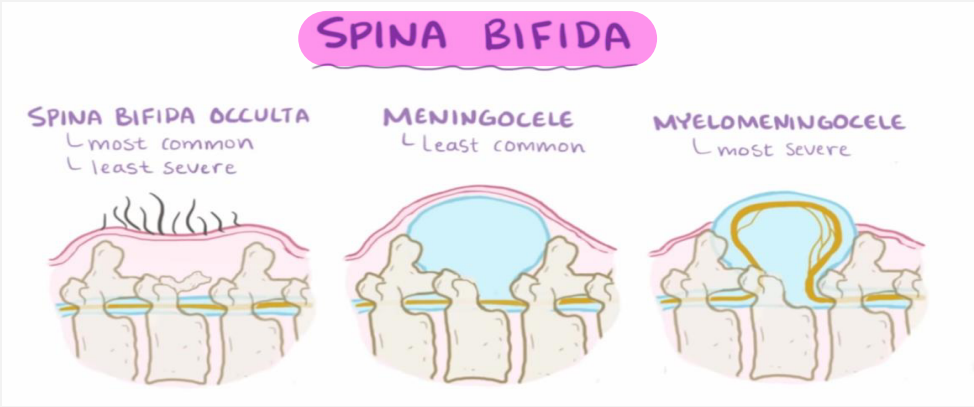
. May associated with other neurological anomalies as : Chiari 2 malformation , hydrocephalus

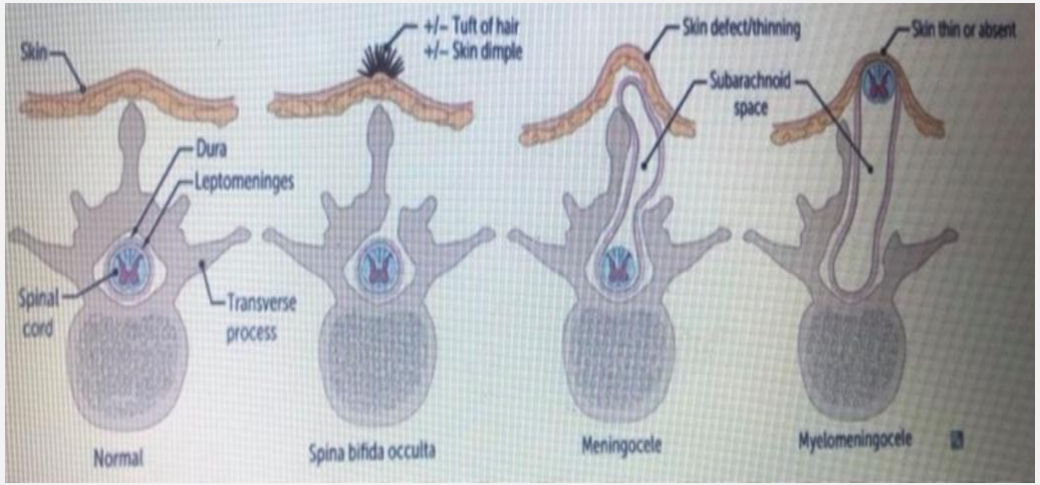
* **Myelocele (Rachischisis)** open back deformity

. Failure of obliteration of the neural tube

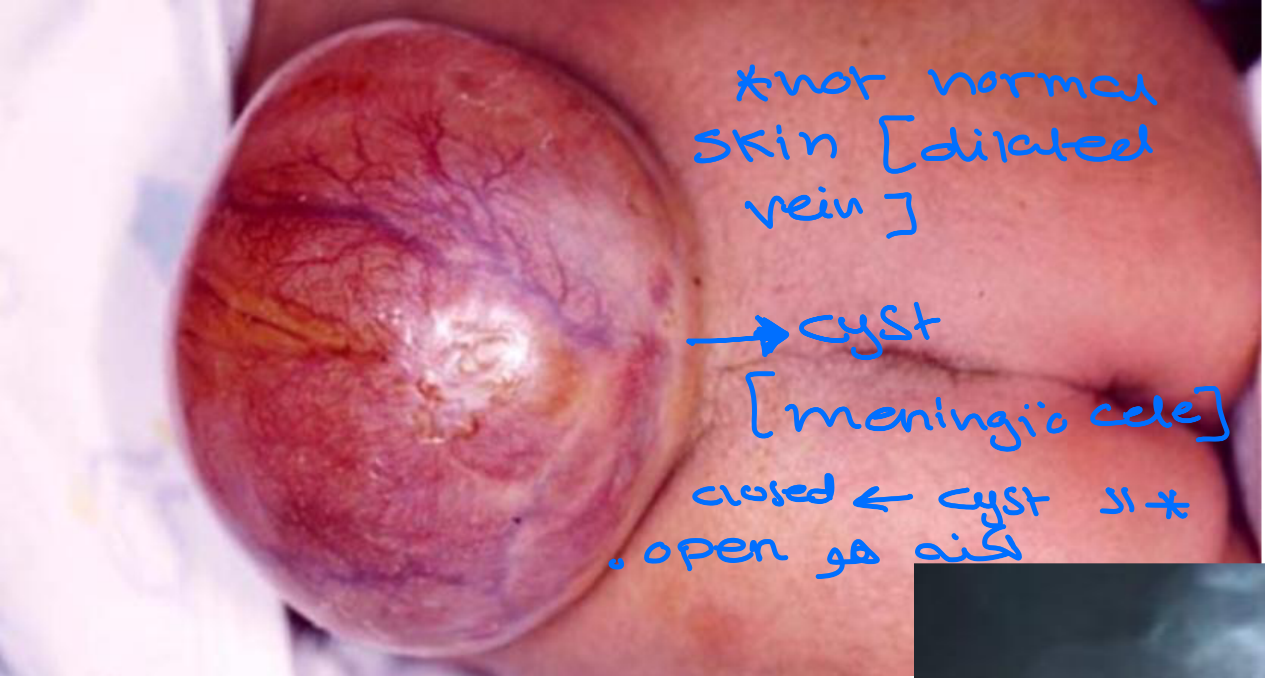
*all patient with myelomeningiocele have chiari 2 [[illegible]]

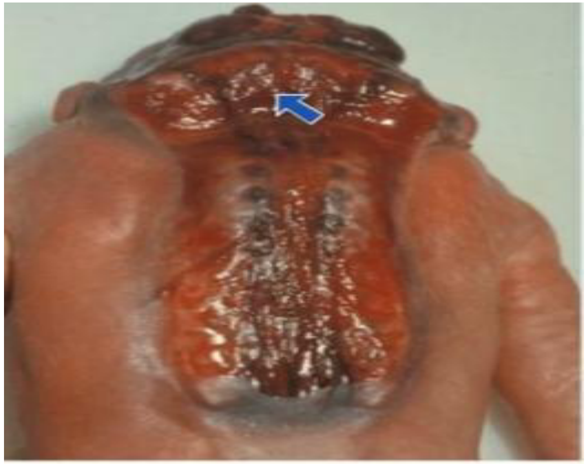

Normal spine
Spina bifida occulta
Meningocele
Myelomeningocele
Bone
Cord


SPINA BIFIDA
SPINA BIFIDA OCCULTA
└ most common
└ least severe
MENINGOCELE
└ least common
MYELOMENINGOCELE
└ most severe

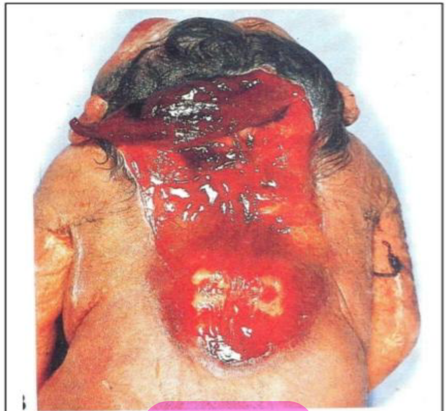

Skin
+/- Tuft of hair
+/- Skin dimple
Skin defect/thinning
Skin thin or absent
Subarachnoid space
Dura
Leptomeninges
Spinal cord
Transverse process
Normal
Spina bifida occulta
Meningocele
Myelomeningocele

	Meningiocele	Myelomeningocele
Incidence	Least common	More common
Prognosis	Less serious	Most serious
Sac	meninges and cerebro-spinal fluid	meninges and cerebro-spinal fluid & nerves and spinal cord.
Nerves	not badly damaged	badly damaged
Ability	❖ Limited disability is present.	bowel and bladder problems.

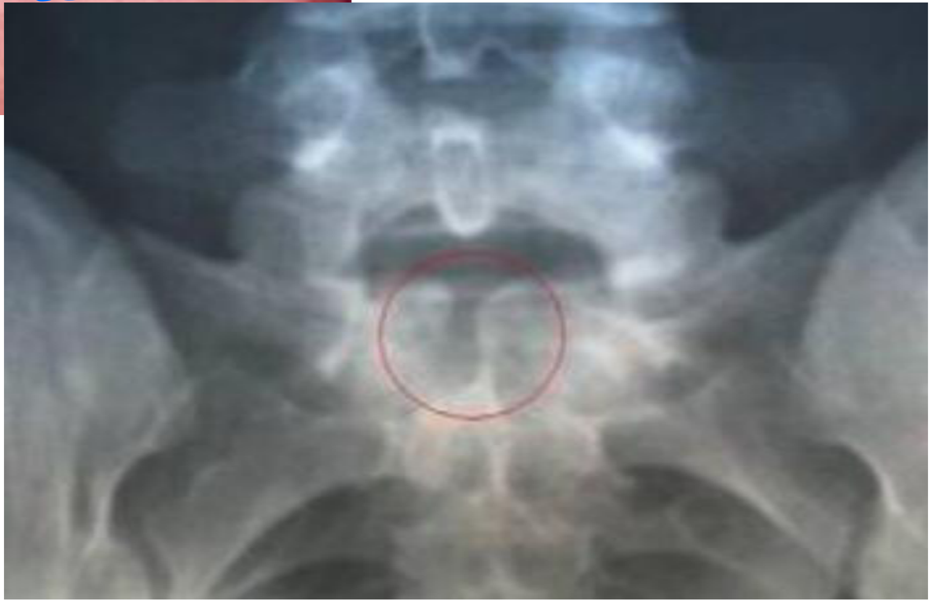




Rachischisis + anencephaly



Rachischisis



Spina Bifida

* داليا مع د موجود Spina bifida

· Rachischisis.

Unknown cause , but the risk factors include :

1. Folate deficiency (Vitamin B9)
2. Obesity
3. Poorly controlled Diabetes
4. Medications that interfere with folate metabolism as :
Anti- Seizure medications
5. Genetic predisposition and Family history
6. Previous pregnancy with same condition

Diagnosed :

1. **Prenatally** ⊛

. Screening (increased Alpha Fetoprotein " AFP " in mother's serum , Acetylcholinesterase)

. US most accurate - → in amniotic Fluid.

. Amniocentesis

indicate → congenital anomely

Confirm that this is CNS anomily and open anomaly

2. **Postnatally**

✓ . Neonatal Examination " clinically " + imaging (MRI)

✓ . Get complications → weakness / paralysis.

✓ . Incidentally by imaging

Treatment :

1. **Prenatal** (with very high risk)

*لازم تتعمل اذا repair خلال اول ٢٤ ساعة (infection)
48 hours.

2. **Postnatal** surgical repair shortly after birth (starting initially by covering the lesion with Gauze soaked with normal saline or ringer solution to prevent dryness , sac excision , emptying of CSF , exposure of the contents , tissue repair)
   - avoid antiseptic (neurotoxic agents) ex: iodine . alcohol
   - avoid mechanical trauma

General assessment

- Assess whether lesion is ruptured or unruptured
  - Ruptured lesions start prophylactic antibiotic
  - Urgent surgery
- Measure size and site of defect for proper planing for Closure
- Evaluation by neonatologist
  - Other anomalies (average 2-2.5% additional anomalies)
  - Condition oppose with surgery e.g lung immaturity
- Bladder
  - Start with on regular urinery catheterization
  - Urological consultation
- Orthopedic consultation for sever deformities

Neurological preop. assessment

- Watch for spontaneous movement of lower limbs which associated with better outcome.
- Assess lowest level of neurological function
  - Response to painful stimuli
  - Differ between **voluntary movement** from **reflex movement** which is stereotyped and not persist after stimulus
- Evaluate other neurological associations
  - A. Hydrocephalus
    - Anterior fontanel
    - Head circumference
  - B. Chiari II
    - Check for inspiratory stridor and apneic episodes

Lipomyelomeningocele : 2ry neurolation

. Lipoma attached to dorsal surface of incompletely closed spinal cord

. Covered by skin

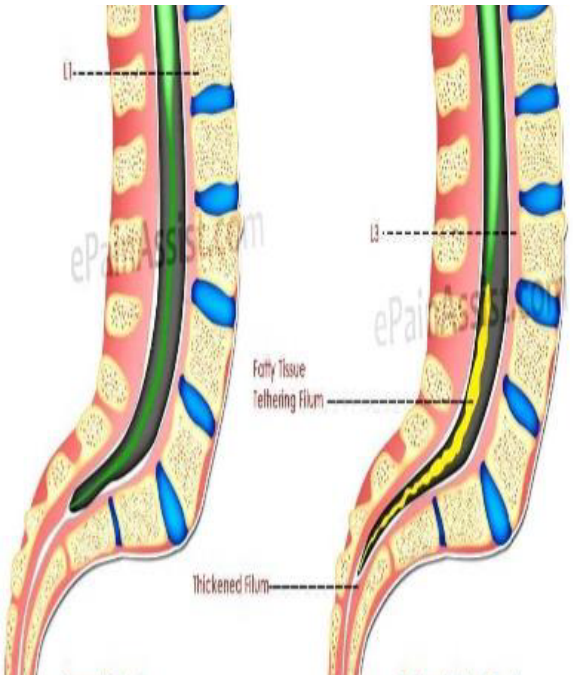
*normal conus medullaris at birth at the level of L3-L4, during growth: ascend

↳ normal skin *closed.

* هون بصير تحت ال L2 . اللي هو الطبيعي بكون لحد

Tethered cord syndrome (tight filum terminale syndrome) :

The spinal cord could be caught against the vertebral by tight band , fibrosis or adhesions (abnormally attached to tissue around) resulting in abnormally low conus medullaris



Symptoms usually at age of puberty including (5 Ds) :

1. Discomfort , back and leg pain
2. Deformities of spine (scoliosis , kyphosis , lordosis) and feets (high arches , foot turned inward)
3. Dysfunctions (weakness , numbness , " kidney , bladder and bowel dysfunctions ")
4. Decline of abilities
5. Dysraphisms of spine

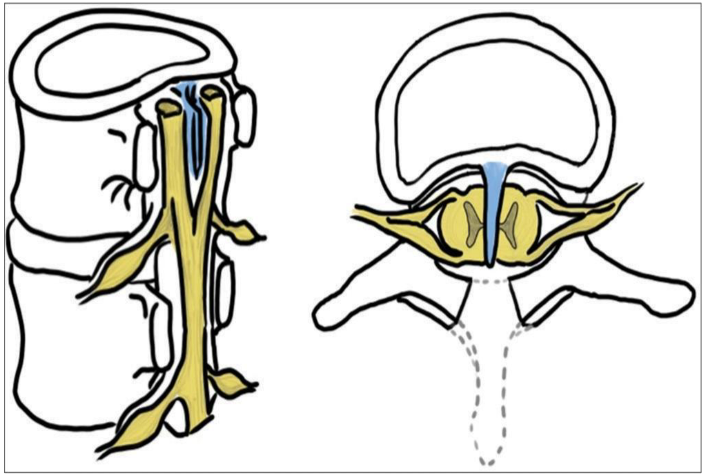
neck pain, back pain & sciatica before age 4? red flag.

Diagnosed by MRI to confirm the diagnosis and rule out Neoplasia

↳ abnormal diameter > 2 mm

Treated surgically by Un- tethering surgery by cutting the filum terminale

Split Cord Malformation (SCM) :



Rare anomaly characterized by a split along the midline of the spinal cord by fibrous or bony septum , which divides it into 2 symmetric or nonsymmetric entities

* بتفضل جزء معين

It NOT involving the whole length of spinal cord , but segment of it mostly at level of lumber or thoracic area

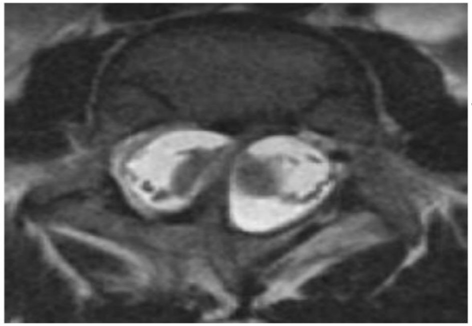
Mostly associated with Tethered cord , so share it's symptoms

Diagnosed mainly by MRI

Treated by Un- tethering surgery **(but after removal of the bony septum and reconstitution of dura into single tube of affected segments until reaching the normal segments)**

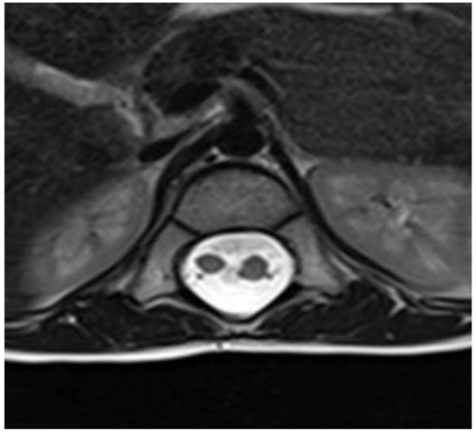
* اول شي بنشيل ال bony septum ، بعدين بنعمل untethering • عشان هيك بحسن contusion •

Type 1



hemicord لكل *
بجدار كانه
cord لحاله.

Type 2



Two types of SCM :

1. Type 1 (**Diastematomyelia**) by bone

. Two hemicords , each with it's **own** central canal , surrounding pia and dural tube , separated by **rigid osseocartilaginous** (bony spur) median septum

. With abnormalities of spine at the level of split (absent disc)

. ⅔ associated with overlying skin abnormalities

2. Type 2 (**Diplomyelia**) by Fibrous

. Two hemicords within a **single dural tube** , separated by a **nonrigid fibrous median septum**

. Without abnormalities of spine at the level of split , but usually with spina bifida occulta

Impact of life :

1. Financial (cost)
2. Physical (paralysis , hydrocephalus , learning disabilities , bowel and bladder control problems , other congenital anomalies)
3. Emotional (miscarriage , stillbirth , infant mortality , disability , feelings)


Thank you