Development of the Spine and Spinal Cord

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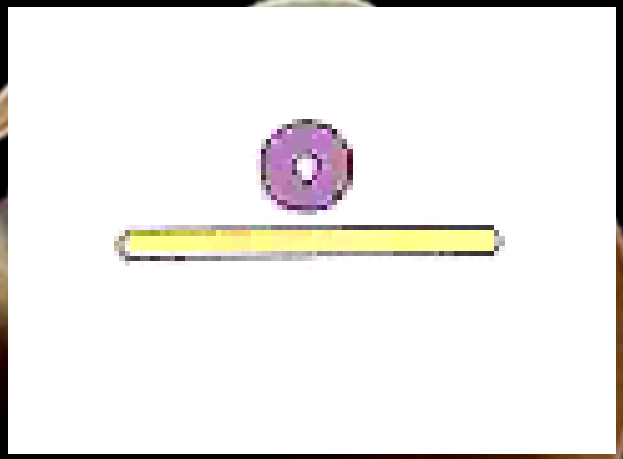
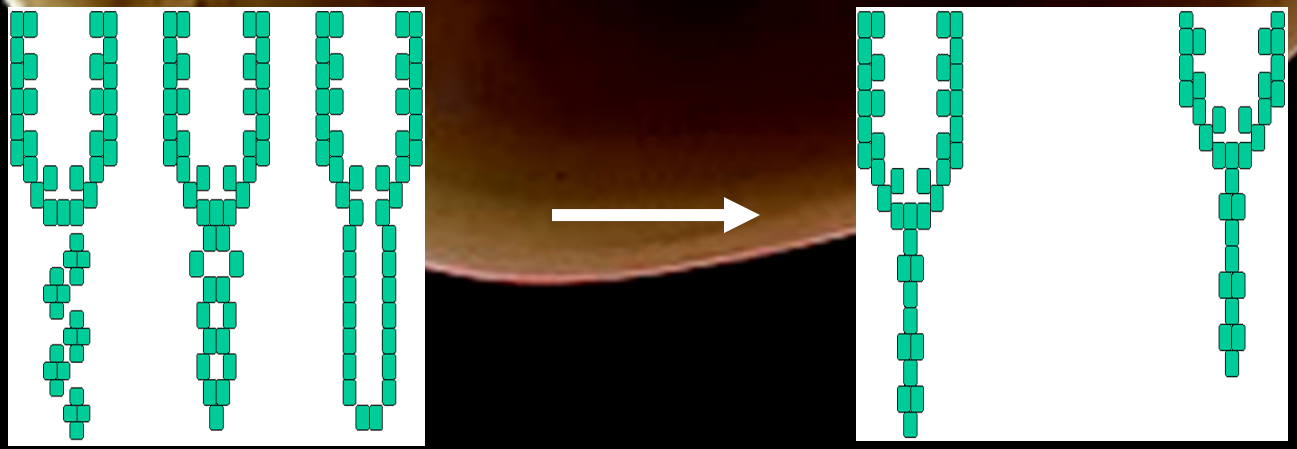
Development of the spinal cord

- Formed in 2 parts
  - Neurulation

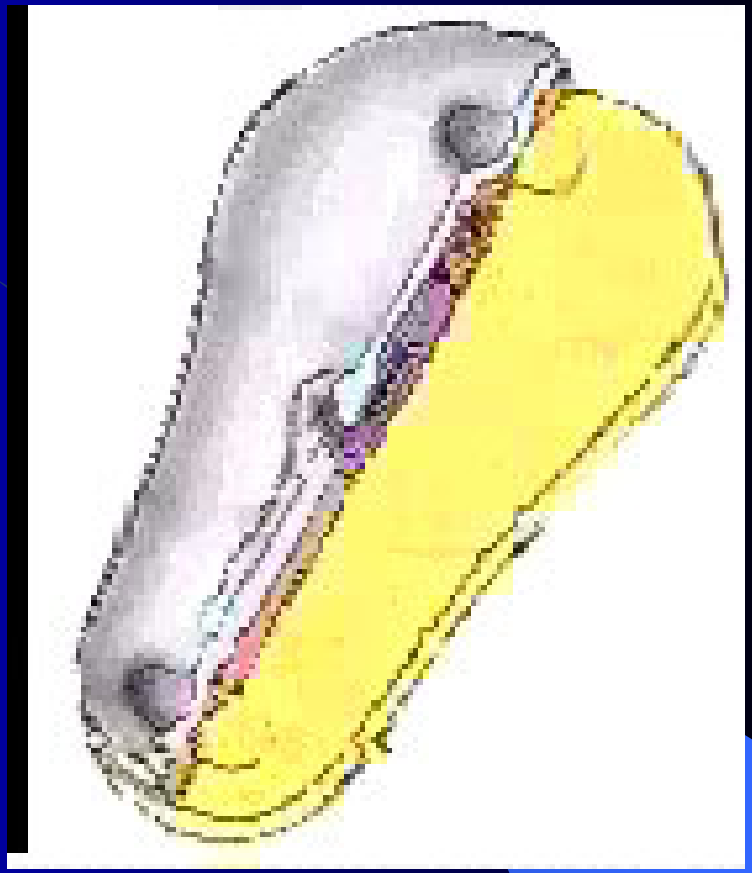


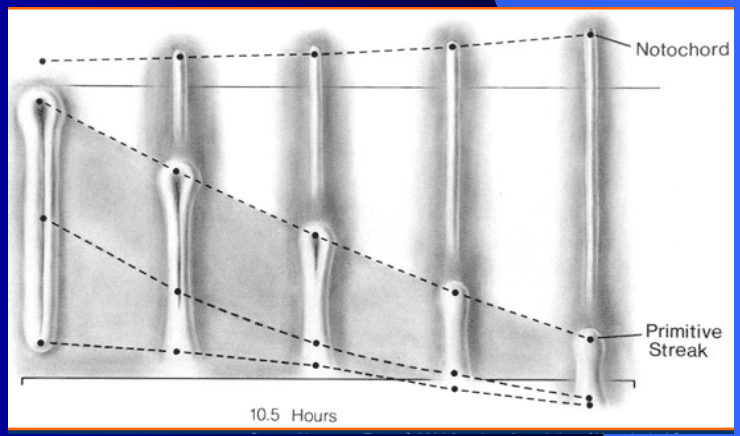
  - Canalization / Retrogressive differentiation



Neurulation

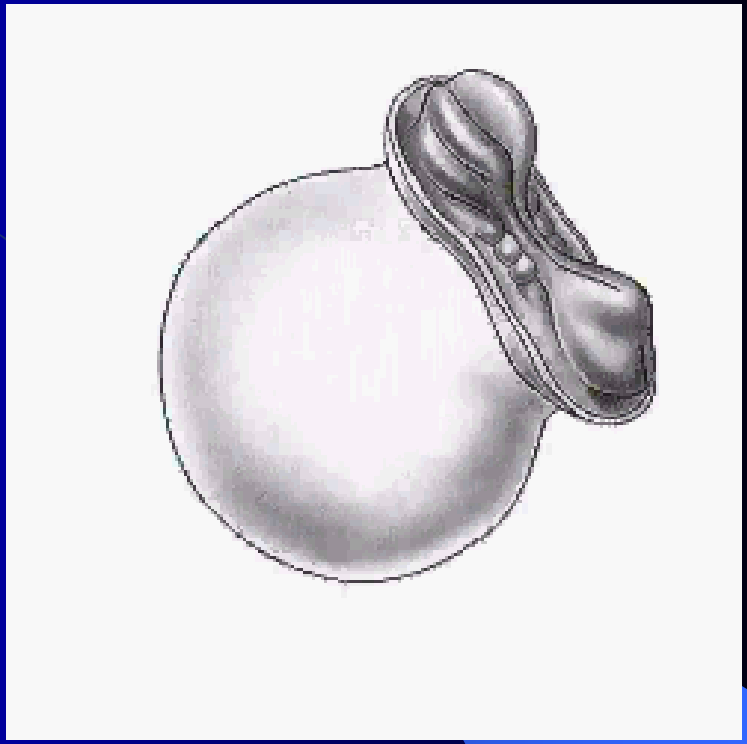
- DOL 5- ectoderm proliferates--> primitive streak
  - Cephalic end of p. streak--> rapid prolif cells--> prim. pit (Henson's node)
- DOL 15-cells entering prim. pit-->form notochord
  - Promotes neuroectoderm--> neural plate

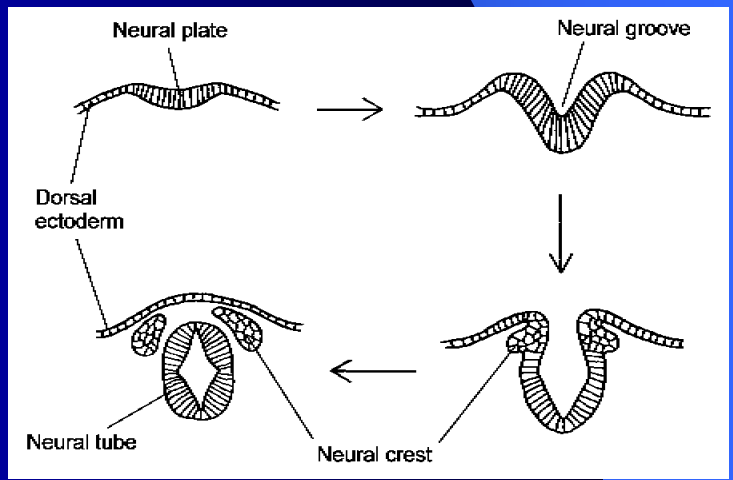




Neurulation

- DOL 17-lateral portion of neural plate thicken--> neural folds
  - Neural folds contract --> neural tube
  - Closure of neur.tube (neurulation)-->cephalad(24d) and caudal(27d) directions
- DOL 30- neurul. complete--> >dysjunction
  - Ectoderm separates fr/ neur tube
  - Neur. crest migrate lat.--> dorsal root gang/symp plexus





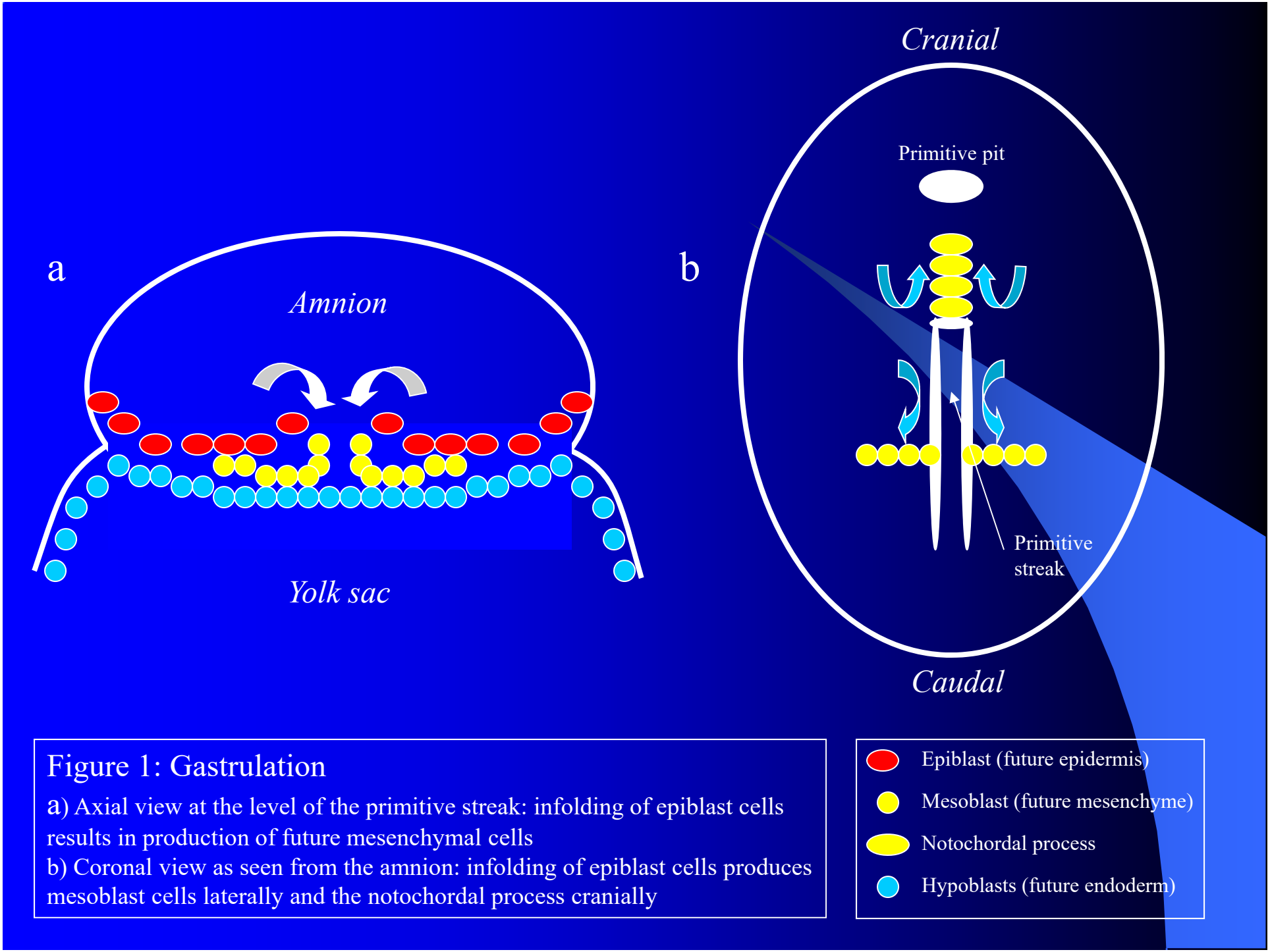


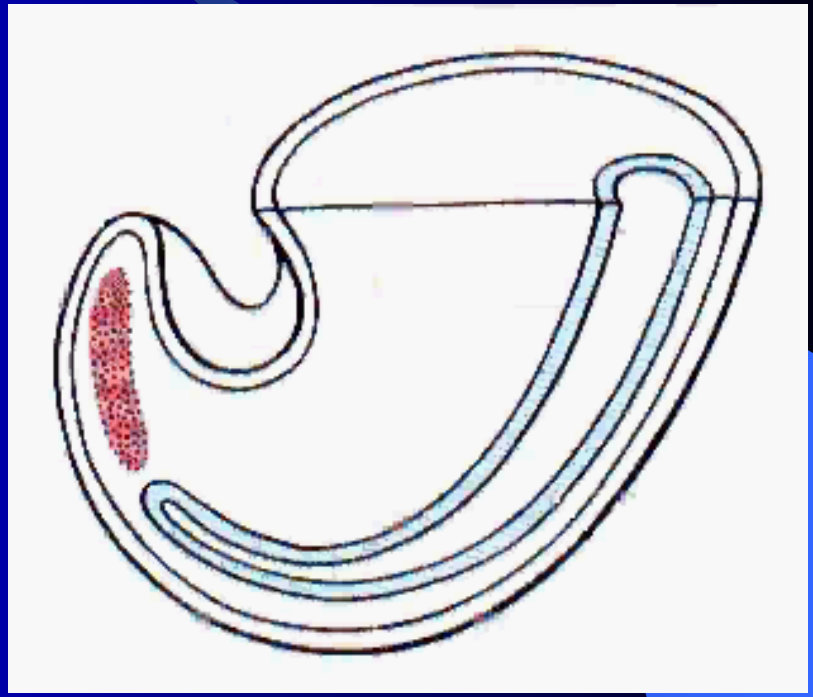
Figure 1: Gastrulation

a) Axial view at the level of the primitive streak: infolding of epiblast cells results in production of future mesenchymal cells

b) Coronal view as seen from the amnion: infolding of epiblast cells produces mesoblast cells laterally and the notochordal process cranially

Canalization

- Neural tube elongates caudal to post neuropore
  - Notochord & neur. epi fuse--> caudal cell mass
- DOL 30 -microcysts coelesce within cell mass-->tubular stucture -->fusing w/ n tube above
- DOL 38 - retrogressive differentiation--> filum terminale, distal conus, terminal ventr.



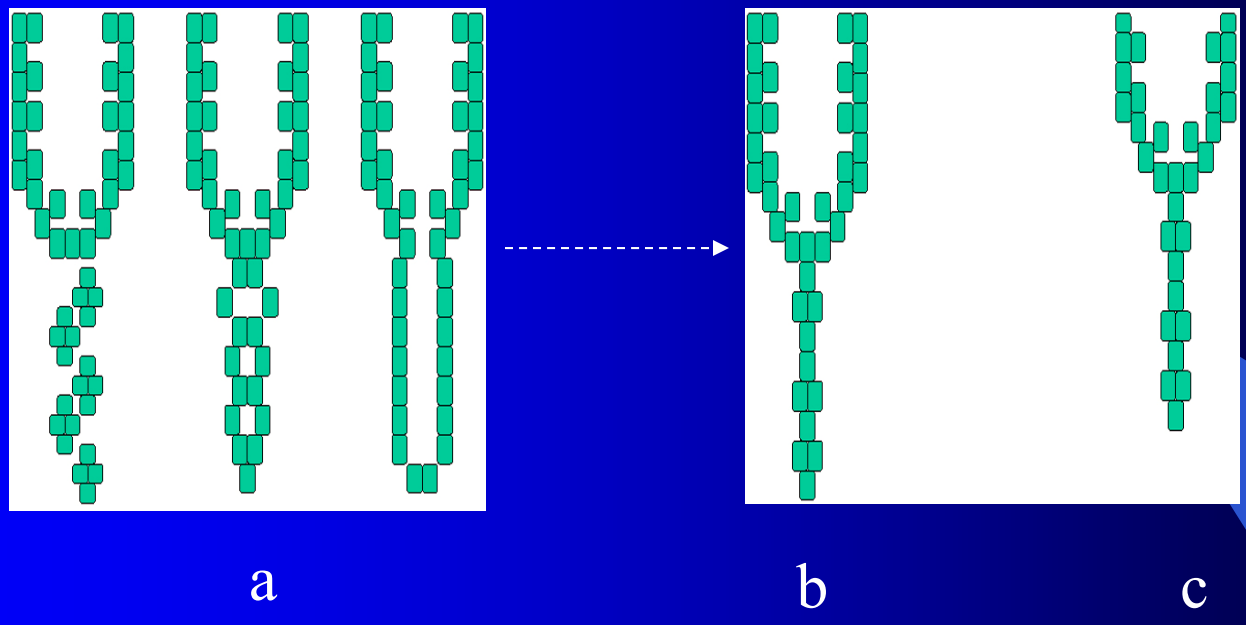
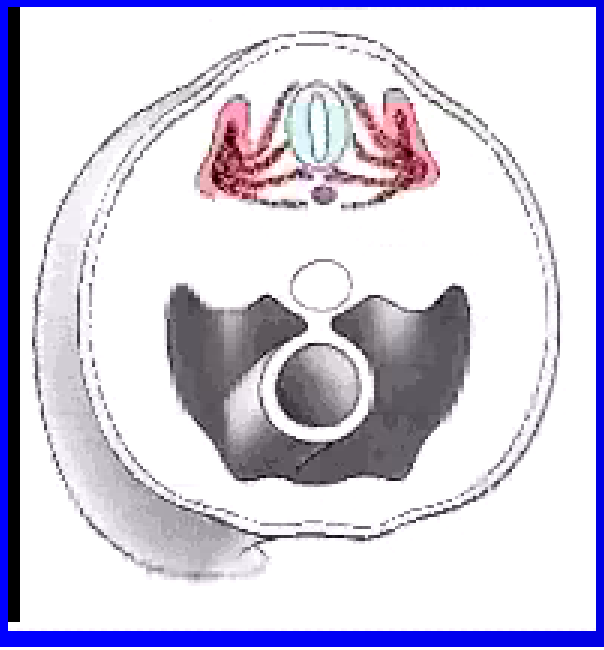


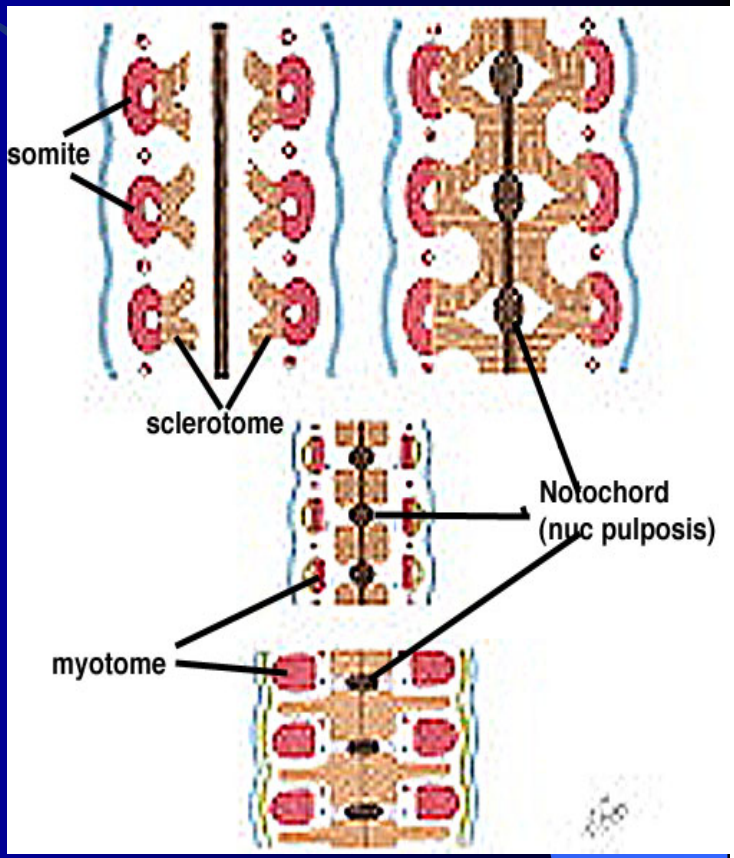
Figure 3: ***Canalization and regression***
(a) Canalization: Undifferentiated cell mass coalesces to form neural cord. Fusion with cephalad neural tube produces confluent neural tube extending from cervical to sacral levels.
(b) Regression, initial phase: Retrogressive differentiation leads to atrophy of the distal neural tube to form atretic pial band of the filum terminale.
(c) Regression, second phase: Differential growth of the supporting structures of the spine leads to ascent of the conus to the L2-3 disc space at birth, and L1-2 disc space by 3 months of age.

Development

Vertebral Column

- Membrane Development
  - DOL 25 - notochord separate fr/gut & n tube
  - Mesenchymal cells migrate ventral/dorsal-->somites separ. by interseg fissures



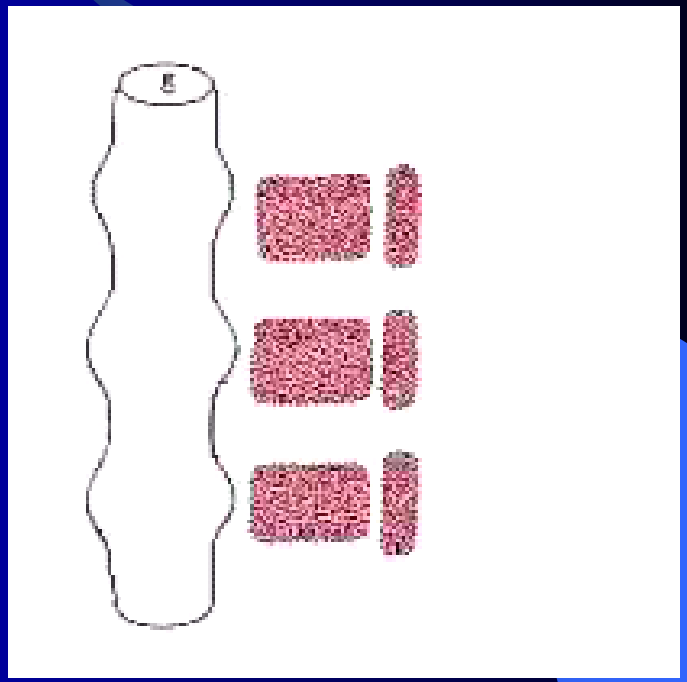


Each somite -->
med. Sclerotome
-->vtb body
lat. myotome
-->paravtb mscl

After dysjunction: post mesenchyme--> post elem.

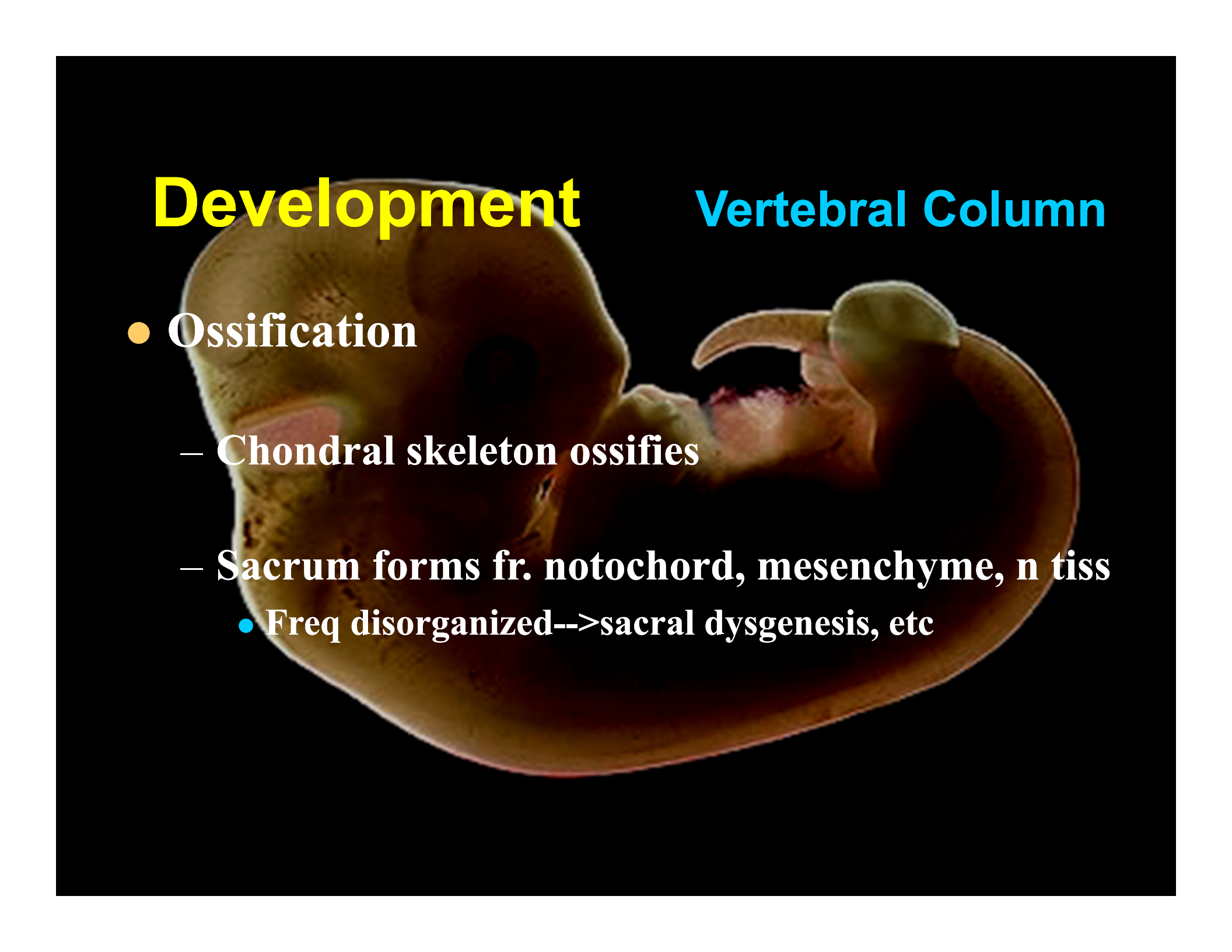
Development Vertebral Column

- Chondrification
  - Sclerotomes divide in half
    - Bottom half fuses with half from above--> vtb body
    - Occurs bilaterally forming entire vtb body
  - Notochord remnants-->nuc pulposis within disc
  - Lat thoracic sclerotomes--> ribs



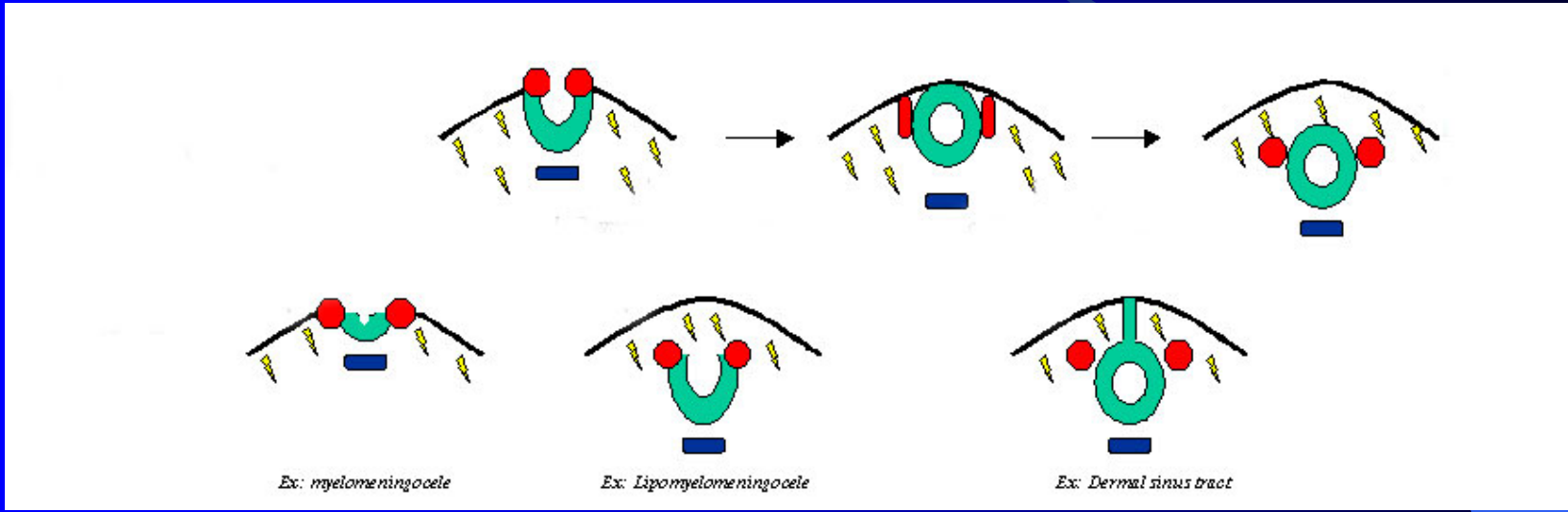
Development

Vertebral Column



- Ossification
  - Chondral skeleton ossifies
  - Sacrum forms fr. notochord, mesenchyme, n tiss
    - Freq disorganized-->sacral dysgenesis, etc

Clinical correlation



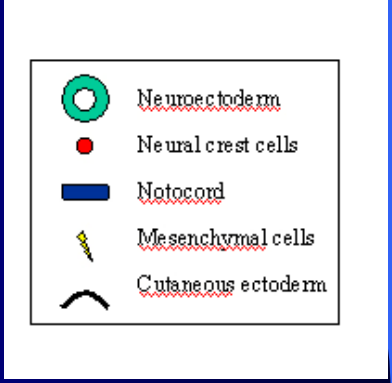
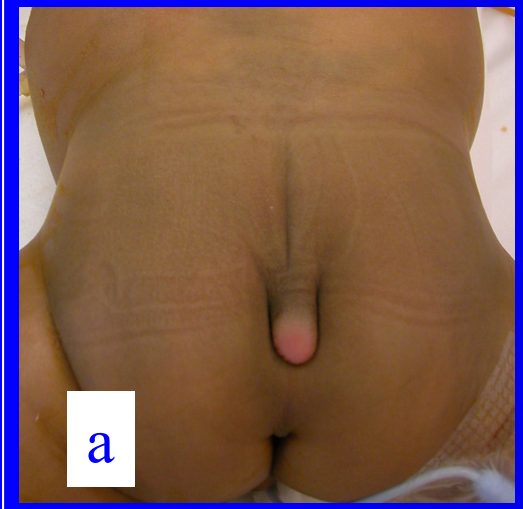
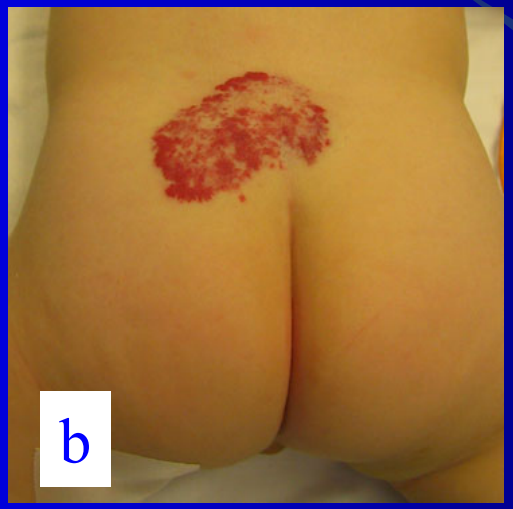


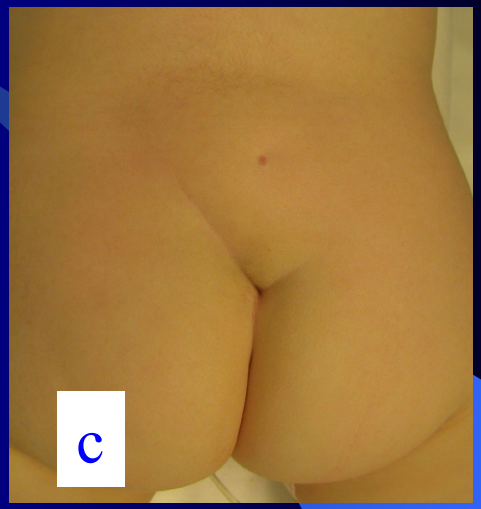
Figure 2: ***Normal and pathologic neurulation***
(a) *Normal embryologic development*: Typical neurulation proceeds with induction of cutaneous ectoderm to neuroectoderm, infolding of the neuroectoderm to form a closed neural tube, and dysjunction of the completed neural tube from the overlying ectoderm
(b) *Pathologic embryologic development*: Errors in neural tube closure or abnormal dysjunction can result in production of conditions capable of producing the tethered cord syndrome.

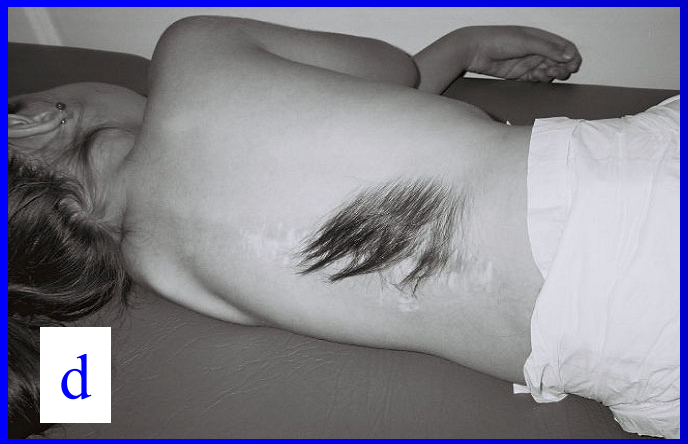
Figure 4: Cutaneous manifestations of occult spinal dysraphism

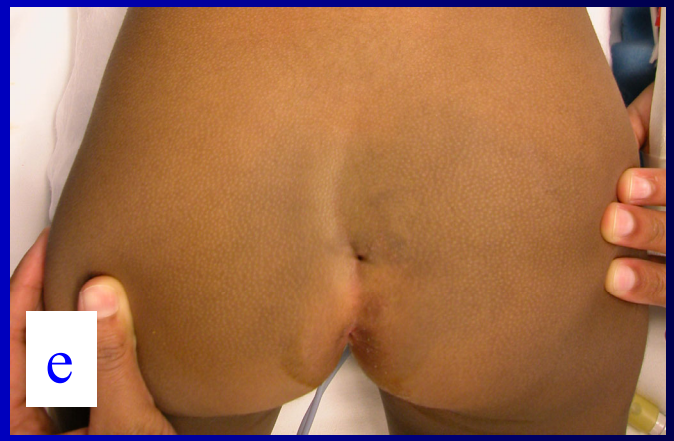
Cutaneous signs of spinal dysraphism include (a) vestigial tail formation, (b) cutaneous hemangioma, (c) subcutaneous lipoma, (d) hypertrichosis, and (e) pathologic sacral dimple.





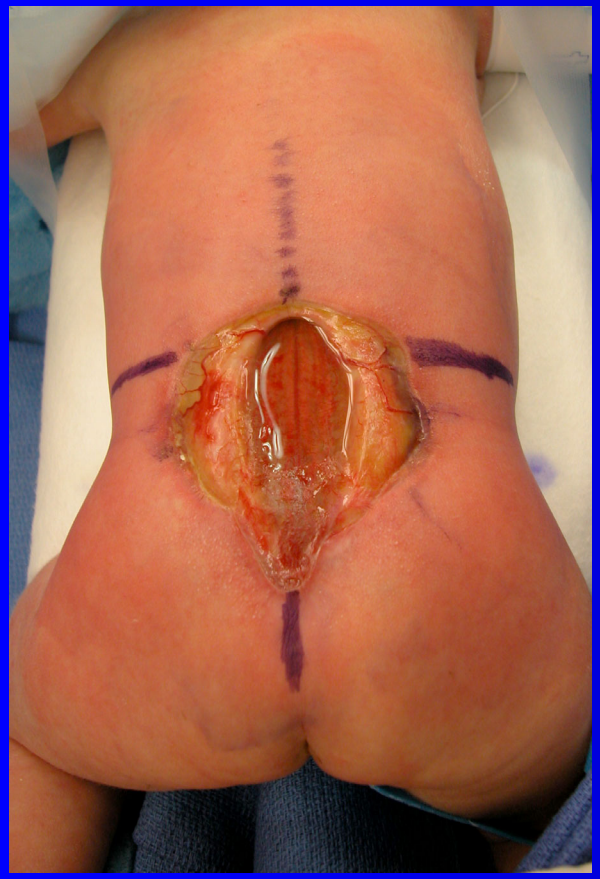


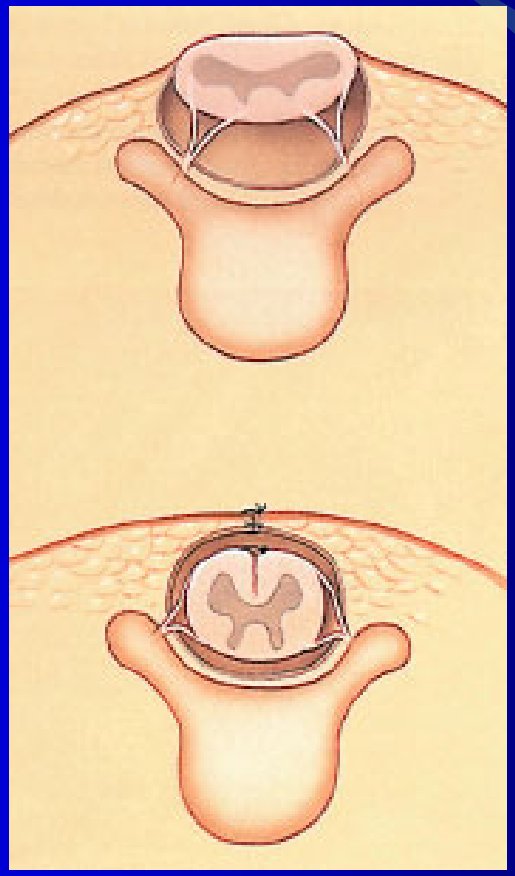


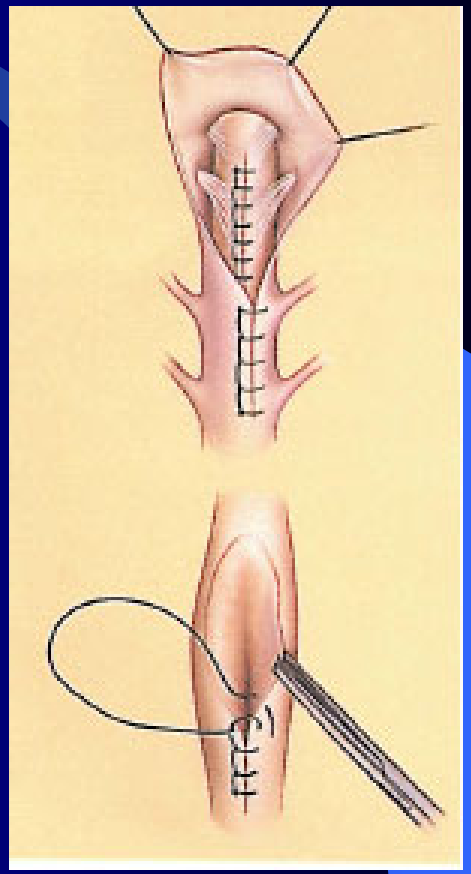


Errors of Neurulation

Myelomeningocele



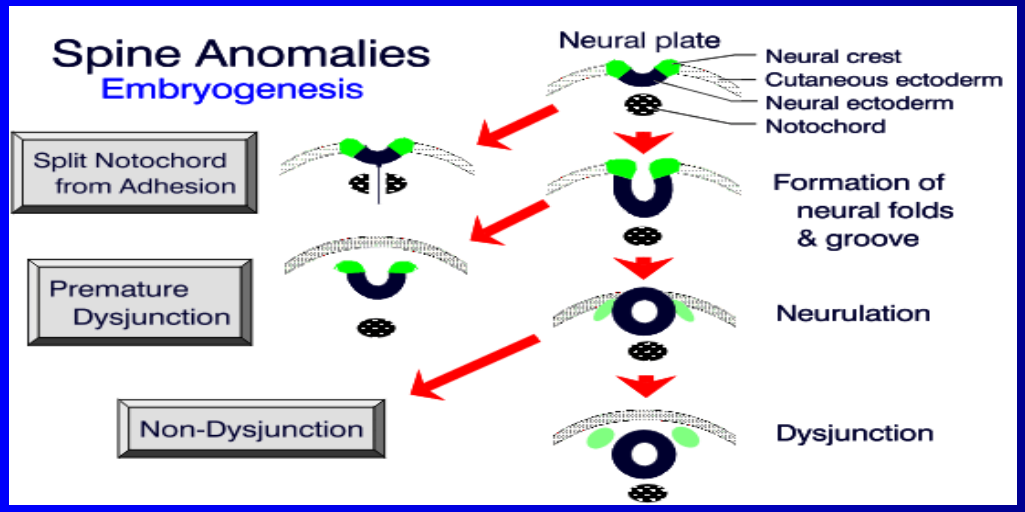


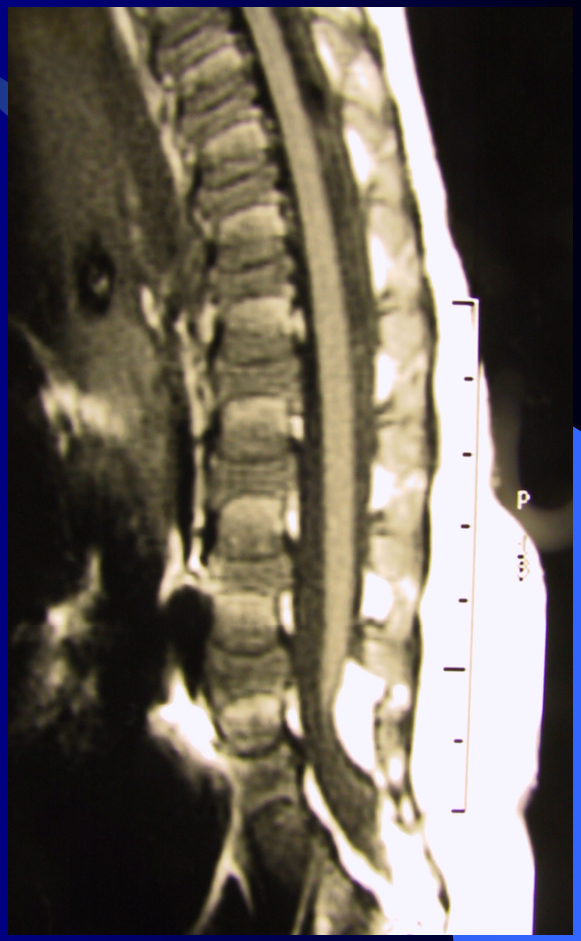


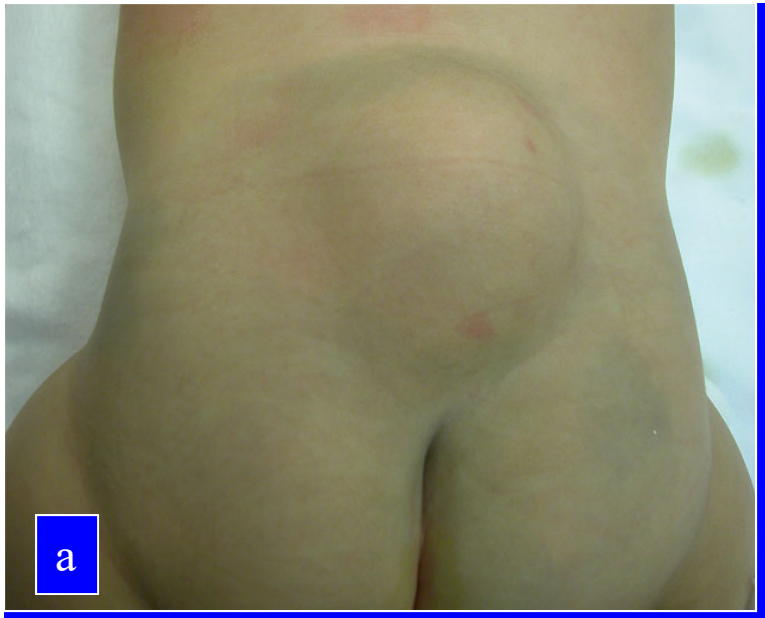
Errors of Dysjunction

Lipoma

- Incomplete separation of ectoderm from neural tube leading to inclusion of dermal elements
- Not a variant of myelomeningocele
  -NO HYDRO / CHIARI
- Often with abnl filum terminale







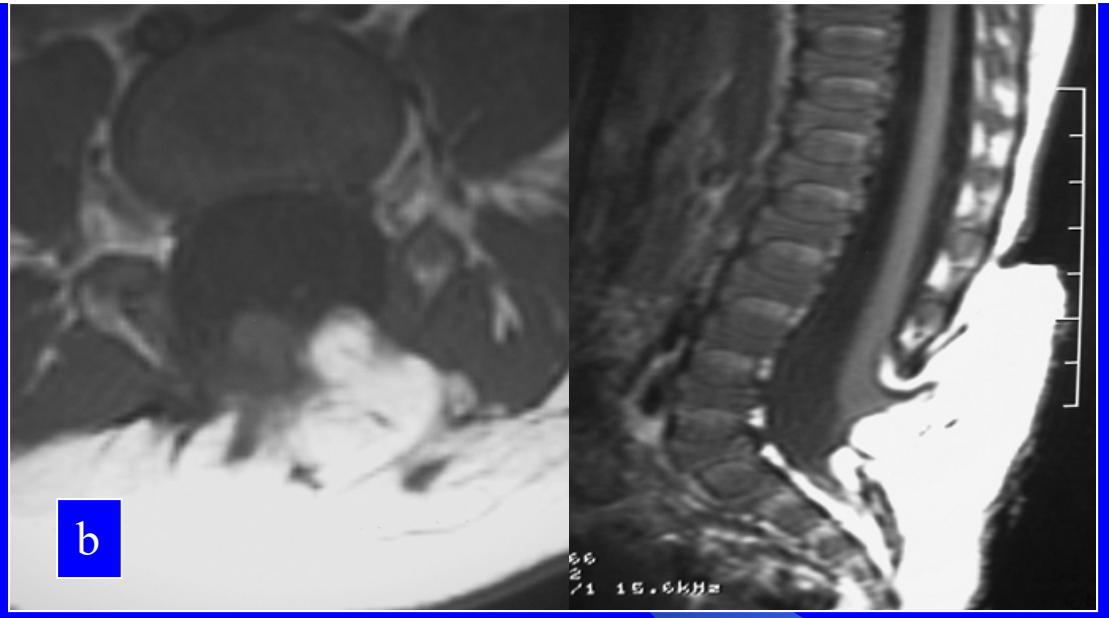
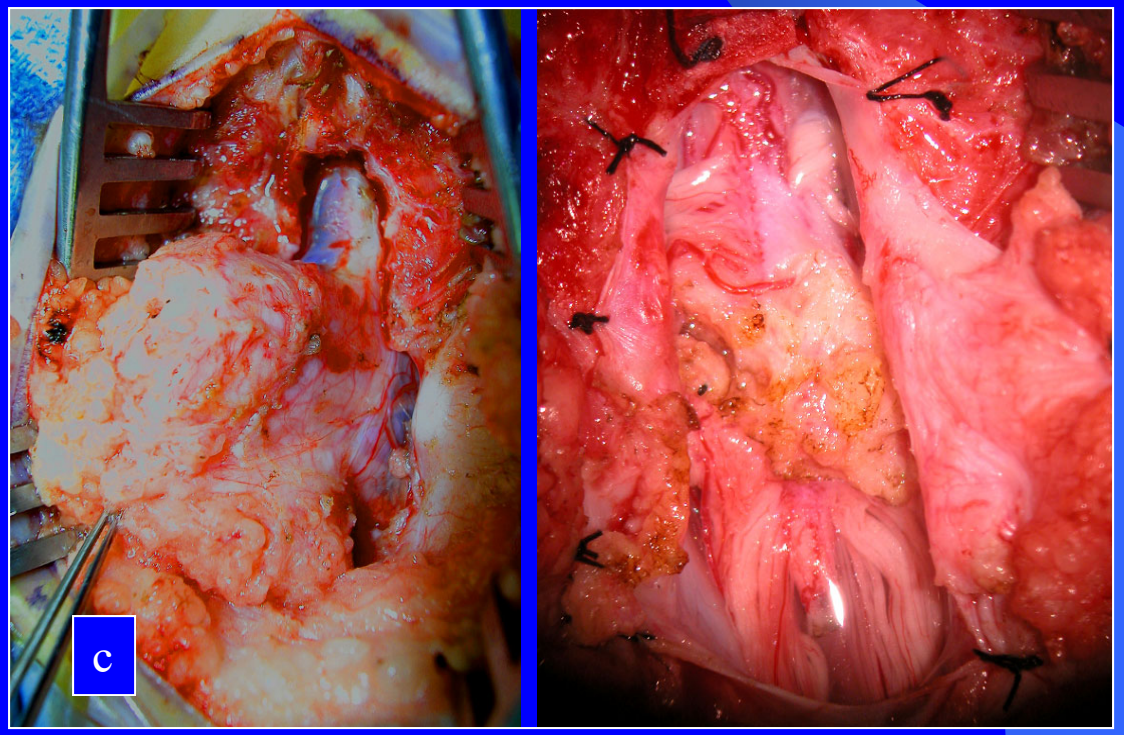
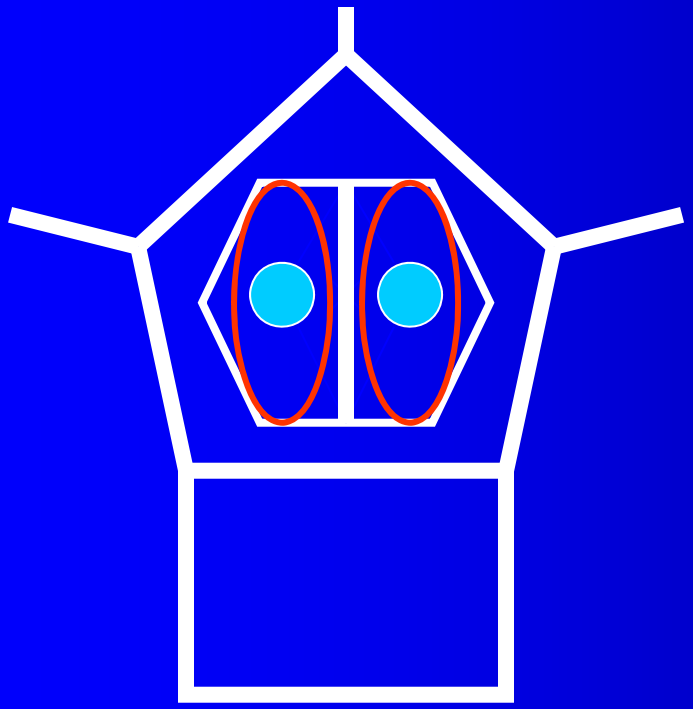


Figure 10: Lipomyelomeningocele
a) Preoperative image demonstrating large ballotable mass of the dorsal lumbosacral area
b) T1 axial and sagittal MRI showing low lying conus with lipomatous mass extending from cord through the dorsal elements into the subcutaneous tissue
c) Intraoperative images following isolation of lipoma (left) and resection to the liponeural junction (right).

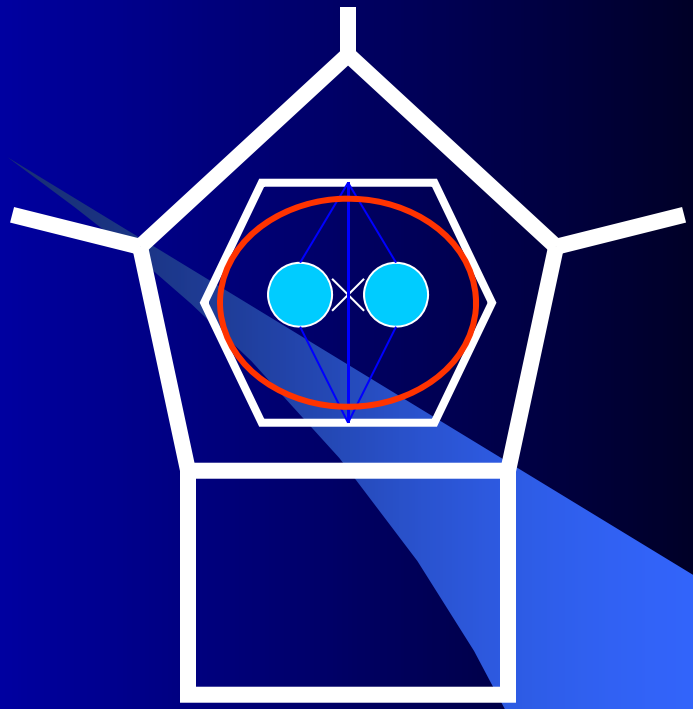


Errors of Dysgenesis

Split cord malformation



Type 1



Type 2

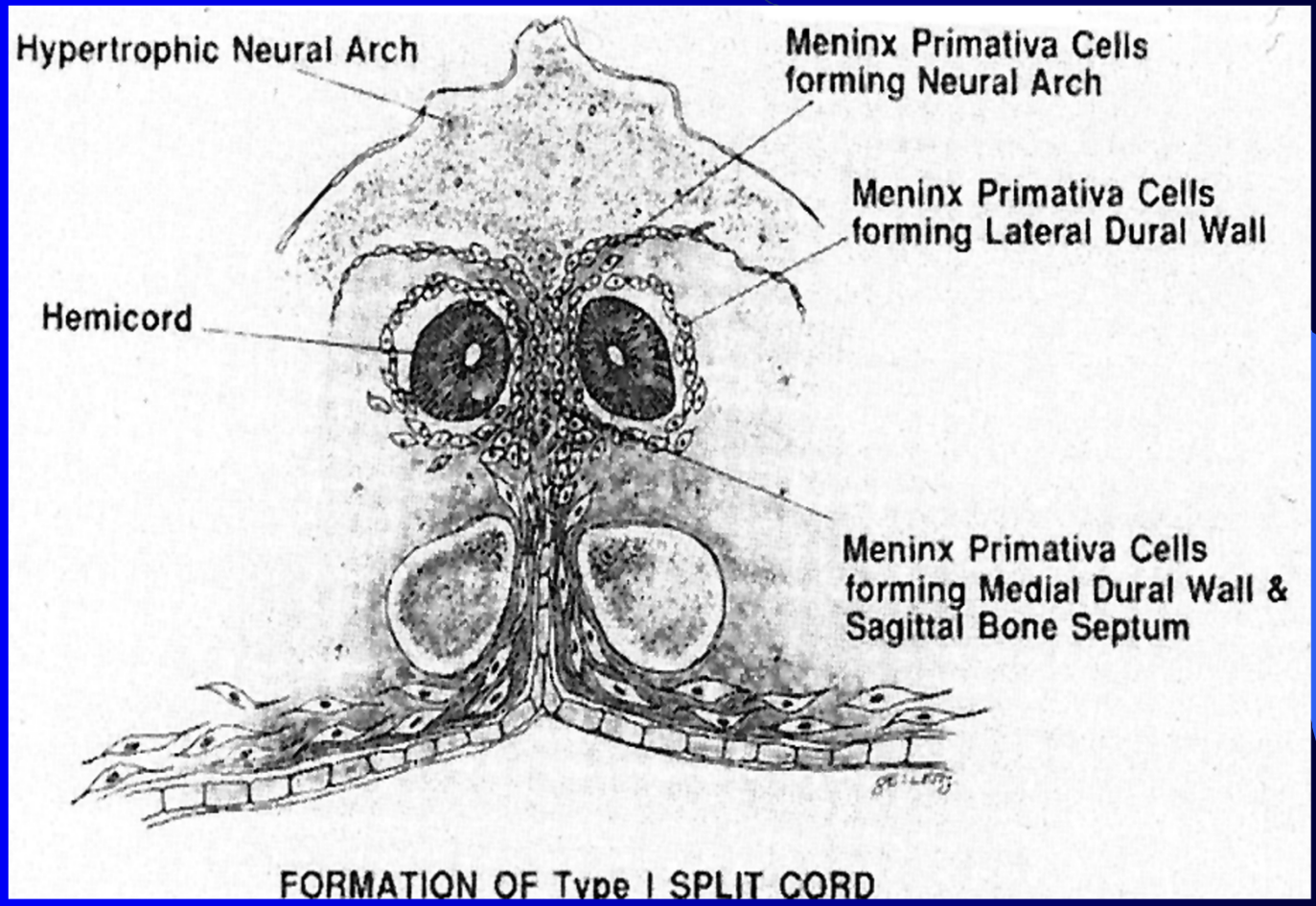
Figure 9: Classification of split cord malformations

-Type 1 malformation: Two hemicords housed in separate dural tubes. The intervening septum is comprised of bone, fibrous tissue, and dura

-Type 2 malformation: Two hemicords housed in a single dural tube. The interventing septum is comprised of fibrous tissue only

Embryogenesis

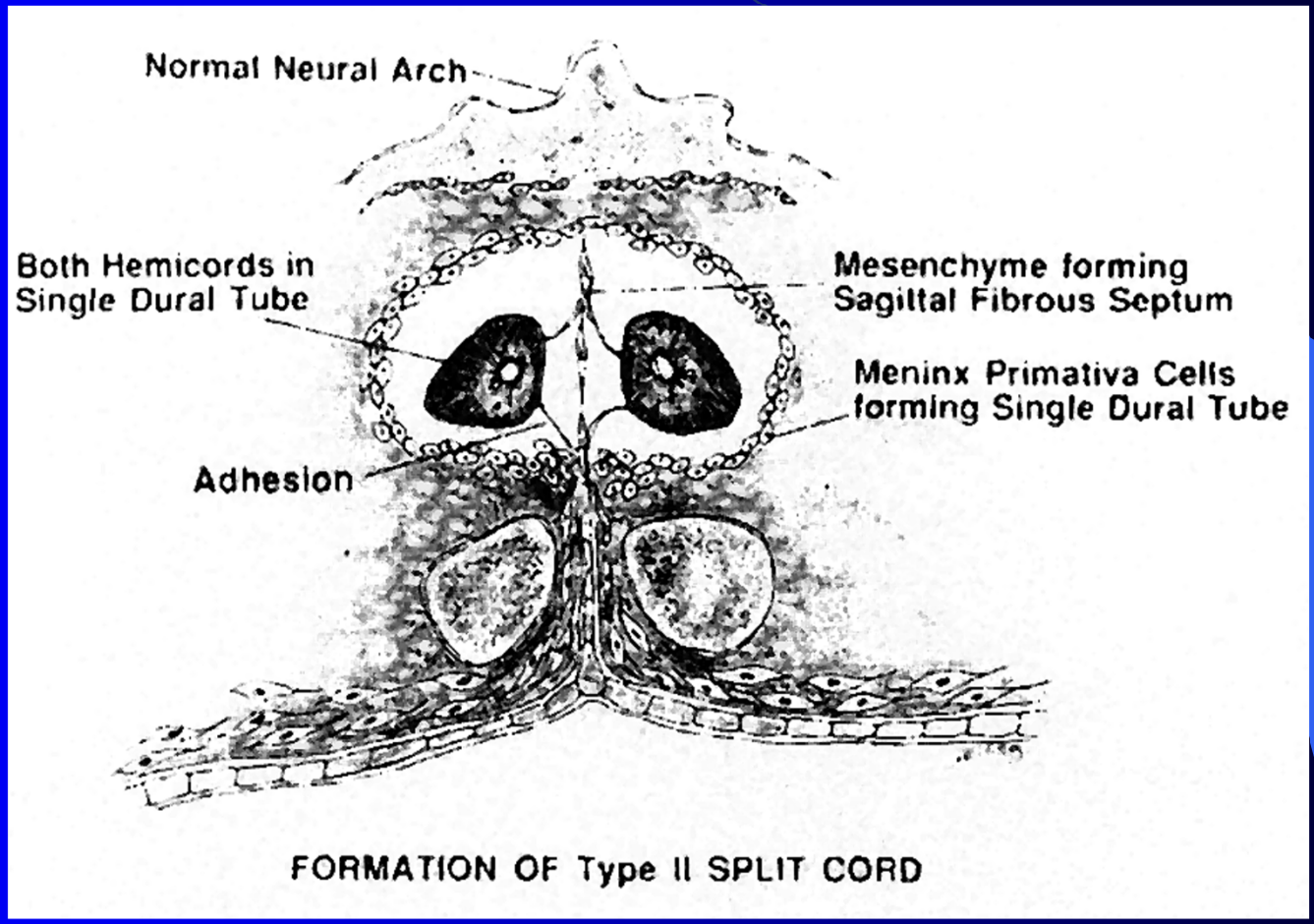
Type I SCM



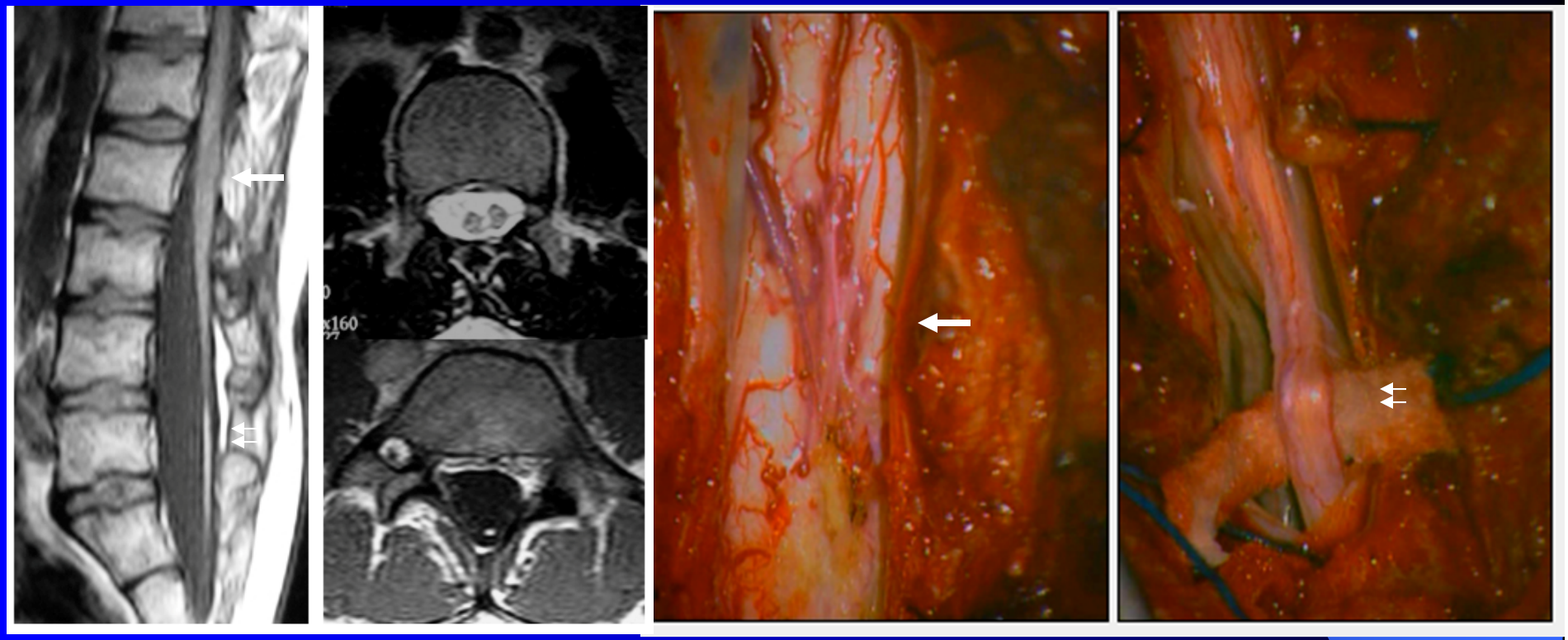
Pang D, Dias M, Ahab-Barmada M, *Neurosurg* 31(3) 451-80,1992

Embryogenesis

Type II SCM



Pang D, Dias M, Ahab-Barmada M, *Neurosurg* 31(3) 451-80,1992

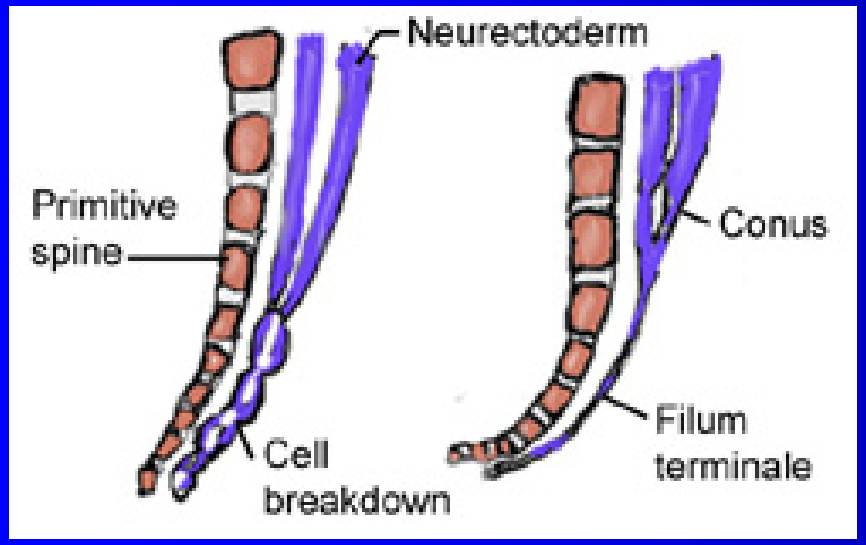


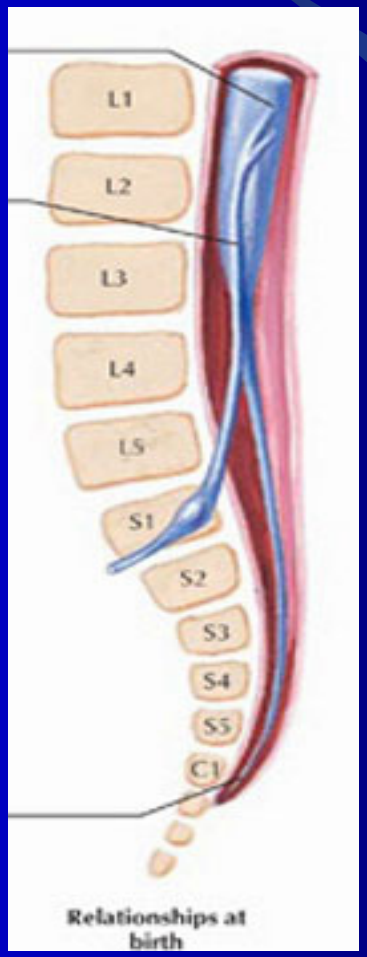
a b c d

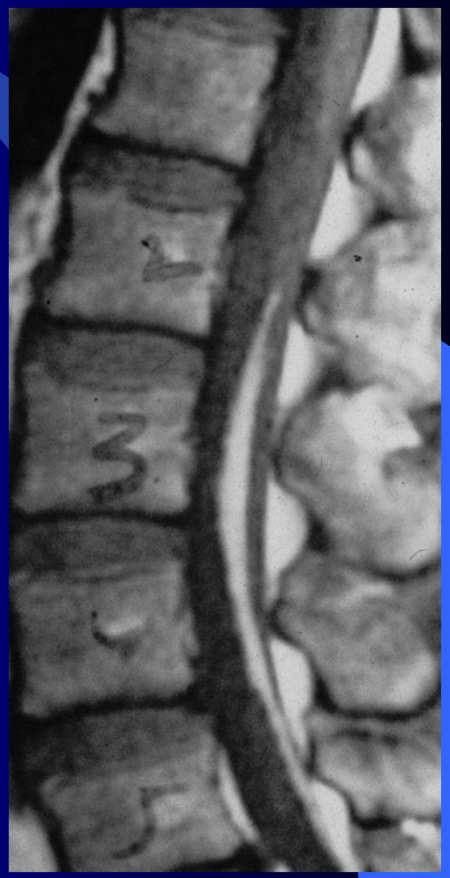
Figure 8: Type 2 split cord malformation

a) Sagittal T1 MRI demonstrates a low lying conus medularis (L3-4 disc space), fatty filum terminale (double arrow), and mixed signal in the distal cord concerning for split cord malformation (single arrow)
b) (above) Axial T2 at the level of the distal cord demonstrating duplicated cords contained within a single dural tube. (below) Axial T1 image at the level of the filum demonstrating lipomatous filum terminale
c) Intraoperative image at the split cord malformation. Note the prominent vasculature within the cleft of the split as well as adhesions at the distal end of the split cord
d) Intraoperative image of the fatty filum prior to division

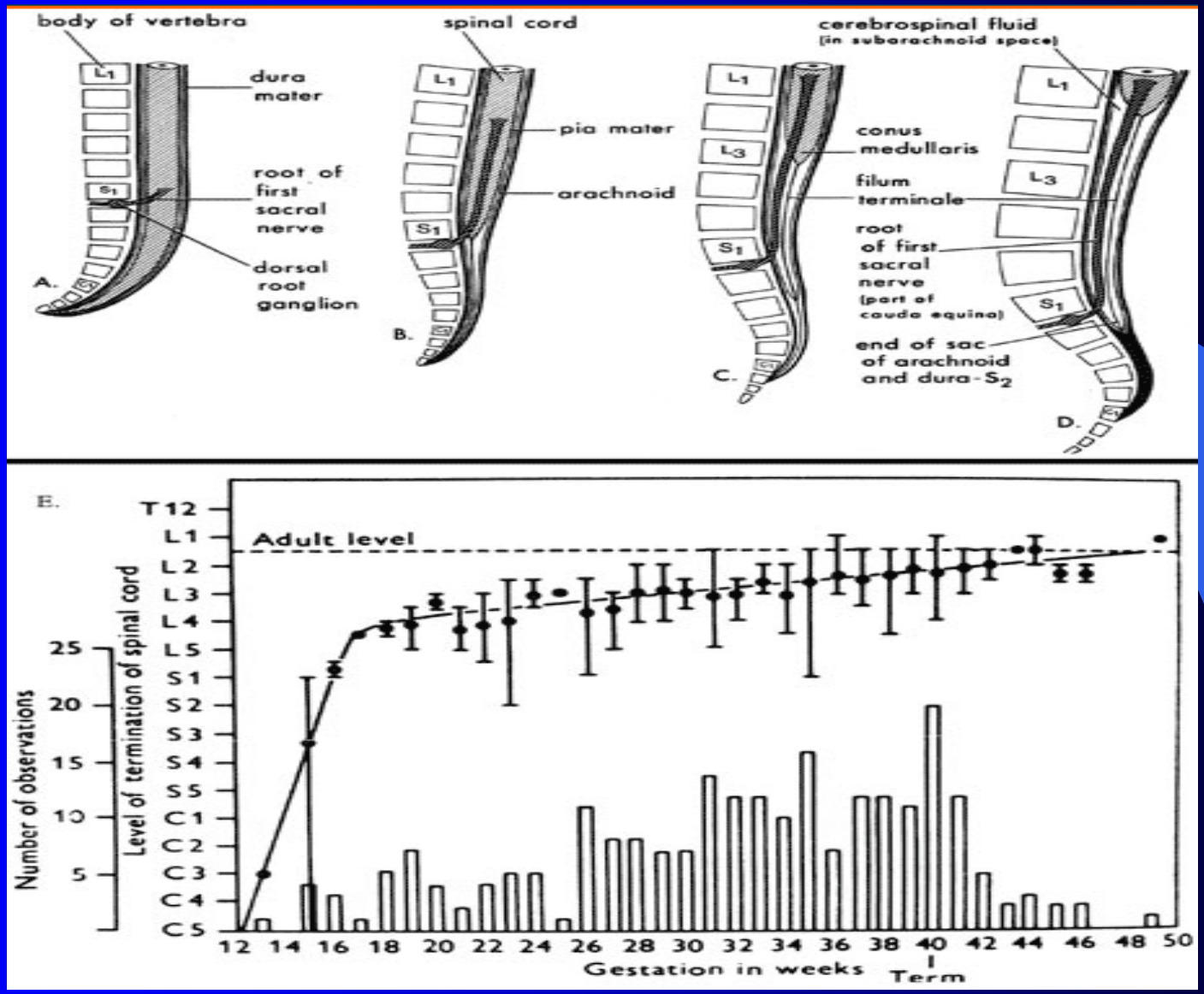
Errors of Dysgenesis Filum







Spinal cord relative to spine

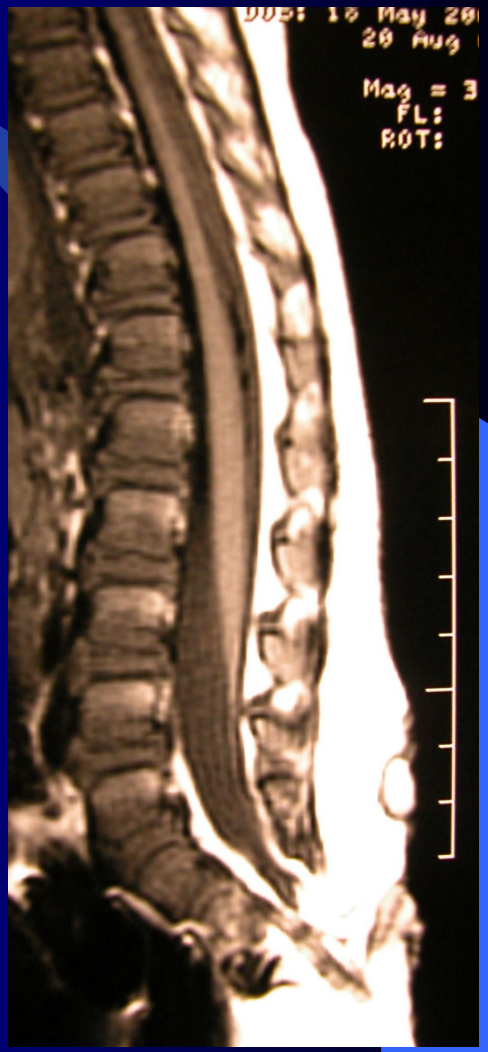


TETHERED SPINAL CORD

DEFINITION

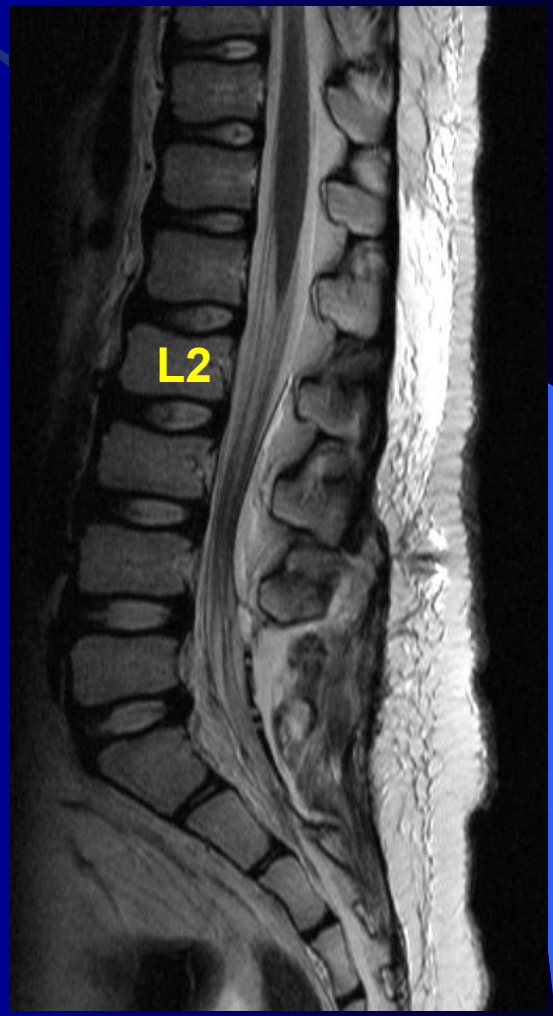
AN ABNORM. LOW LYING CONUS WHOSE NL ASCENT IS TETHERED SECONDARY TO A MULTITUDE OF DYSRAPHIC CONDITIONS.

RADIOGRAPHIC DX DOES NOT ALWAYS CORRELATE w/ CLINICAL PRESENTATION.



POSITION OF THE CONUS

- 20 wk embryo L4-5
- TERM L3
- 2 months postnatal L1-2 (95%)



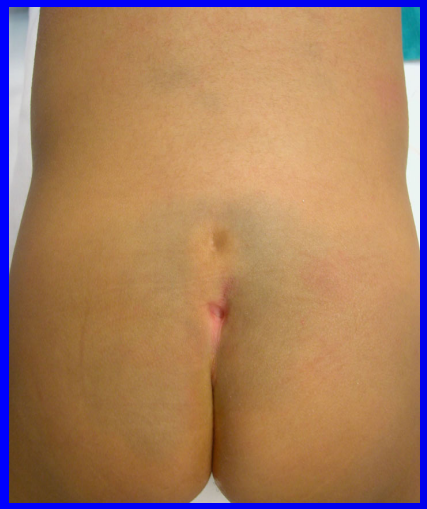
PATHOPHYSIOLOGY

- **STRETCH PLACED ON THE CONUS WILL LEAD TO PROGRESSIVE ISCHEMIA....**
  YAMADA 1981
- **WITH FLEXION THE NORMAL SPINE MAY MOVE CEPHALAD AS MUCH AS 2 CM'S....**
  BREIG 1970,YAMADA
- **SUDDEN TRACTION ON THE CORD MAY PRODUCE SYMPTOMATIC ONSET EVEN IN THE OLDER PATIENT AFTER CESSATION OF CORD GROWTH**

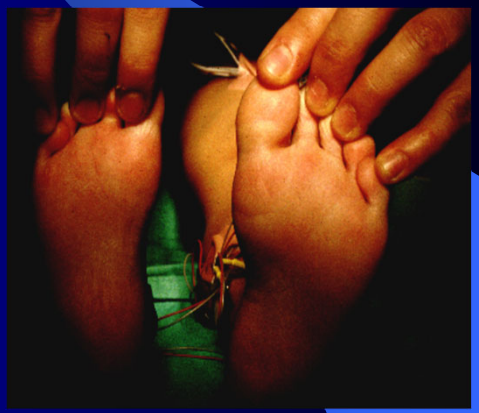
DIFFERENTIAL DIAGNOSIS

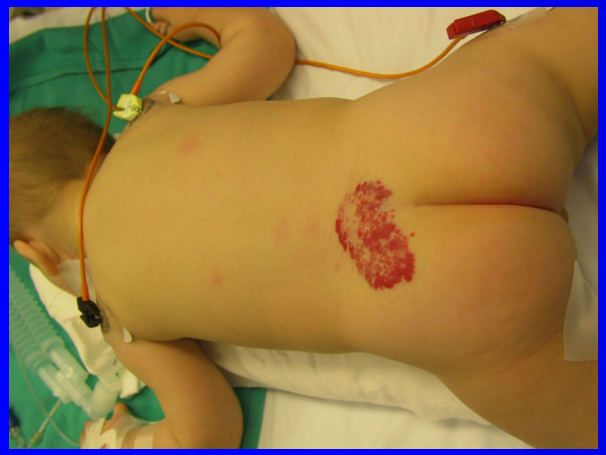
- MYELOMENINGOCELE REPAIR
- LIPOMYELOMENINGOCELE
- SPLIT CORD MALFORMATIONS
- HYPERTROPHIC FILUM TERMINALE
- DERMAL SINUS TRACT
- SLOW GROWING CORD TUMORS

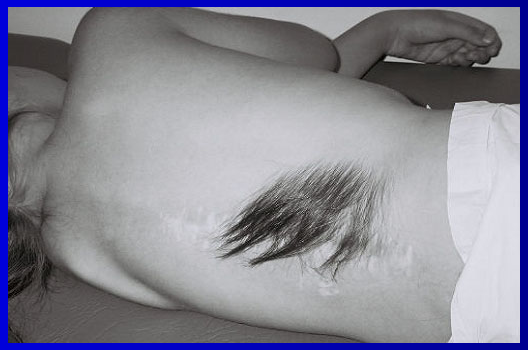
CLINICAL PRESENTATION

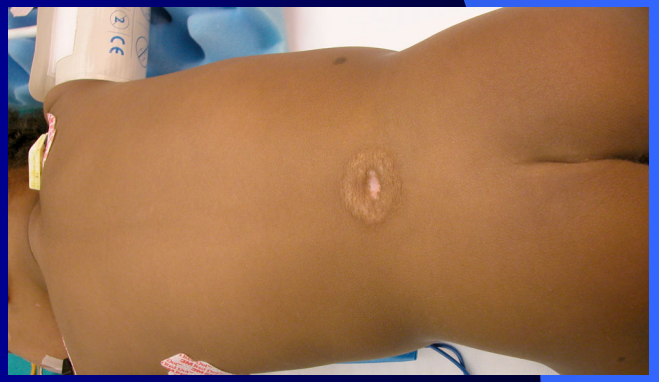


- CUTANEOUS MANIFESTATIONS
  - 30% OF TETHERED PTS --> SKIN LESIONS
    - DIMPLE, HEMANGIOMA, HAIRY PATCH, SQ LIPOMA, CUT. APLASIA
- LOWER EXT.
  - ASYMMETRY
- SCOLIOSIS









NEURO MANIFESTATIONS

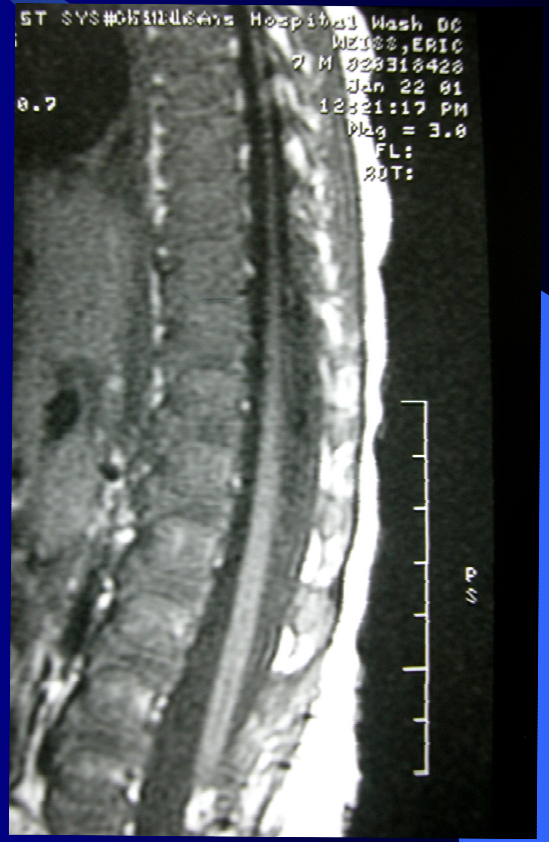
- BACK OR LEG PAIN / CRAMPING
- GAIT DISTURBANCE (fr. equinis / pes deform.)
- PROGRESSIVE SCOLIOSIS
- PROGRESSIVE WEAKNESS / NUMBNESS
- BOWEL / BLADDER HABIT CHANGES

ONSET OF SYMPTOMS

- USUALLY DURING GROWTH SPURTS
  - –7-10 YRS OLD, ADOLESCENTS
- MAY BE BROUGHT ON BY SUDDEN FLEX. MM IN THE SETTING OF TETHERING
  - –EXPLANATION FOR SXS IN THE OLDER PATIENT

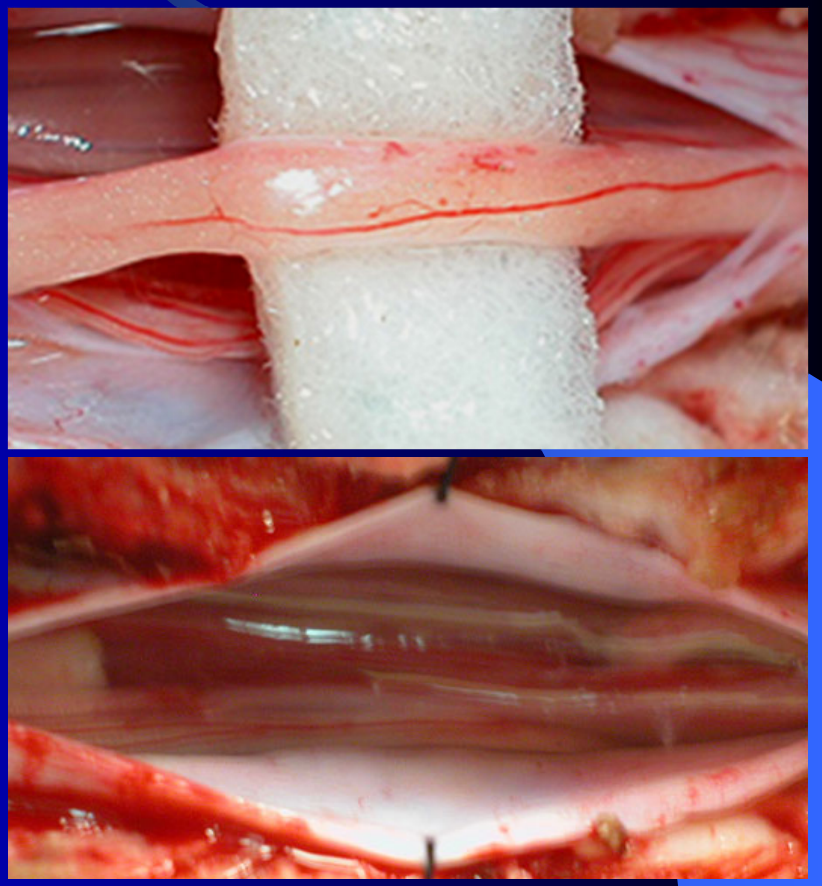
RADIOGRAPHIC INVESTIGATION

- PLAIN RADIOGRAPHS
  - SPINA BIFIDA BELOW L3
- MRI
  - LOW CONUS
  - THICKENED FILUM (> 2 MM)
  - LIPOMATOUS TISSUE
  - DYNAMIC STUDIES
- CT / MYELO
  - ALL OF THE ABOVE
  - HELPFUL



TREATMENT

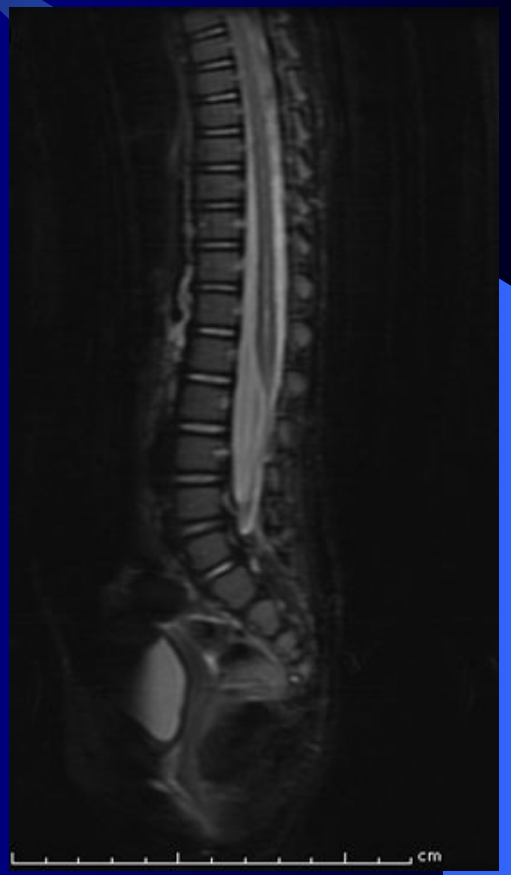
- FUNDAMENTAL SHIFT IN PHILOSOPHY
  - PROPHYLACTIC
  - R:B RATIO ADVANTAGE
- OBJECTIVE-->RELEASE THE CORD
  - MYELOMENING.--> FREE ADHESIONS
  - DIASTEMATO-->REMOVE SPURR/ADHESIONS
  - THICKENED FILUM-->CUT
  - LIPOMYELO.--> RESECT LIPOMA/ FREE ADHESIONS



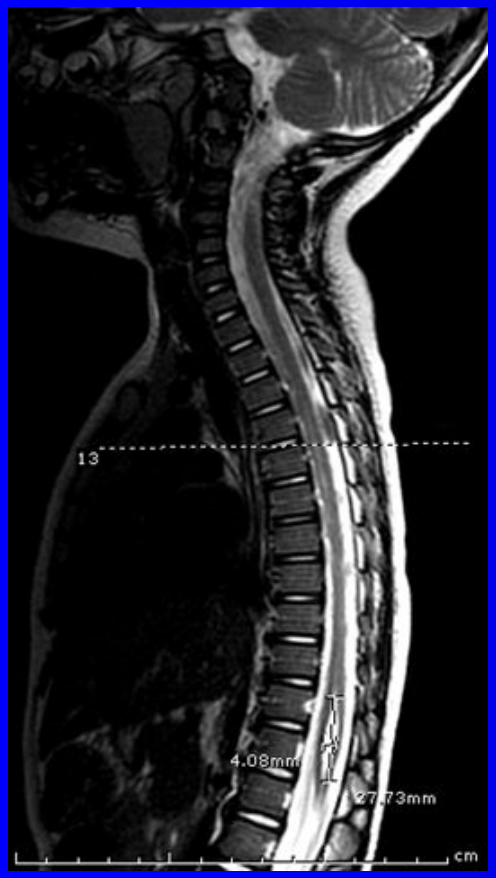
Uncharted territory

Equivocal position of conus

- Normal level above L2 95%
  - Not infrequent to see L2/3 or L3 w/ chromosomal deletion pts
- If no physical abnl as well as normal filum---> conservative course and follow



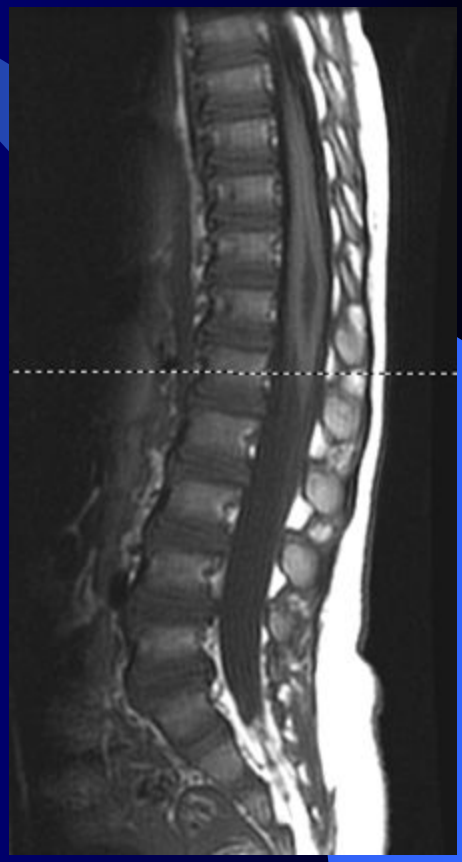
Terminal syrinx



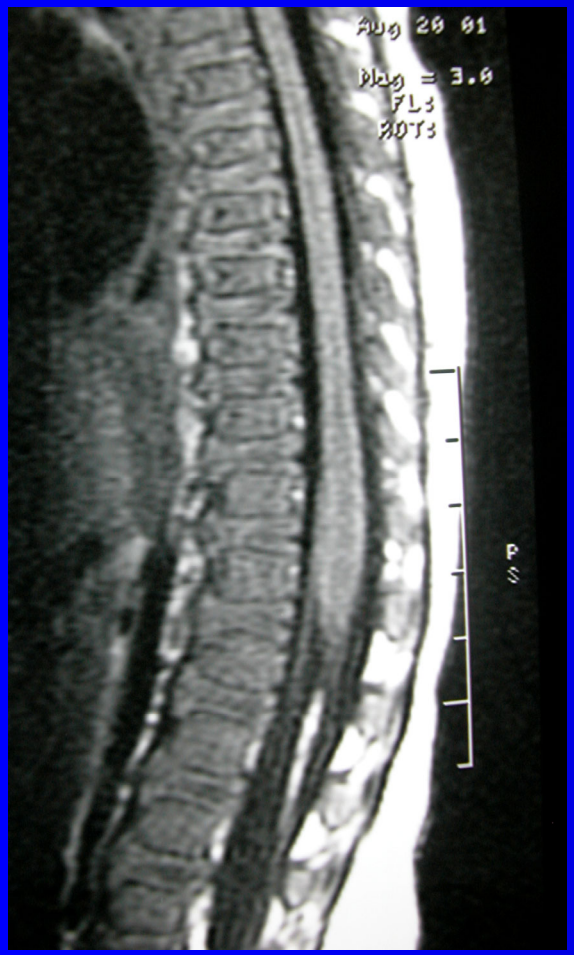
May represent disordered retrogressive differentiation

Often normal patients

Important to r/o any tethering and to follow for any potential B/B difficulties



Thickened filum w/nl conus



- Normal filum < 2mm in diameter
- Fatty infiltration may increase in size w/age
- Follow closely observing for teth cd signs/ sx

Conclusion

- Spinal cord embryogenesis consists of 2 parts:Neurulation and Canalization / Retrogressive Differentiation
- Spinal cord development in concert w/ spinal column (cord ends at L1/2)
- Errors of embryogenesis frequently affect both arenas (ie tethered cord)
- Many atypical MRI findings benign in long run but should be monitored


THANK YOU
R
MP
Page: 1 of 1
Compressed 27:1
IM: 1