SPINAL DYSRAPHISM
EVALUATION AND MANAGEMENT

Spinal dysraphism: classification

- Spinal dysraphism : Group of congenital abnormalities resulting from defective closure of the neural tube in the spinal region of neuraxis
- Encompasses all forms of spina bifida
- Classified into
  - Spina bifida aperta : Open communication of NT with environment
  - Spina bifida cystica : Myelomeningoceles
  - Spina bifida occulta : Covered with normally developed skin.

Myelomeningoceles

- Disorder resulting from defective primary neurulation
- Incidence
  - 0.4 per 1000 live births
  - Racially variable
- 85% caudal thoraco lumbar spine, 10 % in the thorax and the rest cervical
- 80-90 % associated with hydrocephalus and Chiari

Myelomeningoceles

- Neurological deterioration can occur
  - Symptomatic HCP or Chiari-II
  - Hydrosyringomyelia
  - Retethering of cord

  Mostly due to hydrocephalus from shunt malfunction

Risk Factors for NTD

Risk factors	Risk %
Previous pregnancy with NTD	2-3%
Partner with NTD	2-3%
Diabetes mellitus type 1	1 %
Seizure disorder	1 %
Close relative with NTD	0.3-1 %
Post pregnancy obesity	0.2%

Risk Factors for NTD

- Agricultural pesticides and chemicals
- Cleaning solvents and disinfectants
- Nursing
- Radiation
- Anesthetic agents
- Hot tubs, saunas and fever (hyperthermia)
- Lead
- Tobacco smoke

Recommendations for dietary Folate supplementation

Circumstance	Dose
Before pregnancy	
•Women with no risk factors	0.4 mg/ day
•Women at high risk	4 mg day
During pregnancy	
•Women with no risk factors	0.6 mg /day
•Women at high risk	4mg /day
Post partum with breast feeding	
•Women with no risk factors	0. 5 mg /day
•Women at high risk	4mg /day

Prenatal Diagnosis

- Maternal serum Alpha feto protein : initial screening test
- High resolution Fetal Ultrasonography.
  - Can also demonstrate hydrocephalus and Chiari-II abnormality (Lemon and Banana sign)
- Amniocentesis : if MSAFP and USG are suggestive
  - Ach esterase levels along with AFP
- AFP can increase in other developmental anomalies of the gut and kidneys.

Preop evaluation

- General
  - Repaired within 72 hrs
  - Enteral feeding avoided to prevent fecal soiling of placode
  - Prone position, saline dressings
- Neurosurgical
  - Sensory level determined
  - Motor evaluation – Distal most voluntary motion evaluated. Limb abnormalities documented.
  - Anal tone and anal reflex evaluated

Preop evaluation

- Ventricular size documented with preop USG and NCCT head.
- Observe for symptoms of Chiari II
- Renal evaluation
  - 90 % have neurogenic bladder.
  - All should have preop Renal ultrasound for detecting severe anomalies.
  - CIC if fails to void.

MMC repair

- Timing
  - Within 72 hrs
  - Delayed repair – high risk of meningitis/ ventriculitis/ shunt infection. Mortality 13 %
    - Obtain placode culture
  - VP shunt placement along with MMC repair recommended if e/o HCP at birth

MMC repair

- Surgical procedure
  - Patient positioned prone
  - Contact of Povidone iodine solution with neural placode to be avoided.
  - Goal of surgery
    - Protect the functional tissue in the neural placode
    - Prevent CSF loss
    - Minimize risk of meningitis by reconstructing neural tube and coverings

Post op care

- Per- operative antibiotics
- Prevention of fecal contamination of wound
- Nurse in Trendlenberg's
- Observe for HCP – shunt if HCP present
- Complications
  - Superficial wound dehiscence
  - Meningitis
- Symptomatic Chiari
  - Ensure functioning shunt
  - Hindbrain decompression

Occult spinal dysraphism

- Congenital spinal defects covered by intact skin
- Causative lesions
  - Fatty filum terminale
  - Lipomyelomeningocele
  - Split cord malformations type I and II
  - Inclusion lesions (dermoid, dermal sinus tract)
  - Neurenteric cyst
  - Myelocystocele

Manifestations of occult spinal dysraphism

Cutaneous stigmata	Orthopedic deformities	Urologic problems
Asymmetric gluteal cleft	Foot or leg deformities	Neurogenic bladder
Capillary hemangioma	Scoliosis	UTIs
Subcutaneous lipomas	Sacral agenesis	Incontinence
Hypertrichosis		Delay in toilet training
Dermal sinus tract		
Cutis aplasia		

Neurological signs and symptoms in different age groups

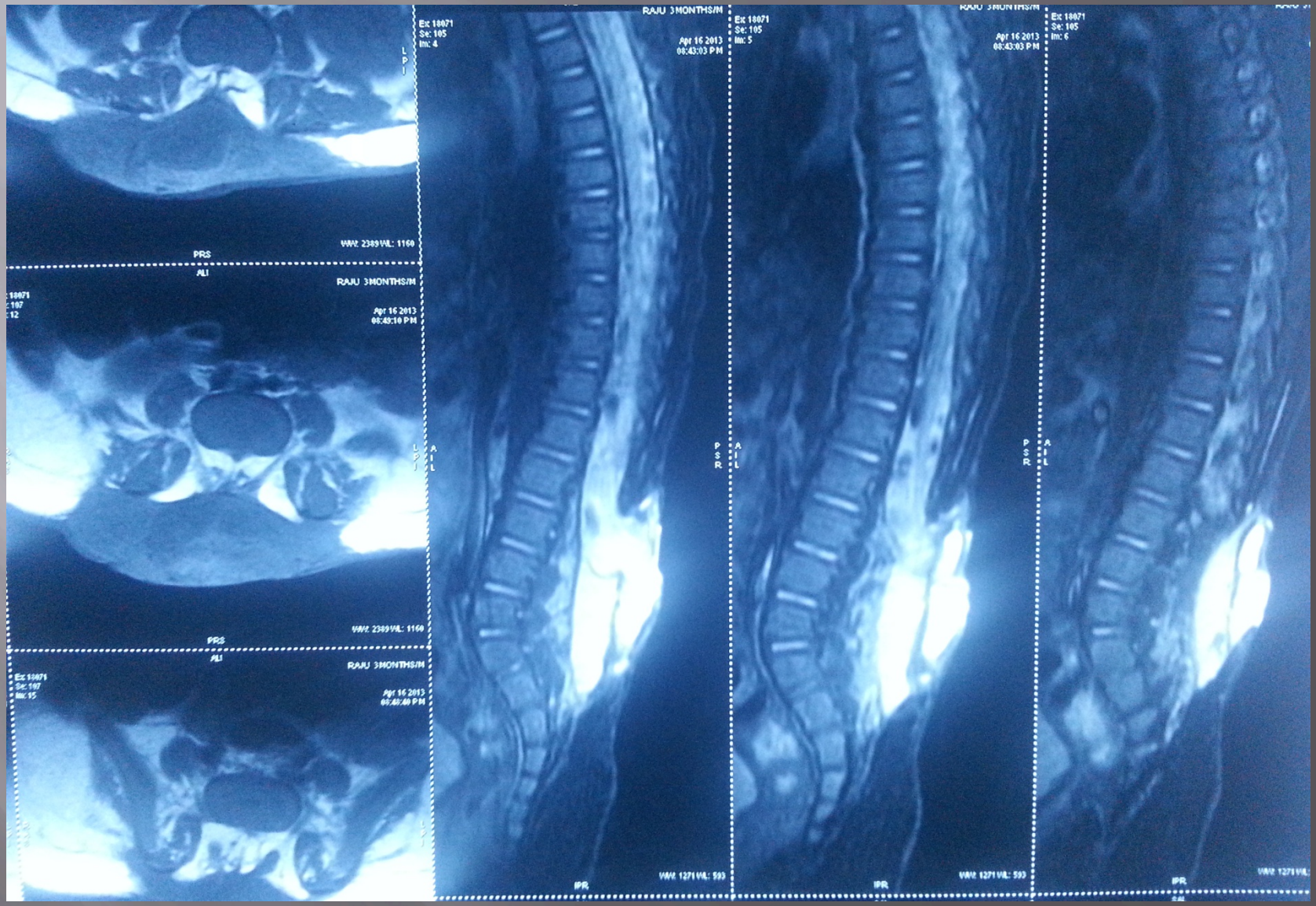
Infants	Toddler	Older children	Young adults
Decreased spontaneous leg movements	Delayed walking	Asymmetric motor/ sensory development	Back pain
Absent reflexes	Abnormal gait	Back/leg pain	Leg cramping/ pain
Leg atrophy		UMN signs	Spasticity
Foot asymmetry		Painless ulceration	Hyperreflexia
Decreased urinary stream			Bowel/bladder incontinence.

Plain radiological findings

Structure	Findings
Lamina	Fusion defects,midline defects,abnormal spinous processes
Vertebral bodies	Hemivertebrae,Butterfly vertebrae,Block vertebrae,Midline cleft defects, canal stenosis
Disk space	Congenital narrowing
Pedicles	Flattening, thinning
Widening of spinal canal	Interpedicular widening, scalloping of posterior border, Midline bony spur.
Failure of development	Reduced number of vertebral bodies, Absence of parts of vertebrae, sacral dysjunction
Spinal curvature	Scoliosis, kyphosis, lordosis.

Lipomyelomeningoceles

- Lipoma tethering the cord to the subcutaneous tissue
- Fascial, spinous and dural defect
- Lipoma cord interface distracted out of the spinal canal by traction created by tethering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- Classification
  - Type I (Dorsal lipoma)
  - Type II (Transitional lipoma)
  - Type III (Terminal lipoma)

- Dorsal lipoma (Type I)
  - Fibrolipomatous stalk tethering cord proximal to conus
  - Usually at middle lumbar to lumbo sacral level
  - Dorsal spinal cord dysraphic at site of attachment of lipoma
  - Site of attachment medial to the dorsal root entry
  - Normal spinal cord distal to myeloschisis.
  - Roots lie within the subarachnoid space.

- Caudal or terminal lipomas (Type III)
  - Directly from conus medullaris or filum terminale
  - Largely or wholly intradural
  - Nerve roots entangled in the lipoma
  - Lipoma cord interface caudal to the dorsal root entry zone.
  - Filum may be fatty, thickened and sometimes attached to subcutaneous tissue (sacral dimple).

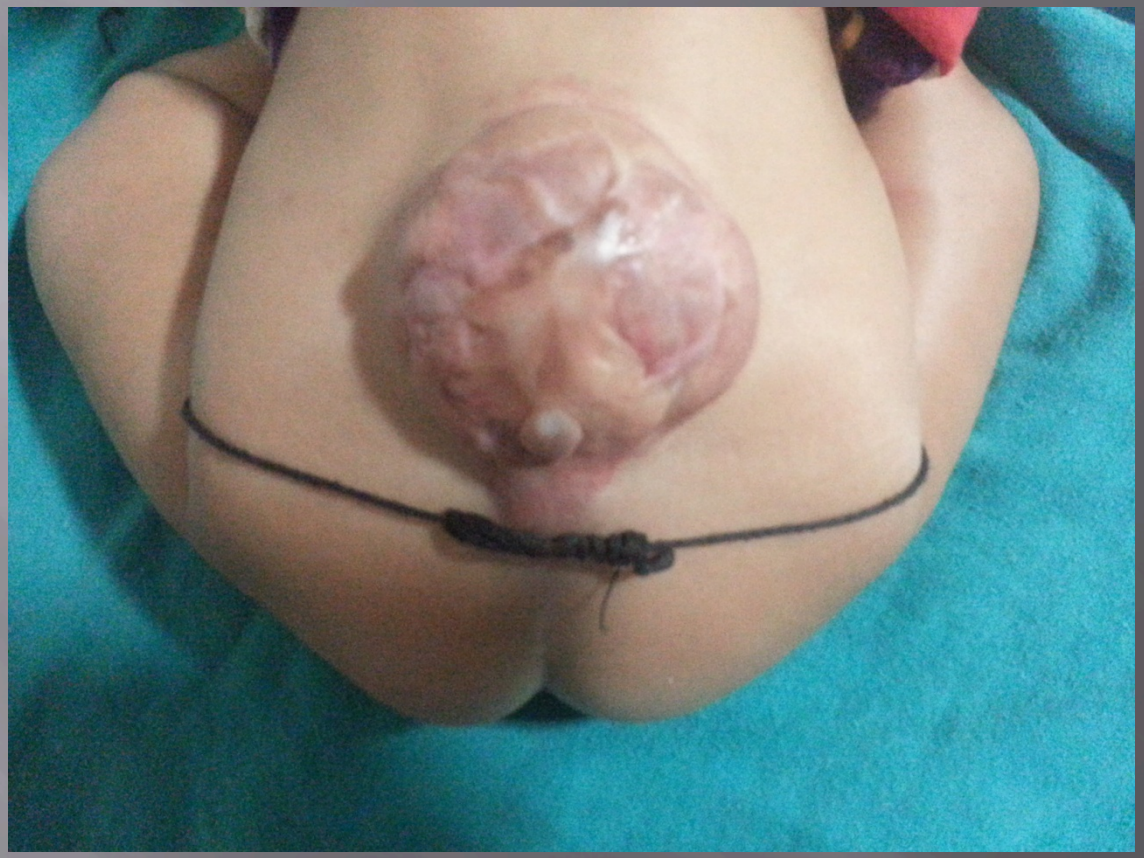
- Transitional lipomas
  - Share the characteristics of both Type 1 and type 2.
  - No normal spinal cord distal to lipoma attachment
  - Initially dorsal roots may be separate but caudally become enmeshed into the lipoma.
  - Frequently asymmetric attachment to cord.

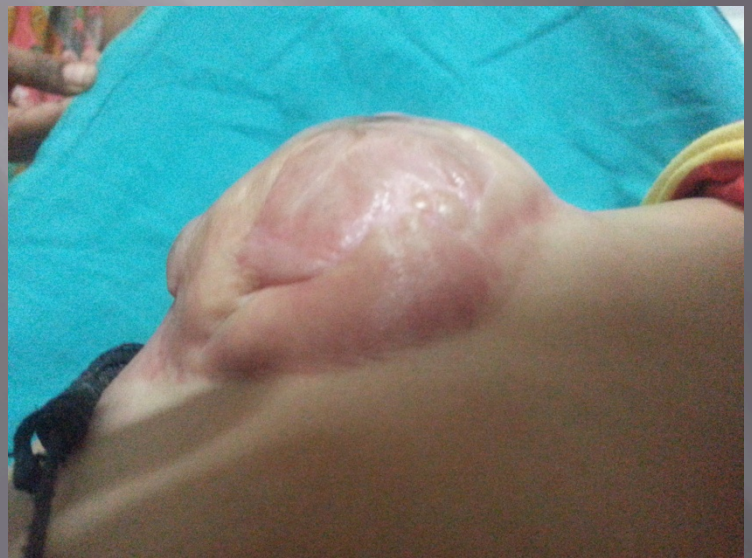
Abnormal embryology LMMC

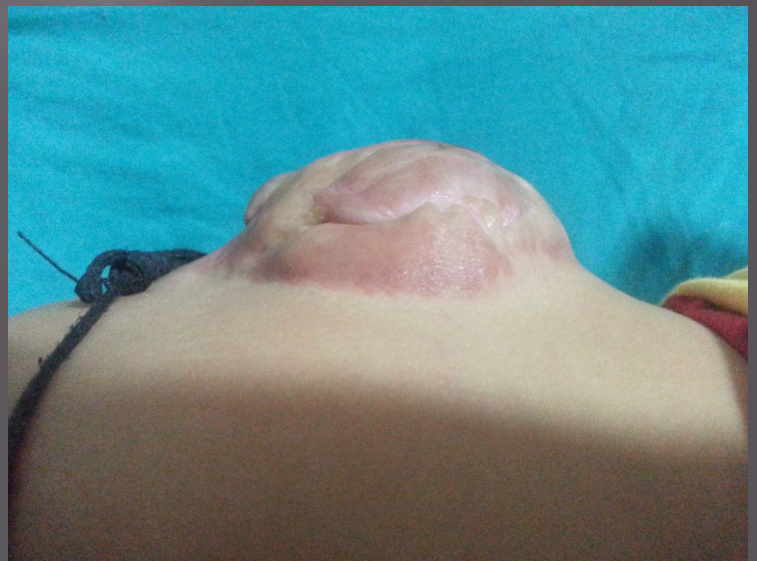
- Usually a disjunction in timing of neural tube closure and cutaneous ectoderm closure
- Elements of the ectoderm become incorporated into the incompletely closed neural tube.

Clinical features

- Subcutaneous masses over the back
- Stigmata of occult dysraphism
  - Hypertrichosis
  - Hemangioma
  - Hypo/ hyperpigmented patch
  - Dermal pit or sinus
  - Atretic meningocele
  - Asymmetric gluteal cleft







- Inexorable symptomatic progression seen in untreated cases
- Risk of precipitous neurologic deterioration
  - Orthopedic syndrome
    - Limb length discrepancy, high pedal arches, hammer toes, calcaneovarus/ valgus foot deformity.
  - Urologic syndrome
    - Urinary incontinence, post void dribbling, urgency, frequency
  - Intractable pain in the legs, back, pelvis or perineum.

Indication for surgical repair

- Asymptomatic infant older than 2 months
- Presence of orthopedic, pain or urologic syndrome
- Neurological symptoms
- Prior to corrective spinal surgery.

- Goals of surgery
  - Detethering of spinal cord
  - Decompression of the cord by removing as much lipoma as possible
  - Preservation of the functional tissue
- Surgical principles
  - Relationship between the lipoma-cord interface and dorsal roots to be established
  - Conservative excision of the lipoma to avoid injury to the cord/ exiting roots.

- Complications of surgical repair
  - Early – CSF leak/ pseudomeningoceles
  - Late – retethering of the cord
    - Mostly presenting between 3-8, 11-22 months after surgery
    - Upto 20% cases may demonstrate retethering
    - Diagnosis primarily clinical.

Lipoma of the terminal filum

- Less severe form of OSD
- More than 2 mm thickness of the filum on MR imaging
- Frequently associated with sacral/gluteal cleft dimples.
- May be associated with VATER association, imperforate anus, cloacal extrophy and other urogenital abnormalities.
- Sometimes a/w sacral agenesis
- Reflects defective secondary neurulation

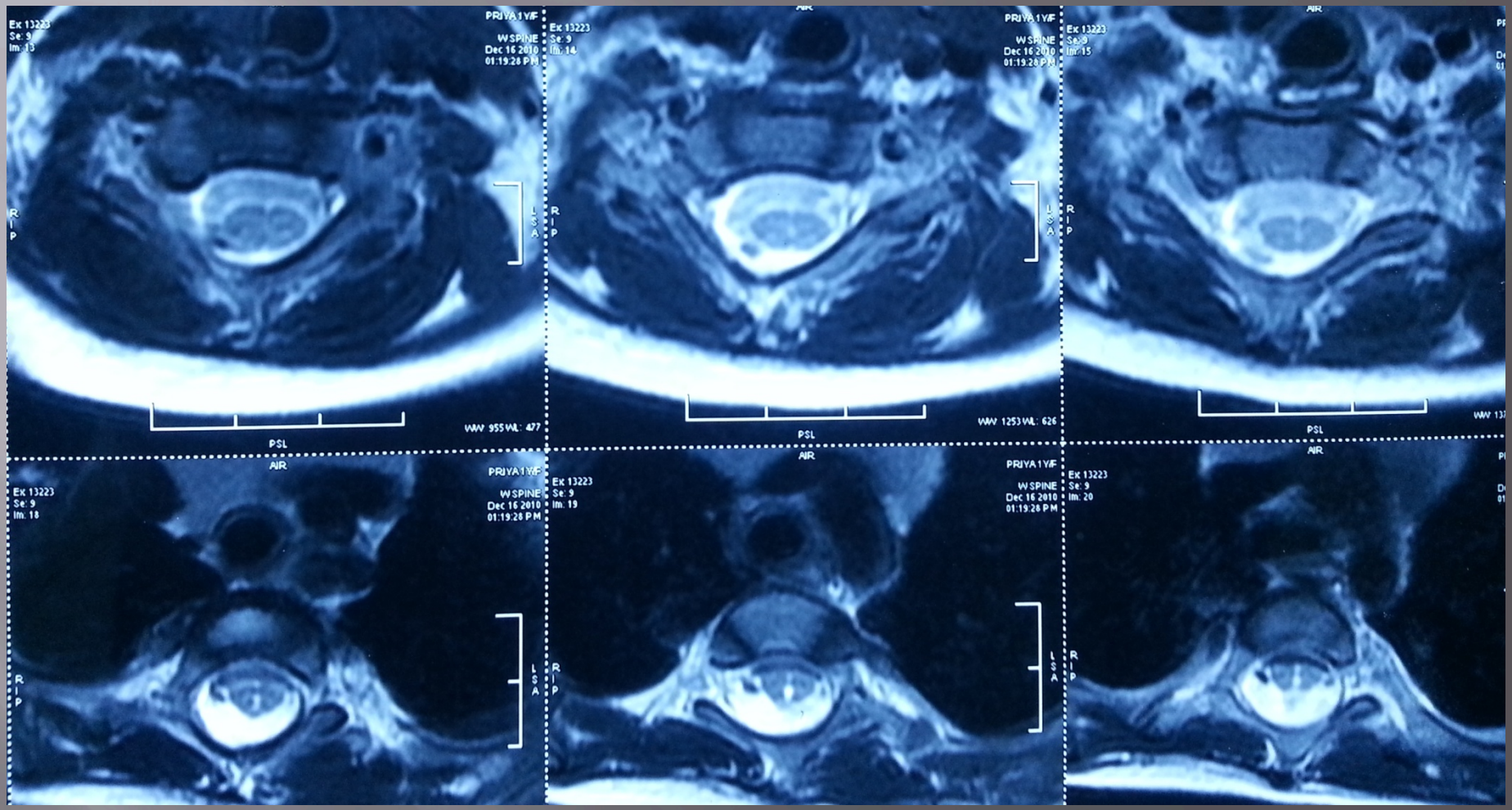
- Clinical presentation
  - Orthopedic
  - Urologic
  - Pain
- All asymptomatic infants and symptomatic adults are surgical candidates.
- Surgical procedure is the exposure of filum through lumbosacral laminectomy or interlaminar approach
  - The filum identified separated from nerve roots and cut.

Split cord malformation

- Longitudinal developmental splitting of the cord over one or multiple levels
- Type I split –separate dural sheaths for the two hemicords separated by intervening osseous/ fibrocartilaginous septum.
- Type II split – single dural sheath, hemicords separated by fibrotic bands.
- Both types may be present simultaneously at different levels

- SCM more common in females (M:F=1:3)
- Average age of presentation is 4-6.5 years
- Cutaneous stigmata most specifically hairy patch
- Orthopedic deformities including scoliosis also commonly seen
- Clinical course similar to other patients with tethering of spinal cord.

- In Type I – Bony septum prevents ascent of spinal cord
- In Type II – Fibrous bands tether the cord to the dural sheath
- Coexistent neuroepithelial cysts, lipomas, dermoids and syrinxes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- Operative management
  - Surgical detethering of cord by excision of the bony spur/ division of the fibrous bands
  - Caution exercised to avoid damage to the hemicords during excision of the spur.
  - Type I split further classified into A- D types (Gupta et al Pediatric neurosurg 2006,42, 341-46)
    - Location of the bony spur
    - Free space available around it
    - Correlating with complexity of surgical repair.

Neurenteric cysts

- Persistent neurenteric canal communicating between yolk sac and amniotic cavity
- Intradural, extramedullary mucosa lined cysts
- Formed from persistent tracts communicating with respiratory and gut epithelia.
- Associated with vertebral anomalies
- MRI- demonstrates non- contrast enhancing intradural extramedullary cyst

- Presentation usually in late years (50-60 years)
- May also present in pediatric age group
- Most common location is cervico- thoracic
- Usually postero-lateral surgical approach
- Complete excision of cyst – long term symptom free survival.

Dermal sinus tracts

- Abnormal tracts communicating between the skin and intraspinal compartment.
- Most common- lumbosacral location
- May occur anywhere from nasion to coccyx in midline
- May be accompanied by other cutaneous stigmata.
- Tract terminates within thecal sac mostly
- Half may have associated dermoids, epidermoids, teratoma at termination.

- Potential pathway for spread of infection
- Repeated episodes of meningitis with atypical organisms
- Operative repair consists of complete excision of the track under prophylactic antibiotic cover.
- Gram positive and gram negative coverage

Meningoceles

- Distinguished from MMCs by absence of hydrocephalus , Chiari malformation or lower limb abnormalities
- Dural defect through with CSF space and meninges herniate.
- Concomitant neurocutaneous lesions such as lipomas, dermal sinus tracts may cause tethering
- Surgical repair of defect at 4-6 months of age.

- Anterior meningoceles
  - Herniation of meninges and CSF in ventral location
  - Commonly in presacral and lumbosacral region
  - Female predominance
  - Currarino's triad- anorectal abnormalities, presacral mass, sacral bony abnormalities.
  - Presacral tumor may be epidermoid, dermoid or teratoma
  - Meningitis by atypical organisms may also occur.
  - Posterior surgical approach to repair the dural defect and dether the cord.

Myelocystoceles

- Terminal dilatation of the central canal that herniates through defective posterior elements
- Expanded spinal cord, CSF, fibrous bands and meninges and lipomatous elements
- Result from disordered development of the caudal cell mass
- Associated anomalies of the anorectal system, lower GI tract and spinal column

Myelocystoceles

- Surgical repair attempted because of the resultant tethering of spinal cord.
- Repair within 6 months
- Per-op trumpet shaped distended conus often adheres to the superficial fat
- Detethering may be difficult.

Thank you