Tethered cord syndrome and occult spinal dysraphism

-Spinal dysraphism is a term that refers to all forms of developmental abnormalities occurring in the midline of the back—from the skin externally to the vertebral bodies internally.
-Spinal dysraphism incidence: 0.05 to 0.25 per 1000 births

Embryogenesis:

Nurulation (day 18–48): neural plate folds > forms neural tube, starts with upper cervical region and then extends caudally and cephalically, which is covered by cutaneous ectoderm.the caudal portion of the neural tube closes at the level L-1 or L-2. It gives rise to the spinal cord only down to the lumbar spine region.

Abnormalities at this stage give rise to: MMC, meningocele, lipomyelomeningocele, SCMs, dermal sinus, and intraspinal tumors such as dermoids and epidermoids

Canalization of the nail bud (Days 28–48): The tail bud forms after completion of neurulation. The formation of the neural tube caudal to that formed during neurulation occurs by canalization of the tail bud, This process consists of the development of vacuoles within the tail bud, then coalescence of these vacuoles to form the canal, which then connects with the rostral neural tube formed during neurulation.

Abnormalities at this stage give rise to the thick terminal filum, terminal myelocystocele, and lipomyelomeningocele.

Regression: The terminal filum and cauda equina are formed from the caudal portion of the neural tube. The ventriculus terminalis marks the level of the future conus medullaris and is a dilation of the central canal that can be identified (Days 43–48) at which time it lies at the coccygeal level. The tip of the vertebral coccygeal segments contains an epidermal cell rest, the coccygeal medullary vestige is formed when the caudal neural tube regresses between the ventriculus terminalis and the coccygeal medullary vestige (Day 52).

-During the fetal period, the vertebral canal grows faster than the neural tube, >“ascent” of the spinal cord.
-At the time of birth, the conus medullaris has reached the L2–3 space, reach the adult level by age 3 months.

Associated Clinical Features of Spinal Dysraphism

Cutaneous Stigmata	Neurological Orthopedic Changes	Vertebral Anomalies	Anorectal Anomalies
In approximately 50% -Midline lumbosacral -Cutaneous hemangioma -Lumbosacral hypertrichosis, -Dermal sinus -Midline lumbosacral -Subcutaneous lipoma -Lumbosacral skin appendage	-Progressive radicular pain -Progressive weakness -Asymmetric hyporeflexia -Spasticity -Sensory changes. -Bowel/bladder dysfunction occur in 75% of patients with TCS.	-Bifid vertebrae -Laminar defects -Hemivertebrae, -Sacral aplasia, -Sacral agenesis and -Multiple segmentation errors.	-Cloacal exstrophy. -Imperforate anus -Anal atresia -Tracheoesophageal fistula -Radial limb -Renal dysplasia -Bladder exstrophy.

Lipomyelomeningocele/Spinal Lipoma/Fatty Filum:
a subcutaneous lipoma within the spinal cord that extends through a defect of the lumbosacral fascia, lamina, dura, and pia into a low-lying spinal cord
-70% of the lesions associated with tethering

1-intradural lipoma : intramedullary lesion usually within the thoracic spinal cord. It is not associated with cutaneous or bone anomalies and often presents with symptoms of spinal cord compression.
2-The fatty filum: involves fatty infiltration the whole length or part of the terminal filum. The fat within the short, thick filum is discernible by unenhanced CT or MR imaging
3-Lipomyelomeningoceles: diagnosed by the associated subcutaneous lumbosacral mass that is found in approximately 90% of patients

-59% asymptomatic
-41% symptomatic (60% urological symptoms , 58% with neurological symptoms, 58% orthopedic abnormalities)
-For motor deficit 50 to 70% do not improvement With surgical intervention.
-After surgery: 39% improved, 58% stabilized, and 3% worsened.

-Early prophylactic untethering in patients with asymptomatic lipoma of the conus medullaris is recommended because of the low rate of neurological worsening (3–4%) resulting from the operation and because of the better neurological outcome at follow up of the asymptomatic patients as compared with symptomatic patients.

Diastematomyelia,
Splitting of the spinal cord, conus medullaris, or terminal filum in the sagittal plane into two not necessarily equal halves.
Type 2: split cord residing in a common dural tube
Type 1: split cord divided by a bone spur with each hemicord residing in a separate dural tube

A thick cutaneous hairy patch usually overlies the region of the diastematomyelia.
Diastematomyelia may account for up to 25% of OSD cases.
Bone anomalies are present in 85% of cases, and scoliosis is present in 50% of cases of diastematomyelia

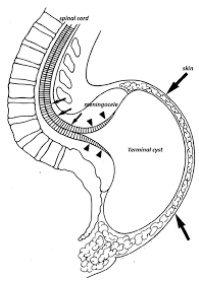
Surgical intervention may involve resecting the median septum and dividing a thickened filum and dorsal tethering bands.
surgical intervention may:
-Stabilize progressive neurological and urological symptoms
-Prevent the onset of neurological and urological deterioration
-Not affect the emergence of the neuroorthopedic syndrome of limb-length asymmetry and foot deformities.

Dermal sinus:
Tracts appear as midline dimples in the lumbosacral region and may extend from the skin surface to the dura, subarachnoid space or the spinal cord ,thereby causing tethering. pathway for an infection, meningitis

Tight Filum Terminale Syndrome
TCS in a patient with a low-lying conus medullaris, a terminal filum greater than 2 mm in diameter and no other tethering agents.
In 86% of the patients, the tip of the conus medullaris lies inferior to L-2. respond to surgery

Terminal Myelocystoceles
expansion of the central canal of the caudal spinal cord by a CSF containing terminal cyst, which itself is surrounded by an expanded dural sheath. The inner terminal cyst communicates with the central canal of the spinal cord, whereas the outer dural sac communicates with subarachnoid space. The outer and inner fluid spaces usually do not communicate.
Tethering results from the attachment of the myelocystocele to the inferior aspect of the spinal cord.

Associated with multiple congenital defects including: cloacal exstrophy, imperforate anus, omphalocele, pelvic deformity, talipes equinovarus, renal abnormalities, and ambiguous genitalia



Neurosurgical intervention involves:
-Separating the spinal cord from the fluid-filled terminal myelocystocele,
-Reconstructing the neural tube,
-incising and reapproximating the dura to recreate the subarachnoid space

Patients usually have no bowel or bladder control and possess poor lower-extremity function

Neurenteric Cysts
congenital malformations lined by alimentary tract mucosa, formed by entrapment of endodermal tissue between a split notochord.
-The cyst may be extraspinal with mediastinal or abdominal extension, or it may be intramedullary.
- frequently associated with anterior or posterior spina bifida, it may be found without any associated dysraphic anomalies.
- present with progressive signs of spinal cord compression that may be acute.
-Neurosurgical intervention involves gross-total resection or partial excision of the cyst

Meningocele Manqué
Dysraphic element of dorsal tethering bands composed of fibrotic or atretic neural tissue connecting the spinal cord to dura or surrounding structures
-usually found incidentally during surgical exploration for other elements of OSD and may exist distant from the site of obvious tethering. These tethering bands are usually found at the site of diastematomyelia; surgical treatment involves the lysis of the bands

Tethered Cord Syndrome and the Conus in a Normal Position
A low-lying conus medullaris below the L1–2 disc space , below the inferior aspect of the L-2 vertebral body
Tethered cord syndrome may occur in the presence of a conus in the normal position with its associated clinical features, urologic dysfunction was not the only abnormality

Tethered Cord Syndrome In Adults
Precipitating events preceding symptoms in approximately 60% of the adult patients.
factors include: heavy lifting, traumatic injury, and the lithotomy position.
Tethered cord syndrome in the adult population is similar to that in the pediatric population with respect to the incidence of

cutaneous stigmata of OSD, neurological abnormalities at presentation, vertebral and orthopedic anomalies, and specific dysraphic elements. In the adult population, however, TCS is accompanied by nondermatomal low-back and leg pain in 50 to 78% of patients. improve bowel and bladder dysfunction in 38% of patients

The Normal Conus and the Hyperreflexic Neurogenic Bladder

Development of a neurogenic hyperreflexic bladder from a tethered spinal cord with a conus in a normal position and an otherwise normal status is a diagnosis of exclusion and has been postulated.
Surgerical management has been performed with varying results,
Different results have led controversy regarding sectioning of the filum

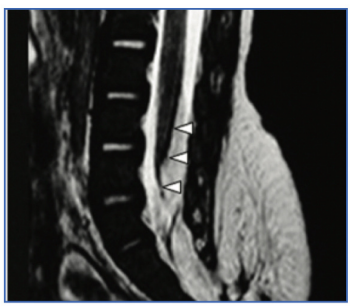
New classification of spinal lipomas based on embryonic stage

Primary neurulation (17–18): > Fusion of the neural folds > separation of the cutaneous ectoderm from the neuroectoderm (dysjunction)

Secondary neurulation (26–27): condensation and formation of the caudal eminence > cavitation of the caudal eminence and connection to the closed primary neural tube (spinal cord)> fusion of the hollow secondary neural tube and the primary neural tube> regression of the secondary neural tube to the filum terminale

Anorectal and Urogenital Organs develop simultaneously with secondary neurulation.

Type 1: Primary Neurulation Failure



-Associated anomalies : none

-Presentation: neonatal period

-Skin stigmata: Present

-Spina bifida: Pathalogical

-fascia, dural defect

-Location: dorsolateral to spinal cord

-Resection: moderately difficult, but radical resection is possible,fusion line exists

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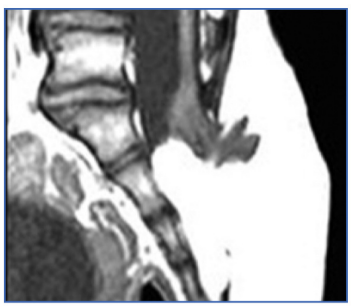
Type 2: Failed Neurulation Between the Primary and Secondary Stages



-Associated anomalies: Present

-Presentation: neonatal period

-Skin stigmata: Present

-Spina bifida: Pathalogical

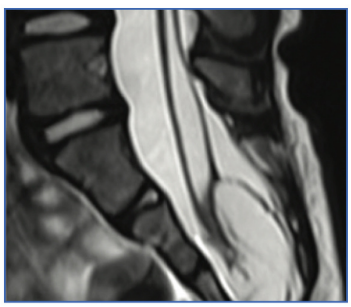
-fascia, dural defect

-malformed conus medullaris

-Location: dorsolateral to spinal cord with neuronal element

-Resection: difficult, radical resection impossible

Type 3: Early Phase Secondary Neurulation Failure



-Associated anomalies: frequent

-little or no skin stigmata.

-Presentation:late infancy or school age

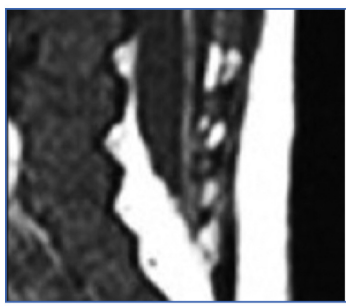
-Spina bifida: physiological

-No dural defect

-Location: attaches to conus medullaris distal to the caudalmost nerve roots.

-Resection: not difficult, but the adhesion of nerve roots to the surface of the lipoma can poses a challenge. radical resection not recommended

Type 4: Late Phase Secondary Neurulation Failure



-Associated anomalies: none

-Skin stigmata: dimple

-Presentation: infancy

-Spina bifida: Physiological

-Location: filum

-Resection: straightforward