Pediatric Tethered cord Syndrome
Izabela Tarasiewicz MD. FRCS(C)
Pediatric Neurosurgery

Disclosure
• None
• Izabela, Tarasiewicz, MD, FRCS(C), has no relationships with commercial companies to disclose.

Overview
• Definition
• Conus level
• Embryology
• Classification: subgroups – nat hx
• Examples
• Multidisciplinary approach

Learning Objectives
At the end of this presentation the participant will be able to:
1. Understand TCS
2. Recognize signs and symptoms to look for
3. Start appropriate work up and referral
4. Understand importance of multidisciplinary approach
5. Provide appropriate f/u

Definition
• Any pathology causing excessive tension on spinal cord
• Wide variety of pathologies
• Dx at birth: skin stigmata
• Dx later due to neurological orthopedic urological gu/gi

Hoffman 1976
Hoffman HJ, Hendrick EB, Humphreys RP
The tethered spinal cord: its protean manifestations, diagnosis and surgical correction.
Childs Brain 2: 145-155, 1976
Conus level:

- L-1/L-2


Inferior margin of L-2:

James M. Drake FRCS(C): Surgical management of tethered spinal cord - walking the fine line. Neurosurg Focus 23 (2): E8, 2007

- Low-lying conus medullaris: its tip is located below the L-2 vertebral level

---

The conus medullaris time of ascension to normal level

A. Malek, S. Schneidler, J. Slinger

Department of Pediatric Radiology, Children's Hospital, University of Illinois, 1989

Abstract. This study was intended to determine the level of the conus medullaris in normal babies. We examined 154 healthy infants using high-resolution scans which identified the spinal cord and the tip of the conus medullaris. This method provided a good analysis of the level of the conus medullaris so that we could ascertain the rate of ascent to L-3. The range of the conus level for all children was at TH 13/L-1 in 39.78% of babies aged between the 30th and 36th postmenstrual week, the tip of the conus medullaris between L-2 and L-3 in 40.41% of babies aged between the 40th and 46th postmenstrual week. In one child aged 15 weeks the tip was found at L-4. Ultrasound is a reliable method to observe the development of the conus level in young infants and to identify a tethered cord.

---

embryology
Failure of Primary Neurulation

- Non-disjunction: open NTD, mm
- Premature disjunction: lipomm
- Focal disjunction failure: dermal sinus tract

Secondary neurulation

pathophysiology

Tight fixation of spinal cord induces oxidative metabolic changes

<table>
<thead>
<tr>
<th>TABLE 206-1: Spinal Dysraphic States</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>LESION</strong></td>
</tr>
<tr>
<td>----------------</td>
</tr>
<tr>
<td>Spina bifida occulta</td>
</tr>
<tr>
<td>Lipoma</td>
</tr>
<tr>
<td>Spina bifida epidermolysis bullosa</td>
</tr>
<tr>
<td>Myelocele</td>
</tr>
</tbody>
</table>

Youmans Neurological Surgery 5th edition ch 206 p3258
symptoms

- Wide variety of pathologies
- Signs and symptoms caused by excessive tension on spinal cord
- Neurological dysfunction:
  - LE motor deficits, spasticity
  - Sensory deficits
- Orthopedic deformities:
  - Contractures
  - Scoliosis
- GU/GI: Urinary dysfunction/sphincter dysfunction

Urinary dysfunction

- Incontinence in gral population: 10%
- Associated with TCS... up to 75% dysfct
- Most common finding: detrusor hyperreflexia
- Importance pre and postop urodynamics
- Formal urodynamics testing recommended in all TCS patients

classification

<table>
<thead>
<tr>
<th>Adult</th>
<th>Pediatric</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at presentation</td>
<td>Depending on underlying dx</td>
</tr>
<tr>
<td>Most frequent presentation</td>
<td>Back pain &amp;/or precipitating event, scoliosis/sphincter dysfunction</td>
</tr>
<tr>
<td>Associated syndromes</td>
<td>Retethering if known dysraphism</td>
</tr>
<tr>
<td>Cutaneous stigmata</td>
<td>If Yes; missed in most cases</td>
</tr>
<tr>
<td>Precipitating event(s)</td>
<td>Yes in most cases</td>
</tr>
</tbody>
</table>

subgroups based on the origin of tethering
1. Postrepair mm
2. Terminal filum lipoma & tight terminal filum
3. Lipomm & conus lipoma
4. SCM
Nat hx: mm

Clinical Signs and Sx of Tethering
- 40% progressive scoliosis
- 47% LE strength decline
- 30% LE contractures
- 25% gait changes
- 17% back pain
- UNIQUE FEATURE: teratoma

Mm outcome Bowman & al

- 70-75% improved gait, LE M strength
- 78% contractures stable
- 64% urological improvement vs 36% stable
- 96% improved or stable scoliosis
- Complications: overall risk of LE motor decline is 3.5%

Group 2. Terminal Filum Lipoma & Tight Terminal Filum

- OSD = skin covered
- Low lying conus +/- fatty filum or thick filum
- Fatty filum true incidence unknown
- 3.7% to 5% of population found on autopsy
- 1.5-5% in MRI imaging studies (Finn-Walker) (Warder)
- 5-7% of adult worked up for back pain: fatty filum on imaging
- Incidence of 0.24% in ax pts (MRI in 1691 patients Japan) (Uchino & al)
- A 2006 study reporting the prevalence of occult tethered cord syndrome in primary school in Turkey, estimated it at 0.1% in primary school children.
Nat hx

- Asx
- Skin stigmata:
  - Dimple
  - Deviated gluteal fold
  - Hemangioma
  - discoloration

International survey on the management of skin stigmata and suspected tethered cord.

Froggatt P, Ben Sira L, Ben-Adar I, Sternbock E, Constantini S
Sacral dimples: 44% recommended imaging
Deviated Gluteal Folds: 75%
Hemangiomas: 92%
discoloration + non-symptom dimple: 30%
Lumbar sinus tract: 38%
Filar cyst: 84%

The value of sacral skin lesions in predicting occult spinal dysraphism in children with voiding dysfunction and normal neurological examination.

- 31% of patients w voiding dysfct & N neuro exam: skin lesion with OSD
Group 3. Lipomm / conus lipoma

- those lesions occur in approximately 1 in 4000 births in the US. There is female prevalence.
- They are mainly diagnosed in childhood due to presence of cutaneous features (50% associated with lipomm) or non tender fatty mass, as in 90% of the cases.
Occult Spinal Dysraphism + Low Conus

A Type I SCM consists of two hemicords, each contained within its own dural tube and separated by a dura-sheathed rigid osseocartilaginous median septum.

A Type II SCM consists of two hemicords housed in a single dural tube separated by a nonrigid, fibrous median septum.

• 50% sx or will become sx
• 50% will stay asx

Group 4. Diastematomyelia/ scm

• 48% asx at presentation
• Most common manifestation: subcut mass, skin dimples, hemangioma, hair patch, skin tags (50-90%) [image]
• Neuro deterioration very common (70% progression within 2-24 months...67 months)
• most authors advocate detethering at time of presentation

Tethered cord syndrome in occult spinal dysraphism: Timing and outcome of surgical release
Cornette, L. MD; Verpoorten, C. MD; Lagae, L. MD, PhD; Van Calenbergh, F. MD; Piets, C. MD, PhD; Versweyen, R. MD, PhD; Casaer, P. MD, PhD.
Neurology
Volume 50(6), June 1998, pp 1761-1765

50% sx or will become sx
50% will stay asx

Tethered cord syndrome: a review of the literature from embryology to adult presentation

• 48% asx at presentation
• Most common manifestation: subcut mass, skin dimples, hemangioma, hair patch, skin tags (50-90%) [image]
• Neuro deterioration very common (70% progression within 2-24 months...67 months)
• most authors advocate detethering at time of presentation
SCM: 75% occult urological abnormalities
6% of patients with mm also have SCM
Neurocutaneous stigmata in 50% (hair 50-60%)
2 patients had previous “cosmetic” surgery without untethering.
Presented later in life with back pain
Asymmetrical limbs

Authors quote Pang: all pts with SCM harbor a 2nd lesion
Other authors: 50-85%
Tandem lesions: 69%

Most common 2nd lesion:
fatty filum (6/16)
lipoma (4/16)
Chiari with syrinx (1/16)

15/16 pts improved/stabilized neuro fct postop (mainly pain relief)
Retethering in SCM is significantly lower than in LipoMM
Scoliosis: >40 degrees: 86% will require spinal stabilization after untethering
Changes in urinary tract: 75%, that stabilized after untethering
outcomes

Surgical management of tethered cord syndrome in adults: indications, techniques, and long-term outcomes in 60 patients

- Pts with tight filum & SCM: prognosis of surgery is excellent, no worsening of condition on most recent f/u
- Pts with previous mm repair & pts with conus lipoma or adhesions had a 9-50% chance of worsening pain & sensorimotor deficits on last f/u

Tethered cord

- Not that rare
- Look for it
- Skin findings
- Asymmetry
- Dimple
- Discoloration/hemangio ma
- masses

Multidisciplinary approach

- Neurosurgery
- Physiatry
- Urology
- Ortho
- Gi/gen surgery

Work up

- Lumbar u/s
- Great screening tool
- MRI
- Definitive imaging

Pediatr Radiol July 2014

The simple sacral dimple: diagnostic yield of ultrasound in neonates

Jennifer N. Kucera & Ian Coley & Sara O’Hara & Edward J. Kosnik & Brian D. Coley

3991 children 3.4% had abn us, after MRI was obtained 2.1% incidence of surgical detethering in N with simple sacral dimple 0.13%