TETHERED CORD SYNDROME in EHLERS-DANLOS –
Diagnostic criteria, Surgical Indication and Filum pathology

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Outline

- What is “tethered cord” – the classic concepts
- Tethered cord in EDS – introduction of the “occult tethered cord syndrome”
- Translation of the occult tethered cord syndrome in EDS – presentation and surgical indication
- Surgical technique and surgical outcome
- Filum HISTOpathology in EDS - cause of neurological manifestations in EDS
What is tethered cord?

An embryological problem of the differentiation of the spinal cord leading to fixation of the cord to the Mesoderm (=Fibrous tissue, Muscle, Fat and Bone)

- Around 6 weeks of gestation:
  Caudal extension of the spinal cord and “more” neural tube formation
  _“Secondary neurulation”_

- Around 9 to 10 weeks of gestation:
  Cell necrosis causes a decrease in the size of the caudal neural tube and will form the Filum Terminale _Retrogressive differentiation_
If all goes well we have a filum terminale…

...a fibrous, collagenous band that “connects” the CONUS with the DURAL SAC
Variants and spectrum of tethered cord!

- Fatty filum
- Filum lipoma
- Intradural lipoma
- Intra-extradural lipoma
- Lipomeningocele
- Lipomyelomeningocele
- Lipomyelomeningocystocele
- Dermal sinus

☞ AKA Spina bifida occulta!
Diagnosis of “Spina bifida occulta”

- CUTANEUS SIGNS
- IMAGING (MRI)
- SYMPTOMS
Imaging of tethered cord:

*Gold standard:* LUMBAR MRI

? Level of the conus:
T2 sagittal and axial MRI

? Pathology of Filum:
T1 sagittal and axial

WHAT is NORMAL?

< 6 months: Level L2-L3
> 6 months: Level Th12/L2

No FAT SIGNAL at and below the conus!
What is not normal on lumbar MRI?

- LOW LYING CONUS (Endplate L2 or L2/3 = *borderline low*)
- THICKENED FILUM > 2mm
- FAT in FILUM
**THE Symptoms TRIAD**

**SUBJECTIVE**: Back pain and leg pain(s) and complain(s):
- “aches” and “burns” *not sharp*
- “heavy”
- “stiff and tight”
- “traveling” *no radiating*

1. **NEURO**: LE signs
   - Infants: Decreased leg movement, absent reflexes
   - Toddler: Abnormal gait or delayed walking (“intoeing”, “loss of ankle control”)
   - Teenager: Pain, Asymmetrical motor dysfunction
   - Young adults: Pain, Spasticity and Hyperreflexia

2. **UROLOGY/GI**: Neurogenic bladder*, Frequent Urinary tract infection (> 3 per year), Incontinence, Constipation

3. **ORTHO**: Foot and leg deformities and asymmetry, Scoliosis, delayed or plateau in growth
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“Occult tethered cord”

Symptoms consistent with tethered cord (as reviewed) and exclusion of other conditions and co-morbidities

No imaging evidence of fatty or thickened filum and conus at “normal” position

Essentially, a normal standard MRI
## History of occult tethered cord

### Occult tethered cord syndrome: a review.

**Tu A**, Steinbok P.

**Table 1: Details of published series of OTCS**

<table>
<thead>
<tr>
<th>Author</th>
<th>Duration of follow up (months)</th>
<th>Number of patients in study</th>
<th>Number with urologic dysfunction</th>
<th>Number with improved urologic symptoms post op</th>
<th>Number with worse symptoms post op</th>
</tr>
</thead>
<tbody>
<tr>
<td>Khoury et al. (1990)</td>
<td>Mean 13.3</td>
<td>31</td>
<td>31</td>
<td>22</td>
<td>0</td>
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<tr>
<td>Warder and Oakes (1993)</td>
<td>Range 6–72</td>
<td>13</td>
<td>13</td>
<td>4</td>
<td>0</td>
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<tr>
<td>Nazar et al. (1995)</td>
<td>Range 2–48</td>
<td>32</td>
<td>32</td>
<td>31</td>
<td>0</td>
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<td>Selucky et al. (2000)</td>
<td>Mean 18</td>
<td>17</td>
<td>17</td>
<td>3</td>
<td>4</td>
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<tr>
<td>Selucky et al. (2003)</td>
<td>Range 3–84, mean 31.7</td>
<td>8</td>
<td>8</td>
<td>7</td>
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<tr>
<td>Komagata et al. (2004)</td>
<td>Not reported</td>
<td>37</td>
<td>33</td>
<td>26</td>
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<tr>
<td>Webb et al. (2004)</td>
<td>Mean 13.9</td>
<td>60</td>
<td>60</td>
<td>58</td>
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<td>Metcalfe et al. (2006)</td>
<td>Mean 49</td>
<td>36</td>
<td>36</td>
<td>26</td>
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<tr>
<td>Selden et al. (2006)</td>
<td>Range 3–30, mean 15.6</td>
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<td>Steinbok et al. (2007)</td>
<td>Range 12–120, mean 37.2</td>
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<tr>
<td>Fabiano et al. (2009)</td>
<td>Mean 16</td>
<td>22</td>
<td>22</td>
<td>16</td>
<td>2</td>
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<tr>
<td>Fukui et al. (2011)</td>
<td>Mean of over 240 months</td>
<td>10</td>
<td>10</td>
<td>9</td>
<td>1</td>
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<td>Comips et al. (2012)</td>
<td>Mean 30.4</td>
<td>9</td>
<td>5</td>
<td>5</td>
<td>0</td>
</tr>
</tbody>
</table>

**Childs Nerv Syst.** 2013 Sep;29(9):1635-40. Occult tethered cord syndrome: a review. **Tu A**, Steinbok P.
Occult tight filum terminale syndrome: results of surgical untethering.

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The entity of an occult tight filum terminale syndrome, characterized by clinical findings consistent with a tethered cord syndrome, but with the conus ending in a normal position and Filum > 2mm but no fatty infiltration.

N= 60 children (ages 3-18 years) followed for more than 6 months (mean 13.9 months).

The criteria for surgical intervention were (1) Bifid lumbar spine (2) progressive bladder instability unresponsive to conservative measures, (3) urological/nephrological evaluation to confirm neurogenic etiology, and (4) two or more of the following: (a) bowel involvement (fecal incontinence or chronic constipation), (b) lower extremity weakness, (c) gait changes, (d) reflex/tone abnormalities, (e) sensory disturbances, (f) back/leg pain, (g) orthopedic abnormalities/limb length discrepancy, (h) scoliosis/lordosis, (i) recurrent urinary tract infections, (j) abnormal voiding cystourethrogram/ultrasound, (k) syringomyelia, and (l) neurocutaneous stigmata.

Postoperatively, urinary incontinence/retention showed complete resolution in 52%, marked improvement (>95% resolution) in 35%, moderate improvement (>75%) in 6%, minimal improvement (> 50%) in 6%, and no improvement (<50%) in 2%. Fecal incontinence completely resolved in 56%, improved in 41%, and was unchanged in 3%. Weakness, sensory abnormalities, and pain improved or resolved in all patients.
The occult tethered cord syndrome and surgical indication - PEDs

¾ clinical qualifiers mandatory for surgical indication!

Less qualifiers are acceptable with “indirect” imaging findings

“Progressive” Syringohydromelia

Filum > 2mm in axial section
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The occult tethered cord syndrome - EDS

Muscular symptoms?  
Peripheral Neuropathy?  
**M. Castori, N. Voermans: Neurological manifestations of Ehlers Danlos syndrome(s): A review. Iran J Neurol 2014**

Increase in hip or other LE joint subluxation?

Pevlic weakness and instability?

Predictor for occult tethered cord in EDS?

"Progressive" Syringohydromelia

FIlum > 2mm in axial section
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Case presentations:

# 1 M, 5yo
- Inconsolable from pain
- Progressive weakness in ankles and knees (upgrade in braces)
- Progressive Urinary leakage
- Urodynamic studies under Anesthesia “grossly normal”

# 2  F, 18 y0
- Progressive “Chiari pain” (3mm Chiari)
- Leg “aches” and weakness
- Progressive scoliosis
- Reduced bladder capacity and low compliance (Urodynamic studies)
#1 MRI

L2 – 3 Interlaminectomy
#1 intraop findings
#2 MRI

NORMAL T2 MRI conus position

NO T1 MRI axial “fat” signal

L2 – 3 Interlaminectomy
"In connective tissue disorders, tethered cord has little radiographic evidence but shows intraoperative abnormal findings"
Technique

Association of Chiari malformation type I and tethered cord syndrome: preliminary results of sectioning filum terminale‡

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Fig. 3. A: Intraoperative photograph of lumbar lica at L4 in 46-year-old female with CM-4TCS showing semitransparent dura with midline clefts defined by arrowheads. Arrow identifies thin, intact, dorsally positioned FT. B-E: Intraoperative CDS assessments in 3-year-old male with CM-4TCS. B. Axial image at L4 before opening dura showing dorsal position of FT (arrow) and lateral packing of cauda equina roots (asterisks). The FT measures 0.8 mm in transverse diameter. C. Axial image at L4 after SFF and closing dura showing even distribution of cauda equina roots. D. Midcannal image at L4 before opening dura showing 5.1 to 2.0 cm2 CSF flow with arterial pulsations, low power, and high resistance. Laterally packed cauda equina roots are not visualized in midline plane. Arrow identifies stretched arachnoid band. Arrowhead identifies dorsally positioned FT. E. Midcannal image at L4 after SFF and closing dura showing 3.5 to 4.0 cm2 CSF flow in multiple streams with arterial, venous, and respiratory variations, bilateral movement, high power, and reduced resistance.
Filum pathology in EDS

Is translucent, taught, thin and dorsally positioned a requisite in tethered cord syndrome in EDS?
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Histology of an abnormal filum - classical

Pathologically, an abnormal filum appears histologically disorganized and can be composed of adipose tissue, elastic fibres, vascular lacunes, nervous or muscle tissues and/or inflammatory cells.
Histology of an abnormal filum  EDS #1

PMNs at blood vessel
Histology of an abnormal filum – EDS #2

PMNs
Histology of an abnormal filum – EDS #3
Histology of filum in EDS: Proposal!

- EM studies of Collagen
- Trichrom stain **to quantify** Elastin
- CD 68 (Microglia/Macrophages/Leukocytes)
Summary and THANK YOU!

- Need to study the mechanism of tethered cord in EDS?
  - Hypermobility?
  - Filum pathology (PMNs)?

- Need to study prospectively the clinical criteria for occult tethered cord in EDS?