Ehlers-Danlos Organisation e.V.
Wir klären auf!

Die beiliegende Präsentation

"Okkultes Tethered Cord Syndrome"

wurde uns freundlicher Weise von
Frau Petra Klinge, MD, PhD,
Professor of Neurosurgery,
für die Veröffentlichung auf unserer Homepage
zur Verfügung gestellt.

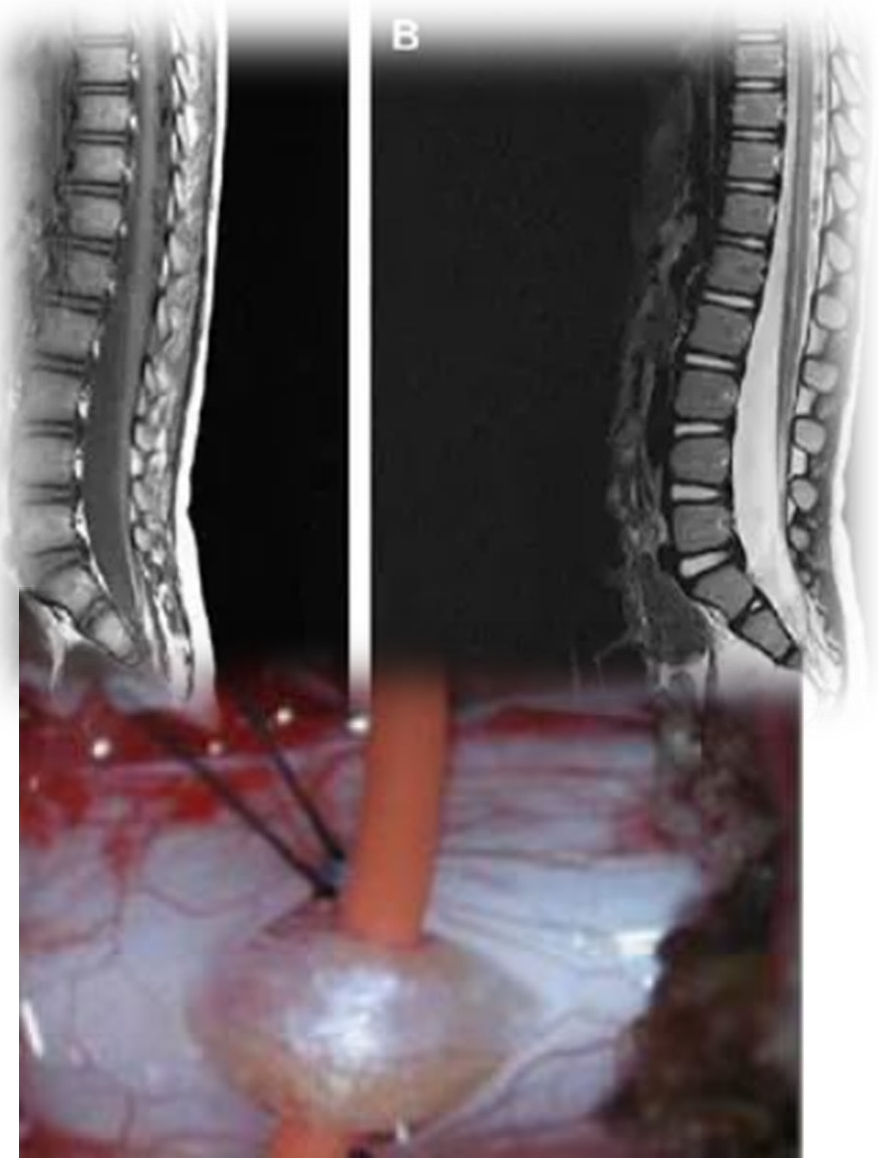
Rhode Island Hospital
BROWN Health
UNIVERSITY

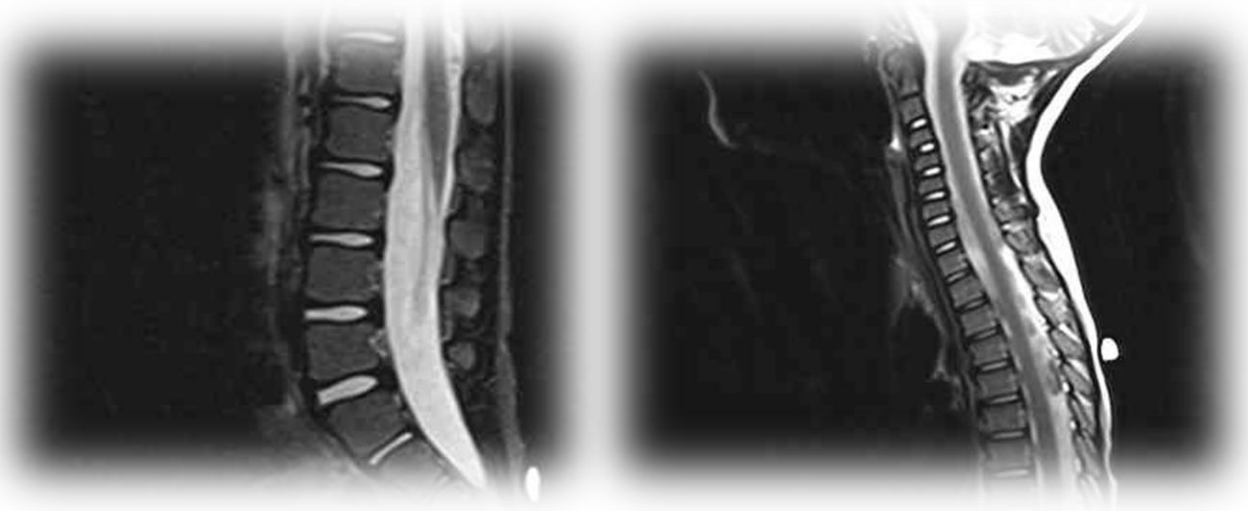
Okkultes
Tethered Cord Syndrome

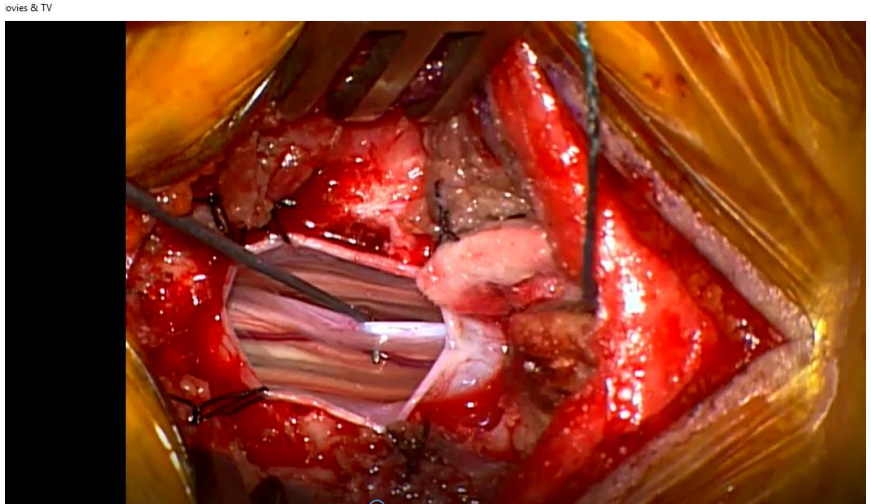
Petra Klinge

Professor fuer Neurochirurgie,
Direktor Paediatrische Neurochirurgie,
Rhode Island Hospital und Hasbro Children's Hospital,
Medizinische Hochschule, Brown University

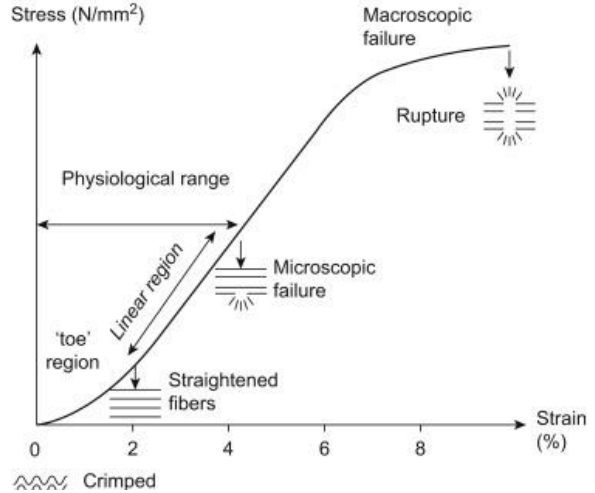
TC vs. *Okkultes* TC



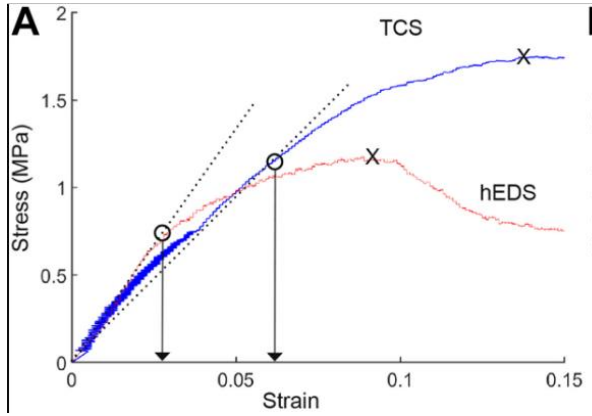


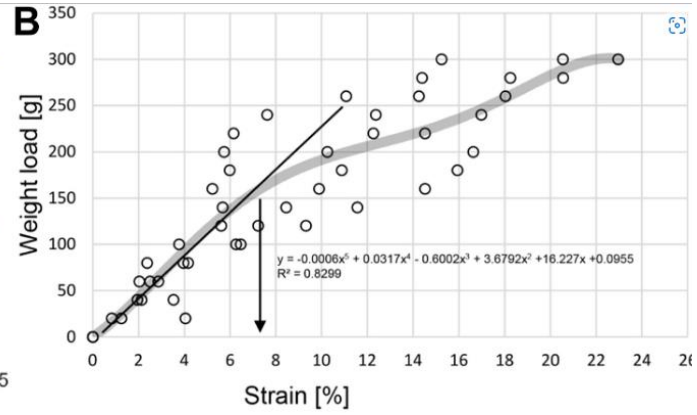


Biomechanik
(TC = okkultes TC)



entspricht chronisch ueberlasteter Muskelsehnen **T*endinopathie***!





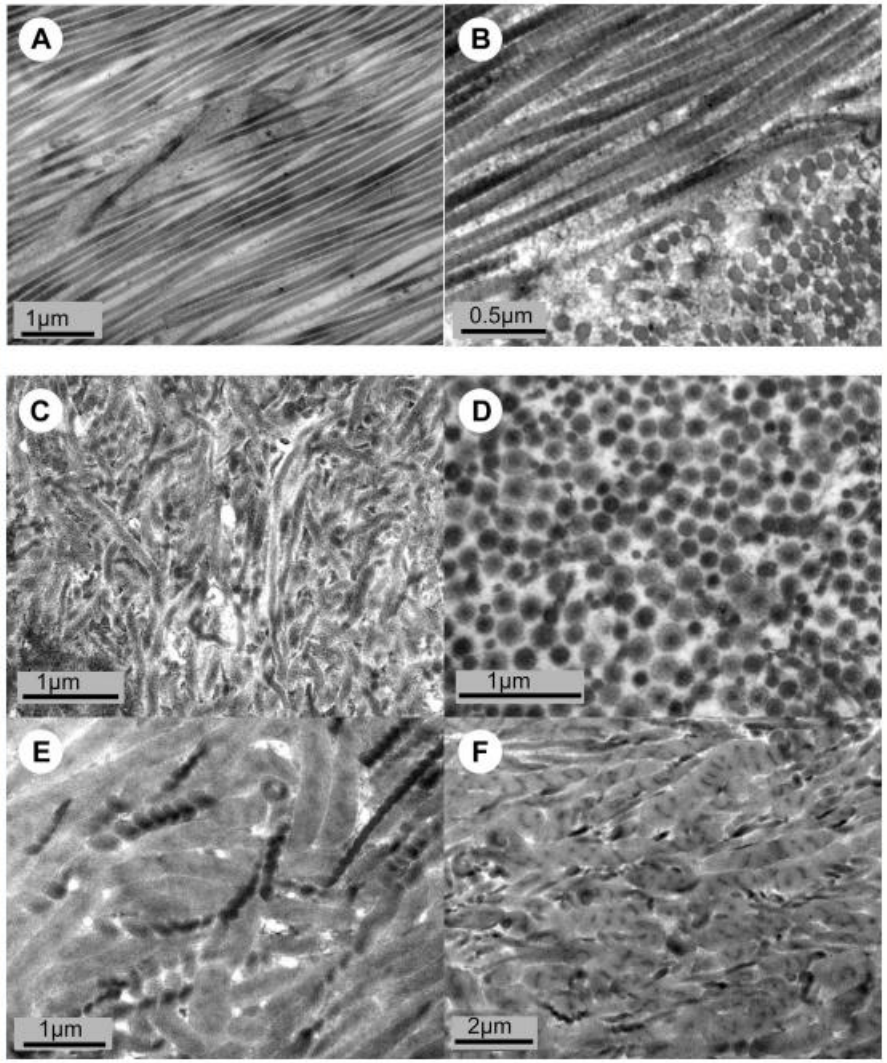


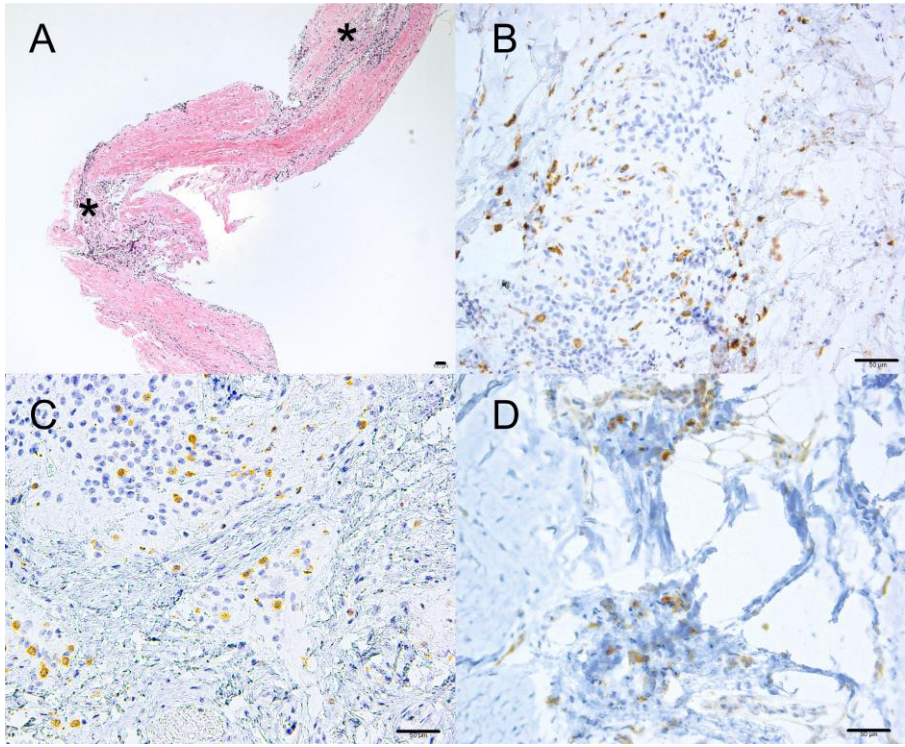
Figure 1. Transmission electron microscopy (TEM) of the filum terminale (FT) specimens without hypermobile Ehlers-Danlos syndrome (hEDS) comorbidity typically showed a regular cross-sectional and longitudinal arrangement of collagen fibrils (**A**). Higher magnification shows the physiologic D-period banding of the fibrils and a uniform fibril diameter, as well as regular distribution of the fibrils (**B**). TEM of hEDS specimens revealed findings consistent with both hereditary and acquired components of FT damage. As compared with part **A**, parts **C, E,** and **F** illustrate the loss of longitudinal alignment and appearance of irregular interwoven fibril structures, which has been recognized as pathology associated with hereditary connective tissue disorder.[17,18] As compared with part (**B**), parts **D, E,** and **F** show acquired damage indicating the mechanical overload of the FT.[19] (**D**) Swollen fibrils with varying diameter often display a halo, most likely indicating denatured collagen. (**E** and **F**) Collagen fibril microdamage in greater detail. The loss of the physiologic D-period banding is apparent in **E**. Dark spots posing as a "string of beads" indicate the kinking of fibrils representing local unwinding of fibrils occurring in response to a mechanical overload.[20,21] (**F**) Fibril swelling with aberrant, "zebra-like" D-period banding.

Table 1. Transmission Electron Microscopy Findings of the Filum Terminale

	hEDS-TCS (n = 78)	TCS (n = 10)	Chi-Square with Bonferroni Correction
Congenital collagen abnormalities			
Fibril disorganization	69.6%	10.0%	$P < 0.001$
Variation in fibril diameter in cross section	40.5%	11.1%	$P < 0.001$
Acquired collagen microdamage			
Kinking fibrils	63.3%	30.0%	$P < 0.05$
Loss of D-period banding	68.4%	30.0%	$P < 0.01$

The filum ultrastructure of the EDS-TCS and typical TCS cases was evaluated in longitudinal and cross sections. Markers of EDS collagen structural abnormalities such as fibril disorganization and variation in fibril diameter in cross section were assessed as well as markers of collagen microdamage such as kinking fibrils and loss of D-period banding.

hEDS, hypermobile Ehlers-Danlos syndrome; TCS, tethered cord syndrome.



Pathologie TC/okkultes TC

(Klinge et al. World Neurosurgery 2022)

Diagnostik TCSyndrom

- ❑ **Fluktuierende Rücken und Sacrale- und nicht-dermatomale dumpfe und ziehende Beinschmerzen und Beinschwäche (“Kraempfe", "Müdigkeit", "Muskelkater", "Steifheit")**
- ❑ **Fuer Patienten schwer zu lokalisieren und schwer zu beschreiben**
- ❑ **NEUROLOGISCHE Befunde in den Beinen oft asymmetrisch und betont im symptomatischeren Bein**
  - ❑ **verstärkter LE-Knöcheltonus**
  - ❑ **Fußklonus**
  - ❑ **Hyperreflexie 4+ und verlängerte Reflexzone**
  - ❑ **Bein-Schwäche 4/5**
- ❑ **=Dysfunktion des oberen Motorneurons, aka Spastizitaet**

Diagnostik *okkultes* TCSyndrom (n=149_60% hEDS)

Klinge et al.

TABLE 2. Prevalence of symptoms on the 15 item-scale

Category or Sx	No. of Patients (%)
Neurological scale	
Increased muscle tone legs*	42 (28)
Hyperreflexia legs*	98 (66)
Foot clonus*	83 (56)
Fatigue legs	127 (85)
Paresthesia legs	84 (56)
Pain scale	
Leg	132 (89)
Sacral	62 (42)
Low-back	119 (80)
Cramps in leg & back	84 (56)
Fluctuating pain Sxs	42 (28)
Bowel & bladder scale	
Urinary leakage	85 (57)
Urinary urgency	87 (58)
Urinary frequency	78 (52)
Urinary hesitation	72 (48)
Bowel Sxs	92 (62)

* Objective clinical findings.

JNS SPINE

CLINICAL ARTICLE

J Neurosurg Spine 40:758–766, 2024

Clinical criteria for filum terminale resection in occult tethered cord syndrome

Petra M. Klinge, MD, PhD,[1] Owen P. Leary, BS,[1] Philip A. Allen, PhD,[2] Konstantina Svokos, DO, MS,[1] Patricia Sullivan, MD,[1] Thomas Brinker, MD, PhD,[3] and Ziya L. Gokaslan, MD[1]

[1]Department of Neurosurgery, Warren Alpert Medical School of Brown University, Rhode Island Hospital, Providence, Rhode Island; [2]Department of Psychology, University of Akron, Ohio; and [3]Department of Neurosurgery, Medical School Hannover, Germany

OBJECTIVE Tethered cord syndrome (TCS) comprises three symptom categories: back/leg pain, bowel/bladder, and neurological complaints. MRI typically reveals a low-lying conus medullaris, filum terminale (FT) pathology, or lumbosacral abnormalities. FT resection is established in TCS but not in radiologically occult TCS (OTCS). This study aims to identify patients with OTCS who are likely to benefit from FT resection.

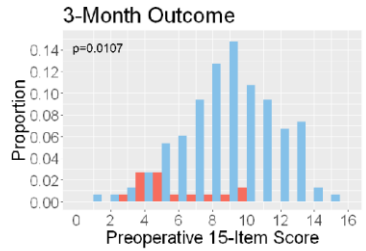
METHODS The authors recruited 149 patients with OTCS (31 pediatric, 118 adult) treated with FT resection—including only cases with progressive TCS, negative spine MRI, and no concurrent neurological/urological conditions. A comprehensive questionnaire collected patient self-reported symptoms and clinical findings at the preoperative and at 3- and 12-month follow-up examinations. Based on questionnaire data, the authors extracted a 15-item symptoms and findings scale to represent the three TCS symptom categories, assigning 1 point for each item present.

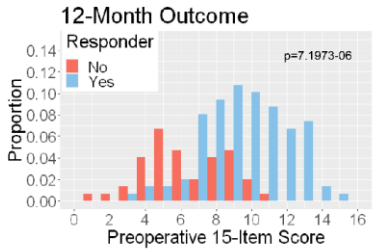
RESULTS OTCS presents without radicular/segmental sensorimotor findings, but with leg/back pain and conus dysfunction, in addition to leg fatigue and spasticity; the latter indicating an upper motoneuron pathology. The 15-item scale showed clinical improvement in 89% of patients at the 3-month follow-up and 68% at the 12-month follow-up. Multivariate analysis of the scale revealed that it accurately predicts outcome of FT resection in 82% of cases. Patients with a preoperative score exceeding 6 points are most likely to benefit from surgery.

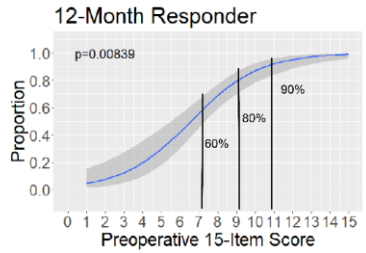
CONCLUSIONS By applying the study's inclusion criteria and incorporating the novel 15-item scale, surgeons can effectively select candidates for FT resection in patients with OTCS. The observed outcomes in these selected patients are comparable to those achieved in degenerative spine surgery.

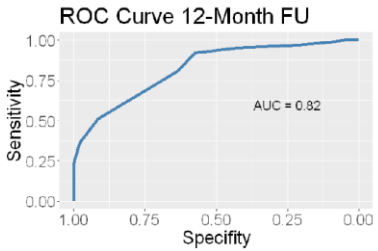
https://thejns.org/doi/abs/10.3171/2024.1.SPINE231191

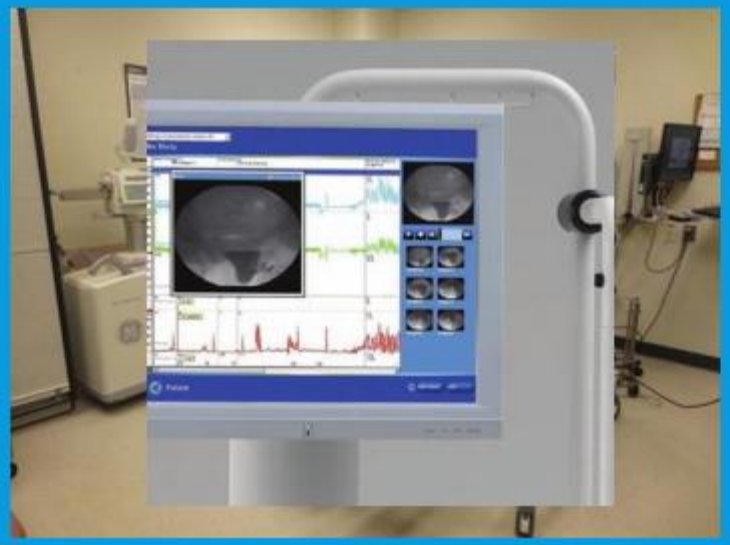
KEYWORDS occult tethered cord syndrome; outcome assessment; symptom scale; congenital; lumbar; sacral; pain

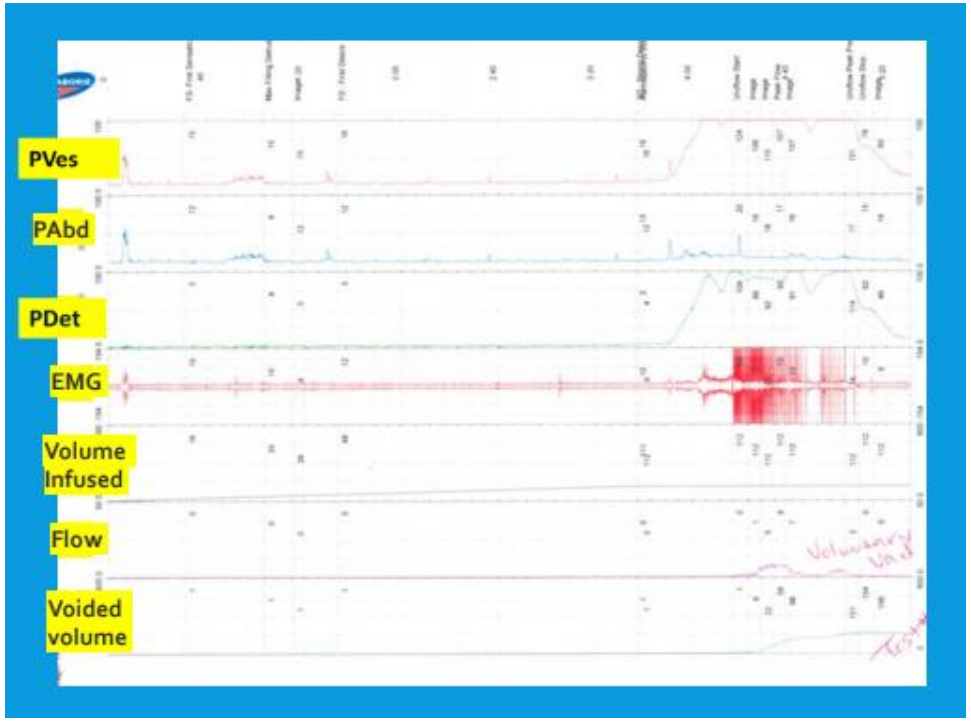










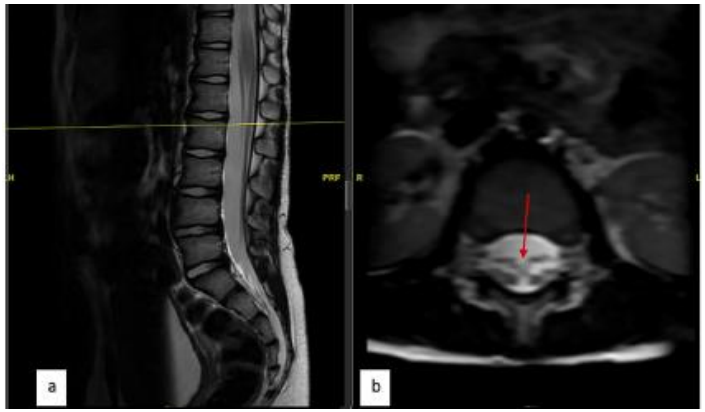


Urodynamik= Konus dysfunktion

N= 116 (78 okkult, 38 klassisch)

- Detrusor-Schließmuskel-Dyssynergie oder Detrusor-Drücke verbunden mit Stressinkontinenz (53 %)
- Erhöhte Blasenkapazität in Verbindung mit unvollständiger Entleerung aufgrund verminderter Empfindung und beeinträchtigter verringerter EMG-Detrusor Kontraktilität =Blasen Unteraktivität (22 %)
- Verringerte Blasenkapazität (16 %)
- Unvollständige Entleerung mit erhöhtem postvoider Restschicht, einschließlich Stressinkontinenz (9 %).

World Neurosurgery, Klinge et al. 2022



HOW I DO IT

Microsurgical approach for resection of the filum terminale internum in tethered cord syndrome—a case demonstration of technical nuances and vignettes

H. Abdulrazeq[1] · B. Shao[1] · R. A. Sastry[1] · P. M. Klinge[1]

Received: 27 February 2023 / Accepted: 19 March 2023

Abstract

Background In tethered cord syndrome due to filum terminale pathology, the surgical approach to achieve detethering of the spinal cord may vary. Traditionally, sectioning the filum through a laminectomy at the lumbosacral level is performed.

Method A microsurgical technique at a higher level to approach the filum below the conus tip is performed. This allows for removal of the entire distal portion of the filum through a limited interlaminar approach and dural opening.

Conclusion We propose a technique to transect the filum terminale below the conus tip and extract the distal filum by releasing it from its intradural attachments to minimize any remnants of the filum terminale.

Keywords Filum terminale pathology · Lumbosacral level · Tethered cord syndrome

Introduction

Traditionally, sectioning the filum through a laminectomy at the lumbosacral level is performed for detethering in patients with tethered cord syndrome (TCS) [5, 9]. Some surgeons would resect a small portion of the filum for histopathological evaluation. The rationale for a lower surgical approach to the filum is to avoid being in the vicinity of the conus medullaris. Furthermore, the increased width of the thecal sac and the loss of cauda equina density in the lumbosacral space facilitate filum isolation and sectioning.

The procedure has been proposed as fairly straightforward with minimal surgical risks; however, the main challenges remain the fairly high rate of retethering of up to 8% in some studies, particularly in a thickened filum, where the residual filum may re-attach to the dura proximal to the surgical site [6, 10, 11].

The authors perform a microsurgical technique at a higher level to approach the filum below the conus tip. This minimizes any residual filum and allows for removal of the entire distal portion of the filum terminale through a limited interlaminar approach and small dural opening. We discuss ways to optimize the surgical technique and limit short- and long-term complications, such as cerebrospinal fluid (CSF) leak, pseudomeningocele, and adhesive arachnoiditis.

Relevant surgical anatomy

The conus medullaris-filum terminale transition zone typically lies at the lumbar 1–2 disc space. The conus medullaris marks the end of the spinal cord proper and the beginning of the cauda equina nerve roots. The filum terminale usually consists of fibrovascular tissue that extends from the conus medullaris and attaches to the end of the thecal sac [1]. The conus-filum interface may be determined on a preoperative magnetic resonance image (MRI) of the lumbar spine which to facilitate allows for surgical planning of the target levels for laminotomy and, therefore, filum resection. Identifying this level will guide the surgical approach and point of resection of the filum (Fig. 1).

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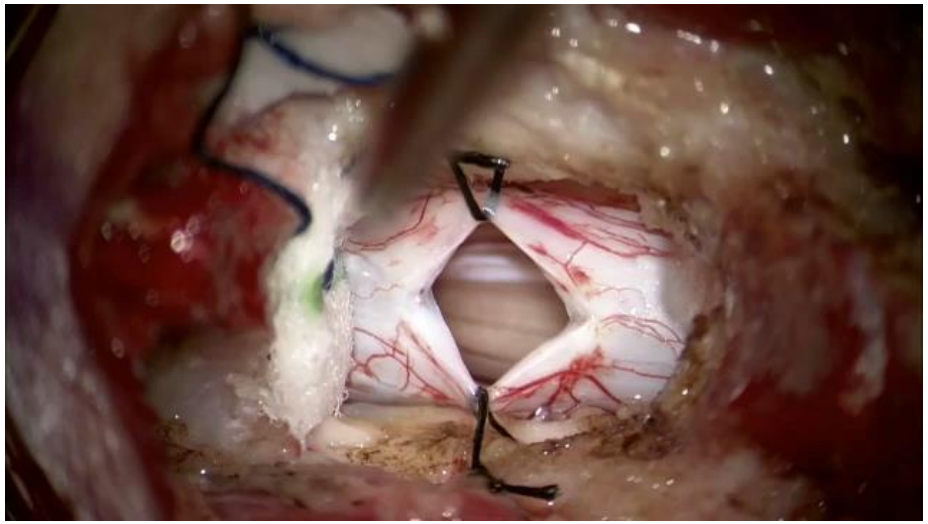


- **OPERATIONSTECHNIK bei TCS und okkultem TCS mit Entfernung des intraduralen Filums**
- **V**erbessert 3 Monats Operationsergebnisse im Vergleich zur herkoemmlichen Filumsektion (n= 606 laufende retrospektive Studie RIH 2015 -2025, *unveroeffentlichte Daten*)

Komplikationen und Operationserfolg

- **Keine wesentlichen Unterschiede zwischen TCS und okkultem TC** (laufende Studie, *unveroeffentlichte Daten*): kumulative Risiken 7 -8%

- Retrospective Studie

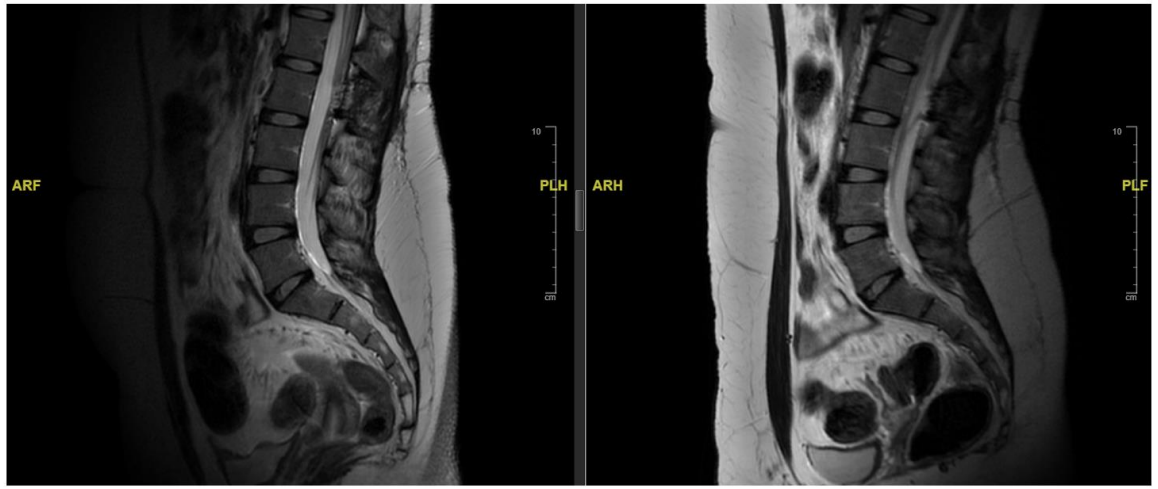
n	
Liquorleck	
Serom	
Subkutane Infektion	
Tiefe Infektion	
Chirurgische Revision bei o.g. Komplicationen	
Chirurgische Revison bei fehlender Wundheilung	

Arachnoidale “Verklebungen” der Cauda equina: 45/561 ~ 8%

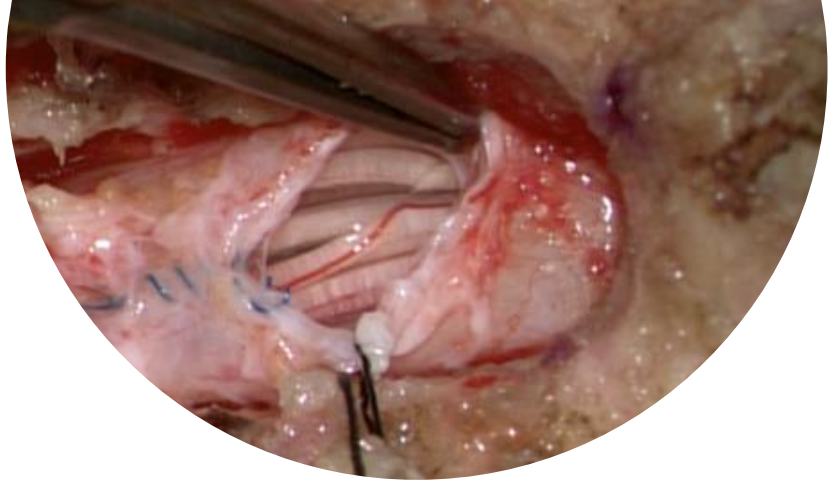
- ✓ *Keine Unterschiede zwischen TCS und okkulten TCS*
- ✓ *Keine erhoehten Befunde bei EDS*
- ✓ ***Laufende Studie_ vorlaeufige und unveroeffentlichte Daten!***

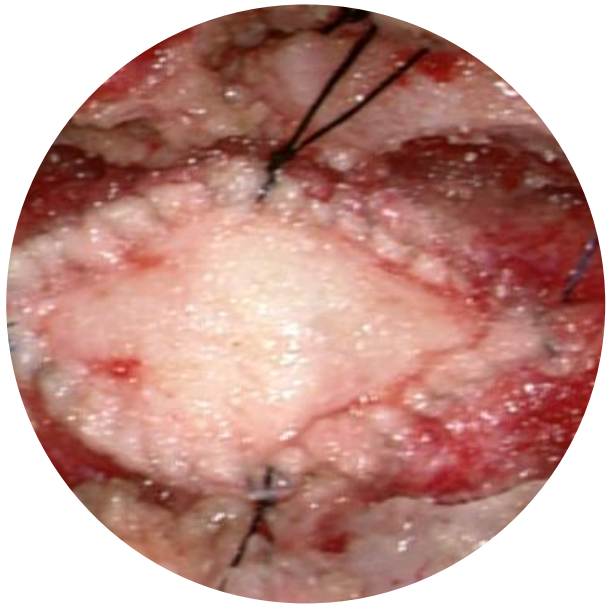
“Symptomatisches Retethering” > 6 Monate (3 bis 5 Jahre) nach OP

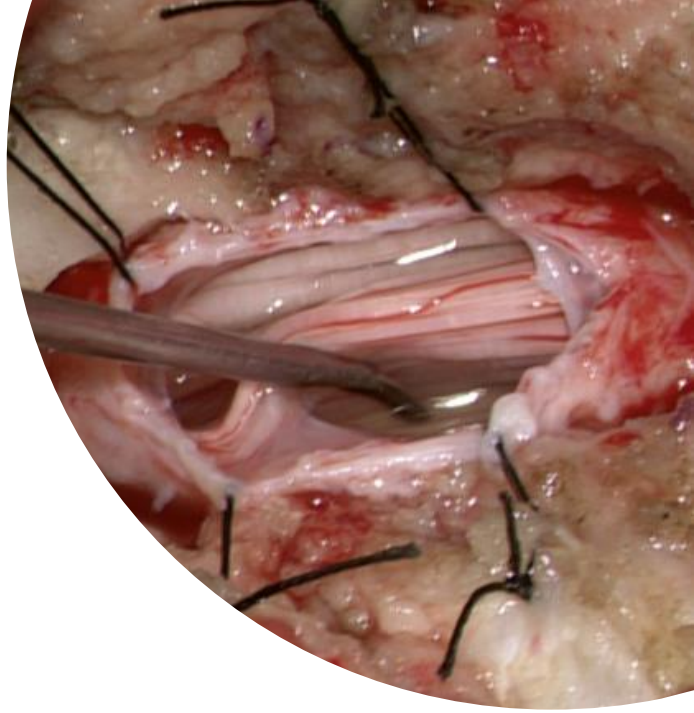
Cauda equina tethering, supine vs. prone MRI, T2 weighted imaging

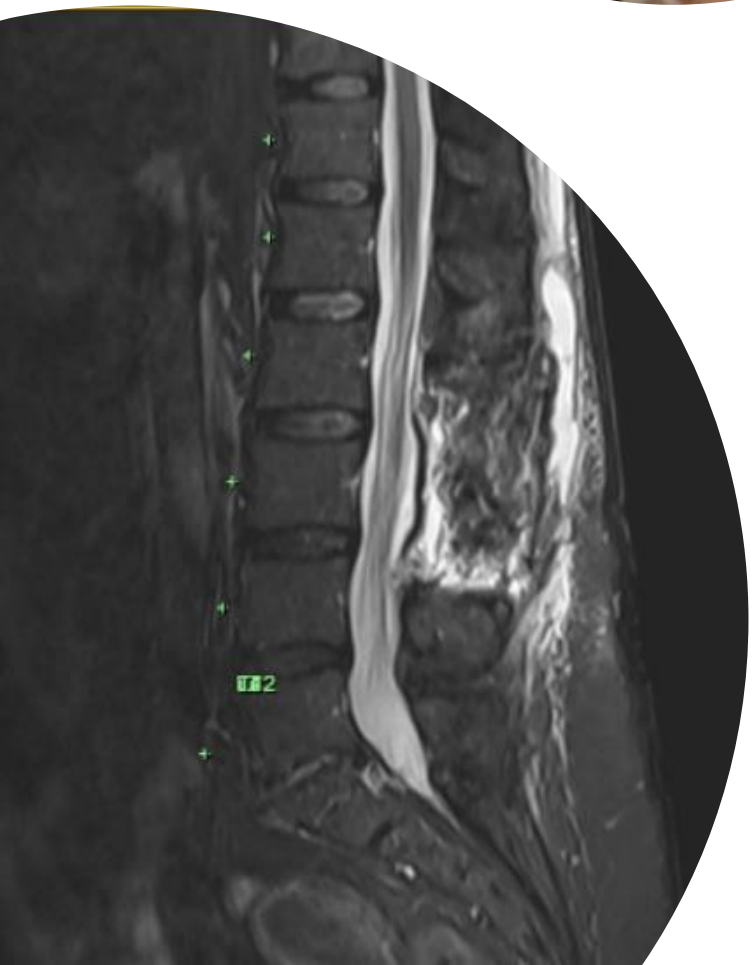


Bauchlagen MRT

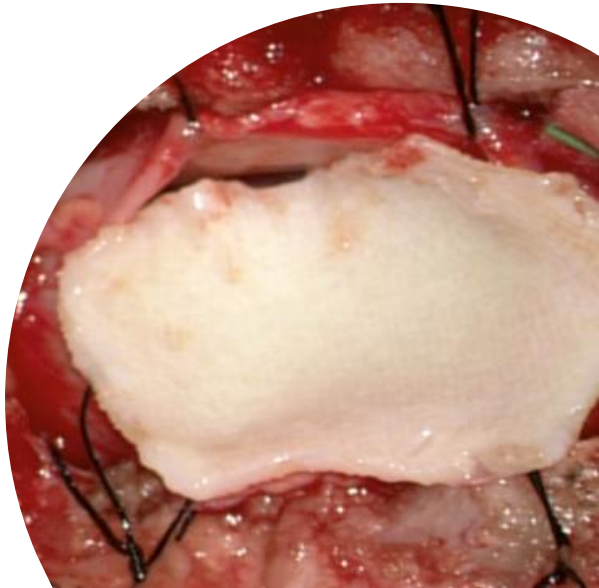


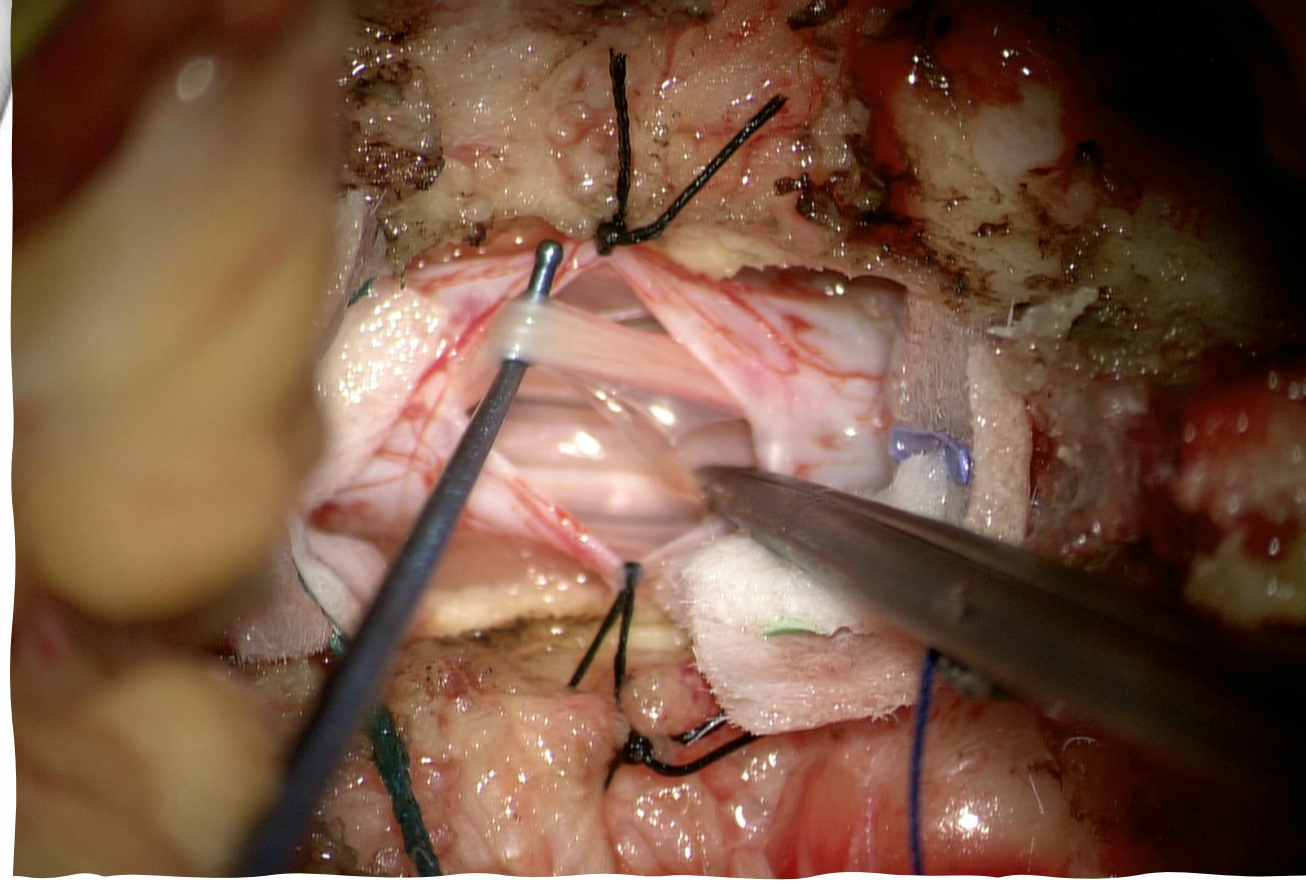






Mikrochirurgische Loesung der Arachnoidalen Verklebungen , Cauda equina *Untethering* und Duraerweiterungsplastik



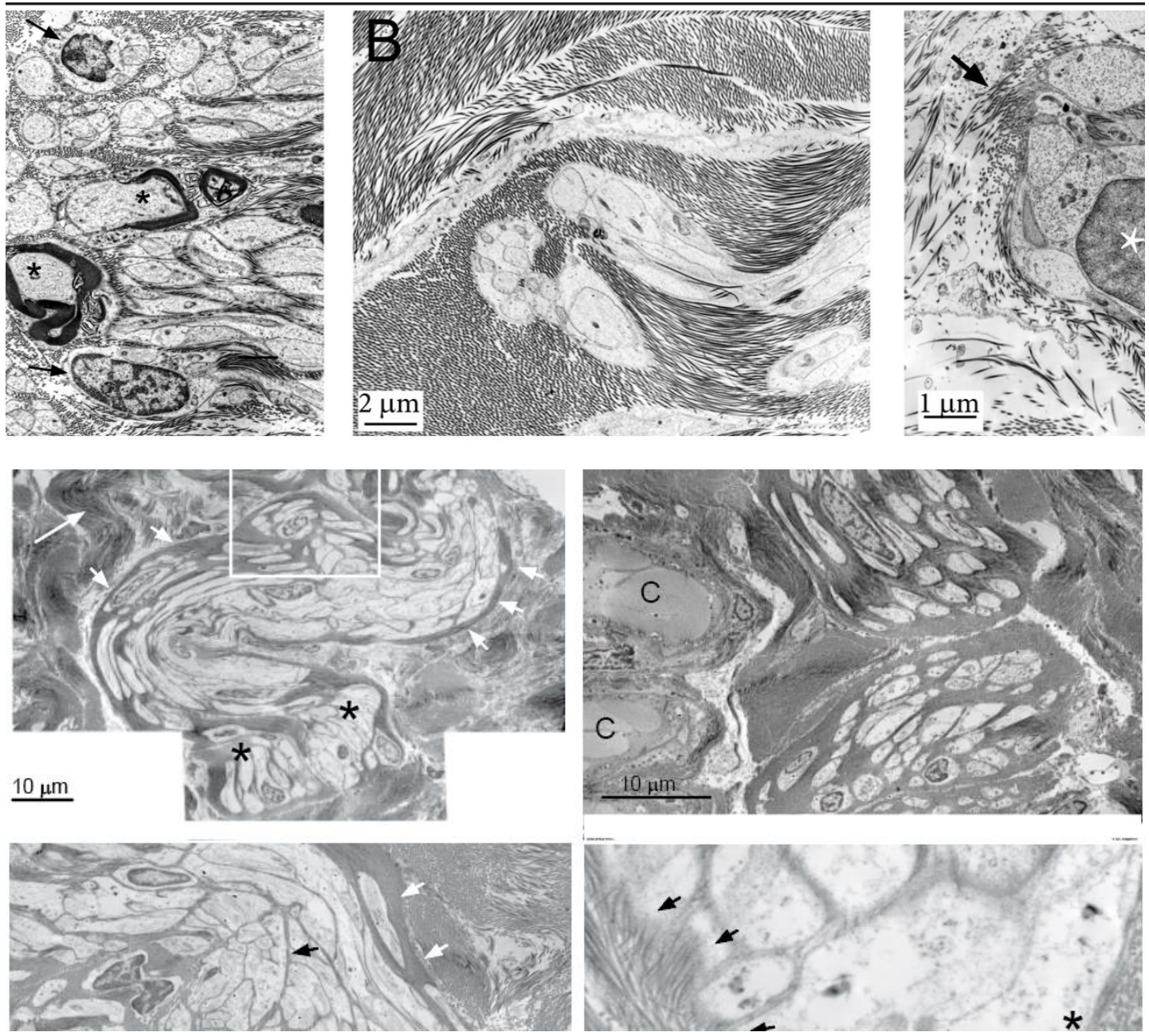


Das Filum tut weh!

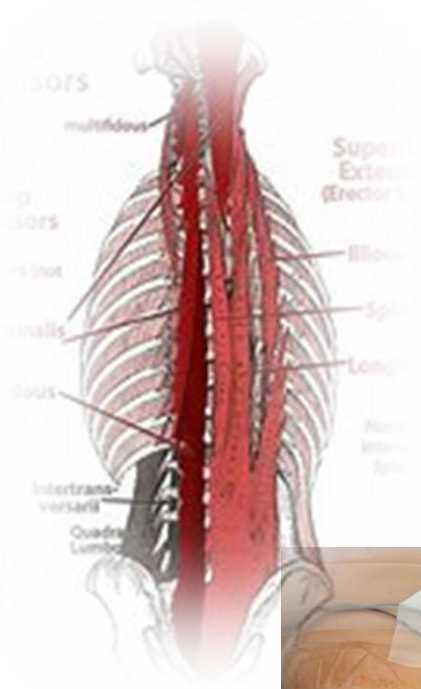
Not Just an Anchor: The Human Filum Terminale Contains Stretch Sensitive and Nociceptive Nerve Endings and Responds to Electrical Stimulation With Paraspinal Muscle Activation. Petra M. Klinge, Abigail McElroy, Owen P. Leary, John E. Donahue, Andrew Mumford, Thomas Brinker, Ziya L. Gokaslan, Neurosurgery. 2022 Oct; 91(4): 618–624

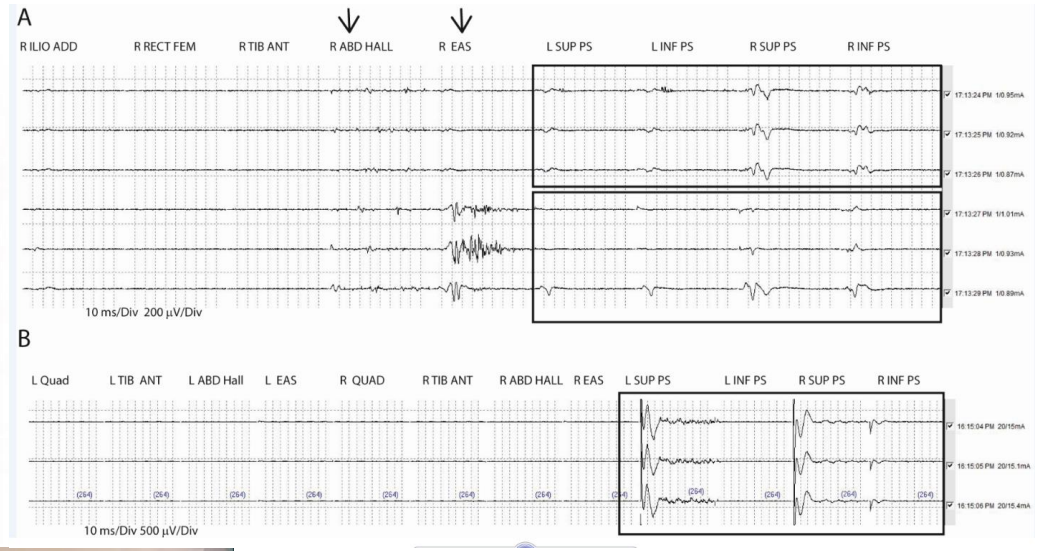
Schmerz und Mechanorezeptoren im Filum (Elektronenmikroskopie)

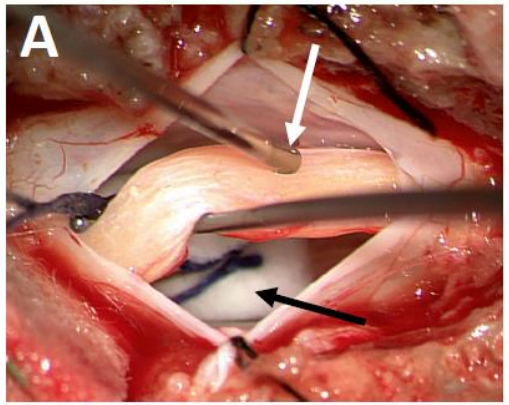
- A-C: Remak-Zellen =nichtmyelinisierte Schwannzellen. Die Nervenenden sind streng am Verlauf der umliegenden kollagenen Fasern ausgerichtet, wodurch möglicherweise Dehnungen des Filums auf die Axone übertragen werden und Schmerzen ausgeloest werden.
- Ruffinikorpuskel (klassische Dehnungsrezeptoren)

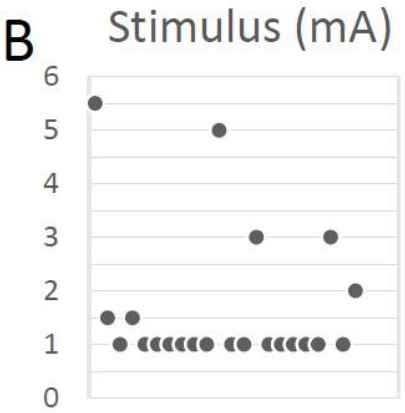


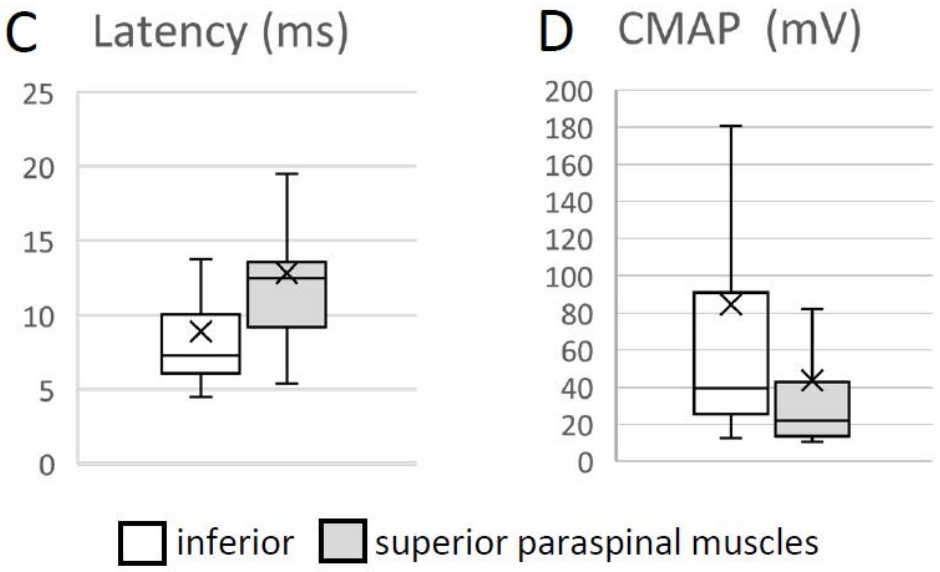
Intraoperatives EMG monitoring: Erklaert Rueckenschmerzen







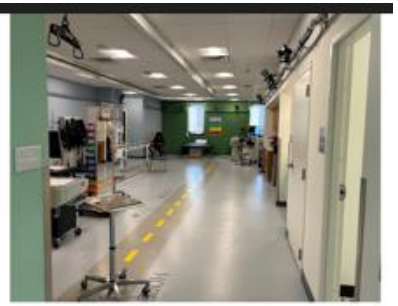




BROWNHealth
UNIVERSITY
Department of Neurosurgery | Brown Neuroscience

RIH IRB Approval #2250795

UNRAVELING SPINAL CONTROL IN EDS



WHAT ARE WE INTERESTED IN?

We're researching movement problems linked to the spine and tethered spinal cord in Ehlers-Danlos Syndrome (EDS). We've discovered that the filum terminale sends signals to a network of nerves along the spine that control spinal posture, movement, and coordination. We assume this process is altered in EDS due to spinal hypermobility and tethered cord, and is linked to back pain and spinal cord neurological symptoms in EDS.

We study why and how tethered cord surgery can be effective.

WHO CAN PARTICIPATE?

All patients over the age of 12 who have no diagnosis of EDS, tethered cord syndrome, or other spine anomalies, and who are able to walk without the use of assistive devices

WHAT DOES THE RESEARCH ENTAIL?

In our Center for Innovative Neurotechnology and Neural Repair (CINNR), our team of clinicians and neuroscientists will assess your movements while walking a short distance alongside an application of skin surface EMG and EEG to monitor your muscle activation and brain activity. During a max. 2 hour visit, you will perform simple reflex tasks, and you will be asked to walk along a path in the lab (max. 14 yards in total). There will only be 1 visit.

QUESTIONS?

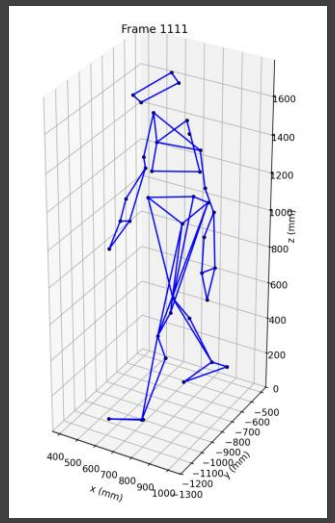
- Please contact the Neurosurgical Research Team by email:
- tcsresearch@brownhealth.org

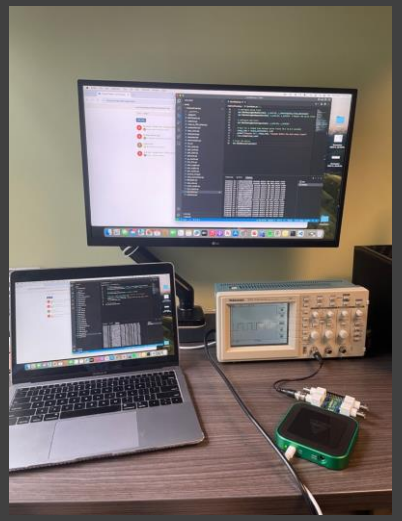
LOCATION:

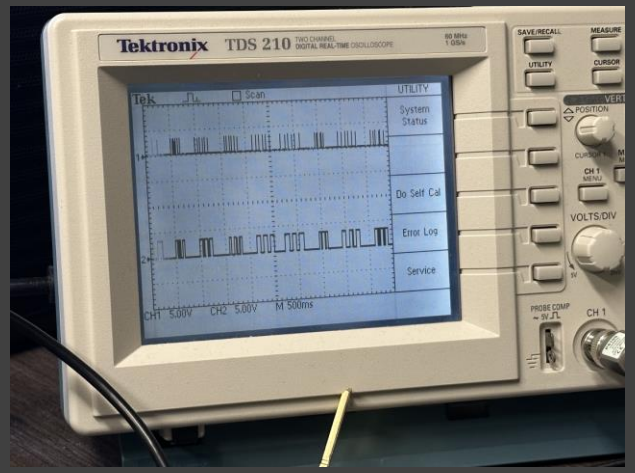
CINNR

- 117 Chapman Street., Providence RI
- Parking is free!

Scan this QR code to send an email indicating interest


Frame 1111
x (mm)
y (mm)
z (mm)

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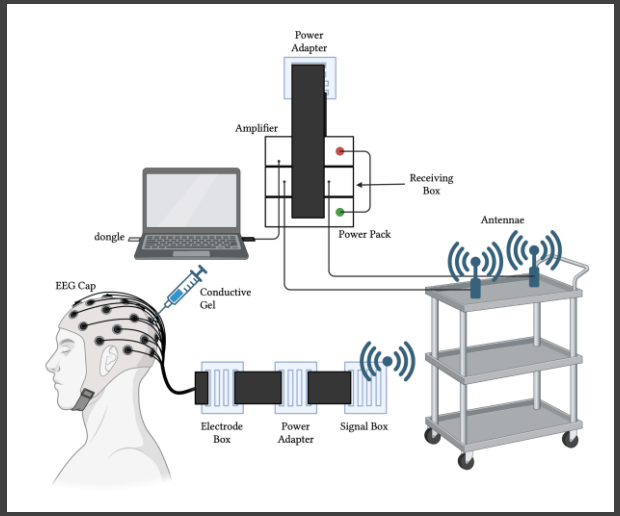
TDS 210
SAVE/RECALL
MEASURE
UTILITY
CURSOR
Tek
Scan
System Status
Do Self Cal
Error Log
Service
VOLTS/DIV
CH 1


Power Adapter
Amplifier
Receiving Box
dongle
Power Pack
Antennae
EEG Cap
Conductive Gel
Electrode Box
Power Adapter
Signal Box