

Adult Onset Tethered Cord Syndrome: Spinal Pathology, Clinical and Histological Characteristics and Post-Operative Outcomes



Matthew J. Hagan BS¹, Owen P. Leary ScB¹, Abigail McElroy DVM MS¹, David D. Liu ScB¹, Jared S. Fridley MD¹, Albert E. Telfeian MD PhD¹, Alexios G. Carayannopoulos DO MPH³, John E. Donahue MD², Ziya L. Gokaslan MD¹, Adetokunbo A. Oyelese MD PhD¹, Petra M. Klinge MD PhD¹



[1] Warren Alpert Medical School of Brown University, Dept. of Neurosurgery, Providence RI 02903

[2] Warren Alpert Medical School of Brown University, Dept. of Pathology, Providence RI 02903

[3] Comprehensive Spine Center, Department of Neurosurgery and Division of Pain and Rehabilitation Medicine, Department of Neurosurgery at Rhode Island Hospital.

Introduction

Tethered Cord Syndrome (TCS) is a rare neurological condition thought to be caused by tightening of the filum terminale. Most of what we understand about TCS is grounded in literature in pediatric populations, when the condition usually presents. However, the typical clinical course for the sub-population who presents in adulthood is not as well understood. The symptoms of so-called "adult-onset TCS" (ATCS) are more non-specific and more gradual in onset compared to pediatric populations.¹ Coupled with the fact that TCS is rarely considered a diagnostic option in adults makes the diagnosis and neurosurgical referral challenging. Unfortunately, delayed time to treatment of TCS has been associated with worsening neurologic function following surgical therapy.^{1,2} While surgical outcomes data have been encouraging following a de-tethering procedure, they highlight the need for further subcategorization of patients and identification of prognostic factor for improved outcomes.

Methods

Electronic medical records (EMR) of consecutive ATCS patients who underwent surgical detethering with a single provider were retrospectively reviewed. Clinical data were centrally collected and summarized using descriptive statistics. Available filum terminale specimens from included subjects were sectioned and stained with Haematoxylin and Eosin (H&E) for general histology and, when possible, analyzed using transmission electron microscopy (TEM). Correlation of pre-operative and pathologic characteristics with outcome were assessed using regression analysis.

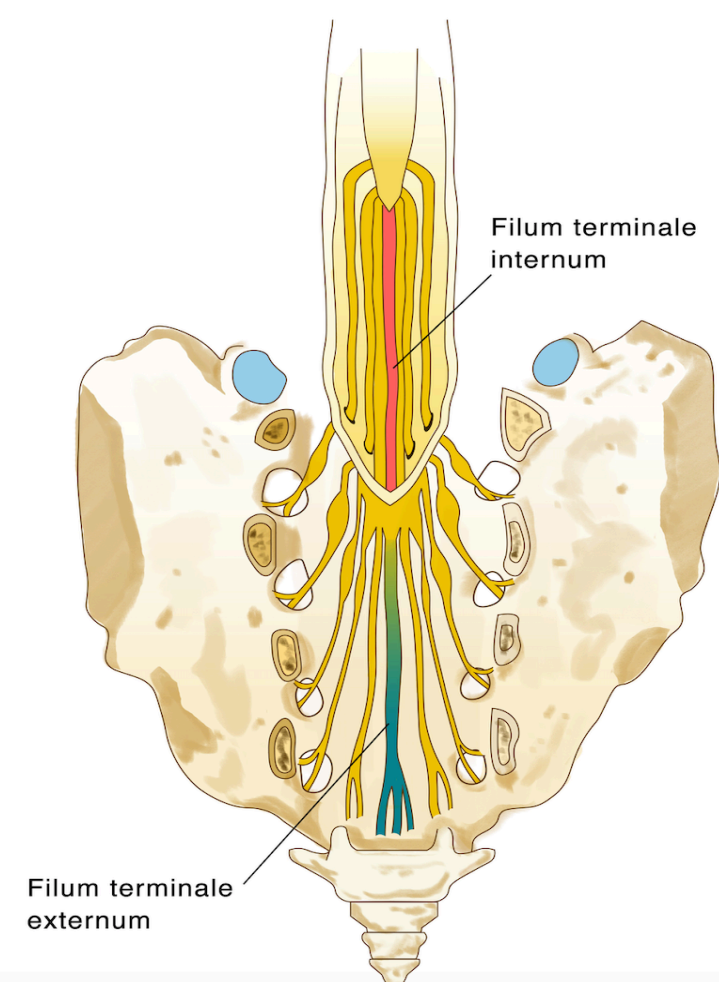


Figure 1: Illustration of filum terminale. During detethering surgery the dura is opened and the thickened filum is removed.

Results

Figure 2: TEM showing collagen disorganization and a 'moth-eaten' myelinated axon.

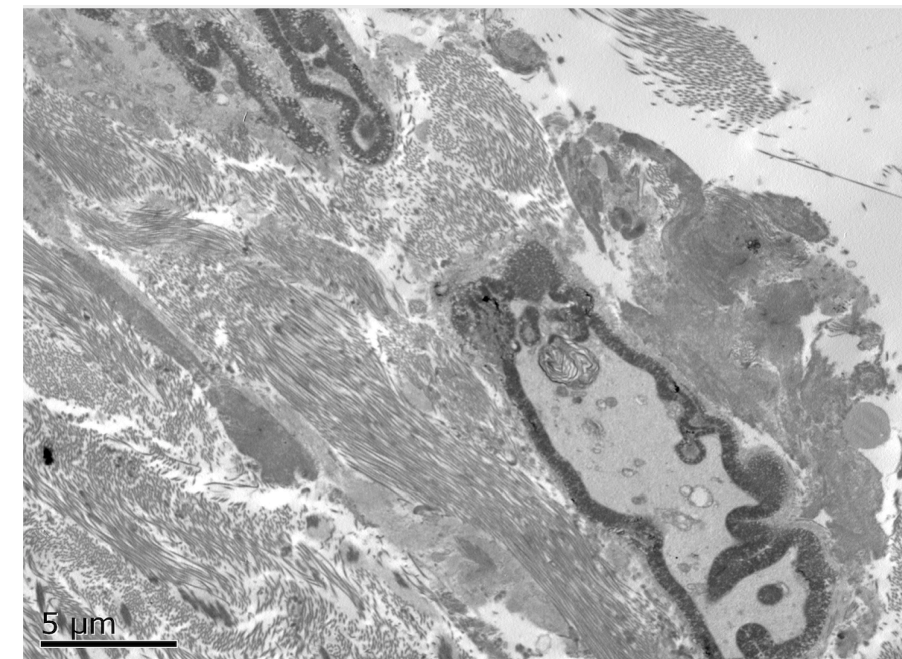


Figure 3: H&E image demonstrating a peripheral nerve fiber, ependyma, blood vessels, and collagen.

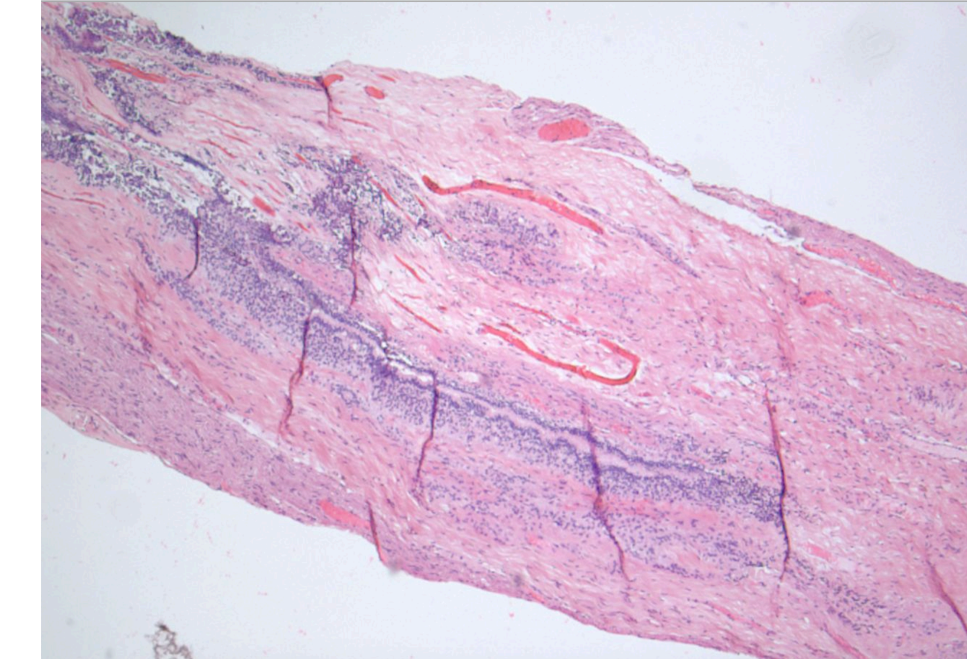
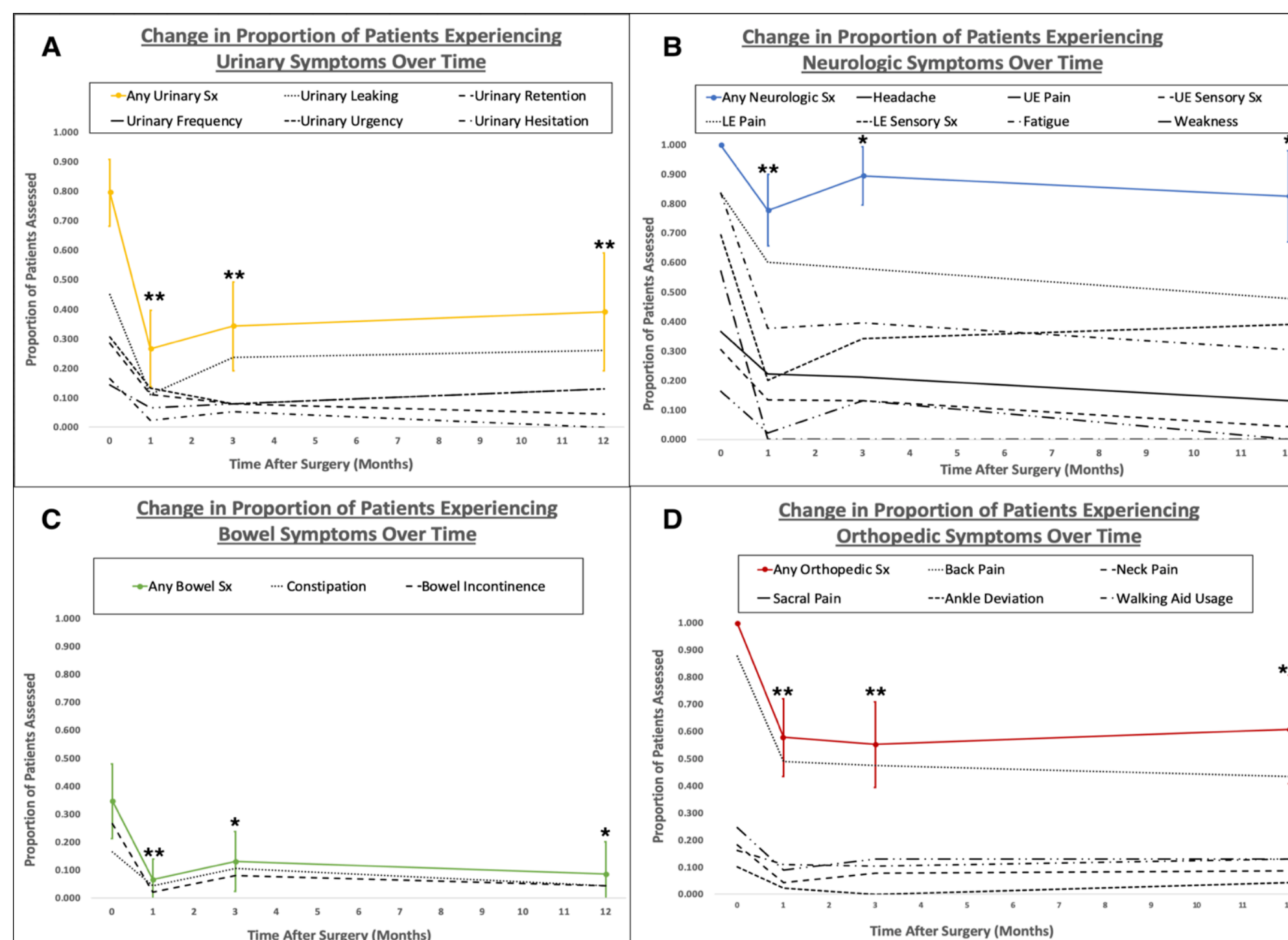


Table 1: Univariate regression found that presence of prominent blood vessels, neuropil / ependyma, neurons, nerve twigs, external nerve fibers, elastin abnormalities, and higher adipose score were all associated with symptomatic improvement by 3 months post-op, while the presence of Dorsal Root Ganglion (DRG) cells and collagen abnormalities within the filum were associated with worse outcomes.

Outcome Variables	Prominent Blood Vessels	Adipose Score	Thickened Filum	Neuropil / Ependyma	DRG Cells	Nerve Twigs	Presence of Neurons	External Nerve Fibers	Collagen Abnormality	Collagen Fibril Corkscrewing	Elastin Abnormality
Resolution of Urinary Sx	+	+		+		+	+	+	-		+
Resolution of Bowel Sx	+			+		+		-	-		-
Resolution of Orthopedic Sx	+	+		+	-	-	+	+	-	+	+
Resolution of Neurologic Sx		+						+			
Symptom-Free @ 3 Months		+						+			

Figure 4: The most common presenting symptoms were neurologic (100%) and orthopedic (100%), with lower proportions of patients reporting urinary (79.6%) or bowel (34.7%) symptoms. The proportion of patients experiencing each category of symptoms decreased significantly by the 1-month postoperative visit ($p < 0.001$) and remained significantly lower at 3-months and 12-months post-op. Urinary and orthopedic symptoms remained highly significant ($p < 0.001$) through the 3 and 12 month follow up visits while the changes in bowel and neurological symptoms became less significant at the further time points ($0.05 > p > 0.001$).



Results (cont.)

Table 2: On MRI, evidence of low lying conus medullaris was seen in 75.5% of patients, fatty filum terminale in 8.2%, filum lipomas in 69.4%, and degenerative spinal disease in 87.7%.

Radiographic Characteristics (Magnetic Resonance Imaging)	Pre-Op (N=49)	Post-Op (N=41)	P-Value
Signs of Tethered Cord Syndrome	49 (100.0%)		
LLC (L2 – L3)	5 (10.2%)	6 (14.6%)	0.528
LLC (L3 – L4)	2 (4.1%)	2 (4.9%)	0.857
LLC (L4 – L5)	30 (61.2%)	2 (4.9%)	<0.001*
Syrinx	9 (18.4%)	9 (22.0%)	0.676
Fatty Infiltration in the Filum	4 (8.2%)	12 (29.3%)	0.009*
Filum Lipoma	34 (69.4%)	0 (0.0%)	<0.001*
Signs of Degenerative Disease	43 (87.8%)	26 (63.4%)	0.006*
Lumbar Degenerative Findings	32 (65.3%)	31 (75.6%)	0.293
Canal Stenosis	17 (34.7%)	-	-
Foraminal Stenosis	20 (40.8%)	-	-
Listhesis	12 (24.5%)	-	-
Disc Disease	16 (32.7%)	-	-
Disc Herniation	5 (10.2%)	-	-

Conclusions

Surgical intervention resulted in favorable symptom improvement for many patients with evidence of ATCS, even in the presence of comorbid degenerative pathology. The correlation of clinical outcome with filum adipose content corresponds to previous findings in pediatric TCS.

Future Directions

- This research highlights the need for prospective studies aimed at the efficacy of detethering surgery
- Further analysis of pre-operative MRI findings that could confer improved prognostic outcomes

References

1. Yamada S. Tethered cord syndrome in children and adults. **Thieme**: 2010
2. Lew SM, Kothbauer KF. Tethered cord syndrome: an updated review. **Pediatr Neurosurg** 43(3): 236–248, 2007

Acknowledgments

This project received funding from the Alpert Medical School Summer Assistantship. Figure 1 was provided by Kendall Rivera-Lane, the Medical Illustrator in the RIH Neurosurgery Department.