SNI
SURGICAL NEUROLOGY INTERNATIONAL
SNI: Spine
OPEN ACCESS
For entire Editorial Board visit : http://www.surgicalneurologyint.com
Editor: Nancy E. Epstein, MD Winthrop Hospital, Mineola, NY, USA

Review Article

A review of the disagreements in the prevalence and treatment of the tethered cord syndromes with chiari-1 malformations

Nancy E. Epstein[1,2]

[1]Professor of Clinical Neurosurgery, School of Medicine, State University of N.Y. at Stony Brook, [2]Chief of Neurosurgical Spine and Education, NYU Winthrop Hospital, NYU Winthrop NeuroScience, Mineola, New York, USA

E-mail: *Nancy E. Epstein - nancy.epsteinmd@gmail.com
*Corresponding author

Received: 07 July 18 Accepted: 09 July 18 Published: 14 August 18

Abstract

Background: The tethered cord syndrome (TCS) accompanying Chiari-1 (CM-1) malformations and the occult tethered cord syndrome (OTCS) syndrome accompanying the low lying cerebellar tonsil (LLCT) syndrome may be treated with sectioning of the filum terminale (SFT).

Methods: Utilizing PubMed, we reviewed the neurosurgical literature to determine how frequently spinal neurosurgeons diagnosed the TCS (e.g., conus terminating below the normal L1-L2 disc level) on lumbar magnetic resonance (MR) studies in patients with CM-1 malformations [e.g. tonsils >5–12 mm below the foramen magnum (FM) warranting SFT]. In addition, we assessed how frequently spinal neurosurgeons encountered the OTCS (e.g., conus normally located at L1-L2 on MR) accompanying the LLCT (e.g., tonsils herniated <5 mm below the FM) also requiring SFT.

Results: According to the neurosurgical literature, the incidence of TCS accompanying CM-1 requiring SFT ranged from 2.2% to < 6%, and up to 14%. Few studies additionally highly correlated the OCTS accompanying the LLCT syndrome warranting SFT.

Conclusions: Given the differences in the literature, more studies are needed to assess the risks (complications) vs. benefits (improved neurological outcomes) of SFT surgery for TCS with CM-1 and SFT for OCTS with LLCT.

Key Words: Chiari-1 malformations, definitions, indications, low lying cerebellar tonsil syndrome, occult tethered cord syndrome, prevalence, section of filum terminale, tethered cord syndrome

Access this article online
Website: www.surgicalneurologyint.com
DOI: 10.4103/sni.sni_230_18
Quick Response Code:

INTRODUCTION

Utilizing PubMed, we reviewed the neurosurgical literature regarding the prevalence of the tethered cord syndrome (TCS) with Chiari-1 malformations (CM-1) requiring sectioning of the filum terminale (SFT). Additionally, we assessed how often the occult tethered cord syndrome (OCTS) was correlated with the low lying cerebellar syndrome (LLCT) also warranting SFT.

How to cite this article: Epstein NE. A review of the disagreements in the prevalence and treatment of the tethered cord syndromes with chiari-1 malformations. Surg Neurol Int 2018;9:161.
http://surgicalneurologyint.com/A-review-of-the-disagreements-in-the-prevalence-and-treatment-of-the-tethered-cord-syndromes-with-chiari-1-malformations/

Further, we asked how frequently the TCS, defined on magnetic resonance (MR) by the conus located below the L1-L2 level, correlated with CM-1 (tonsils >5 mm–12 mm below the foramen magnum FM), and with the LLCT [tonsils herniated < 5 mm below the FM on MR] [Tables 1 and 2]. Further, we queried how often the OCTS, defined on MR by the normal conus location at the L1-L2 level, corresponded with the LLCT (tonsils herniated <5 mm below the FM on MR) [Tables 1 and 2].

DEFINITIONS OF NORMAL TONSIL LOCATION VERSUS CHIARI-1 MALFORMATIONS

Normally, the cerebellar tonsils are located 2.9 mm ± 3.4 mm above or up to 3 mm below the FM [Table 1].[1] Borderline CM-1 are defined by the tonsils located between 3 and 5 mm below the FM [Table 1].[1] According to the neurosurgical literature, CM-1 are typically defined on MR scans by a >5 mm of tonsillar descent below the FM e.g.,[1,2,4,8]

FREQUENCIES OF CHIARI-1 MALFORMATIONS IN ADULTS AND CHILDREN

Different frequencies of CM-1 have been reported in the adult and pediatric age groups [Tables 1 and 2].[4,8,6,9] In 2000, Meadows *et al.* reviewed 22,591 MR studies (over 43 months); only 0.78% (175 patients) had CM-1 malformations (e.g. tonsils >5 mm below FM) [Table 1].[4] Using this definition, in 1999, Milhorat *et al.* diagnosed CM-1 in 332 patients, and in 2009, diagnosed CM-1 in 2987 patients [Tables 1 and 2].[6,8] In 2018, Passias *et al.* found the increased diagnosis of CM-1 in the pediatric population from 45 to 96 per 100,000 [e.g. 5432 Kid Database (2003–2012)], in patients averaging 10.5 years of age (range, 0–20) [Table 2].[9]

SYMPTOMATIC CHIARI-1 MALFORMATIONS

According to the literature, the onset of symptoms correlated with differing degrees of CM-1 tonsillar herniation [Table 1].[1,2,4] In the study by Aboulezz *et al.*, CM-1 patients became symptomatic once the MR demonstrated an average of 10.3 ± 4.5 mm (13 patients) of tonsillar herniation below the FM.[1] In the study by Elster *et al.*, out of 68 patients with CM-1, 70% were symptomatic when the tonsils were 5–10 mm below the FM, while 100% were symptomatic with tonsils >12 mm below the FM [Table 1].[2] In 2000, Meadows found only 175 CM-1 malformations (0.78%) out of 22,591 MR studies performed; nevertheless, only 25 (14% of the 175) were symptomatic from tonsils averaging 11.4 mm ± 4.86 mm below the FM [Table 1].[4]

Table 1: Definitions of normal tonsil location, chiari-1 malformations, and low lying cerebellar tonsil syndrome

Author (ref) year	Number of Patients Pathology	Clinical Data	Additional Criteria	Results	Conclusions
Aboulezz *et al.*, 1985[1]	Normal tonsils 2.9±3.4 above FM (82 patients)	Normal tonsils Up to 3 mm below FM	Borderline CM-1 tonsils >3-5 mm below FM	CM-1 MR >5 mm cerebellar tonsils below FM	Symptomatic Chiari-1 10.3=/- 4.6 mm below FM-13 Patients Symptoms
Elster and Chen 1992[2]	CM-1 MR 68 Patients	3 female: 2 male ratio 24% skeletal abnormality 40% Syrinx	5-10 mm tonsil below FM - 70% Symptomatic	>12 mm tonsils below FM 100% Symptomatic	More common incidental CM-1 malformations
Milhorat *et al.*, 1999[8]	CM-1 Tonsils at least 3-5 mm below FM - 364 Patients Reduced MR Volume Posterior Fossa Mean 13.4 mm New LLCT	275 female: 89 male Age 24.9±15.8 24% trauma 65% Syrinx 42% Scoliosis 12% BI	Obliterated Retrocere-bellar fluid spaces Symptoms Headache Pseudo-tumor	Symptoms Meniere's Lower cranial Nerves Spinal cord deficit	CM-1 Tonsils Down >5 mm (332/364 Patients New LLCT 34 patients Tonsils down <5 mm
Meadows *et al.*, 2000[4]	CM-1 on 22,591 MR over 43 months	Defined CM-1 >5 mm tonsil herniation below FM	0.78% incidence CM-1 on MR 175 patients >5 mm below FM	Only 25 (14%) Patients Symptomatic	Average 11.4 mm=/- 4.86 mm for Symptomatic CM-1
Milhorat *et al.*, 2007[7]	Occipital-atlantal Hyper-mobility CM-1	Cohort 2813 patients with CM-1	EDS/HDCT 357 (12.7%) of 2813 Patients	Cervical spine reduced with traction	Diagnosed EDS/HDCT 357 patients with CM-1 (12.7%)

CM-1: Chiari-1 Malformations, CSF: Cerebrospinal fluid, HDCT: Hereditary connective tissue disorder, CT: Computed tomography, MR: Magnetic Resonance Imaging, EDS: Ehlers-Danlos Syndrome, KID: Kids Inpatient Database, TCS: Tethered cord syndrome, OCTS: Occult Tethered Cord Syndrome, LLCT: Low Lying Cerebellar Tonsils, FM: Foramen Magnum, HC: Hydrocephalus, BI: Basilar invagination

Table 2: Tethered cord and occult tethered cord syndrome with Chairi-1 Malformations and low lying cerebellar tonsil syndrome

Author (ref) year	Number of Patients Pathology	Clinical Data	Additional Criteria	Results	Conclusions
Metcalfe *et al*., 2006[5]	Normal MR 36 patients 0.04% Pediatric Urology Clinic Referred for SFT for OTCS	SFT Criteria Failed 2 years nonsurgical management 83% Daytime Incontinence	47% Encopresis 55% Urodynamic < Bladder Capacity	72% SFT Clinically improved 42% better Incontinence 88% better Bowel function	57% Urodynamics Improved No Single Factors predict SFT outcome
Steinbok *et al*., 2007[10]	Persistent Urinary Incontinence Normal MR Conus location OCTS SFT versus No Surgery	SFT 8 children Ages 4.4-9.8 1-2 operations Urological improvement 7 at 3.1 years 4 Improved urodynamic	No surgery 7 Children Ages 3.1 to 13.5 Followed 3.3 years-2 GU improved 3 Ureters Reimplanted 1 SFT 8 years later (better)	Hypothesis SFT in children with OCTS may improve	Requires true RCT very small numbers Not significant Findings
Steinbok and MacNeily 2007[11]	Occult TCS (OTCS) MR: Normal Conus Location	Proposed OCTS with "Clinically" Tethered Filum	Controversial Symptoms Pediatrics; Neurogenic Urinary Incontinence	Unresponsive to no surgery Suggest SFT to Improve Symptoms	Poor quality evidence need RCTs
Milhorat *et al*., 2009[6]	CM-1/LLCT TCS/Occult TCS Requiring SFT Prevalence TCS 2987 CM-1 289 LLCT LLCT	74 children 244 adults CM-1 with 14% TCS (408 Patients)/SFT LLCT (289) 63% TCS/OTCS (182 patients SFT	More MR data SFT 318 No difference size Posterior Fossa Elongation brain stem (8.3 mm) Downward Displacement Medulla (mean 4.6 mm)	Symptoms Improved 69 (93%) KIDS 83%(203) Adults Symptoms Unchanged 5 (7%) KIDS 39 (16%) Adults 2 Symptoms Worse Adults	Followed 6-27 most mean 16 most MR 1-18 mos. PostoperativeUpward Migration Tonsils 3.8 mm
Valentini *et al*., 2011[12]	With 110 CM-1 TCS with SFT<6% Incidence	TCS due to Conus or Filum Lipomas	TCS with CM-1 Rare <6%	SFT No change in Tonsillar Position	No Correlation Between TCS and CM-1Tonsil Location
Massimi *et al*., 2011[3]	Is SFT indicated for isolated TCS SFT used for OCTS	CM-1 No Correlation CM-1 and TCS	Old Caudal Traction Theory NOT Supported by Evidence	No Relationship CM-1 and OCTS	CM-1/OCTS Questionable Indications for SFT
Passias *et al*., 2018[9]	CM-1 5432 Ages 0-20 Kid database 2003-2012 Average age 10.5 55% Female CM-1 increased 45 to 96/100,000 Other Findings 23.8% Syringomyelia syringobulbia	Other findings 11.5% scoliosis 5.9% HC 2.2% TCS Surgery Increased 2003-2012 66% -72%	Adverse events 2003-2012 7%-3% 7% mortality same 2003-2012 Minimum one AE Neurologic, Dysphagia, Respiratory	Surgery 2003-2012 70% Decompression ONLY Cranial Decompressions Decreased 42.2%-30% Spinal Decompression increased 73.1%-77.4%)	Fusion Rates Increased 0.45%-1.8% >> Complication For Fusion 11.9% versus 4.7% Decompression Alone

CM-1: Chiari-1 Malformations, VAS: Visual Analog Score, CSF: Cerebrospinal Fluid, HDCT: Hereditary connective Tissue Disorder, CT: Computed Tomography, MR: Magnetic Resonance Imaging, EDS: Ehlers-Danlos Syndrome, KID: Kids Inpatient Database, TCS: Tethered Cord Syndrome, OCTS: Occult Tethered Cord Syndrome, LLCT: Low Lying Cerebellar Tonsils, SFT: Section Filum Terminale, AE: Adverse Event, HC: Hydrocephalus, IM: Intramedullary, RCT: Randomized Controlled Trials, OTCS: Occult TCS, GU: Urological

DEFINITION AND FREQUENCY OF THE LOW LYING CEREBELLAR TONSIL SYNDROME (LLCT)

The neurosurgical literature revealed few references to the LLCT [Table 1].[6,8] Milhorat *et al*. in 1999 found 34 patients with MR-documented tonsils <5 mm below the FM who exhibited "Chiari-1 like" clinical syndromes; this led to their definition of the LLCT [Table 1].[8] In 2009, Milhorat *et al*. evaluated 2987 patients with CM-1, and identified another 289 (9.7%) patients with LLCT.[6]

ANOMALIES ASSOCIATED WITH ADULT/PEDIATRIC CHIARI-1/LOW LYING CEREBELLAR TONSIL SYNDROME (LLCT)

The neurosurgical literature demonstrated multiple anomalies accompanying the CM-1/LLCT syndromes

in adult and pediatric populations [Tables 1 and 2].[2,7-9] Several studies defined the following; a 24% incidence of skeletal abnormalities, a 40%-65% frequency of syrinx formation, a 42% incidence of scoliosis, a 12% frequency of basilar invagination, and 12.7% incidence (357 out of 2813 patients with CM-1) with occipital-atlantal hypermobility [hereditary connective tissue disorder (HCTD)/Ehlers–Danlos syndrome (EDS)] [Table 1].[2,8,6] In Passias *et al*., pediatric series of 5432 patients with CM-1 (kid database 2003–2012; ages 0–20), 23.8% had accompanying syringomyelia, 11.5% exhibited syringobulbia, and 5.9% had hydrocephalus [Table 2].[9]

DIFFERENT FREQUENCIES OF TETHERED CORD SYNDROME WITH CHIARI-I

A review of the neurosurgical literature revealed a variable correlation between CM-1 and the TCS (e.g. conus below the L1-L2 disc level on MR) requiring SFT [Table 2].[6,9,12] In 2009, Milhorat *et al*. described a 14% incidence of TCS with CM-1 (2987 patients) requiring SFT [Table 2].[6] Valentini *et al*., in 2011, evaluating 110 patients with CM-1 malformations, and found a <6% incidence of TCS requiring SFT: they concluded there was no significant correlation between the two [Table 2].[12] In 2018, Passias *et al*. reported that, for 5432 children with CM-1 (e.g., kid database, 2003–2012), only 2.2% had associated TCSs; they considered this a minimal correlation at best [Table 2].[9]

CRANIAL VS. SPINAL DECOMPRESSION VS. FUSION FOR CHIAIR-I MALFORMATIONS IN THE PEDIATRIC AGE GROUP

In the pediatric neurosurgical literature, the study by Passias *et al*. (2018) was the most prominent one discussing the successive 2003–2013 frequencies of cranial vs. spinal decompression vs. fusion for CM-1 malformations [Table 2].[9] They found that, out of 5432 children undergoing surgery for CM-1 malformations, the incidence of cranial decompression decreased from 42.2% to 30%, while the frequency of spinal decompression increased from 73.1% to 77.4%. Over the same period, fusion rates increased from 0.45% to 1.8%, but correlated with more complications (e.g., 11.9% adverse events for fusion vs. 4.7% for decompression alone) [Table 2].

DEFINITION OF THE OCCULT TETHERED CORD SYNDROME (OCTS)

A review of the neurosurgical literature showed that few spinal neurosurgeons highly correlated the OCTS (normal location of the conus at the L1-L2 level on MR) warranting SFT with the LLCT [Table 2].[6] In 2009, Milhorat *et al*. observed that, for 289 patients with LLCT (289), there was a 63% frequency of the OTCS (182) warranting SFT [Table 2].[6] Patients exhibited a clinical "Chiari-like syndrome" attributed in part to the elongation/downward displacement of the hindbrain/cerebellar ectopia, reduced cerebrospinal fluid (CSF) flow in the lumbar theca, and other factors, relieved by SFT.

DIFFERENT FREQUENCIES OF SECTIONING OF THE FILUM TERMINALE FOR OCCULT TETHERED CORD SYNDROME IN PATIENTS WITH CHIARI-I/LOW LYING CEREBELLAR TONSIL SYNDROMES (LLCT)

Multiple studies in the neurosurgical literature summarized the results of SFT performed for OTCS in patients with CM-1/LLCT syndromes [Table 2].[3,5,10] Metcalfe *et al*. in 2006 diagnosed 36 pediatric patients with OCTS; this represented just 0.04% of all pediatric urological clinic visits. Signs and symptoms included 83% daytime urological incontinence, 55% reduced bladder capacity, and 47% encopresis [Table 2].[5] Following SFT, 72% of patients improved, 42% had improved urinary incontinence, 57% had improved urodynamics, and 88% exhibited improved bowel incontinence. Steinbok *et al*. in 2007 similarly defined OCTS in patients with neurogenic urinary incontinence [Table 2].[11] In a separate study, they further compared the results of SFT in children with urological symptoms and signs attributed to OCTS. Results of SFT performed in 8 children were compared vs. 7 managed without surgery [Table 2].[10] Up to 3.1 years postoperatively, 7 of 8 patients undergoing SFT improved (e.g., urodynamic confirmation in 4 patients). Over an average of 3.3 years, of the 7 children treated nonsurgically, 2 had urological improvement, 4 had bilateral ureteric reimplantation, and 1 underwent a delayed SFT 8 years later with improvement. The authors concluded that future randomized controlled studies (RCTs) were required to better document the safety/efficacy of SFT for OCTS. In 2011, Massimi *et al*. found no correlation between CM-1 and the frequency of the TCS or OTCS syndromes [Table 2].[3] They could not identify any clinical or experimental evidence to support the "caudal traction theory" for performing SFT in patients with OCTS (e.g., releasing the tethered cord allowing the tonsils to migrate cephalad/ascend). They also concluded that more studies were warranted and that the value of this treatment was "still under debate" [Table 2].

CONCLUSION

Reviewing the neurosurgical literature revealed a 2.2% to <6%, up to a higher 14% frequency of TCS requiring

SFT in patients with CM-1 malformations [Table 2].[6,9,12] Few studies highly correlated the OTCS requiring SFT with the LLCT syndrome [Table 2].[6] Given the differences in the literature reviewed, further studies are warranted to determine the risks (complications) vs. benefits (improved clinical outcomes) for performing SFT for TCS with CM-1, and OTCS with the LLCT syndrome.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Aboulezz AO, Sartor K, Geyer CA, Gado MH. Position of cerebellar tonsils in the normal population and in patients with Chiari malformation: A quantitative approach with MR imaging. J Comput Assist Tomogr 1985;9:1033-6.
2. Elster AD, Chen MY. Chiari I malformations: Clinical and radiologic reappraisal. Radiology 1992;183:347-53.
3. Massimi L, Peraio S, Peppucci E, Tamburrini G, Di Rocco C. Section of the filum terminale: Is it worthwhile in Chiari type I malformation? Neurol Sci 2011;32 Suppl 3:S349-51.
4. Meadows J, Kraut M, Guarnieri M, Haroun RI, Carson BS. Asymptomatic Chiari type I malformations identified on magnetic resonance imaging. J Neurosurg 2000;92:920-6.
5. Metcalfe PD, Luerssen TG, King SJ, Kaefer M, Meldrum KK, Cain MP, *et al.* Treatment of the occult tethered spinal cord for neuropathic bladder: Results of sectioning the filum terminale. J Urol 2006;176:1826-9.
6. Milhorat TH, Bolognese PA, Nishikawa M, Francomano CA, McDonnell NB, Roonprapunt C, *et al.* Association of Chiari malformation type I and tethered cord syndrome: Preliminary results of sectioning filum terminale. Surg Neurol 2009;72:20-35.
7. Milhorat TH, Bolognese PA, Nishikawa M, McDonnell NB, Francomano CA. Syndrome of occipitoatlantoaxial hypermobility, cranial settling, and Chiari malformation type I in patients with hereditary disorders of connective tissue. J Neurosurg Spine 2007;7:601-9.
8. Milhorat TH, Chou MW, Trinidad EM, Kula RW, Mandell M, Wolpert C, *et al.* Chiari I malformation redefined: Clinical and radiographic findings for 364 symptomatic patients. Neurosurgery 1999;44:1005-17.
9. Passias PG, Pyne A, Horn SR, Poorman GW, Janjua MB, Vasquez-Montes D, *et al.* Developments in the treatment of Chiari type I malformations over the past decade. J Spine Surg 2018;4:45-54.
10. Steinbok P, Kariyattil R, MacNeily AE. Comparison of section of filum terminale and non-neurosurgical management for urinary incontinence in patients with normal conus position and possible occult tethered cord syndrome. Neurosurgery 2007;61:550-5.
11. Steinbok P, MacNeily AE. Section of the terminal filum for occult tethered cord syndrome: Toward a scientific answer. Neurosurg Focus 2007;23:E5.
12. Valentini LG, Selvaggio G, Visintini S, Erbetta A, Scaioli V, Solero CL, *et al.* Tethered cord: Natural history, surgical outcome and risk for Chiari malformation I (CM1): A review of 110 detethering. Neurol Sci 2011;32 Suppl 3:S353-6.