Review Article

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

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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., 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 OTCS 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 OTCS 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

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.
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 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-I 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]

**FREQUENCIES OF CHIARI-I MALFORMATIONS IN ADULTS AND CHILDREN**

Different frequencies of CM-1 have been reported in the adult and pediatric age groups [Tables 1 and 2].[4,6,8] 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-I 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 (15 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].[3] 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) | Number of Patients | 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 More common incidental CM-1 malformations |
| 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 | |
| Milhorat et al., 1999[3] | 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 | Symptomatic Menière'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 | Only 25 (14%) Patients Symptomatic Average 11.4 mm=/. 4.6 mm for Symptomatic CM-1 |
| Milhorat et al., 2007[5] | Occipital-atlantal Hyper-mobility CM-1 | Cohort 2813 patients with 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-I 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.
Additional Criteria

Table

Conclusions

Results

Table 2: Tethered cord and occult tethered cord syndrome with Chiari-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[6] | Normal MR 36 patients 0.04% Pediatric Urology Clinic Referred for SFT for OTCS | SFT Criteria Failed 2 years nonsurgical management | 47% Encopresis 55% Urodynamic < Bladder Capacity | 72% SFT Clinically improved | 57% Urodynamics Improved |
| Steinbok et al., 2007[7] | 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 More MR data | Unresponsive to no surgery Suggest SFT to Improve Symptoms | Poor quality evidence need RCTs |
| Milhorat et al., 2009[9] | CM-1/LLCT TCS/Occult TCS Requiring SFT Prevalence TCS 2987 CM-1 289 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 Worsened 83% 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 CM-1 | TCS with CM-1 Rare <6% | SFT No change in Tonsillar Position | No Correlation Between TCS and CM-1 Tonsil Location CM-1/OCTS Questionable Indications for SFT |
| 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 | No relationship CM-1 and OCTS Questionable Indications for SFT |
| Passias et al., 2018[63] | 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 | Adverse events 2003-2012 70% Decompression ONLY 7% mortality 7% 2003-2012 Minimum one AE Neurologic, Respiratory | Surgery 2003-2012 70% Decompression ONLY 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 |

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

The neurosurgical literature revealed few references to the LLCT [Table 1].[63] 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].[3] 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-I/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].

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].

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].

### 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].

In 2009, Milhorat et al. described a 14% incidence of TCS with CM-1 (2987 patients) requiring SFT [Table 2].

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].

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].

### CRANIAL VS. SPINAL DECOMPRESSION VS. FUSION FOR CHIARI-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].

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].

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]. 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-I/LLCT syndromes [Table 2].

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 85% daytime urological incontinence, 55% reduced bladder capacity, and 47% encopresis [Table 2]. 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].

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]. 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].

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.

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Conflicts of interest
There are no conflicts of interest.

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