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Sectioning of the Filum Terminale in Patients with Chiari Malformation Type 1 Associated with Occult Tethered Cord Syndrome: Literature Review

Secção do filum terminale em pacientes com malformação de chiari tipo 1 associado a síndrome oculta da medula presa: revisão de literatura

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Abstract

Approximately 125 years ago, a group of pathologies now known as Chiari malformations was described for the first time. However, some mechanisms of its formation still remain unknown. A bibliographic survey was performed through a search in PubMed. In 1938, it was already theorized that an increase in spinal cord tension could be the cause of Chiari malformation type 1 (CM1) tonsillar herniation. In 1953, a condition known for the anchoring of the filum terminale to the vertebral canal was described for the first time and would later be known as tethered cord syndrome (TCS). Some studies have shown that it is associated with increased tension in the spinal cord, and this formed the basis for a possible pathophysiological explanation of tonsillar herniation. Case series emerged reporting that treatment for TCS with the sectioning of the filum terminale (SFT) could provide clinical improvement of patients with CM1. A new pathological entity emerged when it was realized that patients with the clinical picture of TCS could have the medullary cone in its normal position, differing from the caudal migration expected for the TCS. This condition became known as occult tethered cord syndrome (OTCS). Case series attempted to demonstrate its association with the origin of CM1, a non-intuitive association, since the cone in the normal position contradicts traction as a

Keywords
- Arnold-Chiari malformation
- cauda equina
- tonsillar herniation
- tethered cord syndrome; spinal cord

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source of tonsillar herniation. To this day, the absence of randomized control trials limits any conclusions regarding the effectiveness of SFT for the treatment of patients with CM1.

Resumo

Há cerca de 125 anos, era descrita pela primeira vez um grupo de patologias hoje conhecidas como malformações de Chiari. No entanto, alguns mecanismos de sua formação ainda permanecem desconhecidos. Um levantamento bibliográfico foi feito através do PubMed. Em 1938, já se teorizava que um aumento da tensão medular poderia ser a causa da herniação tonsilar da malformação de Chiari tipo 1 (MC1). Em 1953, foi descrita pela primeira vez uma condição conhecida pelo ancoramento do filum terminale ao canal vertebral e que mais tarde viria a ser conhecida como síndrome da medula presa (SMP). Alguns estudos demonstraram que ela estava associada à tensão aumentada na medula espinhal, e a partir disso estava formada a base para uma possível explicação fisiopatológica da herniação tonsilar. Séries de casos surgiram relatando que o tratamento para a SMP com a secção do filum terminale poderia proporcionar melhora clínica aos pacientes com MC1. Uma nova entidade patológica surgiu quando se percebeu que pacientes com o quadro clínico de SMP poderiam ter o cone medular em sua posição normal, diferente da migração caudal esperada para a SMP. Essa condição ficou conhecida como SMP oculta. Séries de casos tentaram demonstrar sua associação com a origem da MC1, uma associação nada intuitiva, visto que o cone na posição normal contradiz a tração como fonte da herniação tonsilar. A ausência de ensaios randomizados controlados até o dia de hoje não permite concluir a eficácia do método de secção do filum no tratamento de pacientes com MC1.

Palavras-chave

- Malformação de Arnold-Chiari
- Cauda Equina
- Herniação Tonsilar
- Síndrome da Medula Presa
- Medula Espinhal

Introduction

The idea that Chiari malformation (CM) could be associated with increased levels of spinal cord tension appeared around 1938, when Penfield and Coburn[1] introduced the "traction theory" to explain the formation of cerebellar herniation.

In the 1990s, studies have theorized the possibility of tethered cord syndrome (TCS) in individuals with their spinal cord in the normal position, a condition that has been called occult tethered cord syndrome (OTCS).[2]

The use of the sectioning of the filum terminale (SFT) in the treatment of patients with CM type 1 (CM1) associated with symptoms compatible with TCS and conus medullaris in the normal position is not supported by the literature. There are few published studies on this topic, and no randomized controlled trial that provides evidence of the benefit from this intervention.[3]

The present study aimed to review the published works involving patients diagnosed with CM1 and TCS with conus medullaris in the normal position who underwent SFT surgery and to highlight the impact of this procedure on the clinical picture of these patients.

Methods

A Pubmed bibliographic survey was performed. Additionally, articles were obtained through cross-reference. No time limit was established. The articles obtained ranged from historical records to the most current approaches on this topic. The descriptors used were: *Arnold Chiari malformation, type 1*, *tethered cord syndrome*, *occult tethered cord syndrome*, *filum terminale*, *sectioning of filum terminale*, and its variations, alone or in combination. For this study, articles were limited to those written in the English language in which humans were defined as the subjects.

The inclusion criteria were original articles describing the association between CM1 and OTCS and the evolution of knowledge of both pathologies and studies presenting results of the SFT in patients with CM1 and TCS with conus medullaris in the normal position (OTCS). Articles that did not report any of the correlations cited in the inclusion criteria; those that focused on other types of CM; those that exclusively addressed classical TCS, and/or duplicates between the databases were excluded. All bibliographic material included in this review was analyzed by a critical reading of the texts to assess their eligibility and to interpret and compare their findings.

Chiari Malformation Type 1

Chiari malformation comprises a group of pathologies that result from a defective formation of the rhombencephalon and posterior fossa and have cerebellar herniation in common through the foramen magnum and result from a defective formation of the rhombencephalon and posterior fossa.[4] Arnold[5] and Chiari[6] described these changes in 1894 and

1895, respectively, and some of the mechanisms involved in the formation of this disease remain unclear to this day. Chiari malformation can present in several ways and is even associated with other types of malformation, such as syringomyelia and basilar invagination. The incidence of CM1 is estimated at 1/1,000 births[7] and results from an abnormal development of the occipital bone leading to hypoplasia of the posterior fossa and tonsillar herniation.[8] If the herniation of the tonsils towards the upper medullary canal is greater than 5 mm, the condition is known as CM1.[9] This internal pressure in the spinal canal, especially in the presence of basilar invagination, which is often associated with Chiari, because it shares defects in the same anatomical region, leads to a blockage of the flow of cerebrospinal fluid (CSF) at the craniospinal junction.[10]

Treatment is directed towards patients who exhibit symptoms, and it can be done with surgical and non-surgical approaches. If the symptoms are severe, surgical intervention may be necessary. The most used technique consists of the decompression of the posterior fossa with or without duraplasty.[11–14] Other procedures used in surgical treatment in patients with CM1 are: craniectomy, meningeal repair, other brain excisions, cranial nerve decompressions, spinal decompression, fusion surgeries, and other spinal surgeries.[15]

Tethered Cord Syndrome

Tethered cord syndrome (TCS) is a widely known neurological disorder. Its concept has been developed over time among neurologists and neurosurgeons and is gaining space as a factor involved in the etiopathogenesis of other pathologies of the central nervous system. It is believed that its origin, in the congenital form, is related to defects in embryological development during secondary neurulation, leading to the infiltration of fat in the filum and a simultaneous reduction in its elasticity.[16–18] This last factor is extremely important for the development of the condition, since the filum must be elastic. The loss of this elasticity can lead to increased tension over spinal segments, resulting in the appearance of the characteristic signs and symptoms of the syndrome.[19,20] Errors in the embryological process of filum development can lead to structural changes that favor the adherence of neural tissue to adjacent structures, making it difficult for the spinal cord to rise and causing tension in the segments. This explanation for pathophysiology became known as "traction theory"[1]. The clinical findings affect the neurological, neurocutaneous, neuro-urological, and neuro-orthopedic systems.[17] The SFT surgery has been used as the standard procedure to prevent the progression of neurological signs and symptoms and to alleviate existing ones.[21–23]

Radiological Aspects

The normal filum terminale is often so thin that it can barely be detected. In the diagnosis of TCS, the thick filum terminale is often defined as greater than 2 mm in diameter.[24] This cutoff point has been cited in the literature for years and used as the upper limit of normal during intraoperative, myelographic, and magnetic resonance measurements.[25] In addition, a spinal cord that ends below the levels of the vertebral body of L2 or L3 and a conus medullaris subsequently displaced with the filum in contact with the dural sac on or near the L5 blade have been established to aid in the diagnosis of this condition.[26,27] Fat appears as low attenuation on computed tomography (CT) and brilliant in T1-weighted magnetic resonance imaging (MRI) sequences.[28] However, a fatty filum can be an incidental finding,[29,30] and it is not considered a diagnosis of TCS, as it is present in 5.8% of the normal population in the postmortem examination.

The Traction Theory

The idea that CM could be associated with increased levels of spinal cord tension appeared around 1938, when Penfield and Coburn[1] introduced the *traction theory* to explain the formation of cerebellar herniation. This theory is based on the observation of CM1 in children with some condition that causes the adhesion of lower segments of the spinal cord to the spinal canal, such as myelomeningocele, which, due to the uneven development rate between the spinal canal and the spinal cord would lead to an increase in the lowering tension in the spinal cord and consequently cerebellar displacement through the foramen magnum. However, several studies have reported cases of patients who developed CM1 as an adult or in children who did not have any condition that would lead to spinal cord anchorage.[31–34] In addition, Barry et al.[35] studied fetuses and babies with lumbosacral myelomeningocele and concluded that there is a degree of tension resulting from the anchoring of the spinal cord caused by this condition. However, they stated that they do not believe that this tension extends to the upper portion of the spinal cord, having more influence on the portions close to the adhesion site, which would make the theory of traction improbable as a cause of tonsillar herniation. Even so, there is no study that evaluates spinal cord traction in vivo alone since the studies contain other variables that may function as a potential mechanism of tonsillar herniation, such as hydrocephalus, myelomeningocele, caudal displacement of the brainstem and cerebellum, hypoplasia of the posterior fossa of the skull, and enlargement of the foramen magnum.[36,37] In an experimental study using a fresh cadaveric model, Tubbs et al.[38] observed the movements of the spinal cord, brain stem, and cerebellar tonsils after applying tension to the conus medullaris. As a result, there was less than 1 mm of movement in the caudal portion of the brainstem and cervical spinal cord and no change in the position of the cerebellar tonsils, suggesting that the SFT is an unlikely method of reversing tonsillar herniation in cases of CM1.

Occult Tethered Cord Syndrome and Chiari Malformation Type 1

In the 1990s, studies theorized the possibility of TCS in individuals with the spinal cord in the normal position. These patients would have the clinical characteristics of a patient with TCS associated with a conus in the normal

position and a filum with structural changes in the MRI. The occurrence of this condition was first proposed by Warder and Oakes[2] in 1993, and, after that, several case series of surgical intervention were published with consequent clinical improvements in patients with this condition, which came to be called occult spinal cord syndrome.[17,39–43] However, with the cone in the normal position, the etiology of this condition began to be questioned since the theory of spinal cord traction assumed that due to a spinal cord attached through its filum terminale, there was a downward movement of the spinal cord. In their work, Milhorat et al.[36] provided data that would support the hypothesis of a tethered spinal cord even in patients with a cone in the normal position. In this study, it was observed that the filum terminale's width decreased steadily during growth and development, with most patients with a normal spinal cone position presenting positive filament traction tests at the time of surgery. Immediately after the SFT, there was a marked distraction of the divided extremities, in addition to improvement in the regional flow of the CSF.

Yet, based on the previously mentioned findings, the question arises as to how traction would act in the formation of cerebellar herniation in a patient in which the conus medullaris is in the normal position. Some studies have presented a series of cases reporting the outcome in patients with OTCS and tonsillar herniation who underwent SFT (►**Table 1**).

Main Studies on SFT in Patients with CM1 and OTCS

Royo-Salvador et al.[44] evaluated 20 patients who underwent their service between 1993 and 2003, among whom 8 had only scoliosis, 5 only syringomyelia, 2 only CM1, and 5 with a combination of these. Among the five mentioned, two had CM1 among their pathologies, totaling four patients with CM1. All were symptomatic and underwent SFT. Age ranged from 14 to 51 years old. In all patients, signs and symptoms disappeared or spontaneously improved after SFT. Three of the four with CM1 considered the surgery to be helpful, and the fourth patient's opinion was unknown. The author states that the improvement in symptoms in these patients must be associated with the disappearance of tension on the spinal cord after surgery. However, he admits that animal experiments have failed to produce CM1 by pulling the filum down. Recently, a systematic review[3] that included this study concluded that it has low methodological quality, and it is not possible to indicate SFT as a treatment for CM1, being, therefore, considered an experimental treatment.

Yuan Zhou et al.[45] published a case of a 14-year-old child admitted with intermittent pain and numbness in his right upper limb. He also had increased urinary frequency, pain in the neck and in the spine. Imaging exams revealed CM1 and syringomyelia and a medullary cone at the level of L1, a clinical and imaging diagnosis compatible with OTCS. He underwent SFT and had pain relief, improvement in the urodynamic test and urinary dysfunction, but there was no change in the position of the conus medullaris. After 1 year, he was readmitted with numbness in his right upper limb and right side of his back and occasionally pain in his waist. He performed posterior fossa decompression and evolved in the postoperative period with retraction of the cerebellar tonsils and 6 months later with improvement of symptoms. The cone remained in the normal position.

Glenn et al.[4] reported 17 cases (mean age 7.0 years) of CM1 associated with TCS in which they underwent SFT. Of these, 8 had the conus medullaris ending up to the lower portion of L2. The most common symptoms were headache and sensory disturbances of the lower extremities. Many of them were initially referred for posterior fossa decompression; however, due to a condition incompatible with classical CM1, further investigation was carried out. After SFT, 16 patients had the level of tonsillar herniation unchanged, and one had a rise in the cerebellar tonsils. According to the authors, all patients had some level of neurological improvement, and no worsening was detected. Therefore, they suggest that patients with CM1 and TCS who do not demonstrate classic symptoms of CM1 may benefit from SFT. The study does not specify how the clinical improvement of these patients was measured and does not correlate the level of improvement with the position of the conus medullaris, which makes it difficult to conclude whether there is a relationship between the clinical improvement and SFT in patients with CM1 associated with the conus medullaris in the normal position.

Selcuki et al.[46] presented 7 patients diagnosed with CM1 and TCS and who underwent SFT. As a diagnostic criterion for CM1, the clinical and radiological findings of tonsillar herniation were used, and for TCS, the clinical findings and the position of the conus medullaris below L2 and a fatty or thickened filum were used. Patients who had the conus in the normal position were referred for evaluation using the spinal somatosensory evoked potential (SSEP).T who had delayed or blocked nerve conduction were considered compatible with the diagnosis of TCS and underwent SFT before an approach to tonsillar herniation. All seven patients had CM1 and symptoms of TCS associated with a conus medullaris in a normal position and all underwent surgery. In the postoperative follow-up, there was no change in tonsillar herniation. On the other hand, there was a significant improvement in the symptoms of TCS and CM1, which leaves open the question of the contribution of tonsillar herniation alone to the symptoms in CM1.

Abel et al.[47] reported the case of a 3-year-old child with progressive imbalance and torticollis, in addition to anomalies compatible with Klippel-Feil syndrome and progressive scoliosis since birth. The CT showed the fusion of cervical vertebrae. Medical history also revealed a cleft palate and an extra-numerical finger in the hand, both surgically corrected, in addition to developmental delay. There were no changes in bladder or bowel function. A comparison of MRI images differing in three years revealed that CM1 signs were identified in the second MRI that were not identifiable in the first, suggesting that the pathology was not congenital, but acquired throughout life. In the MRI, fatty filum and conus medullaris were identified ending at the level of L2. The

Table 1 Patients with CM1 and TCS with conus medullaris in normal position who underwent SFT

	AGE/SEX	DIAGNOSIS	CMD	OUTCOME	POST OP MRI	FOLLOW-UP
Royo-Salvador et al., 2005[44]	Age range: 20–67 years; mean age: 38 years; sex: F (2) M (2)	CM1 (2)* CM1 + syringomyelia + scoliosis (2)**	L1-L2	40–100% of improvement	Rise of CMD (1)	4–11 years
Abel et al., 2006[47]	F, 3 years.	CM1 + fatty FT + Klippel- Feil Syndrome.	L2	Improvement of imbalance and stiff neck; Klippel- Feil and scoliosis stable;	CM1 and CMD unchanged	15 months
Milhorat et al., 2009[36]	CM1 + TCS: 31.5 ± 12.4 years LLCT + TCS: 31.0 ± 12.5 years 318 total (74 children, 244 adults)	CM1 (TH >= 8 mm) or LLCT (TH 0–7 mm) + TCS with NLCM	Above L2 in 60 (81%) children and 240 (98%) adults	Signs and symptoms: complete resolution in 27 (36%) children and 44 (18%) adults and improvement in 42 (57%) children and 159 (65%) adults.	Complete improvement or reduction of syringes in 76 (55%) of 138 patients; Complete resolution or improvement of scoliosis in 27 (38%) of 71 patients.	Children: mean of 14.8 ± 4.38 months Adults: mean of 16.5 ± 5.04 months
Glenn et al., 2015[4]	Mean age: 7 years	CM1 (17) TH mean of 10 mm	L1 (1) L1–2 (5) L2 (2) L2–3 (7) L3 (2)	100% had some level of improvement	TH unchanged (16) TH reduction (1)	Mean of 21.3 months
Selcuki et al., 2016[46]	Age range: 14-51 years; mean age: 30.7 years; sex: F (5) M (2)	Only CM1 (3) CM1 + syringomyelia (4)	Normal position	60–100% of improvement	TH unchanged	6–33 months
Zhou et al., 2017[45]	14, M	CM1 + Syringomyelia	L1	Clinical and image improvement	CM1 resolution, 6 months after surgery	2 years

Abbreviations: CM1, Chiari malformation type 1; CMD, conus medullaris; F, female; FT, filum terminale; LLCT, low-lying cerebellar tonsils; M, male; MRI, magnetic resonance imaging; NLCM, normal level *conus medullaris*; SFT, sectioning of the filum terminale; TCS, tethered cord syndrome; TH, tonsillar herniation.

*(cases 4 and 11 in the study)

** (cases 5 and 16 in the study)

authors mention that during SFT, a high-tension filum was observed by the surgeon, which was relieved after the sectioning. In the 15-month follow-up, the child showed improvement in his imbalance and torticollis and the other changes remained stable. Postoperative MRI showed no change in CM1 or in the position of the conus medullaris. The parameters used to assess clinical improvement were not mentioned. Milhorat et al.[36] in their retrospective study gathered a large cohort of 2,987 patients with CM1 (defined as tonsillar herniation greater than or equal to 5 mm below the FM) and 289 patients with small tonsillar herniation (LLCT - defined as tonsillar herniation between 0 and 4 mm below the FM) between January 2002 and July 2007. Tethered cord syndrome was present in 14% of the patients with CM1 and 63% of the patients with LLCT, which, according to the authors, reveals an association between these diseases. Of the total patients in the sample, 46% were referred due to previous surgical failure to treat CM1. Among the patients who had CM1 and TCS or LLCT and TCS, 318 underwent SFT. Among these, the age ranged from 12 months to 60 years (mean $\pm$ SD: 29.5 ± 4.1 years) and were divided into 2 groups —children (between 0–18 years old) and adults (18 years old or older). Previous surgical failure for CM1 was observed in 26% of children and 55% of adults. The authors mention that after CM1 surgery, the patients improved for several months, but the symptoms returned or worsened after that period and many had to undergo a second procedure, such as a review of posterior fossa decompression, cranioplasty, and a CSF shunt. These previous interventions can influence patients' response to SFT, and it is not possible to attribute the results only to the sectioning of the filum. All of them underwent a complete physical and neurological examination and a series of imaging tests. The diagnosis of TCS was made based on clinical and radiological criteria and included patients with a medullary cone ending below (19% of children and 7% of adults) and above (81% of children and 98% of adults) the lower edge of L2, with a statistically significant difference between adults and children in both cases. The postsurgical result was followed by a period ranging from 6 to 27 months (16.1 ± 4.6 months). A complete resolution of symptoms occurred more frequently in children than in adults (36% compared to 18%, $p < 0.001$, respectively), improved in 57% of children and 65% of adults, did not change in 7% of children and in 16% of adults, and worsened in 1% of adults. According to the authors, the low rate of complete symptom resolution was due to herniations of the rhombencephalon, the presence of syringomyelia, scoliosis, and interference from previous surgical treatments. Cranial migration of the conus medullaris was detected at an average distance of 5.1 mm ($p < 0.001$), being greater in children than in adults, in addition to a reduction in tonsillar herniation (average of 3.8 mm, $p < 0.001$) and a reduction in distance of the fourth ventricle below the Twining line (average of 2.6 mm, $p < 0.01$). The increase in medullary height, the reduction of tonsillar herniation, and the distance from the fourth ventricle to the Twining line was taken as evidence of cranial migration of the brainstem and cerebellum. The authors mentioned that after SFT of these patients, it was possible to notice a rapid removal of the ends of the filum, suggesting that they were under strong traction, and, through the transdural Doppler ultrasound, it was possible to detect a statistically significant increase in the CSF flow speed.

Conclusion

The evaluation of the benefits of SFT for the correction of the symptoms of CM1 is extremely difficult, since to date it is supported only by observational studies. These studies also have important limitations, which include: a lack of objective parameters for improvement, a relatively short follow-up in most of the studies, a lack of homogeneity in the samples (patients with several other pathologies and previous surgeries) that may interfere with the surgical results, and the absence of a clear correlation between the clinical improvements reported in the studies and the patients' anatomy, which often did not change. Nevertheless, these studies are important milestones of these pathologies. Basic science studies looking for associated molecular mechanisms and randomized controlled clinical trials are needed for further understanding.

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Acquisition of data: J.N.P.O.B., P.A.D., P.B.B.J.,
Analysis and interpretation of data: J.N.P.O.B., P.A.D., P.B.B.J.
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Conflict of Interests
The authors have no conflict of interests to declare.

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