**GUIDELINES IN FOCUS**

**Chiari malformation Type I - effect of the section of the filum terminale**

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**METHODOLOGY FOR EVIDENCE COLLECTION**

This guideline followed the standards for a systematic review with evidence collected based on the Evidence-Based Medicine movement. We used the structured method to formulate the question, synthesized by the P.I.O. acronym, in which: P - corresponds to patients diagnosed with Chiari malformation type I; I - section of the filum terminale, O - robust measures of relevant clinical prognosis. The clinical question was: “What is the effect of the section of the filum terminale in the treatment of Chiari malformation type I symptoms?” From this structured question, we identified the descriptors used to search for evidence in the Medline-Pubmed databases. A total of 21 abstracts and titles were considered eligible for analysis, in addition to 10 studies obtained through cross-references. After applying the eligibility criteria (inclusion and exclusion), only two studies were included to answer the structured question (Annex 1).

**CLINICAL QUESTION**

Does the section of the filum terminale improve the functional prognosis of patients with Chiari malformation type I?

**GRADE FOR RECOMMENDATION AND LEVEL OF EVIDENCE**

A: Experimental or observational studies of higher consistency.
B: Experimental or observational studies of lower consistency.

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The Guidelines Project, an initiative of the Brazilian Medical Association, aims to combine information from the medical field in order to standardize producers to assist the reasoning and decision-making of doctors. The information provided through this project must be assessed and criticized by the physician responsible for the conduct that will be adopted, depending on the conditions and the clinical status of each patient.
OBJECTIVE
This guideline aims to analyze the effect of the section of the filum terminale in the treatment of Chiari malformation type I symptoms.

CONFLICT OF INTEREST
There is no conflict of interest related to this review that can be declared by any of the authors.

INTRODUCTION
Chiari malformation type I (CM) is a congenital dysplasia of the posterior cranial fossa which results in herniations of the cerebellar tonsils through the foramen magnum (Beijani, 2001). The clinical scenario may involve headache, which worsens with the Valsalva maneuver, dizziness, tinnitus, dysphagia, dysphonia, caused by compression of the lower cranial nerves, in addition to the impairment of sensory and motor tracts, which manifests as unbalance, ataxic gait, paresthesias, or paresis. Magnetic resonance imaging (MRI) is the gold standard to confirm the diagnosis, demonstrating the absence of the cisterna magna due to tonsillar herniation (McRae, 1960; Nishikawa, 1997).

The most widely accepted theory to explain the physiopathology of CM is based on the disproportion between the continent, represented by the posterior cranial fossa, delimited by the clivus, the petrous portion of the temporal bone, occipital bone, and cerebellar tentorium, and the content, comprising the cerebellum, brainstem, cranial nerves (III to XII), and vascular structures. Thus, the cerebellar tonsils migrate caudally and impact the foramen magnum, compromising the flow of cerebrospinal fluid between the cranium and spinal canal (Schady W. et al., 1987; Nishikawa M. et al., 1997; Karagöz F. et al., 2002; Milhorat TH. et al., 2010).

The widely accepted treatment of CM is the decompression of the posterior fossa through suboccipital craniectomies, opening the foramen magnum, with or without magnification of the dura mater, associated with resection of the posterior arch of the atlas and, more rarely, of the axis lamina to decompress the cerebellar tonsils and restore the cerebrospinal fluid flow through the foramen magnum (Oliveira et al., 2018; Zhao et al., 2016; Steinmetz et al., 2003).

However, some authors have proposed the theory that the caudal migration of the cerebellar tonsils occurs due to the caudal traction of the spinal cord and, consequently, of the brainstem and cerebellum, resulting in occult tethered cord syndrome (Tubbs et al., 2004; Wehby et al., 2004). Therefore, the section of the filum terminale was proposed as a therapeutic approach for CM. According to the proponents of this theory, this technique, which is already used on the treatment of filum terminale lipomas and other spinal dysraphisms, could improve CM symptoms, with lower risks of complications than the classical technique (Royo-Salvador, 1997; Tubbs et al., 2004; Wehby et al., 2004; Royo-Salvador et al., 2005).

RESULTS OF THE SELECTED EVIDENCE
Does the section of the filum terminale improve the functional prognosis of patients with Chiari malformation type I?

This systematic review was based on two case series published by the same group of authors (Royo-Salvador, 1997; Royo-Salvador et al., 2005). It was not possible to define if the cases of the first study were included in the second. Thus, both studies were evaluated. The methodological qualities of both studies, according to the criteria proposed by MINORS, were low (3 and 4, respectively, considering the 16 points) (Slim et al., 2003). These are retrospective studies with small samples of non-consecutive patients, without a standardized analysis of outcomes, with data collection carried out by the surgical team, with a follow-up time not clearly defined.

Therefore, considering the scientific literature available, it is not possible to determine if the section of the filum terminale improves the functional prognosis of patients with Chiari malformation type I.

SYNTHESIS OF EVIDENCE
The theory that presents occult tethered cord as the genesis of CM, as well as the section of the filum terminale as the treatment for this condition, is controversial (Massimi et al., 2011). In addition, the fact that the classically established treatment for this disease, which consists in the decompression of the posterior fossa, demonstrates clinical outcomes that are satisfactory...
and reproducible in several centers reinforces as the
physiopathology of CM the theory of reduced volume
of the posterior fossa during its formation in the embry-
onic stage (Zhao et al., 2016; Oliveira et al., 2018; Beijani
G, McRae, 1960, Nishikawa et al., 1997, Karagöz F. et
al., 2002, Pang et al., 2011).

The analysis of the 31 excluded studies obtained in
the initial search (21) and from cross-references (10),
resulted in the exclusion of 29. These studies included
patients with a diagnosis of tethered spinal cord or other
spinal dysraphisms, case reports or review studies, in
addition to the studies in which it was not possible to
specify whether the authors treated patients with Chiari
malformation type I or Type II (Millorat et al., 2010).

Both studies included present evidence level 4
(case series of low quality according to the criteria
proposed by Oxford) (available on Http://www.cebm.
net/oxford-centre-evidence-based-medicine-levelsevi-
dence-march-2009; Royo-Salvador, 1997; Royo-Salva-
dor et al., 2005).

**RECOMMENDATION**

It is not possible to recommend the section of the *filum terminale* in the treatment of Chiari malforma-
tion type I based on the findings of this system-
atic review.

The section of the *filum terminale* for treating
Chiari malformation can be considered an experi-
mental treatment.

**ANNEX I**

**Structured question**

P - patients with Chiari malformation type I
I - section of the *filum terminale*
O - robust measures of clinical prognosis

**Methodology for Evidence Search**

PubMed-Medline

(arnold chiari malformation OR (chiari 1) OR (type
1 chiari)) AND (filum terminal*)

First batch of studies retrieved: 25 titles of original
studies

**Studies retrieved**

The evidence used was retrieved by the following
steps: elaboration of the clinical question, structuring
of the question, search for evidence, presentation of
results, and recommendations.

We reviewed articles from the MEDLINE (PubMed)
databases, with no time limit.

The studies retrieved during the search were
initially evaluated based on their titles, then their
abstracts, and, finally, the studies selected were evalu-
ated in full. Two authors were responsible for the
independent evaluation of the results and all disagree-
ments were resolved through discussions between
them (JWD and FO). Cross-references obtained from
the primary articles were evaluated.

The search was conducted on 1st January 2019 and
21 papers were obtained, in addition to 10 obtained
through cross-references, which had their abstracts
evaluated. Of this total of 31 papers, 13 were excluded
because their content was not related to the object of
study or they were case reports (Figure 1). Among the
18 papers evaluated in full, 16 were excluded for vari-
ous reasons (Table 1). Only two studies were included
for the final analysis.

**Inclusion criteria**

1. According to study designs

The search primarily targeted randomized clinical
trials; in their absence, non-randomized clinical trials,
controlled comparative studies, and, finally, a case
series, successively.

2. Language

We included articles in English, Spanish,
and Portuguese.

3. According to publication

Only studies with texts available in its entirety
were considered for critical evaluation.

**Method for critical evaluation**

For the review protocol, the PRISMA flowchart
(REF) was used to describe the flow of tracking, eli-
gibility, and final selection of papers (Figure 1).

**Extraction of results**

The results extracted are described in Annex II
and the recommendations were drawn based on their
discussion according to the Oxford grade for recom-
mandation (REF).

**Quality assessment**

The methodological quality was assessed with the
aid of the MINORS (Methodological Items for Non-Ran-
domized Studies) instrument; Slim et al., 2003).
**APPENDIX II**

**FIGURE 1. FLOWCHART OF THE SEARCH MECHANISM ACCORDING TO THE PRISMA RECOMMENDATIONS FOR SYSTEMATIC REVIEWS (SLIM ET AL., 2003; MOHER ET AL., 2009). PRISMA 2009 FLOW DIAGRAM**

**TABLE 1. EXTRACTION OF DATA ON THE SECTION OF THE FILUM TERMINALE FOR TREATING CHIARI MALFORMATION TYPE I**

| Study/ Type of study | Patients | Follow-up/ Prognosis* | Conclusion |
|----------------------|----------|-----------------------|------------|
| Royo-Salvador, et al./ 1997/ Case series | N = 5  
Scoliosis 1 (20%); Syringomyelia 2 (40%); Chiari type I 1 (20%), Associated 1 (20%)  
Mean age: 33.8 years | Improvement in 5 patients (100%)  
Follo-up not informe | SFT is a useful strategy in the treatment of scoliosis, syringomyelia, and Chiari malformation type I |
| Royo-Salvador, et al./ 2005/ Case series | N = 20  
Scoliosis 8 (40%); Syringomyelia 5 (25%), Chiari type I 2 (10%), Associated 5 (25%)  
Mean age: 33.5 years | Improvement in 9 (45%), Without improvement in 7 (35%), Unknown in 4 (20%)  
Follow-up of 4 months to 11 years (mean 4.8 years) | SFT is a useful strategy in the treatment of scoliosis, syringomyelia, and Chiari malformation type I |

*only intervention carried out; there was no control group for comparison
**TABLE 2. MINORS (METHODOLOGICAL ITEMS FOR NON-RANDOMIZED STUDIES) OF THE PAPERS INCLUDED IN THIS SYSTEMATIC REVIEW (SLIM ET AL., 2003)**

| Study / Items                                                                 | Royo-Salvador (1997) | Items Score | Royo-Salvador (2005) | Items Score |
|-------------------------------------------------------------------------------|----------------------|-------------|----------------------|-------------|
| Objective clearly established                                                  | Yes. The objective was to report (evaluate) the results of cases operated with a surgical technique (an intervention) | 2           | Yes. The objective was to report (evaluate) the results of cases operated with a surgical technique (an intervention) | 2           |
| Consecutive inclusion of patients                                             | No. Non-consecutive patients | 0           | No. Non-consecutive patients | 0           |
| Prospective data collection                                                   | No. Retrospective collection | 0           | No. Retrospective collection | 0           |
| Appropriate outcomes for the objective of the study                          | No. The author described the clinical improvements of each patient, without standardization of data collection | 0           | No. The author described the percentages of clinical improvement for each patient, without standardization of data collection | 1           |
| Unbiased analysis of the study outcome                                        | No. Although it was not described, it is suggested that the surgical team collected the data | 0           | No. Although it was not described, it is suggested that the surgical team collected the data | 0           |
| Appropriate follow-up time for the objective of the study                    | Uncertain. There is no description of long-term follow-up. | 1           | Uncertain. Patients operated between 1993 and 2013. Table 1 suggests that the formal outcomes were collected in September and October 2014. | 1           |
| Prospective calculation of study sample size                                  | No. This is a case series with a small sample of patients. Only patients 3 and 4 were suggestive of or compatible with Chiari malformation type I | 0           | No. This is a case series with a small sample of patients. Only patients 4, 5, and 11 had Chiari malformation type I | 0           |
| Total score                                                                  | 3'                    |             | 4'                   |             |

*The maximum MINORS score for non-randomized studies is 16 points. Therefore, the methodological quality of both studies selected is low.

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