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

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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.

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.

C: Uncontrolled studies/case reports.

D: Opinion deprived of critical evaluation, based on consensus, physiological studies, or animal models.

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 embryonic 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-levelsevidence-march-2009; Royo-Salvador, 1997; Royo-Salvador et al., 2005).

RECOMMENDATION

It is not possible to recommend the section of the *filum terminale* in the treatment of Chiari malformation type I based on the findings of this systematic review.

The section of the *filum terminale* for treating Chiari malformation can be considered an experimental 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 evaluated in full. Two authors were responsible for the independent evaluation of the results and all disagreements 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 various reasons (Table 1). Only two studies were included for the final analysis.

Inclusion criteria

4.1. 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.

4.2. Language

We included articles in English, Spanish, and Portuguese.

4.3. 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, eligibility, 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 recommendation (REF).

Quality assessment

The methodological quality was assessed with the aid of the MINORS (Methodological Items for Non-Randomized 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

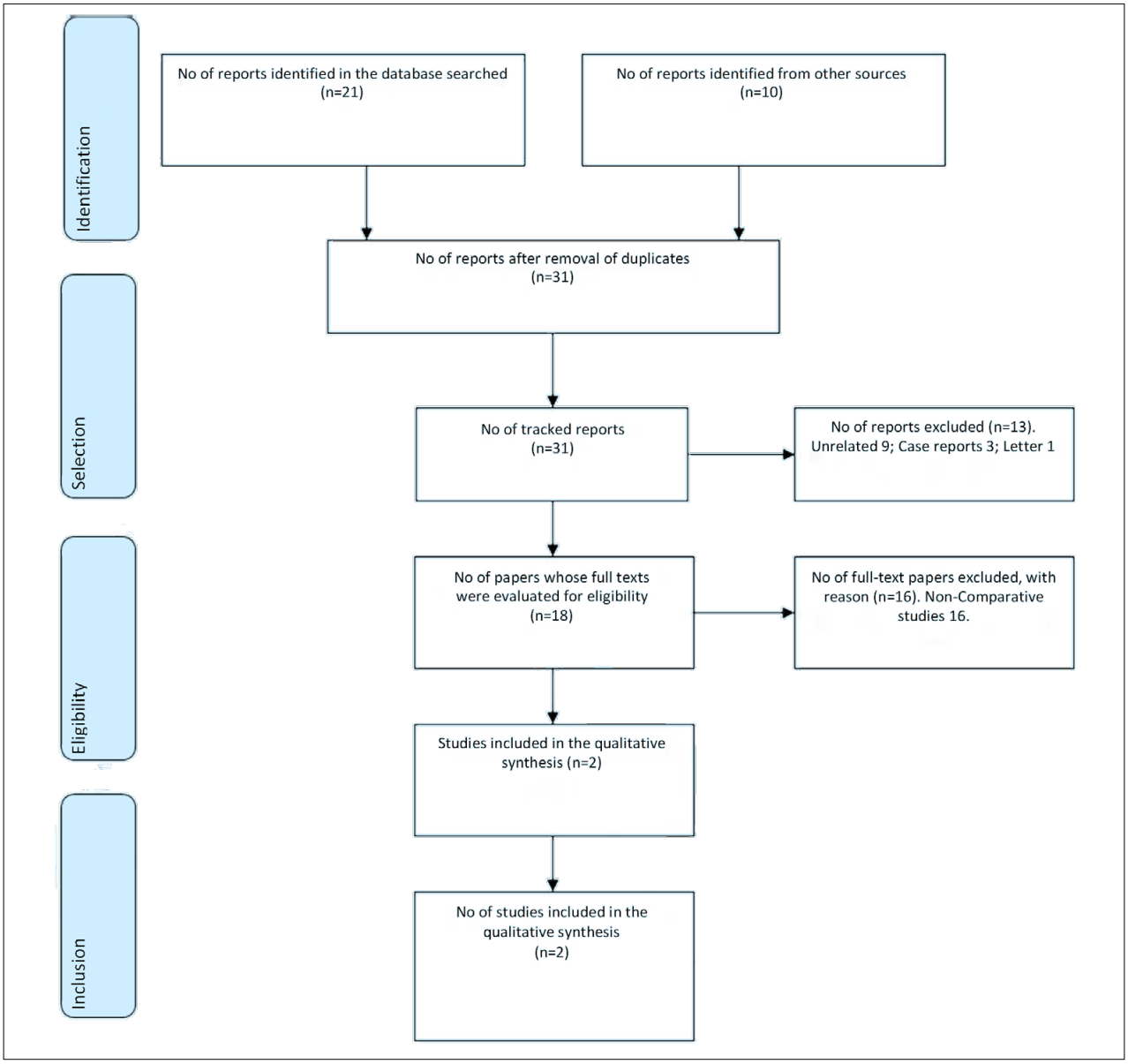

TABLE 1. EXTRACTION OF DATA ON THE SECTION OF THE FILUM TERMINALE* FOR TREATING CHIARI MALFORMATION TYPE I

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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