Management of Chiari I deformity in Children and Adolescents: A report from the Consensus Taskforce of the Brazilian Society of Pediatric Neurosurgery

Avaliação e manejo da deformidade de Chiari I em crianças e adolescentes: Recomendações e resultados da Reunião de Consenso sobre Controvérsias da Sociedade Brasileira de Neurocirurgia Pediátrica

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Abstract

Keywords
► chiari I deformity
► craniocervical junction
► filum terminale
► guidelines
► hindbrain hernia
► tonsillar ectopia

Much controversy remains on the current management of Chiari I deformity (CID) in children, with many clinical, surgical and ethic-legal implications. The Brazilian Society of Pediatric Neurosurgery (SBNPed, in the Portuguese acronym) has put together a panel of experts to analyze updated published data on the medical literature about this matter and come up with several recommendations for pediatric neurosurgeons and allied health professionals when dealing with CID. Their conclusions are reported herein, along with the respective scientific background.

Resumo

Palavras-chave
► deformidade de Chiari I
► diretrizes
► ectopia
► herniação tonsilar
► phyllo terminal
► junção craniocervical

Atualmente, ainda persiste muita controvérsia sobre o manejo da deformidade de Chiari I (CID) em crianças, com diversas implicações clínicas, cirúrgicas e ético-legais. Desta forma, a Sociedade Brasileira de Neurocirurgia Pediátrica (SBNPed) reuniu um painel de especialistas para analisar dados atualizados publicados na literatura médica sobre essa entidade clínica e formulou uma série de recomendações, a fim de padronizar as condutas e auxiliar a tomada de decisões clínicas de neurocirurgiões pediátricos e demais profissionais de saúde que tratam de crianças com CID. Suas conclusões são relatadas aqui, juntamente com o respectivo respaldo científico.

Introduction

Chiari type I deformity (CID) is relatively common in children, and the ever-increasing availability of high-definition diagnostic imaging has resulted in a growing number of referrals for this condition to pediatric neurosurgeons.[1] Much controversy still remains on the management of Chiari type I deformity in children, especially in asymptomatic or oligosymptomatic patients. To overcome this, the Brazilian Society of Pediatric Neurosurgery (SBNPed, in the Portuguese acronym) has organized, in August 2019, a consensus meeting for the discussion of CID, held in the city of Londrina, state of Paraná, Brazil, to analyze the most updated medical literature regarding this topic, gather the opinions of national and international experts and standardize evaluation and management strategies. This consensus report is addressed to pediatric and general neurosurgeons, pediatricians, allied professionals, health managers and decision makers who might get involved in the treatment of this condition.

Methods

A panel composed of members of the Board of Directors and Scientific Committee of the SBNPed put together the scientific program of the consensus meeting, which comprised the main clinical and surgical issues concerning the management of CID patients. Brazilian and foreign specialists were invited to present data available in the literature and to provide a critical summary and analysis of such information. All of the authors were asked to strictly follow guidelines of the Oxford Centre for Evidence-based Medicine, grading published studies in accordance with their levels of evidence and grades of recommendation.[2,3]

All of the statements described herein have taken into consideration this hierarchy, and priority has been given to the highest levels of data quality available. Data was obtained through standard clinical searches in the Medline, Cochrane, Scielo and LILACS databases, with correspondent MeSH terms. By the end of the meeting, an algorithm for the management of CID was formulated (in strict accordance with the scientific data discussed) and approved by all members of the consensus taskforce (►**Fig. 1**). Also, a letter

to the editor of an international pediatric neurosurgery publication has been sent,[4] on the grounds that such journal had recently published a full edition solely about CID,[5] and that this consensus report would further expand the discussion in the subject and underline the standards recommended by the SBNPed.

Clinical Questions

1) What is the definition and best nomenclature of CID?

Ever since the first descriptions of this clinical entity by Hans Chiari himself in 1891[6], it has been referred to as "malformation." Nevertheless, this term does not seem to be directly applicable, for there are many cases without a proper malformation, with normal neurovascular structures of the posterior fossa,[6] and a reappraisal of this terminology has been published recently, as it relates to an acquired rather than developmental pathology.[5,7] Therefore, for the pathophysiological aspects discussed below, the consensus taskforce has reached a decision to call it Chiari I deformity (CID), and this denomination will be used henceforth.

Regarding its definition, CID is usually characterized by a descent of the cerebellar tonsils below the level of the foramen magnum; the extent of this descent is what differs in the literature, with authors considering it significant from 3 to 8 mm.[8,9] Furthermore, the description of the so-called Chiari 0 and Chiari 1.5, while referring to recognizable radiological entities, has added further confusion rather than clarifying the situation.[5,7] Analyzing all these papers, the consensus group has defined CID as "a tonsillar ectopia of 5 (five) millimeters or more below the level of the foramen magnum."

2) What is the physiopathology of CID?

Several authors have addressed this issue.[10–15] The prevailing idea that CID children have smaller posterior fossa volumes has been reported,[15] but this finding could not be replicated by many authors,[10–12] except in children with craniosynostosis, who form a distinct subgroup of patients.[13] Milhorat et al,[14] thus, have subdivided the pathophysiological features of CID into five categories: cranial constriction, spinal cord tethering, cranial settling, intracranial hypertension and intraspinal hypotension. There also seems to be an overlapping among these causative factors in any given patient.[15]
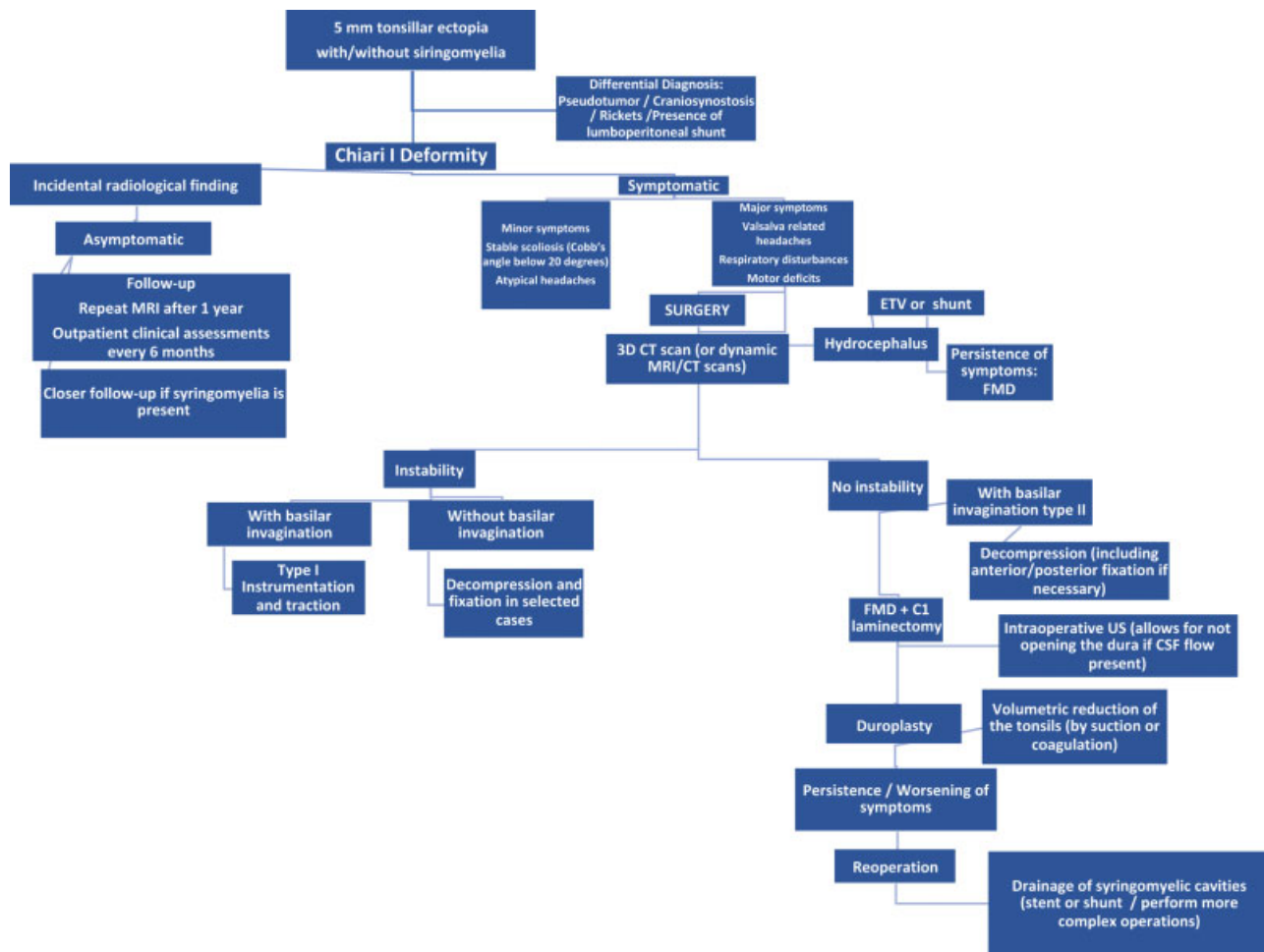
Fig. 1 Algorithm for the Management of Chiari I Deformity in Children according to the recommendations of the Consensus Taskforce of the Brazilian Society of Pediatric Neurosurgery.

Poretti et al[15] have published a comprehensive review of the pathological aspects of CID in children, dividing the possible mechanisms of the development of CID in children into three groups: 1) Primary CID, produced by abnormal growth or mis-segmentation of the craniocervical junction (CVJ); 2) Secondary CID, occurring in patients with abnormal brain expansion (megalencephaly, macrocerebellum) or reduction of the posterior fossa (basilar impression, craniosynostosis); and 3) tonsillar herniation, represented by acquired causes (spontaneous or idiopathic intracranial hypertension, venous congestion due to vascular malformations, expanding intracranial masses).

3) What is the natural history of CID in asymptomatic patients?

In 2008, Novegno et al reported on the natural history of 22 untreated patients with CID: 16 patients (72%) remained unchanged, and only 3 cases had clinical and or/radiological progression after a mean follow-up of 5.8 years.[16] Strahle et al, in 2011, studied 142 patients, of whom 133 (90.5%) did not require treatment (mean follow-up: 3.8 years).[17] Volpon Santos et al, in 2012, reported on a series of 62 CID children with a longer follow-up (10 years), revealing stability of symptoms with conservative treatment in 72.6%, spontaneous regression in 2.4%, and progression/worsening of symptoms requiring

surgery in 27.4%.[18] Similarly, Pomeraniec et al studied 95 CID pediatric patients and concluded that 92.9% of the patients managed conservatively did not experience clinical or radiological progression, whereas 41.7% of those who had presented with symptoms improved during follow-up.[19] Lastly, Langridge et al have recently performed a systematic review of the natural history and conservative management of adult and pediatric patients with CID. Fifteen papers were included in their meta-analysis, allowing for these authors to conclude that the natural history of mild symptomatic and asymptomatic CID is relatively benign and nonprogressive, and that it is reasonable to observe asymptomatic patients and subjects with mild symptoms even in the presence of significant tonsillar descent or syringomyelia.[9]

The consensus panel acknowledged that asymptomatic patients with incidentally found tonsillar ectopia can be followed-up clinically; a magnetic resonance imaging (MRI) scan should be obtained after 1 year, and clinical assessments should be performed every 6 months. Serial MRI scans are not required as long as the patient remains clinically stable; patients with syringomyelia should be observed more closely.

4) What are the clinical indications for surgery in patients with CID?

The consensus taskforce has reviewed clinical and radiological indications of surgical treatment for children with CID. For this purpose, clinical manifestations were divided into major and minor, in accordance with several published studies.[1,20–24] Major clinical symptoms and signs are considered strong indicators for surgery, whereas minor ones should be accompanied by other indicative signs and dealt with on an individual basis.

Major clinical indications include occipital headaches, most often exertional and related to coughing and Valsalva maneuvers, respiratory disturbances (snoring, gagging, sleep apnea), truncal ataxia and corticospinal tract dysfunction (upper and/or lower limb weakness, hyperactive tendon reflexes, Babinski sign, clonus, unilateral and/or bilateral). Minor symptoms and signs listed were atypical headaches (diffuse, migraine-like) and stable scoliosis, especially those with Cobb angle under 20°.

From a radiological standpoint, as indications for surgery, along with the presence of a $\geq$ 5 mm tonsillar ectopia,[25,26] the following findings could help in the surgical decision[21]: presence of syrinx, presence of hydrocephalus, and cerebrospinal fluid (CSF) flow disturbance around the foramen magnum (especially on Cine-MRI scans and flow-related sequences).

5) Which operative techniques and adjuncts should be adopted for the surgical management of CID patients?

Traditionally, the standard surgical modality for the treatment of pediatric CID is a posterior fossa craniectomy for foramen magnum decompression (FMD); several studies have confirmed its utility and good results.[1,22,25–27] It has been used by the majority of pediatric neurosurgeons with only a few distinct technical nuances, which may give rise to some controversy and are discussed herein.

Caldarelli et al have analyzed the required extent of bony resection in 30 pediatric patients and found out that a limited midline suboccipital craniectomy 2.0 cm long and 2.5 cm wide with C-1 laminectomy is sufficient for efficient decompression of the FM, yielding good results in 28 (93%) patients and requiring reoperation in only 2 (7%).[28] It is also important to highlight that, in patients with craniosynostosis, the treatment should be aimed at the primary pathology, as stated by many authors.[13,29–31]

The need for dural opening and duraplasty has been initially discussed by Durham et al in 2008,[32] who concluded that FMD with duraplasty carries a lower risk of reoperation than FMD alone, but a greater risk for CSF–related complications. They also state that the available data on the surgical treatment of CID in children do not allow identification of particular patients who may benefit from the less invasive surgical technique of FMD. In 2011, Hankinson et al[33] performed a systematic review of this subject, concluding that, at that time, there was no level I or IIa evidence comparing FMD with and without duraplasty; the notions that FMD with duraplasty has a lower rate of reoperation and that bony decompression alone has a lower rate of CSF-related complications are both based on IIb/B evidence. In 2018, Lin et al[34] have performed another systematic review and meta-analysis, including 13 articles published until then (and also including adult subjects for a total 3,481 patients in the meta-analysis). Their results showed that duraplasty is an optimal surgical strategy, leading to higher clinical improvement and lower recurrence rate, especially in patients with syringomyelia. However, the authors pointed out that, in patients without syringomyelia, FMD without duraplasty might be the technique of choice, providing similar clinical improvement at lower costs. Lastly, Lu et al[35] have performed a meta-analysis of 3,455 pediatric patients, 1,492 (43%) with and 1,963 (57%) without duraplasty. The authors concluded that the addition of duraplasty to FM decompression in children with CID may improve surgical and performance outcomes, particularly regarding parameters of overall clinical improvement, length of stay, and postoperative complications.

Narenthiran et al have published their experience in 19 patients using intraoperative ultrasound to assess intraoperatively the indication for duraplasty.[36] They did not open the dura if there was adequate cerebellar tonsillar and/or CSF pulsation following suboccipital craniectomy. Eight patients underwent dural decompression and 11 patients had bony decompression only. Clinical outcomes and complication rates were very similar between these groups. In line with this, the role of intraoperative neurophysiological monitoring (IOM) in the surgical treatment of CID has been also described; however, its clinical implications are yet to be defined.[37]

6) What are the best treatment options when CID patients also have syringomyelia?

Some controversy remains whether syringomyelia in patients with CID is a strict surgical indication, since the medical literature has several descriptions of spontaneous resolution of the syrinx in such cases or at least stability/absence of progression, even in cases of large syringomyelic cavities.[38,39] Magge et al studied 48 children with an idiopathic syrinx and found out that 91% of them remained clinically asymptomatic, stable or improved over a mean follow-up of 23.8 months.[40] In those who had follow-up imaging, 87.5% remained radiologically stable or the syrinx reduced over time, with no apparent correlation between changes in size of the syrinx and evolution of symptoms.

Nevertheless, Wetjen et al[41] recommended surgical treatment by means of FMD whenever syringomyelia is associated with CID in children. Their study revealed that resolution of syringomyelia could be achieved after FMD in 86% of the cases. In unfortunate cases where symptomatic syringomyelia does not resolve after FMD, shunting might be an alternative solution, with reported good results for both syringoarachnoid and syringopleural shunt techniques.[42,43]

7) When should patients with CID be reoperated?

Hidalgo et al,[44] reviewing their experience with 105 consecutive children who underwent surgical decompression of symptomatic CID with duraplasty, found that symptoms had resolved by the time of discharge from the hospital in the majority (57%) of children, and syrinx had resolved or decreased in two-thirds of the patients by 3 months of follow-up. By 6 months, headaches had resolved in all cases, and syrinx had resolved or decreased in 79% of the cases.

In the cases where symptoms did not subside, indications for reoperation included persistence of syringomyelia,[20–22]

persistent cranial nerves palsies[24,25] and other neurological symptoms. Children < 3 years old appear to have worse outcomes.[25] Furthermore, Goel et al[45] have published their experience with atlantoaxial fixation for Chiari 1 deformity in the pediatric age group (33 cases), reporting gratifying and sustained clinical improvement in all patients.

The conclusion of the consensus taskforce is that reoperation should be considered particularly in cases of persistent symptoms and/or of neurological deficits, especially when no CSF flow or a compressed brainstem are still visible on postoperative scans. Failure of syringomyelia remission, along with clinical features, is another strong indicator for reoperation. Nonetheless, another agreement of the taskforce important to highlight is that, if clinical improvement has been achieved, a persistent syringomyelia can be closely followed-up. Volumetric reduction of the tonsils is another possibility if the bony and dural decompression are deemed satisfactory. The further need for fixation is addressed below.

8) Is there any evidence to support sectioning of the filum terminale as a first line therapy for CID?

To the best of our knowledge, only two peer-reviewed clinical studies have dealt with this topic so far. Royo-Salvador et al[46] published in 2005 the results of 20 patients with CID treated through section of the filum terminale, reporting clinical improvement in all cases. A second study, published by Milhorat et al[47] in 2009, examined 318 individuals with spinal cord and tonsillar herniation who underwent section of the filum, compared with matched controls and subjects with Chiari without spinal cord tethering, showing clinical and/or radiological improvement in 85% of them. However, it should be emphasized that these were patients with the diagnosis of tethered spinal cord, who would have usually undergone section of the filum as first-line treatment anyway. Therefore, the heterogeneity of the studied subjects with presentation of mixed pathologies precludes reliable statistical analysis and further recommendations based on these results.

From an experimental standpoint, a project conducted by Tubbs et al[48] studied 12 adult cadavers submitted to the application of distal tension (75 N) to the spinal cord, with simultaneous observation of the cervical spinal cord, the brainstem and the rhombencephalon and their relation to the foramen magnum after occipital craniectomy and removal of the posterior arch of C1. These authors did not observe any movement of the cerebellar tonsils and only 2 to 3 mm caudal descent of the brainstem and cervical spinal cord, concluding that, in the cadaveric model, caudal fixation of the distal spinal cord is unlikely to result in inferior displacement of the cerebellar tonsils, and therefore, transection of the filum terminale is likewise unlikely to reverse tonsillar ectopia.

Lastly, Massimi et al[49] stated that there is still no convincing evidence on the association between CID and tethered spinal cord. Nevertheless, the authors concluded that section of the filum terminale may play a role in a small selected subgroup of patients with CID with poorly symptomatic tonsillar herniation, normal posterior fossa volume and evident symptoms of a tethered spinal cord.

As demonstrated, the consensus taskforce has performed a thorough and insightful analysis of the medical literature on the subject and found that there is no strong scientific evidence to support this therapeutic option, which remains, to our knowledge, experimental and with no scientific background. Therefore, the taskforce understands that only exceptional cases can benefit from this strategy and, thus, there is no generally accepted surgical indication of section of the filum terminale for the treatment of CID in children.

9) When should CID patients undergo craniocervical fusion and instrumentation? What are the current recommended approaches? How many levels should be included?

The work of Goel[50–52] has introduced a new paradigm for treating CID; the author has recently suggested that C1–2 fixations should be performed for all cases, considering that tonsillar herniation is secondary (and a protective and compensatory mechanism) to the subtle or gross instability that seems to be present in cases of CID. Although the outcome has been reported as positive, this method of treatment has not been generally accepted yet as a standard mode of treatment for CID.[53]

Brockmeyer et al reviewed the medical literature in 2011 and concluded that C1-C2 fixation is indicated in complex CID patients with basilar invagination (BI) associated with C1-C2 instability.[54,55] It also depends on the type of BI: type I requires instrumentation and fusion plus traction; in BI type II, fixation is recommended for selected cases, and some cases are to be decompressed only. Posterior C1-C2 fusion is also recommended in cases in which wide bone resections are necessary and should include as few levels as possible.[56,57]

Menezes, in 2012, analyzed his large series (> 850 procedures) of posterior instrumentation for CID children and reported a 98% rate of successful fusion.[58] The author recommended that rigid fixation with screws and rods could be performed in patients older > 6 years old, and in younger children rib grafts should be harvested. Similar results were reported by Mackel et al[59] and Kim et al.[60] Kennedy et al have also demonstrated that most young children undergoing atlatoaxial and occipitocervical fusion with rigid internal fixation continue to have good cervical alignment and continued growth within the fused levels during a prolonged follow-up period.[61]

10) Is there any evidence to support a genetic component in the genesis of CID?

Only a few studies have addressed this issue,[62–66] with most data coming from familiar clusters of CID and syringomyelia patients.[60,61] Nevertheless, a genetic component has been shown particularly in CID cases associated with connective tissue diseases[64] and in some cases had mutations in the WNT pathways.[65] Also, a positive familial history is present in 12% of patients with newly-diagnosed CID.[64] Urbizu et al have found single nucleotide polymorphisms in 14 genes (*CDX1*, *FLT1*, *RARG*, *NKD2*, *MSGN1*, *RBPJ1*, *FGFR1*, *RDH10*, *NOG*, *RARA*, *LFNG*, *KDR*, *ALDH1A2*, and *BMPR1A*), suggesting that common variants in genes involved in somatogenesis and fetal vascular development may confer susceptibility to CID.[66]

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