Child's Nervous System (2019) 35:1905–1909
https://doi.org/10.1007/s00381-019-04162-8

SPECIAL ANNUAL ISSUE

Management: opinions from different centers—the Istituto Giannina Gaslini experience

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Received: 17 March 2019 / Accepted: 16 April 2019 / Published online: 9 May 2019

Abstract

Purpose Describe presentation and management of Chiari type 1 malformation. We report our surgical case series proposing a decision making scheme for helping surgeons decide which surgical procedure to perform and when.

Methods We retrospectively examined a series of surgically treated patients with Chiari type 1 malformation. Treatment of associated anomalies, surgical complications, and need for reintervention for insufficient decompression at first surgery are discussed.

Results A total of 172 patients have been surgically treated for Chiari type 1 malformation at the Neurosurgery Unit of IRCCS Giannina Gaslini Children Hospital of Genoa, Italy, in a period between 2006 and 2017. The first treatment addressing Chiari type 1 malformation was bone and ligamentous decompression alone in 104 patients (65%), associated with dural delamination in 3 patients (1.9%) and associated with duraplasty with autologous graft in 53 patients (33.1%). Postoperative complications occurred in 5 patients (2.9%). Reintervention for insufficient decompression at follow-up was needed in 6 patients (3.5%).

Conclusions Surgical decompression of the posterior cranial fossa (PCF) is indicated in symptomatic patients while asymptomatic patients must be followed in a wait and see fashion. Different types of surgical decompression of different invasiveness have been proposed from only bone and ligamentous decompression to coagulation of cerebellar tonsils. Intraoperative ultrasonography is a useful tool to define when a decompression is sufficient. We did not find correlation between the need for reintervention for insufficient decompression and different invasiveness of the techniques. We believe that this finding suggests that our proposed scheme leads to the best tailored treatment for the single patient.

Keywords Chiari malformation · Reintervention · Surgical decompression

What symptoms are considered typical

Chiari malformation is a heterogeneous group of malformations characterized by a disproportion between the posterior cranial fossa (PCF) and nervous structures. Chiari type 1 malformation (CM1) is defined by a downward displacement of one or both cerebellar tonsils through the foramen magnum. By definition, there is no displacement of the brainstem. The bulbar variant of Chiari 1, so-called Chiari 1.5 (CM1.5) is defined by the presence of a caudal displacement of both cerebellar tonsils and brainstem [1]. As long as to our knowledge and in our experience there is no clinically significant difference between CM1 and CM1.5, we here present them as a unique pathology [2].

Signs and symptoms of CM1 may derive from altered cerebrospinal fluid (CSF) flow, compression of neural structures, or associated syringomyelia. Headache is the most frequently reported symptom, and it may be either referred as frontal or suboccipital and is usually worsened by efforts, cough, or other Valsalva-like maneuvers [3]. Cervical pain is often referred in relation with Valsalva or neck extension [4].

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Even if it is not usually stated as a typical CM1 symptom, we found frequently reported pain at lower limbs after efforts. We hypothesize this derives from compression of the posterior fascicles.

More rare but warning and suggesting the need for a more prompt treatment are brainstem compression symptoms like sleep apnea and dysphagia. Central nystagmus and dizziness may be present.

Compression of the brainstem may also result in motor deficits and signs of upper motor neuron lesion usually found at lower limbs, more rarely at upper limbs.

Criteria for surgical indications

Precise radiological evaluation and careful neurological examination are of critical importance in CM1 patients. Surgical treatment is indicated only for symptomatic patients and the decision of the surgical procedure of choice is based on radiological and clinical findings.

There is general agreement in using 5 mm from McRae line as the diagnostic cut-off of the caudal displacement of cerebellar tonsils through the foramen magnum. Anyway, given the fact that up to 35% of patients with a downward displacement of tonsils between 5 and 10 mm are asymptomatic [4], this measurement alone is not a diagnostic criterion; therefore, its exact measurement is not of critical importance. More important are radiological findings of compression of neural structures at the level of the foramen magnum [5]. In our center, we routinely use magnetic resonance imaging (MRI) with CSF flow measurement at the level of the foramen magnum to detect absence or reduction of radiologically evident CSF flow that indirectly indicates compression at that level.

There are several conditions like hydrocephalus, craniostenosis, or tethered cord that may be associated with CM1 and may need to be first addressed by treatment because their treatment may resolve CM1 symptoms [3, 6, 7].

In 10% of CM1 patients that are described with concomitant hydrocephalus [6], careful examination of the MRI is mandatory to detect hydrocephalus to correctly plan the surgical treatment.

In our center, in all patients with evidence of CM1 at MRI, we also routinely perform a full spine MRI to detect eventually present syringomyelia or other associated anomalies (i.e., tethered cord).

Neurophysiological studies have shown no correlation with clinical and radiological findings [8]; therefore, we do not routinely use them in the process of CM1 diagnosis and management.

CM1 natural history is still nowadays not well understood. As a general rule, we think that treatment of incidentally discovered asymptomatic CM1 patients without evidences of cord tethering or syringomyelia should be limited to follow-up in a wait and see fashion. Care must be taken to detect mild symptoms that may be underestimated by the parents or not referred by the patient.

While there is a wide consensus on operating symptomatic CM1 patients with syringomyelia, there seems not to be the same consensus on operating symptomatic CM1 patients without syringomyelia [9].

Considering that surgical outcomes in pediatric CM1 patients are better than those in adult CM1 patients [10], in our pediatric neurosurgical center, we always offer surgical treatment to CM1 symptomatic patients regardless of the presence of syringomyelia.

Type of surgery commonly used

Surgical options for CM1 are bony and ligamentous decompression with suboccipital craniectomy and C1 laminectomy alone and bony and ligamentous decompression combined with dural delamination or with duraplasty using autologous or heterologous graft. Coagulation of the tonsils is a reported option after dural opening. C2 and C3 laminectomies may also be needed depending on the grade of tonsillar descent.

We think that the procedure of choice must be tailored on the single patient; the more relevant the PCF compression and symptoms, the more invasive the treatment will be likely needed by the patient. To define which treatment is sufficient for the single patient, we routinely use intraoperative ultrasound, performed after bony and ligamentous decompression to evaluate the presence of a CSF layer between the dura and tonsils and the pulsation pattern of the tonsils. We consider a sufficient decompression when ultrasound demonstrates the presence of a CSF layer behind tonsils and an anterior to posterior physiologic pulsation of the tonsils. Conversely, a rostral to caudal pattern of pulsation and the absence of CSF behind the tonsils suggest the presence of ongoing compression and therefore indicate the need to further decompress. In these cases, we perform duraplasty with autologous graft.

Opening of the arachnoid with exploration of the fourth ventricle and obex or coagulation of the tonsils is not routinely performed at our institution as long as this procedure is not superior to duraplasty without violation of the arachnoid while carrying higher risks of pseudomeningocele and CSF fistulas [11]. These procedures play a role in revision surgery when symptoms persist despite previous decompression.

When the result is considered good

The main purpose of surgical treatment for CM1 patients is relief or improvement of symptoms; this depends on

several factors like symptom severity at presentation, presence of syringomyelia, and postoperative complications [7]. Success of the treatment resides in symptom relief independently from radiological improvement. Anyway, the absence of radiological improvement in cases of partial clinical improvement may indicate an insufficient decompression that may need revision surgery.

Clinical series

We retrospectively examined 172 pediatric patients with CM1 and CM1.5 surgically treated between 2006 and 2017 at the Neurosurgery Unit of IRCCS Giannina Gaslini Children Hospital of Genoa, Italy. The mean age of the patients was 8.1 years (range, 4 months–19 years). 58.1% were males (male/female ratio, 1.39). 84.9% were defined by MRI as CM1 and 15.1% as CM1.5.

All patients underwent brain and spine MRI; 35 patients (20.3%) who presented with sleep apneas underwent polysomnography to evaluate if these were of central origin or obstructive.

At diagnosis, syringomyelia was present in 26.2% and hydrocephalus in 9.3% of all cases; we did not find statistically significant differences between Chiari 1 and 1.5 in the association with syringomyelia and hydrocephalus (Table 1).

Associated neuraxis anomalies were present in 31 patients (Table 2). In patients with craniostenosis, the first treatment was cranial vault remodeling, and 3 (50%) of these did not need subsequent FMD. Patients in which a tethered cord was present at diagnosis were first treated with section of filum terminale (SFT), and of these, 7 (77.8%) were reoperated for FMD because symptoms of CM1 did not improve (Table 3).

In 11 patients (6.4%), FMD for CM1 was not needed after treatment of the associated conditions. Of these, 6 patients were treated only for hydrocephalus, 3 patients with concomitant craniostenosis were treated only with skull remodeling procedures, and in 2 patients, cord detethering was enough for symptom relieve.

Table 1 Associated anomalies and comparison between Chiari types 1 and 1.5. *P* values are for Fisher's exact test

	Chiari 1	Chiari 1.5	
Syringomyelia			
Yes	39	6	$P = 0.8116$
No	107	20	
Hydrocephalus			
Yes	11	5	$P = 0.0713$
No	135	21	

Table 2 Associated neuraxis anomalies

Nonsyndromic craniostenosis	6
Craniovertebral junction malformations	10
Scoliosis	6
Spinal dysraphisms with tethering of the cord	9

Using our flowchart for decision of the treatment for the single patient (Fig. 1), the first treatment addressing CM1 was bone and ligamentous decompression alone in 104 patients (65%), associated with dural delamination in 3 patients (1.9%) and associated with duraplasty with autologous graft in 53 patients (33.1%).

Postoperative follow-up was available for 171 patients because 1 patient died 1 month after surgery for cardiac disease not related to Chiari malformation. Follow-up ranges from 2 months to 10.9 years (mean = 5.1 years).

Postoperative complications occurred in 5 patients (Table 4), 3 major complications that needed rescue surgery were 1 subdural hematoma following duraplasty, 1 acute postoperative hydrocephalus following duraplasty with unintended fenestration of the arachnoid, and 1 rupture of subfascial drainage positioned after bony decompression. The minor complications were 2 postoperative pseudomeningoceles that showed spontaneous resolution at follow-up.

Syringomyelia at follow-up was reduced in 91.5% of cases and stable in the remaining 8.5%.

Reintervention for insufficient decompression at follow-up was needed in 6 patients (Table 4); 1 patient was reoperated twice. Three patients treated initially with only bony and ligamentous decompression needed reinterventions and underwent duraplasty; 1 of these needed reintervention after duraplasty and was reoperated with coagulation of the tonsils. Three patients first treated initially with duraplasty needed reintervention for insufficient decompression and were reoperated with coagulation of the tonsils.

Statistical analysis show that occipito-cervical decompression with duraplasty is significantly associated with increased complications compared with decompression without duraplasty. We have found no correlation between one of these two procedures and the need for reintervention for insufficient decompression. This finding suggests that the procedure of choice was well tailored for single patients.

Table 3 Associated anomalies that needed FMD after first treatment

	Treated (*n*)	Needed FMD (*n*)
Hydrocephalus	16	6 (37.5%)
Craniostenosis	6	3 (50%)
Tethered cord	9	2 (22.2%)

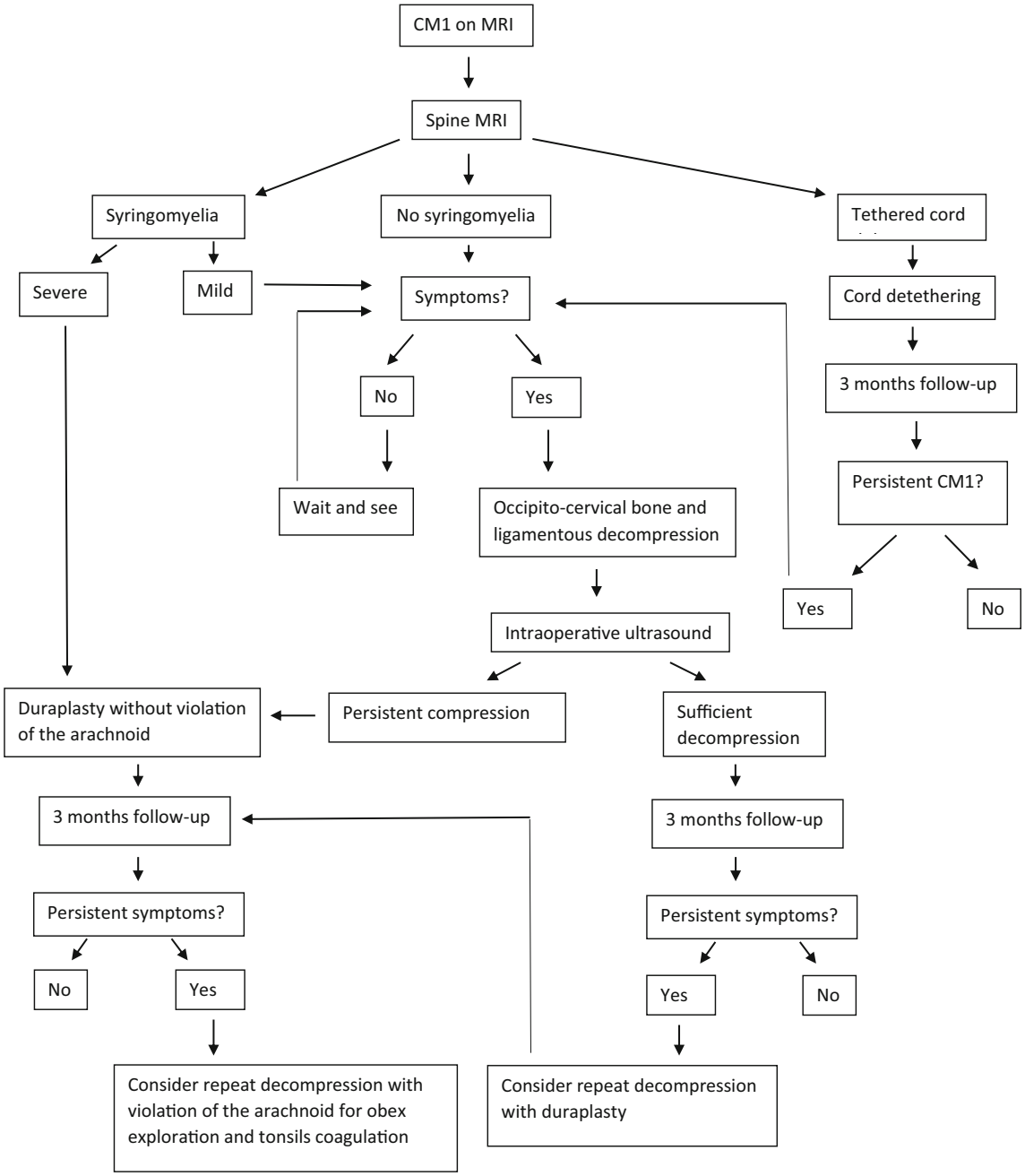

Fig. 1 Flowchart for management of CM1 patients

Table 4 Postoperative complications and reinterventions for insufficient decompression. *P* values are for Fisher's exact test

	Bone and ligamentous decompression (± dural delamination $n = 3$) $n = 107$	Bony decompression and duraplasty $n = 53$	
Complications			
Yes	1	4	$P = 0.0417$
No	106	49	
Reintervention			
Yes	3	3	$P = 0.3986$
No	104	50	

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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