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Chiari type I and hydrocephalus

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Abstract

Purpose The association between Chiari type I malformation (CIM) and hydrocephalus raises a great interest because of the still unclear pathogenesis and the management implications. The goal of this paper is to review the theories on the cause-effect mechanisms of such a relationship and to analyze the results of the management of this condition.

Methods A review of the literature has been performed, focusing on the articles specifically addressing the problem of CIM and hydrocephalus and on the series reporting about its treatment. Also, the personal authors' experience is briefly discussed.

Results As far as the pathogenesis is concerned, it seems clear that raised intracranial pressure due to hydrocephalus can cause a transient and reversible tonsillar caudal ectopia ("pressure from above" hypothesis), which is something different from CIM. A "complex" hypothesis, on the other hand, can explain the occurrence of hydrocephalus and CIM because of the venous engorgement resulting from the hypoplasia of the posterior cranial fossa (PCF) and the occlusion of the jugular foramina, leading to cerebellar edema (CIM) and CSF hypo-resorption (hydrocephalus). Nevertheless, such a mechanism can be advocated only in a minority of cases (syndromic craniosynostosis). In non-syndromic CIM subjects, the presence of hydrocephalus could be explained by an occlusion of the basal CSF pathways, which would occur completely in a minority of cases (only 7–10% of CIM patients show hydrocephalus) while it would be partial in the remaining cases (no hydrocephalus). This hypothesis still needs to be demonstrated. As far as the management is concerned, the strategy to treat the hydrocephalus first is commonly accepted. Because of the "obstructive" origin of CIM-related hydrocephalus, the use of endoscopic third ventriculostomy (ETV) is straightforward. Actually, the analysis of the literature, concerning 63 cases reported so far, reveals very high success rates of ETV in treating hydrocephalus (90.5%), CIM (78.5%), and syringomyelia symptoms (76%) as well as in giving a radiological improvement of both CIM (74%) and syringomyelia (89%). The failures of ETV were not attributable to CIM or syringomyelia. Only 11% of cases required PCF decompression after ETV.

Conclusions The association between CIM and hydrocephalus probably results from different, multifactorial, and not yet completely understood mechanisms, which place the affected patients in a peculiar subgroup among those constituting the heterogeneous CIM population. ETV is confirmed as the best first approach for this subset of patients.

Keywords Chiari I malformation · Hydrocephalus · Posterior cranial fossa · Endoscopic third ventriculostomy

Introduction

The association between Chiari type I malformation (CIM) and hydrocephalus is known since the first description of the disease provided by Hans Chiari in 1891 [11, 12]. The author actually reported on a 17-year-old girl, dead of typhoid fever, whose autopsy revealed a hindbrain herniation associated to chronic hydrocephalus. On these grounds, Chiari postulated that the hydrocephalus was responsible for the caudal ectopia of the cerebellar tonsils. Since there, the relationship between the two diseases has been repeatedly investigated and several cases have been reported.

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CIM is prone to present with concurrent anomalies, syringomyelia and scoliosis being the most common ones [27, 37, 58]. Currently, hydrocephalus is described in about 7–10% of CIM patients in large series [52, 74]. The rate can vary from 2.2% to 18%, with higher percentages in case of syndromic subjects [3, 20, 71]. Such an association still raises a great interest among the scientific community because of the partially unsolved pathophysiological and management issues. In particular, CIM is considered to result from the underdevelopment of the posterior cranial fossa (PCF) although the hypoplasia of PCF does not necessarily lead to CIM and CIM can be present in normal volume PCF [47, 65]. Unexpectedly, only a minority of CIM subjects develop hydrocephalus. However, the diagnosis of hydrocephalus is becoming more and more frequent in CIM patients and burdens the postoperative outcome with an increased rate of complications [58]. Indeed, the pre-existing hydrocephalus has been found to complicate by 6.8% and 1.6%, respectively, the postoperative surgical and medical course of CIM patients, with following increase of the surgical and medical costs [46]. Therefore, the management of these patients is a delicate matter to deal with.

On the other hand, hydrocephalus can be present as postoperative complication of PCF decompression in CIM subjects (especially, in case of osteo-dural decompression). Actually, hydrocephalus complicates surgery for CIM from about 1 to 2.5% up to 18% of cases [15, 19, 30, 77]. Of course, in such an instance, the physiological mechanisms and the management implications are different from those concerning the topic of this article.

Physiopathological considerations

According to the daily clinical practice, the relationship between CIM and hydrocephalus seems to be statistically in favor of causative role of CIM in inducing hydrocephalus, because of the obliteration of the basal subarachnoid spaces, more than the vice-versa. However, although not frequent and not evident in a great proportion of cases, the mechanism exerted by hydrocephalus (or other causes of raised intracranial pressure (ICP)) seems to be more obvious and easier to understand (Fig. 1). The observation of CIM following the progression of hydrocephalus has been actually demonstrated even in utero [40]. The main limitation of this hypothesis, the so-called pressure from above theory, is that the resulting CIM is only transient and regresses after the treatment of hydrocephalus, while it is well known that CIM can radiologically persist even after a successful treatment of the resulting hydrocephalus [18, 78]. Similarly, CIM resulting from other causes of raised ICP, like large tumors (even is located in the supratentorial space) or arachnoid cysts, disappears as its cause is removed [44, 67, 70]. Therefore, it should be simply considered as a transient caudal tonsillar descent more than a chronic disease, as CIM is.

On the other hand, the overcrowding of the foramen magnum by the PCF neural structures represents a more reliable pathophysiological mechanism, to explain the associated hydrocephalus, because such an overcrowding is present in all CIM subjects, being a condition necessary to define this disease. The effacement of the vallecula and the cisterna magna, the compression of the fourth ventricle (or, even, the prepontine and premedullary cisterns), further worsened in case of narrow foramen magnum and/or basilar invagination, give reason of the altered cerebrospinal fluid (CSF) dynamics documented by the flow-sensitive phase-contrast MRI investigations [9, 35]. These studies, indeed, together with the cine phase sagittal MR imaging, demonstrate an abrupt, downward systolic displacement of the brainstem and cerebellar tonsils causing plugging and narrowing of the CSF pulsations with a forced CSF transmission through the crowded foramen magnum [2]. It is not clear yet if these phenomena lead to an occlusion of the fourth ventricle foramina or a direct impairment of the CSF exit from the intracranial space or both. Alternatively, some authors postulated that the dissociation of CSF circulation at level of the foramen magnum would stop the pressure transmission from the pulsating vessels to the CSF, with subsequent decreased CSF flow compliance of both the brain (hydrocephalus) and spinal cord (syringomyelia) [28]. These "mechanical" interpretations do not solve the question of the small proportion of CIM patients (7–10%) developing hydrocephalus, because a mechanical obstacle to CSF circulation within the PCF may be hypothesized in nearly all cases. In such a regard, some authors postulated that, in spite of the obstruction of the foramen of Magendie, an adequate CSF outflow from the IV ventricle is provided by patent foramina of Luschka in the majority of patients [16].

Anyway, the compression on the fourth ventricle would be the result of a "cephalocranial disproportion" following a PCF hypoplasia or a hyperplasia of its content (Fig. 1). This concept is borrowed from the observations provided by Hoffman and Tucker who reported on the progressive thickening of the skull in children with chronic extra-thecal shunt [34]. In these subjects, the early decrease in the intracranial volume may lead to secondary CIM [10] as well as it happens in children with craniosynostosis as the consequence of a cephalocranial disproportion resulting from a small skull compared with normal cerebral hemispheres and ventricles [76]. Once again, in spite of the frequency of CIM in subjects with syndromic craniosynostosis (70–100%), only a minority of them develops hydrocephalus (12–15%), thus suggesting that, probably, there is not a direct cause-effect relationship [13, 14]. However, the high rate of children with hydrocephalus presenting CIM in syndromic craniosynostosis (up to 90%) would indicate, at least, a common physiopathological pathway, based on the early fusion of the basi-occipital and lambdoid sutures [18]. This observation justifies the higher

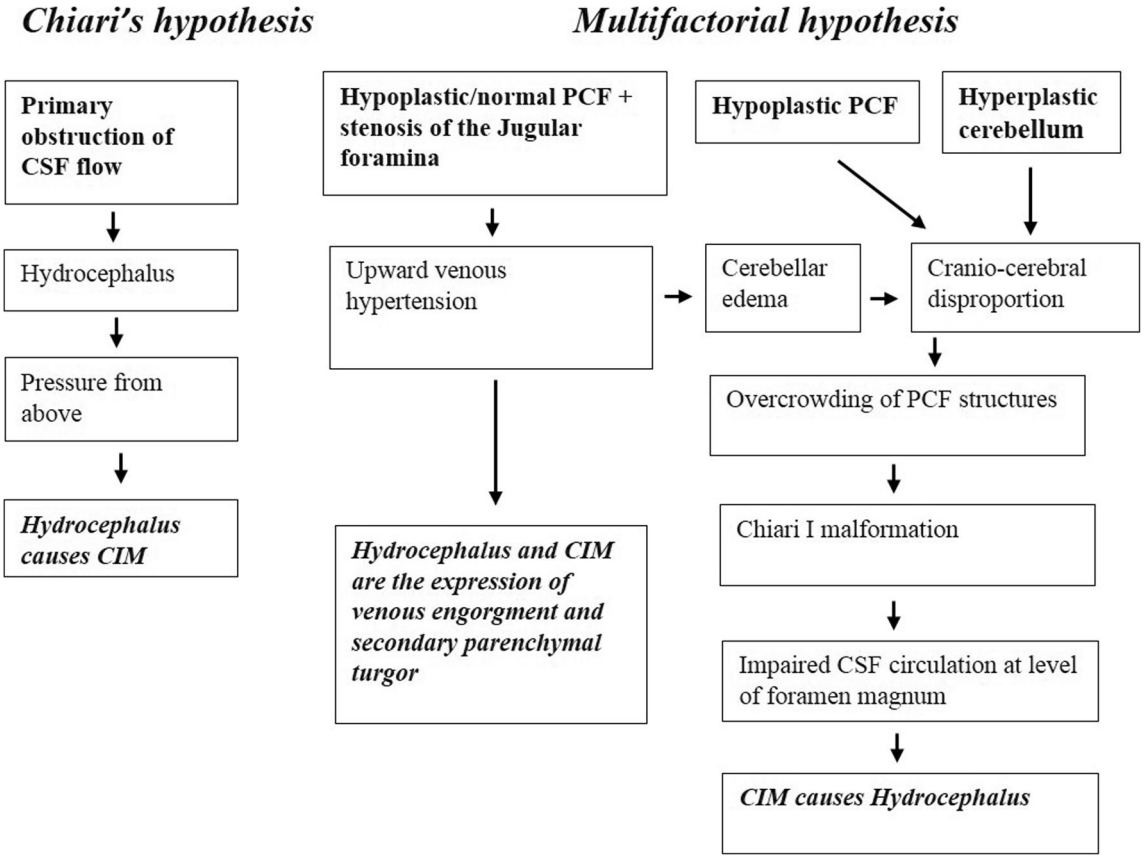

Fig. 1 Pathophysiological hypotheses (modified from 18)

frequency of such association in Crouzon patients compared with other syndromes. Indeed, the occipital synchondroses involved in the growth of the posterior cranial fossa close completely in several subjects with Crouzon's syndrome already during the first year of life [43]. Moreover, the chondrogenic and osteogenic action of the FGFR2 (and FGFR3) gene leads to the early fusion of both anterior and posterior intra-occipital synchondroses in Crouzon and Pfeiffer syndromes but only the posterior ones in Apert syndrome, thus justifying the differences among these syndromes (the only three ones where such a premature fusions has been demonstrated so far) in terms of CIM and hydrocephalus [60]. As a result, the premature closure of the posterior cranial vault sutures further increases the disproportion between the insufficient expansion of the PCF and its content, worsening the PCF overcrowding and the CSF circulation.

If the theory of the cephalocranial disproportion is pertinent for the secondary CIM in craniosynostosis and the resulting hydrocephalus, it fails to explain the occurrence of hydrocephalus before CIM in these subjects. For this reason, some authors hypothesized, a couple of decades ago, a possible role of the bilateral stenosis of the jugular foramina in increasing the venous pressure and decreasing the CSF flow in Crouzon patients [24]. More recently, Cinalli and coworkers confirmed such a causative mechanism in syndromic craniosynostosis where the fusion of the lambdoid suture (usually completed by the first 2 years of life) involves the petro-occipital synchondroses [14]. Accordingly, the impaired venous drainage and the subsequent venous hypertension resulting from the stenosis of the jugular foramina would cause both CIM (venous engorgement, followed by cerebellar "enlargement," followed by tonsillar descent) and hydrocephalus (combination of CSF hypo-adsorption and CIM), independently from each other. In case of no stenosis of the jugular foramina, CIM would result from the early lambdoid closure (small PCF) and the hydrocephalus would be secondary to the impaired CSF circulation (overcrowded PCF) (Fig. 1).

Finally, the cephalocranial disproportion can occur in case of cerebral and, in particular, cerebellar overgrowth in a normal-sized PCF [29]. The occurrence of CIM and resulting hydrocephalus in macrocephaly-associated conditions, like macrocephaly-cutis marmorata teleangectatica congenita, Costello syndrome, Noonan syndrome, CFC syndrome, and NF1, support this hypothesis [36, 42, 48, 72, 83] (Fig. 1).

In spite of the aforementioned observations, it is still a matter of debate whether the anatomical and/or functional obstruction of the CSF flow by CIM is able to cause a secondary hydrocephalus. This is because hydrocephalus occurs only in a minority of CIM patients and a functional stenosis of the Magendie foramen can be demonstrated on cine-MRI only in

some cases. Therefore, some authors postulated that the overcrowding of the foramen magnum induces a "dissociation of the pressure" between the cranial and the spinal compartments, with an abnormally high pressure response to body movements [31, 80]. The obstruction of the foramen magnum would account for the compression of the veins, which lose their ability to distend to compensate an increased intracranial pressure. Subsequently, according to the unifying hypothesis for hydrocephalus, Chiari malformation, and syringomyelia proposed by Williams on 2008, the hydrocephalus could be a condition of edema of brain depending on the venous insufficiency [79]. Such a failure of the venous drainage could act as a transient "pseudotumor cerebri" (raised ICP without ventricular enlargement) that, if severe and repeated, would contribute to the hydrocephalus as well as to the rare case of abrupt clinical deterioration in CIM patients [4, 8, 22, 38, 82]. Unfortunately, this venous insufficiency is hard to demonstrate radiologically or experimentally.

In summary, all the theories here described fails in giving a unique explanation for the association of CIM and hydrocephalus. Probably, such an association results from the relationship among congenital and acquired factors, potentially able to create the combination of the two pathological conditions.

Management

The optimal management of CIM and associated hydrocephalus still raises some debates just because of missing definite pathophysiological mechanisms explaining this association. An initial matter of discussion is what disease should be addressed first. Currently, most of the neurosurgeons agree on to operate hydrocephalus as first step [49, 57, 63, 66, 73], although the reduction of the ventricular dilatation is followed by the persistence of the clinical signs and symptoms of CIM in a certain number of cases [26, 33, 50]. This policy results from three main observations. (1) In the clinical practice, it is often hard to demonstrate which disease comes first (Fig. 2). Therefore, it is straightforward to deal first with the "more symptomatic" one that, usually, is the hydrocephalus, also eliminating the raised ICP [41]. Subsequently, this option frequently allows to improve another associated, CSF flow/raised ICP-depending, "symptomatic" condition, which is the syringomyelia [49]. (2) Similarly, being the hydrocephalus-related raised ICP able to worsen the CIM, it is preferable to eliminate this cause first and then to decompress the PCF, if a symptomatic CIM persists. Actually, by analyzing the clinical history of asymptomatic CIM children, we noticed a worsening only in those affected by hydrocephalus [56]. This option carries a further advantage too, which is to reduce the risk of postoperative pseudomeningocele (possibly resulting from the combination of untreated hydrocephalus-related raised ICP and open space subsequent to PCF decompression) or other CSF-dynamics-related complications [75]. (3) Sometimes, the cause of the hydrocephalus (e.g., aqueduct stenosis) may not be clearly evident at the diagnosis. In this instance, the PCF decompression may not produce a clinical improvement, as demonstrated by Bartoli et al. in a 39-old man with unspecific symptoms who underwent PCF osteodural decompression for CIM (tonsillar descent 10 mm) without clinical advantages [5]. Postoperative MRI revealed both aqueduct stenosis and foramina of Monro hypoplasia that required a specific treatment, finally leading to the clinical recovery.

On the other hand, an early treatment of hydrocephalus is not recommended in children with obvious craniocerebral disproportion due to syndromic craniosynostosis, because it could further worsen the already impaired growth of the skull, contributing to the PCF overcrowding. In these instances, a posterior cranial vault expansion (possibly together with PCF decompression) seems to be more appropriate [18, 64]. It is worth noting, however, that such a worsening generally results from the insertion of VP shunt, while it seems to be less evident by using ETV, which is the treatment of choice of hydrocephalus in this subset of patients. Moreover, in single-suture craniosynostosis, it seems to be reasonable to relieve from raised ICP with ETV before carrying on with cranial remodeling [66].

An exception to the aforementioned strategies of management can be applied to patients harboring only a ventriculomegaly (enlarged ventricles without signs/symptoms of raised ICP) and CIM, which is a further possible combination. As demonstrated by Deng and coworkers in a series composed by 38 adults, the ventricular dilatation (Evan's index >0.30 without papilledema, headache, and vomiting) remained stable and asymptomatic after PCF decompression and duraplasty [17]. The cervico-medullary decompression was effective, and no complications (namely, pseudomeningocele) occurred.

The type of treatment of the hydrocephalus is a second topic for discussion. Since the studies and the clinical findings are in favor of an obstructive hydrocephalus, resulting from the compression/occlusion of the fourth ventricular outlets, with or without concurrent aqueduct stenosis, ETV is currently considered the gold standard therapy [47, 49]. The effectiveness of ETV in the management of obstructive hydrocephalus and its superiority, in terms of reduced rate complications, compared with VP shunt have been largely demonstrated in the last 2–3 decades and does not need any further arguments in support. On the other hand, the effectiveness of ETV in the context of obliterated basal subarachnoid spaces (namely, the prepontine and premedullary cisterns) could be questioned. The answers to this question are

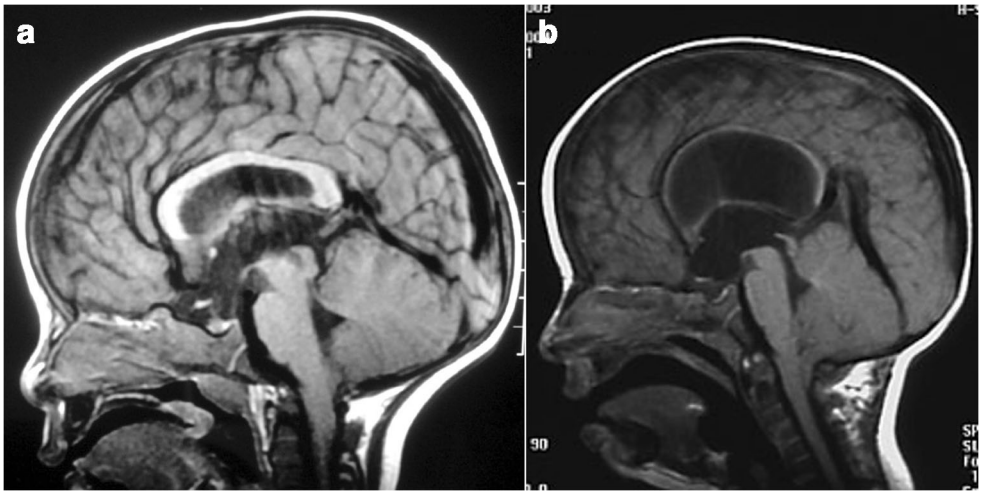

Fig. 2 Two sagittal T1 MRIs showing a similar picture of triventricular hydrocephalus and mild caudal descent of the cerebellar tonsils. However, in **a**, the cerebellum is accommodated inside a normal PCF, and despite that the severity of hydrocephalus is inferior than in **b**, it can be hypothesized that hydrocephalus (aqueduct stenosis) causes CIM in this instance. Instead, in **b**, the cerebellum is inside a smaller posterior fossa (look at the upward herniation of the culmen with displacement of the Galen venous system) and the aqueduct is patent; therefore, it is likely that CIM causes hydrocephalus in this case

mainly factual because there is evidence of the success of ETV in high rate of cases (Table 1). However, according to our personal experience based on the PCF measurements in subjects with hydrocephalus and CIM, the success of ETV can be predicted with a sufficient reliability in two subgroups of patients, which are children without overcrowding of the posterior cranial fossa and adults with large cystic dilatation of the fourth ventricle [50]. In this study, we performed a measurement of PCF volume (PCFV), PCF brain volume (PFBV), and PCF overcrowding (PFBV/PCFV ratio), borrowed from the Nishikawa et al. method [55], in 11 children and 4 adults with hydrocephalus (symptomatic in all cases) and CIM (symptomatic in 2/3 of cases). ETV was effective in treating hydrocephalus (all cases, both clinically and radiologically), improving CIM symptoms (all cases, disappearing in six out of ten patients), improving the tonsillar descent (from 12.7 mm to 8.3 mm mean caudal ectopia), and treating the syringomyelia (disappeared in three out six 6 cases and improved in two cases). Such good results were obtained in a pediatric population where PCFV, PFBV, and PCF overcrowding did not differ from their age-matched healthy controls, and in an adult population where only the PCF overcrowding was increased compared with the controls. This overcrowding resulted from a clear obstruction of the fourth ventricle (cystic dilatation), as already found by Decq and coworkers [16]. Therefore, we concluded the CIM with associated hydrocephalus is a particular subset among the different "diseases" grouped under the name "CIM," which is characterized by a prevalent presence of some kind of obstruction in the CSF circulation that is susceptible of treatment by means of ETV. The reduced volume of the prepontine/premedullary cistern is not a contraindication to ETV.

Clinical series and conclusions

As mentioned, ETV is being more and more utilized to treat the combination of hydrocephalus and CIM/syringomyelia. Indeed, good results have been reported so far in several isolated cases or small series [16, 21, 25, 26, 33, 38, 41, 50, 51, 53, 54, 66, 68, 69, 78, 81]. In the past, satisfactory results were obtained also by a direct approach on the posterior cranial fossa to open the foramina of the fourth ventricle and/or by ventriculoperitoneal shunt [1, 32, 39, 59]. Nowadays, however, these techniques are almost abandoned, considered too invasive (craniotomy approach to the fourth ventricle), or poorly used because they are burdened by the risk of shunt dependency and complications (shunts).

The main aspects of current series available from the literature are summarized in Table 1. According to their results, the following conclusions can be taken. (1) ETV is an effective treatment of the raised ICP resulting from the hydrocephalus associated to CIM. Overall, a 90.5% success rate is reported among the 63 patients described in the literature independently from their age, sex, characteristics of CIM, and presence of syringomyelia. This success rate is even higher than that commonly reported for other types of obstructive hydrocephalus (average 75%, range 50–90%) [7, 23, 61, 62], although the CIM patient sample is too small for a reliable comparison. These evidences reinforce the belief about the "obstructive" character of the CIM-related hydrocephalus. Moreover, the improvement of the hydrocephalus is evident also on postoperative MRI. Actually, a significant reduction of the transverse diameter of the third ventricle after ETV has been demonstrated in two series where this topic was addressed [50, 81]. (2) The failure of ETV (9.5%, 6 cases) does not seem to be correlated with the presence of CIM/

Table 1 Synopsis of the results from the literature[a]

Authors	*Year*	*No. patients*	*Raised ICP improvement*	*CIM improvement*	*Syringomyelia improvement*	*Failed ETV*
Nishihara et al. [54]	1996	1	1/1	Symptoms: NA Radiology: NA	Symptoms: 1/1 Radiology:1/1	–
Fukuhara et al. [26]	2000	5	2/5	Symptoms: NR Radiology: NR	Symptoms: NR Radiology: NR	3/5 (60%)
Suehiro et al. [68]	2000	1	1/1	Symptoms: NA Radiology: NA	Symptoms: 1/1 Radiology: 1/1	–
Decq et al. [16]	2001	5	5/5	Symptoms: 5/5 Radiology: 5/5	Symptoms: NA Radiology: NA	–
Ersahin and Gockai [21]	2002	1	1/1	Symptoms: 1/1 Radiology:1/1	Symptoms: NA Radiology: NA	–
Metellus et al. [51]	2002	1	1/1	Symptoms: 1/1 Radiology:1/1	Symptoms: 1/1 Radiology:1/1	–
Mohanty et al. [53]	2005	2	2/2	Symptoms: 2/2 Radiology:2/2	Symptoms: 2/2 Radiology:2/2	–
Teo et al. [69]	2005	1	1/1	Symptoms: 1/1 Radiology: NR	Symptoms: NA Radiology: NA	–
Fuentes et al. [25]	2006	1	1/1	Symptoms: 1/1 Radiology: 1/1	Symptoms: NA Radiology: NA	–
Hayhurst et al. [33]	2008	16	15/16	Symptoms: 10/16 Radiology: NR	Symptoms: 3/5 Radiology: 5/5	1/16 (6.2%)
Kandasamy et al. [38]	2008	1	1/1	Symptoms: 1/1 Radiology: 0/1	Symptoms: NR Radiology: 0/1	/
Massimi et al. [49]	2011	15	15/15	Symptoms:10/10 Radiology: 8/15	Symptoms: 4/4 Radiology: 5/6	–
Wen et al. [78]	2014	1	1/1	Symptoms:0/1 Radiology:1/1	Symptoms: 0/1 Radiology: 1/1	–
Sgulò et al. [66]	2017	1	1/1	Symptoms: NR Radiology:1/1	Symptoms: NA Radiology: NA	–
Konar et al. [41]	2018	1	1/1	Symptoms: 1/1 Radiology: 0/1	Symptoms: NA Radiology: NA	–
Wu et al. [81]	2018	10	8/10	Symptoms: 0/2 Radiology: 9/10	Symptoms:1/2 Radiology: NR	2/10 (20%)

NA not applicable, *NR* not reported

[a] Only the literature specifically concerning this topic has been considered. Both pediatric and adult cases are included

syringomyelia. Actually, in the only patient where the missed improvement is attributable to the persistence of hydrocephalus, the cause was probably the dependence on a previous shunt (a new shunt was actually required in spite of a patent ETV) [33]. Two other cases were considered as failed although the late deterioration was attributed to CIM progression rather than to the missed resolution of hydrocephalus [81]. Finally, no information is available on the remaining three cases (described about 20 years ago) because it was not the goal of that study, provided by Fukuhara et al. [26]. However, on multivariate analysis, CIM did not reach a statistical significance in precluding the success of ETV. (3) ETV favorably affects also the course of CIM in this subset of patients. Actually, 34 out of 42 subjects with symptomatic CIM (78.5%) showed an improvement/resolution of their symptoms. Such a good rate probably results from the disappearance of the raised ICP other than from a real improvement of CIM. A reduction of the tonsillar descent is demonstrated in 74% of cases on postoperative MRI (29 out of 39 patients where this data is available) as possible result of the reduction of the cephalocranial disproportion due to the CSF bypass related to ETV. As expected, however, such a reduction is not impressive, the mean improvement ranging from 0.6 mm to 4.4 mm [33, 50, 81]. Among the nine patients who did not improve clinically, seven required PCF decompression because of persistent symptoms. Six of them belong to the largest series published so far (16 cases), provided by Hayhurst and coworkers [33]. The authors noticed the persistence of CIM symptoms in four cases and syringomyelia symptoms in two cases who, therefore, required surgery mainly 6 months after ETV (37.5% of the whole series). The remaining patient belongs to the Wu et al. series: they were operated on 7 years after ETV [81]. In the personal series, the second largest in the literature (15 cases), we did not have patients requiring surgery for CIM or syringomyelia after ETV (35-month mean follow-up) [50]. The volumetric analysis of the PCF allowed us to explain such a good result. As mentioned, indeed, our patients are probably part of a subgroup of CIM subjects with normal volumes of PCF where the local compression/overcrowding is able to cause

hydrocephalus and the resolution of hydrocephalus is enough to improve the CIM symptoms (reversible local compression/overcrowding thanks to the reduced craniocerebral disproportion) in spite of its radiological persistence. Similar findings were observed by Decq and coworkers in five adult patients with characteristics overlapping those of our adult patients (namely, cystic dilatation of the IV ventricle with resulting overcrowding of the foramen magnum) [16]. In spite of the reduced PCF volume at the bi-dimensional measurements carried out by the authors, none of their patients required PCF decompression thanks to the post-ETV improvement of the overcrowding. The confirmation of these observations comes from the Hayhurst et al. experience where, because of the small PCF and the missed improvement of the overcrowding after ETV, some patients required PCF decompression [33]. (4) Similar considerations can be applied to the syringomyelia. Indeed, a significant proportion of patients experienced a clinical (13 out of 17 cases, 76%) and/or a radiological improvement (16 out of 18 cases, 89%). In particular, syringomyelia responds very well to ETV as far as its reduction or even disappearance on MRI is considered. This could result from the possible contribution of hydrocephalus in its formation in both communicating and non-communicating forms [6]. Actually, according to some of the most recent theories on syringomyelia, it would result from the subarachnoid obstruction at the foramen magnum and the following difference in pressure above and below the site of obstruction, leading to a disruption of the spinal cord blood barrier with ultrafiltration of crystalloids and accumulation of protein-poor fluid [45]. In this context, the hydrocephalus would be able to increase the differential pressure above and below the foramen magnum, thus favoring the syringomyelia formation. Therefore, the latter improves or disappears as soon as the former is treated.

Compliance with ethical standards

Conflict of interest No conflict of interest to disclose.

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