Childs Nerv Syst (2009) 25:443–452
DOI 10.1007/s00381-008-0760-z

ORIGINAL PAPER

A late complication of CSF shunting: acquired Chiari I malformation

Massimo Caldarelli · Federica Novegno · Concezio Di Rocco

Received: 28 October 2008 / Published online: 5 December 2008

Abstract
Purpose Acquired Chiari I malformation developing after cerebrospinal fluid (CSF) shunting is an intriguing late complication of CSF shunt surgery and not only raises questions as to its pathogenesis but also poses many queries about the possible adverse effects on the subsequent child development as well as on the indications and possibilities of surgical correction.
Materials and methods We report a series of 17 patients with the neuroradiological evidence of an "acquired" Chiari type I malformation. These patients, 3 to 24 years (mean 11.3 years), had been treated previously with an extrathecal CSF shunt. The follow-up varied from 2 to 12 years (mean, 6).
Results Neuroradiological investigation demonstrated a small posterior cranial fossa, small ventricles, markedly reduced periencephalic subarachnoid spaces, and thickening of the skull vault and base. The degree of tonsillar herniation did not correlate with clinical manifestations. All patients with severe clinical manifestations or with progressive worsening of neuroradiological findings were considered for surgical treatment. Decompressive supratentorial craniotomy seemed to fit with the purpose of enlarging the intracranial volume without the risk of aggravating the hindbrain herniation. An immediate relief of clinical manifestations was observed in all patients. Not surgically treated patients did not show any clinical or radiological modification during all the follow-up.
Conclusions This study contributes to the understanding of the underlying pathogenetic mechanisms of acquired Chiari type 1 malformation in cases of long-lasting supratentorial CSF shunting and provides a base for planning the best management, whether conservative or surgical.

Keywords Acquired Chiari 1 malformation · Hydrocephalus · Arachnoid cyst · CSF shunting late complications

M. Caldarelli · F. Novegno (✉) · C. Di Rocco
Department of Pediatric Neurosurgery,
Catholic University Medical School,
Largo A. Gemelli 1
00168 Rome, Italy
e-mail: federicanovegno@hotmail.it

Introduction

The Chiari type I malformation is characterized by a peg-like elongation of the cerebellar tonsils and medial aspect of the inferior vermis outside the cranial cavity into the cervical canal. It can occur either as an isolated malformation or associated with some congenital or acquired craniovertebral junction abnormalities or with selected craniofacial dysostoses, like for instance Crouzon, Carpenter, and, less commonly, Apert syndromes, and even with apparently nonsyndromic craniosynostoses [5, 7, 23].

Considering the acquired forms, cerebellar tonsil herniation has been described following lumbo-peritoneal shunting performed for communicating hydrocephalus or for relieving "benign" intracranial hypertension [1, 4, 8] and quite recently as late complication of the early treatment of supratentorial arachnoid cyst by means of cysto-peritoneal shunt [7, 9, 13, 14, 16]. Chiari I malformation, developing after early treatment of congenital hydrocephalus by means of ventriculo-peritoneal shunting, is an intriguing late complication (or evolution?) of this surgery and not only raises the question of its pathogenesis but also poses many queries about the possible adverse effects on the subsequent child development as well as on the indications and possibilities of surgical correction. Actually, if tonsil herniation can be explained pre-operatively as an expression of

the increased intracranial pressure (ICP), it is much more problematic to explain the novel appearance of a Chiari I malformation as an effect of a satisfactorily working cerebrospinal fluid (CSF) shunt; furthermore, even giving the pathogenesis as explained, the more appropriate surgical treatment of this complication remains questionable.

Among about 1,700 patients treated with ventriculoperitoneal CSF shunting over the last 30 years and followed at our outpatient clinic, we found retrospectively 17 patients who developed an acquired Chiari I malformation as late complication/evolution of CSF shunting. Five of them were symptomatic, whereas the other 12 presented with minimal or absent clinical manifestations in spite of similar neuroradiological pictures. This observation prompted us to report on this unusual late complication of CSF shunt surgery and to analyze the possible underlying pathogenetic mechanisms, as well as to suggest which might be, in our opinion, the most appropriate surgical treatment.

Materials and methods

Seventeen patients (13 boys and four girls) aged 3 to 24 years (mean 11.3 years) were evaluated at the Pediatric Neurosurgery Unit, the Catholic University Medical School, Rome, for the neuroradiological evidence of an "acquired" Chiari type I malformation (one of these cases has been previously reported [8]); all of them had been treated previously with an extrathecal CSF shunt. Some of the patients who underwent extrathecal CSF shunting at our Institution were lost at follow-up; this could underestimate the incidence of acquired Chiari I malformation. All of the patients here considered had undergone repeated computed tomography (CT) scans or magnetic resonance imaging (MRI) studies, either before or after the CSF shunt placement as control for the postoperative clinical observation, which did not demonstrate any kind of cerebellar tonsil herniation before the actual detection of the acquired Chiari type I malformation. In particular, among the 17 patients considered, 12 had performed cranial MRI prior to the shunt implantation. Out of the remaining five children, whose diagnosis was obtained by a preoperative CT scan, two were evaluated also by reconstructed sagittal images. All the 17 patients here considered were evaluated in the postoperative phase according to our protocol which consists of CT and, in the last two decades, MRI scans to be performed immediately after the operation and subsequently at 6 months, 1, 2, and 3 year intervals. As to the initial diagnosis, 13 of these patients were affected by neonatal hydrocephalus. This was secondary to aqueductal stenosis in five cases associated with semilobar holoprosencephaly in one case; in seven children, it was secondary to intra-/periventricular hemorrhage; postmeningitic in one case. Other two boys had been treated by means of a cysto-peritoneal shunt for a supratentorial arachnoid cyst localized over the fronto-parietal convexity in one case and in the chiasmatic cistern in the other. One patient had been treated for a posttraumatic hydrocephalus. Finally, one girl had been shunted for an obstructive hydrocephalus secondary to an optic-hypothalamic glioma. CSF shunting had been performed in the early neonatal period in case of hydrocephalus and later on in case of arachnoid cyst, anyway not beyond 6 months of age (mean 3.1 months) in all but two children, namely, the patient with posttraumatic hydrocephalus (who was 3 years) and the girl affected by optic pathway glioma who was 5 years old at the time of CSF shunting. Clinical manifestations at the first operation were mainly those of increased intracranial pressure in all the cases, although milder in case of arachnoid cyst. The subsequent clinical histories were unremarkable, apart from CSF shunt revision procedures (including CSF shunt elective lengthening) in all but one children; furthermore, one child underwent the drainage of a subdural hematoma; on the other hand, no revision had been performed in one case of cyst-to-peritoneal shunt.

Even though computed tomography could already suggest the diagnosis, neuroradiological demonstration of Chiari I malformation relied mainly on magnetic resonance imaging in all the cases. Basically, the diagnosis of Chiari I malformation was based on the neuroradiological demonstration of the displacement of one or both cerebellar tonsils 5 mm or more below the basion–opisthion line. Other aspects taken into consideration were the following: amount of subarachnoid spaces over the cerebral hemisphere at the level of foramen magnum, "tightness of posterior fossa," appearance of protruding cerebellar tonsils, and presence of syrinx.

Results

Tables 1, 2, and 3 summarize the main data from this series of patients.

Clinical manifestations

Among the 17 patients, eight were asymptomatic at the time of the acquired Chiari malformation diagnosis, and four complained only occasional mild headache without neurological deficits; the remaining five patients presented different symptoms and signs. The most frequent complaint was headache and neck pain, which was present in five patients, and less frequently, vertigo which was observed in one case and nausea and vomiting in one case. Clinical signs were bilateral papilloedema in two cases, VIth nerve palsy in two cases, ataxia in two cases, and upper limbs

Table 1 Summary of the patients who were not operated on

Number	Sex	Diagnosis	Clinical manifestations	Initial treatment	Age at operation (months)	Age at Chiari I diagnosis (years)	Tonsillar herniation degree (mm)	Clinical manifestations	Clinical evolution	Follow-up (years)
1	F	Post-IVH Hy	Markedly increased ICP	VP shunt (mp)	1	10	5	Mild headache	Asymptomatic	8
2	M	Congenital Hy	Moderately increased ICP	VP shunt (mp)	3	17	>10	none	Asymptomatic	8
3	M	Congenital Hy	Markedly increased ICP	VP shunt (mp)	3	21	5–10	none	Asymptomatic	5
4	F	Obstruct Hy (tumor)	Moderately increased ICP; hypo-thalamic dysfunction	Partial tumor resection + VP shunt (mp)	60	10	5	none	Multiple shunt malfunctions; asymptomatic for Chiari I	5
5	M	Congenital Hy	Moderately increased ICP	VP shunt (mp)	2	12	>10	none	Asymptomatic	6
6	M	Congenital Hy	Moderately increased ICP	VP shunt (mp)	1	11	>10	none	Multiple shunt malfunctions; asymptomatic	8
7	M	Postmeningitic Hy	Mildly increased ICP and r-hemiparesis	VP shunt (mp)	2	13	5–10	Sporadic mild headache	Asymptomatic	11
8	F	Posttraumatic Hy	Moderately increased ICP	VP shunt (mp)	36	6	5–10	none	Asymptomatic	5
9	F	Post-IVH Hy and DW variant	Markedly increased ICP	VP shunt (mp)	0.3	8	5	none	Multiple shunt malfunctions; asymptomatic	10
10	M	Post-IVH Hy	Moderately increased ICP	VP shunt (mp)	0.6	14	>10	none	Asymptomatic	4
11	M	Post-IVH Hy	Moderately increased ICP- right hemiparesis	VP shunt (mp)	1	12	5–10	Sporadic mild headache	Asymptomatic	7

HY hydrocephalus, *IVH* intraventricular hemorrhage, *ICP* intracranial pressure, *VP* ventriculo-peritoneal, *mp* medium pressure, *DW* Dandy Walker

Table 2 Summary of patients who underwent surgical procedure

Number	Sex	Diagnosis	Clinical manifestations	Initial treatment	Age at operation (months)	Age at Chiari I diagnosis (years)	Clinical manifestations	Tonsillar herniation degree and syrinx (mm)	Surgical treatment (age-years)	Postop evolution	Total follow-up (years)
12	M	Post-IVH Hy	Markedly increased ICP	VP shunt (mp)	0.3	3	Headache	>10	Bilateral frontal cranioplasty (5)	Worsening of epilepsy; new shunt malfunctions. Asymptomatic	5
13	M	Post-IVH Hy	Markedly increased ICP	VP shunt (mp)	1	3	Sporadic mild headache	5	Suboccipital craniectomy, C1 laminectomy and duroplasty (3)	Asymptomatic: after 1 yr new onset of syrinx on MRI-second operation	3
14	M	Hy Semilobar holoprosencephaly	Markedly increased ICP	VP shunt (mp)	0.6	11	Headache, upper limbs tremor, ataxia	>10, Cervical syrinx	Bilateral parieto-occipital cranioplasty (17)	Improvement of headache; after 1 yr one bone flap sunk-replacement with high pressure shunt	3
15	M	Fronto-T arachnoid cyst	Mildly increased ICP	Cysto-peritoneal shunt (mp)	3.5	5	Papilloedema with retinal hemorrhages; right VIth nerve palsy; headache	5	Lateral parietal cranioplasty (5)	Improvement	6
16	M	Hy + chiasmatic cyst	Moderately increased ICP	Cyst marsupialisation + cysto-ventriculo-peritoneal shunt (mp)	5	12	Headache; bilateral papilloedema and bilateral VIth nerve palsy	5–10	Bilateral parietal cranioplasty (14)	Persisting headache-ICP monitoring revealing high values-second and third operations	6
17	M	Post-IVH Hy	Moderately increased ICP	VP shunt (mp)	0.6	24	Vertigo, ataxia, spontaneous nistagmus	5–10	Bilateral parieto-occipital cranioplasty (24)	Mild improvement	2

HY hydrocephalus, *IVH* intraventricular hemorrhage, *ICP* intracranial pressure, *VP* ventriculo-peritoneal, *mp* medium pressure

Table 3 Two patients required further surgical procedure

Number	Initial surgical treatment	Postop evolution	Further major operations	Postop evolution	Follow-up (years)
13	Suboccipital craniectomy, C1 laminectomy and duroplasty	Asymptomatic: after 1 yr new onset of syrinx on MRI-second operation	Bilateral frontal cranioplasty	Asymptomatic; improvement of syrinx	2
16	Bilateral parietal cranioplasty	Persisting headache-ICP monitoring reviling high values-after 3 years second and after 6 more months third operations	2nd) Bilateral frontal cranioplasty; 3rd) Occipital cranioplasty	Asymptomatic	2

tremor, upper limbs hypoesthesia, and nystagmus in one case each.

In all the nine symptomatic patients, the first suspect was obviously a shunt malfunction which was ruled out by neuroradiological evidence of unchanged shape and size of the ventricles in comparison with the more recent exams performed during the routine follow-up control, by X-ray scans which excluded any mechanical interruption along the shunt system or malposition, by ultrasonography studies which did not demonstrate any abdominal fluid collections, and by blood serum exams which did not show any inflammatory signs.

Neuroradiological investigation

A small posterior cranial fossa was demonstrated in all the cases; this finding was proportional to patients' age, as the older patients disclosed the smallest posterior fossae. Small ventricles (though not properly "slit-like") were present in all the cases, as well as reduced periencephalic subarachnoid spaces (Fig. 1), except one patient who disclosed diffuse cerebral atrophy coexistent with normal posterior fossa neural structures. Cerebellar tonsil displacement below the basion–opisthion line varied from mild (5 mm) in five cases to moderate (5–10 mm) in six cases and severe (>10 mm) in six cases. The degree of tonsillar herniation did not correlate with the clinical manifestations. In fact, it was mild (5 mm beyond the Chamberlain's line) in five patients, among whom three underwent surgical procedures (Fig. 2); a moderate herniation (5 to 10 mm) was detected in six patients, of whom only two were symptomatic (Fig. 3); finally, the most severe degrees of tonsillar herniation (>10 mm) were observed in six patients (four asymptomatic and two symptomatic). Concomitant to tonsillar herniation, thickening of the skull vault and base was observed, as well as an increase in paranasal sinus volume; these modifications, which were interpreted as an effect of a satisfactorily working CSF shunt, did correlate with patient's age as they were more pronounced in the older ones.

Syringomyelia, was detected in only one patient, localized at the cervical region.

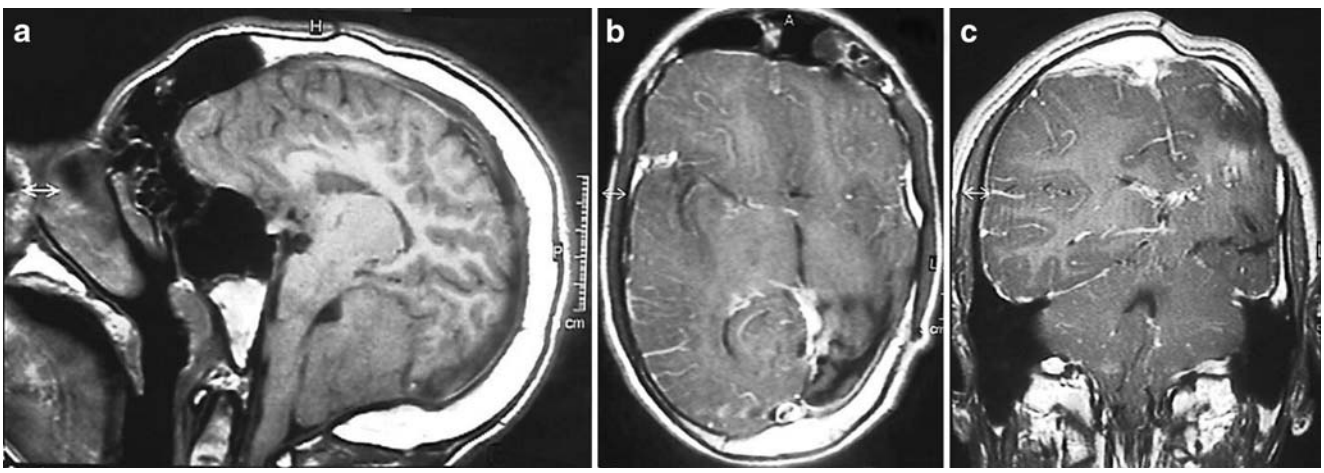

Fig. 1 MR sagittal T1-weighted image (**a**) and post-contrast axial and coronal T1-weighted images (**b**, **c**). This MRI, performed 13 years after ventriculo-peritoneal-shunt implantation for postmeningitic hydrocephalus, shows tonsillar ectopia and displacement of the superior vermis into the great vein of Galen cistern, thickening of the skull vault and base, and increased paranasal sinus volume. In the left hemisphere, the neonatal meningitis results can be observed

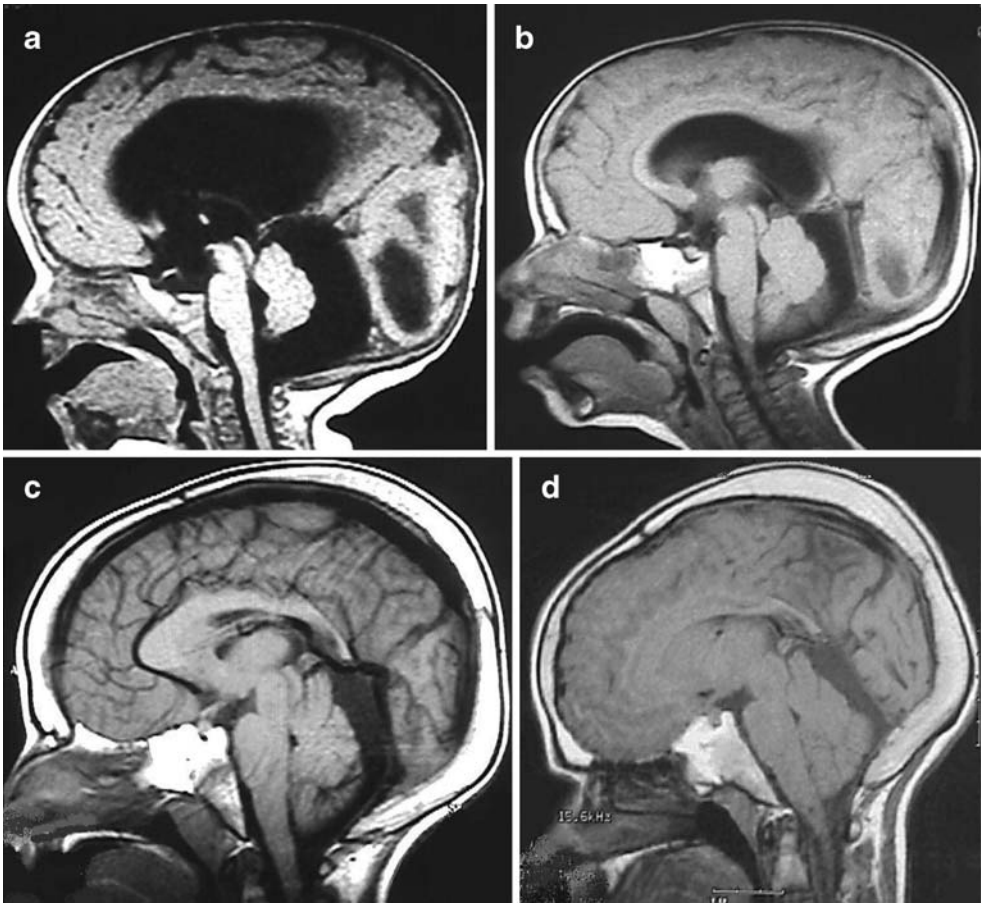

Fig. 2 MR sagittal T1-weighted images (**a**–**d**). Infant with congenital hydrocephalus who underwent a ventriculo-peritoneal CSF shunt placement at the age of 9 days. The preoperative MRI study, carried out in 1991 (**a**), does not show any evidence of cerebellar tonsils descent in spite of the obvious enlarged lateral cerebral ventricles. A progressive reduction of the volume of the subarachnoid space and ventricular system is associated with a progressive thickening of the calvarium. A progressive upward and downward displacement of the cerebellar tonsils is also evident (**b**–**d**)

ICP recording

Only three patients (cases 14, 15, and 16) underwent prolonged ICP recordings (48, 72, and 144 h, respectively) utilizing an ad hoc personally devised computerized system, already described in previous papers [7, 21, 22]. In two cases (patients 14 and 15), the ICP values were slightly superior to the normal range for about 43% and 56% of the recording time, respectively. The other patient (case 16) underwent ICP recording in a severely compromised clinical condition. Abnormal values were recorded only for a few minutes after positioning of the intraparenchymal sensor; thereafter, his clinical condition reversed rapidly to normal in the following hours, with an improvement in papilloedema and strabismus; concomitantly, ICP values reversed to normal and remained within normal ranges for the following 96 h. After a few days, a second prolonged ICP monitoring was performed when he was newly symptomatic; under these circumstances, ICP values were within the ranges of normality for 62% of the recording time but with markedly elevated peaks (>40 mmHg) during rapid eye movement phases of sleep.

Surgical treatment

Among the entire series, six patients underwent surgical treatment (Table 2). Four patients presented clinical manifestations, whereas two, who were mildly symptomatic at diagnosis, were judged to need an operation on the basis of MRI appearance and operated on in the care of other institution. Out of these two, one patient, who presented mild headache at diagnosis, underwent suboccipital craniectomy, C1 laminectomy, and duraplasty. The operation did not prevent the later occurrence of syringomyelia, which was the cause of referral to our center. The second patient underwent a bilateral frontal enlarging cranioplasty. The postoperative course was uneventful.

Four patients, three of them evaluated by ICP recording at our department, underwent cranial vault enlargement procedures. In two cases, a coronal incision was performed, and the calvaria was widely exposed; parietal flaps (unilateral in one case), anchored inferiorly to the temporal bones, were created and mildly displaced outwards; the new position was maintained by means of the interposition of osteoinductive material. The other two patients underwent a parieto-occipital decompression which included also

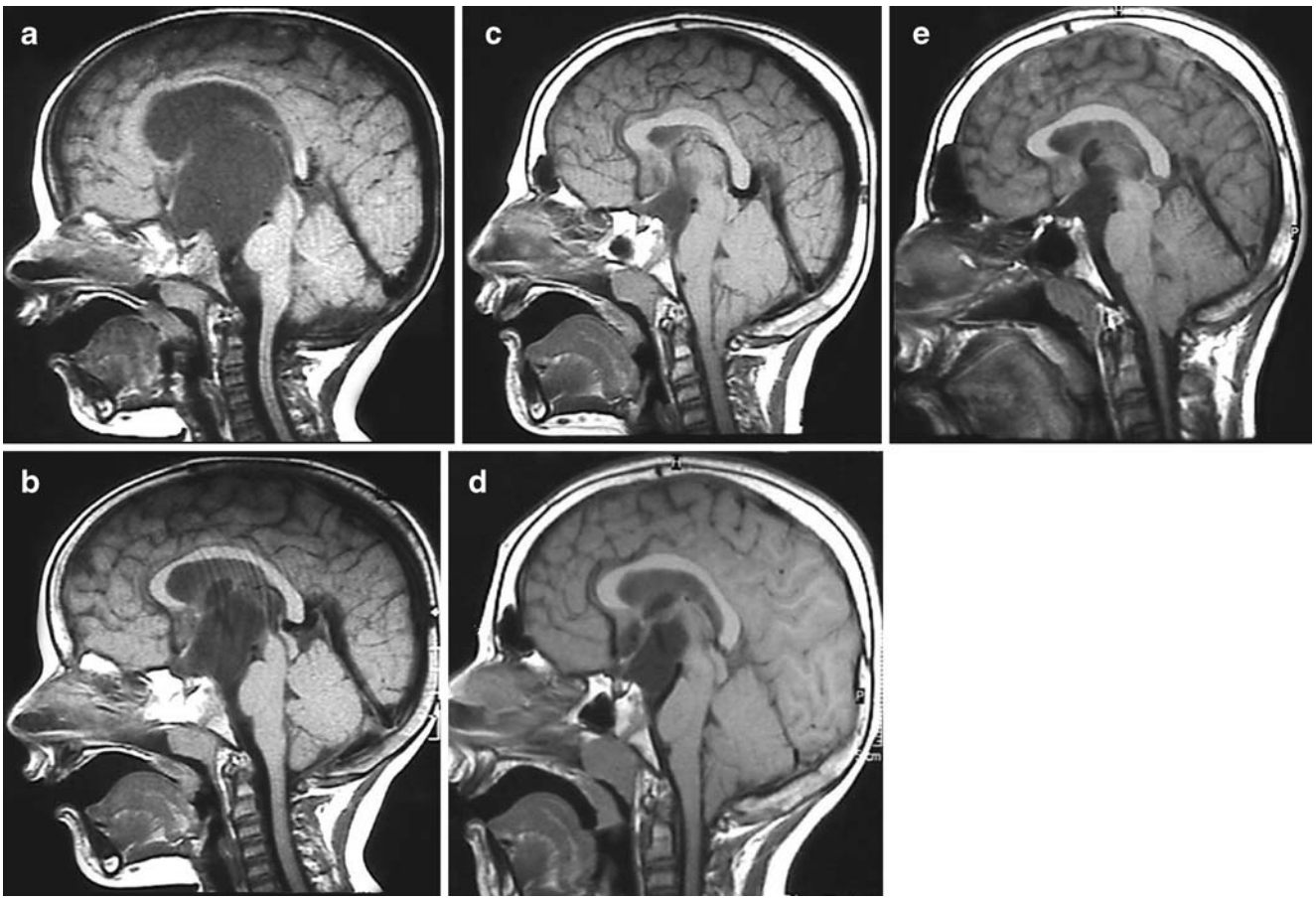

Fig. 3 MR sagittal T1-weighted images (**a**–**e**). **a**–**c** were already published [8] with kind permission of Springer Science+Business Media. This patient underwent cysto-ventriculo-peritoneal CSF shunt at 5 months of age for a huge suprasellar arachnoid cyst with associated hydrocephalus (**a**). After 1 year from the operation, the MRI (**b**) showed the progressive reduction of both ventricular and cystic volumes. During the follow-up, a progressive reduction of the intracranial volume, with thickening of the skull vault and base and increased paranasal sinus volume (**c**–**e**), was observed

a reduction of the tightness of the bone flap, obtained by drilling the inner bone. Postoperative course was uneventful in all the cases.

Clinical follow-up

Overall, the follow-up varied from 2 to 12 years (mean, 6).

Among the five patients who underwent the surgical procedure, an immediate relief of clinical manifestations was observed in all the cases. Headache subsided almost immediately, whereas ocular signs disappeared within 2 weeks. Nonetheless, normalization of clinical picture persisted at present in four out of the six patients. In fact, in two cases, further surgical treatment was performed for the new onset of cervical syringomyelia in one case previously treated with suboccipital decompression and for recurrent headache in a patient whose new ICP recording revealed high values. These repeated surgical procedures were performed after 1 and 3 years after the first operation, respectively.

The other 11 patients, either asymptomatic or those complaining only mild headache, maintain a stable clinical condition at the most recent follow-up examination and remain under strict neurological observation looking for the appearance of any clinical manifestation that should suggest surgical treatment.

Radiological follow-up

All patients underwent radiological evaluation periodically. Early postoperative MRI did not demonstrate significant variations of the neuroradiological picture.

On late neuroradiological investigations, the degree of tonsillar herniation resulted stable in all the patients.

Syringomyelia, which was present in one patient preoperatively, presented a significant reduction at last

follow-up. In contrast, the de novo appearance of a cervical syrinx was observed in one patient who had undergone suboccipital craniectomy, C1 laminectomy, and duraplasty in another institution.

Reoperations

After a period from the surgical treatment which ranged from 1 to 3 years (mean 2 years), two out of the six patients required further surgical procedures (cases 13 and 16; Table 3).

In one case, after 1 year of follow-up from the first operation (suboccipital craniectomy, C1 laminectomy, and duraplasty), the new onset of cervical syringomyelia was detected on neuroradiological exams. The patient, who presented only mild headache at diagnosis, did not show any clinical change; however, for the suspect of CSF dynamics, abnormality due to the new onset of cervical syringomyelia, the patient underwent bilateral frontal cranioplasty with progressive improvement of the syrinx. The second patient, who had undergone bilateral parietal cranioplasty, presented after 3 years recurrent headache; new ICP recording revealed high values again, and bilateral frontal cranioplasty was performed. Occipital decompressive cranioplasty was, then, performed after 6 months. At last follow-up (2 years), the patient is asymptomatic.

Discussion

Out of the various forms of hindbrain herniation first described by Hans von Chiari in 1894 [15], that which is referred to as "Chiari type I malformation" is characterized by a peg-like elongation of the cerebellar tonsils and medial aspect of the inferior vermis outside the cranial cavity into the cervical canal. The neuroradiological picture of Chiari type I malformation can be found in many clinical conditions. It can simply translate an increased ICP; it can manifest as an isolated malformation (with or without syringomyelia), or it can be associated with some congenital or acquired craniovertebral junction abnormalities, such as Klippel–Feil anomaly, atlas assimilation, platybasia, etc., or with some craniofacial dysostoses (like for instance Crouzon, Carpenter, and more rarely, Apert syndromes), and even with apparently non-syndromic craniosynostoses, all characterized by occipital bone involvement secondary to precocious fusion of the lambdoid suture [2]. In general, in the cases of the just mentioned clinical association, the caudal descent of the cerebellar tonsils may progress in time.

In adjunct to congenital forms, the neuroradiological picture of Chiari type I malformation has been also described as an acquired form, namely as a complication of lumbo-peritoneal shunting (performed either for communicating hydrocephalus or for relieving "benign" intracranial hypertension) [3, 12, 19], as well as procedures in which a chronic spinal leakage may be recognized [12, 23]. In these cases, in fact, the tonsillar descent seems to be justifiable by the presence of a pressure gradient across the cranial and spinal compartments, with a lower intraspinal pressure induced in some particular conditions like spinal cerebrospinal fluid drainage [19].

Quite recently, Chiari type I malformation has been reported as late complication of early treatment of supratentorial arachnoid cyst by means of cysto-peritoneal shunting [6, 9, 13, 14, 16]. This acquired form of Chiari I malformation following early CSF shunting (either ventriculo-peritoneal or cysto-peritoneal), is an intriguing late complication (or evolution?) of this surgery, and first of all, raises the question of its pathogenesis; actually, if tonsil herniation can be easily explained pre-operatively as an expression of increased ICP, it is much more problematic to explain its novel appearance as an effect of a satisfactorily working CSF shunt.

A possible explanation of this "developmental" anomaly might be sought in the modifications of skull growth induced by CSF shunting. Thickening of the cranial vault, with inward growth of the calvaria, was a usual finding in children affected by congenital hydrocephalus treated by means of extrathecal shunting. In fact, the abnormal pre-operative growth of head circumference led to a disproportion between the skull and its contents as a result of correctly functioning CSF shunt and consequently to the induction of inwards cranial growth to compensate for it. Even though nowadays surgical indication for congenital hydrocephalus is much more timely and consequently macrocrania only exceptionally reaches the levels recorded previously, the above radiological modification can still be appreciated on postoperative CT scan. Beside cranial vault modifications, other changes occur at the cranial base with thickening of the sphenoidal plane, lesser sphenoidal wings and posterior clinoids, and with volume increase of the paranasal sinuses. All these modifications are aimed at reducing the cranial-parenchymal disproportion brought about by CSF venting from the cranial cavity.

Although uniformly distributed, skull modifications with thickening of the occipital and temporal bones and increased volume of the middle ear and mastoid cells, appear to be relatively more prominent at the level of the posterior cranial fossa, probably due to the major crowding of the nervous structures at this level. In fact, if these modifications are usually devoid of clinical significance, an almost normal posterior cranial fossa like that observed pre-operatively in triventricular hydrocephalus may become significant in cases where the reduced supratentorial volume may impact on the volume of the posterior cranial

fossa. This condition eventually succeeds in an incompetent posterior fossa as demonstrated not only by cerebellar tonsil herniation into the spinal canal but also by the upward progressive displacement of the superior vermis into great vein of Galen cistern and even into the supratentorial space through the tentorial notch.

This phenomenon of brain tissue constrained within the skull in shunted children was initially described by Hoffman et al. [10] in 1976. Actually, some authors [18] do not agree completely with this theory, preferring the description of a "posterior cranial fossa disproportion" rather than "cephalocranial disproportion" because, comparing the supratentorial volume of shunted and nonshunted patients, no significant differences were detected. Thus, the main pathology seems to be restricted to an arrested posterior cranial fossa growth [17] even in cases of extrathecal shunts. They speculate that shunting may cause anyway an alteration of CSF flow dynamic, impairing consequently the mechanism involved in the brain development [18].

Actually, all but three of our patients had been treated in their early infancy with an extrathecal CSF shunt, and all of them exhibited the neuroradiological modifications of skull base and vault above reminded; all of them had small ventricles and "normal" volume of the brain. These features, which remind us of the so-called "slit ventricle syndrome"[20], could suggest to consider the neuroradiological and clinical findings observed in the series here considered as a "variant" of such a condition in which the caudal descent of the cerebellar tonsils is the most obvious sign together with the abnormal thick cranial vault. What appears more problematic to interpret is the lack of any relationship between the degree of tonsillar herniation or skull modifications (and the severity of Chiari malformation) and clinical manifestations. In fact, the most severe degrees of tonsillar herniation occurred more frequently in asymptomatic rather than symptomatic patients. The same applies to the development of an associated syringomyelia. The presence of syringomyelia, detected in one patient initially, and which appeared later on in another patient who had undergone initial suboccipital craniectomy, is usually considered a more reliable sign of the condition gravity and an indicator for surgical therapy, differently from what might be observed in purely Chiari I malformation.

What seems even more difficult to explain (even considering the limited number of our patients) is the association of a "symptomatic" Chiari I malformation with the shunting of a supratentorial cyst (either arachnoid cyst or that associated to semilobar holoprosencephaly). Data in the literature are in agreement with our observation. In fact, the neuroradiological picture of a Chiari I malformation was evident in two of ten shunt-dependent patients operated on (by means of a cyst-to-peritoneal shunt) for an intracranial arachnoid cyst by Kim et al. [13].

Surgical treatment of Chiari I malformation is traditionally based on the relief of the compression exerted by the bony and dural envelopes on neural structures at the level of foramen magnum.

In our belief, in patients with acquired Chiari I malformation developing after CSF shunting, once shunt malfunction and slit ventricle syndrome can be ruled out on the grounds of normal cerebral ventricles or those only minimally reduced in size, the presence of aspecific symptoms like headache is better explained on the grounds of an acquired microcrania secondary to progressive thickening of the vault.

On these grounds, for five patients, we considered that a classical foramen magnum decompression could succeed in favoring further downwards herniation of posterior fossa contents and eventually resulted in further and abrupt clinical deterioration, as described in the literature [11]. Consequently, our first surgical option was decompressive supratentorial skull enlargement. Indeed, decompressive craniotomy seemed to fit with the purpose to enlarge the intracranial volume without the risk of aggravating the hindbrain herniation. The procedure was successful in alleviating the clinical symptomatology in all the cases. Moreover, the patient, who had undergone initial suboccipital decompression and after 1 year presented the new onset of cervical syringomyelia requiring a second operation, also experienced the disappearance of the clinical symptomatology following a bifrontal cranioplasty as well as a progressive reduction of the syringomielic cavity at the radiological control examinations.

Even though the normalization of clinical manifestations was not associated with any significant modification in the postoperative neuroradiological exams, by excepting a slight increase in subarachnoid spaces, the clinical improvement which remained stable in our patients during a relatively long follow-up observation appears, in our opinion, sufficient to justify the surgical treatment.

Conclusion

The aim of this study is to serve as a contribution to the understanding of the underlying pathogenetic mechanisms of acquired Chiari type 1 malformation in cases of long-lasting supratentorial CSF shunting and to provide a base for planning the best management, whether conservative or surgical. However, due to the small number of this series, further studies are required to achieve a better knowledge of this condition.

Acknowledgement This study was supported by the Association "Ali di Scorta onlus, per la lotta alle malformazioni e ai tumori cerebrali infantili", www.alidiscorta.it.

References

1. Arnett BC (2004) Tonsillar ectopia and headaches. Neurol Clin 22:229–236
2. Caldarelli M, Di Rocco C (2004) Diagnosis of Chiari I malformation and related syringomyelia: radiological and neurophysiological studies. Childs Nerv Syst 20(5):332–335
3. Chumas PD, Kulkarni AV, Drake JM, Hoffman HJ, Humphreys RP, Rutka JT (1993) Lumboperitoneal shunting: a retrospective study in the pediatric population. Neurosurgery 32(3):376–383
4. Chumas PD, Armstrong DC, Drake JM, Kulkarni AV, Hoffman HJ, Humphreys RP, Rutka JT, Hendrick EB (1993) Tonsillar herniation: the rule rather than the exception after lumboperitoneal shunting in the pediatric population. J Neurosurg 78(4):568–573
5. Cinalli G, Spennato P, Sainte-Rose C, Arnaud E, Aliberti F, Brunelle F, Cianciulli E, Renier D (2005) Chiari malformation in craniosynostosis. Childs Nerv Syst 21(10):889–901
6. Di Rocco C, Tamburrini G (2003) Shunt dependency in shunted arachnoid cyst: a reason to avoid shunting. Pediatr Neurosurg 38 (3):164
7. Di Rocco C, Tamburrini G, Caldarelli M, Velardi F, Santini P (2003) Prolonged ICP monitoring in Sylvian arachnoid cysts. Surg Neurol 60(3):211–218
8. Di Rocco C, Velardi F (2003) Acquired Chiari type I malformation managed by supratentorial cranial enlargement. Childs Nerv Syst 19(12):800–807
9. Hassounah MI, Rahm BE (1994) Hindbrain herniation: an unusual occurrence after shunting an intracranial arachnoid cyst. Case report. J Neurosurg 81(1):126–129
10. Hoffman HJ, Tucker WS (1976) Cephalocranial disproportion. A complication of the treatment of hydrocephalus in children. Childs Brain 2(3):167–176
11. Holly LT, Batzdorf U (2001) Management of cerebellar ptosis following craniovertebral decompression for Chiari I malformation. J Neurosurg 94(1):21–26
12. Johnston I, Jacobson E, Besser M (1998) The acquired Chiari malformation and syringomyelia following spinal CSF drainage: a study of incidence and management. Acta Neurochir (Wien) 140:417–427
13. Kim SK, Cho BK, Chung YN, Kim HS, Wang KC (2002) Shunt dependency in shunted arachnoid cyst: a reason to avoid shunting. Pediatr Neurosurg 37(4):178–185
14. Lazareff JA, Kelly J, Saito M (1998) Herniation of cerebellar tonsils following supratentorial shunt placement. Childs Nerv Syst 14:394–397
15. Loukas M, Noordeh N, Shoja MM, Pugh J, Oakes WJ, Tubbs RS (2008) Hans Chiari (1851–1916). Childs Nerv Syst 24(3):407–409 Mar
16. Mendonça R, Lima TT, Oppitz PP, Raupp SF (2006) Herniation of the cerebellar tonsils after suprasellar arachnoid cyst shunt: case report. Arq Neuropsiquiatr 64(2B):523–525
17. Nishikawa M, Sakamoto H, Hakuba A, Nakanishi N, Inoue Y (1997) Pathogenesis of Chiari malformation: a morphometric study of the posterior cranial fossa. J Neurosurg 86(1):40–47
18. Osuagwu FC, Lazareff JA, Rahman S, Bash S (2006) Chiari I anatomy after ventriculoperitoneal shunting: posterior fossa volumetric evaluation with MRI. Childs Nerv Syst 22(11):1451–1456
19. Payner TD, Prenger E, Berger TS, Crone KR (1994) Acquired Chiari malformations: incidence, diagnosis, and management. Neurosurgery 34(3):429–434
20. Rekate HL (2008) Shunt-related headaches: the slit ventricle syndromes. Childs Nerv Syst 24(4):423–430
21. Tamburrini G, Di Rocco C, Velardi F, Santini P (2004) Prolonged intracranial pressure (ICP) monitoring in non-traumatic pediatric neurosurgical diseases. Med Sci Monit 10(4):MT53–MT63
22. Tamburrini G, Caldarelli M, Massimi L, Santini P, Di Rocco C (2005) Intracranial pressure monitoring in children with single suture and complex craniosynostosis: a review. Childs Nerv Syst 21(10):913–921
23. Tubbs RS, Law C, Oakes WJ, Grabb PA (2004) Acquired Chiari I malformation following baclofen pump placement in a child. Case report. J Neurosurg 101(2 Suppl):211–3