Childs Nerv Syst (2003) 19:800–807
DOI 10.1007/s00381-003-0837-7

ORIGINAL PAPER

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Acquired Chiari type I malformation managed by supratentorial cranial enlargement

Received: 16 July 2003
Published online: 28 October 2003

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Abstract *Introduction:* Acquired Chiari type-I malformation in hydrocephalic patients who have undergone surgical treatment was initially thought to depend on a craniocephalic disproportion induced by the cerebrospinal (CSF) shunt. However, most of the reports in the literature deal with children with lumbo-peritoneal shunts and emphasize the pathogenic role of the cranio–spinal pressure differential across the foramen magnum brought about by this type of shunt. *Method:* In the present report, the authors describe two further cases of symptomatic acquired Chiari type-I malformation observed in two adolescents operated on for correction of pseudotumor cerebri in one (lumbo-peritoneal shunt) and of a suprasellar arachnoid cyst (cysto-ventriculo-peritoneal shunt) in the other. *Results:* In both subjects, both the clinical manifestations and the cerebellar tonsillar herniation regressed after supratentorial cranial expansion, without the need for any manipulation of the shunt devices implanted earlier. *Discussion:* These results, together with the observation of the concomitant upward and downward herniation of the cerebellum in both patients, indicate that secondary craniocephalic disproportion plays a relevant role in the genesis of acquired Chiari type-I malformation in children bearing extrathecal CSF shunts.

Keywords Chiari malformations · Acquired Chiari type-I malformation · Lumbo-peritoneal shunt · Ventriculo-peritoneal shunt · Craniocephalic disproportion · Cranial enlargement

Introduction

Caudal displacement of the cerebellar tonsils with foramen magnum impaction, identical morphologically to the congenital anomaly usually referred to by the eponymous name of Chiari type-I malformation, was described for the first time in 1976 by Hoffman and Tucker [1], who reported it as a late complication of extrathecal shunt drainage in eight hydrocephalic patients. Six of these eight subjects had undergone placement of a valveless lumbo-peritoneal (L-P) shunt and 2, of a ventriculo-peritoneal (V-P) shunt. Cephalocranial disproportion caused by post-shunting secondary craniosynostosis and resulting in a small skull that was unable to accommodate for brain growth was specified by the authors as the explanation for the phenomenon. Posterior fossa cranial decompression was the therapeutic procedure utilized to resolve this complication.

In 1981, Welch et al. [2], describing a similar course in five hydrocephalic subjects each operated on to place a spinal cysto-peritoneal (1 case) or a lumbo-ureteral (4 cases) shunt pointed out the possible pathogenetic role of spinal subarachnoid shunts in generating negative pressure gradients between the spinal and the cranial compartments. To treat their patients the authors utilized a combination of posterior fossa decompression procedures and conversion of the lumbo-ureteral shunts to ventriculo-ureteral or V-P shunts.

The gradient pressure hypothesis was further emphasized by Payner et al. [3], who 13 years later commented on the high incidence of acquired Chiari type-I malformation in patients treated with L-P shunts (seven cases, including four symptomatic cases, out of a series of ten).They suggested that conversion from L-P to V-P shunting should be tried before proceeding to posterior fossa decompression.

In the present report we describe two further cases of acquired Chiari type-I malformation in patients bearing extrathecal cerebrospinal fluid (CSF) shunts; both the patients presented with clinical symptoms and signs and neuroimaging evidence of so-called slit ventricle syndrome. The first patient had had an L-P shunt implanted because of benign intracranial hypertension, while the second had undergone placement of a cysto-ventriculoperitoneal shunt to drain a suprasellar arachnoid cyst associated with hydrocephalus. In both patients an operation for supratentorial cranial expansion successfully normalized the clinical picture without any modification of the CSF shunting device already in place; in one of them the cerebellar tonsils resumed their normal intracranial position.

Case reports

Case 1

This 15-year-old girl was first admitted to our institution at the end of 1993, with a few months' history of worsening headache and gradually progressive visual impairment. At the time of her first referral, neurological examination disclosed no focal neurological deficit; however, bilateral papilledema was detected by ophthalmological examination. Magnetic resonance imaging (MRI) studies ruled out intracranial space-occupying lesions but demonstrated abnormally small lateral cerebral ventricles. The cerebellar tonsils were above the rim of the foramen magnum, and the cisterna magna was patent (Fig. 1a, b). Idiopathic intracranial hypertension was diagnosed on the grounds of intracranial pressure (ICP) recordings. Because various attempts to normalize ICP during repeated hospital admission periods had failed, a medium-opening-pressure L-P shunt was placed in March 1994. After the procedure, the patient's headaches subsided. A progressive improvement of visual function was then observed, and she became completely asymptomatic less than 4 months after surgical treatment. The girl did well until August 2000, when she reported the onset of episodic headaches, typically associated with an orthostatic posture and decreasing in severity during clinostatism. No associated changes in visual function were observed. MR imaging demonstrated normal-volume cerebral ventricles and showed that the cerebellar tonsils had descended into the foramen magnum (Fig. 2a–c) with no signs of associated syringohydromyelia (Fig. 2d). Because the clinical picture had become progressively worse, brain and spinal MRI examinations were repeated in January 2002, confirming the tonsillar herniation and also revealing a slight enlargement of the spinal central canal at the thoracic level. At that time, the patient was hardly capable of maintaining a standing position for more than 30 min before suffering the onset of headache, which could be controlled only by adopting a recumbent position. No focal deficits were detectable, but examination of the fundus oculi again revealed bilateral papilledema. Prolonged ICP monitoring showed that ICP was moderately above the normal range, with waveform morphology indicating venous engorgement (Fig. 3a). Prolonged periods of ICP elevated at levels markedly over 50 mmHg were observed during the recordings (Fig. 3b, c).

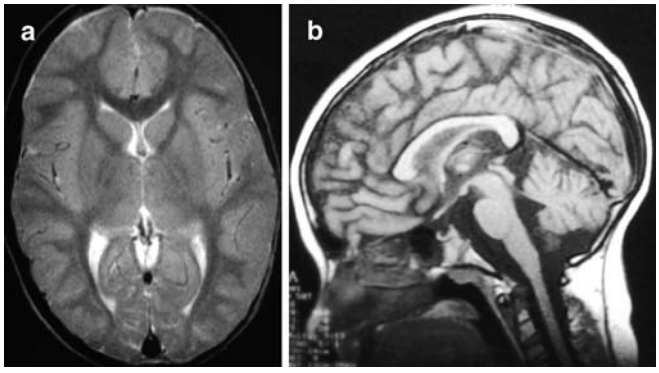


Fig. 1a, b Case 1. MRI study at the time of insertion of the lumboperitoneal shunt. Note the relatively small cerebral ventricles (**a**) and the normal aspect of the posterior cranial fossa (**b**)

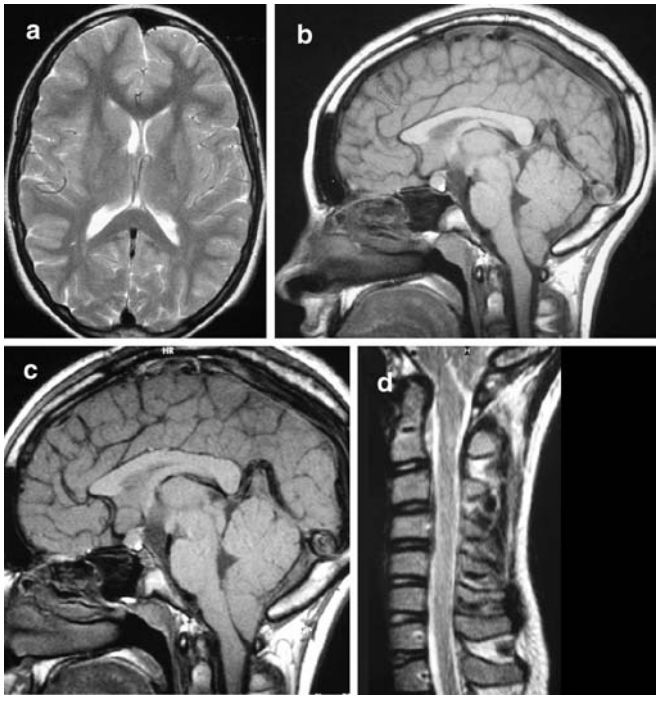
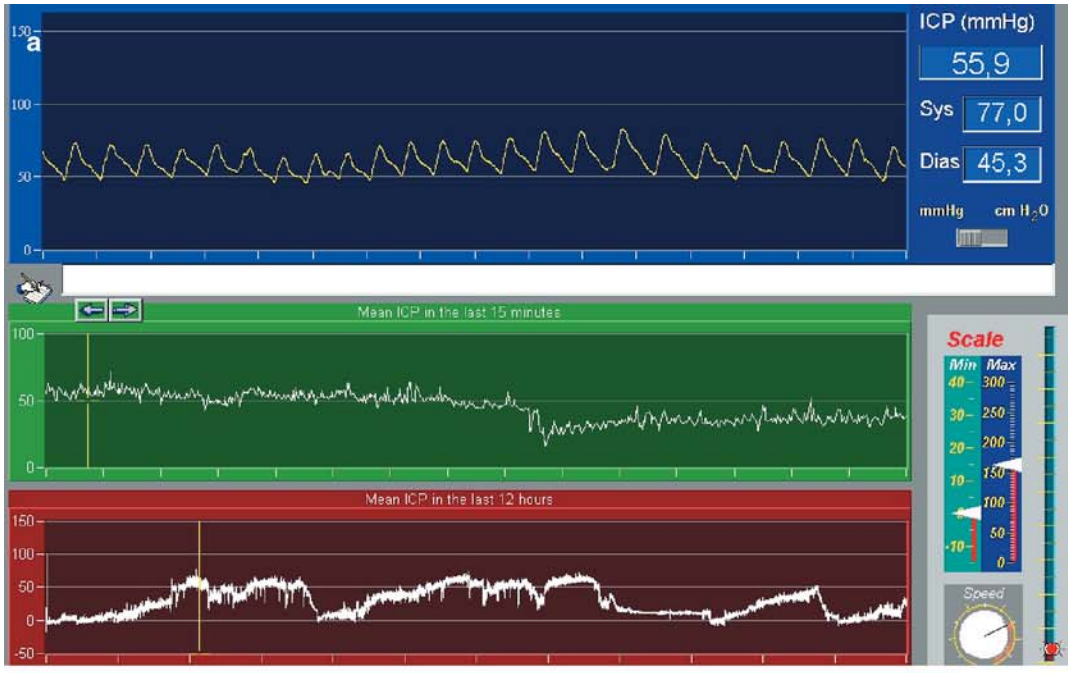


Fig. 2a–d Case 1. Six years after the spinal shunt insertion: the ventricular system is normal (**a**), but the cerebellar vermis appears to be growing upward while the cerebellar tonsils are dislocated caudally (**b**, **c**). There is no associated hydromyelia (**d**)

On 20 March 2002 the patient underwent a surgical procedure of supratentorial cranial decompression. A temporo-parietal bone flap was raised on each side (Fig. 4a). The bone flaps were elevated to increase cranial volume and maintained in the elevated position by the interposition of two small bone pieces derived from the posterior parietal margin of the bone flap (Fig. 4b). To minimize the cosmetic impact of the expanding craniotomy, accurate surface recontouring of the skull was realized (Fig. 4c, d).


a
ICP (mmHg)
55,9
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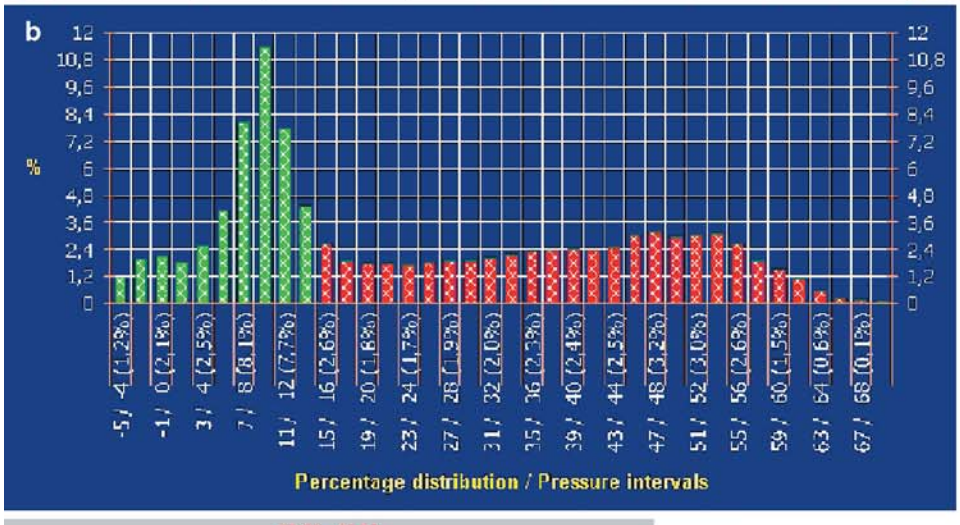
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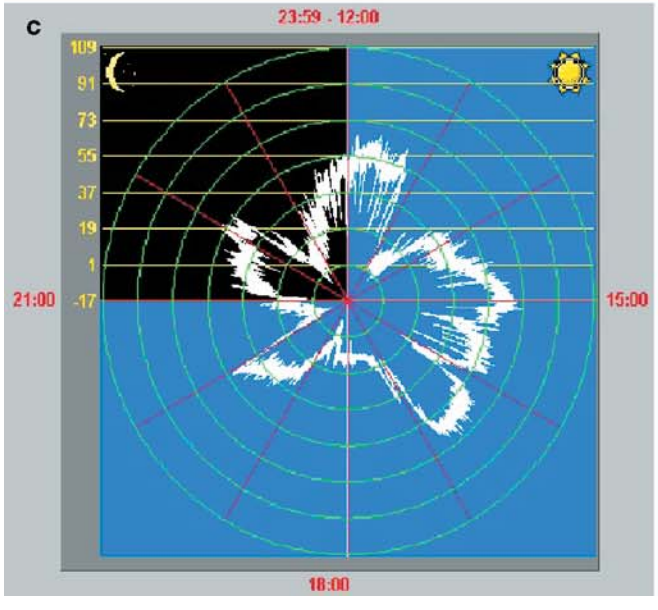
c
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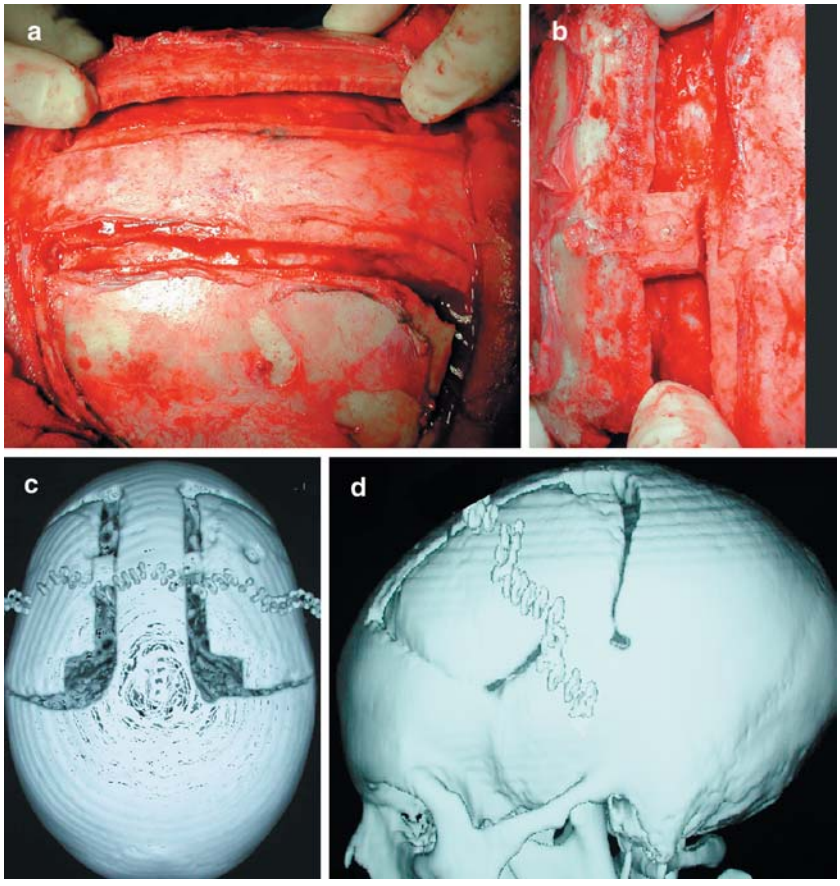

Fig. 4a–d Case 1. **a**, **b** Peroperative images during the operation for cranial enlargement: **a** creation and **b** elevation of the parietal bone flaps. **c**, **d** Postoperative 3D CT study demonstrating the satisfactory cosmetic outcome obtained by recontouring of the skull

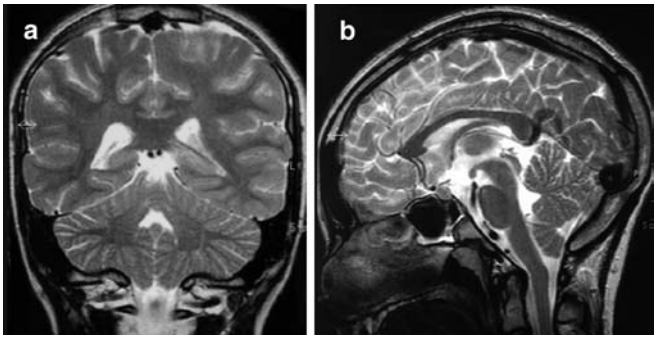

Fig. 5a, b Case 1. Three months after the operation for cranial enlargement: **a** antero-posterior and **b** lateral MRI studies showing regression of the caudal tonsillar herniation

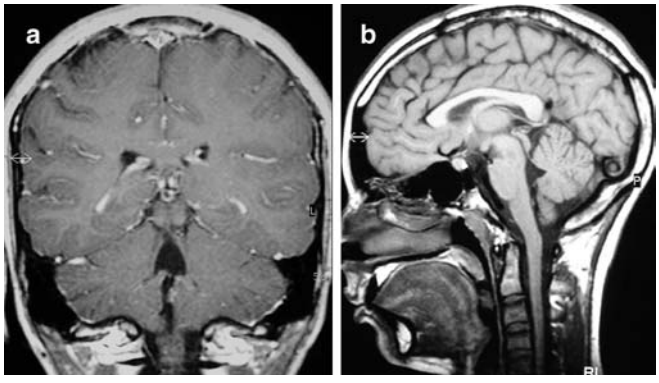

Fig. 6a, b Case 1. Six months after the operation for cranial enlargement: **a** antero-posterior and **b** lateral MRI studies demonstrating further "ascent" of the cerebellar tonsils within the posterior cranial fossa

Fig.3 a–c Case 1. Long-time intracranial pressure (IPC) recording at the time of the clinical symptomatology. The mean ICP is above normal limits, and the pulse wave morphology suggests increased venous pressure (**a**). During the recording, numerous episodes of abnormally elevated ICP are ascertained (**b**, **c**)

After surgery the patient experienced a progressive reduction of standing position headache. Ocular symptoms receded as well. Brain MRI study, performed at 3 months post-operatively showed a significant regression of tonsillar caudal descent (Fig. 5a, b). The cerebellar tonsils reached their normal intracranial position 6 months after the operation, as documented by repeated MRI study (Fig. 6a, b).

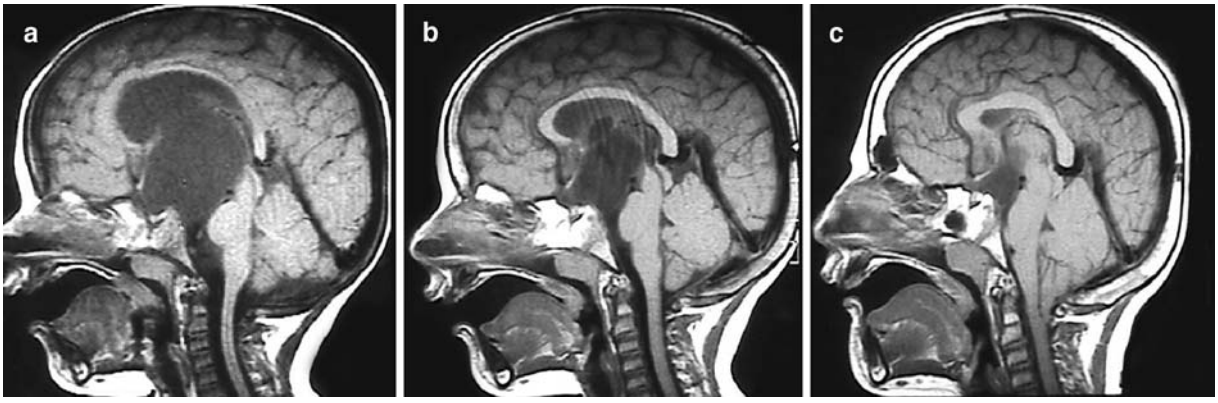

Fig. 7a–c Case 2. MRI studies at different ages of the patient. **a** MRI at 5 months, showing a huge suprasellar arachnoid cyst with associated hydrocephalus. The posterior cranial fossa morphology is normal. **b** One year after implantation of a cysto-ventriculo-peritoneal CSF shunt device, when the volume of both the arachnoid cyst and the ventricular system is obviously diminished; the cisterna magna is also slightly smaller. **c** Twelve years later, the intracranial volume is reduced because of thickening of the calvarial bone. Crowding of neural and vascular structures within the small cranial posterior fossa, with both upward and downward herniation of the cerebellum

Case 2

This 15-year-old boy presented with signs and symptoms of intracranial hypertension soon after birth. At 1 month of age he underwent a computed tomography (CT) scan, which revealed the presence of a large suprasellar arachnoid cyst with associated hydrocephalus (Fig. 7a). Marsupialization of the cyst lining through a pterional approach and placement of a V-P CSF shunt were carried out elsewhere. While apparently normalizing the ICP and decreasing the size of the lateral cerebral ventricles, the association of these two procedures failed to reduce the volume of the cyst significantly. At the age of 5 months, the child was admitted to our hospital for further treatment. A fenestrated catheter tailored to connect the cyst with the left lateral cerebral ventricle was implanted stereotactically in place of the standard ventricular catheter of the pre-existing V-P shunting device in April 1988. A subsequent reduction in the volume of the cyst and of the lateral cerebral ventricles was documented by serial CT and MRI scans (Fig. 7b). The boy's psychomotor development was satisfactory. The child's clinical condition remained substantially stable over the next few years, even though progressive asymptomatic upward herniation of the cerebellar vermis and caudal descent of the cerebellar tonsils were observed on control MRI after the age of 11 years. At the same time, a further reduction in the volume of the ventricular and peripheral subarachnoid spaces and progressive thickening of the cranial vault due to bone deposition at the inner cranial surface (Fig. 7c) were observed. In February 2002 the patient started to complain of intermittent diplopia and episodes of severe headache. An ophthalmological examination revealed bilateral papilledema with retinal hemorrhages in the left eye. In spite of medical treatment, the alterations to the ocular fundi were confirmed 2 months later, when neurological examination showed right-sided mild hemiparesis and mild dysmetria of the left upper limb. The boy was readmitted to our department for long-time ICP recording. This investigation demonstrated that mean ICP levels were within the normal limits. However, transient phases of intracranial hypertension (up to 40 mmHg) were recorded, occurring prevalently during nocturnal sleep and occasionally during the day.

On these grounds, a decompressive supratentorial craniotomy was carried out with the creation of two parietal bone flaps, which were maintained open and elevated relative to the cranial surface by the interposition of bone segments at the craniotomy sides. The procedure was followed by the immediate and lasting (follow-up period: 24 months) disappearance of the clinical manifestations. The fundus oculi examination revealed the disappearance of the retinal hemorrhages and improvement of the papilledema. Postoperative MRI examination demonstrated a mild ascent of the cerebellar tonsils 3 months after surgery, and a further ascent to the level of the occipital rim 1 year after the operation.

Discussion

Clinical and MRI observations have unequivocally demonstrated the possibility of hindbrain herniation late in life in subjects without congenital Chiari type-I malformation. Such an event has been found to occur in particular in some forms of faciocraniosynostosis, namely Crouzon's and Pfeiffer's syndromes, in cases of cranial bone disorders, such as osteopetrosis or congenital posterior fossa hypoplasia, that result in a diminished cranial volume, and in patients in whom an extrathecal CSF shunt has been implanted for the treatment of communicating hydrocephalus or pseudotumor cerebri [1, 2, 3, 4, 5, 6, 7, 8].

In 1994, Payner et al. [3] reviewed a series of 26 cases of acquired Chiari malformation described in the literature up to then, for 19 of which imaging studies and clinical histories adequate for the analysis were available, plus 4 personal cases. They found that 21 patients had each received a spinal subarachnoid shunt and 2, a ventricular shunt before demonstration of the hindbrain anomaly. In their discussion of this series the authors point out the two main theories available to explain the phenomenon: the "craniospinal pressure gradient theory "and the "cephalocranial disproportion theory."

The craniospinal pressure gradient theory postulates that the driving force behind the descent of the cerebellar tonsils in previously normal subjects is generated by a pressure gradient across the cranial and spinal compartments. The prototype of a such a pathogenetic interpretation might be seen in the early explanations of Chiari malformations in subjects with myelodysplasia, whose caudal hindbrain displacement was thought to depend on the antenatal escape of CSF through the spinal defect, resulting in lower intraspinal pressures [9, 10, 11].

According to the various authors who have evoked the craniospinal pressure gradient to explain the downward movement of the cerebellar tonsils, spinal pressure levels that are lower than the ICP values can be the effect of (i) impaired arachnoid granulation CSF absorption at cranial level in the presence of preserved arachnoid function at spinal level [3]; (ii) repeated lumbar punctures [12] or excessively prolonged continuous lumbar drainage [13], or (iii) spontaneous spinal CSF leakage [14] and the action of spinal subarachnoid CSF draining devices [1, 2, 5, 6, 15, 16].

Just prior to the study by Payner et al. [3], Chumas et al. [5, 6] had elaborated the concept of acquired Chiari I as a specific complication of L-P shunts and a possible cause of sudden death. The observation that a child who had died because of shunt failure had been found to have severe narrowing of the neural structures at the foramen magnum [17] had motivated the authors to review their own series of 143 L-P shunts in infants and children with regard to the position of the cerebellar tonsils. Thirty-eight (70%) out of 54 subjects with late CT scans demonstrated neural crowding at the cranio–cervical junction. Five patients with symptoms and signs of hindbrain dysfunction required posterior cranial fossa decompression and, in the whole group, about one third of the patients had their L-P shunts converted to V-P shunts [6].

In a recent paper, Rekate and Wallace [18] challenged the association of acquired Chiari I and L-P shunts as they had observed no cases of such a complication in a series of 26 patients (2 cases of symptomatic pseudomeningocele after posterior fossa craniotomy for brain tumors, 12 cases of pseudotumor cerebri, and 12 cases of slit-ventricle syndrome) they had treated by L-P shunts. The authors, however, apparently subscribed to the pressure gradient theory. In fact, they explained the absence of acquired Chiari I in their series on the basis of the surgical technique and hardware utilized and suggested incorporation of a high-resistance valve in the CSF-draining system to reduce the pressure differential across the foramen magnum.

Rekate and Wallace pointed on two further differences between their series and that published by Chumas et al. [5, 6], namely the etiology of the CSF dynamics disorders and the older age of their patients at the time of the L-P shunt insertion, as all of them had the shunt implanted after brain growth was complete.

The corollary of the pressure gradient theory as far as treatment is concerned is that the pressure differential between the cranial and spinal compartments needs correction to counteract the neural crowding at the foramen magnum. Consequently, three main surgical options have been utilized: posterior cranial fossa decompression, removal of the L-P and substitution of a V-P shunt, and a combination of both these procedures. The combination of posterior fossa decompression and conversion of the L-P shunt to a V-P shunt is reported to be associated with better and more lasting results [3].

Though the favorable results obtained with the above procedure seem to support the pressure gradient theory, it is evident that this hypothesis cannot explain the caudal descent of the cerebellar tonsils, which has been observed in children with ventricular shunts [1, 7, 19] or more generally in those with shunts draining the intracranial cavity, as in our second case. For these cases, the cephalocranial disproportion theory seems to provide an effective pathogenetic hypothesis. The shunting procedure is, in fact, commonly associated with an arrest of cranial growth. In the case of excessive CSF withdrawal from the ventricular subarachnoid cortical spaces the presence of the CSF shunt may interfere with the expansion of the neurocranial capsule and lead to secondary microcrania so that further brain growth can no longer be accommodated. Cephalocranial disproportion is the obvious cause of acquired Chiari I in subjects with faciocranial synostoses, which mean that the postnatal growth of the cerebellum cannot be contained within the hypoplasic posterior cranial fossa [4].

The main argument propounded against the cephalocranial disproportion theory is the time interval between the arrest of cranial growth induced by the insertion of the ventricular shunt and the occurrence of symptomatic tonsillar herniation, which usually occurs some years after the shunt procedure [3]. This objection is easily rejected with reference to the observation that in several patients placement of the extrathecal shunt has been followed not only by an immediate arrest of cranial growth, but also by progressive thickening of the cranial vault as the result of progressive inner apposition of bone, which extends over years, as in the two cases reported in this paper.

A second observation that supports the cephalocranial disproportion theory as an explanation for acquired tonsillar ectopia in shunted children is that the hindbrain caudal herniation is associated with upward displacement of the upper cerebellar vermis to occupy the quadrigeminal cistern. In other words, the overcrowding of the neural structures within the posterior cranial fossa not only induces the descent of the cerebellar tonsils into the upper cervical canal in these children, but also determines upward progressive growth of the cerebellum within the

"empty" spaces of the quadrigeminal plate and the great vein of Galen cisterns (Figs. 2c, 7c). These cisterns clearly offer an additional space for cerebellar expansion besides the cisterna magna.

The pathogenesis of acquired cerebellar vermian and tonsillar ectopia is probably multifactorial. However, the two cases described here seem to demonstrate the relevant role played in the genesis of this complication by the progressive reduction in intracranial volume induced by the presence of the CSF shunt, which results in a relatively hypoplasic skull that is unable to accommodate to the brain growth.

Should the cephalocranial disproportion prove to be the main, or at least one of the main, factors responsible for acquired Chiari I "malformation," the therapeutic options currently available will need to be challenged. Posterior cranial fossa decompression, though sufficient to relieve the symptomatology, at least at in the short term, could not be an adequate procedure in the long run. This type of procedure is addressed at correcting the effects rather than the cause of the condition and could actually result in a further descent of the cerebellar tonsils. Postoperative cerebellar ptosis has in fact been described in patients with suboccipital decompression for Chiari I malformation, though it was interpreted as the result of an excessively large suboccipital craniectomy [20].

The favorable results obtained by the conversion of the L-P shunt to a V-P shunt in the case of acquired cerebellar ectopia might, then, be explained not only on the basis of the elimination of the pressure gradient across the foramen magnum but also on the grounds of the additional intracranial volume created by the ventricular distension. This interpretation is supported by the observation that tonsillar herniation regresses after endoscopic third ventriculostomy [21], embolization of Galen vein aneurysms [22] and supratentorial tumor removal [23, 24].

If the craniocephalic disproportion theory is accepted, cranial expansion would be a logical further surgical option in the management of acquired cerebellar upward and caudal herniation, as apparently demonstrated by the results obtained in our two cases. It is noteworthy that the resolution of tonsillar herniation and of the associated clinical symptomatology was obtained in these cases with no need for any manipulation of the CSF shunt devices already in place. Even in the girl with the L-P shunt, the spinal shunt device was left in place and did not impede the ascent of the cerebellar tonsils after the supratentorial cranial enlargement. (We are not aware of previous similar cases reported in the literature.) The advantages of cranial expansion are evident: the procedure is not burdened by significant complications, it can be utilized regardless of whether children have L-P or V-P shunts, and it does not require any manipulation of CSF shunt devices that are already in place.

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