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Treatment of failed Adult Chiari Malformation decompression with CSF drainage: observations in six patients

G. K. Bejjani[1], **K. P. Cockerham**[2], **W. E. Rothfus**[3], **J. C. Maroon**[1], and **M. Maddock**[1]

[1] Tristate Neurosurgical Associates-UPMC, University of Pittsburgh Medical Center, PA, USA
[2] Department of Ophthalmology, Allegheny General Hospital, PA, USA
[3] Department of Radiology, University of Pittsburgh Medical Center, PA, USA

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Summary

Objective. We report the use of CSF drainage for the management of failed Adult Chiari Malformation (ACM) decompression.

Methods. All patients with more than one year follow-up after treatment of their failed ACM were included in this study. They underwent initial decompression between September 1998 and April 2000. Clinical and radiological data were collected initially and at recurrence. Lumbar punctures (LP) were done at recurrence for diagnostic and therapeutic purposes. Opening pressures and symptomatic relief were recorded. Therapeutic options included intermittent LP and ventriculo-peritoneal shunting (VPS).

Results. There were 6 patients (5 females and one male). Their age ranged from 19 to 43 years. Tonsillar descent ranged from 5 to 21 mm. The symptoms recurred 1.5 to 9 months postoperatively (average 5.6 months). Postoperative imaging revealed the presence of CSF flow behind the tonsils and the formation of a retrotonsillar neocistern in all patients. On LP, the opening pressure ranged from 17 to 31 cm of water (average 23 cm). All patients improved after CSF drainage, and four patients underwent VPS. The other patients were treated with repeat LP $\pm$ Acetazolamide. There was significant improvement in all patients, with 18 months follow-up after CSF drainage (range 16–21 months).

Conclusion. Our results suggest a role for CSF drainage in the treatment of some patients with failed ACM surgery. Possible explanations for the failure of ACM surgery in this subgroup include: surgical complications leading to neural hydrodynamic alteration, inadequate initial surgery, and coexistence with another pathology, possibly a mild form of intracranial hypertension. More prospective and hydrodynamic studies are needed to further clarify these issues.

Keywords: Adult Chiari Malformation; CSF drainage; IIH-WHOP; tonsillar ectopia.

Introduction

The treatment of Adult Chiari Malformation (ACM) has consisted mostly of sub-occipital decompression with various technical modifications [8, 24, 27, 57, 77]. Some authors have also proposed ventricular shunting for syringomyelia associated with hindbrain anomalies [10, 31, 32, 49], especially when hydrocephalus is present [76]. In some of the large published series, postoperative improvement was encountered only in 46 to 65% of patients, with clinical stabilization in another 16 to 31% [23, 35, 50, 53]. With over 3500 decompressions for ACM performed annually in the USA (AANS Procedural Statistics), surgical failure becomes a significant problem. The reasons for failure are numerous, as are the therapeutic alternatives [5, 26, 42, 77]. We report here the use of CSF drainage in the form of ventriculo-peritoneal shunting (VPS) or intermittent lumbar punctures (LP) to treat failed decompression for adult Chiari malformation without accompanying syringomyelia.

Methods

We included in this study all the patients with ACM who had more than one year follow-up after instituting the proposed treatment for their failed decompression. They consisted of six patients who underwent initial surgical treatment by the primary author between September 1998 and April 2000 at Allegheny General Hospital and the University of Pittsburgh Medical Center. Preoperative MRI's and medical records were reviewed. All these patients had Type I Chiari Malformation as defined by the radiological criteria used by Aboulezz [1], Barkovich [4], and Mikulis [43]. Their symptoms were thought to be consistent with the ACM as described by Milhorat *et al.* [44]. Due to the severity of symptoms and their effect on the patients' quality of life, surgery was recommended.

Initial surgery consisted in suboccipital craniectomy and C1 laminectomy with duraplasty using allografts. Every attempt was made to preserve the arachnoid membrane intact to avoid blood spillage which could lead to an inflammatory reaction. Intra-arachnoid dissection was avoided in all cases. Adequate CSF flow was assessed intra-operatively by inspection of the arachnoid membranes and the subarachnoid spaces using the operating microscope and looking for systolic expansion of the CSF spaces around the tonsils. Adequate CSF flow was documented in each case before duraplasty. There was no change in surgical technique during that period.

Treatment failure was defined as symptomatic recurrence. This occurred usually after a period of transient relief. After symptomatic recurrence, clinical data was collected again, and 4 of the 6 patients underwent formal ophthalmological evaluation by a neuro-ophthalmologist. All patients also underwent repeat MRI's and cine-flow studies. The pre and postoperative radiological studies were reviewed by a neuro-radiologist who was blinded to the clinical results and the aim of our study. He evaluated the amount of preoperative tonsillar herniation, the adequacy of decompression, and the presence of cerebellar and hindbrain sag. The technique advised by Duddy and Williams [14] was used to quantify the amount of cerebellar sag. On a mid-sagittal MRI view, a line was drawn between the middle of the posterior edge of the hard palate and ran tangential to the upper surface of the anterior arch of the atlas. From this baseline, the height of the median dorsal recess of the fourth ventricle or fastigium (F) was used to assess the level of the cerebellum, and the height of the upper surface of the pons (P) was measured to assess the level of the brainstem. The pre (F1 and P1) and postoperative (F2 and P2) values were determined. The average of the difference between the postoperative and preoperative values was used to determine the average ascent using the following formula:

$$\text{Ascent} = [(\text{P2} - \text{P1}) + (\text{F2} - \text{F1})] \times 0.5.$$

A positive value was considered superior movement or ascent while a negative value was indicative of inferior movement or descent. Other imaging characteristics were recorded: the change in the shape of the tonsils postoperatively, the presence of flow behind the tonsils, the formation of a retrocellebar neo-cistern, and any difference in ventricle size postoperatively. Postoperative CSF flow was evaluated radiologically using cine-flow MRI, and looking for evidence of retrotonsillar CSF flow. No quantitative CSF flow studies were performed.

After confirming the presence of CSF flow behind the tonsils at the level of the foramen magnum, a LP was performed in the lateral decubitus position. Enough CSF was drained to decrease the opening pressure by approximately 50%. The opening and closing pressures were recorded, as well as symptomatic relief in the following days. Clinical follow-up information was based on patient interview and input from the various physicians involved in their care (primary care physician, neurologist, ophthalmologist).

In this article we have used the term "neural hydrodynamics" when addressing issues related to the various hydraulic parameters (including pressure, volume and compliance) of the nervous system. The term CSF hydrodynamics has been used previously by various authors, including some studying syringomyelia associated with hindbrain anomalies [10, 15, 17, 18, 61]. However the CSF is not the only compartment involved in the hydrodynamics of the nervous system: the vascular and the cerebral compartments also play a significant role [7]. Therefore the term neural hydrodynamics was chosen.

Another term we will be using is "Idiopathic Intra-cranial Hypertension WithOut Papilledema" (IIHWOP). This is a concept that was introduced in the neurological literature in the past three decades when it became clear that some patients with Idiopathic Intracranial Hypertension (IIH) lack frank papilledema [2, 28, 35, 37, 59, 66, 75]. The diagnostic criteria used were symptomatic patients with CSF pressure higher than 200 mm of water on two occasions, no papilledema, and normal neurological evaluation [75].

Results (Tables 1, 2)

There were 5 females and one male (F/M ratio of 5/1). Their age ranged from 19 to 43 years with an average of 34 years (Table 1). No patient had associated syringomyelia. One patient had a previously resected tentorial meningioma (Case 3), and another one had a small incidental anterior communicating artery aneurysm (Case 1). The amount of tonsillar descent ranged from 5 to 21 mm (average = 11.7 mm) (Fig. 1). No patient had associated ventriculomegaly.

Table 1. *Patients characteristics*

Case #	Age	Sex	Ectopia (mm)	Interval to recurrence	O.P. (mean)	Coexistent factors	Treatment	Follow-up*
Case 1	29	F	21	4 months	18 cm	pseudomeningocele requiring lumbar drainage	VPS	16 months
Case 2	40	M	10	7 months	23 cm		VPS	18 months
Case 3	43	F	11	8 months	29 cm	– sinus involvement – previous surgery – overweight	VPS	21 months
Case 4	19	F	17	9 months	26 cm	– overweight – CSF leak	ILP	18 months
Case 5	39	F	5	4 months	23 cm	– overweight	VPS	16 months
Case 6	36	F	6	1.5 months	20 cm		Acetazolamide/ILP	19 months

O.P. Opening Pressure, *VPS* Ventriculo-peritoneal shunting, *ILP* Intermittent Lumbar Punctures.
* Follow-up since CSF drainage instituted, i.e. VPS for Cases 1, 2, 3, 5, and ILP for Cases 4 and 6.

Table 2. *Initial and recurrent clinical presentation*

Case #	Initial clinical presentation	Clinical presentation at recurrence	Findings on fundus exam on recurrence
1	retro-orbital headaches, cervical tightness, dizziness, ataxia, BUE numbness, fatigue	retro-orbital headaches, dizziness, ataxia, BUE numbness, fatigue	absent SVP
2	occipital headaches, cervical pain and stiffness, BUE numbness, fatigue, decreased hearing in left ear	occipital headaches, fatigue, facial numbness	N/A
3	occipital headaches, neck stiffness, bilateral shoulder pain, numbness of the hands, nausea	severe occipital headaches, neck stiffness, nausea	N/A
4	headaches, transient visual obscuration, papilledema	headaches, visual difficulty, BUE numbness	absent SVP
5	cervical pain into BUE, worse with coughing, BUE numbness and tingling, hypertension, dizziness, intermittent UE weakness	cervical pain into BUE, BUE paresthesias, BUE numbness and tingling, hypertension, dizziness	hyperemic discs, absent SVP
6	cervical pain, left arm and leg numbness, neck and facial numbness, swallowing difficulty, dizziness, throat numbness, fatigue, weakness, blurred vision	neck pain, occipital headaches, fatigue, ear pressure, choking, dizziness	thickening of disc margins, telangiectatic vessels consistent with prior edema, absent SVP

BUE Bilateral Upper extremity numbness, *SVP* spontaneous venous pulsations, *N/A* Not available.

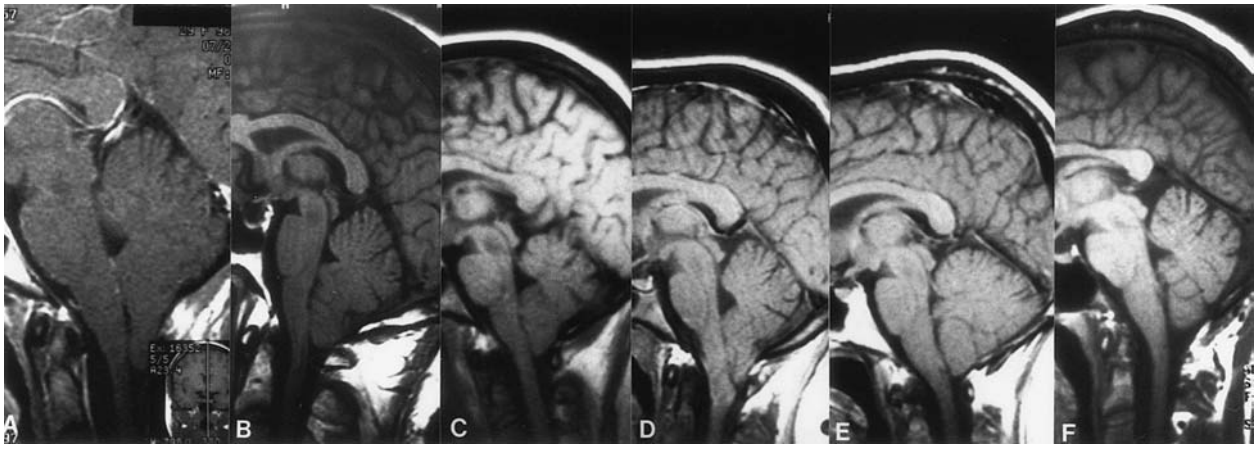

Fig. 1 (A–F). Preoperative sagittal T-1 weighted MRI in Patient 1, 2, 3, 4, 5, and 6 respectively. All patients had tonsillar ectopia equal to or greater than 5 mm

The symptoms consisted mostly of headaches, acral numbness, and various visual and otological symptoms (Table 2). The occurrence of cervical pain, retro-orbital headaches and visual symptoms have been reported in the recent series of Adult Chiari malformation. For example the incidence of retro-orbital headaches and visual symptoms was 63% and 55% respectively in the recent series published by Milhorat *et al.* [44]. During that approximate period, another 17 patients had successfully undergone surgery for ACM, and in this group, neck pain was present in 6 patients (35%), and visual symptoms in 12 patients (70%).

Operative results and complications

There was symptomatic relief for a variable duration postoperatively in all patients. One patient developed CSF leak postoperatively that was treated with repeat oversuturing of the incision and finally lumbar drainage (Case 4). Four had a fluid collection in the surgical bed on early postoperative imaging. In three patients it resolved spontaneously before recurrence of the symptoms; one patient was symptomatic and required lumbar drainage (Case 1). No other complications were seen.

Recurrence

The interval from surgery to symptoms recurrence was 1.5 to 9 months (average 5.6 months). The symptoms at time of recurrence were similar to the preoperative complaints (Table 2); although some symptoms were milder or absent, the patient's quality of life was still significantly affected, warranting further investigations.

Formal ophthalmological evaluation was obtained in four patients at recurrence. All four patients lacked spontaneous venous pulsations. One patient had preoperative papilledema, another had hyperemic disks suggestive of early papilledema, and a third had thickening of disc margins, and telangiectatic vessels consistent with prior papilledema.

After symptoms recurrence, all patients underwent repeat MRI (Fig. 2) and cine-flow MRI studies. There was formation of a retro-tonsillar neocistern and presence of retro-tonsillar flow in all cases. The tonsils were rounder postoperatively in four cases. Hindbrain and cerebellar migration was minimal and observed in both directions (superior and inferior) with an average 0.4 mm ascent and a range between 0.5 mm descent and 1.5 mm ascent (Table 3). The ventricles were not enlarged in any case, and there was no change in ventricular size postoperatively. The cervical spine was examined using MRI, and there was no evidence of any compressive pathology to explain the upper extremity or cervical symptoms.

LPs were then performed in all patients. On LP, the ICP ranged from 17 to 31 cm of water (average 23 cm). Enough CSF was drained to decrease the pressure by half its initial value. All six patients improved clinically after the LP. All the symptoms improved but mostly headaches and paresthesias. The improvement lasted for 2 to 7 days (average 5 days). Although the symptoms returned within one week, they were more tolerable than before the LP, and the duration of that partial relief was variable. We were

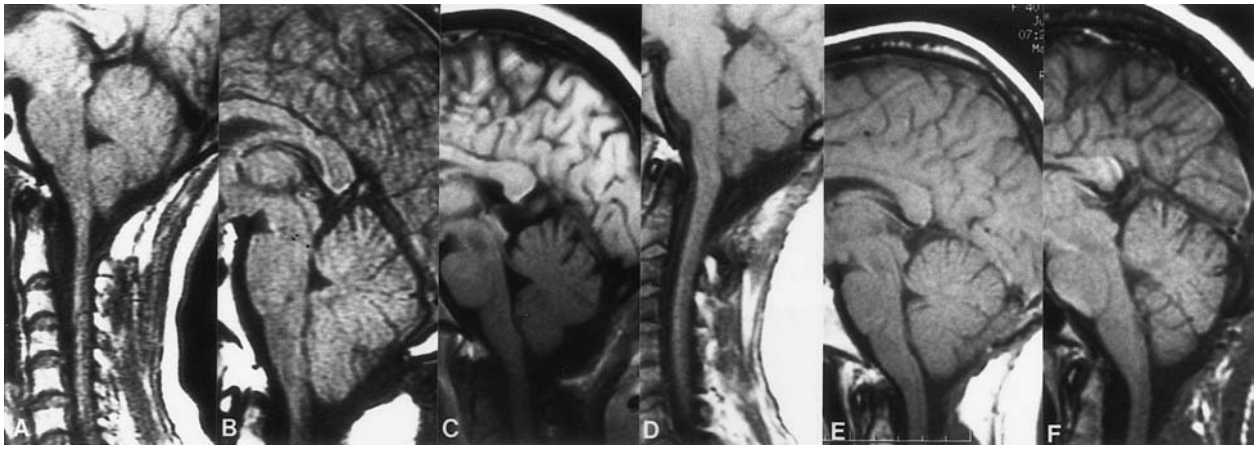

Fig. 2 (A–F). Postoperative sagittal T-1 weighted MRI in Patients 1, 2, 3, 4, 5, and 6 respectively. Adequate decompression is seen, without any evidence of significant cerebellar sagging

Table 3. *Radiological characteristics*

Case #	P1/P2 (mm)	F1/F2 (mm)	Ascent (mm)	Tonsils shape	Retrotonsillar neo-cistern	Retrotonsillar flow	Change in vent. size
1	40/40	21/23	+1 mm	more round	present	present	none
2	52/53	40/41	+1 mm	no change	present	present	none
3	48/47	31/31	−0.5 mm	more round	present	present	none
4	42/43	32/34	+1.5 mm	more round	present	present	none
5	51/51	41/41	0 mm	more round	present	present	none
6	44/44	29/28	−0.5 mm	no change	present	present	none

P1 Pontine height pre-operatively, *P2* Pontine height post-operatively, *F1* Fastigium height pre-operatively, *F2* Fastigium height post-operatively, *Ascent* $[(P2 - P1) + (F2 - F1)] \times 0.5$, *Vent*. Ventricle.

able to retrieve the CSF data at recurrence for five patients. Routine analysis failed to show any significant anomalies: glucose ranged from 56 to 59 mg/dl, protein from 24 to 52 mg/dl, red cell count ranged from 0 to 58 cells/cubic mm and white cell count form zero to 1 cell/cubic mm with neutrophils ranging from 0 to 20%.

After the CSF drainage was found successful, the pros and cons of the various options were discussed with the patients, with counseling over multiple clinic visits. Four patients chose ventriculo-peritoneal shunting using the Codman-Hakim Programmable Valve (CHPV), while two opted for intermittent LP $\pm$ Acetazolamide. The follow-up period after CSF drainage ranged from 16 to 21 months (18 months average). The symptoms were significantly relieved in all six patients, with major improvement in their quality of life. One patient had to undergo multiple revisions for shunt failure during the follow-up period (case 5). No other complications of shunting were observed. For the four patients who underwent shunting, the pressure setting at operation ranged between 90 to 120 mm H2O. One to 5 adjustments were required to reach the final pressure setting resulting in sustained relief. That final pressure setting was within 40 mm of the initial setting and ranged from 70 to 120 mm H2O. Most patients required lowering of the setting except for the one patient who had initial setting at 90 mm H2O: she was upgraded to 120 mm H2O during her hospital stay secondary to postural headaches.

Discussion

Shunting in the management of ACM associated with syringomyelia has been reported previously, even in the absence of frank hydrocephalus. Conway was the first to report the use of a ventriculo-jugular shunt to treat syringomyelia in a patient who was thought to be a poor risk for posterior fossa exploration [10]. For primary treatment of 31 patients with syringohydromyelia, Krayenbuhl used ventriculoatrial shunting in 22 and posterior fossa decompression in the remaining 9 patients [32]. Williams suggested VPS as an alternative treatment for Chiari malformation [76]. Peerless reported the successful use of ventricular shunting in four patients with syringomyelia and mild increase in ventricular size [49]. We are reporting the use of VPS or repeat LP for symptomatic treatment of recurrent ACM, without the presence of hydrocephalus or syringomyelia. Other surgeons have also used VPS after failed ACM decompression in patients with small ventricles (Milhorat T., Iskandar B. personal communications). On the follow-up range that we have (18 months), CSF drainage was successful in alleviating the recurrent symptoms of the ACM.

There are many potential explanations for the recurrences and their successful treatment with CSF drainage. These include surgical complications leading to aseptic meningitis, arachnoid scarring or cerebellar sagging; technical failure during the initial surgery; and finally, initial misdiagnosis with possible co-existence of the tonsillar herniation with another pathology with or without causal relationship.

Surgical complications

It is possible that surgery caused an aseptic meningitis with subsequent alteration of the neural hydrodynamics and need for CSF drainage [42]. The presence of a pseudo-meningocele in four patients, one of which leaked, is a potential cause of aseptic meningitis. On the other hand, increased ICP, as seen in case 4 (who had papilledema preoperatively) will lead to pseudo-meningocele formation and CSF leak. It would be difficult to ascertain what came first. The absence of even a minor increase in ventricular size postoperatively does not favor CSF absorption disorder as an etiology for the alteration of the neural hydrodynamics, although it does not rule it out completely. The normal findings on routine CSF examination rule out persistent aseptic meningitis as the etiology of symptoms at recurrence.

Arachnoid scarring is another possible explanation for the recurrence of symptoms, with recurrence of the block at the foramen magnum. However, improvement would be expected to occur with ventricular drainage only and not with lumbar puncture. The same holds true for cerebellar slumping or sagging. The latter has been reported to occur after generous suboccipital craniectomy [26, 42, 76, 77], and some authors have suggested ventriculo-peritoneal shunting as a potential treatment [26, 76]. However, the LP would be expected to worsen the symptoms of cerebellar slumping, which was not the case in our small group of patients. Radiologically, in three patients there was an ascent of the hindbrain. Only two patients had radiological slumping in the range of 0.5 mm, which is a minor degree of slumping compared to that usually encountered after posterior fossa decompression [14].

Technical failure

An inadequate surgery can cause recurrence of the symptoms. It is hard to determine technical inadequacy, since there are currently no strict technical standards for what constitutes an adequate decompression. There are many technical variations in the surgical decompression of ACM [5, 23, 27, 77]. ACM is thought to be secondary to a small posterior fossa [3, 38, 44, 46, 47, 67] or a disproportion between the intracranial contents and the skull [6, 9, 16, 33, 44, 48, 53, 60, 62, 70]. This disproportion leads to tonsillar herniation as well as an altered compliance of the system. Heiss *et al.* demonstrated the existence of altered compliance in ACM and its correction with decompression [25]. Decompression enlarges the skull and corrects the altered compliance [63]. Similarly, the dural tear secondary to the LP, as well as shunting, by providing an outlet for the CSF, transforms the closed system into a relatively open one and changes the compliance. This explains the success of CSF drainage in cases with technically inadequate surgery. However, there are no standard radiological criteria on what constitutes an adequate decompression; we used the presence of retrotonsillar CSF flow and the formation of a neo-cistern behind the tonsils as signs of adequate decompression. These signs were present in all six patients. In four, the tonsils became round, and the hindbrain ascended in three. Laboratory studies and prospective controlled studies looking at the compliance before and after surgery, and before and after shunting in failed ACM patients, will be needed to definitively answer this question.

Initial misdiagnosis

It is possible that there was an initial misdiagnosis in some patients. Although the tonsillar herniation was diagnostic of ACM by the classic radiological criteria [1, 4, 43], the initial symptoms could have been caused by another coexistent pathology. Most of the patients in our series fit the criteria for IIHWOP, as mentioned in the method section:

- Their symptoms are similar to those observed in patients with increased intracranial pressure as described by various authors [8, 12, 19, 21, 40, 54, 58, 72, 73, 74]. Giuseffi *et al.* found that many symptoms of IIH were missed in retrospective studies where the attention was directed on its cardinal symptoms, mainly headaches and visual loss [19]. These symptoms were: diplopia, visual loss, retrobulbar pain, shoulder/arm pain, dysco-ordination, intracranial noises, decreased smell, numbness, visual obscurations, motor weakness and headaches [19].
- There were no neurological findings on exam.
- On fundus exam, there was absence of spontaneous venous pulsations (SVP) in all cases where an exam was performed. None of the 9 patients treated by Spence for IIHWOP had SVP. SVP are present in 87.6% of the normal population, but absent in patients with increased intracranial pressure without papilledema, and in patients with IIH with papilledema [34]. Other ophthalmological findings in our last two patients (Case 5 and 6) also favor increased ICP. After our initial experience with these six patients we started performing a comprehensive visual examination on all Chiari patients before and after surgery and have found that venous pulsations are absent preoperatively in 90% of patients with ACM and that their return postoperatively correlated with symptomatic improvement. (Cockerham K. P., Bejjani G. K.: Neuro-ophthalmological manifestations of the Adult Chiari Malformation, North American Neuro-Ophthalmology Meeting 2002, Denver, Colorado).
- The ICP was higher than 200 mm H2O on at least two occasions in all 4 patients who underwent two LPs or more. The remaining two patients underwent only one LP since both had significant relief and opted for shunting. One of them had a pressure of 23 cm H2O and hyperemic discs. The other had a pressure of 18 cm H2O. Continuous ICP recordings in patients with IIH have shown that the pressure varies widely during the day, reaching normal values on occasions [2, 22, 66]. Spence described seven patients with IIHWOP that had high pressure waves detected only by 24 hours CSF pressure monitoring: five of those had baseline pressure under 15 mm Hg [66].
- There was no evidence of ventriculomegaly or intracranial mass lesion in any patient.
- The response to treatment, especially with more than one year follow-up in each case, is another strong argument for intracranial hypertension [66].

The issue is whether IIH was present before the initial surgery or was induced by the surgery itself. Most recurrent symptoms were similar to the preoperative symptoms, suggesting that IIH was present from the

beginning. An argument could be made that the blockage of the CSF flow by the tonsils at the foramen magnum creates increased pressure within the cranial cavity and symptoms similar to IIH. In fact, numerous studies clearly demonstrate the presence of increased ICP and cranio-spinal pressure dissociation in patients with ACM [25, 78]. However, if the problem in our patients was persistent or residual obstruction to CSF flow at the foramen magnum from inadequate surgery, the symptoms would be expected to improve only with ventricular drainage. Lumbar drainage would worsen the symptoms. In contrast, in all six patients in this series, symptoms improved after the LP.

As we have mentioned previously, it is possible that this state of intracranial hypertension could have been secondary to postoperative aseptic meningitis, with scarring of the arachnoid granulations and CSF pathways, leading to altered CSF absorption. The absence of even a minor increase in ventricle size does not favor this hypothesis, although it is still possible.

Another explanation is that IIH or IIHWOP were initially present, at least in some patients. It improved transiently secondary to the suboccipital decompression and fluid drainage, then recurred once the tissues healed and fibrosed, leading to the restricted compliance. Elements in favor of this hypothesis are the similarity of the recurrent symptoms to the initial symptoms, the high incidence of pseudo-meningocele, and the time course of the recurrence. In an animal model, Sklar *et al.* found that decompressive craniectomies cause a significant decrease in the elasticity slope and pulse pressure [63]. One possible explanation for the delayed recurrences is scarring that will alter the compliance. Tensile strength in aponeurotic wounds increases quickly over the first few months then at a slower rate for over 1 year [13], similar to the temporal pattern observed in our patients. With time, the dural graft and exposed dura fibrose and become stiff, altering the compliance of the brain-skull complex and allowing some of the symptoms to recur, sometimes milder, despite adequate CSF flow on the cine-MRI. This would be similar to what is seen in patients with pseudo-tumor cerebri, in whom the headaches may recur after subtemporal decompression, leading to shunting, even though the papilledema resolved [30].

Other factors, such as obesity or alteration in venous drainage might have contributed to the increased intracranial pressure. Three of our patients were overweight (Case 3, 4, 5). Obesity is associated with IIH, and it has been proven that central obesity raises the intra-abdominal pressure, increasing the cardiac filling pressure and impeding the venous return from the brain, leading to increased intra-cranial venous pressure and increased intra-cranial pressure in patients with IIH [68]. In Case 3, the partial venous outflow obstruction caused by the meningioma might have contributed to the increased ICP.

Tonsillar herniation and IIH: coincidence, association, or cause-effect relationship?

The association between IIH and tonsillar herniation is not new. Some authors have found that a significant percentage of patients with IIH have evidence of tonsillar herniation on imaging, before lumbar shunting. Johnston [29], in his review of acquired Chiari Malformation after spinal drainage, found that 3 out of 51 patients with IIH (6%) had definite ACM on MRI before treatment, much higher than the 0.77% incidence of tonsillar descent found in the selected Meadows [41] population of patients undergoing brain MRI's in a tertiary care center. This argues against a pure coincindental association between tonsillar ectopia and IIH. Other scattered reports of patients with increased ICP and ACM are also found in the literature [69, 71].

Although it would be nothing more than a speculation to hypothesize a cause-effect relationship between IIH and tonsillar ectopia based on our findings alone, it cannot be ruled out. Tonsillar herniation can be seen after disorders that increase the volume of the intracranial contents, such as supratentorial tumours [48, 62] and infratentorial mass lesions [36, 70]. Acute tonsillar herniation is a common finding in rapidly enlarging intracranial processes like acute brain edema, intracerebral hemorrhage, and acute hydrocephalus. Various pathological [56] and radiological studies [45, 52, 65] point to engorged brain as the underlying disorder in IIH, with secondary alteration of CSF absorption and the vascular bed [20, 39, 51, 64]. In animal models of intracranial hypertension, venous occlusion was associated with increase in brain volume [11]. It is logical that with the increase in the volume of the intracranial contents, the tonsils will occasionally herniate into the cervical canal, especially if the posterior fossa is small, explaining the 6% incidence of ACM in patients with IIH [29]. This was recently demonstrated in an animal model of venous hypertension leading to tonsillar ectopia (Milhorat *et al.*, personal communication).

It is hard to find one hypothesis to explain the findings in all our patients, and we do not pretend to do so. It is possible that the mechanism is different for each patient. However, we would like to report our encouraging results with CSF drainage in these patients. Elucidating the reasons for its success and clarifying the indications will need more prospective, clinical, hydrodynamic and animal studies, some of which are underway by our group, as well as by others.

Conclusion

Failed decompression for ACM is a significant problem. CSF drainage, in the form of repeat LP $\pm$ Acetazolamide or ventriculo-peritoneal shunting is successful in some patients who fail suboccipital decompression. The potential reasons behind the failure of the decompression and the success of CSF drainage are numerous, and include a technically inadequate surgery, postoperative complications leading to altered hydrodynamics, and coexistence of tonsillar ectopia with IIHWOP. However there is a need for long-term follow-up and further hydrodynamic studies to clarify these issues.

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Disclosure

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Comment

This is a very interesting paper by Bejjani *et al.* discussing an important issue: what should we recommend to patients with Chiari I malformation in whom decompression at the foramen magnum has failed. The authors suggest to consider CSF shunting or repeated lumbar punctures as a possibility and report remarkable success in their six patients treated in this manner. However, long-term results need to be studied before this policy can be recommended. If evidence of csf-flow obstruction at the foramen magnum is obtained by cine-MRI, revision surgery should be done at the foramen magnum including arachnoid dissection and opening of the 4th ventricle.

However, a few additional points merit a comment:

1. The surgical technique of foramen magnum decompression without arachnoid opening has its problems: csf fistulas are not avoided and csf collections in the wound occured in 1 and 4 of the 6 cases presented here, respectively. We open the arachnoid routinely in every Chiari patient and close the soft tissue layers as well as the dural layer meticulously. In this way we avoided csf fistulas completely in the past 100 foramen magnum decompressions.
2. Foramen magnum decompression without arachnoid opening did have a significantly worse outcome in our series compared to patients with arachnoid dissection and opening of the 4th ventricle [1]. In the series presented here, 6 out of 23, i.e. 26%, of patients decompressed without arachnoid opening developed clinical problems postoperatively after an initially good clinical result! In our series of 141 patients, 61% reported sustained improvement and 34% stabilization, while only 4% deteriorated.
3. I agree completely that cine-MRI is the method of choice for establishing the postoperative result of a foramen magnum decompression. A free csf-pathway and formation of a cistern should be the aim of surgery. In our own series we have documented 20 patients after a failed first decompression with intraoperative evidence of csf-flow obstruction during revision surgery in whom arachniod dissection improved csf-flow with good clinical success (Klekamp & Samii 2001).
4. The significance of absent venous pulsations needs further studies to establish its significance in Chiari patients.
5. The fact, that patients developed clinical problems despite a good postoperative result after foramen magnum decompression as evident on cine-MRI does raise the question of a coexistent pathology as the authors discuss.

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J. Klekamp

Correspondence: Ghassan K. Bejjani, M.D., Tristate Neurosurgical Associates-UPMC, 200 Lothrop Street, Suite 5C, Pittsburgh, PA 15213.