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The Acquired Chiari Malformation and Syringomyelia Following Spinal CSF Drainage: A Study of Incidence and Management

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Summary

Firstly, 14 patients are described who developed either an acquired Chiari malformation (ACM) alone (7 cases) or ACM and syringomyelia (7cases) after lumbar subarachnoid space (SAS) shunting or in one case, epidural anaesthesia with SAS penetration. Four groups are considered: 3 cases with craniofacial dysostosis and communicating hydrocephalus (CH), 4 cases with CH alone, 3 cases with pseudotumour cerebri (PTC) and a miscellaneous group (4 cases). Initial treatment was varied: resiting the shunt to ventricle or cisterna magna [6], adding an H-V valve [1], syrinx shunting [4] and posterior fossa decompression [3]. Further treatment was required in 6 cases. Secondly, incidence was examined in 87 patients with PTC initially treated either by lumbar SAS shunting [70] or cisterna magna shunting [17]. In the first sub-group, 11 cases (15.7 per cent) developed an ACM, 3 symptomatic (as above) and eight asymptomatic with 1 case also having syringomyelia whereas 1 case occurred in the second group with a questionanably symptomatic ACM. While accurate for symptomatic lesions, these figures are tentative with respect to asymptomatic lesions due to inadequate pre-treatment radiology and detailed MR follow-up. The main conclusions are, first, that the incidence of symptomatic ACM and/or syringomyelia is not high enough to warrant abandoning SAS shunting; second that asymptomatic lesions need not necessarily be treated and third, that when treatment is required, shunt resiting is the first choice.

Keywords: Acquired Chiari malformation; syringomyelia; lumbar shunting.

Introduction

Over the past two decades there have been a number of reports of a Chiari malformation (chronic cerebellar tonsillar herniation) and syringomyelia occuring either singly or together following CSF shunting from the lumbar subarachnoid space [12, 17, 21, 33]. These reports have raised important issues concerning the mechanism of development of such secondary abnormalities, their incidence and the most appropriate method of management, as well as calling into question the use of lumbar subarachnoid CSF shunting in general. These complications have been thought of as rare, there being only a few reports of single or small numbers of cases and no mention of such sequelae in several reviews of patients treated by lumbo-peritoneal shunting [1, 5, 10, 30]. Recently, however, Chumas *et al.* [7] have reported the contrary in a retrospective study of 143 cases treated by L-P shunting at the Hospital for Sick Children in Toronto over a 16-year period. Despite quite significant limitations due to incomplete follow-up and somewhat inadequate radiology (i.e. non-purpose directed CT scan only) they felt able to conclude that these complications might be much more prevalent than previously supposed. The purpose of this paper is twofold: Firstly, to provide more detailed information on incidence by reviewing a series of patients with L-P shunts studied by MR scanning with specific attention to the craniospinal junction and secondly, to examine the matter of management.

Clinical Material

Four groups of patients treated at either the Royal Prince Alfred Hospital or the Royal Alexandra Hospital for Children, Sydney between 1974 and 1996 will be considered. Details of the groups are summarised below. The first three groups comprise patients who had lumbar CSF drainage for a variety of reasons and who subsequently presented with either an ACM or syringomyelia, or both. All patients were necessarily symptomatic. The fourth group, those with pseudotumour cerebri (PTC), included all patients with this condition treated by one of the authors (Johnston) between 1974 and 1996. Where possible, all patients had detailed pre-treatment radiology (in the last eight years, MR scanning) and patients treated by

shunting have again, where possible, had post-treatment MR scanning looking specifically for an ACM or syringomyelia.
The groups are as follows:
(i) Three patients with craniofacial dysostosis and communicating hydrocephalus treated by lumbo-peritoneal shunting.
(ii) Four patients with simple communicating hydrocephalus who developed a symptomatic ACM with or without syringomyelia after lumbar CSF shunting.
(iii) Four patients with other, disparate conditions who developed a symptomatic acquired ACM with or without syringomyelia after lumbar CSF drainage.
(iv) A group of 131 cases of PTC treated by one of the authors between 1974–1996. Forty-four of those patients had medical treatment only, while 87 were treated by CSF shunting from the lumbar subarachnoid space, cisterna magna, lateral ventricles, or a combination of these.

Results

Group I: Craniofacial Dysostosis and Communicating Hydrocephalus (Table 1)

The three cases in this group comprised two patients (one male, one female) with Crouzon's disease and one female with Apert's Syndrome, all of whom developed progressive hydrocephalus within the first three years of life. In no case was there adequate radiology sufficient to exclude a prior ACM, although none was demonstrated on routine head CT in two cases and on ventriculography in one case. Certainly no case had symptomatic evidence of an ACM. All three cases subsequently developed both a symptomatic ACM and evidence of progressive syringomyelia with presentation at an average time of 6.7 years after L-P shunt insertion (3 to 13 years). In the first case, no syrinx was demonstrated at the initial presentation although as myodil myelography was the only investigation carried out, a syrinx may undoubtedly have been overlooked. It is of note that in the two cases initially treated by a syrinx shunt there was only partial or no improvement, whereas in the one case treated by conversion to a ventricular shunt, there was relief of both the ACM and the syrinx. In follow-up over an average period of 5 years, the situation has remained stable in all three cases.

Case 2. This male patient was first admitted at the age of 5 months for investigation of Crouzon's disease. His head circumference was 43.6 cm. X-rays of the cranium, CT scan and ophthalmological assessment were all normal. He was re-evaluated at 2 1/2 years after presenting with symptoms of raised intracranial pressure. His head circumference was then 50.5 cm. While the CT scan was again normal, overnight ICP monitoring via a lumbar subarachnoid catheter showed quite definite intracranial hypertension. In November 1984, bilateral central craniotomies were carried out with relief of symptoms. He presented five months later with recurrent intracranial hypertension (head circumference 52 cm) and again ICP monitoring was abnormal (plateau waves greater than 20 mm/Hg). There was some ventricular enlargement on CT scan so a percutaneous L-P shunt with a James medium-pressure valve was inserted. Over the ensuing 18 months he required 4 shunt revisions because of blockage and infection plus a subtermporal decompression for persistent intracranial hypertension. He remained well until March 1989 (age 6 years 11 months) when he presented with a history of back pain and difficulty walking (head circumference 54 cm). An MR scan demonstrated an extensive syrinx and a Chiari malformation (Fig. 1a). A right ventriculo-peritoneal shunt was inserted and the L-P shunt removed. In the following 6 months he had 3 shunt revisions (ventricular catheter malfunction) but symptoms and signs of his syrinx resolved and in August 1989, a further MR scan showed the syrinx had collapsed (Fig. 1b). In October 1989 he had a craniofacial reconstruction. Since that time he has remained well.

Group II : Communicating Hydrocephalus (Table 2)

The four patients in this group (three males, one female) all had uncomplicated communicating hydrocephalus, two of unknown aetiology, one post-meningitic, and one associated with a chromosomal

Table 1. *Cases with Craniofacial Dysostosis and L-P Shunt*

Case	Presentation	Time from L-P shunt	Radiology	Treatment	Outcome
1. F. Crouzon's	(i) mild (R) limb weakness	13 years	ACM	none	gradually worse
	(ii) kyphoscoliosis, myelopathy	19 years	ACM & syrinx	syrinx to SAS shunt	some improvement
2. M Crouzon's	back pain and leg weakness	4 years	ACM & syrinx	L-P shunt to VP	syrinx resolved ACM persisted
3. F Apert's	(i) bilateral leg weakness	3 years	ACM & syrinx	syrinx to SAS shunt	persistent symptoms
	(ii) worsening leg weakness	4 years	ACM	cervical & FM decomp	improved

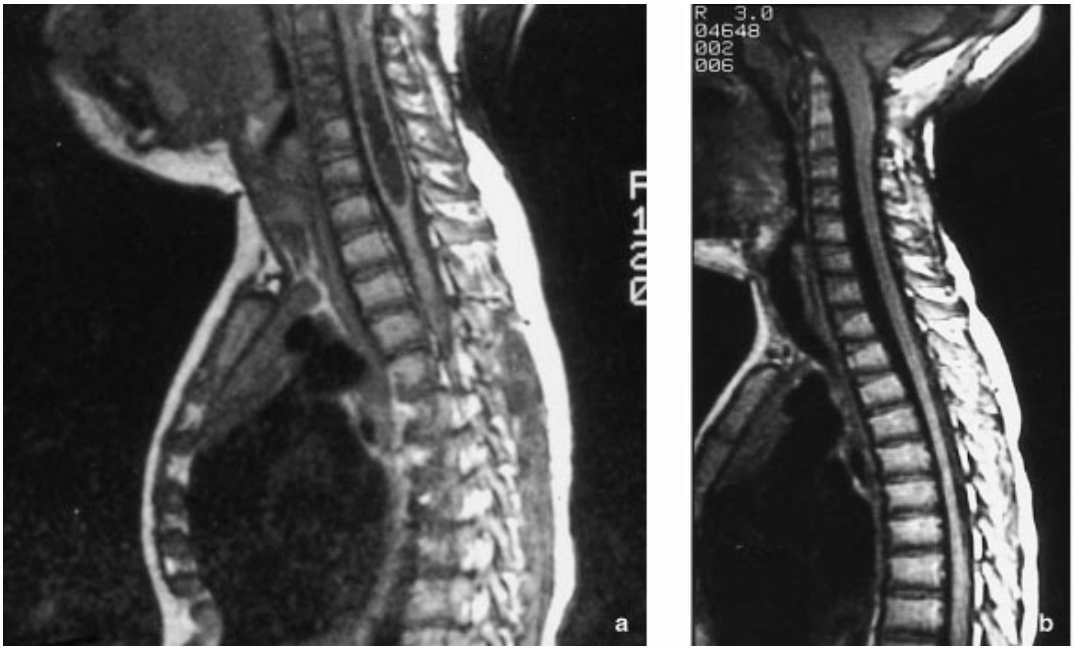

Fig. 1. MR scans for Case 2 before (a) and after (b) conversion of L-P to V-P shunt for treatment for ACM and syringomyelia presenting 4 years after shunting for hydrocephalus

Table 2. *Cases with Communicating Hydrocephalus and L-P Shunt*

Case	Presentation	Time from L-P shunt	Radiology	Treatment	Outcome
4. M	severe interscapular pain	4.5 years	ACM	L-P to V-P	resolved
5. M	drowsiness, bilateral motor signs	1 year	ACM syrinx	ACM decomp, HV valve to L-P shunt	improved
6. F	(i) neck pain, cranial nerve palsies, ataxia	16 years	ACM	(i) ACM decomp	trans. imp ☑ worse
	(ii) recurrence of above	6 months	ACM worse	(ii) L-P to V-A	resolved
7. M	headache, neck pain	9 years	ACM	(i) L-P ☑ V-P (ii) ACM decomp	persisted resolved

abnormality and developmental delay. In three cases a James percutaneous L-P shunt (medium-pressure) was used, and in one a Hoffman T-tube and Raimondi distal catheter (medium pressure). As with Group 1, the onset of later problems was typically long delayed (average 7.6 years range 1–16 years) although in one patient, the only case with both an ACM and a syrinx, the onset was relatively rapid. Symptoms varied with local pain predominating. In case 6 (details below) the clinical presentation was particularly severe. The treatment was again varied. In all cases the shunt was changed: in three from L-P to V-P, and in one by the addition of an HV valve to the existing L-P shunt. In two cases the shunt conversion was clearly the critical manoeuvre, whereas one case still required an ACM decompression after shunt conversion. In the remaining case, the shunt change and ACM decompression were carried out simultaneously so it is impossible to determine the relative efficacy of the two procedures.

Case 6. A female patient presented at 12 years of age with raised intracranial pressure due to communicating hydrocephalus of unknown aetiology. An L-P shunt was inserted using a Hoffman T-tube and Raimondi distal catheter (medium-pressure). Apart from

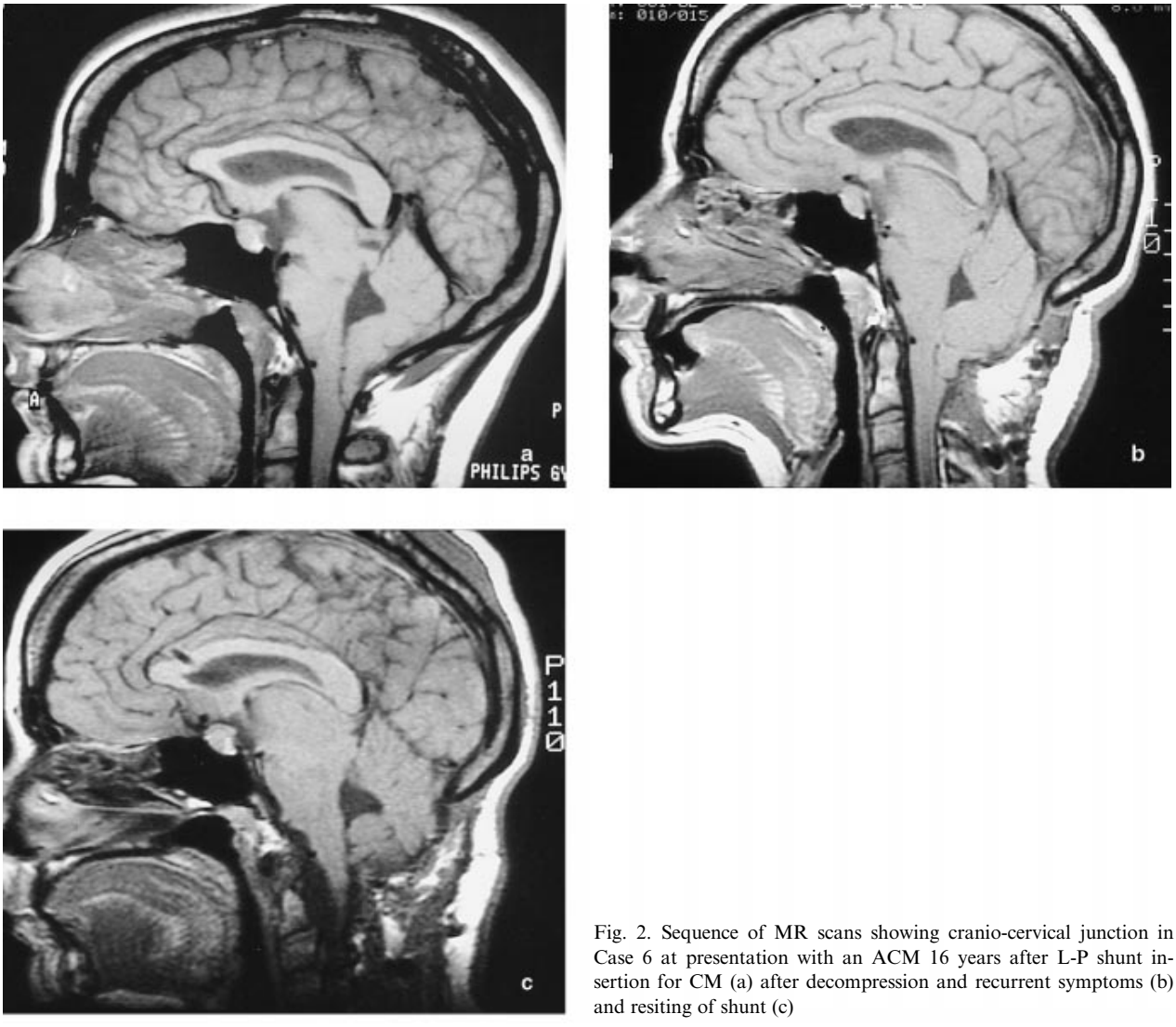

Fig. 2. Sequence of MR scans showing cranio-cervical junction in Case 6 at presentation with an ACM 16 years after L-P shunt insertion for CM (a) after decompression and recurrent symptoms (b) and resiting of shunt (c)

one early shunt revision and a short psychiatric disturbance following her initial treatment, she remained well for 16 years. She then presented to another institution with a short history of headache, neck pain, ataxia and multiple lower cranial nerve palsies. A CT scan was reported as unremarkable apart from some enlargement of the IV ventricle. An MR scan (Fig. 2a) showed a marked Type 1 Chiari malformation. A posterior fossa and upper cervical decompression to C3 was carried out and a wide duroplasty done. The shunt, which was presumed to be non-functioning, was left in situ. Approximately two months later she developed a chest infection with a severe and persistent cough. Shortly after this, her previous brain stem symptoms recurred together with ataxia, abnormal posturing and grimacing which was suggestive of a basal ganglia disturbance. A diagnosis of hysterical conversion reaction was made, an MR scan having been thought to show adequate decompression. She did not respond to psychiatric treatment and, of her own volition, returned to Sydney. The series of MR scans from her initial presentation to her first assessment in Sydney (a 5-month period) showed progressive downward displacement of the posterior fossa contents and also distortion of the upper brain stem and diencephalon (Fig. 2b). The lumbar shunt, which was found to be quite patent, was surgically ligated with immediate clinical improvement. After one week, she again developed intracranial hypertension. A ventriculo-atrial shunt (Sophy programmable valve on medium setting) was then inserted with rapid amelioration of all symptoms and progressive improvement on MR appearances (Fig. 2c). She has remained entirely well for 3 1/2 years.

Group III: Miscellaneous (Table 3)

Within this disparate group, case 3 is particularly interesting in that a mullti-compartment syrinx for which two separate syringo-pleural shunts were inserted at different times, developed before the ACM. The shunting of the second (and higher) syrinx apparently precipitated the development of the ACM. (Details of this case are given below.) Of the others, the first developed symptoms five years after insertion of an L-P shunt for a persistent occipital pseudo-

Table 3. *Miscellaneous Case with L-P Shunt*

Case	Initial condition	Shunt type	Presentation	Time from shunt	Radiology	Treatment	Outcome
8. F	occipital pesudo-meningoceole	L-P (James)	interscapular pain	5 years	ACM syrinx	L-P to V-P	resolved
9. F	?BIH (gliomatosis cerebri)	L-P (non-valved)	medullary compression	immed	nil	L-P out	died
10. F	lumbar pseudo-meningoceole	cyst peritoncal (no valve)	(i) dissoc. sensory loss	9 years	syrinx	syringopleural shunt	improved
			(ii) severe neck pain	17 years	ACM	ACM decomp	resolved
11. F	small prolactinoma	epidural anaesthetic	severe headache	immed	ACM	ACM decomp	improved

meningoceole which followed removal of a posterior fossa astrocytoma. Conversion of the L-P shunt to a V-P shunt gave early and sustained improvement. The remaining two cases were rapidly symptomatic following drainage of lumbar CSF. The first after insertion of a non-valved L-P shunt for what was thought to be benign intracranial hypertension, but turned out at post mortem to be gliomatosis cerebri, and the second after dural puncture during an epidural anaesthetic for abdominal surgery. This patient had a prior MR scan for galactorrhoea showing no significant ACM. A further MR scan following the onset of symptoms showed a quite marked ACM. The first of these two cases may be seen simply as an example of acute tonsillar herniation following lumbar CSF drainage in the presence of an intracranial mass lesion, but is included for its relevance to the issue of mechanism.

Case 10. This female patient, now 22 years old, was born with a lumbosacral lipomeningoceole which was excised at the age of 3 months. At 3 1/2 years she was explored for possible cord retethering and developed, post-operatively, a pseudomeningocoele and CSF leak which were eventually controlled by a cyst-peritoneal shunt (non-valved). The shunt was removed after three months because of persistent low-pressure symptoms. She remained well for nine years apart from a static deficit associated with her spinal dysraphism, before presenting with bilateral leg weakness and distal sensory loss. An MR scan showed a large syrinx extending from T8 to the conus medullaris which was treated by insertion of a syringopleural shunt (non-valved). There was significant improvement, but not complete resolution of her new neurological deficit. On MR there was marked improvement with almost complete disappearance of the syrinx three months after shunting (June 1988). On further MR scan in June 1989, the syrinx remained collapsed and there was no evidence of a Chiari malformation. In April 1990, (aged 15 years) due to a deterioration in her neurological state distally, attributed to cord re-tethering, she was re-explored with some clinical improvement. Again a cyst-peritoneal shunt (non-valved) was required for a troublesome CSF leak. She next presented in February 1992 (aged 17) with upper limb signs and radiological recurrence and extension into the cervical region of her previous thoracic syrinx. Unfortunately after revision of her syringo-pleural shunt, there was no clinical improvement with reduction of the thoraco- lumbar component of her syrinx only on MR scan, suggesting compartmentalisation of the syringomyelia. As her upper limb and thoracic disturbance was progressive, a separate syringo-pleural shunt was inserted into the cervical syrinx. There was some clinical improvement and substantial radiological resolution, but still no evidence of a Chiari malformation. Two months later, she complained of increasingly severe neck pain. A further MR scan showed a definite Type 1 Chiari malformation together with a sizable extra-dural fluid collection in the cervical region. Initially this was managed conservatively but because of persistent symptoms and failure of neurological resolution after 3 months, a foramen magnum decompression for her ACM was carried out and the extra-dural spinal fluid collection drained. Her neck pain disappeared and she has remained well, with a stable neurological deficit related to her spinal dysraphism and syringomyelia, for five years.

Group IV: Pseudotumour Cerebri

The details of the 131 cases in this group are summarised in Table 4. There were 44 patients treated medically only and 87 patients shunted, the latter subgroup being further divided according to the type of shunt initially inserted.

1. Medical treatment (44 Cases): There were no patients with either a symptomatic Chiari malformation or syrinx before or after treatment. However, of 19 patients having a pre-treatment MR scan, 2 had an asymptomatic Chiari malformation.
2. Initial lumbar shunt (70 cases): In this group there were 3 patients with a symptomatic ACM presenting after an average interval of 3.2 years from shunt insertion. Two were treated by conversion of the L-P shunt to a cisterno-atrial shunt, one with resolution (Figs. 3a and b) and one going on to suboccipital decompression. The third patient initially

Table 4. *Treated Pseudo-Tumour Patients*

Case	Initial shunt	Presentation	Time from shunt	Radiology	Treatment	Outcome
12. F	L-P (James)	(i) dissoc sensory loss	3 years	syrinx	syrinx to SAS shunt	improved
		(ii) dizziness, nack pain	7 years	ACM	ACM decomp	resolved
13. F	L-P (James)	headache, nausea, ataxia	2.5 years	ACM	L-P to C-A	resolved
14. F	L-P (James)	headache, ataxia	3 years	ACM	L-P to CA ACM decomp	resolved recurred, rpt decomp

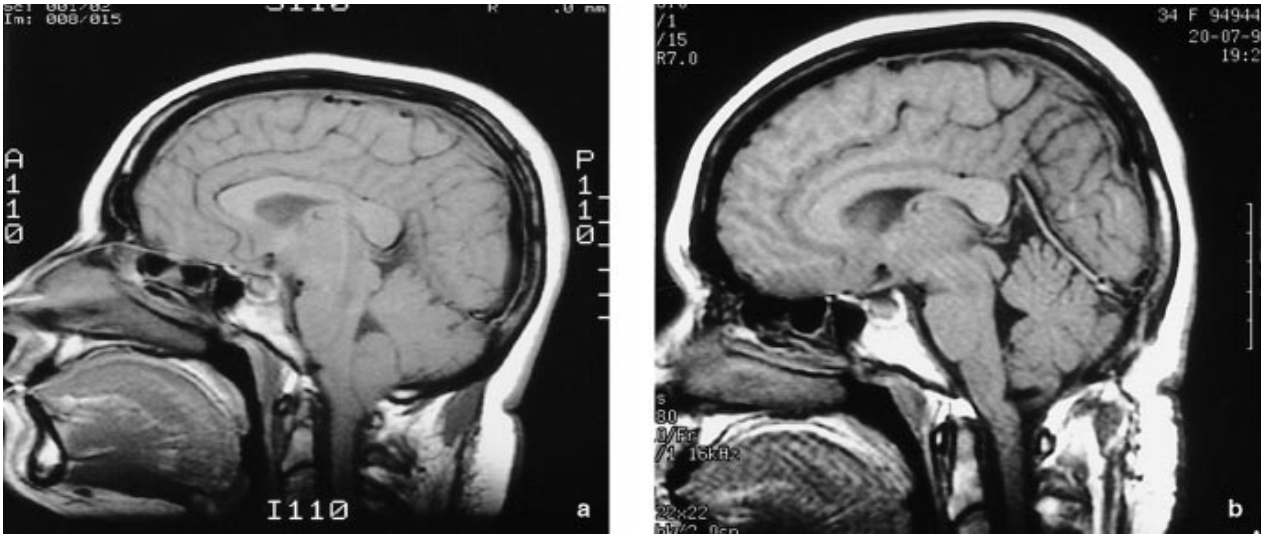

Fig. 3. MR scans before (a) and after (b) conversion of a lumbar to cisternal shunt in a patient with PTC

presented with a cervical syrinx which was treated by syringostomy (she had a functioning L-P shunt). She subsequently required decompression of an ACM not present at the time of her presentation with syringomyelia. There were eight patients with entirely asymptomatic Chiari malformation detected on routine MR follow-up at an average interval of 3.6 years after L-P shunt insertion. In contrast there were 21 patients with a normal MR scan (no ACM) after L-P shunting at an average interval of 6.6 years from shunt insertion. Other relevant figures in this group are as follows: The average overall follow-up period was 7.8 years, with 3 patients only lost to follow up and 3 patients dying from unrelated conditions. Fifteen patients had the L-P shunt removed for a variety of reasons, after an average interval of 3.6 years, while 26 were converted to another form of shunt (cisternal in 25, ventricular in 1) after an average interval of 2 years. Sixteen patients in this group had no evidence of a Chiari malformation on a pre-treatment MR scan.

3. Initial cisternal shunt (17 cases): All patients in this group had a pre-treatment MR scan. One of the patients had a definite Type 1 Chiari malformation while the remainder were quite normal at the craniocervical junction. To date, post-treatment MR scans have been carried out in 12 patients. The one patient with a pre-treatment Chiari malformation has maintained the same degree of asymptomatic tonsillar displacement over five years since shunt insertion. The one patient with some degree of tonsillar descent post-shunt had the shunt changed to a ventricular-atrial shunt without significant change in symptomatology or radiological appearance over a two-year follow-up period. Apart

from this patient, no case has had any suggestion of a symptomatic ACM and the other 10 post-treatment MR scans show a normal craniocervical junction. The average follow-up in this group is 3.8 years and the average time from treatment to post-treatment MR scan 2.5 years.

Summary of PTC Group

Of the total of 131 patients with PTC, 51 had pre-treatment MR scans of whom 3 had a definite Type 1 Chiari malformation. No specific treatment was given for the Chiari malformation in these 3 cases, 2 having medical treatment only for their PTC and 1 a cisterno-atrial shunt without any change clinically or radiologically in the Chiari malformation. Of 70 patients treated initially with an L-P shunt (using a variety of shunts) 3 developed a symptomatic ACM requiring treatment, 1 of whom also had a syrinx prior to the ACM, treated by syringostomy and maintenance of the lumbar shunt. (Details of these patients are shown in Table 5.) In addition there were eight patients with asymptomatic ACM in this group. Of 17 patients treated initially with a cisterna-magna shunt, 1 developed a slight and questionably symptomatic ACM for which the shunt was converted to a ventriculo-atrial shunt without clinical or radiological change.

Summary of Treatment Methods

Overall 14 patients were treated. The number is, of course, too small and the treatment methods too varied to draw any incontrovertible conclusions. With the Chiari malformation there are two possible strategies: Resite the shunt or decompress the craniocervical junction. In 6 cases, shunt resiting alone was used, in 4 to lateral ventricles and in 2 to cisterna magna. There was a significant failure rate with two patients having ventricular shunts showing persistent symptoms (1 requiring further treatment) and 1 with a cisternal resiting requiring further treatment. Three patients had an initial craniocervical decompression (1 with a concomitant change of the lumbar shunt valve to reduce drainage). In one of these 3 cases there was a very significant failure of treatment with marked progression of brain stem displacement (Case 6).

Four patients whose presentation was with syringomyelia had this treated primarily; 3 with a syringopleural shunt and 1 by syringostomy (with a functioning L-P shunt). This treatment was successful in only 2 cases, the other two requiring further treatment. It is noteworthy that two cases developed a Chiari malformation after treatment of the syrinx. One patient had a lumbar shunt urgently removed after acute tonsillar herniation , but to no avail.

Discussion

CSF diversion from the spinal subarachnoid space was first reported by Ferguson in 1898 [11], but like other forms of shunting out of the CSF compartment utilising implanted tubing, did not come into general use until the 1950's. Since then, there has been a num-

Table 5. *Treatment Summary*

			Outcome			Re-Treatment
			Worse	Persistent	Improved	
Add H-V value to L-P	initial	1	0	0	1	0
	subs	0	–	–	–	–
L-P to V-P	initial	4	0	2	2	1
	subs	1	0	0	1	0
L-P to C-A	initial	2	0	0	2	1
	subs	0	–	–	–	–
Syrinx shunt	initial	4	1	1	2	2
	subs	0	–	–	–	–
ACM decompression	initial	3*	1	0	2	1
	subs	5	0	0	5	1
Other	initial	1	1	0	0	1
	subs	0	–	–	–	–

ber of reports of large series of patients treated by lumbar CSF shunting (usually to the peritoneum, but also to other sites such as the ureter) [1, 5, 10, 30]. Predominantly, these have been patients with communicating hydrocephalus but the method has also been used for PTC. [19] and several other conditions [5]. Between 1971 and 1993 there were 6 reports of an acquired ACM and 1 of syringomyelia associated with prolonged shunting from the lumbar subarachnoid space [6, 12, 17, 21, 31, 33]. In all there were 18 cases the initial description being that of Kushner *et al.* in 1971 [21] who reported the post mortem finding of syringomyelia in a patient who had a lumbar-peritoneal shunt in place for communicating hydrocephalus for 13 years converted to a ventricular shunt 18 months prior to death and who died due to a shunt infection. This patient had a chronic scoliosis. No mention was made of an ACM. This was from a series of 34 patients followed up in an overall study of 80 children treated with lumbar shunting. Hoffman *et al.* in 1976 [17] reported 8 cases with an ACM after shunting for hydrocephalus 6 of whom had lumbar shunts inserted for communicating hydrocephalus and became symptomatic from an ACM 2 to 7 years after the initial shunt. All were treated by posterior fossa decompression with a reported good outcome. Two of the patients also had lumbar shunts converted to ventricular shunts but this was due to shunt malfunction and not part of the management of the ACM. The authors grouped the LP and VP shunt cases together and attributed the presumed acquired ACM to ''cephalo-cranial disproportion''. Fischer *et al.* [12] then reported 3 cases of syringomyelia without identification of an ACM after lumbar shunting and Welch *et al.* [33] 5 cases with an ACM 2 of whom also had syringomyelia. Subsequently 2 individual case reports appeared. The first was that of Sullivan *et al.* in 1988 [31] of a patient with a lumbar shunt for PTC which was notable for the demonstrated radiological resolution of the ACM after conversion to a ventricular shunt. The second was that of Chumas *et al.* [6] who attributed death in a 2 1/2 year old child with Crouzon's disease who had been shunted from the lumbar subarachnoid space at 6 months for ''progressive ventriculomegaly'', to an acquired ACM. Although the child clearly had an ACM, its role in her demise is less certain. Two other cases have since been reported; one by Sathi and Stieg [29] after multiple lumbar punctures only and one after shunting of an arachnoid cyst [16].

Over the same period of time, several papers appeared reporting on and advocating the use of lumbar CSF shunting for communicating hydrocephalus in particular and for otherconditions also [1, 5, 10, 30]. In a total of 462 cases from the 4 series referred to, there was mention of an ACM in only 2 [13] and syringomyelia in none. In both ACM cases the symptomatic presentation was shortly after shunt insertion and the presumption was that the ACM antedated the shunt. Thus the complications of chronic tonsillar herniation and syrinx formation could at that time be viewed as rare sequelae of chronic lumbar CSF shunting not necessarily precluding the use of this treatment method.

There then appeared 2 reports [7, 26] which suggested, on the contrary, that these complications might be much more common than hitherto suspected and might in fact represent an important contraindication to lumbar CSF shunting. First Chumas *et al.* [7] concluded that the incidence of acquired ACM might be as high as 70% and that the association of hydrosyringomyelia with the Chiari malformation (presuming the latter to be causative) ranges from 30 to 50%. As a true indication of incidence, the study however is seriously flawed. Thus although there were 143 cases in the report, only 54 had what was described as a suitable post-shunt CT (difficulties of diagnosis of a Chiari malformation by non-purpose directed CT notwithstanding) and only 17 asymptomatic patients had an MR scan. Ideally, of course, a study of incidence should be based on pre- and regular post- lumbar shunt MR scanning specifically directed at the craniocervical junction. Indeed, the incidence of asymptomatic tonsillar descent in normal patients may be as high as 14 per cent [2]. In addition, it is entirely possible, given the nature of the conditions, that patients with communicating hydrocephalus, PTC and CSF leaks may have a higher incidence of both Chiari malformation and spinal syrinx than the normal population. Subsequently Payner *et al.* [26] reviewing 10 children treated by lumbo-peritoneal shunting (7 for hydrocephalus, type unspecified, and 3 for intracranial hypertension without hydrocephalus – presumably PTC), reported 4 cases with a sympromatic ACM 0.75 to 4.5 years after shunt insertion and a further 3 cases with anasymptomatic acquired ACM 1.5 to 4.5 years after the initial shunt. The authors took these findings as a basis for recommending against lumbar shunting in communicating hydrocephalus.

Certainly these reports raised several important issues with regard to lumbar CSF shunting. First,

what is the true incidence of such complications and how does this incidence bear on the use of this form of shunting considering the accepted advantages over ventricular shunting? Second, what is the mechanism of acquired ACM and of syrinx formation after prolonged lumbar CSF drainage; how do the two conditions relate to each other and what implications do these findings have on theories of mechanism of both conditions in general? Third, how should these complications be treated in established cases in situations where patients are permanently dependent on the maintenance of CSF drainage?

We have attempted to address some of these problems in the present study. First on the issue of incidence the figures from the PTC group are relevant in that the reason for shunting is uniform and there has been a long (average 7–8 years) and detailed follow-up, in many cases with MR scanning. The incidence in this group has been, to date, 3 cases (4.3%) of symptomatic ACM, 1 case of syringomyelia (1.4%) and 8 cases (11.4%) of asymptomatic ACM. These figures, while suffering the same drawbacks as those of Chumas *et al.* [7], are nonetheless much less alarming and hardly sufficient grounds for abandoning lumbar shunting. Secondly, what light do the findings of this and the related prior studies cast on the mechanism of formation of the Chiari malformation and syringomyelia? Considering the Chiari malformation first, the various theories have been well reviewed by a number of writers [4, 22, 23, 27]. Marin-Padilla and Marin-Padilla [22] provide a useful summarising tabulation which also lists the major proponents of the various theories of which they identify four: CSF circulation disturbances, mechanical traction, focal overgrowth of neural tissue and focal dysgenesis of neural tissue, themselves adding a fifth; disturbance of mesenchymal development giving reduction of the size of the container as the cause rather than overgrowth or displacement of contents. It is probable, on general grounds, that a combination of factors may be operative in any one case, with the importance of each of the particular mechanisms listed depending on the precise prevailing circumstances. Whether these are sufficient for differentiating a developmental form of the malformation from an acquired form on the basis of morphology as Williams has done [34] remains contentious. What the development of the malformation after lumbar CSF drainage does suggest is, of course, that downward movement of the posterior fossa contents is encouraged by the creation of a postural pressure differential, a fact already well established by the known occurrence of acute tonsillar herniation after lumbar puncture in situations of raised intracranial pressure due to a mass lesion. What is not known is what exactly the pressures are in the intracranial and spinal compartments in the presence of a functioning lumbar CSF shunt.

It seems entirely probable that all the factors listed may be contributory, and that the severity and chronicity of the causative factors, including the presumed pressure differential, determine the observed morphological changes on which Williams based his distinction, but which, in reality, consist only of degree of deformity and the associated reactive changes to this.

The cause of syringomyelia is a somewhat more vexed question and has been extensively discussed in the literature without resolution. The authors agree strongly with the observations of Charles Drake in his foreword to the 1973 monograph on the subject by Barnett *et al.* [3] where he writes: "It seems doubtful that syringomyelia could be regarded as a single entity, either from the point of view of the clinical presentation, from the point of view of pathogenesis or, most importantly, from the standpoint of rational therapy." Existing theories on pathogenesis have been well summarised by Hall *et al.* [15] and are, in brief, as follows:

1. Dysraphic (Greenfield, 14) in which it is postulated that there is late degeneration of embryonal cells incorporated into lines of fusion of the developing spinal cord.
2. Ischaemic (Joffroy [18], Netsky [24]) in which there is presumed cavitation following oedema and necrosis due to ischaemia which may itself be secondary to a variety of insults.
3. Neoplastic (Poser [28]) based on the known association of syringomyelia with spinal cord tumours.
4. Hydrodynamic: a group of theories relating syrinx formation to disorders of CSF circulation and associated particularly with the names of Gardner [13], Williams [35] and Conway [9].

The present report clearly emphasises the last group of theories, as all cases described occurred in patients with an established CSF circulation disturbance (assuming PTC is accepted as such) [20] to which an additional abnormality of CSF circulation was added in the form of a subarachnoid space shunt. Particular points of interest to emerge from the present study are that the Chiari malformation may follow, rather than

precede syrinx formation and may possibly be precipitated by drainage of the latter, and that both conditions may occur despite prior decompression of the craniocervical junction and indeed may be aggravated by continued lumbar CSF drainage after posterior fossa decompression. In addition, there is, as with the Chiari malformation, the question of pressure differentials. At present the findings remain controversial and Williams' concept of craniospinal dissociation [35] is by no means universally accepted [25]. In the one case in the present study in which intracranial and spinal pressure were measured simultaneously in the presence of a Chiari malformation and syrinx, there was no difference. Finally, in the presence of ventricles that are small, decompressed and not in communication with the syrinx, and a spinal subarachnoid space drained by a shunt, whence originates the syrinx fluid? Related to this is the question of whether, in fact, an underlying problem of CSF circulation is necessary for the formation of a spinal cord syrinx following the development of a Chiari malformation.

On the third question, that of management, some initial comments on prophylaxis are apposite. Prior to shunting from the subarachnoid space, whether it be lumbar or cisternal, it is desirable to have MR imaging of the cranio-cervical junction and radiological information on the relevant skeletal structures. The findings after lumbar shunting in patients with craniosynostosis, both in the present study and elsewhere [6, 8], precludes this group and any other cases with incidental findings of similar skeletal abnormalities. In the face of an established Chiari malformation plus or minus a syrinx with a functioning, and necessary lumbar shunt, the important step is to change drainage to the ventricular site. This will at least alleviate the problem (and has done so in most reported cases) and may, indeed, reverse the radiological abnormalities [26, 31]. There is possibly no case for craniocervical decompression. It has been characteristically ineffective in reported cases and may, as in case 6 above, aggravate the problem. When a syrinx, alone or in combination, is present the situation is more complicated. There is a paucity of information on shunt site change alone in the management of syringomyelia, although a not unreasonable presumption would be that the treatment of the Chiari malformation by resiting the shunt may lead to resolution of the syrinx as in the case of Sullivan *et al.* [31]. If a syrinx persists, it should be treated in its own right, probably by a syringostomy. Patient 12 who had a lumbo-peritoneal shunt for PTC in the present study has a good long-term result from decompression of the Chiari malformation after syringostomy for the earlier developing syrinx, with preservation of the L-P shunt. Clearly, a rational approach to treatment depends on further information both on clinical outcome and if possible, pressure characteristics. However, a pragmatic approach, based on present data, would make resiting of the shunt to a ventricular or, possibly cisternal site the cornerstone of management with other options such as syringostomy or syrinx shunt and occipito-cervical decompression held in reserve for exceptional circumstances. A further point of therapeutic relevance is that claims of efficacy of shunting from the lumbar subarachnoid space in the treatment of syringomyelia itself, based on a very short follow up should be viewed with considerable circumspection [32].

Conclusions

In conclusion, what are the implications of these findings for the continued use of lumbar CSF shunting? Should this technique be abandoned altogether or can the characteristics of the drainage systems be modified to obviate these problems such that despite such complications as listed for example by Chumas *et al.* [5] the benefits still outweigh the disadvantages? Also, does an alternative site in the SAS e.g. the cisterna magna, confer the same advantages while minimising the problems. The data is not yet available on which to base any firm conclusions on these matters. Clearly, more controlled studies need to be done to establish the incidence of these complications and to establish the differences between long-term symptomatic and asymptomatic changes. Further, information needs to be obtained as to whether significant pressure differentials are created by chronic lumbar shunting, what relevance these have, and whether thay can be minimised by choosing appropriate valve characteristics. In addition, basic comparative clinical outcome studies on subarachnoid space versus ventricular shunting would be helpful, as would additional information on long-term outcome in cisterna magna shunting.

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Comments

The authors ought to be congratulated with this beautiful report on a well conducted study. The subject is most fascinating, the patient material is enormous, taking into account the rarity of the condition, the literature is well reviewed and the discussion is thorough.

C. Avezaat

The authors are to be congratulated for bringing again to our attention this well recognized complication of lumbar CSF shunting. their conclusion that shunt resiting is the first choice in symptomatic cases is important. However, the present study does not improve our knowledge about the prevalence and clinical significance of ACM following lumbar CSF shunting which are critical points for recommending or not against this form of treatment and planning the neuroradiological follow-up of the patients.

Patients in the present study represent a variegated group of cases selected among the total population undergoing lumbar CSF shunting at their institution, but the selection criteria are poorly defined. Though the authors state in the INTRODUCTION that "the first purpose of the paper is to provide a detailed information on the incidence of this complication" the methodology used does not allow to reach this goal. They provide an idea about the incidence of ACH in patients treated for pseudotumor cerebri (the fourth group included all patients treated by one of the authors between 1974–

1996), but we do not know the rate of acquired hindbrain herniation in patients treated for communicating hydrocephalus or other conditions such as cranial deformities (Crouzon, s syndrome). On the other hand, a substantial proportion of patients did not have MR before lumbar shunting (''where possible patients had detailed pre-treatment radiology'') and neuroradiological follow up was far from systematic. How may patients sucessfully treated for ACH by conversion to a ventricular shunting showed resolution of tonsillar herniation ?. Is resolution of acquired hindbrain herniation a requisite for clinical improvement ?.

As the authors say, other investigators have reported the incidence of tonsillar descent into the foramen magnun to be as high as 70% and have suggested that the rate of hindbrain herniation may be even higher in patients with functioning shunts. Though some authors believe that very few patients become symptomatic from such a complication, thus questioning its clinical importance, others clearly prefer ventriculoatrial or ventriculoperitoneal shunting to lumbar shunting for treating communicating hydrocephalus in children as they believe that tonsillar herniation is the rule rather than the exception after lumboperitoneal shunting in the pediatric population.

I agree with the authors conclusion that for assessing the actual risk of developing acquired tonsillar descent into the foramen magnum subsequent to lumbar CSF shunting we need prospective controlled studies including all patients with normal hindbrain configuration in the preoperative MRI study who undergo lumboperitoneal shunting for different pathologies (communicating hydrocephalus, benign intracranial hypertension, etc). Apart from detecting the time lapse of occurrence of ACM, sequential MRI would also reveal the proportion of patients who remain asymptomatic after developing such a change and the proportion of symptomatic patients showing ascent of cerebellar tonsils after conversion to a ventriculoperitoneal shunt. Careful analysis of CSF hydrodynamics and repeated checking of lumboperitoneal shunt function would also contribute to resolve the numerous unanswered questions and apparent paradoxes related to this complication. The whole issue is complicated by the fact that the incidence of asymptomatic tonsillar herniation in normal individuals may be as high as 14% (see Discussion).

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