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Acquired Chiari 1 malformation and syringomyelia following lumboperitoneal shunting for pseudotumour cerebri

An important but not widely recognised complication of lumboperitoneal shunting is the development of a Chiari 1 deformity and syringomyelia. We present a case of a patient who developed symptomatic cerebellar tonsillar descent and syrinx formation following treatment of pseudotumour cerebri with lumboperitoneal shunting.

Case report

A 31 year old woman was diagnosed with pseudotumour cerebri following development of headaches, loss of vision, and papilloedema, in association with a cerebrospinal fluid (CSF) opening pressure of 36 cm H₂O. Cranial imaging showed an attenuated ventricular system and no other abnormality. In particular, the posterior fossa was satisfactory in appearance. She was treated with lumboperitoneal shunt insertion, with resolution of her symptoms.

Twelve months later, the patient reported a 6 month history of left hemisensory loss, left arm weakness, and unsteadiness. Neurological examination revealed wasting and reduced power of the intrinsic muscles of the left hand, and left-sided hyperaesthesia to pin-prick. Magnetic resonance (MR) imaging showed the development of cerebellar tonsillar descent and syringomyelia throughout the cervico-thoracic spinal cord. The patient underwent insertion of a low pressure ventriculoperitoneal shunt and removal of the lumboperitoneal shunt, with subsequent symptomatic improvement. There was, however, no resolution of the syrinx on follow up MR imaging.

Discussion

The development of cerebellar tonsillar descent is a recognised but rarely reported complication following lumboperitoneal shunting, 1-5 usually in the treatment of communicating hydrocephalus. 1-3 It has been reported to occur in a large proportion of paediatric patients undergoing this procedure, with Chumas *et al* reporting a 70% incidence in this age group, 1 but its incidence in the adult population is undefined. The development of secondary syringomyelia appears to be much less common, with the

above paediatric patients reporting an incidence of syrinx formation of 4%. The development of Chiari 1 and syringomyelia formation following lumboureteral shunting for the treatment of pseudotumour cerebri is recognised but has been less commonly reported.^{2 4 5}

There is a small number of papers reporting chiari development following lumbar shunting for communicating hydrocephalus in children, but only two case reports of syringomyelia formation.

The association of syrinx formation and cerebellar tonsillar descent through the foramen magnum is well described, and is postulated to occur as a consequence of a cranial-spinal CSF pressure gradient and diversion of CSF down the central canal of the spinal cord rather than over the cerebral convexities. 4 It would seem remarkable that this complication is not seen more commonly in the treatment of pseudotumour cerebri.

The non-resolution of the syrinx, in our case following lumboperitoneal shunt removal, is consistent with other workers' experiences, although resolution has been reported in one instance.⁷

In conclusion, we describe the development of Chiari 1 deformity and syrinx formation as an important but otherwise poorly recognised complication of lumboperitoneal shunting in patients with pseudotumour cerebri.

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Effect of posture on levels of arousal and awareness in vegetative and minimally conscious state patients: a preliminary investigation

Moderate to severe brain injury is estimated to occur in 25 individuals per 100 000 population every year. Of these, 10–20%

never fully regain consciousness but remain in a vegetative or minimally conscious state.¹ Patients in the vegetative state may appear at times to be wakeful, with cycles of eye closure and eye opening resembling those of sleep and waking, but show no sign of awareness or of a functioning mind.² In contrast, patients considered to be in a minimally conscious state are said to show inconsistent but definite evidence of awareness despite profound cognitive impairment.³

At present, the pathophysiology underlying the vegetative and minimally conscious states is unclear, a standard treatment approach is lacking, and very little has been discovered to advance rehabilitation techniques. It is widely acknowledged that active rehabilitation should begin early in the intensive care setting, and should be applied to all patients (including those who remain mechanically ventilated). However, this is not yet routine practice. Several reports have highlighted the generic benefits of early rehabilitation,4 however, the benefits of specific interventions remain to be demonstrated. Over the last year, our group has investigated the effects of postural change on levels of arousal and awareness.

A total of 12 patients (eight men, four women; mean age 49 years, range 19-71) classified as either vegetative (n = 5) or minimally conscious (n = 7) according to international guidelines^{2 3} were assessed using the Wessex Head Injury Matrix (WHIM), a 62 point score, which records the recovery of behaviours in brain injured patients.5 Patients were assessed lying in bed, during a 20 minute period of standing using a tilt table at 85°, and again while lying in bed. During the observations blood pressure was measured using an oscillometric cuff. The observations were repeated over a one week period, and the median highest ranked behaviour and median total number of behaviours observed were recorded. The local research ethics committee approved all investigations. Informed assent was obtained from the next of kin

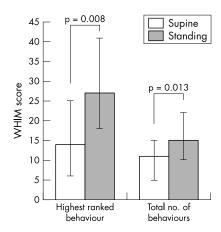


Figure 1 The median highest ranked behaviour and total number of behaviours observed in the lying and standing positions for both vegetative and minimally conscious patients. The highest rank (p=0.008) and total number of behaviours (p=0.0013) observed increased significantly in the standing position. Error bars indicate the interquartile range. WHIM, Wessex Head Injury Matrix.

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Table 1 Highest ranked behaviours recorded in the supine and standing positions for each patient

Patient	VS/MCS*	Supine score	Behaviour observed	Standing score	Behaviour observed
1	VS	43	Smiled	43	Smiled spontaneously
2	VS	4	Eyes held by painful stimulus <2 s	4	Eyes held by painful stimulus <2 s
3	VS	5	Looked at person briefly	26	Frowned/grimaced during physio
4	VS	1	Eyes opened briefly	49	Vocalised in response to pain
5	VS	14	Yawned, sighed	26	Frowned/grimaced during physio
6	MCS	13	Looked at person moving limbs <3 s	16	Turned eyes to look at person talking
7	MCS	20	Vocalised during physio	36	Switched gaze from one person to another
8	MCS	26	Frowned/grimaced in response to pain	34	Monosyllabic response to questions
9	MCS	14	Yawned, sighed	14	Yawned, sighed
10	MCS	18	Tracked for 3–5 seconds	28	Looked at object when requested
11	MCS	8	Made eye contact	23	Showed selective response to preferred people
12	MCS	42	Could find a card from four	43	Smiled spontaneously

*Patient classification at the time of recruitment is denoted VS (vegetative state) or MCS (minimally conscious state).

We found that eight patients (three vegetative and five minimally conscious) showed consistent improvements in the highest ranked behaviours (table 1; p = 0.008) and total number of behaviours (p = 0.013)observed in the standing position (fig 1). Three patients (two vegetative and one minimally conscious) showed no change and one minimally conscious patient showed only an increase in the highest ranked behaviour observed. Although WHIM scores in three vegetative patients increased during standing, the behaviours observed did not reach a level suggesting awareness of self and/or environment. After standing the WHIM scores in the supine position were equal to or below those acquired before standing. No change in blood pressure was observed (p = 0.3).

Our preliminary results suggest that positional changes may have a significant impact on behaviours in vegetative and minimally conscious patients. Although the benefit of this phenomenon in rehabilitation remains unproved, these findings have clear implications for the assessment and categorisation of patients. Neurological assessments used to classify patients according to international guidelines relating to the vegetative and minimally conscious states typically take place with the patient lying in bed. Where physical constraints permit, it may be important to also observe patients in the standing position.

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D Badwan-Patient recruitment and data analysis. D Menon-Project supervision. J Pickard—Project supervision.

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BOOK REVIEWS

Neurological disorders in pregnancy

Edited by Jacqueline M Washington. Published by the Parthenon Publishing Group, 2004, £87.00 (hardcover), pp 150. ISBN 1-84214-

Many neurological disorders occur in women of childbearing age. This small book is designed to bridge the wide gap existing between the disciplines of neurology and obstetrics. It provides a concise overview of the most common neurological disorders that may be seen during pregnancy. Three categories of problems are encountered by obstetricians and neurologists: management of neurological pre-existing disorders during pregnancy, neurological disorders directly due to pregnancy, and neurological affections that require special treatment considerations during pregnancy. The book covers these three categories with eight chapters devoted to migraine, cerebrovascular disease, epilepsy, back pain, multiple sclerosis, peripheral nerve disorders, myasthenia gravis, and central nervous infections. Each chapter includes consideration of the influence of pregnancy on the disorder, the effect of the disorder on pregnancy, and potential effects of proposed therapies on the developing foetus-all concerns shared by every clinician who care for pregnant women. The different chapters provide a useful resource with lists of dosages, contraindications, monitoring guidelines, and side effects of drugs in pregnancy

Two other chapters, covering muscle diseases (in particular myotonic dystrophy) and brain tumours, could have been useful. One also could regret the nearly complete absence of figures or diagrams for a book intended not only for neurologists but also for obstetricians. In contrast, most chapters contain many useful tables.

A few remarks are also worth mentioning. For example, the section covering the course of migraine during and after pregnancy is sometimes redundant and could have been summarised. Post partum angiopathy should also be added to the aetiologies of postpartum headaches. In the chapter on cerebrovascular disease, eclampsia and hemorrhage sections could have been better detailed.

On the whole, this book represents a useful concise text (more than in-depth literature summary or detailed analysis of complex issues) written in a balanced, practical, and informative way. It can be used by a wide audience and will facilitate understanding and treatment of neurologic problems in pregnant women.

C Lamy

The neuropathology of dementia, 2nd edition

Edited by M Esiri, M-Y Lee, J Q Trojanowski. Published by Cambridge University Press, Cambridge, 2004, £195.00, pp 563. ISBN 0-5218-1915-6

Inspecting the hardcover graphics of this new edition your reviewer was startled to find blazoned his comments on the previous, and first, edition to encourage your purchase. It is therefore clear that I am a supporter of this enterprise in principle - though modesty will prevail and I will not flaunt my prescience in