ORIGINAL ARTICLE

Treatment of Secondary Tonsillar Herniation by Lumboperitoneal Shunt Revision

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ABSTRACT: ***Background:*** Idiopathic intracranial hypertension (IIH) is a condition that usually affects young, obese women. Management is aimed at controlling symptoms of increased intracranial pressure (ICP) and prevention of visual failure due to papilledema. A common surgical treatment for IIH is the insertion of a lumboperitoneal shunt (LP shunt). Secondary symptomatic tonsillar herniation is an uncommon side effect following lumbar cerebrospinal fluid diversion. ***Methods:*** We present two cases of symptomatic secondary tonsillar herniation, one associated with a syrinx, in patients with IIH following valved LP shunting. Treatment options for this side effect may include transplanting the shunt to the ventricular system or decompression of the foramen magnum. ***Results:*** In our cases we elected to alter the construct of the LP shunt by inserting a programmable valve which led to clinical and radiological reversal of the tonsillar herniation as well as a dramatic reduction in an associated syrinx. ***Conclusions:*** When faced with LP shunt induced symptomatic secondary tonsillar herniation, consideration may be given to altering LP shunt dynamics, prior to inserting a ventricular catheter into normal sized ventricles or decompressing the posterior fossa.

RÉSUMÉ: Une modification d'une dérivation lombo-péritonéale peut faire rétrocéder une hernie amygdalienne secondaire. ***Contexte :*** L'hypertension intracrânienne idiopathique (HII) est une maladie qui atteint habituellement de jeunes femmes obèses. Le traitement vise à contrôler les symptômes dus à l'augmentation de la pression intracrânienne (PIC) et à prévenir l'atteinte visuelle due à l'?dème papillaire. Le traitement chirurgical courant de l'HII est la mise en place d'une dérivation lombo-péritonéale (DLP). La hernie amygdalienne secondaire symptomatique est un effet secondaire rare de cette intervention. ***Méthodes :*** Nous décrivons deux cas de hernie amygdalienne secondaire symptomatique, dont une associée à un syrinx, chez des patientes atteintes d'HII après la mise en place d'une dérivation LP avec valve. La transplantation de la dérivation au système ventriculaire ou la décompression du trou occipital constituent le traitement de cet effet secondaire. ***Résultats :*** Nous avons choisi de modifier la dérivation LP en insérant une valve programmable, ce qui a fait rétrocéder la hernie amygdalienne tant au point de vue clinique que radiologique et qui a entraîné une diminution dramatique du syrinx qui y était associé. ***Conclusions :*** Quand il existe une hernie amygdalienne secondaire symptomatique, on peut envisager d'abord une modification de la dynamique de la dérivation LP avant d'avoir recours à la mise en place d'un cathéter ventriculaire dans un ventricule de taille normale ou à la décompression de la fosse cérébrale postérieure.

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Idiopathic intracranial hypertension (IIH), or pseudotumour cerebri, was first described over a century ago by Quincke.[1] This syndrome commonly affects young, obese women of childbearing age with a prevalence reaching as high as 20 cases per 100 000.[2] Patients with IIH present with symptoms of increased intracranial pressure (ICP) in the absence of ventriculomegaly and/or a mass lesion, and normal cerebrospinal fluid (CSF) studies, as described by Dandy.[3] This entity has been expanded to include the absence of localizing signs on neurological exam, normal computed tomography (CT) / magnetic resonance imaging (MRI) findings without evidence of dural sinus thrombosis, and ICP of greater than 250 mm water with normal CSF cytology. Once known as benign intracranial hypertension, the reports of papilledema and subsequent vision loss in this patient population led to the renaming of the syndrome, replacing 'benign' with the term idiopathic.[4]

Treatment of IIH is primarily aimed at preventing vision loss and symptoms of increased ICP. Dietary management of obesity with resolution of papilledema has also been well documented.[5,6] Associations with various medications such as vitamin A, antibiotics (tetracycline/minocycline, nalidixic acid, fluoroquinolones, and sulfa drugs), oral contraceptives, progesterone, and danazol, have also been linked to IIH as a spectrum of so-

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called secondary pseudotumour syndromes.[7] Carbonic anhydrase inhibitors are, to date, the only effective medications for treating papilledema by decreasing the production of CSF. Furosemide is a second line medication for those who cannot tolerate carbonic anhydrase inhibitors.[8] Controversy revolves around the use of corticosteroids in the treatment of papilledema, and is generally not advised for routine nor chronic treatment.[9] Serial lumbar punctures and drainage of CSF may offer temporary alleviation of symptoms, but is not a long-term solution.[10]

Surgical management of IIH is considered in the setting of progressive vision loss despite maximal medical therapy; or recalcitrant headaches.[4,11] Optic nerve sheath fenestration and CSF diversion are potential surgical treatments. The mechanism whereby optic nerve sheath fenestration acts is unclear, and may be secondary to filtering of CSF thereby improving peripapillary circulation.[12]

The most commonly used CSF diversion techniques include lumboperitoneal (LP) and ventriculoperitoneal (VP) shunting.[13] Utilization of LP shunts over the last 40 years has waxed and waned. The main advantage of LP over VP shunts in the pseudotumor population center around the avoidance of cannulating normal sized ventricles. Unique complications such as shunt migration, scoliosis, arachnoiditis and secondary tonsillar herniation have been associated with LP shunts. In the last decade, with the development of image guided neurosurgery, the safety and efficacy of cannulating normal sized ventricles has improved and the pendulum has shifted towards VP shunting.

Lumboperitoneal shunts have an overall revision rate of 52%, related to complications of catheter obstruction, catheter migration, infection, arachnoiditis and lumbar radiculopathy.[4,14,15] Lumbar CSF diversion may also lead to the entity of secondary tonsillar herniation (acquired Chiari malformation) which with the advent of MRI has become well recognized. The true incidence of this disorder is unknown, and the exact pathogenesis unclear. Thankfully the majority of these clinical situations involve an asymptomatic patient population.[16,17] When symptoms do arise, a constellation of findings such as neck pain, vomiting, photophobia, sixth nerve paresis, ataxia, motor weakness and sensory disturbances may exist.[17-20] In addition to tonsillar herniation associated MRI findings may include syringomyelia, leptomeningeal enhancement, and subdural effusions.[18-20]

Treatment options when faced with symptomatic secondary tonsillar herniation focus on repositioning the shunt to the ventricular system or an intracranial subarachnoid space thereby abandoning the lumbar subarachnoid space. Other options include decompression of the foramen magnum with or without shunt transplantation.[17] Repositioning the shunt to the ventricular system however does not necessarily preclude any further problems, and foramen magnum decompression alone has been reported with worsening of neurological function.[17] An attractive treatment option focuses on preserving the LP shunt but changing the dynamics such that the shunt can accomplish the necessary CSF diversion to treat the primary disorder and prevent the development of tonsillar herniation.

The use of programmable valves have been reported in the treatment of hydrocephalus secondary to idiopathic normal pressure hydrocephalus, aqueductal stenosis, Chiari malformation, and meningitis.[21]

We herein report, the first case series demonstrating the reversal of secondary tonsillar herniation by a simple alteration in the construct of our LP shunt. The insertion of a programmable valve in our series allowed us to preserve lumbar CSF diversion as a primary treatment modality for IIH. Hopefully this case series will contribute to the literature by allowing clinicians to recognize this potential option before abandoning the lumbar subarachnoid space.

Methods

A case series of two females, ages 20 and 35, who presented to our clinic and were followed over 4.5 years between June 2002 to January 2007.

Results

Case 1

This patient was initially diagnosed with IIH in 1997 at the age of 12. Clinical presentation included headaches and ICP values in the 20-40 mm H_2O range and was managed with serial lumbar punctures, acetazolamide, and steroids. In 2000, at the age of 16, progressive vision loss prompted bilateral optic nerve sheath fenestrations. She presented to our clinic in June 2002 with recalcitrant headaches despite daily lumbar punctures, and maximal medical therapy including acetazolamide, furosemide, and steroids. Her MRI at that time was normal (Figure 1a). With high opening pressures of 30-40 cm H_2O and failing medical treatment, we elected to perform a non-valved LP shunt procedure (Medtronic lumboperitoneal catheter system). Her symptoms improved, and at eight weeks following her surgery, both the acetazolamide and furosemide were discontinued with improvement of her blindspot and optic disc edema. Several months later in October, she presented with high-pressure headache symptoms secondary to shunt migration, prompting revision of her LP shunt with insertion of a low-pressure valve, which subsequently improved her symptoms.

In January of 2004, she presented to our emergency department with headaches and ocular pain. It was hard to ascertain if her headaches were high pressure or low pressure in nature and her ocular pain was non specific. Her shunt was explored and was found to be working properly An MRI scan identified the development of a secondary Chiari malformation (Figure 1b). There was no evidence of a syrinx, venous thrombosis, or leptomeningeal enhancement. Throughout the year, she continued to have symptoms of chronic occipital headaches and possibly low pressure headache symptoms. In addition, by the fall of 2004 she began to develop symptoms of weakness and burning pain in her upper limbs that were felt to be secondary to her acquired Chiari malformation. The decision was made at the time to replace the valve of her LP shunt with a Codman-Hakim programmable valve. We increased the resistance of her LP shunt by setting the valve at 150 mm Hg and she was closely followed with clinical reversal of her symptoms and radiological reversal of her acquired Chiari malformation without evidence of syrinx formation (Figure 1c).

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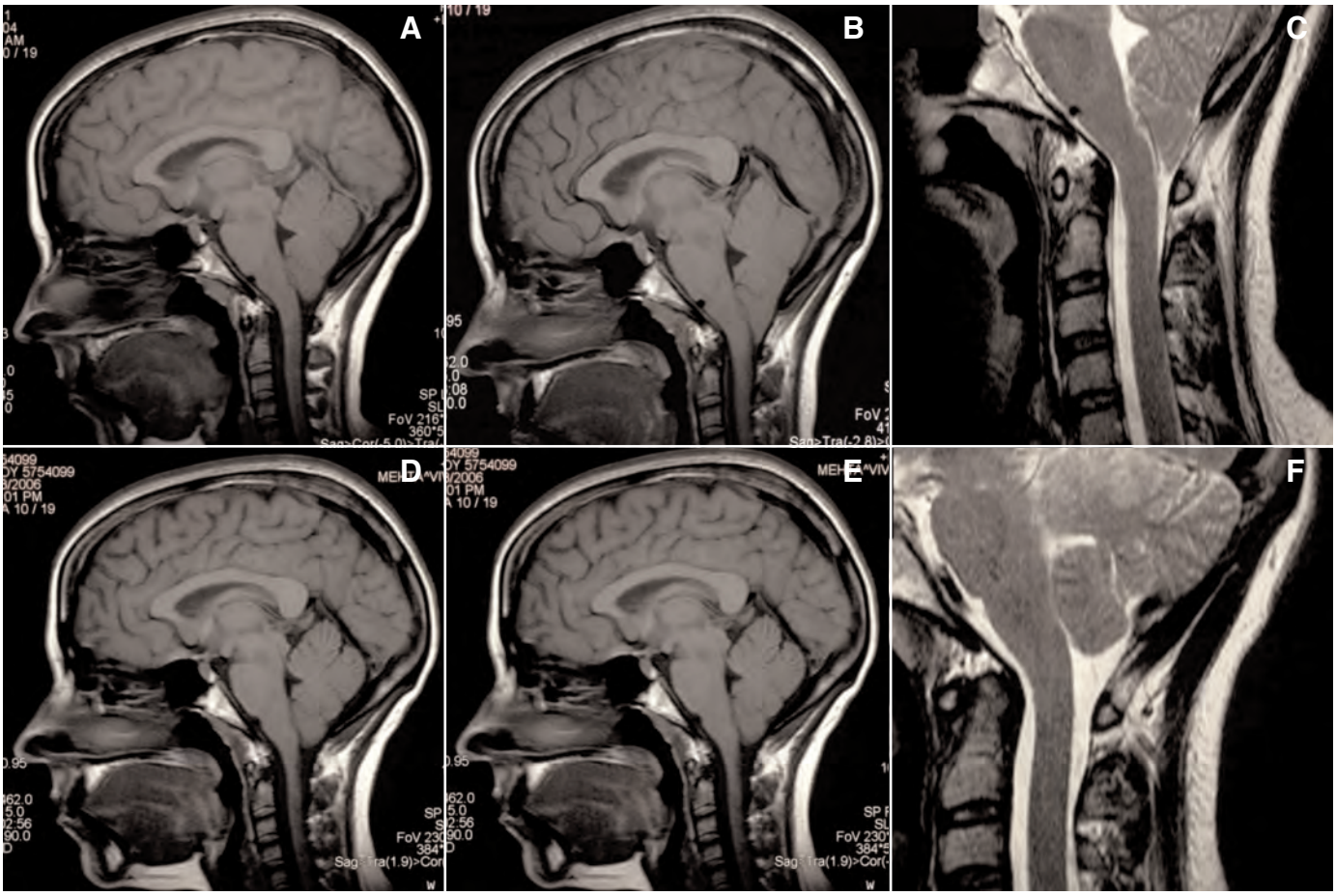

Figure 1: *MRI images of a female with IIH presenting with a symptomatic acquired Chiari malformation. A) A normal MRI at the initial time of presentation at the age of 12 in 1997. B, C) Patient developed an acquired Chiari malformation in 2004 following multiple LP shunt revisions using nonprogrammable valves with significant and symptomatic tonsillar herniation. D) Improvement of symptoms and reversal of herniation at 8 months following insertion of a programmable Codman-Medos valve. E, F) Follow-up at 18 months post-insertion of the programmable valve demonstrates further reduction of her Chiari malformation and total resolution of symptoms.*

Case 2

This 38-year-old right-handed female first presented to our clinic in 2002 with symptoms of pseudotumour cerebri and a normal MRI scan (Figure 2a). At that time, she had undergone repeated lumbar punctures by her neurologist with pressure readings from 18 to 30 cm H_20. She was initially managed using acetazolamide, a trial of steroids, and had successfully lost 40 pounds. She did not have visual field changes or significant papilledema. She had an ICP monitor inserted that confirmed raised intracranial pressure. Given the persistence of her headaches despite medical therapy, a LP shunt with a low pressure PS medical valve was inserted in January, 2003, with subsequent marked improvement of her headaches. Her headaches returned at the end of 2003, but were managed using over-the-counter analgesia, massage, and acupuncture. In September, 2004, following the birth of her child, she developed weakness in her hands with nerve conduction studies showing mild bilateral carpal tunnel syndrome. She was followed conservatively, but returned to her neurologist in October 2005 with an eight week history of progressive weakness and muscle atrophy in her hands, left worse than right. More significantly, she had clawing of some of the fingers in her left hand with weakness in the hand muscles innervated by the median, ulnar, and radial nerves, again left worse than right. This was accompanied by proximal weakness of the left deltoid and brisk stretch reflexes. An MRI scan showed an acquired Chiari malformation with 8 mm of tonsillar herniation associated with a large syrinx extending from the base of the dens to the level of the fourth thoracic vertebra (Figure 2b). There was no evidence of venous sinus thrombosis or leptomeningeal enhancement. The patient with taken to the operating room and her shunt revised using a PS Medical Strata (Medtronic) programmable valve set at 2.5 (the highest resistance). Post-operative imaging one month later showed mild regression of the herniation to 6 mm below the

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foramen magnum but dramatic reduction in the size and diameter of the syrinx, now evident between the C1-C2 levels to the inferior aspect of C3 (Figure 2c). More significantly, the patient's claw-hand deformity resolved and she was pain-free. Repeat imaging at three months demonstrated no change in the degree of tonsillar herniation or further extension of the syrinx (Figure 2d). The patient has continued to remain asymptomatic.

Discussion

The etiopathogenesis of IIH is unknown. Various hypotheses surrounding altered CSF production and flow dynamics have been postulated to account for the symptoms of increased ICP within the setting of normal neuroimaging.[7] Surgical decision making can often be difficult with these patients. What operation to utilize, minimizing complications and surgical timing are often subtle nuances that come into play?

Our two cases of an acquired symptomatic Chiari malformation were reversed by altering the construct of our LP shunt. The first case demonstrated a dramatic reversal of an acquired Chiari malformation using the Codman-Hakim programmable valve system. This valve consists of a ball-cone mechanism whereby a motor adjusts the opening pressure on a spiral staircase-shaped cam, and was first reported by Black et al, for the treatment of slit ventricles, chronic hydrocephalus with aqueductal stenosis, normal pressure hydrocephalus, and normal pressure hydrocephalus with subdural hematoma.[21] Our second case involved an acquired Chiari malformation with syringomyelia. A Medtronic Strata valve was inserted in this patient. The valve utilizes a ball-spring mechanism activated by an external magnet to adjust the flow of CSF at the new desired pressure. There was a significant reduction in the extension and size of the syrinx with complete resolution of her myelopathy and hand deformity.

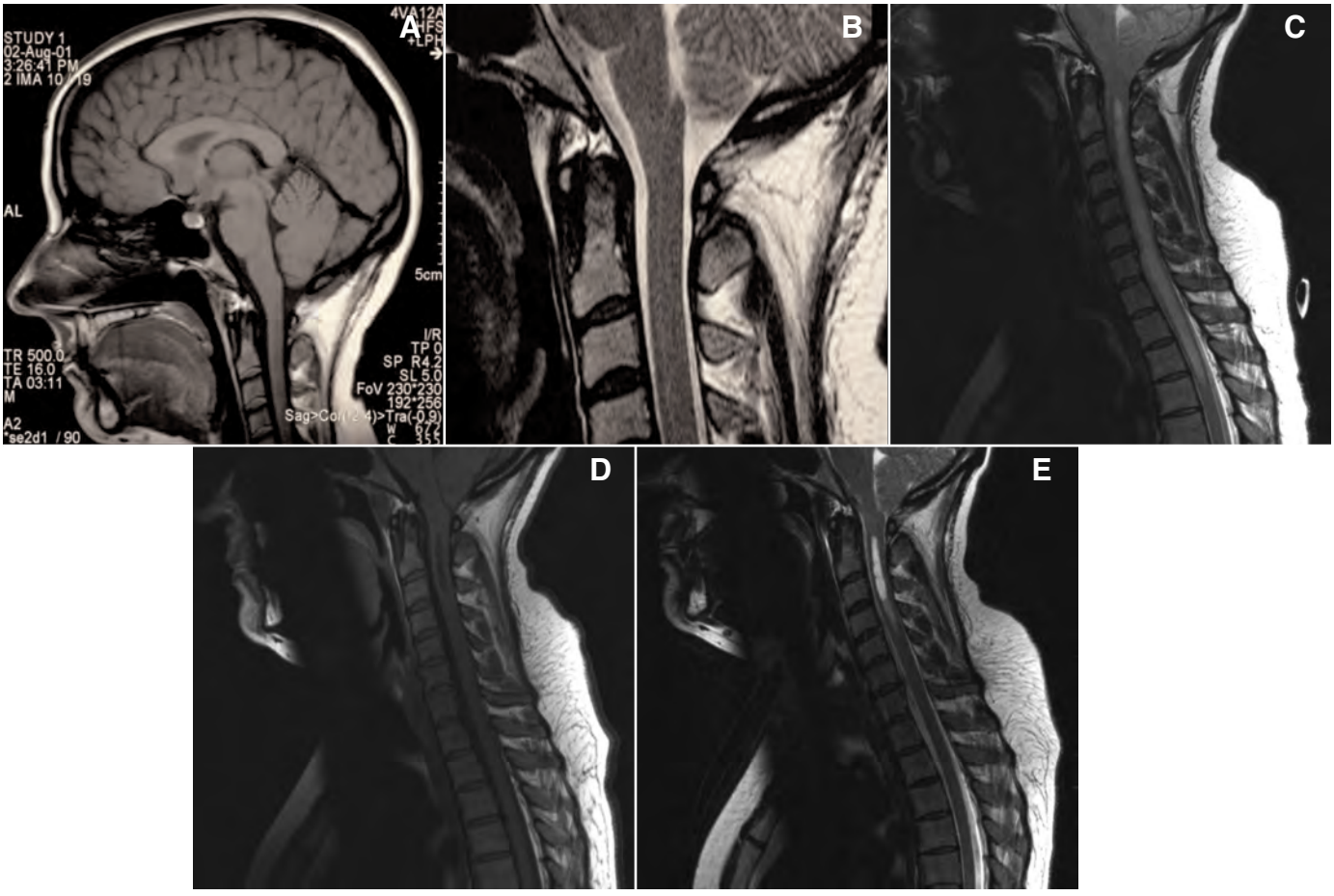

Figure 2: *MRI images of a 38-year-old female presenting with an acquired Chiari malformation and syringomyelia following LP shunting for IIH. A) Normal MRI at time of initial presentation in August, 2002. B) MRI scan in November, 2002 showing a normal cervicomedullary junction at the time of initial LP shunting with a low-pressure, nonprogrammable valve. C) Follow-up scan in October 2005 showing an acquired Chiairi malformation and an extensive syrinx extending from the C2 to T3. Patient presented with signs of syringomyelia and refractory headaches. D) Dramatic reversal of the syrinx and resolution of the symptoms of syringomyelia in November 2005, 1 month following shunt revision with a programmable Strata valve. E) Scan at 3 months in January 2006 following insertion of the Strata valve showing stable regression of the syrinx. The patient has remained free of headaches and has no signs of syringomyelia.*

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To the best of our knowledge, this is the first reported case series of reversal of an acquired Chiari malformation using an LP shunt with a programmable valve system. The main advantage of a programmable valve centers on the ability to non-operatively tailor valve function in order to control the symptoms of unpredictable CSF dynamics among IIH patients. It has been argued that the higher cost of the programmable valve is offset by limited revisions and the ability to fine tune these valves without surgery.[22]

Unfortunately standard complications associated with LP shunting are also applicable when utilizing a programmable valve.[5] In addition a recent retrospective comparison study among the pediatric population demonstrated that programmable valves may be more vulnerable to primary valve failure. Within this series a shunt malfunction occurred in 35 of its 100 subjects who received programmable valves, nine owing to primary valve malfunction, translating into an 11.1% annual valve failure rate. This was in contrast to a 20.2% shunt malfunction in the nonprogrammable valve group, none of which were due to valve failure.[23] Postulations as to why this programmable valve may malfunction include possible blockage secondary to accumulation of proteinaceous debris in the valve, wear and tear on the valve, and added mechanical complexity of the programmable valve.

Tonsillar herniation following LP shunting has been well documented although it was initially considered to be a rare event.[4,11,16,24-28] This rarity of this occurrence has since been debated in studies suggesting that the incidence of herniation is in fact higher than previously suspected.[4,24] Earlier postulates that cephalocranial disproportion may have attributed to secondary tonsillar herniation (as seen in the pediatric population) are difficult to comprehend as a single pathogenesis since this type of herniation can occur in adults.[28] It is now thought that the mechanism is secondary to formation of a craniospinal CSF pressure gradient from LP shunting, causing overshunting of CSF and subsequent tonsillar decent.[24,29] Regardless, tonsillar herniation secondary to LP shunting is often an asymptomatic condition that is recognized on surveillance imaging.

Johnston et al have published one of the largest series of acquired symptomatic Chiari malformations following spinal CSF diversion.[17] Authors of this series divided their 14 cases over a 22-year-period into four groups; craniofacial dysostosis and communicating hydrocephalus, simple communicating hydrocephalus, a miscellaneous group and pseudotumor cerebri. Within their 14 patients some had tonsillar herniation alone, tonsillar herniation with syringomyelia and one patient had isolated syringomyelia. A robust group of 70 patients with pseudotumor were treated with lumbar CSF diversion. Eight patients developed entirely asymptomatic secondary tonsillar herniation at an average of 6.6 years post shunting. Three patients had symptomatic secondary tonsillar herniation. Overall, five surgical procedures were required to treat these three patients (syrinx to subarachnoid shunt, Chiari decompression, LP shunt conversion to cisternal-atrial shunt).

The experience of Johnston's[17] group in Australia is quite different from the experience described by Chumas et al[16] at the Hospital for Sick Children and Payner et al.[24] The group from Toronto felt the rate of secondary tonsillar herniation may be as high as 70%. Serious flaws within this paper center around the number of patients followed up and the number of CT scans used to make the diagnosis of an acquired Chiari. Payner et al[24] presented a small series of ten patients treated by lumbo-peritoneal shunting, of which four cases developed symptomatic secondary Chiari malformations and a further three cases developed asymptomatic secondary Chiari malformations.

It is important to note that the overshunting of CSF in the LP shunt system does not occur through siphoning of CSF, unlike that seen in ventriculoperitoneal (VP) shunts, as the LP shunt is not exposed to gravity to the same extent as VP shunts are. Rather, overdrainage of CSF is due to the effects of positive inlet pressures of the vertical lumbar subarachnoid fluid column on the distal slit valves of the shunt. As a result, all currently manufactured LP shunts are configured for high-pressure systems in an attempt to manage this inlet hydrostatic pressure. We propose that using an LP shunt system with a programmable valve allows for further control of this pressure gradient, thus preventing and reversing tonsillar herniation.

Syringomyelia associated with a Chiari I malformation has been well documented.[27,30] This phenomenon has also been documented as a complication of LP shunting.[26,31-33] An isolated secondary symptomatic syrinx in the absence of tonsillar herniation has also been reported with lumbar CSF diversion.[17] Our second case demonstrated reversal of a syrinx associated with an acquired Chiari malformation using the Medical® Strata programmable valve. In addition to the imaging changes, there was a profound regression of her myelopathic signs and symptoms.

There are many limitations within this case series, namely the small number of patients reported and the short period of follow-up. Having a sample size of two does not allow us to extrapolate about how commonly this problem exists nor whether a programmable valve will be successful in every case at resolving the malformation. It is also possible that initial use of a different construct of the LP shunt (i.e. flow controlled valve, high differential pressure valve, differential valve with an anti-siphon device, horizontal-vertical valve) may have avoided the formation of an acquired Chiari malformation, thus precluding the need for shunt revision with a programmable shunt. Within our case series, it should be noted that the Codman-Hakim system utilized is a completely open or closed differential system that is always open in the erect position. We did not utilize the Siphonguard with this valve and therefore it could be argued that any higher resistance valve would have precluded the development of this complication. Another criticism that is valid pertains to our second case, were we utilized the Delta mechanism, which in theory would be unable to prevent siphoning since it was downstream hydraulically from the source fluid. In addition the programmable aspect of the valve was not used significantly however the availability for the potential use of this option was why this valve was initially placed into the construct.

Furthermore, one may argue that the resurgence of ventriculoperitoneal shunting for IIH in the era of image-guided surgery may negate the need for LP shunting, given the high complication rate associated with the procedure. We currently feel that if CSF diversion through the lumbar subarachnoid space can be utilized it has benefits over ventricular catheters as there

are still concerns with the proximal ventricular catheter becoming occluded and the subsequent need for revision. As such, we favour using a valved LP shunt to allow for ease in assessing shunt function and sampling of CSF.

In summary, we report two cases of symptomatic secondary tonsillar herniation successfully treated by inserting a programmable valve into the LP system. Knowledge of this potential therapeutic option may avoid transplantation of the shunt into the ventricular system or a foramen magnum decompression. This novel case series suggests that altering the construct of a LP shunt with a higher resistance component may allow one not to abandon the lumbar subarachnoid space.

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