ORIGINAL ARTICLE

A Possible Role for Temporary Lumbar Drainage in the Management of Idiopathic Intracranial Hypertension

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ABSTRACT

This paper reports 14 patients with idiopathic intracranial hypertension (IIH) who experienced immediate and sustained resolution of their IIH; 13 in the setting of a low-pressure headache and 1 who underwent lumbar drainage for 4 days draining the cerebrospinal fluid (CSF) at a rate of 5–15 mL/h. These observations, if confirmed, suggest that draining CSF using a temporary lumbar drain draining CSF at a rate greater than it is produced may potentially have a pivotal role in the management of IIH.

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Introduction

Idiopathic intracranial hypertension (IIH) is the syndrome of headache due to raised intracranial pressure (>25 cm H₂O in adults and >28 cm H₂O in children) where the cerebrospinal fluid (CSF) is normal and there is no alternative pathology on imaging.¹ These criteria have supplanted the Dandy criteria since Whiteley et al. demonstrated that normal CSF pressure is between 10 and 25 cm H₂O.² The major consequence of untreated or undertreated IIH is visual loss. A similar clinical picture of raised intracranial pressure with papilloedema occurs with intracranial venous occlusive disease.

The aetiology is unknown, and what triggers the initial rise of intracranial pressure is also unknown. Transverse sinus “obstruction” is intermittent and secondary to the raised pressure.³ The transverse sinus “obstruction” must play a role in perpetuating the raised pressure, as stenting of the transverse sinuses^{4–8} has been used with success to treat IIH. Collapsibility of transverse sinuses has been demonstrated to greatly influence intracranial pressure in an experimental model using a Starling-like resistor.⁹

Initial treatment after confirmatory lumbar puncture (LP) is acetazolamide (Diamox) and a low-salt

weight loss diet in obese individuals.¹⁰ Although IIH can resolve after a single^{11,12} or multiple¹³ LPs, how this occurs is unclear.

Lumboperitoneal shunts, first described by Vander Ark et al. in 1971,¹⁴ despite the potential for complications,^{15,16} have been employed in refractory cases of IIH. The IIH often recurs when they occlude, indicating that they do not cure the problem but only ameliorate the symptoms.

Although there is no large prospective series describing the natural history of IIH, a protracted course lasting months to years appears to be common.^{17,18}

This paper describes a series of patients with IIH who experienced an immediate and sustained resolution of the IIH in the setting of prolonged CSF drainage at a rate greater than CSF production: 13 in the setting of a low-pressure headache (11 after an LP, 1 with a lumbar drain, and 1 with a shunt) and 1 patient who was intentionally treated for 4 days with a lumbar drain.

Materials and methods

This report is based on patients with IIH who fulfilled the Dandy criteria¹⁹ and who would also fulfil

the more recent criteria² seen over 10 years between 1998 and 2008. The patients were retrieved from the principal author's (co-located at Barwon Health) private practice personal electronic database (where a diagnosis has been recorded on all patients between 1984 and 2016), the medical records of Barwon Health, and 1 case treated prospectively with a temporary lumbar drain by the second author.

Some of the cases reported in this paper have been published previously in the *World Journal of Neurology*.²⁰

Results

A total of 31 patients with IIH were identified. Eleven patients had an immediate and sustained resolution of their IIH after an LP. In 4 the closing pressure was <10 cm H₂O, whereas in 7 the closing pressure was either >10 cm H₂O or had not been recorded. All 11 had developed a low-pressure headache, and in 2 patients the low pressure was confirmed at LP (<5 cm H₂O). The minimal duration of the low-pressure headache was 48 hours. Two other patients had an immediate and sustained resolution of their IIH: one in the setting of a lumbar drain and the other when a lumboperitoneal shunt caused low-pressure headache and had to be removed.²¹

All but 2 of these patients remained free of symptoms for up to 10 years, average 3.5 years. One had a recurrence 4 months later; the other patient, who had developed a low-pressure headache in the setting of over shunting remained free of symptoms for 2 years, was lost to follow-up for a further 5 years, at which stage she returned with a recurrence of IIH symptoms.

The 14th patient had a temporary lumbar drain inserted by the second author when her vision continued to deteriorate despite optic nerve fenestration. The CSF was drained at a rate of 5–15 mL per hour for 4 days. The patient experienced severe symptoms of low pressure for the duration of drainage. The IIH resolved, and she has remained in remission 16 months after the procedure.

Discussion

An immediate and sustained resolution of IIH does not reflect the “natural history” of patients treated with drugs such as acetazolamide, optic

nerve fenestration, lumboperitoneal shunt, or transverse sinus stenting where recurrence often occurs with occlusion of the shunt²² or recurrent stenosis adjacent to the stent.²³

We are not the first to observe resolution of symptoms in the setting of low-pressure headache,^{24,25} the reversal of transverse sinus narrowing,^{11,25} nor to suggest that IIH consists of a vicious cycle of raised pressure perpetuated by transverse sinus compression.¹¹ Pickard et al.²⁶ have suggested that the vicious cycle can be interrupted by draining CSF.

The principal author developed an interest in IIH when he encountered a patient with low-pressure headache after an LP in the setting of clinically definite IIH. How could one go from a very high to a very low pressure? It was if someone had thrown a switch. A low-pressure headache represents a CSF leak where the rate of leakage must be greater than CSF production. Aware that the transverse sinus narrowing is secondary to the raised pressure³ and that stenting can lead to a resolution of IIH, the principal author subsequently studied the effect of lowering CSF pressure on the transverse sinus narrowing and observed reversal of the narrowing of one then both transverse sinuses, with lowering of CSF pressure at LP initially to 11 cm H₂O and then at a second LP to 8 cm H₂O.²⁷ The principal author hypothesised that if the CSF pressure was lowered enough to reverse the transverse sinus narrowing, this could possibly lead to a resolution of IIH. This proved not to be the case; several patients had the CSF pressure reduced by LP to <10 cm H₂O and the IIH did not resolve unless they developed a low-pressure headache.²¹ There were other patients in whom the CSF pressure was not lowered at LP to <10 cm H₂O and yet when they developed a low-pressure headache the IIH resolved.

In his paper of 1937, Dandy elegantly argued that the only place that the increased pressure could be was in the substance of the brain.¹⁹ There has been controversy regarding the presence of cerebral oedema, but the principal author agrees with Dandy for the following reasons. The pressure inside a closed space reflects the rigidity of the wall, in this case the rigid skull and the contents within that space. The ventricles are slit-like and there is no CSF collection over the hemispheres; thus, it cannot be excess CSF within the ventricular

system or cerebral spinal fluid space. The intermittent “obstruction” of the veins is a physiological obstruction secondary to raised intracranial pressure,³ and the proximal venous sinuses are not dilated. Arterial blood flow is increased due to the raised pressure.²⁸ The logical conclusion is that the only possible place that the increased fluid volume causing increased pressure can be is within the substance of the brain.

If the transverse sinus narrowing is secondary to the raised pressure and yet stenting relieves the IIH, the only logical explanation is that the transverse narrowing must play a role in perpetuating the raised pressure.

Although it is unclear what initially triggers the raised intracranial pressure that results in IIH, we hypothesise that that IIH consists of raised intracranial pressure with secondary compression of the transverse sinuses, thus perpetuating a vicious cycle of raised intracranial pressure. A low-pressure headache is simply where the CSF leak is greater than CSF production, and this could be reproduced by a lumbar drain. The breaking of this vicious cycle could explain the occasional response to LP. If the lumboperitoneal shunt is set at a pressure of 15 cm H₂O, this could possibly relieve symptoms but may not be low enough to break the vicious cycle, and this could be a possible explanation for recurrence of the IIH when the shunt occludes.

We are suggesting that this cycle could possibly be interrupted by prolonged drainage of CSF at a rate greater than that of production of CSF, emulating what is seen in low-pressure headache.

The potential implications of these observations are:

- (1) It may be possible to treat IIH with a lumbar drain anticipating a complete resolution of the problem rather than just alleviation of the symptoms.
- (2) If a patient develops a low-pressure headache in the setting of a lumboperitoneal shunt, the shunt should be removed and not revised, it is possible that the IIH has resolved.
- (3) A potential reason that recurrence occurs when a lumboperitoneal shunt occludes is that the shunt simply drains the CSF at a pressure that decreases symptoms but does

not break the vicious cycle and lead to a resolution of IIH.

- (4) If this approach is effective, then it would eliminate the need for long-term drug therapy, venous sinus stenting, and lumboperitoneal shunts with their associated complications.

A great deal needs to be done to assess the possible utility of a lumbar drain in the management of patients with IIH. The duration of drainage is unclear; the minimal duration of low-pressure headache was 2 days. The successful lumbar drain was continuous drainage over 4 days at the rate of 5–15 mL per hour. The risk of a subdural haematoma occurring in the absence of antiplatelet or anticoagulant therapy is small with a lumbar drain; however, does reducing the pressure lower increase the risk? It is also unclear what percentage of patients will respond to this therapy. The recurrence rate after lumbar drain is also uncertain, and long-term follow-up data will be needed.

Further evaluation requires a multi-centre study with a centralised database. One option would be to drain for 24 hours, occlude the drain for a period of time, for example, 12 hours, and then re-measure the pressure. If it is not elevated, the problem may have resolved; if it remains elevated, then a further period of more prolonged drainage would seem appropriate. If centres were to collaborate and contribute their patients to the database, the question of whether lumbar drainage is a therapeutic option in idiopathic intracranial hypertension would be resolved more rapidly.

One could argue that since immediate and sustained resolution has not been a feature of treated IIH to date, a randomised trial may not be necessary if resolution can be confirmed by measuring the CSF pressure and finding that it has returned to normal.

Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the article.

References

- [1] Friedman DI, Liu GT, Digre KB. Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. *Neurology* 2013;81:1159–1165.

- [2] Whiteley W, Al-Shahi R, Warlow CP, Zeidler M, Lueck CJ. CSF opening pressure: reference interval and the effect of body mass index. *Neurology* 2006;67:1690–1691.
- [3] King JO, Mitchell PJ, Thomson KR, Tress BM. Manometry combined with cervical puncture in idiopathic intracranial hypertension. *Neurology* 2002;58:26–30.
- [4] Arac A, Lee M, Steinberg GK, Marcellus M, Marks MP. Efficacy of endovascular stenting in dural venous sinus stenosis for the treatment of idiopathic intracranial hypertension. *Neurosurg Focus* 2009;27:1–8.
- [5] Donnet A, Dufour H, Levrier O, Metellus P. Exertional headache: a new venous disease. *Cephalalgia* 2008;28:1201–1203.
- [6] Brazis PW. Clinical review: the surgical treatment of idiopathic pseudotumour cerebri (idiopathic intracranial hypertension). *Cephalalgia* 2008;28:1361–1373.
- [7] Higgins JN, Owler BK, Cousins C, Pickard JD. Venous sinus stenting for refractory benign intracranial hypertension. *Lancet* 2002;359:228–230.
- [8] Puffer RC, Mustafa W, Lanzino G. Venous sinus stenting for idiopathic intracranial hypertension: a review of the literature. *J Neurointerv Surg* 2013;5:483–486.
- [9] Stevens SA, Stimpson J, Lakin WD, Thakore NJ, Penar PL. A model for idiopathic intracranial hypertension and associated pathological ICP wave-forms. *IEEE Trans Biomed Eng* 2008; 55 (2 Pt 1): 388–398.
- [10] Wall M, McDermott MP, Kiebertz KD, et al. Effect of acetazolamide on visual function in patients with idiopathic intracranial hypertension and mild visual loss: the idiopathic intracranial hypertension treatment trial. *JAMA* 2014;311:1641–1651.
- [11] De Simone R, Marano E, Fiorillo C, et al. Sudden re-opening of collapsed transverse sinuses and long-standing clinical remission after a single lumbar puncture in a case of idiopathic intracranial hypertension. Pathogenetic implications. *Neurol Sci* 2005;25:342–344.
- [12] Johnston I, Paterson A, Besser M. The treatment of benign intracranial hypertension: a review of 134 cases. *Surg Neurol* 1981;16:218–224.
- [13] Weisberg LA, Chutorian AM. Pseudotumor cerebri of childhood. *Am J Dis Child* 1977;131:1243–1248.
- [14] Vander Ark GD, Kempe LG, Smith DR. Pseudotumor cerebri treated with lumbar-peritoneal shunt. *JAMA* 1971;217:1832–1834.
- [15] Niotakis G, Grigoratos D, Chandler C, Morrison D, Lim M. CSF diversion in refractory idiopathic intracranial hypertension: single-centre experience and review of efficacy. *Childs Nerv Syst* 2013;29:263–267.
- [16] Wang VY, Barbaro NM, Lawton MT, et al. Complications of lumboperitoneal shunts. *Neurosurgery* 2007;60:1045–1048; discussion 1049.
- [17] Gans MS. Idiopathic intracranial hypertension. Medscape. 2016.
- [18] Lee AG, Wall M. Idiopathic intracranial hypertension (pseudotumor cerebri): prognosis and treatment. 2015. UpToDate.
- [19] Dandy WE. Intracranial pressure without brain tumor: diagnosis and treatment. *Ann Surg* 1937;106:492–513.
- [20] Gates PC. Resolution of idiopathic intracranial hypertension after sustained lowering of cerebrospinal fluid pressure. *World J Neurol* 2015;5:47–51.
- [21] Gates PC. Immediate resolution of idiopathic intracranial hypertension with prolonged drainage of CSF at low pressure. Presented at the 65th American Academy of Neurology Meeting; San Diego; March 16–23 2013.
- [22] Sinclair AJ, Kuruvath S, Sen D, Nightingale PG, Burdon MA, Flint G. Is cerebrospinal fluid shunting in idiopathic intracranial hypertension worthwhile? A 10-year review. *Cephalalgia* 2011;31:1627–1633.
- [23] Ahmed RM, Wilkinson M, Parker GD, et al. Transverse sinus stenting for idiopathic intracranial hypertension: a review of 52 patients and of model predictions. *AJNR Am J Neuroradiol* 2011;32:1408–1414.
- [24] Loh Y, Labutta RJ, Urban ES. Idiopathic intracranial hypertension and postlumbar puncture headache. *Headache* 2004;44:170–173.
- [25] McGonigal A, Bone I, Teasdale E. Resolution of transverse sinus stenosis in idiopathic intracranial hypertension after L-P shunt. *Neurology* 2004;62:514–515.
- [26] Pickard JD, Czosnyka Z, Czosnyka M, Owler B, Higgins JN. Coupling of sagittal sinus pressure and cerebrospinal fluid pressure in idiopathic intracranial hypertension—a preliminary report. *Acta Neurochir Suppl* 2008;102:283–285.
- [27] Lee SW, Gates P, Morris P, Whan A, Riddington L. Idiopathic intracranial hypertension; immediate resolution of venous sinus “obstruction” after reducing cerebrospinal fluid pressure to <10cmH(2)O. *J Clin Neurosci* 2009;16:1690–1692.
- [28] Gross CE, Tranmer BI, Adey G, Kohut J. Increased cerebral blood flow in idiopathic pseudotumour cerebri. *Neurol Res* 1990;12:226–230.