ORIGINAL ARTICLE

Cervical puncture and perimedullary cistern shunt placement for idiopathic intracranial hypertension: An alternative to lumbar cistern or cerebral ventricular catheter placement a report of two cases

Jeremiah N. Johnson, Mohamed Samy Elhammady, Christian B. Theodotou, Ramsey Ashour, Mohammad Ali Aziz-Sultan[1]

Department of Neurosurgery, University of Miami, Miller School of Medicine, Miami, FL, [1]Department of Neurosurgery, Brigham and Women's Hospital, Boston, MA, USA

ABSTRACT

Objective: Idiopathic intracranial hypertension (IIH) is a syndrome characterized by increased intracranial pressure (ICP) in the absence of an identifiable cause, and if untreated, can result in permanent vision loss. In symptomatic IIH patients, cerebrospinal fluid (CSF) diversion can lower ICP and protect vision; however, currently used CSF diversion systems are prone to malfunction in this population.

Materials and Methods: In two IIH patients with histories of numerous prior shunt revisions that presented with proximal ventriculoperitoneal shunt malfunction, ICP reduction was achieved by an alternative surgical cerebrospinal fluid (CSF) diversion technique: Fluoroscopically guided, percutaneous placement of a catheter in the premedullary cistern and subsequent connection to the valve and distal shunt system.

Results: Postoperatively, both patients' papilledema resolved, headaches improved, and the shunts were working well at 3-month follow-up. At 1-year follow-up, one patient was well without papilledema or symptom recurrence, and the second patient had the shunt system removed by an outside surgeon.

Conclusion: This technique may hold promise as an alternative shunting strategy in IIH patients with numerous proximal shunt failures or who are poor candidates for ventricular and lumbar shunts.

Key words: Cerebrospinal fluid, cervical vertebrae, cerebrospinal fluid shunts, idiopathic intracranial hypertension, pseudotumor cerebri

Introduction

Idiopathic intracranial hypertension (IIH) also known as pseudotumor cerebri is a condition characterized by increased intracranial pressure (ICP) with normal cerebrospinal fluid (CSF) composition occurring in the absence of large ventricles or an explainable cause. For patients failing lifestyle modification and medical therapy, insertion of a CSF shunt can improve papilledema and halt visual decline. However, both ventriculoperitoneal (VP) and lumboperitoneal (LP) shunts are prone to malfunction in this population necessitating surgical revisions.[1-4] We report two cases of CSF shunting in IIH patients through percutaneous insertion of a premedullary cistern catheter, connection to programmable valve and distal shunt system. To the best of our knowledge, one previous report from 1977 describes a similar CSF diversion strategy.[5]

Access this article online	
Quick Response Code:	Website: www.asianjns.org
	DOI: 10.4103/1793-5482.142735

Address for correspondence:
Dr. Mohammad Ali Aziz-Sultan, Department of Neurosurgery, Brigham and Women's Hospital, 75 Francis Street, PBB-311, Boston 02115, MA, USA. E-mail: sultan@partners.org

Surgical Method

The patient is positioned supine position with the head resting on a horseshoe shaped head holder. The neck is kept in a neutral position. The retro-auricular region is shaved and then the neck, chest and abdomen are prepped and draped in a standard fashion. Two sterilely draped C-arm fluoroscopic machines are positioned to obtain anteroposterior (AP) and lateral images of the C1 and C2 vertebrae [Figure 1a]. It is important to obtain true fluoroscopic AP and lateral views. The skin incision site is identified on lateral fluoroscopy at the C1-C2 interlaminar space [Figure 1b]. A 1 cm skin incision is made at the puncture site and dissected to a depth of 1cm. A 14-gauge ×9-cm Tuohy needle (Integra Neurosciences, Plainsboro, New Jersey) is placed in the incision. Under AP/lateral fluoroscopy, the needle is advanced toward the spinal canal. As the needle nears the dura, it should be advanced under live AP/lateral fluoroscopy to ensure puncture at the appropriate site: On the lateral plane, between the C1 and C2 lamina at the junction of the anterior 2/3 and the posterior 1/3 of the spinal canal [Figure 1c and d]. On AP images, the subarachnoid space is frequently entered when the tip of the Tuohy needle is very near the midline. Once CSF is encountered, the needle bevel is rotated cranially, the stylet is removed, and a hermetic closed-tip lumbar catheter (Integra Neurosciences Plainsboro, New Jersey) with an indwelling guide wire is inserted through the Tuohy needle. The catheter is advanced under fluoroscopic guidance until reaching the caudal clivus [Figure 1c and d]. The Tuohy needle is then removed over the catheter [Figure 1e], and then the internal guide wire is carefully withdrawn. The drain tubing is anchored to the cervical subcutaneous tissue using an encircling plastic clip provided in the lumbar drain kit.

A second horizontally oriented skin incision is then made below the clavicle and a superficial pocket, not deeper than 1 cm, is made for the programmable valve [Figure 1f]. The cervical tubing is then tunneled from the cervical to the chest incision. A step-up connector is used to connect the smaller-than-usual proximal tubing to the valve. A standard distal shunt catheter is then tunneled, connected to the distal end of the valve. If the system is draining properly, the distal catheter is inserted into the peritoneal or pleural cavity in the usual fashion.

Illustrative Cases

Case 1

A young obese female with a history of IIH and numerous VP shunt revisions presented with constant severe headache, papilledema and visual changes. A computed tomography (CT) demonstrated slit ventricles. Appropriate counseling and informed consent were obtained prior to surgical intervention. The patient underwent surgical exploration of the shunt and was found to have unreliable, position-dependent flow through the proximal catheter suggesting intermittent obstruction, possibly due to collapsed slit ventricles. The proximal catheter and valve were removed. Using the technique described in this report, a C1-C2 puncture was performed, and a premedullary cistern drain was placed and connected to a Codman Hakim programmable valve below the clavicle. Due to her complex abdominal surgical history, the distal end of the new valve was connected to the existing functional peritoneal catheter. The patient's headaches improved, and her papilledema resolved post-operatively. At 1-year follow-up, she continued to do well without recurrence of severe headaches or papilledema.

Case 2

A young patient with a history of IIH, schizoaffective disorder and multiple prior shunt revisions, presented with incapacitating headaches, declining vision and papilledma. The patient had a pleural shunt because of numerous previous abdominal surgeries, including a bowel injury. A head CT demonstrated small ventricles and a shunt tap revealed no proximal flow. After appropriate counseling and informed consent had been obtained, surgical exploration revealed a proximal shunt failure. The ventricular catheter was removed, and a premedullary cisternal shunt was placed and connected to a programmable valve, and subsequently connected to the existing pleural distal catheter [Figure 2]. The patient's papilledema and visual symptoms completely resolved, but he experienced contralateral neck pain for several weeks post-operatively.

Post-operative course

The patient did well for 3-months, but returned with low-pressure like, positional headaches and small convexity subdural hygromas on CT, presumably from over-drainage of CSF. His Hakim shunt valve pressure setting was increased from 90 to 110, the symptoms improved, and he was discharged. However, 2 weeks later, he returned to an outside hospital complaining of recurrent headaches. An outside surgeon was unable to adjust the valve, and replaced the Hakim valve with a high-pressure valve. The patient was subsequently found to have a long history of chronic narcotic seeking behavior. One-month after the high-pressure valve was placed, the patient returned to an outside surgeon complaining of continued headaches. Brain CT showed improvement in the subdural hygromas. The surgeon did an LP with an opening pressure of 18, and then explored the C1-C2 shunt valve; however, the proximal catheter was inadvertently pulled out of the subarachnoid space, and thus the system was removed. Ultimately, a new VP shunt was placed, the hygromas eventually resolved, the severity of his headaches improved, but his baseline daily headaches continued.

Discussion

Idiopathic intracranial hypertension, also known as pseudotumor cerebri, is a condition of raised ICP without a known underlying cause. Headache is the most common presenting symptom, and visual deterioration is the most serious consequence.[6,7] Patients often undergo CSF diversion procedures to lower ICP and protect vision.[2,6,8-10] The two most commonly performed CSF shunting procedures are VP and LP shunts; however, both types of shunts have disadvantages in the IIH patient population. For VP shunts, the typical small ventricles of IIH patients can make ventricular catheter placement difficult and predispose the system to

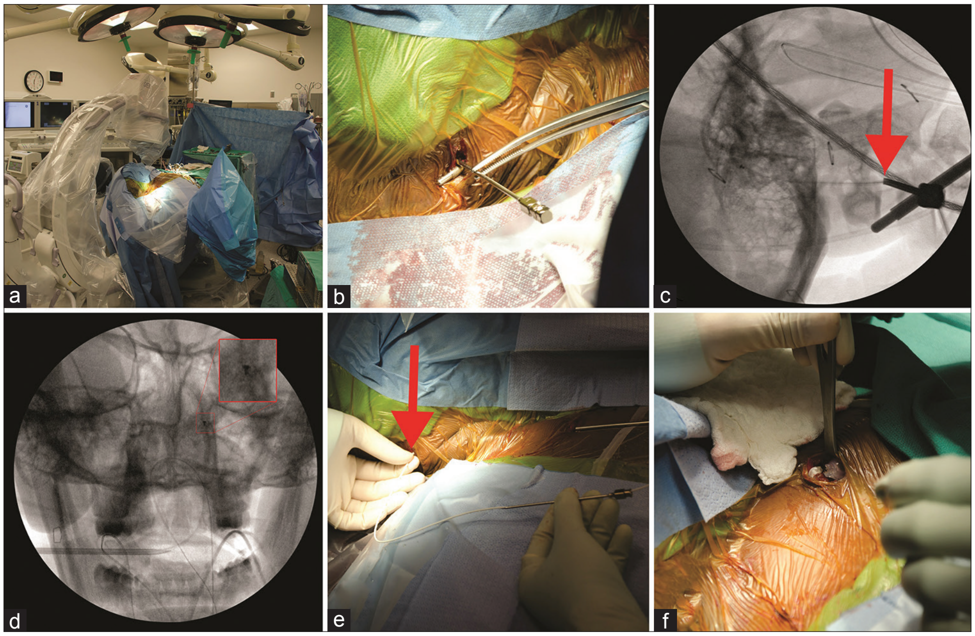

Figure 1: (a) Operative set-up with sterile biplane fluroscopy set around patient. (b) Localization of needle entry point and trajectory with lateral fluroscopy. (c) Intraoperative image of the Tuohy needle (arrow) entering the dorsal 1/3 of the spinal canal. The catheter is aimed rostrally with the tip resting in the inferior 1/3 of the clivus. (d) Tuohy needle in the spinal canal, catheter directed upward and the tip resting in the contralateral perimedullary cistern (magnified). (e) Removal of Tuohy needle post-catheter placement. Shunt passer crosses from subclavicular incision (left) to the post-auricular incision (arrow) for proximal tunneling. (f) Valve placement into subclavicular subcutaneous pocket

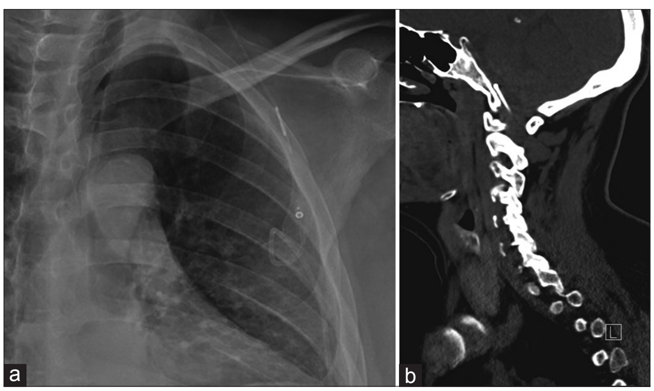

Figure 2: (a) Post-operative sagittal computed tomography reconstruction showing the catheter (arrow) in the generous subarachnoid space of the premedullary cistern. (b) Post-operative anteroposterior chest X-ray showing the placement of the shunt valve in the infraclavicular chest wall (arrow) and distal shunt tubing in the interpleural space (case number 2)

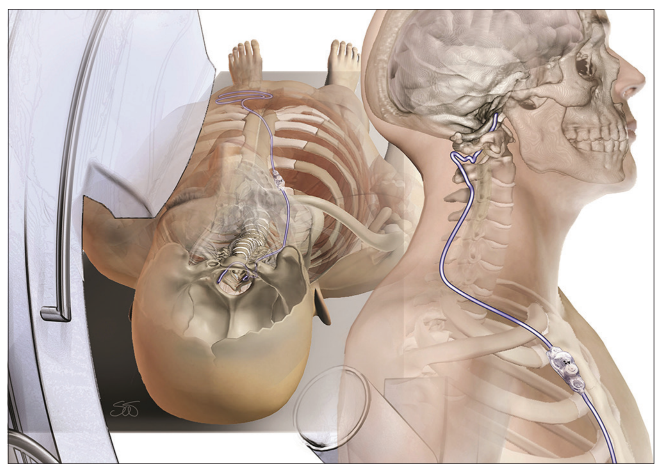

Figure 3: An artist's depiction of the course a perimedullary shunt system takes from the perimedullary subarachnoid space to the peritoneal cavity

malfunction.[11-13] LP shunts are effective at lowering ICP in IIH patients,[4,13,14] but they have high-malfunction rates, up to 85% in one series.[1,3,10,15,16] In addition, LP shunt case series have reported a unique type of complication: Spinal CSF over drainage that may result in a secondary Chiari malformation, thoracic syrinx, and even hindbrain herniation and death.[3,13,17] Given the limitations of VP and LP shunts in the setting of IIH, better CSF diversion strategies would be welcome for this population.

Five authors have previously described open surgical placement of subarachnoid shunt catheters into the cisterna magna or cervical subarachnoid space under direct vision.[18-22] The three largest cisternal shunt placement series reported a high rate of shunt revision, wound infections, and several cranial nerve palsies.[18,19,21] In 1977, Spetzler *et al.* described fluoroscopically guided needle puncture at C1-C2, placement of a shunt catheter in the intracranial cisterns and distal CSF drainage to the cardiac atria in 6 patients.[5] They reported no procedural complications and one distal catheter malfunction 14 months post-procedure. In complicated IIH patients, we propose to explore a similar shunting technique, but with the addition of modern programmable valves [Figure 3].

Although uncommonly employed, CSF drainage of the premedullary cisterns could have several advantages to VP and LP shunts in IIH patients including A small catheter in generous CSF spaces leading to fewer proximal malfunctions than ventricular catheters. Compared with lumbar catheters, there is a smaller hydrostatic pressure column in premedullary cistern when the patient is upright, thus potentially decreasing the propensity for positional over-drainage.[23-26] However, there are also potential disadvantages of premedullary cistern shunts including: The small risk of cervical spinal cord or vertebral artery injury during placement, cranial nerve irritation, the need for intra-operative biplane fluoroscopy, and unproven shunt durability.

The safety of the lateral C1-C2 approach to the cervical subarachnoid space has been established in the cervical myelography literature; yet, there is little data regarding the safety of cervical cisternal catheter placement. A large survey by Robertson and Smith including 55,419 myelograms performed via C1-C2 puncture reported a complication rate of 0.045%.[27] In 2011, Farhat *et al*. reported no complications in a series of 24 cisternal catheters placed via C1-C2 puncture for temporary CSF diversion.[28] Of the >70 C1-C2 temporary CSF drainage catheters placed at our institution from 2006 to 2013, there was one complication: An angiogram negative subarachnoid hemorrhage after catheter removal. Although literature on this topic is limited, properly trained physicians can place cisternal shunts with low-complication rates.[5,28-30]

Our two IIH cisternal shunt patients each had small lateral ventricles, multiple previous abdominal surgeries, numerous proximal VP shunt catheter failures, papilledema, and elevated ICP in the setting of acute shunt malfunction. In both patients, proximal ventricular catheter malfunction was confirmed at surgery, and it was felt prudent to avoid placing another proximal catheter in small lateral ventricles. Similarly, LP shunt placement was undesirable in the face of multiple previous abdominal surgeries. The premedullary cistern shunts placed in these two patients were working at 3-month follow-up, and the papilledema resolved in both patients. Patient number 1 remains at her baseline at 1-year follow-up. However, patient number 2 had a low valve setting at shunt placement, the system was over-draining, another surgeon had difficulty adjusting the shunt valve settings and the C1-C2 shunt was ultimately removed. The authors feel that patient number 2's shunt was likely draining at the time of removal and that the pertinent questions are as follows: (1) Would the over-draining have been halted by simply dialing the Hakim valve to a high setting, thus avoiding revision altogether? (2) Did the system need an anti-siphoning device? (3) Did the pleural location of the distal catheter contribute to siphoning during the repetitive negative intra-thoracic pressure of inspiration, contributing to over draining? The answer to these questions remains unclear. Despite the two good short-term outcomes and one good outcome at 1-year, we reserve this shunting strategy for patients where ventricular and lumbar shunts are difficult, undesirable or contraindicated.

Conclusions

In patients IIH needing CSF diversion that are poor candidates for ventricular or lumbar cistern shunt placement, we propose an alternative shunting strategy: Fluoroscopically guided premedullary cistern shunt placement with distal catheter placement in the peritoneum, or atria. Although this shunting technique is feasible, more experience and longer follow-up is needed to assess the long-term safety, efficacy, and durability.

References

1. Bynke G, Zemack G, Bynke H, Romner B. Ventriculoperitoneal shunting for idiopathic intracranial hypertension. Neurology 2004;63:1314-6.
2. Johnston I, Besser M, Morgan MK. Cerebrospinal fluid diversion in the treatment of benign intracranial hypertension. J Neurosurg 1988;69:195-202.
3. Karabatsou K, Quigley G, Buxton N, Foy P, Mallucci C. Lumboperitoneal shunts: Are the complications acceptable? Acta Neurochir (Wien) 2004;146:1193-7.
4. Wang VY, Barbaro NM, Lawton MT, Pitts L, Kunwar S, Parsa AT, *et al.* Complications of lumboperitoneal shunts. Neurosurgery 2007;60:1045-8; discussion 1049.
5. Spetzler RF, Weinstein PR, Chater N, Wilson CB. Cisternoatrial, ventriculocisternal, and other cisternal shunts simplified. A percutaneous technique. J Neurosurg 1977;47:299-302.
6. Wall M, George D. Idiopathic intracranial hypertension. A prospective study of 50 patients. Brain 1991;114 (Pt 1A):155-80.
7. Radhakrishnan K, Ahlskog JE, Cross SA, Kurland LT, O'Fallon WM. Idiopathic intracranial hypertension (pseudotumor cerebri). Descriptive epidemiology in Rochester, Minn, 1976 to 1990. Arch Neurol 1993;50:78-80.
8. Ball AK, Clarke CE. Idiopathic intracranial hypertension. Lancet Neurol 2006;5:433-42.
9. Banta JT, Farris BK. Pseudotumor cerebri and optic nerve sheath decompression. Ophthalmology 2000;107:1907-12.
10. Burgett RA, Purvin VA, Kawasaki A. Lumboperitoneal shunting for pseudotumor cerebri. Neurology 1997;49:734-9.
11. Brodsky MC, Vaphiades M. Magnetic resonance imaging in pseudotumor cerebri. Ophthalmology 1998;105:1686-93.
12. Abu-Serieh B, Ghassempour K, Duprez T, Raftopoulos C. Stereotactic ventriculoperitoneal shunting for refractory idiopathic intracranial hypertension. Neurosurgery 2007;60:1039-43; discussion 1043.
13. Aoki N. Lumboperitoneal shunt: Clinical applications, complications, and comparison with ventriculoperitoneal shunt. Neurosurgery 1990;26:998-1003; discussion 1003.
14. Choux M, Genitori L, Lang D, Lena G. Shunt implantation: Reducing the incidence of shunt infection. J Neurosurg 1992;77:875-80.
15. Padmanabhan R, Crompton D, Burn D, Birchall D. Acquired Chiari 1 malformation and syringomyelia following lumboperitoneal shunting for pseudotumour cerebri. J Neurol Neurosurg Psychiatry 2005;76:298.
16. Eggenberger ER, Miller NR, Vitale S. Lumboperitoneal shunt for the treatment of pseudotumor cerebri. Neurology 1996;46:1524-30.
17. Chumas PD, Armstrong DC, Drake JM, Kulkarni AV, Hoffman HJ, Humphreys RP, *et al.* Tonsillar herniation: The rule rather than the exception after lumboperitoneal shunting in the pediatric population. J Neurosurg 1993;78:568-73.
18. Rekate HL, Nadkarni T, Wallace D. Severe intracranial hypertension in slit ventricle syndrome managed using a cisterna magna-ventricle-peritoneum shunt. J Neurosurg 2006;104:240-4.

19. Johnston IH, Sheridan MM. CSF shunting from the cisterna magna: A report of 16 cases. Br J Neurosurg 1993;7:39-43.
20. Lundar T, Nornes H. Pseudotumour cerebri-neurosurgical considerations. Acta Neurochir Suppl (Wien) 1990;51:366-8.
21. Lee MC, Yamini B, Frim DM. Pseudotumor cerebri patients with shunts from the cisterna magna: Clinical course and telemetric intracranial pressure data. Neurosurgery 2004;55:1094-9.
22. Beatty RA. Cervical-peritoneal shunt in the treatment of pseudotumor cerebri. Technical cerebri. Technical note. J Neurosurg 1982;57:853-5.
23. Chapman PH, Cosman ER, Arnold MA. The relationship between ventricular fluid pressure and body position in normal subjects and subjects with shunts: A telemetric study. Neurosurgery 1990;26:181-9.
24. Deininger MH, Weyerbrock A. Gravitational valves in supine patients with ventriculo-peritoneal shunts. Acta Neurochir (Wien) 2009;151:705-9; discussion 709.
25. Higashi S, Futami K, Matsuda H, Yamashita J, Hashimoto M, Hasegawa M, *et al.* Effects of head elevation on intracranial hemodynamics in patients with ventriculoperitoneal shunts. J Neurosurg 1994;81:829-36.
26. Nadkarni TD, Rekate HL, Wallace D. Concurrent use of a lumboperitoneal shunt with programmable valve and ventricular access device in the treatment of pseudotumor cerebri: Review of 40 cases. J Neurosurg Pediatr 2008;2:19-24.
27. Robertson HJ, Smith RD. Cervical myelography: Survey of modes of practice and major complications. Radiology 1990;174:79-83.
28. Farhat HI, Elhammady MS, Levi AD, Aziz-Sultan MA. Cervical subarachnoid catheter placement for continuous cerebrospinal fluid drainage: A safe and efficacious alternative to the classic lumbar cistern drain. Neurosurgery 2011;68:52-6; discussion 56.
29. Wahlig JB, Welch WC, Kang JD, Jungreis CA. Cervical intrathecal catheter placement for cerebrospinal fluid drainage: Technical case report. Neurosurgery 1999;44:419-21.
30. Cox TC, Stevens JM, Kendall BE. Vascular anatomy in the suboccipital region and lateral cervical puncture. Br J Radiol 1981;54:572-5.

How to cite this article: Johnson JN, Elhammady MS, Theodotou CB, Ashour R, Aziz-Sultan MA. Cervical puncture and perimedullary cistern shunt placement for idiopathic intracranial hypertension: An alternative to lumbar cistern or cerebral ventricular catheter placement a report of two cases. Asian J Neurosurg 2014;9:153-7.

Source of Support: Nil, **Conflict of Interest:** None declared.

Author Help: Online submission of the manuscripts

Articles can be submitted online from http://www.journalonweb.com. For online submission, the articles should be prepared in two files (first page file and article file). Images should be submitted separately.

1) **First Page File:**
Prepare the title page, covering letter, acknowledgement etc. using a word processor program. All information related to your identity should be included here. Use text/rtf/doc/pdf files. Do not zip the files.
2) **Article File:**
The main text of the article, beginning with the Abstract to References (including tables) should be in this file. Do not include any information (such as acknowledgement, your names in page headers etc.) in this file. Use text/rtf/doc/pdf files. Do not zip the files. Limit the file size to 1 MB. Do not incorporate images in the file. If file size is large, graphs can be submitted separately as images, without their being incorporated in the article file. This will reduce the size of the file.
3) **Images:**
Submit good quality color images. Each image should be less than 4096 kb (4 MB) in size. The size of the image can be reduced by decreasing the actual height and width of the images (keep up to about 6 inches and up to about 1800 x 1200 pixels). JPEG is the most suitable file format. The image quality should be good enough to judge the scientific value of the image. For the purpose of printing, always retain a good quality, high resolution image. This high resolution image should be sent to the editorial office at the time of sending a revised article.
4) **Legends:**
Legends for the figures/images should be included at the end of the article file.