Benefits of using Programmable Lumboperitoneal Shunt in the Management of Idiopathic Intracranial Hypertension

Omar Mohsin Mohammed Ghazal , Ali Kamil Al-Shalchy***

ABSTRACT:

BACKGROUND:

Idiopathic intracranial hypertension (IIH) is a neurological disorder that requires the use of lumboperitoneal (LP) shunt which is the procedure of choice for most surgeons.

AIM OF THE STUDY:

To assess the benefits of using a programmable LP shunt in the treatment of IIH, and how to reach the optimal final performance level with minimal shunt-related complications.

PATIENTS AND METHODS:

A prospective single-center study of 30 patients diagnosed with IIH, treated with adjustable LP shunt in the neurosurgical department at the teaching Martyr Ghazi Al-Hariri Hospital for Specialized Surgery for a period extending from August 30th, 2017 till November 21st, 2018.

RESULTS:

Among the included patients in the study (30 patients), the female:male ratio was 9:1 with an age range of 17-50 years. Most of the cases (40%) were presented with BMI more than 35. The opening pressure was within the range of 280-585 mmH_2O. For the final performance level (P/L), 67% ended up with P/L 1.5 and 27% ended up with P/L 2.0. The outcome recorded as headache improvement 97% and visual improvement 85%. The complications were CSF collection 10%, infection 3%, and the need for shunt revision 7%.

CONCLUSION:

Using the adjustable Medtronic Strata NSC valve has the possibility of avoiding shunt-related complications with better outcomes.

KEYWORDS: Idiopathic intracranial hypertension (IIH), Strata NSC valve, adjustable LP shunt

INTRODUCTION:

Idiopathic intracranial hypertension (pseudotumor cerebri) Idiopathic intracranial hypertension (IIH), sometimes referred to by an old name, "pseudotumor cerebri (PTC)", is a chronic neurological disorder, which can mimic the symptoms of a brain tumor.[1] The condition was first described in 1897.[2]

Idiopathic intracranial hypertension (IIH) is characterized by increased intracranial pressure with no evidence of intracranial mass, hydrocephalus, infection, or hypertensive encephalopathy. Its diagnosis is made by systematically ruling out other disorders. The Modified Dandy Criteria is used to assist physicians in establishing the diagnosis.[3]

Epidemiology

Within the general population, approximately 1 - 2 people in 100,000 suffer from IIH. Among obese women of childbearing age, the incidence is 19 - 21 in 100,000 people.[3] About 2 per 100,000 people are newly affected per year.[4]

Diagnosis

There are four criteria for diagnosing IIH (Modified Dandy Criteria)[5]

1. High intracranial pressure (ICP). Cerebrospinal fluid (CSF) pressure of more than 25 cm H_20,[6] although pressures of more than 40 cm H_20 are not uncommon
2. Normal composition of CSF
3. Papilledema and headaches with no focal findings (In a small percentage papilledema is not always present)
4. Normal CT or MRI with the exceptions of slit ventricles, partial or total empty sella, or alterations in cerebral venous sinus visualized by MRI venography or cerebral angiography

*Tikrit Emergency Hospital / Saladdin, Iraq.

**Ghazi Al-Hariri Teaching Hospital for Specialized Surgery / Baghdad Medical City/ Baghdad, Iraq.

TREATMENT

Surgical Procedures

Surgery is employed when patients initially present with severe optic neuropathy or when other forms of treatment have failed to prevent visual loss. It is not recommended for the treatment of headaches alone.[7]

The surgical therapeutic modalities includes optic nerve sheath fenestration (ONSF), venous sinus stenting, lumboperitoneal (LP) and ventriculoperitoneal (VP) shunting, with the latter two being the most commonly used for the idiopathic intracranial hypertension (IIH) management.[8,9]

Optic nerve sheath fenestration despite relieving papilledema, but it often fails to relieve the headaches of IIH. Moreover, 32% of the patients in one series, vision continued to deteriorate after optic nerve sheath fenestration.[10] It is usually not recommended to repeat ONSF in failed cases owing to the low likelihood of improvement and the additional risks for higher morbidity.[10,11]

Placement of the VP shunt catheter for patients with IIH is challenging, due to the small ventricles, with an increased risk of intracranial injuries (including hemorrhage, infection or seizure) and/or shunt malfunctions. Furthermore, non-programmable VP shunts are typically associated with over- or underdrainage of CSF. These complications were reduced using programmable VP shunts with neuro-navigation.[12,13,14,15] But, recent studies have shown failure of VP shunts in managing 14% of the patients.[16]

Alternative method for the treatment of IIH patients is by using standard LP shunting which avoids the risk of intracranial injuries. Yet, another concern with the use of LP shunts without valves is overdrainage.[17] Laurent Riffaud et al. observed an increased risk of developing symptomatic acquired Chiari malformation and syringomyelia if a valveless LP shunt is used. So, They suggested LP shunt with an adjustable valve to prevent such complications.[18] Besides, high failure rates, which can reach up to 11%, are associated with the use of the standard LP shunts.[16]

The adjustable Strata valve from Medtronic appeared to have the same advantages as the programmable VP shunt without the intracranial complications. At the same time, it is similar to the standard LP shunt with the same advantages in addition to decreasing the risk of developing the acquired Chiari malformation.[12,19]

AIM OF THE STUDY:

To assess the benefits of using the Strata NSC adjustable lumboperitoneal shunt in the treatment of idiopathic intracranial hypertension, and how to reach the optimal final performance level with minimal shunt-related complications.

PATIENTS AND METHODS:

This is a prospective single-center study of 30 patients with idiopathic intracranial hypertension treated in the neurosurgical department at the Martyr Ghazi Al-Hariri teaching Hospital for Specialized Surgery for a period extending from August 30th, 2017 till November 21st, 2018.

Inclusion criteria:

- Confirmed diagnosis of IIH by the Modified Dandy Criteria.
- Failure of medical therapy and/or severe optic neuropathy.
- No contraindication for LP shunt placement (e.g., no congenital or acquired spinal deformity at the implantation site, nor infection in any areas in which the various components of the shunt system will be implanted like arachnoiditis from previous tapping).
- OR, Complicated previous conventional valveless LP shunt (due to shunt obstruction, underdrainage, or overdrainage).

Exclusion criteria:

- Secondary pseudotumor cerebri (in which treating the underlying condition usually produces improvement).
- Patients who refused shunting surgery, or refused to participate in the study or follow-up visits.

Surgical technique

The valve is adjusted to the proper initial performance level (P/L) setting before implantation. The whole length of the peritoneal catheter (120 cm) is used without cutting the end as it may affect the pressure setting.

Beginning with a small low back midline incision (about 2.5 cm) in the lumbar region between L3-L4, cutting the skin and subcutaneous tissue down to the lumbar fascia. A 14-gauge Tuohy needle is then used to insert the tip of the lumbar catheter (after being flushed with normal saline) into the subarachnoid space for about 10 cm length cephalad (in addition to the length of the needle). Using fixation tab as close as possible to the fascia entry point and sutured to the fascia by a non-absorbable suture material. Then, a left abdominal paramedian incision (about 4 cm) to expose the peritoneum. Lastly, a left flank incision (about 3 cm) is made over a rib at the mid-axillary line cutting the skin

and undermining the edge of the skin for the valve placement.

Postoperative follow-up

The first follow-up visit a week later and a second follow-up visit 3 weeks postoperatively are advised. During these visits, the following parameters were assessed & recorded:

- Patient's presenting symptoms (improved or not).
- Complications (if any).
- Magnetic field exposure.
- P/L checked (adjusted during follow-up visits according to status of the patient).

The patients were also instructed to avoid magnetic field exposure to the valve. Otherwise, an additional visit is needed for the valve re-adjustment to the last recorded performance level.

All possible complications were explained to the patients. In the scenario of developing any complication during the postoperative period, the patient was constructed that further visits are recommended.

RESULTS:

The age of the patients was in the range of 17-50 years with mean value of 32 years as detailed in "figure (1)". Twenty-seven (90%) of our patients were female, while 3 (10%) of them were male with female:male ratio of 9:1.

The body mass index (BMI) for this study was within the range of 23.9-44.6 with the mean value of 33.3. As for the male group of patients, they were normal or overweight. While, overweight and obesity were noticed in all the female gender. "figure 2"

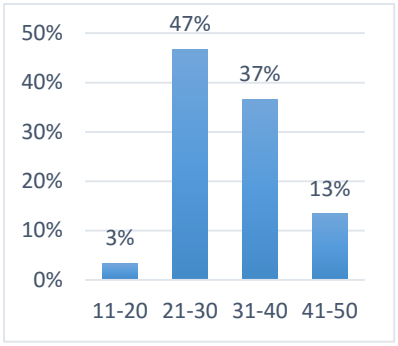

Figure 1: Age distribution of the patients

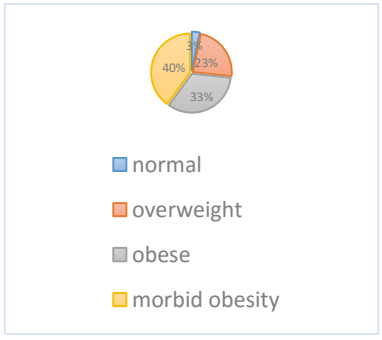

Figure 2: BMI of the patients

Valve pressure setting:

The valve pressure was set pre-operatively as the initial performance level (P/L). One week later, during the 1st (follow-up visit), the valve pressure setting was adjusted according to the patient's condition as the 2nd P/L. Further two weeks later (during the 2nd follow-up visit), a third adjustment of the valve pressure was applied (when needed). Some patients required more than these two adjustments, and the final P/L was recorded. In other instances, the patient exposed to magnetic field and the final P/L was restores by re-adjusting the valve to the final recorded setting "table (1)".

Table 1: The performance level (P/L) for each patient.

Case no.	Age	Gender	BMI	Opening pressure (mmH_2O)	Initial P/L	Second P/L	Final P/L
1	47	Female	23.9	380	2.5	2	1.5
2	34	Female	26.5	360	2.5	2	1.5
3	25	Female	26.9	295	2.5	2	1.5
4	23	Female	27.9	440	2.5	2	1.5
5	50	Female	27.9	400	2.5	2	1.5^+
6	21	Female	29.4	350	2.5	2	1.5^+
7	45	Female	29.7	350	2.5	2	
8	36	Female	29.7	365	2.5	2	
9	37	Male	30.4	335	2.5	1.5	2.5^+
10	27	Female	30.7	400	2.5	1.5	2^+
11*	40	Female	30.8	540	2.5	1.5	2
12	32	Female	31.1	300	2.5	1.5	2
13	27	Female	31.2	390	2.5	1.5	2
14	17	Female	31.2	420	2.5	1.5	
15	25	Male	32.9	320	2.5	1.5	
16	28	Female	33.1	330	2.5	1.5	
17	27	Female	33.2	375	2.5	1.5	
18*	29	Female	34.8	320	2.5	1.5	
19	38	Female	35.1	285	2.5	1.5	
20	46	Female	35.4	585	2.5	1.5	
21~	21	Female	36.0	410	2.5	1.5	
22*	32	Female	36.1	395	2	1.5	1
23	31	Female	36.1	400	2	1.5	
24	24	Male	36.3	280	2	1.5	
25	38	Female	36.7	330	2	1.5	
26~	26	Female	37.3	420	2		
27	37	Female	40.4	310	1.5	2	
28	35	Female	40.8	420	1.5		
29~	28	Female	41.8	340	1.5		
30~	26	Female	44.6	390	1.5		

* Patient exposed to magnetic field

$^+$ more than 2 adjustments were needed (after the initial setting)

~ patients who were already on a previous valveless LP shunt

Table 2: Number of the cases according to the final P/L.

Final performance level	Pressure range	No. of cases	Percentage
0.5	0-30 mmH_2O	0	0%
1	10-60 mmH_2O	1	3%
1.5	55-115 mmH_2O	20	67%
2	105-170 mmH_2O	8	27%
2.5	155-255 mmH_2O	1	3%

Table 3: Clinical improvement.

Gender	Headache improvement (%)	Visual improvement (%)
Male	2 (67%)	2 (100%)
Female	27 (100%)	21 (84%)
Total	29 (97%)	23 (85%)

Table 4: Complications.

Complication	No. of cases	Percentage
Overshunting	6	20%
Undershunting	6	20%
CSF collection	3	10%
Infection	1	3.33%
Revision	2	6.67%

DISCUSSION:

A total of 30 patients were included in our study, in which 27 of them were of female gender and the remaining 3 of them were male with female:male ratio was 9:1. Toma et al found the same female:male ratio of 9:1 with 18 female patients and only 2 males.[20] While, Yadav et al included 24 patients in their study, in which 22 of them were female with a female:male ratio of 11:1.[21] This goes with the general concept of that there is female predominance concerning the idiopathic intracranial hypertension.

The age of the patients was in the range of 17-50 years with mean value of 32 years. Approximately, the same age range which was from 17-58 years (mean value of 39 years) was reported by Yadav et al in their series.[21] The only difference was that one of the patients was of 58 years old. While, Alkherayf et al reported an age range of 23-46 years with a mean value of 33 years.[22] But, their study included only 7 patients. In our study, we also found that the majority of the patients were in the age between 20-40 years (84%).

The range of the body mass index (BMI) was 23.9-44.6 with an average of 33.3. Data collected by Jusué-Torres et al was BMI average of 36 (range 30-41).[23] We found that the incidence of the IIH is increasing with weight (73% of cases presented with BMI value of greater than 30).

Choosing the initial performance level was tricky. We tried different ways to reach the optimal results, but no relationship has been established between the CSF opening pressure and choosing an appropriate initial performance level and its effect on establishing the final performance level. Another problem is setting the final performance level depending on the patient preference and according to their complaints of over- or underdrainage, but dealing with some patients was difficult enough to explain the difference between high-pressure and low-pressure headache, that we even had to measure the ICP from the valve itself using an 25-gauge needle while pressure was applied on the occluder. To overcome this issue, we recommend to start with 2.5 performance level as the initial setting (as we did in 70% of the cases in the study), so as any postoperative headache would be considered as underdrainage and described as the headache is relieved in severity but still there. Most of the cases ended up with the final setting of 1.5 performance level (67%). While, the final setting of 2.0 performance level was in 27% of cases with the 2.5 & 1 performance levels were in 3% of cases for each of them. Among the 30 patients who were included in the study, adjustments of the valve pressure setting were needed in 12 patients at the 3rd follow-up visit. Some of them required more than one adjustment due to magnetic field exposure or the difficulty in addressing the optimal final setting. Alkherayf et al, in their limited study to 7 female patients, recommended to start with the 1.5 performance level as the initial pressure setting. But, four patients (57%) required adjustment at the 4th postoperative week. Adjustment to the 2.0 performance level was required in 2 of the cases (28.5%), and to the 0.5 & 1.0 performance level was required in one case (14.25%) for each. While the remainder 3 cases (43%) remained at the same 1.5 performance level.[22] Unfortunately, this was the only available study to compare with regarding the performance levels.

Headache improvement was observed in 97% of cases and visual improvement was recorded in 85% of cases. For the valveless LP shunts, the improvements for headache were 93.8%, 91.7%, 86.4% & the visual 76.9%, 55%, 72.7% in the Abd-Ali et al, Yadav et al, El-Saadany et al studies respectively.[21,24,25] Using a programmable valve LP shunt, the results were 100% & 86.7% regarding the visual improvement in the Alkherayf et al & Toma et al studies respectively. Meanwhile, the headache improvement was observed in 100% of cases in both latter mentioned studies (Toma el al also observed late postoperative headache in 4 patients, in which extensive workup including ICP monitoring & neurology opinion to exclude shunt related headache).[20,22] These results implies the concept that utilizing a programmable LP shunt leads to better outcome.

The need of a second adjustment for the valve, during the 2nd follow-up visit 3 weeks postoperatively or later on, is considered either overshunting or undershunting (40%) and were treated by adjusting the device. CSF collection was encountered in 3 cases (10%), whereas 2 of them were treated by lowering the pressure setting while the other case required surgical shunt revision. Wound infection over the valve incision was developed in one case (3%) and followed by meningitis (fever, signs of meningismus, nausea & vomiting, and worsening of the headache). Measuring the ICP through the valve using a 25-gauge needle (while pressing the occluder), the ICP measurement was within normal range (145 mmH_2O), and the obtained CSF through this procedure, that was turbid in

appearance, was sent for analysis and culture. Thus, confirming diagnosis by the results of the analysis and culture. Shunt removal was the first step of management followed by antibiotics according to the culture and sensitivity results. Shunt revision was needed in 2 cases (6%), first case due to CSF collection, previously described, and a disruption in catheter was noted. The later, due to distal shunt obstruction, in which a pseudocyst around the peritoneal catheter was found to be the cause. Finally, we did not encounter other complications like: subdural collection, tonsillar herniation or acquired Chiari I malformation, seizure, pneumoencephalus, catheter malposition.

Yadav et al, using valveless shunt system in 24 cases for 11 years, found CSF leak in 4% of cases, overdrainage 62.5% & shunt obstruction 8% (which was difficult to evaluate using intrathecal dye).[21] El-Saadany et al, using also valveless shunt in 22 cases for 3 years, found infection in 9%, overdrainage 13.6% & shunt obstruction 27% (most of them due to migration of distal catheter).[25]

Jusué-Torres et al, using horizontal-vertical (H-V) lumbar valve in 26 patients for 20 years, found that 69% of the cases developed at least one complication (infection 8%, CSF leak 15%, overdrainage 35%, shunt obstruction 19%). Shunt revision was needed for 58% of cases (mostly due to overdrainage and tonsillar herniation), in which multiple revisions was required in 39%, conversion to other type of shunt (e.g., VP, VA, lumbopleural or lumboatrial) in 42% and changing valve type 8%.[23]

Wang et al, compared 46 patients treated with Integra H-V valve versus 21 patients treated with conventional valveless Silastic LP shunt, found that overdrainage was 8.6% in the first versus 33% in the latter.[22]

Toma et al, using strata NSC programmable valve in 20 cases for 2 years, found distal end obstruction in 15%, proximal end obstruction 5%, CSF leak 5% (due to damage to the valve perioperatively). Shunt revision was done for 35% of cases (for the previous complications in addition to 10% of cases due to either valve malfunction "inability to adjust the valve", or suspected shunt disconnection by radiograph in which exploration revealed intact shunt system).[20]

Comparing with other studies, the programmable LP shunt when used to treat IIH has the same postoperative complication rates as the valveless shunt system regarding the infection and the CSF leak. But, the latter may be managed by lower the performance level of the valve. Overdrainage could be easily reversed by readjusting the valve. In term of the need for shunt revision, many causes could be prevented or reduced like migration of the distal or proximal end (as those 2 ends are connected to a valve), distal end obstruction (as the valve permits using a different tube width for the peritoneal end).

CONCLUSION:

- Using the adjustable Medtronic Strata NSC valve as LP shunt in treating idiopathic intracranial hypertension is a good and promising modality of treatment, with the possibility of avoiding shunt-related complications (underdrainage and overdrainage) due to the ability to adjust the performance level of the valve according to the patient's condition and need.
- It has been noticed that there is no relationship between the opening ICP and the final performance level used.
- Better outcome was noted when compared to other studies which used valveless LP shunt or even H-V LP valve.
- It is also found to have less possibility of shunt obstruction due to the use of the long wide-bore small lumen peritoneal catheter provided in the set which makes it more resistant to kinking.
- It is easier to evaluate the complications using a 25-gauge needle through the valve for intracranial pressure measurement, CSF sampling, dye injection to check the shunt patency in respect to the valveless LP system.

RECOMMENDATIONS

- Encourage the use of a programmable valve LP shunt to treat idiopathic intracranial hypertension.
- Further studies in other centers are required to outweigh the benefits versus the pitfalls of using the programmable valve in such condition.

REFERENCES:

1. Friedman DI. The pseudotumor cerebri syndrome. Neurol Clin. 2014; 32:363-96.
2. Wall M. Update on Idiopathic Intracranial Hypertension. Neurologic Clinics. 2017; 35:46.
3. Greenberg MS. Handbook of Neurosurgery. 8th ed. New York: Thieme Medical Publishers. 2016; 766.
4. Wakerley BR, Tan MH, Ting EY. Idiopathic intracranial hypertension. Cephalalgia: An International Journal of Headache. 2015: 35:248–61.
5. Gary YS, Stephanie KM. Benign Intracranial Hypertension: A Diagnostic Dilemma. Case Reports in Otolaryngology. 2012; 2012:1-8.
6. Lee SC, Lueck CJ. Cerebrospinal fluid pressure in adults. J Neuroophthalmol. 2014; 34:278-83.
7. Winn HR. Youmans & Winn Neurological Surgery. 7th ed. Elsevier. 2017;164:1360-1.
8. Brazis PW. Clinical review: the surgical treatment of idiopathic pseudotumour cerebri (idiopathic intracranial hypertension). Cephalalgia. 2008; 28:1361-73.
9. Binder DK, Horton JC, Lawton MT, et al. Idiopathic intracranial hypertension. Neurosurgery. 2004;54:538-52.
10. Giridharan N, Patel SK, Ojugbeli A, et al. Understanding the complex pathophysiology of idiopathic intracranial hypertension and the evolving role of venous sinus stenting: a comprehensive review of the literature. Neurosurgical Focus FOC. 2021; 45:e10.
11. Plotnik JL, Kosmorsky GS. Operative complications of optic nerve sheath decompression. Ophthalmology. 1993; 100:683–90.
12. Wang VY, Barbaro NM, Lawton MT, et al. Complications of lumboperitoneal shunts. Neurosurgery. 2007;60:1045-49.
13. 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.
14. Chang CC, Kuwana N, Ito S. Management of patients with normal pressure hydrocephalus by using lumboperitoneal shunt system with the Codman Hakim programmable valve. Neurosurg Focus. 1999;7:e8.
15. Aoki N. Lumboperitoneal shunt: clinical applications, complications, and comparison with ventriculoperitoneal shunt. Neurosurgery. 1990; 26:998-1004.
16. Abubaker K, Ali Z, Raza K, et al. Idiopathic intracranial hypertension: lumboperitoneal shunts versus ventriculoperitoneal shunts - case series and literature review. Br J Neurosurg. 2011; 25:94-9.
17. El-Saadany WF, Farhoud A, Zidan I. Lumboperitoneal shunt for idiopathic intracranial hypertension: patients' selection and outcome. Neurosurgical review. 2012; 35:239-44.
18. Riffaud L, Moughty C, Henaux PL, et al. Acquired Chiari I malformation and syringomyelia after valveless lumboperitoneal shunt in Infancy. Pediatr Neurosurg. 2008; 44:229-33.
19. McGirt MJ, Woodworth G, Thomas G, et al. Cerebrospinal fluid shunt placement for pseudotumor cerebri associated intractable headache: predictors of treatment response and an analysis of long-term outcomes. J Neurosurg. 2004; 101:627-32.
20. Toma AK, Dherijha M, Kitchen ND, et al. Use of lumboperitoneal shunts with the Strata NSC valve: a single-center experience. Journal of neurosurgery. 2010; 113:1304-8.
21. Yadav YR, ParIhar V, Agarwa M, et al. Lumbar peritoneal shunt in idiopathic intracranial hypertension. Turkish neurosurgery. 2012;22:21-6.
22. Alkherayf F, Al-Shaar HA, Awad M. Management of idiopathic intracranial hypertension with a programmable lumboperitoneal shunt: Early experience. Clinical neurology and neurosurgery. 2015;136:5-9.
23. Jusué-Torres I, Hoffberger JB, Rigamonti D. Complications of lumboperitoneal shunts for idiopathic intracranial hypertension. Cureus. 2014; 6:e188.
24. Abd-Ali BS, Abdulnabi AY, Radhee HK. Lumboperitoneal Shunt Clinical Applications & Complications. Mustansiriya Medical Journal. 2013; 12:19-22.

25. El-Saadany WF, Farhoud A, Zidan I. Lumboperitoneal shunt for idiopathic intracranial hypertension: patients' selection and outcome. Neurosurgical review. 2012; 35:239-44.