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Cerebrospinal Fluid Diversion Procedures for Treatment of Idiopathic Intracranial Hypertension: Single Center Experience

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Abstract

Idiopathic intracranial hypertension (IIH) is a disorder characterized by increased intracranial pressure without any identifiable etiology with normal brain imaging and normal cerebrospinal fluid (CSF) content. CSF diversion procedures are commonly used for treatment if medical treatment failed. The aim of this study is to report our experience in treatment of IIH with lumboperitoneal (LP) and stereotactic guided ventriculoperitoneal (VP) shunts. The clinical data of 43 consecutive patients with IIH refractory to medical treatment and underwent CSF diversion procedures between 2009 and 2014 were analyzed. 29 patients underwent LP shunts and the remaining 14 patients underwent stereotactic guided Ventricular shunts. All patients underwent clinical, imaging and CSF manometry evaluation. 38 (88.4%) patients were female and the remaining 5 (11.6%) patients were male. The mean age was 27.2 years. The opening pressure was above 300 mm H_2O in 26 (69.8%) patients. Headache (100%) and blurring and/or diminution of vision (81.4%) were the commonest clinical presentation. 36 (83.7%) patients reported recovery of their headache and 30 (69.7%) patients showed complete resolution of papilledema. The clinical outcome between both procedures was not significant. The incidence of perioperative complications (20.7% vs. 0%) and shunt revisions (27.6% vs. 7.1%) were higher in patients with LP shunt than patients with stereotactic Ventricular shunts. The results of this study demonstrate that both LP and Ventricular shunts are valid diversion procedures for treatment of IIH. Stereotactic guided Ventricular shunt has lower incidence of complications and revisions and seems to be safe, effective and feasible alternative procedure for treatment of IIH.

Keywords

Idiopathic Intracranial Hypertension, Lumboperitoneal Shunt,

Ventriculoperitoneal Shunt, CSF, Complications

1. Introduction

Idiopathic intracranial hypertension (IIH) also known as pseudotumor cerebri, is a disorder of increased intracranial pressure with normal neuroimaging and CSF composition of unknown cause [1]. It usually affects obese women of childbearing age but it may be seen in patients of any age, gender and without obesity [2]. The annual incidence of IIH is approximately 1 per 100,000 of the general population [1]. The incidence increases to 13 - 14.8/100,000 for women aged 20 - 44 years with 10% or more above the ideal body weight [3]. The exact mechanism of IIH remains to be evaluated. Proposed mechanisms of pathogenesis revolve around CSF physiology such as increased cerebral blood volume, excessive CSF production and/or compromised resorption [4]. Recently, vascular causes have been proposed to contribute to the underlying pathology, with emphasis on arterial inflow and venous outflow anomalies [4]. Risk factors for IIH rather than obesity are the use of exogenous substances such as lithium, abnormal metabolism of vitamin A, tetracyclines and corticosteroid intake or more likely following steroid withdrawal [1]. Diagnosis of IIH is largely dependent on modified Dandy criteria, which include raised lumbar puncture opening pressure in the absence of an intracranial lesion or ventricular dilatation, normal CSF composition, normal neurological examination except for papilledema and occasional VI nerve palsy and normal level of consciousness [5]. IIH does not currently have a well established management algorithm due to absence of clear pathophysiology [1]. Treatment options include medical and surgical modalities. Patients often received medical treatment and surgery is reserved for patients with severe and acute visual deterioration or in whom intractable headache and visual deficits are refractory to medical treatment [2]. Surgical treatment of IIH includes CSF diversion procedures, optic nerve sheath fenestration, and the newly introduced transverse sinus stenting [2] [6] [7]. CSF diversion procedures are the most commonly used owing to relative technical simplicity, minimal invasiveness and easily managed perioperative complications [2]. Both Lumboperitoneal shunting and ventriculoperitoneal shunting with or without stereotactic or navigation guidance are used with variable efficacy and shunt failure [8]. This study describes the treatment of IIH in our center with both lumboperitoneal and stereotactic guided ventricular shunting as regard the efficacy, perioperative complications and the need for shunt revisions.

2. Materials and Methods

This study included 43 consecutive patients with IIH treated with CSF diversion procedures in the Department of Neurosurgery, Zagazig University at the period between 2009 and 2015. All patients underwent a comprehensive clinical and radiological evaluation that include through medical, neurological and ophthal-

mological examination. Clinical and neurological examination evaluate the analysis of symptoms of increased intracranial pressure through the same questionnaire for headache, vomiting, visual impairment, tinnitus and associated medical co-morbidity such as obesity and contraceptive pills intake. Ophthalmological examination included visual acuity using best corrected Snellen, bilateral fundus examinations and visual field using automated Humphrey perimetry (Zeis, Jena, Germany). All patients received an initial medical treatment in the form of acetazolamide and/or corticosteroids and at least lumbar puncture once with reporting of the CSF opening pressure. The CSF pressure was measured with the patients in the lateral decubitus position and classified into mild, moderate, severe and fulminant according to El-Saadany *et al*. [2] study and as shown in Table 1. CSF chemistry was analyzed. Diagnostic modalities included Computed tomography of the brain, MRI and MRV for the patients as shown in Figures 1-4. The surgical indications in our patients included patients with acute and severe visual deterioration and patients with persistent intractable headache despite adequate medical treatment and repeated lumbar CSF drainage. The surgical treatment involved placement of a lumbopritoneal shunt in 29 patients and ventriculoperitoneal shunt in 14 patients. The commercially available (Codman & Shurtleff, Inc. Raynham, MA, USA) devices were used for the patients. All patients were operated in the lateral decubitus position for lumpoperitoneal shunt and the stereotactic guided Ventricular shunt was performed in supine position through a frontal Burr hole. In patients with ventricular shunt; 9 patients received ventriculoperitoneal shunt and the remaining 5 patients had ventriculojugular shunt.

Patients were evaluated postoperatively clinically for improvement of headache and visual complains. Headache assessment before and after treatment was conducted using visual analogue scale which measures the intensity of pain by making a 10 point scale describing the intensity of headache (0 = no pain and 10 = worst pain). Headache analysis before and after surgery included onset, duration, site, character, frequency and also aggravating and relieving factors. Fundus examination was evaluated after surgery for improvement of papilledema, and the visual field for improvement of field defects. All patients completed at least one year follow up after surgery.

3. Results

The study group included 38 (88.4%) and 5 (11.6%) male patients. Their age ranged between 18 and 47 years with mean age of 27.2 years. The peak incidence was at the third decade of life in 19 (44.2%) of the patients. Headache was reported in all patients followed by visual obscuration in 35 (81.4%) of the patients as shown in Table 1. Fundus examination showed advanced papilledema in 29 (76.4%) and optic atrophy in 8 (18.6%) patients. Obesity was reported in 25 (58.1%) of the patients and contraceptive pills in 11 (25.6%) of the patients. The study included two sisters of IIH treated at our department with LP shunt. Visual field examination preoperatively revealed scotoma in 21 (48.8%) and concentric

Table 1. Demographic and clinical characteristics of the patients.

Age
Range: 20 - 48-year
Mean: 27.2-year
Sex
Female: 38 (88.4%)
Male: 5 (11.6%)
Associated co-morbidity
Obesity: 25 (58.1%)
Contraceptive Pills: 11 (25.6%)
Anemia: 5 (11.6%)
DM: 6 (14%)
Hypertension: 8 (18.6%)
Clinical Presentation
Headache 43 (100%)
Vomiting 9 (21%)
Blurring/diminution of vision 35 (81.4%)
Diplopia 11 (25.6%)
Tinnitus 6 (14%)
Fundus examination
Early papilledema 6 (14%)
Advanced papilledema 29 (67.4%)
Optic atrophy 8 (18.6%)
Visual field (Preoperative)
Normal 5 (11.6%)
Enlarged blind spot 4 (9.3%)
Scotoma 21 (48.8%)
Concentric concentration 13 (30.2%)
Intracranial pressure
Mild (250 < 300) 13 (30.2%)
Moderate (300 < 400) 14 (32.5)
Severe (400 < 500) 3 (6.9%)
Fulminant (>500) 9 (20.9%)
CSF diversion Procedure
Lumboperitoneal 29 (67.4%)
Ventricular shunts 14 (32.6%)
Imaging Finding
Slit and narrow ventricle 32 (74.4%)
Normal ventricle 11 (25.6%)

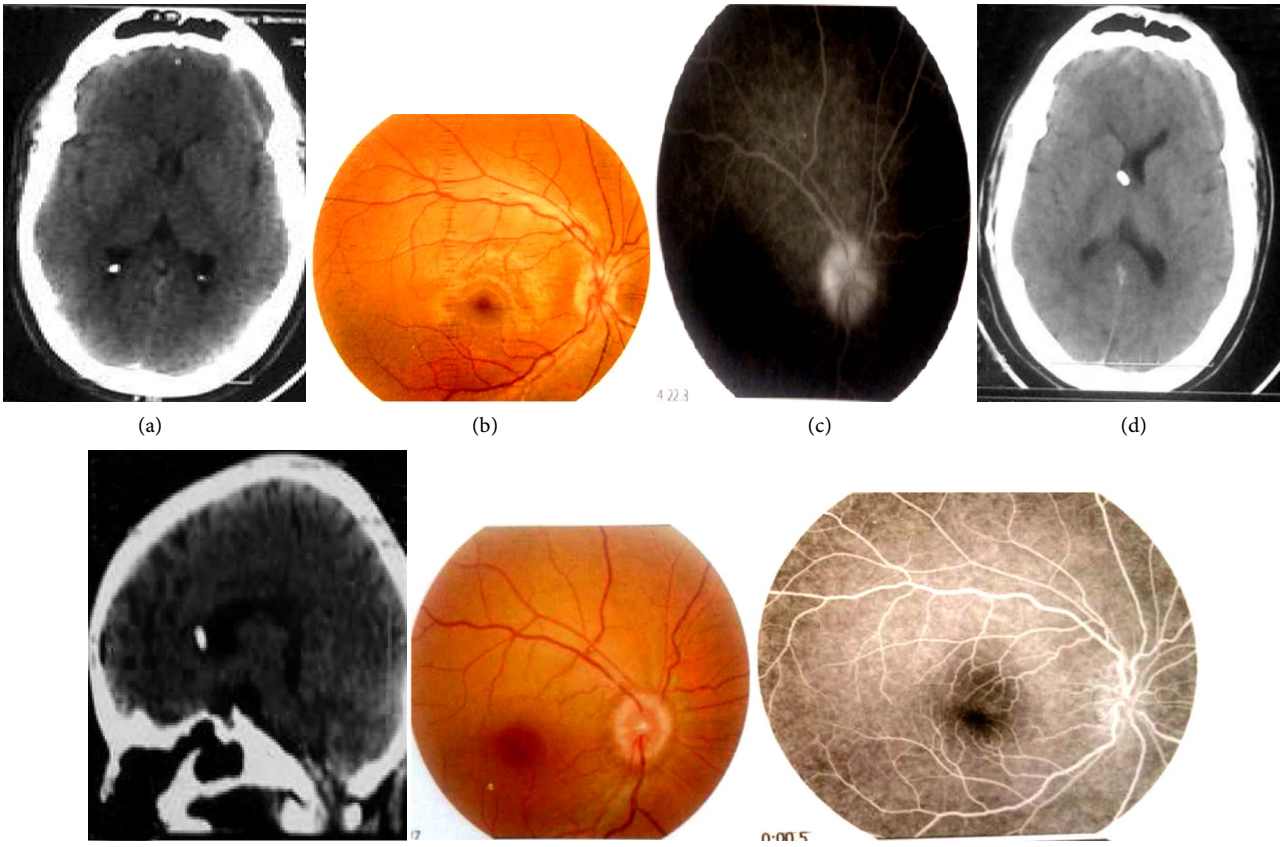

Figure 1. Female patient treated with ventriculojugular shunt (a) Axial CT brain showed slit ventricle, (b) Preoperative fundus examination showed mild venous engorgement of the optic head, blurred margin, obliteration of the cup, (c) Preoperative fluorescein angiography showed hyperflorescence overlying the optic head extending to the juxta papillary retina, (d) Axial CT brain showed the accurate ventricular insertion of the proximal catheter, (e) Sagittal CT brain showed the ventricular catheter, (f) Postoperative fundus examination showed resolution of papilledema, (g) Postoperative fluorescein angiogrpahy showed no leakage.

field contraction in 13 (30.2%) of our patients. Among all patients, the opening CSF pressure for the first lumbar tap was > 300 mm H_2O in 26 (69.8%) patients. 6th nerve palsy was reported in 5 patients from increased intracranial pressure. Computed tomography and MRI of the brain had been showed normal size ventricle in 11 (25.6%) of the patients and narrow or slit ventricle in the remaining 32 (74.4%) patients.

The clinical and functional outcome in the early postoperative period during hospital stay included improvement of headache in 36 (83.7%) of the patients. 3 patients had CSF hypotension headache which improved with fluids and corticosteroids for few days, and the other 4 patients reported some improvement in severity and frequency of headache. Fundus examination on late follow up revealed resolution or improvement of papilledema in 30 (69.7%) of the patients, no improvement in the severity of papilledema in 5 (11.6%) of the patients. All the 8 patients with optic atrophy remained the same.

Periprocedural Complications

All LP shunts were placed with the lumbar needle successfully except 3 patients

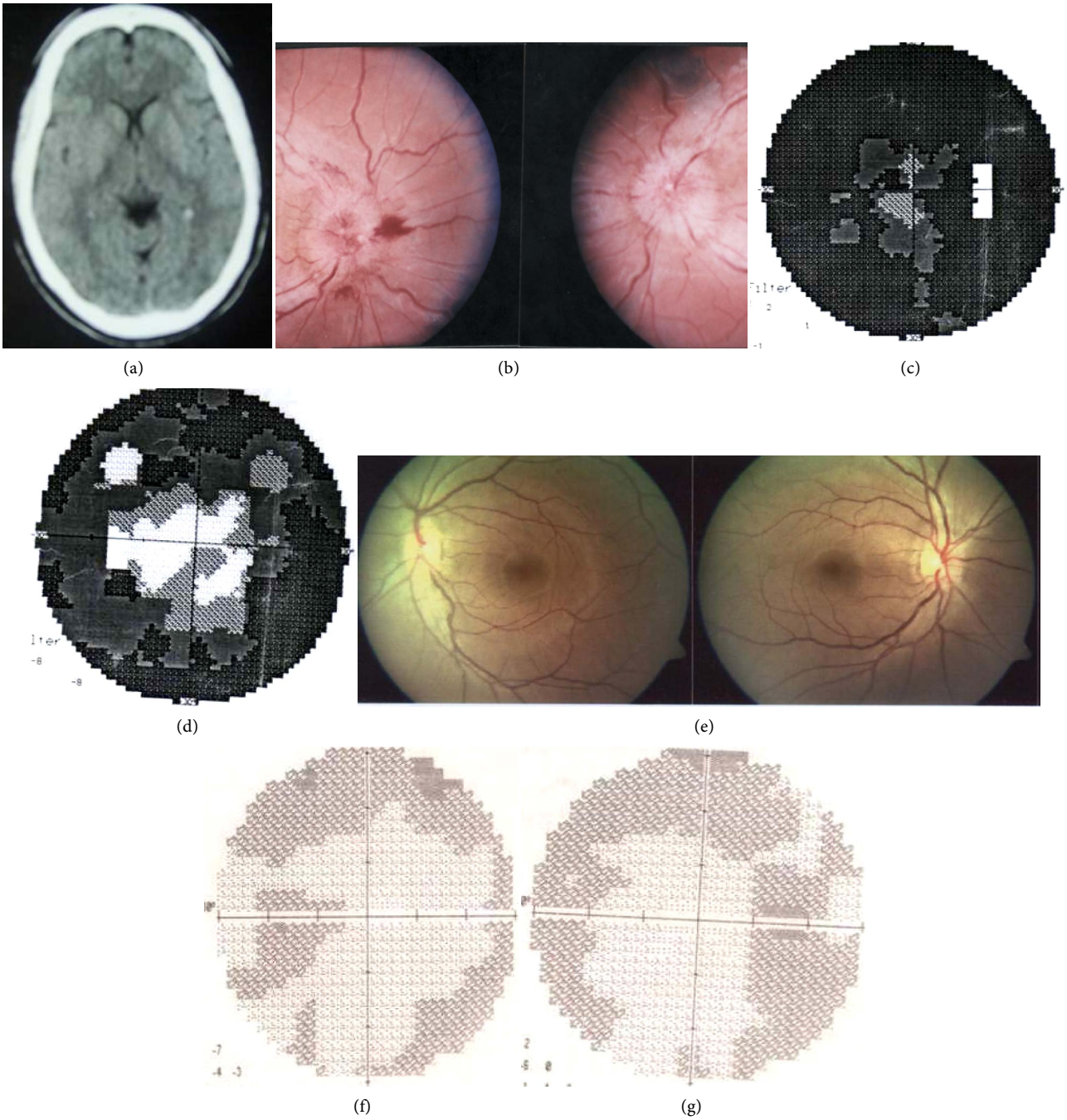

Figure 2. Female patient with lumboperitoneal shunt, (a) Preoperative Axial normal CT brain of the patient, (b) Preoperative fundus examination showed advanced papilledema, (c) Preoperative Visual field of the patient showed concentric contraction of the field in the left eye, (d) Preoperative Visual field of the patient showed tubular field of the right eye, (e) Postoperative fundus examination showed complete resolution of papilledema, (f) and (g) postoperative visual field showed bilateral improvement of the field.

who required interspinous exposure and minimal laminectomy for shunt placement. ALL stereotactic guided Ventricular shunts were placed safely with adequate intraoperative CSF flow from the distal end before peritoneal insertion. Early periprocedural complications included wound infection treated conservatively with antibiotics only occurred in two patients with LP shunt. Extera-peritoneal

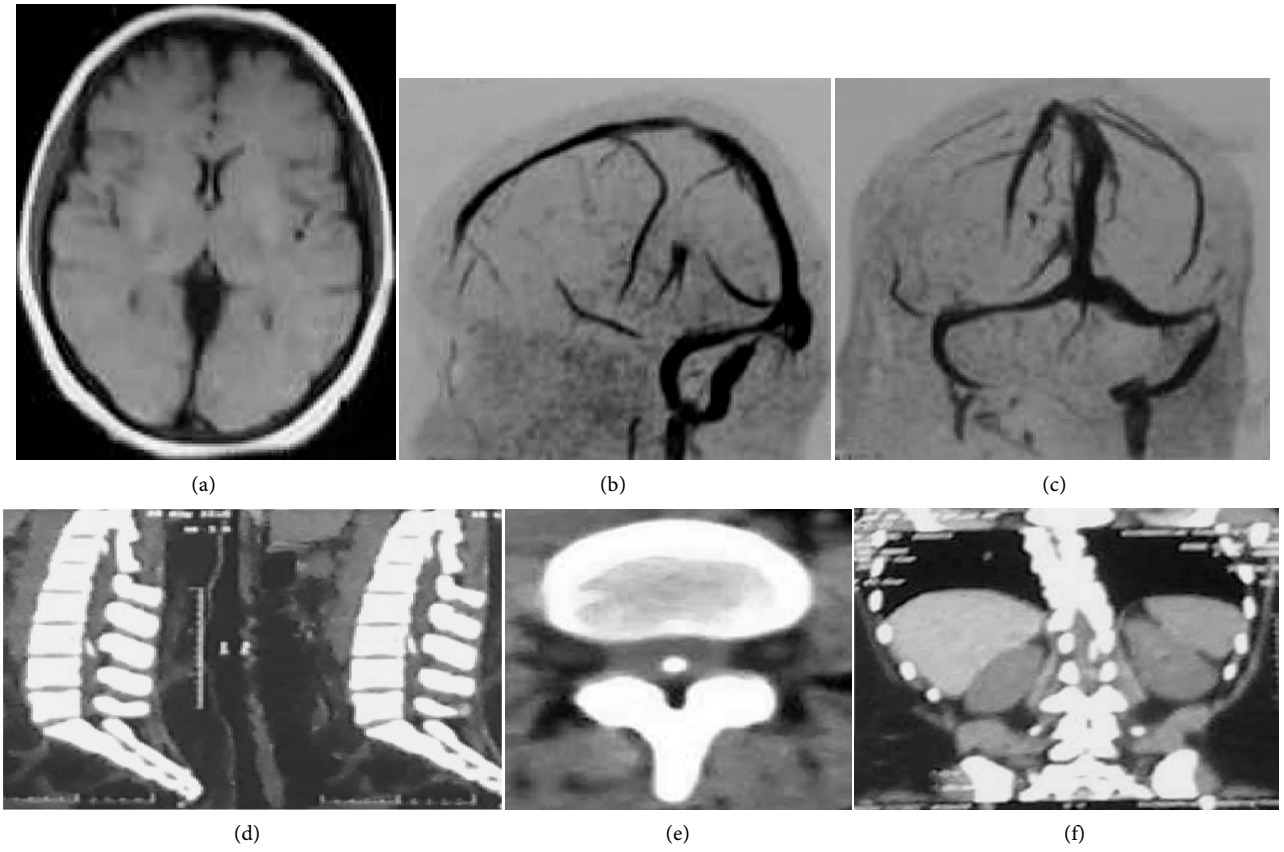

Figure 3. Female patient with lumboperitoneal shunt and subcutaneous abdominal wound collection, (a) Ti weighted image axial MRI showed slit ventricle, (b) and (c) Magnetic resonance venography of the patient with apparent stenosis or thrombosis, (d) CT lumbosacral spine with sagittal reconstruction showed the intra-thecal location of the catheter, (e) CT Lumbar spine, axial view showed the location of the proximal catheter, (f) CT abdomen showed the intraperitoneal location of the distal catheter.

shunt insertion was reported in one patient with LP shunt which required reoperation with correct insertion of the distal end of the same shunt in the early postoperative period as shown in Table 2. Subcutaneous collection was reported in one patient with LP at the abdominal incision treated conservatively after Abdominal CT which showed intra-peritoneal insertion of the shunt as shown in Figure 4. Radiculopathy was reported in two patients on the early postoperative period that improved with analgesia. We do not report postoperative lower limb weakness from LP shunt insertion that may require MRI.

On late follow up of the patients; 9 (20.9%) patients required shunt revision due to recurrence of symptoms. 8 patients operated with LP peritoneal shunt and one patient only with stereotactic guided Ventricular shunt. The most common cause for shunt revision was distal migration and/or recurrence of severe headache and visual obscuration. The only patient with revision in the Ventricular shunting group had proximal obstruction that required shunt revision.

4. Discussion

IIH is a well known but under-investigated clinical entity with unclear pathophysiology; hence it does not have a well-established management algorithm,

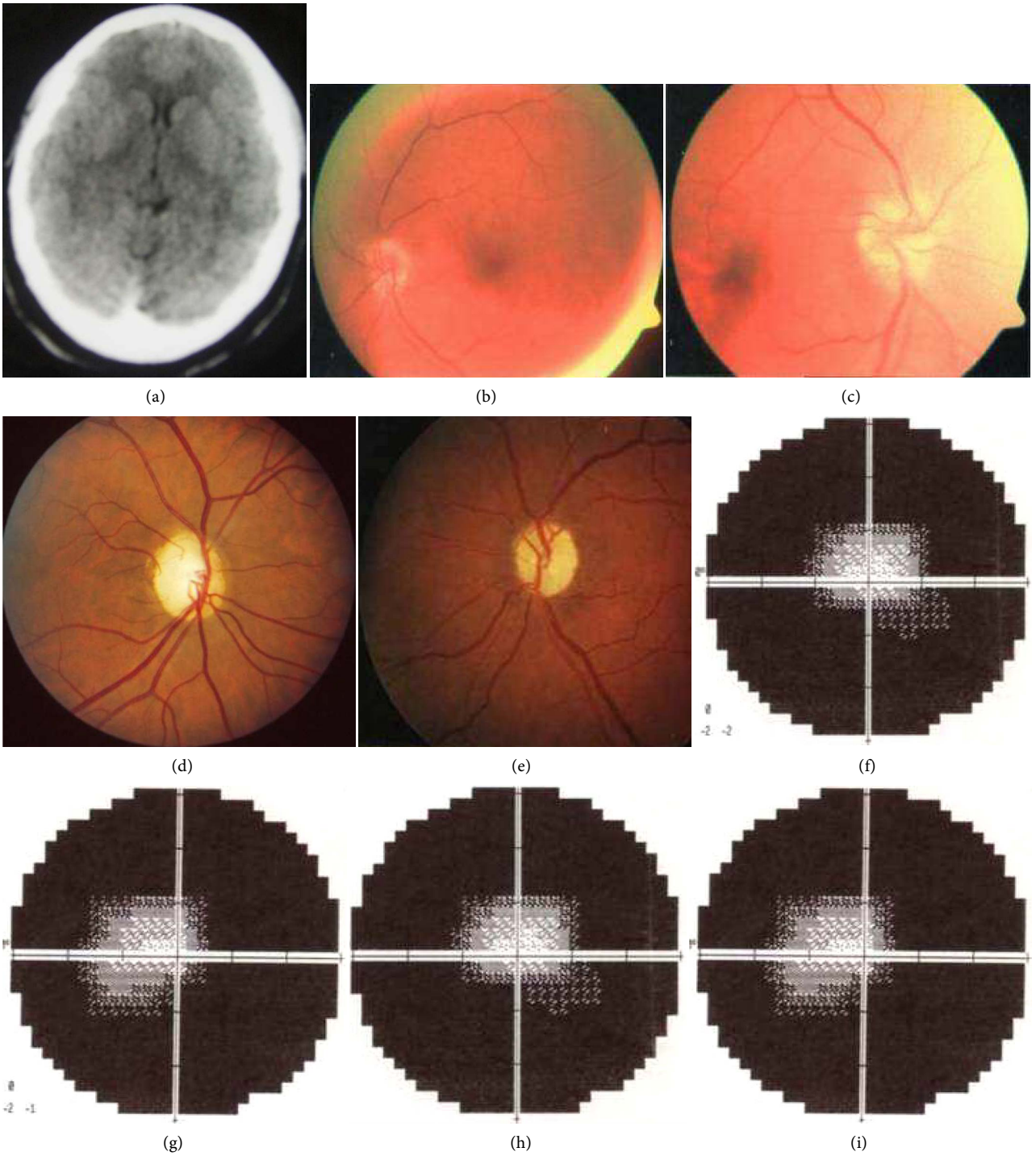

Figure 4. Female patient with lumboperitoneal shunt with no visual improvement, (a) Axial CT brain showed the slit ventricle, (b) and (c) Colored fundus showed chronic advanced preoperative papilledema, (d) and (e) postoperative fundus picture showed optic atrophy, (f) and (g) preoperative visual tubular field of the patient, (h) and (i) Postoperative visual field show no improvement.

given the paucity of studies on that pathology [1]. Headache was the most commonly reported symptom in 100% of our patients followed by visual obscurations in 81.4%. This is in agreement with previously published data of Sinclair and others [9], who reported headache in 96% and visual obscurations in 78% of

Table 2. Periprocedural complications and outcome of the patients.

Clinical outcome
Improved headache 36 (83.7%)
Papilledema 30 (69.7%)
Procedure success
Ventricular shunt 14 (100)
LP shunt 26 (89.65%)
Perioperative complications
Ventricular shunt no complications
LP shunt 6/29 (20.7%)
Wound infection 2
Extraperitoneal insertion 2
Subcutaneous collection 1
Radiculopathy 1
Shunt revisions
Ventricular shunt 1 (7.1%)
LP shunt 8 (27.6%)

their patients. The goal of treatment of IIH is directed for preservation of vision and reduction of symptoms usually headache [10]. Multiple factors should be considered when selecting the appropriate treatment. The presence and degree of symptoms, the severity of visual obscuration and any apparent progression should be considered [11]. Identification of underlying factors such as obesity, causative medication, anemia, and venous sinus thrombosis should be considered [1]. Treatment of IIH includes multiple modalities that include carbonic anhydrase inhibitors, loop diuretics, corticosteroids, topiramate, repeated lumbar punctures and surgery [2]. Surgical approaches for treating patients with refractory or rapidly deteriorating IIH include both CSF diversion techniques and non-CSF diversion techniques. The non-CSF diversion techniques include optic nerve sheath decompression (ONSD), bariatric surgery to reduce weight and recently intracranial venous sinus stenting [2] [6] [10]. ONSD effectively treats patients with papilledema and severe visual deterioration but does not improve headache in most patients. ONSD rapidly improves papilledema on the operated side and rarely on the contralateral side. The procedure has a little effect on ICP and the clinical improvement is likely due to local reduction in pressure on the optic nerve by decreasing the intra-sheath pressure [6]. Patients with improved visual acuity after ONSD should be monitored carefully as long-term visual decline may occur [6].

There are cumulative data recently reported that some patients with IIH have bilateral transverse sinus stenosis [4] [7]. This vascular theory supported venous sinus stenting in some patients with IIH with variable results [4] [7]. Puffer and others in their review of literatures including 143 IIH patients treated with venous sinus stenting revealed that most of the patients had improvement of headache,

papilledema or visual acuity after the procedures [12]. Restenosis may occur after the procedure [7]. Moreover, transverse sinus stenosis as revealed by venography, persists in patients with IIH after normalization of CSF pressure showing the lack of a direct relationship between the caliber of transverse sinus and CSF pressure [7]. This leads to a debate on whether such a transverse sinus stenosis is the cause or effect of IIH [7].

CSF diversion procedures are the most widely performed surgical treatment for IIH and can treat papilledema, headache and visual deterioration [2]. The procedures have a technical simplicity, minimal invasiveness, tolerable and with easily managed complications [11] [13] [14]. El-Saadany and his colleagues reported recovery of headache in (86.4%) of their patients and complete resolution of papilledema in 72.7% of their patients treated with lumboperitoneal shunts [2]. Woodworth *et al*. [15], reported significant improvement of headache immediately after shunting in 100% of their patients. Kandasamy and others reported clinical improvement of all of his 17 patients treated with electromagnetic stereotactic ventriculoperitoneal CSF shunting for idiopathic intracranial hypertension [11]. The authors now preferentially perform custom-designed electromagnetic image-guided VP shunt placement instead of LP shunting in treating medically refractory IIH. Ventricular shunts, such as ventriculoperitoneal shunts, ventriculojugular and ventriculoatrial shunts are less frequently used due to the potential difficulty of ventricular catheter insertion into the slit ventricle in patients with IIH [16]. However stereotactic or navigation guided ventricular placement of the proximal catheter may help to overcome such problem [15] [17]. In our study, the ventricular cannulation was successful in all of our patients with ventricular shunting with less incidence of perioperative complications and shunt revisions. This is in consistent with previously published data. Tarnaris and others in their study of difference in outcomes in patients with IIH with the choice of CSF diversion site reported 20.5% rate of complications and the need for revision was 35% [18]. The authors reported that the patients with lumboperitoneal shunt suffered more complications and first time revision than patients with VP shunt; however the difference was not statistically significant [18]. Kandasamy and his colleagues evaluated 17 consecutive patients treated with electromagnetic stereotactic ventriculoperitoneal CSF shunting for IIH and reported clinical improvement in all patients at the last follow-up compared to their preoperative conditions [11]. In a nationwide study, Menger and others evaluated 4480 patients with IIH, with 2505 undergoing first-time VP shunt placement and 1754 undergoing initial LP shunt placement. Revision surgery occurred in 3.9% of admissions for VP shunts and in 7% of admissions for LP shunts [19]. Woodworth and others in their study of 21 patients underwent 32 ventricular shunting procedures. All patients experienced significant improvement of headache immediately after shunting. 10% of ventricular shunting failed at 3 months after insertion, 20% failed by 6 months, 50% failed by 12 months and 60% failed by 24 months [15]. However, all shunt revision was due to distal obstruction, over drainage, distal catheter migration or CSF leak and there were

no shunt revisions due to proximal catheter obstruction [15]. In contrast to the aforementioned studies, Abubaker *et al*. [10], Reported slightly higher failure for VP shunts (14%) than LP shunts. However, revision rates were higher with LP shunts (60%) than VP shunts (30%).

Although this study evaluated adult group with IIH, in pediatric group of patients, Lumboperitoneal shunt may carry worse prognosis and complications in pediatric group of patients. Niotakis *et al*. [17], in their 7 pediatric patients reported shunt revisions in all patients and 6/7 had persisting headaches at their last follow up. In addition, delayed Chiari malformation, spinal intradural hematoma and permanent paraparesis and leakage of CSF from the thecal sac after lumboperitoneal shunt placement were previously reported [17] [19] [20].

This study is limited by being retrospective study and by the small number of patients treated with steriotactic guided VP shunts compared whom treated by lumpoperitoneal shunts. However, the study demonstrates that ventricular shunts can be effectively performed with stereotactic guided ventricular cannulation with less perioperative complications and shunt revisions.

5. Disclosure Statement

The authors have no personal or institutional interest in the materials or supplies used in the study.

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