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REVIEW ARTICLE - NEUROSURGICAL TECHNIQUES

# Efficacy, complications and cost of surgical interventions for idiopathic intracranial hypertension: a systematic review of the literature

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**Abstract**
*Background* To define the efficacy, complication profile and cost of surgical options for treating idiopathic intracranial hypertension (IIH) with respect to the following endpoints: vision and headache improvement, normal CSF pressure restoration, papilloedema resolution, relapse rate, operative complications, cost of intervention and quality of life.
*Methods* A systematic review of the surgical treatment of IIH was carried out. Cochrane Library, MEDLINE and EMBASE databases were systematically searched from 1985 to 2014 to identify all relevant manuscripts written in English. Additional studies were identified by searching the references of retrieved papers and relative narrative reviews.
*Results* Forty-one (41) studies were included (36 case series and 5 case reports), totalling 728 patients. Three hundred forty-one patients were treated with optic nerve sheath fenestration (ONSF), 128 patients with lumboperitoneal shunting (LPS), 72 patients with ventriculoperitoneal shunting (VPS), 155 patients with venous sinus stenting and 32 patients with bariatric surgery. ONSF showed considerable efficacy in vision improvement, while CSF shunting had a superior headache response. Venous sinus stenting demonstrated satisfactory results in both vision and headache improvement along with the best complication profile and low relapse rate, but longer follow-up periods are needed. The complication rate of bariatric surgery was high when compared to other interventions and visual outcomes have not been reported adequately. ONSF had the lowest cost.
*Conclusions* No surgical modality proved to be clearly superior to any other in IIH management. However, in certain contexts, a given approach appears more justified. Therefore, a treatment algorithm has been formulated, based on the extracted evidence of this review. The traditional treatment paradigm may need to be re-examined with sinus stenting as a first-line treatment modality.

**Keywords** Idiopathic intracranial hypertension · Pseudotumour cerebri · Surgical interventions · Efficacy · Complications · Cost of intervention

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**Abbreviations**

| | |
|---|---|
| BMI | Body mass index |
| CSF | Cerebrospinal fluid |
| HDU | High dependency unit |
| HRQOL | Health-related quality of life |
| ICP | Intracranial pressure |
| IIH | Idiopathic intracranial hypertension |
| LAGB | Laparoscopic adjustable gastric banding |
| LOS | Length of hospital stay |
| LPS | Lumboperitoneal shunting |
| LRYGB | Laparoscopic Roux-en-Y gastric bypass |
| MRI | Magnetic resonance imaging |
| MRV | Magnetic resonance venography |

| | |
|---|---|
| NHS | National health service |
| ONSF | Optic nerve sheath fenestration |
| PICOS | Participants, Interventions, Comparisons, Outcomes, Studies |
| PRISMA | Preferred Reporting Items for Systematic Reviews and Meta-Analyses |
| RCT | Randomised control trial |
| RPPR | Revision per patient rate |
| SVPS | Stereotactic ventriculoperitoneal shunting |
| VPS | Ventriculoperitoneal shunting |

## Introduction

Idiopathic intracranial hypertension (IIH) is a syndrome of obscure aetiology resulting in elevated intracranial pressure (ICP). Overall incidence is low (0.9/100,000), though nearly 20 times higher in obese women of childbearing age [23]. Although symptoms are not life-threatening, they can be sight-threatening and also include incapacitating headaches.

Several theories have been proposed to explain the pathophysiology of IIH but none has adequately done so. Elevated intra-abdominal pressure (as caused by obesity), sleep apnoea (a sequelae of obesity), decreased conductance to CSF outflow and venous sinus stenosis have been implicated as potential causes of IIH [8, 11, 51, 65]. However, this syndrome likely represents the common final pathway of several different mechanisms. Certain drugs (such as tetracyclines, vitamin A) and systemic illnesses (such as lupus erythematosus, uraemia, hypothyroidism) have also been associated with IIH [15, 33, 48, 50, 57].

Headache, visual disturbances and pulsatile tinnitus are the most common symptoms, but asymptomatic presentations can occur, with patients being diagnosed after routine ophthalmological examination [32]. Papilloedema is the cardinal sign and the cause of visual loss in IIH. The majority of IIH patients experience visual field defects, the most common being enlargement of the blind spot. Arcuate scotomas, nasal defects and general field constrictions can also be present [71]. Visual acuity is typically normal, except for fulminant or long-term syndromes, or in the presence of a detached retina [71]. Diagnosis is based on the "modified Dandy criteria", which were initially proposed in 1985 and amended by Friedman (Friedman's criteria) in 2002 [30].

Treatment options range from conservative to interventional and surgical. Conservative options include weight loss, drugs (acetazolamide, topiramate, furosemide, octreotide) and serial lumbar punctures. Interventional and surgical options are CSF diversion procedures (ventriculoperitoneal or lumboperitoneal shunting), optic nerve sheath fenestration (ONSF), venous sinus stenting and bariatric surgery.

Apparently, no consensus has been reached regarding the optimal management of IIH. In the majority of cases, medical treatment supplemented by weight reduction often suffices, halting the progression of IIH [9]. However, one quarter of patients require surgical/interventional management due to visual deterioration or persistent headaches [18]. Medically refractory patients have traditionally been treated with CSF diversion or ONSF [31]. Recently, venous sinus stenting and bariatric surgery have also been used. Randomised, prospective studies, along with systematic reviews, comparing and comprehensively analysing the outcomes of the various surgical procedures are currently lacking [44]. There are, however, some reviews [9, 21, 26, 31, 42, 58, 69, 70] discussing the pathophysiology behind IIH along with the various medical and surgical treatments. Thus, our objective was to assess available data related to the effects and complications of different surgical interventions for IIH, with a view to generating an evidence-based treatment algorithm. Furthermore, we set out to propose specific guidance on data collection and design that will improve the impact and comparability of future studies assessing outcomes of surgical management of IIH.

## Methods

A systematic review concerning the surgical treatment of IIH was conducted according to recommendations of the "Preferred Reporting Items for Systematic Reviews and Meta-Analyses" (PRISMA) statement [47].

### Search methods for identification of studies

The Cochrane Library, MEDLINE and EMBASE were systematically searched from 1985 ("Modified Dandy Criteria" were first introduced in March 1985) to January 2014 to identify all relevant articles published in English. Moreover, the references of retrieved papers and relative narrative reviews were searched to identify additional articles. Table 1 includes details of the search strategy.

### Inclusion criteria

Inclusion criteria in terms of PICOS (Participants, Interventions, Comparisons, Outcomes, Studies) are outlined in Table 2. This review was designed to include randomised controlled trials, non-randomised trials or observational studies. Only studies in which the "modified Dandy criteria" were met (either nominally or descriptively) were considered for inclusion. Moreover, the syndrome is often stated as idiopathic even in the context of a known associated factor. These conditions were considered as secondary intracranial hypertension and were excluded from this review. Studies including combined outcome data for both VPS and LPS were carefully scrutinised, and only those with unambiguous extractable

**Table 1** MEDLINE search strategy (via OVID)

| |
|---|
| 1 exp pseudotumor cerebri/ |
| 2 exp intracranial hypertension/ |
| 3 1 or 2 |
| 4 intra?cranial.tw. |
| 5 intra cranial.tw. |
| 6 inter?cranial.tw. |
| 7 inter cranial.tw. |
| 8 or/4–7 |
| 9 (hypertens$ or pressur$).tw. |
| 10 (increas$ or elevat$ or high$).tw. |
| 11 (benign$ or idiopathic$).tw. |
| 12 8 and 9 and 10 and 11 |
| 13 ((pseudotumor or pseudo tumor$) adj3 cerebr$).tw. |
| 14 (pseudoabscess$ or pseudo abscess$).tw. |
| 15 ((otitic or toxic$) adj5 hydroceph$).tw. |
| 16 (meningeal adj3 hydrop$).tw. |
| 17 3 or 12 or 13 or 14 or 15 or 16 |
| 18 (surger$ or surgical or operative or operation$).tw. |
| 19 (CSF diverting procedure$ or CSF diversion$ or CSF shunt$ or CSF shunting).tw. |
| 20 (lumboperitoneal shunt$ or ventriculoperitoneal shunt$).tw. |
| 21 (optic nerve sheath fenestration$ or optic nerve sheath decompression$).tw. |
| 22 (venous sinus stent$ or dural sinus stent$ or venous sinus stenting or dural sinus stenting).tw. |
| 23 bariatric surger$.tw. |
| 24 or/18–23 |
| 25 17 and 24 |

Similar modified searches were performed for EMBASE and Cochrane

outcome data relevant to the individual shunting modality were included in this review.

## Data collection and extraction

Suitability for inclusion of studies (titles/abstracts initially, full texts subsequently) was independently assessed by two reviewers (according to the aforementioned inclusion criteria). Disagreements between reviewers were settled by consensus with the exception of three cases, where the issue was resolved by reference to a third party. A predefined extraction form was used for data acquisition. Definition of visual improvement was based upon the investigators' criteria. Relapse rate was defined as recurrence of headache or visual symptoms. This definition can be easily applied to all interventions except for CSF diversion procedures, in which IIH relapse could be attributed to the dysfunction of the shunt system (shunt obstruction, infection, low pressure function). Concerning VPS and LPS, shunt revisions were recorded.

## Cost of interventions

The cost of intervention was defined as an approximate tariff for each intervention calculated by combining data from two hospitals (Evangelismos Hospital, Athens, Greece, and the Western General Hospital, Edinburgh, UK) along with data from "NHS reference costs for 2012-2013" and "2013/14 Scottish Tariffs for Cross Boundary Flow Costing" [20, 59].

We estimated an approximate cost of the surgical procedures by combining the following data: Average respective

**Table 2** Inclusion criteria in terms of PICOS

| | |
|---|---|
| Participants | All patients of any age and gender clinically diagnosed with IIH. Patients with secondary intracranial hypertension (such as venous sinus thrombosis) were excluded |
| Interventions | 1. CSF diversion procedures (ventriculoperitoneal or lumboperitoneal shunt) |
| | 2. Optic nerve sheath fenestration (ONSF) |
| | 3. Venous sinus stenting |
| | 4. Bariatric surgery |
| Comparisons | Placebo or other therapeutic modalities or none (if efficacy has been examined with inclusion of pertinent quantitative data) |
| Outcomes | Primary |
| | • Improvement of vision |
| | • Restoration of normal CSF pressure |
| | • Improvement of papilloedema |
| | • Improvement of headache |
| | • Relapse rate/revisions |
| | • Complications of the procedure |
| | Secondary |
| | • Cost of intervention |
| | • Quality of life |
| Study design | Randomised controlled trials, non-randomised trials, observational studies |

*PICOS* Paricipants, Interventions, Comparisons, Outcomes, Study design length of hospital stay (LOS), daily cost of a hospital bed (data from NHS reference costs), average cost of hardware used (e.g. the shunt itself, the laparospopic tray/kit in bariatric procedures, the stent and other interventional disposables used in venous sinus stenting), the length of operative time (in minutes) as well as the respective hourly cost per theatre session.

Approximate cost per intervention was calculated as follows: [Average length of hospital stay (days) × Daily cost of a hospital bed] + Cost of hardware used + [Minutes of operative time × (Cost of an operative hour/60)].

Length of hospital stay (LOS), hardware cost and operative time were estimated by combining data from two hospitals (Evangelismos Hospital, Athens, Greece, and Western General Hospital, Edinburgh, UK). Length of hospital stay and operative time were estimated as the average of that reported for the specific procedure by retrospectively examining the hospital's medical records. Average hardware cost was provided by the hospital's billing office. The daily cost of a hospital bed was retrieved from "NHS reference costs for 2012-2013", which is £273 [20]. The HDU bed per day cost was reported in "2013/14 Scottish Tariffs for Cross Boundary Flow Costing" [59]. Running costs for an operating theatre average approximately £1200 per hour [49]. The venous sinus stenting procedure occupies the angiography suite for a session, which costs approximately £2500.

### Statistical analysis

Characteristics and outcome data of the included studies were collected, pooled and compared for each type of surgical modality. Pooled estimates (meta-analysis) were not performed for the following reasons: considerable heterogeneity of the patient populations, large number of treatment subgroups and outcome measures, and the small number of patients/data in specific treatment subgroups.

## Results

### Search results and description of studies

The search yielded 1304 studies (Cochrane Library: 195, MEDLINE: 693, EMBASE: 393, References/bibliography: 23). After removing the duplicates, 1122 studies remained. The majority referred to cases of intracranial hypertension due to a known cause, such as tumour or injury, and thus were excluded (995 studies). A total of 84 full-text articles were assessed for inclusion. Of these, 16 were excluded because of the "wrong topic", 2 because of the "wrong intervention", 16 because of "not enough quantitative data", 6 because "data were not extractable" and 3 because of the "wrong study type" (see Table 3 for characteristics of excluded studies). No RCT was identified. Finally, 41 studies (36 case series and 5 case reports), totaling 728 patients, were included in this review (Fig. 1). Three hundred forty-one patients underwent ONSF (525 optic nerves), 128 patients underwent LPS, 72 patients underwent VPS, 155 patients underwent venous sinus stenting, and 32 patients underwent bariatric surgery.

Female patients constitute 85.7 % (600/700) of this cohort. Previous treatments were defined in 438. Specifically, 348 patients had previously received medical treatment (mainly acetazolamide), 82 patients medical and surgical treatment, 6 only surgical treatment and 2 patients no treatment. Weight status was reported in 289 patients, 226 of whom were obese (78.2 %). We extracted data for all outcomes described, except "quality of life". Quality assessment was not conducted because included studies were only case series and case reports.

### Effects of interventions

*Characteristics and primary outcomes*

Given the small number of patients in specific subgroups/outcomes, we present the various values in both numbers of patients/eyes and percentages, thus avoiding misleading extreme results. In every intervention/outcome category, the number of patients for which data exist (pre- and post-operative data) is reported. In almost all cases, this is smaller than the total number of participants in this category. In general, reported values represent patients except for the ONSF category in visual acuity, visual fields and relapse rate outcomes, where values represent eyes. CSF pressure was measured either by lumbar puncture in the lateral decubitus position or ICP monitoring. In all studies, papilloedema was evaluated by fundoscopy. Improvement of headache was defined as subjective pain relief following intervention. The complication profile of the surgical interventions is presented in Table 4. For LPS/VPS procedures, we report the indications for revision of the shunt. The majority of sinus stenting and ONSF complications were transient and benign.

**Optic nerve sheath fenestration (ONSF)** ONSF was examined in 15 studies, 12 case series and 3 case reports (see Tables 5 and 10) [3, 7, 10, 16, 17, 28, 34, 38, 40, 53, 55, 60, 63, 64, 68]. In particular, 341 patients underwent 525 ONSFs. The mean age of the patients was 31.7 years, and 285/341 of them were female (83.6 %). The weight status of the patients was provided only in three studies [53, 55, 63], in which 37/39 (94.9 %) patients were obese. Average follow-up was 42.3 months. In 75 % of patients (102/136), ONSF was the first surgical treatment performed. Several techniques have been used. The medial transconjuctival approach was performed in 342 eyes, the lateral transconjuctival approach in 53 and a combined approach (medial and lateral transconjuctival) in 3, while the superomedial lid slit approach (vertical) was performed in 1 eye.

**Table 3** Excluded studies

Main reason for exclusion: Wrong topic (e.g. secondary intracranaial hypertension, not meeting "Modified Dandy criteria", examining other outcomes)

Barnett, M., et al. (2013). "Intracranial hypertension presenting with severe visual failure, without concurrent headache, in a child with nephrotic syndrome." BMC Pediatrics 13(1)

Dorman, R. B., et al. (2012). "Risk for hospital readmission following bariatric surgery." PLoS ONE [Electronic Resource] 7(3): e32506

Green, J. P., et al. (1996). "Normal pressure" pseudotumor cerebri." Journal of Neuro-Ophthalmology 16(4): 241–246

Guy, J., et al. (1990). "Treatment of visual loss in pseudotumor cerebri associated with uremia." Neurology 40(1): 28–32

Karahalios, D. G., et al. (1996). "Elevated intracranial venous pressure as a universal mechanism in pseudotumor cerebri of varying etiologies." Neurology 46(1): 198–202

Khan, M. U., et al. (2013). "Idiopathic intracranial hypertension associated with either primary or secondary aldosteronism." American Journal of the Medical Sciences 346(3): 194–198

Lee, J. K., et al. (2012). "Incidence and risk factors of ventriculoperitoneal shunt infections in children: a study of 333 consecutive shunts in 6 years." Journal of Korean Medical Science 27(12): 1563–1568

Michaelides, E. M., et al. (2000). "Pulsatile tinnitus in patients with morbid obesity: The effectiveness of weight reduction surgery." American Journal of Otology 21(5): 682–685

Nagasaka, T., et al. (2010). "Subcutaneous migration of distal ventriculoperitoneal shunt catheter caused by abdominal fat pad shift—three case reports." Neurologia Medico-Chirurgica 50(1): 80–82

Nithyanandam, S., et al. (2008). "Optic nerve sheath decompression for visual loss in intracranial hypertension: report from a tertiary care center in South India." Indian Journal of Ophthalmology 56(2): 115–120

Stannard, M. W. and N. K. Rollins (1995). "Subcutaneous catheter calcification in ventriculoperitoneal shunts." AJNR: American Journal of Neuroradiology 16(6): 1276–1278

Taban, M., et al. (2001). "Histopathology and ultrastructural examination of optic nerve sheath biopsies after optic nerve sheath decompression with and without mitomycin." Ophthalmic Plastic and Reconstructive Surgery 17(5): 332–337

Thomas, S., et al. (2004). "Optic nerve sheath fenestration for intracranial hypertension associated with chronic inflammatory demyelinating polyneuropathy." Ophthalmic Plastic and Reconstructive Surgery 20(4): 325–327

Traynelis, V. C., et al. (1988). "Cerebrospinal fluid eosinophilia and sterile shunt malfunction." Neurosurgery 23(5): 645–649

Traynelis, V. C., et al. (1991). "Millipore analysis of valvular fluid in sterile valve malfunctions." Neurosurgery 28(6): 848–852

Yoshida, S., et al. (2000). "Migration of the shunt tube after lumboperitoneal shunt—two case reports." Neurologia Medico-Chirurgica 40(11): 594–596

Main reason for exclusion: Wrong intervention

Zada, G., et al. (2010). "Cushing's disease and idiopathic intracranial hypertension: case report and review of underlying pathophysiological mechanisms." Journal of Clinical Endocrinology and Metabolism 95(11): 4850–4854

**Table 3** (continued)

Karaman, K., et al. (2003). "Familial idiopathic intracranial hypertension." Croatian Medical Journal 44(4): 480–484

Main reason for exclusion: Not enough quantitative data

Alleyne Jr, C. H., et al. (1996). "Cranial migration of a lumboperitoneal shunt catheter." Southern Medical Journal 89(6): 634–636

Cabezudo, J. M., et al. (1990). "Infection of the intervertebral disc space after placement of a percutaneous lumboperitoneal shunt for benign intracranial hypertension." Neurosurgery 26(6): 1005–1009

Caron, E., et al. (2011). "Pediatric Emergency Department experience with rapidly progressing idiopathic intracranial hypertension." Journal of Investigative Medicine 59 (2): 377–378

Gates, P. and J. Christensen (2013). "Immediate resolution of idiopathic intracranial hypertension with drainage of CSF at low pressure." Neurology 80 (1 MeetingAbstracts)

Golnik, K. C., et al. (1999). "Visual loss in idiopathic intracranial hypertension after resolution of papilledema." Ophthalmic Plastic and Reconstructive Surgery 15(6): 442–444

Hay, S. A., et al. (2004) Endoscopic implantation and patency evaluation of lumboperitoneal shunt: an innovative technique. Surgical Endoscopy 18, 482–484 DOI: 10.1007/s00464-003-9038-4

Kabeya, R., et al. (2000). "Cerebral blood flow during plateau waves in a patient with benign intracranial hypertension—case report." Neurologia Medico- Chirurgica 40(5): 287–292

Lim, M., et al. (2005). "Visual failure without headache in idiopathic intracranial hypertension." Archives of Disease in Childhood 90(2): 206–210

Narula, P., et al. (2006). "Visual loss and idiopathic intracranial hypertension in children with Alagille syndrome." Journal of Pediatric Gastroenterology and Nutrition 43(3): 348–352

Perez, M. A., et al. (2013). "Primary spontaneous cerebrospinal fluid leaks and idiopathic intracranial hypertension." Journal of Neuro-Ophthalmology 33(4): 327–334

Riggeal, B., et al. (2012). "Does the presence of transverse sinus stenosis (TSS) influence the clinical presentation and outcome of idiopathic intracranial hypertension (IIH)." Neurology 78 (1 Meeting Abstract)

Rohr, A., et al. (2007). "Reversibility of venous sinus obstruction in idiopathic intracranial hypertension." AJNR: American Journal of Neuroradiology 28(4): 656–659

Russo, R. R., et al. (2010). "Progressive visual loss due to obstruction of an optic nerve sheath fenestration demonstrated on SPECT/CT radionuclide cisternography." Clinical Nuclear Medicine 35(3): 208–210

Suri, A., et al. (2002). "Subarachnoid hemorrhage and intracereebral hematoma following lumboperitoneal shunt for pseudotumor cerebri: a rare complication." Neurology India 50(4): 508–510

Tulipan, N., et al. (1998). "Stereotactic ventriculoperitoneal shunt for idiopathic intracranial hypertension: technical note." Neurosurgery 43(1): 175–176; discussion 176–177

Zagardo, M. T., et al. (1996). "Reversible empty sella in idiopathic intracranial hypertension: an indicator of successful therapy?" AJNR: American Journal of Neuroradiology 17(10): 1953–1956

Main reason for exclusion: Data not extractable (e.g. no distinct data in the subgroups)

Chumas, P. D., et al. (1993). "Lumboperitoneal shunting: a retrospective study in the pediatric population." Neurosurgery 32(3): 376–383; discussion 383

McGirt MJ, et al. (2004). "Cerebrospinal fluid shunt placement for pseudotumor cerebri-associated intractable headache: predictors of treatment response and an analysis of long-term outcomes." J Neurosurg;101:627–32

Rosenberg, M. L., et al. (1993). "Cerebrospinal fluid diversion procedures in pseudotumor cerebri." Neurology 43(6): 1071–1072

**Table 3** (continued)

Tarnaris A, et al. (2011). "Is there a difference in outcomes of patients with idiopathic intracranial hypertension with the choice of cerebrospinal fluid diversion site: a single centre experience." Clin Neurol Neurosurg;113:477–9
Thambisetty, M., et al. (2007). "Fulminant idiopathic intracranial hypertension." Neurology 68(3): 229–232
Wilkes, B. N. and R. M. Siatkowski (2009). "Progressive optic neuropathy in idiopathic intracranial hypertension after optic nerve sheath fenestration." Journal of Neuro-Ophthalmology 29(4): 281–283

Main reason for exclusion: Wrong study type (e.g. review, meta-analysis)
Feldon, S. E. (2007) "Visual outcomes comparing surgical techniques for management of severe idiopathic intracranial hypertension (Structured abstract)." Neurosurgical Focus 23, E6
Fridley, J., et al. (2011) "Bariatric surgery for the treatment of idiopathic intracranial hypertension." Journal of Neurosurgery 114, 34–39
Martin, R. L., et al. (2011). "Incidence and correlates of failure for ventriculoperitoneal and lumboperitoneal shunts in patients with idiopathic intracranial hypertension." Journal of Investigative Medicine 59 (1): 179

Visual fields improved in 64 % of cases (247/384 eyes) while visual acuity improved in 67 % of them (275/411 eyes). Visual fields and visual acuity worsened in 8 % (31/384 eyes) and 11 % (44/411 eyes), respectively. Only 41 % of patients (47/114) reported improvement in headache while the remaining 59 % (67/114) reported no change. Papilloedema improved in 95 % of patients (93/98). Furthermore, 63/397 eyes (15.9 %) deteriorated regarding visual acuity and fields after a seemingly successful initial procedure. In 44/63 cases (69.8 %), a second fenestration (re-operation) was performed. Only three studies provide data on pre- and post-operative CSF pressure [28, 54, 62], in which two-thirds of patients achieved restoration of CSF pressure. Seventy-seven complications were reported in 72/278 (26 %) patients with diplopia

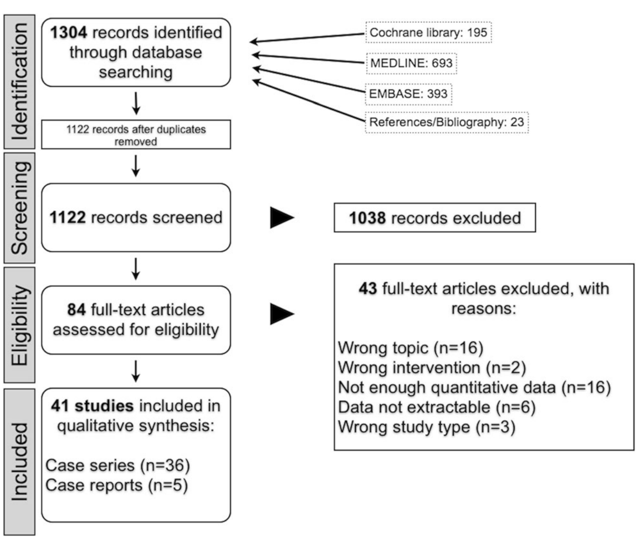


**Fig. 1** Study's "flow of information" diagram according to the PRISMA statement

being the most common of them (see Table 4). Most of these complications were transient.

## CSF diversion procedures

### *Lumboperitoneal shunting (LPS)*

LPS was assessed through seven studies, six case series and one case report, involving a total of 128 patients, 90.1 % (100/110 patients) of which were females (see Tables 6 and 10) [1, 12, 24, 25, 35, 61, 74]. Weight status was specified in 3/7 studies, in which 49/60 (81.7 %) patients were obese (12, 24, 61). The mean follow-up was 44.7 months while the mean age of patients was 32.5 years. In almost all cases (59/60, 98.3 %), LPS was the first surgical procedure performed. Among the included studies, there was no report on the equipment used (e.g. valve-mediated or valve-less type) while the technique was specified in only two studies [25, 35]. Specifically, Tuohy needle insertion was utilised in 18, laminotomy in 4 and laparoscopic-assisted LP insertion in 4 procedures.

Visual acuity improved in 67 % of patients (35/52) and visual fields in 71 % of them (42/59). Five patients experienced deterioration in visual acuity (10 %) and none experienced deterioration of visual fields (0 %). Improvement of headache and papilloedema was reported in 96 % (87/91) and 91 % (49/54) of cases, respectively. Deterioration in papilloedema was reported in one 36-year-old woman who subsequently underwent bilateral ONSFs [61]. No study reported both pre- and post-operative CSF pressure measurements. Forty-nine patients (49/128, 38 %) underwent 211 revisions of the LPS. Consequently, the revision per patient rate (RPPR) was 4.3. The indications for revision are presented in Table 4, with the most common being shunt obstruction (66/100, 66 %).

### *Ventriculoperitoneal shunting (VPS)*

VP shunting was studied in 6 case series totaling 72 procedures in an equal number of patients (see Tables 7 and 10) [1, 2, 14, 45, 72, 73]. The mean age of patients was 35.2 years, 74.2 % (46/62) were female and 51.1 % (24/47) obese. The mean follow-up of the included studies was 39.6 months. In 59.1 % (26/44) of cases, VPS was the first surgical modality performed. In 53 cases, a stereotactic ventriculoperitoneal shunting technique (SVPS) was used (35 frame-less vs. 18 frame-based), while in the remaining 19, the standard procedure was followed. No adequate quantitative data on the type of implanted valves (such as flow-regulated or programmable) nor the use of anti-siphon devices were reported.

Visual fields and visual acuity improved in 69 % (18/26) and 55 % (16/29) of patients, respectively. However, 7 % (2/29) of patients reported deterioration in visual acuity while no patient deteriorated in the visual field category.

**Table 4** Complications

| CSF diversion | | ONSF[‡] | Sinus stenting[§] | Bariatric surgery[II] |
|---|---|---|---|---|
| LPS revisions[*] RPPR = 4,3 | VPS revisions[†] RPPR = 1,83 | | | |
| Shunt obstruction ($n = 66$) | Shunt obstruction ($n = 26$) | Diplopia ($n = 35$) | Transient headache lateralised to the side of treatment ($n = 8$) | Incisional hernia ($n = 9$) |
| Catheter migration/-malposition ($n = 12$) | Infection ($n = 7$) | Corneal dellen ($n = 6$) | Transient partial hearing loss lateralised to the side of treatment ($n = 2$) | Stomal stenosis ($n = 8$) |
| Overdrainage ($n = 12$) | Overdrainage ($n = 6$) | Anisocoria (tonic pupil) ($n = 27$) | Transient unsteadiness ($n = 1$) | Marginal ulcer ($n = 6$) |
| Infection ($n = 4$) | Abdominal wall cyst ($n = 4$) | Orbital apex syndrome ($n = 1$) | Intraluminal thrombus requiring thrombolytic therapy ($n = 2$) | Anastomotic leak ($n = 2$) |
| Sciatica/radiculopathy ($n = 4$) | Catheter migration/-malposition ($n = 4$) | Traumatic optic neuropathy ($n = 1$) | Subdural haematoma ($n = 1$) | Major wound infection ($n = 2$) |
| CSF fistula ($n = 1$) | Valve malfunction ($n = 2$) | 3rd and 6th cranial nerve palsy ($n = 2$) | Subdural and intracerebral haematoma ($n = 1$) | Small bowel obstruction ($n = 1$) |
| Abdominal pain ($n = 1$) | CSF leak ($n = 1$) | Orbital haematoma ($n = 2$) | Subdural haematoma and subarachnoid haemorrhage ($n = 1$) | Upper gastrointestinal bleeding ($n = 1$) |
| | | Perilimbal conjunctival filtering bleb ($n = 1$) | Retroperitoneal haematoma ($n = 1$) | Wound seroma ($n = 1$) |
| | | Disk haemorrhage ($n = 1$) | Femoral pseudoaneurysm ($n = 1$) | |
| | | Conjunctival abscess ($n = 1$) | | |

*RPPR* Revisions per patient rate

[*] $n = 211$ in 49 out of 128 patients. In 100/211 the indication has been stated in the table.

[†] $n = 53$ in 29 out of 70 patients. In 50/53 the indication is stated in the table

[‡] $n = 77$ in 72 out of 278 patients

§$n = 18$ in 18 out of 152 patients

[II] $n = 30$ in 28 out of 32 patients

Improvement of headache was noted in 93 % of patients (63/68) and improvement of papilloedema in 90 % (27/30) of them, while no patient deteriorated in both categories. Fifty-three revisions of VPS were reported in 29/70 (41 %) patients, with a RPPR equal to 1.83. The most common indication for VPS revision was shunt obstruction, in 52 % of cases (26/50). No study provided data on CSF pressure restoration.

**Venous sinus stenting** Eleven studies, ten case series and one case report (involving a total of 155 patients) were reviewed

**Table 5** Characteristics and outcomes of ONSF studies

| No. | Author and year | Pts/ eyes | Female | Follow-up, months (range) | SD | Mean age, years (range) | SD | Primary outcomes | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | | | | | | | Improved VF (worsened) | Improved VA (worsened) | Improved HA (worsened) | Impr. papp. (worsened) | Relapse rate/revisions | Restored CSF pressure |
| 1 | Brourman et al. 1988 [10] | 6/10 | 6 | NR (4–11) | NE | 38.5 (28–62) | 11.74 | 10/10 | 4/5 (0) | NR | 8/10 (0) | NR | NR |
| 2 | Corbett et al. 1988 [17] | 28/40 | 21 | 26.9 (2–90) | 19.61 | 30 (14–62) | 11.62 | 21/38 (7) | 12/40 (6) | 11/17 | 36/36 | 1/28 | NR |
| 3 | Sergott el al. 1988 [60] | 23/29 | 22 | 21.5 (3–45) | 12.3 | 36.8 (18–63) | 11.26 | 29/29 | 24/24 | 13/17 | 24/29 | 2/29 | NR |
| 4 | Kelman et al. 1992 [38] | 17/21 | 15 | 17.3 (11–26) | 4.57 | 39.7 (16–73) | 15.5 | 20/21 (1) | 15/16 (1) | 9/10 | 21/21 | NR | NR |
| 5 | Smith et al. 1992 [63] | 1/1 | 1 | 3 | – | 34 | – | 1/1 | 0/0 | 1/1 | 1/1 | 0/1 | 0/1 |
| 6 | Acheson et al. 1994 [3] | 11/15 | 8 | 37.1 (12–84) | 25.81 | 37 (23–53) | 10.79 | 10/15 (2) | 8/10 (2) | NR | NR | NR | NR |
| 7 | Spoor et al. 1993 [64] | 54/75 | 47 | 16.4 (2–60) | NE | 28.9 (16–37) | 4.66 | 37/75 (3) | 10/13 (2) | NR | 47/51 (3) | 24/75 | NR |
| 8 | Goh et al. 1997 [34] | 86/158 | 72 | 20 (1–108) | NE | 32.1 (NR) | NE | 71/81 (10) | 148/158 (10) | 8/61 | NR | 16/158 | NR |
| 9 | Banta and Farris 2000 [7] | 19/29 | 15 | 16.8 (1–50) | 15.58 | 33.1 (16–52) | 13.93 | 16/27 (4) | 4/29 (3) | NR | NR | 4/29 | NR |
| 10 | Thuente et al, 2005 [68] | 12/17 | 6 | 39.6 (2.4–105.3) | 29.36 | 10.1 (4.4–16) | 4.42 | 6/11 (0) | 9/14 (0) | 5/8 | NR | NR | NR |
| 11 | Knapp and Sampath 2005 [40] | 13/27 | 9 | 9.6 (1–32) | NE | 27 (14–49) | 9.88 | 4/27 (0) | 18/24 (4) | NR | NR | 4/27 | NR |
| 12 | Chandrasekaran et al. 2006 [16] | 32/51 | 29 | 27.6 (0–121) | NE | 33.4 (17–65) | NE | 13/31 (1) | 13/39 (7) | NR | NR | NR | NR |
| 13 | Prabhakaran et al, 2009 [55] | 1/1 | 1 | 28 | – | 34 | – | 1/1 (0) | 1/1 (0) | NR | 1/1 | 0 | 1/1 |
| 14 | Fraser et al. 2012 [28] | 1/1 | 1 | 1.5 | – | 15 | – | NR | 1/1 (0) | NR | 1/1 | 0 | 1/1 |
| 15 | Pineles et al, 2013 [53] | 37/50 | 32 | 210 (4–700) | 200 | 33 (19–74) | 11 | 8/17 (3) | 8/37 (9) | NR | NR | 12/50 | NR |

Reported values represent patients except for the ONSF category in visual acuity, visual fields and relapse rate outcomes, where values represent eyes

*CSF* cerebrospinal fluid, *HA* headache, *Impr. papp.* improved pappiloedema, *Pts* patients, *VA* visual acuity, *VF* visual field

**Table 6** Characteristics and primary outcomes of LPS studies

| No. | Author and year | Patients | Female | Follow-up, months (range) | SD | Mean age, years (range) | SD | Primary outcomes | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | | | | | | | Improved VF (worsened) | Improved VA (worsened) | Improved HA (worsened) | Impr. papp. (worsened) | Revisions (patients) | Restored CSF pressure |
| 1 | Shapiro et al. 1995 [61] | 4 | 4 | 24.1 (3–60) | 25.85 | 26 (22–36) | 6.73 | 3/3 | 3/4 (1) | 4/4 | 1/2 (1) | 0/4 | NR |
| 2 | Eggenberger et al. 1996 [24] | 26 | 24 | 78.5 (21–278) | 66.17 | 31 (8–51) | 7.79 | 9/13 (0) | 9/13 (0) | 15/15 (0) | NR | 65 (14/26) | NR |
| 3 | Burgett et al. 1997 [12] | 30 | 28 | 35 (0–143) | 51 | 32.9 (11–68) | 14.83 | 10/15 (0) | 7/11 (1) | 13/17 (0) | 16/17 (0) | 126 (19/30) | NR |
| 4 | Hammers et al. 2008 [35] | 4 | 4 | NR (7–24) | NE | 27.5 (20–34) | 6.53 | NR | NR | NR | 3/3 | 0 (0/4) | NR |
| 5 | Abubaker et al. 2011 [1] | 18 | NR | 48 (6–96) | NE | NR (25–65) | NE | 10/10 (0) | 6/6 (0) | 11/13 (0) | 11/12 (?) | 12 (10/18) | NR |
| 6 | Yadav et al. 2011 [74] | 24 | 22 | 51 (18–137) | 20.91 | 39 (17–58) | 7.88 | 10/18 (0) | 10/18 (3) | 22/24 (0) | NR | 2 (2/24) | NR |
| 7 | El-Saadany et al. 2012 [25] | 22 | 18 | 12 (NR) | NE | 28.5 (20–38) | NE | NR | NR | 22/22 (0) | 18/20 (0) | 6 (4/22) | NR |

*CSF* Cerebrospinal fluid, *HA* headache, *Impr. papp.* improved pappiloedema, *VA* visual acuity, *VF* visual field

**Table 7** Characteristics and primary outcomes of VPS studies

| No. | Author and year | Patients | Female | Follow-up, months (range) | SD | Mean age, years (range) | SD | Primary outcomes | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | | | | | | | Improved VF (worsened) | Improved VA (worsened) | Improved HA (worsened) | Impr. papp. (worsened) | Revisions (patients) | Restored CSF pressure |
| 1 | Maher et al. 2001 [45] | 13 | 10 | 12.4 (1–38) | 13 | 31.9 (6–54) | 12.57 | 0/1 (0) | 5/11 (0) | 9/13 (0) | NR | 3 (3/13) | NR |
| 2 | Bynke et al. 2004 [14] | 17 | 12 | 77.8 (22–154) | 47.31 | 33.9 (13–63) | 13.27 | 12/16 (0) | 4/8 (2) | 15/15 | 16/16 | 9 (7/17) | NR |
| 3 | Woodworth et al. 2005 [73] | 21 | 17 | 20 (NR) | 17 | 42 (NR) | 10 | NR | NR | 21/21 | NR | 29 (11/21) | NR |
| 4 | Abu-Serieh et al. 2007 [2] | 9 | 5 | 44.3 (6–110) | 33.4 | 26.4 (4–63) | 17.37 | 1/3 (0) | 3/5 (0) | 9/9 | 3/5 (0) | 9 (6/9) | NR |
| 5 | Abubaker et al. 2011 [1] | 10 | NR | 48 (6–96) | NE | NR (25–65) | NE | 4/4 | 4/4 | 9/10 (0) | 6/7 (0) | 3 (2/10) | NR |
| 6 | Warden et al. 2011 [72] | 2 | 2 | 36 (24–48) | 16.97 | 37 (25–49) | 16.97 | 1/2 (0) | 0/1 (0) | NR | 2/2 | NR | NR |

*CSF* cerebrospinal fluid, *HA* headache, *Impr. papp.* improved pappiloedema, *VA* visual acuity, *VF* visual field

**Table 8** Characteristics and primary outcomes of sinus stenting studies

| No. | Author and year | Patients | Female | Follow-up, months (range) | SD | Mean age, years (range) | SD | Primary outcomes: Improved VF (worsened) | Improved VA (worsened) | Improved HA (worsened) | Impr. papp. (worsened) | Relapse rate, patients | Restored CSF pressure |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| 1 | Higgins et al. 2003 [36] | 12 | 12 | 14.2 (2–26) | 6.75 | 33.1 (19–52) | 10.8 | 3/5 (0) | 0/3 (0) | 7/12 (0) | 6/7 (0) | 0 | 1/1 |
| 2 | Owler et al. 2003 [52] | 4 | 3 | 9.8 (5–12) | 3.2 | 27.3 (17–38) | 8.57 | 2/2 | 1/2 (0) | 3/3 | 2/2 (0) | 0 | 3/3 |
| 3 | Donnet et al. 2008 [22] | 10 | 8 | 17.2 (6–36) | 10.12 | 41 (28–60) | 9.79 | NR | 7/8 (0) | 8/10 (0) | 10/10 | 0 | 10/10 |
| 4 | Bussiere et al. 2010 [13] | 10 | 10 | 20.1 (3–60) | 18.95 | NR (16–65) | NR | NR | 4/4 | 10/10 | 9/9 | 0 | NR |
| 5 | Ahmed et al. 2011 [4] | 52 | 47 | 24 (2–108) | NE | 34 (10–64) | NE | 23/30 (0) | 9/13 (0) | 35/43 (0) | 45/45 | 6/52 | NR |
| 6 | Albuquerque et al. 2011 [5] | 18 | 15 | 20 (2–40) | 13.13 | 31 (12–51) | 11.89 | NR | NR | 12/15 (0) | NR | 1/18 | NR |
| 7 | Fields et al. 2011 [27] | 15 | 15 | 14 (1–49) | 12.18 | 34 (20–56) | 10.27 | NR | 6/14 (1) | 10/15 (2) | 15/15 | NR | NR |
| 8 | Kumpe et al. 2012 [41] | 18 | 12 | 43.7 (11–136) | 32.02 | 38 (16–62) | 15 | NR | NR | 10/12 (0) | 15/16 (0) | 2/18 | NR |
| 9 | Lazzaro et al. 2012 [43] | 3 | 3 | 11. 3 (1–21) | 8.17 | NR | NE | 3/3 | 3/3 | 2/3 | 3/3 | 0/3 | NR |
| 10 | Mei et al. 2012 [46] | 1 | 1 | 12 | – | 45 | – | 0/1 (0) | 0/1 (0) | 1/1 | 1/1 | 0 | 1/1 |
| 11 | Radvany et al. 2013 [56] | 12 | 11 | 16 (9–36) | NE | 39 (21–55) | 9.6 | 9/12 (1) | 8/11 (1) | 7/12 (0) | 11/12 (0) | 3/12 | NR |

*CSF* cerebrospinal fluid, *HA* headache, *Impr. papp.* improved pappiloedema, *VA* visual acuity, *VF* visual field

**Table 9** Characteristics and primary outcomes of bariatric surgery studies

| No. | Author and year | Patients | Female | Follow-up, months (range) | SD | Mean age, years (range) | SD | Primary outcomes | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | | | | | | | Improved VF (worsened) | Improved VA (worsened) | Improved HA (worsened) | Impr. papp. (worsened) | Revisions (patients) | Restored CSF pressure |
| 1 | Sugerman et al. 1995 [66] | 8 | 8 | 33.8 (4–72) | 24.01 | 33.4 (26–43) | 5.57 | 6/7 (?) | 0/0 | 8/8 | 7/7 | NR | 8/8 |
| 2 | Sugerman et al. 1995 [67] | 24 | 24 | 62 (8–168) | 52 | 34 (15–53) | 10 | NR | NR | 18/19 (0) | 12/12 | 2/19 | NR |

*CSF* cerebrospinal fluid, *HA* headache, *Impr. papp.* improved pappiloedema, *VA* visual acuity, *VF* visual field

(see Tables 8 and 10) [4, 5, 13, 22, 27, 36, 41, 43, 46, 52, 56]. The mean age of the patients was 34.6 years, 88.4 % (137/155) of whom were females and 75.7 % (84/111) were obese. The mean follow-up was 22.2 months. Sinus stenting was the primary surgical procedure in 80.3 % (110/137) of cases. The preoperative planning for the selection of patients consisted of venography, manometry and measurement of the pressure gradient across the respective sinus stenosis, with a predefined threshold to be met (4–10 mmHg) in order to proceed to stenting. All patients were treated in tertiary neuro-interventional centres, were heparinised during the procedure and received dual antiplatelet therapy pre- and post-procedure. The stents used were either balloon-expandable or self-expandable.

Improvement in visual acuity and visual fields was reported in 65 % (38/59) and 75 % (40/53) of cases, respectively, with two patients deteriorating in visual acuity and only one patient in visual fields category. Headache improved in 77 % (105/136) and deteriorated in 2 % (2/105) of patients while improvement of papilloedema was noted in 98 % of cases (117/120). Twelve patients (8 %) experienced recurrence of symptoms. A second sinus stenting (re-treatment) in the area of the re-stenosis (diagnosed after venography) was performed in ten of them. Pre- and post-procedural CSF pressure measurement was documented in four studies with 15/15 (100 %) patients achieving CSF pressure restoration [22, 36, 46, 52]. Eighteen complications were documented in 18/152 (12 %) patients (Table 4). The majority of them were transient and benign with the most common being headache lateralised to the side of stenting (8/18, 44 %). However, six serious complications related to subdural haematomas ($n = 3$), retroperitoneal haematoma ($n = 1$) and intraluminal thrombus ($n = 2$) were also reported.

**Bariatric surgery** Bariatric surgery was reviewed through data derived from 2 case series involving 32 patients (see Tables 9 and 10) [66, 67]. All included patients (100 %, 32/32) were female and obese. The mean age was 33.8 years. The

**Table 10** Summary of studies comparing outcomes between surgical interventions

| Intervention | Studies | Pts | Female % | Obese % | Mean follow-up, months | Mean age, years, | First surgery performed% | Impr VF % | Primary outcomes | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | | | | | | | | Impr VA % | Impr HA % | Impr papp % | RR % | Complications % | Revisions (RPPR) |
| ONSF | 15 | 341 | 83.6 | 94.9 | 42.3 | 31.7 | 75 | 64 | 67 | 41 | 95 | 15.9 | 26 | NA |
| LPS | 7 | 128 | 90.1 | 81.7 | 44.7 | 32.5 | 98.3 | 71 | 67 | 96 | 91 | NA | 38 | 211 (4.3) |
| VPS | 6 | 72 | 74.2 | 51.1 | 39.6 | 35.2 | 59.1 | 69 | 55 | 93 | 90 | NA | 41 | 53 (1.83) |
| Sinus stenting | 11 | 155 | 88.4 | 75.7 | 22.2 | 34,6 | 80.3 | 75 | 65 | 77 | 98 | 8 | 12 | NA |
| Bariatric surgery | 2 | 32 | 100 | 100 | 46.8 | 33.8 | 80.3 | 87.5 | NR | 96 | 100 | 10 | 87 | NA |

*HA* headache, *Impr* improved, *LPS* lumboperitoneal shunting, *ONSF* optic nerve sheath fenestration, *Papp* pappiloedema, *Pts* patients, *RPPR* revision per patient rate, *RR* relapse rate, *VA* visual acuity, *VF* visual field, *VPS* ventriculoperitoneal shunting

**Table 11** Approximate estimated cost of interventions

| Intervention | Approximate estimated cost | Details |
|---|---|---|
| LPS | £ 2716 or £ 4227[a] | • Hardware cost ≈ £ 624 or £ 2135*<br>• LOS cost ≈ 4 (days) × 273 ≈ £ 1092<br>• Cost of operative time (≈50′) ≈ £ 1000 |
| VPS | £ 4698 | • Hardware cost ≈ £ 2406<br>• LOS cost ≈ 4 (days) × 273 ≈ £ 1092<br>• Cost of operative time (≈60′) ≈ £ 1200 |
| LAGB | £ 5348 | • Hardware cost ≈ £ 3202<br>• LOS cost ≈ 2 (days) × 273 ≈ £ 546<br>• Cost of operative time (≈80′) ≈ £ 1600 |
| LRYGB | £ 9608 | • Hardware cost ≈ £ 4516<br>• LOS cost ≈ 4 (days) × 273 ≈ £ 1092<br>• Cost of operative time (≈ 200′) ≈ £ 4000 |
| ONSF | £ 873 | • Hardware cost ≈ £ 0<br>• LOS cost ≈ 1 (days) × 273 ≈ £ 273<br>• Cost of operative time (≈30’) ≈ £ 600 |
| Venous sinus stenting | £ 4690 | • Hardware cost ≈ £ 1200<br>• LOS cost ≈ [1 (HDU days) × 717] + [1 (days) × 273] ≈ £ 990<br>• Cost of operative time ≈ £ 2500 |

*LOS* length of hospital stay, *HDU* high dependency unit, *LAGB* laparoscopic adjustable gastric banding, *LRYGB* laparoscopic Roux-en-Y gastric bypass

[a] If a programmable valve is used

mean follow-up was 46.8 months. In 80.3 % (110/137) of patients, bariatric surgery was the primary surgical procedure performed. The techniques utilised were specified in all cases. Twenty-nine gastric bypasses (with several technical variations including 22 Roux-en-Y, 6 long limb, 1 distal gastric by-pass), two gastroplasties (1 horizontal, 1 vertical banded) and one laparoscopic adjustable gastric banding (LAGB) were performed.

Visual acuity and visual field data were provided by only one study [66]. Visual acuity was not affected in any patient while visual fields improved in 7/8 (87.5 %) and deteriorated in 1 patient (12.5 %). Papilloedema resolved in all patients and headache improvement was documented in 96 % of patients (31/32), with no patient deteriorating. Two out of 19 patients (10 %) relapsed. Thirty complications were reported in 28/32 patients (87 %) with the most common being incisional hernia (see Table 4).

*Secondary outcomes*

**Cost of intervention** Costs according to the principles clarified in the methods section are presented in Table 11. For bariatric surgery, costs for laparoscopic adjustable gastric banding (LAGB) and laparoscopic Roux-en-Y gastric bypass (LRYGB) are presented. This is only an estimation of the cost and not a cost-effectiveness study.

**Quality of life** Pertinent quality of life scores (such as HRQOL) were not utilised in any of the included studies. Consequently, data for quality of life outcome/endpoint were not extractable

## Discussion

This systematic review assesses surgical interventions used to manage IIH with respect to efficacy, complications and cost. Our review highlights the lack of robust evidence on the surgical management of IIH. Unfortunately, none of the surgical interventions has yet been examined in an adequately controlled trial. Forty-one (41) retrospective observational studies, 36 case series and 5 case reports were included. Weight status and gender demographics demonstrated high participation of obese (78.2 %) and female patients (85.7 %). However, only half of the VPS category patients are obese. The follow-up of studies was adequate, ranging between 39 and 46 months in all categories except sinus stenting where follow-up approximates 22 months. The most likely explanation for this difference is that sinus stenting is a relatively new modality in the treatment of IIH and, as a result, studies with long follow-up periods have not yet been published. In the majority of cases the studied intervention was the first-line surgical management to be utilised. Specifically, LPS was the first surgery performed in almost all cases. Furthermore, sinus stenting, although a recently adopted procedure, was the primary surgery in 8/10 patients. Of note, VPS was the first choice in only 6/10 patients. This significant divergence may be partially explained by the fact that, until recently, VPS was considered a more demanding procedure when compared to LPS because of the difficult cannulation of small-to-normal sized ventricles. However, the use of image-guided stereotaxy helped in overcoming the previous difficulties in the ventricular access of IIH patients [2, 6, 39].

Visual improvement was adequately achieved in all intervention categories. However, valid conclusions cannot be derived from the bariatric surgery cohort, as visual outcomes were not satisfactorily reported. Quantitative visual function outcomes, not unexpectedly, were better documented in the ONSF category, since ONSF is performed by ophthalmologists, who can independently and routinely monitor visual function (acuity and fields), as opposed to neurosurgeons or neurovascular interventionalists [26].

Headache satisfactorily improved in all intervention categories except for ONSF, where only minimal improvement was reported. Possibly, this can explain the established tendency towards CSF shunting when headache is the principal symptom. Papilloedema improved or resolved in all intervention categories. Only one patient deteriorated (in one eye) after LPS, despite a working shunt. Both pre- and post-operative CSF pressure measurements were reported in only eight studies (4 in sinus stenting, 3 in ONSF and 1 in the bariatric surgery category); thus, it is difficult to perform any comparisons.

The relapse rate was reported for ONSF, sinus stenting and bariatric surgery. Sinus stenting seems to have the lowest relapse rate. However, re-stenoses around intravascular stents at other sites have been documented in the literature [62]. Moreover, intraluminal thrombi requiring thrombolytic therapy were reported in two cases in our review. Consequently, longer follow-up periods are required to assess the durability of sinus stenting results. ONSF is normally performed once per optic nerve [31]. However, 15.9 % of operated eyes deteriorated with respect to visual acuity and visual fields after a seemingly successful initial procedure. In 69.8 % of these cases a second fenestration was required. Notably, some authors suggest that eyes that undergo more than one ONSF procedure are less likely to improve and are prone to vascular complications [54].

Sinus stenting seems to have the best complication profile in terms of affected/non-affected patients. Moreover, the majority of the complications were minor and transient, with headache (lateralised to the side of treatment) being the most common and attributed to the dural stretching by the stent [22]. However, serious complications were also reported. ONSF seems to have the second best complication profile and also transient complications. Distinct from other interventions, no foreign materials are implanted, thus reducing infection rates. The complication rate of bariatric surgery was high when compared to other treatment options with 87 % of patients developing a post-operative complication. Given that gastric bypass has a higher morbidity rate compared to the other weight-loss procedures (such as LAGB or sleeve gastrectomy), and given that gastric bypass was performed in almost all cases (29/32), this high rate may be partially misleading [19]. Both CSF diversion procedures are prone to a wide range of complications, with shunt obstruction being the most common for both LPS and VPS. Fifty-three VPS revisions were needed to treat 29 patients with complications, while 211 LPS revisions were performed in 49 patients. The "revision per patient rate" is therefore far higher for LPS than for VPS (4.3 vs. 1.83).

ONSF has the lowest cost, probably due to the fact that no foreign material/hardware is used. LPS is lower cost than VPS. However, preoperative selection of the LPS appropriate valve opening pressure is difficult. Moreover, it has recently been proposed that the use of programmable LPS systems may result in a lower incidence of shunt obstruction or symptomatic intracranial hypotension [9]. When programmable valves are used for LPS the cost approximates that for VPS. The cost of sinus stenting approximates that of VPS, while the cost of bariatric surgery is higher than for all other modalities.

This study reviewed the efficacy, complication profile and cost of the current surgical and interventional treatments for IIH, aiming at an evidence-based treatment algorithm. Common and simple treatment variants such as CSF shunting and ONSF have been investigated along with bariatric surgery and venous sinus stenting, as the newest options added to the IIH surgical armamentarium. It has to be specifically stressed that with regards to the aetiological correlation of venous sinus stenosis and IIH, current evidence is rather ambiguous as to whether sinus stenosis leads to intracranial hypertension due to compromised venous return (cause) or is the result of sinus compression from persistent intracranial hypertension (symptom) [31, 41, 56]. We have, therefore, included venous sinus stenting as a "symptomatic" treatment for IIH in view of new data. However, if IIH proves to be the aetiological outgrowth of venous sinus stenosis in a proportion of IIH patients, then the term "idiopathic" in these cases should be omitted and this clinical condition should be analysed and treated as a discrete entity.

## Suggested treatment algorithm

A tentative treatment algorithm is shown in Fig. 2. Bariatric surgery might be considered earlier (than depicted in the algorithm) if patients do not present with acute or rapidly progressive visual loss, where interventions such as ONSF, CSF shunting and venous sinus stenting may be more appropriate. Achieving a crucial, for symptom relief, weight loss through

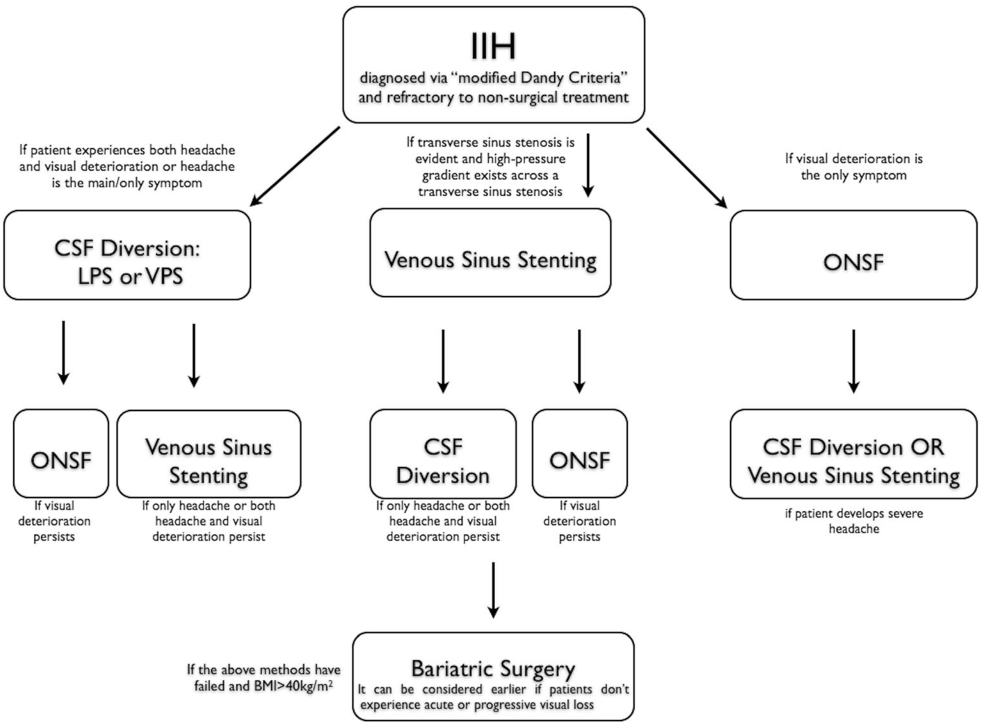


**Fig. 2** Suggested treatment algorithm

bariatric surgery occurs over time. Consequently, bariatric surgery may not be the best option in instances of acute or progressive visual loss [29]. Bariatric surgery should be a treatment option only in morbidly obese patients (patients with a BMI greater than 40 kg/m$^2$ or with BMI greater than 35 kg/m$^2$ combined with severe obesity-related co-morbidities, such as diabetes, hypertension and cardiovascular disease) [37]. The additional benefits of bariatric surgery (long-term weight loss, improvement of diabetes, hypertension, hyperlipidaemia and obstructive sleep apnoea) are worth noting [37]. Venous sinus stenosis and a given pressure gradient across the stenosis (usually >8 mmHg), evident by venography and manometry, are considered special criteria for patients undergoing venous sinus stenting. Moreover, dual antiplatelet therapy is required, pre- and post-procedurally, in almost all of the reviewed studies. Hence, it must be confirmed that the patient does not have any known contraindication to dual anti-platelet therapy to proceed to venous sinus stenting.

## Limitations of the review

This systematic review is based on class IV evidence studies (case series and case reports). Systemic biases (selection, detection, performance, attrition, reporting and publication) make retrospective observational studies the weakest study design for assessing the effects of interventions. However, these are currently the only available evidence for surgical management of IIH. Moreover, only two studies (32 patients in total) on bariatric surgery for obese patients with IIH were included in the present review, both of which were conducted by the same scientific group and therefore patient overlapping cannot definitely be ruled out. This issue makes meaningful comparisons between bariatric surgery and other treatments difficult.

Although measures used to assess vision were appropriate and homogeneous across studies (best corrected Snellen test for visual acuity, Goldmann perimetry or automated Humphrey visual field test for assessment of visual fields, fundoscopic examination for assessment of papilloedema and CSF opening pressure or ICP monitoring for measuring CSF pressure), the definition of improvement or worsening of specific outcomes, such as visual acuity and visual fields, was not consistent across different studies. Regarding headache relief, no details were provided on the methods used to define improvement or worsening (such as self-administered questionnaires or other standardised assessment tools).

Each study emphasised different outcomes and many of our review outcomes were not adequately reported in many studies. Furthermore, there were varying follow-up periods across studies.

Variations in surgical techniques (or modifications of standard ones) were employed across studies. This may influence outcomes (particularly the revision rate following the introduction of stereotaxy in the placement of VPS).

Lastly, LOS costs, hardware costs and operative time costs for the different procedures may vary significantly between countries or even hospitals of the same country and thus potentially affect the costs of the different procedures. Hence, it needs to be highlighted that this study provides only a rough estimation of cost.

## Conclusions

### Recommendations for practice

None of the available surgical treatments has been shown to be significantly superior. However, in certain IIH presentations, a given surgical approach appears more justified.

CSF diversion seems more appropriate when both headache and visual deterioration are present, while ONSF may prove a better choice when the cardinal symptom is visual deterioration, or if visual symptoms progress after CSF shunting. ONSF has an appealing complication profile and a lower cost.

Deciding between VPS and LPS is difficult. Though VPS has a better complication profile in terms of revision per patient, it is technically more difficult to cannulate the small-to-normal sized ventricles seen in IIH. Consequently, the expertise of the surgeon and the available technological adjuncts, such as stereotaxy and neuronavigation, play an important role. Moreover, VPS is generally more expensive, yet, when programmable valves are used for LPS, the cost differences are eliminated.

Venous sinus stenting is a relatively new treatment option for IIH and can be utilised when transverse sinus stenosis and a given pressure gradient across the stenosis are confirmed. It is the most promising treatment modality given the remarkable results in terms of both vision and headache along with the best complication profile and low relapse rate. Consequently, the treatment paradigm may need to be re-examined with sinus stenting as a first-line treatment modality. Possible superiority of this particular treatment modality needs to be examined with longer follow-up studies.

The available data support the use of bariatric surgery in morbidly obese IIH patients without acute or rapidly progressive visual loss. This treatment option can be particularly useful in cases with obesity-related co-morbidities (such as diabetes and hypertension). Studies with better documentation on visual outcomes are needed.

### Recommendations for research

This review highlights that robust evidence for the surgical treatment of IIH is lacking. It is crucial to design prospective randomised controlled trials in order to better determine the efficacy of surgical interventions for IIH treatment. We recommend the following parameters to be appreciated and included in future studies assessing IIH:

- Reporting gender/age, weight status, indications for surgery, previous treatments and surgical techniques.
- Pre- and post-operative quantitative assessment of visual acuity via best corrected Snellen acuity scale, visual fields through automated Humphrey programmes, categorisation of papilloedema according to the Frisen scale, CSF pressure measured in the lateral decubitus position and headache response according to self-administered questionnaires (such as the visual analogue scale to record the pain intensity and HIT-6 to record the impact of headache in everyday activity).
- Documentation of relapse rate and follow-up/loss to follow-up, complications and their management with pertinent tables.

**Compliance with ethical standards**

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**Conflict of interest** None

**Ethical approval** For this type of study formal consent is not required.

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## Comments

Kalyvas and co-workers provide a systematic review of the literature about “Efficacy, complications and cost of surgical interventions for idiopathic intracranial hypertension”. I have read the paper with great interest and congratulate the authors for this—in my opinion—important review. The limitation of difficulties in comparing lengths of hospital stay, hardware costs and operative time costs in different hospitals and even more in different countries has been adequately discussed.

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