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Review

Visual outcomes and headache following interventions for idiopathic intracranial hypertension

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ABSTRACT

The optimal surgical management for medically refractory idiopathic intracranial hypertension (IIH) is not well established. Few studies have directly compared headache and visual outcomes across treatment modalities. A systematic analysis of case series was conducted to compare therapeutic efficacies among currently available interventions. The electronic databases from EMBASE (1980–17 September 2013), Medline (1980–17 September 2013), Cochrane databases, and references of review articles was searched. All publications reporting headache and visual outcomes following intervention for IIH were included. A total of 457 manuscripts were selected and full text analysis produced 30 studies with extractable data. All studies constituted Class III evidence. Overall, 332 patients treated by optic nerve sheath fenestration (ONSF), 287 by lumboperitoneal shunt (LPS), 61 by ventriculoperitoneal shunt (VPS), and 88 by dural venous sinus stenting, were identified. Visual acuity improved in 49.3%, 56.6%, 67.2% and 84.6% of patients following VPS, LPS, ONSF, and stent placements, respectively. Resolution of papilledema was noted in 59.9% to 97.1%. Postoperative headache improved in 36.5%, 62.5%, 75.2%, and 82.9% of patients treated with ONSF, VPS, LPS, and stenting, respectively. Shunt revision was more frequent for LPS compared to VPS (46% *versus* 36%; $p < 0.2$). Among the LPS revisions, 87.5% occurred within the first 12 months following initial surgery. Our pooled analysis indicated an overall similar improvement in visual outcomes across treatment modalities, and a modest improvement in headache following cerebrospinal fluid shunting and endovascular stent placement. Based on currently available literature, there is insufficient evidence to recommend or reject any treatments modalities for IIH.

1. Introduction

Idiopathic intracranial hypertension (IIH) is an uncommon disorder characterised by raised intracranial pressure without an intracranial mass [1]. The incidence in both adolescents and adults directly correlates with the prevalence of obesity [2]. Onset can occur at any age but is most frequent between 20 and 40 years of age [3]. Diagnosis of IIH requires the demonstration of (1) symptoms and signs referable only to elevated intracranial pressure; (2) cerebrospinal fluid (CSF) opening pressure >25 cm H_2O measured in the lateral decubitus position; (3) normal CSF composition; and (4) no evidence for an underlying structural cause (using MRI or contrast-enhanced CT scan for typical patients and MRI and MR venography for all others) [4,5].

Most patients with IIH respond to maximal non-operative therapy such as weight loss, repeat lumbar punctures, and medical treatments. Surgical intervention for IIH may be necessary in circumstances of failed medical treatment, or progressive visual loss or intractable headache despite maximal medical therapy. Procedures that are available include the placement of a lumboperitoneal shunt (LPS) [6–9], ventriculoperitoneal shunt (VPS) [10–13], optic nerve sheath fenestration (ONSF) [14–18], or the insertion of a dural venous sinus stent [19–25]. Surgical decision-making is confounded by a lack of clear and robust outcome data across currently available treatment modalities [3,26]. The therapy of choice depends, in part, on local availability and expertise, and the biases of the treating physician [27].

To our knowledge, there are no evidence-based studies in which the risks and benefits of various interventions are assessed. The purpose of the current study was to review the existing literature to compare the efficacy and complications of ONSF, LPS, VPS and dural venous sinus stent placement in the management of IIH.

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2. Methods

A systematic review of published literature was performed for the primary outcome of visual function and headache improvement following interventions for patients with medically refractory IIH. A secondary outcome analysis was performed for treatment related complications and the incidence of shunt revisions among the LPS and VPS cohorts. A Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA; www.prisma-statement.org) style was adhered to where possible but quality assessment was not conducted, as the target study types were case series and cohorts.

2.1. Eligibility criteria

Included studies were reviewed and carefully scrutinised for study design, methodology, patient characteristics, and primary findings. Only manuscripts reporting original data on patients undergoing ONSF, LPS, VPS, and dural venous sinus stent placement were considered in the analysis. To minimise selection bias, we included only studies of 10 or more patients that reported on perioperative outcomes. Trials comprised subjects of any age, with any co-morbidity, and with varied duration of follow-up. To be considered in the analysis, patients had to meet the modified Dandy criteria for IIH [28]. Review articles were used only to extract any additional articles not found within the initial search and were not included as separate articles. Attempts were made to eliminate redundant descriptions of the same patient series. Only studies pertaining to IIH treatment outcomes were considered. A few series included combined outcome data for both LPS and VPS. In these cases, care was taken to extract only information relevant to the individual treatment modality.

2.2. Search criteria

The Medline database was searched from January 1980 to 17 September 2013, and the EMBASE database was searched from January 1980 to 17 September 2013. The Cochrane Collaboration database and the UK National Health Service Evidence Health Information Resources website were also searched. The bibliographies of identified manuscripts were reviewed for additional data sources. No unpublished trials were included. We designed a search strategy to include manuscripts relevant to any aspect of treatment outcomes following interventions for IIH. The search strategy used for EMBASE and Medline databases is shown in Table 1.

Studies were selected in an un-blinded standardised manner once the searches were completed. The publications extracted were grouped by title and duplicates were excluded. The abstracts were then reviewed to ascertain whether they met the inclusion and exclusion criteria as described above.

2.3. Data extraction

Standardised data sheets were used for each study. The primary outcomes were recorded as postoperative improvement in headache, visual acuity (VA), visual fields (VFs) and papilloedema. Secondary outcome measures focused on treatment related complications for each type of intervention. For outcomes related to shunt procedures, the number of patients, the perioperative morbidity, the duration of follow-up, the number of recurrent cases and the time taken to recurrence was recorded. Where duplicate publication was anticipated from centres re-publishing updated reports on their surgical experience over time, the most recent or largest published data was included for analysis in the current study.

2.4. Statistical analysis

Statistical assessments were performed primarily with descriptive data. Data from the individual studies were combined by cohort and then compared using chi-squared and Fisher's exact tests as appropriate. Where data extraction on the VA outcome was obtainable, the average VA from Snellen/Decimal equivalent was converted to the logarithm of the minimum angle of resolution (logMAR) value [29]. This enabled the comparison of standardised VA outcomes among studies. Significance was set to a probability value of 0.05. The modified Wald method was used to calculate the 95% confidence intervals (CI) for a proportion (GraphPad Software, La Jola, CA, USA). Data were analyzed using Statistical Package for the Social Sciences version 19 (SPSS Inc., Chicago, IL, USA).

3. Results

3.1. Literature review results

The search of EMBASE and Medline produced a total of 456 studies written in English. Additional records identified through the Cochrane database yielded one further study that was included in the analysis, totalling 457 studies. After exclusion of duplicates and irrelevant records, 307 studies remained. A title search found 169 articles on interventions for IIH. Those studies captured in the search that were technical notes (n = 34), reviews (n = 21), commentaries (n = 15) and case reports (n = 27) were excluded from the analysis. This selection process is outlined in Figure 1.

The remaining 73 articles reporting treatment outcomes following interventions for IIH were subjected to full-text assessment. Of these, 19 contained samples of fewer than 10 patients and four described only outcomes following medical treatments. These manuscripts were further excluded.

Perioperative outcomes were recorded for 50 studies. An additional 20 studies were excluded due to non-extractable data. Thirty studies with reports of post-treatment visual and headache outcomes following interventions for IIH were included in the final analysis.

3.2. Primary outcome: Visual outcomes and headache

ONSF is generally considered when visual function is threatened and cannot be stabilised with medical options and headache is not a significant symptom. Quantitative analysis revealed a total of 11 clinical studies related to ONSF, encompassing 332 patients with 508 eyes treated [14–18,30–35] (Table 2). The mean age was 31.2 years (range 4.4 to 74.0 years) and the mean follow-up was 38.9 months (range 0.1 to 700 months). Females accounted for 81.9% of the patients (272/332 patients). Following treatment with ONSF, VA was reported to be stable in 24.2% (114/472 eyes) and improved in 67.2% (317/472 eyes). Five studies contained extractable outcome data comparing the preoperative and postoperative mean VA. A total of 97 patients with 134 eyes treated by ONSF were included in the analysis. The preoperative mean VA improved from 0.31 ± 0.10 logMAR (Snellen 6/12) to 0.28 ± 0.25 logMAR (Snellen 6/11) in the postoperative period [30–33,35]. VF was stable in 13.4% (39/291 eyes) and improved in 71.8% (209/291 eyes). Of the two studies that reported serial Humphrey VF outcome data, the mean preoperative mean deviation (MD) improved from −15.5 dB to a mean postoperative MD of −9.1 dB [32,35]. Two studies (43 patients with 50 eyes treated) reported