DIOPATHIC intracranial hypertension is a syndrome consisting of elevation in intracranial pressure without evidence of structural lesion or alteration of CSF. Symptoms and signs must be attributable only to increased intracranial pressure or papilledema.

The most common manifestations of the disease are headache and an assortment of visual symptoms, including transient obscurations of vision, diplopia from abducent nerve paresis, and visual loss associated with the effects of papilledema. Pulsatile tinnitus may also be present. Most commonly occurring in obese women,\textsuperscript{11,35} this syndrome has also been called pseudotumor cerebri and benign intracranial hypertension.

As the term IIH connotes, the etiology of this entity is unknown, although several hypotheses exist. An increase in CSF secretion or a decrease in CSF absorption caused by unknown hormonal or proinflammatory mediators has been suggested. Another concept is based upon increased intracranial venous pressure due either to transmission of high intraabdominal pressure or to relative (or functional) stenosis of the intracranial venous sinuses. These concepts are presented and discussed in an excellent review by Friedman.\textsuperscript{15}

Multiple therapeutic alternatives are available for management of IIH. Medical therapy is directed toward reducing CSF secretion and increasing CSF absorption with diuretics such as furosamide and carbonic anhydrase inhibitors such as acetazolamide.\textsuperscript{9,41} Weight loss, through diet or surgical intervention, is advocated to reduce intraabdominal pressure or change secretion of offending but unknown mediators.\textsuperscript{21,25,40} Cerebrospinal fluid diversion procedures address the effects of IIH rather than purported causes, simply draining excess CSF out of the subarachnoid space. Such procedures include placement of LP shunts,\textsuperscript{5,11,31} VP shunts,\textsuperscript{5,29} and foramen magnum–atrial shunts,\textsuperscript{22} as well as subtemporal decompressions. Optic nerve sheath decompression may work in part through CSF diversion and in part by blocking transmission of intracranial pressure to the optic disc at the level of the lamina.
cribrosa. Based upon the concept that there is functional or actual stenosis of the intracranial venous outflow, intravascular stenting of the venous sinuses has been advocated as yet another mode of treatment. Primary indications for the treatment of IIH are intractable headache and vision loss. Unfortunately, there are no evidence-based studies in which the risks and benefits of various interventions are assessed. Furthermore, the relative rarity of severe complications of IIH and the variability in presentation make prospective clinical trials very problematic. Thus, in the absence of a scientifically demonstrated advantage for any single treatment, the indications for a particular therapy differ widely depending upon the biases of the treating physicians. Although there may be some advocates of primary surgical therapy, the consensus is that maximum tolerated medical therapy (diuretics and weight loss) should be tried before invasive options are considered. Case studies suggest that ONSD is generally effective for managing visual loss, whereas diversion and perhaps stent placement procedures may be more beneficial for managing headache. One reason for this impression might be that ophthalmologists follow up patients with visual loss and, therefore, are more likely to suggest a procedure performed by ophthalmologists. By similar reasoning, neurologists would then be more likely to see patients with headache and recommend a neurosurgical or neurovascular approach. All interventions are associated with known risks, of which the most likely is a failure to sustain benefit.

The purpose of the present study is to directly compare the published experience of surgical techniques commonly advocated for treatment of visual loss in IIH. This analysis is intended to investigate the visual benefit of LP shunts, VP shunts, and venous sinus stents, relative to ONSD. A further intention is to recommend improvements in data collection that will increase the clinical value of future studies assessing visual outcomes following surgical management of IIH.

Clinical Material and Methods

Peer-reviewed published articles were retrieved using online MEDLINE searches. Key words used in the primary search included the following: idiopathic intracranial hypertension, pseudotumor cerebri, and benign intracranial hypertension. Subsidiary searches were conducted for lumbo-peritoneal shunts, ventriculoperitoneal shunts, optic nerve sheath decompression and intracranial venous stents. Web of Science Citation Index searches and reference reviews were conducted on relevant recovered articles. Series or cases that did not include vision data were not considered. Excluded from analysis of ONSD were individual case reports, series of less than 10 cases, topic reviews, and articles not written in English. One series was excluded because publication dates suggested overlapping cohorts.

Because of a paucity of reports, individual cases and small series in the nonpediatric age group were included if they were related to stents and CSF diversion techniques and if there were vision data available. Articles providing summary data only (no individual case data given) were excluded. Cases involving venous sinus occlusion or other secondary causes of increased intracranial pressure within an included series were excluded from the analysis.

For ONSD, summary data were combined to develop an aggregate of experience with the procedure. For all other surgical interventions, data available for the individual cases in each series were combined in order to assess the cumulative reported experience with each procedure. The authors of one report of a series of cases in which ONSD was performed subcategorized cases as acute and chronic. The data for chronic IIH were evaluated separately from data pertaining to acute cases.

For all studies in which multiple time points were reported for visual outcome, the 6-month data were utilized. Criteria for visual improvement or deterioration were based upon the investigators’ definitions. If no definition was provided, any reported change was included. Transient obstructions of vision and enlarged blind spots on visual field testing, when specified, were not considered to constitute vision loss.

Results

Optic Nerve Sheath Decompression

Seven retrospective series were identified which met the inclusion criteria for the study. Seven retrospective series were identified which met the inclusion criteria for the study. In all, 423 eyes of 252 patients were evaluated with a mean follow-up of 21.1 months (range 0–121 months). The patients’ mean age was 33.6 years (range 6–72 years), and 83% of the patients were female. Of interest, the majority (59%) of surgeries were bilateral. In 90% (226) of cases, ONSD was the first surgery performed. Visual acuity was improved in 50% of eyes (113) and visual field was improved in 72% of eyes (226). Visual acuity or visual field worsened in only 42 cases (11%). The reoperation rate was 12%, with repeated ONSD being the most common procedure (6%) followed by CSF diversion procedure (4%). Results are summarized in Table 1. Note that not all data was available for all cases; thus, denominators used in calculating percentages differed.

Intracranial Venous Sinus Stents

Stent placement was the least frequently reported of all surgical procedures advocated for management of IIH. Only 17 cases, followed up for an average of 11.8 months (range 5–24 months), met the inclusion criteria. Indications for surgery included both headache and vision problems in 12 cases (70.6%), vision problems only in 1 case (5.9%), and headache in 4 (23.5%). The average age was 30 years with a range of 15–52 years. Fifteen (88.2%) were women and all were obese, with a mean BMI of 33.9 (the threshold for obesity being an index greater than 25 being defined as obese). In the majority of cases (58.8%), stent placement was the first procedure performed. Resolution of papilledema occurred in 8 cases (47.1%), improvement in papilledema occurred in 1 case (5.9%), and there was no change in papilledema in 5 cases (29.4%). In 3 cases, information regarding disc swelling was unavailable.

Headaches resolved in 8 cases (47.1%) and improved in 2 others (11.8%). Headaches persisted without change in 4 cases (23.5%), and no information was available regarding headache severity in 1 case. Vision recovered completely in
### TABLE 1

*Summary of articles on ONSD for treatment of IIH*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Pts</th>
<th>No. of Eyes</th>
<th>Mean Age in Yrs (range)</th>
<th>M</th>
<th>F</th>
<th>Surgery</th>
<th>Mean FU in mos (range)</th>
<th>Acuity Impr or Same</th>
<th>Acuity Impr</th>
<th>Field &amp;/or Impr</th>
<th>Acuity Worse</th>
<th>Subsequent Procedures</th>
</tr>
</thead>
<tbody>
<tr>
<td>Corbett et al., 1988</td>
<td>28</td>
<td>40</td>
<td>30 (14–62)</td>
<td>8</td>
<td>20</td>
<td>12</td>
<td>26.9 (0–90)</td>
<td>34</td>
<td>12</td>
<td>21</td>
<td>13</td>
<td>2 diversions</td>
</tr>
<tr>
<td>Sergott et al., 1988</td>
<td>23</td>
<td>29</td>
<td>38.1 (18–63)</td>
<td>1</td>
<td>22</td>
<td>6</td>
<td>21.5 (3–45)</td>
<td>23</td>
<td>21</td>
<td>29</td>
<td>1</td>
<td>2 rpt ONSD</td>
</tr>
<tr>
<td>Spoor et al., 1991</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>35</td>
<td>44</td>
<td>68</td>
<td>0</td>
<td>rpt ONSD (16 in 13 eyes)</td>
</tr>
<tr>
<td>chronic cases</td>
<td>18</td>
<td>32</td>
<td>32.7 (7–57)</td>
<td>5</td>
<td>13</td>
<td>5</td>
<td>14.6 (3–46)</td>
<td>32</td>
<td>14</td>
<td>7</td>
<td>0</td>
<td>1 rpt ONSD</td>
</tr>
<tr>
<td>Acheson et al., 1994</td>
<td>11</td>
<td>15</td>
<td>37.1 (23–53)</td>
<td>3</td>
<td>8</td>
<td>4</td>
<td>24 (12–84)</td>
<td>4</td>
<td>9</td>
<td>8</td>
<td>2</td>
<td>4 diversions; 1 sub-temp decompr</td>
</tr>
<tr>
<td>Goh et al., 1997</td>
<td>19</td>
<td>29</td>
<td>33.1 (16–52)</td>
<td>6</td>
<td>13</td>
<td>10</td>
<td>15.7 (1–50)</td>
<td>15</td>
<td>9</td>
<td>2</td>
<td>2</td>
<td>4 rpt ONSD</td>
</tr>
<tr>
<td>Banta &amp; Farris, 2000</td>
<td>86</td>
<td>158</td>
<td>32.1 (NA)</td>
<td>13</td>
<td>73</td>
<td>72</td>
<td>20 (1–108)</td>
<td>148</td>
<td>71</td>
<td>16</td>
<td>1</td>
<td>4 rpt ONSD &amp; diversion; 2 diversion only; 2 ONSD only</td>
</tr>
<tr>
<td>Chandrasekaran et al., 2006</td>
<td>32</td>
<td>51</td>
<td>33.4 (17–65)</td>
<td>3</td>
<td>29</td>
<td>18</td>
<td>27.6 (0–121)</td>
<td>32†</td>
<td>13†</td>
<td>13‡</td>
<td>8§</td>
<td>11 diversions</td>
</tr>
<tr>
<td>summary</td>
<td>252</td>
<td>423</td>
<td>33.6 (6–72)</td>
<td>43</td>
<td>209</td>
<td>209</td>
<td>21.1 (0–121)</td>
<td>357</td>
<td>113</td>
<td>226</td>
<td>42</td>
<td>26 rpt ONSD (6%); 19 diversions (4%); 1 subtemp decompr (&lt;1%); 4 ONSD &amp; diversion (1%); Total: 12% reop rate</td>
</tr>
</tbody>
</table>

* decomp = decompression; FU = follow-up; Impr = improvement; pts = patients; rpt = repeated; subtemp = subtemporal; NA = data not available.
† Based upon 39 eyes (limited follow-up data available).
‡ Based upon 31 eyes.
§ Based upon 39 eyes for acuity and 31 eyes for visual field.
3 cases (17.6%) and improved in 5 others (29.4%). There was no change in vision in 4 cases (23.5%), and no information regarding vision change was available in 4 others. Results are summarized in Table 2.

**Ventriculoperitoneal Shunts**

Reports of 2 case series and 2 single cases were identified in which VP shunts were utilized for treatment of IIH and the criteria for inclusion in the study were met. The mean age of the 31 patients was 32.4 years (range 6–63 years), and 23 (74.2%) were female. Average follow-up was 48.3 months (range 1–153.6 months). The mean BMI was 31.1 for the 18 cases for which information was provided. Placement of a VP shunt was the primary surgical procedure in 16 cases (51.6%). Papilledema resolved in all of the 17 cases for which data pertaining to that condition were reported. Headache resolved in 1 case and improved in another, but information regarding this symptom was not available for 29 patients. Vision was noted to improve in 12 cases (38.7%). In the majority of patients, there was no change in vision (19 cases, 61.3%). Results are summarized in Table 3.

**Lumboperitoneal Shunts**

Reports of 2 case series and 2 single cases were identified in which LP shunts were used for treatment of IIH the criteria for inclusion in the study were met. Average follow-up was 58.7 months (range 0.2–278 months). The 44 patients' mean age was 30.7 years (range 19–52 years), and 92.9% were women, but in 1 series of 16 cases, the age and sex of individual patients was not reported. The BMI was provided only in the 2 case reports and was 27 in each. Placement of an LP shunt was identified as the primary surgical intervention in 27 (96.4%) of 28 cases for which information was provided.

Papilledema resolved in 10 cases (22.7%) and improved in another 7 (15.9%), but information was not available for the majority of patients (27 cases [61.4%]). Headache resolved in 8 cases (18.2%) and improved in another 12 (27.3%). Again, information was not provided in the majority (24 cases [54.5%]). Vision symptoms or signs resolved in 7 cases (15.9%) and improved in 13 (29.5%). There was no change in vision in 7 cases (15.9%) and worsening occurred in 2 (4.5%). Results are summarized in Table 4.

**Discussion**

Surgical procedures are commonly utilized for treatment of IIH when medical treatments fail or are poorly tolerated. However, this review demonstrates that when one excludes the data on ONSD (252 cases), there is a paucity of data regarding the effect of various surgical interventions on vision (17–44 cases, depending on the intervention). The most likely explanation for the difference in data collection between ONSD and other interventions is that ONSD is performed by ophthalmologists who routinely monitor visual function, whereas the other surgeries are performed by neurosurgeons or neurovascular interventionists, who depend upon others for visual assessment. In studies other than those on ONSD, either the visual function of patients may not be readily available to the investigators or visual outcome may not be the primary thrust of the study.
### TABLE 3

**Summary of articles on VP shunt surgery for treatment of IIH***

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Pts</th>
<th>Mean Age in Mos (range)</th>
<th>M (%)</th>
<th>F (%)</th>
<th>Mean BMI</th>
<th>Primary Surgery</th>
<th>Symptoms</th>
<th>Mean FU in Mos (range)</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maher et al., 2001</td>
<td>13</td>
<td>31.9 (6–45)</td>
<td>3 (23)</td>
<td>10 (77)</td>
<td>—</td>
<td>0</td>
<td>B: 3</td>
<td>12.4 (1–31)</td>
<td>—</td>
</tr>
<tr>
<td>Owler et al., 2003</td>
<td>1</td>
<td>16</td>
<td>0</td>
<td>1 (100)</td>
<td>34</td>
<td>0</td>
<td>H</td>
<td>11</td>
<td>R</td>
</tr>
<tr>
<td>Bynke et al., 2004</td>
<td>16</td>
<td>34.0 (13–63)</td>
<td>5 (31.3)</td>
<td>11 (68.7)</td>
<td>30.8</td>
<td>16 (100%)</td>
<td>B:14</td>
<td>77.9 (21.6–153.6)</td>
<td>R: 16</td>
</tr>
<tr>
<td><strong>summary</strong></td>
<td>31</td>
<td>32.4 (6–63)</td>
<td>8 (25.8)</td>
<td>23 (74.2)</td>
<td>31.1†</td>
<td>16 (51.6 %)</td>
<td>B: 18 (28.1%)</td>
<td>48.3 (1–153.6)</td>
<td>R: 17 (54.9%)</td>
</tr>
</tbody>
</table>

* — = not reported.
† Calculated on the basis of 18 cases.

### TABLE 4

**Summary of the literature concerning response to chemotherapy in optic pathway gliomas***

<table>
<thead>
<tr>
<th>Authors, Year</th>
<th>Chemotherapy Regimen</th>
<th>Median FU Duration</th>
<th>Response to Tx</th>
</tr>
</thead>
<tbody>
<tr>
<td>Packer et al., 1988 (24)</td>
<td>vinc/AD</td>
<td>4 yrs</td>
<td>stable disease (14)</td>
</tr>
<tr>
<td>Petronio et al., 1991 (15)</td>
<td>TPDVL</td>
<td>67.3 wks</td>
<td>stable disease (3)</td>
</tr>
<tr>
<td>Packer et al., 1993 (30)</td>
<td>car/vinc</td>
<td>14 mos</td>
<td>stable disease (10)</td>
</tr>
<tr>
<td>Janss et al., 1995 (32)</td>
<td>AD/vinc (31), VP16/vinc (1)</td>
<td>72 mos</td>
<td>stable disease (9)</td>
</tr>
<tr>
<td>Sutton et al., 1995 (10)</td>
<td>AD/vinc</td>
<td>3 yrs</td>
<td>stable disease (10)</td>
</tr>
<tr>
<td>Aquino et al., 1999 (12)</td>
<td>car</td>
<td>38.6 mos</td>
<td>stable disease (6), partial response (4)</td>
</tr>
<tr>
<td>Mahoney et al., 2000 (50)</td>
<td>car</td>
<td>18 mos</td>
<td>39/50 stable or better</td>
</tr>
<tr>
<td>Silva et al., 2000 (14)</td>
<td>car (8), car &amp; vinc (4), other (2)</td>
<td>15 mos to 8 yrs</td>
<td>5-year progression-free survival 63%</td>
</tr>
<tr>
<td>Mitchell et al., 2001 (12)</td>
<td>monthly car</td>
<td>not applicable</td>
<td>stable radiologic disease (9)</td>
</tr>
<tr>
<td>Gnekow et al., 2004 (123)†</td>
<td>vinc/car</td>
<td>22.5 mos</td>
<td>progression-free survival 61%</td>
</tr>
</tbody>
</table>

* All numbers in parentheses are numbers of patients. Abbreviations: AD = actinomycin-D; car = carboplatin; FU = follow-up; TPDVL = 6-thioguonine, procarbazine, dibromodulatol, vincristine, lomustine; Tx = treatment; vinc = vincristine; VP16 = etoposide.
† Patients had a median age of 3.7 years.
Difficulties in interpreting the results of this study are numerous and include the following: retrospective case series and case reports, small sample sizes, incomplete and variable data collection, differing definitions for meaningful change in outcome, differing indications for surgery, varying duration of postoperative assessment. Despite the considerable limitations of the present analysis, direct comparisons of differing surgical techniques may provide some insights into patient selection as well as beneficial or deleterious effects of treatment. All the case series report on patients with remarkably similar demographics, with a high percentage of women (59–88%) of similar age (average range 30.4–33.6). Although outcome data was reported at 6 months as a rule, the mean duration of follow-up was much longer for VP and LP shunt studies (47.3 and 57.2 months, respectively) compared to studies of stent placement (11.8 month) and ONSD (21.1 months) (Table 5). Shunt placement procedures may be more prone to failure or in other ways have more complicated courses, perhaps explaining the longer follow-up periods. The use of stents for treatment of IIH is relatively new, so follow-up is expected to be of shorter duration.

Optic nerve sheath decompression seems to have been a primary procedure in nearly all cases (90%) in which it was performed, whereas placement of a shunt or stents was as a primary procedure in just over half the cases in which it was performed (range 52–58.8% of cases). Although there are instances of poor visual outcome following surgical procedures for treatment of IIH, optic nerve sheath fenestration in pseudotumor cerebri and related conditions.

### Conclusions

One important conclusion from the current analysis is that better data are needed if a rational management plan is to emerge for the surgical treatment of IIH. Although there is always a clamor for randomized, prospective clinical trials to assess visual outcomes following surgery, practical issues including sample size and expense make an evidence-based approach unlikely. In the absence of a prospective clinical trial, more case series may provide data that will be helpful to clinical decision-making. Such series should include careful preoperative and postoperative documentation of visual function and optic disc appearance. Recommended functional measures include best corrected visual acuity, color vision, and computerized visual fields. Despite many caveats, in the absence of a more definitive study, the current analysis suggests that ONSD is the preferred primary surgical technique for managing the visual symptoms and signs of IIH resistant to medical therapy.

### References

Visual outcomes in surgical management of IIH


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Address correspondence to: Steven E. Feldon, M.D., 210 Crittenden, Box 659, Rochester, New York 14642. email: Steven_Feldon@urmc.rochester.edu.