Internal cranial expansion surgery for the treatment of refractory idiopathic intracranial hypertension

Clinical article

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Object. Idiopathic intracranial hypertension (IIH) may be refractory to available medical and surgical therapies. Patients with this condition may suffer from intractable headaches, experience visual deterioration, or have other symptoms related to elevated intracranial pressure. Internal cranial expansion (ICE) is a novel surgical procedure that the authors have developed for the treatment of patients with this condition. Here, they describe ICE and present their initial experience in using this surgical procedure for the treatment of patients with refractory IIH.

Methods. The authors conducted a retrospective review of 10 consecutive patients who underwent ICE for the treatment of IIH during a 5-year period. Preoperative and postoperative clinical parameters including patient symptoms, presence of papilledema, and available ICP or CSF opening pressures were compared. Procedural details and complications were noted. Intracranial volume increases were calculated using available pre- and postoperative CT scans.

Results. Follow-up for the 10 patients in this series ranged from 1 to 39.6 months (mean 15.5 months). Technically successful ICE was performed in all patients within the cohort. Surgical complications included a single postoperative seizure in one patient and a sagittal sinus tear with no clinical sequelae in another patient. At the time of last follow-up, 7 (70%) of 10 patients were either symptomatically improved or asymptomatic. Six (67%) of 9 patients with preoperative headaches had reduction or resolution of this symptom, and all patients (4 of 4) with preoperative papilledema had a reduction in or complete resolution of this sign. Postoperative ICP or CSF opening pressures were normal in all patients (4 of 4) tested. Postoperative intracranial volume expansion ranged between 3.8% and 12%.

Conclusions. Internal cranial expansion is a safe and effective surgery for the treatment of patients with refractory IIH. This surgery expands the intracranial volume and thus promotes ICP normalization, which may lead to the reduction or complete resolution of the signs and symptoms of IIH. Internal cranial expansion may be used as part of a multidisciplinary management approach in the treatment of refractory IIH.

KEY WORDS • idiopathic intracranial hypertension • craniotomy • headache • papilledema • pseudotumor cerebri • technique

Abbreviations used in this paper: BMI = body mass index; ICE = internal cranial expansion; ICP = intracranial pressure; IIH = idiopathic intracranial hypertension; LP = lumboperitoneal; ONSF = optic nerve sheath fenestration; VP = ventriculoperitoneal.

This article contains some figures that are displayed in color online but in black-and-white in the print edition.
Internal cranial expansion for intracranial hypertension

nally described by our group for the treatment of Camu- 
rati-Engelmann disease and subsequently modified for 
the treatment of slit-ventricle syndrome. ICE involves 
removal of the skull’s inner calvarial table and cancell-
ous bone to effect an increase in the volume of the intra-
cranial compartment (Fig. 1). This increased intracranial 
volume enables expansion of the intracranial contents 
(brain, blood, and CSF) and thus decreases ICP. Here, we 
describe our initial experience with ICE for the definitive 
surgical treatment of patients with refractory IIH. The 
surgical technique is presented with delineation of com-
plications and clinical outcomes. We also discuss how our 
results may shed light on the etiology of this heretofore 
poorly understood condition.

Methods

Patient Population

Between 2007 and 2011, ICE was performed in 10 
consecutive patients diagnosed with IIH that was refrac-
tory to medical treatment and, in some cases, other sur-
gical treatment. The clinic chart, electronic medical re-
cords, and available radiographic studies for each patient 
were retrospectively reviewed. Baseline clinical charac-
teristics, prior surgical interventions, perioperative de-
tails, available pre- and postoperative ICP measurements, 
and clinical assessment at the last follow-up clinic visit 
were noted.

Image Analysis

Preoperative and postoperative noncontrast head CT 
scans were obtained for volumetric analysis in 4 patients. 
Similar to methodology previously described, we 
used the public domain image processing program ImageJ 
(1.44p, National Institutes of Health) to calculate 
intracranial volume change. Briefly, CT image stacks 
composed of raw DICOM files were opened within ImageJ. 
Scale and slice thickness were set for each stack based 
on metadata contained within each DICOM file. Using 
bone windows, automated selection and calculation of the 
intracranial compartment area were performed for each 
slice. All intracranial compartment area selections were 
visually confirmed to be bound by the inner table of the 
skull. An intracranial compartment slice volume was cal-
culated by multiplying the intracranial compartment area 
(A) by the slice thickness (T) as follows: \( V_{\text{slice}} = A_{\text{slice}} \times T_{\text{slice}} \). 
The total intracranial compartment volume (\( V \)) was 
calculated by summing individual intracranial compartment 
slice volumes from the skull vertex to the foramen 
= \( \sum V_{\text{slice}} \).

Surgical Procedure

Although it has been previously described, the 
ICE procedure is elaborated here for the benefit of the 
reader. The patient is positioned supine with the head 
neutral or slightly flexed on a horseshoe headrest. Alter-
natively, rigid cranial stabilization using 3-point fixation 
is used. The scalp is prepared in a usual sterile fash-
on, and craniotomy drapes are placed. An extended zig-
zag coronal incision encompassing the retroauricular re-

gions bilaterally is then fashioned. The temporalis fascia 
is left intact, and the muscle is not taken down. The scalp 
is then reflected anteriorly and posteriorly, providing 
wide exposure of the cranium. Using a surgical marking 
pen, large bilateral craniotomies are outlined and num-
bered anterior and posterior to the coronal suture (Fig. 
2). Bony struts (1.5–2 cm wide) are maintained along the 
coronal suture and orthogonally over the sagittal sinus. 
The 4 outlined craniotomies are turned, and the flaps are 
taken to a sterile side table where another surgeon works 
to remove the inner cortical and cancellous layers using a 
drill, osteotome, and oscillating saw as needed. The inner 
table and cancellous layer of the struts and of the remain-
ing frontal, temporal, and parietal bones are drilled off as 
low as possible to the skull base. Entrance into the frontal 
sinus is avoided. Care is also taken to protect the under-
yling dura with a malleable brain retractor during drill-
ing. The thinned bone flaps from the side table are then 
replaced using titanium plates and screws, thus preserving 
the integrity and rigidity of the skull while expanding the 
intracranial volume (Fig. 3). A Jackson-Pratt drain is 
placed in the subgaleal space, and the scalp is closed in the 
usual fashion.

Results

Clinical Characteristics

The study cohort includes 3 male and 7 female pa-
tients who ranged in age from 11 to 61 years at the time of 
ICE surgery (Table 1). All but 1 patient (Case 4) pre-
sented with intractable headaches, and most patients (7 
of 10) also had visual symptoms including blurry vision,
transient visual obscurations, or vision loss over a period ranging from 2 months to 31 years prior to ICE. Elevated lumbar CSF opening pressures, typically greater than 30 cm H2O, were noted in all patients. Papilledema was observed in 4 patients preoperatively. All female patients were noted to be either overweight with a BMI of 25–29 (1 of 7 patients) or obese with a BMI of 30 or more (6 of 7 patients). The 3 male patients each had a normal BMI.

Prior to surgical referral, all patients were initially treated medically with 1 or more carbonic anhydrase inhibitors (acetazolamide or topiramate), a loop diuretic (furosemide), and/or corticosteroids. Additionally, pain management was initiated by a multidisciplinary team, and weight loss was encouraged for overweight or obese patients. In 5 patients (Cases 1–3, 5, and 10), surgical therapies including LP, VP, and/or lumbopleural shunt treatment, and/or ONSF were previously performed for medically refractory IIH (Table 1). In these cases, often after multiple revision surgeries, the signs and symptoms of intracranial hypertension persisted prior to ICE.

Perioperative Details and Surgical Complications

Select perioperative details have been summarized in Table 2. Technically successful internal cranial expansion was achieved in all patients within the cohort. All previously placed shunt hardware (valves, proximal, and distal catheters) was removed without replacement at the time of ICE. The ICE procedure duration varied between 3 and 6.2 hours (mean 4.6 hours), and intraoperative blood loss ranged from 200 to 1000 ml (mean 456 ml). Procedure-related complications were encountered in 2 patients. In one of these patients (Case 3) a single, self-limited episode of secondarily generalized tonic-clonic seizure occurred on postoperative Day 8. Small, localized hyperintense T2/FLAIR signals were noted on postoperative MRI in the right frontal and parietal lobes. No MRI evidence of stroke or electroencephalographic seizure activity during continuous electroencephalography monitoring was seen. In the other patient (Case 9), a tear in the sagittal sinus resulted in blood loss requiring intraoperative transfusion of 1 U of packed red blood cells. The dural defect was repaired with an autologous pericranial patch, and no clinical sequelae were observed.

Clinical Outcomes

Follow-up for the 10 patients in this series ranged from 1 to 39.6 months (mean 15.5 months). In total, 7 (70%) of the 10 patients were either symptomatically improved or asymptomatic at the time of the last follow-up visit. The remaining patients included 2 (Cases 3 and 8) who had only temporary postoperative symptomatic improvement and 1 (Case 1) who had no postoperative improvement. Importantly, these 3 patients had documentation of normal ICP or lumbar CSF opening pressure postoperatively, indicating that adequate decompression was accomplished. Headaches were completely resolved or reduced in intensity and frequency in 6 of 9 patients with this symptom at presentation. Similarly, a reduction or complete resolution of papilledema was noted in all patients (4 of 4) with this sign at presentation. Improvement or stabilization of other symptoms including visual complaints, tinnitus, retroorbital pain, and ear pain were also noted for some patients within the cohort (Table 3).

Intracranial Volume Measurements

Similar to previously reported values in patients un-
Internal cranial expansion for intracranial hypertension

TABLE 1: Patient characteristics*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Sx</th>
<th>Duration of Sx Prior to ICE</th>
<th>Papilledema</th>
<th>Opening Pressure (cm H2O)</th>
<th>BMI</th>
<th>Management Prior to ICE</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>16, M</td>
<td>HA, vertigo</td>
<td>2.6 yrs</td>
<td>no</td>
<td>36</td>
<td>22.8</td>
<td>medical, LPS (removed due to persistent Sx), VPS (w/ multiple revisions; removed due to persistent Sx)</td>
</tr>
<tr>
<td>2</td>
<td>20, F</td>
<td>HA, blurry vision, retroorbital pain, tinnitus</td>
<td>10 yrs</td>
<td>no</td>
<td>30</td>
<td>27.4</td>
<td>medical, LPS (w/ multiple revisions &amp; placement of programmable valve dialed up to highest setting; removed due to low pressure HAs)</td>
</tr>
<tr>
<td>3</td>
<td>13, M</td>
<td>HA</td>
<td>2.4 yrs</td>
<td>no</td>
<td>36</td>
<td>22.4</td>
<td>medical, VPS (w/ multiple revisions, valve adjustment to highest setting, &amp; placement of antisiphon device; removed due to low pressure HAs)</td>
</tr>
<tr>
<td>4</td>
<td>15, F</td>
<td>blurry vision, intermittent transient visual obscurations, vision loss</td>
<td>2 mos</td>
<td>yes</td>
<td>high</td>
<td>38.3</td>
<td>medical</td>
</tr>
<tr>
<td>5</td>
<td>17, F</td>
<td>HA, vision loss</td>
<td>1 yr</td>
<td>yes</td>
<td>56</td>
<td>45.8</td>
<td>medical, ONSF</td>
</tr>
<tr>
<td>6</td>
<td>13, M</td>
<td>HA, vision loss</td>
<td>3 yrs</td>
<td>yes</td>
<td>low 40s</td>
<td>33.7</td>
<td>medical</td>
</tr>
<tr>
<td>7</td>
<td>20, F</td>
<td>HA, blurry vision</td>
<td>1.4 yrs</td>
<td>no</td>
<td>31</td>
<td>40.9</td>
<td>medical</td>
</tr>
<tr>
<td>8</td>
<td>11, M</td>
<td>HA</td>
<td>2 yrs</td>
<td>no</td>
<td>38</td>
<td>23.5</td>
<td>medical</td>
</tr>
<tr>
<td>9</td>
<td>17, F</td>
<td>HA, blurry vision, ear pain, pulsatile tinnitus</td>
<td>2.6 yrs</td>
<td>yes</td>
<td>54</td>
<td>36.7</td>
<td>medical</td>
</tr>
<tr>
<td>10</td>
<td>61, F</td>
<td>HA, blurry vision, tinnitus</td>
<td>31 yrs</td>
<td>no</td>
<td>40</td>
<td>38.0</td>
<td>medical, LPS (removal due to failure), LPluS (removal due to failure), VPS</td>
</tr>
</tbody>
</table>

* HA = headache; LPS = LP shunt; LPluS = lumbopleural shunt; Sx = symptoms; VPS = VP shunt.

dergoing ICE for slit-ventricle syndrome, volumetric analysis shows postoperative intracranial volume expansion ranging between 3.8% and 12% in this series. The expanded epidural compartment created during the ICE procedure was seen to be nearly completely obliterated in all patients who underwent postoperative head CT scanning. This nearly complete obliteration was seen as early as 2 weeks after ICE and occurred due to progressive dural relaxation, with circumferential expansion of the intradural contents until they became apposed to the skull. Computed tomography evidence of brain relaxation with increased sulcation was also observed (Fig. 4).

TABLE 2: Perioperative data*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Length of Op (hrs)</th>
<th>Blood Loss (ml)</th>
<th>Transfusion</th>
<th>No. of PODs in Hospital</th>
<th>Perioperative Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>4.7</td>
<td>500</td>
<td>none</td>
<td>6</td>
<td>none</td>
</tr>
<tr>
<td>2</td>
<td>4.3</td>
<td>300</td>
<td>none</td>
<td>3</td>
<td>none</td>
</tr>
<tr>
<td>3</td>
<td>4.1</td>
<td>300</td>
<td>none</td>
<td>9</td>
<td>single episode of secondarily generalized seizure on POD 8 (no cEEG correlate)</td>
</tr>
<tr>
<td>4</td>
<td>5</td>
<td>200</td>
<td>none</td>
<td>3</td>
<td>none</td>
</tr>
<tr>
<td>5</td>
<td>4.5</td>
<td>800</td>
<td>none</td>
<td>5</td>
<td>none</td>
</tr>
<tr>
<td>6</td>
<td>4.75</td>
<td>200</td>
<td>none</td>
<td>3</td>
<td>none</td>
</tr>
<tr>
<td>7</td>
<td>5.3</td>
<td>400</td>
<td>none</td>
<td>4</td>
<td>none</td>
</tr>
<tr>
<td>8</td>
<td>4.1</td>
<td>400</td>
<td>2 U PRBC</td>
<td>3</td>
<td>none</td>
</tr>
<tr>
<td>9</td>
<td>6.2</td>
<td>1000</td>
<td>1 U PRBC</td>
<td>4</td>
<td>intraop sagittal sinus tear, repaired w/ pericranium</td>
</tr>
<tr>
<td>10</td>
<td>3</td>
<td>200</td>
<td>none</td>
<td>6</td>
<td>none</td>
</tr>
</tbody>
</table>

* cEEG = continuous electroencephalography; POD = postoperative day; PRBC = packed red blood cells.

Discussion

Walter Dandy was the first neurosurgeon to routinely use cranial decompression surgery in the treatment of patients with IIH. In his landmark series of 22 cases published in 1937, Dandy explained, “This condition is both immediately and permanently controlled (possibly with rare exceptions) by a subtemporal decompression.” However, this procedure has been largely abandoned due to the development of first-line pharmacological therapies and because more contemporary subtemporal decompression series have indicated that many patients will eventually...
require CSF diversion. Our experience and success with using ICE to treat the rare disorder Camurati-Engelmann disease and the more common slit-ventricle syndrome suggested that the same surgical principles may be applied in the treatment of patients with refractory IIH.

In this series, we show that ICE enabled improvement or complete resolution of symptoms in the majority (7 of 10) of patients undergoing the procedure. At least temporary symptomatic improvement was observed in all but 1 patient. Importantly, most patients (6 of 9) had improvement or complete resolution of intractable headaches, and all patients had improvement or complete resolution of papilledema (4 of 4) after undergoing ICE. Postoperative normalization of ICP was directly demonstrated in 2 patients by extended parenchymal (Case 1) or intraventricular (Case 8) pressure monitoring, while a significant reduction in lumbar CSF opening pressure was observed in 2 additional patients (Cases 3 and 5). Of note, the 3 patients (Cases 1, 3, and 8) who did not have postoperative reduction or resolution of symptoms also did not have evidence of continued intracranial hypertension measured by ICP monitoring or CSF opening pressure (Table 3). For these patients, management of symptoms is challenging and must be undertaken by a multidisciplinary team consisting of neurologists, pain specialists, mental health practitioners, and social workers.

While all current surgical therapies for IIH have been used effectively in select patients, we believe that ICE offers several distinct advantages in a broader cohort of patients. Optic nerve sheath fenestration has been shown safe and effective for preventing deterioration of vision in patients with IIH; however, postoperative complication rates as high as 40% per eye have been reported. These complications may include temporary motility or pupillary dysfunction, as well as vascular complications such as central retinal artery occlusion, resulting in poor visual outcome. Optic nerve sheath fenestration is also not effective for reducing ICP or for relieving headache and has a reported failure rate of 35% by 3–5 years. On the other hand, shunting (LP, VP, and ventriculopleural) procedures more effectively address increased ICP and headaches but are similarly plagued by high failure rates, necessitating repeat surgery. In a review by Rosenberg et al. of CSF diversion procedures in patients with IIH, it was shown that 64% of shunts lasted for shorter than 6 months. Ventriculoperitoneal shunting can create shunt dependence, cause overdrainage symptoms, and subject the patient to infection due to permanent hardware implantation. The completely extradural surgical approach of ICE enables avoidance of the hemorrhagic and cortical scarring complications associated with intraventricular catheter placement. Challenging placement and maintenance of patency of the intraventricular catheter inside slit-like ventricles is also avoided. Nonetheless, 1 patient in our series had a perioperative seizure, and another patient sustained a sagittal sinus tear; both complications underscore the general risks of any extra-

### Table 3: Patient outcomes

<table>
<thead>
<tr>
<th>Case No.</th>
<th>FU (mos)</th>
<th>Clinical Status (at last FU)</th>
<th>Intracranial Vol in ml</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Preop</td>
<td>Postop</td>
</tr>
<tr>
<td>1</td>
<td>13.3</td>
<td>1408</td>
<td>1483</td>
</tr>
<tr>
<td>2</td>
<td>39.6</td>
<td>1333</td>
<td>1420</td>
</tr>
<tr>
<td>3</td>
<td>26.6</td>
<td>1370</td>
<td>1422</td>
</tr>
<tr>
<td>4</td>
<td>33.7</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>5</td>
<td>19.5</td>
<td>1216</td>
<td>1366</td>
</tr>
<tr>
<td>6</td>
<td>3.4</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>7</td>
<td>10.8</td>
<td>—</td>
<td>—</td>
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<tr>
<td>8</td>
<td>2.6</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>9</td>
<td>4.3</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>10</td>
<td>1</td>
<td>—</td>
<td>—</td>
</tr>
</tbody>
</table>

* FU = follow-up; — = not available.

**Fig. 4.** Case 1. Computed tomography scans obtained after ICE brain relaxation. **Left:** Sulcal effacement is seen on a noncontrast image obtained immediately after VP shunt revision in a patient with IIH, suggesting that the persistent intracranial crowding may be due to a congenitally small supratentorial compartment. **Right:** Repeat image obtained 2 weeks after ICE and VP shunt removal, showing less effacement with more open sulci, indicating that significant intracranial expansion and brain relaxation are achieved.
Internal cranial expansion for intracranial hypertension

dural surgery and the specific risk of working around the midline.

The etiology of IIH is not understood and for this reason IIH has been termed a “disease of theories.” However, decreased CSF absorption due to increased venous pressure is a widely cited pathogenic mechanism.\textsuperscript{5,7,11,12,27} It is possible that by increasing the space for the sagittal sinus and other cortical veins, ICE reduces intracranial venous pressure, restoring a more physiological pressure gradient across the arachnoid granulations and promoting CSF absorption. This hypothesis could be addressed in future studies in which sinus pressures are either directly measured during catheter angiography or calculated with the use of noninvasive quantitative MR angiography technology such as NOVA (VasSol, Inc.).\textsuperscript{3,4,22}

Another intriguing yet untested hypothesis for the etiology of IIH is that it results from having a congenitally small cranial vault. It is conceivable that patients with relatively small intracranial volumes may have an increased likelihood of intracranial hypertension. Indeed, our observation of continued sulcal effacement in a patient with IIH despite the presence of a functional VP shunt suggested that a relatively small supratentorial compartment might have contributed to his symptoms (Fig. 4). Thus, analogous to the “small posterior fossa” theory of Chiari malformation Type I,\textsuperscript{3,17,19,20,28,29} we propose the “small supratentorial compartment” theory of IIH. For patients with IIH, ICE decompresses the supratentorial space much in the way that a suboccipital craniectomy decompresses the posterior fossa in patients with Chiari malformation Type I.

Conclusions

Internal cranial expansion is a safe and effective surgery for the treatment of patients with refractory IIH. This procedure results in increased intracranial volume enabling ICP normalization, which obviates the need for CSF diversion in this population. Improvement or complete resolution of headaches, visual complaints, papilledema, and other signs and symptoms related to intracranial hypertension may be seen in patients who undergo ICE. A multidisciplinary approach should be used for the optimal management of patients with IIH and is especially important in patients who remain symptomatic despite the ICP normalization conferred by ICE. Additional clinical and basic research is needed to clarify the pathogenesis of this vexing disorder.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Ghatan. Acquisition of data: Ghatan, Ellis, O’Hanlon. Analysis and interpretation of data: Ghatan, Ellis. Drafting the article: Ghatan, Ellis. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Ghatan.

References


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