Idiopathic intracranial hypertension (IIH), formerly known as pseudotumor cerebri, is a disease of elevated intracranial pressure that is thought to develop due to impaired CSF absorption related to elevated venous sinus pressure in the setting of increased intraabdominal and thoracic pressures. Symptoms can be disabling and, if left untreated, can lead to permanent visual loss. Previous treatments directed toward vision preservation include CSF diversion through shunting and optic nerve sheath fenestration. Recently, attention has been turned toward surgical weight loss strategies as an alternative to shunt treatment. The authors present a report of 3 patients with adolescent-onset IIH that was treated at the authors’ institution (Duke University) in whom bariatric surgery was pursued successfully. The patients had previously undergone CSF shunting at ages 12, 15, and 23 years. They were shunt dependent for a collective average of 3.3 years prior to bariatric surgery. All patients reported “low-pressure” or postural headaches after bariatric surgery that correlated with dramatic reduction in their weight. Two of the 3 patients had their shunts removed and continued to be shunt free 1.5 years later at last follow-up; the third patient remained shunt dependent with the pressure set at 200 mm H2O. Given the significant complications inherent to multiple shunt revisions, earlier intervention for weight loss, including bariatric surgery, in these patients might have prevented complications and the associated health care burden. The authors recommend a multidisciplinary approach for IIH treatment with early consideration for weight loss interventions in select patients.

https://thejns.org/doi/abs/10.3171/2017.6.PEDS17145

KEY WORDS  idiopathic intracranial hypertension; pseudotumor cerebri; weight loss surgery; bariatric surgery; gastric bypass; shunt; hydrocephalus

©AANS, 2017
ing dramatic weight loss. Before committing children and teenagers to possible long-term implanted hardware for CSF diversion, the utility of weight loss surgery should be explored. We now advocate for early consideration of weight reduction surgery as a means to avoid the many unnecessary headaches and complications associated with CSF diversion.

**Case Reports**

In our series, patients were able to achieve symptom resolution with increasing shunt settings or shunt freedom through significant weight loss after bariatric surgery. In Table 1 we attempted to summarize the shunt-related points of contact these patients had with the health care system, although the encounters are likely incomplete because of the use of multiple hospital systems and practitioners outside the Duke University system. Preoperative findings on imaging in all of these patients were unremarkable and without evidence of cerebral sinus thrombosis.

**Case 1**

This female patient presented at 16 years of age with polycystic ovarian syndrome, obesity, and headaches. She was found to have elevated opening pressures on lumbar puncture and was initially treated medically with attempted weight loss and acetazolamide. Multiple lumbar punctures, failed medical therapy, severe daily intractable headaches, and persistent elevated ICP led to the placement of a VP shunt when she was 23 years old. After 4 shunt revisions including conversion to a ventriculoatrial shunt for persistent abdominal pain and catheter migration out of the peritoneum, the patient elected to undergo gastric sleeve surgery 14 months after her initial shunt placement. Although she did have complications related to her

**Table 1. Case series summary**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Sex</th>
<th>Weight Loss Method</th>
<th>No. of ED Visits</th>
<th>Shunt Type</th>
<th>No. of Shunt-Related Ops</th>
<th>Other IIH-Related Ops*</th>
<th>Shunt Freedom</th>
<th>Notable Shunt-Related Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>Surgical</td>
<td>13</td>
<td>VP</td>
<td>4</td>
<td></td>
<td>VP shunt at 200 mm H2O</td>
<td>Prolonged externalization for infection, multiple malfunctions</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>Surgical</td>
<td>4</td>
<td>VP, LP</td>
<td>13</td>
<td>Bilat subtemporal decompression, separate prolonged ICP monitoring</td>
<td>Yes</td>
<td>Multiple malfunctions</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>Surgical</td>
<td>&gt;15</td>
<td>VP, VA</td>
<td>4</td>
<td></td>
<td>Yes</td>
<td>Multiple malfunctions</td>
</tr>
</tbody>
</table>

* Does not include lumbar puncture.
gastric sleeve surgery that necessitated reoperation, which led to a gastric bypass, she was successfully weaned from her ventriculoatrial shunt, and it was removed over the course of 2 years. Her BMI decreased from 49.8 before bariatric surgery to 21.3 at the time of shunt removal. She remains shunt free 1.5 years later.

Discussion

IIH is a disease defined by clinically symptomatic elevation of ICP in the setting of normal anatomy and no apparent underlying cause. The estimated annual incidence of ICP in the pediatric population ranges from 0.47 to 1.2 per 1000,000 in developed countries, although there is no precise incidence estimate in the United States.11,33 In older children and adolescents there is an increasing incidence, particularly in obese adolescent females.28 If left untreated, the long-term sequelae of elevated ICP is vision loss due to optic nerve damage. Surgical treatment options of CSF diversion and optic nerve sheath fenestration are aimed at preserving vision. All patients in our series underwent neurological and neurosurgical clinical evaluations, and all had multiple documented accounts of lumbar punctures revealing elevated ICP. The decision to place a VP or LP shunt was made in the presence of chronic headaches and/or papilledema/visual deficits after failure to control symptoms with conservative and medical therapy. Although not completely understood, the pathophysiology of IIH is thought to result from impaired CSF absorption in the setting of elevated venous sinus pressure with increased intraabdominal and intrathoracic pressures associated with obesity.4,19,31 While only 2 case reports currently exist in the pediatric literature,7,22 numerous studies in adults have shown improvement in symptoms and ICP through weight loss.18,21,26 In patients with CSF shunts, there is a documented risk of overdrainage headaches after weight loss.29 Interestingly, all 3 of our patients demonstrated symptoms of low-pressure headaches correlating directly with weight loss. We advocate for the use of programmable valves in this patient population, as the programmable shunt valves allow the shunt to be adjusted to higher pressures as patients continue to lose weight (Figs. 1 and 2). We monitored neurological symptoms after gastric surgery with interval outpatient follow-up every 3 months. No invasive testing to confirm ICP was done, but improvement in symptoms after reprogramming the shunt to higher pressures confirmed diagnosis in addition to providing therapy.

CSF shunting is obviously not without cost or risk. It is estimated that 20%–25% of all patients requiring a

FIG. 1. Case 1. Graphic representation of BMI by age. Points represent clinic visits. Depicted is the process of increasing the shunt valve setting because of overdrainage symptoms, which correlated with dramatic weight loss. Figure is available in color online only.
shunt will have at least 1 complication. The complications related to shunt placement and revision in a population undergoing multiple revisions in a relatively short period of time are well demonstrated in this series. The published literature echoes this; in a large cohort of patients undergoing shunt surgery, the incidence of infection was 6.1%, and the revision rate was 22.0%.25 When patients with IIH are evaluated, shunt revision rates tend to be higher than those of other types of hydrocephalus and have been reported to be as high as 60%–100% for VP shunts and even higher for LP shunts.1,24,27 In our series of patients who underwent shunting, the revision rate was 100% with all patients requiring multiple revisions within 1 year. With the many complications requiring frequent evaluations, shunts in this population can place the patient at significant risk. Specifically, particular considerations regarding the details of shunt removal should be noted. Prior to removal in our series, the shunts were set at the highest pressure settings in consultation with a headache specialist. These patients underwent removal of the distal catheter and shunt valve. Because of the risk of hemorrhage and small ventricles associated with this disease, the ventricular catheter was tied off and sutured to the periosteum. If symptoms of ICP ever returned, the CSF diversion system could be easily reimplemented. After shunt removal, these patients were monitored overnight in the hospital, followed by outpatient follow-up for recurrence of symptoms related to elevated ICP and assessment for immediate postoperative complications. Although we did not leave a reservoir, in hindsight this may be a prudent technical strategy as the effects of bariatric surgery might be transient or not always effective, and ICP evaluation via a reservoir may be significantly easier than through a lumbar puncture in this population.

Furthermore, the economic and resource burden of the IIH population to the health care system should not go unmentioned. The median cost for shunt revision for failure is approximately $5000, plus an additional estimated $40,000 if the shunt becomes infected.12,30 In addition, the average number of emergency department visits requiring some combination of neurosurgical consultation, lumbar puncture, shunt flow study/shuntogram, and shunt reprogramming was approximately 10 per patient, which is a substantial additional cost to the patient and health care system.

In our series we noted an immediate improvement in IIH symptoms after bariatric surgery, which directly correlated with dramatic weight loss as noted on the BMI charts. As a result of initial successes such as those reported here, our institution now offers weight loss surgery to adolescents. To qualify, patients must be at least 14 years old, have a BMI > 35 with at least one “serious” obesity-related comorbidity or a BMI > 40 with any obesity-related comorbidity. “Serious” comorbidities include Type 2 diabetes, severe obstructive sleep apnea, pseudotumor cerebri, and documented steatohepatitis. After referral to our weight loss center, patients undergo a 6-month trial of healthy lifestyle and conservative weight loss strategies and clinical and psychological evaluation before they are allowed to undergo any bariatric operation.
In general, bariatric surgery has been felt to be low risk and the most clinically effective and cost-effective treatment of obesity-related comorbidities in adults.\textsuperscript{17,20,32} The most common side effects include abdominal pain and changes in bowel habits, including constipation. With regard to adolescents compared with adults, pediatric patients differ biologically, psychologically, and socially. First, pediatric patients may have potential linear growth, and some surgical techniques may result in malabsorption and vitamin deficiencies important in a growing population.\textsuperscript{5} Pediatric patients also have a higher incidence of eating disorders, depression, and anxiety, which are associated with greater noncompliance after surgery.\textsuperscript{10,16} Lastly, bariatric surgery performed in an adolescent patient raises the question of informed consent for operations that have life-altering results.\textsuperscript{6}

Despite these differences, bariatric surgery has been demonstrated to improve weight loss, health, and psychosocial outcomes in adolescents. Because of the increase in positive results, we present a new algorithm that involves early referral to a multidisciplinary weight reduction team. For IIH, we present an algorithm for the management of patients, both obese and nonobese (Fig. 3). For obese patients, unless severe IIH with focal neurological findings or vision loss is present, we recommend referral to weight loss specialists before permanent neurosurgical intervention. All 3 patients at initial presentation would have met criteria for mild to moderate IIH and would have likely benefited from earlier referral to weight loss specialists for bariatric surgery consideration. For nonobese patients, once secondary causes have been evaluated, traditional treatment such as CSF diversion or optic nerve sheath fenestrations are considered.

As weight loss surgery continues to expand to younger populations, the use of bariatric surgery and effective nonsurgical weight loss strategies should be part of the neurosurgeon’s armamentarium for patients with IIH. Currently, there is a paucity of long-term follow-up data, limited to 2 case reports,\textsuperscript{7,22} but there is an abundance of literature in the adult population that has supported the use of bariatric surgery as a treatment modality for IIH.\textsuperscript{9,23} As with our small series, gastric bypass successfully helped resolve IIH symptoms (headaches and vision loss) in the other 2 case reports in the pediatric literature. With increasing obesity rates, we envision a multidisciplinary approach to this patient population. With the aid of general surgeons, weight loss physicians, psychologists, psychiatrists, nutritionists, neurologists, and ophthalmologists, IIH patients would ideally be referred to and reviewed by a multidisciplinary team to streamline care and avoid unnecessary procedures. Long-term results of this treatment should be followed prospectively with registries such as the Intracranial Hypertension Registry.\textsuperscript{2}

Conclusions

IIH is increasing in the pediatric population. Pediatric neurosurgeons must be aware of the various treatment paradigms. We demonstrate the success of bariatric surgery in the adolescent population and advocate for early referral to weight loss physicians. The traditional treatment algorithm with all roads leading to CSF diversion surgery must be revisited.

References

4. Bloomfield GL, Ridings PC, Blocher CR, Marmarou A, Sugerman HF: A proposed relationship between increased...

Disclosures
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
Conception and design: all authors. Acquisition of data: Hooten, Hoang. Analysis and interpretation of data: all authors. Writing the article: Hooten, Hoang. Critical revising the article: all authors or methods used in this study or the findings specified in this paper. Correspondence
Kristopher G. Hooten, Department of Neurosurgery, Duke University, 4544 Hospital South, Box 3807, Durham, NC 27710. email: kristopher.hooten@neurosurgery.duke.edu.