This updated review of pediatric idiopathic intracranial hypertension focuses on epidemiology, clinical presentations, diagnostic criteria, evaluation, clinical course, and treatment. General guidelines for the clinical management of idiopathic intracranial hypertension are discussed. A new algorithm outlines an efficient management strategy for the initial diagnostic evaluation of children with signs or symptoms of intracranial hypertension. This algorithm provides a systematic approach to initial evaluation and management, and identifies important decision-making factors. The risk of permanent visual loss with idiopathic intracranial hypertension necessitates a prompt, thorough collaborative approach in the management of patients.

Increased intracranial pressure is classified into two main categories, depending on its idiopathic or secondary etiology. In the literature, the two categories have not always been clearly delineated, leading to difficulties in understanding associations or drawing inferences about the syndrome. Categorizing etiologies is necessary, because characteristics, management, and outcomes may vary considerably. Idiopathic intracranial hypertension is defined as increased intracranial pressure without a space-occupying lesion or hydrocephalus and normal cerebrospinal fluid composition [1]. Initially termed “serous meningitis,” this syndrome was first recognized by Quincke in 1897 [2]. With its unclear etiology, the syndrome has assumed different names, including pseudotumor cerebri and benign intracranial hypertension. Especially in children, questions continue to surround the diagnostic criteria and medical management.

A review of the literature indicates that the diagnostic evaluation of these patients often involves several medical subspecialties. Articles have been written by different physicians in many subspecialties, including general practice, emergency medicine, radiology, psychiatry, neurology, ophthalmology, neuro-ophthalmology, and neurosurgery [3-8]. Although the approach to patients with idiopathic intracranial hypertension is often similar among subspecialties, differences do exist. Coordinating the patient’s care from presentation to follow-up is an enormous challenge. Because a risk of permanent visual loss exists with idiopathic intracranial hypertension, patients require a prompt and thorough collaborative approach in their management.

Wide diversity is evident in the published approaches, and little has been written about children. Therefore, an urgent need exists for a coherent approach to the evaluation and management of children. This updated review of idiopathic intracranial hypertension offers a focus on children, including epidemiology and pathogenesis, clinical presentations, common clinical course, diagnostic criteria and challenges, and treatments. An algorithm for the initial diagnostic evaluation of children with intracranial hypertension is proposed. This algorithm provides a systematic approach to management, identifies important decision-making factors, and illustrates the importance of a coordinated multidisciplinary approach.
Epidemiology and Pathogenesis

Epidemiology

The true incidence of idiopathic intracranial hypertension in children is unknown. In the past, children were often included within adult studies, leaving the pediatric incidence unclear. Moreover, several previous pediatric studies did not exclude children in whom a secondary etiology was discovered for their intracranial hypertension signs. Hence the number of patients with idiopathic intracranial hypertension may have been falsely elevated [9,10]. Although the incidence of idiopathic intracranial hypertension in children is unknown, we are gaining a better understanding of this syndrome. With this knowledge and further research, we will achieve a better understanding of the trends of idiopathic intracranial hypertension in children.

Limited evidence points to possible associations with gender and obesity in older children. Babikian et al. reported that nearly 60% of children with idiopathic intracranial hypertension were aged more than 10 years [11]. This finding illustrates that increasing age may be a risk factor for children with idiopathic intracranial hypertension. Baker et al. reported a female prevalence in postpubertal children [3]. Weisberg and Chutorian reported that 14 of 30 (47%) patients below age 13 years were female, in contrast with six of eight (75%) patients older than 13 years [12]. Although Babikian et al. [11], Baker et al. [3], and Weisburg and Chutorian [12] included patients over a long duration of time (ranging from 10-30 years), their studies were retrospective chart reviews with a possible selection bias, based on potential case severity and referral patterns. Younger children may have been unable to communicate their symptomatic complaints. This limitation could have led to underdiagnosis or diagnosis of mild severity without referral. Balcer et al. demonstrated a significant relationship between sex and age in patients with idiopathic intracranial hypertension: 50% of patients aged 3-11 years, 88% of patients aged 12-14 years, and 100% of patients aged 15-17 years were female [13]. That same study also demonstrated a similar relationship between obesity and age in patients with idiopathic intracranial hypertension: 43% of patients aged 3-11 years, 81% of patients aged 12-14 years, and 91% of patients aged 15-17 years were obese [13]. The analysis by Balcer et al. suggested that the likelihood of obesity in older children was not solely dependent on female predominance in older children [13]. One challenge regarding the possible associations of sex and obesity in older children involves determining a definition of “older.” Many authors in the pediatric literature recommend categorizing children according to their sexual maturation (prepubertal and pubertal/postpubertal). As recognized by Rangwala and Liu [14], one difficulty with categorizing children based on sexual maturation involves deciding whether to base categories on secondary sexual characteristics or predeter-
were 20% or more over their ideal weight [20]. Although that study helped physicians realize that idiopathic intracranial hypertension was not a rare syndrome, the incidence rates could have been lowered by the short timeframe and the survey method for gathering data.

Other adult-based studies reported the similar finding that obesity and female sex are significant risk factors for this syndrome in adults. Based on population studies in Minnesota, Radhakrishnan et al. [21] estimated a similar annual incidence for adults. They reported an annual age-adjusted incidence rate of 1.6 for women. In females aged 15-44 years with a body mass index >26, the incidence rate rose to 7.9 [21]. Although that study evaluated a population over a 15-year period, the small number of patients (n = 9) may have limited the ability to generalize its findings to a larger population. Some authors stated that annual incidence rates are now higher because of an increased awareness of the syndrome and the increasing prevalence of obesity [22,23].

Pathogenesis

The exact mechanism causing idiopathic intracranial hypertension is unknown, and many theories have been proposed. Most of the literature focuses on cerebral hemodynamics, including increased cerebral blood volume, increased cerebrospinal fluid production, or increased brain volume, along with decreased cerebrospinal fluid resorption or venous flow [7,24,25]. Although any of these factors could raise intracranial pressure, one of the most discussed possibilities involves the obstruction of cerebrospinal fluid or venous flow [26-28]. Cerebrospinal fluid pressure must remain higher than cerebral venous pressure to maintain normal pressures between cerebrospinal fluid and blood. A rise in cerebral venous pressure leads to increased cerebrospinal fluid pressure [22]. Karahalios et al. [28] demonstrated that dural sinus pressure was elevated in patients with idiopathic intracranial hypertension during venography. They hypothesized that the elevated venous pressure led to decreased cerebrospinal fluid resorption, and subsequently to increased intracranial pressure [28]. However, Baryshnik and Farb argued that increased intracranial venous pressure is an effect of increased intracranial pressure, because intracranial venous pressure normalizes when elevated intracranial pressure is lowered [29].

Diagnostic Challenges

Diagnostic criteria were originally outlined by Smith [30] (Table 1), based on characteristics described by Dandy in 1937 [31]. Over the past several years, physicians have proposed further expansions of the criteria, primarily as a result of further research and knowledge, as well as advances in diagnostic technology such as magnetic resonance imaging. For example, physicians have recommen-ded further clarification that a patient is not required to exhibit signs of idiopathic intracranial hypertension. However, if patients do, these signs must be consistent with intracranial hypertension or papilledema [1,32]. Physicians have also specified the positioning in the lumbar puncture as well as the diagnostic opening pressure [1,14,32,33]. Further recommendations regard appropriate neuroimaging [1,5,32]. Some added criteria stating that no other cause of intracranial hypertension should be identified [1,32,33].

The original modified criteria of Dandy provide the acceptable minimal guidelines for a diagnosis of idiopathic intracranial hypertension. However, applying primarily adult-derived diagnostic criteria to all age groups of children remains a challenge. One goal of this review is to discuss different recommendations proposed in both the adult and the pediatric literature, and to focus on areas in need of further prospective research, to clarify the criteria that would help lead to a systematic approach to patients.

Differences of opinion exist regarding the criteria for signs. The criteria of the International Headache Society suggest there must be a progressive headache with at least one of the following characteristics: daily, diffuse, or constant nonpulsating, or pain aggravated by Valsalva maneuvers. However, several reports in the literature describe patients, particularly children, who are not symptomatic but who have papilledema diagnosed on a routine eye examination, and who meet the other required criteria for idiopathic intracranial hypertension [16,34,35]. The International Headache Society, in concordance with other authors, stated that a patient diagnosed with idiopathic intracranial hypertension may have normal results of a neurologic examination, including the absence of papilledema [1,33,36,37]. Digre et al. [38] performed a cross-sectional analysis comparing patients with idiopathic intracranial hypertension and papilledema to those without papilledema. Although that study involved mostly adults and only a few pubertal and postpubertal children, Digre et al. [38] estimated the prevalence of patients with idiopathic intracranial hypertension but without papilledema to be 5.7%. They stated that those without papilledema more often reported photopsias and nonphysiologic visual field constriction than did patients with papilledema. They also reported that although all patients in their study exhibited elevated intracranial pressure, the mean opening pressure in patients without papilledema was lower than in patients with papilledema (309 mm H$_2$O and 373 mm H$_2$O, respectively) [38]. That

<table>
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<th>Table 1. Modified criteria of Dandy [31]</th>
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<tr>
<td>1. Signs of increased intracranial pressure.</td>
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<tr>
<td>2. No localizing neurologic signs otherwise, with the single exception of unilateral or bilateral paresis in nerve VI.</td>
</tr>
<tr>
<td>3. Cerebrospinal fluid can exhibit increased pressure, but no cytologic or chemical abnormalities otherwise.</td>
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<tr>
<td>4. Normal to small symmetric ventricles must be demonstrated.</td>
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Standridge: Idiopathic Intracranial Hypertension 379
cross-sectional analysis offers a current perspective on the prevalence of patients with idiopathic intracranial hypertension but without papilledema. However, it is limited because a causal relationship cannot be inferred between the absence of papilledema and a lower opening pressure.

Another criterion that remains controversial involves the diagnostic cerebrospinal fluid opening pressure. Earlier literature regarding adults considered opening pressures above 200 mm H2O to be elevated [39-42]. However, later studies produced evidence of opening pressures ranging from 200-250 mm H2O in normal healthy subjects. Gilland et al. studied 31 healthy subjects, and found five with opening pressures between 200-240 mm H2O [42]. Tourtellotte reported an average opening pressure of 150 ± 33 mm H2O, reaching a maximum of 216 mm H2O using two standard deviations in 105 healthy subjects [43]. These findings suggest that opening pressures of more than 200 mm H2O, but less than 250 mm H2O, may not be abnormal. Therefore, in adults, most of the literature is in agreement that an opening pressure of at least 250 mm H2O is diagnostic, and that opening pressures of 201-249 mm H2O are nondiagnostic [6,44,45].

In children, the cerebrospinal fluid opening pressure that fulfills the diagnostic criteria for idiopathic intracranial hypertension continues to be debated. Similar to the situation in adults, a range of recommendations has been proposed without the support of clinical data. However, studies have been widely accepted regarding recommended opening pressures in children. In infants, Welch reported that up to 82 mm of H2O constituted a normal opening pressure in children 8 years and older begins to resemble the previously established normal adult upper limit of 200 mm H2O [46]. Some authors suggest that normal opening pressure evolves with age. Normal opening pressure increases from 82 mm H2O in infancy to 180 mm H2O in children less than 8 years of age, and the opening pressure in children 8 years and older begins to resemble the previously established normal adult upper limit of 200 mm H2O [40,47]. These published studies and recommendations suggest that mean opening pressure in children is lower than in adults, but varies with age, and in older children tends to approach adult values. Until prospective studies can produce reliable and valid measures of opening pressures in children, guidelines must be based on previous studies and expert opinion. In the proposed algorithm, a diagnostic opening pressure of 200 mm H2O in children less than 8 years of age and 250 mm H2O in children 8 years and older is recommended for the diagnosis of idiopathic intracranial hypertension.

Several studies indicate that cerebrospinal fluid pressure fluctuates widely in individuals with idiopathic intracranial hypertension [48-50]. Because cerebrospinal fluid pressure can fluctuate, a child with signs of idiopathic intracranial hypertension may not meet the suggested diagnostic opening pressure criteria. Performing at least a second lumbar puncture may be necessary in a person with consistent idiopathic intracranial hypertension signs [1,51]. Because intermittent increases of cerebrospinal fluid pressure may be missed, some authors suggest transducer monitoring through a lumbar drain, or intracranial pressure monitoring, to confirm the diagnosis of idiopathic intracranial hypertension within patients suspected of manifesting idiopathic intracranial hypertension [1,6,51,52].

Another criterion for diagnosing idiopathic intracranial hypertension involves the necessary imaging to exclude secondary etiologies of intracranial hypertension. Computed tomography provides an adequate modality for excluding hydrocephalus, hemorrhage, or mass effect. However, magnetic resonance imaging is superior to computed tomography in detecting isodense tumors, meningeal infiltrations, posterior fossa pathologies, or other subtle intracranial abnormalities. One area of debate involves whether magnetic resonance venography should be included in the initial evaluation, to exclude cerebral venous sinus thrombosis as a secondary cause of intracranial hypertension. This etiology is important to consider, because the treatment for cerebral venous sinus thrombosis may involve anticoagulation therapy. Magnetic resonance imaging is often sufficient to detect a complete occlusion of the cerebral veins or sinuses, but with partial occlusions, magnetic resonance venography is superior [53]. Although magnetic resonance venography may be more sensitive than magnetic resonance imaging at detecting venous occlusions, it presents limitations. Studies indicate that hypoplastic venous sinuses, smaller cortical veins, and artifactual signal loss in the venous system may be difficult to distinguish from a cerebral venous sinus thrombosis according to venography [5,53,54]. However, combining the information from magnetic resonance imaging with magnetic resonance venography, or using contrast-enhanced venography, may confirm a diagnosis of cerebral venous sinus thrombosis more definitely than either modality separately [5,55].

Although magnetic resonance venography is noninvasive, its role in children with signs of intracranial hypertension is undefined. To date, no randomized, controlled studies have evaluated diagnostic imaging, and particularly magnetic resonance venography, in children with intracranial hypertension. A recent retrospective study reported no significant clinical differences in children presenting with signs of intracranial hypertension, based on the completion of venography or the presence of thrombosis [56]. Because a history of recent sinus or otitis media infection was reported as an associated risk for thromboses in children, some authors suggest that a physician consider magnetic resonance venography in children with this history [1,57,58].

Baker et al. reported that children with dural sinus thrombosis were at a higher risk for serious visual loss than children with different etiologies for intracranial hypertension [3]. However, that study was retrospective, and therefore a direct link between sinus thrombosis and visual loss cannot be confirmed. Phillips et al. retrospectively reviewed 35 children with intracranial hypertension over a 20-year period [58]. Of the 35 patients, five were diagnosed with dural sinus thrombosis. The authors reported
that none of those five patients with dural sinus thrombosis exhibited residual deficits in their visual acuity or visual fields during follow-up [58]. Although that study was retrospective, it suggests that cerebral sinus venous thrombosis may not be a risk factor for poor visual outcomes.

In the adult literature, studies indicate that up to 37% of patients with cerebral venous sinus thromboses present with signs of idiopathic intracranial hypertension [59,60]. However, other studies of typical patients with idiopathic intracranial hypertension (young, obese, and female) who were studied using magnetic resonance venography were negative for thromboses [61]. At present in the adult literature, two different opinions predominate regarding magnetic resonance venography during initial evaluation: either perform this additional imaging in all patients who present with signs of intracranial hypertension, or perform this imaging only in patients described as “atypical” including males, nonobese females, the elderly, and prepubescent children [1,59-61].

Clinical Presentations

In children, the most common presenting symptom is headache [11,16,58]. However, the characteristics of these headaches are variable. A headache may begin as moderate and episodic and progress to severe, chronic pain [62]. Patients often report a headache to be exacerbated by Valsalva and postural changes [62]. Patients may describe a headache similar to a migraine, with a pulsatile quality as well as photophobia, nausea, and vomiting [62]. Differentiating between headaches caused by idiopathic intracranial hypertension and other headache syndromes remains a challenge, especially in young children who may not be able to describe their symptoms clearly. Moreover, a patient may manifest both a primary migraine and headaches as a result of idiopathic intracranial hypertension [63,64].

Another symptom that patients may describe is pulsatile tinnitus, which is present in approximately 50% of patients with idiopathic intracranial hypertension [22]. Patients may express complaints of decreased vision, vision loss, or visual changes such as transient visual obscurations. Idiopathic intracranial hypertension constitutes one of the few conditions to cause transient visual obscurations [7,22]. In addition, patients often voice nonspecific complaints, such as shoulder and neck pain, ataxia, and paresthesias.

The patient may or may not be able to realize the presence of visual deficits. The visual field is usually affected early in the course of idiopathic intracranial hypertension, resulting in the enlargement of blind spots, inferonasal loss, and generalized constriction of the visual fields [25,35,38,65]. Visual acuity is usually unaffected early in the course of idiopathic intracranial hypertension, but any loss of visual acuity is considered ominous and necessitates immediate treatment [22]. In children, up to 90% demonstrate visual field deficits, and up to 20% present with a loss of visual acuity loss [11,15]. Papilledema, the typical sign of intracranial hypertension, may be present bilaterally, unilaterally, asymmetrically, or not at all [36,66]. Patients may develop cranial neuropathies. Abducens palsy is the most common, and can lead to a complaint of double vision [17,67].

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![Algorithm-based approach for the initial evaluation of pediatric idiopathic intracranial hypertension](image)

**Figure 1.** An algorithm-based approach for the initial evaluation of pediatric idiopathic intracranial hypertension. CSF = cerebrospinal fluid; HCT = head computed tomography; IHH = idiopathic intracranial hypertension.
Proposed Pediatric Algorithm

Approach

The need exists for a systematic approach to the initial diagnostic evaluation of a child with signs of intracranial hypertension. The proposed algorithm provides a systematic approach to initial evaluation and management, and identifies important decision-making points (Figs 1, 2). It also illustrates the importance of a coordinated multidisciplinary approach. Although Class I evidence is lacking, this algorithm could support more consistent practice and provide a foundation for larger collaborative studies. These studies will be necessary to help develop more appropriate pediatric diagnostic criteria and management, for improved accuracy of diagnoses and outcomes.

Based on the modified criteria of Dandy, in addition to a complete history and physical examination, the neurologist must evaluate patients for normal to small symmetric ventricles, increased cerebrospinal fluid pressure, and other cytologic or chemical abnormalities [31]. The pediatric neurologist may be able to arrange urgent head imaging and an outpatient spinal puncture procedure, or collaborate with a facility that provides these procedures.

The common pathway of idiopathic intracranial hypertension and intracranial hypertension caused by a secondary etiology involves an elevation of intracranial pressure that requires appropriate medical attention [68]. Patients suspected of manifesting intracranial hypertension and who are taking medications or who exhibit comorbidities anecdotally associated with idiopathic intracranial hypertension should receive appropriate diagnostic evaluation and continue in the algorithm. This procedure will help affirm a diagnosis of intracranial hypertension, exclude other secondary etiologies, and help direct management of the signs of intracranial hypertension.

Because certain medications are reportedly associated with intracranial hypertension, any suspicious treatments should be discontinued if possible [25,51]. Several alleged associations between idiopathic intracranial hypertension and different medications or certain comorbidities are

Figure 2. An algorithm-based approach to the initial evaluation of pediatric idiopathic intracranial hypertension. CSF = cerebrospinal fluid; CSVT = cerebral sinus venous thrombosis; HA = headache; HCT = head computed tomography; ICH = intracranial hypertension; IHH = idiopathic intracranial hypertension; LP = lumbar puncture; MRI = magnetic resonance imaging; MRV = magnetic resonance venography; ONSF = optic nerve sheath fenestration; Ophtho = ophthalmology.

Δ = if symptoms/signs persist, consider repeat LP and/or continuous pressure monitoring

# = if no response to therapies, consider MRV to rule out CSVT

LP, lumbar puncture  MRV, magnetic resonance venography  HA, headache  Ophtho, ophthalmology
CSVT, cerebral sinus venous thrombosis  ONSF, optic nerve sheath fenestration  ICH, intracranial hypertension  CSF, cerebrospinal fluid
IHH, idiopathic intracranial hypertension  HCT, head computed tomography
derived from retrospective case reports or series, and therefore causation or pathogenesis cannot be proven [69-78]. Giuseffi et al. [79] and Ireland et al. [80] demonstrated that several previously reported associations were questionable, e.g., antibiotics (tetracycline, nalidixic acid, and nitrofurantoin), oral contraceptives, and corticosteroids [79,80]. In addition, Ireland et al. reported strong associations between obesity and weight gain in the 12 months before a diagnosis of idiopathic intracranial hypertension, and their findings are widely referenced today [80]. Although that study involved questionnaires with a small number of patients, it is one of the first case-control studies to evaluate several of the reported associations proposed by many earlier case reports. Jacobson et al. [81] reported on an association between a high serum concentration of vitamin A in the form of retinol and patients with idiopathic intracranial hypertension compared with healthy control subjects. The limitations of that study include the small sample size of patients and the lack of a pathogenesis linking elevated serum retinol levels and idiopathic intracranial hypertension [81].

The proposed algorithm is designed for the neurologist to collect the minimal yet necessary information for a diagnosis and initial management. Because not all neurologists or medical institutions have immediate magnetic resonance capability, the cranial computed tomography image is considered adequate and is acceptable as the initial head imaging. However, if cranial magnetic resonance imaging can be completed within the initial presentation and evaluation, magnetic resonance imaging is recommended. A lumbar puncture must be performed whenever idiopathic intracranial hypertension is suspected, with patients in the lateral decubitus position and with legs relaxed [40,42,52]. This procedure is critical in evaluating the opening pressure, and in excluding secondary etiologies of intracranial hypertension such as infections and malignancy [45,52].

Subsequent to history and physical examination, head imaging, and lumbar puncture, the pivotal decision-making point in the algorithm concerns the findings of a visual examination (Fig 2). This step is appropriate because the most serious morbidity for idiopathic intracranial hypertension involves the possibility of permanent visual loss. The findings that ophthalmologists and neuro-ophthalmologists evaluate for visual loss include visual function (visual acuity and visual fields) and the appearance of optic discs [3,65,66,68]. Therefore, the cornerstone of management constitutes the degree of papilledema and resulting changes in visual function [22,25,52,82]. The severity of findings of a visual examination, including changes in visual acuity, visual field loss, and grade of papilledema, determines the direction and speed of the diagnostic evaluation in the algorithm.

Experts in idiopathic intracranial hypertension such as Friedman have proposed specific categories for the findings of visual examinations that correlate with different management pathways, depending on degree of severity (Table 2) [52]. Visual acuity is measured with a Snellen

<table>
<thead>
<tr>
<th>Stage</th>
<th>Normal optic disc</th>
</tr>
</thead>
<tbody>
<tr>
<td>(A)</td>
<td>Blurring of the nasal, superior, and inferior poles in inverse proportion to disc diameter</td>
</tr>
<tr>
<td>(B)</td>
<td>Radial NFL without tortuosity</td>
</tr>
<tr>
<td>(C)</td>
<td>Rare obscuration of a major vessel, usually on the upper pole</td>
</tr>
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Stage 1: Very early papilledema

<table>
<thead>
<tr>
<th>Stage</th>
<th>Moderate papilledema</th>
</tr>
</thead>
<tbody>
<tr>
<td>(A)</td>
<td>Obscuration of nasal border of the disc</td>
</tr>
<tr>
<td>(B)</td>
<td>No elevation of disc borders</td>
</tr>
<tr>
<td>(C)</td>
<td>Disruption of normal radial NFL arrangement, with grayish opacity accentuating nerve fiber bundles</td>
</tr>
<tr>
<td>(D)</td>
<td>Normal temporal disc margin</td>
</tr>
<tr>
<td>(E)</td>
<td>Subtle grayish halo with temporal gap</td>
</tr>
<tr>
<td>(F)</td>
<td>Concentric or radial retinochoroidal folds</td>
</tr>
</tbody>
</table>

Stage 2: Early papilledema

<table>
<thead>
<tr>
<th>Stage</th>
<th>Severe papilledema</th>
</tr>
</thead>
<tbody>
<tr>
<td>(A)</td>
<td>Obscuration of all borders</td>
</tr>
<tr>
<td>(B)</td>
<td>Elevation of all borders</td>
</tr>
<tr>
<td>(C)</td>
<td>Increased diameter of optic nerve head</td>
</tr>
<tr>
<td>(D)</td>
<td>Obscuration of one or more segments of major blood vessels leaving the disc</td>
</tr>
<tr>
<td>(E)</td>
<td>Peripapillary halo: irregular outer fringe with finger-like extensions</td>
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</tbody>
</table>

Stage 3: Moderate papilledema

<table>
<thead>
<tr>
<th>Stage</th>
<th>Very severe papilledema</th>
</tr>
</thead>
<tbody>
<tr>
<td>(A)</td>
<td>Dome-shaped protrusions, representing anterior expansion of optic nerve head</td>
</tr>
<tr>
<td>(B)</td>
<td>Peripapillary halo is narrow and smoothly demarcated</td>
</tr>
<tr>
<td>(C)</td>
<td>Total obscuration of a segment of a major blood vessel may or may not be present</td>
</tr>
<tr>
<td>(D)</td>
<td>Obliteration of optic cup</td>
</tr>
</tbody>
</table>

Adapted from Friedman [52].

Abbreviation:

NFL = Nerve fiber layer
A child whose findings are consistent with the severe category of visual examination at any point in the evaluation should be admitted to a hospital with resources for aggressive medical or possibly surgical management. Severe visual loss (either marked loss of visual acuity or visual field loss) and marked to severe papilledema provide indications for possible surgical intervention [6,22,84]. After a patient is admitted, medical management must be initiated. An emergent ophthalmology consultation is necessary. This procedure will allow for a swift confirmation of the severity of findings from the visual examination, and will help indicate further recommendations for acute medical management. The ophthalmologist may recommend a neuro-ophthalmology consultation for possible optic nerve sheath fenestration. A neurosurgery consultation should be ordered to evaluate the patient for other possible surgical options such as cerebrospinal fluid shunting. At this point, the child may have undergone only the suggested minimal head imaging. Because the child may be a surgical candidate, further magnetic resonance imaging and magnetic resonance venography are necessary. These studies may provide additional information that could acutely influence the need for surgery or its timing. A child in normal, mild, or moderate categories after a visual examination can be managed as an outpatient. The decision after obtaining the results of a visual examination is based on cerebrospinal fluid opening pressure. Children with a normal opening cerebrospinal fluid pressure can be managed based on the presence or absence of signs. Children with signs should be treated accordingly. Further investigation should be pursued for other etiologies such as headache. Chronic daily headache or headaches associated with analgesic overuse may manifest with some similarity to idiopathic intracranial hypertension without papilledema [37,52]. In children with signs, symptomatic management, including treatment of intracranial hypertension, may be warranted [25]. A child with signs of idiopathic intracranial hypertension should also be examined promptly by an ophthalmologist, and should undergo further magnetic resonance imaging. Children with signs only should also be examined promptly by an ophthalmologist. Depending on the ophthalmologic evaluation, if a cause for the papilledema is diagnosed, further imaging may not be warranted.

Children in normal, mild, or moderate categories after a visual examination and with elevated opening cerebrospinal fluid pressure should receive appropriate management for intracranial hypertension. These children also need prompt ophthalmologic consultation for baseline evaluation.
and monitoring, as well as cranial magnetic resonance imaging for the detection of secondary etiologies of intracranial hypertension.

Collaboration

Collaboration among physicians within the algorithm is very important. The speed of the initial diagnostic evaluation and management depends on a well-coordinated multidisciplinary approach. If urgent outpatient cranial imaging and lumbar puncture are not possible, the neurologist may need to coordinate with a nearby, appropriately equipped facility for a rapid evaluation. This procedure may be particularly beneficial if sedation is necessary to complete cranial imaging and lumbar puncture. Collaboration with radiology departments can be instrumental in completing magnetic resonance imaging, instead of following the minimal recommendations of computed tomography and the imaging of morbidly obese patients. Coordinated care with an ophthalmologist or neuro-ophthalmologist is instrumental for efficient diagnostic evaluation and management, especially in patients with severe findings of their visual examination [6,22,84]. Collaboration with an ophthalmologist is also instrumental in helping evaluate subtle findings of a visual examination in a patient suspected of manifesting idiopathic intracranial hypertension. Direct ophthalmoscopy may not be sensitive enough to diagnose papilledema, especially at stages 0, 1, and possibly 2 [22,52]. In patients with normal results of a direct ophthalmoscope examination but suspected of manifesting idiopathic intracranial hypertension, a dilated examination of the optic disc with indirect ophthalmoscopy and stereoscopic viewing may be required to detect subtle forms of papilledema [3,38]. In asymptomatic patients with swelling of the optic disc, ophthalmologists can undertake further evaluations, e.g., with fluorescein angiography. Additional evaluations can help exclude entities that mimic the papilledema caused by intracranial hypertension, such as drusen, disc crowding, and pseudopapilledema [22]. Ophthalmologists may also be helpful in evaluating uncooperative children. The visual examination, including visual acuity, the visual fields, and a direct ophthalmoscopy examination, can be particularly difficult in these individuals. The ophthalmologist may supplement the visual examination with different methods of testing such as tangent screening, color vision, or confrontation with toys that are more developmentally appropriate [3].

Because visual loss can occur any time in idiopathic intracranial hypertension and is often insidious as well as asymptomatic, the appropriate monitoring of vision by an ophthalmologist is necessary [65]. Wall and George demonstrated that visual acuity testing with a Snellen chart and visual field testing with finger confrontation are insensitive to worsening papilledema [35]. In conjunction with monitoring visual acuity, ophthalmologists can follow the visual fields with quantitative perimetry, and optic discs with stereoscopic viewing and stereo photographs. Ongoing communication and collaboration between the ophthalmologist and the neurologist in the chronic management of children with idiopathic intracranial hypertension is strongly encouraged, because collaboration may lead to more efficient care and better visual outcomes.

Discussion and Limitations

One limitation of the algorithm pertains to a lack of recommended time frames for diagnostic procedures. At present, no evidence-based data provide guidance for a specific timeline of evaluation. Because visual loss can occur any time during the course of idiopathic intracranial hypertension, the physician should undertake a diagnostic evaluation promptly, to initiate management for preserving vision and reducing signs.

Another limitation involves the lack of evidence regarding magnetic resonance venography in the evaluation of children with normal, mild, or moderate findings of a visual examination. Currently, no Class I guidelines pertain to appropriate initial cranial imaging in children presenting with signs of idiopathic intracranial hypertension. Future prospective, randomized, controlled studies are needed to identify the risk factors for thromboses in children with presumed idiopathic intracranial hypertension. Friedman and Jacobson suggested that a physician consider magnetic resonance venography in children with a recent history of sinus or otitis media infections, in those presenting with rapidly deteriorating visual changes, or in patients who do not respond to therapy [1]. An argument to perform magnetic resonance venography during the initial evaluation centers on the idea that visual outcomes may worsen with a longer duration of signs. However, two studies demonstrated no association between duration of signs and visual loss [67,85]. Rush [67] reviewed 63 patients with intracranial hypertension, both idiopathic and secondary. That study contained six patients with cerebral sinus venous thrombosis causing intracranial hypertension. No significant differences were evident in the median duration of signs in patients with poor visual outcomes (84 days) compared with patients with good visual outcomes (105 days)
[67]. Clearly more data are needed to determine which children need magnetic resonance venography in the initial evaluation of presumed idiopathic intracranial hypertension. Until further data are available to support the contention that every child should receive a magnetic resonance venogram during the initial evaluation of presumed idiopathic intracranial hypertension, the recommendations of this algorithm for venography during an initial evaluation pertain to children who are potential surgical candidates, who present with fulminating visual loss or a history of increased risk for veno-occlusive disease, or who do not respond to therapy.

A third limitation concerns the lack of evidence regarding a repeated lumbar puncture or continuous intracranial pressure monitoring in children who are suspected of manifesting idiopathic intracranial hypertension, but whose opening cerebrospinal fluid pressure is normal. As discussed previously, an elevated opening pressure may be missed because of fluctuations in cerebrospinal fluid pressures [48-50]. Therefore, in patients with signs of idiopathic intracranial hypertension, diagnostic testing, including transducer monitoring through a lumbar drain or intracranial pressure monitoring, should be considered. Soler et al. recommended monitoring in the preoperative evaluation of children with persistent signs and an inability to perform visual field testing, or in children without papilledema and unresponsive to medication [7].

Treatment and Clinical Course

Chronic Management

The goal of treatment for idiopathic intracranial hypertension is primarily twofold: to prevent visual loss and to eliminate symptoms. At present, no randomized, controlled, double-blind, prospective studies have evaluated possible treatments for children. Because the etiology of idiopathic intracranial hypertension remains unclear, a specific treatment tailored to the cause has not yet developed. Therefore, current treatments are tailored to treat signs. Further studies are desperately needed to develop better treatment plans.

Treatments that have been used in children include but are not restricted to a single lumbar puncture at diagnosis, repeated lumbar taps, carbonic anhydrase inhibitors, diuretics, weight loss management, and surgical procedures [7,25,52,57,86]. Just as the initial diagnostic evaluation requires collaboration among physicians, the ongoing management often involves multiple caregivers, including a primary care physician, neurologist, ophthalmologist, nutritionist, and psychologist or counselor. These physicians must collaborate and communicate any concerns, recommendations, and changes regarding a patient’s management. Because the needs of the patient often overlap caregiver domains, collaboration will help increase the efficiency of care and decrease redundancy.

In some reports, the signs of idiopathic intracranial hypertension resolved in a few children after a single, diagnostic lumbar puncture [14,44]. The reason for this response is unknown. Repeated lumbar punctures to decrease cerebrospinal fluid pressure are generally not recommended in the management of children with chronic idiopathic intracranial hypertension [7,25]. Lumbar punctures are not only difficult to perform in children, but often require sedation and are of questionable benefit, because the amount of cerebrospinal fluid drained will quickly be replenished.

Most children respond to typical medical treatments. Currently, the first line of therapy often involves a carbonic anhydrase inhibitor such as acetazolamide. Acetazolamide is thought to reduce the rate of cerebrospinal fluid production and therefore reduce cerebrospinal fluid pressure [87]. In younger children, most recommendations include treatment with 15-25 mg/kg/day divided into 2-3 doses [14,16]. A few studies reported higher doses of up to 100 mg/kg/day divided into 2-3 doses [19,57]. Postpubertal children are generally treated with adult dosing from 1-4 g/day, divided into 2-3 doses [7,52]. Side effects, including paresthesias, altered tastes of food and carbonated beverages, nausea, abdominal complaints, and fatigue, are generally dose-dependent, and may limit the effectiveness of the medication.

If acetazolamide is ineffective or intolerable, furosemide may be supplemented or substituted. The exact mechanism of lowering cerebrospinal fluid pressure with furosemide is not completely understood [88]. Schoeman reported that a combination of furosemide and acetazolamide reduced intracranial pressure more effectively in childhood idiopathic intracranial hypertension [19]. Another carbonic anhydrase inhibitor, topiramate, was recently described as effective in a case report [89]. Topiramate is similarly thought to reduce the production of cerebrospinal fluid at clinically relevant doses [90]. In a recent open-label study comparing acetazolamide and topiramate treatment for idiopathic intracranial hypertension, no significant difference was evident in visual outcomes between the two medications. However, Celebisoy et al. reported a significant weight loss in the topiramate group [91]. Although that report was of an open-label pilot study in a small group of patients, further studies may affirm that topiramate is as effective as acetazolamide in treating visual signs, and more effective in aiding weight loss.

Regarding weight loss, several case reports suggest that weight reduction in obese patients may help resolve idiopathic intracranial hypertension, and should be encouraged as a treatment modality [86,92]. Kupersmith et al. reported that weight loss was associated with a faster recovery from papilledema and visual field abnormalities [86]. Although that study was retrospective, it suggests that weight loss may constitute an important treatment in obese patients with idiopathic intracranial hypertension. With the trend toward increasing obesity, further studies are needed to analyze links between idiopathic intracranial hypertension...
and obesity, and the effects of weight loss on visual outcomes.

In general, the modalities used to treat visual abnormalities via lowering cerebrospinal pressure also improve headache. However, this is not always true. Other prophylactic medications such as beta blockers, tricyclic antidepressants, or antiseizure medications may be needed to treat chronic headaches [22,63]. Weight should be monitored closely, because weight gain is a possible side effect of some of these medications [63]. Acute medications such as acetaminophen or ibuprofen are appropriate for abortive therapy. Patients with idiopathic intracranial hypertension may be at risk for medication-overuse headaches, and should be encouraged to use these treatments only intermittently [37]. In patients with concomitant migraines, additional antimigraine medications may prove helpful [63,93].

Although most studies maintain that idiopathic intracranial hypertension is usually self-limited, no duration of standardized treatment has been established. The duration of treatment varies, and is driven by the presence and progression of visual loss and the disability attributable to headache [25]. A patient should be examined by an ophthalmologist at least monthly as treatment begins, to monitor closely for any visual field changes or loss, and to reinforce the patient’s comprehension of the potential for visual loss and to encourage the patient’s compliance [94]. Friedman and Jacobson recommended that when a patient’s visual status and optic nerve appearance have stabilized or after the disease has been in remission for at least 6 months, the medications used to lower cerebrospinal fluid pressure can be gradually withdrawn [44].

### Acute Management

Indications for admission for management and surgical evaluation include initial severe visual findings (Table 2) or a worsening of the results of a visual examination or a nonresponsive headache despite maximal treatments (Table 4) [52,94]. These patients may need acute measures to preserve their vision before possible surgery. Intravenous steroids are used as an acute short-term treatment, in addition to chronic management medications such as acetazolamide. Because steroids are thought to decrease intracranial pressure rapidly, intravenous steroids are often used as a presurgical stabilizing measure [96,97]. However, with the detrimental side effects of weight gain and rebound idiopathic intracranial hypertension during gradual withdrawal, steroids are not recommended as part of the chronic management of idiopathic intracranial hypertension in children.

For those refractory to maximal treatments with worsening eye examinations or in those initially diagnosed with severe visual findings, an emergent referral for consideration of neuro-ophthalmologic or neurosurgical procedures is indicated. Common surgical procedures include optic nerve sheath fenestration, lumbar peritoneal shunting, and ventriculoperitoneal shunting. At present, no prospective, randomized, clinical trials have compared the outcomes of these different therapies. Therefore, no procedure has been proven to be most efficacious.

Optic nerve sheath fenestration may be beneficial in preventing further acute visual deterioration [98,99]. Fenestration may prevent further visual loss, and has also helped relieve headache in up to two thirds of patients [51]. Proponents of fenestration claim that it is the superior treatment. However, up to 33% of patients receiving fenestration who demonstrate visual improvement initially may manifest later deterioration in visual function [100,101].

Cerebrospinal fluid shunting may be beneficial in treating acute visual loss and symptoms of intractable headache [102,103]. Although considered the superior treatment for headache relief, shunting also significantly improved or stabilized visual function [102,104]. Baker et al. recommended shunting for intractable headaches in the setting of progressive visual loss [57]. Complications of shunting may include dysfunction, infections, overdrainage, and iatrogenic Chiari I malformations [102,104]. Curry et al. reported that the number of new shunt placements for idiopathic intracranial hypertension increased by 320% from 1998-2002 [105]. The estimated proportion of patients receiving a shunt procedure during the course of idiopathic intracranial hypertension approaches 20% [102,106]. The procedure chosen for therapy may depend on several factors, such as the availability and expertise of the physician performing the procedure, the patient’s wishes, and the predominating signs. Each type of procedure has advantages and disadvantages, and neither procedure alone may resolve both the visual deterioration and intractable headaches.

### Prognosis

Permanent loss of visual function, regardless of treatment, was reported in children. Permanent decreased visual acuity occurs in up to 10% of children [15,58,107]. Permanent visual field defects occur in up to 17% [58]. In mixed adult and pediatric studies, blindness in one or both eyes developed in up to 5% of patients [35,65]. Findings of visual examinations or complaints such as transient visual obscurations, severity of papilledema, duration of signs, or the presence of cranial neuropathies have not been associated with worse visual outcomes [35,65,67]. One risk for worse visual outcomes may

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**Table 4. Surgical indications for patients undergoing medically treatment for idiopathic intracranial hypertension [94,95]**

<table>
<thead>
<tr>
<th>Indication</th>
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<tr>
<td>1. Recent visual field loss*</td>
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<tr>
<td>2. Progressive visual field loss in a preexisting visual field defect*</td>
</tr>
<tr>
<td>3. Reduction in visual acuity not attributable to macular edema</td>
</tr>
<tr>
<td>4. Severe headache not responsive to standard medical therapy</td>
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* Enlargement of blind spot is not considered a significant visual loss.
involve the patient’s weight. Wall and George reported a significant association between weight gain in the 12 months preceding a diagnosis of idiopathic intracranial hypertension and deterioration of vision [35]. Regarding symptoms of headache, it is not uncommon for patients to continue to require long-term headache management [93].

Although idiopathic intracranial hypertension is thought to be monophasic, recurrences can happen. The recurrence rate of increased intracranial pressure is reported as 6-22% [15,58,107]. It is highly recommended that patients are monitored for recurrence of visual changes, particularly in those who initially lose and later regain weight [15,44,58,79]. Several authors indicated that patients may manifest consistently elevated cerebrospinal fluid pressures despite the resolution of papilledema. However, the risk to vision attributable to chronically increased intracranial pressure without papilledema is not well understood [65,108].

Future Directions and Conclusions

Although the etiology of idiopathic intracranial hypertension currently eludes the medical profession, the continued search for that etiology is imperative, because identifying the cause could lead to better treatments. Idiopathic intracranial hypertension is an uncommon problem, and an algorithm to outline an efficient management strategy for children should be helpful. This algorithm serves as a first step toward the consistent management of children with this condition. The lack of prospectively collected data for such a critical problem illustrates the need for a collaborative examination of this condition, its risk factors, its rates of recurrence, and treatments. Physicians practicing in a more consistent manner across the country will provide a foundation for larger collaborative studies. These studies may provide information supporting modifications of the proposed algorithm and the subsequent development of better diagnostic criteria, standardized approaches to imaging, and management strategies.

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References


