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Idiopathic intracranial hypertension: Diagnosis, monitoring and treatment

Hypertension intracrâniennne idiopathique : diagnostic, suivi et traitement

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ABSTRACT

Introduction. – Idiopathic intracranial hypertension (IIH) is a disorder typically affecting young, obese women, producing a syndrome of increased intracranial pressure without identifiable cause.

State of the art. – Despite a large number of hypotheses and publications over the past decade, the etiology of IIH is still unknown. There continues to be no evidence-based consensus or formal guidelines regarding management and treatment of the disease. Treatment studies show that the diagnostic lumbar puncture is a valuable intervention beyond its diagnostic importance, and that weight management is critical. However, many questions remain, regarding the efficacy of acetazolamide, cerebrospinal fluid (CSF) shunting procedures, optic nerve sheath fenestration, and cerebral transverse venous sinus stenting. Identification of subgroups of patients at high-risk for irreversible visual loss, such as black patients, men, morbidly obese patients, and patients with fulminating IIH, helps determine management approaches and refine follow-up strategies.

Perspective. – Better understanding of the pathophysiology and ongoing clinical trials will hopefully help inform treatment strategies over the next few years.

RÉSUMÉ

Introduction. – L’hypertension intracrânienne (HIC) idiopathique affecte le plus souvent les femmes jeunes et reste sans étiologie claire. Elle consiste en un syndrome d’HIC isolée sans cause reconnue.

État des connaissances. – Malgré un grand nombre d’hypothèses et de publications au cours des dix dernières années, l’étiologie de l’HIC idiopathique reste mystérieuse. Il n’y a pas de consensus ou de recommandations officielles concernant la prise en charge et le traitement de l’HIC idiopathique. Certaines études ont montré que la ponction lombaire réalisée pour le diagnostic d’HIC idiopathique entraîne souvent une rémission prolongée des symptômes d’HIC. De plus, la perte de poids est essentielle dans la prise en charge des patients obèses. Cependant, de nombreuses questions restent posées au sujet de

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Idiopathic intracranial hypertension (IIH) is a syndrome characterized by isolated increased intracranial pressure (ICP) of unknown cause (Box 1). By definition, intracranial disorders such as a meningeal process, or cerebral venous thrombosis must be ruled-out to make a diagnosis of IIH (Friedman and Jacobson, 2002, 2004). Rarely, certain medications may induce a syndrome of isolated raised ICP, and these patients are still given the diagnosis of “IIH” according to the English nomenclature. Similarly, patients with isolated raised ICP who have cerebral transverse venous sinus stenoses (not thrombosis) are still conventionally classified as having “IIH” (Farb et al., 2003). Although imperfect, the term “IIH” is currently the preferred designation for this disorder in the English literature (“hypertension intracrânienne idiopathique” in the French literature (Biousse and Bousser, 2001), rather than “pseudotumor cerebri”, which often includes patients with other causes of raised ICP such as cerebral venous thrombosis, and “benign intracranial hypertension”, which is falsely reassuring considering that a number of IIH patients irreversibly lose vision (Fig. 1) (Digre, 2010; Karahalios et al., 1996; Wall, 2010).

This review emphasizes recent studies regarding the diagnosis, prognosis, monitoring, and management of IIH.


1. Signs and symptoms of increased intracranial pressure (headaches, nausea, vomiting, tinnitus, transient visual obscurations, papilledema) in an awake patient.
2. No localizing, focal neurologic signs, except unilateral or bilateral sixth nerve paresis.
3. Cerebrospinal fluid opening pressure more than or equal to 25 cm of water, but without cytologic or chemical abnormalities.
4. Normal neuro-imaging adequate to exclude cerebral venous thrombosis, (ideally magnetic resonance imaging of the brain, often with magnetic resonance venography).

The number of 25 cm of water is not an absolute cut-off, especially in children in whom cerebrospinal fluid opening pressures as high as 28 cm of water have been documented to be normal (Avery et al., 2011).

1. Diagnosis of idiopathic intracranial hypertension

The diagnosis of IIH is made in patients presenting with isolated increased ICP in whom an intracranial abnormality and a meningeal process have been ruled-out (Box 1) (Friedman and Jacobson, 2002).

1.1. Clinical features

Headache is the most common symptom at presentation and is reported by more than 90% of patients with IIH. It is often associated with tinnitus, typically pulsatile, which is described by at least 50% of IIH patients (Friedman and Rausch, 2002; Wall and George, 1991; Wall, 2010).

Papilledema is the most common sign of IIH and should be present (unless the patient has developed secondary optic atrophy) to make a diagnosis of IIH. Patients with chronic headaches from raised ICP, but who have normal-appearing optic nerves, are usually considered to have so-called “IIH without papilledema” and should be differentiated from IIH patients since the diagnosis and management of IIH without papilledema are very different (IIH patients without papilledema have no risk of visual loss) (Digre et al., 2009). It is not uncommon to diagnose IIH after papilledema is detected on a routine ophthalmic examination on asymptomatic patients, explaining why ophthalmologists often first diagnose this disease. This is particularly common in men who often do not complain of headaches (Bruce et al., 2009).

Transient visual obscurations are very brief episodes of blurred vision, which may be uni- or bilateral and are classically precipitated by bending over. They occur in about two-thirds of patients with papilledema from IIH, and they are not associated with poor visual outcome (Wall and George, 1991; Wall, 2010). They do not occur in IIH patients without papilledema. Papilledema is associated with visual field changes in almost all patients, ranging from reversible enlarged blind spots (Fig. 2) to devastating permanent severe constriction of visual fields (Fig. 1). Central visual acuity remains normal until late in the course of the disease, unless there is macular edema or exudates. Because visual field loss is progressive and often insidious, formal perimetry is essential in the initial evaluation and follow-up of IIH patients (Wall and George, 1991; Wall, 2010).
Binocular diplopia is less common, and is usually related to a uni- or bilateral sixth nerve palsy (or to divergence insufficiency) secondary to raised ICP; it usually improves or resolves immediately after the diagnostic lumbar puncture (Wall, 2010).

1.2. Optical coherence tomography

The role of optical coherence tomography (OCT) in the evaluation of papilledema remains unclear. A few studies have proposed an adjunctive utility of optic nerve imaging by

Fig. 1 – Very severe bilateral papilledema with profound visual loss in a young obese woman with fulminant idiopathic intracranial hypertension (IIH). This patient presented with 10 days history of severe unusual headaches and developed decreased vision in both eyes 1 week later. She had visual acuities of “no light perception” in the right eye and “count fingers” in the left eye at initial presentation. Funduscopic examination showed severe bilateral hemorrhagic papilledema (A). There were white exudates on the discs, and the optic nerve heads could not be seen (the right eye is on the left side of the picture and the left eye is on the right). The patient immediately underwent brain MRI and MRV with contrast which showed only bilateral transverse sinus stenosis (and no thrombosis); a lumbar puncture showed normal CSF contents with CSF opening pressure of 55 cm. A lumbar drain was placed immediately and she received 1 g of intravenous methylprednisolone. Her headaches dramatically improved, and her vision improved to count fingers in the right eye within a few hours. A ventriculoperitoneal shunt was placed the following day. Vision continued to improve in the right eye, and a few weeks later, she had developed severe bilateral optic atrophy (B); visual acuity was 20/40 in the right eye with very severe visual field constriction and “hand motion” in the left eye with a very small island of residual vision (C). She remained legally blind, but able to ambulate by herself and use low vision aids to read a few words at-a-time.
OCT in monitoring the thickness of the peripapillary retinal nerve fiber layer during the course of IIH (Heidari and Rizzo, 2010). However, the reductions in the retinal nerve fiber layer observed, as disc edema resolves cannot be easily differentiated from axonal loss reflecting disease worsening rather than improvement. At this time, quantification of visual function by ophthalmic examination and automated perimetry remains much more helpful than anatomical characterization of the optic nerve with OCT in the monitoring of patients with IIH.

1.3. **Brain imaging**

Although brain imaging must be “normal” in order to diagnose IIH (Friedman and Jacobson, 2002), imaging changes associated with raised ICP are commonly observed in IIH patients (Degnan and Levy, 2011). These include an empty sella or flattening of the pituitary gland, flattening of the posterior globes, protrusion of the optic nerve heads, enhancement of the prelaminar portion of the optic nerve heads, distension of the optic nerve sheaths, and vertical tortuosity of the optic nerves. Although cerebral venous thrombosis must be ruled-out in patients with suspected IIH (Biousse et al., 1999), numerous studies from the last decade have shown that unilateral stenosis of the dominant transverse sinus or bilateral stenoses of the transverse sinuses, often with measurable hemodynamic gradients, are common in typical IIH patients (Farb et al., 2003).

1.4. **Cerebrospinal fluid (CSF) opening pressure**

The normal limit for CSF opening pressure remains controversial and varies from one patient to another. The classic IIH criteria (Friedman and Jacobson, 2002) require a CSF opening of at least 25 cm of water. However, this limit must be adjusted to the clinical presentation, and must take into consideration that in some asymptomatic patients (particularly children), a pressure as high as 28 cm of water might be normal (Avery et al., 2011). Similarly, a CSF opening pressure of 21 cm of water in a patient with symptoms of raised ICP and frank papilledema on funduscopic examination should still be

Fig. 2 – Moderate bilateral papilledema with enlarged blind spots on automated perimetry in a young woman with idiopathic intracranial hypertension. This obese young woman presented with new-onset chronic daily headaches for the past 2 months and reported a few episodes of black-out of vision in both eyes when bending over (transient visual obscuration); she also had mild bilateral tinnitus. Funduscopic examination showed bilateral moderate papilledema with a few hemorrhages (A). The optic nerves were diffusely swollen and there were choroidal folds in the left eye (the left eye is on the right side of the picture and the right eye is on the right). Visual acuity was normal and Humphrey visual fields performed using the 24-2 SITA Fast strategy, showed enlarged blind spots in both eyes (B). Brain MRI was normal and the lumbar puncture showed normal CSF contents with CSF opening pressure of 37 cm of water. The patient's headaches improved dramatically after the diagnostic lumbar puncture; she was treated with acetazolamide 500 twice a day and was given recommendations for weight loss. The papilledema resolved within a few months and her visual function remained normal. She discontinued the acetazolamide 7 months later.
considered compatible with a diagnosis of IIH. Diagnostic confusion often results from anomalous optic discs in patients with borderline increased ICP, and CSF opening pressures less than 25 cm of water in patients with presumed IIH should always be interpreted with caution.

2. **Factors influencing the prognosis of idiopathic intracranial hypertension**

### 2.1 Prognosis of idiopathic intracranial hypertension

In most patients, IIH has a self-limited course, and symptoms and signs resolve with the diagnostic lumbar puncture, weight loss and medical treatment. In others, chronic symptoms (mostly headaches) result in altered quality of life. About 25% of IIH patients have some irreversible visual loss, sometimes resulting in blindness, despite normalization of the ICP. Long-term follow-up of a small North American cohort (Shah et al., 2008) suggested that IIH can also be a chronic disease, with delayed worsening and recurrence.

Many IIH patients suffer from persistent headaches, even after normalization of the ICP, and frequently require the continued involvement of a neurologist for adequate treatment (Kleinschmidt et al., 2000). Patients with IIH also experience depression, anxiety, and reduced quality of life (Kleinschmidt et al., 2000). These reductions of quality of life are more than would be expected based on the patients’ persistent headaches and co-morbid obesity (Daniels et al., 2007; Kleinschmidt et al., 2000).

Papilledema can result in insidious and slowly progressive visual loss, which is usually reversible with appropriate treatment. However, up to 25% of IIH patients develop secondary optic atrophy and associated permanent visual loss (Fig. 1) (Corbett et al., 1982; Digre, 2010; Wall and George, 1991; Wall, 2010). Visual loss is usually relatively mild at presentation but progresses insidiously. In fact, most patients have visual field defects on automated perimeter at presentation, but are unaware of their visual dysfunction (Wall, 2010). Visual field abnormalities typically progress from enlargement of the physiologic blind spot, to nasal and arcuate defects, and ultimately to severe visual field constriction (Figs. 1 and 2). All IIH patients must be monitored carefully with formal automated perimeter, with intervention taken whenever deterioration of visual fields is documented (Corbett and Thompson, 1989; Wall, 2010).

Predicting which patient might be at risk for severe visual loss when evaluating new patients with presumed IIH is challenging, but essential, as it determines the strategies chosen for monitoring and management (Fig. 3). Several demographic risk factors for visual loss in IIH have been identified (Box 2).

### 2.2 Obesity

IIH occurs most frequently among obese women of childbearing age. Daniels et al. (2007) showed a dose relationship of higher body mass index (BMI) associated with a greater risk of IIH. Interestingly, this study also showed that even non-obese patients (BMI < 30) were at greater risk for IIH if they had a recent moderate weight gain as low as 5 to 15% of their weight. Another recent study (Ko et al., 2011) showed that a group of 26 women with recurrent IIH had a greater BMI at the time of recurrence compared to their BMI at the time of initial diagnosis. This study also suggested that even moderate weight gain might be a risk factor for recurrence. Additionally, Szewka et al. (2012) demonstrated that increasing degrees of obesity were associated with an increased risk of severe visual loss. The exact relationship between obesity and IIH remains poorly understood, with several etiologic hypotheses proposed, including increased central venous pressure and various hormonal and metabolic changes commonly found in obese patients (Ooi et al., 2008).

### 2.3 Gender

Only about 10% of IIH patients are men (Bruce et al., 2009; Digre and Corbett, 1989). While affected men have a similar BMI when compared to affected women, they are, on average, about a decade older than women at the time of presentation. Although men develop IIH less frequently than women, their visual prognosis is worse, perhaps because they have less headaches to alert them to the problem (Bruce et al., 2009). In one study by Bruce et al. (2009), men were more likely to have worse visual acuity and visual fields at presentation and follow-up, with the odds of severe visual loss for men at least double those of women. Thus, men also likely need more aggressive intervention when visual loss ensues, and closer follow-up given their lower propensity for headache (Bruce et al., 2009).

### 2.4 Race

Although race does not seem to influence the incidence of IIH, race appears to be an important determining factor of a patients’ visual prognosis, worse in black patients compared with whites living in the US (Bruce et al., 2008), and worse in white US IIH patients compared with white French IIH patients (Mrejen et al., 2009). Bruce et al. (2008) showed that black patients with IIH were three times more likely than non-black patients to have severe visual loss in at least one eye and were nearly five times more likely to be blind in both eyes. In this study, this difference in visual prognosis was likely unrelated to racial differences in diagnosis, treatment, or access to care, suggesting that black patients with IIH appear to have a more aggressive disease and, therefore, need closer follow-up and a lower threshold for early aggressive

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Box 2. Factors independently associated with a worse visual outcome in idiopathic intracranial hypertension.

- Male gender
- Race (black patients)
- Morbid obesity
- Anemia
- Obstructive sleep apnea
- Acute onset of symptoms and signs of raised intracranial pressure (fulminant IIH)
2.5. **Medications**

While various medications have been proposed to cause or, more likely, to precipitate IIH (such as tetracycline and its derivatives, cyclosporine, lithium, nalidixic acid, nitrofurantoin, oral contraceptives, levonorgestrel, danaxol, and tamoxifen) (Wall, 2010), there are compelling test-retest data only in a few individual patients (such as in patients treated with tetracyclines, particularly minocycline) (Friedman, 2005). The prognosis of IIH precipitated by medications tends to be good, with a self-limited course once the drug is discontinued (Kesler et al., 2004). However, fulminant IIH has been described in patients with severe increased ICP precipitated by minocycline (Fraser et al., 2012) and visual loss should not be overlooked in these patients.

2.6. **Obstructive sleep apnea**

Obstructive sleep apnea (OSA) is also associated with IIH (Wall, 2010). It is unclear, however, whether obesity is the common pathophysiological link, or whether OSA is another factor capable of triggering IIH in predisposed patients. It has also been suggested that OSA might portend a worse prognosis for IIH patients (Bruce et al., 2009; Wall and Purvin, 2009).

2.7. **Anemia**

Anemia at the time of diagnosis has been associated with worse visual outcome in IIH patients (Bioussé et al., 2003; Bruce et al., 2008). In a few patients, prompt treatment of the anemia resulted in dramatic improvement of visual function (Bioussé et al., 2003).
2.8. Hypertension

Although no study has definitely linked high blood pressure with a worse outcome in IIH, a few studies have suggested a trend towards worse visual outcome in IIH patients with poorly controlled hypertension (Bruce et al., 2008; Wall and George, 1991).

2.9. Absence of headaches

Almost all patients with IIH first seek medical attention because of headaches. Patients without headaches at presentation are likely to be diagnosed at a much later stage, when asymptomatic papilledema is already responsible for irreversible visual field changes. This might explain why the absence of headache is associated with a worse visual outcome in IIH, and might contribute to the worse outcome of men with IIH, who are less likely to report headaches than women with IIH (Bruce et al., 2009).

2.10. Fulminant course

Rarely, patients with IIH may have an acute onset of symptoms and signs of raised ICP (so-called “fulminant” or “malignant” IIH), with rapid worsening of visual loss over a few days, but meet the criteria for IIH with normal brain MRI and venography (Fig. 1). In a study of 16 such “fulminant IIH” cases (Thambisetty et al., 2007), surgical treatment was required because of ongoing severe visual loss in all patients; 50% remained legally blind and the visual fields remained severely impaired in all cases. Prompt recognition and early aggressive treatment of patients with fulminant IIH is therefore essential.

3. Management of idiopathic intracranial hypertension

The two goals of treatment in IIH are to alleviate symptoms of increased ICP, particularly headaches and diplopia, and to preserve vision (Fig. 3).

General recommendations include evaluation and treatment of potential contributing factors (such as weight gain and obesity, medication use, anemia, and OSA), weight loss, and medical headache management. Routine screening of IIH patients with the Berlin questionnaire for OSA (brief and validated screening questionnaire which includes questions about snoring, daytime somnolence, BMI and hypertension) has been shown to be a reliable metric in the young population commonly affected by IIH (Thurtell et al., 2011). Overnight, polysomnography is usually obtained when the clinical suspicion for OSA is high in IIH patients (Thurtell et al., 2011; Wall and Purvin, 2009).

Immediate management is primarily based on the duration of symptoms, evaluation of visual function and patients’ characteristics, and include the diagnostic lumbar puncture as well as a number of medical and surgical approaches, although most proposed treatments have not been evaluated in controlled randomized studies.

3.1. Diagnostic lumbar puncture

Not surprisingly, the diagnostic lumbar puncture often results in dramatic improvement of symptoms and signs; however, it is not uncommon to observe a lasting clinical remission following a single lumbar puncture in some IIH patients (De Simone et al., 2005), obviating the need for further medical or surgical treatment. Because the CSF, removed at the time of the lumbar puncture, is renewed within a few hours, this lasting improvement cannot be simply explained by the amount of CSF drained, or by the hole made in the dura by the needle used for the lumbar puncture. Instead, the interaction of various factors, such as CSF formation, compliance, cerebral blood flow, and outflow resistance, which can lead to multiple stable and unstable equilibrium levels of ICP in IIH, most likely explains this phenomenon, as suggested by mathematical models of CSF hydraulics (Bateman et al., 2009). The most classic hypothesis is that if a single lumbar puncture lowers the CSF pressure beyond the threshold level of an unstable equilibrium, the ICP must settle into a lower stable pressure state until other factors cause the pressure to again exceed that threshold. This observation emphasizes the importance of the initial lumbar puncture as a therapeutic procedure in IIH in addition to its diagnostic importance. It also explains why some patients dramatically improve after two or three lumbar punctures or require a repeat lumbar puncture every few months to remain asymptomatic.

3.2. Weight loss

Weight loss is a critical part of the treatment of overweight and obese IIH patients. Although it is likely that aggressive weight control is an efficient way to improve the overall quality of life of obese IIH patients, only a modest degree of weight loss (about 5–10% of total body weight) is usually required for improvement in symptoms and signs (Wong et al., 2007). A recent study of 25 obese women with IIH showed that weight loss effectively reduces not only headaches and papilledema, but also ICP (Sinclair et al., 2010).

Although, weight loss is not an effective short-term treatment, it is essential to emphasize the importance of weight loss and the prevention of weight fluctuation in the long-term management of obese or overweight IIH patients in order to minimize the risk of recurrence (Ko et al., 2011). Bariatric surgery can be considered in morbidly obese IIH patients in whom attempts at weight loss have been unsuccessful or in whom other medical morbidities of obesity already coexist (Egan et al., 2011; Fridley et al., 2011).

3.3. Medical treatment

Carbonic anhydrase inhibitors, such as acetazolamide (500 mg to 1 g twice a day), are the most commonly prescribed medications for IIH. Although no trial data are currently available to confirm its effectiveness, acetazolamide has always been considered the treatment of choice for IIH because it decreases the production of CSF in humans (Rubin et al., 1966). The preliminary results of the IIH Pilot Trial from Birmingham, UK, (Ball et al., 2011) in which 50 patients were randomized to receive acetazolamide or no acetazolamide,
emphasized the practical difficulties of performing such a study because of poor recruitment and medication side
effects, particularly paresthesias, altered taste sensation,
and lethargy. A multicenter, double-blind, placebo-controlled
clinical trial, called the Idiopathic Intracranial Hypertension
Treatment Trial (IIHTT), is currently enrolling patients in the
USA (http://www.nordicclinicaltrials.com/). This trial compa-
res the efficacy of acetazolamide and placebo in the treatment
of IIH patients with moderate visual field defects. All patients
are also treated with a low-sodium diet and participate in a
standardized weight loss program. This trial will hopefully
clarify the efficacy of acetazolamide and weight loss in IIH
patients with moderate visual loss.

Topiramate has weak carbonic anhydrase inhibition pro-
erties and has also been suggested for the treatment of IIH,
particularly for the treatment of headaches (Celebisoy et al.,
2007). A small study (Celebisoy et al., 2007) of 40 patients
comparing acetazolamide with topiramate did not show any
difference in the outcome between the two groups, but
demonstrated significant weight loss in the topiramate treated
group, suggesting that topiramate may have advantages in the
treatment of patients with IIH, although a different side-effect
profile.

Oral steroids were used in the past to treat IIH, but are
associated with long-term side effects, such as weight gain,
and therefore should not be prescribed. However, high-dose
intravenous steroids are still occasionally used in patients
with severe visual loss from fulminant IIH while a surgical
treatment is organized (Thambisetty et al., 2007).

3.4. Surgical treatment and venous stenting

A surgical procedure is required in patients with a fulminant
onset of disease or when other treatments have failed to
prevent progressive visual loss. More rarely, surgery may be
performed for refractory headaches related to chronically
elevated ICP. The choice of procedure depends on local
resources, as well as the patient’s symptoms and signs
(Table 1 and Fig. 3).

In patients with papilledema who have severe visual loss,
but minimal or no headache, optic nerve sheath fenestration
is often advised (Alsuhaibani et al., 2011), while in those with
visual loss, papilledema, and headache, a CSF diversion
procedure, such as ventriculoperitoneal or lumboperitoneal
shunting, is often preferred. Aggressive management with
CSF shunting is usually required to prevent catastrophic
visual loss in those with acute and rapidly progressive visual
loss (Thambisetty et al., 2007). These patients might benefit
from a transient lumbar drain while awaiting a more
definitive surgical procedure. Although no prospective study
has effectively compared ventriculoperitoneal shunts with
lumboperitoneal shunts, a large number of retrospective
series (Abubaker et al., 2011; El-Saadany et al., 2012; Sinclair
et al., 2011; Tarnaris et al., 2011; Ulivieri et al., 2009) have
suggested that both techniques are equally efficient in
controlling clinical manifestations of IIH as well as reducing
the ICP in the immediate postoperative period. Despite the
high rate of failure and complications, CSF shunting pro-
cedures remain the most widely performed surgical treat-
ment for IIH and are usually very useful acutely to prevent or
treat devastating visual loss in selected patients. It is,
however, important to emphasize that most IIH patients
have a relatively mild and self-limited course, and will not
require any surgical treatment.

Stenting of transverse venous sinus stenoses reduces
cerebral venous pressure, reduces ICP, and improves symp-
toms and signs in selected IIH patients (Ahmed et al., 2011a,b;
Albuquerque et al., 2011). However, endovascular venous
sinus stenting can result in serious complications, such as
stenosis migration, venous sinus perforation, in-stent thrombo-
sis, subdural hemorrhage, and the development of recurrent
stenoses immediately proximal to the stent (Ahmed et al.,
2011a). Until more data are available on the safety and long-
term outcome of venous stenting in young IIH patients, this
procedure should be limited to selected patients with bilateral
TSS, and refractory symptoms and signs of increased ICP, who
cannot undergo (or have failed) more conventional surgical
treatments.

<table>
<thead>
<tr>
<th>Table 1 - Surgical treatments in idiopathic intracranial hypertension.</th>
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<tbody>
<tr>
<td><strong>CSF shunting procedures (performed by neurosurgeons)</strong></td>
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<tr>
<td>Derivation of CSF drainage into the peritoneum</td>
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<tr>
<td>Lumboperitoneal shunt or ventriculoperitoneal shunt</td>
</tr>
<tr>
<td>Preferred when headaches are severe</td>
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<tr>
<td>Obstruction or disconnection require a revision in about 50% of CSF shunts</td>
</tr>
<tr>
<td><strong>Optic nerve sheath fenestration (performed by ophthalmologists)</strong></td>
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<tr>
<td>Decompression of the optic nerve by making a window into its dural sheath from a transconjunctival medial or lateral approach</td>
</tr>
<tr>
<td>Done on the eye with worst visual function first—often needs second eye surgery (although fellow eye improvement may occur after operation on the first eye)</td>
</tr>
<tr>
<td>Preferred when visual loss is predominant and headaches mild</td>
</tr>
<tr>
<td>The fenestration fails in one-third of cases within 3 years</td>
</tr>
<tr>
<td><strong>Endovascular transverse venous stenting</strong></td>
</tr>
<tr>
<td>Treatment of intracranial venous hypertension by stenting a stenosed transverse sinus.</td>
</tr>
<tr>
<td>Only one transverse sinus is stented in patients with bilateral transverse sinus stenosis (or stenosis of a dominant venous sinus and contralateral hypoplastic transverse sinus)</td>
</tr>
<tr>
<td>Preferred in selected patients with bilateral transverse sinus stenosis, and refractory symptoms and signs of increased ICP, who cannot undergo (or have failed) more conventional surgical treatments</td>
</tr>
<tr>
<td>The stent can result in serious complications, such as stent migration, venous sinus perforation, in-stent thrombosis, subdural hemorrhage, and recurrent stenosis may later develop immediately proximal to the stent</td>
</tr>
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4. Special situations

4.1. Fulminant idiopathic intracranial hypertension

Rarely, IIH may present acutely over a few days to 2 weeks. These patients usually complain of very severe headaches associated with rapid loss of vision. In this setting, IIH is a diagnosis of exclusion and venous sinus thrombosis must be definitely ruled-out. Because vision deteriorates quickly (and often irreversibly), treatments aimed at decreasing the ICP must be initiated immediately. The diagnostic lumbar puncture often stabilizes vision for a few hours; intravenous methylprednisolone is often administered in association with a lumbar drain until more definitive treatments (most often a CSF shunting procedure; sometimes an optic nerve sheath fenestration) can be arranged (Thambisetty et al., 2007).

4.2. Pregnancy

Because it is a disease which predominantly affects young women of childbearing age, IIH is often seen in pregnant women; however, it is important to emphasize that IIH is not more common in pregnant compared to non-pregnant women (Digre et al., 1984; Huna-Baron and Kupersmith, 2002). IIH can be diagnosed at any time during pregnancy, but most new cases occur in the first or second trimester. Any symptom of presumed “IIH” at the end of pregnancy or in the postpartum period should be concerning for cerebral venous thrombosis which can mimic IIH (Biousse et al., 1999).

The management of IIH can be challenging during pregnancy. Weight gain should be limited to recommended healthy levels. If medical therapy is deemed necessary, consideration must be given to potential teratogenicity of medications. Acetazolamide has shown adverse effects in pregnant animals, including rare limb anomalies in offsprings. However, this medication has not been documented to cause any adverse effect on pregnancy in humans, or to cause congenital malformations, including when started in the first trimester (Lee et al., 2005). Loop diuretics can be used with caution for a short period during pregnancy but thiazide diuretics are contraindicated. A short course of corticosteroids can be used in pregnancy, but should be reserved for patients with rapidly progressive visual loss (fulminant IIH) and should not be prescribed chronically. More invasive measures are necessary for intractable headaches or when severe papilledema results in visual field deficits. Serial lumbar punctures are often performed during pregnancy. Indeed, it is usually best to try to delay any surgical intervention until after delivery, or at least until the third trimester. Optic nerve sheath fenestration can be performed without difficulty in pregnant patients, as long as the risk directly related to anesthesia is deemed minimal by the obstetrical team. Cerebrospinal fluid shunting procedures (lumboperitoneal or ventriculoperitoneal shunts) are usually avoided during pregnancy. However, it is usually safe to leave indwelling shunts in place when a woman with IIH becomes pregnant. IIH does not interfere with pregnancy and these patients have the same rate of spontaneous abortion as the general population. Method of delivery, anesthesia and analgesia should be based only on obstetrical considerations (Digre et al., 1984). Indeed, although vaginal delivery is associated with a severe increase in intracranial pressure, even in normal patients, it is transient and does not alter the prognosis of IIH.

4.3. Children

Children may develop IIH similar to adults (Ko and Liu, 2010). Boys are affected as often as girls before puberty, and obesity is not as common in pre-pubertal children as it is in post-puberty or in adults. Therefore, recommendations for weight loss should be based on the age and BMI of the child. However, the effect of papilledema on vision is the same in children as in adults, and fulminant IIH may also occur in children with devastating visual loss. The medical and surgical treatment is similar to that of adults, although endovascular stenting of transverse sinus stenosis is not performed in children (Ko and Liu, 2010).

5. Conclusion

Although the pathophysiology of IIH remains unknown, identification of subgroups of patients at high-risk for irreversible visual loss, such as black patients, men, morbidly obese patients, and patients with fulminant IIH helps determine management approaches and follow-up strategies. All overweight IIH patients should receive counseling regarding weight loss, which is important in the long-term prognosis of IIH and may reduce the risk of recurrence. Co-morbidities such as hypertension, chronic anemia and OSA likely increase the risk of visual loss and need to be detected and appropriately treated. In the absence of evidence-based guidelines, medical and surgical treatments are individually tailored to the patient’s clinical presentation, initial visual function and estimated risk of visual loss. Finally, many IIH patients continue to complain of chronic headaches despite resolution of papilledema and normalization of the intracranial pressure. Analgesic abuse-induced headaches, chronic tension headaches and depression are particularly common in this population and should be specifically managed.

Disclosure of interest

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References


