Learning Objectives:
At the end of this program, participants will be able to:

1. Describe normal fluctuation in cerebrospinal fluid pressure
2. Explain the development of papilledema in response to intracranial hypertension based on animal and human studies
3. Convey the natural history of IIH with respect to the persistence of headaches

CME Questions:

1. Which of the following is NOT required to make a diagnosis of idiopathic intracranial hypertension?
   A. CSF opening pressure > 250 mm water (adults)
   B. Normal CSF contents
   C. No evidence of space occupying process or abnormal enhancement on neuroimaging studies
   D. Papilledema
   E. Normal sensorium

2. The most common presenting complaint of patients who have IIH without papilledema is:
   A. Chronic daily headaches
   B. Transient obscurations of vision
   C. Intracranial noises
   D. Episodic migraines
   E. Awareness of physiologic blindspot

3. Which of the following statements about IIH without papilledema is FALSE?
   A. It tends to occur in young, overweight women
   B. Chronic headaches are common
   C. The usual treatments for IIH (acetazolamide, shunting procedures) usually produce relief of symptoms
   D. Analgesic overuse must be excluded as the cause of headaches
   E. Prolonged CSF pressure monitoring may be needed to confirm the diagnosis

Key Words: Papilledema, Idiopathic intracranial hypertension, chronic daily headache

What is Normal Cerebrospinal Fluid Pressure and How Much Does it Fluctuate?

Normal cerebrospinal fluid (CSF) pressure in adults, measured by lumbar puncture with the patient in the lateral decubitus position, varies between 80 and 200 mm of water. Contrary to popular belief, CSF pressure is not dependent on weight or height (1), although pressure readings may be spuriously elevated when the patient coughs, strains, or holds his or her breath during the procedure. Measurements between 201-249 mm water are not diagnostic and those greater than 250 mm water are elevated. Normal values have not been well established in children but are generally accepted to be 200 mm water or less. Additionally, ICP in normal persons and in patients with increased ICP can vary widely from one moment to the next.

Adson and Lillie (2) introduced a needle into the lateral ventricle of a patient with a glioma of the frontal lobe and made continuous readings of the ICP at hourly intervals for four days and recorded variations in pressure that ranged from 130 to 980 mm of water! Fifty years later, Gucer and Viernstein implanted a small epidural pressure sensor in four patients with pseudotumor cerebri (PTC) and continuously monitored the patients before and after medical and surgical treatment. ICP before treatment showed irregular variations ranging from 50 to 500 mm of water over a 24 hour period (3). Their findings support the claim of Ford and Murphy in 1939 that “… one determination of intracranial pressure in a case of disease of the central nervous system (CNS) is no more instructive than one determination of a patient’s temperature during the course of a fever” (4).

Diagnostic Criteria for Idiopathic Intracranial Hypertension (IIH) and Rationale for 2002 Revisions

The diagnostic criteria for IIH were revised by Friedman and Jacobson in 2002 to reflect several aspects of diagnosis that were not adequately addressed.
by J. Lawton Smith’s “modified Dandy criteria” of 1985 (5). Specifically, the revised criteria allowed for diagnosis of IIH in an asymptomatic patient. While a somewhat strange concept for neurologists, ophthalmologists occasionally encounter patients who have incidental papilledema discovered on a routine eye exam. In criteria 3, the authors, frustrated with their own experience of treating patients who were diagnosed with no lumbar puncture (LP) or with a cerebrospinal fluid (CSF) pressure measured with the patient in a seated or prone position, specified and strengthened the language regarding the requirements of the LP. The neuroimaging requirements were also modernized, as a plain computed tomography is inadequate to exclude many of the secondary causes of PTC, particularly venous sinus thrombosis. Finally, the wording of criterion #2 also allowed the diagnosis of IIH without papilledema although the authors cautioned that a second lumbar puncture or prolonged intracranial pressure (ICP) monitoring might be necessary to confirm the diagnosis.

**Diagnostic Criteria for Pseudotumor Cerebri**

1. Symptoms, if present, reflect only those of increased intracranial pressure or papilledema
2. Signs, if present, reflect only increased intracranial pressure or papilledema
3. Documented elevated intracranial pressure during lumbar puncture measured in the lateral decubitus position
4. Normal cerebrospinal fluid composition
5. No evidence of ventriculomegaly, mass, structural, or vascular lesion on magnetic resonance imaging or contrast-enhanced computed tomography for typical patients, and magnetic resonance imaging and magnetic resonance venography for all others
6. If no other cause (including medication) of intracranial hypertension is identified, the syndrome is termed “idiopathic intracranial hypertension”.

**Pathogenesis of Papilledema**

Any unifying mechanism for papilledema must account for (1) transient unilateral or bilateral visual obscurations, (2) edema localized to the optic nerve head, (3) flattening of the posterior sclera, as demonstrated by neuroimaging techniques, and (4) venous distension and cessation of venous pulsations (6). Various hypotheses have been proposed over the years, including direct infiltration of the optic nerve head by CSF (7,8), obstruction of posterior flow of intraocular fluid (9), inherent turgescence of prelaminar nerve substance (10), swelling of glia (11), absence of the restraining influence of Müller cells in the peripapillary retina (12), disequilibrium of hydrostatic pressure in the tissue and blood stream (13), and compression of the central retinal vein as it traverses the subarachnoid space (13) or cavernous sinus (14) with elevated central venous pressure (6).

An animal model of experimental papilledema, developed in the 1960s, provided a means to test most of the theories regarding the pathogenesis of papilledema. Increased ICP has been produced in monkeys by cranial irradiation (15) and by the introduction of an inflatatable balloon into the subarachnoid space (16-18). Using these models, there are some general points of agreement regarding the pathogenesis of papilledema. First, papilledema occurs only when there is patency of the subarachnoid space surrounding the optic nerve and intracranial structures (19-21). Blockage of these spaces by adhesions or tumor prevents papilledema from occurring on the side of the obstruction. Second, papilledema does not occur when antecedent optic atrophy has destroyed most or all of the nerve fibers. Finally, and most important, axonal transport is clearly abnormal in patients with papilledema as well as in patients with disc swelling from all other causes (e.g., ischemia, inflammation, hypotony or low intraocular pressure).

Othograde transport is studied using autoradiography by injecting labeled amino acids (usually leucine or proline) into the vitreous cavity. Both the fast (+ 400 mm/day) and slow (+ 1 mm/day) components of axon transport accumulate in the regions of the lamina cribrosa of monkeys with experimentally produced increased ICP and papilledema (22,23). The role of central retinal venous pressure in papilledema is controversial. If the CSF pressure in the optic nerve sheath constantly fluctuates, as it does in the brain, the sheath, like the spinal dural sac, could function as an expansion vessel (6). Unlike the extradural veins of the spinal sac, the central retinal vein is located within the subarachnoid space. With markedly elevated ICP, the venous pressure in the optic nerve sheath could be double that of the eye, accounting for the loss of spontaneous venous pulsations and the presence of choroidal folds, acquired hypermetropia, shortening of the globe on MRI scanning and
forward bowing of the lamina cribrosa (6). However, experimental compression of the central retinal vein at its site of exit from the optic nerve does not produce optic disc edema (24). Similarly, monkeys exposed to raised CSF pressure for up to two hours showed increased ophthalmic vein pressure but not disc edema (25).

Research to date suggests that there are three components are required for the development of papilledema: (1) increased and fluctuating pressure in the distal optic nerve sheath, (2) elevated central retinal venous pressure, and (3) impaired perfusion of the neurons traversing the lamina cribrosa (6). Despite the findings described above, numerous questions still exist regarding the pathogenesis of papilledema (26).

How Quickly Does Papilledema Develop with an Acute Rise in Intracranial Pressure?

The rapidity of developing papilledema depends to a large extent on the etiology and degree of the increased ICP. Experimental models using inflat- able balloons in the subarachnoid space of monkeys showed papilledema in 30% of the animals in 24 hours, in 50% after 2 days and in 90% after 5 days when papilledema was assessed using direct ophthalmoscopy, stereoscopic photography and fluorescein angiography (27,28). Most of the animals developed papilledema from the surgery even prior to the balloon being inflated (27) and papilledema took 1 to 3 weeks to re-develop after the post-operative edema resolved and the balloon was inflated (27).

Clinical experience with humans suggests that the development of optic disc edema may be protracted. The largest study investigating this process was performed in 37 patients with acutely elevated ICP from cerebral hemorrhage or trauma receiving continuous ICP monitoring (29). Using both direct and indirect ophthalmoscopy, no papilledema was observed in the 13 patients with mildly elevated ICP (20-30 mm Hg) and only two patients had venous congestion by day six. In seven patients with ICP ranging from 30-70 mm Hg, papilledema developed in one patient with a subarachnoid hemorrhage who developed fatal cerebral edema and carotid artery distribution infarction post-operatively. No optic disc or fundus abnormalities were seen in the other 6 patients. Within the first three days of developing transient increased ICP (prior to successful treatment), none of 17 patients had papilledema. This experience suggests that clinically detectable papilledema is not usually present with acute elevations of ICP, and may take over a week to develop.

Cases have been reported of papilledema within 2 to 4 hours after intracranial hemorrhage (30). Fulminant, hemorrhagic papilledema was also seen within 5 to 8 hours in the context of other conditions (metastatic frontal lobe tumor, epidural hematoma) (31). In addition, minimal papilledema may exist and suddenly become fully developed in several hours in certain settings, such as encephalitis in the setting of a cerebral abscess (32,33). Occasionally, a paradoxical development or progression of papilledema several days to a week after normalization of increased ICP may occur (34).

Papilledema usually produces swelling of the inferior and superior poles of the optic disc prior to the nasal and temporal poles (27). Blurring of the disc margins by direct ophthalmoscopy is only apparent after optic disc elevation occurs (27).

With respect to IIH, the timing of papilledema in the course of the disease process is fascinating from an intellectual standpoint but also rises to the fore in the legal arena. It is unfortunately common that the diagnosis is missed acutely, leading to a delay in treat- ment and permanent visual loss, sometimes devastating blindness. The crux of a case is often related to the timing and detection of papilledema. Many a prosecuting attorney, defense attorney and expert witness have spent hours arguing whether or not “it was there”. Imagine an overweight teenage girl coming into the emergency department with a severe headache of four days duration that is the “worst of her life”. It is pounding, bifrontal, with photophobia, phonophobia, nausea, vomiting, blurred vision, and transient obscu- rations of vision. She is treated with narcotics, has a CT scan that is normal, and sent home with acetamin- ophen with codeine for migraine. Two days later the headache is no better and she is also seeing double. She goes back to the E.D. and gets a prescription for antibiotics for a sinus infection. Three days later she suddenly loses vision in one eye and the vision in the other rapidly deteriorates. By now someone looks in her eyes and she has florid papilledema with hemorrhages, exudates. The diagnosis is made but, despite aggressive management, she sustains permanent visual loss and will never be able to drive.

Suppose you are reviewing the records as an expert witness. On the first ED note it says PERRL, EOMI, discs sharp. During the second ED visit the “normal” box next to “HEENT” is checked off. The
visual acuity was not recorded on either visit. What are the possibilities?
1. There really wasn’t any papilledema on the first visit, the ED physician was correct.
2. There must have been papilledema on the first visit, maybe it was subtle and not easily seen with a direct ophthalmoscope, the ED physician was incorrect and should have called an ophthalmologist to examine the patient.
3. By the second visit there was certainly papilledema, no one looked.
4. If the ED physician says in deposition that he/she examined the eyes on the second vision and the discs looked normal, could that be possible or is he/she incompetent to view the ocular fundus (with a direct ophthalmoscope, under those circumstances, etc.)?

Some neuro-ophthalmologists used to think (and may still think) that the papilledema always precedes the symptoms of IIH. This probably arises from the finding of “asymptomatic papilledema” in some patients (this can also happen in ischemic optic neuropathy – the disc abnormality may precede the visual loss). However, it’s clear from the work done on the neurosurgical ward above, and from the experience of those patients who have acute “IIH without papilledema” that occasionally the symptoms may herald the optic nerve swelling by days to a week or more. That’s when the “package deal” of symptoms and the “headache red flags” become important.

How High Does the ICP Have to Be Before Papilledema Develops?

The absence of spontaneous retinal venous pulsations is thought by some investigators to be an early sign of papilledema. According to several authors, pulsations cease when ICP exceeds about 200 mm of water (35-37). Thus, if spontaneous venous pulsations are present, ICP should be below this figure. However, as noted above, marked fluctuations in ICP can occur in patients with increased ICP (3,38,39), and in such patients, the ICP may occasionally drop into the normal range. A patient could therefore have increased ICP but be examined at a time when ICP was transiently reduced at the trough of a pressure wave, at which time spontaneous venous pulsations might be observed. In addition, spontaneous venous pulsations occur in only about 80% of normal subjects (3,40,41). Thus, 20% of patients with normal ICP also lack spontaneous venous pulsations. For these reasons, the absence of spontaneous venous pulsations does not always support a diagnosis of papilledema. The observation of spontaneous venous pulsations suggests only that the ICP is probably below 200-250 mm of water at that moment.

How high is “too high”? Like most physiologic values, there is likely a spectrum of normal. Many aspects of intracranial pressure are similar to intraocular pressure, which there is much more experience with. As in glaucoma, there may be a range of tolerable intracranial pressures among individuals. Perhaps some patients’ brains adjust to a chronic ICP averaging in the high 200s or low 300s without developing papilledema. Others will develop papilledema when their normal pressure averaging 100 mm CSF rises to 200 mm CSF (similar to “normal tension glaucoma”).

IIH without Papilledema: Experience in the Literature

Most cases of IIH without papilledema in the literature occur in overweight women with chronic daily headaches (42-44). I believe that it is important to distinguish acute or subacute IIH without papilledema from “chronic IIH without papilledema” – they are probably not the same entity.

Acute
One early report was of two women who presented with new migrainous headaches, accompanied by “red flags” – daily headaches increasing in severity, sometimes associated with a recent weight gain (42). One patient had symptoms for two months and another had symptoms for four months. In another series of nine patients, 7 had headaches for six months or less, and another patient with a 2 year history of headache had an increased frequency and severity of headaches over the previous six months that led to the diagnosis (45). Seven patients had CSF pressure monitoring to confirm the diagnosis after a single LP opening pressure reading was normal. All patients had “red flags” in their history suspicious for increased ICP. Eight had transient obscurations of vision. The headache worsened with coughing, straining or Valsalva in 7 patients. All but 2 patients had a history of recent mild head trauma.

Chronic
Eighty-five patients with refractory transformed migraine at the Houston Headache Clinic had lumbar
puncture to exclude chronic meningitis or increased intracranial pressure (46). Elevated CSF pressure was found in 12 patients, ages 13-54 years, with duration of headaches from 3 to 30 years. CSF pressures ranged between 270 and 450 mm CSF. Four patients reported pulse synchronous tinnitus and one had an empty sella. All 12 patients had a history of migraine without aura and chronic tension type headache, 5 also had migraine with aura, and 6 had medication overuse headache. Acetazolamide and furosemide was added to the anti-migraine therapy of patients after the intracranial hypertension was detected with “a reduction in the number of days with severe headache, reduced consumption of abortive agents and overall improvement of quality of life.” Follow-up LP 3-20 months later ranged from 210-360 mm CSF; statistical analysis was not performed. The CSF pressure decreased in 8 patients, increased in 3 and was unchanged in one patient.

The largest case-control study is from the Jefferson Headache Center (43). Case subjects were 25 patients diagnosed between 1989 and 1996. The diagnosis of IIH was considered if they had intractable headaches which did not respond satisfactorily to treatment. Diagnostic opening pressure was 200 mm CSF on two occasions (at least one was over 240 mm CSF). Control subjects 60 patients with intractable chronic daily headache who had a normal CSF opening pressure, seen between 1992 and 1996. The average duration of headache was 7 ± 8 years in cases and 5 ± 7 years in controls. Statistical predictors of increased ICP were:

<table>
<thead>
<tr>
<th>Univariate Analysis</th>
<th>Cases</th>
<th>Controls</th>
<th>OR (95% CI)</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>&quot;Obesity&quot;*</td>
<td>20 (80)</td>
<td>27 (45)</td>
<td>4.9 (1.6-14.7)</td>
<td>0.003</td>
</tr>
<tr>
<td>Hx of seizure</td>
<td>4 (16)</td>
<td>2 (3)</td>
<td>4.6 (1.0-34.1)</td>
<td>0.03</td>
</tr>
<tr>
<td>Blurred vision</td>
<td>19 (76)</td>
<td>31 (52)</td>
<td>3.6 (1.2-10.8)</td>
<td>0.02</td>
</tr>
<tr>
<td>Tinnitus</td>
<td>11/18 (61)</td>
<td>14/44 (32)</td>
<td>4.5 (1.1-10.5)</td>
<td>0.03</td>
</tr>
<tr>
<td>Pulsatile tinnitus</td>
<td>9/18 (50)</td>
<td>4/44 (9)</td>
<td>12.9 (2.5039.8)</td>
<td>0.000</td>
</tr>
</tbody>
</table>

| Multivariate Analysis | | |
| Pulsatile tinnitus | 13.0 (2.8-60.9) | 0.01 |
| "Obesity"*          | 4.4 (1.1-18.6) | 0.04 |

*Quotation marks added, DF
The body height of cases and controls was similar (64 ± 3 vs. 65 ± 4 inches).

There are many interesting findings in the data. Note that both groups were obese (204 ± 46 vs. 173 ± 49 lb), although the cases were considerably more overweight than the controls. Although unknown at the time of publication in 1998, recent work by Scher and colleagues assessing risk factors for chronic daily headaches (CDH, 180+ headaches per year) showed that obesity was associated with developing CDH. The adjusted odds ratio rose with increasing BMI (1.26 for overweight, 1.34 for obese). The headache profiles were similar in both groups: severe, causing functional disability, throbbing, unilateral in just over 40%. Most (60-62%) had a history of migraine, over 90% had chronic daily headaches and 68% had transformed migraine. Analgesic overuse was common (80%) in both groups.

Patients were treated with lumbar punctures (56% response in cases, not reported in control group), dihydroergotamine (similar response in both groups), and diuretics (31% of cases improved). Five patients had shunt surgery; two had one operation, two had three operations and one had four operations. Three of the five consistently responded to shunting although diuretics and steroids conferred no additional benefit in shunted patients.

Their disappointing experience approaching the headache treatment by lowering the ICP is not unusual. A recent paper by the group at Johns Hopkins reviewed the records of shunt placement procedures performed for intractable headache of IIH between 1973 and 2003 (47). Forty-two patients underwent 115 shunt procedures (79 lumboperitoneal shunts, 36 ventriculoperitoneal or ventriculo-atrial shunts). Although 95% of patients experienced significant improvement of headaches immediately after surgery, severe headache recurred despite a properly functioning shunt in 19% of patients by 12 months and 48% by 36 months. The poorest prognosis for recurrent headache was in patients who had no papilledema (RR 5.2, 95% CI 1.5-4.3) or those with headaches longer...
than two years (RR 2.5, 95% CI 1.5-5.7) pre-operatively. Lumboperitoneal shunts were associated with a 2.5 fold risk of revision due to a threefold risk of shunt obstruction, although over-drainage, distal catheter migration and shunt infection occurred with equal frequency between the two types of shunts.

A retrospective study from a neuro-ophthalmic practice at the University of Utah found that of 353 patients with IIH, 20 had IIH without papilledema (48). The most common presenting complaint was headache. 18 of the patients were obese women (mean BMI 34.5) and the mean age at 28 years. Unlike patients with papilledema who presented within a year of symptom onset, patients without papilledema presented, on average, five years after symptom onset. Their headaches were migraine-type, and they were less likely than patients with papilledema to experience transient visual obscurations, pulsatile tinnitus, diplopia, or visual loss. If visual field loss was present, it was usually functional (non-organic). Their headache response to therapy was no different than patients with papilledema although 3 of 4 patients had improvement in their headaches with shunting. Most patients had observable spontaneous venous pulsations, consistent with their generally lower CSF pressures than patients with papilledema (260-420, mean 312 mm CSF vs. 260-550, mean 330 mm CSF in patients with papilledema, p<0.01, T-test).

**Is Increased ICP in a Headache Patient Specific to IIH?**

We reviewed the records of adults evaluated in the ED at SUNY Upstate Medical University Hospital from 1997-2001 who met the following criteria: (1) a lumbar puncture was performed, and (2) the discharge diagnosis, regardless of the patient’s disposition, was migraine, tension-type headache, vascular headache, cluster headache or unspecified headache (49). Patients known to have IIH or later diagnosed with IIH were excluded. We also excluded headache patients with a severe intracranial process. The study was approved by the SUNY Upstate Institutional Review Board for the Protection of Human Subjects and the information was collected anonymously.

We reviewed 168 charts and collected the following data: age, gender, opening pressure, closing pressure, patient positioning during the procedure, final diagnosis, and specialty of the physician performing the lumbar puncture.

The CSF opening pressure was recorded in 30 of the 168 charts reviewed, and 28 patients met the inclusion criteria. The range of opening pressures in 28 patients was 85 to 370 mm of water. Fourteen patients had opening pressures of 200 mm of water or greater and ten patients had OP above 249mm water. The patient positioning was only documented in 7 of 28 patients. The discharge diagnoses were migraine (n=12), unspecified headache (10), tension-type headache (3), post-traumatic headache (2) and basilar migraine (2). There was no correlation between headache type and opening pressure.

<table>
<thead>
<tr>
<th>Headache Type</th>
<th>OP (cm water)</th>
<th>Headache Type</th>
<th>OP (cm water)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Migraine</td>
<td>19</td>
<td>Tension type</td>
<td>25</td>
</tr>
<tr>
<td>Migraine</td>
<td>18</td>
<td>Tension type</td>
<td>10</td>
</tr>
<tr>
<td>Migraine</td>
<td>18</td>
<td>Tension type</td>
<td>26</td>
</tr>
<tr>
<td>Migraine</td>
<td>24</td>
<td>Post-traumatic</td>
<td>18</td>
</tr>
<tr>
<td>Migraine</td>
<td>27</td>
<td>Post-traumatic</td>
<td>27</td>
</tr>
<tr>
<td>Migraine</td>
<td>16</td>
<td>Basilar migraine</td>
<td>70</td>
</tr>
<tr>
<td>Migraine</td>
<td>14</td>
<td>Basilar migraine</td>
<td>18</td>
</tr>
<tr>
<td>Migraine</td>
<td>26</td>
<td>Basilar migraine</td>
<td>20</td>
</tr>
<tr>
<td>Migraine</td>
<td>18</td>
<td>Basilar migraine</td>
<td>18</td>
</tr>
</tbody>
</table>

**What is the Natural History of Headaches in Patients with IIH? What are the Implications for Treatment?**

The longest natural history study of IIH was a retrospective cohort study published in 1982, following 57 patients from 5 to 41 years (50). This was a “pre-CT” era study, so it is possible that some of the patients may have had other disorders. The aim of the study was to determine visual function and it did not address headache. The only other prospective series of 50 patients did not assess headache as an outcome variable (51). Our retrospective study of 82 patients with IIH diagnosed before age 50, 68% had headaches classifiable by International Headache Society Criteria after their intracranial pressure was successfully treated (52). Twenty-three percent had more than one type of headache. The headaches were identified by the patients as being different from their IIH headaches. Eleven of the adults had a history of episodic headaches since childhood. The most common headache types were episodic tension type headache (30 patients), migraine without aura (20), chronic tension type headache (10), anal-
gesic overuse headache (8), idiopathic stabbing headache (3). All 3 patients who had migraine with aura had a prior history of it, preceding the symptoms of IIH. If anything, the study underestimated the incidence of headache disorders both subsequent and prior to the development of IIH because of its retrospective nature.

I have subsequently been impressed by the number of my IIH patients who (1) have a pre-existing history of migraine and (2) despite the resolution of their papilledema and visual symptoms, continue to have headaches. This has several implications for treatment:

- IIH patients with otherwise stable disease don’t need a spinal tap/shunt revision/trip to the emergency department every time they get a headache – they can get migraines too.
- If the papilledema is gone and the vision is stable, treat the patient for chronic headaches, with headache prophylaxis
- Watch out for analgesic overuse headaches in IIH patients – it is a problem
- It is possible that frequent analgesic use may increase the intracranial pressure – it happens in rats, we’re not sure about humans (and there is no straightforward way to test the hypothesis)
- When using prophylactic medications in an obese IIH patient, use medications that cause weight gain and peripheral edema with caution – monitor closely (valproate, verapamil, tricyclic antidepressants). If there are no contraindications, consider using topiramate (but avoid concurrent use with acetazolamide).
- Treat headaches medically, not surgically, especially if there is no papilledema.

Conclusions:

1. IIH without papilledema is uncommon. In general, the diagnosis of IIH should be made as a “package deal” with the appropriate symptoms and signs, using the diagnostic criteria.

2. However, IIH without papilledema may infrequently occur in the acute to subacute stage. There are several possible mechanisms:
   - Although the increase in pressure is not high enough to produce papilledema, it is high enough to produce symptoms.
   - There is a time lag between the pressure rise and the development of papilledema (e.g., the neurosurgical ICU experience)
   - There is anatomical variability in the micro-architecture of the optic nerve or the continuity of subarachnoid space precluding the development of papilledema (or in some cases, producing unilateral or asymmetrical papilledema)
   - The pressure head is deflected elsewhere (empty sella, Chiari malformation, spinal compartment).

3. “IIH without papilledema” persisting beyond 6 months is probably not IIH. Differential diagnosis:
   - Medication overuse headache (transformed migraine, etc.) – most common
   - Chronic tension type headache
   - Chronic migraine
   - Remember, obesity is a risk factor for chronic daily headache

4. The papilledema from IIH does not always completely resolve. In a stable patient (with chronic residual papilledema, or without papilledema) whose main problem is headache, treat them medically, not surgically. Do not shunt for headache!

Selected References

44. Lipton HL, Michelson PE. Pseudotumor cerebri syndrome without papilledema. JAMA 1972;220:1591-1592.

CME Answers:
1. D
2. A
3. C