ROSENBERG SCHOOL of OPTOMETRY

Optometric Clinical Practice

Volume 5 | Issue 2

2023

Idiopathic Intracranial Hypertension: A Case Report

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Recommended Citation

Bugajski C. Idiopathic Intracranial Hypertension: A Case Report. *Optometric Clinical Practice*. 2023; 5(2):74. https://doi.org/doi.org/10.37685/uiwlibraries.2575-7717.5.2.1015

Idiopathic Intracranial Hypertension: A Case Report

Abstract

Background: Optic nerve head edema is a serious finding for which multiple etiologies need to be considered. When optic nerve head edema is observed bilaterally, among the top differentials is idiopathic intracranial hypertension (IIH), also known as primary pseudotumor cerebri. An emergent magnetic resonance image with and without contrast as well as venography and lumbar puncture with opening pressure measurement must be performed to diagnose and determine management. **Case Report**: This case specifically features a 30-year-old Caucasian female with bilateral optic disc edema secondary to IIH. In addition to discussing the details regarding this patient's case, this report highlights pertinent imaging, visual testing, and co-management. **Conclusion**: Important caveats to treatment and management of the condition, and other underlying complications of IIH are discussed. Close monitoring of papilledema, visual acuity, and visual field, along with co-management with neurology and neuro-ophthalmology will provide optimum care.

Keywords

idiopathic intracranial hypertension, pseudotumor cerebri, optic nerve edema, magnetic resonance imaging, lumbar puncture

INTRODUCTION

Elevated intracranial pressure can result from two main categories: non-mass related conditions and intracranial mass lesions.[1,2] Pseudotumor cerebri (PTC) is a condition where increased intracranial pressure is present in the absence of any mass, lesion, dilation of ventricles, or other disease states and can be separated into two categories: primary (90%) and secondary (10%).[3-6] Primary is a diagnosis of exclusion. Causes for secondary PTC may include venous sinus thrombosis and medication usage, among others.[1-3] Primary PTC, also called idiopathic intracranial hypertension (IIH), characteristically is a condition that affects obese women of childbearing age with an average age between 25-36 years old.[3,7-9] Of all cases of IIH, the prevalence of men with the condition, when considering modern neuroimaging techniques, ranges from as low as 6% to as high as 19%.[10] Thus, while possible, the condition is less frequent in males.[10-12] PTC can also occur in children, but the condition may not follow the characteristic presentation. PTC has no predilection for females in pre-pubescent children, as it is found equally among males and females.[5,13] In pubescent children, however, predilection is similar to the characteristic adult presentation.[13] As indicated earlier, primary IIH is a diagnosis of exclusion where normal neuroimaging and abnormal lumbar puncture opening pressure measurement are present. Neuroimaging is necessary to rule out any mass, lesion, or hemorrhaging. Magnetic resonance imaging (MRI) with and without contrast and magnetic resonance venography (MRV) are the preferred method over computed tomography (CT) imaging.[1,2,14] A lumbar puncture opening pressure measurements above 25 cm H_2O is considered diagnostic for elevated intracranial pressure.[3,9] Cerebral spinal fluid is of normal composition in IIH patients.[3,8,15] This information is used within the criteria for determining the presence of IIH. In 2013, Friedman et al. released pseudotumor cerebri syndrome (PTCS) diagnostic criteria, which is an attempt at being all-encompassing and designed for use with children and adults in cases with papilledema present or absent. It builds on previously developed criteria. Acetazolamide is commonly given to patients with IIH, along with recommendations for weight loss. The Idiopathic Intracranial Hypertension Treatment Trial (IIHTT) concluded that acetazolamide use in combination with weight loss, in comparison to weight loss alone, proved more effective in improvements of visual field function.[17] Neurosurgical treatments are reserved for severe and sudden onset IIH, including patients with rapid vision loss, continued vision loss with standard treatment, the presence of a RAPD, and macular edema with severe papilledema.[3,4,7-9] Co-management with neurology, neuro-ophthalmology, neurosurgery, and neuroradiology may occur, and the patient should be followed regularly to evaluate visual acuity, visual field, and papilledema, as well as possible systemic concerns such as headaches and pulsatile tinnitus.

CASE DATA

INITIAL EXAMINATION

A 30-year-old Caucasian female presented to the Department of Veterans Affairs (VA) Eye Clinic with a chief complaint of loss of vision in the right eye. The vision loss had occurred intermittently, transiently, and multiple times throughout the day for three weeks prior to the appointment. She reported associated moderately severe headaches concentrated around and behind her eyes that had worsened over the previous three months. She denied any dizziness, eye pain, or diplopia. Personal ocular history was unremarkable. The patient's family history was negative for any ocular conditions. Her medical history included migraine headache, sinusitis, major depressive disorder, post-traumatic stress disorder, and hysterectomy. The patient was not diagnosed with hypertension or diabetes. Her current medications included propranolol HCL 40 mg, sumatriptan succinate 50 mg, hydroxyzine HCL 10 mg, sertraline HCL 100 mg, and riboflavin 100 mg. She denied use of oral contraceptives and denied any medical allergies. She was oriented to person, place, and time.

Corrected distance visual acuities were 20/20 OD and 20/20 OS with a spectacle prescription of -3.50 -1.00 x 164 OD and -3.75 -0.75 x 005 OS. Ocular motilities were full and smooth in all gazes in each eye, and no pain was noted on eye movement. A cover test demonstrated orthophoria at distance and near. Confrontation visual fields were full to finger count in each eye. Pupils were equal, round, and reactive to light with no relative afferent pupillary defect (RAPD).

An examination of the anterior segment evaluation in each eye revealed normal lid appearances, clear and intact corneas, clear and quiet conjunctiva, deep and quiet anterior chambers, and normal, flat irides. Intraocular pressures were measured using non-contact tonometry at 14 mm Hg and 16 mm Hg in the right and left eye respectively.

A dilated (1% tropicamide) posterior segment evaluation demonstrated a clear crystalline lens and a normal vitreous in each eye. The optic discs displayed blurry margins 360° depicting bilateral moderate disc edema, graded 3 on the Frisén scale. No papillary or peripapillary hemorrhages were observed, and the maculae appeared normal and flat. Retinal blood vessels were in a normal ratio of ⅔, and the periphery revealed an intact retina with no holes, tears, or detachments 360° noted in either eye.

Fundus photographs (Figure 1) and optical coherence tomography (OCT, Figures 2 & 3) were ordered and performed the same day.

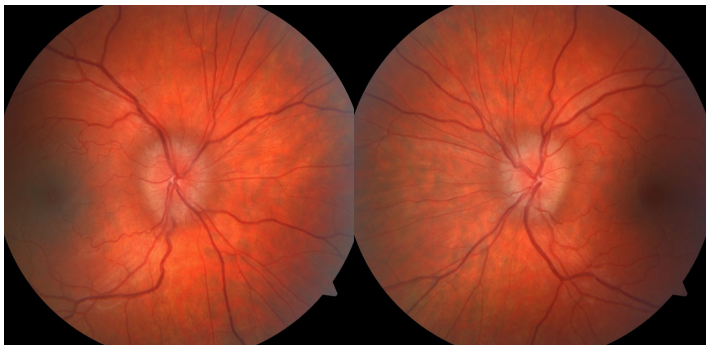

Figure 1 is a side-by-side fundus image of OD and OS. The image displays the blurred margins of the optic nerve head (ONH) OD and OS. No hemorrhages at the disc or elsewhere are observed in either eye.

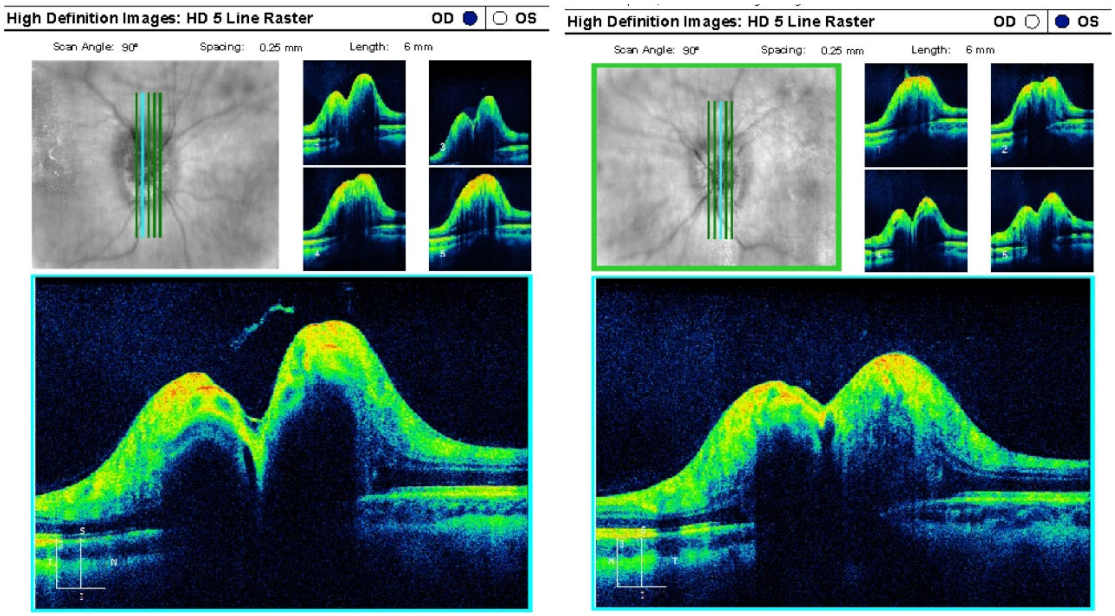

Figure 2 is a side-by-side high-definition OCT image of OD and OS. The image displays a 90° scan angle through the ONH OD and OS. The optic nerve edema is clearly confirmed in both eyes.

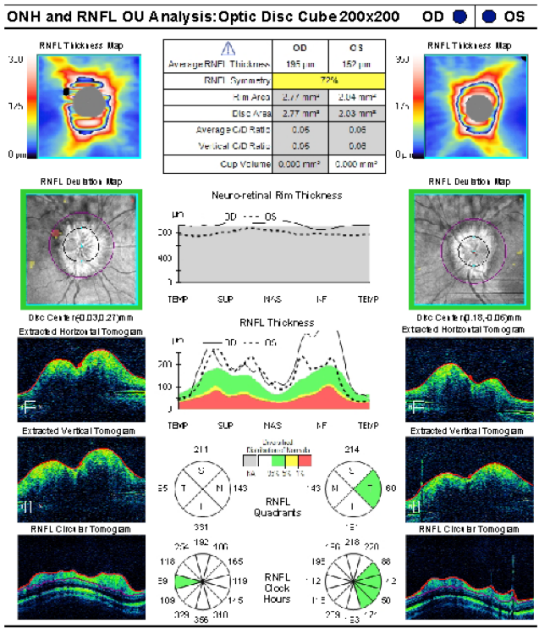

Figure 3 is an ONH and retinal nerve fiber layer analysis of OD and OS. The OCT image clearly displays the ONH edema observed funduscopically in both eyes. The average retinal nerve fiber layer thickness was 195 and 152 OD, OS respectively.

The patient was diagnosed with papilledema with pending imaging and testing for determination of etiology with a high suspicion of IIH. Extensive counseling was performed regarding the findings and need for immediate imaging and lumbar puncture. The patient was sent to the local emergency department (ED) as the Veterans Affairs (VA) Medical Center was unable to provide testing required that day. The attending physician at the local ER was contacted via phone, and the patient's case was reviewed. The necessary VA paperwork was completed including an order for an urgent non-VA care MRI of the brain and orbits with and without contrast and lumbar puncture with opening pressure measurement.

Prior to leaving for the emergency room, the patient was scheduled for a two-week follow-up with the eye clinic for a Humphrey Visual Field (HVF) 30-2, color testing, and repeat dilation. VA Medical Center Neurology was also consulted to evaluate the patient within one week. She was prescribed acetazolamide 500 mg twice daily, and the need for diet control and weight loss was discussed as the likely mechanism of the findings was IIH.

EMERGENCY DEPARTMENT VISIT

At the emergency department , the patient was initially triaged by the nursing staff before being taken for an MRI and lumbar puncture. MRI images with and without contrast were performed and reviewed as normal with no evidence of intracranial mass, lesion, or hemorrhage. Opening pressure was measured at 27 cm water. Closing pressure was measured at 15 cm water. The diagnosis of idiopathic intracranial hypertension was confirmed.

EYE CLINIC FOLLOW-UP #1

The patient presented for follow-up one week after initial presentation with little to no improvement in her symptoms. Corrected visual acuities were 20/20 OD and 20/20 OS with her habitual distance correction. Motilities were full and smooth in all gazes OU. A cover test demonstrated orthophoria at distance and near. HVF testing was performed and was within normal limits (Figure 4). Pupils were equal, round, and reactive to light with no RAPD. She was oriented to person, place, and time.

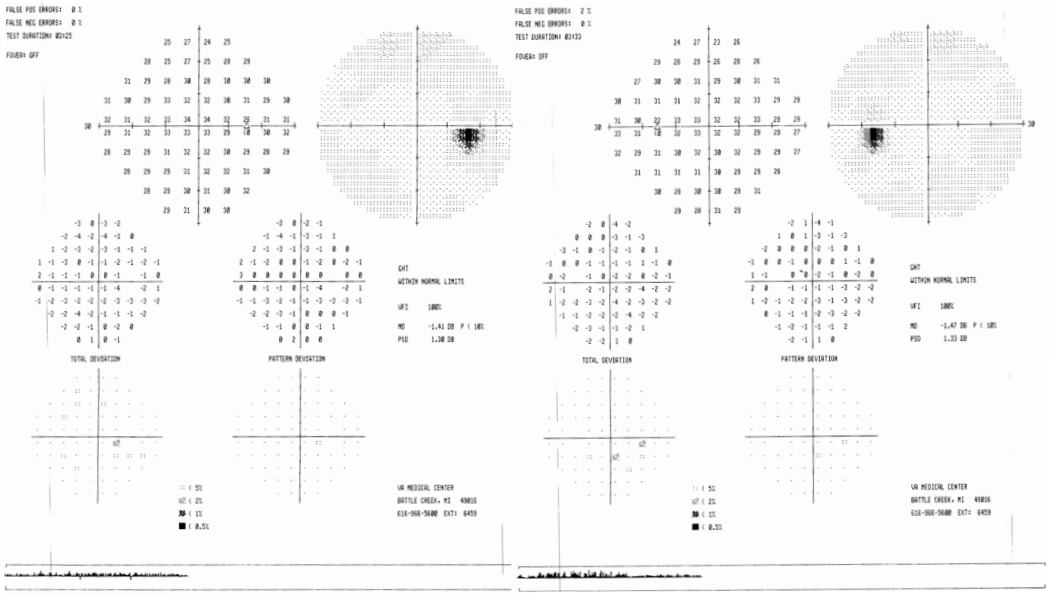

Figure 4 is a side-by-side analysis of the HVF 30-2 OD, OS. The tests are reliable and show no significant defects. Specifically, there is no enlargement of the natural blind spot.

Color vision testing with Ishihara plates revealed normal color vision in both eyes. Anterior segment evaluation revealed normal lid appearance OU, clear and intact corneas OU, clear and quiet conjunctiva OU, deep and quiet anterior chambers OU, and normal, flat irides OU. The patient's intraocular pressures were measured using non-contact tonometry at 15 mm Hg OD, 16 mm Hg OS.

The dilated posterior segment evaluation showed a clear lens and normal vitreous in each eye. Bilateral disc edema (Frisén Grade 3) with non-distinct disc margins and no discernible cupping in both eyes appeared stable from her initial presentation two weeks prior. No papillary or peripapillary hemorrhages were observed in either eye. Normal, flat maculae were depicted. The retinal blood vessels in each eye were of normal ratio of ⅔, and the periphery was intact retina with no holes, tears, or detachments 360°.

The patient was educated on the results of the Humphrey visual field and color vision testing as well as the stable fundus findings. She was instructed to continue using acetazolamide 500 mg twice daily, and a neurology consultation was established for one week with VA Medical Center Neurology. A three-month follow-up was scheduled for dilated fundus evaluation, ONH OCT, and HVF 30-2.

VETERANS AFFAIRS MEDICAL CENTER NEUROLOGY FOLLOW-UP

The patient presented to the VA Medical Center Neurology as scheduled for further evaluation of IIH and to transfer care from the previous ED physicians. The neurologic examination was within normal limits with the exception of the disc edema observed funduscopically. A CT venography was performed and revealed no evidence of deep vein thrombosis. The patient was instructed to continue acetazolamide 500 mg twice daily with slow and steady increase to 750 mg twice daily, then 1000 mg twice daily. The patient was instructed to continue care with the VA Eye Clinic every three months for HVF testing and dilated fundus exam.

PATIENT SELF-SCHEDULED EXAMINATION

The patient presented to the VA Eye Clinic two months later with a chief complaint of intermittent, transient blurred vision of both eyes with an onset of 1-2 weeks prior to the appointment. She said it occurred most mornings upon awakening and would last about 15-20 minutes; however, throughout the day, the blurred vision was not an issue. There were no changes to her medical or ocular history, and she was oriented to person, place, and time. She was using 1000 mg of acetazolamide twice daily.

The corrected visual acuities were 20/20 OD and 20/20 OS with habitual distance correction. Motilities were full and smooth in all gazes OU. Cover test demonstrated orthophoria at distance and near. Confrontation visual fields were full

to finger count in each eye. Pupils were equal, round, and reactive to light with no RAPD. The anterior segment evaluation of both eyes revealed a normal lid appearance, clear and intact corneas, clear and quiet conjunctiva, deep and quiet anterior chambers, and normal, flat irides. Intraocular pressures were measured using applanation tonometry at 12 mm Hg in each eye.

On a dilated posterior segment evaluation, the optic discs displayed blurry margins 360° depicting bilateral moderate disc edema, graded 2 on the Frisén scale. No papillary or peripapillary hemorrhages were observed OU. Normal, flat macula were observed OU. Retinal blood vessels were in a normal ratio of ⅔ OU, and the periphery revealed an intact retina with no holes, tears, or detachments 360° noted OU.

Fundus photographs (Figure 5) and OCT (Figures 6 & 7) were ordered and performed the same day.

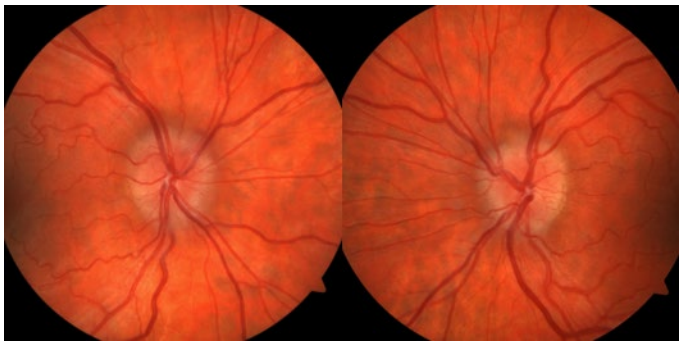

Figure 5 is a side-by-side fundus image of OD and OS. The image displays the blurred margins of the ONH OD and OS. No hemorrhages at the disc or elsewhere are observed in either eye. Improvement in papilledema appearance can be appreciated.

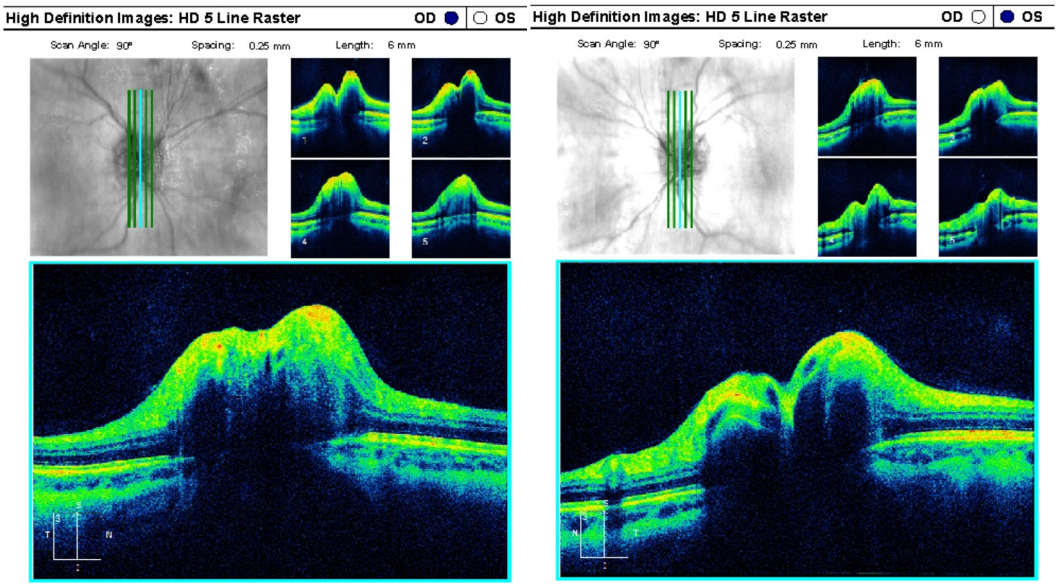

Figure 6 is a side-by-side high-definition OCT image of OD and OS. The image displays a 90° scan angle through the ONH OD and OS. The optic nerve edema is clearly confirmed in both eyes.

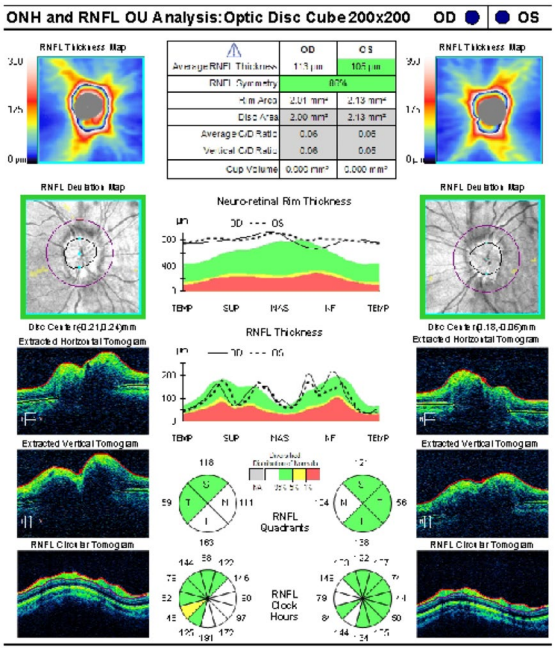

Figure 7 is an ONH and retinal nerve fiber layer analysis of OD and OS. The OCT image clearly displays the ONH edema observed funduscopically in both eyes. The average retinal nerve fiber layer thickness was 113 and 105 OD, OS respectively. These values are reduced compared to the original presentation, depicting less ONH edema.

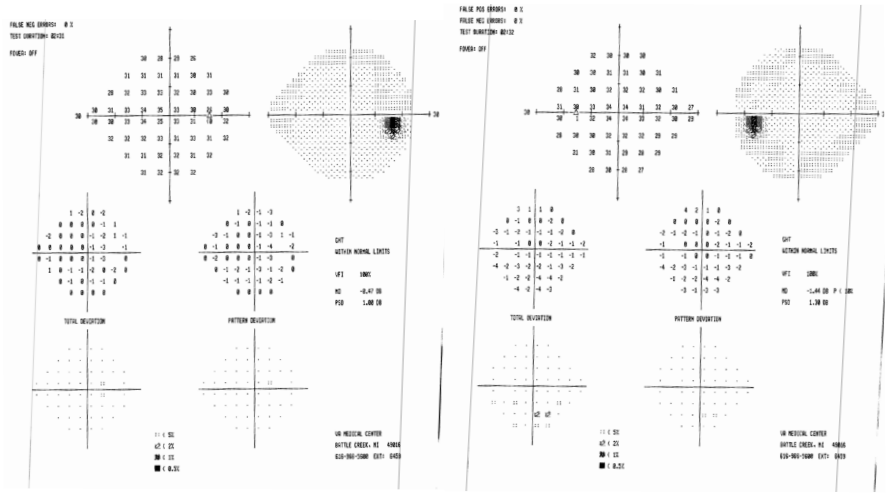

Figure 8 is a side-by-side analysis of the HVF 24-2 OD, OS. The tests are reliable and show no significant defects. Specifically, there is no enlargement of the natural blind spot.

The patient was reassured based on the normal visual acuity and visual field seen that day as well as the decrease in optic nerve swelling OU observed on funduscopic evaluation and confirmed by OCT imaging. She was instructed to continue using acetazolamide 1000 mg twice daily and diet control. The patient was reminded of an upcoming VA Medical Center Neurology appointment, and a three-month follow-up appointment was scheduled at the VA Eye Clinic.

DISCUSSION

Pseudotumor cerebri is a condition where increased intracranial pressure is present in the absence of any mass, lesion, dilation of ventricles, or other disease states and can be separated into two categories: primary (90%) and secondary (10%).[3-6] PTC is a diagnosis of exclusion. Causes for secondary PTC may include venous sinus thrombosis and medication usage, among others.[1-3] Primary PTC, is commonly referred to as idiopathic intracranial hypertension. Signs and symptoms of primary and secondary PTC are the same.[3] Symptoms of PTC include headaches, transient visual obscurations, diplopia, and pulsatile tinnitus; papilledema is classically observed.[3,4,7,9,15] Headaches are commonly associated with PTC and are reported 75-94% of the time with some relief seen after lumbar puncture and CSF drainage.[7,9] Frequently, the headaches are bilateral, typically located in the frontal or retro-bulbar region, are worsened by Valsalva maneuvers, and may present with nausea, vomiting, phonophobia, and photophobia.[3,9,15]

Transient visual obscurations (TVOs) are reported 68-72% of the time and can be unilateral or bilateral.[3,4,7,9] A TVO is thought to occur because of temporary ischemia caused by ONH circulation interference.[3,4,9] A patient with PTC will report binocular diplopia 18-40% of the time.[3,9] Typically, the diplopia is horizontal and due to a unilateral or bilateral cranial nerve six (abducens nerve) palsy.[3,4,7] Diplopia is infrequently observed as a vertical deviation.[3,7]

Pulsatile tinnitus, which is thought to be caused by cerebral venous pathway interference, can be unilateral (⅓) or bilateral (⅔), is reported 52-60% of the time, and is typically worse in the evenings.[3,7,9] Increased intracranial pressure in PTC patients results in papilledema, a distinctive and noticeable feature of the condition, is seen on funduscopic examination. Papilledema will appear with non-distinct ONH margins, ONH elevation, and sometimes blood vessel obscuration at the ONH. It may also appear with peripapillary hemorrhages, exudates, and macular edema in more severe presentations.[3,9] Papilledema is a significant finding observed in PTC patients as it is the most common sign of the condition.[3,4,7,9] Usually symmetric, papilledema in PTC patients can appear asymmetric, unilateral, or not at all.[3,4,7,9] A unilateral presentation is observed 3.6-10.0% of the time, and a lack of ONH edema in the presence of PTC is rare.[3,4,9] The most common system of grading papilledema is the Frisén scale (Table 1), which ranges from normal (stage 0) to severe papilledema (stage 5) and can provide a good prediction of visual outcome as well as possibly assist with treatment management.[3,13]

Modified Frisén scale for grading papilledema[18,19]

Stage 0: Normal optic disc or not a disc but no edema/swelling
A. Prominence of the retinal nerve fiber layer at the nasal, superior, and inferior poles in inverse proportion to disc diameter
B. Radial nerve fiber layer striations, without tortuosity
Stage I: Minimal
A. C-shaped halo that is subtle and grayish with a temporal gap; obscures underlying retinal details
B. Disruption of normal radial nerve fiber layer arrangement striations
C. Temporal disc margin normal

Stage II: Low degree
A. Circumferential halo
B. Elevation – nasal border
C. No major vessel obscuration
Stage III: Moderate
A. Obscuration of 1 or more segments of major blood vessels leaving disc
B. Circumferential halo
C. Elevation – all borders
D. Halo – irregular outer fringe with finger-like extensions
Stage IV: Marked
A. Total obscuration on the disc of a segment of a major blood vessel on the disc
B. Elevation – whole nerve head, including the cup
C. Border obscuration – complete
D. Halo – complete
Stage V: Severe
A. Partial obscuration of all vessels on disc and total obscuration of at least 1 vessel on disc

TABLE 1

Bidot et al. report that higher Frisén stages and subsequent visual effects are seen in patients with larger optic canal sizes.[20] Frisén stage 1 and higher may show retinal-choroidal folds indicating optic nerve head stress.[3,21,22] Sibony et al. report peripapillary wrinkles, retinal folds, and choroidal folds were present 26%, 19%,

and 1% of the time respectively on fundus photography, with higher percentages for each found using OCT because of increased definition.[21] Another term sometimes used in place of PTC is benign intracranial hypertension; however, this term should not be used as frequently due to the potential non-benign impact PTC can have on vision, as permanent vision loss can occur in 23-40% of patients with PTC.[7,9,13,23] Piper et al. report vision is affected in 75-87% of individuals affected by PTC.[5] A patient's visual field can also be affected in PTC. A Humphrey visual field 24-2 or 30-2 should be performed in cases of PTC. The most common visual field defect noted in 80% of patients is an enlarged physiologic blind spot, thought to be due to the papilledema typically present in patients with PTC.[7,9] Other visual field defects that can be present include nasal field and generalized constriction at 72% and 54%, respectively.[9] If progression on HVF testing is noted in a patient with stability or improvement in PTC, then Cello et al. recommend retesting the HVF.[24] While not yet formally completed in clinical testing of PTC, the pupillary light reflex has been shown as abnormal in these patients.[25]

Primary pseudotumor cerebri, commonly called idiopathic intracranial hypertension , is a condition that characteristically affects obese women of childbearing age with an average age between 25-36 years old.[3,7-9] Piper et al report a slightly large age range of 20-40.[5] In clinical observation, an 8:1 female-to-male ratio exists.[3] Obesity can be found in 57-100% of the total individuals with IIH.[9] Handley et al., report around 94% of patients are obese.[26] Examining the characteristic epidemiology, the incidence globally is 12-20 per 100,000 people;[9] however, in the general global population, the incidence is only 0.5-2 people per 100,000.[4,9] IIH is estimated to occur in the general United States' population 0.9 per 100,000 individuals.[3] For women who are 10% above ideal body weight, the incidence rises to 13 per 100,000 and to 19 per 100,000 for women 20% above ideal body weight.[3] Thurtell et al. report that the incidence in the United States is estimated to occur 19 per 100,000 females 20-44 years-old that are obese.[7] Wakerley reports a high IIH incidence of up to 323 per 100,00 in the United States.[4] In the United Kingdom, incidence was 11.9 per 100,000 in an obese female population. Per The Idiopathic Intracranial Hypertension Treatment Trial, "The diagnosis of IIH should be made with caution in nonobese patients, men, and those without typical symptoms such as headache, transient visual obscurations, and pulse synchronous tinnitus."[27]

Idiopathic intracranial hypertension is a diagnosis of exclusion where neuroimaging and lumbar puncture with open pressure measurement are required for work-up. Neuroimaging is necessary to rule out any mass, lesion, or hemorrhaging. MRI with and without contrast is the preferred method over CT imaging.[14] Magnetic resonance venography or CT venography should be performed in conjunction, especially for atypical presentations.[3,14] There are findings observed on MRI that are seen in the following percentage in patients with raised intracranial

pressure: posterior globe flattening (98%), enlarged optic nerve sheath (89%), and empty sella (70-100%).[14,28] Sivasankar et al. report empty sella being present 75-92% of the time.[28] However, it is important to note that an empty sella is also commonly found in non-IIH patients.[14] A lumbar puncture with an opening pressure measurement is also required in suspected cases of IIH. Opening pressure measurements above 25 cm H_2O are considered diagnostic for elevated intracranial pressure.[3,9] Pressures between 20 and 24.9 cm H_2O are considered non-diagnostic.[1] Cerebral spinal fluid is of normal composition in IIH patients.[3,8,15] The above information is used within the criteria for determining the presence of IIH.

B-scan ultrasonography can provide an initial alternative to more invasive methods of determining elevated intracranial pressure.[29,30] Mohson and Auday report, "Transorbital ultrasonography is very accurate, available, noninvasive, safe, and easily performed procedure for the detection of raised ICP."[29] Kanski states, "B-scan ultrasonography can be used to aid in distinguishing between papilloedema and other causes of a swollen or apparently swollen optic disc with 80-90% sensitivity and specificity by measuring the external diameter of the optic nerve sheath (ONSD), which is substantially distended (5.0-5.7mm or greater at 30.mm behind the globe."[30] The crescent sign and bulging optic disc appearance on ultrasound may be observed. "The crescent sign refers to an anechoic area in the anterior intraorbital nerve usually representing the separation of the nerve and its sheath by cerebrospinal fluid," per Mohson and Auday.[29] However, this testing is unable to determine the cause of the elevated intracranial pressure; additional testing may still be needed.

In 1985, Smith released the "Modified Dandy Criteria" to help determine if a patient has PTC.[31] In 2002, Friedman and Jacobson released new criteria for diagnosing PTC building from the "Modified Dandy Criteria" given the updates in imaging capabilities.[31] In 2013, Friedman et al. released pseudotumor cerebri syndrome (PTCS) diagnostic criteria (Table 2), which is an attempt at being all-encompassing and designed for use with children and adults in cases with papilledema present and absent.[16]

Criteria for Diagnosing Pseudotumor Cerebri Syndrome[16]

Papilledema
Normal neurologic examination except for cranial nerve six abnormalities

Criteria for Diagnosing Pseudotumor Cerebri Syndrome[16]
Neuroimaging: Normal brain parenchyma without evidence of hydrocephalus, mass, or structural lesion and no abnormal meningeal enhancement on MRI, with and without gadolinium, for typical patients (female and obese), and MRI, with and without gadolinium, and magnetic resonance venography for others; if MRI is unavailable or contraindicated, contrast-enhanced CT may be used
Normal CSF composition
Elevated lumbar puncture opening pressure (≥250 mm CSF in adults and ≥280 mm CSF in children [250 mm CSF if the child is not sedated and not obese]) in a properly performed lumbar puncture

TABLE 2

The criteria above provide guidelines to the clinician in determining if PTC is the most appropriate as a diagnosis. From Bandyopadhyay and Jacobson, "older patients, diagnosed based on the Modified Dandy Criteria, consisted of a higher proportion of men, were less frequently obese, were generally less symptomatic, and more often had identifiable (non-idiopathic) causes of PTC."[32] When compared to the characteristic primary PTC patient, older age and normal weight patients had similar or better visual outcomes. Men with primary PTC, however, suffered worse visual outcomes.[33] While less common, PTC can occur in children, and the condition may not follow the characteristic presentation. PTC has no predilection for females in pre-pubescent children, as it is found equally amongst males and females.[5,13] In pubescent children, however, the predilection is similar to the characteristic adult presentation.[13] Typically, PTC in children is secondary to another cause and includes tetracycline use, hypervitaminosis A, and cerebral venous sinus thrombosis.[8] Gospe et al., report that papilledema Frisén grade 3 and higher on presentation is a major risk factor in visual outcomes in pediatric patients with 50% of eyes having potential visual impact.[13] Aylward et al. report 17.8% of the PTC pediatric population researched did not have papilledema associated.[34] A separate criterion has been proposed for diagnosis in children (Table 3).

Criteria for Prepubertal Intracranial Hypertension[35]
CSF opening pressure of >18 cm water in children <8 or >25 cm water in children >/= 8 years or <8 years without optic edema
No focal neurological signs (with the exception of cranial nerve IV or VI palsy)
Normal CSF composition
Exclusion of recognized secondary causes
Bilateral optic disc edema
Symptoms suggestive of elevated intracranial pressure (headache, nausea, vomiting, transient visual obscurations, tinnitus) that improve following CSF drainage

TABLE 3

Acetazolamide, a carbonic anhydrase inhibitor, is used as a common treatment in patients with IIH. The use of the mediation in IIH is thought to affect the production of CSF at the choroid plexus by reducing the transport of sodium ions.[17] A reduction in CSF from 6-50% can be expected.[17] While obesity is commonly associated with the condition, even a 6% reduction in weight has shown improvement in IIH.[17] Thurtell et al. report a 5-10% reduction in weight can show improvement in the overall condition.[7] The Idiopathic Intracranial Hypertension Treatment Trial concluded that acetazolamide use in combination with weight loss in comparison to weight loss alone proved more effective in improvements of visual field function.[17] The IIHTT was important because it was the first landmark study to demonstrate the importance of using acetazolamide in patients with IIH. Visual outcomes of the patients studied with acetazolamide-use improved more than just weight loss alone.[17] Improvement in retinal nerve fiber layer thickness and other measurements calculated using OCT can be seen in patients undergoing acetazolamide and weight-loss treatment as suggested by the IIHTT.[36] Acetazolamide use not only improved visual outcomes of patients, it also improved quality of life measurements.[17] Amongst others, common adverse reactions to acetazolamide include: paresthesia, nausea, fatigue, headache, dysgeusia, and diarrhea.[17] Typical dosing begins at one gram daily, divided equally at 500 mg twice daily or 250 mg four times daily.[7,15] Slow increases in dosages, as tolerated, may occur leading to total daily doses of 2-4 grams.[4,7,15,17] Other medications that have been used in patients with IIH include: methazolamide, topiramate, and furosemide.[3-5,7,9] Corticosteroids have been used in patients with IIH but is controversial because of side effects such as weight gain.[3,4,7]

Neurosurgical treatments are reserved for severe and sudden onset IIH, including patients with rapid vision loss, continued vision loss with standard treatment, the presence of a RAPD, and macular edema with severe papilledema.[3,4,7-9] Neurosurgical procedures include optic nerve sheath fenestration (ONSF), dural venous sinus stenting, and cerebral spinal fluid diversion. ONSF is completed by creating slits in the dural covering of the optic nerve immediately behind the globe.[3,7] ONSF is typically reserved for patients with severe papilledema and vision loss without headaches.[7] This allows for a lowering of pressure around the nerve head and subsequent reduction in papilledema appearance and improvement in visual field and function in 70-90% of patients.[3,4,9] Complications surrounding ONSF can occur in 4.8-45% of cases and include: ischemic optic neuropathy, vascular occlusion, diplopia, dilated pupil, and infection.[3,8,9] Dural venous sinus stenting has been shown to reduce intracranial pressure while improving signs and symptoms of IIH, including improvement in headaches.[3,7,9,28] Cerebral spinal fluid diversion, which is typically reserved for patients with severe papilledema, vision loss, and headaches due to IIH, involves a lumboperitoneal shunt or ventriculoperitoneal shunt.[3,4,7-9] Complications surrounding cerebral spinal fluid diversion occur in around 50% of performed procedures and include: infection, shunt blockage, and intracranial hypotension amongst others.[3,4,7-9]

Papilledema and intracranial pressure reduction have been observed in patients with weight loss from lifestyle modifications.[3,4,9,26] Because obesity is commonly associated with IIH, and weight loss has demonstrated improvements in the condition, it is thought that bariatric surgery would be beneficial in these patients, especially in morbidly obese individuals where lifestyle modifications are insufficient.[3,7,26] Bariatric procedures that are typically performed include: laparoscopic gastric bypass and laparoscopic adjustable gastric banding.[9] According to the research by Handley et al. bariatric surgery was beneficial to patients with IIH, having alleviated IIH symptoms.[26] Markey et al. report 92% of bariatric surgery patients showed improvement in IIH symptoms following the procedure.[9] It is suggested that bariatric surgery could prove to be a potential replacement for neurosurgeries in future.[26]

Co-management with neurology and general ophthalmology or neuro-ophthalmology is frequently seen in patients with IIH. Unless chronic or severe, patients are typically followed every three months monitoring papilledema, visual acuity, and visual field.

Conclusion

This case demonstrates the role of case history, clinical observations, and proper imaging in the diagnosis and management of primary pseudotumor cerebri, which is known as idiopathic intracranial hypertension. It is important for clinicians to remember that headaches are commonly associated and reported 75-94% of the time, diplopia can occur and is typically horizontal as a result of a unilateral or bilateral cranial nerve six (abducens nerve) palsy, and the common system of grading papilledema is the Frisén scale, which ranges from normal (stage 0) to severe papilledema (stage 5) and can provide a good prediction of visual outcome, as well as possibly assist with treatment management. Furthermore, it is important for clinicians to remember that magnetic resonance imaging with and without contrast, magnetic resonance venography, and lumbar puncture with opening pressure measurement are critical diagnostic aids and should be obtained in all cases of suspected PTC. No mass, lesion, or hemorrhage should be present, though there are findings observed on MRI that are seen in patients with raised intracranial pressure and include posterior globe flattening, enlarged optic nerve sheath, and empty sella. Opening pressure measurements above 25 cm H_2O are considered diagnostic for elevated intracranial pressure. In 2013, Friedman et al. released pseudotumor cerebri syndrome diagnostic criteria, which is an attempt at being all-encompassing and designed for use with children and adults in cases with papilledema present and absent.[16] It provides the clinician with details to formally diagnose a patient with idiopathic intracranial hypertension. The Idiopathic Intracranial Hypertension Treatment Trial concluded that acetazolamide use in combination with weight loss in comparison to weight loss alone proved more effective in improvements of visual field function. Neurosurgical treatments are reserved for severe and sudden onset PTC, including patients with rapid vision loss, continued vision loss with standard treatment, the presence of a RAPD, and macular edema with severe papilledema. Close monitoring of papilledema, visual acuity, and visual field along with co-management with neurology and neuro-ophthalmology will provide optimum care.

REFERENCES

1. Lee AG, Wall M. Idiopathic intracranial hypertension (pseudotumor cerebri): clinical features and diagnosis. UpToDate. Updated March 2021. Accessed July 15, 2022. https://www.uptodate.com/contents/idiopathic-intracranial-hypertension-pseudotumor-cerebri-clinical-features-and-diagnosis?search=idiopathic%20intracranial%20hypertension%20clinica%20features%20and%20diagnosis&source=search_result&selectedTitle=1~150&usage_type=default&display_rank=1.
2. Bienfang DC. Overview and differential diagnosis of papilledema. UpToDate. Updated May 2019. Accessed May 27, 2020. https://www.uptodate.com/contents/overview-and-differential-diagnosis-of-papilledema?search=overview%20and%20differential%20diagnosis%20of%20papilledema&source=search_result&selectedTitle=1~150&usage_type=default&display_rank=1.
3. Winn HR. *Youmans Neurological Surgery*. 6th ed. Saunders/Elsevier. 2011:1700-1712.
4. Wakerley BR, Tan MH, Ting EY. Idiopathic intracranial hypertension. *Cephalalgia*. 2015;35(3):248-261. doi: 10.1177/0333102414534329
5. Piper RJ, Kalyvas AV, Young AMH, Hughes MA, Jamjoom AAB, Fouyas IP. Interventions for idiopathic intracranial hypertension. *Cochrane Database Syst Rev.* 2015;8. doi: 10.1002/14651858.CD003434.pub3
6. Aylward SC. Pediatric idiopathic intracranial hypertension: a need for clarification. *Pediatr Neurol*. 2013;49(5):303-304. doi: 10.1016/j.pediatrneurol.2013.05.019
7. Thurtell MJ, Bruce BB, Newman NJ, Biousse V. An update on idiopathic intracranial hypertension. *Rev Neurol Dis*. 2010;7(2-3):e56-e68.
8. Friedman DI, Jacobson DM. Idiopathic intracranial hypertension. *J Neuroophthalmol*. 2004;24(2):138-145. doi: 10.1097/00041327-200406000-00009
9. Markey KA, Mollan SP, Jensen RH, Sinclair AJ. Understanding idiopathic intracranial hypertension: mechanisms, management, and future directions. *Lancet Neurol.* 2016;15(1):78-91. doi: 10.1016/S1474-4422(15)00298-7
10. Bruce BB, Kedar S, Van Stavern GP, et al. Idiopathic intracranial hypertension in men. *Neurology*. 2009;72(4):304-309. doi: 10.1212/01.wnl.0000333254.84120.f5
11. Kesler A, Goldhammer Y, Gadoth N. Do men with pseudotumor cerebri share the same characteristics as women? a retrospective review of 141 cases. J Neuroophthalmol. 2001;21(1):15-17. doi: 10.1097/00041327-200103000-00004

12. Digre KB, Corbett JJ. Pseudotumor cerebri in men. *Arch Neurol*. 1988;45(8):866-872. doi:10.1001/archneur.1988.00520320056015
13. Gospe SM, Bhatti MT, El-Dairi MA. Anatomic and visual function outcomes in paediatric idiopathic intracranial hypertension. *Br J Ophthalmol*. 2016;100(4):505-509. doi: 10.1136/bjophthalmol-2015-307043
14. Bidot S, Saindane AM, Peragallo JH, Bruce BB, Newman NJ, Biousse V. Brain imaging in idiopathic intracranial hypertension. *J Neuroophthalmol.* 2015;35(4):400-411. doi: 10.1097/WNO.0000000000000303
15. Idiopathic intracranial hypertension/pseudotumor cerebri. In: *The Wills Eye Manual: Office and Emergency Room Diagnosis and Treatment of Eye Disease*. 6th ed. Lippincott Williams & Wilkins. 2012:273-274.
16. Friedman DI, Liu GT, Digre KB. Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. *Neurology.* 2013;81(13):1159-1165. doi: 10.1212/WNL.0b013e3182a55f17
17. NORDIC Idiopathic Intracranial Hypertension Study Group Writing Committee, Wall M, Mcdermott MP, et al. Effect of acetazolamide on visual function in patients with idiopathic intracranial hypertension and mild visual loss. *JAMA.* 2014;311(16):1641-1651. doi: 10.1001/jama.2014.3312
18. Frisén L. Swelling of the optic nerve head: a staging scheme. *J Neurol Neurosurg Psychiatry*. 1982;45(1):13-18. doi: 10.1136/jnnp.45.1.13
19. UpToDate. Modified Frisén scale for grading papilledema. Accessed July 15, 2022. https://www.uptodate.com/contents/image?imageKey=NEURO%2F73887
20. Bidot S, Clough L, Saindane AM, Newman NJ, Biousse V, Bruce BB. The optic canal size is associated with the severity of papilledema and poor visual function in idiopathic intracranial hypertension. *J Neuroophthalmol.* 2016;36(2):120-125. doi: 10.1097/WNO.0000000000000318
21. Sibony PA, Kupersmith MJ, Feldon SE, Wang J, Garvin M, OCT Substudy Group of the NORDIC Idiopathic Intracranial Hypertension Treatment Trial. Retinal and choroidal folds in papilledema. *Invest Ophthalmol Vis Sci*. 2015;56(10): 5670-5680. doi: 10.1167/iovs.15-17459
22. Sibony PA, Kupersmith MJ, OCT Substudy Group of the NORDIC Idiopathic Intracranial Hypertension Treatment Trial. "Paton's Folds" revisited: peripapillary wrinkles, folds, and creases in papilledema. *Ophthalmology*. 2016;123(6):1397-1399. doi: 10.1016/j.ophtha.2015.12.017
23. Chen JJ, Thurtell MJ, Longmuir RA, et al. Causes and prognosis of visual acuity loss at the time of initial presentation in idiopathic intracranial hypertension. *Invest Ophthalmol Vis Sci*. 2015;56(6):3850-3859. doi: 10.1167/iovs.15-16450

24. Cello KE, Keltner JL, Johnson CA., Wall M, NORDIC Idiopathic Intracranial Hypertension Study Group. Factors affecting visual field outcomes in the idiopathic intracranial hypertension treatment trial. *J Neuroophthalmol*. 2016;36(1):6-12. doi: 10.1097/WNO.0000000000000327
25. Park JC, Moss HE, McAnany JJ. The pupillary light reflex in idiopathic intracranial hypertension. *Invest Ophthalmol Vis Sci*. 2016;57(1):23-29. doi: 10.1167/iovs.15-18181
26. Handley JD, Baruah BP, Williams DM, Horner M, Barry J, Stephens JW. Bariatric surgery as a treatment for idiopathic intracranial hypertension: a systematic review. *Surg Obes Relat Dis*. 2015;11(6):1396-1403. doi: 10.1016/j.soard.2015.08.497
27. Wall M, Kupersmith M, Kieburtz K, et al. The idiopathic intracranial hypertension treatment trial. *JAMA Neurol.* 2014;71(6):693-701. doi: 10.1001/jamaneurol.2014.133
28. Sivasankar R, Pant R, Indrajit IK, et al. Imaging and interventions in idiopathic intracranial hypertension: a pictorial essay. *Indian J Radiol Imaging.* 2015;25(4):439-444. doi: 10.4103/0971-3026.169464
29. Mohson KI, Auday N. Role of orbital ultrasound in the assessment of clinically detected papilledema. *J Med Ultrasound*. 2019;27(3):135-40. doi: 10.4103/JMU.JMU_70_18
30. Salmon JF. Neuro-ophthalmology. In: *Kanski's Clinical Ophthalmology.* 9th ed. Elsevier; 2020:745-825.
31. Friedman DI, Jacobson DM. Diagnostic criteria for idiopathic intracranial hypertension. *Neurology*. 2002;59(10):1492-1495. doi: 10.1212/01.wnl.0000029570.69134.1b
32. Bandyopadhyaya S, Jacobson D. Clinical features of late-onset pseudotumor cerebri fulfilling the modified dandy criteria. *J Neuroophthalmol.* 2002;22(1):9-11. doi: 10.1097/00041327-200203000-00003
33. Bruce BB, Kedar S, Van Stavern GP, Corbett JJ, Newman NJ, Biousse V. Atypical idiopathic intracranial hypertension. *Neurology*. 2010;74(22):1827-1832. doi: 10.1212/WNL.0b013e3181e0f838
34. Aylward SC, Aronowitz C, Roach ES. Intracranial Hypertension Without Papilledema in Children. *J Child Neurology*. 2016;31(2):177-183. doi: 10.1177/0883073815587029
35. Optical Coherence Tomography Substudy Committee, NORDIC Idiopathic Intracranial Hypertension Study Group. Papilledema outcomes from the optical coherence tomography substudy of the Idiopathic Intracranial Hypertension Treatment Trial. *Ophthalmology.* 2015;122(9):1939-1945. doi: 10.1016/j.ophtha.2015.06.003