

THE SWOLLEN OPTIC DISC WITH AN EMPHASIS ON THE PEDIATRIC PATIENT

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Abstract

The decision to determine whether a swollen disc is caused by a vision or life-threatening condition, or if the disc appearance is physiological is an important clinical responsibility. A 4-year-old black female presented with sudden onset esotropia, head turn, decreased visual acuity, swollen discs, and disc hemorrhages secondary to papillitis. The differential diagnoses of blurred optic discs are reviewed, with emphasis on the most common pediatric etiologies, and the manifestations and prognostic implications. When making a differential diagnosis of blurred disc border(s), the doctor should consider several factors such as history, optic nerve head and fundus appearance, age, symptoms, and results of in-office optometric tests. Often it is necessary to refer the patient for further testing.

Key Words

drusen, esotropia, head turn, hyperopia, intracranial pressure, magnetic resonance imaging, optic neuritis, papilledema, papillitis, pediatric patient, pseudopapilledema, pseudotumor cerebri, retrobulbar optic neuritis

INTRODUCTION

When examining the optic nerve, eye care practitioners place a strong importance on its appearance. At times its border (or margin) does not appear distinct. An important clinical decision is to determine whether a swollen disc margin is caused by a life or vision threatening disease, or if the abnormal disc is physiological, warranting documentation, monitoring, and patient education.

A key factor in determining whether an abnormal finding is physiological or pathological is “the company it keeps,” e.g., signs and symptoms. It is likely that a patient experiencing headaches, transient visual obscurations (TVOs), changes in visual acuity (VA), and/or eye pain has a pathological cause to the optic disc swelling. In contrast, it is likely that an asymptomatic and healthy patient has a physiological cause to the swelling.¹

The differential diagnosis of blurred disc margins in a child can be different than that of an adult for several reasons. First, the most common pathological causes of blurred disc margins in children may have different etiologies, manifestations and prognostic implications for visual recovery than in adults. Second, the differential diagnosis in a child may be more challenging because of the likely absence of a reliable history and reported symptoms. Third, a child will likely have a shorter attention span than an adult, po-

tentially limiting the duration and detail of the practitioner’s view of the nerve. Finally, the child may not be able to respond to some optometric tests that are easier to administer and interpret with adults. This circumstance often requires referral for further specialized evaluation. I present the case report of a pediatric patient with blurred disc margins, and then proceed to discuss the differential diagnoses, manifestations, and prognostic implications of a swollen disc in an adult versus a child, with an emphasis on the latter.

CASE REPORT

A 4-year-old black female, accompanied by her mother, presented at the State University of New York, State College of Optometry’s Pediatric Vision Clinic with a chief complaint of a left eye turn of four days duration and a leftward head turn for three days. The mother was not sure whether the eye was turning in or out. She stated that her daughter complained of “darkening vision” one week prior to the visit. The mother claimed that her daughter was in good health and that since the onset of these symptoms, there were no changes in her behavior, motor skills, cognitive skills, hearing, or speech. She reported that the patient had no recent illnesses or vaccinations and had not been taking any medications.

The habitually uncorrected entering Snellen VA was OD 20/50 and OS 20/40(-). During the acuity measurement,

the patient demonstrated a marked head turn to the left. Her pupillary reactions and extraocular motilities did not indicate any neurological implications. An afferent pupillary defect was not present. On distance and near cover test (CT), the patient manifested an 18 prism diopter intermittent left esotropia. Noncycloplegic static retinoscopy revealed low hyperopia with a slight amount of with the rule astigmatism OU. This correction did not improve the monocular acuities. She demonstrated gross line stereo acuity and no random dot stereo acuity. Using Ishihara plates, her color vision (CV) was normal. The anterior segment seen under slit lamp was unremarkable. The cycloplegic retinoscopy result was similar to that of the noncycloplegic retinoscopy.

Examination of the posterior segment of the patient's eyes revealed mildly blurred disc margins 360 degrees OU and flame shaped disc hemorrhages OU. Her cup to disc ratio was 0.2 with pink rim tissue. She had an artery to vein ratio of 1:2 with tortuosity. We made a diagnosis of bilateral swollen discs, and the patient was referred for a pediatric neurological consultation with imaging recommended. The magnetic resonance imaging (MRI) results were negative, and the neurologist's impression was normal intracranial pressure (ICP), but that the patient should be monitored for any headache symptoms which might relate to pseudotumor cerebri (PTC).

On her three-week follow-up examination, she no longer manifested a head turn or strabismus. At this time the mother remembered that the child had experienced viral symptoms some two months previously, but they had not sought medical attention. The remaining history was otherwise unremarkable. The HOVT letter chart distance VA was 20/25 OD, 20/30+ OS. On CT, she manifested a 3 prism diopter intermittent esophoria at distance and orthophoria to slight exophoria at near. Her discs were slightly elevated with blurred margins OU. The vessels were of normal caliber, and there were no hemorrhages. The assessment was a resolving papillitis and esotropia caused by a viral infection.

On her six-week follow-up examination, distance VA using the HOVT chart was 20/20 OD and OS. The CT indicated orthophoria at distance and near. Her optic nerves were pink with distinct margins

OU. The patient was diagnosed with a resolved papillitis and was scheduled for a three-month follow up visit.

DISCUSSION

In this paper we divide the condition of swollen optic disc into four etiologies, namely:

1. Inflammation of the optic nerve

Included are the terms optic neuritis, papillitis and retrobulbar neuritis.

2. Elevated intracranial pressure

The clinical conditions include papilledema and pseudotumor cerebri

3. Neurological or systemic disease

4. **Physiological** Included are pseudopapilledema, optic disc drusen, hyperopia and embryonic remnants.

In order to provide completeness, we have included adult presentations; however, the pediatric population is the primary focus of the discussion.

1. Disc swelling caused by Inflammation of the optic nerve

a. General information

Much information about optic neuritis was obtained from the Optic Neuritis Treatment Trial (ONTT), a multicenter study enrolling adult patients with acute unilateral optic neuritis.² This trial provided useful information regarding the efficacy of corticosteroids in treating optic neuritis, its clinical profile, natural history, and relationship to multiple sclerosis (MS). MS is a progressive disease in which disseminated demyelination of the central nervous system (CNS) occurs.

The pathogenesis of optic neuritis is demyelination of the optic nerve.³ Swelling of the nerve tissue results from a combination of inflammatory cell action, myelin sheath and nerve tissue degeneration³ and impaired axonal transport.^{1,4} In *papillitis, or anterior optic neuritis*, the swelling is located at the disc, and thus is visible to the clinician. In *retrobulbar optic neuritis*, the swelling is posterior to the globe and hence the disc appears normal.¹

The typical patient presentation with optic neuritis is a young Caucasian female (mean 32 +/- 7 years in the ONTT) with an acute monocular central vision loss and ocular pain that worsens on eye movement.^{2,3} The loss of vision is commonly abrupt, occurring over several hours to several days. VA may range from minimally reduced vision to complete blind-

ness with no light perception. Patients frequently describe their vision loss as a diffuse blur. Typically the ocular pain lasts only a few days, and it can range from mild to debilitating. An additional symptom which may occur is photopsia (spontaneous black squares, light flashes, or showers of sparks).³

Less common presentations of optic neuritis can also occur. Acute optic neuritis in adults can rarely present as bilateral and simultaneous. Patients with acute optic neuritis may maintain normal vision yet complain of a peripheral vision loss. Acute optic neuritis is not age dependent and can occur in childhood (discussed extensively below) and into the 6th and 7th decades. This disease can also present in chronic and subclinical patterns.³

Patients with acute optic neuritis typically demonstrate optic nerve dysfunction, which includes reduced VA, contrast sensitivity and CV. Although the contrast sensitivity deficit often parallels the decrease in VA, the CV deficit is greater than expected when compared to the VA reduction. If the optic neuritis is unilateral, a relative afferent pupillary defect will most likely be detectable, regardless of whether it is anterior or retrobulbar. Patients with optic neuritis frequently have a reduced sensation of brightness in the involved eye.³ A visual field (VF) loss will often exist. Although some authors report that the VF loss is most commonly central or cecentral,^{1,5} the ONTT study found the VF losses to have varied patterns and prevalence, including diffuse (48%), altitudinal, arcuate, or nasal step (20%), central or cecentral (8%), or other types (24%).^{2,3}

The clinical appearance of the optic disc in optic neuritis varies from swollen (anterior involvement) to normal (retrobulbar). In the ONTT, 35% of patients presented with swollen discs.⁶ Although the appearance of the swelling can vary from slightly to markedly blurred, the degree of swelling does not correlate with the severity of VA or VF loss. Disc hyperemia and dilated surface capillaries may also be seen. It is rare to see disc or peripapillary hemorrhages in acute optic neuritis; however, in severe cases they may be observed along with disc hyperemia and elevation. Occasionally a few vitreous cells may be observed overlying the disc.³

The natural history of acute optic neuritis is worsening of the disease over several days to two weeks, and then improvement within one month.^{2,3} Initially the improvement is rapid, and then it typically levels off; however, visual acuity may continue to improve even one year after the initial symptoms began. Other measures of visual function, such as contrast sensitivity, color perception, and VF improve along with the improvement in VA. It is expected that the worse the initial vision loss, the poorer the eventual recovery. Factors that do not appear to affect visual acuity outcome are adult age, gender, optic disc appearance, pattern of VF loss, and laterality. The prognosis for VA recovery after an acute optic neuritis episode is overall extremely good, and most patients recover to normal or near normal vision. However, some patients may experience a severe visual loss after a single event, and others who have recovered to "normal" visual acuity may complain of movement induced photopsias and may have visual deficits when tested with more sensitive techniques.³

In adults there is a strong relationship between MS and acute optic neuritis, which is a presenting sign in 20% of MS patients, while 50% of MS patients will at some point experience optic neuritis. Optic neuritis is thought to be a "forme fruste" of MS, due to the similarities between these two diseases in terms of demyelination pathogenesis, incidence, cerebral spinal fluid (CSF) findings, histocompatibility data, MRI results, family history, and other features.³

Other systemic diseases causing optic neuritis in adults include sarcoidosis, syphilis, HIV/ AIDS, Systemic Lupus Erythematosus (SLE) and other vasculitides, Lyme Disease, and Sinus Disease. Adult optic neuritis can rarely present as a manifestation of other underlying systemic infections (viral more commonly than bacterial), or can present after vaccinations. Finally, idiopathic cases of optic neuritis occur in the absence of signs of MS or other systemic disease.³

b. The pediatric patient

Acute optic neuritis in children is considered fundamentally different than in adults. Pediatric optic neuritis is commonly bilateral and anterior.⁷ It is usually associated with a postinfectious condition that is classically considered unassociated

with MS.¹ In contrast, the adult presentation of acute optic neuritis is usually unilateral and retrobulbar,⁶ and is closely related to MS, as described above.³ Headaches are a more common symptom in patients with pediatric optic neuritis.^{7,8} Pediatric patients may not be the best historians; thus it may be more difficult to obtain an accurate history of onset of symptoms in children.¹ Also, children may not notice unilateral vision loss, and may tolerate bilateral vision loss until it becomes so severe that it is incapacitating. Older children may deny symptoms if they sense panic.⁷

In pediatric optic neuritis, the initial visual loss is commonly severe and can include a lack of light perception. Despite the profound visual loss, there is generally an excellent prognosis for visual recovery.¹ Commonly, improvement begins by the third week after symptoms first appeared. Maximum improvement is usually achieved by six months.⁷ Kriss et al suggested that the greater potential for remyelination in younger patients may explain the improved visual prognosis among children as compared to adults.⁸ In most children vision recovers to 20/20, but some optic pallor commonly persists.^{8,9} Generally, a slower onset of visual loss is followed by a slower and less complete visual recovery.¹

A febrile or flu like illness often precedes pediatric optic neuritis by days or weeks. Additionally, DPT vaccines and other immunizations have been found to precipitate optic neuritis in children.¹ A list of infectious and post infectious causes, along with noninfectious causes of pediatric optic neuritis is given in Table 1.

Research from the 1980s has demonstrated that the relationship between post-infectious childhood optic neuritis and MS may not be as absolute as was once thought.¹ Other research indicates that the delayed onset of pediatric optic neuritis after infection or immunization, in addition to the bilateral involvement in the majority of such cases, suggests a systemic disease condition, such as systemic autoimmune demyelination, rather than a local viral infection of each optic nerve.⁹

Riikonen, et al., proposed that a combination of abnormal immunological responses (i.e., demyelinating autoimmunity), precipitated by infectious agents (e.g., viral or bacterial disease), in genetically susceptible individuals, may

Table 1.
Infectious, Postinfectious, and Noninfectious Causes of Pediatric Optic Neuritis^{1,7,10-15}

Infectious and Postinfectious

Rubella
Rubeola (measles)
Paramyxovirus (mumps)
Varicella zoster (chicken pox)
Pertussis (whooping cough)
Boriella burgdorferi (Lyme disease)
Epstein-Barr virus (infectious mononucleosis)
Rochalimaea (cat scratch disease)
Treponema pallidum (syphilis)
Toxocara canis
Toxoplasmosis
Tuberculosis
Rickettsia
Coxsiella burnetti
Brucella
Q fever
Vaccinations (DPT, etc.)

Noninfectious

Multiple Sclerosis
Devic Disease
Sarcoidosis
Bee Venom
Vasculitis (e.g., lupus)

lead to either optic neuritis or MS.¹⁶ One study showed that a significant number of children with optic neuritis eventually developed MS and over half of these children had a bacterial or viral infection within two weeks before the symptoms of optic neuritis appeared. Vaccinations with live or attenuated virus (e.g., polio, vaccinia, rubella, influenza) had been given to several of these patients, and subsequent vaccinations caused the optic neuritis to exacerbate.¹⁷ In contrast, killed virus components have not been shown to cause optic neuritis.^{1,17} Another study describes similar findings, and therefore Brodsky concluded that postinfectious optic neuritis may be a harbinger of MS in children.^{1,18}

It is thought that there is a greater chance for a child with unilateral optic neuritis to develop MS than a child with a bilateral presentation.^{8,17} However, the risk of developing MS for children with bilateral optic neuritis is not negligible.⁸

When pediatric optic neuritis is accompanied by several neurological signs, the three primary etiologies are MS, acute disseminated encephalomyelitis (ADE),

and Devic Disease. ADE, also known as postinfectious encephalomyelitis, is an uncommon inflammatory demyelinating disease of the CNS that usually follows a viral illness or vaccination by days or weeks. Devic disease is an acute and self-limiting disease caused by necrotizing myelopathy of the spinal cord, optic nerves and chiasm (but sparing the rest of the brain).¹

Some specific differential signs regarding optic neuritis exist. As mentioned above, a few vitreal cells may be occasionally observed overlying the optic disc in demyelinating anterior optic neuritis. However, if there is a significant cellular reaction observed in eyes with papillitis, causes other than demyelination should be considered, such as sarcoidosis, tuberculosis, syphilis, or Lyme disease. Additionally, when macular or peripapillary exudates accompany the disc swelling, neuroretinitis should be considered as a diagnosis. Disc or peripapillary hemorrhages are uncommon in eyes with papillitis. Nevertheless, the presence of disc hemorrhages along with a swollen disc is a common appearance in anterior ischemic optic neuritis.³

Summary for disc swelling in the pediatric patient caused by inflammation of the optic nerve

When examining a child who may have optic neuritis, a thorough history of recent infection, immunization, systemic disease, bee sting, tick bites, or neurological symptoms should be obtained.¹ Important tests in addition to the primary optometric exam include CV, light brightness sensitivity, and VF testing. However, these tests might not produce reliable results with children. MRI testing might be needed to rule out an intracranial mass. MRI is a powerful tool in predicting the risk of developing MS in both pediatric and adult patients who have optic neuritis.¹⁹ A referral for further testing to rule out additional causes of blurred discs, such as other local ocular diseases (see Table 2), elevated ICP (discussed next), and systemic or neurological disease may also be indicated. Table 2 summarizes the most common ocular diseases that cause disc swelling in pediatric patients.

Table 2.
Disc Swelling Due to Local/
Primary Ocular Disease in
Children^{1,4}

Optic Neuritis
Uveitis
Hypotony (e.g., after glaucoma surgery)
Trauma
Optic Disc tumors
Optic disc hemangioma
Tuberous sclerosis
Optic disc glioma
CHRPE
Retrolbulbar Tumors
Leber Idiopathic Stellate Neuroretinitis

2. Disc Swelling Caused by Elevated Intracranial Pressure

Papilledema is the term used for disc swelling caused specifically by elevated ICP.^{1,20} Elevated ICP is transmitted throughout the CNS via the cerebrospinal fluid (CSF), affecting various neurological structures. For example, the brainstem may be displaced downward, stretching the VI cranial nerve and causing a palsy of that nerve.²⁰ Since the CNS is a relatively closed system, the increased ICP is transmitted via the nerve sheath to both optic nerves, resulting in bilateral disc swelling. Specifically, the elevated ICP causes optic nerve swelling by impairing retrograde axoplasmic transport.⁴ Since only the anterior portion of the optic nerve is directly visible to the clinician, the effect of the elevated ICP is observed as optic disc swelling.

Papilledema can occur at any age. There is no upper age limit, and there is evidence that it occurs frequently in infants and children.³

There are four stages or types of papilledema: early, fully developed, chronic, and atrophic.

In the *early phase*, typical signs are disc hyperemia, blurring of the peripapillary nerve fiber layer (NFL), slight or partial optic disc swelling, blurring of the disc margins, peripapillary flame-shaped hemorrhages, and absent spontaneous venous pulsation (SVP).³

SVP ceases when ICP rises above 200 mm water.^{1,3} It has been found that ICP, measured by the opening pressure during lumbar puncture (LP), can vary within wide limits from one moment to the next among both normal patients and those with elevated ICP. Thus, the observation

of SVP only indicates that at that moment, the ICP is below 200 mm water. Therefore, Miller concluded that a patient who presents with swollen discs and a positive SVP may still have papilledema if the ICP was not elevated at the moment of observation.³ Brodsky, however, believes the presence of (+) SVP is sufficient to rule out elevated ICP.¹

Conversely, the absence of SVP may also provide limited information since only 80 % of the normal population actually has SVP.^{3,20} In other words, 20% of normal individuals (patients without increased ICP) lack SVP. Therefore, the absence of SVP is not pathognomonic for elevated ICP. In the absolute sense, looking for SVP in an individual who is suspect for papilledema can only be helpful in the following situation: A patient who had SVP in a past exam and now presents with bilateral swollen discs and no SVP; this patient is likely to have papilledema.

In the *fully developed stage* of papilledema, disc swelling becomes more obvious, veins become engorged, and numerous splinter hemorrhages occur next to the disc margin. During further progression, the surface of the disc becomes elevated above the retinal surface, along with capillary dilation on the disk surface, blurring of the disc margin, opacification of the nerve fiber layer and resulting surface vessel obscuration, along with the formation of microaneurysms, cotton wool spots (CWS), papillary and peripapillary tortuous vessels, and flame-shaped hemorrhages. In severe cases of papilledema, circumferential retinal folds (Paton's lines), hard exudates and hemorrhages may develop in the peripapillary and macular area. Since the nerve fibers in the macula have a radial fan shaped appearance, hemorrhages and exudates in this area become fan or star shaped. Also, since vascular compromise on and around the disc is responsible for these macular changes, the star figure is usually more prominent on the nasal side of the fovea, towards the disc. Also, subhyaloid and vitreous hemorrhages may develop, especially if the rise in ICP is rapid. The central cup is usually retained even in the most severe cases of acute papilledema.³

In the *chronic papilledema* of several months, the hyperemia and hemorrhages resolve, the disc develops a round, compact shape, the central cup ultimately be-

comes obliterated, and the disc takes a milky, gray appearance as hard exudates become apparent in the superficial disc substance. These exudates resemble optic disc drusen and hence may be misdiagnosed as pseudopapilledema. Slit-like or diffuse NFL atrophy is also often observed.³

In *postpapilledema atrophy*, the disc becomes atrophic, retinal vessels become narrow and sheathed, and the NFL can no longer be visualized. Persistent pigmentary changes or choroidal folds are often observed in the macula as well.³

Papilledema is usually bilateral, especially in advanced cases, and symmetric between both eyes. Occasionally papillitis can present as unilateral or asymmetric, and although this is not uncommon in adults, it is rare in children.^{3,21} Papilledema may be unilateral in the early stages.¹ Other causes of unilateral papilledema include unilateral optic disc dysplasia (in which a congenital anomaly of the optic nerve sheath prevents transmission of the CSF pressure), asymmetrical cases in which signs of papilledema are overlooked in the "normal" disc, and the Foster Kennedy syndrome. In this syndrome, unilateral optic atrophy occurs due to a frontal lobe or olfactory groove tumor. The atrophic nerve appears unswollen due to the loss of nerve fibers and lack of elevated CSF pressure transmission. The other optic nerve swells since the NFL is intact and the elevated CSF pressure is transmitted, resulting in unilateral papilledema. Papilledema may be unilateral or asymmetrical in the cases of other intracranial mass lesions, especially abscesses. In such cases the papilledema is usually ipsilateral to the lesion.³

The course of development of papilledema can be variable depending on the etiology; it can develop as quickly as a few hours. Fully developed papilledema can also disappear within hours, days, or weeks, depending on the method in which the ICP is lowered.³

Nonvisual symptoms are generally severe and bothersome to patients with papilledema. These symptoms are a result of the increased ICP affecting various brain structures. Common nonvisual symptoms include a headache which increases upon coughing or straining, nausea, and vomiting.³ These symptoms are frequently present upon awakening.¹

Diplopia can appear as a symptom and is usually the result of a bilateral or unilateral incomplete sixth nerve palsy. Third or fourth nerve palsies rarely occur, and are likely due to mass lesions.¹

Typically, patients with early and fully developed papilledema are visually asymptomatic, and neither VA nor VF are affected. When visual symptoms are present, they can indicate an impending permanent visual dysfunction. When a VF defect is present, it is most commonly a mild or moderate concentric enlargement of the physiologic blind spot, and the patient may be symptomatic or asymptomatic. Arcuate scotomas and nasal steps may also occur. Other patients may have a variable loss of VA, VF, or both. Hemorrhages or macular pathologies such as choroidal folds, exudates, macular edema, or macular pigment changes may cause patients to lose central vision in the early stages.³

An important differentiating test used in suspected cases of early or fully developed papilledema with vision loss is CV testing. In cases where vision is compromised, it is necessary to differentiate between macular and optic nerve pathology. As mentioned above, papilledema does not usually affect vision; however, vision can be affected in patients with papilledema due to secondary macular pathology. In such cases, the CV will be spared despite the decreased VA. In contrast, if there is an evolving optic neuropathy due to chronic papilledema, CV will be affected along with VA.¹

In severe cases of papilledema, patients may experience TVO episodes lasting a few seconds and have visual experiences ranging from mild blurriness to total blindness with a rapid and complete recovery. Some patients experience a rapid gray-out of vision, whereas others describe positive visual phenomena (phosphenes, photopsias, and scintillating scotomas) that obscure their vision. Typically these obscurations can occur 20-30 times a day. TVO's are thought to be caused by momentary ocular or brain ischemia.¹ TVO's are usually precipitated by changes in posture, especially from lying down to sitting, or from sitting to standing.³ They can also be caused by a Valsalva maneuver, which is any forced expiratory effort against a closed airway such as coughing, vomiting, or lifting heavy objects.²² Here, the muscles of the

Table 3.
Causes of Elevated Intracranial Pressure and Resulting Symptoms^{4,20}

Causes

- 1) Intracranial Mass
- 2) Obstructed Cranial Venous Outflow
- 3) Decreased CSF Outflow (PTC)
- 4) Excess CSF Production (choroidal plexus papilloma)

Symptoms

- 1) HA upon wakening
- 2) Transient Visual Obscurations
- 3) Nausea, vomiting
- 4) Diplopia due to VI > IV nerve palsies

chest, abdomen and diaphragm contract while the glottis remains closed, resulting in increased intrathoracic or intraabdominal pressure.^{23,24} This pressure is transmitted to the eye and brain, causing increased venous pressure in these structures,²⁴ and resulting in transient ocular and brain ischemia.

In chronic papilledema, constriction of the VF may occur. The constriction is usually worse nasally than temporally, and progresses to leave a temporal island of vision before complete blindness occurs.³ In cases of chronic papilledema, children share the same risk as do adults for permanent visual loss.¹

The visual prognosis in patients with papilledema can be uncertain. A general overview of papilledema is: the more rapid the development, the greater the danger to sight; and the more severe, the poorer the visual prognosis. Ominous retinal signs include narrowing of retinal arteries, often with sheathing, loss of peripapillary NFL, and disc pallor that is concurrent with papilledema. The above signs indicate that irreversible changes in the optic nerve tissue have occurred, and these patients often have clinical evidence of visual dysfunction.³ In severe cases loss of vision can occur over a period of weeks, and hence the finding of decreased VA demands aggressive and urgent intervention. Signs that do not appear to affect visual prognosis include severely engorged veins, retinal hemorrhages, and hard and soft exudates.^{1,3}

Table 3 lists the causes of elevated ICP leading to papilledema, along with symptoms consistent with elevated ICP. One such cause of elevated ICP is

Table 4.
Criteria for Primary Pseudotumor Cerebri Diagnosis²⁰

- 1) Signs of elevated intracranial pressure (papilledema)
- 2) Symptoms of elevated intracranial pressure (Table 3)
- 3) Normal size ventricles on MRI, CT
- 4) Normal CSF composition
- 5) Normal neurological exam (VI nerve palsy allowed)
- 6) Alert and oriented patient
- 7) All other causes of PTC/ elevated ICP have been ruled out

Pseudotumor Cerebri (PTC). This is thought to be caused by decreased CSF outflow.¹ Primary PTC is characterized by signs and symptoms of elevated ICP without evidence of mass lesion or hydrocephalus.³ Synonymous terms for PTC are idiopathic intracranial hypertension and benign intracranial hypertension. The latter diagnosis is less preferred because of the recognized visual morbidity potentially accompanying chronic papilledema.¹ The criteria for diagnosis of primary PTC are listed in Table 4.

PTC has a very different presentation in adults than in children. Whereas adult PTC is considered a disease of obese women of childbearing age, pediatric PTC has no gender predilection, nor is obesity considered an important epidemiological factor. Children with PTC manifest behavioral changes and neurological deficits more often than do adults (see Table 5). In contrast, children with PTC have the same potential for visual loss as adults, and hence surgical intervention in cases of VA loss is not age dependent.¹

In children the most common causes of PTC are secondary to dural venous thrombosis, steroid withdrawal, and malnutrition associated with refeeding. Here, the previously nutritionally deprived child receives nourishment and experiences accelerated brain growth which is excessive for skull volume. This growth results in ICP elevation.²⁵ Additional causes of PTC among children include neurological disease (e.g., arteriovenous malformation drainage into venous sinus, meningitis), systemic disease (e.g., SLE, Addison disease, anemia), and ingestion of exogenous agents (e.g., vitamin A intoxication from acne medication, tetracycline or minocycline therapy for acne, thyroxine replacement in hypothyroidism).¹

Table 5.
Contrasting Pseudotumor Cerebri in Adults and Children¹

	Adults	Children
Gender predominance	10:1 female predominance after puberty	50:50 before puberty
Obesity as factor	yes (rare in non-obese females)	not a factor under age 10
Spontaneous remission	rare	common, may also occur after LP
Behavioral/ neurological Sx's	rarely manifested	more commonly manifested (listlessness, somnolence, dizziness, ataxia, seizures, VI nerve palsy/ other pareses)

Summary for disc swelling caused by elevated ICP in the pediatric patient

When a pediatric patient is suspected of having papilledema, it is important to obtain a detailed history regarding symptoms consistent with elevated ICP, and symptoms of and risk factors for PTC. Important additional testing to the primary optometric exam include CV if a decrease in VA is found, and VF testing (depending on the child's ability to respond reliably). Seeking optic nerve and fundus signs consistent with the stages of papillitis will help guide the diagnosis. Also, referral for MRI to rule out a mass lesion or ventricular involvement suggestive of hydrocephalus will likely be necessary for the diagnosis. Measurement of the ICP should be done along with analysis of the CSF to determine if any infectious, infiltrative, or neoplastic causes accompany the papilledema. In difficult cases, fluorescein angiography and/ or ultrasonography may help detect papilledema. Additional testing to rule out systemic and neurological disease may also be necessary.³

3. Disc Swelling Caused by Neurologic or Systemic Disease

One neurological disease causing disc swelling is hydrocephalus, a disease in which there is excess CSF in the cerebral ventricles.¹ Some common pediatric causes of hydrocephalus include choroid plexus papilloma, stenosis or gliosis of the sylvian aqueduct, Chiari malformations, Dandy-Walker malformation, cerebral hemorrhage, and cerebral infection.¹ Other pediatric neurological and systemic diseases which may cause disc swelling are listed in Table 6.

4. Physiologic Causes of Disc Swelling

Pseudopapilledema is the umbrella term in which normal variations of the disc cause the border to appear swollen.¹ A hyperopic, small disc can appear swollen because the nerve tissue is crowded as it passes through the small scleral canal. Also, myelinated NFL and other embryonic remnants can cause the disc margin to appear blurred. Additionally, disc drusen can crowd the nerve head tissue, causing a swollen and blurred disc appearance. Other causes of pseudopapilledema are listed in Table 7.

One way to distinguish between a swollen disc and pseudopapilledema is the absence of the previously mentioned signs of papilledema. In pseudopapilledema there should be no CWS, exudates, or hemorrhages (except in the case of disc drusen discussed below). The papillary vessels should not be dilated, and the disc should not be hyperemic. The central retinal vessels should not be obscured, the peripapillary NFL light reflex should be sharp, and a circumpapillary light reflex should be present.¹

In cases where disc drusen are suspected to cause the pseudopapilledema, it may be possible to distinguish the latter from true papilledema by examining the disc vasculature. A disc with buried disc drusen may have increased major retinal vessels with early branching and anomalous trifurcations and quadrifurcations. In contrast, in true pupilledema without drusen, the branching of the disc vasculature should be normal despite the congestion.¹ More importantly, a patient with drusen is asymptomatic, whereas a patient with papilledema will likely complain of the symptoms associated with elevated ICP, as discussed earlier. Another useful sign associated with buried disc drusen is the fluorescein angiographic ap-

Table 6.
Neurological and Systemic Disease Causes of Disc Swelling¹

Neurological Diseases

- 1) Hydrocephalus
- 2) Neurofibromatosis
- 3) Spinal cord tumors
- 4) Subacute sclerosing panencephalitis

Systemic Diseases

- 1) Diabetic Papillopathy
- 2) Sarcoidosis
- 3) Malignant Hypertension
- 4) Leukemia
- 5) Cyanotic Congenital Heart disease
- 6) Child Abuse (Shaken Baby Syndrome)
- 7) Cysticercosis
- 8) Mucopolysaccharidosis
- 9) Infantile Malignant Osteopetrosis
- 10) Craniosynostosis Syndromes

pearance: ophthalmoscopically prominent drusen may exhibit autofluorescence in the preinjection stage, followed by hyperfluorescence at the location of the drusen during the arteriovenous and late stages.²⁷ Also in a fluorescein angiogram of drusen, there is no visible leakage along the major vessels, whereas in true papilledema, there is leakage.²⁷ Additionally with drusen, any disc elevation which may occur is confined to the disc, whereas in papilledema, the elevation extends beyond the disc.¹ Some confounding signs associated with drusen, i.e., signs that appear with both true papilledema and disc drusen, make this differential diagnosis more difficult. Notably, solitary disc hemorrhages and VF losses may appear in both these instances.^{27,28} In the presence of these confounding signs, or when a diagnosis of drusen is not clear from ophthalmoscopic findings alone, a definite diagnosis of drusen can be obtained through CT or ultrasound B scanning.³

MANAGEMENT AND TREATMENT

Often it is not possible to make a differential diagnosis based solely on the optic nerve head appearance. Therefore, symptoms and certain *differentiating* signs are important in making the differential diagnosis. For example, a patient with headaches, TVOs, and unaffected VAs is most likely to have papilledema. On the other hand, a patient with affected

Table 7.
Causes of Pseudopapilledema¹

- 1) Buried disc drusen
- 2) Small scleral canal (hyperopia, nanophthalmia)
- 3) Dysplastic, tilted discs
- 4) Optic nerve hamartoma
- 5) Embryonic remnants
 - Persistent anterior hyaloid artery
 - Epipapillary glial tissue
 - Bergmeister's papilla
 - Juxtapapillary myelinated nerve fibers
- 6) Associated systemic disorders
 - Down Syndrome (early blood vessel bifercation)
 - Alagille Syndrome
 - Kenny Syndrome
 - Leber Hereditary Neuroretinopathy
 - Mucopolysaccharidosis

VAs and eye pain most likely has optic neuritis. In contrast to both the above examples, an asymptomatic and healthy patient with an incidental finding of "disc swelling" is likely to have pseudopapilledema.¹

In order to make the definitive diagnosis, further testing and/or imaging will ultimately be needed in nearly every swollen disc suspect, except in the case of certain pseudopapilledema. An MRI of the brain will rule out a space occupying lesion, hydrocephalus, and demyelination. Measuring the opening pressure on LP will rule out elevated ICP. CSF analysis will rule out infectious, inflammatory, infiltrative, demyelinating, or neoplastic disease as a cause of the swollen disc(s). Blood testing will similarly rule out infectious, inflammatory, infiltrative, metabolic, or neoplastic disease as a cause of the swollen disc(s).

Treatment is directed at the cause of the disc swelling. Patients with optic neuritis should be treated with intravenous steroids, followed by oral corticosteroids, per the ONTT.² A reasonable treatment regimen for an adult might be 250 mg IV methylprednisone q6h for 3 days or 1 gm/day in a single dose x 3 days, followed by a 2 week course of oral prednisone 1 mg/kg/day, with a rapid taper.³ In a child, a recommended regimen is IV methylprednisone 1 mg/kg/day for 3-5 days, followed by a slow taper of oral prednisone, since a higher relapse rate was

found among children whose steroid regimen was rapidly tapered.³ It is noteworthy that controlled studies to determine the efficacy of oral or IV steroids on pediatric optic neuritis have not yet been done.²⁶

If a patient is found to have elevated ICP, treatment is directed toward controlling this pressure. Papilledema with unaffected vision is monitored, but in cases where VA is affected, surgical or pharmaceutical intervention is indicated. Pediatric cases of PTC resolve spontaneously or following one LP more often than do adult cases. Pharmaceutical intervention found to be effective for pediatric PTC includes oral steroids, or a combination of furosemide and high-dose acetazolamide. Surgical intervention for PTC includes optic nerve sheath fenestration and lumboperitoneal shunt, the former of which is preferred for its lower risk and greater effectivity in restoring or preserving vision. Surgical and/or other intervention is almost certainly necessary in the case of an intracranial mass.¹

The present case report concerns a pediatric patient who presented with sudden onset esotropia, head turn, decreased VA, swollen discs, and disc hemorrhages. In order to make a diagnosis, several differentials were considered. The medical history (initial negative report of recent infection, systemic disease, trauma, or behavioral changes), along with the sudden onset of symptoms, the fundus presentation, and the patient's young age limited the list of differentials to papillitis due to an unreported viral infection, and, less likely, to papilledema (e.g., caused by an intracranial tumor, PTC, etc). A referral for examination and imaging by a pediatric neurologist resulted in a normal MRI and impression of normal ICP, tentatively ruling out papilledema. Also, a subsequent history positive for a recent viral infection, and a recovery period consistent with papillitis resulted in this diagnosis.

CONCLUSION

The appearance of blurred optic disc border(s) presents a challenge to the doctor. He or she does not want to unduly alarm the patient or the patient's agent, but at the same time caution is paramount. There is a useful method of clinical thinking. First, observe the nerve head and the surrounding NFL and note the presence or absence of signs consistent with

papilledema, papillitis and pseudo-papilledema. These factors are not conclusive since some signs are overlapping, and in nearly all cases it is not possible to make a differential diagnosis based on these factors alone. Second, consider the patient's age: the most common etiologies of swollen discs, with their manifestations and prognoses for visual acuity recovery are sometimes different for children than adults. Third, determine differentiating symptoms, such as headaches, TVO, or eye pain, even though these may be somewhat unreliable with pediatric patients. Fourth, use differentiating tests such as VA, VF, and CV. At this point the clinician might deem the cause as physiological and elect to monitor the patient. However, if there is even slight uncertainty that the condition is benign, it is necessary to seek further testing that includes MRI and/or CT scan, LP, CSF analysis, and/or blood tests.

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References

1. Brodsky MC, Baker RS, Hamed LM. Pediatric Neuro-Ophthalmology. New York:Springer, 1996:76-116.
2. Beck RW. The optic neuritis treatment trial. In: Kertes PJ, Conway MD, eds. Clinical Trials in Ophthalmology. Baltimore:Williams & Wilkins,1998:271-82.
3. Miller NR, Newman NJ, eds. Walsh and Hoyt's Clinical Neuro-Ophthalmology, vol I, Baltimore, MD:Williams and Wilkins Co; 1998.
4. Acheson JF, Sanders MD, eds. Common Problems in Neuro-ophthalmology. Philadelphia:WB Saunders Co., Ltd, , 1997:78-84.
5. Farris BK. The Basics of Neuro-ophthalmology. St. Louis:Mosby-Year Book, 1991:257-70.
6. Optic Neuritis Study Group. The clinical profile of optic neuritis, experience of the Optic Neuritis Treatment Trial. Arch Ophthalmol 1991;109:1673-78.
7. Kennedy C, Carroll FD, Optic Neuritis in Children. Arch Ophthalmol 1960;63:747-55.
8. Kriss A, Francis DA, Cuendet F, et al., Recovery after optic neuritis in childhood., J Neurol Neurosurg Psychiatry 1988;51:1253-8.
9. Selbst RG, Selhorst JB, Harbison JW, Myer EC. Parainfectious optic neuritis, report and review following varicella. Arch Neurol 1983;40:347-50.
10. Brazis PW, Stokes HR, Ervin FR. Optic neuritis in cat scratch disease. J Clin Neuroophthalmol 1986;6:677-8.
11. Abd Elrazak M. Brucella optic neuritis. Arch Intern Med 1991;151:776-8.
12. Farris BK, Pickard DJ. Bilateral postinfectious optic neuritis and intravenous therapy in children, Ophthmo 1990;97:339-45.
13. Kazarian EL, Gager WE. Optic neuritis complicating measles, mumps, and rubella vaccination. Am J Ophthal 1978;86:544-7.
14. Purvin V, Herr GJ, De Meyer W. Chiasmal neuritis as a complication of Epstein-Barr virus infection. Arch Neurol 1988;45:458-60.
15. Pickens S, Sangster G. Retrobulbar neuritis and infectious mononucleosis. Br Med J 1975; 4:729.
16. Riikonen R, Donner M, Errkila H. Optic neuritis in children and its relationship to multiple sclerosis, a clinical study of 21 children. Dev Med Child Neurol 1988;30:349-59.
17. Riikonen R. The role of infection and vaccination in the genesis of optic neuritis and multiple sclerosis in children. Acta Neurol (Scand) 1989;80:425-31.
18. Bye A, Kendall B, Wilson J. Multiple sclerosis in childhood: a new look. Dev Med Child Neurol 1985;27:215-22.
19. Riikonen R, Ketonen L, Sipponen J. Magnetic resonance imaging, evoked responses, and cerebrospinal fluid findings in a follow-up study of children with optic neuritis. Acta Neurol Scand 1988;77:44-9.
20. Kline LB, Bajandas FJ. Neuro-ophthalmology Review Manual, 4th ed. Thorofare, NJ:Slack, Inc.:1996:85, 137-45.
21. Lepore FE. Unilateral and highly asymmetric papilledema in pseudotumor cerebri, Neurology, 1992; 42:676-78
22. Mosby's Medical, Nursing, and Allied Health Dictionary. 5th ed. St. Louis: Mosby, 1998.
23. Stedman's Medical Dictionary. Baltimore: Williams and Williams, 1961:898.
24. Albert DM, Jakobiec FA. Principles and Practice of Ophthalmology. 2nd ed. Philadelphia:W.B. Saunders, 2000:2225,4224.
25. Couch R, Camfield PR, Tibbles JAR. The changing picture of pseudotumor cerebri in children. Can J Neurol Sci 1985;12:48-50.
26. Hedges TR. Bilateral vision loss in a child with disc swelling. Surv Ophthalmol 1992; 36:424-28.
27. Friedman AH, Beckerman B, Gold DH, et al. Drusen of the optic disc. Surv Ophthal 1977;21:375-90.
28. Savino PJ, Glaser JS, Rosenberg MA, A clinical analysis of pseudopapilledema II: visual field defects. Arch Ophthal 1979; 97:71-75.

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