Tip of iceberg: when unusual vision complaints with a normal examination prompt a closer look

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Key Clinical Message
Nonorganic vision loss (NOVL) is a relatively common condition in pediatric patients. Prompt diagnosis can prevent costly, time-consuming, and frustrating workups. It is valuable for general practitioners and specialists alike to include NOVL in their differential when evaluating patients with visual complaints that are inconsistent with normal examination findings.

Keywords
Conversion disorder, functional vision loss, nonorganic vision loss, temporal hemianopsia

Introduction
Nonorganic vision loss (NOVL), also referred to as functional vision loss, is characterized by the onset of visual deficit or disturbance that cannot be explained by organic pathology. Typically, patients present with subjective visual complaints, but extensive diagnostic imaging, labs, and exams reveal no abnormalities. The first published case of NOVL was in 1865, however, despite its relatively long history there are few studies and case reports on the topic.

This case describes a patient with an unusual presentation of NOVL, reviews the important features of diagnosis and management, and highlights the need for a heightened awareness of this condition. Diagnosis can usually be made from noninvasive tests. Prompt diagnosis can prevent extensive workups that are costly to the patient, as well as time-consuming and frustrating for both the patient and the clinician. A diagnosis of NOVL may be a clue of underlying psychological, academic, or social stress in a pediatric patient. As a consequence, NOVL offers an opportunity for the ophthalmologist and primary care physician to collaborate and address psychiatric, psychosocial, and emotional issues uniquely presenting as visual deficits. In situations where visual disturbances are the sole clinical manifestation, those psychiatric or social issues may otherwise remain undetected and unaddressed.

Patient Presentation
A 12-year-old girl was referred to a pediatric ophthalmologist for evaluation of abrupt onset of monocular visual field loss. Ten days prior to presentation, she had experienced a headache and blurry vision, and 1 day later “half of the vision blacked out.” She saw a local optometrist who performed Humphrey Visual Field (HVF) testing and diagnosed her with monocular temporal hemianopsia of her left eye. She was referred to a community ophthalmologist who confirmed the results with HVF testing. She underwent magnetic resonance imaging (MRI) of the brain and orbit 6 days prior to presentation, which was repeated twice more prior to presentation due to metallic artifacts from dental braces, and all yielded normal results. The patient was given one course of intravenous methylprednisolone with no improvement of her symptoms.

The patient was the product of a full-term, uncomplicated pregnancy and had no significant past medical history. She did not take any medications. There was no reported family history. She was a seventh-grade student...
with no recent changes in her academic progress. She lived with her mother, stepfather, brother, sister, brother-in-law, and nephew. She denied sexual activity and any use of illicit substances. On review of systems, the patient denied any other neurological or visual disturbances.

Upon examination, she had 20/20 visual acuity without correction in both eyes at distance and near. Intraocular pressures were within normal range. The pupils were equal, round, reactive to light without an afferent pupillary defect. She was orthotropic with full ocular motilities. She had

Figure 1. Automated Humphrey Visual Field (HVF) test results. (A) An example of a normal HVF result of the right and left eyes with the physiological blind spot. (B) Our patient’s HVF test showing a temporal visual field defect of her left eye respecting the vertical meridian. (C) Our patient’s left eye HVF compared side-by-side with her left eye manual perimetry results. (D) Of note, binocular visual field testing appeared identical to monocular testing confirming the diagnosis of nonorganic vision loss.
normal color discrimination bilaterally on Ishihara plates. Objective slit-lamp examination of her lids, lashes, conjunctiva, cornea, anterior chamber, lacrimal system, and vitreous did not reveal any abnormalities in either eye. Her dilated fundoscopic examination showed normal optic nerves with no observable retinal pathology. Optical coherence tomography (OCT) was performed to analyze the macula and the retinal nerve fiber layer, and the results were normal. Monocular automated visual field testing revealed clear-cut temporal visual field defect of her left eye respecting the vertical meridian, which is consistent with her prior evaluations (Fig. 1).

On the basis of this history, a number of possible etiologies were considered. The completely intact visual field in her fellow eye made a post-chiasmal lesion very unlikely. An anterior optic tract lesion, optic nerve compression, optic disk pathology, or monocular retinovascular disease can all produce a clinical picture similar to this patient, however, these were unlikely given her lack of afferent pupillary defect, normal color discrimination, and normal retinal exam/OCT. Her remarkable examination and the absence of any structural abnormalities raised some suspicion that the etiology of her visual defect may be nonorganic. The visual fields were tested using manual perimetry, which involves a skilled technician testing each point of the patient’s visual field using illuminated targets of different sizes. This was repeated with each eye separately, and then with both eyes open. Although the manual perimetry of the right eye remains normal, the left eye demonstrates a temporal visual field defect with blurred vertical meridian. When tested with both eyes open, the left temporal defect persists, which defies organic explanation. This confirmed the diagnosis of NOVL. The patient was managed conservatively, primarily with reassurance and follow-up. Within 3 months, her visual defect had completely resolved.

Discussion

NOVL is relatively common, occurring in 1–5% of pediatric patients in general ophthalmology practice [1, 2]. Many of these patients initially present to their primary care physician before seeing an ophthalmologist, so there is significant value in promoting more awareness about this disease. NOVL occurs most frequently in girls aged 8–13 [3, 4]. The most common clinical presentation is reduction in visual acuity with symmetric, bilateral eye involvement [5]. Of NOVL patients with visual field deficit, tunnel vision is the most common [6]. Patients with NOVL are frequently subjected to protracted evaluations and workups that are expensive, frustrating, and time-consuming for both the provider and the patient. It is valuable, therefore, for general practitioners and specialists alike to include NOVL in their differential when evaluating patients with visual complaints that are inconsistent with normal examination findings.

Importantly, NOVL is not a diagnosis of exclusion. Diagnosis of NOVL requires the absence of findings that are attributable to the vision loss, as well as the demonstration of positive test results that are physiologically inconsistent with the presenting visual complaints. In the example of our patient, persistence of the hemianopsia during binocular visual field testing and the absence of a relative afferent pupillary defect distinguish functional hemianopsia (i.e., nonorganic) from organic etiologies of visual field loss [7, 8]. On binocular visual field testing, a normal-functioning fellow eye will compensate for the defect in the field of the affected eye. Additionally, in the case of organic hemianopsia of this size and depth, one would expect to find an ipsilateral afferent pupillary defect. Electro-physiological testing can also be extremely useful in detecting and diagnosing NOVL. For example, visual-evoked potential (VEP) provides the practitioner with objective assessment of a patient’s visual acuity [9]. In some cases, NOVL may be accompanied by organic disease, but in those instances, patients will rarely have a completely normal examination [10, 11].

Psychological and social stressors are thought to play a role in the development of NOVL in children [4, 5, 10, 12]. As a result, a diagnosis of NOVL should prompt clinicians to investigate possible triggers and associated psychopathologies that may have precipitated the child’s ophthalmologic symptoms. Psychiatric conditions, such as conversion disorder, are also implicated as a cause of NOVL, however, psychiatric therapeutic management has not been shown to improve outcome or decrease the period of time the child experiences the visual deficit [2, 13–15]. In a small percentage of children with NOVL, there is a history of physical or sexual abuse [2, 12]. While this association is not common, it is important to consider it and the possibility should be carefully explored. In our patient, the addition of her nephew to the home was the likely trigger for the NOVL as there were no signs of physical or sexual abuse. Recognition and diagnosis of NOVL offers an opportunity for clinicians to identify and address issues in a child’s life that may otherwise remain obscured, benefiting both the patient and the provider.

Malingering is an alternative psychogenic cause of NOVL. In a case of malingering, the patient feigns an illness to achieve a secondary goal. In children, the secondary goal associated with malingering may be as benign and simple as a desire to wear glasses. Unlike conversion disorder, malingering is intentional and purposeful. Malingering children are typically less cooperative with the examination and their findings are less likely to fluc-

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tuate over time. Often times the distinction between a maling erer and a patient with a somatoform/conversion disorder can be quite difficult.

Regardless of the severity of the patient’s clinical presentation, several studies have shown that the most effective management of NOVL is reassurance and follow-up [6, 13, 16–18]. Psychotherapy is rarely indicated and has not been shown to improve outcome [2, 14, 15]. However, in situations where psychotherapy is specifically desired or is likely to confer a clear benefit to the patient, it should be discussed with the family as well as the primary care physician. Overall, the prognosis for these patients is excellent and the vast majority will have full recovery within several months to a year after diagnosis [1, 19].

The clinical approach to a pediatric patient with newonset visual deficits should begin with referral for a basic ophthalmologic examination that includes visual acuity, slit-lamp, and fundoscopic exams, ocular motility, pupillary function, and cycloplegic refraction. If all of those tests yield normal results, more specific ophthalmologic tests should be undertaken prior to referral for a full neurologic workup. If NOVL is suspected, it must be positively supported by examination findings that establish normal visual function, and organic lesions should always be suspected and ruled out. This is important because some severe organic disorders can cause concomitant neuropsychiatric and visual manifestations (e.g., central nervous system neoplasms), so caution must be taken to not automatically dismiss visual complaints when psychiatric features are present. If NOVL is indeed supported by positive test results, the child’s pediatrician and ophthalmologist should collaborate to rule out the presence of an underlying psychogenic cause. Referral for psychiatric evaluation may be helpful in cases with a high suspicion of a psychosocial stress or traumatic event, however, reassurance and follow-up are typically sufficient treatment measures in NOVL.

**Conflict of Interest**

None declared.

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