A Monocular Precaution: A Case of Unilateral Optic Nerve Hypoplasia

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Abstract:
Optic nerve hypoplasia (ONH) is a congenital anomaly that can impact visual acuity over a wide spectrum. We present a 13 year old with unilateral ONH, and the importance of patient education and ocular protection.

Case Report:

❖ Case history
   ➢ Patient demographics
     ▪ 13 year old African American Female
   ➢ Chief complaint
     ▪ Decreased vision OS sc
   ➢ Ocular, medical history
     ▪ Extra ocular muscle surgery at 3 years of age
     ▪ Longstanding severe decrease in visual acuity OD
     ▪ Negative history of patching
     ▪ Poor compliance with previous spectacle correction
   ➢ Medications
     ▪ None
   ➢ Other salient information
     ▪ No known drug allergies
     ▪ No familial history of strabismus, amblyopia, or other eye disease

❖ Pertinent findings
   ➢ Exam findings
     ▪ VA sc
       OD: 20/200
       OS: 20/40-2
     ▪ Adnexa: unremarkable
     ▪ Pupils: PRRL OU, +2APD OD
     ▪ EOMs: Full OU
     ▪ DCTsc: orthophoria
     ▪ NCTsc: orthophoria
     ▪ Retinoscopy
       OD: +0.50+0.75x090
       OS: -1.25DS
• Manifest Refraction:
  OD: +0.50+0.75x090  20/200
  OS: -1.25DS  20/20
• Anterior segment: unremarkable
• Posterior segment:
  OD: small nerve with 360 pale peripapillary ring
  OS: normal optic nerve with pink and health rim tissue

➢ Physical
  ▪ Patient appeared alert and oriented (as appropriate for age)

❖ Differential Diagnosis
  ➢ Primary/leading
  ▪ Unilateral optic nerve hypoplasia
  ➢ Others
  ▪ Optic nerve atrophy
    • Genetic
    • Tumor
  ▪ Optic nerve aplasia
  ▪ Optic pit
  ▪ Optic disc coloboma

❖ Diagnosis and Discussion
  ➢ Our patient was diagnosed with unilateral optic nerve hypoplasia OD, and a
    myopic refractive error OS. Diagnosis was based on the following:
    ▪ Ophthalmoscopy findings of a small optic nerve with a double ring sign.
    ▪ History of stable longstanding decrease in visual acuity in the right eye.
    ▪ Retinoscopy and manifest refraction results.

  ➢ Elaborate on the condition
    Optic nerve hypoplasia (ONH) is a congenital anomaly with a reported
    prevalence of 6.3 out of 100,000 children. The presence of ONH can be noted
    clinically as an abnormally small and often pale nerve, and histologically as a
    decrease in number of optic nerve axons. The maldeveloped retinal nerve
    fiber layer results in varying affects on visual acuity, from 20/20 to the
    absence of light perception. ONH can present as an isolated anomaly,
    however it is often associated with significant central nervous system and
    endocrine abnormalities such as septo-optic dysplasia, hydranencephaly, and
    other growth hormone deficiencies. The exact etiology is unknown and
    thought to be multifactorial, given the large array of associated findings. Most
    authors agree that the resulting clinical picture of ONH is dependent on the
    location and timing of the gestational CNS injury.
Optic nerve hypoplasia most commonly occurs bilaterally, and often asymmetric but may occur as a unilateral case. The clinical presentation of unilateral ONH, can include a relative afferent pupillary defect, strabismus, amblyopia, and poor fixation with the affected eye. Due to the large morphological spectrum of ONH, diagnosis of mild unilateral cases can be challenging. A list of funduscopic criteria has been postulated to aid in the clinical diagnosis. More specifically, the list includes detecting a small nerve, a peripapillary halo surrounding the disc (double ring sign), the size of normal disc if the halo is included in approximation, the presence of mild vessel tortuosity, a dim foveal reflex, and an abnormally thin retinal nerve fiber layer.

Our patient had a longstanding history of unilateral ONH and presented with the classic findings of a grossly small optic nerve, and the double ring sign. However, no additional CNS or systemic conditions were noted on exam or through patient history. Due to the age at presentation, patient history and previous ophthalmic care no additional testing was performed at our visit.

- **Expound on Unique features**
  While performing binocular indirect ophthalmoscopy (BIO) with a 20D lens, the hypoplastic nerve appeared relatively normal in size secondary to the presence of the double ring surrounding the disc. Upon evaluation under higher magnification with a 90D lens, the appearance of the peripapillary halo surrounding the disc and the mild pallor of the nerve became more evident. Our patient exhibited a high level of cooperation on exam, which allowed for further analysis of the disc morphology. However, in younger or less cooperative patients the subtle distinctions may be more readily evaluated with a 14D lens during BIO.

- **Treatment and Management**
  - Optic nerve hypoplasia is a self-limited condition often requiring no treatment. However, bilateral ONH is often associated with systemic anomalies that may require further evaluation. From an ocular standpoint, children within the critical period of visual development should be considered for the treatment of any superimposed amblyogenic factors, including unilateral ONH, and asymmetric bilateral ONH. Our recommendations included prescribing polycarbonate lenses for full time wear that included the myopic refractive error in the left eye. Additionally, extensive patient education regarding ocular protection was given to both the patient and parent.
Protective eyewear is a necessary prophylactic treatment for patients whom suffer from severely decreased vision in one eye. Unfortunately, this patient population has also demonstrated a higher risk of vision loss when compared to the general population. According to Prevent Blindness America (PBA), 2.4 million eye injuries occur annually in the United States and approximately 35% of these injuries affect children under the age of 17. The PBA also implies that most of these ocular injuries were preventable. Thus, our recommendations for safety eyewear in the optometric setting can further aid in the prevention of this common occurrence.

Compliance with safety eyewear can be of particular concern for patients with minimal or no refractive error. Nevertheless, prophylactic treatment should still be advised in these patients. The compliance of monocular patients with safety eyewear was reviewed in a study of 33 children. Results indicated good compliance, and 45% reported protection of the sound eye during a potentially serious incident.

Due to the nature of full time wear, monocular children must wear their protective eye wear during all activities, including sports. Annually over 40,000 ocular related injuries are related to sports activities. In attempt to prevent this occurrence the American academy of Ophthalmology and American Academy of Pediatrics strongly recommend the use of protective eyewear during any sport activity that may result in eye injury. Further, the American Society for Testing and materials (ASTM) has established criteria for protective eyewear in most organized sports. These standards apply to all children participating in organized sports activities. However, an additional subset of criteria has been set for the functionally one eyed athlete; who is defined as having 20/40 best corrected visual acuity in the poorer seeing eye. Our consultations should therefore include recommendations to all patients in sports related activities as well as relaying any additional precautions to the functionally one-eyed athlete.

Contact lenses are not strictly contraindicated in patients with reduced visual acuity in one eye. However, patient education regarding associated risk factors with contact lens use is warranted. The risk for development of microbial keratitis has been strongly associated with the use of contact lenses. Microbial keratitis, although occurs rarely can have devastating visual consequences. Conversely, several individuals use contact lenses safely as an alternative to spectacle lenses. If contact lenses are pursued, although highly discouraged, the patient management should include a discussion on lens hygiene, replacement schedule and demonstration of proper insertion and removal techniques.
 ¶ Conclusion:
   ➢ Clinical Pearls
     ▪ Patient education on protective eyewear is essential in the prophylactic treatment of patients with a moderate to severe unilateral decreased in vision.
     ▪ As a monocular precaution protective eyewear is still required if the patient has minimal or no refractive error.
     ▪ Discourage any activity that may increase the risk of visual loss in the sound eye, including contact lens use.

 ¶ Bibliography