Article ▸ Bringing Unilateral High Myopia into Focus

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ABSTRACT

Background: Myopia is a relatively common refractive error that typically occurs bilaterally. In rare cases, myopia may present unilaterally, with one eye having minimal refractive error and the other eye being highly myopic. In some patients, there is no identifiable cause for this unique form of anisometropia, but commonly there is an associated ocular or systemic pathology. The visual acuity in the more myopic eye typically ranges from 20/20 to 20/200, with refractive amblyopia frequently being present. The increased myopia also puts these patients at a higher risk for retinal complications.

Case Series: This article reviews potential underlying causes for unilateral high myopia, clinical findings, beneficial testing, and proper treatment and management. Several case examples of unilateral high myopia are presented. Age, medical/ocular history, examination findings, refractive error, unaided and best-corrected visual acuities, treatment/management, and potential underlying etiologies are discussed.

Conclusion: The visual prognosis can vary greatly, but current research has shown that improvement in the visual acuity can often be made with full correction of the refractive error alongside amblyopia treatment. Viable refractive correction options include spectacle correction, contact lens wear, orthokeratology, corneal refractive surgery, and intraocular lens implantation. These patients also require annual dilated fundus exams due to the higher risk of retinal complications associated with high myopia.

Keywords: anisometropia, myopia, myopia progression, refractive amblyopia

Background

Myopia is a common visual condition affecting approximately 22.9% of the world population. The prevalence is expected to increase to roughly 49.8% by 2050.[1] High myopia, defined as a spherical equivalent greater than -5.00 diopters (D), is far less common, affecting only 2.7% of the world population.[1] An even more rare optical phenomenon is high unilateral myopia, where one eye is relatively emmetropic and the fellow eye has a large amount of myopic refractive error. Two different studies reported the occurrence as roughly 1.5% in Caucasian eyes and 4% in Taiwanese eyes.[2,3]

Despite its rare nature, there are common findings seen in the majority of patients with anisomyopia. Weiss reported that 94% of the children with unilateral high myopia studied had axial elongation of the more myopic eye.[4] Amplitude scans (A-scans) have shown that there is minimal difference in corneal thickness, anterior chamber thickness, and crystalline lens thickness between the more and less myopic eyes. The main contributor to the extended axial length in the more myopic eye is increased vitreous chamber depth (Figure 1).[5] This suggests that anisomyopia may be due to a difference in the rate or degree of posterior chamber growth. Over time, axial elongation of the more myopic eye can cause stretching and thinning of the choroid and retina (Figure 2). This can lead to complications such as peripheral retinal degenerations or tears, retinal detachment, lacquer cracks, choroidal neovascularization (CNV), chorioretinal or retinal pigment epithelial atrophy, posterior staphyloma, or macular hemorrhage.[6] A study by Lai et al. based out of Hong Kong found that 56.1% of adult subjects with greater than 6.00 D of myopia had one or more peripheral retinal lesions, and 11.3% had one or more posterior pole lesions.[7] This demonstrates that patients with unilateral high myopia should receive annual dilated fundus exams (DFEs) in order to assess for these potential ocular complications. Beneficial examination

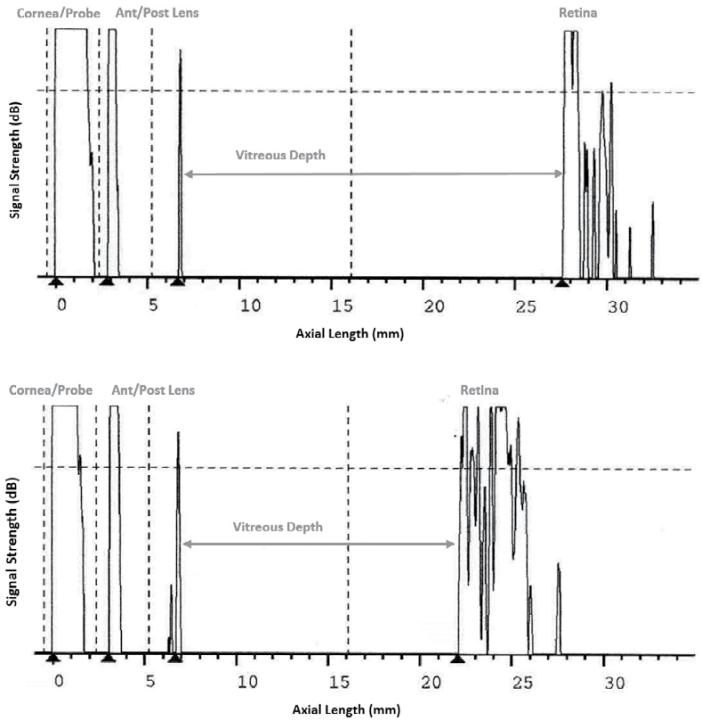

Figure 1. *A-scan measurements of the right eye (top image) and the left eye (bottom image) in Patient #8. The refractive error of this patient was -18.00 OD and +1.00-2.00x005 OS, and axial measurements were 27.76 mm OD and 21.88 mm OS. Note the increased vitreous chamber depth OD.*

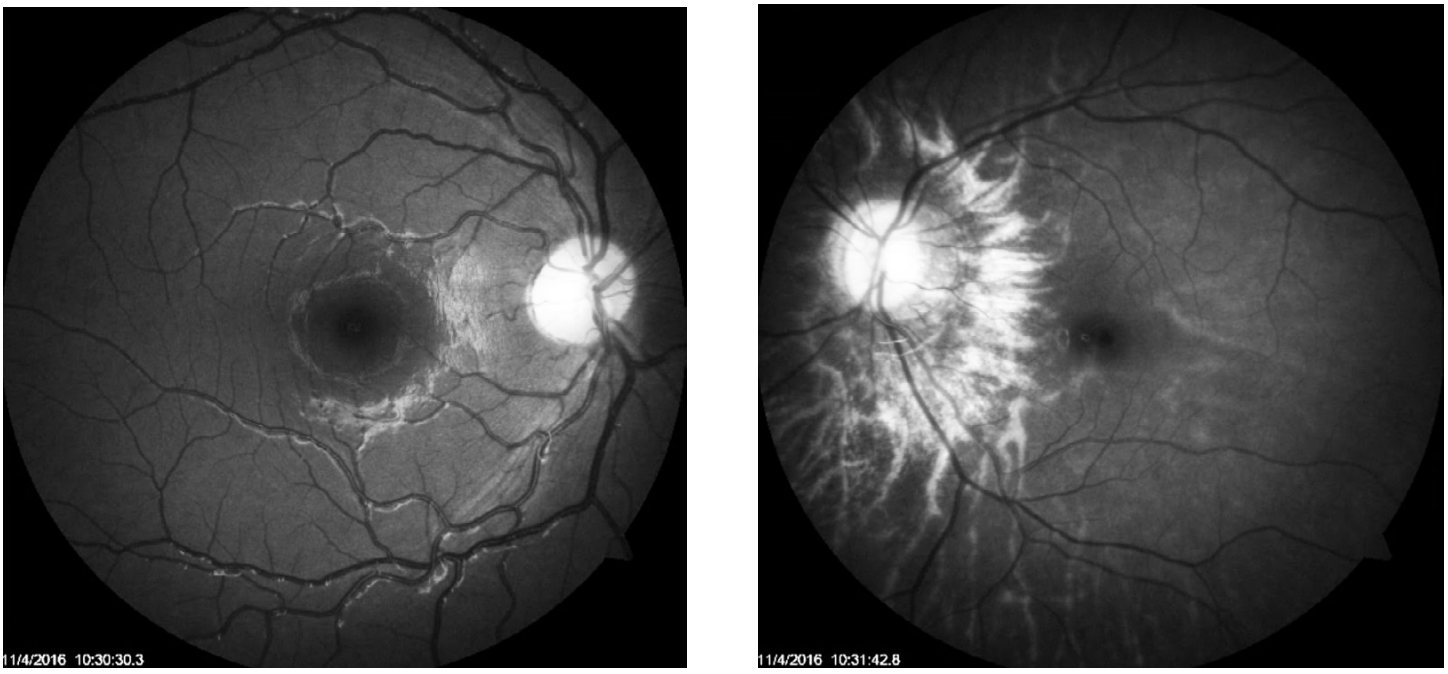

Figure 2. *Fundus photographs of the right eye (left image) and the left eye (right image) in Patient #1. The refractive error of this patient was +0.50 OD and -13.00 OS, and axial measurements were 22.69 mm OD and 28.01 mm OS. Note the highly myopic apperance to the retina OS and the nearly absent neuroretinal rim temporally OS. This patient was diagnosed with segmental optic nerve hypoplasia OS.*

testing include A-scan measurements, color vision testing, pupil testing, visual field testing, visual evoked potential (VEP) testing, optical coherence tomography (OCT) testing, and a thorough evaluation of the macula, optic nerve, and retina.

Visual acuity in the more myopic eye typically ranges from 20/20 to 20/200.[4] Strabismus is a relatively common finding among patients with anisomyopia.[8] A study by Tanaka et al. found that approximately 56% of the subjects with unilateral high myopia had strabismus.[9] Exotropia was found to be the most common type of strabismus, followed by vertical heterotropia and esotropia.

Underlying Causes

There is currently conflicting research as to what can cause this unique form of anisometropia. How is it that two eyes with presumably the same genetic and environmental influences develop such asymmetric refractive errors? As more research on the matter is published, primarily from Asian countries where myopia prevalence is highest, it appears that the cause may be multifactorial.[5] Some studies suggest that biomechanical differences between the ocular structures of the two eyes may lead to anisomyopia, such as differences in corneal hysteresis, ciliary body thickness, and intraocular pressure (IOP).[10,11] Other studies have suggested that optical properties, such as hyperopic defocus, may lead to the development of unilateral high myopia.[12] Several case reports involving twins have suggested that there may be a genetic component to the etiology.[13,14] It is likely that the actual cause of unilateral high myopia involves a combination of different factors.

In some cases, the anisomyopia is due to an ocular or systemic pathology. Weiss reviewed the medical records of 48 children with unilateral high myopia and found that 45 of the 48 children had an ocular disorder associated with reduced acuity, a central nervous system abnormality, or a family history of high myopia.[4] Only 3 of his subjects had no apparent ocular or systemic cause for the anisometropia. The top five causes for unilateral high myopia in his study were: 1) optic nerve disorders including optic nerve hypoplasia (complete or segmental), myelinated nerve fibers, atrophy, and coloboma; 2) central nervous system abnormalities including congenital hydrocephalus and meningitis; 3) lens abnormalities including lens subluxation; 4) retinopathy of prematurity; and 5) family history of high myopia. This study emphasizes the importance of ruling out a pathological cause for unilateral high myopia and the importance of reviewing the patient's perinatal and family history.

Below is a listing of various ocular and systemic pathologies that can cause unilateral high myopia:

- Optic nerve hypoplasia (ONH) – a congenital condition in which the optic disc is underdeveloped with a decrease in the number of axons. The entire optic disc may be small, in the case of diffuse ONH, or only a portion of the optic disc may be affected, as seen in segmental ONH. Common characteristics include retinal vessels that are either tortuous or abnormally straight, visual field constriction, and an APD.[15,16]
- Myelinated nerve fiber – a condition that results in myelination of the retinal nerve fiber layer. It is typically peripapillary and appears as white, striated, feathery patches. It is usually benign but may cause axial myopia, amblyopia, or strabismus.[17]
- Optic nerve coloboma – a congenital anomaly of the optic disc that is characterized by an excavation of the inferior portion of the nerve. It is frequently associated

with colobomas of the iris and/or ciliary body as well as a high refractive error.[18]

- Retinopathy of prematurity – a disease that can occur in prematurely born infants who required oxygen therapy. It causes abnormal blood vessel growth that can eventually lead to scarring and blindness if left untreated.
- Congenital hydrocephalus – a buildup of cerebrospinal fluid (CSF) in the brain that is present at birth. It can lead to permanent brain damage as well as strabismus and high refractive errors.

In order to identify the underlying cause for the unilateral high myopia, it may be necessary to perform ancillary testing. This can include A-scan measurements; color vision, pupil, visual field, VEP, and OCT testing; and a thorough evaluation of the optic nerve and retina. A-scans measure the axial length of each eye; typically the more myopic eye will have an increased axial length.[4] Color vision, pupil, visual field, and VEP testing will allow a better assessment of the visual pathway. Disorders of the optic nerve or central nervous system may result in abnormal findings on these tests. OCT testing of the optic nerve can also aid in identifying optic nerve abnormalities, such as optic nerve hypoplasia. Once the etiology of the unilateral high myopia is identified, it may be necessary to refer the patient for additional assessment. Optic nerve hypoplasia, for example, can frequently be associated with endocrine or cranial abnormalities and may require neurological or endocrinological work-ups.[15]

Visual Management

It was previously believed that patients with unilateral high myopia had poor visual prognosis, and often, the more myopic eye was undercorrected to allow the patient to be more balanced cosmetically and optically.[19,20] More recent studies show that patients with refractive amblyopia due to unilateral high myopia can have significant improvement in their visual acuity. Pang et al. performed a study in which all of the subjects with amblyopia associated with myopic anisometropia received 16 weeks of full-time spectacle and/or contact lens wear followed by 16 weeks of part-time patching with near activities. They found that 88% of the subjects had 2 or more lines of visual improvement, and that the visual acuity improved more when patching plus near activities was added.[20] Viable amblyopia treatment options for patients with unilateral high myopia include part-time patching of the better-seeing eye, atropine penalization, and vision therapy.[21] These treatments should be added alongside full-time correction of the refractive error.

Identifying the proper refractive correction will depend on the amount of anisometropia present, as well as the patient's compliance. Most people with greater than 3.00 D of anisometropia cannot tolerate the aniseikonia, and possible double vision, that is induced by spectacle correction.[22] For these patients, contact lens correction, corneal refractive surgery, or intraocular lens implantation may be better options. Several studies have investigated each of these treatment options for patients with unilateral high myopia, and all have had good visual outcomes when additional amblyopia therapy was added.[22-24] These corrective treatments reduce the cosmetic concern that can arise from wearing a highly myopic spectacle correction and also reduce the aniseikonia. If amblyopia is present, though, it is still recommended that these patients wear safety frames/lenses.

When the unilateral myopia is due to an ocular pathology, such as optic nerve hypoplasia, the visual prognosis may be more guarded. Many of these conditions can cause permanent vision loss, but a component of the reduced vision may also be from amblyopia associated with the high refractive error.[25] Amblyopia treatment should still be recommended to patients with an ocular pathology and a high refractive error.

Aside from attempting to reduce the refractive amblyopia, another method for improving the visual outcome for patients with unilateral high myopia is slowing down or stopping the myopia progression. There are currently numerous studies looking into the process of myopia progression and how it can be better controlled.[26-30] Promising treatment options for these patients include corneal reshaping with orthokeratology (Ortho-k) lenses and multifocal contact lenses. The design of both Ortho-K and multifocal contact lenses induces peripheral myopic defocus.[29,30] This allows peripheral light rays to focus anterior to the retina and act as a stimulus to slow down or to stop axial elongation, and therefore to reduce myopia progression. Both of these lens options can be beneficial for patients with unilateral high myopia in order to prevent the anisometropia from becoming even greater.

Case Examples

Table 1 presents 8 different patients with unilateral high myopia that were seen at The Eye Center at Southern College of Optometry. They have an age range of 6 to 20 years old, and none of the patients are related. Their ages, medical/ocular histories, examination findings, refractive errors, unaided and best-corrected visual acuities, treatment/management plans, and potential underlying etiologies are listed.

As seen in Table 1, strabismus is a common finding among patients with unilateral high myopia, especially exotropia. Five of the eight patients had strabismus, and the majority of these patients showed suppression of the more myopic eye on stereo testing. Four of the eight patients had no apparent origin for their anisomyopia, but the remaining four had systemic and/or ocular abnormalities that could be causative. These included segmental optic nerve hypoplasia, premature birth, developmental delay, and optic nerve atrophy following a viral infection. On dilated fundus exam, many of these patients had a tigroid appearance to the fundus with myopic tilting of the nerve and an associated choroidal and/or scleral crescent (Figure 2). None of the patients had peripheral or posterior pole lesions. Full-time spectacle and/or contact lens correction

Table 1. History, Examination Findings, and Treatment/Management of Patients with Unilateral High Myopia

Pat #	Age	Medical/ocular history	Unaided acuity	Exam findings	Refractive error	BCVA	Management	Anisomyopia etiology
1	10	• Born with "7% of the vision in her left eye" • Born full-term • Met milestones appropriately	OD: 20/15 OS: CF @ 3ft	• (+)APD • Constricted VF • ILXT • OS suppression • Axial length = 22.69 mm OD, 28.01 mm OS • Segmental optic nerve hypoplasia OS	OD: +0.50 OS: -13.00	OD: 20/20 OS: 20/125 •Achieved with CL OS	• Contact lens (CL) OS • Vision therapy(not yet initiated)	Segmental optic nerve hypoplasia OS (Figure 2)
2	7	• Born at 30 weeks and required oxygen • Met milestones appropriately	OD: 20/500 OS: 20/20	• IRXT • OD suppression	OD: -8.00 OS: -0.25	OD: 20/250 OS: 20/20 • Achieved with spectacles	• Full-time spectacle correction • Attempted patching with poor patient compliance • Consider CL OD in the future	Possibly due to premature birth
3	7	• Developmental delay in speech (started talking at age 4) otherwise unremarkable • Born full-term	OD: 20/20 OS: 20/800	• IAXT	OD: -0.25 OS: -5.00	OD: 20/20 20/30 OS: • Achieved with spectacles	• Full-time spectacle correction and vision therapy • Vision therapy (not yet initiated)	Possibly due to developmental delay
4	20	• Born full-term • Hospitalized for 3 months after birth because of a viral infection • Intellectual disability • Seizures throughout childhood	OD: 20/40 OS: CF @ 1ft	• ILET and LHyperT • OS suppression • Optic nerve pallor and atrophy OS	OD: +2.00-1.25x045 OS: -7.25	OD: 20/20 OS: 20/500 • Achieved with spectacles	• Full-time spectacle correction	Optic nerve atrophy, likely due to the viral infection
5	10	• Unremarkable • Born full-term • Met milestones appropriately	OD: 20/350 OS: 20/20	• Orthophoria • OD suppression	OD: -6.50-2.50x180 OS: Pl-1.00x180	OD: 20/60 OS: 20/20 • Achieved with spectacles	• Full-time spectacle correction and vision therapy • Consider CL OD in the future • Vision therapy (not yet initiated)	No apparent cause
6	6	• Unremarkable • Born full-term • Met milestones appropriately	OD: 20/200 OS: 20/20	• IAXT	OD: -5.00-2.00x180 OS: Plano	Before VT: OD: 20/125 OS: 20/20 After VT OD: 20/40 OS: 20/20 w/spectacles)	• Full-time spectacle correction and vision therapy • Completed 27 sessions of VT	No apparent cause
7	10	• Unremarkable • Born full-term • Met milestones appropriately	OD: 20/20 OS: 20/600	• Orthophoria • OS suppression	OD: Plano OS: -8.50	OD: 20/20 OS: 20/50 • Achieved with spectacles	• Full-time spectacle correction and vision therapy • Consider CL OS in the future • Vision therapy (not yet initiated)	No apparent cause
8	9	• Unremarkable • Born full-term with C-section • Met milestones appropriately	OD: 20/800 OS: 20/40	• Orthophoria • OD suppression • Axial length = 27.76 mm OD, 21.88 mm OS (Figure 1)	OD: -18.00 OS: +1.00-2.00x180 Patient reported diplopia through this and could only tolerate: OD: -5.00 OS: +1.00-2.00x180	Before VT: OD: 20/200 OS: 20/20 After VT: OD: 20/125 OS: 20/20 • Achieved with spectacles	• Full-time spectacle correction and vision therapy • Attempted contact lens wear with poor patient cooperation • Completed 35 sessions of VT	No apparent cause

alongside vision therapy was the treatment of choice for the majority of these patients in order to improve both the acuity and the strabismus. Patients #6 and #8 completed vision therapy and showed several lines of visual acuity improvement, with patient #6 reaching a BCVA of 20/40. Patient #6 also gained binocularity and no longer suppressed his myopic eye on stereo testing.

Conclusion

Unilateral high myopia may have no apparent etiology but can also be due to an underlying ocular or systemic pathology. A thorough evaluation of the patient's ocular and medical history is necessary in order to identify potential causes. Beneficial examination testing includes A-scan measurement; color vision, pupil, visual field, OCT, and VEP testing; and a thorough evaluation of the macula, optic nerve, and retina. These patients require annual DFEs due to the high risk of retinal complications associated with high myopia. Regardless of the underlying cause for the unilateral high myopia, amblyopia treatment options should always be presented to these patients. Although an ocular or systemic pathology can cause a permanent reduction in vision, a portion of the decreased visual acuity may be due to refractive amblyopia. As research continues to identify the many causes of unilateral high myopia, additional knowledge may be gained regarding how to slow down or stop the unilateral myopic progression.

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