

Case Report

Severe Anisometropic Myopia in Identical Twins

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ABSTRACT

High anisometropic myopia is a rare condition in twins. Genetic factors have been implicated in its development and there may be an association with vision-threatening complications. A pair of 11-year-old twins presented with poor distance vision in both eyes. Detailed ocular examination was performed including slit lamp examination, dilated funduscopy, cycloplegic refraction, keratometry and axial length measurement. The objective refraction was -6.50DS -2.00DC × 180 (right eye), -1.00DS (left eye) for the first twin; -13.75DS -2.25DC × 180 (right eye), -0.50DS -0.75DC × 04 (left eye) for the second twin. This case suggests an underlying genetic defect in the development of myopia.

Key words: Anisometropia, Myopia, Twins

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INTRODUCTION

Myopia has a multifactorial origin with interaction between genetic and environmental factors.¹ Genetic factors play a major role in the development of myopia. For example, studies of twins indicate that genetic factors may account for up to 80% of the variance in myopia irrespective of the age of onset.² The prevalence of myopia has been steadily increasing over the last few decades^{3,4} with wide racial differences.^{4,5} A study from England, reported the prevalence of myopia in 10-11 year-old children as 25.2%, 10.0%, and 3.4% in South Asians, African-Caribbeans and white Europeans, respectively.⁵ In developing countries in Asia, high myopia accounted for 10-20% of the over 80% of high school dropouts who were myopic.⁶ Fitzgerald *et al.*⁷ evaluated 145 children with high myopia and found bilateral high myopia to be more common with a prevalence of 64%. Severe myopia is a significant public health problem because it may be associated with myopic macular degeneration, cataract, glaucoma, peripheral retinal changes (such as lattice degeneration), retinal tears and holes as well as retinal detachment.⁸

Anisometropia, by definition, exists when there is a difference in refractive status between an individual's eyes.⁹ Myopic anisometropia is rare in twins. In this case report, I present the presence of severe anisometropic myopia in a pair of 11-year-old female identical twins.

CASE REPORT

A pair of 11-year-old identical twin sisters were seen in the eye clinic in April, 2012 with poor distant vision of one year duration. They had no associated complaints such as deviation of the eyes and there was no history of prior spectacle use. Their past medical histories were unremarkable. They were delivered at full term and there was no history suggestive of myopia in the family. The first twin had a distance visual acuity (VA) of counting fingers at two meters (0.03) in the right eye and near vision of N14 (0.4) and the left eye had a distance VA of 6/18 (0.3) and near vision of N4.5 (1.0). Cycloplegic (cyclopentolate 0.5%) refraction revealed an error of -6.50DS -2.00DC × 180 in the right eye and -1.00DS in the left eye. Best corrected VA was 6/12 (0.5) and N5 (0.9) with -6.00DS -2.00DC × 180 for the right eye and 6/6 (1.0) and N4.5 (1.0) with -1.00DS for the left eye. The difference in the spherical equivalent between the two eyes was - 6.5 diopters (D). On ocular examination there was no abnormality in the anterior segment of either eye. Funduscopy examination revealed a pink disc with peripapillary chorioretinal atrophy in the right eye and a normal fundus in the left eye. The patient was prescribed spectacles with full correction for the left eye and slight under-correction in the right eye.

The second twin had a distance VA of counting fingers at one meter (0.02) in the right eye and a near VA of N36 (0.1) while the

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left eye had a distance VA of 6/18 (0.3) and near vision of N4.5 (1.0). Cycloplegic refraction revealed an error of - 13.75DS - 2.25DC \times 180 in the right eye and - 0.50DS -0.75DC \times 04 in the left eye. There was no improvement in the VA with refraction in the right eye while the best corrected VA was 6/6 (1.0) and N4.5 (1.0) with - 0.50DS -0.50DC \times 030 in the left eye. The difference in the spherical equivalent between the two eyes was -14D. On ocular examination, the anterior segments in both eyes were normal. Funduscopy revealed a hypoplastic disc with peripapillary chorioretinal atrophy in the right eye while the left fundus was normal. The patient was prescribed spectacles with full correction for the left eye and balanced for the right eye. There was no strabismus, and systemic examination did not reveal features of any genetic defect suggestive of syndromic high myopia. Genetic studies could not be performed due to the lack of the necessary facilities.

The refraction, radius of anterior corneal curvature and corneal refractive power were measured using the autorefractometer (Accuref-K 9001; Ajinomoto Trading, Inc., Tokyo, Japan). Axial length was measured with an A-scan ultrasonographer (Storz Alpha II; Bausch and Lomb Storz® Instruments, Rochester, NY, USA). The results of these measurements are presented in Table 1.

DISCUSSION

The occurrence of anisometric myopia in identical twins with degenerative myopia in the same eye and simple myopia in the other suggests an underlying genetic defect in this condition. Genetic factors have been shown to have a major role to play in the development of myopia. Twin studies in European-derived populations have reported high heritability for myopia, in the 90% range, with similar figures for the various biometric factors that control refractive error, such as axial length, anterior chamber depth and corneal curvature.¹⁰ Recently, a genome-wide association study identified a susceptibility locus for myopia on chromosome 15q14.¹¹ The genetic basis for the dissimilar growth between eyes in cases of unilateral high myopia has been attributed to the skewed lyonization in females heterozygous for X-linked myopia.¹² Skewed X-chromosome inactivation in the eye has been found in monozygotic twins and eye pairs of individuals discordant for color vision deficiency.¹³ The difference in the right eyes of the sisters in this report may be due to variable expression of the underlying genetic defect.

Table 1: Biometry and refraction results

	Twin 1		Twin 2	
	Right eye	Left eye	Right eye	Left eye
Axial length (mm)	28.02	26.05	30.39	25.06
Keratometry (D)	41.60	41.12	41.37	40.42
Corneal curvature (mm)	8.12	8.20	8.16	8.36
Spherical equivalent (D)	-7.5	-1.00	-14.87	-0.87

Myopia can occur as an isolated finding or as a part of specific genetic syndromes.¹⁴ These syndromes include the syndromic high myopias, in which high myopia is associated with other symptoms that define the disease, such as connective tissue disorders (e.g., Marfan and Stickler syndromes), and complete and incomplete congenital stationary night blindness.³ Unlike these syndromes, however, the vast majority of individuals with moderate or severe myopia, have no associated defects.¹⁴ The absence of associated defects in these sisters suggests a non-syndromic high myopia.

Myopic anisometropia is a rare type of anisometropia.⁹ If it occurs with intact near vision it seldom results in amblyopia. When ordinary vision is however defocused due to high myopia, refractive amblyopia may occur.⁹ Mild myopic anisometropia (less than -3D) usually does not cause amblyopia but unilateral high myopia (-6D or more) often results in severe amblyopic visual loss.¹⁵ Unilateral high myopia with a difference of over -6D between the two eyes of the sisters in this report would account for the occurrence of amblyopia in the right eyes.

Severe myopia may be associated with posterior segment pathology. There was chorioretinal atrophy in the right eyes of both sisters with optic disc hypoplasia in the twin with the greater axial length. This could also have been a contributory factor in the development of amblyopia. Weiss¹² evaluated 48 children with anisometric myopia and reported a high prevalence of unilateral optic nerve hypoplasia with associated visual deficits.

Amblyopia due to anisometropia is difficult to detect and is essentially asymptomatic unless accompanied by a squint.^{16,17} Diagnosis is therefore usually late¹⁶ as in the current case report. The standard treatment for anisometric amblyopia is correction of any refractive error with occlusion therapy.¹⁶ Refractive correction is performed first using spectacles and/or contact lenses with subsequent patching and near activities once visual acuity plateaus.¹⁸ High anisometric amblyopia is however challenging to manage^{16,18} and has been reported to be the least responsive subtype of anisometric amblyopia to treatment.¹⁹ As a result, many clinicians would undercorrect the more myopic eye of these patients in order to allow the lenses to be more balanced optically and cosmetically, and do not pursue further treatment.¹⁸

Contact lenses with occlusion therapy can be used safely in children with amblyopia due to high anisometropia.¹⁶ This would have been the preferred treatment option in this case but compliance and regular follow-up were identified as likely problems. Contact lenses may also be problematic.²⁰ Contacts can be difficult to insert and remove, are frequently lost, can be irritating to the eye or may be too costly.²¹ Both sisters were boarding students and adequate supervision to ensure compliance could not be guaranteed in the absence of their

parents. Compliance with amblyopia treatment has been found to be the most critical factor for predicting a successful outcome.²²

Conventional treatment with glasses and occlusion therapy is usually unsuccessful but may be worth instituting as initial management.¹⁶ Refractive correction alone using spectacles was performed for both sisters with undercorrection of the more myopic eye. Full refractive correction was not possible as the difference in the spherical equivalents would result in aniseikonia.

Refractive surgery, although controversial, is another option in the management of pediatric refractive errors.²³ The main indications are bilateral high ametropia and anisometropia where conventional treatment with spectacles or contact lens is not tolerated.²⁴ The most commonly performed procedures currently are surface ablation procedures using excimer laser.²⁴ The major drawback of surface ablation procedures is refractive regression, which is more pronounced in higher degrees of ametropia.^{21,24} When corneal refractive surgery is not possible, such as in extremely high myopia, phakic intraocular lens may be an option.^{23,25} Long-term endothelial cell loss remains a source of concern with phakic intraocular lens.²⁴ The advantages of these lenses are reversibility, predictability, and lack of regression.²⁴ Refractive surgery however has a limited role to play in the management of refractive errors in our environment as the equipment required are very expensive and are not readily available.

Regular monitoring of refraction in these sisters is mandatory as the myopia may still progress with an increase in the axial length of the eyes. For children between the ages of 6 and 15 years with myopia, increased progression in younger children and in those with greater myopia at the baseline examination has been a consistent finding.²⁶

In conclusion, high anisometric myopia may result in severe amblyopia especially when there is an associated abnormality of the eye. This case report is highly suggestive of an underlying genetic defect in its development.

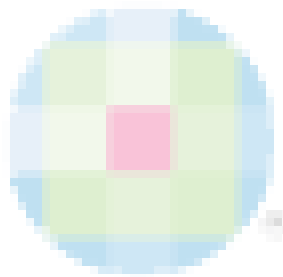
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