Anomalies of binocular function in patients with longstanding asymmetric keratoconus

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Abstract

Aims—To study binocular function in patients with longstanding asymmetric keratoconus.

Methods—In 20 adult patients with longstanding asymmetric keratoconus managed with a scleral contact lens a full clinical and orthoptic assessment was performed with and without the scleral contact lens in the poorer eye.

Results—All 20 patients had a corrected acuity of at least 6/9 in their better eye. With the scleral lens in situ the acuity of the poorer eye ranged from 6/6 to 6/60 and without the lens from 6/18 to hand movements. Patients were aged from 18 to 68 years and had worn a scleral contact lens for between 3 and 106 months. Without the contact lens in their poorer eye all patients had a small exotropia and all showed suppression, with the exception of one patient who had a right hypertropia with diplopia. With the scleral lens in situ 12 patients had an exophoria or esophoria, six a micro-exotropia, and two a manifest exotropia with suppression.

Conclusions—Binocular function breaks down in some adult patients with longstanding asymmetric keratoconus. This is probably caused by longstanding unilateral visual deprivation. There are similarities to the breakdown of binocular function seen in some patients with a longstanding dense unilateral adult onset cataract who can develop intractable diplopia following cataract surgery.

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This study was prompted by the clinical observation that some patients with longstanding asymmetric keratoconus chose to wear a scleral contact lens in their poorer eye, even though the corrected vision of this eye was still substantially worse than that of their better eye. This suggested that their binocular function was improved by wearing the lens. A group of such patients was therefore investigated using clinical tests of binocular function. These demonstrated striking abnormalities in their binocular function, which were reduced with the contact lens in situ. These changes are probably due to the longstanding, mainly unilateral, visual impairment resulting from their asymmetric keratoconus. There appear to be no previous reports of abnormalities of binocular function in patients with keratoconus.

These data were presented at the 25th Annual Conference of the European Strabismological Association, Jerusalem, September 1999.¹

Methods

From a database of approximately 350 patients with keratoconus being managed with a scleral contact lens, 35 were identified as having a corrected acuity with the scleral lens in their more affected eye which was two or more Snellen lines worse than their fellow eye. Twenty of these 35 patients were recruited for this study. Visual correction was not possible with conventional contact lenses in these patients with very marked keratoconus, as a stable fit could not be achieved. However, a stable fit had been achieved with a rigid gas permeable scleral lenses and these were well tolerated.² Because of the irregularity of the cornea in advanced keratoconus and because scleral lenses use the fluid lens between the posterior lens surface and the corneal surface to produce emmetropia, it is not possible to give meaningful refractive data for the patients. However, all were emmetropic with their lens in situ.

Snellen visual acuities were measured with and without the contact lens. Binocular function was assessed with and without the scleral lens in their poorer eye using the cover test, prism cover test, Bagolini glasses, and prism fusion range at 6 metres and ½ metre. Stereopsis was initially tested using the TNO test; if a patient failed to demonstrate any stereopsis with this method the Titmus test was used to look for the presence of gross stereopsis. The 4 dioptre base-out prism test was used to test for central suppression and aniseikonia was assessed using the Awaya Aniseikonia test and/or Rubens slides on the synoptophore. A microtropia was defined as a manifest squint of

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Table 1 Details of patients with keratoconus

Patient	Sex	Age (years)	Time from diagnosis (years)	Fixing eye	Fixing eye correction	Duration scleral lens wear in affected eye (months)
1	M	18	6	Right	Corneal CL	30
2	M	18	3	Left	Corneal CL	31
3	M	18	6	Left	Corneal CL	40
4	M	26	8	Right	Corneal CL	8
5	M	27	12	Left	Corneal CL	92
6	F	28	10	Right	Scleral CL+PK	27
7	M	29	10	Left	Scleral CL	40
8	F	30	8	Right	Corneal CL	7
9	M	32	18	Left	Scleral CL+PK	41
10	M	35	12	Right	Corneal CL	9
11	F	36	18	Left	Corneal CL	27
12	M	36	17	Right	Unaided	48
13	M	37	11	Right	Corneal CL	106
14	F	40	14	Right	Corneal CL	62
15	M	44	19	Right	Spectacle	30
16	F	47	32	Right	Corneal CL	60
17	F	51	28	Right	Corneal CL	90
18	F	64	34	Left	Spectacle	3
19	F	65	50	Left	Corneal CL	48
20	F	68	51	Left	Scleral CL	66

CL = contact lens; PK = penetrating keratoplasty.

Figure 1 Age at diagnosis of keratoconus, duration of disease, and duration of scleral contact lens wear for 20 patients.

10 prism dioptres or less, with evidence of anomalous binocular function.

Results

The 20 patients studied were from 18 to 68 years old. The diagnosis of keratoconus had been made between 3 and 51 years previously and the patients had worn a scleral lens for between 3 months and almost 9 years (Table 1, Fig 1). All patients had a corrected acuity of at least 6/9 in their better, fixing eye, in most cases by using a corneal contact lens (Tables 1 and 2). Acuities with the scleral lens in their poorer eye ranged from 6/6 to 6/60 and without the lens from 6/18 to hand movements (Table 2).

Without the scleral lens all 20 patients had a manifest squint with absent binocular function (Table 2). Nineteen patients showed suppression of their poorer eye when tested with Bagolini glasses and one had diplopia and a small manifest vertical deviation. All except one were

exotropic, although the angles were small and not cosmetically obvious. In most patients it was not possible accurately to measure the angle of the squint by cover test as fixation with the affected eye was poor. An abnormal angle kappa was recorded in 15 cases. The corneal reflections were either temporally displaced giving rise to a pseudo-esotropia, inferiorly displaced giving rise to a pseudohypertropia or a combination of both.

With the scleral lens in their poorer eye, 16 of the 20 patients showed evidence of binocular single vision with Bagolini glasses and 14 had measurable stereoacuity. In four patients this was only gross stereopsis and only one patient achieved a stereoacuity of 60 seconds of arc (Table 2). No patient overcame a 4 dioptre prism placed in front of the more affected eye, indicating the presence of a central suppression area. On cover test 12 patients had a phoria, six a microexotropia, and two a manifest exotropia (Table 2). There was an associated vertical deviation on distance testing in 12 of the cases and for near in 10 cases (Table 3). Sixteen patients had a measurable motor fusion range at 6 metres and 17 at $\frac{1}{3}$ metre (Table 3). Only one patient gave a history of any previous orthoptic treatment as a child and this patient had one of the best stereoacuities.

No patient reported significant aniseikonia when tested using their scleral lens; this could not be tested without the contact lens because of suppression.

Discussion

This study describes 20 patients with long-standing asymmetric keratoconus who all showed evidence of grossly abnormal binocular function by standard orthoptic tests. With a scleral contact lens in their poorer eye most achieved a degree of binocular single vision. Without the scleral lens no patient showed any evidence of binocular function and all except one suppressed the deviating eye. The patients thus gained a substantial functional advantage

Table 2 Visual acuity and binocular function with and without scleral lens in situ

Patient	Fixing eye acuity	Affected eye with scleral lens in situ				Affected eye without scleral lens		
		Acuity	Binocular function	Bagolini glasses	Stereopsis (seconds of arc)	Acuity	Binocular function	Bagolini glasses
1	6/6	6/24	Exophoria	Suppression	200 (w)	3/60	Exotropia + R/L	Suppression
2	6/9	6/36	Microexotropia	BSV	NIL	CF	Exotropia	Suppression
3	6/6	6/18	Microexotropia	BSV	NIL	1/60	Exotropia	Suppression
4	6/9	6/36	Exophoria	BSV	200 (w)	CF	Exotropia	Suppression
5	6/9	6/60	Microexotropia	BSV	Gross (w)	1/120	Exotropia	Suppression
6	6/6	6/24	Microexotropia	BSV	NIL	CF	Exotropia	Suppression
7	6/6	6/36	Microexotropia	BSV	480	1/120	Exotropia	Suppression
8	6/9	6/24	Exophoria	BSV	NIL	HM	R/L	Diplopia
9	6/5	6/18	Exophoria	BSV	120	1/120	Exotropia	Suppression
10	6/5	6/18	Exophoria	BSV	60	1/60	Exotropia	Suppression
11	6/6	6/18	Esophoria	BSV	240	CF	Exotropia	Suppression
12	6/4	6/18	Exotropia	Suppression	NIL	CF	Exotropia	Suppression
13	6/5	6/18	Esophoria	BSV	Gross (w)	1/60	Exotropia	Suppression
14	6/9	6/12	Exophoria	BSV	480	CF	Exotropia	Suppression
15	6/5	6/6	Exophoria	BSV	480	6/18	Exotropia	Suppression
16	6/6	6/36	Exotropia	Suppression	NIL	CF	Exotropia	Suppression
17	6/6	6/12	Exophoria	BSV	240	1/60	Exotropia	Suppression
18	6/9	6/24	Exophoria	BSV	200 (w)	CF	Exotropia	Suppression
19	6/6	6/24	Exophoria	Suppression	Gross (w)	1/60	Exotropia	Suppression
20	6/9	6/24	Microexotropia	BSV	Gross (w)	HM	Exotropia	Suppression

BSV = binocular single vision; (w) = Wirt stereotest, otherwise TNO; CF = counting fingers; HM = hand movements; R/L = right over left.

Table 3 Details of orthoptic findings

Patient	Prism cover test with scleral lens				Prism fusion range with scleral lens		4 Dioptre base-out prism	Previous ophthalmic history
	6 metres (prism dioptres)		½ metre (prism dioptres)		6 metres (prism dioptres)	½ metre (prism dioptres)		
1	1XT	1R/L	4XT	1R/L	10 PBO-4 PBI	40 PBO-4 PBI	Not overcome	Patched
2	4XT		16XT		4 PBO-6 PBI	16 PBO-14 PBI	Not overcome	None
3	12XT		8XT		NIL	NIL	Not overcome	None
4	2XT		8XT		12 PBO-2 PBI	25 PBO-8 PBI	Not overcome	None
5	16XT	2L/R	16XT	2L/R	6 PBO-2 PBI	6 PBO-2 PBI	Not overcome	None
6	1XT	1L/R	4XT	1L/R	NIL	18 PBO-8 PBI	Not overcome	PK
7	4XT	2R/L	10XT		6 PBO-1 PBI	6 PBO-1 PBI	Not overcome	None
8	4XT	2R/L	6XT		16 PBO-6 PBI	35 PBO-10 PBI	Not overcome	None
9	2XT	2L/R	8XT	2L/R	10 PBO-8 PBI	20 PBO-16 PBI	Not overcome	PK
10	1XT		4XT		16 PBO-8 PBI	35 PBO-10 PBI	Not overcome	None
11	2ET	1R/L	4XT	1R/L	20 PBO-2 PBI	45 PBO-16 PBI	Not overcome	None
12	6XT		16XT		NIL	NIL	Not overcome	None
13	1ET	2L/R	2XT	3L/R	16 PBO-4 PBI	30 PBO-14 PBI	Not overcome	None
14	1XT		1XT		20 PBO-8 PBI	40 PBO-16 PBI	Not overcome	None
15	0	4R/L	6XT	2R/L	16 PBO-6 PBI	30 PBO-14 PBI	Not overcome	None
16	6XT		4XT		NIL	NIL	Not overcome	None
17	2XT	1R/L	8XT	2R/L	6 PBO-6 PBI	25 PBO-14 PBI	Not overcome	None
18	20XT	6R/L	4XT	2R/L	20 PBO-10 PBI	20 PBO-12 PBI	Not overcome	None
19	0	1L/R	6XT	2L/R	12 PBO-4 PBI	30 PBO-10 PBI	Not overcome	None
20	1XT		10XT		2 PBO-4 PBI	4 PBO-4 PBI	Not overcome	None

XT = exodeviation; ET = esodeviation; PBO = prism base out; PBI = prism base in; PK = penetrating keratoplasty to fellow eye; R/L = right over left; L/R = left over right.

from wearing their scleral lens and this almost certainly explains their enthusiasm for doing so.

To our knowledge there are no previous reports of abnormal binocular function in patients with keratoconus. The patients in the present study all had longstanding keratoconus in which both the uncorrected and corrected acuity of one eye was substantially poorer than that of the other. They were recruited from a pool of about 350 patients with keratoconus managed with scleral contact lenses. All 20 patients subjectively found an advantage in wearing a scleral lens in their worse eye and were found to have anomalous binocular function on formal testing. The anomalies in these patients were not readily apparent without formal testing of binocular function because the squint angles were small and the corneal distortion found in keratoconus or the presence of a contact lens may give rise to abnormally aligned corneal reflexes. Most patients suppressed the deviating eye when manifest and so did not complain of diplopia. It is possible that anomalous binocular function is not uncommon in patients with longstanding keratoconus.

Uniocular optical blur alone reduces stereoacuity in normal subjects.^{3,4} These studies used induced anisometropia to cause blur, which would also produce aniseikonia. Because of the advanced keratoconus and the way in which scleral lenses produce their correction by using the fluid lens between the scleral lens and a very irregular cornea, it was not possible to obtain meaningful refractive data for the present patients. However, all were emmetropic with their scleral lenses in situ and were shown not to be aniseikonic. Nevertheless, for an equivalent reduction in acuity of the poorer eye, stereoacuity was more reduced in the keratoconic patients than in normal subjects with optically reduced acuity in one eye (Fig 2). Of the 14 normal subjects studied by Goodwin and Romano,⁴ none lost stereoacuity altogether until the acuity of the worse eye was reduced to 20/200, even though they also had

induced anisometropia. By comparison, six of the present 20 keratoconic patients had no stereopsis with their contact lens in situ at acuities of between 6/18 and 6/36 (20/60 to 20/120) (Table 2). A statistical comparison of the stereoacuities of the 18 keratoconic subjects who had acuities of between 6/12 and 6/36 with the values for the normal subjects with the nearest equivalent acuities of between 20/40 and 20/100 in Goodwin and Romano's study,⁴ show the keratoconic subjects to have a significantly worse stereoacuity (keratoconic median 3000 seconds of arc, normal 400 seconds of arc; $p < 0.02$, Mann-Whitney U test). The mean stereoacuities of the normal subjects studied by Levy and Glick³ were substantially better at each Snellen acuity than the median values of Goodwin and Romano and so the keratoconic subjects were even more abnormal compared to this group (Fig 2). Although such comparisons must be treated with considerable caution, it would appear that stereoacuity in the keratoconic subjects is substantially worse than can be accounted for by reduced acuity alone.

More striking is the observation that six of the 20 keratoconic patients had a microexotropia (with anomalous binocular function)

Figure 2 Comparing the median stereoacuity for Snellen acuities of the poorer eye between 6/12 and 6/60 (20/40 and 20/200) in 20 patients with longstanding asymmetric keratoconus with those for 14 normal subjects taken from table 1 of Goodwin and Romano⁴ and the mean stereoacuities for 10 normal subjects with a comparable acuity difference from Levy and Glick.³

and two a manifest exotropia (with suppression) with their contact lens in situ. Goodwin and Romano⁴ specifically looked for tropias during testing of their normal subjects and found none and none were noted by Levy and Glick.³ A further study which examined the relation between the degree of induced anisometropia and binocular function in normal subjects found only occasional minor abnormalities such as brief suppression with an induced difference of 2–3 dioptres,⁵ but nothing close to the degree of binocular disruption found in the present patients.

Although there is no direct evidence that the patients described here had normal binocular function prior to the development of their keratoconus, only one of the patients had a known history of squint, amblyopia or poor vision in the affected eye. Although microtropias may go unrecognised in childhood, they are relatively uncommon within the overall population with squints, and it seems unlikely that at least 20 of the 350 patients with keratoconus all had previously unrecognised microtropias. The abnormalities described here follow a consistent pattern, with central suppression of the deviating eye, reduction or loss of stereopsis with a microtropia when wearing their scleral lens, and a manifest exotropia with suppression when the poorer eye is uncorrected. It seems most likely that some at least of these changes are due to prolonged deprivation of form vision in the poorer eye from the asymmetric keratoconus.

It has generally been assumed that the sensitive period during which abnormal visual experience affects visual development in humans finishes at about 7 years of age and that the adult visual system is not affected by visual deprivation in animals or humans.^{6–10} The earliest age at which keratoconus was diagnosed in our series of patients was 12 years, and in most it was considerably later and well into adulthood (Fig 1). However, it has been shown that patients with longstanding, dense monocular cataracts of adult onset show a marked delay in visual evoked potentials recorded to stimulation of the deprived eye following cataract extraction.¹¹ In most cases this resolved over a period of months, but longstanding, monocular visual deprivation associated with adult onset cataracts can result in an irreversible loss of binocular function, with intractable diplopia following cataract surgery.^{12–14} Suppression is often considered to occur only in squints of childhood onset, but we have observed it in

patients with longstanding adult onset squints from cranial nerve palsies and thyroid eye disease and so its presence does not necessarily demonstrate the onset of a binocular anomaly in childhood. The above findings indicate that the adult visual system is susceptible to prolonged visual deprivation and the present patients would appear to be another instance of this. It will be of considerable interest to see if they have comparable abnormalities in their visual evoked responses to those found in the patients with longstanding unilateral cataracts.

It is probable that the patients described here developed their anomalous binocular function because of prolonged reduced acuity in one eye from their keratoconus. In most instances scleral lenses were introduced into their management at a fairly late stage in the disease. It is possible that earlier correction of their poorer eye would have preserved better binocular function. Even when anomalous binocular function has developed it is clear that even a limited improvement in acuity in the poorer eye from using a scleral lens can have a substantial impact on the overall visual function of a patient.

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