

Role of contact lenses in the management of congenital nystagmus

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SUMMARY Congenital idiopathic nystagmus is usually associated with poor vision which has generally proved resistant to treatment. This study reports the use of contact lenses in 8 patients, 5 of whom achieved an improvement in their visual acuity of 3 lines on the Snellen's chart.

Congenital idiopathic nystagmus is a disorder of eye movement characterised by involuntary oscillations of one or both eyes, usually in a horizontal direction but also on occasion vertically. It is present at or shortly after birth,^{1,2} and the definition is usually taken to exclude any known ophthalmic or neurological cause. The character of the nystagmus may be either pendular or jerk-type, and its incidence in the general population is low. Cogan³ considered that those patients with pendular nystagmus usually had an underlying ocular defect, while those with jerk nystagmus were in the true congenital idiopathic group. This subdivision has not found universal recognition, as either type of nystagmus may be seen in different members of the same family.^{2,4} Moreover, the type of nystagmus seen in any one patient may vary with the position of gaze.² Our experience accords with the view that congenital idiopathic nystagmus is variable in type and that diagnostic conclusions cannot be justified on the basis of the character of the nystagmus alone. The defect may be transmitted genetically, but in most patients there is no known family history.⁵ The nystagmus continues throughout life, but the amplitude may decrease in the adult.⁶ It may be manifest under binocular conditions or become apparent only on occlusion of one eye (latent nystagmus).

Patients with congenital nystagmus usually have diminished visual acuity, though rarely it is normal. The acuity is usually inversely proportional to the intensity of nystagmus—that is, the product of its amplitude and frequency.⁷ It has long been a topic for debate whether poor vision is primary or secondary to the nystagmus. Nettleship⁶ thought that there was

always an underlying primary ocular defect even if it was not discernible, but most workers now consider that in the absence of an ocular defect the diminished visual acuity is due to the constant movement of the target image on the retina. High-speed cinematographic studies have shown that the image of the fixation target falls on the fovea only at one or other peak of the oscillations.²

Because good vision is so important to the academic and social development of children and young adults, every possible step should be taken to ensure they achieve their full potential for vision. We believe that the place of contact lenses in the treatment of poor vision associated with congenital nystagmus deserves wider recognition, and we report here 8 patients, 7 of whom achieved a significant improvement in their visual acuity as a result of their use.

Patients

The patients in this study consisted of 4 males and 4 females, whose ages ranged from 10 years to 43 years at the time of contact lens fitting. Three patients (cases 3, 5, 8) had a minor degree of albinism or iris atrophy represented by iris translucency on retroillumination, but there were no other known ophthalmic or neurological abnormalities. Seven patients were seen over a 3-year period in the Contact Lens Clinic at the Norfolk and Norwich Hospital and one (case 8) was a pupil at the East Anglian School for the Partially Sighted and had been referred to the Contact Lens Department at Moorfields Eye Hospital, London. None of our patients had an abnormal head posture, though one (case 6) did have a small, cosmetically acceptable, convergent squint of 10° for both near and distance. These patients could there-

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fore be considered a selected group in whom surgical treatment was not appropriate and in whom no other form of treatment currently held any prospect of visual improvement.

All patients had an accurate refraction and keratometry before fitting with hard corneal contact lenses, and visual acuity was recorded with a standard Snellen test type.

Table 1 shows details of the patients, their best corrected vision wearing spectacles prior to contact lens fitting, and after at least 6 months' established contact lens wear.

The younger patients were initially fitted with daily-wear soft (hydrophilic) contact lenses, and then after between 3 and 6 months, when tolerance and confidence in handling lenses had been created, they were changed to hard gas-permeable corneal contact lenses for maximum visual benefit. The adult patients were fitted with hard gas-permeable corneal contact lenses from the outset.

Seven patients (13 eyes) showed an improvement in visual acuity of at least one line on the Snellen's chart, and 4 patients (5 eyes) were improved by 3 lines (Table 1). Of these 7 eyes, none had irregular astigmatism and only one had regular corneal astigmatism of greater than 1.50 DC. Thus the visual improvement could not be attributed solely to the optical advantages of contact lenses over spectacles.

Discussion

The precise underlying disorder causing congenital nystagmus is unknown, but it is generally assumed to be a defect in the control of the slow eye movement (pursuit) subsystem.⁸ There may be a defect in the normal damping of these movements analogous to the disorder of extrapyramidal damping of normal

voluntary limb movements in conditions such as Parkinson's disease. The amplitude of the oscillations can often be reduced or even abolished in certain positions of horizontal gaze or on extreme convergence. Thus the patient's near vision is often much better than distant vision, provided he is allowed to hold the book sufficiently close. Similarly, in order to achieve good distant vision the patient may adopt an abnormal head posture so as to turn the eyes into the position of minimal nystagmus.

The foregoing observations form the basis on which most attempts to treat congenital nystagmus are based. Prismotherapy aims by optical means to put the eyes into the favoured position of gaze, either laterally or into convergence, while maintaining the head in the normal straight-ahead position and the object at its normal distance. Surgical treatment, first advocated by Anderson⁹ and Kestenbaum¹⁰ and subsequently modified,¹¹ aims to achieve the same result surgically when there is a horizontal conjugate deviation in the neutral position.

More recently a completely new line of treatment has been suggested utilising a type of biofeedback.¹² By use of special spectacles Abadi and his coworkers have been able to convert information about the amplitude and frequency of nystagmus into an audible tone, the pitch of which varies with the nystagmus. They report an initial success in training adult patients to control the degree of their nystagmus in response to the feedback they get from these spectacles. The improvement achieved is so far limited, but they hope to improve this by training children to use the apparatus.

We have found that the use of contact lenses can produce significant improvement in the visual acuity of some patients suffering from idiopathic congenital nystagmus. Standard ophthalmological tests^{13 14}

Table 1 Visual results in patients with congenital idiopathic nystagmus treated with contact lenses

Patient	Age	Sex	Keratometry	Refraction	Corrected vision:	
					Spectacles	Contact lens
1	43	M	R 9.0:8.6 L 9.0:8.3	R +0.50/+1.50 DC L +0.50/+3.00 DC	6/24 6/24	6/12 6/12
2	24	F	R 7.8:7.7 L 7.7:7.7	R -7.00/-1.00 L -3.50/-0.50	6/24 6/60	6/9 6/36
3	11	F	R 8.0:7.40 L 7.90:7.50	R -7.0/-1.0 DC L -7.0/1.0 DC	6/36 6/36	6/24 6/24
4	10	M	R 7.7:7.9 L 7.7:8.0	R -8.0/-1.5 DC L -10.0/-3.0 DC	6/60 6/18	6/18 6/12
5	21	F	R 7.9:8.1 L 8.1:8.1	R -0.25/+0.5 DC L plano/-0.75 DC	6/18 6/36	6/12 6/12
6	22	F	R 7.4:7.8 L 7.3:7.7	R plano/+1.0 DC L plano/+1.5 DC	6/24 6/12	6/24 6/12
7	15	F	R 8.3:8.4 L 8.2:8.3	R -8.0/-1.0 DC L -8.0/-1.0 DC	6/36 6/36	6/24 6/18
8	20	M	R 7.70:7.15 L 7.75:7.20	R -1.0/-6.0 DC L -2.5/-6.0 DC	6/60 6/60	6/24 6/24

make no mention of this type of treatment, and we have been able to find only 2 reports of their use in patients with no other ocular abnormality. The report by Sedan¹⁵ included only 2 patients, one of whom was said to have had marked albinism, while Abadi¹⁶ in one case obtained a small improvement of vision from 6/18 to 6/12 in one eye when contact lenses were used. He also demonstrated by measuring the contrast sensitivity function that a better retinal image was obtained by contact lenses than by spectacles.

Although the improvement in vision in our patients would seem to be small, in most cases it means a significant improvement in their visual function. Current thinking about education of children is moving away from entirely segregated schooling for the visually handicapped child. An improvement of vision from 6/36 to 6/18, for example, may be sufficient to allow that child to remain in a normal school.

A similar improvement in a young adult may mean a similarly dramatic increase in career opportunities, or make the difference (as it did in case 2, a nurse) between being able to take a driving test or failing purely on the eyesight requirements. The benefits to the patient's morale and independence brought about by such an improvement is immense.

It is all too easy to dismiss a distance vision of 6/36 in an adolescent and say that, because he is well outside the critical first few years of life, no further improvement of vision can be expected. It is well known, however, that patients with congenital nystagmus usually have better near vision than for distance. This suggests that the cortical and subcortical neuronal basis of good visual acuity has been established in early childhood but is not being fully realised in distance vision. What then is the mechanism of the increase in visual acuity seen in our patients?

We have found that the visual improvement obtained is greater than could be expected solely from the correction of any corneal astigmatism in most of our patients. The one exception is case 8. Probably the most important additional factor in bringing about this improvement is the fact that the patient is looking along the visual axis of his correcting lens for a far greater proportion of the time

with contact lenses than with spectacles, since the contact lens moves with the eye. This particularly applies to those patients with a high refractive error.

Abadi¹⁶ showed that the wearing of contact lenses diminishes the amplitude and frequency of the nystagmus, and he suggested the mechanism for this reduction might be the additional vergence and accommodative effort generated by contact lenses. Alternatively the sensory feedback from the edge of the contact lens rubbing against the edges of the lids as the eyes oscillate may achieve the same effect as the rather more expensive and cumbersome equipment reported by Abadi *et al.*¹²

In conclusion, we believe that the use of contact lenses should be considered in all patients who have significant impairment of distance vision due to congenital nystagmus.

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