Pattern of amblyopia and fixation after keratoplasty

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SUMMARY Forty cases of clear corneal grafts in 2 groups of patients who developed corneal opacities either before or after 5 years of age were investigated for the pattern of amblyopia and fixation. A severe irreversible form of amblyopia with eccentric fixation was observed in cases of unilateral corneal opacity when the anomaly occurred before the age of 5 years and corneal grafting was delayed until the second or third decade of life. If the opacity was bilateral, although the onset was before the age of 5 years, the improvement in visual acuity after corneal grafting occurred in the relatively better eye with foveal fixation. Good visual results were seen after corneal grafting when the offending opacity occurred after the age of 5 years. Foveal fixation was also observed in most of these cases.

None of the patients who had a squint preoperatively lost their angle of strabismus. Four cases in group B, however, developed an alternating squint postoperatively.

After a successful corneal graft a patient may not regain vision owing to associated amblyopia. In India such a situation is encountered more frequently because corneal opacities due to keratomalacia, keratoconjunctivitis, and smallpox arise commonly in early childhood. Hence it is important to select cases for surgery to avoid disappointment and the waste of valuable donor material.

Data have been collected from the eye bank clinic at Irwin Hospital, where corneal grafting has been performed without consideration of the age of onset of corneal opacities. This provided an opportunity to study the density of amblyopia in patients who had clear media postoperatively.

Materials and methods

Forty cases of successful corneal grafts with clear optical media for at least 3 months were selected from the eye bank clinic at Irwin Hospital. Patients who had any pathological lesion in the media, retina, or visual pathways which might account for their low visual acuity were discarded. There were 33 cases of penetrating and 7 cases of lamellar corneal grafts.

Cases were divided into 2 groups according to

the age of onset of corneal opacity. Cases with an onset before 5 years and after 5 years of age were labelled as groups A and B respectively. Each group comprised 20 cases (Tables 1 and 2).

Visual acuity was determined after refractive correction, utilising Snellen's whole-line optotypes. In cases of corneal opacity where visual acuity did not improve satisfactorily with glasses because of irregular astigmatism a contact lens was fitted to elicit the maximum visual improvement. Fixation pattern was determined with the help of the projectoscope after full mydriasis. The type of fixation and degree of eccentricity were graded according to the classification of Malik *et al.* (1969).

Results

All the cases in group A had an associated squint of varying degree whereas most of the cases in group B were orthophoric, although seven cases in this group of 19 had a small degree of squint (Fig. 1). The refractive state in both the groups showed astigmatism in almost all cases. The degree and type of refractive error in the two groups were comparable (Table 3).

Most of the cases selected for corneal grafting in both groups had very poor visual acuity, ranging from accurate projection of light to counting fingers. Postoperative visual gain in individual cases of group A and group B is shown in Fig. 2

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Table 1 Corneal opacity cases—onset before 5 years of age

Sl. No. 1	Patient's age (years) 2	Corneal onset age in years 3	Opacity, duration (years) 4	Associated squint 5	Pre-op. vision 6	Post-op. vision with correction 7	Fixation 8
1	8½	5	1½	RCS	RE CF 3M LE 6/6	RE 6/18	RE Parafoveal LE Foveal
2	12	½	11	LDS	RE 6/6 LE CF 1'	LE CF 1'	RE Foveal LE Non-fix
3	11½	½	11	RDS Nystagmus	RE CF 6" LE 6/9	RE CF 6"	RE Non-fix. LE Foveal
4	8	1	6	LCS	RE 6/6 LE CF 1'	LE CF 1'	RE Foveal LE Non-fix.
5	16	½	15	RDS	RE PR LE 6/12	RE HM 6"	RE Paracaeal LE Foveal
6	16	3	10	RCS	RE CF 2' LE 6/18	RE CF 1'	RE Paracaeal LE Foveal
7	19	3	16	RDS	RE CF ½M LE 6/6	RE CF ½M	RE Divergent LE Foveal
8	19	2	14	LCS	RE 6/24P LE PL	RE 6/12P	RE Foveal LE Not seen
9	32	2	30	RDS	RE CF 1M LE: CF 1'	LE: 6/18	RE: Not seen LE: Foveal
10	25	3	20	RCS	RE: PR LE: 6/24P	LE: 6/18	RE: Not seen LE: Foveal
11	20	3	17	RCS	RE: CF ½M LE: 6/6	RE: CF 1M	RE: Paracaeal LE: Foveal
12	22	2	16	RDS Nystagmus	RE: PR LE 6/6	RE: CF 6"	RE: Paracaeal LE Foveal
13	22	1	21	RDS Nystagmus	RE: PR LE: 6/9	RE: CF 6"	RE: Non-fix. LE: Foveal
14	24	1	18	LDS	RE: 6/9 LE: CF ½M	LE: CF ½M	RE: Foveal LE: Paracaeal
15	18	3	14½	RCS	RE: CF ½M LE: 6/12	RE: CF ½M	RE: Paracaeal LE: Foveal
16	22	½	21	RDS	RE: CF 1M LE: 6/6	RE: CF 1M	RE: Paracaeal LE: Foveal
17	40	3	37	RDS	RE: HM LE: 6/12	RE: CF 6"	RE: Paracaeal LE: Foveal
18	40	2	38	LDS	RE: 6/18 LE: PR	LE: CF 1'	RE: Foveal LE: Paracaeal
19	24	3	20½	LDS	RE: 6/12 LE: CF 6"	LE: CF ½M	RE: Foveal LE: Paracaeal
20	12	5	7	RCS	RE: CF 1' LE: 6/6	RE: CF 1'	RE: Paracaeal LE: Foveal

Average age of onset 2.2 years. Average duration of corneal opacity 17.4 years.

and Fig. 3 respectively. In group A 16 cases (80%) did not show any visual gain postoperatively. Four cases (20%) showed slight to moderate improvement. Of these 4 cases, in 3 the operated eye was the only eye; the other eye had corneal opacities (Cases 8, 9 and 10, Table 1). In one case (Case 1) visual acuity improved from 3/60 to 6/18 postoperatively. The corneal opacity in this case developed at the age of 5 years and was operated upon 1½ years later. In group B 19 cases (95%) had visual improvement after corneal grafting; in only 1 case (Case 1, Table 2) did visual acuity not improve. This patient was a 31-year-old male in whom a corneal leucoma developed in the right eye at the age of 17 after injury with a hockey ball. Corneal grafting was done 12 years later, and remained clear for 2 years. The patient had a divergent squint and divergent type of fixation in this eye.

In group A 11 cases in whom no visual gain was observed postoperatively had eccentric fixation, mostly centrocaecal or paracaeal in type; 6 cases had a divergent and 5 cases a convergent squint. Only 1 case had a divergent type of fixation with associated divergent type of squint. Four patients in this group who had associated divergent squint and nystagmus failed to show any particular retinal point of fixation. Nystagmus persisted after surgery. However, they had very widely eccentric areas of fixation. Case 1 (Table 1) had a parafoveal type of fixation and had shown a visual gain from 3/60 to 6/18 postoperatively. All the 3 cases of bilateral corneal opacity mentioned earlier had central fixation in the grafted eye.

In group B 18 cases (90%) had central fixation and 2 cases (10%) had eccentric fixation. The fixation pattern in the grafted eye in the 2 groups is shown in Fig. 4.

Table 2 Corneal opacity cases—onset after 5 years of age

Sl. No. 1	Patient's age (years) 2	Corneal onset, age in years 3	Opacity duration (years) 4	Associated squint 5	Pre-op. vision 6	Post-op. vision with correction 7	Fixation 8
1	31	17	13	RDS	RE: CF 1M LE: 6/6	RE: 2/60	RE: Divergent LE: Foveal
2	34	12	22	RDS	RE: CF 2' LE: 6/6	RE: 6/18	RE: Foveal LE: Foveal
3	25	13	12	Ortho-RCS	RE: 5/60 LE: 6/12	RE: 6/12	RE: Foveal LE: Foveal
4	46	21	19	LDS	RE: 6/36 LE: 6/60	LE: 6/12	RE: Not seen LE: Foveal
5	20	13	7	Ortho.	RE: 6/6 LE: CF 6"	LE: 6/6P	RE: Foveal LE: Foveal
6	25	20	5	Ortho.	RE: CF 1M LE: 6/6P	RE: 6/9P	RE: Foveal LE: Foveal
7	16	7	4	RDS	RE: CF 1M LE: 6/6	RE: 6/18P	RE: Parafoveal LE: Foveal
8	18	11	4	Ortho	RE: 6/60 LE: 6/60	RE: 6/12	RE: Foveal LE: Foveal
9	32	25	4	Ortho.	RE: 6/60 LE: PR	RE: 6/12	RE: Foveal LE: Not seen
10	35	27	3	Ortho.	RE: 6/9 LE: CF 1m	LE: 6/9	RE: Foveal LE: Foveal
11	23	19	2½	Ortho.	RE: 6/6P LE: 6/60	LE: 6/12	RE: Foveal LE: Foveal
12	20	14	1½	Ortho.	RE: 6/6 LE: CF 1M	LE: 6/9	RE: Foveal LE: Foveal
13	35	32	1½	Ortho.	RE: 6/60 LE: CF 6"	LE: 6/12	RE: Foveal LE: Foveal
14	17	10	1	Ortho.	RE: 6/36 LE: 6/12P	RE: 6/18	RE: Foveal LE: Foveal
15	19	18	1	RDS to ADS	RE: 6/60 LE: 6/6	RE: 6/12	RE: Foveal LE: Foveal
16	40	39	1	LDS to ADS	RE: 6/9 LE: CF 1M	LE: 6/9	RE: Foveal LE: Foveal
17	28	27	1	Ortho.	RE: 6/6 LE: CF 6"	LE: 6/9P	RE: Foveal LE: Foveal
18	16	15	1	Ortho.	RE: CF 2' LE: 6/6	RE: 6/9	RE: Foveal LE: Foveal
19	20	19	1	Ortho.	RE: 6/6 LE: CF 1M	LE: 6/6P	RE: Foveal LE: Foveal
20	14	13	1	—	RE: Enucleated LE: 5/60	LE: 6/9P	RE: — LE: Foveal

Average age of onset 19.1 years. Average duration of corneal opacity 5.3 years.

Table 3 Showing the type of refractive error in the grafted eye

	Hypermetropic astigmatism	Myopic astigmatism	Mixed astigmatism	Irregular astigmatism
Group A	6	6	4	4
Group B	8	8	4	—

Discussion

It is known that the structure of the peripheral organ of vision is almost complete at birth but that vision develops gradually. Its development depends on adequate stimulation of the retina by light and objects during early infancy and childhood. When such stimulation is denied to the eyes by opacity in the ocular media early in life a condition of irreversible loss of visual acuity develops which is

termed stimulus-deprivation amblyopia (Von Noorden, 1967). In experiment studies by Chow *et al.* (1957), Wiesel and Hubel (1963a, b, c, 1965a, b), Von Noorden *et al.* (1970), and Goswamy and Agarwal (1971) severe irreversible amblyopia was induced in animals when their eyes were occluded (either by suturing the lids or by plastic occluder or by producing corneal scarification) during the first few months of life. No comparable visual defect was observed when these experiments were repeated in adult animals. Though such data cannot be transferred to man without reservation the results of occlusion appear to be similar to clinical amblyopia.

The present study shows that, when a corneal opacity is unilateral and arises before 5 years of age and corneal grafting is delayed until adulthood, a severe amblyopia develops. If, however, the corneal opacities are bilateral, the amblyopia is not very dense at least in one eye, and good visual

Fig. 1 Nature of associated squint

Fig. 2 Group A: Postoperative visual gain

Fig. 3 Group B: Postoperative visual gain

Fig. 4 Fixation pattern (groups A and B)

improvement takes place (case 9, Table 1). Von Noorden and Maumenee (1968) recorded 3 similar cases. They did not find visual improvement in cases of unilateral corneal opacity, but some visual improvement was seen in two cases of bilateral corneal opacity. Picetti and Fine (1966) also observed good visual gain in 2 cases of bilateral congenital corneal dystrophy even though they were operated on at 7 years of age.

In the present series all the cases of unilateral corneal opacity before 5 years of age developed eccentric fixation, but if the corneal opacity was bilateral then in the relatively better eye foveal fixation was preserved and visual gain obtained (Cases 8, 9 and 10, Table 1).

The present series also clearly indicates that when corneal opacity develops after 7 years of age it has no significant influence on the development of amblyopia and fixation pattern irrespective of the duration of the opacity before surgery. However, if the corneal opacity is unilateral and develops around the border line age of 5 and 7 years (Case 1, Table 1 and case 7, Table 2) there is less improvement in visual acuity with the parafoveal type of eccentric fixation.

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