

FOVEAL ABNORMALITIES IN AMETROPIC AMBLYOPIA

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This paper was awarded "The Emmie Russell Prize" for 1980.

Abstract

A study was performed on 90 children who clinically exhibited some form of foveal and/or macular abnormality. The most significant feature found was the high incidence of ametropia.

An attempt was made to correlate the clinically seen abnormalities of the fovea with:

- 1) The refractive state of the eye*
- 2) The degree of visual reduction.*

Key Words

Foveal abnormalities, ametropia, hypermetropia, astigmatism, foveal/macular hypoplasia, amblyopia.

INTRODUCTION

During a study performed by the author in 1979 on patients with bilateral eccentric fixation, certain specific abnormalities of the fovea were noted.

The foveal abnormalities seen in this study are divided into the following categories:

- 1) decreased or absent foveal reflex
- 2) poorly defined pigmentation in the macular area
- 3) abnormally small vessel-free area.

These categories were described by Curran and Robb² and Yoshizumi et al³ in their discussions on foveal hypoplasia.

METHOD

A total of 90 children were found who exhibited one or more of the following characteristics in one or both eyes:

- 1) absent foveal reflex
- 2) diffuse, mottled, pale, poorly defined or elongated foveal reflex
- 3) displaced foveal reflex (ie. not in its normal central macular position)
- 4) absence of the pigmented macular area
- 5) poorly defined pigmentation in the macular area
- 6) small vessel-free area
- 7) nystagmus (either on observation or on visuscopy).

All these patients had been given a routine ophthalmological examination including refraction under cycloplegia, fundus check and a general orthoptic investigation including visuscopy. All patients had also at some stage been examined by the author.

Except for 2 patients who exhibited some degree of albinism, all the patients studied were considered to be grossly normal by the consultant ophthalmologist.

RESULTS

Sex Distribution — of the 90 cases, 46 (51%) were male and 44 (49%) were female.

Age Distribution — the age range of patients at first attendance was from 10 months to 15 years, the average age at the initial visit was 5.8 years.

Strabismus — some form of strabismus was present in 81 patients in the following proportions:
69 patients had a convergent squint (76.7%)
12 patients had a divergent squint (13.3%)
6 patients had a heterophoria only (6.7%)
3 patients had no demonstrable squint (3.3%)

Area of Fixation — as the foveal reflex was either absent, poorly defined or displaced in many cases, the fixation patterns were classified according to the following criteria:

- 1) "central" or "unsteady central" — around the central macular area — 69 eyes (38.3%)

- 2) "parafoveal" – between the central area and the side of the macula – 75 eyes (41.7%)
- 3) "macular" on the side of, but within the macular area – 21 eyes (11.7%)
- 4) "paramacular" – outside the macular area, but within the vessel-free area – 7 eyes (3.9%)
- 5) "greater than paramacular" – beyond the vessel-free area – 4 eyes (2.2%).

A further 4 eyes have not been classified into the above groups. These were the 2 albinoid patients who demonstrated none of the above landmarks, fixation being nystagmoid movements in the area where the peripheral blood vessels converged.

Refractive Error – the figures used in the following statistics are taken in terms of retinoscopy.

TABLE 1 – Spherical Error (in dioptres)

Amt. of error	No. of eyes	%
+6.00 and over	74	41.1
+2.00 to +5.75	80	44.4
-1.75 to +1.75	14	7.8
-2.00 to -5.75	7	3.9
-6.00 and over	5	2.8

Table 1 shows the distribution of general refractive error (using the spherical equivalent value). It can be seen that there is a significant number of eyes with a high degree of hypermetropia (41.1% with +6.00D and over) and only a very small number of eyes with high myopia (2.8% with -6.00D and over).

TABLE 2 – Astigmatism (in dioptres cylinder)

Amt. of error	No. of eyes	%
nil to 0.25	63	35.0
0.50 to 0.75	43	23.9
1.00 to 1.25	24	13.3
1.50 to 1.75	18	10.0
2.00 to 2.75	21	11.7
3.00 and over	11	6.1

Table 2 shows the distribution of astigmatism. It can be seen that there is a fairly high number of eyes in the higher degrees of astigmatism (18% with 2.00DC and over).

Visual Acuity – Fig. 1 shows the overall distribution of the best achieved visual acuity. Only 44 eyes (24.4%) had normal vision (6/6 or better). 55.6% of eyes had mildly reduced vision (6/9 – 6/12) showing that in these cases, the foveal abnormalities are unlikely to be an indication of a gross ocular anomaly.

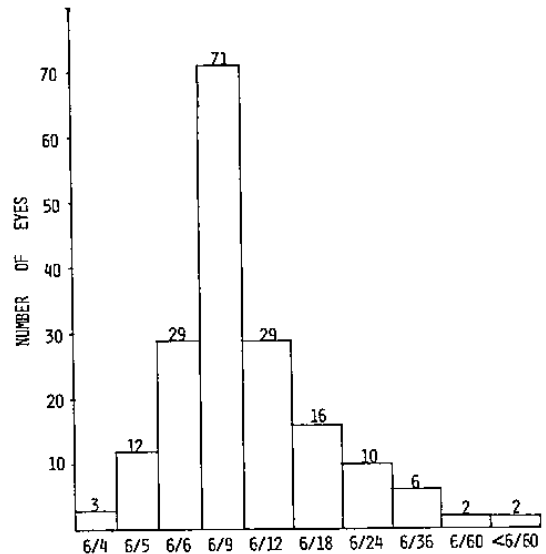


FIGURE 1

Visual Acuity (best achieved) vs. Refractive Error

– Reduced visual acuity of 6/12 or worse was found to occur much more frequently in the higher refractive error groups (51.3%: +6.00D and over, 100%: -6.00D and over) and very infrequently in the lower refractive error group (21.4% between -1.75D and +1.75D). In the latter group, normal vision was more likely to be the rule (42.9%: 6/6 or better).

The picture was similar in the case of astigmatism, where above average acuity (6/5 or better) was only attained in the presence of low degrees of astigmatism (0.75DC or less) and the incidence of reduced acuity (6/9 or worse) increased with an increase in the degree of astigmatism (68%: 0.75DC or less, 100%: 3.00DC and over).

Types of Foveal Abnormality – A wide variety of abnormalities was found, sometimes only one area being affected and sometimes a combination of areas. The most common abnormalities were those involving the fovea itself (50%).

Visual Acuity vs. Foveal Abnormalities – The occurrence of foveal abnormalities was higher in the presence of reduced acuity (92% with 6/9 - 6/12) and lower in the above average vision group (53.3% with 6/5 or better). There was however no further increase in the incidence of foveal abnormalities as the visual acuity decreased. There was no significant difference in the incidence of foveal abnormalities in the dominant and non-dominant eye.

Refractive Error vs. Foveal Abnormalities – A correlation was found between the amount of refractive error (spherical error) and the presence

of foveal abnormalities; the highest incidence being in the higher refractive error groups (89.2%: +6.00D and over, 80%: -6.00D and over) and the lowest incidence in the lower refractive error group (57.1% between -1.75D and +1.75D).

A similar correlation was found in the case of astigmatism (82.9%: 0.75DC and less, 90.9%: 3.00DC and over). It has also been found that astigmatic eyes sometimes demonstrate an elongated foveal reflex as found at ophthalmoscopy rather than the usual pinpoint shape. A similar correlation also exists in this case (10.4%: 0.75DC and less, 27.3%: 3.00DC and over).

DISCUSSION

Helveston and von Noorden⁴ conducted a study on the appearance of the fovea in strabismic amblyopes. They compared the foveal appearance of the amblyopic eye to that of the other so-called "normal" eye, in the same individual. They found the incidence of foveal abnormalities to be almost the same in both eyes. However, it has been found that the dominant or "normal" eye of an amblyope may not indeed be the same as a normal eye of a normal subject, where some degree of amblyopia and eccentric fixation may be demonstrated even in the fixing eye.^{5 6 7} This was also demonstrated in this study where 64 patients (71.1%) had foveal abnormalities in both eyes.

Albinism — A common feature of the albinoid eye is that the macular area is not fully developed and there is absence of the foveal reflex. Mann⁸ states that in foetal development pigmentation of the eye appears just before the differentiation of the retinal layers, and may also determine this. In the absence of pigmentation, it can follow that retinal development may be delayed and never reach its final stage ie. the formation of the fovea. Thus reduced visual acuity would occur.

This feature may be evident in the two patients in this survey who exhibited albinism, where the macular and foveal areas were absent and substandard vision was found.

Refractive Error —

Hypermetropia — In cases of macular hypoplasia, the retina in the region of the posterior pole fails to develop fully, remaining at a stage of differentiation similar to that of a 6th month foetus. These patients usually demonstrate gross amblyopia or even blindness.¹ Duke-Elder¹ and Mann⁸ both state that a similar condition or intermediate stages between hypoplasia and full differentiation

of the macular area may exist in highly hypermetropic eyes. Duke-Elder⁹ also states that the hypermetropic eye is typically small and as such could therefore be considered an underdeveloped eye. If this is the case, then it could follow that the hypermetropic eye might demonstrate a relatively more frequent occurrence of developmental foveal abnormalities.

In this present study, it can be seen that a significant percentage of eyes have a high degree of hypermetropia. 35.7% have an error of +7.00D and over. The significance of this figure can be seen when compared to a survey by Broekema⁹ where only 4.3% of hypermetropes had an error of +7.00D to +10.00D. Also there is a much higher incidence of foveal abnormalities in the most hypermetropic group than in the lower refractive error group (89.2%: 57.1%).

Duke-Elder⁹ states that the visual deficiency in marked hypermetropia may be due in part to a lack of retinal development. Reduced acuity of 6/12 or worse was present in 51.3% of eyes in the most hypermetropic group compared to only 20% of eyes in the next group (+2.00D to +5.75D).

Astigmatism — It is interesting to compare the incidence of astigmatism found in this study to a survey by Cavara et al⁹. The incidence in lower degrees (less than 1.00DC) is similar in the two surveys — 58.9% here compared to 65.5% (Cavara); however, a different picture is seen in the higher degrees (2.00DC and over) - 17.8% compared to only 9.5%. As previously mentioned, in the higher degrees of astigmatism, there is a higher incidence of both amblyopia and foveal abnormalities.

Ikeda and co-workers^{10 11} have found that amblyopia can arise as a result of lack of clearly focused image stimulation of the central retinal ganglion cells during early development. In the case of high hypermetropia and astigmatism there is no position where an object of regard is seen clearly and permanent reduction in acuity may result.

CONCLUSION

The most significant finding in this study is the high incidence of ametropia, especially hypermetropia and astigmatism in children with foveal abnormalities.

The markedly hypermetropic or astigmatic eye may be one that is developmentally deformed and as such could then have a higher incidence of

developmental anomalies of the foveal and macular areas, as these are the last of the retinal areas to fully differentiate.

To possibly determine the effect of these abnormalities on any associated amblyopia, one could perform a foveal electro-retinogram (ERG) to establish if there is some depression of function at the retinal level itself.

The fact that a high percentage of the patients in this study do not have gross amblyopia, as would be expected in true foveal hypoplasia, indicates that in these cases the foveal abnormalities are not the main aetiological factor of the reduced vision; but these abnormalities may be associated with some other causative factor eg. hypermetropia or astigmatism.

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Ed's. note:

The role of unilateral squint and anisometropia in the genesis of amblyopia and the evidence of binocular interaction at cortical level has not been considered in this paper. Useful reviews are those by Gunter K. von Noorden "Current Concepts of Amblyopia" in *Orthoptics Past, Present, Future*, 1976. Stratton Intercontinental Book Corporation New York p37, and Hisako Ikeda "Visual Acuity, its Development and Amblyopia" August 1980 *Journal of the Royal Society of Medicine*, 73, 546.