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ORIGINAL ARTICLE

STEREOACUITY AND FUSIONAL VERGENCE RANGES OF SIBLINGS OF CHILDREN WITH FAMILIAL STRABISMUS

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Abstract

Genetic factors play an integral role in the cause of many cases of strabismus and it is currently believed that the mode of transmission is multifactorial with a threshold effect, that is, that specific genes must be present above a certain threshold before a strabismus is produced. If this is the case, then it is likely that close relatives of those with strabismus may also possess some of these 'abnormal' genes, but to a lesser extent, and therefore have subtler abnormalities of binocular vision without eliciting a manifest strabismus.
This assumption was tested by measuring the stereoacuity and fusional vergence ranges of 16 non-squinting siblings of children with early onset strabismus. Analysis of the data showed that compared with a control group of children with no family history of strabismus, there was a significant decrease in the stereoacuity of the experimental group (p=0.0245) but no significant difference in the fusional vergence ranges.
It is possible that defects of the vergence response may be too subtle to be elicited by this method and more information may be gained by studying the dynamics of the response. Nevertheless, the reduced stereoacuity supports the multifactorial theory for the inheritance of strabismus.
Keywords: *inheritance, genetics, strabismus, stereoacuity, fusional vergence.*

INTRODUCTION

Observations on familial strabismus have been documented since the origins of the medical profession. Hippocrates is said to have stated that:

> "the children of parents having distorted eyes, squint also for the most part"[1]

The relative failure of the Mendelian model to explain the inheritance of strabismus, along with the observation that, in strabismus, a continuum exists among individuals (from orthophoria to heterotropia) gives credence to the current belief that 'familial' strabismus is most probably

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due to multifactorial inheritance with a threshold effect[1,2a,3]. Basically it is a condition which is expressed when independently inherited genes work additively to produce the specific strabismus genotype, but only when a distinct threshold is exceeded[1].

In multifactorial inheritance it is recognised that the parents are unlikely to be genetically uniform in regard to the presence of 'abnormal' genes, therefore, each different parental gene combination will produce a different frequency of affected offspring, as affectation is dependent on how many abnormal genes are present in the combined parental gene pool. Due to this complexity, it is usually impossible to predict the likelihood that an offspring will be affected. However, it is known that with the birth of each affected child the likelihood of future offspring being similarly affected increases[1].

Binocular Single Vision is dependent upon the eyes' anatomy, sensory aspects and refractive state. These factors are thought to be determined polygenically, that is, numerous genes act to encode the development of the eyes and orbits, neurological connections, the grades of binocular single vision, the AC/A ratio and the refractive error[4].

Currently it is not known precisely what genes are involved and what quantities of abnormal genes are required for an individual to have strabismus. "However, whatever factors do exist, it is likely that they will have differing effects depending on their combinations"[1].

According to Spivey[4], the co-occurrence of poor vergence ability and hypermetropia mediate against the full development of binocular vision. Shclossman and Priestly[3] postulated that at least two genes (one affecting the ectoderm and the other affecting the mesoderm in the developing embryo) caused strabismus and that both genes probably had different patterns of inheritance. Richter[2b] also felt that two or more genes were responsible in the expression of strabismus. She held that these genes were independent and dominant; one determining a phoria (motor anomaly) and the other determining an anomaly of binocular vision (sensory anomaly.

It has been determined that phoria[1], sensory anomalies and refractive errors can all be inherited independently but if members of pedigrees with different types of anomalies marry, some of the offspring are more likely to have strabismus[1]. It has been shown[5,6] that many relatives of affected individuals demonstrate various slight abnormalities of binocular function without actually having strabismus, for example, non-squinting parents of children with early onset esotropia tend to have lower fusion ranges[5] and stereopsis[6] that those of the general population.

Although it is impossible to precisely predict the probability of offspring being affected, three conditions have been defined which significantly increase the likelihood of offspring inheriting esotropia[3]:

1. When a parent has an esotropia.
2. When parents are unaffected but there is a strong family history of the condition.
3. When parents are unaffected but have a low fusional ability and/or a significant refractive error.

So if it is possible for parents to pass the genes responsible for strabismus to their offspring, it is feasible to assume that non-squinting siblings of affected children may have inherited some aspects of abnormal binocular function from their parents without manifesting strabismus itself; that is, "there could be subnormal (or submodal)

functioning of one or more of the contributing components of binocular single vision"[7], namely simultaneous perception, fusion and stereoscopic vision.

The research question therefore addressed in this study was:

Do the non-squinting siblings of children with early onset esotropia of presumed genetic origin have reduced stereoacuity and/or reduced fusional vergence amplitudes?

METHOD

Subjects

Two groups of subjects were involved in the study. The control group consisted of 55 subjects between 4 and 13 years of age with no family history of strabismus. They were pupils from primary schools in the Sydney metropolitan region. The experimental group was obtained from orthoptic clinics and consisted of 17 subjects of the same age range, all of whom were non-squinting siblings of strabismic patients. (For the fusional vergence amplitude analysis, only 13 of these subjects were tested).

In order to be included in the study, control group subjects had to meet the following selection criteria:

- Written parental consent.
- No family history of strabismus (no affected siblings, parents, grandparents, cousins, paternal/maternal uncles or aunts.
- Non-corrected visual acuity of 6/6 or better with either eye (this was established during testing).

Stereoacuity

This was conducted using the Titmus Stereotest under good background illumination and held 40 centimetres from the subject's face. Firstly, gross responses, that is, the house fly, were checked. This was then followed by testing the animals, followed by the circles. Stereoacuities were recorded as the level before two consecutive mistakes were made. Any suspicious points were rechecked once. The test plate was always geld by the researches as suggested by the instruction guide accompanying the Titmus Stereotest.

Horizontal fusional vergence amplitudes

Horizontal fusional vergence amplitudes were then measured at ⅓m and 6m using a prism bar. One recording at each distance was taken due to time restrictions imposed by schools and clinics. An average of three measurements would have been preferred. However, all subjects were equally disadvantaged and therefore results would not have been influenced. Divergence range was measured first, followed by convergence range which was measured using a non-accommodative target, to remove as much as possible any effects of accommodation influencing convergence. The fusional vergence amplitude was taken as the power of prism prior to that which caused fusion to break. Subjects were not encouraged to maintain a single image as such fusional exertion does not occur in everyday situations.

RESULTS

Preliminary analyses showed that both the size of the latent deviation, the age of the subjects and the vision of both groups were equivalent.

Stereoacuity

The stereoacuity levels of the control group ranged between 40 and 80 seconds of arc with a mean score of 43.09 and a mode of 40 seconds of arc. The sibling group's stereoacuity had a range of 40 to

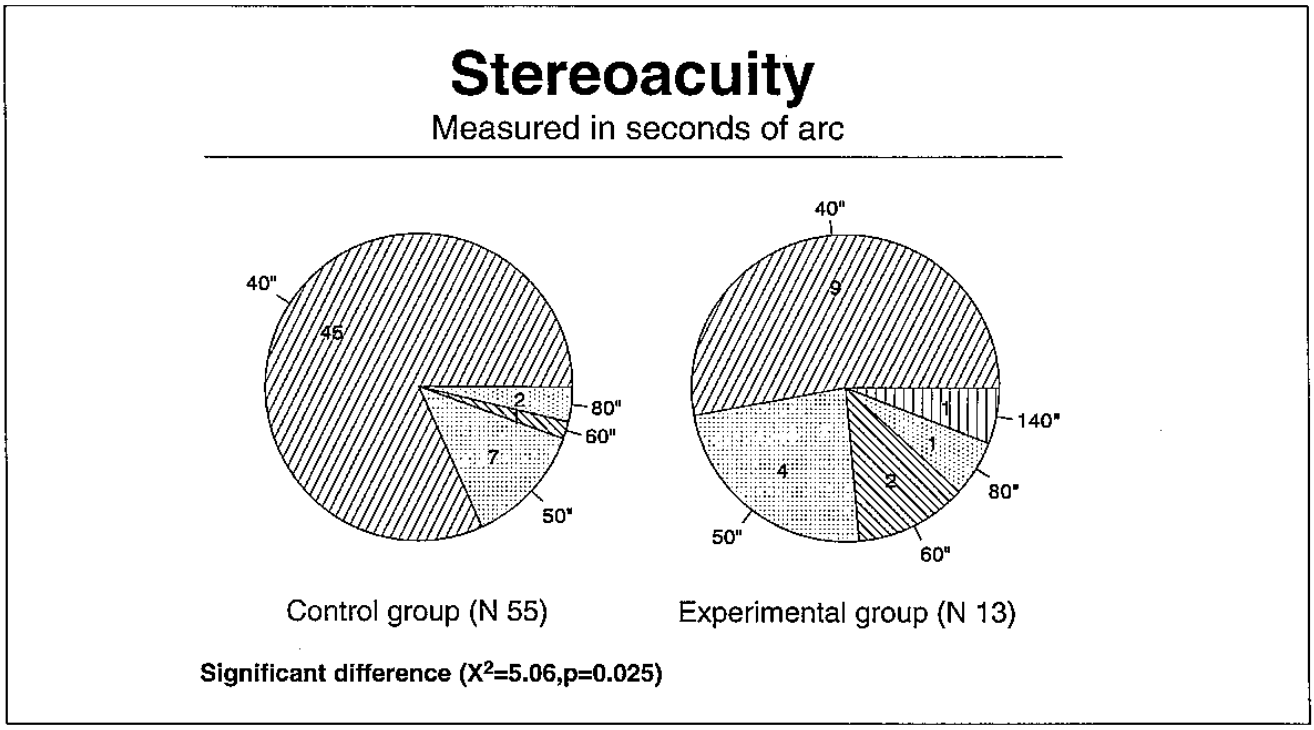

Figure 1. Stereoacuity

140 with a mean of 53.13 and a mode of 40 seconds of arc.

The two groups' stereoacuities were compared using a Kruskal-Wallis one-way non-parametric ANOVA test and were found to be significantly different at the 0.05 level ($X^2 = 5.06$ and $p = 0.025$). (See Figure 1). The two groups appear to have

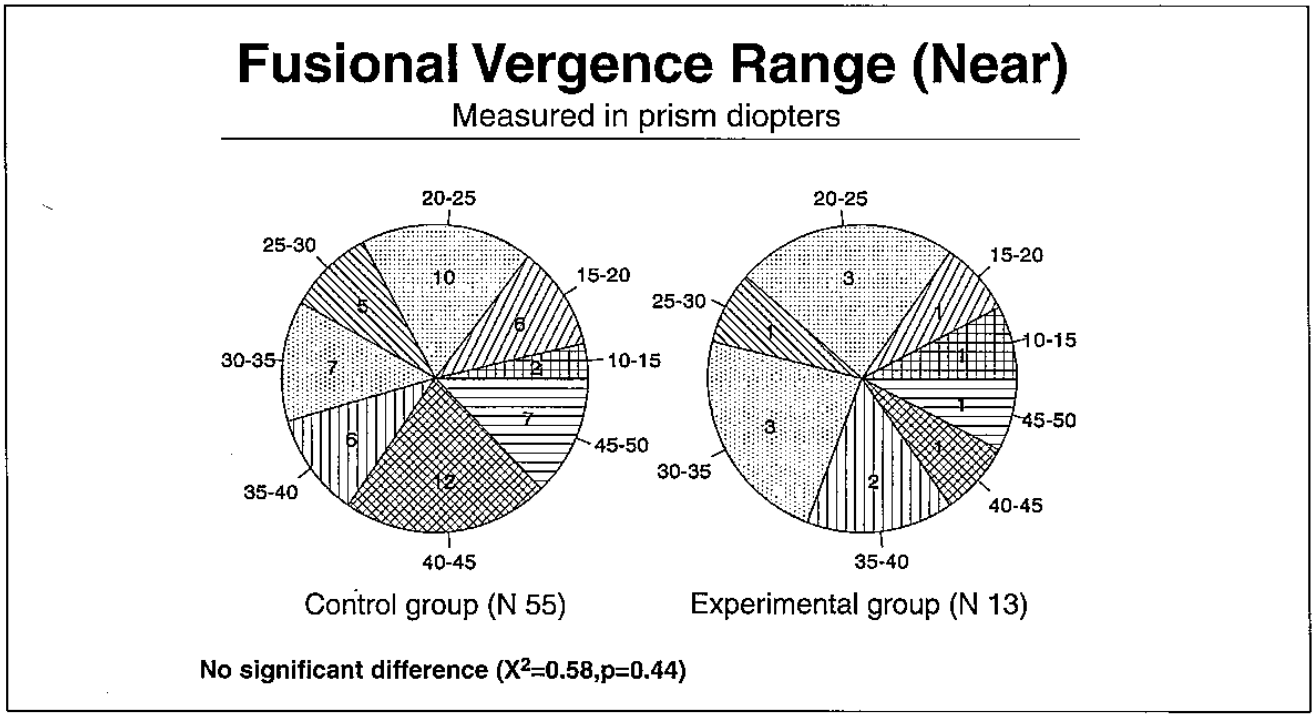

Figure 2. Fusional vergence range (near)

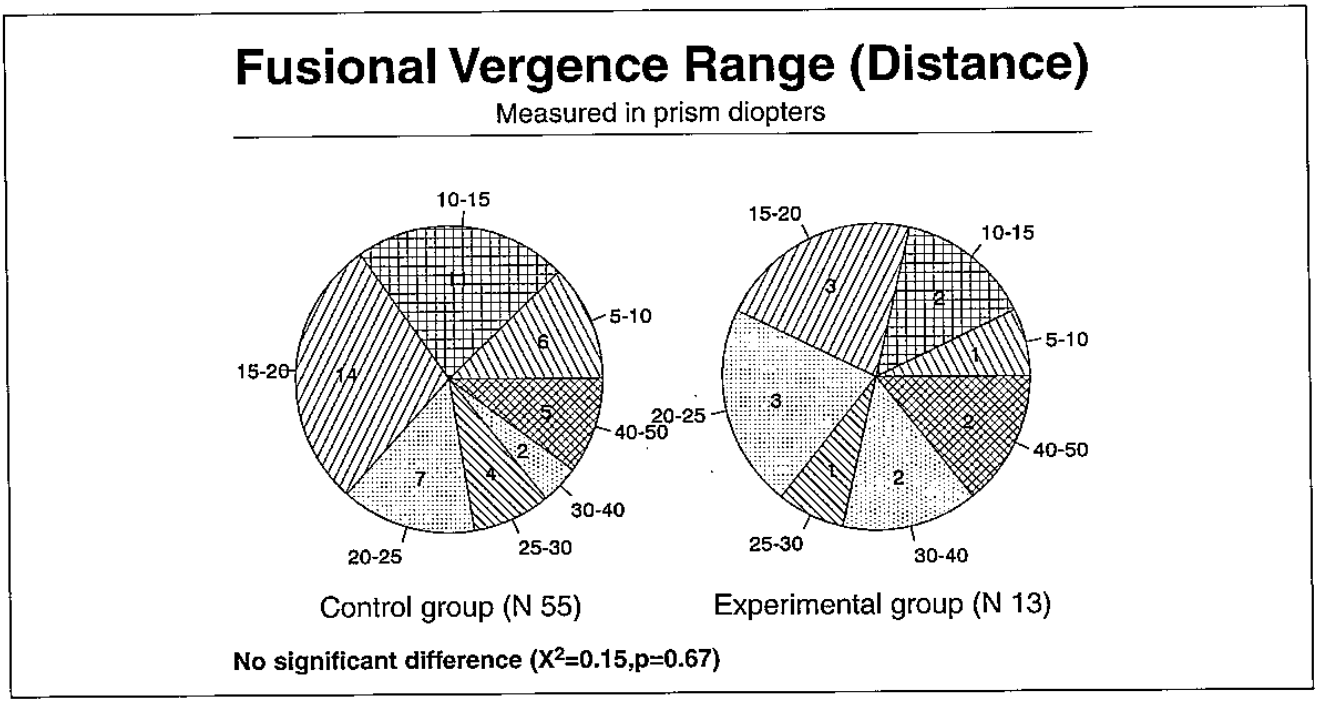

Figure 3. Fusional vergence range (distance)

come from different populations. This analysis indicates that there is a significant difference between the stereoacuity of siblings of children with a strabismus, that is, those children with a suspected affected gene pool, and the general population.

It can therefore be summarised that children who have a genetic predisposition to strabismus are more likely to have a reduced level of stereoacuity compared to a group of children for whom there is no evidence of a genetic predisposition.

Fusional vergence amplitudes

The base in and base out amplitudes of each group were added to give the total range for both near and distance. (See Figures 2 & 3).

A Kruskal-Wallis one-way ANOVA showed that no significant difference existed between the control group and the experimental group for near fusional vergence range ($X^2 = 0.58$ $p = 0.44$) and distance fusional vergence range ($X^2 = 0.0.15$ $p = 0.67$).

DISCUSSION

Stereoacuity

Although the sample size of the experimental group for the research was fairly small, it can be considered that the difference of stereoacuity levels between the two groups is indeed a clinically significant difference. This is due to the power of the experimental design. As the sample consisted of only 16 subjects in the experimental group, it would be expected that a significant difference would be found only if the effect size in the population was larger. If the effect size was only subtle then differences between the two groups would be likely to be found only if the sample sizes were much large, that is, approximately 200 subjects or more[8]. Therefore, for this difference between the two groups to be found with only 16 subjects it must be quite significant.

Fusional vergence amplitudes

The results for this variable would also have been affected by the low number of subjects in the experimental sample. The power of statistical tests to detect population differences depends on the size of the samples taken from that population and the magnitude of the effect within the population. Had there been a large effect within the population, a sample size of 13 should have been adequate to detect it. The fact that the probability levels in this study did not even approach significance suggests that, if the effect does occur in the population, it is unlikely to be a very large effect[8].

It may be that relatives of strabismic individuals demonstrated slight defects of fusional vergence amplitude. Perhaps these abnormalities are too slight to be detected by the methods used in this study. A sample size of approximately 200 would be needed to give a reasonable chance of detecting any subtle effect which may exist in this case. It was not feasible to collect a sample of this size for this study. However, further research should involve much larger samples than those used here.

Perhaps more refined methods of investigating the quality of the fusion reflex may be needed, for example, Schor[9] stated that "fusional vergences are controlled by a fast acting (transient) mechanism that aligns the eyes in response to retinal image disparity and a slow (sustained) mechanism that sustains binocular alignment". Studies such as this could be extended by the use of precise methods of analysing the vergence response to investigate this aspect of the vergence response, or possibly the latency and accuracy of the response.

SUMMARY

Reduced stereoacuity levels have been demonstrated in the siblings of children with early onset esotropia. This has shown support for the theory that several gene pairs are required to produce strabismus in a quantity large enough to reach a threshold level, and that without this threshold level being reached, the result may be a reduced level of binocular single vision.

That the experimental group did not demonstrate significant differences in fusional vergence amplitudes suggests either that this component of binocular single vision is not affected by inherited factors, or, possibly, that the effect is slight and requires a more detailed analysis to reveal any difference in this function.

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