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Clearly describe the purpose of the study. Include methodological information on procedure and design. Outline statistical methods of analysis and demonstrate results using figures and tables. Avoid duplicating information between text and diagrams. Discuss the relevance and implications of the study and provide a brief conclusion.

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Books: Cornsweet TN. Visual perception. 2nd ed. New York: Academic Press Inc. 1971: 6-26.

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Editorial

Strabismus Vision Perimetry

The three pillars of orthoptic practice are presented in this edition of the AOJ. Strabismus is being investigated as it never has before, with an oral tissue swab. The Strabismus Inheritance Study in Tasmania, lead by Robin Wilkinson, is currently divining the genetic DNA mysteries of families with hereditary strabismus. This work has received the mantel of The Patricia Lance Lecture and is printed in full, following presentation at the 54th Annual Scientific Conference of the OAA in Sydney, 1997. This very exciting study has been designed to answer questions on the sequence of motor or sensory defects in the evolution of strabismus and quandaries such as seemingly opposite types of strabismus, convergent and divergent, occurring as a result of a common genetic anomaly in a controller such as the vergence system. This research will be invaluable to our understanding of the mechanism of binocular vision. Santamaria reminds us of the embryological approach to strabismic syndromes in an atypical case of vertical retraction syndrome (without retraction!). An extensive review on vergence eye movements by Luu et al brings the reader in line with current neurophysiological findings concerning the location and behaviour of the vergence control centre. An exhaustive population study by Cornell et al of almost 2000 normals and their accommodative responses to the RAF rule reaffirms the normative data attributed to this instrument nearly a century ago. These authors remind us though, that an average is an average is an average! When determining if accommodative ability is deficient, the range (1-4D) around the average needs to be considered as well as the stated age related near point.

Vision and its relevant assessment features predominantly in this edition. Spirited dialogue is seen in the Letters to the Editor. In the last edition, Duyshart commented on the article by Williams et al, Comparison of crowded single optotypes with linear acuities in amblyopes. 1995; 31:21-27. Issues were raised as to the screening tests that detect different types of amblyopia and the most applicable tests for three and a half year olds. The authors have taken the opportunity to respond to these comments, reminding the reader that the limitations of research are clearly outlined and that composite parts of a research question need often to be considered in isolation before they can be orchestrated in combination. This is a continuing problem in ocular research as psychophysical tests have effects in their separate components and then in interactions. Whitton provides us with statistical confirmation of clinically known entities that SGS elicits a greater VA response than SGL and that four year olds give a better VA level than three and a half year olds. The temptation to embrace such results unequivocally is almost irresistable. Whitton achieved these results by converting VA measures to a ratio scale and applying parametric analyses. She has made many assumptions in the treatment of the data and has even averaged the VA responses for each eye into one score. As this will become a much quoted and landmark study, we need to consider if these assumptions are valid. The possible differing treatments of VA as a variable in analysis are demonstrated in three papers. While Whitton converted VA to a ratio scale, Haynes et al compared their VA data to questionnaire data by treating VA as an ordinal scale and Fitzmaurice et al utilized both parametric and non-parametric analyses. Researchers must outline their assumptions and justify their decisions to embrace such assumptions, while readers need to query this constantly.

Haynes et al raise the question as to whether visual acuity testing pre-operatively in cataract

patients serves any useful purpose when compared to subjective assessment of visual (dis)ability in daily activities. What needs to be considered is, is VA testing a valid method of assessment of visual function in the ageing population with progressive cataract formation? Is the VF 14 a valid measure of visual function? Do we need to design a better questionnaire and query patients on their responses to monocular and not binocular vision? The implications for public health care expenditure based on these results are significant. Judgements on such resource allocations are also raised by Jones who looks at predictors in stroke rehabilitation and this provides useful information for clinicians attending this population. The appropriate use of VA as a measure is also raised by Fitzmaurice et al in the population of intellectually and physically handicapped children. These authors conducted a multicentre international collaborative study using computer generated visual objects and assessed subjects' abilities to respond. The importance of pursuits such as this is clearly outlined by the extensive survey of Tosswill et al which showed that the incidence of visual impairment in the multihandicapped population is 100 times greater than in the general Australian population.

The issues in perimetry that are presented are all significant. Jolly describes three cases with retinitis pigmentosa and illustrates the effect of arbitrary standards in VA and perimetry for driving licencing. She provides a range of non standard tests of visual function and remediation for raising an individual's awareness of the vision sense during driving. Rota-Bartelink provides a review on the perimetric literature in relation to glaucoma. This is a most useful summary which describes the variables; static, kinetic, achromatic, short wavelength, and flicker. Discourse is also given on the role of motion detection, colour vision and the influence of the ageing lens.

Vassar et al investigated the relationship of colour vision sensitivity to contrast sensitvity and discovered that contrary to their hypothesis, diminished function of one ability was related to a diminished, rather than a compensatory increase in the other ability. This appears to hold a key to closely associated neurophysiological substrates. This is akin to the comments of Pollock regarding thresholds in red perimetry included in the Letters to the Editor in this edition. Pollock comments on the article by Piraino and Goodacre, Normal threshold values for red targets in the central 10 degree visual field.1996:32:19-25. The issue is raised as to the values generated by the Humphrey Visual Field Analyser when comparing chromatic and achromatic targets. A case is presented that shows higher dB thresholds for white over red targets, but a greater field loss for a red target in the graytone field. This testing has incorporated white light sensitivity, selective spectral sensitivity, form detection and on/off motion detectors. Pollock advances possible neurophysiological substrates. The authors reply to this comment choosing to focus on the bases of the HVFA calculations and the importance of the interpretation of dB values as being relative or absolute. There will be inherent measurement variability, as well as differences across quadrants. These comments remind us that the majority of our research needs to focus on validation of our clinical tools. Are we measuring what we want to measure? How are changes quantified? How does pathology influence these measurements?

Whitton et al provide a report on the recent glaucoma detection device, the nerve fibre analyser. Nerve fibre layer and ganglion cell loss attributed to glaucoma are deduced by interpretation of deviations in parallel light ray paths. Early detection and early intervention are the aims in the treatment of this disease. We are reminded that this applies to amblyopia as well, in a review with an historical perspective presented by Shippman. Thought processes over the centuries on this disease are revisited and we are mainly reassured of the occlusion rationale but not necessarily the regime. Life goes on.

Errata

Erratum 1.

Piraino, J and Goodacre, H.

Normal threshold values for red targets in the central 10 degree visual field.
AOJ 32; 1996: 19-25.

In the abstract on page 19, the visual threshold for red fields is stated as 26.6 dB with standard deviation + 1.83 dB centrally. This should read 26.2 dB.

In the last paragraph of the Results section on page 23, this is corrected to reflect the differences in results found in one subject using white and red targets. This paragraph now reads as follows: Two of the three red abnormal firelds were both from the same subject. The results for the left eye using a white target show a normal visual field. However the same eye with a red target had an almost complete superior field defect with the superior temporal and nasal quadrants equally affected. Z-test shows that 99.92% of the population scores a better quadrant total than 337 in the temporal quadrant and 99.95% of the population scores a better quadrant total than 346 for the superior nasal quadrant. Other tests performed on this subject showed normal colour vision but borderline contrast sensitivity.

Erratum 2.

Cornell, E, Flanagan, J, Heard, R.

Evaluation of compensatory torsion by blind spot mapping.
AOJ 32; 1996 : 13-17.

The key in Figure 7 should show reversal of the labelling for intorsion and extorsion. The text is quite clear in that the counter intorsional response is larger than the counter extorsional response to passive head tilt. This is graphically demonstrated in Figure 7.

The Australian Orthoptic Journal published an article by Piraino and Goodacre in the last edition on the thresholds for red perimetry. This drew the following letter from Jean Pollock.

Dear Editor,

I noted with interest the welcome normative data for "...threshold values for red targets in the central 10 degree visual field" (Piraino J. & Goodacre H. Aus Orth Jnl 1996; 32:19-25[1]). The authors raised the old clinical conundrum of how and why a chromatic stimulus demonstrates greater apparent sensitivity in revealing pathology compared with an achromatic stimulus.

If we assume that a chromatic stimulus is only behaving as a diminished luminance stimulus, then we should expect that simply decreasing the luminance of an achromatic stimulus will reveal the same distribution of visual field loss as that demonstrated by the red stimulus. Is this the case using the Humphrey Visual Field Analyser (HVFA)?

Amongst their normal population Piraino & Goodacre[1] found that the mean threshold decibel (dB) measurements for white and red stimuli were significantly different. They also identified 3 "abnormal" fields where no defect was seen with a white stimulus, despite the presence of a definite abnormality with a red stimulus. In one of these subjects a borderline contrast sensitivity function was noted. How should we view the potential dB differences between white and red stimuli in the presence of pathology?

I suggest that comparison of the relative defect depths could be more informative of whether actual differences exist between results using these stimuli.

A comparison of this type would accept that the magnitude of the measurements were different, instead comparing the log scale of relative difference. The comparison instead is therefore between *relative* defects in dB.

The following data compare the threshold dB defects between a 30-2 white and a 30-2 red HVFA measurement from the same patient. A diagnosis of left sphenoidal ridge meningioma was confirmed radiologically and surgically. In the interests of brevity only the dB deficits in the central 10° are presented for the left, affected eye. (See Figure 1.)

In 11/16 (69%) of the measurements, the defect depths for the white stimuli were more severe or equal to those of the red stimuli.

11 **10**	12 **11**	12 **8**	12 **7**
12 **15**	14 **14**	14 **10**	5 **11**
12 **14**	14 **16**	14 **31**	13 **36**
12 **16**	14 **19**	13 **29**	13 **36**

Figure 1

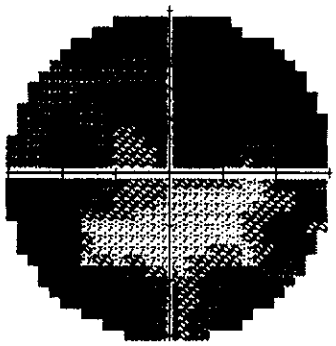
Decibel defect depth with HVFA 30-2 red and ***30-2 white***

However, when viewing the distribution of threshold (graytone) visual field loss (Figures 2. & 3.), we note the evident enlargement and profundity demonstrated by the red compared with the white stimulus. We do not see a similar pattern of field distribution loss with the white stimulus even at high dB defect depths.

There appears to be no controversy regarding chromatic stimuli eliciting responses in their own right and not simply as diminished luminance stimuli, when their luminance is matched to that of the achromatic stimulus (isoluminant). This is acknowledged by Piraino and Goodacre[1]; citing Flanagan & Hovis[2]. What luminance levels does the HVFA actually offer then for each of these test paradigms? According to the 1994 User's Guide the maximum stimulus intensity for the achromatic test is 10,000 apostilbs (apostilbs

Figure 2

Threshold graytone 30-2 white

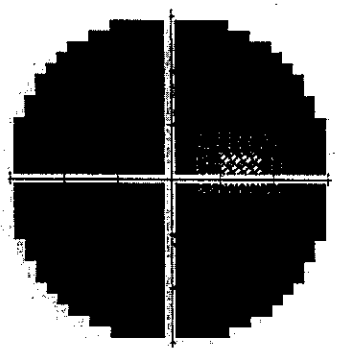


[asb] are measurement units of luminance) which is equivalent to 0 dB or a 4e Goldmann stimulus (i.e. no neutral density filter attenuation). "In color testing, zero decibels still represents the maximum instrument brightness although that maximum brightness is less than 10,000 asb"[3]. The light transmittance of The Hoya R62 red filter is 91.5%.The background luminance of 31.5 asb[3] provides an intensity in the photopic range.

In order to begin to understand the possible mechanisms underlying chromatic and luminance sensitivity we need to consider the neural substrates of the sensory visual system. The primate retino-geniculo striate pathways have been clearly documented[5,6,7]. Two parallel pathways are described according to the differences between their cell structures and functional capacities. The parvocellular (small cell) pathway conveys signals from colour

Figure 3

Threshold graytone 30-2 red.The central reference was 16dB compared with 27dB in the unaffected eye.



opponent retinal ganglion cells (80%). It responds to high contrast stimuli with small receptive field size and weakly to movement. Cell discharges are slow (tonic). The magnocellular (large cell) pathway conveys signals from broadband wavelengths (i.e. not spectrally isolated or "coloured"), representing 10% of the ganglion cells. The remaining ganglion cells project to subcortical areas[7]. The magnocellular pathway demonstrates greater contrast sensitivity compared with the parvocellular pathway and responds to low contrast stimuli with large receptive field size and strongly to movement. Cell discharges are fast (phasic).[1,5,6,7,8]

The parvocellular system (P) projects to layers 3,4,5, and 6 of the lateral geniculate nucleus (LGN) while the magnocellular system (M) projects to layers 1 and 2. The systems appeared to remain relatively separate in their projections and this was supported histologically by their structural differences[7].

This lent credence to an "unashamedly reductionist" theory of functional specialization[8]. In order to describe the terminations of these projections in the cortex it is helpful perhaps to review some nomenclature.

The cortex is a laminated structure that has its projections from the LGN terminating in the striate layers named 2, 3, 4A, 4B, 4Cα, 4Cβ, 5 and 6. These layers occur with the designated smallest number, caudally, through to the highest number rostrally. The visual cortex is further divided by its spread from the primary occipital lobe V1 (Brodmann's area 17), to V2, (Brodmann's area 18), to V3, to V4 and V5, distally, along the convolutions from V1. [7, 8, 10,12]

The P system projects to layer 4Cβ of V1 separating into "blob" and "interblob" layers. The blob or thin stripe pathway conveys information about colour (50% demonstrate double opponency)[7,9,10] and is thought to be only wavelength sensitive rather than colour perceptive[11]. Cells found here are sensitive to luminance at low spatial frequencies but are not orientation selective. The interblob or pale stripe pathway conveys information in response to contours produced by differences in wavelength or luminance and fine detail and form. Cells found here are sensitive to luminance at high spatial frequencies and are orientation selective[7]. In V2 the blobs pass via dark, thin stripes and the interblobs via pale interstripes to V4. Colour perception occurs in V4[11].

There are also connections to V3, an area specialized for orientation and to the Middle Temporal (MT) area, which is the major reservoir for M pathway input. Connections from V4 (P system), then predominantly synapse in the temporal lobe at the rostral superior temporal sulcus and the inferotemporal gyrus (ventral stream)[7,10], where object recognition occurs[12].

The M system projects to layer 4Cα of V1, terminating in layer 4B. All cells here are orientation and motion selective. In V2 passage via thick dark stripes terminates in V3 (orientation centre), V5 (motion and binocular disparity centre) and also to the MT area, before synapsing in the posterior parietal lobe (dorsal stream).

Functionally then, the ventral stream has been associated with object, but not spatial vision and the dorsal stream with spatial, but not object vision[10,12].

The above descriptions understate enormously the abundant inter-connectivity between the P, M systems and the cortical areas V1, V2, V3, V4, V5 and their underlying cell layers of 4Cβ 4Cα and 4B. This necessarily weakens the purely reductionist view of one area or process being responsible for one type of sensitivity/sensation or behaviour[12].

The visual acuity of the patient described above was essentially normal at VR 6/4 VL 6/5. However, contrast sensitivity measurements using the Pelli-Robson Chart demonstrated a low frequency loss: VR 1.65 VL 1.05. Not surprisingly a L RAPD was also noted.

My temptation is to theorize that the parvocellular system with its specialty for chromatic sensitivity and static stimuli at high contrasts, could be seen to be selectively responsible for the difference in dB sensitivity demonstrated with a chromatic rather than an achromatic stimulus. The photopic adaptation created by the HVFA background luminance further supports the notion of a P system response. Piraino & Goodacre's[1] 3 "abnormal" red fields despite normal white fields, especially in a subject who demonstrated borderline contrast sensitivity function, must surely support the contention that a red stimulus is enabling measurement of a different sensitivity in the presence of pathology.

Jean Pollock

Acknowledgments

The patient: for permission to use their visual data. Dr Larry Abel: for scanning the field images.

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Response to a Letter to the Editor: Morabito (Piraino) and Wozniak (Goodacre) have taken the opportunity to comment on the Letter to the Editor by Jean Pollock, concerning their original article on thresholds in red perimetry.

As implied by Pollock in the Letter to the Editor the use of coloured targets in automated perimetry is far from understood. Essentially Pollock is providing a case example to suggest that the use of the chromatic (red) target in automated perimetry may reveal greater defects than those found using conventional white or achromatic targets; and that a possible mechanism for such a finding be related to the parvocellular pathway. While not wishing to comment on the proposed mechanism for any differences, we feel that there are several issues regarding the information provided in the case that warrant further discussion.

Figure 1, the dB readings, outlines the "relative defect depths" for both white and red stimuli in the central 10°. To be able to comment on such "figures", further information is required stating how these "relative defect depths" were calculated. For example when calculating defect depths for white targets the Humphrey Field Analyser uses the subject's actual dB values recorded and compares these to normative data to calculate any differences. For red targets no such normative data is used for the calculation of degree of defect depth. Pollock does not state whether the normative data previously published[1] was used to calculate the values in Figure 1. It is likely that the magnitude of these values for the red targets would be different depending on which method was used. (Infact when the 3 cases mentioned in the published article[1] showing field loss with red targets not apparent with white targets are reviewed; the defect depth calculated by the Humphrey Field Analyser is greater than the defect depth calculated from the normative data. Thus the Humphrey Field Analyser appears to be over-estimating any loss noted with a red target.)

Apart from the above the letter states after Figure 1 that 11/16 (69%) of points show a similar or greater defect depth with white targets. Are white targets therefore considered more sensitive than red? When comparing this data one needs to be mindful of the overall variability or scatter of any automated perimetry threshold measurements. The threshold value obtained on any given measurement location may fall anywhere along the frequency of seeing curve [2,3] and gives rise to a certain amount of variability. This variability occurs during a test as the short term fluctuation, and also occurs between tests known as long term fluctuation. It is believed that this fluctuation is greatest in areas of the field with defects (magnitude in the literature varying from 4dB[4] up to 10dB[5]). As Liebermann and Drake state: "one must not assign too much importance to a 2-4dB change in an isolated value from one test to another."[4] When this variability in the measurement is taken into account, and the data in Figure 1 reviewed, it can be seen that only 7/16 points show a greater than 4 dB difference - where the white target has a greater "relative defect depth". The remaining points show a difference that could be accounted for by variability in the measurement technique alone.

Gray scale printouts are then provided to suggest the opposite; that a greater defect is noted with a red target than a white target. As shown in our article[1] the dB level for red targets will always be lower than that for white, thus explaining the darker "picture" shown in Figure 2. The dB values recorded for red targets and used to create the gray scale printout are those that the white stimulus would have had if the red filter were not in place[6].

What remains to be addressed in the colour perimetry debate is whether or not a chromatic target is able to detect defects that could not be detected with achromatic stimuli. This would be best researched by examining fields that are normal with achromatic targets yet showing apparent defects with chromatic targets. Personal communications that HW has had with clinicians across the globe[7] indicate that chromatic targets

are currently being used to monitor the toxic effects of certain drugs to the visual field, but little has been published to date. Easterbrook who has published widely on the toxic effects of hydroxychloroquine does state that "in some patients with established retinopathy, testing with a red test object and the 10/2 Humphrey perimeter may show larger and deeper scotoma than that determined using a white test object, even when the reduced illumination of the red test object is taken into consideration. However red-field testing has not been standardized with age matched controls and currently is considered a research tool."[8]

Despite what may occur in the future, it is imperative that researchers addressing this issue consider normative data when making comparisons between targets, and also the inherent variability of the threshold measurement; before any conclusions about the effectiveness of chromatic targets and automated visual field testing be made.

In the words of others:(Henson, p.19): "Its (colour perimetry) role in the detection and monitoring of ocular pathologies has not yet reached the level it deserves. This is due to inertia in the perimetric community, lack of commercially available equipment, and lack of standardization."[2]

Helen Wozniak (nee Goodacre)
Josephine Morabito (nee Piraino)

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Response to a letter by R. Duyshart in the AOJ Volume 32.

In response to the letter by R. Duyshart published in the Australian Orthoptic Journal 1996 volume 32. pg 7-8, reference in her letter was made to the article "Comparison of crowded single optotypes with linear acuities in amblyopes" by Williams et al. Although the authors recognise her points regarding the choice of visual acuity tests for screening programs, we feel that the discussion in her letter unfairly reflected the nature of the original research undertaken. We feel that it is necessary to clarify the ideas behind our research.

The aim of the research was to simply assess the validity of the LM test (one we had seen in limited use in the clinical setting) to detect amblyopia, by comparing it to some well known tests which had been more thoroughly investigated as to their validity to detect amblyopia (the SG Linear and SG Singles test). We felt that if the LM test showed an ability to be able to detect amblyopia, then further research would be able to determine if the test was suitable for detecting amblyopia in the younger population undergoing vision screening.

Duyshart states that our results would have had greater clinical significance if the research was performed on children aged 3-5 years, if it included reliability analyses and examined different types of amblyopia separately. Although all these ideas are logical it is often necessary when undertaking clinical research to investigate different issues separately and hence break a research question down into its component parts.

Although the area that this test would clinically be most helpful is in the age bracket of 3-5 years, we were faced with two dilemmas in testing within this age group.

a) Children of this age were not available to use in the numbers that we would need for the study.

b) Children of the age, known to have short attention spans, would most likely find it hard to concentrate for VA testing for 3 tests of each eye in one sitting. We wanted to minimise this confounding factor in our research. Thus the research question investigated the ability of the test to detect amblyopia, not the ability of this age group to perform the test. Further research using the LM test on this population would need to examine the test's reliability.

As far as the amblyopic population tested is concerned, the small sample size made it impossible to consider each type of amblyopia separately, and be able to make valid conclusions from such an analysis.

The shortcomings of the research were clearly outlined in the original paper.

We believe that it is important to design research studies that answer adequately the research question being asked. Often it is necessary to look at a research area and break it up into a series of questions, such that not all outcomes of research have direct clinical significance. This was infact clearly stated in our conclusion: "What remains to be established is whether the LM test is as simple as the Sheridan Gardiner test to comprehend in the population where it is most needed: screening large numbers of perverbal children who can not perform linear tests." p 27

The suitability of this test for vision screening is yet another question. The test does appear to be easy to understand, quick to perform and sensitive to amblyopia. It may be of benefit in the detection of amblyopia or when loss of concentration or lack of co-operation with other tests makes it difficult to perform screening. Although there are many other alternatives for testing vision in the orthoptic setting, these tests may be costly and more difficult to perform by vision screeners who have more limited training and knowledge of amblyopia.

We were not stating that because our research showed the LM test to have a favourable result in the detection of the crowding phenomena that it should be implemented, rather that its availability should be noted, its effectiveness as a test should be considered, and of course after further research its use considered.

Megan Turtle (Williams),
Tiffany Wong,
Helen Wozniak (Goodacre)

The Patricia Lance Lecture 1997 Heredity and Strabismus

This highly distinguished invited lecture was delivered by Robin Wilkinson at the 54th Annual Orthoptic Association of Australia Scientific Conference in November 1997, Sydney, Australia.

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Abstract

Strabismus has long been thought to be hereditary. Studies of the increased prevalence of strabismus in the siblings of probands and studies of the higher concordance of strabismus in monozygotic as opposed to dizygotic twins support this observation. The genetic basis is not known. However, it does not appear to follow a simple Mendelian pattern of inheritance and is thought to be polygenic and multifactorial with environmental factors playing a part. The Strabismus Inheritance Study in Tasmania has been established to discover the gene(s) responsible for hereditary strabismus. The study will examine 300 affected sibling pairs and their nuclear families.

Key Words:
Hereditary strabismus, multifactorial inheritance, sibling pairs.

Introduction

Strabismus has long been noted clinically to have an apparent hereditary nature in some families. The higher prevalence of strabismus in the siblings of affected individuals (probands) together with a positive family history in many cases, have led clinicians to believe there is an underlying genetic predisposition to the disorder. In recent years there have been major advances made in the understanding of the genetic basis of many eye diseases, however the underlying genetic basis of strabismus is unknown. One of the major reasons for this lies in the fact that there are many different phenotypic expressions of strabismus. Strabismus can be congenital or acquired, constant or intermittent, manifest or latent, convergent, divergent or vertical. This variability of phenotypic expression is matched by the many different theories on the aetiology of strabismus. Worth theorised that the underlying basis of strabismus was in fact a defect affecting the fusion mechanism[1]. Chevasse on the other hand theorised that the underlying problem was an interference with the development of conditioned binocular reflexes leading to deviation of the eyes[2]. The more modern approach theorises that some patients may have a congenital lack of ability to develop central fusion[3] (eg congenital esotropia), whereas in others the development of strabismus is due to an interruption to the immature visual system which relies on the proper development of sensory and motor factors to produce binocular single vision[4].

Evidence That Squint Is Hereditary

In the general population the prevalence of strabismus is said to be from 4% to 5%[5,6]. There are several clinical indicators that lend weight to the hereditary nature of strabismus.

1.Prevalence of Strabismus in the Siblings of Probands

If the prevalence of strabismus amongst the siblings of probands is higher than in the normal

population then the defect is likely to have an hereditary component . Several studies have confirmed this. Czellitzer found that 15% of the siblings of affected individuals also had strabismus[7], while a large epidemiological study in America found that for any pair of siblings the odds of esotropia for one sibling more than doubled if the other sibling had esotropia. If one sibling had exotropia then there was only marginal evidence of association[8].

2.Parent/Offspring Correlation

Richter conducted a large study of siblings and their nuclear families and found that the incidence of squint amongst siblings increased if one or more parents were affected[9]. She found that if one or both parents of an index case were affected then about 30% to 50% of the siblings were also affected. If both parents were unaffected then only 20% to 30% of the siblings of a proband were affected. Thus the incidence of strabismus within a nuclear family increased if one or more parents were affected.

3.Presence of a Positive Family History

A familial trait is one which appears more frequently in the relatives of a proband than in the general population. Pratt Johnson reviewed 254 consecutive cases of simple concomitant squint and found that 65% of these probands had a positive family history of strabismus[10]. Schlossman and Priestly found a positive family history in 48% of patients with strabismus, while a Greek study by Chimonidou found a positive family history in 55%[11,12]. An interesting study in Western Australia, looking at children attending for follow up after squint surgery (this is a biased population in that the squints were all severe enough to require surgery), found that in those who had surgery before one year 72% had a history of squint in the parent generation and 24% in the grandparent generation. If the surgery was after two years, then 61% had a positive history in the parent generation and 30% in the grandparent generation[13].

4.Twin Studies

Twin studies are of great value when estimating the hereditability of a disorder. Monozygotic (identical) twins share exactly the same genes or DNA, whereas dizygotic (nonidentical) twins only share the same 50% genetic material that normal siblings do. If a pair of twins both have strabismus then they are said to be concordant for that trait. The influence of hereditary factors can be assessed by comparison of the concordance rates for monozygotic and dizygotic twins. If the concordance rate of a particular trait approaches 100% in monozygotic twins, while the same rate in dizygotic twins is lower, then the trait is said to have an hereditary component[14].

It should be noted that sometimes monozygotic twins show differences due to factors that are not genetic. For example, the effects of external factors during intrauterine life eg the conditions of fetal circulation, the effects of birth trauma/anoxia are not always the same for the two fetuses[15]. Several twin studies have been undertaken to investigate strabismus with the hope of differentiating hereditary and environmental factors, with varying results.

Waardenburg pooled his twin statistics with those in the literature and found a concordance rate in monozygotic twins of 81% , while dizygotic showed only 9%[16]. Richter, in a smaller study, found concordance in 12 pairs of monozygotic twins of 92% and in 27 pairs of dizygotic twins of 26%[9]. A Chinese study found concordance of 77% for monozygotics and 22% for dizygotics[17]. At the other end of the scale Reynolds found monozygotic concordance of 42% and dizygotic concordance of 20%[18]. A study by DeVries of 17 pairs of monozygotics found concordance for squint in 47%[19]. Obviously the statistics can vary enormously, and selection bias, small sample populations, and errors in assessing the true zygosity where monozygotic twins are in fact dizygotic can all lead to erroneous statistics for the concordance of strabismus. Another factor affecting the concordance is the quality of the clinical evaluation itself.

With consideration of these variable results Magli et al conducted a 5 year study of monozygotic twins and compared concordance with the same number of dizygotic twins. They concluded that there was a 72% concordance for monozygotic twins with esotropia as compared with 22% for dizygotic[20]. This lends definite supports to a genetic basis for strabismus.

5. Racial Genetic Variation

If racial differences occur for a given trait then this may provide supportive evidence that a trait is hereditary. It has been shown that there is a significantly lower rate of esotropia in blacks than whites. Eustace examined the prevalence of strabismus and refractive error in second generation West Indian children born in Birmingham and compared the data with previous data collected on white children and found that exotropia was four times more common in black children and that myopia was also more prevalent[21]. In summary, the clinical observation that there appears to be a genetic

component in the aetiology of strabismus is supported by the clinical data from several studies.

Genetics

In order to understand the possible genetic and molecular basis for the inheritance of strabismus and the pattern or model for inheritance, a basic knowledge of genetics and genetic terminology is necessary.

The human cell nucleus contains 46 chromosomes, arranged in 23 pairs based on their morphology on karotyping. The 23rd pair are the sex chromosomes XX in a female and XY in a male. Thus there are 22 pairs of autosomes and one pair of sex chromosomes. The cell nucleus also contains two different types of nucleic acid - RNA which contains the sugar ribose and is called ribonucleic acid and is found mainly in the nucleolus and the cytoplasm, and DNA which contains the sugar deoxyribose and is called deoxyribonucleic acid. It is found mainly in the chromosomes. Thus a chromosome is made up of DNA. Arranged linearly along the chromosomes are discoid elements called genes. These genes are the basis of the development of hereditary characteristics. A gene is in fact a tiny strand of DNA and its primary action is to control a specific cell function by governing the synthesis or manufacture of a specific protein. The DNA of each gene makes a specific RNA by a process called transcription, which then directs production of a protein by a process called translation. Thus DNA directs the synthesis of RNA and RNA is translated to protein[22].The location of a gene on a chromosome is called a locus. Each pair of chromosomes has two copies of a given gene and these corresponding copies of the gene at a given locus on a pair of chromosomes are called alleles. The clinical expression of a gene is called a phenotype. There are approximately 100,000 genes which code for specific proteins. Currently there is a world wide study 'The Human Genome Project' which aims to provide a map of all the human chromosomes and should be completed by the year 2000. A genetic defect or trait may be defined as any condition caused by an error in the genetic material DNA This error may occur as a result of a mutation. A mutation is a permanent change in the constitution of a gene which results in modification of its action. It may have a positive, negative or indifferent effect. If there is no effect of a given variation the term polymorphism may be applied. All genetic disorders may be grouped into three broad categories[23].

1.Chromosomal abnormalities: caused by structural rearrangement of the DNA in the chromosome, or an incorrect complement of chromosomes (eg. trisomy).

2.Molecular alterations: in gene structure: eg point mutation or deletion of a part or all of a gene.

3.Multifactorial abnormalities: where genetic and environmental factors play a part. In many traits multiple genes interact and hence the term polygenic is used.

Models of Inheritance

The discussion of the role of heredity and strabismus must necessarily include a model for its inheritance. The traditional Mendelian models fail to fit the observed pattern of inheritance for strabismus[24]. The three main Mendelian patterns are:

1. Autosomal dominant inheritance
2. Autosomal recessive inheritance
3. Sex linked inheritance

1.Autosomal dominant inheritance

An autosomal dominant disorder occurs when a trait is manifested when only one of the two copies of the gene (alleles) is abnormal. Autosomal dominant inheritance is characterised by:

1.Transmission from one generation to the next. Phenotypically normal parents do not transmit the phenotype to their offspring.

2.The risk of an affected individual having an affected offspring is 50%.

3. Males and females are equally affected.

4. All forms of transmission are observed eg from either male or female to offspring of either sex. With reference to heredity and strabismus, the clinical data on family studies does not support simple autosomal transmission in that often both parents are clinically normal.

2.Autosomal recessive inheritance

An autosomal recessive disorder means that each parent must have a copy of the abnormal gene and two abnormal copies are needed to produce a trait. Autosomal recessive inheritance is characterised by:

1.Individuals in one generation in a single sibship eg brothers and sisters are affected, and it does not occur in previous or subsequent generations.

2.The risk to offspring of a couple who are carriers is one in four or 25%.

3.Males and females are frequently affected.

4.Cosanguinity in parents can be an indicative factor (eg marriage between first cousins).

With reference to strabismus the clinical data on family studies does not support autosomal recessive transmission, in that just as parents may be unaffected, a certain proportion are affected.

3. Sex linked inheritance (X-linked dominant and X-linked recessive inheritance)

There is no evidence for the strabismus pattern being X-linked. This form of inheritance pattern implies differences in inheritance and transmission according to sex. On the contrary, clinical data have shown that males and females are equally affected. There is also no data to support a difference in transmission from either sex.

From the evidence above it is clear that strabismus does not follow a simple Mendelian pattern of inheritance and that we need to look to a different model to explain its familial aggregation.

The model which best fits hereditability of strabismus and in fact many other inherited disorders is called multifactorial inheritance.

Multifactorial Inheritance

The multifactorial model is now the most commonly held theory of the inheritance of strabismus. The term is used to explain the inheritance pattern in which both genetic and environmental factors are thought to play a part. The inherited genes are thought to work *additively* and there may be no single major error in the genetic information but rather a combination or additive effect from either or both parents, such that only when a threshold is reached and exceeded then the combined effect is seen as an abnormality. When a disorder is caused by the additive effect of more than one gene it is termed polygenic. In multifactorial inheritance the expression of a disorder in an individual is determined by:

1. How many of the gene factors responsible for that disorder are present in each parent.

2.What quantities of each factor are transmitted to each offspring[25].

3. Environmental factors.

Given the evidence that strabismus is hereditary with a polygenic multifactorial inheritance model it is important to assess both the contribution made by environmental factors and the genetic risk factors that contribute to the additive threshold effect that appears to cause the phenotypic expression of the trait.

Risk Factors

There are three main risk factors reported from clinical studies that are thought to contribute to the so called additive effect[14].

1.A positive family history and more specifically when either parent has esotropia.

2.If parents are normal but there is between them very low vergence ability.

3.If either parent has significant hypermetropia.

The first two points have been covered in earlier discussion. However refractive errors and their genetic significance may prove to be of vital importance in the inheritance of strabismus.

Refractive Errors

The refractive state of an eye is the result of a combination of different optical components such as corneal curvature, corneal thickness, depth of the anterior chamber, thickness of the lens, anterior and posterior lens curvatures and axial length. None of these components are constant and they vary between individuals so that total refraction may vary according to the combination and interaction of these components[26].

There are three main theories on the causes of refractive errors[27]:

1.The biological theory in which it is postulated that all errors of refraction are due to the way in which the components of the eye combine.

2.The use/abuse theory which explains the onset of myopia as an adaptation to the use of the eyes in prolonged closework.

3.The emmetropisation theory proposed by Van Alphen which postulates that the eye is self focusing and emmetropia is produced by cortical and subcortical control of the tonus of the ciliary muscle and ametropia by factors which interfere with this mechanism. This theory proposes that a stretch factor is exerted on the sclera by the ciliary muscle which originated from the scleral spur[28].

As with strabismus there is clinical evidence to support a genetic basis for refractive errors given that environmental factors can modify the effect.

Twin studies have established that refraction as a whole and its major individual components axial length and the powers of the cornea and lens, are genetically determined. Sorsby found concordance for monozygotic twins to be 71% for refractive errors while for dizygotic twins it was negligible[29]. Hereditability was high for corneal curvature, posterior lens surface and axial length.

A study by Young and Leary found that there was a significant child-parent refractive error correlation[30]. This is especially true for myopia where studies have found that when both parents are myopic the prevalence of myopia in the offspring is at least three times higher than when neither parent is myopic[31].

A study by Hegmann et al looked at three different groups and their nuclear families;

1. 118 families with esotropia.
2. 27 families with exotropia.
3. 163 random families.

They examined the clinical data in relation to the implication that hypermetropia and myopia are a primary factor in the aetiology of strabismus. They found results consistent with the general clinical observation that hypermetropia tends to occur with esotropia and myopia tends to occur with exotropic patients. They concluded that the correlation between hypermetropia and esotropia, and myopia and exotropia was statistically significant and suggested that there were gene differences among the refactive errors and that these may be directly implicated in the aetiology of strabismic phenotypes[32].

Finally a British study by Anker looked at a sample population of 829 infants with a positive family history of strabismus and screened them between the ages of 6 months and three years for refractive errors. They concluded that infants with a positive family history of strabismus are four times more likely to develop significant hypermetropia and that 20% of these hypermetropes will develop a strabismus.[33]

Thus it appears that refractive errors and their inheritance are in some way related to strabismus and its inheritance.

Environmental Factors

A large multidiciplinary study in America examined data to identify risk factors for strabismus in a group of children followed from gestation to age seven years[34]. They found the following factors to be significant:

1. The risk of strabismus increased with low birth weight.
2. The risk of esotropia increased with increasing maternal age until 34 years.
3. Maternal cigarette smoking during pregnancy increased the risk of offspring developing strabismus.

A study in the USA has shown that a dietary intake of Omega 3 fatty acids during the first months of life was important. It showed that at 36 months children fed with human milk containing omega 3 fatty acids had better random dot stereopsis and letter matching skills than formula fed children (the latter being a poor source of this nutrient)[35]. A further study showed that Omega 3 fatty acids played an essential role in retinal development[36].

Thus it can be seen that while there is evidence of a genetic predisposition to strabismus there have been shown to be risk factors and environmental factors that cause an additive effect and a threshold is reached whereby the defect expresses itself clinically as strabismus.

The Strabismus Inheritance Study

The Strabismus Inheritance Study (SIST) commenced in Tasmania in February 1997. The aim of the study is to identify the genes responsible for hereditary concomitant strabismus.Given the clinical evidence that there is a definite hereditary component and that the model which best fits this inheritance pattern is multifactorial, the study will examine in detail 300 affected sibling pairs and their nuclear families, looking at the penetrance of the phenotype in each family and also at possible environmental factors. The study will compare parent/offspring similarities for several clinical measures as well as collecting DNA for analysis.

The SIST team is comprised of three orthoptists who have designed the clinical protocols and who clinically examine all participants. The genetic investigation and laboratory supervision will be managed by an ophthalmologist, and a molecular geneticist.

The study has compiled a list of over 250 sibling pairs in Tasmania from case records in private practices and from public hospital records and will target these families initially.

Methodology

Polygenic inheritance complicates genetic mapping because no single gene locus is responsible for producing a trait. The SIST study has adopted the affected sibling pair method of analysis. This method has been shown to be the method of choice where multifactorial inheritance is the model. The aim is to compare the DNA of a large number of affected sibling pairs looking for genetic markers. Genetic markers are patterns of DNA sequences with a known location on a chromosome and these are mapped and compared, looking for identical segregation of a genetic marker with a disease phenotype that is not simply random segregation.

The closer together two genes are located on a chromosome when they are compared the higher the chance of their being inherited together.

The initial genetic mapping will target a major subgroup of strabismus - congenital esotropia. Investigation will then be carried out on other major subgroups, which may or may not be linked to the same gene.

Two recent ophthalmic genetic discoveries offer an interesting insight into the search for the strabismus gene and where the initial search may be best directed. The location of one of the gene mutations responsible for primary open angle glaucoma has proven to be the same gene responsible for juvenile glaucoma located on chromosome 1. The location of the gene mutation responsible for 16% of age related macular degeneration is known to be on chromosome 2 and is the same gene responsible for the more severe early onset Stargardt's macular dystrophy.

There is no candidate gene for strabismus, but it would appear sensible to look at congenital infantile esotropia initially, as it is arguably the most severe form of strabismus in that it appears in the first six months of life and binocular single vision is rarely if ever achieved. It is also phenotypically a very distinct subgroup of strabismus.

Tasmania, and in particular the north - west coast of Tasmania, provides an excellent population for such a genetic study, in that many families have lived in the area for generations and can be traced back to common ancestors in many cases. This is called the 'founder' effect and provides a gene pool with less external genetic influences and an ideal population to target initially, as far fewer families with the identical genetic disorder may be needed by the laboratory. It is hoped that by early next year a pilot genome search for genetic markers in congenital esotropia may be commenced.

Clinical Examination

Much of the difficulty in studying the hereditability of strabismus lies in the use of varying definitions and measures of strabismus. The study aims to clinically examine, refract and classify every member of each nuclear family. One of the strengths of the study lies in the fact that the same team will examine every family and strict criteria are adhered to for examination and classification.

A comprehensive history is obtained from each family member with particular attention given to the factors outlined previously as risk and environmental factors. The questionnaire looks at all the siblings and the maternal history during pregnancy, birth details, milestones and strabismus history of those affected. This history is sent to each family prior to the clinical appointment and then thoroughly checked by a member of the team on the day.

Clinical examination is then carried out on all members of the nuclear family wherever possible. The following factors are investigated by the same examiner:

1. Visual acuity using the logMar chart where possible.
2. Near and distance cover test with prism bar measurements.
3. Ocular motility.
4. Investigation of binocular vision.
5. Stereoscopic acuity using the Randot and Titmus tests.
6. Measurement of prism fusion range.
7. Measurement of the AC/A ratio (heterophoria and gradient method)
8. Fixation.
9. Refraction using an autorefractor.

Cycloplegic refraction is carried out on those aged 10 years and under.

A comprehensive classification has been designed wherein each strabismus is classified into a very specific subgroup. One of the difficulties has been to classify the strabismus in the subgroup it belonged to *before* treatment as this is the relevant classification genetically. This is essential for the laboratory once genome studies are commenced given that more than one gene is thought to be responsible for strabismus and there are so many different types of strabismus. It may well eventuate that some subgroups will be traced to the same gene or that previously apparently unrelated subgroups may prove to be genetically linked. However, initially, it is essential to classify each subgroup so that they are phenotypically identical within the group.

This comprehensive examination will provide valuable information on the penetrance and phenotypic expression of a gene in each family. Strict guidelines have been established to define the range of normality and thus where abnormality begins. The entire examination takes approximately one hour per family, and the family is required to attend for one visit only.

DNA Collection

DNA is typically isolated from blood samples for genetic analysis. While this is the ideal and more robust method of collection this has only

been used in adults in the SIST study. Buccal mucosal swabs are a non-invasive method of DNA collection. The DNA is contained in cells which are plentiful in the mucosal lining of the mouth. The sampling is obtained with plastic cyto brushes which are gently rubbed on the inside of the cheeks and stored in a fixative until sent to the laboratory. The main disadvantage of this method of DNA sampling is that the yield of DNA is not as high as with blood samples and the yield drops if the DNA is not extracted within three weeks. Blood samples can be kept indefinitely. The yield from 1 ml of blood is about 100 micrograms of DNA and the average yield from one buccal swab per patient is approximately 50 micrograms per swab in total. However this yield will be sufficient given that current genome searches require much less DNA than in the past. Mucosal DNA can be immortalised to make it more stable long term, however this is cost prohibitive given that it is a relatively easy procedure to repeat, and the study is dealing with a relatively young population.

The study is now piloting an updated DNA collection kit, using smaller cyto brushes and small containers called epindorfs which are easier to transport and easier to process, giving a higher DNA yield and only requiring two swabs per person.

Clinical Observations To Date

Having completed the first year of what is anticipated will be a three year study by the SIST team, it is relevant to look at an overview of 70 affected sibling pairs and their nuclear families, 8 of these being families with monozygotic twins. Several observations are of clinical interest.

1. Monozygotic Twins

As previously mentioned, if a trait shows a high concordance in monozygotic twins it provides evidence the trait is hereditary. This study has examined 8 pairs of monozygotic twins to date and found 100% concordance for strabismus, subgroups, and refractive errors.

2. Presence of a Positive Family History

In the 62 families of affected sibling pairs (ie excluding monozygotic twins) there was a positive family history of strabismus in 74% of families.

With regard a positive history: 39% were positive on the mothers side, 16% on the fathers side and 19% on both sides.

This is a higher incidence of a positive family history than in previous studies of consecutive concomitant squint mentioned earlier (65% being the highest). One possible reason for this is the fact the study is looking at families with a higher penetrance, that is, with two affected siblings. Another reason is the detailed history taken by the SIST team, as quite often a definite family history is revealed with more detailed questioning.

3. Is Strabismus Phenotypically the Same in Families?

In the 62 families under discussion some interesting variations occurred within families. Only 60% of families had sibling pairs from an identical subgroup while 40% had affected pairs who fell into different subgroups. Of the 60% with identical subgroups there were 14 pairs of congenital esotropes. The 40% with different subgroups had a variety of combinations of subgroups including those thought to be unrelated eg congenital esotropia with fully accommodative and intermittent divergent squint with convergent microtropia.

Thus it appears that familial strabismus is not phenotypically the same in every family and can be expressed in a variety of subgroups within a nuclear family.

4. Risk Factors

The parents were examined for risk factors thought to contribute an additive effect in the inheritance of strabismus. By far the most significant risk factor in this group of affected sibling pairs was the positive family history of 75% previously mentioned. The incidence of parental strabismus was 26%. The risk factor of a low vergence range in either parent appeared in 23% of families where the limit of 40pd of convergence and 16pd of divergence was taken to be the normal range,and 25pd of convergence to 10/12 pd of divergence or below these limits taken as a reduced vergence range.

There was an overall incidence of parental refractive error of 49% where 2 diopters of hypermetropia and 1 diopter of myopia were taken to be significant. Interestingly, the breakdown shows more parental myopia than hypermetropia with 30% being myopic and 19% being hypermetropic.

Thus it appears the above risk factors do play a significant role in the inheritance of strabismus.

Significance Of SIST

The discovery of the genes responsible for strabismus and the investigation of the nuclear families and their clinical status will provide new and valuable information that will assist in the management and treatment of strabismus.

1. The discovery of the gene(s) will lead to a better understanding of the pathophysiological processes involved in the development of strabismus. This is absolutely essential if we are to progress in the understanding and management of strabismus.

2. An understanding of the underlying genetic basis of strabismus and the genes involved may lead to reclassification of strabismus, as previously apparently unrelated phenotypes are shown to have the same genetic basis. This may lead to the adoption of different management procedures.

3. Genetic identification of the molecular basis of strabismus may give rise to new therapeutic interventions eg medication, or changes to diet or environment to minimise or negate the effects of the trait.

4. The ability to accurately predict its occurrence will lead to earlier intervention.

5. Improved and more economical screening techniques may be adopted once high risk individuals are more readily identified.

Conclusions

Strabismus has been shown by the clinical research data available to have an hereditary component. However the pattern of inheritance does not fit a simple Mendelian model but rather is thought to be polygenic and multifactorial with environmental factors playing a part. The Strabismus Inheritance Study aims to identify the genes responsible for hereditary concomitant strabismus. It is the first study of its kind in the world and the information obtained on the genetic inheritance of strabismus and the familial phenotypic penetrance of strabismus has the potential to change the way we classify, interpret and manage strabismus in the future.

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Visual Assessment in a Developmentally Disabled Population: Marsden Eye Survey

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Abstract

The aim of this study was to assess the visual function, ocular conditions and general diagnoses of 328 Marsden residents who were seen at the Marsden Eye Clinic between July 1985 and July 1997.

A questionnaire was sent to all parents and guardians regarding past medical history, pregnancy and birth history as well as family history. Medical records and Eye Clinic notes were reviewed and correlated with the questionnaire.

Results showed that a significant number (9%) of the Marsden residents are blind (with both eyes open) and many visually impaired, with best vision less than 6/12 (49.9% - 53.3%). Strabismus, nystagmus and refractive error feature predominantly in this group, as well as organic pathology, such as cataract and corneal scarring.

This remarkably stable population has a significantly greater incidence of visual impairment than the general Australian population.

Keywords:
Developmental delay, visual impairment, general disability, strabismus, refractive error, ocular abnormality.

Introduction

Marsden Hospital, located in Westmead, Sydney, is a residential centre for mentally and physically handicapped patients. When it opened in November 1969, it was the first purpose-built hospital in New South Wales for disabled children. Many of the original children, now adults, have remained at Marsden since its opening and reflect a remarkably stable population. These Marsden patients make up part of the 1.86% of Australians who have an intellectual disability[1].

Method

This current study, the Marsden Eye Survey (MES), commenced in July 1985, focuses on the visual function, ocular conditions and general diagnoses of 328 residents assessed at the Eye Clinic. Questionnaires were sent to all parents and guardians regarding background information on each patient at Marsden seen in the 12 year period. Information regarding pregnancy, birth, early childhood and illnesses, and family history were included in the questionnaire. The response rate was 53%. As the average age of the patients is now thirty, many parents have since passed on or were simply unable to be contacted.

Table 1.
The percentage of affected individuals in each category.

Category of General Diag.	% ofpopulation
Prenatal	5.5
Perinatal	8.5
Postnatal	9.5
Genetic	29.5
Unknown	47.0

Table 2.
The most common general conditions.

Condition	Number of Cases
Down syndrome	35
Postnatal infection	20
Rubella	13
Prematurity	11
Birth trauma	8
Angelman syndrome	6
Tuberous sclerosis	5

Table 3.
Predominant associated conditions found in the MES.

** Over half of this group were deaf because of Rubella.*

Condition	% of population
Epilepsy	43.0
Spastic Quadriplegia	12.8
Microcephaly	8.8
Deafness *	4.3
Hydrocephalus	3.0

Results

The youngest resident at Marsden is 13 years of age and the eldest is 45 years, with the average age being 30 years, which follows on from that found by Dr Graham Henry in his study of the patients at Marsden 17 years ago, when the average age was 13 years[2]. Since our study began in July 1985, only 13 residents have died, either from the progressive nature of their disease or pneumonia, and one in an accident. The number of males outnumbered the number of females at 1.8:1. 1995 figures of the Australian Institute of Health and Welfare show that, generally the number of Australian males with a primary disability type of acquired brain injury outweighs that of females by 1.8 : 1.[3]

14.6% of the Marsden patients had a family history of mental retardation and 1.8% had consanguinous parents. 73% were mobile, 22.5% wheelchair bound and 4.5% unknown as it had not been recorded earlier in the case notes and they are no longer at Marsden.

When the general disability diagnosis was known, it was grouped into one of five categories:

a)Prenatal causes: teratogenic, intrauterine infection (e.g. Rubella), trauma, structural abnormalities of the brain.

b)Perinatal i.e. at the time of birth and up to 4 weeks after birth: prematurity (<36/40), birth trauma and asphyxia, kernicterus, intracranial haemorrhage, hydrocephalus.

c)Postnatal, i.e. 4 weeks after birth: infections such meningitis, encephalitis, near-miss sudden infant death syndrome (SIDS), septicaemia, trauma, reactions to immunisation.

d)Genetic: chromosomal disorders (e.g.Down syndrome, fragile X), familial conditions (e.g. Tuberous sclerosis)

e)Unknown aetiology.

The incidence for each category is given in Table 1.

The high percentage in the unknown category reflects the fact that only if the diagnosis was definite, i.e. well documented in the history, was it grouped according to the cause. Further to this, the most common causes of developmental delay amongst the Marsden residents are summarised in Table 2.

Interestingly, 6 patients over the last few years have been detected as having Angelman (Happy Puppet) syndrome whereas previously their diagnosis was unknown. This reflects the greater sophistication of chromosomal testing in recent years, particularly the emergence of the Fluorescence In-Situ Hybridisation (FISH) technique, which looks for a specific gene mutation. It is now known that Angleman syndrome is caused by a mutation of chromosome 15.

It has been observed throughout the study that, in addition to their primary diagnosis, many of the Marsden patients have other significant disabilities. The most common of these conditions are summarised in Table 3.

The visual function and ocular condition of all residents was assessed. Visual acuity was tested using the Catford Drum, Sheridan Gardiner Single Letters and the linear chart. Visual acuity standards were divided into three levels:<6/60, 6/18-6/60, and 6/12 or better. Vision was recorded right eye (RE) and left eye (LE) if possible, otherwise with both eyes open (BEO). Approximately half of the patients had best recorded vision as 6/12 or better in either eye (RE = 50%, LE = 50%) or with BEO (46.7%). A study by Jan Erby[4] showed 50.2% of the same group to have best recorded vision at this level. Between 27.3% (RE) and 30.6% (LE) had monocular vision of 6/18-6/60 and 38.1% with BEO (Erby 43.2%). Visual acuity of less than 6/60 was present in 22.7% RE, 19.4% LE and 15.2% BEO, which is quite different from that found by Erby (6.7%). Since her survey, 18 years ago, many of the patients have had either

progressive neurological impairment, progressive eye disease including glaucoma and cataracts, or ocular trauma including that from self-injurous behaviour.

It must be noted that observation is a major tool used when assessing a population such as that of Marsden. Concentration and co-operation are very limited, making formal testing extremely difficult at times. Because of the difficulty in quantifying accurate visual acuity objectively, vision was also recorded subjectively as either good or poor. "Good" visual acuity meant functional vision allowing the person to move around, recognise people or accurately reach for objects, and "poor" meant blindly having to reach for objects and feel their way when walking. 82.6% had good vision, this group containing 38 people whose vision could not be tested due to lack of co-operation. 16.8% had poor vision, including 9% who had no perception of light with both eyes open. Reasons for poor vision and blindness are summarised in Table 4.

Congenital anophthalmos was present in 2 cases and 6 enucleations were also recorded. 9% of the Marsden population is blind. Current Australian Bureau of Statistics figures show that 0.09% of the Australian population is blind, with a further 1.42% having partial visual impairment[5]. These numbers highlight the fact that reduced vision is significantly more prevalent in this handicapped population. Squint was evident in nearly half of those seen (49.4%), with slightly more exotropias than esotropias (25.9% : 22.9%). 25% of patients had nystagmus with 18% being horizontal in direction. Eye muscle movement disorders were minimal, with no pattern detected.

Refraction and fundoscopy were also performed, using either Cyclogel 1% or Mydriacyl 0.5%. Refractive error was not corrected if it fell within plano to 3.00D of hypermetropia or myopia, or <2.00D of astigmatism, which is a judgement based on clinical experience. Glasses given for those lower amounts generally are not worn and often no obvious visual benefit is gained. It has been observed many times that a patient with uncorrected high myopia or hypermetropia is very mobile and accurate when reaching for an object.

Results showed the highest incidence of refractive error to be myopia and astigmatism (Table 5). 29% of patients have either previously worn or currently wear glasses, 61% have never worn them and 10% are unknown. It should be stressed that there is a need to check the glasses being worn as it is not uncommon to find someone else's glasses being worn by mistake.

Cause of Poor Vision/Blindness	Number of Cases
Cortical visual impairment	15
Optic atrophy	8
Congenital abnormality *	8
Cataract	5
Rubella oculopathy	4
Retrolental fibroplasia	2
Glaucoma	2
Trauma	1
Optic nerve hypoplasia	1
Retinal dystrophy	1

Table 4.
Causes of poor vision and blindness found in the MES.

**Congenital abnormality includes microphthalmos and coloboma.*

Refractive Error	Right Eye	Left Eye
Plano to +/-3.00D	67.4%	68.2%
Myopia>-3.00D	22.2%	21.6%
Hypermetropia>3.00D	10.4%	10.2%
Astigmatism<2.00D	26.3%	26.5%
Astigmatism>2.00D	20.7%	18.6%

Table 5.
Percentages of type of refractive error found in the MES.
** Note: Some patients had both a spherical and cylindrical error combined - therefore totals are greater than 100%*

Throughout the orthoptic and ophthalmological assessments, may ocular abnormalities were noted, the six most common of which are summarised in Table 6 below.

Ocular Abnormality	% of Population
Cataract	17.7
Corneal scarring	7.9
Keratoconus	5.8
Glaucoma	4.8
Retinal detachment	3.4
Uveitis	1.0

Table 6.
The most common ocular abnormalities found in the MES.

Often, corneal scarring was secondary to previous cataract surgery, which led to uveitis and then a phthisical eye. This was particularly evident in the settling of Rubella oculopathy. Keratoconus was noted in 5.8% of patients. Interestingly, unilateral cataract formation has been observed to develop in 2 patients who had keratoconus and episodes of hydrops. It is considered by the authors that keratoconus is often a non-specific finding reflecting the fact that severe rubbing of the eyes causes a significant amount of trauma to induce keratoconus and occasionally a secondary cataract. It is also noted that acute hydrops is often a recurrent problem in this group of developmentally delayed patients.

Conclusion

Overall, the MES showed that this remarkably stable population of developmentally delayed individuals has a significantly greater incidence of visual impairment than the general Australian population. Often the general diagnosis is elusive, but with improved genetic testing, more patients and their families are able to be given a cause for the disability. This is an important consideration as often siblings are at the age of having children of their own. If genetic factors are involved, there are obvious implications of having further affected children.

Approximately 50% of patients had vision that was less than 6/12, but subjectively the majority had good functional vision (82.6%). Squint is a common feature, as is nystagmus. Myopia and astigmatism are the most prevalent refractive errors but glasses are infrequently prescribed.

The ocular status of each patient at Marsden has a significant impact on their daily living skills. Those with poorer vision need extra help and there is a stronger emphasis on stimulating other senses. Music therapy, ball rooms, touch rooms, carpeted walls and tactile toys are an integral part of those wards with the partially sighted and blind residents. At Marsden a homelife environment exists. This means day outings, picnics, and a variety of special functions. Those patients who are better sighted will visually gain more from these activities, but with extra intervention from staff, all patients are helped to ensure maximal gain. When assessing the vision of this population, the visual needs of each individual must be taken into account, and the functional level considered. Acknowledging the overall daily visual requirements of each patient is necessary, and realising that the effects of visual loss can sometimes be compensated for in other ways.

To know the ocular and visual status of a population such as that of Marsden is essential. Maximising the quality of life in the developmentally delayed is often the responsibility of the health professional. To quote Dr Graham Henry :"At grass roots level, if you cannot see you cannot feed or educate yourself and the earlier this is known to helpers [carers], the better".[2]

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A Test of Visual Function Applicable to Children with Severe Cognitive Impairments.

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Abstract

Many tests of vision have been developed to ensure the accurate measurement of this important sense. Some of these tests have been modified to facilitate testing in cases of special need, yet despite these modifications some severely cognitively impaired children are unable to comply with the requirements of visual acuity testing. This paper reports some of the evaluation findings of a new test of visual function designed to facilitate testing of severely intellectually and/or multi-handicapped children. Results from trials of two phases of the new test are presented. Phase 1, validation trials with 96 cognitively normal children and phase 2, evaluation trials with 73 intellectually, multi-handicapped children are presented. Phase 1 trials indicated the new test demonstrated strong positive correlation with standard clinic tests and had good internal validity. Phase 2 indicated the new test was significantly more successful in facilitating testing of the target population than standard clinic tests.

Key words:
Vision test, computer generated test, intellectual and multi-handicap, functional vision.

Introduction

Vision is the major sensory modality through which knowledge is gained with a large area of the brain devoted to analysis of visual information. The eyes are unique as sensory organs in having two types of receptor, thus facilitating analysis of the many facets of visual stimuli presented to them.[1,2]

Many tests have been developed to assess components of vision in order to enhance understanding of the mechanism, and to facilitate early detection of visual dysfunction. These tests are concerned with the three basic components of vision:

- The minimum visible
- The minimum resolvable
- The spatial minimum discriminable

More recently the impact of contrast on these three components has also been considered. The minimum resolvable is the component most commonly tested in the clinical setting and can be further subdivided into minimum separable ie the ability to distinguish two objects as separate, such as Teller acuity or Landolt's C[3-5] or minimum legible which involves the higher processing task of recognition, such as Snellen acuity or Kay pictures[3,6]. Predominantly, test development has reflected methods of obtaining the most accurate levels of visual acuity, necessitating sensitivity to the smallest changes in function. Criteria manipulated included: optotype legibility both in terms of letter similarities and contrast; optotype shape ie grating, C or letter; and progression of optotype size ie arithmetic or geometric.[3,7]

Test development has also reflected changes in the reasons for investigating vision. The need to test pre-verbal children resulted in a series of tests

with optotypes modified as pictograms,[6,8,9] or gratings.[5] Some tests are modified in terms of response mode, such as, matching or visual pointing.[5,6,10] The need to assess low vision patients for the prescription of optical aids resulted in the development of charts more sensitive at lower acuity levels which could be easily manipulated in terms of test distance, such as the logMAR chart.[11] More recently test developers have considered modifications necessary to facilitate testing of multi-handicapped children who are unable to respond to conventional tests of visual acuity.[12-17] Similar to tests for the pre-verbal paediatric population these tests have been developed by modification of conventional tests of vision. The modifications being the subject matter of the optotype and response mode to be used.

Based on clinical observation and reports in the literature some children are unable to respond to these modified tests. Such children have severe cognitive disabilities and or severe multi-handicaps. The problems testing these children include:

The low interest level of the optotypes used.

The lack of cognitive ability to identify or match letters and shapes.

The lack of fine motor skills to provide a pointing or matching response.

The lack of verbal development to provide a response.

Objective tests of vision such as VER have been suggested as providing a measure of vision independent of the patient's ability to respond or co-operate with the test.[18] As an increasing number of researchers acknowledge, there is a lack of relationship between measured acuity and functional vision.[19-21] Consequently the application of such objective test measures is not appropriate as an indication of the level of vision available for daily function. Knowledge of visual function is essential to the development of educational and skill training programs. The test of vision described in this paper was developed to facilitate testing children with severe cognitive impairment and/or severe multi-handicap to provide an indication of vision which might be applied to daily function.

Method

The data presented in this paper relate to the validation trials of the pilot test program, VizAssess and analysis of evaluation trials of the subsequent revised program VizTest. These are computerised tests of visual function.[22]

Subjects:

1. VizAssess validation: 96 children attending a paediatric ophthalmology clinic in Melbourne. The children were of varied intellectual ability with an age range of three to twenty one years. Forty seven subjects were male and forty nine were female.

2. VizTest evaluation: There were three subject populations.

- Seventy three intellectually/multi-handicapped children attending a Special Development School and School for the Visually Impaired in Melbourne, Australia.
- Forty two intellectually/multi-handicapped children attending a school for the deaf blind and two special schools in Sydney, Australia.
- Twenty eight children who were intellectually/multi-handicapped, severely visually impaired or both, attending a blind school and two special schools in London, UK.

Procedure:

1. VizAssess validation: Visual acuity was assessed using the computer test and an appropriate clinical test of acuity. Testing was carried out independently by two examiners, with each examiner blind to the results of the other. Visual acuity on the computer test was considered to be the smallest sized optotype the subject correctly recognised or could follow on screen on at least two occasions. All testing was commenced at 1 metre and if no response was obtained to the 6/30 optotype the test distance was reduced to 0.5 metre and the subject re-tested. Subjects who needed to move closer than 0.5 metre had their results recorded as less than 6/60. Clinical testing was undertaken with a test of acuity appropriate to the age and ability level of the subject. Most subjects were tested with the Medmont Visual Acuity tester: however, some children unable to cope with the logMAR format of the Medmont were tested with Kay Pictures or Sheridan Gardiner Singles. Clinical test distances varied from 6 metres to 0.5 metre depending on the needs of the subject. Clinical testing preceded computer testing in all cases. Computer testing began with module 2 followed by modules 1 and 3. Module 2 was selected first as this facilitated quick determination of optotype size for the subject, this size was then used with modules 1 and 3. If a subject recognised the optotypes at the selected size more easily on subsequent modules the optotype was reduced in size.

2. VizTest evaluation: Test protocol for VizTest was the same at all test sites. Subjects were tested with module 1 followed by module 2, and if a response was gained to module 2 then module 3 was tried. Testing was performed at 1 metre monocularly if the subject cooperated and BEO if

not. If test distance was reduced the result was recorded as less than 6/60. Clinical assessment of acuity varied between test sites.

• Melbourne: Subjects from the Special Development School were assessed using the computer test and Kay picture test. The order of test presentation was alternated between subjects. Kay pictures were presented at three metres, testing being done monocularly where possible or with both eyes open. Both tests were administered by the investigator. Students from the school for visually impaired were unable to co-operate with conventional clinic testing.

• Sydney: Subjects were assessed with the computer test and Catford drum(at half a metre), Cardiff acuity cards or Sheridan Gardiner singles (at 3 metres). VizTest was administered by the investigator and the clinic test by the orthoptist normally working at the location. Each examiner was blind to the results of the other.

• London: Subjects were tested with the computer test by the investigator and the clinical assessment of vision was taken from the most recently recorded clinical measure. The computer test was performed at one metre if a 15 inch display monitor was available and at 0.75 metre if a lap top computer display was used. The reduction in test distance was proportionate to the decrease in display screen area.

The computer tests.

VizAssess was described in a previous paper reporting the results of trials with severely multi-handicapped children[22].

VizTest consists of 3 modules. Each module enables the selection of images from a drop down menu.

Module 1: Visual attention. This module is designed to combine movement with a colourful image to attract the patient's attention. A range of pictures is available including commonly seen images, cartoon images and patterns. Each image is presented within an area 68 x 79 cm and can be made to "jump" from one side of the display to the other. This module is not intended to indicate any measured level of acuity, only to determine if visual attention can be gained.

Module 2: Vision category. This module incorporates the presentations of the three modules of VizAssess. A range of colour images can be presented on screen in one of three sizes. The images selected were considered to be of familiar content to the target subjects and included animals, plants, food, vehicles and people.

• Size. A coloured image is displayed in the centre of the screen at the smallest size of 28 x 23 mm. The image can be increased in size to 56 x 46 mm and 112 x 92 mm. The images do not display specific outline widths or contrast elements similar to Snellen design and outside dimensions are similar to Snellen equivalents of 1/18, between 1/36 to 1/60, and > 1/60 respectively. The pictures are not intended to measure precise visual acuity, rather to indicate a level of function.

• Movement. A coloured image can be selected in one of the three sizes described above. This image is moved horizontally across the screen in either direction. The speed of movement can be varied from fast 4.4cm/sec to slow 1.4cm/sec.

• Display. A coloured image can be selected in one of the three available sizes and displayed at the top, bottom, left or right of the screen. Display position is randomly generated and not predictable.

Module 3: Vision measure. This module has two components, acuity and contrast.

• Acuity: Black on white line drawings are displayed on screen. The drawings are created of line widths equivalent to Snellen line widths of 6/12, 6/24 and 6/60 optotypes. Overall size is not equivalent to Snellen design with each image appearing within a square 9.5 x 9.5 cm. A key square is presented for matching purposes and a blank square can be displayed to test reliability.

• Contrast: A line drawing is presented at high or low contrast in a square 9.5 x 9.5 cm. When the image is selected a random generator places the image to the right or left side of the screen.

Results

VizAssess validation.

Data were divided into groups based on pathology. Subjects whose pathology would be favoured by a near test distance, ie those with nystagmus and myopia, were analyzed separately to one another and to the rest of the paediatric population. Data analysis is based on data from each eye independently. The results of correlations between each module and the standard and between modules are summarized in Tables 1 - 3.

Modules	Spearmans rho	p value
M1: Standard	0.721	0.0005
M2: Standard	0.661	0.0015
M3: Standard	0.661	0.0015
M1: M2	0.959	0.0001
M1: M3	0.959	0.0001
M2: M3	1.0	0.0001

Table 1

Sub group Myopes

N = 24

Table 2

Sub group Nystagmus

N = 43

Modules	Spearmans rho	p value
M1: Standard	0.744	0.0001
M2: Standard	0.765	0.0001
M3: Standard	0.744	0.0001
M1: M2	0.867	0.0001
M1: M3	0.955	0.0001
M2: M3	0.913	0.0001

Table 3

Remainder of paediatric population

N = 125

Modules	Spearmans rho	p value
M1: Standard	0.803	0.0001
M2: Standard	0.790	0.0001
M3: Standard	0.798	0.0001
M1: M2	0.924	0.0001
M1: M3	0.963	0.0001
M2: M3	0.962	0.0001

Correlation between each module and the standard test was positive and of moderate correlation. The normal paediatric population demonstrated the strongest correlation between all modules and the standard, with the myopic group demonstrating the weakest correlation. Correlation between computer modules was positive and strong in all groups.

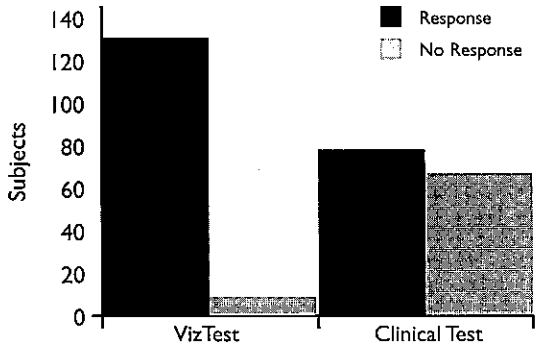


Figure 1 *Number of subjects demonstrating a response to VizTest and the clinic tests (n = 143)*

Figure 1 indicates that 132 (92%) of subjects were able to respond to VizTest where as only 78 (54%) were able to respond to a clinical test of vision and the difference between test response rates was significant *(chi^2 = 4.49, p = 0.034)*. Response rates did vary between trial sites and these data are presented separately in Figure 2.

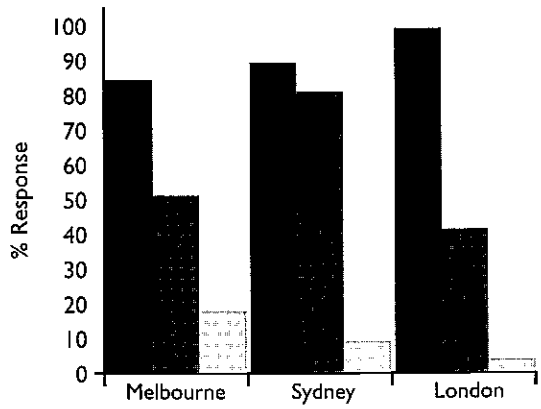


Figure 2 *Responses to VizTest and Standard test at three test locations (n = 143)*

VTest
Clinic
No Response

The Melbourne and London populations demonstrate a much higher response rate to VizTest than to the clinical test and while the Sydney population also demonstrates a higher response rate to VizTest, the difference between response rates to VizTest verses clinical is much less. The difference in response rates for the Melbourne and London populations was significant to a two tail paired t test *(Melbourne t = 6.86, p = 0.0001 and London t = 5.7, p = 0.0001)*. The difference in response rate for the Sydney population was not significant to two tail paired t test at the 0.05 level *(t = 1.78, p = 0.0831)*.

Analysis of the combined data from the three trial sites of subjects able to respond to both VizTest and a standard clinic tests is reported in Table 4.

Discussion

Results were recorded as a vision category, the categories used were based on the WHO disability classification of > 6/12 = normal; < 6/18 = moderate low vision; and < 6/60 = severe low vision (legal blindness in some countries)[23]. This method of recording was chosen as construction of the test shapes did not allow complete equivalence to the Snellen optotypes; and vision category is an appropriate indicator of functional vision which is the purpose of this test.

The original test VizAssess demonstrated good positive correlation when compared to standard clinical tests. Results of analysis from the paediatric population indicate that the computer test is internally consistent across all groups. The weakest correlation was that between modules 1 and 2 in the nystagmus sub-group at 0.867, the strongest being between modules 2 and 3 of the myopic sub-group at 1.0. The remaining correlations were all within the 0.9 range (Tables 1 - 3). Correlations to the standard tests were positive but weaker. The normal population gave the strongest correlations at 0.8 for module 1 and 0.7 for modules 2 and 3; the nystagmus group reduced to 0.7 for all modules; with the myopic group being 0.7 for module 1 and 0.6 for modules 2 and 3. This loss of correlation may be related to the pathology. The myopic group not being heterogeneous in terms of correction, under corrected myopes may have been advantaged by the near distance of the computer test.

A further confounding factor in the paediatric trial population was the use of a range of vision tests for the standard test. This population was heterogeneous in terms of intellectual capacity

and the vision test used in the clinical setting was chosen to suit the child's level of function. The standard test therefore varied in response requirement, complexity of optotype, type of test presentation and test distance. In the clinical setting where this test will ultimately be used, test procedures are not standardized between clinical practices. Comparison of VizAssess to a range of commonly used clinical tests was deemed to provide useful data whilst acknowledging the introduction of a further variable. Analysis of the correlations achieved with the computer test indicate a high p value (.0001 in most cases, Tables 1 - 3) supporting the test being a suitable indicator of visual function in a paediatric population, with the exception of under corrected myopes.

Analysis of evaluation trial data for VizTest indicates that this revised test has successfully attracted attention and facilitated responses from the target population. Results from both the Melbourne and London trials indicate that a significantly larger number of children were assessable using VizTest than with a standard clinic test [Melbourne 63 (86%):37 (51%) and London 27 (96%):12 (43%)], this is seen in Figure 2. Whilst the Sydney trial indicated more children were assessable using VizTest the difference between groups was less 38 (88%):35 (81%). The difference in response rate may reflect some of the differences in the study populations. In both the Melbourne and London trials subjects were in an environment where routine ocular examination did not occur. The Sydney subjects attended schools where an orthoptist provided regular assessment (although this may not be more than an annual assessment).

The subjects in this study are from a population who are noted for liking routine and familiarity of environment. The Sydney subjects were tested in the office of the school nurse and in the presence of the school nurse which gave a high level of familiarity to this test situation. The majority of Melbourne subjects were tested on a withdrawl from class basis, they were tested without the support of a familiar person in an administrative area of the school which would not be well known to them. The London subjects were tested in the presence of a familiar person but not in a particularly familiar environment. In the Sydney study the standard test was administered by the consulting orthoptist who was familiar to at least some of the children, where as in the Melbourne study the standard test was presented by a person unfamiliar to the subjects. The standard vision for the London subjects was obtained from a clinical record and had generally been assessed by a person unfamiliar with the children and often in a strange environment (hospital clinic).

	VizTest	Clinic
Visually Impaired	12	0
Full Sight	15	44

Specificity = 1.0 Sensitivity = 0.44

Table 4 *Specificity and sensitivity of VizTest (n = 71)*

Another variation between studies was the test used as standard. Both the Melbourne and London populations were tested with tests requiring a matching response based on recognition of a pictogram or letter (Kay, Sheridan Gardiner singles). These tests are commonly used in the clinical environment to test multi-handicapped children. In addition, a number of subjects in the London study had been recorded as not previously testable. The Sydney population was tested with the Catford drum, a test which does not require recognition of a pictogram or letter, and requires an ocular following response. The Catford drum is a test which has been criticized in terms of the accuracy of assessment and because of the noise associated with the small motor moving the drum. This noise might be attracting the child's attention resulting in a response, leading to a false assumption that the response is indicative of vision. VizTest provides no such aural cues.

The trials conducted with VizTest with severely intellectually/multi- handicapped children at three trial sites were not intended to be validation trials. However using the combined data from the three trial sites VizTest demonstrated excellent specificity with moderate sensitivity. Based on this data VizTest tends to under-detect visual impairment. Module 3 of VizTest was the only module to give some quantification of vision, in terms of a vision category. Data from module 3 was correlated with data from the standard clinic tests for those subjects responding to both tests. Correlation with the Melbourne and London populations were moderate and positive (0.597 and 0.634 respectively). The Sydney population demonstrated very weak correlation (0.011). See Figures 3, 4 and 5.

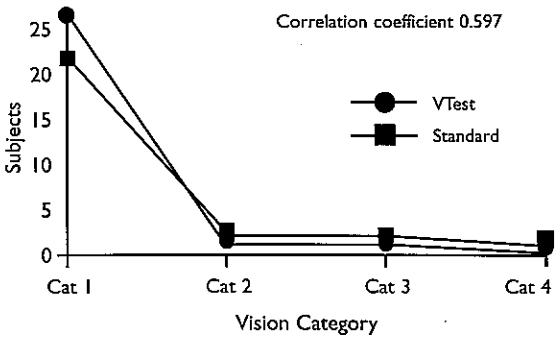
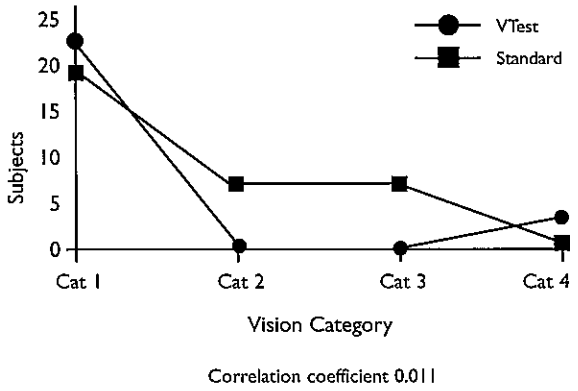


Figure 3

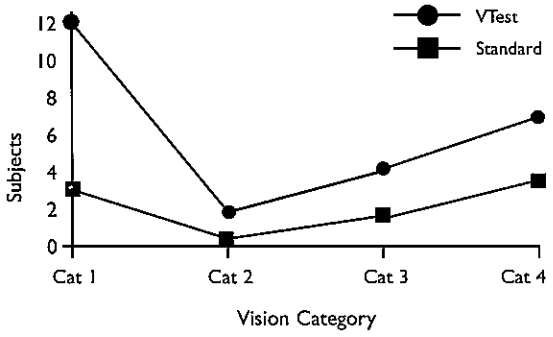
Correlation of vision category VizTest / Standard, Melbourne

This difference between the Sydney and other populations may be a result of the test used as standard. Tests based on optotypes of Snellen design were used in Melbourne and London (Kay, Sheridan Gardiner) where as the standard test with the Sydney population were predominantly the Catford drum or Cardiff Cards. The module of VizTest which was used to indicate vision category in this series of trials was module 3 which was closer in cognitive requirement to Kay or Sheridan Gardiner. The use of module 3 as the indicator of vision category also reduced the number of subjects for whom data was obtained, consequently reducing statistical viability.



Correlation coefficient 0.011

Figure 4 *Correlation of vision category VizTest/Standard Sydney*



Correlation coefficient 0.634

Figure 5 *Correlation of vision category VizTest/Standard London*

Conclusion

In conclusion the initial test, VizAssess, demonstrated that the computer did provide a test medium which was of interest to the target population. The ability to move the stimulus optotypes was successful in gaining subjects' attention and facilitated visual pointing as a response mode when subjects did not have the fine motor skill to finger point or verbal skill to name an optotype. Further trialing with a general paediatric population supported VizAssess as a good predictor of visual category when compared with a standard test of acuity. The revised computer test VizTest on preliminary analysis successfully attracted attention and facilitated responses from the target population and provided a moderately reliable indicator of vision category. These data support the use of computer presentation with colourful and familiar pictures as a target of interest to severely intellectually/multi-handicapped children. The data provide further support of the use of movement to facilitate responses from non verbal multi-handicapped subjects. Validation testing with cognitively normal subjects is continuing.

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The Assessment of Impaired Visual Functioning Due to Cataract

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Abstract

The aim of this study was to investigate the relationship between objective and subjective measures of visual function in patients booked for cataract surgery. Visual acuity is the traditional objective measure of vision and yet this clinical measure can be difficult to equate with patients' symptoms. One hundred subjects who were booked for first eye cataract surgery were recruited from the Royal Victorian Eye & Ear Hospital. Snellens visual acuity was recorded. Subjective visual function was assessed by a questionnaire related to performance of everyday tasks. Results show no straightforward relationship between self-reported visual functioning and visual acuity. No correlation was found between visual acuity in the eye to be operated on (worse eye) and the visual functioning questionnaire. However, a moderate correlation (*rho= -0.403, p= 0.0001*) was found between visual acuity in the better eye and visual functioning. Visual functioning is more closely related to visual acuity in the better eye but cannot be fully explained by it. In assessing the timing of cataract surgery, self-reported visual functioning may be a more important indicator than visual acuity alone.

Key Words:
Cataract, visual acuity, visual functioning.

Introduction

Cataract is the most common cause of blindness and visual impairment in the elderly population, with 95% of the population over 65 years of age having lens opacities[1]. In 1996, 11% of Australia's population were over 65 years old. By 2042 this is expected to double to 22%[2]. With the advances of cataract rehabilitation of the last decades, cataracts are being operated on at ever earlier stages, with the number of operations performed doubling in the last 10 years[2] and cataract surgery now being the second most common major surgical procedure performed[3]. The advances in technology, together with the aging of the population are resulting in an ever increasing strain on the health care budget.

Opacities form in the lens as part of the normal aging process[1]. It is not until these opacities interfere with vision and have an impact on everyday activities that surgical intervention is required. Judging the most appropriate time for surgery is important for the patient so that lifestyle can be maintained while unnecessary surgery is avoided.

The symptoms of cataract are essentially a disturbance, then a diminution and finally a failing of vision[1]. Visual disturbances are varied and include myopic shift, possible astigmatism change, monocular diplopia, polyopia, colour vision change, reduced field of vision and disabling glare[4]. In the clinical setting impaired vision due to cataract is traditionally measured by visual acuity. The limitations of visual acuity as a measure of visual function are well known. Visual acuity uses high contrast black letters on a white background in a well lighted environment at a standard distance. This can be difficult to translate

into functioning in the 'real' world where objects are of different shapes, sizes, contrast and colour. A measure of visual acuity does not necessarily reflect a patient's visual functioning or the symptoms associated with cataract. Indeed it can be difficult to equate a patient's symptoms with the objective measure of visual acuity. Numerous studies[5-11] have shown no straightforward relationship between visual acuity and a patient's symptoms. Although finding a linear trend with visual functioning decreasing as acuity decreases, correlations between the two have been only in the poor to moderate range.

Functional impairment due to cataract may be evaluated by the use of questionnaires. In 1981, Bernth-Petersen[5] devised a Visual Functioning Index. In 1992, Mangione et al[10] developed the Activities of Daily Vision Scale and in 1994 Steinberg et al[11] produced the VF 14. Questions are asked about everyday activities such as reading, driving and watching television, resulting in a score which gives a level or grade of functional disability.

This study aimed to further investigate the relationship between objective and subjective measures of visual function in patients booked for cataract surgery.

Method

Subjects

The subjects were 100 patients, 41 males and 59 females ranging in age from 48 years to 91 years with a mean age of 74 years. All were attending the outpatients department, Royal Victorian Eye & Ear Hospital and had been booked for cataract surgery. Subjects were excluded if they had a previous intraocular lens, myopia greater than 5 dioptres, were booked for a simultaneous ocular procedure or did not have enough English skills to complete the questionnaire.

Apparatus

The questionnaire chosen to assess impairment of visual functioning was the VF 14, an Index of Functional Impairment In Patients with Cataract, developed by the Cataract Patient Outcome Research Team[11] in 1994. Questions relate to everyday activities including seeing steps, writing cheques, playing table games, taking part in sports, cooking, reading small print, doing fine handiwork, reading a newspaper or a book, daytime driving, night driving, reading traffic signs, reading large print and recognising people. Subjects were asked to rate the degree of difficulty they had with each activity because of their vision, with 0 being inability to do the activity and 4 no difficulty at all with the activity. A score out of 100 resulted, with 0 being inability to do any of the activities because of vision and 100 being able to do all the activities without difficulty. Subjects were also asked about their overall satisfaction with their vision and a Satisfaction Score between 1 and 4 was given, with 1 being very dissatisfied with vision and 4 very satisfied with vision. Visual acuity was measured on the Snellen chart, a standard instrument used in the clinical setting.

Procedure

Patients attending clinics for their pre-operative assessment were invited to participate. Informed consent was obtained and the subject was interviewed. Interviews took up to ten minutes to complete. The most recent recording of Snellens visual acuity using the patient's current glasses was taken from the medical records. Demographic details, ocular comorbidity and cataract type when available, were also gained from patient records.

Characteristics	% of Participants
Gender	
Male	41
Female	59
Country of Birth	
Australia	51
Other	49
Ocular Comorbidity(29%)	
ARMD	11
Glaucoma	10
Diabetic Retinopathy	2
Other	6
Cataract Type	
Cortical	8
Nuclear	25
PSC	14
Mixed	20

Table 1 *Demographic and Ocular details*

Design and Analysis

In this study there were 3 variables, the VF 14 questionnaire measured on a 100 point scale, visual acuity converted into a scale of 1 to 10 and Satisfaction Score measured on a 4 point scale. Each of the variables was treated as ordinal data and the correlation coefficient used was Spearman's Rho with p values set at 0.05.

Results

Demographic and ocular details are shown in Table 1.

Visual acuity in the eye to be operated on ranged from 6/9 to PL (perception of light) with a median acuity of 6/24. Visual acuity in the

Table 2 *Distribution of Visual Acuity in Worse and Better Eyes.*

Visual Acuity	Worse eye (Eye Booked for Cataract Surgery) N = 100	Better eye (Eye Not Booked for cataract surgery) N = 100
6/4		2
6/5		8
6/6		16
6/9	17	25
6/12	17	26
6/18	16	11
6/24	12	5
6/36	16	4
6/60	7	1
less than 6/60	15	2

better eye ranged from 6/4 to less than 6/60 with a median acuity of 6/9 (Table 2).

The VF 14 scores ranged from 13 to 100 with a mean of 71 (Table 3). The Satisfaction Scores are shown in Table 4.

Table 3 *Distribution of VF 14 scores pre-op.*

VF 14	No.
0-10	0
11-20	2
21-30	0
31-40	4
41-50	10
51-60	12
61-70	17
71-80	19
81-90	21
91-100	15

Satisfaction and VF 14 Scores

Using Spearman's Rho a statistically significant relationship was found between Satisfaction Scores and VF 14 scores (*rho = 0.631, p = 0.001*).

Visual Acuity and VF 14

Using Spearman's Rho no relationship was found between visual acuity in the eye to be operated on (worse eye) and the VF 14 (*rho = -0.123, p = 0.2215*). However, a statistically significant relationship was found between visual acuity in the better eye and the VF 14 (*rho = -0.403, p = 0.0001*).

Table 4 *Distribution of Satisfaction Scores pre-op.*

Satisfaction Score	No. of subjects
1 (very dissatisfied)	28
2 (dissatisfied)	55
3 (satisfied)	17
4 (very satisfied)	0

Visual Acuity and Satisfaction Score

Using Spearman's Rho no statistically significant relationship was found between visual acuity in the worse eye and the Satisfaction Score (*rho = -0.136, p = 0.1768*). However, a statistically significant relationship was found between visual acuity in the better eye and the Satisfaction Score (*rho = -0.244, p = 0.0152*).

Discussion

Satisfaction Scores & VF 14

This present study found a strong correlation between the two subjective methods of assessing visual function, the Satisfaction Score and the VF 14 scores. This agrees with Mangione et al[10] who also found a high correlation of 0.70 using the Activities of Daily Vision questionnaire. Steinberg et al[11] found a moderate correlation of 0.34 using the VF 14 questionnaire. A good correlation between the two subjective methods of assessing visual function would be expected.

Visual Acuity and Visual Functioning (VF 14)

This study found no statistically significant relationship between visual acuity in the eye to be operated on and visual functioning as measured by the VF 14. This would suggest the ability of subjects to perform daily living tasks is not determined by the vision in the worse eye and it is not until the vision in the other eye becomes impaired that these activities are affected. This finding agrees with Steinberg et al[11] who also found no correlation (*r = -0.08*) between vision in the worse eye and visual functioning as measured by the same questionnaire. In contrast Elliott et al[8] found a statistically significant relationship between vision in the worse eye and reading vision (*r =0.46*) and mobility (*r = 0.52*).

This present study demonstrated that vision in the better eye is significantly correlated with visual functioning (*r = -0.403*), that is, as visual acuity becomes worse so does the subjective reporting of visual functioning. Although the correlation is statistically significant it is only of moderate strength, suggesting that some subjects function well in spite of poor acuity, while others with good acuity have poor functioning.

These findings agree with Steinberg et al[11] who found a correlation of *-0.27* between vision in the better eye and visual functioning. Bernth-Petersen[6] also found a positive relationship between visual acuity in the better eye and reading vision.

Elliott et al[8] found a moderate correlation between vision in the better eye and reading ability (*r = 0.42*) but no significant relationship between vision in the better eye and mobility.

Other studies have compared visual functioning with binocular visual acuity as this presents a more normal state for the patient. Lundstrom et al[9] and Mangione et al[10] found weak but significant relationships between binocular vision and visual functioning (*r = 0.2825 and r = 0.37 respectively*). These studies suggest that binocular visual acuity equates with better eye visual acuity. A blurred image in one eye may well affect visual comfort rather than visual acuity and some patients report clearer vision if one eye is closed.

Visual Acuity and Satisfaction Score

This study found no relationship between vision in the eye to be operated on and Satisfaction Score but did find a statistically significant but weak correlation (*r = -0.244*) between vision in the better eye and Satisfaction Score. This suggests that some people with poor acuity are satisfied with their vision, while others with good acuity are not. Steinberg et al[11] found a similar non-significant relationship with vision in the worse eye, but also found no relationship between vision in the better eye and satisfaction (*r = -0.01*).

Conclusion

This study demonstrated that there is indeed a relationship between visual acuity and subjective visual functioning but the relationship is not straightforward. Satisfaction with vision and visual functioning are more dependent on acuity in the better eye but the relationship is only a moderate one. Poor visual acuity can be reflected by poor visual functioning, but not in all cases. Likewise, not all patients with good acuity function well. When assessing the need for cataract surgery in the absence of lens induced disease, the patient's visual functioning and their satisfaction with vision may be more important indicators than a measure of visual acuity alone. A patient's satisfaction with the outcomes of cataract surgery depend on the level of pre-operative impairment. If there is very little pre-operative impairment, outcomes are likely to be disappointing. Schein et al[12] linked poor outcomes of cataract surgery with a pre-operative VF 14 score of 90 or greater. Patients need to understand the reason for their surgery and the likely outcomes, in order to make an informed decision regarding the need for such surgery and whether the likely benefits outweigh the potential risks, costs and inconvenience of surgery.

These 100 subjects will be interviewed 3 months following cataract surgery and outcomes will be measured by Satisfaction Scores and VF 14 as well as the traditional measure of visual acuity.

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The Use of Predictive Factors in Stroke Rehabilitation

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Abstract

Empirical and clinical predictive factors are measures that can be used to aid in the prognosis of rehabilitation outcomes. It is important for orthoptists to be aware of the background of these factors when assessing patients for rehabilitation. An extensive review of 54 papers on the topic revealed that although predictors can aid clinical decision making, they cannot currently be used exclusively without clinical considerations. An explanation of the most common predictors is included to help clinicians make decisions about their patients' ocular health with their general prognosis in mind.

Key Words:
Prognosis, cerebro-vascular accident, incontinence, training.

Introduction

Factors ranging from continence to cognition have the potential to predict rehabilitation outcomes in stroke survivors. Good prediction of outcomes can be useful for orthoptists when setting treatment goals, calculating clinic budgets, counselling patients or in justifying treatment options. Negative predictors indicate a poor prognosis for rehabilitation, which may be reflected in a longer length of rehabilitation, worse discharge destination (institution rather than home), poor ambulatory status or low scores on functional ability tests. Positive factors indicate better outcomes.

Following the scrutiny of 54 papers concerning predictive factors in stroke rehabilitation, spanning the last 40 years, this review comments on the past and present state of these factors. Their development, limitations and uses are outlined.

Early Predictors

Although research written before the 1970s did contribute significantly to the current body of knowledge about predictors, it was usually substandard in the area of data analysis and design[1]. That literature is now mainly of historical significance, as the data is not comparable due to the anecdotal nature of the papers. Despite large group numbers and adequate information, the research often degenerated into the authors' intuitive feeling about which were the best predictors.

The historical review[2-6] found that at the end of the 1960s, the most common predictors were: bladder incontinence, age, onset-admission interval, mobility status, blood pressure and place of rehabilitation. The proceeding sections follow the fate of some of these factors to discover how they stood up to more rigorous investigation. Several new factors have emerged and they will also be examined. The predictors at the beginning of the discussion have the most support in the literature and those towards the end have the least (see Table 1.).

Bladder Incontinence

Pre 1970s literature supported bladder incontinence as a useful predictor[2,3,6] and this

review has found bladder incontinence to be the most popular predictor with 15 studies[7-21] finding it had predictive value and none disputing this value. One paper[21] revealed initial urinary incontinence to be the second most powerful predictor in the study, with a negative correlation of 0.45 with Barthel ADL (Activities of Daily Living) scores at six months after the stroke. This may have been a biased sample however, as the study lost half its original 162 subjects to death or follow up. Barer's 1989 study[9] had a more representative distribution[15,16], with only 27% of the 363 patients dying in six months and also proved incontinence to be a powerful predictor.

Similarly, Oczkowski and Barreca[19] found that continence had good predictive value for subjects being discharged home, however their use of medians as predictors made the results difficult to compare with other studies or to be used clinically. In contrast, the classification tree of Falconer and co workers[12] is impressive in its simplicity. This tree uses four variables to predict 88% of favourable outcomes, but only needs the first two, toilet management and bladder management to predict 80% of favourable outcomes and 68% of unfavourable outcomes.

Reasons cited for the good predictive ability of bladder incontinence have included non-causative factors such as association with organic medical changes, poor motivation, emotional problems and difficulties in transportation and management.[6] However, Barer[9] was unconvinced about this non-causative hypothesis and contended that lack of awareness due to apathy and urgency of micturition due to bladder instability, were the major problems in stroke patients. Borrie and co workers[11] asserted that inability to communicate the need to urinate, due to dysphasia, was the major cause.

Motor Function

The early work of Bourestom[3] and others[2] first sparked interest in the analysis of motor function as a predictor. As this area is not as clearly defined as age or urinary incontinence, there have been a variety of measures of motor function cited in the literature, creating confusion as to which actually work as predictors. The most obvious sign of motor dysfunction in stroke survivors is the degree of hemiparesis.[22] Feigenson and co workers[13] discovered that although only 74% of subjects with severe hemiparesis on admission were discharged home, 86% of those with mild or moderate weakness returned home. Hemiparesis combined with the presence of homonymous hemianopia has proved to be a strong predictor,[13,23-25] possibly because it infers a larger lesion.[25] Olsen[26] found Barthel

Predictor	For	Against
bladder incontinence	15 [7-21]	0
motor function	8 [10,13,22-27]	0
age	11 [10,15,16,19,21,23,25,28-31]	3 [7,8,32]
ADL score	6 [19,20,29,35-37]	2 [15,29]
perceptual disorder	7 [7,10,13,16,22,29,43]	1 [19]
cognition	5 [13,38-41]	1 [19]
cortical damage	4 [47-50]	1 [51]
onset admission interval	5 [7,13]	3 [19,21,31]
social factors	3 [15,29,30]	2 [15,52]

Table 1. *Most popular predictors and the number of research papers supporting them.*

ADL scores were slightly better predictors of rehabilitation outcome, but the ease of measurement of hemiparesis favoured its use. Upper limb mobility has also been found to have predictive value,[10,27] as has postural control on admission.[19,27]

Although mobility has strong support as a predictor, and few detractors, its value is limited by the varied methods of measurement.

Age

Age was a popular predictor in early literature,[4,5] although not all agreed.[2] More recent studies[21,28] have found that older patients were less likely to have a high Barthel ADL score on completion of rehabilitation. Further studies, using place of discharge as an outcome measure, found older people less likely to be discharged home than younger people1.[9,28,29] However, Lehmann and co workers[29] did find this related to the availability of family support. Age has also been a useful multivariate factor in predictive equations[10,23,30] and further support for age as a predictor has come from Haerer[25] and others.[15,16,31]

Not all studies, found age had predictive value[7,8,32] although Anderson and associates[7] suggested that age may have been excluded from their study due to its high correlation with a stronger predictor, as it was in Bourestom's study.[3]

Regarding the reason for its predictive ability, Oczkowski and Barreca[19] asserted that age was linked to co morbidity, which resulted in worse outcomes, however they provided no evidence. In contrast, Kalra[28] went to great lengths to determine whether it was co morbidity or another factor that made age a negative predictor. He found that although subjects over 75 years old had a higher incidence of degenerative disabilities, deficits due to the present stroke and the initial Barthel ADL scores were comparable with their younger counterparts. Shah et al[30] similarly found that the effect of age on outcome was independent of other factors.

ADL Scores

Activities of daily living (ADL) indices score the ability of patients to care for themselves physically.[33] These scores are used in rehabilitation fields to diagnose, gauge improvement and predict outcomes. Most commonly used is the Barthel ADL Index, a taxonomy of functional activities in the areas of self care, bowel and bladder sphincter control and mobility.[34] This score therefore encompasses many of the predictive factors that have been previously studied and gives them equal weighting.

Wade and co workers[21] claim to have been the first to relate ADL ability after rehabilitation with initial data, however this was investigated 16 years earlier by Bourestom.[3] Their research[21] showed high correlation ($r = 0.48$) between Barthel ADL scores on initial assessment and six months after stroke. Granger et al[35] found that a Barthel score of 60 was pivotal in predicting independence. Initial Barthel scores have also been found to be the most powerful factors in several predictive equations.[30,36] Another commonly used ADL score the Functional Independence Measure, was found by Oczkowski and Barreca[19] to be the most powerful of the factors they used to predict location of discharge after stroke rehabilitation, and others[37] have also found it had good predictive ability. In contrast, neither Lehmann and associates,[29] nor Jimenez and Morgan[15] found predictive value in ADL scores at admission.

Associated Neurological Deficits

In 1977, Feigenson and co workers[13] found perceptual function (neglect, denial, apraxia), cognitive function and motivation to be such strong predictors that they concluded that these were the "only" predictors and that all others were superfluous. Unfortunately, although this was a large, well-implemented study, percentages were used to compare outcomes and the researchers used personal judgement in deciding whether these factors were clinically significant, with no account of statistical significance. Although not making such presumptuous statements, other more reliable studies[38-41] have agreed that cognition is a well correlated predictor, although this has been disputed.[19] Problems in comparing studies or using these factors clinically arise because cognition must be measured in different ways to accommodate its various components.

Perceptual disorders also have support as predictors, however whether they have a causative role or are merely an indication of severity of function is contentious.[42] Stern et al[43] showed that hemisensory losses were found predominantly in patients with poor functional outcomes but used three unrepresentative tests. Anderson and co workers[7] included perceptual disorders in their list of predictors but did not indicate which perceptual disorders they assessed. Kaplan and Hier[22] found a significant correlation between perceptual tasks scores and self care status on discharge. Lehmann and co workers[29] and Lincoln and co workers,[16] used the Weschler neuropsychological tests to analyse perceptual factors as predictors and found good correlation, although Lehmann and co workers[29] did not state follow-up times. This is relevant as Barer and Mitchell[10] found perceptual factors had predictive value three months after hospital admission, but not after six months. Oczowski and Barecca,[19] did not find perceptual factors to be predictors at any time.

Amount of Cortical Damage

The relationship between the extent of cortical damage secondary to stroke, and the functional losses related to that damage was previously thought to be very strong,[44] however the work of Vygotsky and Luria have shown that this is not necessarily the case.[44-46] Recent technological advances have allowed this area to be more thoroughly investigated, in particular with computerised tomography. This was first used in attempting to predict rehabilitation outcomes in stroke patients by Miller and Miyamoto in 1979,[47] who found that those with large superficial lesions had a 50% chance of a good outcome, whereas those with deep lesions, showed a 25% chance of a good outcome. They did not find any outcome changes when the damage was in different areas. Consequently, others[48,49] have investigated the prognostic value of cortical damage by comparing it with Barthel ADL scores in patients undergoing stroke rehabilitation. They found that although size of lesion had predictive value it was not as powerful as initial Barthel ADL scores. Using a four point scale to categorise cortical damage due to stroke, a 1988 study[50] found that those with less damage had a greater chance of going home, although all groups, except those with bihemispheric damage, improved after rehabilitation.

Research by Henley, Petit, Todd-Pokropek and Tupper[51] disputed these claims, finding no significant correlation between outcome and size of the lesion.

Onset Admission Interval

This was one of the first factors advocated for its predictive ability[3-5], and it continues to be investigated, possibly because it is nearly always an easily calculated variable. However, although some researchers[7,13] contend that an increased time

interval between stroke onset and rehabilitation initiation is a negative predictor of good outcome, others[19,21,31] have found no correlation, possibly because what is actually happening is natural recovery.[18]

Social Factors

This is an area where results vary depending on the definition of successful outcome. Although research has shown[15,29,30] that subjects with maximal family involvement were more likely to be discharged home, others[15,52] have found that those with more family involvement had less functional improvement. This is possibly because those returning home need family help if they are not totally independent,[29] but the patients feel overprotected and lose motivation in their rehabilitation when they have too much family involvement.[52]

How are Predictors Devised and Implemented?

Methods for calculating and implementing predictors have varied widely, from simple correlations to clinical decision trees and even quadratic equations.

In the search for an objective method of prediction, Bruell and Simon[5] chose three factors that they as clinicians thought might be useful predictors and then divided those patients with good and bad rehabilitation outcomes. They found, using averages, that the three factors had a statistically significant correlation with rehabilitation outcome, making them useful as predictors. The original measures were then reduced with T-score transformations to a common scale with a mean of 50 and a standard deviation of 10. By adding and averaging these T -scores, three two-factor measures and one three-factor measure were obtained.

Instead of fractions of T-scores, Bourestom[3] used correlation coefficients which form the basis of many later attempts to calculate predictors.[7,22,26,36] These predictors were then used to estimate improvement in ADL scores. The study took a multivariate approach similar to that of Bruell and Simon,[5] but they also did a regression analysis in order to find the most powerful predictors from a field of twelve. Their criterion was that the factor not only had a high correlation coefficient but that it also did not correlate strongly with more powerful factors. Therefore, although age and locomotion status had high correlations, they were excluded as they correlated well with the more powerful ADL scores.

Another popular method has involved using ADL scores themselves as predictors. Shah et al[30] used the Barthel ADL index to develop a quadratic equation that explained 8% more variance than a simple regression equation. Oczkowski and Barreca[19] used the Functional Independence Measure, in a study of 113 stroke survivors and developed a table of likelihood ratios that predicted the probability of discharge home which was based on the average probability of discharge home being 73%.

In 1994, Falconer and co workers,[12] designed a classification tree for simple clinical use. To generate it, they first identified 51 possible predictor variables and measured them in 225 stroke survivors, plus whether the subjects had good or poor rehabilitation outcomes. Computer analysis then identified the predictor that best separated favourable and unfavourable outcomes into two subgroups. These subgroups were also split until a large tree was grown that included all the predictor variables. The tree was then reduced so that it included only the best predictors, because too many predictors increased the chance of error. To determine outcomes, the clinician follows different branches of the tree, based on the patient's characteristics, until the end of a branch is reached and a favourable or unfavourable result is forecast. This tree correctly predicted 88% of the cases with an 18% error rate.

Uses of Predictors

After 40 years of studying predictors, it appears odd that even the most powerful ones are not in common usage. The problem appears to have occurred due to poor statistical analysis, use of different outcome measures and lack of cross validation.[18,53] Researchers are still stressing the same point that was made 30 years ago,[3] that more objective criteria are needed to predict rehabilitation outcomes.[54] In addition, some researchers have contended that predictive factors are not powerful enough to determine an individual's progress and are only useful in large group analyses.[8,10,15,17] Nevertheless, the use of predictors is becoming increasingly popular and proposed uses are numerous.

The most common suggestions for using predictors usually concern efficient use of resources, including the evaluation of the effectiveness and the cost of rehabilitation.[12,30] This is important due to financial constraints and long waiting lists.[19,27] Osberg and co workers,[32]

investigated this area in 1990, when they used retrospective data on stroke patients to determine which groups used the most resources with the least improvements. They found that a small group of patients who do not improve with rehabilitation consume a disproportionate share of inpatient rehabilitation resources. However, as Wade[55] discussed, data collection itself takes resources.

Predictors have been advocated as an aid in the planning of services and in identifying those who will most benefit from rehabilitation.[4,28,30,37,39,40] In selecting appropriate patients for referral, Boyle & Schalzitti[4] suggested only providing rehabilitation to those patients under 60 years old for the first month and then to the one month survivors in the older group. However, the study by Feigenson and co workers,[56] found that excluding negative predictors did not result in better rehabilitation outcomes, partly because their exclusion methods were not good enough. Barer and Mitchell[10] compared multiple and single variable predictors, and found that although multivariate formulas did predict significantly better, this difference was irrelevant clinically. They advocated the use of a few clinical signs such as consciousness level, arm function and continence to make clinical judgements.

A less dramatic form of rehabilitation program planning may involve using the prediction of a patient's discharge destination to direct the focus of their training.[37] This strategy would be particularly relevant for assessment of ocular function and subsequent visual rehabilitation. Patients predicted to be discharged to a nursing home for example, would have visual rehabilitation as a low priority compared to transfers and bladder control. In contrast, a patient discharged to their home, may have a need for an intensive period of visual rehabilitation, especially if treatment after discharge is inconvenient.

Predictors may also have a psycho-social use. They can be used to help allocate support and counselling for those patients and their families with unfavourable prognoses.[57] They also allow clinicians to gauge the risk associated with any proposed treatment.[21] Additionally, predictors suggest direction for research, with strong negative predictors often targeted for extra treatment or further study.[17]

Conclusion

Although they have not been developed to the stage where they are useful as the sole guide for determining rehabilitation potential, predictive factors can be a valuable tool for the orthoptist working with patients who have survived stroke. They allow estimation of a rehabilitation time frame to help set goals and assist the orthoptist in catering to the visual needs of their patients. Experienced clinicians would already employ these judgements informally, however this is not a readily transferable skill, and clinicians inexperienced in this area should particularly benefit from some basic guidelines based on solid research.

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A Comparison of Contrast Sensitivity between People with a Colourvision Defect and those with Normal Colourvision

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Abstract

This study tested the prediction that people with reduced colour vision would show improved contrast sensitivity, as a compensatory adaptation. Twenty eyes with normal colour responses on the Farnsworth-Munsell 28 Hue test and the Ishihara test, and twenty-five eyes which showed colour vision defects on these tests, were compared on the Vectorvision CSV 1000 contrast sensitivity test. The prediction was not supported by the data. Eyes with colour vision defects showed reduced contrast sensitivity for lower spatial frequencies. This effect was strongest in eyes with more severe colour defects. Colour vision defects were not related to contrast sensitivity for higher spatial frequencies. Increased age and reduced visual acuity predicted poorer contrast sensitivity at higher spatial frequencies.

Key Words:

Contrast sensitivity, colour vision defects, Ishihara test, Farnsworth-Munsell 28 Hue test, Vectorvision CSV 1000, spatial frequencies.

Introduction

When we consider occupations such as the defence force, bus drivers, and electricians, there is one thing that all of them have in common. None of these occupations employ people who are colour blind.

In the general population it is seen as a liability to be colour blind, but what if people with a colour vision defect could compensate for their lack of colour appreciation by using an alternative visual function such as shade or contrast?

Colour vision defects can be congenital or acquired. In people with congenital colour vision defects, usually one cone photoreceptor is not functioning normally (anomalous trichromat), or may not be functioning at all (dichromatic). Therefore, in the case of someone with an abnormally functioning cone they will use "abnormal proportions of these [cones] to colour match".[1] In the case of someone with only two types of cone photoreceptors, they are only able to use these two cones to colour match. In both of these cases the result is an altered perception of colour. According to Fitzgerald and Billson,[1] if the green cone, for example, is not functioning normally, the person will be able to distinguish between pure reds and greens, but will have trouble with colours that are a mixture of red and green. These people have 'deuteranomalous colour vision'. If the green cone is not working at all, or is absent, the person will have trouble distinguishing between pure reds and greens, and will see everything in either blue, yellow, grey, or black. These people are termed 'deuteranopic'.

From this it can be seen that people with colour vision defects clearly need to use other visual cues apart from hue to be able to tell certain colours apart. Ravin, Anderson, and Lanthony[2] while discussing the famous artist Charles Meryon stated, "Despite his colour

Accommodation Values in a Normal Sydney Population, is the RAF Rule Still Valid?

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Introduction

The evaluation of the dynamic components of the near response forms a major part of an assessment of a person who has symptoms for near, or who has difficulty in changing focus. The close association of accommodation with age means that measures obtained in a clinical assessment must be compared with age related normal values to determine whether or not any abnormality is present.

In Australia, the most commonly used instrument to assess accommodation is the RAF rule. This instrument has the advantage of being able to determine the amount of accommodation occurring (in dioptres) and to match this against age related values. It also enables a simple measurement of the accommodation and convergence near point (in cms). The 'normal' values indicated on this device are those determined by Duane in 1912[1]. These are taken from a comprehensive study of normal accommodation, where the near blur point was measured and converted to diopters, assuming that any refractive error was corrected. It is likely that the 'mean' values are actually median values as they are always exactly midway between the upper and lower values.
(See Figure 1).

Clinical norms must be matched to those of the relevant population, and, in an urban Australian society at the end of the 20th century, the question must be asked as to whether values determined over eighty five years ago are still appropriate to use as normal for our population.

Figure 1. *Duane's normal values of accommodation for age measured in 1912.*[1]

Borish[2] reports other studies which have been made on age related accommodation values, which are summarised in Table 1. Some measures are notably different from Duane's, possibly due to the different measuring techniques which were used (especially those of Donders in 1864). Even the most recent (Turner, 1958) was published forty years ago. It is likely that the subjects in these studies were mostly of western European origin, whereas the current urban Australian population (Sydney in this study) has significant numbers of citizens with other ethnic origins.

Different clinicians have different measuring techniques; for example, the speed at which the target is moved will influence the recorded near point of accommodation. Some clinicians take only one measure, others measure several times. Unless each examiner uses the exact techniques that were used by Duane, values that differ from his will not necessarily represent abnormalities. In determining general values that can be used by all, there is also merit having input by many examiners in the determination of normal values for a particular population of subjects and examiners.

Table 1. *Summary of accommodation for age studies.*

Age (Years)	Donders (1864)	Duane (1912)	Sheard (1917)	Jackson (1922)	Turner (1958)
0-10					
11-15	19.7	13.4	12.0	14.0	13.0
16-20	16.0	12.3	11.0	12.0	10.6
21-25	12.7	11.2	9.0	10.0	9.5
26-30	10.4	10.0	7.5	9.0	7.9
31-35	8.2	8.7	6.5	8.0	6.0
36-40	6.3	7.3	5.0	7.0	5.75
41-45	5.0	5.7	3.75	5.5	4.4
46-50	3.8	3.9	2.75	4.0	2.5
51-55	2.6	2.1		2.5	1.6
56-60	1.75	1.4		1.25	1.1
61-65	1.0	1.2		0.5	0.7
66-70		1.1			
71-75		1.0			
76-80					
81-85					

For these reasons, a study of accommodation values in the Sydney population was carried out to determine current standards, and to compare these findings with the commonly accepted norms.

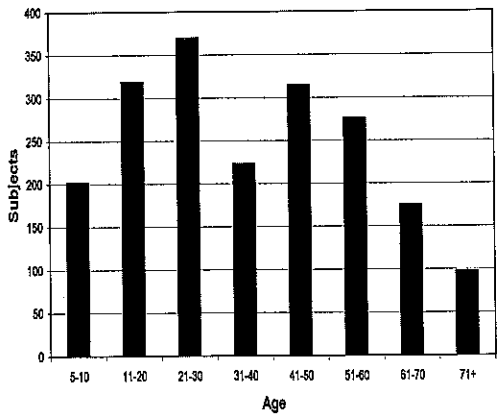

Figure 2. *Subject numbers in age categories.*

Method

The normal values of accommodation were measured in 1,978 subjects over a two year period (1995 -1996) by a total of 40 third year orthoptics students. These students had all been assessed as being competent in the appropriate measuring technique, and, in many cases, were supervised by a clinician during the testing.

Subjects

Subjects were included if there was no known or suspected anomaly of the ciliary muscle or the lens. The following were exclusion criteria:

- aphakia (or pseudophakia)
- cataract
- known anomalies of accommodation (eg, accommodative spasm)
- medication which affects accommodation
- the squinting eye in uniocular strabismus
- amblyopia

As normal values were needed, the examiners were encouraged to take measurements from a non-clinical population, ie from amongst their family and friends, and non patients in the clinics. Although attempts were made to achieve relatively similar subject numbers in all age groups it was inevitable that larger numbers were found from the young to middle age adult population. The presence of cataract also excluded many of the older subjects. Even so, 273 subjects over the age of sixty were assessed. The distribution of subjects is shown in Figure 2.

Procedure

The RAF rule was used where this was available, using the incorporated reduced vision chart or the four lines of different sized near print. Where this was not available, a ruler was placed against the subject's infraorbital ridge and an accommodative target was brought towards the eye along the edge of the ruler. The near point was measured in centimetres and converted to dioptres (using the formula 100/cms).

The subjects wore appropriate distance correction. If there was any known undercorrection, the full distance correction was used. Where progressive lenses (or bifocals) were incorporated in the glasses, care was taken to ensure that the subject was looking through the distance section of the glasses. Where the near point was more remote than 50cms (the length of the RAF Rule), this distance was measured and the results converted to dioptres.

Each eye was assessed separately.

Results were recorded in the age groups, from six to ten years, and thereafter in five year age groups to age 85 years.

The subjects' initials, age and clinic code were recorded to detect any duplicate measurements. At the end of the relevant period, the data was analysed using EpiInfo and Minitab.

Results and Discussion

Valid measures on 1,978 subjects were obtained. When right and left eyes were compared there was a statistically significant difference between the two eyes ($t = -2.4$, $p = 0.017$), however the actual difference of 0.57D is so small and clinically meaningless that it was decided to disregard it. The probable reason for the statistically significant difference was the very large sample size. (This is an example of a difference between practical and statistical significance.) The resulting data from each eye were therefore pooled.

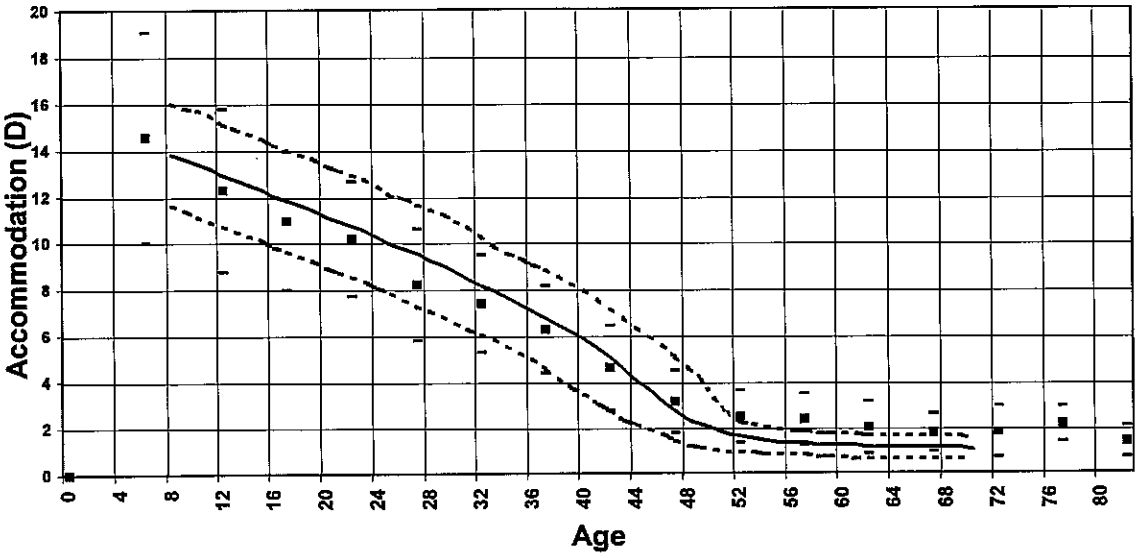

Figure 3. *Mean and SD of current study compared with Duane's values. Continuous and dotted lines show Duane's values. Current values are superimposed, as squares and dashes.*

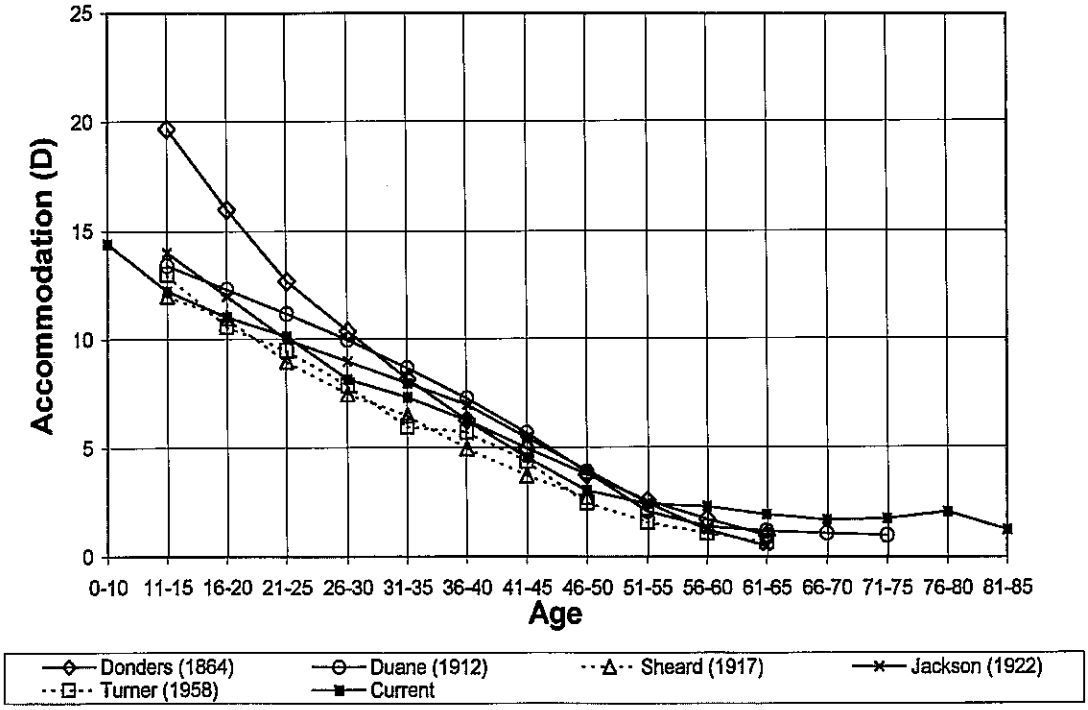

Figure 4. *Mean accommodation for age values in the current study compared with previous work.*

Values for the mean and plus or minus one standard deviation study are shown in Table 2, and are compared with the Duane values ('median' 'minimum' and 'maximum') in Figure 3.

Table 2. *Normative accommodation values in this study.*

	Current Study		
Age	**-1 SD**	**Mean**	**+1 SD**
6-10	10.0	14.5	19.1
11-15	8.8	12.3	15.8
16-20	8.0	10.9	13.9
21-25	7.7	10.2	12.6
26-30	5.8	8.2	10.6
31-35	5.2	7.3	9.4
36-40	4.3	6.2	8.1
41-45	2.7	4.5	6.4
46-50	1.7	3.1	4.4
51-55	1.3	2.4	3.6
56-60	1.2	2.3	3.4
61-65	0.8	2.0	3.1
65-70	0.9	1.7	2.6
71-75	0.7	1.8	2.9
76-80	1.3	2.1	2.9
81-85	0.7	1.3	2.0

It can be seen that the values obtained in this study are close to (although slightly lower than) the Duane values up to around 50 years of age, after which they are consistently better (by about 1.5D). The trends in each study imply that this improvement persists up to the age of 80 (although data are not available after the age of 70 from Duane). It is interesting to note that the values are very similar for subjects in their forties in each study, ie, the onset of presbyopia would appear to remain the same.

There is a greater spread of values in young children in this study, probably due to the difficulty in obtaining accurate data from these subjects. This is particularly evident in those under eight years, suggesting that these values should be considered with caution, although the mean value remains very consistent with Duane's study.

When the mean values are compared with other studies (see Figure 4), values under the age of fifty are very consistent, so that for a general indication of whether a patient's accommodation is normal, the RAF Rule still remains an appropriate measure. However, as this device only gives mean values, one must take care in deciding whether different measures are abnormal or within the normal range as given in Table 2.

Evaluation of accommodation in subjects over 50 is not normally carried out, so the moderately improved values in the older population, which persist when compared with other studies (see Figure 4), and the additional data given for those over 70 years have less clinical relevance. Nevertheless these findings are of interest. They could possibly be due to measurement errors, however the consistency of measures in the younger subjects make this unlikely. Other explanations could include:

- Better health in older age, due to improved medical care and lifestyle, leading to improved ciliary muscle tone and/or delayed hardening of the lens.
- Possible increase in the amount of close work undertaken by the current population group which may also affect the ciliary muscle and lens.
- A different demographic group, with significant numbers of subjects from Asia and Eastern Europe being represented in the Australian urban population.

Each of these possibilities would, of course, require further study to evaluate fully. What is clear from this study is the need for clinicians to be cautious in using data that has not been validated in the current population when determining possible abnormalities.

Conclusion

Measures of accommodation up to the age of 50 are very similar to those of previous studies, however after this age the values are moderately and consistently better than those previously published.

Acknowledgements

Obviously full acknowledgement needs to be made to the forty students who obtained the data for this study. Not only did they gain valuable experience in a common measuring technique; they were able to be involved in a clinical research project, which has real implications for their future clinical practice.

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A Comparison of Contrast Sensitivity between People with a Colourvision Defect and those with Normal Colourvision

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Abstract

This study tested the prediction that people with reduced colour vision would show improved contrast sensitivity, as a compensatory adaptation. Twenty eyes with normal colour responses on the Farnsworth-Munsell 28 Hue test and the Ishihara test, and twenty-five eyes which showed colour vision defects on these tests, were compared on the Vectorvision CSV 1000 contrast sensitivity test. The prediction was not supported by the data. Eyes with colour vision defects showed reduced contrast sensitivity for lower spatial frequencies. This effect was strongest in eyes with more severe colour defects. Colour vision defects were not related to contrast sensitivity for higher spatial frequencies. Increased age and reduced visual acuity predicted poorer contrast sensitivity at higher spatial frequencies.

Key Words:

Contrast sensitivity, colour vision defects, Ishihara test, Farnsworth-Munsell 28 Hue test, Vectorvision CSV 1000, spatial frequencies.

Introduction

When we consider occupations such as the defence force, bus drivers, and electricians, there is one thing that all of them have in common. None of these occupations employ people who are colour blind.

In the general population it is seen as a liability to be colour blind, but what if people with a colour vision defect could compensate for their lack of colour appreciation by using an alternative visual function such as shade or contrast?

Colour vision defects can be congenital or acquired. In people with congenital colour vision defects, usually one cone photoreceptor is not functioning normally (anomalous trichromat), or may not be functioning at all (dichromatic). Therefore, in the case of someone with an abnormally functioning cone they will use "abnormal proportions of these [cones] to colour match".[1] In the case of someone with only two types of cone photoreceptors, they are only able to use these two cones to colour match. In both of these cases the result is an altered perception of colour. According to Fitzgerald and Billson,[1] if the green cone, for example, is not functioning normally, the person will be able to distinguish between pure reds and greens, but will have trouble with colours that are a mixture of red and green. These people have 'deuteranomalous colour vision'. If the green cone is not working at all, or is absent, the person will have trouble distinguishing between pure reds and greens, and will see everything in either blue, yellow, grey, or black. These people are termed 'deuteranopic'.

From this it can be seen that people with colour vision defects clearly need to use other visual cues apart from hue to be able to tell certain colours apart. Ravin, Anderson, and Lanthony[2] while discussing the famous artist Charles Meryon stated, "Despite his colour

vision defect, Meryon was able to depict contrasts in light and dark". This suggests that Meryon was using shade and contrast rather than hue to distinguish between different colours. For example, to someone with a red/green colour vision defect red and green may both appear to be of a similar hue, however the green may be a different contrast than the red. It is likely that a person who has grown up with a colour vision defect has learned that green is a different shade than red. In this situation it is the contrast of the two colours that is the distinguishing factor. It is possible that people with a colour vision defect rely on the appreciation of contrast rather than hue to provide reliable information from their visual input, which can lead to the idea that people with a colour vision defect have a superior contrast sensitivity function.

Clinically, the functions of contrast sensitivity and colour vision can be tested separately. The testing of the contrast sensitivity function is commonly performed following an approach of varying contrast and frequency. The contrast refers to the presentation of gratings (ie. line forms) within a background. These gratings move from levels of high contrast (dark grey on light grey), then decrease in contrast until the shade of the gratings equals the background shade. The frequency refers to the width of the gratings, where low frequency corresponds to fewer and wider gratings, while high frequency corresponds to narrower and more numerous gratings. The gratings are sinusoidal in nature and therefore show a gradual change from the lighter background to the darker gratings.

Testing the colour vision function can be performed using two main methods, colour matching, and figure/background discrimination. The colour matching method used in the Farnsworth-Munsell 28 Hue colour vision test is performed by presenting the patient with a coloured object, and asking them to locate the object that is most similar (in colour) to the original object. The figure/background method used in the Ishihara colour vision test requires the patient to locate a coloured figure within a differently coloured background.

It is evident that the recognition of colour and contrast require different visual mechanisms, colour vision requires specific cones to send an excitatory or inhibitory message to the ganglion cells, while contrast sensitivity is not cone specific.

As the two visual mechanisms clinically can be measured separately, this study addresses the questions: "Do people with colour vision defects have a superior level of contrast sensitivity compared to people with normal colour vision?" and "Does the severity of the colour vision defect have an effect on the contrast sensitivity function?". Research in this area could be very beneficial to people with colour vision defects and the occupations who overlook them.

If it is found that people with a colour vision defect have superior contrast sensitivity, this may indicate that they are in fact able to perform just as competently in some of the occupations mentioned earlier (ie. bus drivers, electricians, and the defence force) as people with normal colour vision. However, if the findings indicate that people with a colour vision defect have equal, or inferior contrast sensitivity compared to people with normal colour vision, then our paper may provide justification to the occupations that choose not to employ people with colour vision defects.

Method

Subjects

Volunteers for this study were found at The University of Sydney (Cumberland Campus), and others known to the researchers.

The results of forty-five eyes were analysed. Twenty-one possessed normal colour vision and twenty-four had a colour vision defect.

The age of subjects included in this study ranged from 15-55 years, which is approximately compatible with the age range of the Australian workforce.

Tests and Procedures

In order to determine whether subjects were suitable for this study, a preliminary eye examination was carried out, comprising:

- a short ocular history
- monocular visual acuity
- cover test

Binocular single vision was not tested as all procedures were carried out monocularly.

History

Subjects were questioned as to whether they possessed any of the following :

- amblyopia
- strabismus
- cataract (or IOL implants)
- stroke or head injury
- eye infection
- any other ocular defect not previously mentioned. Subjects were also questioned regarding ocular surgery, general health, and current medications.

Visual Acuity

Visual acuity was carried out monocularly at a distance of 8 feet, (the standard distance for the Vectorvision LogMAR chart) under normal room illumination, using the Vectorvision CSV 1000 LogMAR chart (which is internally lit). The subject's distance optical correction was worn.

The LogMAR consists of the same number of letters (5) on each line. For statistical purposes, visual acuity was quantified by obtaining the sum of letters (from 20/32 to 20/10) observed by the subject. Therefore, a score of 5 would indicate a level of VA of 20/32, (6/9) while a score of 30 would indicate a level of VA of 20/10 (6/3).

Cover Test

A cover/uncover, and alternative cover test was performed at :

- 1/3 metre with a light and an accommodative target (with and without glasses if appropriate);
- 6 metres (with and without glasses if appropriate).

Ishihara Colour Vision Test

This was carried out monocularly (with glasses if applicable), under normal room illumination, on a uniform black background, at a distance of 75 centimetres from the subject. Localised light (in the form of a lamp with a daylight globe), was used in addition to the general room illumination.

The subject was questioned as to what numbers (if any) they could see in plates 1-17.

Farnsworth-Munsell 28 Hue Colour Vision Test

The 28 Hue was performed monocularly (with glasses if needed) on a uniform black background, under the same lighting conditions as used for the Ishihara colour vision test.

Starting with the reference chip, the subject was asked to place the other 27 chips in order so that there was a gradual change in colour from the first chip to the twenty-eighth. The results of the 28 Hue colour vision test were entered into a specifically designed computer program which gave a numerical value (0 - 200) corresponding to the severity of the colour vision defect. A score of 0-2 indicates normal colour vision, and a score of 4-200 indicates a colour vision defect (4 being mild, 200 being severe).

Contrast Sensitivity

The contrast sensitivity of the subjects was measured monocularly, at the standard distance of 8 feet, using normal room illumination as well as the internal illumination of the Vectorvision CSV 1000. Subjects wore their distance optical correction.

The Vectorvision is made up of 4 rows (A-D), each containing 8 pairs of circles (column 1-8). One circle in each pair contains gratings. The other circle in each pair contains one uniform shade of grey (ie. no gratings).

The frequency of the gratings increases from row A to row D (row A = 3 cycles/degree, row B = 6 cycles/degree, row C = 12 cycles/degree, row D = 18 cycles/degree). The contrast of the gratings decreases between circles 1 to 8.

Starting at row A, column 1, the subject indicates which circle in each pair contained the gratings (ie. top, bottom or neither). Once all columns in row A had been tested, the subject repeats the process for rows B, C, and D respectively.

The test is scored by summing the number of circles seen correctly. Each spatial frequency has a separate score.

Inclusion Criteria

Those included in the control group were subjects who had no current or previous visual defects, visual acuity of 20/32 (approx. 6/9) or better, and had normal colour vision.

Those included in the experimental group were subjects who had a colour vision defect with no other visual problems, and visual acuity of 20/32 (approx. 6/9) or better.

In subjects with a strabismus, in order to avoid a secondary influence from sensory adaptations, only the non-deviating eye was included in the results.

Statistical Analysis

The basic purpose of the research was to explore the relationship between the degree of colour vision defect, and contrast sensitivity at four spatial frequencies. The first analysis correlated the colour defect score with contrast sensitivity scores. Age and visual acuity are potential predictors of the relationship between colour vision defect and contrast sensitivity. Their role was explored by including them with the colour vision defect score as predictors in a multiple regression. Contrast sensitivity was used as the dependent variable. The effect of the multiple regression is to show if colour vision defect, age, and visual acuity taken singly or in combination, are related to contrast sensitivity.

Table 1

Mean and standard deviation of; age, VA, Row A-D (for the experimental and control groups)

	Control		Experimental	
	Mean	Standard Deviation	Mean	Standard Deviation
Age (years)	35.25	15.18	32.36	13.25
Visual Acuity	16.40 (approx. 6/6)	4.48 (approx. 1 line)	16.52 (approx. 6/6)	5.27 (approx. 1 line)
Row A	6.50	0.89	5.12	0.73
Row B	6.15	0.81	5.48	1.08
Row C	6.30	1.34	5.96	1.59
Row D	6.05	2.11	5.64	1.49

Table 2

Correlations (r) between rows A-D, and inverse error, age and VA

	InverseError	Age	Visual Acuity
Row A	0.744	- 0.085	-0.081
Row B	0.383	- 0.299	0.346
Row C	0.103	- 0.468	0.417
Row D	0.099	- 0.361	0.432

Table 3

Single regression data between contrast sensitivity and inverse error.

	Equation	Adjusted R-Squared Values	P-Values
Row A	Row A=4.82 + (3.88 x inv.error)	54.300	0.000
Row B	Row B=5.32 + (1.93 x inv.error)	12.700	0.009
Row C	Row C=5.93 + (0.75 x inv.error)	0.000	0.502
Row D	Row D=5.59 + (0.88 x inv.error)	0.000	0.518

Table 4

Multiple regression data between contrast sensitivity, age, VA, and inverse error.

	Equation	Adjusted R-Squared Values	P-Value	Combined P-Value
Row A	Row A=5.62-(0.00974 x age)-(0.0284 x VA)-(3.88 x inv. error)	54.400	Age: 0.244 VA: 0.238 Inv. error: 0.000	0.000
Row B	Row B=4.86-(0.00141 x age)+(0.0568 x VA)+(1.93 x inv. error)	24.700	Age: 0.176 VA: 0.061 Inv. error: 0.006	0.002
Row C	Row C=5.82-(0.0381 x age)+(0.0853 x VA)+(0.734 x inv. error)	24.500	Age: 0.014 VA: 0.053 Inv. error: 0.449	0.002
Row D	Row D=4.50-(0.0293 x age)+(0.127 x VA)+(0.86 x inv. error)	18.600	Age: 0.125 VA: 0.023 Inv. error: 0.480	0.009

Results

Of the 46 eyes that were tested, one result (in an eye with normal colour vision) stood out as being markedly below the contrast sensitivity range. It was the authors' belief that this result was not indicative of the general population. Consequently, this result was excluded from the statistical calculations.

The mean contrast sensitivity results in the group with colour vision defects were consistently lower than those in the group with normal colour vision (Table 1.).

The results of correlation and multiple regression analyses are most trustworthy if sample data are normally distributed. When the colour vision defect scores were graphed, they were found to be heavily skewed to the right, with many low scores and relatively few high scores. Because the colour defect score is an arbitrary number, it was decided to change the shape of the distribution to normal (a standard practice with colourvision scores). This was achieved by adding 2 to all the scores (to ensure that all were larger than 1) and then taking the inverse of the score, i.e. transformed colour error score = 1/(error score + 2). Once the scores were transformed a lower score represented a greater degree of colour vision defect.

The results showed a significant correlation between contrast sensitivity and inverse total error only for rows A and B of the contrast sensitivity test, i.e. the low frequency test face. The correlations for these two rows were positive, indicating that the lower the inverse total error score, the lower the contrast sensitivity score. As the total error scores have been inverted, a low inverse total error score corresponds with a severe colour vision defect, while a low contrast sensitivity score corresponds with a poor contrast sensitivity function. Therefore, the correlations for row A and B indicate that the more severe the colour vision defect, the worse the contrast sensitivity function.

No significant correlation was found between the inverse total error and row C and D of the contrast sensitivity test, i.e. higher frequency test face (Table 2).

When multiple regressions were performed, it was found that the inverse total error result was able to significantly predict the level of contrast sensitivity for the low spatial frequencies (Row A and B). However, for the higher spatial frequencies, it was age and visual acuity that were the significant predictors of contrast sensitivity.

It was interesting to find in all instances that, while only one independent variable (ie. age, VA, or inverse total error) was a significant predictor of

the level of contrast sensitivity for each row, when combined together the total p-value was more significant than any individual predictor (Tables 3 and 4).

With regard to the severity of the colour vision defect differences were found when measured on the Ishihara colour vision test as opposed to the Farnsworth-Munsell 28 Hue colour vision test. The colour vision defect often appeared more severe when tested on the Ishihara, and occasionally appeared more severe when tested on the Farnsworth-Munsell.

Discussion

The results obtained bring us to the conclusion that the subjects with a colour vision defect have a lower contrast sensitivity function than subjects with normal colour vision. However, this conclusion can only be made with regard to the lower spatial frequencies. Concerning the higher spatial frequencies, it does not appear that colour vision status is significantly linked to the level of contrast sensitivity.

In addition to the above conclusion, it can also be observed that (for the lower spatial frequencies, ie. row A and B) the more severe the colour vision defect, the worse the contrast sensitivity function.

It has been proven in many studies that contrast sensitivity is influenced by age and visual acuity.[3,4] Therefore, it was necessary to calculate multiple regressions that included age and visual acuity along with colour vision status. The results of the multiple regressions allow us to make the conclusion that age and visual acuity do not have a significant effect on the lower spatial frequencies of the contrast sensitivity test. However, when looking at the higher spatial frequencies we can conclude that it is age and visual acuity which are the significant predictors of the level of contrast sensitivity. There was no link between colour vision and the higher spatial frequencies, however, it was interesting to note that the level of contrast sensitivity for each row could be predicted more accurately when the p-values for age, VA, and colour vision status were combined, (ie. when all of the variables were taken into account). This indicates that all three predictors actually did have an effect on each row of the contrast sensitivity test, even though some may not have been statistically significant. Therefore, while age and VA had a large influence on the contrast sensitivity results for the higher spatial frequencies (row C and D), the colour vision status also made a small contribution.

While conducting this experiment, the severity of colour vision defects differed considerably when measured on the Ishihara as opposed to the Farnsworth-Munsell 28 Hue. Littlewood and Hyde,[5] conducted a study comparing the Ishihara colour vision test to the Ohkuma colour vision test. Their research indicated that the Ohkuma colour vision test was superior in detecting and grading colour vision defects. As the Ishihara colour vision test is the most widely used screening test, this research raises the question of whether it is the most appropriate screening method available.

As there was a range of ages, and visual acuities in this study, it was inevitable that they would have an effect on the contrast sensitivity result. Perhaps in prospective studies, less variation in age and visual acuity can be adopted so they will not significantly influence future results.

In order to cause as little inconvenience to volunteers as possible, testing time was kept to a minimum, therefore, the Farnsworth-Munsell 28 Hue colour vision test was used to grade the severity of the colour vision defect. The authors feel that further research in this area of colour vision and contrast sensitivity, using the Farnsworth-Munsell 100 Hue colour vision test would be useful, as this is likely to give a more accurate grading of the severity of the colour vision defect.

Conclusion

The question arises, "Should people with colour vision defects be excluded from certain employment areas?" This study would suggest that an important factor in answering this question lies in the type of work involved. If the job involves a great deal of work with fine detailed objects, then exclusion from these occupations seems unnecessary. However, if the job description involves a lot of work with coarse, less detailed objects, then exclusion from these occupations may be justified only if the colour deficient person is required to work with low contrast objects or images. If the contrast between object and background is high, then at any spatial frequency, the colour deficient person is not disadvantaged when compared to their colleagues with normal colour vision.

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Visual Acuity Testing in Pre-School Aged Children - What Can Be Expected?

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Abstract

Three hundred and sixty pre-school aged children underwent orthoptic screening as part of The Outreach Programme run by The Orthoptic Department of Sydney Eye Hospital. The children were screened at Kindergartens and Child Care Centres in the inner city. Of the 360 children screened 353 (98%) cooperated with monocular Sheridan Gardiner (SG) visual acuity testing at 6 metres. Of the children participating in visual acuity testing 79.9% managed SG linear testing, with 20.1% of the children being tested with SG singles. There was a significant difference in the age of children who completed the SG singles versus SG linear method of testing. SG singles testing was required more often with the younger children. The average visual acuity score for SG singles assessment was 6/6 part, with the majority of children achieving 6/6; whereas for SG linear assessment the average score was 6/9 plus, with the majority of children achieving 6/9. It was found that irrespective of the visual acuity test method used, older children achieved slightly better visual acuity results than younger children. It was also found that irrespective of the child's age SG singles testing produced better visual acuity results than SG linear. Reduced visual acuity was noted in 9.8% of children with SG singles tesing, and 2.5% with SG linear.

Key Words:
Pre school vision screening, visual acuity, Sheridan Gardiner singles, Sheridan Gardiner linear.

Introduction

Numerous reviews of screening studies carried out during the various stages of visual development in childhood are contained in the literature.[1,2,3,4,5] Some authors feel that screening is of minimal value to visual outcomes, whilst others indicate that vision screening is particularly effective when carried out at a young age.[6] Ingram[7] reports that screening at preschool age is supported on the grounds that squint and amblyopia would be identified at an age when remedial treatment might be successful. Amblyopia is described by Taylor[8] as "a preventable visual disability affecting 1-3% of the general population and up to 5% of the preschool age group." It is the "commonest disorder encountered in paediatric ophthalmology clinics"[8]. Edwards[6] reported that Ehrlich et al found strabismus & amblyopia to be the two main defects preventing normal visual development, being present in 5-8% of children at school entry. Elston[9] states that "amblyopia in preschool children is asymptomatic and, if it is not due to, or accompanied by strabismus, will only be detected fortuitously unless screened for."

Beardsell[5] supports these findings stating "it has proved possible to test children quickly and accurately" at a preschool age. The Orthoptic Association of Australia NSW Branch's document titled The Orthoptist's Role in Vision Screening-: Review & Recommendations[10] states that the aim of vision screening is to identify ocular problems that may cause permanent visual loss or interfere with classroom learning. The document also

recommends that vision screening be carried out during a critical period of visual and ocular development.

The Orthoptic Department of Sydney Eye Hospital established an Outreach Vision Screening Programme in April 1995. The programme provides an orthoptic vision screening service to three and four year old children attending eighteen Child Care Centres and Pre-schools in the vicinity of the hospital.

The Outreach Vision Screening Programme is conducted once a week by the Orthoptic Student Supervisor and is also attended by orthoptic students on clinical placement from the University of Sydney. The testing procedure involves assessment of visual acuity, cover testing, convergence and stereoacuity assessment with the Langs I and/or II stereoacuity tests.

The Programme has proved to be worthwhile, as approximately 8% of the children screened have been found to have a visual defect requiring follow-up ophthalmological investigation. Of the children detected with ocular abnormality, the majority had reduced visual acuity in the absence of strabismus. The study to be presented will only examine information relating to visual acuity testing.

Method

Appleboom[11] reported that, since the earliest screening programme which was initiated in 1899, many alterations to original screening procedures have occurred after consideration of test reliability, testing conditions and child maturity. Currently "a variety of screening tests are in use because it is unclear which is the best for the detection of visual deficit" in preschool aged children.[12] The literature describes many forms of visual acuity testing procedures,[1,2,13-22] however it was Fern[19] who concluded that "a well designed preschool visual acuity test should consist of high contrast Snellen optotypes without directional components that progress in 0.1 log steps down to a level of 6/3". Fern also reports that "of the tests that have been standardised Sheridan Gardiner comes closest to meeting these criteria".

Preschools within the vicinity of the hospital were approached. Those interested in participating in the Programme were provided with an explanation of the testing procedures and consent forms which were to be signed by a parent/guardian and collected from each child prior to screening. The consent form contained details of the child's name, gender and date of birth.

The testing procedure used for the Outreach Vision Screening Programme, visual acuity (VA) was assessed monocularly (right eye prior to left) at 6m using a back lit linear Sheridan Gardiner (SG) chart. All children were encouraged to attempt SG linear testing. If the children were unable to perform the SG linear test, they were then assessed using SG single letters. Prior to commencing the test each subject was shown the SG key card and a trial of matching the letters was carried out to ensure that the principle of the test was understood.

Visual acuity results for each eye and the test method used were documented for each subject. VA was assessed down to 6/5 using SG linear and 6/3 using SG singles where applicable. For research purposes the visual acuity was recorded as 'part', when there was plus 2 or minus 2 letters recognised on a line. That is, if the VA was 6/6-2 it was recorded as 6/6 part, and if the VA was 6/6+2, it was recorded as 6/5pt. When there was plus 1 or minus 1 on a line, the VA was recorded simply as that line. For example 6/6+1 would be rounded to 6/6. If the child was unable to perform either of the VA tests monocularly at 6m, no result was recorded.

Results & Discussion

Three hundred and sixty pre-school children from 18 individual inner city child care centres underwent orthoptic screening over a period of approximately 8 months. Roughly equal numbers of males and females were tested. In this study 7 of the 360 (2%) children screened were unable to have their VA assessed. Five of these children would not allow the orthoptist or orthoptic students to occlude either eye. The remaining two children were unable to manage VA testing due to intellectual impairment. Fitzgerald's paper[5] titled the "Incidence of reduced visual acuity and squint in preschool children aged three in Australia" reported that "6.5% of children could not do the SG singles VA test at 6m". The small percentage of children in our study (2%) who were unable to participate in visual acuity assessment may be attributed to the older age of the majority of the children.

The majority of the children tested 282/353 (79.8%) managed to carry out the SG linear test, the remaining 71 (20.1%) of participants having to re-attempt VA assessment with SG singles. The children who required re-assessment with the SG singles tended to lose interest in the SG linear 'matching game' or found the method difficult and refused to continue. Several children claimed that they could not see the letters on the SG linear chart.

The data studied were found to be suitable for parametric statistical analysis. A t-test was carried out to compare the ages of the children performing SG singles versus SG linear. There was a significant age difference found (*t -7.57, df 351, p<0.001*) with younger children requiring the use of SG singles more often than the older children. The average age of children tested with SG singles being 43.5 months (approx 3 1/2 years), and 50.3 months (approximately 4 years) with SG linear.

Visual acuity scores were coded for statistical analysis as:

1= 6/60	7= 6/9 part
2=6/36	8= 6/9
3= 6/24	9= 6/6 part
4= 6/18	10= 6/6
5= 6/12 part	11= 6/5 part
6= 6/12	12= 6/5

There was no significant difference in the visual acuity results for the right and left eyes for either the SG singles (*t -0.48, df 96, p=0.632*) or SG linear test methods (*t 0.21, df 281, p=0.83*). Analysis showed no order, learning or fatiguing effect with either VA assessment method. This finding was also reported by Fitzgerald in the Orthoptic Association of Australia Study[5]. The VA results for each of the right and left eyes were therefore studied together as a group. Subjects scoring different VA results between the two eyes, had their result codes averaged. As a result of this, intermediate codes were created where scores fell between two visual acuity categories.

The average visual acuity score for children assessed with SG singles was 6/6 part, with the most common visual acuity score being 6/6 (29.5%).(Figure 1.)(*Mean 8.84, Std Dev 1.82, Mode 10*)

The average visual acuity score for children assessed with SG linear was 6/9 'plus', with the most common visual acuity score being *6/9 (27%)*.(Figure 2.)(*Mean 8.64, Std Dev 1.53, Mode 8*)

In order to investigate the effect of age differences and the effect of differences in the test method employed on the VA score achieved by preschoolers the group was sub-divided. Group 1 was comprised of preschoolers who were 40 months or younger and Group 2 of children older than 40 months. The result of splitting the group into two age categories meant that there were disproportionate numbers in each group, with the majority of children falling into Group 2. In order for statistical testing to be valid, a random sample of subjects from Group 2 was compiled to equalise the numbers in the two age sub-groups.

Analysis of Variance (ANOVA) of the sub- groups showed that :

1. SG Singles method of testing produced higher (better) VA scores (*x= 8.84*) than the SG Linear method of testing. (*x=8.24*) (*F=1, 131, 4.74, p=0.031*).

2. Preschoolers 40 months and younger achieved lower VA scores (*x=8.21*) than preschoolers older than 40 months (*x= 8.77*) (*F=1, 131, 3.99, p=0.34*).

The two-way interactions ANOVA showed no interaction, that is, SG Single VA scores are higher than SG Linear VA scores for both age groups; and preschoolers 40 months and younger achieve lower VA scores than preschoolers older than 40 months for both of the VA testing methods. (*F=1, 131, 0.93, p= 0.34*) In other words: SG single letters produced VA scores better than SG linear testing irrespective of the subject's age; and subjects older than 40 months achieved a significantly better VA score than subjects 40 months or younger irrespective of the VA test method used.

These results confirm two aspects of visual acuity assessment that have previously been suspected but unconfirmed statistically, namely:

1. Preschoolers find the SG Singles a much easier VA test than SG Linear and achieve better results with SG Singles. This finding is similar to that reported by Shaw[23] who stated that "single letter optotypes are generally considered to overestimate visual acuity," and

2. Mature preschoolers generally achieve better VA results than immature preschoolers on any VA test. This finding is in line with Elston's editorial[9] which stated that "the visual acuity of children increases with age."

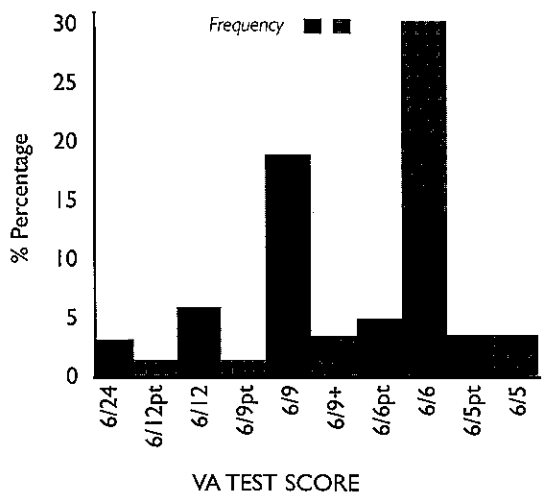

Figure 1

SG Singles Test Scores

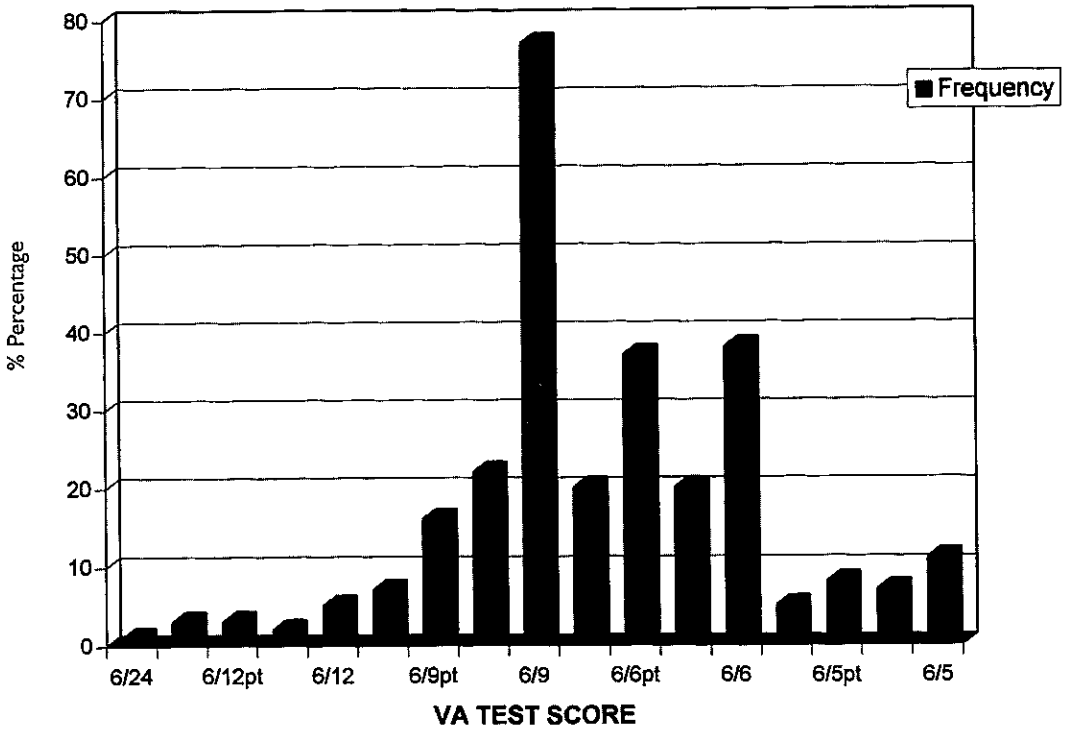

Figure 2

SG Linear Test Scores Intermediate codes eg. 6/9+ (between 6/9 and 6/6pt) are not labelled.

The referral criterion used by the Outreach Vision Screening programme was:

Preschoolers scoring less than 6/9 with SG Singles were referred on for further ophthalmic investigation. Preschoolers scoring less than 6/12 with SG Linear were also referred on for further ophthalmic investigation.

Defective visual acuity (using the referral criterion) was detected in 9.8% of preschoolers assessed with SG Singles and 2.5% of preschoolers assessed with SG Linear. This notable difference between SG Singles and Linear may be due to the research design, whereby children claiming that they could not 'see' the SG Linear test were then re-tested with the SG Singles. This re-testing procedure was carried out for the purpose of this project and ensured that the SG linear test was not being abandoned by the preschooler merely due to the test's complexity.

The percentage of children referred for follow up investigation from the Outreach Vision Screening Programme appears to be in line with other studies. Williamson et al[12] investigated an inner city preschool population & reported that "10% of the children who were screened were referred" to a hospital service, and 58.2% of these children were found to have refractive aetiology.

Ingram et al[7] found that approximately 8% of the preschool population screened by them, required follow-up assessments. Verin[24] suggests that "15% of children under the age of 6 years" were found to have a visual anomaly. Fitzgerald[5] reported an incidence of 14.7% of reduced VA detected during screening of three year old preschoolers in Australia.

The high incidence of reduced vision due to refractive error, independent of strabismus detected by screening programmes, is well cited in the literature.[5,7,12] Of the 30 children referred from the Outreach Vision Screening programme 17 attended the Orthoptic Department at Sydney Eye Hospital for follow up assessment. Of these nine (52.9%) were diagnosed with a significant refractive error in the absence of strabismus, two (11.7%) had strabismus, one of which also had a refractive error, three(17.6%) had other ocular pathology and three (17.6%)children had no apparent visual defect on subsequent testing.

Fern[19] reports that, "the need for visual acuity assessment in pre-school age children has been long recognised, yet there are no standardised visual acuity norms or screening criteria." The findings of this study provide valuable normative data for visual acuity assessment in pre-school aged children.

Conclusion

The results of the Outreach Vision Screening Programme confirm statistically that preschoolers find the SG Singles a much easier test to perform than SG Linear, with better visual acuity results being obtained with SG Singles. Also the age factor is relevant when considering the normal level of visual acuity in preschoolers. The level of visual acuity obtained improves with age.

The Orthoptic Department of the Sydney Eye Hospital will continue to carry on the Outreach Vision Screen Programme which has proven to be a valuable service, well supported by parents and the Child Care Centres.

Acknowledgements

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An Historical Look at Amblyopia - from Patch to Patch.

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Abstract

Amblyopia is one of the major areas of treatment in which the orthoptist is an active participant. As the understanding of the pathophysiology of amblyopia evolved so did the treatment attempt to respond. Reviewing the evolution of amblyopia treatment can help one to appreciate the present day treatment. Though it appears that the treatment has remained the same, ie: patching, this is not really the issue as our understanding of amblyopia increases.

Purpose: this paper reviews the evolution of theories about amblyopia and the treatment responses up to the present day.

Amblyopia Review

Throughout the years, the main treatment of amblyopia has remained the same-patching. Though the treatment appears to have remained unchanged, many treatments have been advocated in response to increased understanding of the pathophysiology of amblyopia. Looking at the changes in thinking about amblyopia and the different treatments tried, allows us to appreciate how much we have progressed in our understanding of amblyopia as well as how much we still have to learn.

In the 1500s, straightening the deviated eye by covering the good eye was thought to correct a squint. In order to straighten the eyes, treatment involved wearing nut shells, horned discs or a strabismus mask, one for esotropia and one for exotropia.[1] The problem became apparent that the covered eye was still turned. Amblyopia at this time was not understood to be associated with strabismus.

By the 1700s it was recognized that amblyopia was associated with strabismus. Strabismus was thought to be the single cause of a decrease in vision. The first treatment of amblyopia was credited to DeBuffon who had amblyopia and strabismus. He stressed the importance of full time occlusion as a treatment.[2] The problem was that only some vision deficits improved since this treatment was done on older patients.

In the late 1700s, amblyopia was regarded as a congenital and hereditary problem because improvement of vision with patching was not very successful. One of the earliest methods of partial occlusion was an oversized artificial nose that covered an eye.[3] It was also thought that since an imbalance of the extaocular muscles caused strabismus and decreased vision, treatment by manipulating the extraocular muscles with surgery might improve vision, even in patients with straight eyes.[4] Amblyopia obviously did not improve with surgery. Patching was not really advocated as a treatment at this time.

By the 1800s, amblyopia was recognized by some as a functional problem of binocular vision, and not residing in the eye and extraocular muscles.[5] In 1850, with the introduction of the Ophthalmoscope, the ability to see a normal retina in amblyopia supported the concept of a functional cause. At this time, Javal devised a treatment involving a series of exercises to train the eye and stressed the correction of refractive errors.[6] Donders established the importance of the correction of the refractive errors in the treatment of strabismus.[7] Again, the problem remained that amblyopia was not always cured and strabismus was still present after treatment.

During the 1800s, it became generally accepted that amblyopia was a functional problem residing in central areas of the nervous system.[8] Therefore treatment was directed toward activities other than patching in order to progress beyond the levels that patching was achieving. Treatment with pharmacological agents such as amyl nitrate inhalation, strychnine injections and even eye massages were advocated.[9] These treatments met with limited success.

During the 1800s, two different types of amblyopia were recognized, these being amblyopia of disuse and congenital amblyopia. Amblyopia of disuse occurred at the onset of the strabismus and was treatable. Congenital amblyopia was the remaining amblyopia and could not be improved with patching.[10] Patching was the accepted treatment at this time but no one could agree as to whether full time or part time was most effective. The problem was that there was no explanation for anisometropic amblyopia with straight eyes. There was also a limited understanding of suppression, and also of congenital amblyopia.

In the early 1900s, the concept of functional amblyopia was fully accepted. Worth emphasized that the age of onset of strabismus and of treatment was very important in improving amblyopia.[10] Chavasse named and described amblyopia of arrest as arrested development of vision at the onset of the strabismus.[11] Patching treatment saw the first adhesive patch. Full time occlusion was stressed even to the point of suturing the lids.[12] Worth was treating amblyopia by atropine and blurring the good eye through dilation.[13] Again, not all vision deficits improved. Organized treatment protocol was not fully accepted since some clinicians still claimed that patching did not help.

By the mid 1900s, it was recognized that organic and functional amblyopia coexisted in some patients. This explained why patching did not always succeed. Suppression, as an active inhibition of an image, was appreciated as more important than the concept of nonuse. Since active treatment was felt to be important, several unusual active treatments were tried. One treatment advocated but never successfully proven was the Master Korrector, a type of rotating placido disc. This treatment was used to stimulate both eyes in special spatial orientations which would be a direct stimulation to the cortex.[14] There was still debate whether full or part time patching was the most effective.[15] Still the problem remained that not all vision deficits improved.

During the mid 1900s there grew an increased interest directed toward amblyopia and its etiology. Burian stressed that amblyopia was an ongoing problem from misuse, not disuse.[16] Eccentric fixation was recognized as the main cause of poor visual results. The superiority of the fovea needed to be restored. Treatment with occlusion of the amblyopic eye called inverse occlusion was thought to break the abnormal fixation. A treatment regimen devised by Bangerter was named Pleoptics and consisted of actively dazzling the peripheral retina so the fovea would regain superiority. The pleoptophore was used to accomplish this dazzling.[17] The main problem was that this treatment could only be done with older patients, was costly, time consuming and had limited success.

By the 1950s the scientific method of organizing a treatment protocol was being used for amblyopia treatment. The concept of regaining foveal superiority for vision rehabilitation was accepted. Cuppers modified Bangerter's concept, adding his treatment method concentrating on regaining straight ahead localization of the fovea.[18] The problem was that even with this modification of treatment, eccentric fixation was not eliminated in many patients. It was most apparent that younger patients regained central fixation and improved vision just with direct occlusion of the non-amblyopic eye.

During the 1960s, early treatment was recognized as essential in the recovery from amblyopia. Costenbader stressed the need to treat patients as soon as the strabismus was apparent.[19] Regaining foveal superiority was still a problem in older patients. Since pleoptics was very time consuming, other methods were advocated to regain foveal fixation. One approach placed a red filter before the amblyopic eye while the good eye was occluded, the premise being that since the cones are more sensitive to red, the cones in the fovea would be forced to work.[20] Another treatment involved using prisms to either move the image to the fovea, the direct method, or away from the fovea, the indirect method.[21] Penalization treatment, using cycloplegia to force the patient to use one eye for distance and the other for near was suggested again. Though foveal fixation improved with these treatments, vision often did not.

By the 1970s, Hubel and Weisel had shown that early monocular deprivation during the sensitive period caused permanent visual loss in the cells of the central nervous system. The abnormal binocular interaction and spatial orientation of cells were the essential part of amblyopia.[22] Von Noorden's classification and description of different types of amblyopia

organized thinking about the treatment of amblyopia.[23] Current treatment stresses early intervention on younger patients and full time occlusion. Only a few suggested special approaches have recently been advocated. One is the CAM stimulator treatment that tried to incorporate the new information of spatial orientation into a stimulus for amblyopia treatment. Part time patching with stimulation of different spatial orientations of the amblyopic eye by a rotating disc was thought to lessen treatment time and improve vision.[24] Follow up studies failed to confirm the claimed results.[25,26] Also, a pharmacological agent Levodopa/carbidopa, is showing some short term effect on functional amblyopia but more investigation needs to be done.[27] An effective treatment is still being pursued so that all types of amblyopia can be improved.

Though patching is still and has been the treatment of choice for amblyopia, much has been learned about the necessity for early treatment, correction of refactive errors and the importance of binocular interaction in the treatment of amblyopia. Most treatments, though abandoned or of limited success, helped to move towards the direction and understanding we have of amblyopia. We still struggle with patients whose vision does not improve, but with new research, new treatments will come.

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An Overview of Recent Developments in Automated Perimetric Techniques Used in the Detection of Glaucoma

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Abstract

The use of automated perimeters in clinical ophthalmic practice forms a crucial element in the detection and management of glaucoma. Publication and advertising on the rapid changes in research and development of perimetric techniques can create an information overload to those in clinical practice. The following paper will attempt to provide an overview of recent research developments in automated perimetric techniques and their potential usefulness as a clinical tool in glaucoma.

Key Words:
Flicker, Blue-Yellow, Motion Detection

Glaucoma is a frequent cause of blindness in the elderly population. Recent population-based studies estimate the incidence of glaucoma at between 1.3 to 2.1 percent in the over forty age group.[1,2] In a recent Visual Impairment Study Project in Melbourne, Australia, a population-based sample of 3270 non-institutionalised people over the age of forty years, 1.6 percent were diagnosed with definite primary open angle glaucoma and a further 1.7 percent were suspected of having glaucoma.[3] With an increasing aged population, it has been estimated that by the year 2000, glaucoma will come second to cataract as the most prevalent cause of blindness in the world.[4] With increased demand on health care services to successfully manage people suffering from cataract and glaucoma, a need is created for further research into the processes which underlie these diseases.

Ultimately, the aim of research into glaucoma is to identify disease susceptibility in people before nerve fibre damage can occur. In such people, the development of preventative management techniques could lead to the ultimate control of glaucoma. At this stage in the development of knowledge on glaucoma, research has concentrated on the early detection of the disease and the careful follow-up of established cases. In the quest for therapeutic success, there is demand for the development of more sensitive testing techniques. The requirement is for reliable testing methods which are quick and easy to administer. These tests must be able to accurately indicate whether the disease is present, stable or progressive.

Histopathological investigations on glaucomatous eyes demonstrate that the extent of "silent loss" of ganglion cells, ie. the percentage lost prior to detection with standard perimetry, can vary according to the type of perimetric test performed, the region of retina tested, and the reliability of both the subject and the examiner. The range of variability in undetected ganglion cell loss has been estimated to lie between 15 to 50%.[5] Up to 40% of the optic nerve fibres may be damaged when a 10 dB (decibel) loss is found in the central 30 degree visual field as measured by standard light threshold perimetry. [6]

Based on these findings, various testing methods have been investigated for their sensitivity in detecting early ganglion cell damage. These have included retinal nerve fibre layer imaging, electrophysical and psychophysical investigations of people with, or suspected of having glaucoma. Electro-diagnostic investigations have include electroretinograms and visual evoked

potentials. Areas of psychophysical investigation include the assessment of deficits in temporal sensitivity, contrast resolution, low frequency colour perception and motion perception. These tests are often compared to standard light threshold perimetry as a gold standard.

The development of computer driven static perimetry (automated perimetry) allowed visual fields to be assessed with greater accuracy and reproducibility. Due to the psychophysical nature of the test, the compromises in accuracy from the patients' viewpoint still remain. Such factors include changing psychological states, fatigue and physical constraints.[7] However, automated perimetry minimises the influence of the test operator on the accuracy of the test results. Other advantages include a standardised test protocol, the mathematical analysis of results and the calculation of patient reliability indices. Consecutive automated visual field tests can be statistically compared and analysed in order to detect progressive decline in threshold values of the entire visual field as a whole, or for any region within the field, or for any individual stimulus location.

The study of visual fields in a patient with glaucoma has become an essential component of glaucoma management. Recent advances in the computing software, testing programs and perimetric hardware are aimed at achieving a greater sensitivity and specificity of visual field results. In the quest of developing a visual field test which is faster to perform clinically, hopefully with minimal compromise to test sensitivity, some manufacturers are producing software modifications to the traditional test strategy program. However, standard automated threshold perimetry may not be the test of choice in detecting early glaucomatous nerve fibre damage.

There is evidence of other techniques which may also reflect early glaucomatous selective nerve damage. Spatial contrast sensitivity involves the presentation of alternating (non-flickering) light and dark sinusoidal bars at different spatial frequencies. The minimum threshold contrast at which the bars can be seen at each frequency is then measured. The results of investigations into spatial contrast sensitivity deficits in glaucoma to date have been equivocal. It has been reported that in the absence of significant field loss in subjects suspected of having glaucoma, spatial frequency performance may be compromised within the central visual field region.[8] In another study of central contrast sensitivity, comparisons were made between people with glaucoma, ocular hypertension and normal eyes. It was found that no significant differences could be found between the groups for any of the spatial frequencies tested.[9]

The measurement of contrast sensitivity within the peripheral field as measured by resolution perimetry (or high-pass perimetry) has been extensively investigated.[10-12] The results of these studies reveal contrast sensitivity losses in the peripheral field of people with glaucoma.

Resolution perimetry has been compared to standard threshold perimetry in the presence of glaucoma in several studies.[13-15] The findings in these studies demonstrate that resolution perimetry results are slightly less, if not, equally sensitive to standard threshold perimetry. These results are confirmed when resolution perimetry results are compared to both standard and flicker threshold perimetry.

Sensitivity to short-wavelength light has been reported to be affected by glaucoma. Previous research has explored the use of blue and blue-yellow colour vision deficits as early indicators of glaucomatous damage. Through use of a FM 100-Hue colour vision test it was found that patients with ocular hypertension and such colour vision deficits stood a much higher risk of developing glaucomatous visual field loss over five years.[16] Such sensitivity to central colour deficits have not been confirmed in subsequent studies by Lachenmayr.[11,17]

Recent studies on specific colour sensitivity losses peripherally across the visual field, have offered more promise. Much development in this area of research is based on the principle that early damage to ganglion cells near the fovea result in disruption to the blue/yellow colour wavelength detecting system.[18] Short wavelength light sources are used as target points in the peripheral visual field. Short wavelength automated perimetry (SWAP), otherwise known as blue-on-yellow perimetry, has been reported to be sensitive to early glaucomatous field.[19-22]

In a three year prospective study, Casson, Johnson, and Shapiro, (1993) compared the progression of standard threshold, to flicker threshold perimetry and blue-on-yellow perimetry in early glaucoma and ocular hypertension.[23] It was concluded that both blue-on-yellow and flicker perimetry show sensitivity to early glaucomatous damage and that both techniques were sensitive and specific testing procedures for the detection of early glaucomatous visual field loss.

Two major disadvantages of blue-on-yellow perimetry are the influence of age upon the optical media and the greater magnitude of long-term fluctuation as compared to standard threshold perimetry. Age-related yellowing of the human lens reduces the amount of short wave-

length light reaching the retina, especially blue light. The amount of light transmission loss produced by the lens must be evaluated prior to blue-on-yellow perimetry in order to establish the amount of sensitivity loss that can be attributed to optical factors. The procedures to evaluate this transmission loss can be complex and time consuming.[24]

Newer techniques have been developed to test motion sensitivity deficits in the presence of glaucoma.[25,26] These studies have demonstrated that glaucoma can affect the perception of motion; however, tests of motion sensitivity were not sensitive to early glaucomatous ganglion cell loss and therefore not effective as an early diagnostic tool.

The development and research of perimetric motion tests such as Motion Automated Perimetry (MAP) in glaucoma, have demonstrated a greater sensitivity to glaucomatous visual field loss than larger foveally centred motion tests. The results however demonstrated that MAP did not have an advantage over standard threshold perimetry in detecting localised visual field defects.[27]

Psychophysical investigations into temporal transfer deficits in the presence of glaucoma have been divided into two major areas; critical fusion frequency (CFF) and modulation (flicker) sensitivity measures. Critical fusion frequency measurement represents a point at which an intermittent (flickering) light stimulus characterised by an increasing frequency is first perceived as a continuous light.[22] This represents the highest resolvable frequency. Modulation (or flicker) sensitivity represents the minimum illumination at which a flickering stimulus at a set frequency is first perceived as flickering.[28] This represents the lowest resolvable illumination for a specific stimulus frequency.

Critical fusion frequency deficits in the presence of glaucoma or ocular hypertension have been studied in the past. The findings of these studies are equivocal. Two papers demonstrated definite critical fusion frequency deficits in the presence of glaucoma.[17,29] In 1992, Tyler, measured significant critical fusion frequency deficits at frequencies greater than 20 Hz in the presence of ocular hypertension.[30] However, in a study in which glaucoma was simulated by artificially increasing intraocular pressure, there were no significant measurable critical fusion deficits.[31] It appears that, although the measurement of critical fusion frequency may demonstrate early glaucomatous ganglion cell damage, there may be a more sensitive test strategy available.

In 1990, Toi, Grounauer and Burckhardt compared critical fusion sensitivity to modulated flicker sensitivity in simulated temporary hypertension. It was found that artificially increasing intraocular in normal human eyes produced loss of flicker sensitivity although central CFF was unaltered.[31] These findings have been confirmed in other studies, although there is some dispute as to the frequency at which significant flicker sensitivity loss occurred.[32] Such comparisons between the two strategies of temporal transfer measurement suggest that the measurement of flicker sensitivity may be preferable to critical fusion frequency techniques in detecting early pathological rises in intraocular pressure.

Another technique used to assess temporal (or flicker) resolving power is called multi-flash capimetry.[33] This technique involves the measurement of the minimum interval required for the detection of flicker in a central target. The stimulus is flickering at a constant 5Hz and the area of the stimulus is varied from a 0.625 degree to 20 degree visual field. This technique is reported to be as sensitive, if not more so than conventional standard threshold perimetry, however comparisons between this modality and that of flicker sensitivity have not been made.

Changes in central flicker sensitivity in the presence of glaucoma and ocular hypertension have been extensively investigated.[29,30,32,34-36] The authors of these papers are in general agreement that the presence of glaucoma is accompanied by flicker sensitivity losses, especially at the higher temporal frequencies of 25 to 50Hz . A small percentage of subjects with ocular hypertension also demonstrated mild sensitivity losses.[36] A study of full-field flicker sensitivity deficits in patients with glaucomatous perimetric defects found that sensitivity was significantly reduced in patients with both diffuse and localised field defects.[37]

The development of a flicker sensitivity measurement system in the peripheral retina has resulted in the creation of flicker threshold perimetry. Several investigators have studied influence of glaucoma and/or ocular hypertension on flicker threshold perimetry results as compared to subjects without glaucoma.[38-41] All studies demonstrated significantly greater flicker sensitivity losses in subjects with early glaucoma as compared to an age-matched normal population at all frequencies.

Longitudinal measurements of flicker threshold perimetry in the presence of glaucoma and ocular hypertension were compared to standard threshold perimetry by Casson and Johnson in 1992.[20] It was reported that the early glaucoma subjects demonstrated equally reduced sensitivity to all flicker frequencies. There were flicker threshold perimetry defects present in all subjects demonstrating defects to standard

threshold perimetry. Flicker perimetry provided consistent results which, in many cases, predicted the onset or progression of standard threshold perimetry deficits and successfully identified which subjects with ocular hypertension would go on to develop glaucoma in the near future.

After reviewing the literature, the use of a flickering stimulus in a psychophysical testing procedure appears to provide a sensitive technique for the early detection of ganglion cell loss due to primary open angle glaucoma. In the past, several different modalities have been investigated for their ability to detect early ganglion cell loss in glaucoma. The overall aim of these investigations is to develop a quick, easy and reliable test which was capable of detecting glaucomatous nerve cell damage earlier than current standard threshold perimetric techniques.

Flicker threshold perimetry remains as one of the more promising of these techniques due to its resistance to the confounding effects of media opacities, optical blur and refractive errors, its comparable levels of subject response variability to standard threshold perimetry and its practicality of use in the clinical setting.[42] Standard threshold perimetry is currently used as the clinical gold standard psychophysical technique by which glaucomatous ganglion nerve fibre damage can be documented. Flicker threshold perimetry can co-exist in this clinical environment, and in some cases co-inhabit the same testing equipment,,to hopefully provide a more sensitive diagnostic test of choice for people with early or suspected primary open angle glaucoma.

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Overview of the GDx Nerve Fibre Analyser

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Introduction

Glaucoma is a multifactorial optic neuropathy in which there is a characteristic acquired loss of optic nerve fibres; this loss of fibres typically eventuates in visual field loss.[1] However, glaucoma is now known to 'cause the loss of a substantial number of optic nerve fibres and ganglion cells **without** detectable anomaly in standard perimetry'.[2] Several clinical trials have demonstrated that thinning of the retinal nerve fibre layer (RNFL) precedes visual field defect.[3]

The Nerve Fibre Analyser instrument referred to in this paper is the updated 'GDx Glaucoma Scanning System.' The GDx is a scanning laser polarimeter. It is a diagnostic instrument which provides sensitive testing for the early detection of glaucoma.[4] Around the region of the optic nerve head, the RNFL is known to be the most vulnerable to damage in the glaucoma process.[5] Therefore, by analysis of the thickness of the RNFL around this region, and by comparing the thickness with an age/race matched normative data base, glaucomatous damage can be identified.

The GDx gives precise, reproducible, quantitative, objective measurements of the RNFL. It is important to understand that the GDx measures the thickness of the RNFL rather than the surface topography of the retina. The birefringent nerve fibres cause a low power infra-red polarised light directed towards the eye to split into two parallel rays that travel at different velocities. The light undergoes a wavelength shift proportional to the thickness of the RNFL. The retardation between the rays emerging from the RNFL directly correlates to the nerve fibre layer thickness. Since the cornea also has birefringent properties, the GDx incorporates a "corneal compensator" to correct for the effect this may have on the retardation measurements.[4]

Image Acquisition

The GDx has a liquid crystal display (LCD) monitor as part of the scanhead, which allows for continuous visual contact of the fundus during the acquisition of the retinal image. (See Figure 1.) The patient is set up in front of the scanhead and asked to fixate a green target light. Acquisition of the retinal image involves aligning the optic disc with the centre of the LCD monitor and adjusting both the intensity and focus controls. The GDx is linked to a Microsoft Windows programme which records and analyses the data. The total time of the procedure, including alignment of the optic nerve can vary from one minute to several minutes. This time is dependent upon the patient's ability to fixate the target light in a steady manner. Once a focused image of the optic nerve is visualised by the examiner, the scan is then recorded in 0.7 sec. Approximately sixty-five thousand measurement points (known as pixels) are taken in a 15x15 degree field. These pixels are not affected by magnification error or media opacities. Pupil dilation is not required provided the pupil diameter is greater than 2mm. There is no discomfort experienced by the patient during acquisition of the RNFL image. The data is then processed. The scanning procedure is repeated so that images are obtained for each eye enabling the best to be selected for analysis.[4,6]

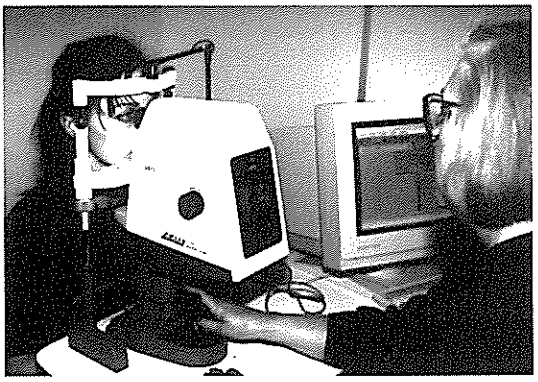

Figure 1. *The GDx Glaucoma Scanning System - Nerve Fibre Analyser.*

Data Analysis

The GDx normative data base was developed by Laser Diagnostic Technologies Inc by collecting data from normal eyes of volunteers between the ages 18 and 80 years of various races. The GDx software automatically compares images to the database and presents the scan results for each eye. Normal ranges were established for a variety of parameters, and the most effective parameters at differentiating normals from those with glaucoma are evaluated.[6]

The final result includes the assignment of an overall score for each eye referred to as "the number". "The number" is an experimental value currently under evaluation. It is derived from the assessment of 200 parameters via the computer's neural network. This network assigns to "the number" a value between 0 and 100, where 0-30 represents normal, 30-70 GDx suspects (ie glaucoma suspects), and 70 and above glaucomatous.[6]

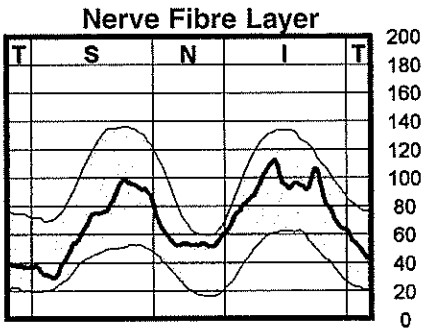

Figure 2. *Double-hump curve of a normal RNFL measured in microns.*

In addition to "the number", the analysis provides a list of values known as "GDx parameters". These parameters include maximum thickness scores, intergrals (ie total area scores) and ratios of the four quadrants examined around the optic nerve head. Parameter results indicate whether the patient is "Within Normal Limits" (p>10%; the chance that the patient has a normal RNFL is more than 10%), "Outside Normal Limits" (p<5%), or "Borderline" (5%<p<10%). Patients in the GDx suspect category may prove to be "Borderline" or "Outside Normal Limits" on some GDx parameters, but not exhibit any visual field loss or other indications of glaucoma.[6]

The crucial step in establishing reliable GDx parameters is the placement of the 'illuminated alignment ring' referred to as the ellipse. The ellipse allows for measurement of the RNFL thickness at a set radial distance from the disc. Physiological cup size is genetically determined and dependant on the size of the disc.[7] This variation in the size of the optic nerve head means the operator must adjust the ellipse using the computer mouse, until best possible placement around the disc is achieved. Once the elipse is in place the operator can obtain statistical analysis by selecting the 'Calculate Analysis Data Button' on the menu bar.[6]

The monitor displays a reflectance/ fundal image and a colour coded thickness map. The dark colours (black and blue) represent thinner areas of RNFL and light colours (red and yellow) represent thicker areas. The reflectance image is like an optic disc photograph. This image is therefore useful in reviewing the cup disc ratio, but more importantly for the purpose of judging the quality of the image displayed. The image shows whether the scan has met the criteria of centration of the optic nerve head, clear focus of the flat peripheral retina and even illumination with correct intensity.

The thickness of the RNFL around the optic nerve head, in the temporal, superior, nasal, and inferior segments is represented by a bi-modal (double hump) curve. (See Figure 2.)
The double hump curve represents an "unrolled view' of the RNFL thickness around the optic nerve head. In the normal subject the superior and inferior RNFL at the retinal rim are thicker than nasal and temporal bundles.[5] Therefore the thicker RNFL in the superior and inferior regions are represented by two peaks on a bi-modal curve. The shaded double hump curve represents age related normal parameters. The line graph which falls within the shaded area is the patient's RNFL thickness in microns.

Summary

In summary, this diagnostic instrument provides a sensitive and quantitative measurement of RNFL thinning prior to visual field loss. The GDx has the advantage of being objective and efficient, with the results being processed and compared to a normative, age related data base.

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The Assessment of Driving Skills in the Presence of Restricted Visual Fields Associated with Retinitis Pigmentosa

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Abstract

The vision standard required by the Roads and Traffic Authority in New South Wales until mid 1997 stated that a driver must have 6/12 visual acuity in the better eye and a peripheral visual field of 130 degrees on the horizontal plane using a standard perimeter with a 3mm target at 1/3 metre.

Three case studies are presented which demonstrate that, in the presence of field loss which fails to meet that standard but matches or exceeds the standard when a larger target is used, the driving skills reflect the standard with the larger target. Assessment of driving performance in the on-road situation is described with reference to the performance of the individual drivers. Remedial strategies are also described.

The outcome for these drivers demonstrates that the visual standard as laid down by the licensing authorities should be used to alert practitioners to the existence of a vision defect and then to refer an affected patient for assessment in the driving situation. The driver's performance can then be evaluated in a realistic setting and where appropriate remediation instigated to achieve a safe driving standard.

Key Words:

Field loss, vision standard, driver performance.

Introduction

Retinitis Pigmentosa is an inherited disease characterised by night blindness and constricted visual fields.[1] Central acuity is often of a good standard. The field loss can be relative, or absolute and when relative, the size of target used demonstrates a range of sensitivity to information in the real world such that large objects are easily seen but small detailed objects are not seen. When driving, the ability to appreciate large objects in the periphery, such as vehicles and people, is important. The identification of detailed information is the role of the fovea and in the presence of adequate central vision this role will continue.

The standard of visual field required by the Roads and Traffic Authority (RTA) of NSW up until mid 1997 was 130 degrees on the horizontal plane using a standard perimeter with a 3mm target at 1/3 metre.[2] This standard on perimeters that have a working distance of 1/3m refers to the Arc perimeter. On the Goldmann perimeter a 3mm size is between the III and IV target size. For both perimeters there are larger targets available and in the presence of a relative field loss the response with the larger targets can be markedly different and approach a normal response. In 1997 the RTA standard was revised and has become 120 degrees using the IV4e (4mm) target on the Goldmann.[3] The target size has slightly increased but the acceptable range decreased which leaves the outcome at a similar level.

It is still not clear what relationship exists between the current standard of the driving authorities and the on-road driving ability. The following case studies demonstrate the inter relationship between the visual standard and driving performance in 3 patients with retinitis pigmentosa and are presented in an attempt to clarify this relationship.

Case Studies

Clinical Information

In summary, each of the 3 patients had a visual acuity level that was within the RTA requirements (6/5 to 6/9), a normal response to contrast sensitivity, bifoveal BSV, appreciation of Lang-Stereotest, and full ocular motility. Each was healthy and alert, and in relation to driving skills had fast reaction time, good insight and excellent planning skills. The response on the visual field assessment by practitioners in the field who used a 3mm target showed an outcome that was less than the medical guideline advises. When a large target such as the V4e Goldmann target (6.4mm) was used the outcome along the horizontal meridian increased to meet or exceed the standard of the guideline. According to the RTA guidelines as the standard had not been met, the licence should be cancelled.

Each of the patients had a strong personal need to continue to hold his/her licence. One was a truck driver who was supporting 3 family members from his earnings, another was an accountant who was required to drive between businesses and the third wished to be able to drive for personal and safety reasons. In the opinion of the referring medical practitioners, all these patients appeared capable of driving safely but their clinical responses supported the cancellation of the licence. For these patients the reality of the RTA standard was called into question. In order to answer this question and with the approval of the RTA medical division the patients were referred for an on-road assessment by the Driver Rehabilitation Team.

The Driver Rehabilitation Process

The team, which consists of the specialist Driver Rehabilitation Occupational Therapist, the Orthoptist and a specialist Disability Trained Driving Instructor conducted an off-road assessment to determine the cognitive, clinical and physical skills of each driver (including an ocular motility and visual field assessment) followed by an on-road assessment with all team members present.

The standard on-road assessment takes up to 50 minutes and follows a specifically planned route that progresses from simple to complex driving situations, namely:

- quiet streets with no lane markings to busy streets with lane markings;
- through quiet shopping centres then busy shopping centres with multiple pedestrian crossings and people crossing.

During this sequence the performance of the driver is evaluated by all team members with the Orthoptist noting the visually related tasks such as:

- the positioning of the car on the correct side of the road, within lanes, when turning corners and on roundabouts;
- the response of the driver to unplanned visual events such as a pedestrian suddenly crossing a road, a car pulling out of a parking spot;
- the use of rear view mirrors and blind spot checking.

Commentary driving, where the driver verbally reports the visual information that is important to that driving situation (road signs, vehicles, traffic lights and pedestrians) is also used. Navigation skills based on visual cues are used to provide good feedback about visual ability and for people with vision problems that change with decreased light conditions. The test includes driving and parking in an underground parking station as well as a driving session at night.

At the completion of the on-road assessment feedback is given to each patient, a report is forwarded to the RTA and, where appropriate, remedial action is taken by the team with the major input from the driving instructor. In the situation where a modification to the vehicle, such as the permanent addition of a convex mirror is required, the patient will have to undertake an additional test with the licensing authority.

Patient Response to the Process

A summary of the main ocular features of each patient and the outcome of the on-road assessment plus follow up information is presented in Table 1.

In each patient there was a marked difference between the field tested with the smaller target size and the largest size available, the 6.4mm size. When tested with the smaller target each patient was well below the required standard and therefore disqualified from driving. When tested with the larger target each patient was able to achieve equal to or better than 130 degrees along the horizontal meridian. One patient gained a full total field and the other two showed a partial loss in the superior and inferior fields. An example from subject 2 of how the effect of a change in the target size can cause a driver to move from licence disqualification (small target) to a normal response (large target) is presented in Figure 1.

In the driving situation each patient drove with an ability that demonstrated visual feedback commensurate with the response for the larger target. There is no routine pattern to these responses but each is typical of driver behaviour in the presence of vision defects. Patient 1 demonstrated a superior field loss yet had problems with road signs in the lower field. This

Table 1. *Summary of Responses for 3 Patients to Driver Rehabilitation Program.*

Procedure	Patient 1 age 43 yrs	Patient 2 age 29 yrs	Patient 3 age 47 yrs
Visual acuity	6/9	6/5	6/6
Field loss	3mm target reported as 30 degrees horizontally, 6.4mm target, 130 degrees horizontally with superior field loss	3mm target reported as 30 degrees horizontally, 6.4mm target; 140 degrees horizontally	automated perimeter 3 degree target 30 degrees horizon, 6.4mm target 140 degrees horizon with inferior quadrant loss on static target
Subjective Opinion about effect of visual standard	driving in reduced light a problem	no effect	safe, he will give up when he is not safe. Avoids car light problems by driving in truck cabin
On-road test performance	**visually:** not using mirrors; position at intersections & roundabouts incorrect; problems with signs in the lower field; satisfactory in dim light **generally:** drives too fast	**visually:** erratic sign identification; using traffic signs to guide actions; no difference in dim light **generally:** bad habits & driving too fast	**visually: daylight** - delayed recognition of internal detail of white background signs. **dusk** - delay for yellow background signs. **night**-internal detail of reflected signs delayed
Remediation	use convex mirrors; increase scanning; commentary driving; lessons to correct bad habits	use convex mirrors; commentary driving; lessons to correct bad habits	during on-road test with conscious scanning delay decreased for detail identification
Outcome	RTA disability test because of mirrors = satisfactory standard gained. On regular review	excellent standard achieved & maintained at 16 month review; personal confidence boost	pending outcome of RTA review.

response is seen in patients who realise that they have a field loss and make an effort to move their eyes to cover the lost vision area.

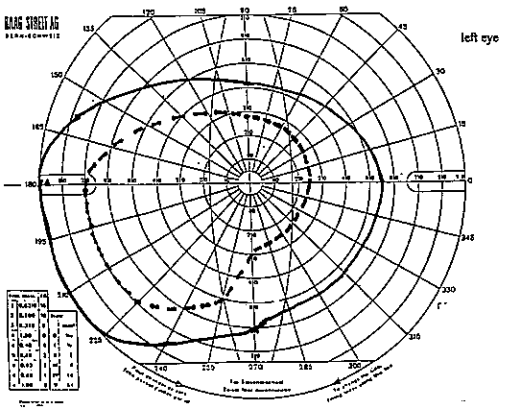

Figure 1. *Goldmann Field of the left eye of subject 2 showing the change in the measurement of the horizontal meridian from 90 degrees with a 2mm target (closet size available to match 3mm) to 140 degrees with the largest 6.4mm target.*

III4e 2mm

V4e 6.4mm

Because they are concentrating on the defective area, in this case the superior field, they fail to retrieve the information in the area directly opposite, i.e. the inferior field. Remediation requires a conscious effort to scan in both areas. This process can be reinforced by verbal reporting (commentary) of all relevant driving information.

Patient 2 with the large sized target had a full field but demonstrated a generalised vision deficiency in the on-road situation shown by failure to report signs but not in any particular segment of the field. She also had a gaze pattern that was restricted to rotation across the horizontal field rather than scan up and down as demonstrated by using the traffic pattern to guide her driving decision rather than look up to traffic lights.

There was a lack of confidence to scan widely which was compounded by driving too fast. High speed reduces the amount of visual information that can be taken in over a given distance.

Patient 3 had an inferior field loss when the large target was used in static presentation. This defect was used to advantage for personal comfort, by placing himself in a driving situation at night so that the glare from oncoming cars would shine into the field defect. This patient also demonstrated difficulties reporting detailed information suggesting that although his visual acuity was clinically satisfactory, his actual skill of reading detail was decreased. Remediation was attempted for this patient by requiring him to consciously move his eyes across the visual field and verbally report all relevant information. His subsequent reporting skill became faster but then decreased as he became tired.

Additional convex mirrors were used for patients 1 and 2. These consist of a Brookstone mirror placed over the rear view mirror and fish eye mirrors that are placed on the side mirrors. The convex structure expands the field of vision seen in the mirror and when accurately positioned will cover the blind spot, thereby eliminating the need for the patient to turn to look to the side. Time saved by avoiding this action can then be given to visually scanning the driving environment. Patients using this approach have demonstrated an increase in driving safety.

The impact of bad driving habits on patient performance compounds the end result of a driving assessment. The longer the time lapse between the gaining of a driver's licence and re-evaluation the greater the opportunity for bad habits to develop. These include driving at inappropriate speeds, failing to observe rules of the road, particularly those rules that have been introduced in recent years. When a patient with bad habits also has a physical defect the effect is to make compensation for the defect more difficult e.g. in the presence of a field loss, approaching a corner at a high speed will reduce the available time to scan to compensate for the loss and reduce the visual information that can be retrieved.

Discussion

The responses of these patients demonstrate several issues. The first is that the visual standard is too simplistic. To cancel a licence because a vision standard is not met does not take into account other factors such as the intellectual and general physical ability of a patient. These factors, which include alertness, insight, reaction time and physical strength, when functioning at a high standard, can assist a patient to compensate for an isolated physical defect such as field loss.

Secondly, this group of patients challenges the validity of the test standard. The standard specifies target size and a numeric response. This group of patients showed that if the procedure is varied slightly the response can increase to approach normal and also that this response better parallels actual driving performance. This favours an approach of using the standard as a guide for recommendation of licence disqualification but also raises the need to consider the standard against the total clinical response of the patient. Where the total response suggests an ability for the patient to cope then the patient should be referred for evaluation by an on-road assessment.

Thirdly, this group of patients supports the value of using the clinical response in conjunction with an on-road assessment. The initial performance of these patients showed that their driving ability was better than the specified standard suggested. It did reveal that the vision defects impacted on the driving performance but at a level that was retrievable. The combination of clinical response with actual skill performance provides a total package which is realistic for the individual driver and the driving community at large.

Fourthly, this group of patients also supports the value of adaptations to the vehicle and driving skills as well as remediation of driving performance in enabling patients with some vision loss to continue to drive but at a level of enhanced performance and safety.

Conclusion

It is considered that vision is the most important sense used in driving. It is therefore important that the standard of vision used is at a level that enables a driver to drive safely. This group of patients raises issues that challenge the practitioner to consider the clinical results in a different light. These patients demonstrate that the standards laid down by the licensing authority serve to alert the practitioner that a vision defect is present which can affect driving performance. The practitioner having been alerted to the existence of a vision defect needs to consider the extent of the defect through a range of clinical procedures and also to evaluate its impact on performance in the driving situation.

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An 'Atypical' Case of Vertical Retraction Syndrome in Association with Klippel-Feil Syndrome

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Abstract

A case is presented of a young child with an 'atypical' vertical retraction syndrome, demonstrated by limitation of ocular movement and lid retraction on depression without any associated globe retraction. The child has Goldenhar syndrome, including a Klippel-Feil anomaly, an external ear malformation and various other congenital anomalies. This case illustrates the wide range of associated anomalies that can present with problems such as Klippel-Feil disorder and Duane's retraction syndrome, suggesting a common teratogenic incident between the fourth and eighth weeks of gestation.

Key words:
Klippel-Feil syndrome, Goldenhar syndrome, Wildervanck syndrome, vertical retraction syndrome, Duane's retraction syndrome.

Klippel-Feil syndrome

Klippel-Feil syndrome was first reported by Klippel and Feil in 1912 [1,2]. The term is currently used to describe persons with congenital fusion of the cervical vertebrae, but this syndrome also has many associated signs. These include cervical problems such as a short neck, limited range of neck motion, low posterior hairline, webbing of the neck, elevation of the scapula, torticollis and scoliosis.[1-3] Both cranial and facial asymmetry may also occur, along with the facial anomalies of cleft lip and cleft palate; ocular anomalies such as coloboma and ptosis; and ear anomalies such as absence of the external auditory canal or ossicle malformation. There may be neurogenic problems such as deafness, facial nerve palsy, synkinesia, spasticity or flaccid paralysis and peripheral abnormalities of syndactyly or thumb hypoplasia. This syndrome may have multiple system involvement, with cardiovascular, renal, urogenital and pulmonary anomalies also reported.[1-3] The disorder is due to a failure of segmentation of somites of the cervical area during embryological development.[1,2]

The association of Klippel-Feil syndrome with Duane's retraction syndrome

With this wide range of associated problems there can be groupings of various anomalies, combining to form 'malformation syndromes'. Wildervanck syndrome (cervico-oculo-acoustic syndrome) describes a condition with the triad of Klippel-Feil anomaly, Duane's retraction syndrome and sensorineural deafness.[4,5] There have been various cases reported of Duane's retraction syndrome with Klippel-Feil anomaly[5-11] and various cases with Wildervanck syndrome have also been described.[5,6,12,13] Goldenhar syndrome (facio or oculo-auriculo-vertebral dysplasia) consists of a similar triad of signs including facial asymmetry, auricular malformations, epibulbar dermoids and vertebral anomalies.[3,5] Many cases of Goldenhar syndrome with Duane's retraction syndrome appear in the literature.[5,6,10,14-19] In a study of 186 cases with Duane's retraction syndrome, Pfaffenbach[5] reported that between 33% and 50% of the cases had one or more other congenital malformations.

Initially Duane's retraction syndrome was thought to be mechanical in origin, but the current theory is that in most instances it would be due to innervational anomalies, a misdirection of the nerve fibres during embryological development [8,12,20] or a spectrum of mechanical, anatomical and innervational factors.[21] Hoyt and Nachtigaller,[22] in a review of the literature, reported various anatomic anomalies, including absence of sixth nerve, extra branches of both the third and sixth nerves to other muscles and anastomoses of the third and sixth nerves.

Aetiology

Both Klippel-Feil disorder and Duane's retraction syndrome have been attributed to a disruption during embryological development.[2,5,7,8] The Klippel-Feil disorder shows a failure of segmentation of the somites of the cervical area. As the vertebrae of the neck are properly segmented in the Klippel-Feil Syndrome, then the teratogenic effect must occur later than the fourth week, when primary segmentation into the caudal and cephalic halves of the body has occurred.[1] At around the same time, the extraocular muscles are developing from three paired, separate masses of mesoderm, each associated with a cranial nerve. Initially, at 26 days, the four oculomotor muscles appear as one mass, then the lateral rectus develops at 27 days and the superior oblique at 29 days. The ocular motor nerves grow from the brain into the muscle masses slightly later, reaching the mesodermal connections at 31 days for the IIIrd and IVth nerves, and at 33 days for the VIth nerve. The muscle cone is in the posterior pole at 5 weeks and the muscles grow forwards towards the globe, fusing at their insertions near the end of the third month. The levator begins to separate from the superior rectus at around 45 to 55 days and is complete during the fourth month.[23,24]

The combination of malformations, ocular, acoustic, auricular, vertebral and other organ systems indicates a disturbance between the fourth and eighth weeks of gestation at the time of development of the ocular muscles and nerves, vertebral segmentation, external ear structures and the formation of organs.[5,6,22,25]

Case details

SM, a 10 month old boy, diagnosed with Goldenhar syndrome, has Klippel-Feil syndrome with the following associated signs:

- fusion of cervical vertebrae
- short webbed neck
- restricted neck motion
- right shoulder higher than left
- left facial hypoplasia, face and jaw
- left external ear malformation
- slight conduction loss in left ear
- hypertonia of upper limbs
- short left leg
- developmental delay
- poor growth
- pelvic kidney
- small muscular ventricular septal defect

Orthoptic assessment

On observation, his left palpebral fissure was wider than the right, with the appearance of left lid retraction. Orthoptic assessment showed the following results:

- Cover Test
  small L hypertropia
- Ocular Movements
  - limitation of left depression in all depressed positions of gaze, only minimal movement below the midline
  - widening of left palpebral fissure on depression
  - elevation of the left eye full, with minimal L/R in elevation
  - horizontal movements, full
  - no apparent globe retraction on depression or elevation

Vertical retraction syndrome

It appears that this child has some form of vertical retraction syndrome in association with the Klippel-Feil syndrome. Vertical retraction syndrome, a much rarer congenital condition than Duane's retraction syndrome, has been variously described in the literature as a limitation of either elevation or depression [26,27] or a limitation of both elevation and depression.[28,29] The retraction of the globe and narrowing of the palpebral fissure may not be as obvious as in Duane's retraction syndrome [28] and again there are opposing descriptions of the retraction, some authors stating that retraction may be seen on either elevation or depression,[27,28] others that retraction may only be evident on depression.[26,29] There may be either orthophoria, hypertropia, hypotropia or an associated horizontal deviation in the primary position.[21,26,28]

Two cases of bilateral vertical retraction syndrome in siblings were reported by

Khodadoust and von Noorden.[30] It was suggested in view of the positive results to forced duction tests, that both of these cases were due to structural rather than neurogenic anomalies, but as electromyography was not performed, paradoxical innervation could not be excluded as the primary cause. Other than a slight ptosis in one case, there were no detectable anomalies of the levator muscle, the changes in lid position occurring with retraction of the globe. The pathogenesis of vertical retraction syndrome is thought to be similar to that of Duane's retraction syndrome.[21]

'Atypical' Vertical Retraction Syndrome

Three cases have been reported of 'atypical' vertical retraction syndromes, the common feature of which appears to be lid retraction on depression.[31-33]

In one case [32] it appears that the levator fires simultaneously with both the superior oblique and the medial rectus, as there is lid retraction of the right eye on both laevodepression and laevoversion, with no muscle underactions. In a second case[33] there appears to be paradoxical innervation involving the levator and the inferior rectus as there is limitation of movement, lid retraction and slight proptosis of the left eye on depression and laevodepression. In a third case[31] the ocular movements appear to be a combination of the 'typical' vertical retraction syndrome (demonstrated by limited elevation of the right eye, limited adduction with downshoot and globe retraction, widening of the palpebral fissure on abduction and a positive forced duction test) and the 'atypical' syndrome which includes the characteristic of lid widening on depression. In this case, electromyographic results showed anomalous innervation of the vertical recti, but the authors were unable to explain the lid widening on depression in the presence of globe retraction.

It seems that the 'typical' vertical retraction syndrome involves some limitation of vertical movement associated with retraction of the globe and narrowing of the palpebral fissure, in a similar manner to the horizontal Duane's retraction syndrome. The two reported cases appear to have structural anomalies, evidenced by forced duction testing. The three reports of 'atypical' vertical retraction syndrome each appear to have the common factor of lid retraction on depression, indicating a paradoxical innervational anomaly between the levator and one or more of the extraocular muscles, and therefore may be somewhat different from the groups of vertical retraction syndrome or Duane's retraction syndrome.

Discussion and conclusion

The ocular motor results indicate that SM may have what others have termed an 'atypical' vertical retraction syndrome,[31-33] with anomalous innervation involving the levator and both the depressor muscles, as the limitation of depression is symmetrical across all depressed positions.

He has been given a diagnosis of Goldenhar syndrome, which includes Klippel-Feil disorder and the left external ear malformation, but there is no evidence of epibulbar dermoids. However, Baum[14] in a review of the literature, found that 24% of the reported cases with Goldenhar syndrome had no epibulbar dermoids. It could also be suggested that SM has an unusual variant of a Wildervanck syndrome, as he has Klippel-Feil disorder, some hearing loss and a variant of a retraction syndrome. It has been stated that a diagnosis of Wildervanck syndrome is still accepted in the absence of one of the three features.[5,6] DeRespinis[21] stated that Wildervanck and Goldenhar syndrome overlap, as can be seen with the multiple cases of Duane's retraction syndrome associated with Goldenhar syndrome.

SM presents as an infant with Goldenhar syndrome, an 'atypical' vertical retraction syndrome with anomalous innervation, a Klippel-Feil disorder and multiple congenital anomalies, presumably due to a common teratogenic effect between the fourth and eighth weeks of gestation. This association of anomalies, including skeletal, auricular, ocular, neural and multi-system involvement illustrates the importance of a general physical examination in each case of Duane's retraction syndrome as suggested by many other authors.[5,6,11,15,18]

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The Physiology and Neurology of Vergence Eye Movements: An Update.

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Introduction

Human eye movements have been classified into three groups: reflex, voluntary and fusional (Bielschowsky, 1956). The reflex and voluntary eye movements are described as conjugate or versional movement which have the characteristics of both eyes moving in the same direction of gaze. Reflex eye movements are controlled by the vestibulo-ocular and optokinetic systems. These two systems have very short latencies and exist to stabilise the eyes for compensatory head movements. Voluntary eye movements are controlled by the saccadic and smooth pursuit systems. The saccadic system has high level velocity eye movements to re-fixate targets from different gaze positions, whereas the smooth pursuit system involves low level eye movements which provide the advantage of tracking slowly moving targets.

The vergence system is the controller for fusional eye movements. Unlike reflex and voluntary eye movements, innervation from the vergence centre will initiate disjugate movements of the eyes, that is, both eyes moving in opposite directions. This type of movement is important in maintaining the state of binocular fixation at various viewing distances. This paper aims to give a summary of the physiology of vergence eye movements and to highlight recent findings concerning the neurology of the vergence system.

Types of Vergence Eye Movements

It has been well known that horizontal fusion has strongest motor control compared to vertical and torsional fusion. This motor fusional component is served by the vergence eye movement system. It has been well documented that horizontal vergence, especially convergence, is composed of several components. In 1893, Maddox (cited in Morgan 1983, p.16) first postulated the four main vergence components including: tonic, fusional, accommodation and proximal vergence. Later, in addition to these four components, Maddox described the additions of disparity, relative and voluntary convergence (Morgan 1980).

Tonic

Due to the structure of the orbit, the anatomical position of the globe is somewhat divergent. The anatomical resting position is demonstrable during deep sleep or anaesthesia as opposed to the physiological resting position of the eyes which is noted in the absence of visual stimuli ie. in the dark. The anatomical divergent angle has been reported to vary from slight divergence (Cogan, 1956) to as much as 68 degrees of divergence (Mann cited in Owens and Leibowitz, 1983), and it is influenced by drowsiness, alcohol, anaesthetic agents, age and stress (Owens and Leibowitz, 1983).

During the state of waking consciousness, there is a continuous tonic innervation to the extraocular muscles to align the eyes in the parallel position, either with or without the presence of a stimulus (Schor, 1980; Morgan, 1980). Maddox (cited in Owens and Leibowitz, 1983) recognised that the magnitude of tonic convergence varied between individuals and that errors in tonic innervation could induce misalignment of the eyes known as heterophoria. Excess tonic vergence resulted in latent overconvergence (esophoria) while latent underconvergence (exophoria)

appeared to be the consequence of a deficiency in tonic vergence.

Disparity versus fusional vergence

The motor response to an abrupt change in retinal disparity can be separated into two stages: the initiation and the completion (Jones and Stephens, 1989; Jones, 1983; Schor,1980). The initial response is referred to as disparity vergence which is a coarse, fast, transient and open-loop mechanism. Disparity vergence is thought to be the primary motor response to disparate targets which can be large or quite dissimilar in characteristics (Stark, Kenyon, Krishnan & Ciuffreda, 1980; Semmlow, Hung, Horng & Ciuffreda, 1994). The completion or fusional vergence is a fine, slow, sustained and feature sensitive mechanism. Its movements bring residual disparity of the eyes to their final accurate bifoveal fixation position under a visual feedback control system (Stark, Kenyon, Krishnan & Ciuffreda, 1980; Semmlow, Hung, Horng & Ciuffreda, 1994). Fusional vergence only responds well to small disparities with the requirements that the images seen by each eye have many features in common (Semmlow, Hung, Horng & Ciuffreda, 1993). These components of the vergence movement system are made almost automatically, but they are not considered reflexes because visual stimuli must be conveyed to the visual cortex in order to induce ocular movements (Bielschowsky, 1956). These types of eye movements are elicited by disparate images and are therefore also considered as psycho-optical reflexes because they occur only when there is sensorial stimulation, that is, disparity of the retinal areas (Bielschowsky, 1956).

Accommodative

The existence of accommodative vergence has been strongly postulated following Muller's experiment (Leigh & Zee, 1991). By asking a subject to fixate a moving target from distance to near with one eye while the other eye was covered, it was reported that the eye under the cover slowly converged as the target moved closer. This meant that the blurred stimulus not only stimulated the accommodation mechanism but also initiated the vergence system, and therefore there must be a link between accommodation and convergence. Accommodative vergence is described as being significantly reduced with age due to the presence of clinical presbyopia (Ciuffreda, Ong & Rosefield, 1993). It has been noted that accommodative vergence is reduced with increasing age as the accommodation becomes weaker (Nuzzi et al. cited in Hokoda 1991). These authors stated that there is also an increase in proximal vergence with age to compensate for the reduction of accommodative vergence.

Proximal

Proximal convergence is the ability to converge the eyes as a result of knowledge or awareness of the nearness of an object. Thus this component is variable in magnitude and involves more of a psychological basis (Hokoda & Ciuffreda, 1983). Proximal vergence accounts for 50% of the near vergence response under open looped conditions. The amount of proximal vergence is estimated by calculating the difference in the AC/A ratios calculated by heterophoria and the gradient methods. This value is thought to range from 0.5-2.0 Δ/D (convergence-Δ/ distance-D) (Hokoda & Ciuffreda, 1983). Studies by Schapero and Nadell (1957) on the changes of accommodation and vergence in 16 subjects aged between 30 and 74 years (30-39 =8 subjects; 40-49 = 6 subjects; >70 = 2 subjects) showed that proximal vergence reduced with age. However, Sheedy and Saladin (1975) claimed that proximal vergence remained constant in their study of twenty three subjects (13 non-presbyopic subjects aged 22-29, and 10 presbyopes aged 51-75). Recently, Hokoda and his co-workers (1991) studied the proximal vergence of 106 subjects aged between 6-47 years (mean age = 28.5). They showed that there was no significant correlation between proximal vergence and increasing age (Spearman's $r = -0.06$). However, the number of subjects in each group was not equal. Most of the subjects were in the 21-30 and 31-40 groups, and there were only 12 subjects over 40 years of age. This therefore reflected the smallness of the presbyopic group which was not able to show a significant correlation between proximal vergence and increasing age.

Relative

Relative convergence is the amount of convergence that can be changed in relation to a set amount of accommodation without producing diplopia (von Noorden, 1990). Conversely, relative accommodation is the amount of accommodation that can be changed in relation to a set amount of convergence without the perception of blur. von Noorden (1990) stated that these amounts could be modified with training. Exercise to produce convergence in excess of accommodation is referred to as positive relative vergence, whereas exercise to train accommodation in excess of convergence is known as positive relative

accommodation or negative relative vergence (von Noorden, 1990).

Voluntary

The ability to perform convergence of the visual axes with effort under the absence of visual stimulus is known as voluntary vergence (Morgan, 1980; Eskridge, 1971). Voluntary vergences have similar characteristics to other voluntary eye movements such as saccadic or smooth pursuit movements. This means that the movements are controllable and can be varied in magnitude (Eskridge, 1971). However, not everybody can converge their eyes voluntarily, and this ability appears to require training or practice (Bielschowsky, 1956; Eskridge, 1971; Morgan, 1980).

Physiology of vergence eye movements

Stimulus for vergence eye movements

The vergence system is comprised of several components and there are therefore several stimuli that can initiate vergence; a blurred stimulus will initiate accommodative vergence, a knowledge of nearness will initiate proximal vergence and the disparity vergence will be induced by retinal disparity. However, the two primary stimuli that evoke the initiation of convergence are the disparity and blur of the stimulus, which ultimately initiates fusional and accommodative vergence (Leigh & Zee, 1991; Zuber, 1971). Accommodative and disparity vergence have generally been considered as the two main components for the alignment of the eyes during convergence, whereas proximal convergence has traditionally been considered as secondary. Wick (1985) claimed that proximal vergence comprised up to 70% of the steady state near vergence response. Joubert and Bedell (1990) found that the contribution of proximal vergence varied from 35-64% of the total vergence responses. The most recent study by North and associates (1993) comprised of 18 subjects, aged 18-23, with normal binocular vision, looked at the contribution of the proximal, accommodative and disparity vergences in the normal vergence responses. Their results showed that proximal vergence was the most important component, contributing 45% to the total vergence response, followed by disparity (41%) and accommodative vergence (14%). Wick and Bedell (1989) studied seven normal binocular vision adult college students and showed that proximal vergence had a greater velocity than disparity and accommodative vergence, and therefore it was the major component for rapid eye alignment. These findings were confirmed by Wick and Bedell (1992). Overall, the initiation of a particular component of vergence of depends upon the nature of the stimulus. In the condition where fixation changes rapidly from distance to near, then proximal vergence is probably the main component to inititate this change. If the fixation stimulus moves slowly towards to the viewer, the accommodative and disparity vergences are thought to be the main components for the vergence response. It is important to note that the total vergence movement is an additive process, therefore all the components are important for maintaining binocular vision. However, one component might be more important than the others under differing conditions.

Dynamic properties of vergence eye movements

Vergence eye movements have relatively long latencies. The average latency for vergence movements is approximately 160 msec, which is the second longest latency after the saccadic system which has a latency of 200 msec (Gay, Newman, Keltner & Straud, 1974). There are also some variations in latency of different types of vergences, such as fusional vergence (160 msec) which has a shorter latency than accommodative vergence (200 msec) (Leigh & Zee, 1991). The vergence system also has the slowest eye movement velocity of approximately 20°/sec (Gay at al., 1974). The dynamic properties of vergence eye movements are that they are very slow and it might take as long as one second to complete. It is thought that the velocity of vergences is increased when the target of interest changes its position across the visual field as well as in depth (Leigh & Zee, 1991). This means that the vergence movement is faster if it is combined with a versional movement. Furthermore, it has been suggested that the command signal for pure vergence movements is a step (tonic) change in innervation to the extraocular muscles. Unlike the saccades, the dynamic properties of vergence responses are considerably more variable. They are related to the type of the stimulus, that is, disparity, blur, size, the condition of viewing and whether or not it is associated with a saccade or a blink (Leigh & Zee, 1991).

Compared to horizonal vergence, vertical vergence is much slower, taking as long as 8 seconds for completion (Kertesz, 1983). Vertical fusion cannot usually overcome disparities of more than a degree, even though part of the fusional response (≈20%) is accomplished by sensory not motor processes (Kertesz, 1983). Cyclodisparities also elicit cyclotorsional movements but like the

vertical vergence, they are slow and have a limited range, and the motor component is very weak because there is a strong sensory component to cyclofusion (Kertesz & Jones 1970, and Kertesz, 1983).

Accuracy of vergence eye movements

There is a constant state of error in vergence eye movements. Vergence movements recorded by direct photographs, corneal reflections and oculography show steady state errors varying from 2 minutes of arc to as large as 2° (Zuber, 1971). Recently, Cornell (1995) claimed that these errors could be as high as 8.2° for peripheral and 5.5° for central targets in normal subjects. These measurements were recorded by the Ober2 infrared eye movement system. It seems that the level of accuracy in vergence eye movements is inversely proportional to Panum's fusional area. When Panum's fusional area is narrow, the vergence movements need to be more precise to keep the targets within Panum's fusional area so that binocular single vision is appreciated. Since Panum's fusional area varies with central and peripheral fixation, and with stimulus parameters, the level of accuracy in vergence eye movements will depend upon the nature of the stimuli and the test conditions.

AC/A and CA/A ratios

The relationship between accommodation (A) and accommodative linked convergence (AC) can be expressed as the AC/A ratio (prism diopters/sphere diopters). As the vergence angle (Vθ) in prism diopters is equal to the IPD (cm) divided by the viewing distance 'd' (m), the calculated ratio would be 6:1, given that the average interpupillary distance is six centimetres. However, the clinical AC/A ratio is usually smaller (4:1). Thus, during binocular viewing of near objects, disparity/fusional vergences must also be functioning to align the axis correctly. It has been noted that the AC/A ratio remains relatively constant with age (Hokoda, Rosefield and Ciuffreda, 1991). Contrary, findings by Bruce, Atchison & Bhoola (1995) show that the AC/A ratio increased (average 0.126 Δ/D per year) with age. Yet, not only is convergence (C) linked to accommodation, but accommodation is also linked to vergence (convergence linked accommodation) (CA/C). The clinical CA/C ratio, the amount of accommodation in sphere diopters induced per prism dioptre of convergence, is typically about 0.1:1. In fact, this ratio should be about 0.16:1 if the amount of convergence linked accommodation were just equal to that required for clear vision at all distances. The CA/C ratio is thought to decrease with age (Bruce at el., 1995). Bruce and his co-workers (1995) indicated that the mean CA/C ratio decreased from 0.1D/Δ at the age of 20 to 0.03D/Δ at the age of 40. Overall, because of the differences in the exactness of AC/A and CA/C ratios, accommodative and fusional drives must work together to enable the acquisition of clear, single objects at all distances.

Neurology of vergence eye movements

Control centre for vergence eye movements

It has been well described that the paramedian pontine reticular formation (PPRF) is the hortizonal gaze control centre for versional eye movements and its major output is to the ipsilateral abducens nucleus. The abducens nuclei have two types of neurons: abducens motoneurons which supply the ipsilateral lateral rectus and abducens internuclear neurons whose axons decussate at the nucleus level and ascend together with the medial longitudinal fasciculus (MLF). These axons synapse with the medial rectus motoneurons of the oculomotor nucleus. The axons from the medial rectus motoneurons supply the ipsilateral medial rectus muscle. Lesions at the MLF give a clinical syndrome known as internuclear ophthalmoplegia (INO) which has a characteristic limitation in adduction of the ipsilateral eye when attempting a versional movement (conjugate eye movements to the oppposite side of the lesion). It is interesting that the vergence eye movements are usually unaffected even in the case of bilateral INO. Based on these phenomena many authors (Leigh & Zee, 1991; Lawler & Cowey, 1986; Cogan, 1956; Mays, 1984) have agreed that the vergence system is neurologically independent of the conjugate eye movement system.

The possible localisation of a vergence control centre has been reported by many authors. A study of the oculomotor nucleus morphology by Buttner-Ennever and Akert (1981) showed that the medial rectus motoneurons has three distinct segregations of cells named A, B & C. Group A cells are located in the ventral and rostral parts of the oculomotor nucleus (OMN). The average diameter of cells in group A is 26μm (Buttner-Ennever & Akert, 1981). B group cells are situated dorsally and caudally, and they have an average diameter of 30μm (Buttner-Ennever & Akert, 1981). Group C cells are located dorsomedially and rostrally. This cell group has its diameter (average diameter = 18μm) significantly smaller than group A and B cells,

and it supplies smaller muscle fibres of the medial recti (Buttner-Ennever & Akert, 1981). Therefore, the group C cells are thought more likely to be involved in generating slow eye movements such as vergence, but there is no physiological evidence to support this hypothesis (Buttner-Ennever & Akert, 1981).

Early studies showed that cells in the mesencephalic reticular formation change their activities during vergence eye movements. Schiller (cited in Mays, 1984) showed a small number of neurons recorded in the dorsal lateral region of the caudal part of the oculomotor nucleus complex, whose activities increased during vergences. Later, Mays (1984) made a thorough study of such neurons, and showed that there were cells located dorsal and dorsal lateral, approximately 2mm within the medial rectus motoneurons of the oculomotor nucleus (OMN), that changed their firing rate approximately linearly to the vergence angle. Recently, Judge and Cumming (1986) showed that most of the near response cells were recorded in a region extending only 1mm above the OMN and 2.5mm above the magnocellular red nucleus (mRN). The majority of these cells were close to the dorsal limit of the oculomotor or red nuclei, and a few cells may have been in the pretectum or rostral superior colliculus. According to the changing activities of these cells, Mays (1984) subdivided these cells into two main groups: convergence and divergence cells.

Convergence cells

Mays (1984) stated that there were neurons (68 cells) near the oculomotor nucleus that increased their firing rate with convergence eye movements, but that there was no change in the activity of these cells for purely conjugate eye movements. During steady fixation at any vergence angle, the firing rate of these cells is constant. Furthermore, the firing rate of convergence cells is a linear function of the vergence angle and is unaffected by the direction of conjugate gaze. The average firing pattern over the vergence angle is 10.4 spikes.s^{-1}.deg^{-1}. Once the near fixation has stabilised, firing rate of these cells remains nearly constant.

Divergence cells

The divergence cells were found intermixed with the convergence cells (Mays, 1984). These cells have a steady firing rate inversely proportional to the vergence angle, that is, the rate decreases during convergence and increases during divergence. There is no change in the firing rate during conjugate eye movements. These cells have an average firing rate of 10.3 spikes.s^{-1}. deg^{-1} (Mays, 1984).

Mays and Porter (1984) also reported that there was a group of cells found in the vicinity of these vergence cells that also changed their activities to vergence movements. The activities of these cells were thought to be related to vergence velocity. Later, this finding was confirmed by Mays, Porter, Gamlin and Tello (1986). According to the activities of these cells in response to vergence movements, they have been classified into three main types; convergence burst cells, burst tonic cells and divergence burst cells (Mays et al., 1986).

Convergence burst cells

These cells give a vigorous burst of activity just before and during all convergence eye movements, but are not modulated during purely conjugate eye movements or during fixation. There are about 58 cells that have this characteristic and they have an average firing rate of 4.45 spikes.s^{-1}/deg.s^{-1} (Mays et al., 1986). The average amplitude, as spikes increase, is about 6.37 spikes/deg (Mays et al., 1986). Most convergence burst neurons are found ventral to the mesencephalic reticular formation, dorsal and lateral to the oculomotor nucleus. Some convergence burst cells are also found in a more dorsal region which may also include some pretectal nuclei, rostral to the superior colliculus (Mays et al., 1986)

Burst tonic cells

Most of these cells burst for a small vergence movement (up to 2°) and show little tonicity unless a large vergence is made (Mays et al., 1986). These tonic rates are indicated to be proportional to the vergence angle (Mays et al., 1986). Most burst tonic cells are located at the area similar to the near response cells, and some are located approximately 5mm dorsal and 5mm lateral to the oculomotor nucleus (Zee & Levi, 1989).

Divergence burst cells

This cell type has a far smaller number (8 cells). Similar to the convergence burst cells, these cells only increase their firing rate just before and during divergence (Mays et al., 1986). The divergence burst cells are often found in the ventral region of the vergence angle cells, and their firing rates are correlated to divergence velocity (Mays et al., 1986).

The activities of medial rectus motoneurons during vergence eye movements

Robinson and Keller (1972) reported that the medial rectus motoneurons displayed a step

change in firing rates in response to vergence movements. This means that at the beginning of each movement, the rates change abruptly to a new level and stay there both during and after the movements. Later studies by Mays and Porter (1984) showed that there was a gradual change during symmetrical vergence. Mays et al. (1986) stated that near response cells (NRCs) could only carry either eye position or velocity signals to the medial rectus motoneurons. Recently, Zhang, Gamlin and Mays (1991) reported that these cells carried both eye position and vergence velocity. This has been confirmed by Gamlin and Mays (1992) by using single unit recording techniques. In addition, Zhang, Gamlin & Mays (1991) reported that NRCs are predominantly related to vergence and not accommodation, as reported by Judge & Cumming (1986). However, Zhang, Mays and Gamlin (1992) suggested that most of NRCs were driven by both the blur and disparity controller but differ in their accommodative and vergence coefficients. None of these cells however, had a high coefficient for both accomodation or vergence.

Internuclear pathways of vergence system

Oculomotor neurons (III CN) change their activities during versional as well as vergence eye movements suggesting that both vergence and versional eye movement commands are joined at this level before the signals are sent to the extraocular muscles. However, it is obvious that the connecting pathways between the vergence control centre and the OMN will certainly not travel through the MLF. Cogan (1956) suggested that the supranuclear pathways for vergence were along the anterior branchia and the superior colliculi rather than by way of the pons. Mays (1984) claimed that abducens internuclear axons carried only versional control signals. However, Gamlin and associates (1989) reported some inappropriate signals sent to the MLF during convergence.

Mays (1984) and Judge & Cumming (1986) suggested that convergence and divergence cells provided a monosynaptic drive to the medial rectus motoneurons. By using an antidromic microstimulation technique, Nakao's (1986) experiment in twenty-four anaesthetised adult cats showed that the neurons in the vicinity of the oculomotor nucleus projected directly to the medial rectus subdivision of oculomotor nucleus. Nevertheless, since the animals were paralysed, there was no evidence to support that these cells being near response cells. Recently, by using a similar technique to the one described in Nakao's experiment, Zhang, Gamlin and Mays (1991) conducted a study on two trained monkeys to identify the cells that project to the medial rectus neurons of the oculomotor nucleus and to see whether these cells were related to the NRCs. Their results showed that there were twenty-eight cells that were antidromatically activated from the ipsilateral medial rectus neurons and that these cells also behaved as NRCs. This was the first electrophysiological evidence to support the direct monosynaptic projection from NRCs to medial rectus motor neurons (MMNs) of the oculomotor nucleus.

There is still a lack of evidence showing whether the projection of the NRCs to the MMNs is ipsilateral, contralateral or bilateral. Zhang et al. (1991) claimed that failure of the NRC to activate from the contralateral oculomotor nucleus suggested that vergence input to the medial rectus neurons was ipsilateral. However, it is already known that symmetrical vergence needs imput to be sent to the MMNs from both sides of the brain. Furthermore, unilateral electrical stimulation of the midbrain where NRCs are located causes adduction of both eyes. If direct input to the medial rectus motorneurons occurs ipsilaterally, then it is possible that the NRCs on both sides of the brain have a common input or are cross-coupled (Zhang et al., 1991).

Many authors have provided evidence that the abducens nucleus decreases its firing rate during convergence and increases its firing rate during divergence (Robinson & Keller 1972, Mays 1984, Judge & Cumming, 1986). If the vergence commands pass directly to the oculomotor nucleus, then how are the abducens motorneurons receiving the vergence signal during disjugate eye movements? Leigh and Zee (1991) suggested the possibility of a projection from the interneurons (dorsal to the oculomotor complex) and projection to the ipsilateral and contralateral abducens nuclei. By using a double retrograde labelling technique in 12 adult cats, Maciewicz and Phipps (1983) provided evidence that a small percentage (16%) of the internuclear oculomotor neurons had bilateral projection to the abducens nucleus. Mays and Porter, (1984) reported that only 2/28 abducens motor neurons had the characteristic related to vergence, that means there was a significant increase in firing rate during vergence movements while decreasing the activity for conjugate eye movements.

Clendaniel and Mays (1994) studying 4 monkeys, identified 18 oculomotor internuclear neurons (OINs). These cells behaved similarly to medial rectus motoneurons during vergence and versional eye movements and non OINs sensitive to vertical eye position. Microstimulation of the

oculomotor nucleus where both the OINs medial rectus motoneurons were located, resulted a large adducting of the ipsilateral eye and a small abducting of the contralateral eye (could near response cells be versional eye movement response cells?). This evidence suggested that most of these cells innervated the contralateral abducens nucleus. Lidocaine injection to the OINs location resulted in hypometric and slowed adducting saccades to the contralateral side of the injection site that suggested crossed OIN pathways are excitatory. However, these signs have not been reported in a complete third nerve palsy clinically. Are OINs only for versions but not vergences?

Supranuclear control of vergence eye movements

Mays (1984) claimed that there was some vergence paralysis due to lesions at the midbrain following trauma but the vergence centre could not be localised because this usually involved multiple lesions. Some of the cortical centres which are thought to have a role in vergence movements include: superior colliculus, cerebellum, frontal eye fields, occipital lobe, parietal lobe.

Leigh and Zee (1991) reported that stimulation of the pontine tegmentum between the two MLF, or in the third nerve nucleus itself, produces vergence movements. Further more, ablations of the cerebellum in monkeys transiently impairs vergences. Electrical stimulation of Brodman's areas 19 and 22 in the occipital cortex gives different combinations of the components of the near triad, and stimulation of the frontal eye fields produces convergence movements (Leigh and Zee, 1991). Bilateral stimulation of the frontal and occipital lobes has been done (Cogan, 1956), but the observations have been too few and too inexact to permit localisation. Weakness of convergence has followed bilateral occipital lobe injuries, ablation of the cortex around the angular gyri has been reported to result in paralysis of convergence with a disturbance of depth perception (Cogan, 1956).

Westheimer and Blair (1973) examined vergence eye movements on four monkeys who had had cerebellectomies. The results showed that no vergence was demonstrated after a complete removal of the cerebellum and suggested that the cerebellum played an important role for vergence as well as versional eye movements.

Lawler and Cowey (1986) studied the effect of bilateral pretectal and superior colliculus lesions on binocular vision. Nine monkeys with different sites of bilateral cortical ablations were included; three monkeys who had areas 5 and 7 of the parietal lobes ablated, three other monkeys who had inferotemporal ablations and another three had frontal eye fields (area 8) ablated. The three monkeys who had frontal eye fields ablated, then had bilateral superior colliculi ablated three months later. These results showed that those monkeys with superior colliculus and frontal eye field ablation demonstrated more missearching errors when they performed under binocular viewing conditions. The authors indicated that searching errors were due to diplopia and were only evident at near, which was thought to be caused by a deficiency in vergences. Since the monkeys had both superior colliculi and frontal eye field ablations, and also there was no visual discrimination task given to the monkeys before the superior colliculi ablation, it could not be stated that limitations in vergences were due to lesions of superior colliculi. There were minimal missearching errors due to parietal lesions and no significant errors resulted from inferotemporal lesions.

Otsuka, Maekawa, Takeda, Uede and Chiba (1988) reported the case of a thirty-one year old man who had blurred vision and diplopia due to accommodative and vergence paralysis following a middle cerebal artery occlusion. Recently (1993), Ostuka reviewed horizontal eye movements of a nineteen year old man with arteriovenous malformation with haemorrhage in the right cerebellum (which was revealed on arteriography and computer tomography). Magnetic resonance imaging showed a haematoma located on the right, medial and inferior cerebellar peduncles and the hemispheres. Apart from gaze evoked nystagmus, cogwheeling smooth pursuit, hypermetric right saccades and sometimes hypometric left saccades, the vergence eye movements were absent. Accommodation was normal and pupils were constricted at near. The peak velocity of horizontal saccades was within the normal range that indicated the burst neurons in the PPRF and motorneurons were still intact.

Studies of the frequency of convergence insufficency in traumatic brain injury (TBI) on 98 patients in the Loewenstein Rehabilitation Hospital (72 follow-up and 26 hospitalised patients) by Cohen, Groswasser, Barchadski and Appel (1989), showed that 42% of follow-up patients and 38% of hospitalised patients had insufficiency of convergence. The authors also noted that convergence insufficiency was significantly associated with dysphasia and cognitive disturbance but was not associated with behavioural disturbances. The authors claimed that long standing vergence insufficency is fairly common following severe TBI.

Conclusion

In conclusion, the existance of the vergence eye movement system facilitates binocular fixation at various distances. The cortical areas that have a role in vergence movements are thought to be the frontal eye fields, Brodman's areas 19 and 22 in the occipital cortex, the superior colliculus, parietal lobe and the cerebellum. The supranuclear pathways that connect these cortical areas to the vergence centre are still unclear, however, it is thought to be via the anterior branchia and the superior colliculi rather than by way of the pons. The vergence centre is located dorsal and dorsolateral to the oculomotor nucleus. Outputs from the vergence centre project ipsilaterally to the medial rectus motoneurons. This projection is thought to be monosynaptic and it is cross-coupled. Unfortunately, there is little understanding about the neurology of vertical and torsional vergence. These types of vergences are very small in amplitude and usually do not exist in normal free-space viewing conditions. Overall, the neurophysiological aspects of horizontal vergence eye movements have been studied more thoroughly over the last decade. However, most of these works have been concentrated on the midbrain area. There are still many doubts about the higher vergence control areas and their projections to the vergence centre. Therefore, more studies related to the supranuclear pathways need to be done in the future to give a better understanding of the neurology of the vergence eye movement system.

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