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CONTENTS

EDITORIAL

THE PATRICIA LANCE LECTURE

In 1987 the Council of the Orthoptic Association initiated an annual lecture to be given at the opening of our Scientific Meetings to honour Patricia Mary Lance on her retirement from her position as Head of the School of Orthoptics at Cumberland College of Health Sciences.

Miss Lance completed her training in 1941 in the second training course in Australia. She was a founding member of the OAA and has held the office of President for a record six terms, in addition to her continuous contributions in other capacities and her presentations at Scientific Conferences. She was made an honorary member of the OAA in 1965, and this was converted to that of Fellow when this award was introduced in 1973. She assisted in the formation of the International Orthoptic Association and became Australia's first representative on its Council of Management. Following a long involvement in orthoptic education she was appointed in 1973 as the first Head of School of Orthoptics at the New South Wales College of Paramedical Studies (shortly to be renamed Cumberland College of Health Sciences). She was honoured in 1979 with an MBE for her services to orthoptics.

I was given the honour of giving the inaugural address in Sydney in 1988, which is included in this edition as an Editorial. Alison Pitt's lecture, given at the 1989 meeting in Brisbane, is also printed in this issue. It is hoped to publish future lectures in subsequent editions of this journal.

Occasionally Pat has described me as one of the few Orthoptists in NSW who was not one of 'her' students. I really think that this is no longer the case. Whereas most of the others were her students for only two or three years, I have had the opportunity to be with her and learn from her for eleven years.

Pat's career in Orthoptics has spanned many developments, and, whilst it has been popular at recent conferences to look to the future, with themes such as 'Orthoptic Horizons', I feel that a great deal can also be learned by looking at our past. What were our origins, why were we needed, what niche did we fill?

Clearly, we were specialists in binocular vision, or, more particularly, in the use of exercises to treat amblyopia and squint. Orthoptists were needed not only because of the many hours needed for treatments in those days, but also because of the complexities of the sciences of binocular vision and ocular motility. To be frank, many ophthalmologists were only too happy to direct responsibilities for the management of these cases to the orthoptist.

However, times have changed, ophthalmologists are now well educated in these areas. This has helped us by providing a more equal base of respect and understanding, as is evidenced by our conjoint scientific meetings. But I feel that this understanding has not always extended to the potential for orthoptic treatment in the management of many patients, and alternatives to surgical treatment are not always considered seriously. This means that the skills of the orthoptist working, as is often the case, in an ophthalmic setting, may be confined to routine measurement.

Certainly this situation has evolved to fit the common pattern, but the question remains, who will give the eye exercises?

We now have a well educated public, educated not only through formal schooling but also through the often more effective media. The status of the medical profession is not as hallowed as it was a generation ago, and patients are more likely to question the doctor and look for alternative methods of treatment, particularly if the treatment involves surgery on their child's eyes. There is also a concomitant and not unrelated rise in the popularity of 'alternative medicine' as orthodox management does not always give the patient what he wants, be it a cure or just a sympathetic ear. Who will provide the alternative to squint surgery, or treatment for symptoms arising from accommodative problems or decompensating heterophoria if the orthoptist doesn't? Others, without the orthoptists education, will, and are doing so already.

But will we still have the skills to provide this treatment, other than a few simple convergence

exercises? We may look to other areas of ophthalmology to provide directions for our future, yet the developments of technology mean that the skills of refraction and visual field assessment may become redundant in the future. What technology is there that will replace the skills of teaching a person with an intermittent exotropia to control his deviation, or the newer areas of therapy such as rehabilitation of the low vision patient? The role of eye movement training has now extended beyond that directly related to squint or heterophoria into the management of visual field problems, eccentric viewing training and null point training.

Before I am branded a complete reactionary, let me make two things quite clear:

- I am in complete agreement with the development of orthoptics into areas of general ophthalmology. This has meant that the graduating orthoptist has a wider knowledge of the eye, its anatomy, physiology, optics and pathology, and of the whole body that the eyes serve. It has meant that orthoptists can choose to specialise in other areas of vision and research if they wish, and there are several examples of such people in our profession. Indeed, a broader education may be one of the answers to the issues I am addressing.
- Secondly, I am not advocating a blinkered return to orthoptics of the past. Certain treatments have been found to be ineffectual, unnecessary and a waste of time and money. There is no doubt that surgery is the only form of effective treatment in many cases of squint. Modern research into the neurology of binocular vision has shown us why it is pointless to attempt to restore binocular single vision in squints where it has never developed. Indeed, I am anxious that we don't fall into the methods of others that we criticise, and dress up unfounded, even quack remedies, in the guise of professionalism. The time has passed when we could give treatment and demand payment (or expect the taxpayer to support us in a publicly funded institution) just because we think that it works. The public has a right to demand evidence of the efficacy of our skills, a concept now known as accountability.

For this reason, we must direct more of our time to systematically evaluating our treatments. Other professions do it, it is one of the hallmarks of professionalism. For 15 years orthoptic students have been studying statistics and research methods as part of their college based education, but why is it that, when final year students are carrying out research projects, I see a reflex dilation of the pupils, a blanching of the skin when I suggest that a statistical analysis of their results may be appropriate? Most submit, resigned because we hold the power of marking the assignment, but I get the distinct impression that many, freed at last from college control, would not consider using simple research methods and statistical applications to their clinical findings. Why is this so, why is it that so much of the research in orthoptics is found outside our profession?

It is hoped that the new degree courses in orthoptics may help some of these issues, but I also feel that a major problem is a lack of a strong professional identity, that is, a lack of identity in what we do, rather than what we are. Are we specialists in our own area, or are we technicians. We started as a profession with admittedly a very narrow focus, but have we blurred the focus by broadening our role?

I started by looking back, and I will close in the same way. My first day as an orthoptic student involved turning up at 127 Collins Street in Melbourne in the early 60s. Those of you privileged to have also been to this address, now unfortunately engulfed in a modern hotel, will know that Beverly Balfour ran her practice there from her delightful flat on the top floor.

The main consulting room was a wonderful mix of orthoptic equipment, Persian rugs and all kinds of bric a brac, including a magic monkey that children looked at if they wanted to see a few extra letters on the vision chart. But the best feature was a large table piled high with old Christmas cards, nail polish, cellophane, scissors, coloured pins and many other bits and pieces. Each patient had a carefully designed exercise made to suit his or her particular needs, to inhibit or stimulate convergence, to discourage suppression or overcome amblyopia. This was my first impression of orthoptics, and I feel that it still represents the core of our identity. Although the methods we use may change with changing knowledge and technology, the skills of orthoptic treatment will always be needed. We can do it, we can do it well, and if we don't, others will.

Elaine Cornell

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ACCOMMODATION DEFICITS IN A GROUP OF YOUNG OFFENDERS*

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*Paper presented as the Patricia Lance Lecture, Brisbane, 1989

The enormity of the honour bestowed on the person invited to deliver the Patricia Lance Lecture is second only to the challenge that it represents in coming up with a paper worthy of the name. I am indeed honoured, and hope that what I have to say is of some practical significance and stimulus for further clinical research.

Patricia Lance for all age-groups of British orthoptists, has been synonymous with orthoptic training in Australia. She has been very active in the international world of orthoptics and ophthalmology, and host to several senior British orthoptists. I was delighted therefore in my early days in Melbourne to have the unequivocal support of Miss Lance. She was in fact one of the few to give me great confidence and inspiration in leading the school in the direction it has subsequently taken. For that I am most grateful. Miss Lance is a clinician, and an inspiration within Australian orthoptics — this annual lecture being a very suitable reminder of her eminence in the field.

The subject I have chosen to speak about has arisen as so much clinical research does, out of looking at multitudinous variables in other projects. In 1988 I was invited to screen a group of young offenders in a local remand youth training centre — the special education teachers believing that there may be some vision deficits. The bench-mark for vision defects for most people of course is the wearing of glasses, and the report was that no boys coming into the centre ever wore glasses. I anticipated of course, that there should be a number with refractive errors. There were, but what became much more obvious, was the number of boys with accommodative deficits with or without convergence insufficiency.

Before looking at the study itself, there are some comments of a general nature I would wish to make. These relate to our testing methods and the populations about whom we have normal data. After briefly reporting on the study, this will be further illustrated. Many of the functions that we assess clinically rely heavily on instrumentation, subjective responses, and something nebulous called clinical judgement. For the most part by good luck, these serve us well for the majority of patients, but still, there are always those difficult ones, and, depending upon our tolerance levels we label them with a diagnosis and a cause — often 'functional'.

However, our clinical judgement is more or less effective based upon the first 2 factors, that is, the reliability of the instrument and the results that it gives us. That part we should have complete and repeatable control over. In addi-

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tion to that, we must know whether the responses are normal or not for an individual, and in the case of a study, such as the one that I shall relate to you, whether the occurrence of a particular result across a sample population is normal or not. On review of a fairly wide range of literature on accommodation as with so many of the visual functions that we test, there is very little in the way of data from normal populations. There is much on pre-school and school-age children, but little beyond that. That is, from non-symptom or sign-producing visual function. It is therefore quite difficult to label aspects of visual function as normal or abnormal if the study does not have an age, and in this case, sex-matched group with which to compare.

ANATOMY OF ACCOMMODATION

In order to understand what is being tested, it is necessary to briefly look at what is known of the pathways for accommodation, and its relationship with the other aspects of the near reflex. The accommodative reflex is activated in the retina by a blurred image, chromatic aberration, and an awareness of proximity. The afferent pathway runs with the visual pathway to area 17 in the cortex, then relays to areas 19 and 22 — the visual association areas. The efferent pathway is via the cortico-tectal tract to the Edinger Westphal nucleus, from where the parasympathetic fibres travel as part of the IIIrd cranial nerve to the ciliary ganglion. After this the short ciliary nerves leave the ganglion entering the eye to supply the ciliary muscle. Whether or not there is a sympathetic supply in opposition is still the subject of some debate. Davson[1] supports the view that the action of the ciliary muscle is opposed purely by the elastic properties of the lens capsule and contents. However, more recent studies, notably Stephens,[2] reporting in 1985, shows sympathetic involvement. In his study, alpha and beta adrenergic stimulation was used in conjunction with measurement of the AC/A response in human subjects. The results indicate a dual autonomic innervation for accommodation.

If you consider the musculature of the ciliary muscle, it can be seen that it is the annular (or sphincter fibres), and the radial fibres which contract causing elongation of the muscle and therefore an increased accommodative state. The presence of the meridional fibres suggests an opposing system. The reason of course for revising the nervous systems involved particularly in this group, is because of the effects of certain ingested agents which stimulate or denervate the autonomic nervous system. Substances such as caffeine and alcohol, through to prescribed and non-prescribed drugs have some effect on the autonomic nervous system, and therefore our measures of autonomic nervous system function — in this case, accommodation.

METHOD

The subjects were all inmates of a remand and youth training centre for periods varying between one month to 3 years, some being repeat visitors. Because of the nature of the institution and their rights within it, the vision screen was on a voluntary basis. However, only one refused over the 6 months of the study — it seems that the attention provided during a vision screening often represented a relief of boredom or was preferable to an anti-smoking film! The sample was gained by my attendance on a regular day each week in one of 3 sections in the centre and as many as possible on that section at the time were screened. Testing took place in room-lighting conditions, without external light, usually in the medical room attached to the section. A number of tests of visual function were carried out which are not reported here, but included convergence and accommodation. The total number of boys seen was 75 between the ages of 13 and 22, with a mean age of 17.37 years. The majority in fact were 17-19 year olds. Accommodation and Convergence was measured using the RAF gauge in the normal manner. In the case of accommodation, the measure was taken subjectively from the point at which the print first became blurred. The suggested normal near point of both convergence and accommodation for this age is 8 cms, representing 12 dioptres of accommodation. I have in these results erred on the side of conservatism, and taken 12 cms or less to be defective in the absence of other data.

In this study, all subjects with defective accommodation had distance visual acuity of 6/6 or better and demonstrable binocular single vision.

RESULTS

Accommodation

Of the 75, 27 (36%) showed accommodation to 12 cms or less, representing 8D or less of available accommodation. Of those, 14 (18.5%) could accommodate to 20 cms or less, 5D or less of accommodation.

Convergence

Of the 75, 13 (17.3%) showed convergence of 12 cms or less. Of those 13, 9 (12%) showed accommodation to be more reduced than convergence, only 1 (1.3%) showed convergence to be less than accommodation, and 3 (4%) showed equal convergence and accommodation.

Symptoms

Whilst remembering that none of these subjects had complained previously about visual/ocular problems, on questioning prior to testing, 19 (70%) of the 27 with reduced accommodation complained of some symptoms often associated with reading and close work. Ten (77%) of the 13 with convergence insufficiency also complained of symptoms.

Drugs — Prescribed/Non-Prescribed

It became evident through voluntary admission that many of the subjects were using drugs — mainly non-prescribed drugs such as cocaine, marijuana, heroin, amphetamines, and Avil, in addition to alcohol and smoking. Many admitted to taking the whole lot or "anything I can get". It was impossible therefore to judge any dosage or frequency of drug use, so it was merely recorded that they did or did not use drugs, or as unknown.

Of the 27 with reduced accommodation, 17 (63%) reported drug abuse. This compared to only 16 (33.3%) reporting drug abuse out of the 48 with normal accommodation. Of the 60 with convergence better than 12 cms, 27 (45%) reported abuse of drugs, compared with 6 (46.1%) of those with reduced convergence of 12 cms or less, i.e. there is no significant difference between the drug users and non-drug users in the levels of convergence.

DISCUSSION AND CONCLUSIONS

First of all it must be emphasised that according to all accounts of what constitutes a normal measurement of accommodation and convergence, the levels used here have been very conservative. If the clinically accepted normal measurement of the near point of accommodation and convergence had been taken to be 8 cms then 46 (61%) of the subjects would be considered to have defective accommodation, and 19 (25%) to have defective convergence. The conclusions that can be drawn from this information in the absence of other data are several:

1. This age-group of males may have a higher than normal incidence of accommodative weakness.
2. The reported synkinesis between accommodation and convergence may very easily be dissociated — the subjects demonstrating a weakness of accommodation were far more frequent than those with defective convergence.
3. The use of non-prescribed drugs may significantly affect accommodation. The specific drugs and their effects on either the parasympathetic or a possible sympathetic innervation cannot be commented on from this data.
4. The possibility that accommodative disuse even in the young adolescent leads to a reduction in its function.
5. The RAF Rule as a measure of accommodation in particular may underestimate the actual capabilities.

There are several comments of a general nature to be made having conducted this survey into accommodation deficits. Firstly, our traditional clinical methods of measurement of accommodation either as a range with the RAF Rule, or amplitude with lenses, fall far short of giving us adequate information about its function. Neither method yields information about the reaction time, maintenance of accommodation, or its real relationship to convergence and the near reflex. This data shows a clear dis-

sociation between the two particularly when accommodation is defective. In addition, it to some extent, refutes the idea that in all subjects accommodation is the major stimulus for convergence. In other words, a disruption of accommodation does not necessarily lead to loss of convergence.

A review of the literature reveals the difficulties with accommodation measurement and certainly provides a strong indication that our techniques of measurement should be evaluated and changed. Firstly, our measurement techniques. Current clinical measures involve total subjectivity and a measurement of near point only, or the total dioptric accommodative power using lenses. As mentioned earlier, no account is taken of response time or maintenance levels.

Secondly, we must consider the influence of ingested substances on the autonomic nervous system and therefore accommodation, the influence of temperature, and diurnal variations.

When the literature is examined, it is easy to see that attempts have been made to increase the objectivity of accommodation measurement. A review of the literature shows a lack of response by orthoptic clinicians to research work done as early as 1937. Specific instruments were designed for experimental purposes to look at fluctuations in accommodation in normal subjects.[3,4] Since then of course, other accommodometers have been devised but there would be few in clinical practice. To the extent that our systems are outdated, we as clinicians are ignoring at least one section of visual function and continuing with unreliable methods. This is the first point I would like to make, that in our efforts to expand our own research, we must not make the fundamental error of separating research from clinical practice. The symbiosis between the two should be a productive one for patients. However, we should be a little quicker to respond to the research than from 1937 to 1989.

The mechanism of accommodation and its relationship to the near reflex is clearly complex. It is complex because its control is dominated by the autonomic nervous system which is highly sensitive to metabolic changes induced by agents produced within the body and by ingested substances. Autonomic ocular changes have been reported with such factors as facial skin temperature, caffeine and alcohol.[5] Similarly eye movement function has been reported to alter with known illegal drugs.[6] The point here I wish to make is that when we assess something as delicately controlled as accommodation, then there are obviously factors which need to be taken into consideration, not only in experiments, but in clinical practice.

Here lies the clinical dilemma. We see patients or clients in the clinic — not subjects! These are people with symptoms and possibly signs of visual or ocular dysfunction — they are individuals and each one demonstrates multitudinous variables. As clinicians our role is quite clearly to produce an accurate diagnosis and subsequently a management strategy which will relieve the symptoms or cure the signs. As we all know too, for some people, it may be quite acceptable to have reduced accommodation and convergence (if we know what is reduced that is), but those that come within the normal limits of our tests, and still complain of symptoms who are the problem. That is why our testing measures must be improved — it should not be based so heavily on clinical judgement. I am convinced that that is one powerful reason why some patients will move from one clinician to another across all professions until someone can resolve their problem.

In conclusion, to return to the study related here, on the basis of the available tests of accommodation, there was a large number demonstrating an accommodative deficit by the standards available. There was also a significant level of drug abuse amongst those with reduced accommodation. But my original problems remain — the lack of data for normal populations, and the inadequacies of our testing methods for accommodation. This function plays such an important role in so much of our work with squint in children, through to whiplash and pregnancy in adults, it is time to consider an improvement of our methods of measurement and subsequently our management strategies. Clearly this sort of data leads us on to further research with more clearly defined and

isolated variables. It has answered one question — that this group of young offenders does demonstrate a high level of accommodation difficulties, and clearly these boys are not high achievers academically, either for reasons of motivation or because of their symptomatic or asymptomatic visual conditions or both. Conversely a disuse of accommodation leads into the hermeneutic cycle where reading and close work produce discomfort so that a commitment to reading is lost and further disuse is the outcome.

One final point that I would wish to make is the alacrity with which authors blame reduced accommodation on so-called functional factors.[7] That is, family background, psychological disturbance, or any other factor which provides an excuse for the difficulties which are encountered in the diagnosis and management of accommodative changes. I am suggesting that our ignorance should be tested first before passing off real symptoms and signs to functional failure. Various researchers have attempted to devise objective methods of accommodation measurement by adaptations of refractometers. We should be evaluating these methods against our current clinical methods, in order to establish the best method of measurement. In addition, the paucity of normal data is an issue that we must attempt to rectify in order to understand and make statements about individuals and populations.

Meanwhile, the decisions about the boys in this study who were found to be accommodatively defective by current standards, were taken out of my hands by the sorts of events that are common in the prison environment and by referral through the medical officer to outside clinicians. The interesting points are that there were a significant number with defective accommodation and when that happened it did not correlate with the convergence response. Secondly, there seems to be a close relationship between the use of non-prescribed drugs and a reduction in accommodative capacity. One could hardly argue that this group had some disturbance in their home or environment, but far be it from an orthoptist to claim that psychological factors were the primary cause.

In this study I have simply presented some very basic results and anecdotal comments on one of our traditionally accepted clinical measures. It has in turn raised a number of questions relating to our fundamental knowledge of clinical events and interpretation of results because as clinicians we rarely if ever take an over-view of a normal population. It is also the case that we rarely, if ever, evaluate our clinical testing procedures which have been designed to substantiate diagnoses. I venture to suggest that there are times when we use those clinical measures to substantiate a clinical punt, instead of basing our diagnosis on sound measures. This is not intended to be a cynical view, it does however, lead me to believe that orthoptics as a profession in Australia is more fortunate than we yet fully understand, to have its educational base in the university system which is there to be exploited for applied research purposes. New ideas can be explored in conjunction with those in clinical practice so that research and clinical work is inextricably intertwined. The universities have access to a range of funds for this purpose — we as a profession must extract our rightful share.

I would like to thank the Orthoptic Association Council once again for inviting me to present this paper under such a prestigious name at the Annual Scientific Conference of the Association, and to once again pay tribute to Miss Patricia Lance. May I also take this opportunity to thank the organisers and wish them luck for a successful conference.

References

1. Davson H. The physiology of the eye. 2nd ed. Boston: Little, Brown and Company, 1963.
2. Stephens KG. Effect of the sympathetic nervous system on accommodation. Am J Optom and Phys Opt, 1985, 402.
3. Collins G. The electronic optometer. Brit J Physiol Optics I. 1937, 30-42.
4. Campbell FW. A high speed infra-red recording optometer. J Physiol 1956, 31.
5. Lewis M McD and Pitt A. Factors affecting intrinsic eye muscles. Proc Aust Physiol Pharmacol Soc 18, 1987, 49.
6. Leigh RJ and Zee DS. The neurology of eye movements. 1983, 1-9, Davis, Philadelphia.
7. Karseras A, Unwin B and Wybar K. Brit Orthop J 31, 1974, 91.

Australian Orthoptic Journal 1990; 26: 6-12

INFANT VISUAL FIELD ASSESSMENT

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Abstract

A screen, called the "Baby Visual Field Screen" has been developed to assist with plotting and describing the visual fields of infants, toddlers and physically or mentally handicapped patients.

The screen is made of transparent perspex, is free standing, and is of similar dimensions to a one metre Bjerrum's screen.

During the examination the infant is seated on the parent's lap, one metre from the screen. Central fixation is elicited by one examiner behind the screen showing the subject a succession of toys, faces and/or talking to the infant as required. A second examiner introduces targets into the infant's peripheral visual field and moves them toward the centre, until the infant fixates the target or makes some sign of recognition. The targets were finger puppets of equal size and luminosity. The two examiners agreed on the point where the infant fixated the peripheral target and plotted this point on the screen. Horizontal, vertical and diagonal meridia were tested.

Monocular fields were plotted for twelve adults and twelve normal infants ranging in age from five to ten months and binocular fields for six infants of two and three months of age. Results indicate that the screen provides an accurate indication of a normal visual field. In a group of 13 infants and children at risk for visual field defects, the method was able to detect and record defects in five cases.

INTRODUCTION

Assessing and describing visual field defects in infants is a difficult task, but, as with adults, it can provide valuable information. Documentation of a visual field is important in describing the disability resulting from lesions of the visual pathway. This has implications in further therapeutic assessments, treatment and educational management.

Previously we have relied on confrontation techniques to estimate visual fields. In the multidisciplinary environment a more quantitative method has become necessary for more effective communication.

Complex technical equipment has been developed to study the visual fields of infants in a controlled environment. Mohn and Van Hof-van Duin[1] assessed the horizontal and vertical extent of the binocular visual field in 99 infants from birth to one year using a kinetic perimetry technique. The monocular visual field of 53 infants from six weeks to one year was also studied. The binocular visual field showed little development between birth and two months of age, but then expanded rapidly to the age of eight months and more slowly until twelve months. By this age the upper visual field had reached adult size while the horizontal and lower fields were still smaller

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than the adult. The development of the monocular visual field in the temporal and vertical direction closely resembled that of the binocular field but the nasal visual field was smaller than the temporal field at all ages.

Schwartz, Dobson, Sandstrom and Van Hof-van Duin[2] noted that neonates showed a larger visual field than 4 to 8 week old infants, however visual field shape was similar in infants and adults. Again a kinetic technique for perimetry was employed.

Mayer, Fulton and Cummings[3] have developed a static perimetry technique with LED stimuli and a forced choice observation procedure. Four central pulsating LED's were used to elicit central fixation and this was maintained with the aid of auditory stimuli. Adult observations of infants' eye movements to fix peripherally illuminated LED's were used to determine the extent of the field. The binocular visual fields of infants aged six to seven months were found to be similar to those of adults tested with the same apparatus. Monocular visual fields were smaller than those of adults. Mayer et al suggested this equipment had the potential to be a valuable clinical tool.

Maurer, Clarke and Lewis[4] used a similar static perimetry technique to demonstrate that the temporal visual field develops more rapidly than the nasal field.

The aim of the current study was to develop a clinical tool to improve the methods of reporting infant visual fields.

METHOD

Apparatus

The Baby Visual Field Screen was constructed by the Biomedical Engineering Department of The Children's Hospital, Camperdown.

The screen is made of a perspex sheet which is one metre square and three millimetres thick. It is mounted in a powder coated steel frame. Light alloy glazing bars locate and stabilize the screen within the frame. The complete assembly is mounted on casters, making the centre of the screen one metre from the ground.

The circles marked on the screen subtend angles of 5, 10, 15, 20, 25 degrees from a distance of one metre. An adjustable compass was used to scribe the circles into the screen about a fixed centre. It was specifically designed for this purpose and incorporated a cutting tool mounted on fixed rollers to engrave the perspex. Radial lines were scribed using the same cutter. Horizontal and vertical lines were first engraved, then the oblique at 15 degree intervals. "Scuff Stuff" boot polish was used to fill the engraved lines. Numbers were marked with "Letra-set".

Subjects

The Baby Visual Field Screen was trialed at Glebe Early Childhood Centre. Eighteen infants from two to eleven months of age with no known ocular defects were assessed following discussion about the trial with the Clinic Sisters and infant's parents.

A brief history of the infant's course to date and family details were obtained from the parents. Visual acuity was tested using Teller Acuity Cards and ocular posture was assessed. The results and data were within normal expectations for all infants.

The 12 infants aged five to eleven months all had monocular visual fields and visual acuity tests. The remaining 6 infants who were two or three months old were tested with both eyes open.

Monocular fields were assessed in 12 adults with corrected visual acuity of 6/6 or better and no history of ophthalmological or neurological problems.

The Baby Visual Field Screen was also used in 13 cases where the patient was "at risk" for having a field defect. There were a variety of neurological and ophthalmological problems involved with these patients.

Procedure

The screen was positioned in a quiet room with constant overhead fluorescent lighting. During the field examination infants were seated on the parent's lap one metre from the screen, with the infant's eyes aligned with the centre of the screen. (See Figure 1).

Figure 1: The infant, mother and examiner positioned behind the screen

The first examiner was positioned behind the screen, to attract the infant's attention and maintain central fixation, with her face or using toys. Auditory stimulation was sometimes employed to assist with a distractible infant. This examiner was the key observer in determining when the infant fixated the peripheral target and differentiated fixation movements from spontaneous eye movements. (See Figure 2)

The second examiner presented the finger puppet targets (green plastic monsters, approximately 3cm x 2cm x 3cm) placed on the end of

Figure 2: The examiner in front of the screen attracts the child's attention, and marks the point where peripheral fixation is made.

a tongue depressor. The targets were presented from behind the patient, into their peripheral field and moved toward the centre until the infant looked towards it. Horizontal, vertical and diagonal meridia were tested, using a kinetic technique similar to confrontation field examination.

When the infant looked towards the target, the first examiner moved to be 'in line' with it, and this peripheral point was marked on the screen using "Blue Tac".

During the testing, if inconsistency or "inaccuracy" was noted by either examiner, then immediate re-examination of that area was performed.

The entire procedure took approximately 20 minutes to complete monocular assessment, both right and left eyes; and 10 minutes when the tests were performed with both eyes open.

The adults in the trial verbally reported peripheral stimulus detection. All adults were tested monocularly.

RESULTS

Normal Infants

The fields resulting from testing with this technique demonstrated an increasing field size with age. These results are similar to the visual field development studies of other groups, who utilized kinetic techniques.[1]

For analysis the trial population has been divided into age groups. The binocular fields of

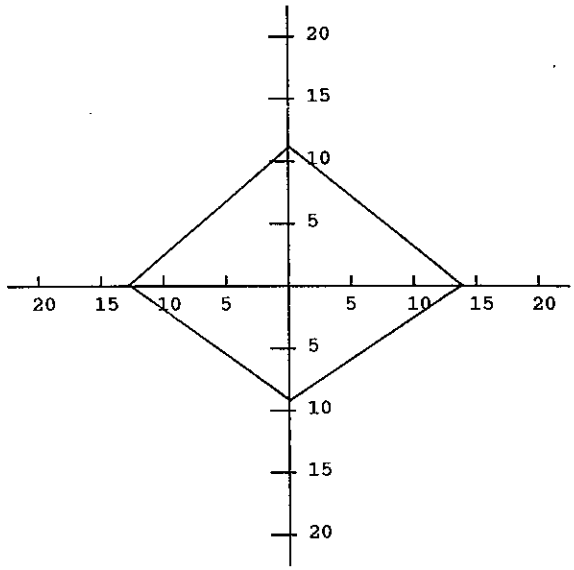

Figure 3: Binocular Visual Fields of Normal Infants less than 3 months old. Mean values at 0 deg, 90 deg, 180 deg, 270 deg.

infants aged less than three months were plotted using the mean values of horizontal and vertical meridia (Figure 3). The monocular fields were grouped into three to six months, six to nine months and nine to twelve months. The right and left eyes were considered separately and plotted, again using the mean values of the horizontal and vertical meridia. (Figure 4)

The mean values for adult eyes are shown in Figure 5.

"At Risk" Patients

The 13 "at risk" patients were tested using the same procedure. In many cases, more vigorous methods were employed by the examiner to ensure central fixation. This was necessary when the patient's state of health was poor or when a defect became apparent, making the test more time consuming.

Visual field defects were demonstrated in 5 of the 12 cases who were considered to be "at risk". A left hemianopia was found in a 4 1/2 year old child with cerebral palsy, a small right cerebral hemisphere and postinfarct and haemorrhagic changes. An 8 month old infant who had suffered a severe head injury causing an occipital intracranial haemorrhage had a right hemianopia. Sturge Weber Syndrome had caused extensive calcification of the left cerebral hemisphere in a 1 year old infant, resulting in a right hemianopia. A post-encephalitis victim was found to have a left hemianopia at 3 1/2 years of age. A 4 year old child who suffered a left cerebral vascular accident during cardiac surgery had a right superior temporal quadrant deficit in the right eye, and a nasal field deficit in the left eye.

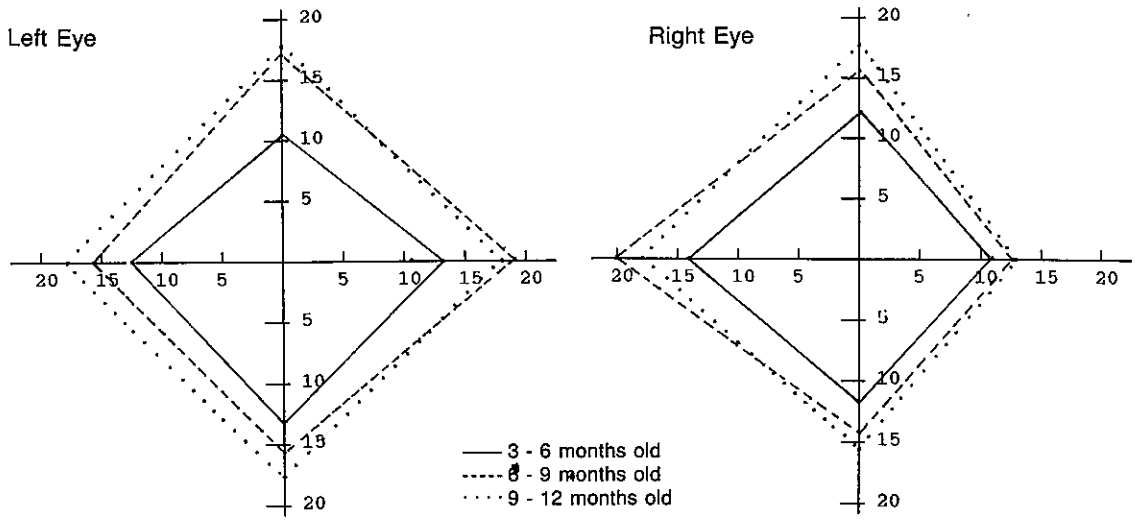

Figure 4: Monocular Visual Fields of Normal Infants 3-12 months old. Mean values at 0 deg, 90 deg, 180 deg, 270 deg.

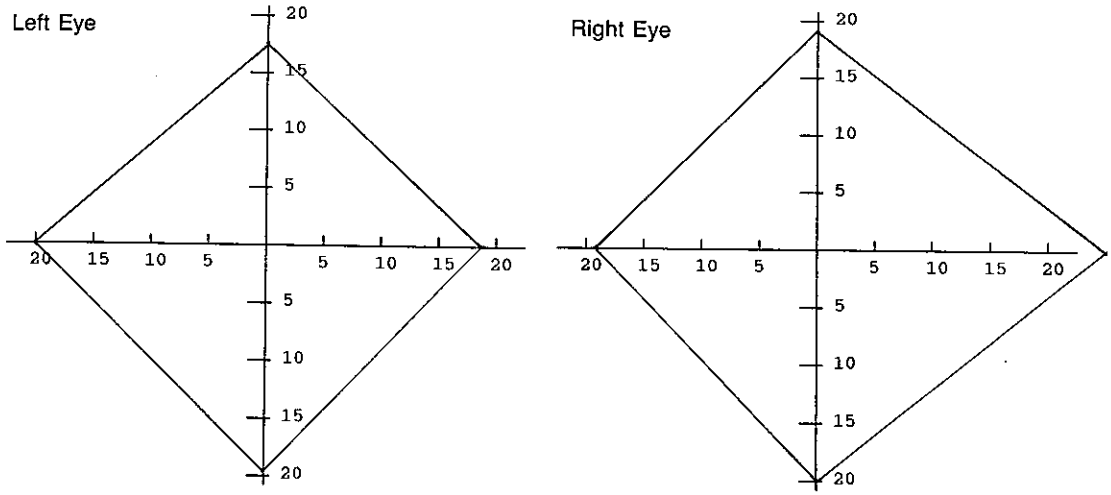

Figure 5: Monocular Visual Fields Normal Adults. Mean values at 0 deg, 90 deg, 180 deg, 270 deg.

The remaining 7 patients tested using the Baby Field Screen were not found to have a visual field defect. This group included a 3 year old with cerebral palsy; two children who had suffered severe head injury, (one aged 7 years, the other aged 11 years); one 3 year old with bilateral colobomas of the disc, retina and choroid; a developmentally delayed 2 year old child; a case of benign intracranial hypertension affecting a 3 year old; and a 7 year old with bilateral optic nerve gliomas.

DISCUSSION

The Baby Field Screen proved to be a convenient clinical tool to assess and document the visual fields of infants and children. Numerous advantages of the system became evident.

The Baby Field Screen is clearly not as refined as the techniques previously discussed.[1,2,3,4] This procedure for infant visual field assessment is designed for easy clinical application to detect and plot "gross" visual field defects in patients unable to co-operate with more sophisticated methods. The major improvements over traditional confrontation testing are the more accurate plotting of the fields enabling demonstration of the field defect to the parent/caregiver, giving them a better understanding of the limitations of the condition.

Two examiners were required to perform the test. This was found to be beneficial. Consultation between the two examiners ensured that the screen was marked accurately at the point of the infants fixation. This allows the opportunity to differentiate spontaneous eye movements and true fixation. The techniques involved in assessing and observing are readily learned by skilled orthoptists.

Co-operation was readily gained from the normal subjects and the 'at risk' patients. This could be attributed to the testing procedure being approached as a game. The importance of this approach was evident when testing acutely or chronically ill children who had previously undergone many invasive procedures. Their confidence was gained when they realised that this procedure was non-threatening. When necessary children in both groups were pacified with the help of a dummy, by holding a toy or by comfort from a parent. Maintaining central fixation was more easily achieved in the normal group.

It is envisaged that the Baby Field Screen test has further applications with patients who are not mentally or physically capable of performing alternative visual field tests. This information may aid further therapeutic assessments, treatment, rehabilitation and educational

management in handicapped children, disabled adults and stroke patients.

The procedure is effective due to its simplicity and speed. The screen itself is portable, durable, requires only a small area for use and does not need electricity for its operation.

Overall, the baby visual fields appeared similar in size to the adult fields by the 9-12 month age group, a finding which is similar to Mohn et al[1] and Schwartz et al.[2] Monocular testing of infants tended to show slightly larger nasal fields which is in contrast to Mohn et al[1] and Maurer.[4] Our results may well be due to the small sample size, and by examining the results too closely in a test where the variables are great and difficult to control. Such variables include the overall alertness of each individual child throughout the test procedure, the exact position of the central fixation target and frequent change of target (which was usually moved a little and swapped with other targets to promote sustained interest). Also, the hand held peripheral target may have varied in its speed of approach and exact distance from the screen on approach to the centre of the visual field. This is due to slight variations in distances from the screen to the peripheral target when examiners try to manoeuvre around different subjects without the examiner becoming too obvious to the subject. Examiner bias is not believed to have contributed to the results, as examiners did not have a fixed role in the testing procedure.

Clearly, the documentation of a visual field defect in the "at risk" patient group provides valuable information of these patients' conditions. The technique allows accurate plotting of defects such as hemianopia and quadrantinopia. Defects, such as those due to enlargement of the optic disc or coloboma, which may be plotted in co-operative patients using sophisticated methods, can not be recorded with the Baby Visual Field Screen.

CONCLUSION

The Baby Visual Field Screen provides a significant improvement in describing the visual field defects of infants and small children. It is useful in the clinical setting, allowing a rapid, effective and non-threatening assessment. The resultant information is meaningful for the examiners, medical team, therapists, educationalists and parents/caregivers.

References

1. Mohn G, Van Hof-van Duin J. Development of the binocular and monocular visual fields of human infants during the first year of life. Clin Vision Sci, 1986; 1: 51-64.
2. Schwartz TL, Dobson V, Sandstrom J, Van Hof-van Duin J. Kinetic perimetry assessment of binocular visual field shape and size in young infants. Vis Res 1987; 27: 2163-2175.
3. Mayer DL, Fulton AB, Cummings MF. Visual fields of infants assessed with a new perimetric technique. Invest Ophthalmol Vis Sci 1988; 29: 452-459.
4. Maurer D, Clarke AL, Lewis TL. The development of peripheral detection during infancy. ARVO Abstracts. Invest Ophthalmol Vis Sci 1986; 27 (Suppl): 264.

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EVALUATION REPORT — THE CAMBRIDGE VIDEO REFRACTOR

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Abstract

Photoscreening is a technique that has been developed in an attempt to refine vision screening programmes, which aim to identify visual disorders in early childhood. Various groups have developed photorefracting tools in recent years. This study evaluates the role of one such tool–the Cambridge Video Refractor (CVR).

The CVR is an isotropic on-axis photorefractor, which utilises a computerised system to evaluate three photographs of a subject's eyes, and predict refractive error.

When the results of video refraction on 101 non cyclopleged eyes were compared with their retinoscopic refraction, 41.6% of the CVR predictions were correct. The video refraction of 83 cyclopleged eyes were compared to the retinoscopic refraction, and the accuracy of the CVR was 51.8%. For this evaluation, the video refraction prediction was within 1.0D and the axis within 20° of the retinoscopic refraction to be considered accurate.

INTRODUCTION

Photoscreening is a method of estimating the refractive state of the eyes by photographing the light returning from a subject's fundi, when the eyes have been illuminated by a light source centred in a camera lens.

It is a technique that has been developed in an attempt to refine screening programmes, whose aim is to identify visual disorders in early childhood. It is well accepted that treatment of refractive and strabismic amblyopia is most effective in the early years of life.

Numerous groups have produced photorefractive tools,[1,2,3] and these appear to have been helpful in dealing with the difficulties of traditional screening methods. An on-axis photorefracting technique was introduced by Howland and Howland[4] in 1974. This has since been modified by the Vision Development Unit of The University of Cambridge, to produce the Cambridge Video Refractor.[5,6]

The Cambridge Video Refractor (CVR) is an isotropic photorefractor in which the light source is mounted on a video camera. Three flash photographs are taken at different focal lengths. The fundal light reflections are compared and analysed on the monitor screen. The refractive status of the eye will determine the size and shape of blurred images. The principal meridians of the images are measured to predict refractive error using photographic data empirically calibrated in a computerised system.

In the study of Atkinson, Braddick, Ayling, Pimm-Smith, Howland and Ingram[7]

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photorefraction was used to screen 1096 infants aged 6 to 9 months. Those cases that were considered to have a significant refractive problem were followed up. Of those reviewed, retinoscopic refraction confirmed the photorefractive findings, with the only significant discrepancy being the anisometropes.

The Cambridge Video Refractor (CVR) was introduced to Australia in April, 1988. The Orthoptic and Ophthalmology Departments of The Children's Hospital, Camperdown were interested in this equipment, as the departments are involved in numerous visual screening programmes, as well as the promotion of preventative eye health and education of other groups performing vision screening.

Members of the departments had no previous experience with the CVR, so there was considerable interest in its effectivity and its ability to significantly improve current screening techniques.

The current study was therefore undertaken to evaluate the CVR's value as a screening tool. Prior knowledge of the demands of large scale vision screening programmes enabled the researchers to establish criteria for an effective screening tool, and it is against the following criteria that the CVR is evaluated:

- It should provide a consistent indication of significant refractive error and the need for formal follow-up.
- It should be effective without cycloplegia.
- It should be suitable for use by non-technical staff.
- The cost of the equipment should be acceptable.
- The equipment should be portable.
- The procedure should be rapid.

During this investigation, only the refraction component of the CVR function was evaluated, although it is acknowledged that it may also be used to detect strabismus.

METHOD

The subjects were patients of The Children's Hospital Eye Clinic undergoing either initial assessment or routine review of visual function and refractive error.

The visual acuity and orthoptic status of all subjects was initially assessed. Patients and parents were informed of the CVR's function and the evaluation project discussed.

A total of 62 subjects were involved in the study but in some cases it was only possible to test one eye. Ages ranged from 4 months to 16 years (mean age 4 years 11 months). (See figure 1). This sample was considered to represent a cross-section of patients seen in The Children's Hospital Eye Clinic who were available for testing during the trial period, including patients with a wide range of paediatric ocular and general conditions.

The results of video refraction on 101 non-cyclopleged eyes were compared with cycloplegic refraction, and an additional comparison of the video refraction of 83 cyclopleged eyes to their cycloplegic refraction was made.

The first series of photos were taken, in accordance with the CVR manual, with the subject seated 75cm from the camera and the lens aperture set at 0.75m to produce the "pupil photo", then repeated with lens settings of 1.5m and 0.5m to create the "blur photos". The subject's attention was directed towards a toy or the examiner's face, positioned just above the camera. The room was dimly lit.

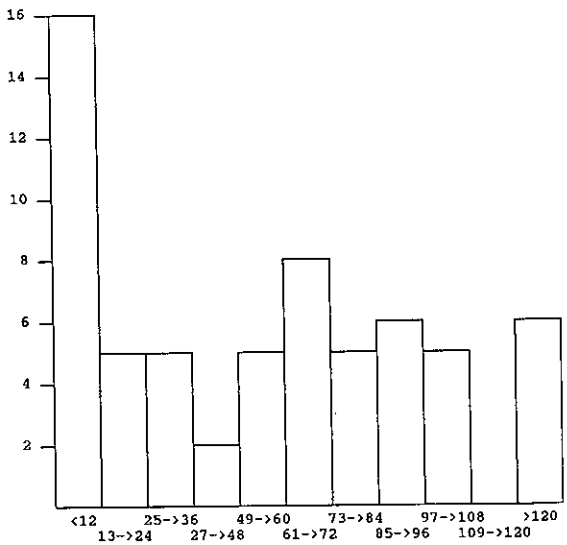

Figure 1: Frequency distribution of subjects by age.

TABLE 1
Video Refraction Predictions
Accuracy to within 1.0D and 20°

Non cycloplegic video refraction vs Retinoscopic refraction N = 101

		Hyper-metropic	Hyper. Astig	Myopic	Myopic Astig	Mixed Astig
Total No.		54	17	14	5	11
Correct	42	30	1	7	3	1
	41.6%	55.5%	5.9%	50%	60%	9.1%
Incorrect	59	24	16	7	2	10
	58.4%	44.4%	94.1%	50%	40%	90.9%

Cycloplegic video refraction vs Retinoscopic refraction N = 83

		Hyper-metropic	Hyper. Astig	Myopic	Myopic Astig	Mixed Astig
Total No.		49	20	1	1	12
Correct	43	34	5	—	—	4
	51.8%	69.4%	25%	—	—	33.3%
Incorrect	40	15	15	1	1	8
	48.2%	30.6%	75%	100%	100%	66.7%

The video and computer equipment was utilised to make the appropriate measurements of the focused and defocused photos. The relevant details of the resulting "video refraction" were recorded.

Where a myopic error was indicated, the procedure was repeated with the subject positioned 1m from the camera and the lens aperture set at 1m, 3m and 0.6m.

Cycloplegia was obtained using two to three drops of Cyclopentolate 1%OU. When possible the video refraction was repeated following cycloplegia. Unfortunately, due to the constraints of a busy Eye Clinic, not all subjects received a non-cyclopleged and cyclopleged video refraction.

Retinoscopic refraction was performed by an ophthalmologist. Retinoscopy was used as the reference, as it is a traditionally accepted method of evaluating refractive errors in children.

Results of non-cyclopleged and cyclopleged video refraction and cycloplegic retinoscopy were charted, with note made of the ophthalmologist's working distance.

RESULTS

Cases of hypermetropia, hypermetropic astigmatism, myopia, myopic astigmatism, mixed astigmatism and anisometropia were considered. Astigmatism was considered to exist when the refraction varied by more than 1.0D between the axes. Anisometropia was defined as a difference in refraction between the eyes of more than 1.0D.

Evaluation was made with acceptable limits of accuracy set at 1.0D and 20° (ie video refraction prediction must be within 1.0D and the axis be within 20° of the cycloplegic refraction to be considered accurate). Using these limits the predictions of the CVR were correct in 41.6% of cases of non-cycloplegged video refraction, and 51.8% of cycloplegged video refractions. (See Table 1).

Correlation co-efficients have been calculated to further compare the results of non cycloplegic and cycloplegic video refraction, to cycloplegic refraction. Table 2 presents these results. For the purpose of detailed analysis, the two meridians and each axis are considered separately.

The extent of variability shown by these figures is rather concerning and is perhaps an indication of the CVR's lack of consistent reliability. Whilst the correlation coefficients for each meridian are around 0.5 or higher, the best, 0.78 is still not sufficient to justify the clinical reliability of the test. We consider a correlation of at least 0.8 for each meridian a minimum requirement. In particular, the low correlations between the axes of astigmatism should be noted.

TABLE 2
Correlation Coefficients for Non cyclopleged & Cyclopleged
Video refraction vs Retinoscopic refraction

	Retinoscopic refraction			
	Meridian 1	Axis 1	Meridian 2	Axis 2
Video refraction non cyclopleged				
Meridian 1	0.69			
Axis 1		0.44		
Meridian 2			0.49	
Axis 2				0.27
Video refraction cyclopleged				
Meridian 1	0.78			
Axis 1		0.17		
Meridian 2			0.58	
Axis 2				−0.06

The CVR predicted anisometropia correctly in 72.7% of the non-cyclopleged video refraction group and 66.7% of the cyclopleged video refraction group. (See Table 3).

DISCUSSION

The comparison of video refraction and retinoscopic refraction produced rather disappointing results. Correlation values for the non cyclopleged video refraction group are below R = 0.69, and in the cyclopleged group R = 0.78 or less. At best, the CVR was found to have a reliability level of 51.8%, working with 1.0D error (ie 2.0D range) and 20° range, on cyclopleged patients.

Our findings do not reflect the result reported by the Visual Development Unit, where results of retinoscopic refraction confirmed the video refraction findings, in their group of 6 - 9 month old, cyclopleged infants.[7] They found a correlation of R = 0.77 or higher when photorefraction was compared to retinoscopic refraction.

There are a number of distinct differences in the Cambridge and Camperdown groups. The smaller number of subjects in the current group may detract from its significance, however, the trends observed in this group left the researchers reluctant to continue. The age range may also have influenced the results as this study made no attempt to restrict the age of subjects to the 6-9 month range used in Cambridge, but we did not observe an improvement in accuracy in the younger subjects. In fact, the reliability fell from 78.3% to 47.3% when cyclopleged infants under nine months only were considered.

Problems encountered during the trial of the CVR were numerous. Controlling the accommodation of the non-cyclopleged subjects during video refraction is an area of concern which is common to any form of non-cycloplegic photorefraction. As suggested by the CVR Manual, the subject's fixation was directed toward a toy or the examiner's face, positioned directly above the camera. However, we observed numerous cases of "over-accommodation" in the younger subjects, resulting in a false indication of myopia in the non-cycloplegic video refraction. It is realised

TABLE 3
Video Refraction Predictions
Anisometropia > 1.0D

	Non cycloplegic video refraction vs retinoscopic refraction.	Cycloplegic video refraction vs retinoscopic refraction
Total No.	11	12
Correct No.	8	8
	72.7%	66.7%
Incorrect No.	3	4
	27.3%	33.3%

that cycloplegia is recommended for CVR, but in view of our evaluation criteria, we felt compelled to assess a non-cycloplegic group.

The subjective nature involved in performing video refraction measurements prevented not only standardisation between examiners, but also between consecutive assessments by a single examiner. The "blur circle" seen in the defocused photos varies in clarity and can have quite a significant indistinct zone. The examiner must make the decision on where to place the measurement cursor on this zone. Error in placement obviously results in inaccuracies in video refraction. The difficulty also occurs in placement of the cursor to determine the axis of astigmatism.

Interesting is the finding that anisometropia was correctly predicted more frequently in the non-cycloplegic group (72.7%) than following cycloplegia (66.7%). This, we believe is also due to the problems of consistently dealing with the blur of the image on the screen.

The evaluation demonstrates that using a cycloplegic agent improves the CVR's ability to predict refractive error from 41.6% to 51.8%. Although the accuracy with cycloplegia is less than adequate, it is apparent that many refractive errors would go undetected if a video refraction screening programme were attempted without cycloplegia.

The accuracy and consistency of the video refractor as demonstrated by these findings, must be considered in terms of the CVR's intended role as a screening tool. It is not designed to replace retinoscopic refraction, but rather as a screening tool to indicate when a significant refractive error is present and formal follow-up is required. Hence it should not be expected to exactly determine refractive error. Taking this into consideration, and even when the evaluation extended the range in which video refraction would be considered correct the results suggest an element of unreliability. When the prediction was allowed to be within 2.0D of retinoscopic refraction, the accuracy only improved to select 67.3% of the non cyclopleged group and 78.3% of the cyclopleged video refraction group. With this generous range, the CVR predictions were incorrect in 22.7% of the subjects tested, which would produce a high level of errors in a screening programme.

The Cambridge Vision Development Unit advocate the suitability of the CVR for use by non-technical staff, after a short training period, though they actually employ an orthoptist for this task. The video refraction examiners in this study were all experienced orthoptists, who have a comprehensive understanding of the optics of refraction and video refraction. The confidence of the examiners did improve during the trial period, but the accuracy of results continued to be disappointing and were not felt to be influenced by a "learning curve".

The cost of the CVR is certainly prohibitive to the organisations involved in vision screening known to The Children's Hospital. Whilst it is well accepted that results of treatment of stimulus deprivation amblyopia will be more effective if the condition is detected and treated early, we were not able to demonstrate that the CVR significantly improved the results of screening to a level which could be equated to its cost.

Many vision screening services have adopted the philosophy of taking the service to the people, in order to reach the largest and most needy population. The screening venue may move frequently. Equipment must therefore be portable in this setting. Immediate access to the photorefractor images is a major advantage of the CVR's video camera and monitor system, which is not possible with many other photoscreening techniques. There is however, no facility to store the images. Other groups are reported to be developing a Polaroid system, that will combine the features of "instant results" and permanent images. The CVR can be moved successfully, but it is fragile, heavy, awkward and difficult.

CONCLUSIONS

The CVR has been evaluated in terms of its intended role as a tool to assist in screening for refractive error using an on-axis technique.

This study found that results obtained by cycloplegic video refraction were closer to retinoscopic refraction than were the results of

non cycloplegic video refraction. However, with a range of 2.0D permitted for video refraction to be considered correct, the level of accuracy of the video refractor was disappointing.

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References

1. Cibis-Tounge A, Cibis GW. Bruchner test. Ophthalmology, 1981; 88: 1041-1044.
2. Kaakinen KA. A simple method for screening of children with strabismus, anisometropia or ametropia by simultaneous photography of the corneal and fundal reflexes. Acta Ophthalmol, 1979; 57: 161-171.
3. Molteno ACB, Hoare-Nairne J, Parr JC, Simpson Anne, Hodgkinson IJ, O'Brien NE, Watts SD. The Otago Photoscreener, a method for the mass screening of infants to detect squint and refractive error. Jnr of the Ophthalmol Soc of New Zealand. 35, 1983; 43-49.
4. Howland HC, Howland B. Photorefraction: a technique for study of refractive state at a distance. J Opt Soc America, 1974; 66: 240-249.
5. Atkinson J, Braddick OJ, Ayling L, Pimm-Smith E, Howland HC, Ingram RM. Isotropic photorefraction: a new method for refractive testing of infants. Doc Ophthal Proc Series, 1981; 30: 217-223.
6. Howland HC, Braddick O, Atkinson J, Howland B. The Optics of photorefraction: orthogonal and isotropic methods. Journal of the Optical Society of America, 1983; 73: 1701-1707.
7. Atkinson J, Braddick OJ, Durden K, Watson PG, Atkinson S. Screening for refractive error in 6-9 month old infants by photorefraction; British J of Ophthalmology, 1984; 68: 105-112.
8. Atkinson J, Braddick OJ. The use of isotropic photorefraction for vision screening in infants. Acta Ophthalmol (Kbn), 1983: 157 (Supp): 36-45.
9. Hubel DH, Weisel TN. The period of susceptibility to the physiological effects of unilateral eye closer in kittens. Journal of Neurophysiology, 1963: 1003-1017.

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LIMITATIONS OF SUPERIOR RECTUS MOVEMENT: A LITERATURE REVIEW AND CLINICAL STUDY

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Abstract

Controversy exists in the literature regarding the differential diagnosis between a primary superior oblique palsy of one eye, and a primary superior rectus palsy of the contralateral eye. As a follow up to our previous study of long standing superior oblique palsies, which showed greater limitation of the contralateral superior rectus muscle, a literature review and a study of previously diagnosed primary superior rectus palsies was undertaken. It was revealed that isolated neurogenic superior rectus palsies are rare and are unlikely to occur without associated ptosis.

Key words: *Fourth cranial nerve palsy, superior rectus palsy, tight inferior rectus, thyroid eye disease, double elevator palsy, Bielschowsky head tilt test.*

Congenital isolated neurogenic superior rectus palsies are uncommon and are usually associated with ptosis on the ipsilateral side.[1]

To understand why this is the case, we wish to emphasise the following anatomical and physiological points.

The levator muscle and the superior rectus arise from the same embryonic striated muscle mass,[2] and both muscles are innervated by the superior division of the third cranial nerve, therefore, any neurological defect or lesion would affect both the levator and the superior rectus muscles.

There are also intimately bound fascial connections between the superior rectus muscle and the levator, (the fascial sheaths of Whitnall). At their origins on the sphenoid bone their tendons are blended together and the levator lies upon the superior rectus during its entire course.[3] In fact, surgery such as recession of the superior rectus muscle will carry the levator back with the superior rectus muscle and so the lid will be raised.[4] Also it has been reported (personal communication) that on giving an injection of Botulinum into the superior rectus muscle, ptosis of that eye often occurs.

With these facts in mind, the presence of muscle sequelae showing the greatest underaction being that of the superior rectus muscle without ptosis should encourage detailed investigation of the contralateral superior oblique muscle to avoid misdiagnosis.

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LITERATURE REVIEW

A literature review of the past 22 years produced some interesting aspects of superior rectus dysfunction. Anatomical absence of the superior rectus muscle has been reported, but this condition is rare and is usually associated with other craniofacial or extra ocular muscle anomalies. Mather and Saunders report that to their knowledge "there has been no surgically or radiographically documented cases of absent superior rectus muscle presenting as an isolated finding".[5]

Many references to double elevator palsies are made, involving symmetrical underactions of both superior rectus and inferior oblique muscles of the same eye.

Jampel and Fells[6] report finding sudden onset symmetrical paresis of elevation of one eye with normal lid function and suggest the cause to be a lesion in the midbrain tectum or pretectum, near or in the ocular motor nucleus. In these cases, Bell's phenomenon was reduced. This work seems to be the only significant publication in the last two decades, involving superior rectus dysfunction without ptosis and suggests a neurological cause.

Metz[7] described a congenitally short and tight inferior rectus muscle giving clinical signs of a double elevator palsy, when in fact, according to saccadic velocity measurements, the elevators were normal. This anomaly elicits poor or absent Bell's phenomenon. This suggests that Bell's phenomenon should be tested routinely in conjunction with a forced duction test, to differentiate between a brain stem lesion and a tight inferior rectus. Both conditions can exhibit absent or reduced Bell's phenomenon, but only the tight inferior rectus muscle will give a positive forced duction test.

Duke Elder states that the congenital adhesion syndrome of the superior rectus and superior oblique muscles at the point of crossing of the muscles can simulate clinical signs of a superior rectus palsy but without ptosis.[8]

Goodier[9] has diagnosed superior rectus palsies which elicited some type of "atypical" positive Bielschowsky head tilt test response but failed to define her "atypical" terminology. We consider it clinically misleading and incorrect to refer to an atypical head tilt response. This test was described as a specific differential diagnostic test to which the response can only be positive or negative to a superior oblique palsy (or an isolated inferior oblique palsy) as described by Bielschowsky.[10]

EXPLANATION OF THE BIELSCHOWSKY HEAD TILT TEST (BHTT)

Misconceptions regarding the complex principles of the BHTT appear to be common. Torsional movements of the eyes on tilting the head are the basis of this differential diagnostic test. Therefore, it must be remembered that physiologically the cycloduction of the oblique muscles is greater than that of the vertical recti muscles, and conversely the vertical action of the vertical recti muscles exceeds that of the oblique muscles.

The BHTT involves forcibly tilting the head to the side *opposite* to the usual compensatory ocular torticollis adopted for the paresis of the vertically acting muscle.

In the case of a superior oblique palsy, on tilting the head to the opposite side to that usually adopted as a compensatory head posture (ie. to the same side as the palsied superior oblique muscle), a vestibulo-ocular reflex of intorsion of the palsied eye occurs. In a synergistic movement the superior oblique and the superior rectus muscles of that eye should bring about the required compensatory intorsion.

However, because the superior oblique is palsied, the superior rectus, now being unopposed in the vertical field, produces an updrift or increased vertical deviation of that eye, ie. a positive BHTT response for a superior oblique palsy. Some authors explain this phenomenon by suggesting that an exaggerated contraction of the superior rectus muscle occurs.

In the case of a suspected superior rectus palsy, on tilting the head to the opposite side to that of the usual compensatory ocular torticollis, there is negligible change of vertical movement as the extorsion of the affected eye, induced by the tilt, is carried out by the inferior oblique and the inferior rectus muscles of that eye, and their antagonistic function in the vertical field is balanced.

The head tilt test may also be applied to the differential diagnosis between palsies of the inferior oblique and the contralateral inferior rectus muscles.

Walsh and Hoyt[11] state there is no increase of the vertical deviation in cases of superior rectus or inferior rectus paralysis on forcible tilting of the head to either side, and have found the BHTT to have great value in the differential diagnosis between a superior oblique and contralateral superior rectus muscle palsies.

Other authorities give similar explanations of the BHTT, and also claim the test to be a valuable differential diagnostic test.[12-16]

CLINICAL STUDY

In 1988 our department reported a series of fourth nerve palsies where the greater underaction was exhibited by the contralateral superior rectus muscle.[17] This finding has led us to undertake a 10 year retrospective study of previously diagnosed superior rectus palsies.

Documentation of only six cases of primary superior rectus palsies was found over this 10 year period.

All patients were contacted but only two returned for follow up. The required clinical information was incomplete in some cases, so the following details when available, were:

Hess charts
Ocular motility
Ptosis
Bielschowsky Head Tilt Test (BHTT)
Bell's phenomenon
Forced ductions

FINDINGS

In our study the following aetiologies were revealed as the contributing factors for the apparent superior rectus dysfunction.

Case 1: Congenital tight inferior rectus syndrome, confirmed by forced duction test on follow up.

Case 2 and 3: Trauma to the same side of skull as the underacting superior rectus muscle resulting in sudden onset diplopia in only that area of gaze, ie. not the usual neurogenic muscle sequelae and no ptosis.

Case 4: True neurogenic superior rectus palsy with ptosis on the same side.

Case 5: Thyroid eye disease

Case 6: Apparent superior rectus palsy, however, on review, a positive BHTT confirmed a contralateral superior oblique palsy.

It will be noted that only two cases were of neurogenic origin: Case 4 which was a true neurogenic superior rectus palsy with ptosis on the same side, and Case 6 which was a superior oblique palsy on the contralateral side confirmed by the BHTT. See Figure 1.

CONCLUSION

We find that evidence from the literature, our clinical data and personal communications support the belief that an isolated primary palsy of the superior rectus muscle without ptosis as a neurogenic entity would be a very rare occurrence.

The following aetiologies are considered to be responsible for an isolated superior rectus dysfunction without ptosis.

- congenital anomalies of the insertion of the muscle, muscle fibrosis or muscle aplasia.
- Thyroid Eye Disease/Thyroid Orbital Disease.
- congenital tight inferior rectus syndrome.
- double elevator palsy.
- direct trauma to the superior rectus muscle.
- orbital pathology.
- sudden onset double elevator palsy.
- post lens extraction which may result in superior muscle palsy with or without ptosis.
- primary palsy of the superior oblique muscle on the contralateral side.

Excluding trauma, pathology and congenital anomalies, we consider the most common cause of isolated limitation of movement of the superior rectus muscle to be a primary superior oblique palsy on the contralateral side. Careful examination must be carried out to establish the correct diagnosis.

References

1. Duke Elder. System of Ophthalmology Vol VI. Ocular motility and strabismus. London: Henry Kimpton, 1973. 709-710.
2. Mein and Harcourt. Diagnosis and management of ocular motility disorders. London: Blackwell Scientific Publications, 1986: 10.

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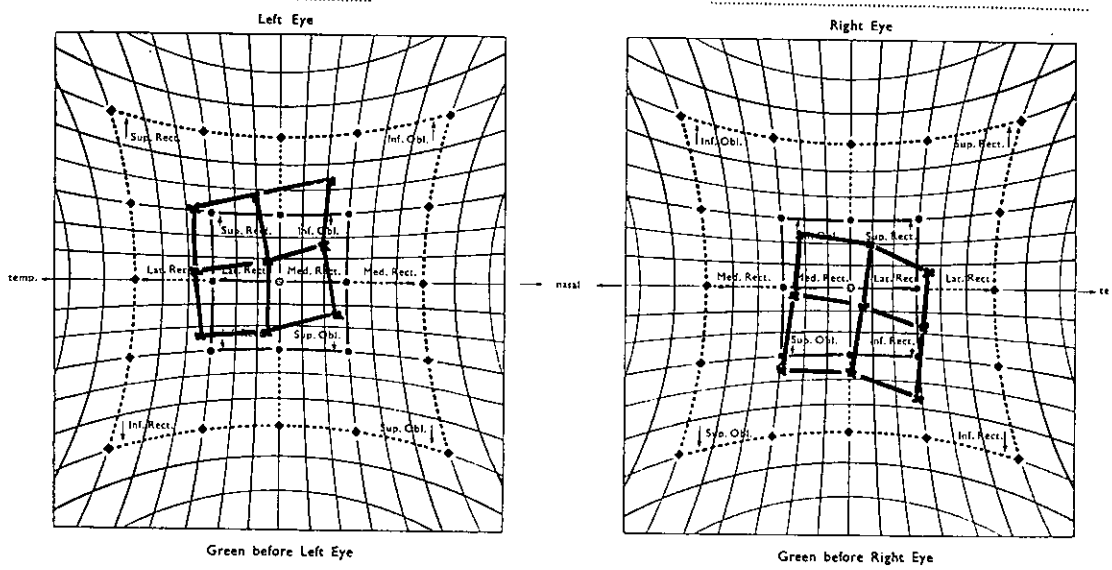

Figure 1: Case No. 6, showing greatest deviation in the area of the right superior rectus muscle but with a positive Bielschowsk Head Tilt Test response to the left side, indicating a primary palsy of the left superior oblique muscle.

3. Whitnall SE. Anatomy of the human orbit and accessory organs of vision. London: Oxford Medical Publications, 1921: 136.
4. Adler. Physiology of the eye, clinical application. 6th Ed. St Louis: CV Mosby Co, 1975; 5: 96.
5. Mather TR, Saunders RA. Congenital absence of the superior rectus muscle, case report. J Pediatr Ophthalmol Strabismus 1987. Nov, Dec; 24 (6): 291-295.
6. Jampel R, Fells P. Monocular elevation paresis caused by a central nervous system lesion. Arch Ophthalmol 1968 July; 80: 45-57.
7. Metz HS. Double elevator palsy. Arch Ophthalmol 1979 May; 79: 901-903.
8. Duke Elder. System of Ophthalmology Vol VI. Ocular motility and strabismus. London: Henry Kimpton, 1973: 737.
9. Goodier HM. The evaluation of the Bielschowsky's head tilting test. Orthoptics, Research and Practice. Trans 4th International Orth Congress. Mein and Moore. London: Henry Kimpton, 1981: 189.
10. Bielschowsky A. Lectures on Motor anomalies. Hanover NH. Darthmouth College Publications 1943.
11. Walsh and Hoyt. Clinical Neuro Ophthalmology. Vol I, 3rd Ed. Baltimore: Williams Watkins Co, 1969: 264.
12. Zee D, Leigh J. The neurology of eye movement. Philadelphia: F A Davis & Co, 1983: 158.
13. Duke Elder. System of Ophthalmology Vol VI. Ocular motility and strabismus. London: Henry Kimpton, 1973: 668.
14. Duke Elder. Textbook of Ophthalmology. London: Henry Kimpton, 1949: 4043.
15. Hugonnier R. Strabismus, Heterophoria, ocular motor paralysis. St Louis: CV Mosby Co, 1969: 346.
16. Burian HM, von Noorden. Binocular vision and ocular motility. St Louis: CV Mosby Co, 1974: 344.
17. Stephens A, Kelly J, Merrick F, Marshall S, Doyle M. Aust. Orthop J 1989; 25: 23.

Australian Orthoptic Journal 1990; 26: 24-27

BROWN'S SYNDROME, CURRENT CONCEPTS AND A CLINICAL REVIEW OF TWENTY CASES

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Abstract

Twenty cases of Brown's syndrome (two of which were acquired) who presented to the Eye Clinic at The Children's Hospital during a 12 month period from 1987 to 1988 are reviewed. The findings, which are similar to other published studies of this condition, show that the syndrome is generally a benign entity with a comparitively low incidence of strabismus and amblyopia. However, the examiner must be alert to the possibility of associated general and ocular conditions.

Key words: *Brown's syndrome, superior oblique tendon sheath syndrome, strabismus.*

The restrictive movement of Brown's syndrome (superior oblique tendon sheath syndrome) is thought to be due to an abnormality of the superior oblique tendon, its sheath or the Trochlea, preventing free passage of the tendon through the trochlea during inferior oblique action. Brown originally postulated in 1950[1] that the course was that the check ligament of the anterior tendon sheath of the superior oblique muscle was short and tight, possibly secondary to a congenital inferior oblique weakness. Electromyography studies, however, have shown that inferior oblique function is normal.[2]

Brown, in 1973 described two main groups: (i) true — which was congenital, permanent, and due to a congenital shortening of the anterior tendon sheath of the superior oblique, and (ii) simulated — where the anomaly could be permanent or intermittent, was acquired and had various aetiologies. Congenital cases which did not appear to be due to a shortened anterior tendon sheath were included.[3]

Parks and Brown describe an abnormal tendon, which is lacking the normal elasticity, to better explain a variety of clinical findings.[4]

Abnormal insertions of the superior oblique have been described in congenital cases where presentation was late in childhood, suggesting progression of the anomaly. Sandford-Smith postulates an abnormal relationship between tendon sheath and trochlea, secondary to an abnormal insertion, causing general "wear and tear" and secondary tendon swelling.[5]

It is also thought that adhesions between the sheath and tendon in its anterior parts, or, swelling or a nodule in the tendon behind the trochlea, is the mechanism leading to the acquired forms of Brown's syndrome.[6] This would allow movement of the tendon through the trochlea when the muscle is actively contracting but would prevent movement in the opposite direction.

Acquired Brown's syndrome is usually due to inflammation or trauma. Inflammatory changes may be due to local inflammation in the orbit; or part of more generalised inflammatory diseases such as rheumatoid arthritis and tenosynovitis.[5] Sinusitis and diseases of the nasopharynx can lead to a Brown's syndrome secondary to fibrous proliferation about the

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trochlea, the tendon and its sheath preventing free movement.[7,8]

Wright reported a clinicopathologic study of a patient with acquired inflammatory Brown's syndrome.[9] The superior oblique tendon, trochlea, and anterior superior oblique muscle were surgically-removed and studied by light microscopy. The entire specimen was normal, without signs of inflammation or intrasheath scarring. The only abnormal finding was perisheath adhesions anterior to the trochlea. This indicated that the cause of acquired inflammatory Brown's Syndrome may, in many cases, be due to scarring around the tendon sheath, rather than an intrasheath pathologic condition.

The acquired form of the syndrome can result from direct trauma to the region of the trochlea. A similar appearance is seen after some orbital floor fractures, frontal ethmoidal fractures, and crush fracture of nasal bone.[10,11] Trimble, Kelly and Mitchell have reported two cases which followed windscreen glass injuries, which initially presented as superior oblique weakness, but at about one month after injury the disorder spontaneously changed to a typical Brown's syndrome.[12]

Brown's syndrome has also followed frontal sinus surgery due to secondary fibrous proliferation about the trochlea and superior oblique tendon, and has been reported after superior oblique tucking.[11,13]

Booth-Mason and Kyle report the presentation of a 62 year old man with Brown's syndrome due to an orbital metastatic deposit.[14] There was a four month history of diplopia in elevation, and bi-frontal headaches. There was no relevant past medical history — although he had been a heavy smoker. There was no history of ocular disease; nor anything else found on clinical examination. General medical and neurological examinations were normal. Routine haematological, biochemical and immunological tests were negative. Skull and orbital X-rays were normal, but a chest X-ray showed an opacity, which on biopsy was found to be an undifferentiated carcinoma. A computerised tomographic scan showed a mass in the region of the superior oblique, with no bony erosion around the mass. This strongly suggested a metastatic deposit. Radiotherapy to the orbit was tried, but its effects on motility could not be accurately assessed because the patient deteriorated rapidly due to cerebral metastases, and died shortly afterwards.

Both congenital and acquired Brown's syndrome may be intermittent when the "click" phenomenon may be present.[5] An audible or palpebral snap may be noted by the patient or examiner during attempted upgaze. Following sustained effort or pressure over the trochlea, full elevation is possible. The "click" phenomenon is regarded as a stage in the resolution of the condition, Waddell postulating that the mechanisms being either enlargement of the trochlea with growth, or "wearing down" of the swelling with time.[15] In no cases in the following study from The Children's Hospital, could a "click" be recognised.

Pittke relates Brown's syndrome to a pseudoparesis of the inferior oblique muscle and terms it the proximal click syndrome due to abnormal tendon, anterior to the trochlea.[16] He has described a case of distal click syndrome which appears as a pseudoparesis of the superior oblique muscle, where the tendon is affected beyond the trochlea. A 25 year old woman presented for evaluation of "haziness of vision" in her right eye. There was no past ocular history, but fundoscopy revealed scarring and irregularity of the right optic disc. The right visual field showed an inferior temporal "horn-like" scotoma probably due to an old chorioretinitic lesion. There was a pseudoparesis of the right superior oblique muscle, which disappeared by blinking forcibly or by slight shaking of the head. When movement of the right eye stopped, the patient reported brief onset of diffuse, blurred vision, but without diplopia. The patient was unaware of the mechanism by which she compensated for the impaired superior oblique.

With regard to inheritance, Moore, Walker and Taylor[17] report bilateral Brown's syndrome in two siblings from their own experience, and report on six other apparent familial cases with more than one member of a family being affected with Brown's syndrome. In all the reported families the numbers of affected

TABLE I
Comparative incidence (in %) of features associated with Brown's Syndrome (% rounded to nearest whole figure)

This study	Male/Female	Bilateral	Unilateral (R, L)	Forced Primary Gaze			CHP, not binocular	No CHP not binocular
				Binocular	Hypotropia	Intermittent exotropia/ hypotropia		
This study (20 cases)	65/35	20	80 (60/40)	50	15	5	15	15
Clarke and Noel[22] (28 cases)	57/43	11	89 (36/64)	46	18	7	29	—

individuals are small, therefore, speculation on the mode of inheritance is difficult. It has been suggested that an embryologic insult occurs early; the finding of Brown's syndrome in more than one member suggesting that this early insult may have a genetic component.

Eleven cases of inferior oblique palsy were presented by Pollard and the benign nature of this entity stressed.[18] The inferior oblique is the least likely of all the extraocular muscles to be involved in an isolated paresis. Burian and von Noorden[19] have reported that it was the rarest extraocular muscle to be paralysed, and this has been reiterated by von Noorden and Oliver.[20] Of Pollard's cases the aetiology was attributed to congenital, traumatic, or presumed vascular nature. No cases of central nervous system tumour, infection, myasthenia gravis or diabetes were diagnosed. Marlow in 1923 reported one case occurring after a sinus infection, another by a small tumour of the orbit, and another with central nervous system infection due to syphilis.[21] There is little literature reporting inferior oblique muscle paresis.

Although no pathology was found in Pollard's study, patients who present with such must have a full work up — for example, a small stroke that causes inferior oblique paresis might be the harbinger of further vascular disease.

Twenty cases of Brown's syndrome presented at the Eye Clinic at The Children's Hospital over a 12 month period from 1987-1988. Of these 65% were female, and 35% male. Eighty percent were unilateral, of which 40% involved the left eye, and 20% were bilateral. This compares with the similar study by Clarke and Noel[22] who reviewed 28 cases of Brown's syndrome and reported a 57% male predominance, with only 10.7% being bilateral. Of the unilateral cases the left eye was affected in 54%.

In this series, difficulty in elevation was the presenting problem in 35% of cases. Reported horizontal strabismus was the next most frequent cause for concern in 15%, although all of these cases were actually heterophoric. Other presenting problems included compensatory head posture (CHP), school medical service referral, eye injury and a family history of eye problems (although not Brown's syndrome).

Eighteen (90%) of the cases were congenital. Of the two acquired cases, one was due to inflammation from juvenile rheumatoid arthritis which responded well to steroids. The second followed a kick to the face, although no associated orbital fracture was found.

Some binocularity could be demonstrated in 13 (65%) cases, and 10 of these had a CHP (neither of the acquired cases developed a CHP). 50% were binocular in forced primary gaze, which is similar to the Clarke and Noel study. Other similar findings in the two studies are detailed in Table 1. In addition, in this series 20% had esotropia with hypotropia in forced primary gaze, and 10% had constant esotropia.

Congenital Brown's syndrome is usually a benign entity. Amblyopia occurred in six (30%) of these cases, of which two could be classified as strabismic amblyopia and the other four ametropic amblyopia, who required correction of small to moderate amounts of hypermetropia and astigmatism. Two cases required surgery for horizontal strabismus, and one to lessen a CHP. This patient interestingly, had good binocular vision both with and without the CHP.

Congenital cardiac anomalies have been reported in association with this syndrome.[23] Conditions associated with the acquired form include rheumatoid arthritis[3] juvenile rheumatoid arthritis,[24] sinusitis and diseases of the nasopharynx,[11] tenosynovitis,[5] metastatic deposits[14] and trauma.[11]

Associated problems occurring in this series included developmental delay, microcephaly, mild dysmorphia, mild pulmonary stenosis (in the congenital cases) and juvenile rheumatoid arthritis and trauma (in the acquired cases).

Very few ocular anomalies are associated with this syndrome, but ptosis and hypertelorism have been reported.[23] Of the 20 cases in this series, one had lid coloboma and another had asymmetric orbits and lids.

CONCLUSIONS

This series shows similarities with other published studies on Brown's syndrome. It can be seen that it is generally a benign entity, but one must be always alert to the possibility of sinister signs which may also occur.

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References

1. Brown HW. Congenital structural muscle anomalies. In: Allen JH, ed. Strabismus Ophthalmic Symposium. St Louis: CV Mosby, 1950; 205-236.
2. Catford VG, Dean Hart JC. Superior oblique tendon sheath syndrome: an electromyographical study. Br J Ophthalmol 1971; 55: 155-160.
3. Brown HW. True and simulated superior oblique tendon sheath syndromes. Doc Ophthalmol 1973; 34: 123-126.
4. Parks MM, Brown M. Superior oblique tendon sheath syndrome of Brown. Am J Ophthalmol 1975; 1: 82-86
5. Sandford-Smith JH. Superior oblique tendon sheath syndrome and its relationship to stenosing tenosynovitis. Br J Ophthalmol 1973; 57: 859-864.
6. Roper-Hall MJ, Roper-Hall G. The superior oblique "click" syndrome. Orthoptics, ICS 245. Excerpta Medica, Amsterdam. 360-366.
7. Brown HW. Strabismus in the adult. Haik GM (ed). Strabismus Symposium of the New Orleans Academy of Ophthalmology. St Louis: CV Mosby, 1962: 248-273.
8. Clarke E. A case of apparent intermittent overaction of the left superior oblique. Br Orthopt J 1966; 23: 116-117.
9. Wright KW, Silverstein D, Marrone AC, Smith RE. Acquired inflammatory superior oblique tendon sheath syndrome. A clinicopathologic study. Arch Ophthalmol 1982 (Nov); 100: 1752-1754.
10. Zipf RF, Trokel SL. Simulated superior oblique tendon sheath syndrome following orbital floor fracture. Am J Ophthalmol 1973; 75: 700-705.
11. Blanchard CL, Young LA. Acquired inflammatory superior oblique tendon sheath (Brown's) syndrome. Report of a case following frontal sinus surgery. Arch Otolaryngol 1984; 110: 120-122.
12. Trimble RB, Kelly V, Mitchell M. Acquired Brown's syndrome. In: Ravault AP, Lenk M (eds). Trans v Int Orthoptic Congress Lyons: LIPS, 1983: 267-273.
13. Expression Y, Hudelo J, Weiss JB. Syndrome et pseudo-syndrome de Brown. Bull Soc Ophthalmol Fr 1969; 69: 812-815.
14. Booth-Mason S, Kyle GM, Rossor M, Bradbury P. Acquired Brown's syndrome: an unusual cause. Br J Ophthalmol 1985; 69: 791-794.
15. Waddell E. Brown's syndrome revisited. Br Orthopt J 1982; 39: 17-21.
16. Pittke EC. The proximal and distal click syndrome of the superior oblique tendon. Graefe's Arch Clin Exp Ophthalmol 1987; 225: 28-32.
17. Moore AT, Walker J, Taylor D. Familial Brown's Syndrome. J Ped Ophthalmol Strab 1988; 4(25): 202-204.
18. Pollard ZF. Inferior oblique paresis: a benign entity. Ann Ophthalmol 1986; 18: 17-180.
19. Burian H, von Noorden G. Paralytic strabismus. In: Burian H, von Noorden G. Binocular Vision and Ocular Motility. St Louis: CV Mosby, 1974: 357.
20. Oliver P, von Noorden G. Results of superior oblique tenectomy in inferior oblique paresis. Arch Ophthalmol 1982; 100: 581-584.
21. Marlow SB. Isolated paralysis of the inferior oblique. Arch Ophthalmol 1923; 52: 12-24.
22. Clarke WN, Noel LP. Brown's Syndrome: fusion status and amblyopia. Can J Ophthalmol 1983; 18(APR): 118-123.
23. Rao VA, Sood GG and Santoshkumar. Brown's syndrome with congenital ptosis. Ind J Ophthalmol 1982; 30: 57-59.
24. Searle CM, Horne SM. Acquired Brown's syndrome: a case report. Aust Orthopt J 1984; 21: 43-46.
25. Jackson OB (Jnr), Nankin SJ, and Scott WE. Traumatic simulated Brown's syndrome: a case report. J Ped Ophthalmol Strab 1979; 16: 160-162.

Australian Orthoptic Journal 1990; 26: 28-31

BILATERAL BROWN'S SYNDROME ASSOCIATED WITH PREGNANCY: A CASE REPORT

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Abstract

Brown's syndrome is recognised to occur in several forms, both congenital and acquired. Similarities have been noted between certain of the acquired forms and stenosing tenosynovitis of the hand. We report a case of bilateral Brown's syndrome with onset related to pregnancy. As stenosing tenosynovitis is known to occur with increased frequency in pregnancy, the observations in this case support the hypothesis that the two conditions share a similar pathogenesis.

Key words: *Brown's syndrome, pregnancy, stenosing tenosynovitis.*

Brown's syndrome or superior oblique tendon sheath syndrome was described by Whaley Brown nearly forty years ago.[1] Since then both congenital and acquired forms have been described. In 1969 Sandford-Smith drew the parallel between certain of the acquired types, often the intermittent forms, and stenosing tenosynovitis (DeQuervain's tenosynovitis).[2,3]

We present a case of bilateral Brown's syndrome associated with pregnancy. We suggest that this link with pregnancy supports the concept of a similarity between some forms of acquired Brown's syndrome and stenosing tenosynovitis, as the latter is known to occur more frequently during pregnancy or in the post-partum period.

CASE REPORT

A 29 year old woman presented with a history of frontal headaches during her first pregnancy, which increased post partum. She complained that on occasion her eyes became fixed in one position after a rapid change in direction of gaze. These episodes lasted only minutes, during which she noticed diplopia. She had no history of arthritis or local trauma. The pregnancy was complicated by hypertension, fluid retention and proteinuria but proceeded to term with a normal delivery. She was also troubled by painful wrists which persisted for a few months after delivery. She was first seen in our clinic nine months after her first child was born.

She had a face turn to the right, and a gross limitation of either eye on elevation in adduction (Fig 1). There was downshooting of the left eye on dextroversion. A V-pattern was noted in direct elevation with divergence of the right eye. Attempted eye movements in the direction of limitation produced epiphora and pain similar to that complained of as headaches. There was no localised tenderness in the trochlear area; the remainder of the ophthalmological examination was normal. Examination of her wrists revealed no signs of tenosynovitis.

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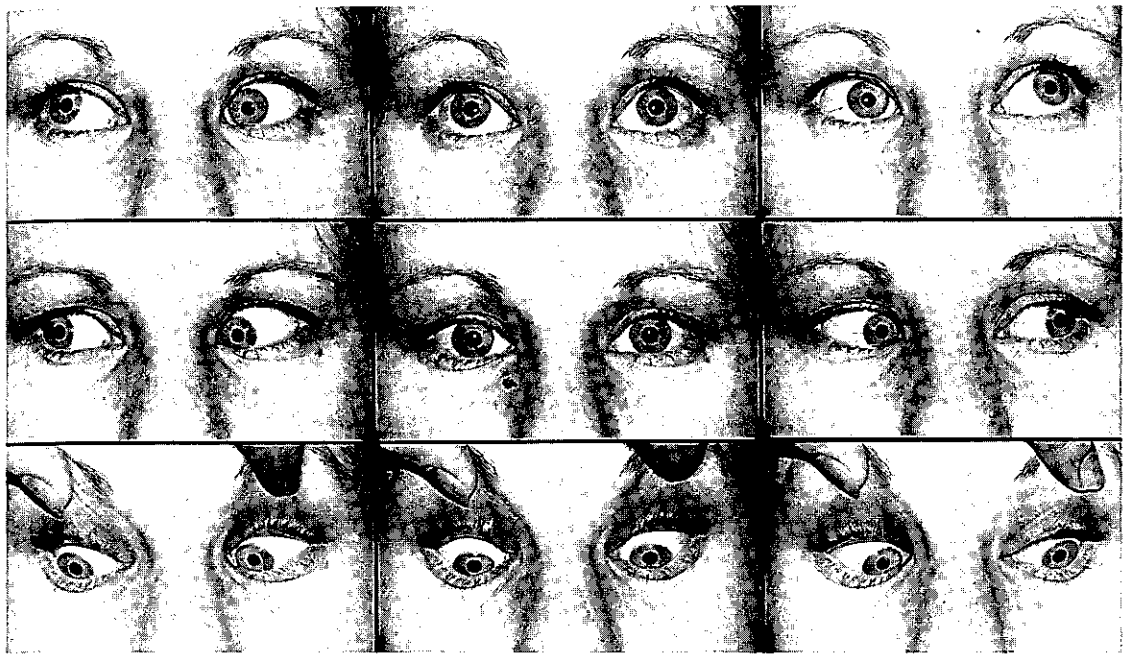

Figure 1: Bilateral Brown's syndrome, with limitation of elevation in adduction more marked in the left eye.

Forced duction testing showed restriction of elevation in adduction. A Hess chart shows a typical bilateral Brown's syndrome, with apparent underaction of the inferior oblique muscles, more marked on the left (Fig 2). There was no overaction of the superior obliques (the ipsilateral antagonists). Antinuclear antibody, rheumatoid factor and erythrocyte sedimentation rate were normal. A CT scan revealed no abnormality of the orbits or adjacent frontal sinuses.

Ocular motility did not change significantly during subsequent visits. Her symptoms increased during a second pregnancy eighteen months later and were only minimally improved by topical dexamethasone eye drops. Post partum, the non-steroidal anti-inflammatory drug ketoprofen 100mg daily produced partial relief of the discomfort. Hess charts showed no significant improvement in motility.

DISCUSSION

PATHOGENESIS

Mein lists seven possible aetiologies for limitation of elevation in adduction:[4]

1. Short anterior tendon sheath
2. Swelling on the tendon
3. Iatrogenic after tucking of the superior oblique tendon
4. Trauma in the region of the trochlea
5. Anomalous innervation
6. Structural abnormality of inferior oblique
7. Congenital inferior oblique palsy

A short anterior tendon sheath is the original theory suggested by Brown in which initial improvement was obtained after surgical division of the sheath. Such a congenital onset is unlikely in our case given the late presentation without previous suggestive symptoms, although it cannot be conclusively disproved in the absence of a surgical exploration.

Swelling on the tendon was first proposed as an aetiology by Girard, who suggested in 1956 that a circumferential constriction of the trochlea and sheath prevented a locally enlarged tendon from sliding freely.[5] Sandford-Smith states that the cause is a swelling of the tendon of the superior oblique just behind the trochlea at the mouth of the fibrous sheath. Despite muscle

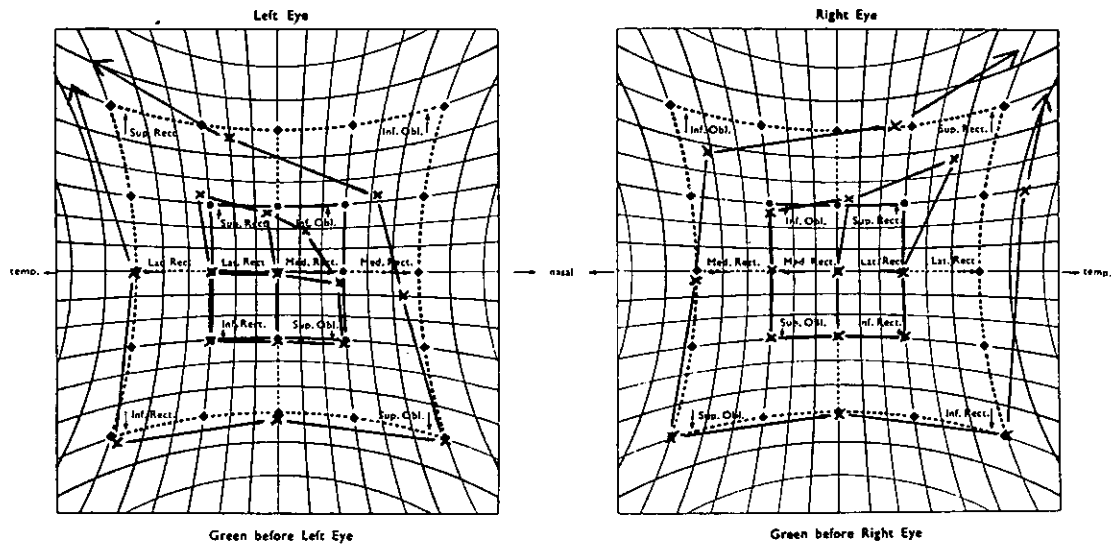

Figure 2: Hess chart showing bilateral Brown's syndrome. Note apparent underaction of the inferior oblique, more marked on the left, but no superior oblique overaction.

relaxation, the swelling is unable to pass into the sheath, causing a restriction of ocular movement. In his original case, the underlying cause was chronic nodular rheumatoid arthritis.[2] However, in 1973 he likened the mechanism of Brown's syndrome to the condition of stenosing tenosynovitis in the flexor tendons of the fingers or abductors and extensors of the thumb.[3] These tendons also pass in fibro-osseous tunnels and develop painful swellings which limit their movement. Sandford-Smith and Waddell[6] hold this to be the cause of the more common type of Brown's syndrome presenting in children, as well as many of the acquired cases, especially those which are intermittent.

Mein describes three other features commonly seen with this aetiology:[4]

(i) Movement of the eye in the direction of action of the inferior oblique may be improved with practice.

(ii) Discomfort or pain may be experienced by the patient on attempted elevation in adduction.

(iii) The patient may describe a clicking sensation resulting in an increase in movement or even an overshoot.

Discomfort and pain when attempting to move either eye into the area of action of the inferior oblique are marked in our patient. She also complained of locking of the eye in one position on several occasions, but a clicking sensation was not noticed. No increase in movement was obtained during our follow up period. We feel this is the likely mechanism in this case.

An iatrogenic Brown's syndrome after tucking of the superior oblique tendon or trauma in the region of the trochlea were ruled out with a negative history.

Anomalous innervation, structural abnormality of inferior oblique or a congenital inferior oblique palsy are unlikely for several reasons. Innervational defects would not result in symptoms of discomfort. Positive forced duction tests confirmed a mechanical rather than an innervational cause in our patient. The usual sequelae following paralysis of a vertical muscle, that is overaction of the ipsilateral antagonist (the superior oblique of the same eye) was not present. Finally the presence of a V-pattern, according to Mein,[4] is a more reliable method of diagnosis than a forced duction test. With a palsy of the inferior oblique muscle, one would expect an A pattern, since the adducting power

of the affected muscle is lost. However in Brown's syndrome, extra innervation is sent to the inferior oblique in an attempt to elevate the eye and as elevation is mechanically restricted by the superior oblique, adduction occurs with production of a V pattern.

ASSOCIATION WITH PREGNANCY

Musculoskeletal problems such as stenosing tenosynovitis, have recently been recognised to occur with greater frequency in pregnant and post partum women.[7] The similarity between Brown's syndrome and stenosing tenosynovitis has already been discussed. Schned found that 25% of female patients with stenosing tenosynovitis were pregnant or postpartum.[8] The reason for this particular susceptibility is not clear. Hormonal effects such as changes in oestrogen and progesterone secretion and the production of relaxin have been implicated in the increased frequency of carpal tunnel syndrome in pregnancy, and Schned suggests these hormonal changes may also play a role in stenosing tenosynovitis.[8] Five of six patients in one series[7] had hand swelling that could have contributed to tendon compression. Mechanical factors involved in caring for newborn babies have also been suggested to play a role.

Our patient had fluid retention and painful wrists during her first pregnancy. Mechanical factors would not seem to be significant in periocular disease, but hormonal factors and fluid retention may have been involved. Ptosis has been reported in pregnancy, thought to be of the aponeurosis defect type caused by a similar mechanism of fluid retention,[9] but we know of no previous reports of the association of Brown's syndrome and pregnancy.

MANAGEMENT

The treatment of Brown's syndrome is controversial and many surgical approaches have been described. While the findings at operation in some cases have helped elucidate some of the causes of the condition, therapeutic results have been disappointing in the long term,[10] probably due to subsequent fibrosis and further restriction of the tendon in its delicate sheath.

We considered medical therapy in our case because the main symptoms were those of discomfort rather than disturbance of functional binocularity. Systemic medications were not used during the pregnancy, and topical steroids produced little effect. Ketoprofen, a non-steroidal anti-inflammatory agent used post partum, has provided moderate relief from symptoms, but no change in ocular motility. Other modalities used for stenosing tenosynovitis in the hand include splinting, surgery and the use of local injection of depot steroid.[8] Splinting is not possible and surgery has been discussed. Depot steroid injections are possibly worth consideration although we have elected not to use them in this case as yet.

CONCLUSION

Stenosing tenosynovitis is recognised to occur with a greater frequency in pregnancy. The observation in our case of the onset of bilateral Brown's syndrome related to pregnancy supports the argument that certain of the acquired forms of Brown's syndrome and stenosing tenosynovitis of the hand share a similar pathogenesis.

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References

1. Brown HW. In Allen JH Ed: Strabismus Ophthalmic Symposium I. St Louis: Mosby, 1950; 205.
2. Sandford-Smith JH. Intermittent superior oblique tendon sheath syndrome. Br J Ophthalmol 1969; 53: 412-417.
3. Sandford-Smith JH. Superior oblique tendon sheath syndrome and its relationship to stenosing tenosynovitis. Br J Ophthalmol 1973; 57: 859-865.
4. Mein J. Superior oblique tendon sheath syndrome. Br Orthopt J 1971; 28: 70-76.
5. Girard LJ. Pseudoparalysis of the inferior oblique muscle. Sth Med J 1956; 49: 342-346.
6. Waddell E. Brown's syndrome revisited. Br Orthopt J 1982; 39: 17-21.
7. Schumacher HR, Dorwart BB and Korzeniowski OM. Occurrence of De Quervain's tendinitis during pregnancy. Arch Intern Med 1985; 145: 2083-2084.
8. Schned ES. DeQuervain tenosynovitis in pregnant and post partum women. Obstet Gynecol 1986; 68: 411-414.
9. Sunness JS. The pregnant woman's eye. Surv Ophthalmol 1988; 32: 219-238.
10. Fells P. The superior oblique: its actions and anomalies. Br Orthop J 1975; 32: 43-53.

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VISUAL AND OCULAR MOTILITY PERFORMANCE OF ONE HUNDRED CRICKETERS*

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Abstract

Evidence from the U.S.A. suggest that good standards of vision and general ocular co-ordination are factors in higher levels of skill of baseball players. A similar study has not been conducted on a group of Australian sportsmen. The purpose of this study was to establish the relevance that various ocular standards have on performance skills of a group of cricketers of varying capabilities. The sample population comprised 100 cricketers from 1st grade to 10th grade. The tests performed were: visual acuity, cover test, ocular rotations, fusion, stereopsis and colour vision. The different grades of cricketers were compared with respect to their performance on these tests to ascertain whether or not visual and ocular motility defects were influential factors in performance levels.

The results showed that there was no statistically significant difference ($p < 0.001$) between the ocular status of cricketers in the higher grades as compared to those in the lower grades on the tests performed.

Key words: *Visual performance of sportsmen, vision, ocular rotations, fusion, stereopsis, colour vision, cricketers, ocular muscle balance.*

INTRODUCTION

Recently there has been an increasing interest in sports medicine and within that speciality, an increasing interest in visual function and sport.[1,2] The areas of visual function that have been investigated include static, kinetic and dynamic visual acuity, ocular muscle balance, stereopsis and visual fields, and claims have been made that athletes have better visual abilities than non-athletes, and that good standards of vision and good ocular co-ordination are factors in higher levels of skill.[3,4,5,6] It has been reported that more successful athletes have better ocular muscle co-ordination and stereopsis than less good athletes.[2]

Current research also indicates that certain ocular functions can be improved through visual therapy, as indicated by Stine, Artberton and Stern[2] (with depth perception and phorias), and Burian and Von Noorden[7] (with accommodation). But does this enhance an athlete's performance?

Thus, the question of visual training of athletes using ocular motility exercises needs to be addressed. Publications such as "Sports Vision Highlights"[1] recommend that vision specialists become part of a sport's teams required coaching panel.

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One would expect that the basis of visual training would be based on three assumptions:

1. that athletes have better visual ability than non-athletes;
2. the visual abilities are trainable;
3. that visual training is transferable to improving the performance of athletes.

The aim of this study was not to investigate the effect of visual training but to address the first question — do athletes have better visual ability than non-athletes?

In order to do this, one hundred district and sub-district cricket players were assessed to try to establish the relevance, if any, that various ocular standards have on performance skills and how the visual status of this group of sportsmen compared to the normal population.

SUBJECTS

The study was conducted on one hundred cricketers from several clubs with district and sub-district grades. In Melbourne, district clubs are those from which state players are selected, while sub-district players take part in a suburban competition of a lower standard. However, according to the various coaches, the level at which players participate is not necessarily an indication of their skill level as some players, for example, choose to play in the sub-district competition when they have the ability to be in the district competition. In this study were forty players from a district club from first to fourth grades, with ten from each grade, and sixty players from sub-district level. There are also four grades at sub-district level, so there were ten players at each level and two composite grades of twenty players. They ranged in age from eighteen years to thirty-four years with an average of twenty-six years.

METHOD OF VISUAL ASSESSMENT

One hundred cricketers were surveyed. The tests performed were:

* visual acuity with the Snellens chart at 6 metres
* cover test at 1/3 metre and 6 metres
* smooth pursuit movements to nine positions of gaze at 1/3 metre
* saccadic movements both vertically and horizontally at 1/3 metre (both smooth pursuit and saccadic movements were assessed subjectively by the examiner. Equipment was not available to test these functions objectively).
* convergence measured by the RAF Gauge
* stereoacuity using the T.N.O. and
* colour vision using the Ishihara test.

All tests were administered as defined by Burian and Von Noorden[7] and Stein and Slatt.[8]

Tests were performed with optical correction if the correction was worn during competition.

RESULTS

A major problem in a survey of this type involves the definition of skill ranking. The assumption is that if a player is playing in first grade he is of greater skill than a fifth grader. It was decided for the purpose of this study the best method was to rely on the club's existing selection criteria to grade their players on a normal competitive basis.

A spearman's table correlation was chosen to analyse the results. The SPSS-X version 2.1 statistics model was used to give point biserial correlation to show any significance between grade and performance on ocular testing, the dependent variable being the cricket grade versus performance on visual tests. Percentages of the sample population (cricketers) who failed or those who over achieved the visual tests were also compared to the "normal" population standards.

The criteria for normal values were selected from existing studies and a combination of accepted standards.[9,10,11,12] As Jolly[13] commented in 1985, the number of studies conducted on general populations is limited, and so no direct comparison can be made with a group of similar age. However, it is interesting to also compare these results with Brown[9] who reported on the visual screening of 5,000 kindergarten age children and with those of Jolly[13] who tested forty-three junior athletes.

1. VISUAL ACUITY

The visual acuity results showed that 95% had vision 6/6 or better with both eyes (See Table 1).

TABLE 1
Visual Acuity

	This Study	Jolly	Brown
Vision 6/6 or better both eyes	95%	91.5%	80%
Vision 6/6 or better one eye 6/9 other	2%	4.25%	5.6%
Vision 6/9 both eyes	2%	0	8.1%
Vision one eye less than 6/9	1%	4.25%	6.3%

While these results may suggest better that average vision when compared to the Brown study, there was no statistically significant correlation ($p<0.01$) between the vision of a cricketer and his grading. These results are similar to those of Jolly.

2. OCULAR MOTOR DEVIATIONS

The cover test showed that 4% of the cricketers had strabismus which is consistent with the Brown and Jolly studies, the generally accepted population norm. (see Table 2).

While the incidence of heterophoria (40%) is similar to the Brown study, it is less than that found by Jolly. There are no known population norms for this age group, however as this study is consistent with the larger of the two studies, and as there is no proof that the incidence of heterophoria alters with age, it is argued that this study possibly reflects more accurately an incidence which is similar to that of a normal population. It is also possible that different diagnostic methods could explain some of the differences.

Because the incidence of strabismus and heterophoria is consistent with the Brown study, then the incidence of orthophoria is also similar. However, when cricketers of the various grades were compared, the incidence of orthophoria was significantly higher ($p<0.01$) in players in the top three grades.

An examination of ocular rotations, including saccadic movements and convergence, showed no significant abnormalities as compared to the Brown and Jolly studies nor statistically significant difference ($p<0.01$) between grades. (See Table 3).

TABLE 2
Ocular Motility Defects

	This Study	Jolly	Brown
Orthophoria	56%	21.25%	40.7%
Heterophoria	40%	74.5%	55.8%
Strabismus	4%	4.25%	3.5%

TABLE 3
Ocular Rotations

	This Study	Jolly	Brown
Full movements	60%	56%	—
Abnormal movements	40%	44%	—
Normal conv.	71%	68%	86.4%
Reduced conv. (>6 cm)	29%	32%	13.6%

3. STEREOACUITY

Most subjects (56%) achieved 60 seconds of arc, with the mean value being 90 seconds of arc. Romano, Romano and Puklin[14] have stated that 40 seconds of arc is normal stereoacuity. The most common score of 60 seconds of arc as tested on the TNO can be explained by the fact that this test jumps from 60 to 30 seconds of arc with no 40 seconds of arc test plate. If a player failed one of the two plates at 30 seconds of arc, it was deemed an overall fail, and recorded as 60 seconds of arc.

These results are inconsistent with those of Jolly who found a mean level of 47 seconds of arc, with the largest single group (47%) achieving 30 seconds of arc. The higher levels of stereoacuity were particularly found amongst the tennis players, and it was suggested that "the natural selection operating in sporting activities could be influenced by visual standards." The results of this study are more consistent with those of Frisby, Neilson and Parker[12] who examined sixty-

TABLE 4
Stereoacuity (TNO Results)

	This Study	Jolly
25 seconds	—	5 (10.6%)
30 seconds	22 (22%)	22 (46.8%)
60 seconds	56 (56%)	15 (31.9%)
120 seconds	10 (10%)	3 (6.4%)
240 seconds	—	—
480 seconds	8 (8%)	—
Nil	4 (4%)	2 (4.3%)
Total	100 (100%)	47 (100%)

TABLE 5
Colour Vision

	This Study	Normal Population
Normal	97%	92%
Defective	3%	8%
Total	100%	100%

eight university students whose "mean age was about 20 years". The mean response was 82 seconds of arc.

As with the cover test results it is argued that these findings are more consistent with and therefore reflect the stereoacuity levels found in the "normal" population. There were no significant different ($p > 0.01$) in stereoacuity levels between cricketers in the different grades.

4. COLOUR VISION

Results of the Ishihara test showed 3% of cricketers had defective (red-green deficiency) colour vision, which is less than the 8% reported in an average male population.[16] It is interesting to note that a 2nd grade district player failed the red-green Ishihara section completely. (See Table 5).

CONCLUSION

In conclusion, this study shows that there is no statistically significant difference on visual tests between the different grades. When assessed by the methods stated above, the statistical analysis revealed that the incidence of ocular defects found was similar to that of the normal population and that they were evenly spread over all grades.

The results did show that higher grade players have higher than normal incidence of orthophoria and that cricketers in this sample population had a lower incidence of red-green defects than the normal male population.

While these findings are useful for describing a relationship between two variables, i.e. the cricketing grade and the results of the visual tests, further investigation is required to demonstrate a causal relationship.

However, the tests performed were predominantly of an ocular motor nature, and so far there is no evidence of what level of visual status is required of a cricketer.

Cricketing pundits are often fond of quoting that great batsman "see" or "pick-up" the ball sooner. However, while the demands on the integration of sensory and ocular motor apparatus is high, the importance of ocular motility to this process is questionable. In the closed environment of a cricket ground, the visual demands of a player are defined; the player knows/assumes the moving target is the ball. Does the player's vision system really need the ability to count the stitches on the ball before it triggers the physical response of body movements?

The results of this study indicate that this sample population of cricketers has a similar incidence of ocular motility disturbances to those found in the "normal" population. It found a significant ($p < 0.01$) correlation only between orthophoria and higher grade players.

References

1. Sports Vision Highlights — a newsletter dedicated to sports vision professionals. 1982; vol. 1, No. 2.
2. Stine CD, Arterburn MR and Stern AS. Vision and sports: a review of the literature. J Am Optom Assoc 1982; 53 (8) August: 627-633.
3. Sherman A. Overview of research information regarding vision and sports. J Am Optom Assoc 1980; 15(7) July: 661-665.
4. Watanabe Y. Effect of 15 minute bicycle workload on static and kinetic visual acuities. J Sports Med Phys Fitness 1983; 23(4) December: 373-381.
5. Hoffman LG, Rouse M and Ryan JB. Dynamic visual acuity: a review. J Am Optom Assoc 1981; 52(11) November: 883-887.
6. Winograd S. The relationship of timing and vision to baseball performance. Res Q Am Assoc Health Phys Educ 1942; 13: 481-493.
7. Burian H and Von Noorden G. Binocular vision and ocular motility: theory and management of strabismus. 3rd ed. St Louis: C.V. Mosby Company, 1974.
8. Stein and Slatt. The ophthalmic assistant fundamentals and clinical practice. 4th ed. St Louis, Toronto, London: CV Mosby Co, 1983.
9. Brown S and Jones D. A survey of the incidence of defective vision and strabismus in kindergarten age children — Sydney 1976. Aust Orth J 1977; 15: 24-28.
10. National Health & Medical Research Council (Australia) Occupational Health Guide. Vision & eye sight tests for visual display terminal (VDT) operators. Canberra: Dept of Health, 1980(?).
11. Tredici Thomas J. Aerospace Ophthalmology. United States Air Force School of Aerospace Medicine (n.d.).
12. Frisby JP, Nielsen P and Parker J. Clinical tests of stereoacuity: do they measure the same thing? Orthoptics Research & Practice. Mein & Moore (eds.) London: Henry Kimpton, 1981; 211-214.

13. Jolly N and Jolly R. Visual standards of the participants in the Shaklee Junior Sports Development Programme. Aust Orth J 1985; 22: 41-47.
14. Romano PE, Romano JA and Puklin JE. The development of normal binocular single vision in childhood. Orthoptics Past, Present and Future. Moore, Mein, Stockbridge (eds). New York: Stratton Intercontinental Medical Book Corporation, 1976: 11-16.
15. Cruz-Coke R. Colour blindness an evolutionary approach. Illinois, U.S.A.: Charles C. Thomas, 1970.

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APPLICATIONS OF MICROCOMPUTERS TO ORTHOPTIC MEASUREMENT

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Abstract

This communication emphasises the advantages of direct and objective measures of visual functions in the clinical setting. The applications of microcomputers in the role are outlined and an example of such a system as developed in the School of Orthoptics at Cumberland College of Health Sciences, Sydney, is described. This system utilises the basic IBM microcomputer for analysis and storage of patient data. Additional hardware enables the system to generate complex visual stimuli and to record the visual responses evoked by these stimuli.

Current interests are centred on visual evoked responses, electroretinograms and eye movement studies. The interfacing of standard ophthalmic testing equipment, eg perimeters and ultrasound, adds a further dimension to the system. The relatively low cost and simplicity of operation of microcomputer/based systems suggests they are likely to proliferate within the eye clinic environment.

INTRODUCTION

Many measures of visual function used by orthoptists tend to be subjective in that they rely upon verbal responses from subjects and therefore their co-operation and ability to communicate effectively. In some patient groups, such as infants, these inbuilt variables may limit the reliability and accuracy of measurements to the extent that their clinical value is questionable. There is therefore a strong incentive to develop techniques capable of direct or objective measurement of visual parameters.

To date the advantages of objective measurement of visual responses have been more than offset by the technical nature of the necessary equipment, its cost, a lack of technical support personnel and the time required and complexity of testing procedures. Due to these limitations, such equipment has been largely restricted to a few specialist clinics and research laboratories.

The purpose of this communication is to draw attention to the current and potential impact of microcomputers on measurement.

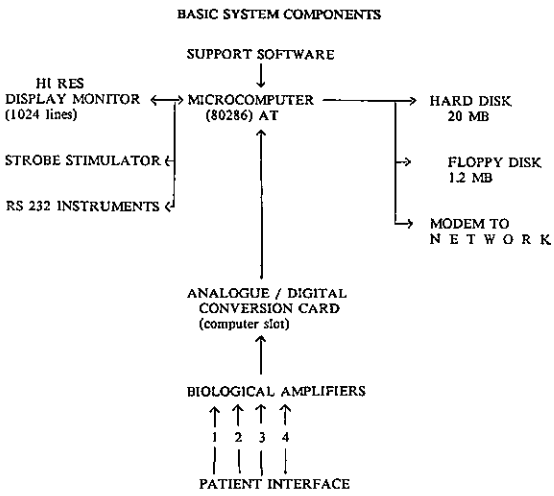

Figure 1: Flow diagram of major system components.

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BASIC SYSTEM COMPONENTS

In recent years, we have begun to develop microcomputer-based systems for selected measurement applications. These have been used in teaching and collaborative clinical research. The current system is shown in Fig. 1. To the basic IBM AT computer, additional hardware can be added via slots provided in the rear of the machine. In our system a variety of biological signals are amplified and then fed to the computer via a slot-mounted analogue to digital converter (IO) board. Recordings, the results of data analysis and patient details are then stored on the large capacity hard disk and backed up on floppy disk.

VISUAL STIMULATION

Currently VER, ERG and EOG signals are processed on the above equipment. The properties of these responses are largely stimulus dependent. The value of the test is substantially increased by tailoring of the stimulus to suit the test design, the ideal being complete control over all stimulus parameters. Microcomputer systems offer advantages in this area. With suitable interfacing to stimulator devices important stimulus parameters may be set from the keyboard. By way of example, patterned stimuli are generated by the computer and presented to the patient on a high quality monitor with reasonable control of intensity, contrast, size, spatial and temporal frequency and drift (movement). A bright strobe flash stimulator is needed for conventional ERG testing. In this case, appropriate computer interfacing enables control of stimulus intensity and temporal frequency.

CURRENTLY SUPPORTED TESTS

Fig. 2 summarizes a range of tests supported by the system as described above. These fall under the broad headings of evoked responses, eye movement analysis and psychophysical assessments. The latter rely upon verbal/subjective responses rather than electrical measures, however computers offer the advantages of improved control over stimulus parameters and presentation.

1. Evoked Responses
   - Pattern visual evoked responses

   Which support the following assessments:
   - — lesions
   - — grating acuity
   - — contrast sensitivity
   - — field loss and abnormal projections

   - Flash ERG

   Which support tests of:
   - — retinal integrity
   - — rod/cone function
   - — pigment abnormalities

   - Pattern ERG

   Which supports assessment of:
   - — localised retinal lesions
   - — ganglion cell/optic nerve function (eg. optic neuritis, glaucoma)
   - — retinal contrast sensitivity

2. Eye Movements
   - Electro-oculogram

   To investigate:
   - — retinal adaptation
   - — peripheral disorders of ocular motility
   - — central nervous system pathology/nystagmus

3. Behavioural/Psychophysical Measurements
   - Contrast sensitivity
   - Grating acuity
   - Snellen acuity
   - Colour vision

Figure 2: List of tests supported by the current system.

Figure 3 illustrates the potential for generating a haploscopic stimulus by the addition of a second monitor. A project has commenced at this college to use such an arrangement to generate Hess screen measurements.

RS232 INSTRUMENT INTERFACE

Increasing numbers of instruments are supplied with the above computer interface (eg. Allergan Humphrey perimeter, Allergan ultrasound, some autorefractors). In general this facility enables transfer of test measurement data files to the PC. The advantages of this facility include the establishing of normative data banks, centralising of

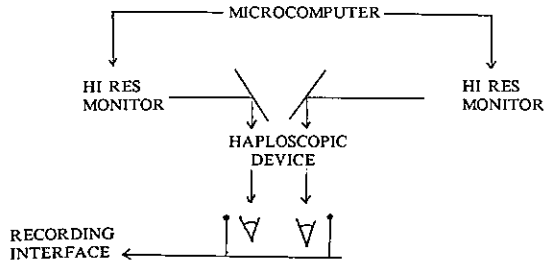

Figure 3: Illustrates use of dual monitors with suitable optics to provide a haploscopic stimulus.

patient files and tailoring of evaluation and reporting protocols.

IMPLICATIONS OF MICROCOMPUTERS FOR CLINICAL PRACTICE

The ability of microcomputers to rapidly store and retrieve large volumes of information enables them to readily accumulate a data bank of test measurements and patient data. This raises the possibility of improved statistical evaluation of individual results against normative data. These features can be incorporated into the software used for report generation so that they are fully automated and become a routine part of the patient assessment. The result is improved confidence and reliability in testing.

The addition of a modem (interface to the telephone) to the system raises the possibility of transferring test data, patient details and completed reports between clinics. Networking of clinics offers a number of exciting prospects including the pooling of normative test data so as to increase the size of the bank.

CONCLUSIONS

The applications described above probably represent only a small fraction of those with potential in orthoptic practice. The analysis and information handling capabilities of the modern systems provide an opportunity to include valuable but previously too complex and time consuming procedures into routine patient assessments.

The flexibility of microcomputer systems is such that the number of applications is limited only by the imagination and the availability of suitable computer interfaces. It is now very apparent from the most preliminary cost/benefit analysis that the applications will continue to proliferate through most areas of practice within a relatively short time. The challenge for Orthoptists will be to join the initiaters rather than the recipients of this technology.

ABSTRACT

This communication emphasises the advantages of direct and objective measures of visual functions in the clinical setting. The applications of microcomputers in the role are outlined and an example of such a system as developed in the School of Orthoptics at Cumberland College of Health Sciences, Sydney, is described. This system utilises the basic IBM microcomputer for analysis and storage of patient data. Additional hardware enables the system to generate complex visual stimuli and to record the visual responses evoked by these stimuli.

Current interests are centred on visual evoked responses, electroretinograms and eye movement studies. The interfacing of standard ophthalmic testing equipment, eg perimeters and ultrasound, adds a further dimension to the system. The relatively low cost and simplicity of operation of microcomputer/based systems suggests they are likely to proliferate within the eye clinic environment.

J. A. Burne — Applications of microcomputers to orthoptic measurement

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HOMONYMOUS HEMIANOPIA: TRAINING COMPENSATORY STRATEGIES

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Abstract

SEETEC is a project funded by the Royal Guide Dogs Associations of Australia. It provides mobility training for people with visual disabilities.

Over the past two years, SEETEC has developed a programme for people who have homonymous hemianopia. Most people are referred following a stroke, head injury or removal of a tumour.

To date 169 people in NSW have been assessed for this service. Of these, 100 undertook a programme of visual training.

This paper summarises the objectives, method and results of the programme.

Key Words: *Homonymous hemianopia, cerebral vascular accident (CVA), attention deficit, rehabilitation, vision training.*

Homonymous hemianopia is a visual impairment which involves an attentional deficit as well as sight loss.[1] It is usually due to a single lesion of the optic pathway at or post chiasm. The visual defect is that of half field loss in the vertical meridian of the field. Homonymous hemianopia may affect any age group following a cerebral vascular accident (CVA), head injury or tumour removal.

There are many functional problems which may be associated with the field loss, these problems are determined by the extent and location of the lesion. Generally if the lesion occurs within the occipital lobe the client will compensate for any deficit unaided. Lesions occurring in cortical areas outside the occipital lobe will have a variety of problems. Some of these problems include; difficulty when moving in a confined space, neglect of people and objects within the area of field deficit and problems with scanning activities such as reading.

These functional problems cause extreme handicap and frustration to the person. An important aspect of rehabilitation is to address these problems. Diller and Weinberg[2] formulated procedures in 1977 aimed at providing structured and intensive training to people with hemi inattention. Hill[1] restructured this program in 1981 and extensively evaluated the revised program with 350 patients.

The revised program is currently being used in the rehabilitation of clients with homonymou hemianopia.

The aim of the program is to assist people t travel safely. This is achieved by defining th perimeter of the left or right field, overcomin perceptual rivalry, establishing an automati seeking response into the affected field an improving the speed and accuracy of scanning Client characteristics include an ability to lear (ie. no frontal lobe damage and an awareness o the visual loss). Training should commence a least one month after onset to allow time t develop cortical stability.

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METHOD

Apparatus:

A Diller and Weinberg Tracking Machine described by Diller and Weinberg,[2] is used during the static training phase.

Firstly, the visual loss is demonstrated to the patient using the machine with a central fixation light and a light at both the left and right extremities. The patient is then instructed to look to the affected field and locate illuminated lights. This task increases in complexity by increasing the number of lights to be located and the speed at which they are presented. The patients ability to problem solve and retain visual cues is also assessed by sequencing the presentation of the lights. At the completion of this phase the patient should be able to scan the blind field rapidly.

The next phase is dynamic. The client is required to scan and locate objects presented vertically and horizontally within the field of view whilst walking. Scanning activities are repeated in a variety of outdoor situations until the client is visually safe or the level of visual safety is ascertained.

The following case study illustrates the positive value of this training to the client.

CASE STUDY

Mrs R, 63 years of age was referred for a tracking and scanning program five years after her CVA. Her medical report showed that she had a dense left hemianopia, left hemiplegia, chronic headaches and a chronic anxiety state.

Mr R informed me that his wife's main problem areas were that she missed food on the left hand side of her plate, was startled when people approached her from the left and was unable to enjoy being a passenger in a car because the environment made no sense to her.

Mrs R's hemianopia presented as severe when assessed on the Tracking Machine as she missed all the lights to the left of the centre and made no attempt to search for them. Perceptual rivalry was severe — Mrs R made no attempt to look left first when she was told there were lights on the left and right of the machine.

Over a three week period Mrs R was given nine sessions on the Tracking Machine. In these sessions she was taught how far she needed to turn her head to see the light on the far left periphery. Once she learned this, she was trained to automatically look to the lights on the left before she looked to the lights on the right. Different combinations and the numbers of lights were continually presented to Mrs R until she was able to look to the light on the far left perimeter first and then systematically and accurately name the lights on from left to right.

Reading, static tracking and scanning exercises were then given to Mrs R. Dynamic exercises were not relevant due to Mrs R's severely restricted mobility.

Following the program Mr R told me that his wife ate all the food on her plate at meal times and looked to the left to avoid objects she had previously ignored.

Follow up visits at six weeks, three months and six months revealed that Mrs R had maintained the skills she had been taught.

Mrs R was able to enjoy going driving with Mr R and practised by reading the number plates of cars in front of them. Mrs R was also able to have her visual acuity accurately tested for the first time in four years because she now turned her head far enough left to see the entire chart.

SUMMARY

The rehabilitation program undertaken by Mrs R was part of the program developed by SEETEC for the Royal Guide Dogs Associations of Australia. To date 169 people have been assessed for such programs in NSW, of these people one hundred have undertaken visual training. The tracking program is a vital rehabilitation technique and should be used whenever possible. Further research and development should be encouraged to continue improvement of these techniques.

ACKNOWLEDGEMENT

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References

1. Hill B. Cortical Defects of Vision and Disorders of Space Exploration. SEETEC paper, National Mobility Centre, Royal Guide Dogs Associations of Australia, 1983.
2. Diller L , Weinberg J. Hemi-inattention in rehabilitation: the evaluation of a rational remediation program. In: Weinstein EA, Friedland RP (eds). Advances in Neurology, vol. 18. New York: Raven Press, 1977.

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CORTICAL BLINDNESS IN MULTIHANDICAPPED CHILDREN

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Abstract

Three case studies of diagnosed cortical blindness are presented, and their patterns of recovery are compared, with particular emphasis on length of recovery time and current level of vision. All three were found to progress to a level of "panoramic vision", with any further improvement being limited to within these boundaries, that is, vision primarily concerned with spatial relationships and movement, by orienting the eyes and limbs to sudden peripheral movements.

Key Words: *Permanent cortical visual impairment, panoramic vision.*

Cortical blindness is a phenomenon commonly encountered when working in the field of developmental disabilities. Hoyt[1] found that on examination, the cortically blind child failed to show any visual fixation or following movements retained pupillary responses to light, had no nystagmus and no abnormalities of the ocular structures. These findings were generally associated with widespread neurologic disease.

Denny Brown and Chambers[2] concluded that two distinct visual systems may exist in humans. This conclusion was reached through experiments on Macaque monkeys and through the review of literature on cortical blindness in adult patients. The first system which they labelled "object vision" involves the macula, the lateral geniculate body and the striate cortex, it also requires interaction with the superior colliculus. The second system which they labelled "panoramic vision" is primarily concerned with spatial relationships and movement, orienting the eyes and limbs to sudden movements registered by the peripheral retina. This second visual pathway includes the peripheral retina, the inferior pulvinar nucleous and areas 18 and 19. They found that each system can function without the other, though normally they must be integrated at all levels. This research would indicate therefore that destruction of the striate cortex (area 17) alone should not cause complete loss of vision.

Whiting, Jan, Wong, Flodmark, Farrell and McCormick[3] have identified two forms of cortical blindness which they prefer to call cortical visual impairment (CVI). The first they call transient CVI and the second permanent CVI where little or incomplete recovery occurs. They have also reported that the general pattern of recovery from cortical blindness is that the patient perceives and follows light, then moving objects. Later, visual acuity improves but poor vision and visual-perceptual difficulties may remain permanently.

It is this pattern of recovery that has been observed in a series of multihandicapped children. The conclusions reached by Whiting et al[3] would therefore have implications regarding the extent of visual recovery that can be expected in these children.

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They also found that transient CVI most often is reported following meningitis and minor head injury, with the major causes of permanent CVI being perinatal asphyxia and shunt malfunction.

Hodgson[4] in her study of 22 children with "pure" cortical blindness (ie. with no optic nerve or retinal pathology) found that seventeen showed apparent improvement in vision before the age of two years with no improvement occurring after that. Hoyt[1] in his study of 43 children found that improvement was variable but generally quite slow with visual recovery continuing to occur in some patients more than two years after the original insult.

This study shows how "permanent CVI" has presented in three multihandicapped children currently being seen at the Royal New South Wales Institute for Deaf and Blind Children. All three children have shown improvement in their functional vision with improvement occurring up to six years after the initial insult.

CASE 1

The first child contracted encephalitis at four months of age resulting in a diagnosis of cortical blindness, abnormal muscle tone and developmental delay. He was first seen at 15 months where he was noted to be aware of light by turning towards a diffuse light source such as a window, he did not however, respond to a torchlight. A small to moderate right exotropia was present, optokinetic nystagmus (OKN) was elicited when the drum was rotated with the stripes moving from right to left and when it was rotated in a downwards direction. He was then seen at six monthly intervals, during which a steady improvement was evidenced with the child being able to locate brightly coloured objects that were held at one third of a metre (1/3 m). This progressed to his being able to follow a slowly moving object when he was two and a half years old. At this stage no central fixation was evident, optokinetic nystagmus was now present in all directions. When last assessed at three and a half years he saw and reached for the stycar toys when they were held fifteen to twenty (15-20) centimetres from his eyes. Responses were more consistent if the object was moved slightly. When tested with the stycar balls he was again noted to be more reliable when the balls were moving rather than stationary. Cover testing showed that both eyes were in a divergent position with a right hypertropia. All smooth pursuit movements were irregular with the object of regard having to be moved slowly for him to be able to follow it.

CASE 2

The second child has diagnosed cortical blindness, bilateral optic atrophy, spastic quadriplegia and epilepsy as a result of placental insufficiency and premature birth (34 weeks gestation). He was first seen at 11 months of age at which stage he was noted to be attracted to diffuse sources of light. Occasional horizontal jerk nystagmus was present.

He was seen again at 19 months of age, at this assessment, he displayed searching eye movements in the general direction of a bright torchlight. He also saw and followed a thin white wooden rod as it was moved across his line of vision. Intermittent horizontal jerk nystagmus was present.

He was not seen again until he was four years of age. At this assessment he followed a penlight torch in all directions. If a brightly coloured object was held approximately 30 cm from his eyes he would reach for it only after it had been moved slightly. He followed a slowly moving object in all directions. On cover testing he was found to have a large left hypertropia, the left eye was also slightly exotropic. Fine horizontal nystagmus and some wandering eye movements were present.

CASE 3

The third child suffered severe anoxic brain damage at birth resulting in cortical blindness, microcephaly, global developmental delay, spasticity and epilepsy. He was first seen when he was four years old, on this assessment he was noted to look in the general direction of a light, both eyes were in a divergent position and horizontal OKN was present. He was reassessed at five years, four months of age, where he was found to be reaching on sight for large bright toys at 33 cm, and showing some irregular

following movements. At the age of seven years four months, he would crawl towards patterned objects (such as the OKN Drum) at approximately 1½-2 m where he would inspect them visually. There was no evidence of central fixation.

The most recent assessment was at seven years and seven months. Any signs of improvement at this stage were minimal. It was noted that he responded to a moving object immediately either by reaching for one that was being moved slightly, or by visually following an object. Central fixation was not present.

DISCUSSION

Of the three cases presented, two were the result of birth difficulties and problems during pregnancy. Both of these children have severe developmental delay and visual improvement of any significance was not apparent until they were over three years of age. In the first case the problems did not commence until four months of age. This child is improving in all areas both cognitively and physically and visual improvement was first noted at the age of two years.

None of the children have firmly established central visual fixation and they have all been noted to respond more readily to moving rather than stationary objects. It would appear that they all have "permanent cortical visual impairment".[3] All three present with a similar pattern of recovery and all have progressed to the stage of "panoramic vision",[2] ie. vision primarily concerned with spatial relationships and movement, where they eyes and limbs are oriented to sudden movements registered by the peripheral retina.

Whilst all three children have an intellectual impairment it is felt that this would not account for the type of recovery that has been documented, ie. the apparent visual improvement was not due to the delayed improvement in other functions. The causes in all three are quite diverse as is the recovery time which ranges from three to seven years after the initial insult. The three subjects, whilst having shown a continual steady improvement, have all reached the stage of detecting movement before stationary objects and have limited, if any, central fixation. Once having reached this stage, improvement has been within these limitations suggesting that improvement in visual acuity may not be a realistic expectation.

CONCLUSION

Cortical blindness is commonly encountered when working with multihandicapped children. This paper has presented three subjects who, whilst making a steady improvement in the use of their vision, have all seemed to plateau at the level of "panoramic vision". The implications of this study are therefore, that in cases of cortical blindness visual improvement can occur up to seven years after the initial insult and that the level of improvement may be limited to "panoramic vision".

References

1. Hoyt CS. Cortical Blindness in Infancy. Paediatri Ophthalmology and Strabismus. Trans of the Nev Orleans Academy of Ophthalmology, Raven Press, Nev York, 1986.
2. Denny Brown D, Chambers RA. Physiological Aspect of Visual Perception. 1. Functional Aspects of Visua Cortex. Archives of Neurology 1976; 33: 219-227.
3. Whiting S, Jan JE, Wong P, Flodmark O, Farrell K McCormick A. Permanent Cortical Visual Impairmer in Children. Developmental Medicine and Chil Neurology 1985; 27: 730-739.
4. Hodgson L. Visual Improvement in the Cortically Blin Child. Student Research Papers, Cumberland College c Health Sciences, Sydney, 1986.

Australian Orthoptic Journal 1990; 26: 45-47

AN UPDATE IN GENETICS FOR THE ORTHOPTIST
A BRIEF REVIEW OF GENE MAPPING

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Abstract

Genetics in the field of medical research is an area which is rapidly developing. The process of mapping the human genome via the method of reverse genetics is resulting in the chromosomal location and identification of genes responsible for various inherited disorders. The specific location of genes on specific chromosomes will allow for prenatal diagnosis, presymptomatic diagnosis, carrier detection, confirmation of individuals affected and, later on, possible treatment. In some disorders this has already been the case.

This paper gives a brief overview of one of the techniques used in gene mapping known as Recombinant DNA Technology in relation to inherited eye disease.

Key words: *Recombinant DNA technology, molecular genetics, reverse genetics, chromosomal locations, inherited eye diseases.*

INTRODUCTION

Hereditary conditions until recently have been diagnosed largely on a descriptive basis. This included the pattern of inheritance (family trees), clinical evidence and the natural history of the disorder. It is now, however, possible to describe and diagnose these conditions on a molecular (genetic) basis via reverse genetics and chromosomal locations.

In 1979 the editor of the American Journal of Human Genetics, David Coming, described a new approach to gene mapping and referred to it as the "new genetics". There are today many methods of gene mapping, of which recombinant DNA technology is only one.

To date there has been in the vicinity of 1,200 autosomal genes localised, and 150 X-linked genes mapped.[1] These advances are now playing major roles in the areas of prenatal diagnosis, presymptomatic diagnosis, carrier detection, confirmation of an affected individual and treatment.[2]

Medicine is becoming more and more involved in genetics with these new procedures. The advances made in the treatment of various diseases and the new technology available has meant that many conditions have "cures", but it is the genetic conditions that are still a mystery in many areas of medicine. It has been estimated by Burn[1] that when single gene disorders, chromosomal disorders and structural malformations are grouped together, that they will account for half of all miscarriages, a quarter of perinatal deaths and three-quarters of severe handicaps. As well, one in eighty adults will be affected by a late onset genetic disorder, ranging from Alzheimer's and Huntington's Chorea to diabetes and heart disease. The area of ophthalmology and inherited eye conditions is no different, with genetic disorders ranging from

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retinoblastoma to colour blindness, aniridia and retinal dystrophies. Thus it is the responsibility of all clinicians (including orthoptists) to be aware of the new technology in genetics so that it can be used to the best advantage and care of our patients, especially when it comes to counselling and the services available to them.

REVIEW OF THE CELL AND ITS GENETIC MAKE-UP

In order to understand the process of gene mapping and recombinant DNA technology, it is necessary to review the components of the cell and its genetic make-up.

The nucleus of the cell contains lengths of deoxyribonucleic acid (DNA). These are known as chromosomes, which are grouped into 23 pairs, 22 pairs of autosomes and one pair of sex chromosomes. The DNA consists of a phosphate and sugar band with nucleic bases, bonded loosely together in pairs. There are four bases: Adenine (A), Guanine (G), Cytosine (C) and Thymine (T). These bases are always paired: A = T and G = C. A specified sequence of these bases will lead to the production of a protein.

It is on the chromosome that the genetic information is stored. A gene therefore is a length of DNA containing the information for a specific protein. These specified sequences are not continuous and are known as exons. The intervening sequence which breaks the code between genes and the proteins they produce are known as introns.

The chromosomes, being loosely bonded, separate easily and become single stranded in the process of cell division, providing templates for two new molecules. The DNA after separating is complemented by a matching single strand of ribonucleic acid (RNA), only the base Uracil (U) is used instead of Thymine (T).

These 23 pairs of chromosomes, consisting roughly of six million base pairs, with more than 50,000 genes, constitute the Human Genome. There is a huge task involved in mapping it and it will take many years before it is completely mapped.

GENE MAPPING — RECOMBINANT DNA TECHNOLOGY

Before discussing the procedure involved in locating the chromosome and therefore an abnormal gene, further definitions of new terminology are required.

1. Restriction Enzymes: These are enzymes called restriction endonucleases. When a specific enzyme is applied to DNA it will read and cut the DNA at a specific base sequence, thus producing fragments of DNA of different lengths. The recognition sites vary in length from 4-8 base pairs.
2. Restriction Fragment Length Polymorphisms (RFLPs): These are normal variations in the base sequences of DNA They are inherited and have no apparent clinical effect.[2] These polymorphisms can be detected by subjecting DNA to a restriction endonuclease which will produce a specific length of DNA This is known as a Restriction Fragment Length Polymorphism (RFLP).[3]
3. Gene Probes: A gene probe is an identical copy of a sequence of a single strand of DNA which has been cloned. The copy can be made from messenger RNA by the enzyme reverse transcriptase or directly from DNA The result is a complementary strand of DNA (cDNA). The cDNA also has a radio-active base known as p^{32} included in its phosphate-sugar band. This addition of p^{32} allows the probe to be located by autoradiography and therefore aids in the detection of abnormal genes.

PROCEDURE INVOLVED IN GENE MAPPING

DNA is obtained from any available tissue. The most common and easily accessible form is a blood sample of at least 5 mls. This is sufficient to analyse any normal or mutant gene for which the probe is available. The white blood cells are then extracted, which contain a high proportion of DNA material.

A restriction enzyme of choice, depending on the chromosome and region under analysis, is then added, generating fragments of DNA of specific lengths. These fragments are then

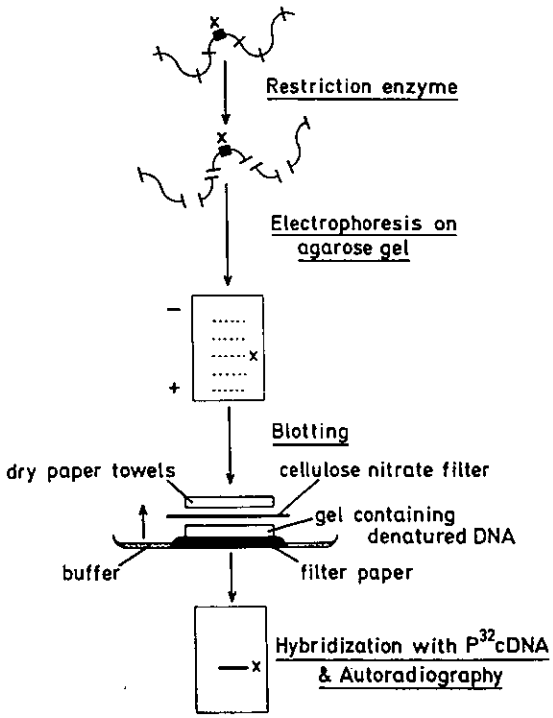


Figure 1: Showing the technique of Recombinant DNA Technology (from Emery AEH. Elements of medical genetics, 6th ed. Edinburgh, Churchill Livingstone, 1983).

subjected to a process known as electrophoresis in agarose gel. This separates the fragments of DNA, which migrate distances according to their molecular weight. Smaller fragments migrate faster than larger ones. The DNA then undergoes a process of denaturisation by an alkali which separates the double strand of DNA into a single strand. This makes it capable of bonding to a probe (cDNA). The single strands are then transferred to a cellulose nitrate filter by a process known as "Southern Blotting".

Next, is the addition of a radioactive probe to the filter. When the cDNA finds its complementary sequence on the filter it 'bonds' and a double strand of DNA is formed. In order to detect an RFLP, a probe within the restriction site of the polymorphic region must be used. The position of the probe can be localised and visualised by auto-radiography and any abnormalities noted.

The radioactive bands highlight the variations and appear at different places as blots of DNA from different individuals. A single blot will indicate that the person is homozygous and a double blot heterozygous.[4] In some cases the band may be missing indicating an abnormality (Figure 1).

SUMMARY

This in a 'nutshell' is a brief description of gene mapping via recombinant DNA technology which is used to localise genes responsible for inherited conditions.

It is hoped that this brief article has helped bridge the gap a little in regard to this rapidly developing area of medicine and genetics.

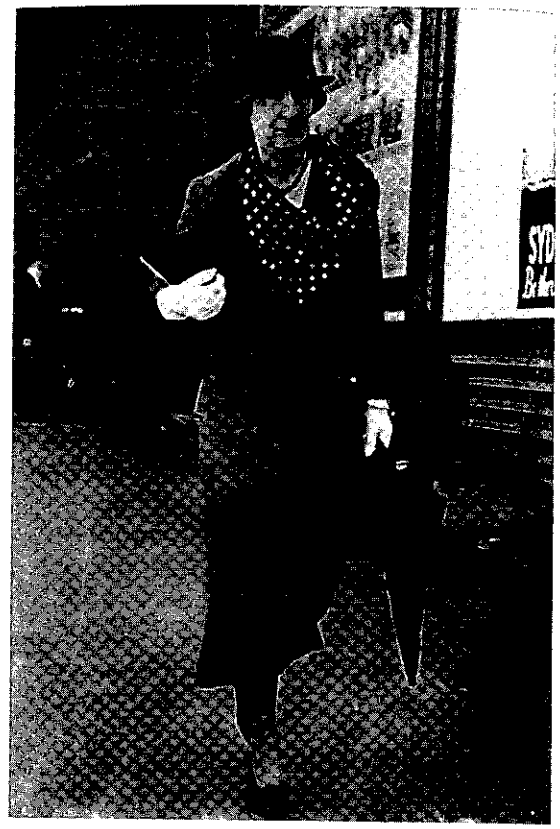
ACKNOWLEDGEMENTS

I wish to thank Dr F B Halliday and Mrs C Toohey, without whose help and support this paper would not have been possible.

References

1. Burn J. Clinical genetics - a review. Journ of Royal College of Phys of London, 1988, Vol. 22: No 4: 212-225.
2. Emery AEH. Methodology in medical genetics. An introduction to statistical methods. 2nd ed. Great Britain: Churchill Livingstone, 1986.
3. Bolstein D, White RL, Skolnick M and Davis RW. Construction of a genetic linkage map in man using restriction fragment length polymorphisms. Am J Hum Genet. 1980, 32: 314-331.
4. White R and Lalovel JM. Chromosome mapping with DNA markers. Sci Am. 1988; 258(2): 20-28.
5. Emery AEH. Elements of medical genetics, 6th ed. Edinburgh. Churchill Livingstone 1983; page 215.

A TRIBUTE TO EMMIE RUSSELL



There are now few working members of the Orthoptic Association of Australia who had the privilege of knowing and working with Emmie Russell, who was one of Australia's first orthoptists. Born in 1892, she was educated at PLC Strathfield, and during the First World War was a voluntary worker for the Red Cross. She was a close family friend of Dr J Ringland Anderson, an ophthalmologist with a strong interest in strabismus, who was also a pioneer in the development of orthoptics in Australia. When, in 1932, he started a training program for orthoptists at the Alfred Hospital in Melbourne, based on the British syllabus, Emmie Russell (NSW), along with Ethel Southey and Margaret Fox (Victoria) Lucy Willoughby (South Australia) and Lena Gilchrist (Tasmania) were his first students. She registered with the Orthoptic Council of NSW when it was initiated in 1938, and, in 1944 when the Orthoptic Board of Australia was formed, she was its first registrant.

On completion of her training, Emmie Russell (always known as Miss Russell to her colleagues) started a professional career marked by the highest ethical standards that was to continue up to, and beyond her retirement in 1956. She established her own private practice at the BMA building in Macquarie St in January 1933, and started the Orthoptic Clinic at the Royal Alexandra Hospital for Children a few months later. During the second world war she, and other orthoptists gave voluntary service to the RAAF in training stereoscopic vision for their pilots. Throughout her career, she had à long working relationship with Sir Norman Gregg, the ophthalmologist who recognised and reported on the relationship between maternal rubella and congenital abnormalities.

At the same time she was a crucial figure in developing orthoptics as a profession in Australia. She founded the NSW Orthoptic Association and, soon after, with Diana Mann (Craig), also an early student of Dr Ringland Anderson, she founded the Orthoptic Association of Australia in 1944. As an active member of the Association she was its first President from 1945-47, as well as from 1950-51, and was a member of Council from 1945 to 1953. After her retirement she was made an Honorary member in 1959, and, in 1961 became the first orthoptist to be appointed Patron of the Association. Her strong interest in developing the profession took a tangible form in 1957, when she donated a sum of money to the Association to encourage younger members to undertake their own research in orthoptics. The Emmie Russell Prize, for the best paper presented at an Annual Scientific Meeting by a member who has graduated in the previous five years still remains the most prestigious award for our newer members.

These facts, although outlining a dedication to her profession, only begin to reveal her special personal qualities. All who knew her speak fondly of her intelligence, charm, love of the arts, gentle sense of humour and dedication to her family. In adversity, such as the time when, in 1965, her mother, Sir Norman Gregg and Thea Proctor (a close relative) all died within a short space of time, she maintained her quiet dignity

and concern for others. It is obvious that she was loved by those who had the privilege of knowing her. Children in particular loved her, and in her latter years she was frequently visited by the (now adult) children of her friends in whose lives she still maintained an active interest.

Right up until her death in 1987, at the age of 95, she remained alert and interested in others. Even those who knew her only in the last years of her life recognised in her a very special person. Orthoptics lost not only a pioneer of our profession, but also a truly exceptional woman. A legacy from her estate to the orthoptic clinic at The Royal Alexandra Hospital for Children (shortly to be renamed The Emmie Russell Department of Orthoptics) will be invested to promote the development of orthoptics and young orthoptists. It is appropriate that her name will live on for future members through awards which represent excellence and a commitment to orthoptics.

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CUMULATIVE INDEX: VOLUMES 21-26 (1984-1990)

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Begin with a title page giving a title which should be concise, followed by author(s) name, degrees or qualifications, name of place or institution where work was conducted and an address for communication.

On a separate page give a brief abstract of no more than 150 words, giving specific facts, findings, conclusions or opinions. Key words (about 5) or short phrases to assist indexers in cross-indexing the article, should follow the abstract on the same sheet. Key words should not duplicate words in the title but should be mentioned in the abstract.

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* Refer Med J Aust 1982; Dec 11/25, or apply to the Editor for a copy.