Bifocals in children with Down syndrome (BiDS)

by

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Authors declaration

I hereby declare that I am the sole author of this thesis. This is a true copy of the thesis, including any required final revisions, as accepted by my examiners.

I understand that my thesis may be made electronically available to the public.

Abstract

Down syndrome (DS) is the most common genetic cause of mental challenge in individuals and is associated with many ocular disorders. One of these anomalies which is frequently present in this population is reduced accommodation and many studies have reported this. Accommodation is the ability of the crystalline lens in the eye to focus for objects at different distances. Prescribing bifocals could potentially help in correcting the resultant inaccurate focus, although this modality of treatment is not very commonly practiced. The impact of bifocals on reading and literacy skills (academic skills) as well as visual-perceptual skills in individuals with DS has not been studied previously.

The aim of this study was to investigate the impact of bifocals on the educational attainment of children and young adults with DS who have reduced accommodation and monitor their performance longitudinally. This is the first time that the impact of bifocal provision on the functional performance of children and young adults with DS has been studied. Also for the first time in children with DS, frequent measures of performance have been used to control for progression with time before and after bifocal prescription. A battery of tests comprising early literacy and visual-perceptual skills was administered before and after bifocal prescription. Accommodation and printing skills were also measured periodically. It was expected that the prescription of bifocals would help to improve near visual acuity and that the improved near acuity would result in educational achievements at school. Compliance with spectacle wear and school reports were also considered.

A longitudinal observational study design was utilized with each child acting as his/her own control. Fourteen children and teenagers aged 8-18 with DS were recruited and underwent a basic optometric exam including measurement of their accommodative ability and a cycloplegic

refraction. Seventy nine percent required a change in their spectacle prescription and were prescribed single vision (SV) lenses. One hundred percent had reduced accommodation both before and after new SV glasses were prescribed. Distance visual acuity did not significantly improve with SV lenses ($p>0.05$) but near visual acuity showed a significant improvement (p-=0.015) from 0.64±0.25 logMAR to 0.54±0.20 logMAR. A high prevalence of high refractive errors, including both hyperopia and myopia, was observed t andnear visual acuity even with a habitual correctionwas reduced compared to distance VA.

A full battery of reading and visual-perceptual tests was administered with SV lenses. Thereafter the participants were followed for 6 months and monthly subtests (probes) of literacy skills and printing tasks were administered. These “probes” acted as immediate indicators of the child’s performance with his/her correction and change in performance over this time period was monitored. Over the 6 months the participants showed no noteworthy progression in their literacy skills. The group of participants performed at an age-equivalent between 3-10 years. The quality of printing formation in this population has been studied for the first time and showed no significant change over time. It was observed that some aspects of visual-perceptual and early literacy skills could be measured in all the participants. Chronological age and receptive vocabulary were significantly correlated with visual motor integration and Word Identification.

Eighty five percent of the participants were prescribed bifocals with additions ranging from +1.00D to +3.50D at the 6th month after the provision of SV lenses. Post-bifocal measures of visual acuity, accommodation, visual-perceptual and early literacy skills were taken 1-2 weeks, and finally 5 months, after bifocal correction. Throughout the pre- and post-bifocal period, verbal compliance with spectacle wear was assessed through school and parental reports. The mean near logMAR VA improved with bifocals ($p=0.007$) compared to SV lenses.

Accommodative accuracy improved with bifocals (less accommodative lag) compared to SV lenses ($p=0.002$) but there was no change in the accommodation exerted through the distance portion of the lens compared to SV lenses ($p=0.423$).

There was a main effect of bifocals on sight words ($p=0.013$), Word Identification ($p=0.047$), and 2 out of 3 tests of visual perception ($p<0.05$). It was observed that bifocals have a positive impact on the children's visual and school performance and this was supported by reports of improved performance in school for nine out of eleven individuals who were prescribed bifocals. The children adapted to bifocals more readily than the SV glasses, wearing them for the majority of their waking time.

All the sessions of early literacy and visual-perceptual skills administered throughout the duration of the study were videotaped and were then analyzed by a naïve examiner. The time taken to perform each task was calculated and compared between the main single vision and bifocal visits. There was a significant decrease in the completion times on the test battery with bifocals for Word Identification ($p=0.0015$) and the Dolch sight words ($p=0.048$). All participants who completed the monthly probes took less time to complete the Dolch sight words ($p= 0.025$) and the number writing task ($p=0.001$) with bifocals. Similar results were not observed for the visual-perceptual tests.

Performance in the monthly probes was compared before and after bifocal prescription in terms of the average raw scores and time taken. The rate of improvement in performance with bifocals was calculated by plotting the test scores against time and determing the regression lines. There was an overall significant improvement in the monthly probe scores of Word Identification ($p=0.050$), Dolch sight words ($p=0.025$) and the number test ($p=0.023$) with bifocals. The rate of progression in scores increased with bifocals for the Word Identification ($p=0.008$). Evidence of improved and faster performance with bifocals on some literacy skills was seen. It was concluded that bifocals, which result in improved near focusing, help individuals with DS to maximize their educational potential. It is suggested that more children and teenagers with DS will benefit from bifocal prescription, as they were observed to improve near visual acuity and enable better focusing for near.

This thesis has provided a comprehensive analysis of the some tests of literacy, visual-perceptual and early printing skills before and after a bifocal prescription in a group of children and teenagers with Down syndrome. This is the first study to longitudinally monitor the educational impact of bifocals in a population with Down syndrome. Furthermore, the quality of printing formation in this population is a previously unstudied area and was studied longitudinally prior to and after bifocal intervention. The impact of bifocals on printing skills is also discussed. Another novel approach was that all the literacy, writing and visual-perceptual tasks sessions were videotaped to calculate the time taken to complete each task pre- and post-bifocals.

This thesis is an addition to the existing literature on bifocal prescription in Down syndrome populations. From the findings in this thesis, the following recommendations are made in order to improve the standard of clinical eye care in this population. Measurement of accommodation should be considered a routine test in the clinical ocular examination for young individuals with DS, now that it is known that many of them present with accommodative deficits. When accommodation is found to be reduced, prescription of bifocals is indicated and should also become the standard of care in this population.

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Dedication

To my mom, dad and sister.

Mohan , to you for always being my inspiration even when you are not with us.

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List of Abbreviations

AI	Arithmetic instruction
ANOVA	Analysis of variance
BCC	Binocular cross cylinder
BF	Bifocals
BiDS	Bifocals in Down syndrome Study
BVMI	Beery Visual Motor Integration
cm	Centimetre
D	Dioptre
DS	Down syndrome
Hsa21	Human Chromosome 21
IEP	Individual Educational Plan
IQ	Intelligence quotient
LI	Letter Identification
LS	Literacy skills
logMAR	Logarithm of Minimum Angle of Resolution
m	Metre
MEM	Monocular estimation method
NRA	Negative relative accommodation
PPVT	Peabody Picture Vocabulary Test
PRA	Positive relative accommodation
RI	Reading instruction
SV	Single vision
TA	Teaching assistant

TVPS	Test of Visual - Perceptual Skills
VA	Visual acuity
VEP	Visually evoked potential
WA	Word Attack
WI	Word Identification
WrI	Writing instruction
WRMT	Woodcock Reading Mastery Test

1 Introduction

1.1 Introduction to Down syndrome

Down syndrome (DS) was first described in 1866 by an English physician, John Langdon Down and hence was named after him. In one of the earliest descriptions of DS, Down described a group of individuals seen by him, as having particular physical manifestations accompanied by moderate mental challenge (Down. 1866). The genetic aspects of DS were largely described later in 1959 by Jerome Lejeune and his team and they attributed the condition to trisomy of the chromosome 21 (Hsa21) (Lejeune et al. 1959). DS is the most commonly identified cause of mental retardation, (Smith-Bindman et al. 2001) and is known to affect approximately 1 in 800 live births (or 12.5 per 10,000 live births). It has been postulated that the trisomy of a few proteins associated with the gene may be what influences learning in individuals with DS (Best et al. 2007, Best et al. 2008, Sago et al. 1998, Wiseman et al. 2009).

1.1.1 Diagnosis and testing for Down syndrome:

Down syndrome can be diagnosed by genetic testing although in many cases a newborn baby's physical features can give an initial indication that the child has Down syndrome. However some traits can be subtle in a newborn, depending on the type of Down syndrome that is present. In general, a preliminary diagnosis can often be made from observation of physical features although a confirmed diagnosis necessitates a genetic testing. Cytogenetics is a branch of genetics that is concerned with the study of the structure and function of the cell, especially the chromosomes. Profiles of the chromosomes, called karyotypes are made and observed as a part of the standard procedures in cytogenetics to study the structure and pattern of the chromosomes. The normal human karyotype contains 22 pairs of autosomal chromosomes and one pair of sex chromosomes. The karyotype for females contain 2 X chromosomes, denoted 46, XX, and males have both an X and a Y chromosome denoted 46, XY (Figure 1-1). Any variation from the standard karyotype may lead to developmental abnormalities. These karyotypes can be used to identify a genetic abnormality eg; Prader-Willi syndrome or Down syndrome. This involves applying an appropriate dye to stain the cell under observation, to understand the abnormal pattern by counting the number of chromosomes and looking at structural abnormalities.

Roizen and Patterson (2003) report that the current health care practice in the USA offers pregnancy screening by means of blood tests for chromosomal anomalies followed by cytogenetic testing if needed. Early testing of unborn babies helps to identify DS or similar conditions, preparing the families for the special needs of the child. According to Smith-Bindman (Smith-Bindman et al. 2001), 80-85% successful detection of the condition is possible by using the maternal serum and an ultrasound testing of the fetus. The incidence of DS depends on maternal age along with other factors such as health of the mother, previous history of abnormal pregnancies etc. According to the National Institutes of Health, “the chance of having a baby with Down syndrome increases as a woman gets older—from about 1 in 1,250 for a woman who gets pregnant at age 25, to about 1 in 100 for a woman who gets pregnant at age 40”. Thus pre-natal testing for the anomaly is recommended in mothers aged 35 years or more.

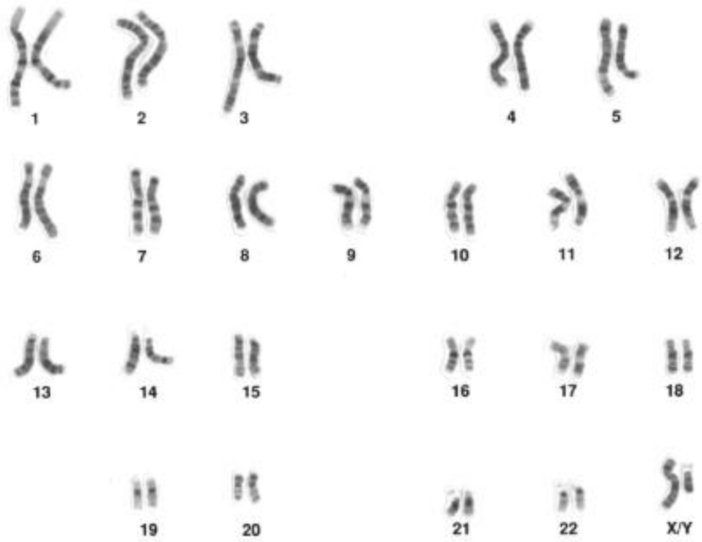


Figure 1-1: A normal male human karyotype *(Image courtesy: National Human Genome Research Institute)*

Based on the karyotype, there are three types of DS identified and these are described below.

1.1.2 Types of Down syndrome:

1.1.2.1 Trisomy 21:

This is the most common type of DS, occurring in almost 94% of this population. An anomaly in the cell division occurs, where a pair of the 21st chromosomes from either of the parents does not separate properly either before or at the time of conception and an extra chromosome 21 is then found in every cell in the body causing the characteristics of Down syndrome (Figure 2-2), hence the name Trisomy 21.

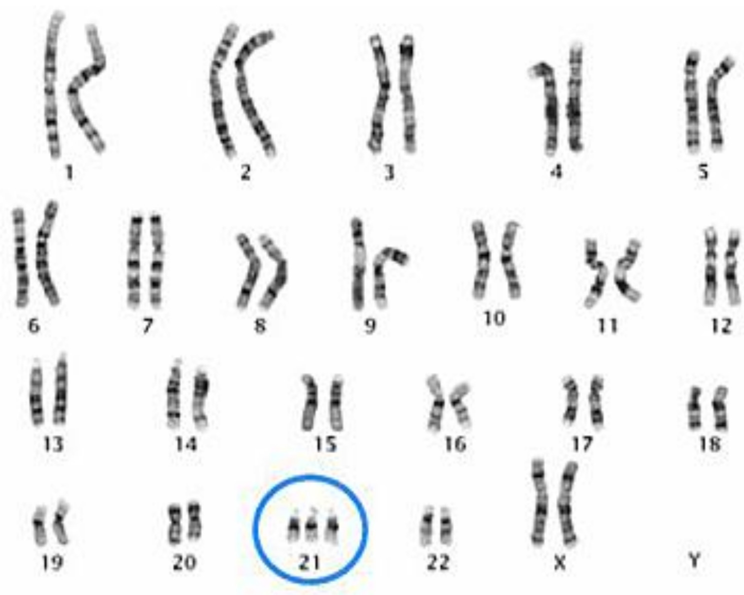


Figure 1-2: Karyotype of Trisomy 21 individual *(Image courtesy: National Human Genome Research Institute)*

1.1.2.2 Mosaicism:

In this type of Down syndrome, constituting about 2% of this population, the error in separation of the 21st chromosome occurs during the first few cell divisions after fertilization. Thus some cells of the individual have 46 and the others have 47 chromosomes, resulting in the name mosaicism. The physical attributes in this condition depend on the ratio of cells with 46 chromosomes to those with 47 chromosomes.

1.1.2.3 Translocation:

In this type of Down syndrome, seen in around 4% of this population, a part of chromosome number 21 breaks off and attaches itself to another chromosome. Mostly it is seen to attach to chromosome number 14. This causes all cells in the body to have the extra piece of the 21st chromosome. In this type of DS, it is usual that one of the parents is a carrier for the unusual chromosomal material, hence transferring it to the child.

1.1.3 Associated conditions in Down syndrome:

Individuals with DS present with typical physical features/characteristics which are listed below (Korenberg et al. 1994, Caputo et al. 1989, Catalano. 1992, da Cunha & Moreira. 1996, Down. 1866, Haugen et al. 2004, Pueschel & Gieswein.1993 , Pueschel. 1981, Shapiro & France. 1985).

- Short stature
- Microcephaly (smaller circumference of the head)
- Brachycephaly (disproportionate or sometimes flattened head)
- Flat nasal bridge
- Vaulted palate
- Furrowed tongue
- Tendency to open mouth
- Malpositioned ears
- Small/dysmorphic ears
- Short neck
- Duodenal stenosis (a defect where a portion of the small intestine is narrowed which prevents the stomach contents from flowing through at a normal rate)
- Broad short hands
- Brachydactyly (shorter fingers and toes)
- Clinodactyly of the 5th finger (bend or curved 5th finger towards the 4th finger)

- Wide gap between toes 1 & 2
- Abnormal dermatoglyphics (finger prints)
- Palmar crease (single crease in the palm)
- Hypotonia (reduced muscle tone)
- Lax ligaments (loose ligaments)

Individuals with DS are at a greater risk of the following associated physiological anomalies in comparison to typically developing population:

- Congenital heart disease
- Moderate mental challenge
- Leukemia
- Susceptibility to infectious diseases particularly pneumonia
- Dementia
- Ear abnormalities and decreased hearing (70-90%)
- Thyroid abnormalities
- Gastrointestinal problems
- Skin problems, very commonly atopic dermatitis (eczema) and folliculitis
- Alzheimer's disease over 60 years of age

Ocular disorders in individuals with DS have been reported to increase in frequency with age. (Roizen et al. 1994) The following ocular conditions are more commonly seen in individuals with DS than in typically developing populations and the range of percent prevalence between studies (Caputo et al. 1989, Cregg et al. 2001, Cullen & Butler. 1963, da Cunha & Moreira J. 1996, Kim et al. 2008, Liza-Sharmini et al. 2006, Lowe. 1949, Paudel et al. 2010, Shapiro &

France. 1985, Tsiaras et al. 1999, van Splunder et al. 2003, Woodhouse et al. 1993) is given in brackets (%).

- Up-slanted palpebral fissures (63%-89%)
- Congenital glaucoma (1-7%)
- Epicanthal folds (24%-96.7%)
- Brushfield spots (36-81%)
- Blepharitis (10%-47%)
- Hypoplasia of the iris (42%-95%)
- Strabismus (27-57%), esotropia being more common than other types of strabismus
- Nasolacrimal duct obstruction (3.3%-30%)
- Nystagmus (20-33.3%)
- High refractive errors (35-76%)
- Reduced visual acuity (24-46%)
- Reduced accommodation (55%-80%)

The last three disorders will be discussed in more detail later, but a general background to accommodative function will be given first.

1.2 Accommodation

Accommodation is defined as the process by which the optical system of the eye varies its focal length in response to a visual stimulus. The process of accommodation primarily involves the ciliary muscle, the crystalline lens and zonules as shown in figure 1-3. When viewing a distant object, the ciliary muscle relaxes, increasing the tension on the zonules in order to flatten the lens and reduce its thickness (figure 1-4a). This is the unaccommodated state of the eye. The farthest point at which the eye can maintain a clear retinal image is called the ‘far point of the eye’. The ciliary muscle fibers run tangentially and when accommodating for a near object, the contraction of the ciliary muscle releases the tension on the zonular fibers, which move forward and inward, thereby causing the lens to assume its natural shape and become more spherical (Figure 1-4b). This increase in the curvature of the lens results in an increase in the refractive power of the lens and of the whole eye (Helmholtz theory or classical theory (1962), cited in Garner) (Garner. 1983). Both the anterior and posterior curvatures of the lens increase, but there is a greater change in the anterior surface. Along with these changes, there is an increase in the thickness of the lens, a decrease in its equatorial diameter and a reduction in pupil size (Brown. 1973). The ‘near point’ is described as the object distance for which a clear retinal image is achieved when maximum accommodation is exerted.

The accommodative response is measured in dioptres (D) and is the reciprocal of the distance at which the emmetropic eye is focused (an emmetropic eye is one with a refractive state such that, when unaccommodated, parallel incident rays of light are brought to focus on the retina). Thus if an emmetropic eye is focused at a distance of 1 m, the accommodation is said to be 1 D; if it is 0.5 m or 0.33 m, the accommodation is 2 D or 3 D, respectively.

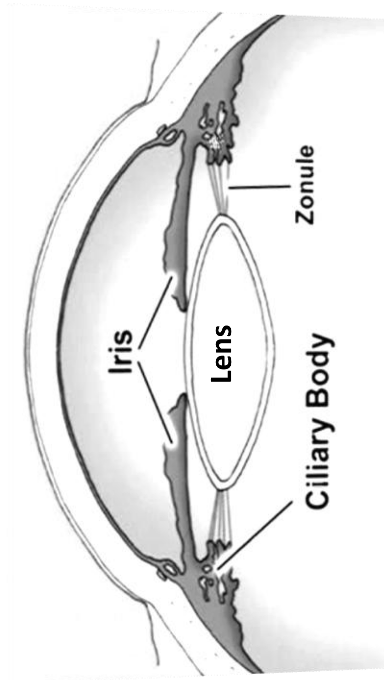
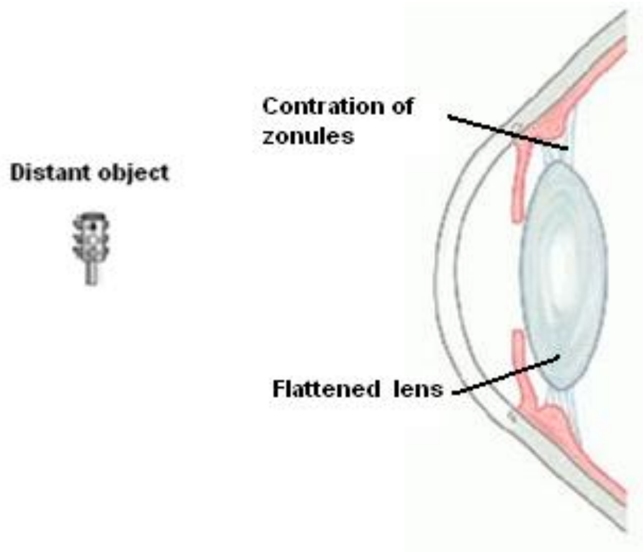
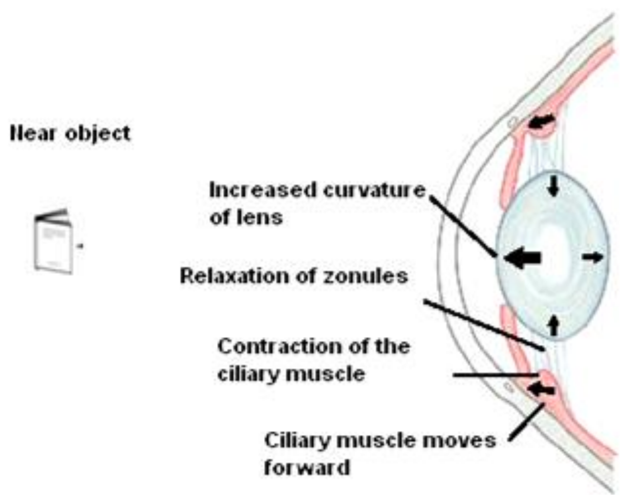


Figure 1-3: The accommodative apparatus (*Image adapted and modified from Fisiologia/neurofisiologia/Clayman91c.jpg)*



(a) **Viewing a distant target** (Unaccommodated state of an emmetropic eye)



(b) **Viewing a near target** (Accommodating eye)

Figure 1-4: Unaccommodated (a) and accommodated eye (b) *(adapted from a248.e.akamai.net/.../accommodation(1).jpg)*

1.2.1 Accommodative stimulus-response:

The stimulus to accommodation can be defined as the accommodation demand required to focus the image of a particular object on the retina, and, for an emmetropic eye, is given by the dioptric distance of the object in metres. Accommodative response is the actual amount of accommodation exerted by the eye.

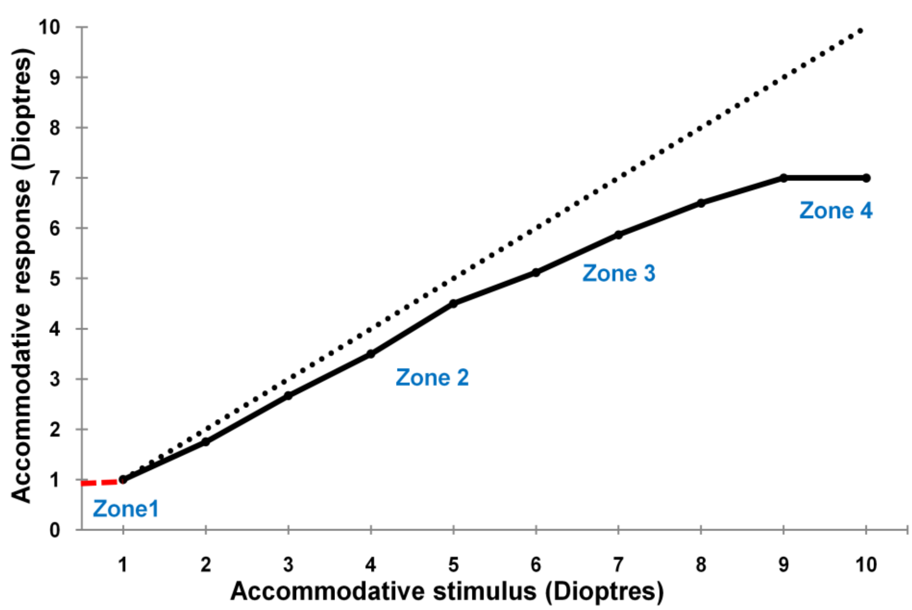


Figure 1-5: Accommodative stimulus response curve *(adapted from values in Cuiffreda and Kenyon, 1983).* The solid line is the typical response curve from empirical measures of the young human eye and the dotted line indicates a perfect relationship between stimulus and response (1:1)

The accommodative response is related to the distance of the stimulus. This is described by the accommodative stimulus-response curve, as seen in figure 1-5. The solid line shows the normal stimulus-accommodative response for a young individual. It can be divided into four zones (Ciuffreda & Kenyon. 1983):

Zone 1 (0-1D approximately) shows that there is a ‘lead’ accommodative response for the given stimulus i.e. an over-accommodation. Through this zone, the response is almost constant and is induced by the tonicity of the ciliary muscle, occurring for lower stimulus demands.

Zone 2 shows a lower response than the stimulus, thereby producing a ‘lag’ of accommodative response, at these intermediate stimulus levels.

Zone 3 shows that with further increase in the stimulus, there is a change in the accommodative response, but this response is progressively smaller than for a similar change in stimulus compared to Zone 2.

Zone 4 describes the region of saturation i.e., any further increase in the stimulus does not produce an increased response. This zone also defines the ‘amplitude of accommodation’, the maximum accommodation that can be produced for any given stimulus.

1.2.2 Components of accommodation:

There has been considerable debate regarding what are the physiological components of the stimulus to accommodation for an eye and it is conceded that ‘blur’ is the primary stimulus for accommodation (Fincham. 1951, Ciuffreda. 1991, Heath. 1956, Phillips & Stark. 1977). There are a number of different, other cues to accommodation which stimulate accommodation to varying degrees (Gray et al. 1993, Kruger & Pola. 1985, Kruger et al. 1997, Mathews & Kruger. 1994, McLin et al. 1988, Rosenfield & Cohen. 1995, Siderov & Johnston. 1990, Sivak & Bobier. 1978).

These include:

- Proximity of the target
- Changing target size
- Chromatic aberration
- Convergence of the eyes

According to Heath (Heath. 1956), accommodation response can be divided into four functional or operational components:

Reflex accommodation: This can be described as an automatic adjustment of the refractive status in order to maintain a sharply focused retinal image in response to a blurred input. According to Fincham (Fincham. 1951) reflex accommodation occurs for smaller amounts of blur, up to 2.00D. Reflex accommodation is the largest and most important component of accommodation under both monocular and binocular conditions (Hung et al. 1996).

Vergence accommodation: Vergence accommodation can be described as the component of accommodation induced by the binocular disparity of the retinal images and the resultant convergence movement of the eyes. This can be demonstrated by presenting a target free of blur (e.g, using binocular pinholes or a blur-free difference of Gaussian [DoG] target with a lower central spatial frequency) in order to negate the possible reflex accommodation. Vergence accommodation is the second major component of accommodation (Ciuffreda. 1998).

Proximal accommodation: This component of accommodation is initiated by an awareness of the nearness of an object. According to Rosenfield (Rosenfield et al. 1991), proximal accommodation is stimulated by objects present 3m or less from an individual.

Tonic accommodation: This is referred to as the resting state of accommodation in the dark. In other words, tonic accommodation is present in the absence of blur, disparity and proximal cues and is measurable by removing all these other inputs. It could also be defined as the equilibrium state of the accommodative system. Under such conditions, the mean tonic accommodation value in an individual is about 1D (ranging from 0-4.4D). (Maddock et al. 1981, Robert et al. 1984).

Neural innervations due to the aforesaid components of accommodation, either individually or in unison, act to drive the accommodative response.

1.2.3 Neural pathways of accommodation:

Accommodation is evoked by the sensory system and starts with the stimulation of the retinal photoreceptor cells, by means of a defocused retinal image (blur) as shown in the flowchart Figure 1-6. The blur signals pass through the visual pathway and are transmitted to Area 17 of the visual cortex and then to the parieto-temporal regions for further processing. The signal is then transmitted to the midbrain-oculomotor nucleus complex where it is transformed into a motor command at the Edinger-Westphal nucleus. The motor command in transmitted by the efferent pathway via the oculomotor nerve, the ciliary ganglion and the short ciliary nerves to the ciliary muscle. The ciliary muscle is mainly innervated by the parasympathetic nerve fibers although there are some innervations from the sympathetic system. The origin of the parasympathetic pathway is at the Edinger-Westphal nucleus and it follows the course of the oculomotor nerve and synapses at the ciliary ganglion. The efferent pathway ends at the ciliary muscle where a change in the state of contraction alters the refractive power of the crystalline lens and thus attains a focused image on the retina.

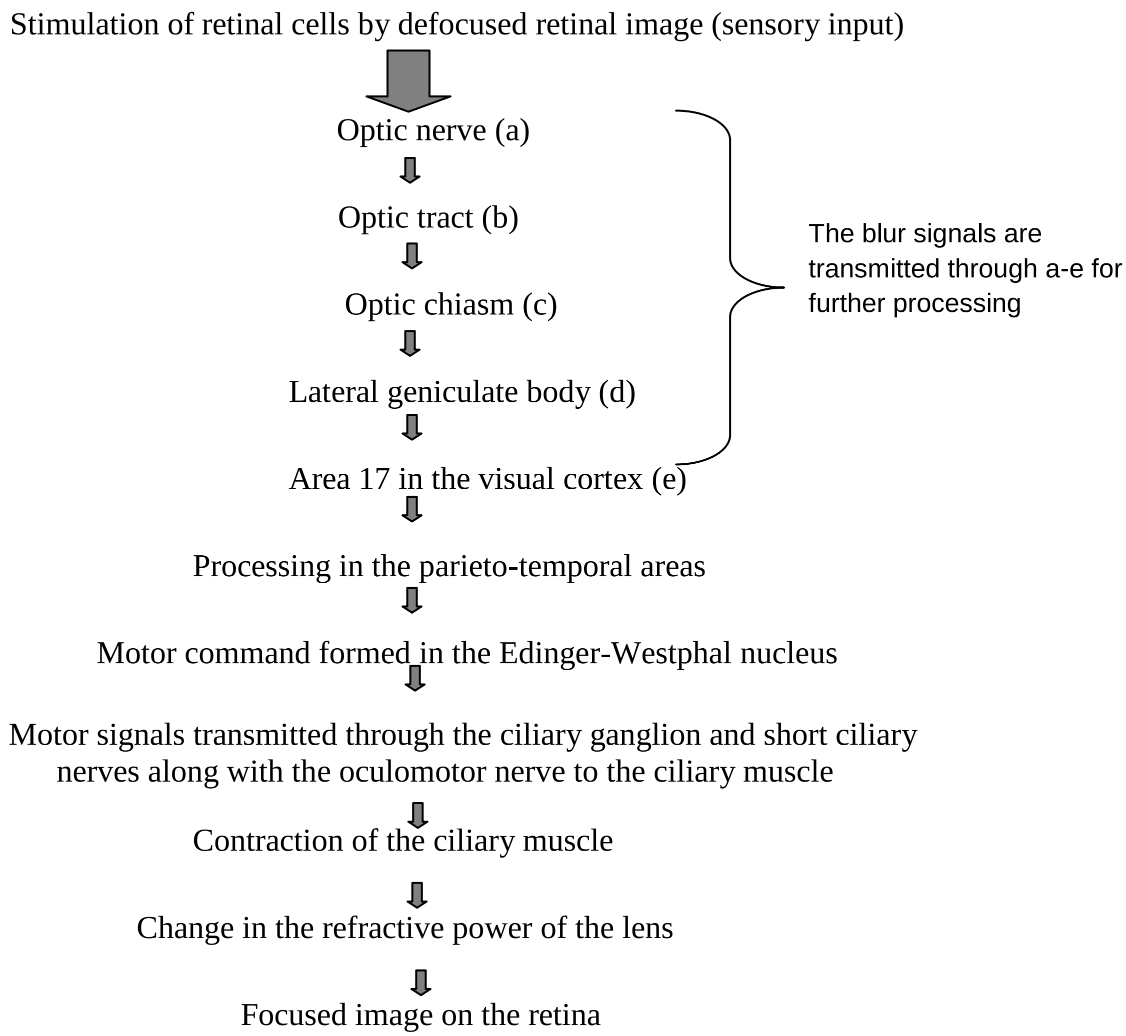


Figure 1-6: Flow chart of sensory and motor pathway for blur-driven accommodation

(adapted from (Borish et al. 1998)***)***

1.2.4 Clinical measures of accommodation:

There are different aspects of the accommodative response that can be measured clinically and these are measured by different methods. The best measurement technique in any situation depends in part on how the accommodative response is being manipulated. e.g., changing the stimulus distance or changing the stimulus characteristics or introducing lenses. Both subjective and objective tests exist to determine the accommodative function.

The subjective tests described briefly here include:

1.2.4.1 Accommodative Amplitude tests

Donders push-up method (Grosvenor. 1996)

With the patient's best distance refractive correction in place , the near point card is placed at 40 cm from the patient and is adequately illuminated. The patient's attention is drawn to the 6/12 row of letters and he/she is instructed to indicate the 'first sustained blur' as the examiner slowly moves the card towards the patient. The distance at which the first sustained blur is reported is noted from the accommodation rule (or with a tape measure in the absence of a rod) and the dioptric equivalent gives the amplitude of accommodation.

Minus lens method (Grosvenor. 1996)

In this method, the reading card is placed at 40 cm with patient wearing his/her best distance refractive correction and looking at the 6/12 on the near point card placed at 40 cms from the patient. The patient is instructed to report when the letters in the line first start to blur as negative lenses are introduced monocularly (with the other eye occluded). The lens which produces the first sustained blur is noted. For example, if -2.75D did not produce blur, but a –

3.00D lens did produce blur, the amplitude of accommodation is calculated by 3+ 2.50 (for the 40 cm working distance) = 5.50D.

1.2.4.2 Accommodative facility tests

Plus/minus flippers (Elliott. 2003)

This test involves measuring the speed (facility) of changes of accommodation by introducing positive and negative lenses in the form of flippers (usually ± 2.00D) to decrease or increase the accommodation stimulus when looking at a near point card at 40cm. The patient is instructed to flip over from one lens pair (e.g. +2.00D) to the other (-2.00D) every time the letters clear and the number of cycles (flip from one lens to the other) per minute (cpm) gives a measure of the accommodative facility.

1.2.4.3 Relative accommodation

Negative Relative Accommodation (NRA) (Theodore Grosvenor. 1996)

NRA gives a measure of the maximum ability to relax the accommodation while maintaining clear, single binocular vision. The patient's attention is drawn to the 6/12 row of letters on the near point card. As plus lenses, 0.25D at a time are added binocularly, the patient is instructed to report the first sustained blur. The lens with which the first sustained blur is reported gives the value of NRA. Since the NRA is a relaxing test (as plus lenses are introduced), it is always done before the PRA in practice.

Positive Relative accommodation (PRA)

Similarly PRA measures the maximum ability to increase accommodation while maintaining clear, single binocular vision. This test is the same as the NRA except that negative lenses are used instead of positive lenses.

1.2.4.4 Measures of accommodation accuracy

Binocular cross cylinder (BCC) at near distance (Rosenfield & Logan. 2009)

This is a subjective measure of the accommodative response at 40 cm and is a helpful measure in addition or as an alternative to dynamic retinoscopy (described below) to estimate the lag of accommodation (Theodore Grosvenor. 1996). In this test, a pattern of horizontal and vertical intersecting lines is used as a target and is viewed by the patient through a cross-cylinder lens (usually ±0.50D) such that the minus axes are at 90º. The patient is then asked to report if the horizontal and/or vertical lines are equally clear/blurred. If the patient is underaccommodating (lag of accommodation) before the introduction of the cross-cylinder, they would ideally report that the horizontal lines are clearer and vice versa (vertical lines clear) if over accommodating. In case of a lag, plus lenses are added (in cases of lead, negative lenses are added) until both the sets of lines appear equally clear. The power of the lens that gives rise to the equally clear lines gives the measure of the error of accommodation - lag or lead.

The only objective clinical test for accommodation is dynamic retinoscopy:

Dynamic retinoscopy

The most common method for measuring accommodation in a clinical setting is by the push-up method which is described previously which determines the subjective amplitude of accommodation. This method, however, cannot be used with many non-verbal patients and with very young children. Dynamic retinoscopy, the most widely practiced objective clinical measure of accommodation, has been shown to be a useful measure of accommodative response in non-communicative individuals (Leat. 1996, Woodhouse et al. 1993). In this technique, the subject/patient with his/her best distance correction is asked to look at a detailed target and retinoscopy is performed This is used clinically to establish the accuracy of the accommodative response, e.g., the degree of lag or lead of accommodation (Leat & Gargon. 1996). There are several versions of dynamic retinoscopy and there are three different techniques that have been developed for young children, Nott, MEM, and Bell retinoscopy and these are described in detail in the next section.

Nott dynamic retinoscopy

Nott retinoscopy is a version of dynamic retinoscopy in which the subject binocularly views a near point test card and the examiner shines the retinoscope light through a hole in the card (Nott. 1925). The observer moves with the retinoscope in order to find the neutral point. The advantage of this method is that trial lenses need not be introduced and hence this reduces the distraction that could occur in young children by the lenses.

Woodhouse et al. (Woodhouse et al. 1993) made a modification to Nott retinoscopy by which they were also able to assess leads of accommodation. The arrangement of the typical Nott retinoscopy made it difficult to move closer in order to measure any lead of accommodation. Hence in the modification by Woodhouse et al., the stimulus was arranged such that both leads and lags could be measured. Also, the stimulus used was made more interesting for children as it was comprised of pictures. The target was a white box, internally illuminated to gain more attention. There was a choice of pictures to maintain attention, and these had both coarse and fine detail in order to provide an adequate stimulus for eyes with a range of visual acuity. This modified Nott retinoscopy (Figure 1-7) has been used in many studies in children with special needs in order to measure their accommodative responses (Woodhouse et al. 1993,Woodhouse et al. 2000, Leat. 1996). Using the modified Nott method, consistent under-accommodation at all tested distances was seen in many children and young adults with DS (Woodhouse et al. 2000). This method has also been used in children with cerebral palsy, with 42% of the children showing an accommodative response which was reduced compared to the normal control group for their age (Leat. 1996).

Figure 1-7: Modified Nott dynamic retinoscopy

Bell retinoscopy

Bell retinoscopy is another form of dynamic retinoscopy used for assessing accommodation in young children and is described by Apell (Apell. 1975). Originally this technique was performed by dangling a small bell in front of the examiner's forehead but this was later replaced by a 0.5 inch chrome steel ball attached to a thin metal rod instead of a bell. The examiner is positioned with the retinoscope in one hand at 50 meters from the patient's face and the examiner holds the steel ball suspended at the patient's eye level in the other. Similar to the Nott method, trial lenses are not used in this procedure. The patient is instructed to look at his/her reflection on the ball, while the examiner performs retinoscopy to observe the direction of the motion of the reflex. The ball is moved closer to the patient's face until a neutral reflex is observed in each principal meridian. The distance between the patient and the position of the bell (when neutrality is observed) gives a measure of the endpoint. According to Apell (Apell. 1975), the neutrality using this technique is usually observed when the ball is located at about 37-40 cm from the patient resulting in a lag of 0.5-0.75D.

Monocular Estimation Method

The monocular estimation method (MEM) as described by Bieber (Bieber. 1974) utilizes a white reading card containing a 0.5 inch hole with letters, numbers or pictures appropriate to the patient's age level printed around the hole. The card is attached by means of a clip to the retinoscope such that the retinoscope beam can pass through the hole. The examiner is positioned slightly below the patient's eye level in order for the patient to have a moderate downward gaze as would occur while reading. The patient is asked to read the letters/numbers or describe the pictures aloud. The estimation of lag or lead of accommodation is determined with the brief introduction of minus or plus lenses (minus lens for an “against” reflex indicating a lead of accommodation and plus lens for a “with” reflex indicating a lag of accommodation) so as to not influence the accommodation.The spherical lens that produces neutrality is noted. It is called the monocular estimation method as the accommodation is determined monocularly when the patient is viewing the target binocularly. The expected range is between +0.25 to +0.75 (Saladin.1998).

1.2.4.5 Validity and repeatability of Dynamic retinoscopy:

A few studies have compared these different dynamic retinoscopy methods. Garcia and Cacho (Garcia & Cacho. 2002) studied the accommodative response with MEM and Nott retinoscopy. Their results showed that there was a statistically significant difference between the techniques, with MEM showing greater response values than Nott retinoscopy. There was a high correlation between the results with the two techniques ($r=0.90$) and a co-efficient of agreement between the 2 techniques of ±0.53 D which the authors concluded is clinically significant and hence the two techniques are not interchangeable. Locke and Somers (Locke & Somers. 1989) compared values obtained by two experienced examiners using all three dynamic retinoscopy techniques described in the earlier section along with the BCC. The accommodative lags of 10 young adult subjects measured using these techniques showed no significant differences between the results of the two examiners ($p=0.267$). Results obtained by the MEM, Cross (target similar to MEM at 40 cm but plus lenses are added binocularly to determine neutrality), and Nott techniques were not significantly different, but those obtained by Bell retinoscopy and BCC were significantly different from the other three techniques. The results of this study suggested that an examiner may use MEM, Cross retinoscopy, or Nott retinoscopy interchangeably to evaluate accommodative lag of the young adult subject. The Garcia and Chacko study (Garcia & Cacho. 2002) concluded that there was lack of agreement between the 2 techniques (MEM and Nott).

Nevertheless in comparison with a later report on repeatability of distance retinoscopy (Smith. 2006), the limits of accuracy of retinoscopy measurement were between ± 0.35 and 0.76D. This would mean that the former study (Garcia & Chacko, 2002) in fact indicates good agreement between the 2 measures of accommodative response. Therefore, from the 2 earlier studies on Nott retinoscopy, it appears that Nott retinoscopy is in agreement with the MEM.

McClelland and Saunders (McClelland & Saunders. 2003) undertook a study to examine the repeatability and validity of modified Nott dynamic retinoscopy compared with the Shin-Nippon SRW 5000 autorefractor at 4, 6 and 10 D distances. Their results showed no significant difference between two measures of dynamic retinoscopy at any distance (paired t-test, $p>0.1$) and the co-efficient of repeatability for the dynamic retinoscopy was

±1.34 D for the 10 D stimulus, ±1.09 D for the 6 D stimulus and ±0.56 D for the 4 D stimulus. Similary there was no significant difference between Nott retinsoscopy and the auto-refractor at any distance ($p>0.1$). The coefficient of agreement suggested that accommodative responses measured with the dynamic retinoscopy technique could be expected to be within ±1.58 D of those obtained with the autorefractor at a stimulus distance of 10 cm and within ±1.16 D for stimulius distance of 16.7 cm. A similar finding was not seen at the 4D distance as the autorefractor gave higher measures than the dynamic retinoscopy method as the accommodative responses increased and this was attributed to the presence of 2 outlying points. Therefore a co-efficient of agreement was not calculated for this stimulus level. However overall it was seen that that there was no significant difference between the mean results obtained with the two methods (paired t-test, $p > 0.1$) and they concluded that the dynamic retinoscopy technique is valid. In another study, dynamic retinoscopy (Nott method), binocular cross-cylinder (with and without fogging lenses) and a near red-green duo-chrome test for determining the accommodative response were compared with the findings of an infrared autorefractor (Rosenfield et al. 1996). The results show that, under binocular conditions, the mean accommodative responses for all the tests were clinically equivalent. The Nott dynamic retinoscopy showed the closest agreement with the autorefractor, whereas the two dynamic cross-cylinder procedures (with and without fogging) exhibited the greatest variability in findings compared with the autorefractor. Leat and Mohr (Leat & Mohr. 2007) also show good inter-observer repeatability for the modified Nott dynamic retinoscopy method (co-efficient of repeatability = 0.637 D overall with a mean difference of 0.008D between observers). The co-efficients of repeatability at 4, 6 8 and 10D were 0.372, 0.667, 0.708 and 0.764D respectively showing that the modified dynamic retinoscopy technique is repeatable, particularly at the lower stimulus demands. In addition, they report no significant difference in the measurements depending on the order of measurement i.e. increasing or decreasing accommodation demand (repeated measures ANOVA, $p = 0.15$) and no effect of observer ($p = 0.67$).

These studies show that Nott dynamic retinoscopy is a repeatable and valid objective technique for measurement of accommodative response when other conventional methods cannot be used. The repeatability and validity of dynamic retinoscopy makes it a suitable technique to be used in populations with special needs and it is very easily applicable in a clinical setting. In addition there have been several sets of data of age-related norms age for the Nott retinoscopy technique and not as many for the other techniques. Hence Nott retinoscopy has been chosen to measure accommodative responses in the current study with the modifications discussed earlier.

1.2.4.6 Other measurement methods

Laboratory studies have used instruments such as autorefractors and photorefractors and their modified versions, for measurements of refractive error and accommodative response. In particular, photorefactors, owing to their photographic basis, rapid measuring time and more remote working distance, have been particularly useful in refracting children in research settings (Howland. 1985). Ocular biometry is a technique that measures the changes in the accommodative components (anterior chamber depth, lens thickness, refraction) during accommodation. This is used to understand the anterior segment changes which occur during accommodation.

1.2.5 Accommodative response with age:

In young adults a clear retinal image is achieved for a wide range of viewing distances by the process of accommodation (Currie & Manny. 1997). There is an increasing volume of literature on when the normally developing human accommodation system responds in an adult-like fashion. One of the early studies describing this development showed that accommodation was adult-like between 2-4 months (Haynes et al. 1965). Although there are some differences between the later reports, overall it has been established that the normal accommodative system responds fairly accurately at least by about 2-3 months of age (Banks. 1980, Bobier. 1990, Braddick et al. 1979, Brookman. 1983, Howland et al. 1987). The majority of studies involving measurement of accommodation in infants have used binocular measurements. Most recently, Bharadwaj and Candy (2008) have shown that binocular accommodation is adult-like at 2 months of age but monocular accommodation is not adult-like until 8 years.

There are not many studies describing the accommodative function between the ages of 1-4.5 years. This is likely because of the difficulty posed by this age group in assessing the responses either due to inattention or lack of understanding of the procedure. Leat and Gargon (1996) and McClelland and Saunders (2004) studied accommodative responses in children including 3 and 4 year olds respectively. Leat and Gargon (1996) describe accommodative responses in children and adults ranging from 3-35 years old. They showed that the 3-10 year olds had accommodative accuracy within 0.50D over the stimuli range used. They also add that the 3-5 year olds showed slight over accommodation, the 6-10 year olds had accurate accommodation, while the 11-26 year olds under accommodated. It was also noticed that individuals greater than 10 years old showed more under accommodation with increasing accommodative demand. Figure 1-8 shows dynamic retinoscopy values (norms) from Leat and Gargon (1996). In the two older age groups, there is increasing lag with increasing age.

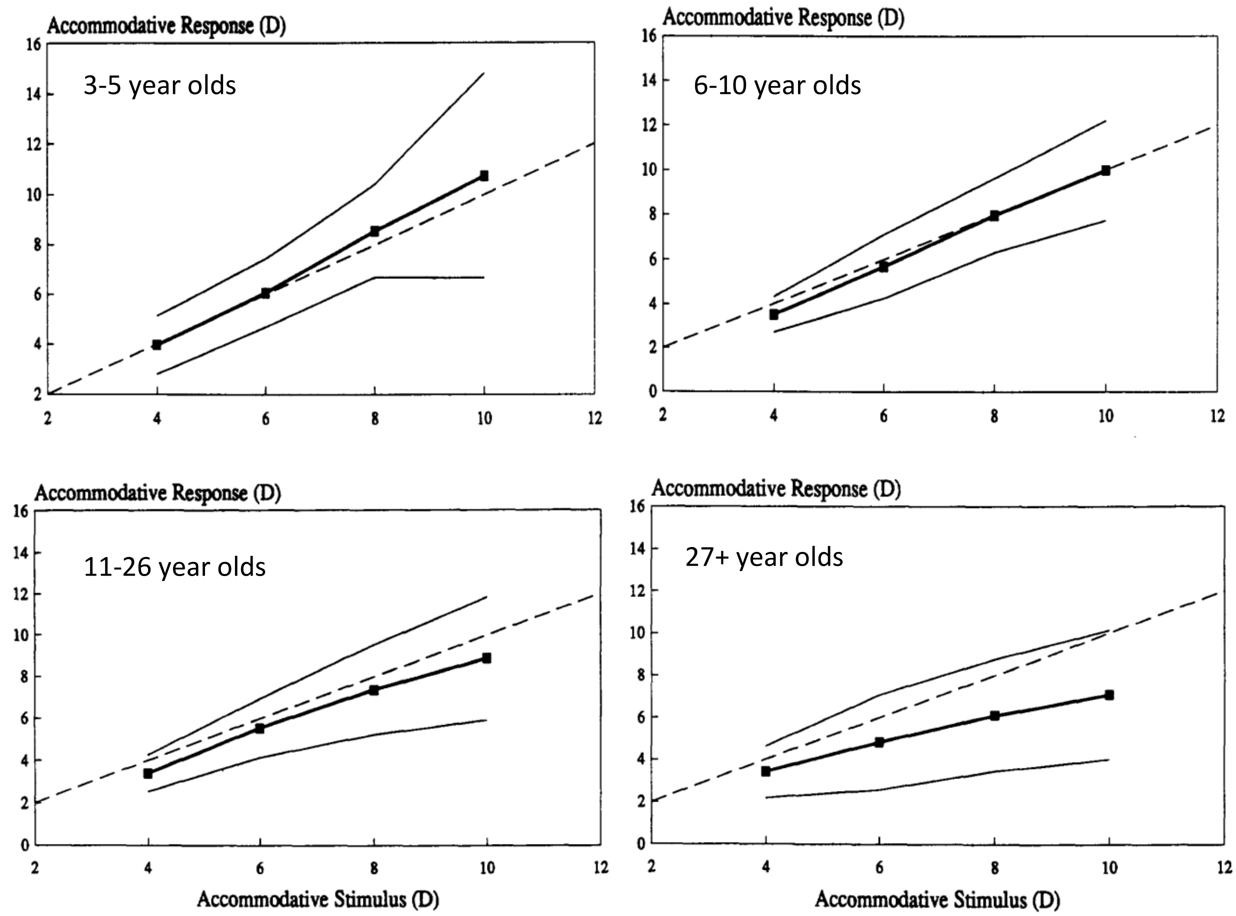


Figure 1-8: Mean accommodative responses in normally sighted children and young adults
Reprinted from Ophthalmic and Physiological Optics. Leat SJ & Gargon JL. Accommodative response in children and young adults using dynamic retinoscopy (1996): *Ophthalmic Physiol Opt*,16: 375-384,1996; with permission from Wiley-Blackwell publishing. The thin solid lines show ± 1.96 x standard deviation (95% confidence range). The dotted line shows the perfect accommodative response. The dark black solid line shows the actual accommodative response obtained in the group.

Table 1-1 Mean accommodative response and 95% confidence limits (mean ± 1.96 × SD) of accommodative response for each age group at each accommodative demand

Age (yrs)	n	Accommodative Response (D)					
		4 D Demand		6 D Demand		10 D Demand	
		Mean (D)	Normal Range (D)	Mean (D)	Normal Range (D)	Mean (D)	Normal Range (D)
4	11	3.70	2.52–4.88	5.57	4.28–6.86	7.54	3.82–11.26
5	10	3.86	3.37–4.35	5.32	4.48–6.14	7.67	5.36–9.98
6	17	3.73	2.75–4.71	5.22	3.98–6.44	7.85	5.01–10.69
7	10	3.77	3.21–4.31	5.65	4.77–6.53	8.12	6.06–10.18
8	10	3.73	3.24–4.22	5.36	3.97–6.75	7.36	4.42–10.30
9	9	3.73	3.08–4.38	5.06	4.29–5.81	7.09	4.39–9.79
10	7	3.42	2.87–3.97	4.82	4.01–5.62	6.92	5.63–8.21
11	9	3.71	3.02–4.39	5.30	4.10–6.50	7.71	6.09–9.33
12	12	3.79	3.07–4.51	5.29	4.31–6.25	7.17	4.94–9.40
13	11	3.75	3.04–4.46	5.28	4.10–6.46	6.98	5.59–8.37
14	9	3.49	2.78–4.20	4.82	3.80–5.84	7.25	4.82–9.68
15	10	3.60	2.66–4.54	5.19	3.99–6.39	8.10	5.65–10.55

Reprinted from Optometry and Vision Science with permission from Wolters Kluwer Health. McClelland JF& Saunders KJ. Accommodative lag using dynamic retinoscopy: Age norms for school-age children. *Optom Vis Sci*.2004; 81:929–933.

Similarly, McClelland and Saunders (2004) describe accommodative responses in 4-15 year olds using the same modified Nott dynamic retinoscopy method and their results are shown in Table 1-1. They report that the widest accommodation ranges were observed in the 4 year olds and discuss that there is a possibility of attention or concentration difficulty in this youngest group which might have caused the differences. The mean lag at the 4D in the 4-year olds was 0.3 D which increased to 2.46D at the 10D demand. In addition the lag at 4D also increased from 0.30 in the 4 year olds to 0.40 D in the 15 year old groups. Leat and Gargon (1996) showed that at the 4D demand, the lag increased from 0 in the 3-5 year olds, to 0.48 ± 0.42 in the 6-10 year olds and 0.60 ± 0.44 in the 11-26 year olds. McClelland and Saunders discuss that the small number of subjects in the former study and the higher age of the oldest subjects may explain the differences between the studies. Nevertheless, both studies found greater accommodative lags for the greatest accommodative demand. Sterner et al. (2004) report lower amplitudes of accommodation than expected for their age in children (6-10 year olds) using the Donder's push up technique.

With increase in age, the amplitude of accommodation reduces (a recession in the near point of accommodation) eventually causing symptoms such as blur and ocular discomfort at the habitual reading distance (Ciuffreda. 1998). This progressive aging change, caused due to the reduced focusing ability for near objects, is called 'presbyopia' ('aged eye'). This reduction has been shown to be caused due to lenticular changes (increase in lens thickness or hardening of the lens) with increased age (Fisher. 1973, Glasser & Campbell. 1998, Glasser & Campbell. 1999, Koretz et al. 1997, Pau & Kranz. 1991). Presbyopia is first clinically reported between 40 and 45 years of age although its onset is seen to be anywhere between 38 to 48 years (Ciuffreda. 1998, Kleinstein. 1987). Presbyopia can be corrected by the use of additional plus power provided by reading glasses, bifocals or multifocals which compensate for the reduced accommodation and provide clearer near vision.Table 1-2 shows the Donders' table of accommodative amplitude with age (Borish , 1970) as adapted by Ciuffreda (1998).

Table 1-2 Donders age–related accommodative amplitude

Age (in years)	Amplitude of accommodation (in Dioptres)
10	19.70
15	16.00
20	12.70
25	10.40
30	8.20
35	6.30
40	5.00
45	3.80
50	2.60
55	1.80
60	1.00

1.3 Visual acuity and refractive errors in Down syndrome

Reduction in visual acuity with no associated ocular or physiological condition has been observed commonly in this population (Courage et al. 1994, Tsiaras et al. 1999). Woodhouse et al., (1996) using the Cardiff Acuity and Teller Acuity Cards, measured visual acuity in individuals with DS, aged 12 weeks - 4.75 years. They found that the visual acuity in this population was similar to that in the typically developing population from infancy to 2 years of age and then fell below normal with increasing age. This was independent of refractive error.

Alternatively, Courage et al., reported unexplained reductions in visual acuity in children with DS who were 6 months of age (Courage et al. 1994).The etiology of poor visual acuity in the DS population is still not completely understood although there have been a few studies investigating the factors that could be associated. One approach was to use an objective measure such as visually evoked potential (VEP) to see if the reduced visual performance was due to behavioral or motivational factors as compared to an actual reduction of VA (John et al. 2004). This approach showed that the decreased vision could not be attributed to behavioural factors i.e. it was still present with the VEP measurement. The same group of researchers also found significantly reduced contrast sensitivity and used this to support the idea that a sensory deficit is present in the DS population (John et al. 2002). Furthermore they suggest that this deficit in acuity could occur at any location in the visual pathway extending up to the visual cortex but is not likely to be located in the higher areas (John et al. 2002). The VEPs reflect the integrity of the visual pathway up to the level of the primary visual cortex (Little et al. 2007), whereas behavioral measures involve higher and more complex areas of of visual and cognitive processing (Teller. 1997). Since the reduction of VA is seen to be independent of the behavioral or motivational factors as shown earlier, it could mean that the reduced VA may be due to some deficit in the visual pathway up to or including the visual cortex. Suttle and Turner (2004) supported this view by finding a cortical deficit in their participants with DS using VEPs. Recently, Little et al. (2007) also showed that both grating and interferometric resolution acuities are significantly reduced in these individuals and suggested that a neural deficit is partly accountable for the reduced visual acuity. This suggestion has been supported by histological findings of lower brain weight, poor maturation and less organization of the visual cortices in these individuals compared to typically developing controls (Becker et al. 1991).

There have been numerous studies that report the prevalence of high refractive errors in this population, the only difference between studies being whether myopia or hyperopia is more common. The majority of reports support hyperopia being more common than myopia although a few reports are equivocal (Bailey et al. 1989, Berk et al. 1996, Doyle et al. 1998, Fanning. 1971, Jaeger. 1980, Lowe. 1949, Sriubienė et al. , Woodhouse et al. 1997,Cregg et al. 2001, Cregg et al. 2003, Gardiner. 1967, Paudel et al. 2010, Stewart et al. 2005, Stewart et al. 2007, van Splunder et al. 2003). Significant levels of astigmatism have been reported, involving 12.7-56.5% of these individuals (Bailey et al. 1989, Berk et al. 1996, Caputo et al. 1989, Cregg et al. 2003, da Cunha & Moreira. 1996, Gardiner. 1967, Paudel et al. 2010, Stewart et al. 2005). High prevalences of anisometropia have also been reported ranging from 11.6-22% (Cregg et al. 2001, Fanning. 1971, Paudel et al. 2010, Tsiaras et al. 1999). Refractive error percentages from various studies are given in Table 1-3.

There is also a high prevalence of strabismus in this population, involving almost 29-42% of the population (Cregg et al. 2003, Haugen & Hovding. 2001). According to Cregg et al.(2003) the classic assumption is that accommodative esotropia, with its onset at about 3 years in typically developing children, is usually associated with high hyperopia. The children tend to over accommodate to obtain a clear retinal image, leading to a high degree of accommodative convergence which is believed to cause esotropia (convergent strabismus). They also report that this did not seem to be the case in individuals with DS as the presence of strabismus is irrespective of the sign and magnitude of refractive error.

Table 1-3 Refractive errors in Down syndrome

Authors	Participants (n)	Myopia (D)	Astigmatism (in dioptres D)	Hyperopia (D)
Lowe (1949)	35	34% (>-6 D)		8.5% (> 2.62D)
Gardiner (1967)	19	50% 10.5 % (>-6D)	37 % (>2D)	15 %
Fanning (1971)	24	8.3 % (> -2D) 4.25% (> -6D)		54.2 % hyperopia (> 1D) 12.5 % hyperopia (> 6D)
Jaeger (1980)	75	19.7 % (> -4D) 12 % (> - 8D)	15.5 % (>2.5D) 2.7 % (> 5D)	18.3 % (> 4D) 1.3 % (> 8D)
Shapiro & France (1985)	54	10% (< -5D) 27 % (> -5D)	25 % (>3D)	29 % (< 5D) 6 % (> 5D)
Bailey et al. (1989)	116	12.1 % (< 6D) 2 % (>6D)	17.2 % (>-2D)	30.3 % (>2D)
Berk et al. (1996)	55	9% (<-5D) 3.6% (>-5D)	30.9 % (<3D) 12.7 % (>3D)	50.9 % (<5D) 1.8 % (>5D)
Doyle et al. (1998)	50	18% (-0.5 to -8D)		80 % (0.5 – 7.5 D)
Sriubienė et al. (2002)	393	23.9 %	29.3 %	67.4%
Cregg et al. (2003)	99	13.3% (>-1D)	21.8 % (>1D)	32% (>3D)
Sharmani et al. (2006)	60	29.2 %	8.3 %	25 %
Paudel et al. (2010)	36	25% (>-0.50D)	44% (>1D)	55% (>1D)

Emmetropia is the condition in which, in the unaccommodated state, parallel rays of light from an object at infinity are focused on the retina of the eye. Any condition where this does not occur is called ametropia. Myopia and hyperopia are examples of ametropia. Many human infants are born with significant refractive errors. Human infant studies confirm that typically developing infants are mostly hyperopic (Atkinson et al. 1984, Atkinson et al. 2007, Ehrlich et al. 1997, Howland.1993). Typically developing children become more emmetropic or even slightly myopic as they get older (Gwiazda et al. 1993). This decrease in ametropia toward emmetropia is called emmetropization. This process of emmetropization is well established in the typically developing population (Atkinson et al. 2000, Ehrlich et al. 1997, Rabin et al. 1981, Saunders et al. 1995) .

The mechanism of refractive error development and emmetropization is still unclear in individuals with Down syndrome. The process of emmetropization seems to be lacking in many individuals with DS (Cregg et al. 2003). These individuals do not show the typical pattern of refractive error development and according to Cregg et al., 'children with DS who have a refractive error in the early months of life are much more likely to maintain or increase the refractive error rather than outgrow it.' In other words, many individuals with DS fail to emmetropize. Some of them do emmetropise while others may start with low refractive errors and then develop higher errors with time. In other words it is difficult to predict this development in individuals with DS. It has been suggested that typically developing children with strabismus do not emmetropize without or with a spectacle correction and have a poor appreciation of blur (Ingram et al. 1991, Ingram et al. 2000). According to Cregg et al. (Cregg et al. 2003) this could be similar in some DS individuals with strabismus.

Refractive error and biometric studies in DS have shown correlations between axial length and spherical equivalent refractive error (Doyle et al. 1998, Haugen et al. 2001). In this sense they are like the typically developing population (Hosaka. 1988). However factors such as reduced central corneal thickness, thinner lenses and steeper corneal curvatures have also been reported in individuals with DS (Haugen et al.).Whether these optical factors predispose to the failure of emmetropization is still not known (Woodhouse et al. 1997, Doyle et al. 1998, Haugen et al. 2001, Cregg et al. 2003). This implies that children with DS should be monitored at regular intervals throughout their childhood for onset of refractive error and strabismus. As they are more prone to such conditions, they require frequent and more stringent screening criteria in comparison to typically developing individuals.

1.4 Impact of refractive errors on function in typically developing children and those with Down syndrome

Refractive errors are also associated with amblyopia. Amblyopia is defined as 'a non specific loss of visual acuity of at least two lines that is not caused by pathology, nor correctable by ordinary refractive means' (Schapero et al. 1980). Amblyopia caused by hyperopic anisometropia (the difference in the refractive error between both the eyes) is possibly the most common form of refractive amblyopia (London & Wick. 1998) and it is known that uncorrected high hyperopes may develop bilateral amblyopia (Leat et al. 1999, Wallace et al. 2007). Amblyopia is a risk in individuals with Down syndrome, many of whom present with high uncorrected amounts of refractive error, mainly hyperopia. It is likely that individuals with DS, who present with constant reduced accommodation as well as high levels of hyperopia, have a possibility of developing amblyopia. This may also be the case for lower levels of hyperopia because of the constant near blur due to reduced accommodation. In probably one of the only studies that specifically studied amblyopia in individuals with DS, Tsiaras et al.,(Tsiaras et al. 1999) reported amblyopia to be present in 22% of their study group and it was mainly associated with strabismus and high refractive errors. The percentage of visual acuity less than 20/50 associated with other conditions in their DS cohort is given in figure 1-9.

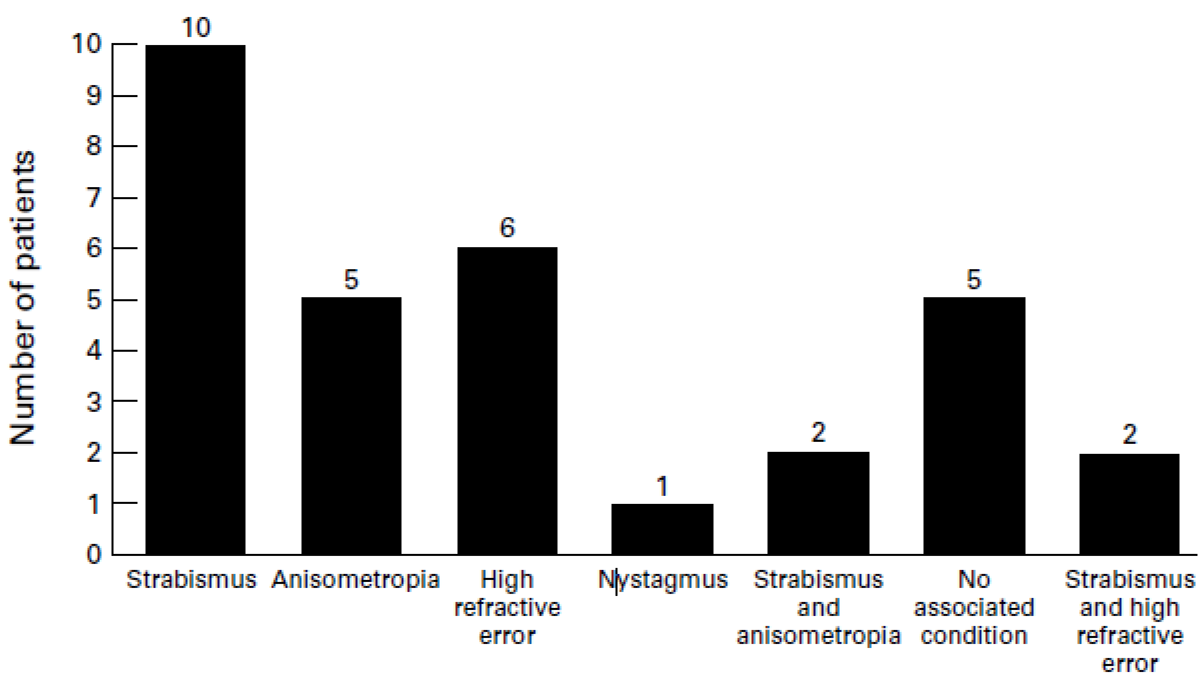


Figure 1-9 Conditions associated with amblyopia and/or bilateral vision < 20/50 (6/15)

Reprinted from British journal of Opthalmology. With permission from BMJ Publishing Group Ltd. William G Tsiaras, Sigfried Pueschel, Charles Keller, et al. Amblyopia and visual acuity in children with Down's syndrome. *Br J Ophthalmol* 1999; 83: 1112-1114.

Uncorrected hyperopia has been shown to be associated with poor VA. Mutti et al.(Mutti. 2007) report that distance visual acuity can steadily worsen by about 2 lines (0.2 logMAR) for the highest amounts of hyperopia (>4D) in children who remain uncorrected. They also suggest that a distance hyperopic correction improves visual acuity and would likely have an impact on near acuity.

A number of studies have linked uncorrected hyperopia, that could result in poor near focusing, with poor visual-perceptual, cognitive, motor and attention skills in typically developing individuals (Atkinson et al. 2002, Atkinson et al. 2007, Rosner & Rosner. 1989, Shankar et al. 2007, Williams et al. 1988). This may also be true in children with DS considering the high amounts of refractive errors with which they present. There are several studies of refractive errors and educational attainments in typically developing populations and these encompass various aspects of literacy functioning. Atkinson et al., (2002) reported that that children between 14 months and 3.5 years of age with larger amounts of hyperopia (>3.5 D) performed significantly worse than a control group on several spatial cognitive and motor tests. In addition, they also found that this early hyperopia was associated with a range of developmental deficits that persisted at least up to the age of 5.5 years. This was still the case even if children with strabismus and amblyopia were removed from the analysis. Similarly in another report, preschoolers with significant hyperopia were also found to have attention deficits (Atkinson et al. 2004). In a recent pilot study, Shankar et al., (Shankar et al. 2007) reported that uncorrected hyperopic children, aged 4 to 7 years, show reduced performance on tests of letter and word recognition, receptive vocabulary, emergent orthography and crowded VA. They mention that it is difficult at this point to know if these hyperopes will catch up to the performance of emmetropes with time. Furthermore, Rosner and Rosner (1997) compared academic achievement in children with and without vision deficits and reported that lower academic scores were present in children who had more than +1.5D of hyperopia. Interestingly, hyperopia has also been found to be an accompanying factor in cases of reduced IQ, reading and academic performance (Shankar et al. 2007). Recently, French et al.(2009) reported that children with uncorrected hyperopia spent significantly less time engaged in near-work in comparison with children without refractive error or those with hyperopia who wore a correction. Hyperopia is also seen to be more prevalent than myopia in children with learning disorders (Rosner & Rosner. 1987). Also hyperopic children have been shown to demonstrate poorer reading performance when compared with emmetropic and myopic children (Eames. 1955, Garzia & Nicholson. 1990, Rosner & Rosner. 1997, Williams et al. 2005, Young. 1963).

There is an increasing amount of literature linking myopia and higher IQ or reading abilities. In one of the early reports on comparison of refractive errors in children with learning disability, the difference in academic performance among learning disabled myopic, hyperopic, and emmetropic children was studied (Wharry & Kirkpatrick. 1986). The results showed that myopic learning disabled children out-performed hyperopic and emmetropic children on a mathematics test and that myopic children also scored better than hyperopic children on some reading subtests and an oral comprehension test. Saw et al., (Saw et al. 2004) supported the view that non-verbal IQ is highly correlated with myopia and those with higher non-verbal IQs had significantly higher myopia than the others. In other studies, myopic children were seen to perform better academically (Grosvenor. 1970, Teasdale et al. 1988, Young. 1963), than hyperopes when controlled for IQ (Ingram et al. 2000) even when they were prescribed for their hyperopia. In the randomized clinical trial by Atkinson et al, prescribing a partial correction for hyperopic infants did not interfere with emmetropization (Atkinson et al. 2000). In fact, Atkinson and colleagues (Atkinson. 1993, Atkinson et al. 1996) found improved visual acuity as well as a reduced chance of development of strabismus by a factor of 4 times, although this reduction in strabismus was not significant in a later study from the same group (Atkinson et al. 2007). Thus it is seen that early correction may be beneficial in many children to correct visual problems. According to Mutti (Mutti. 2007), if the majority of emmetropization occurs in the first year of life and if there is a little change occurring in the refractive status from then on, then the chance of interrupting emmetropization with a prescription at or after one year is minimal. This supports the view that correction of hyperopia in infants of 1 year and older will have little effect on emmetropization. Several studies, however, show that some emmetropisation still occurs after 1 year (Atkinson et al. 1996, Ehrlich et al. 1997, Gwiazda et al. 2005).

There is a need to develop guidelines regarding hyperopic correction, in particular, the timing of when to correct and the amount of hyperopia to correct. There is still a lack of consensus on this aspect, especially for low to moderate hyperopes as it is believed that the children can 'focus through' their hyperopia (Robaei et al. 2006). Leat et al., (Leat et al. 1999) recommend prescribing for hyperopia of 2D or more from 4 years of age. Cotter (Cotter. 2007) suggests prescribing for hyperopia > 1.25D in school children. The Orinda criteria for referral ftom screening hyperopia in school age children was + 1.50D or more hyperopia (Blum et al.1968). Dwyer & Wick (1995) used the Orinda criterion to correct school age hyperopes in a longitudinal study and found that 60 % of the children showed improvements in accommodation and vergence.

These recommendations could be applied to children with DS. There are no existing guidelines for refractive correction in children with DS. The Cardiff group (Stewart et al. 2005, Woodhouse et al. 1997) suggest prescribing for hyperopes >3D. This would be a less proactive approach than suggested above for typically developing children (a common approach in children with developmental disorders). Indeed, the large numbers of children with DS who are not given a spectacle prescription or who are under corrected may still be evidence of this approach.

1.5 Accommodation in Down syndrome

In the last few decades there has been a considerable focus on reduced accommodation in this population (Cregg et al. 2001, Haugen et al. 2001, Haugen & Hovding. 2001, Haugen et al. , Lindstedt. 1983, Stewart et al. 2007, Woodhouse et al. 1993, Woodhouse et al. 1996, Woodhouse et al. 2000) and it has been shown that reduced accommodation is prevalent in 55-92% of the population with DS.

Marked decreases in the amplitudes of accommodation have been reported in children with DS as young as 6 years old. In the first study to document this reduced accommodation, 80% of the children had reduced amplitudes (Woodhouse et al. 1993). In a later study, almost 92% of participants aged 12 weeks – 57 months had accommodation that was reduced and the infants did not show the typical improvement of accommodation with age (Woodhouse et al. 1996). Under-accommodation, as much as 5.00 D for a 10 cm target, was reported and large lags of accommodation were consistently present at all tested distances (Cregg et al. 2001). It was also seen that greater lags of accommodation were associated with higher amounts of uncorrected hyperopia (Woodhouse et al. 2000), while some children with lower amounts of hyperopia had normal accommodation (Haugen et al. 2001).

What causes this reduction in accommodation is still an area that needs work. Woodhouse and colleagues in the Cardiff group (Woodhouse et al. 1993) initially suggested that the reduced accommodation could be caused by premature aging (early presbyopia) of the lens. Later on they showed that the decrease in the amplitude of accommodation in individuals with Down syndrome was not due to physical changes in the lens as occurs in the presbyopic population (Cregg et al. 2001). When presbyopes are asked to view a range of near targets, the total accommodative response is similar for each target distance representing their amplitude of accommodation. In contrast, Cregg at al. found that children with Down syndrome showed constant under-accommodation i.e. the amount of accommodative response varied with target distance and did not improve with spectacle prescription.

Originally, reports from the Cardiff group concluded that correcting this accommodative lag with reading addition lenses would not be useful. In one of their reports, Cregg et al.(Cregg et al. 2001) observed that addition of positive lens power in the distance prescription did not improve accuracy and hence it was concluded that bifocals would not be beneficial in individuals with DS. At the same time, other workers (Leat, personal communication , 2006) were successful in prescribing bifocals clinically in some children with DS who were seen at the Pediatric and Special Needs Clinic at the School of Optometry, University of Waterloo. There were significant improvements in near and distance VA as well as improvements in the accommodative response such that the bifocal addition could be reduced, although not enough improvement to remove the bifocals altogether. Significant improvements in learning were seen in some children as indicated by parental reports that included (1) has jumped two grades since got new bifocals, (2) improvements in fine motor skills, (3) reads well with bifocals, (4) reading at grade 5 level and (5) loves to read. The Cardiff group then placed their cohort into bifocal lenses and published two studies showing that prescribing bifocals in DS populations improved accommodative accuracy (Al-Bagdady et al. 2009, Stewart et al. 2005). In the first report, 34 children (ages 5-11) with DS were assigned in equal numbers to form two matched groups and were followed up thrice over a 5 month period. The treatment group was prescribed bifocals with a +2.50 addition and the control group with SV lenses to correct for any refractive errors. They found that the bifocal treatment group showed consistently more accurate accommodation compared to the controls ($p<0.001$) over the entire period. In the second, recent report, the clinical records of 40 children from the Cardiff Down Syndrome Vision Research Unit, who had been previously prescribed bifocals, were reviewed. The duration of the follow up review was between 1-7.8 years in this group of patients. They reported that 14 children showed accurate accommodation according to their critera, 12 of them showed improved accommodation and the rest (16) did not show any change. In addition, both these reports showed that the accuracy of accommodation improved through the distance portion of the lens with time in many children. The 2009 study even suggested that bifocals can be used as a temporary treatment modality in many children and that they can be discontinued with time. These studies, however, are observational studies reported over the last few years and discuss their findings from a clinical population with DS and do not include any functional measures.

1.6 Literacy and Visual - perceptual skills in Down syndrome

There is now a large body of literature on the development and teaching of reading and literacy skills in children with DS. Over many decades, the attitude towards the education of children with challenges, such as seen in DS, has changed. In one of the earliest attempts to impart basic literacy skills, Hayden and Dmitriev (Hayden & Dmitriev. 1975) developed an educational program and showed that children with DS could be trained to read. Since then, there has been surmounting evidence showing that children with DS are capable of learning to decode words (Boudreau. 2002, Byrne et al. 2002, Lorenz et al. 1985) and that many can achieve foundational levels of reading (Casey et al. 1988, Laws. 2000, Lorenz et al. 1985, Sloper et al. 1990) and basic skills like writing and numerals (Duffen. 1976, Shepperdson. 1994, Turner & Alborz. 2003). Lorenz et al., (Lorenz et al. 1985) administered a series of questions to teachers about reading-related skills in order to understand the early pre-reading and reading abilities of 58 children with Down syndrome aged 5 to 7 years. Their results showed that 19, 32 and 44 percent of the 5, 6 and 7 year olds respectively were able to read 5 to 10 words. Buckley (Buckley. 2001) quotes , from studies done in Australia and the UK, that 60-70% of individuals with DS are able to achieve functional reading by their adult life. Van Kraayenoord et al. suggest that many of these individuals can still develop 'functional levels' of literacy in their later years and continue to develop these skills with appropriate instruction (Van Kraayenoord et al. 2000). 'Functional reading skills', as explained by Buckley, (Buckley. 2001), is a 'reading age' of 8-9 years that would be adequate to read books, newspapers and write letters. This functional achievement in children and young adults with DS was associated with factors such as early instruction, parental attitudes and the type or nature of skill sets administered at different ages (Lorenz et al. 1985). Other studies have reported similar evidence on the attainment of literacy skills in children with Down syndrome (Fowler et al. 1995, Kay-Raining Bird et al. 2000, Kay-Raining Bird et al. 2008).

Most of the studies described above were cross-sectional studies. There have been some longitudinal studies that describe developmental aspects of reading, literacy and academic ability in children with DS (Cupples & Iacono, 2000 [this study is both cross sectional and longitudinal], Laws & Gunn, 2002, Kay-Raining Bird et al. 2000). The advantage of a longitudinal study is that it gives information on the development of one or more characteristics being measured across a certain duration of time. Cross-sectional studies, on the other hand, involve studying one or more groups of participants for certain characteristics at the same point in time. These longitudinal studies show that children with DS, although showing variability in performance, still demonstrate steady progress in reading accuracy (word identification skills) but their reading comprehension, phonological awareness, language, spelling and memory skills are still constrained (Byrne et al. 2002, Kay-Raining Bird et al. 2000, Kay-Raining Bird et al. 2008).

It has been suggested that individuals with DS have specific areas of relative strength while they may lack in the others. This relative comparison is within their own areas of strength and weaknesses i.e. within the Down syndrome population – they still may show deficits in all areas in comparison with typically developing individuals. With respect to early literacy, Word Identification (Fowler et al. 1995, Kay-Raining Bird et al. 2000) and vocabulary comprehension (Kay-Raining Bird et al. 2008) are areas of strength in individuals with DS. Phonological awareness, (Cossu et al. 1993, Fowler et al. 1995, Kay-Raining Bird et al. 2000) (which is defined as the ability to focus consciously on the sound structure of a language (Cupples &

Iacono. 2000), as well as decoding ability (Boudreau. 2002, Kay-Raining Bird et al. 2000) is seen to be deficient in children with DS. Other areas of strength and weakness in abilities such as visual perception are discussed in the following sections. From an educational stand point, sight words have been shown to be an effective teaching tool in individuals with moderate to severe mental challenge (Browder & Xin. 1998). These sight words are high frequency words that are difficult to decode and are recognized by sight (Juel. 1980). This method of sight word teaching could be useful in individuals with DS, many of who present with varying levels of mental challenge (Browder & Xin. 1998).

Although there are many studies of reading in children with DS, there is only one study of writing. This study focuses on written narratives in this population (Kay-Raining Bird et al. 2008). The few studies that describe writing characteristics in typically developing individuals concentrate on either the development of writing and/or the association of developmental skills with writing (Berninger et al. 1992, Berninger et al. 1994, Berninger et al. 2002, Swanson & Berninger. 1996). Kay-Raining-Bird et al. (2008) studied writing fluency and written narratives in DS by first administering an alphabet-fluency task which involved writing or typing as fast as possible in lowercase letters without a sample provided to copy. They measured written narratives by asking the children to read their own written story which was glossed, the correct spelt words being placed above the children's words in the story. However they did not report the results of the data in terms of the legibility of letters and concentrated on the written narratives. They reported that many of the school-age children with DS showed written narrative abilities which were comparable to the reading-matched controls. Furthermore, they found the written narratives to be predicted by the vocabulary comprehension skills. This shows that there is a link between vocabulary comprehension and attainment of literacy skills. This is important, as vocabulary comprehension has been shown to be a strength in these individuals.

A literature search found no studies on printing or the formation of letters in any groups of children. Printing needs to be differentiated from writing, which according to Berninger & Swanson (Berninger & Swanson. 1994) involves transcription, which is spelling and handwriting, plus the text generation process. Both printing and writing require fine-motor skills in order to generate text. Thus there is a paucity of studies assessing printing or writing skills in the DS population and Kay-Raining Bird et al. (2008) comment that written language has generally been an area of neglect in the literature, specifically in individuals with intellectual disability. It should be noted that motor delays are also present in individuals with DS (Carr. 1970, Henderson et al. 1981, Spano et al. 1999). It has been suggested that these neuro-motor difficulties could result in poor finger coordination and poor formation of letters in children with DS (Cowie. 1970). Therefore deficits in a complex function such as printing, which involves fine motor skills, would be anticipated. This constraint in fine-mottor skills has also been suggested by Kay-Raining Bird et al.(2008), who find increased constraints when the individuals with DS were asked to write text. It is apparent that there is a lack of research in this area and studies of printing development in the DS population may be a useful area of study, in order to understand this facet of literacy skills.

Along with literacy skills, visual-perceptual skills have been studied in the Down syndrome population and it is known that many of these individuals can perform these tasks (Dykens et al.2001 , Spano et al. 1999, Wang et al. 1995). Dykens et al. (2001) reported that the children with DS who participated in their study showed significant strengths on visual motor integration, in excess of their IQ expectations. There are similar reports of these individuals demonstrating stronger visual or visual motor skills than performance on verbally guided tasks. Specifically, Klein and Mervis (1999) showed that children with DS showed strengths in areas of visuospatial construction compared to auditory or verbal tasks. Similarly, Rohr and Burr (1978) reported that children with DS showed lower verbal abilities compared to visual motor abilities and that their verbal – auditory levels were weaker in comparison to two other cognitively-matched groups (based on IQ) with cognitive delays who were described as biologically brain damaged or having delays due to unknown causes.

Auditory short term memory in individuals with DS has been studied by a few researchers and it has been concluded that these individuals perform better when provided with visual rather than auditory information (Marcell & Armstrong. 1982, Marcell et al. 1988, Rohr & Burr. 1978, Varnhagen et al. 1987). This has been attributed to the high incidence of middle ear disorders and auditory difficulties that are present in the DS population (Balkany et al. 1979, Brooks. 1972, Cunningham & McArthur. 1981, Dahle & McCollister. 1986, Roizen et al. 1993, Shott et al. 2001). Jarrold and Baddeley (1997) dispute that the poor short term memory for verbal as opposed to visuospatial tasks was due to hearing abnormalities but argue that it is rather due to the selective impairment of the phonological loop in working memory. According to Baddeley 'the phonological loop stores and rehearses speech-based information and is necessary for acquisition of both native and second language vocabulary' (Baddeley. 1998).

It is quite clear that Down syndrome is a multifaceted condition in which individuals may have visual, auditory and cognitive processes affected to different degrees. Thus in an educational setting, it is important to understand and work with these individuals in an individualized manner specifically suited to their needs. From the few studies mentioned above, it is clear that some of these individuals may have strengths in visual tasks (such as Word

Identification) in comparison to verbal and auditory skills. From an educational stand point, sight words have been shown to be an effective teaching tool in individuals with moderate to severe mental challenge (Browder & Xin. 1998). This would mean that emphasizing visual information in the educational program for children with DS may be more fruitful. For a higher performance on such tasks, it would be anticipated that optimal visual acuity and accommodation is required, which can be provided by appropriate refractive correction. Since individuals with DS need to rely more on visual, rather than auditory information, the importance of optimizing the clarity of vision becomes more apparent.

2 Aims and objectives

2.1 Rationale

From the background in the previous chapter, it can be seen that only one group of researchers in Cardiff (Al-Bagdady et al. 2009, Stewart et al. 2005) have studied the effect of bifocals in correcting for the reduced accommodation in individuals with DS. These studies, however, are observational studies reported over the last few years and discuss their findings from a clinical population with DS. Although there is some evidence of improved accommodation with bifocals, their effect on academic attainment and visual pereptual skills in this population has not been studied to date. It is commonly accepted that a visual correction will help an individual when there is a significant refractive error. This would be expected to apply to populations with Down syndrome. In fact, this may be even more the case in populations with Down syndrome, as reduced visual acuity and accommodation may be an additional barrier to academic performance than in typically developing children. One way to understand how children and young adults with DS can perform in their everyday lives with a spectacle correction would be to follow them over longer time periods using everyday skills tests to evaluate reading, writing and perceptual skills. This would enable the gathering of information of their progress (if any) with a visual correction, in comparison to the absence of a correction. Hence this study was designed to measure this impact. A visual correction, in the form of bifocals was provided to a group of school children and teenagers with DS and they were followed before and after the bifocal correction over a 12-18 month study period. Visual acuity, accommodative ability, early literacy and visual-perceptual skills were also assessed. The aims and objectives of this thesis are given below.

2.2 Aim

The aim of the study described in this thesis was to investigate the impact of bifocals (to correct reduced accommodation), on the visual function and educational attainment in children and young adults with Down syndrome.

2.3 Hypotheses

1. The prescription of bifocals in children and young adults with DS will facilitate the improvement of near visual acuity as compared to VA without a bifocal
2. Bifocals will result in education gains as indicated by
    a. Improvements in standardized tests of early literacy and visual-perceptual skills
    b. Improvements in printing skills as measured by the average size and position of the letters on the line and the variability of these measures - a smaller and more uniform positioning of letters may indicate a more developed or mature writing
    c. Improvements in school reports before and after the prescription of bifocals
3. Bifocals will result in improved efficiency of performance on the tasks administered, that is, there will be faster completion times of the tasks with bifocals.
4. The improvements in vision will result in better compliance with spectacle wear for bifocals as compared to single vision spectacles.

2.4 Study design

The present study, utilized a longitudinal study design to evaluate the efficacy of bifocals. It could argued that a randomized clinical trial study design would be an ideal choice to study the efficacy of bifocals. However, if rightly designed, this study design would require a larger sample size, in order to balance the many factors that influence performance in children with DS, such as cognitive ability and level and type of educational programme. This would most likely require multiple centers, and would therefore become expensive in nature. Therefore the present study used a longitudinal study design with delayed intervention to evaluate the efficacy of bifocals with participants acting as his/her own control. Single vision (SV) lenses were prescribed to all participants at the initial optometric visit based on the criteria described in Chapter 3. A battery of literacy and visual-perceptual skills (described in Chapter 4) were administered at the SV baseline,visit and at the 6th month with bifocals.

Because a control group was not used, and therefore improvements in the battery of tests might be due to natural progression over time, monthly subtests or probes were administered. Each participant acted as his/her own control and was followed up monthly for attributes such as early literacy and printing skills. The progression of skills was measured by the scores at each month and regression lines were plotted to measure the rate of progress pre- and post- bifocals. Hypothetical graphs of the possible outcomes after bifocal prescription are given in Figure 2-1. It was expected that there could be (a) a gradual improvement prior to bifocals and then a faster progression in performance after bifocals would be seen as a change in slope (Figure 2-1a) or (b) a sudden jump in performance immediately after bifocals and then a steady progression thereafter (Figure 2-1b). The group of participants were followed over a 12-14 month period to track their educational progress and monitor for any progression with bifocals on (a) Early literacy skills, (b) Visual-perceptual skills and (c) Early printing skills. Periodic measurements of accommodation and visual acuity were included. All the monthly and main sessions were videotaped to observe any changes in the time taken to complete all the tasks, that is, whether the literacy, perceptual and printing tasks were completed faster with bifocals.

The detailed study design and methodology and baseline results with single vision lenses in the group of participants is described in Chapter 3.

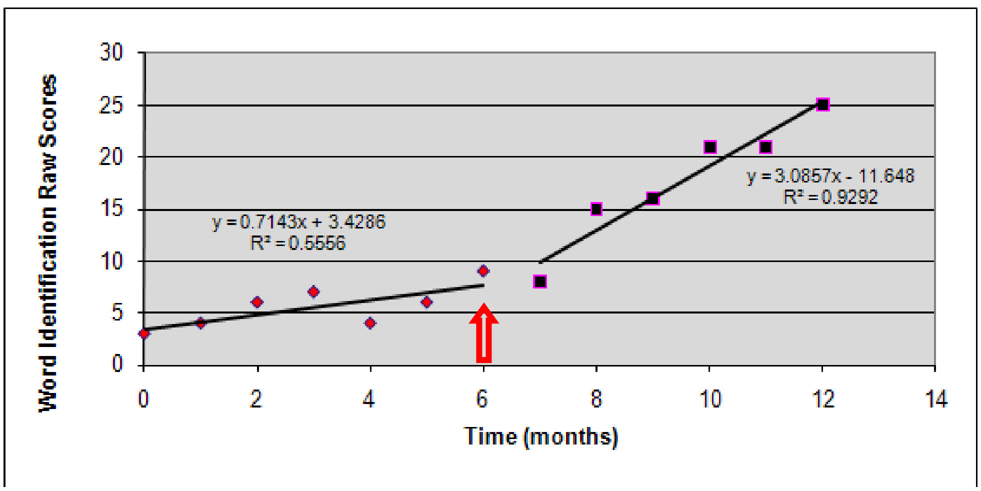


Figure -2-1a : Gradual improvement and then faster progression with bifocals.

(Arrow indicates the month when bifocal is prescribed)

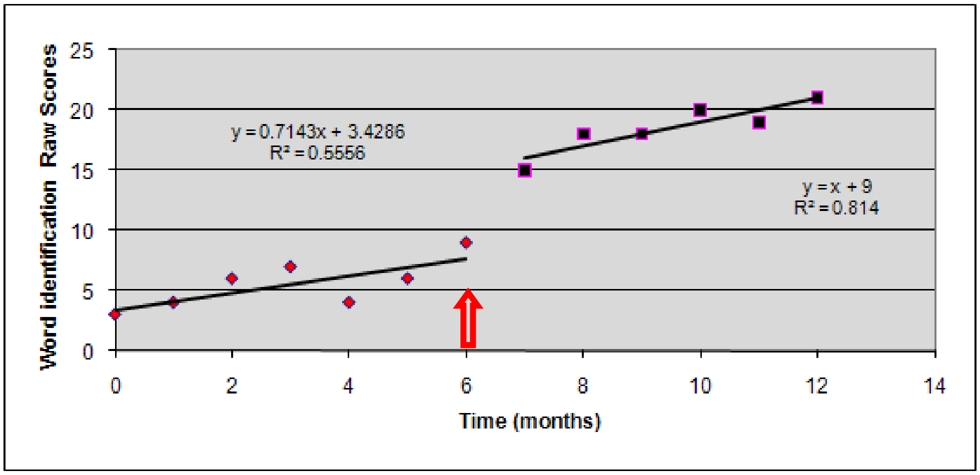


Figure -2-1b : Sudden jump and then a steady progression after bifocals

Figure 2-1 Hypothetical graphs of progression on the WI score after bifocal provision

2.5 Key novel aspects of the study

This current study is the first and only study to measure the functional impact of bifocal correction in children and young adults with DS, using both a battery of tests and monthly subtests. The following are specific novel aspects of this study:

2.5.1 Printing tasks

Rather than deal with the more complex task of writing composition, it was decided to consider the more basic task of printing formation as a measure of the impact of bifocals. So the printing tasks were made simple by asking the child to print his/her name, any ten letters and any ten numbers. Printing was evaluated by measuring the size and position of each of these letters. The instruction did not specify if the children had to print or use cursive writing but the children all chose to print. Hence these tasks are referred to as ‘printing’ tasks in the majority of this thesis.

2.5.2 Compliance

Compliance was measured indirectly through parental reports and from observations in the laboratory i.e., if the participant was more ready to keep his/her glasses on or came in with their glasses, to determine if there was a difference in this compliance before and after bifocals.

2.5.3 Progress at school

The teachers were requested to complete a questionnaire (given in Appendix C) at the end of each 5 month period on the child's progress at school. Similar information was also obtained by requesting a copy of the child's school progress report every term to be compared before and after bifocal provision, as another measure of progress.

2.5.4 Videotaping the sessions

All the sessions of early literacy and visual-perceptual skills were videotaped and were randomized and analysed by an observer who was naïve as to the visit (SV, first or second bifocal visit). Unfortunately, it was not possible to always blind the observer to whether the child had a SV or a bifocal (as the line was occasionally visible in some of the videos). However, the , the videotapes were randomized such that the observer was unaware of the exact visit. The time taken by each child to complete each page in every task was calculated to determine differences before and after bifocals.This was used as an additional outcome measure to understand if there any change in the efficiency of performance after bifocals, even if there was no improvement in actual scores.

3 Bifocals in Down syndrome study (BiDS) – Study design and baseline results of visual function

This chapter is published as follows:

Krithika Nandakumar, Susan J Leat. Bifocals in Down syndrome study (BiDS): Design and Baseline Visual Function. *Opt Vis Sci 2009; 86(3):196-207.*Reproduced with permission from *Optometry and Vision Science.*

Roles

	Concept / design	Recruitment	Acquisition of data	Analysis	Write-up / publication
Nandakumar	Modifications, development of specific protocol for the administration of tests	Y	(1) Refraction to determine SV correction (2) Dynamic retinoscopy for determining lag (3) All other data	Analysis of the data	Write up
Leat	Original concept created for the grant application	-	(1) Dynamic retinoscopy for determining lag. (2) Rechecked refraction and signed prescription	Guidance and suggestions	Editing and suggestions

3.1 Overview

Purpose*:* Among children and young people with Down syndrome (DS) there is a high prevalence of reduced accommodation. Prescribing bifocals for these patients has not become fully clinically accepted, although it would be anticipated to improve visual acuity (VA). The aim of this study is to investigate the impact of bifocal correction on visual acuity, visual perceptual skills and early literacy development in children with DS who have reduced accommodation and who are provided with a bifocal correction. This paper describes the study design and the baseline optometric findings.

Methods: We have chosen a longitudinal design with frequent measures of subtests of performance to control for progression with time. The main outcome measures are early literacy and visual perception skills. Secondary outcomes are visual acuity and accommodative function. These are measured at baseline, the participant followed for 6 months when bifocals are prescribed if necessary and the participants followed for another 6 months with bifocals.

Results: Fourteen participants with DS aged 8-18 years were enrolled. At baseline 79% required a change in their distance spectacle prescription. 100% had reduced accommodation both before and after new single vision glasses were prescribed. None had an adverse reaction to 0.5 or 1% Cyclopentolate. All of the subjects were able to perform either a distance or near crowded Patt-pics matching test. There was a significant improvement of near VA with the new single vision spectacles ($p=0.015$). The mean binocular distance VA was 0.362+/-0.17 logMAR while binocular near VA was 0.489+/-0.235.

Conclusion: This study confirms previous findings of a high prevalence of reduced accommodation and shows that near VA is reduced compared to distance VA. The present results indicate that all subjects might benefit from bifocal provision.

Key words: Down syndrome, bifocals, accommodation, visual perceptual skills, early literacy skills, refractive error, visual acuity

3.2 Introduction

Down syndrome (DS) is one of the most common genetic anomalies, occurring in about 1 in 1000 live births. The syndrome was first described by John Langdon Down in 1866 (Down. 1866), as having particular physical manifestations accompanied by moderate mental challenge (Haugen & Hovding. 2001). There have been numerous studies that have described the increased incidence of various ocular findings in individuals with DS (Caputo et al. 1989, da Cunha & Moreira. 1996, Haugen et al. 2004, Shapiro & France. 1985).These ocular disorders include epicanthus, Brushfield spots, high refractive errors, reduced visual acuity, strabismus, nystagmus, keratoconus, cataracts and hyperplasia of the iris (Catalano. 1992, Pueschel & Gieswein.1993). Reduced accommodation is another common manifestation in children with DS which has now been described in a number of studies (Haugen et al. 2001, Haugen & Hovding. 2001, Lindstedt. 1983, Woodhouse et al. 1993) with about 55- 80% of children with DS having reduced accommodation (Haugen et al. 2001, Woodhouse et al. 1993). It is likely, therefore, that there would be reduced visual acuity for near work, resulting in decreased performance for near tasks in these cases.

It was a common misconception until a few decades ago that children with DS could not achieve many reading, writing or educational skills. Until recently, many children with Down syndrome had not been introduced to fundamental literacy skills (Bochner. 2001), but it has now been shown that children with DS can attain some basic levels of reading and writing skills. Early on, Hayden and Dmitriev (1975) devised an educational program and successfully showed that children with DS could be taught to read. Most children with DS are capable of learning to read single words (Boudreau. 2002, Byrne et al. 2002) and many can attain useful levels of reading (Laws. 2000) plus other foundational skills such as writing and learning numbers (Turner & Alborz. 2003). Laws et al. (2000) reported that placing children with DS in mainstream education gave important gains such as development of language and memory.

This being the case, the blur caused by the presence of reduced accommodation, could be an additional barrier in these children achieving their maximum potential in early literacy and numeracy skills, and could also impact their development of cognitive and/or visual perceptual skills. There are a number of studies that have linked uncorrected hyperopia (which may also result in near defocus or asthenopia) with poorer visual cognitive/perceptual tests and/or reading skills (Atkinson et al. 2002, Rosner & Rosner. 1997, Shankar et al. 2007, Williams et al. 1988). Shankar et al. (2007) recently showed that normally developing children with uncorrected hyperopia were delayed in their early literacy and writing skills. Clinically, a distance correction is generally provided to children with DS, by means of single vision lenses, but prescribing bifocals has not become the standard. Earlier, Cregg et al. (2001) found that the addition of a positive distance prescription did not improve the accuracy of accommodation and concluded that, therefore, bifocals would not be beneficial (the child would further relax his/her accommodation). More recently, Stewart et al.(2005) prescribed bifocals for children with DS and found accommodation to be more accurate both with and without the bifocal in place. Clinically, Leat has prescribed bifocals in children with DS since being involved in the initial study (Woodhouse et al. 1993), which described the deficits of accommodation and has seen anecdotal evidence of significant improvements in learning, as well as visual acuity, in some children. Parents reported such changes as “improvements in fine motor skills”; “reads well”; “has jumped two grades since got new bifocals”; “reading at grade 5 level”; “loves to read”. She noted changes such as improvements in near and distance visual acuity and improvements in the accommodative response so that the power of the bifocal could be reduced (although not eliminated). Therefore the motivation behind this study is to investigate the impact of bifocal provision in a controlled study.

Another common finding in children with DS is reduced visual acuity, with no obvious physiological cause. It is common to find acuity values between 6/30 and 6/9 for DS children between the ages of 6-12 years (Courage et al. 1994, Tsiaras et al. 1999). It has recently been shown that this is not due to behavioral differences in test performance between children with and without Down syndrome – the differences remained when objective testing was used (John et al. 2004). It is possible that the constant under-accommodation for distances closer than infinity might result in a subtle bilateral amblyopia. Indeed, children with bilateral aphakia do not attain a full 6/6 visual acuity and lag behind in their acuity development even after surgery (Maurer et al. 1989). These aphakes are typically corrected with bifocals, but would still experience defocus for object distances other than infinity and the dioptric distance of the reading addition. Also, children with moderate hyperopia may not obtain such high levels of VA as those with emetropia or myopia (Atkinson et al. 2002, Shankar et al. 2007) and it is well known that high hyperopes may develop bilateral amblyopia (Leat et al. 1999, Wallace et al. 2007).

The aim of this study, therefore, is to investigate the impact of bifocal correction on visual acuity, visual perceptual skills and early literacy development in children with DS who have reduced accommodation and who are provided with a bifocal correction. The hypothesis is that:

1. The prescription of bifocal lenses in children with DS will result in an improvement of near corrected visual acuity as compared to visual acuity without the bifocal.
2. Bifocal lenses in children with DS will result in visual perceptual and educational gains.
3. The prescription of bifocals (or multifocals) will facilitate the development of improved corrected absolute levels of visual acuity.

We have designed a longitudinal study to evaluate the efficacy of bifocals in providing visual and educational gains to children with DS. This paper describes the study outline and baseline optometric results, (including visual acuity and accommodation with habitual and newly prescribed single vision spectacles) of the ongoing study. Also, since there have been suggestions of an increased sensitivity to atropine and possible other similar drugs (Berg et al. 1959, Sacks & Smith. 1989), we will report our results of the use of Cylopentolate in this population. The baseline results and first 6 months of probes of the early literacy and visual perceptual skills will be described in a subsequent paper.

3.3 Methods

This is a preliminary longitudinal study designed to investigate the efficacy of bifocals where each child acts as his/her own control. The ideal study would be a randomized, masked clinical trial. However, if correctly designed, such studies are very expensive. Because of the variability of cognitive abilities and levels and types of educational programmes among these children, a fully case-controlled randomized clinical trial would require a large subject sample. Since this is the first study of its kind, we have chosen to undertake a longitudinal natural history study initially, with each child acting as his/her own control and with detailed and frequent measures taken on fewer subjects. The format of the study is shown in Figure 3-1. The main outcome measures are standardized tests of early reading - Letter Identification, Word Identification and Word Attack, Visual-motor integration, Visual-perceptual skills, and visual acuity (distance and near). Accommodative response is also monitored. The full battery of outcome measures are administered at baseline (after correct single vision spectacles have been prescribed) and at 1-2 weeks, 6 months and 12 months after bifocals are prescribed and the pre- and post- results will be compared. The children in the study are attending school and therefore some natural progress of skills is expected with time. In order to control for and measure this natural progression, we will use a delayed intervention plus monthly measures of sub-tests of reading and perceptual skills to measure possible changes in the rate of development when bifocals are prescribed. These sub-tests will allow statistical analysis within subjects e.g., give information on the variability of performance (mean and standard deviation) and rate of change over time, before and after bifocal provision. These frequent measures (or probes) consist of two sub-tests which are given at more frequent intervals, so that the progress of the child may be tracked more systematically.

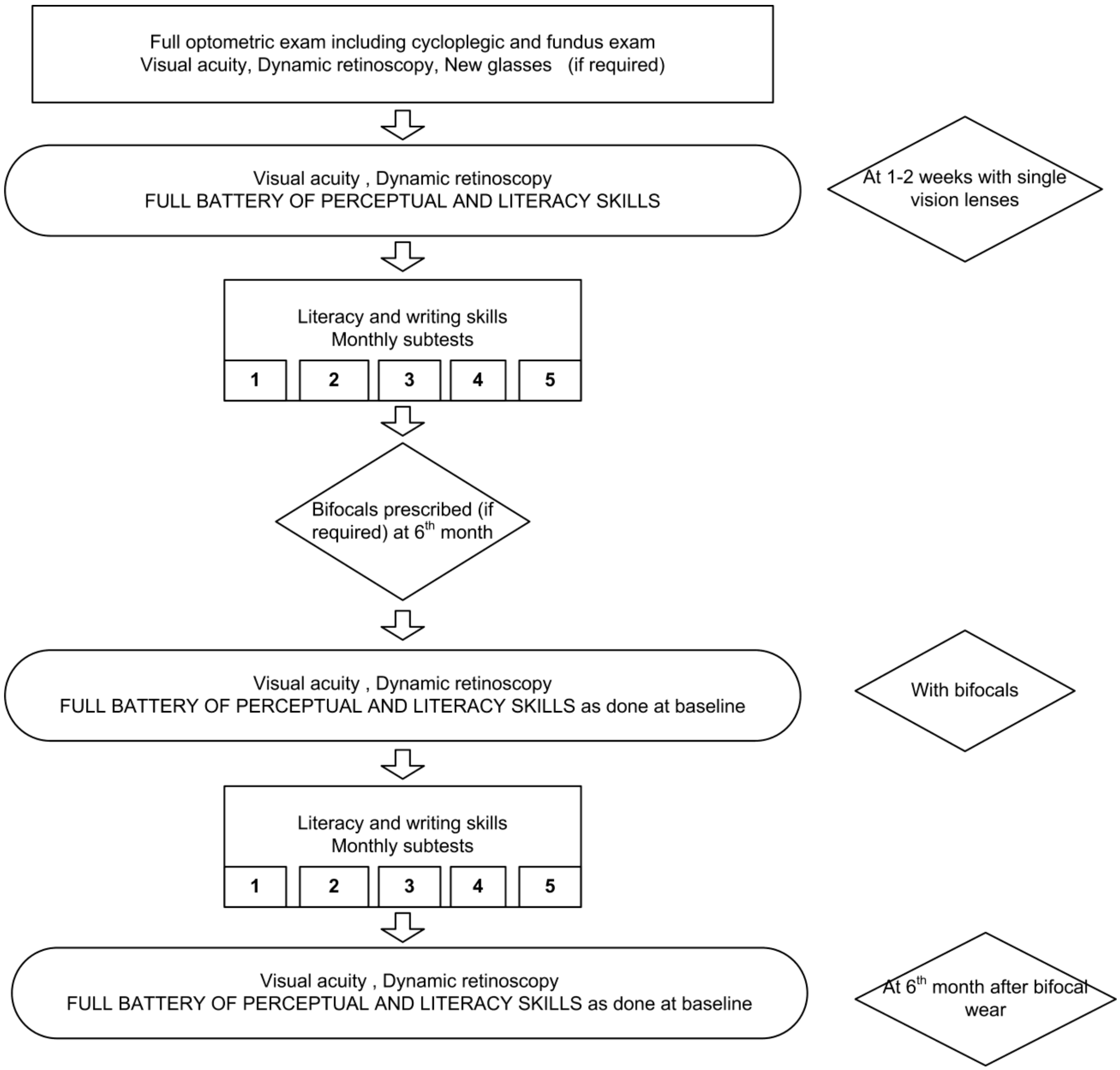


Figure 3-1: Flow chart of the study protocol

3.3.1 Participants:

The study participants were recruited from the Paediatric and Special Needs Clinic, School of Optometry at the University of Waterloo and from local Public and Catholic School boards, the local Down Syndrome Society and through advertisements in local newspapers. The study conformed to the declaration of Helsinki and was approved by the Office of Research Ethics at the University of Waterloo.

Inclusion criteria were:

- Diagnosed with Down syndrome
- Age between 5 years to 21 years i.e. school age. This lower age limit was chosen because the child must be enrolled in a school program that involves reading and other academic pursuits. The upper limit is higher than normal school leaving age, because, in Canada, most children with DS remain in school until about 21-22 years of age, a little older than the normal school leaving age.
- Engaged in academic instruction, at least pre-literacy learning of letters
- Speaks English as first language
- Not already wearing bifocals
- Be verbal or able to understand instructions
- No other significant eye conditions, such as keratoconus or cataract
- No other diagnosed neurological, sensory or behavioural disorders such as autism, microcephaly or significant hearing loss
- Children with strabismus were not excluded. Since strabismus is very common among Down syndrome children, excluding these would significantly reduce our numbers.

The prevalence of strabismus in DS children has been reported to be between 20-47% (Deacon et al. 2005, Kim et al. 2002, Stephen et al. 2007, Tsiaras et al. 1999).There is no reason why children with strabismus would not benefit from bifocals – in fact they may have an added benefit of decreased strabismus for near vision (Haugen et al. 2001) although Cregg et al. (2003) did not find that strabismus in DS is related to refractive error as is common in other children.

Fourteen children were recruited to the study, aged 8-18 years at the beginning of the study, 6 males and 8 females (Table 1). Two potential participants were not included, as they were deemed not able to co-operate with the testing.

3.3.2 Initial optometric visit:

Binocular distance and near logMAR VA were measured using crowded Patti-Pics symbols (Precision Vision.2008). The distance version had a central symbol surrounded by crowding bars and the near version was the MassVAT version with a row of 5 letters surrounded by a box. The distance and near tests were started above the acuity threshold and the method of limits was applied, showing one shape at each acuity level until the first error was made. The matching cards were used, unless the child was able to verbalize all the shapes. On making the first error, the examiner went back to the previous line and completed 4 presentations at each level. The test was stopped when 3 errors were made in a line (3/4) i.e. the lower stopping criteria was 3 out of 4 errors. We also ensured, for an upper limit, that the subject was correct on at least 3 out of 4 presentations. Visual acuity was calculated as a by-letter score, each letter being worth 0.025 logMAR. Distance VA was tested at 3m and near VA at the habitual working distance, which was found by asking the parents about the usual distance at which the child reads. These habitual working distances ranged from 15-40 cm. The unilateral cover test (distance and near) was undertaken or Hirschberg test for less compliant children. Photopic pupil size measurements were taken with a pupil gauge. Dry refractive error was measured with streak retinoscopy and a trial frame while a DVD cartoon was played at 3 metres.

Accommodative response was measured using the modified Nott dynamic retinoscopy, as described by Leat and Mohr (2007). Responses were measured at 4, 6, 8 and 10 D of accommodative demand by two observers (SL and KN) with or without habitual correction and the responses were averaged. The meridian was selected by considering the dominant eye and the meridian which had the least uncorrected positive error (least uncorrected hyperopia or most uncorrected myopia). The same meridian was used throughout the study. The dynamic retinoscopy bar had a white acrylic cube with 4 different pictures to keep the child's attention and with a chinrest for better positioning. A thin measuring tape was attached to the retinoscope at the sight hole to measure the distance of the neutral reflex more accurately. The examiner used a bracketing technique to determine the neutral point. The dioptric distance of the peephole of the retinoscope from the eye being measured gave the accommodative response. The accommodative response values were compared with the control data from Leat and Mohr (2007).

The openness of the anterior chamber angles was estimated with a penlight in order to determine any contraindication for cycloplegia. One or two drops of Cyclopentolate hydrochloride 0.5 or 1% were used, depending on the age and pigmentation of the participant. The 1% drops were instilled only once. A dilated fundus examination was performed after 20 minutes and a cycloplegic retinoscopy after 30 minutes and pupils were measured prior to and after the instillation of drops in most of the participants. Single vision glasses were prescribed based on the cycloplegic refraction, if the difference between the measured refraction and the habitual glasses was more than 0.5D in any meridian. If the child had no current correction, glasses were prescribed if there was hyperopia ≥1D, astigmatism ≥1D, or myopia >1D in either eye, except for an eye which demonstrates a constant strabismus. It was considered important at this point in the study that the child had an accurate distance prescription, so that changes of outcomes after prescription of the bifocal would not be compounded by changes in distance glasses. Thus the criteria for prescribing new glasses were fairly strict. The glasses were provided at no charge to the parent and were dispensed with polycarbonate Essilor Airwear® lenses and a Crizal® coat.

3.3.3 Baseline Visit (After new single vision spectacles had been worn for 1-2 weeks):

Visual acuities (distance and near) were measured with the new single vision lenses. Dynamic retinoscopy was repeated in the same meridian and eye as at the previous visit by both observers at 4, 6, 8 and 10 D and the results of the two observers were averaged.

The following battery of tests for visual perceptual skills was done at baseline in the following order:

- The Beery Visual Motor Integration (BVMI) (Beery. 1997)
- Woodcock Reading Mastery Test (WRMT-R) 1; *Subtests:* Letter Identification (LI), Word Identification (WI), Word attack (WA) (Woodcock.1998)
- The Test For Visual Perceptual Skills (TVPS) *Subtests:* Visual Discrimination, Visual Form Constancy , Visual Closure (Gardiner. 1996)
- Peabody Picture Vocabulary Test (PPVT) 1(Dunn & Dunn. 1997)
- Dolch sight word list (2008)

➢ For children unable to read any letters, a list of numbers was devised and used instead of the Woodcock and Dolch.

The criteria for selecting each literacy or visual perception test was a) it should be a standardized and known test b) ideally, there should be at least 2 versions of the test, so that a different version could be used for the baseline and the outcome measures, to reduce repetition effects c) ideally, there should be previous data indicating that children and teenagers with DS would be able to perform the test. The BVMI, TVPS, WRMT-R and PPVT are all standardized tests and the BVMI, WRMT-R, PPVT have been used previously with subjects with DS (Cupples & Iacono. 2000, Fidler et al. 2005, Kay-Raining Bird et al. 2000, Klein. 1999, Spano et al. 1999, Wang et al. 1995). The children were videotaped while performing the tests and the DVD files will be used to determine the time taken for each components or subtest of each test. The videos were labeled in a random order and will be analyzed by a naïve observer who will be blind as to the type of lens (single vision or bifocal) the child is wearing. A more detailed description of these tests and the development of the Dolch and number tests will be given in the subsequent paper.

The order of the tests was not randomised, but was established in advance. The order was chosen to optimise the performance of the child. Thus the tests that the child would tend to enjoy more and that were less threatening (e.g. the BVMI) were done earlier and were interleaved with the more demanding reading tests. The Peabody was last, since it was not an outcome measure, but would be used to correct for the mental age of the child. The order was not randomized, as we were interested in comparing measures before and after bifocal provision (rather than an absolute measure of the child's performance), and wished to eliminate as much as possible other factors, such as fatigue or inattention, due to different ordering of the tests.

Two subtests of these tests, including basic writing skills were done monthly for 6 months before and after bifocal prescription to monitor for variability and progress in skills. A few sub-tests only were chosen, so that the monthly visits would be short and not too onerous and could be undertaken at the child's home. Otherwise, it would be more difficult to retain the participants in the study. Specific subtests were chosen for each child such that the child could at least perform some level on the test and so that a ceiling would not be reached, i.e. so that changes can be measured. For the writing test, the child was asked to write his/her name, write numbers from 1 to 10 and write any 10 letters.

The participant was videotaped during all these tests of visual perception and literacy skills. Thus, even if there is no improvement in raw scores, we may be able to measure change with the time taken to perform elements of the test and this will be a secondary outcome measure. The parents were asked to encourage their child to wear the glasses as much as possible, ideally for full time wear, but if that was not possible, at least for school classes and all near work. The parents and the participant were given a diary with smiley stickers to keep track of the number of hours of spectacle wear in the following categories: a) Wore glasses the whole day; b) Wore glasses for half a day; c) Wore glasses for near work/school; d) Glasses not worn at all. The teachers were requested to complete a questionnaire at the end of each 6 month period on the child's progress at school. This information was also obtained by asking for the child's termly progress report which will be compared along with other measures, before and after bifocal provision to observe for any progress.

3.4 Results

Of the 14 children and teenagers who were enrolled, 13 had a diagnosis of Trisomy 21 and one had Robertsonian translocation. Table 3-1 shows the demographic data and the number without spectacles at the first visit.

Table 3-1 Demographic data of participants

Gender	Number	Average age	Uncorrected at 1st visit	New Rx prescribed
Female	8	11.12± 3.2	4	6
Male	6	14.5 ± 3.4	3	5

At baseline 11 participants (79%) required a change in their distance prescription or were prescribed with new glasses. We observed that 6 participants (43%) had incorrect prescriptions. Thus 5 (36%) with a significant prescription (according to our criteria) had no glasses. Notably, 3 hyperopes between +1D and +2D had no previous prescription, one high myope (10.5D) was overcorrected by 1D, and 2 astigmats with 2.5DC and 4.5DC were uncorrected. The distribution of cycloplegic spherical refractive error is shown in Figure 3-2 as compared with the Down syndrome data of van Splunder (van Splunder et al. 2003) and data for normally-developing children from Gwiazda et al. (1993) and the distribution of cylinder power shown in Figure 3-3 is compared to normal data from Gwiazda et al. (Gwiazda et al. 1984). Anisometropia greater than 1D of mean sphere was present in two participants (14%). One participant was antimetropic.

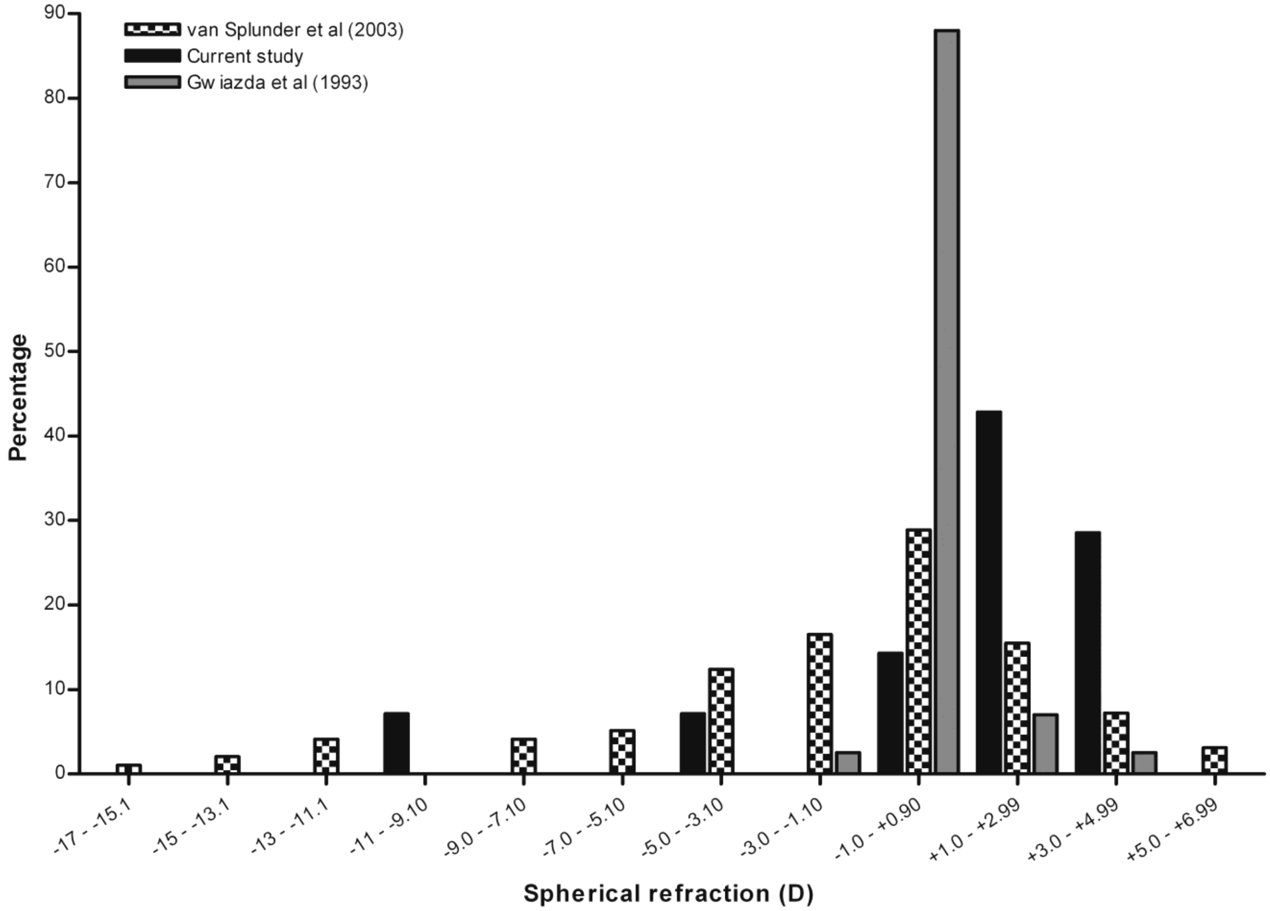


Figure 3-2: Distribution of cyloplegic spherical equivalents of the right eye compared with the estimations taken of spherical refractions from van Splunder et al.(van Splunder et al. 2003) **and Gwiazda et al.** (Gwiazda et al. 1993)

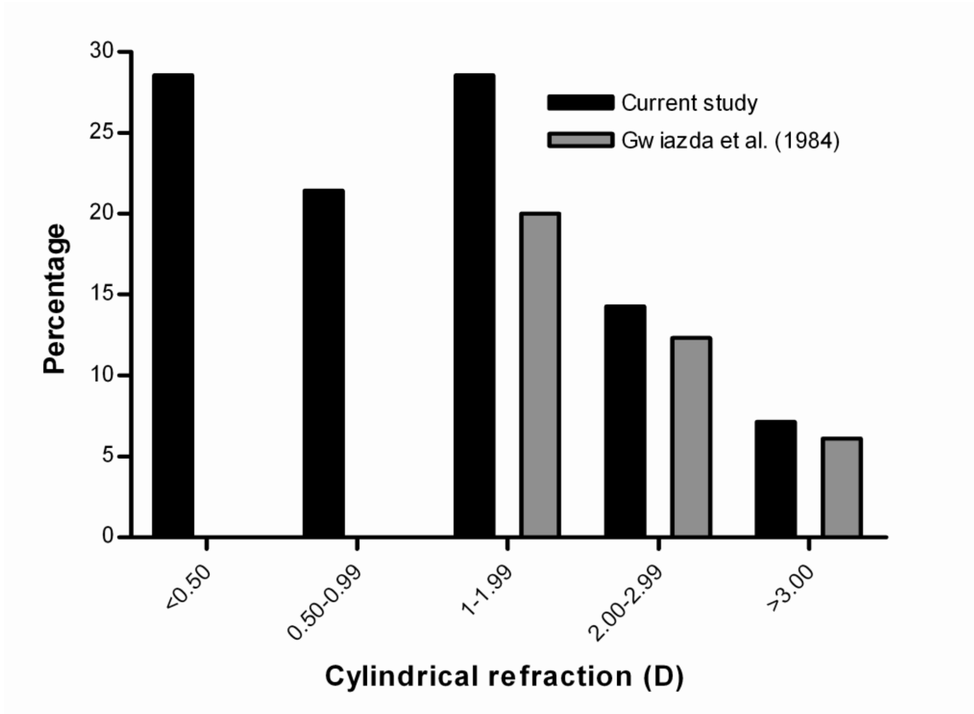


Figure 3-3: Distribution of cycloplegic cylindrical refraction compared with normal data of 6 year olds from Gwiazda et al.(Gwiazda et al. 1984)

All children were found to be compliant with either concentration of Cyclopentolate and there were no adverse reactions. The pupillary diameter in our study ranged from 5-9.5 mm in photopic conditions and 9-11 mm after pupillary dilation. Since accommodation is known to be reduced in young people with DS, it may be thought that a cycloplegic refraction is not necessary, as they will be more likely to relax their accommodation during dry retinoscopy. Therefore, we examined the difference in mean sphere, most hyperopic meridian and cylinder power between the dry and wet refractions for the right eye. The mean differences, and coefficients of agreement are shown in Table 3-2. This shows that overall there was not a large increase of plus power for either the most hyperopic meridian or the mean sphere in either eye (ranging from 0.1 to 0.18D). However, there was a fairly large variability between subjects, shown by the co-efficient of agreement being 1.5D in some cases. This tended to be influenced by one or two subjects in each case (not the same subject for each eye). The mean difference for cylinder power is even smaller, but again the co-efficient of agreement is notable, particularly in the right eye.

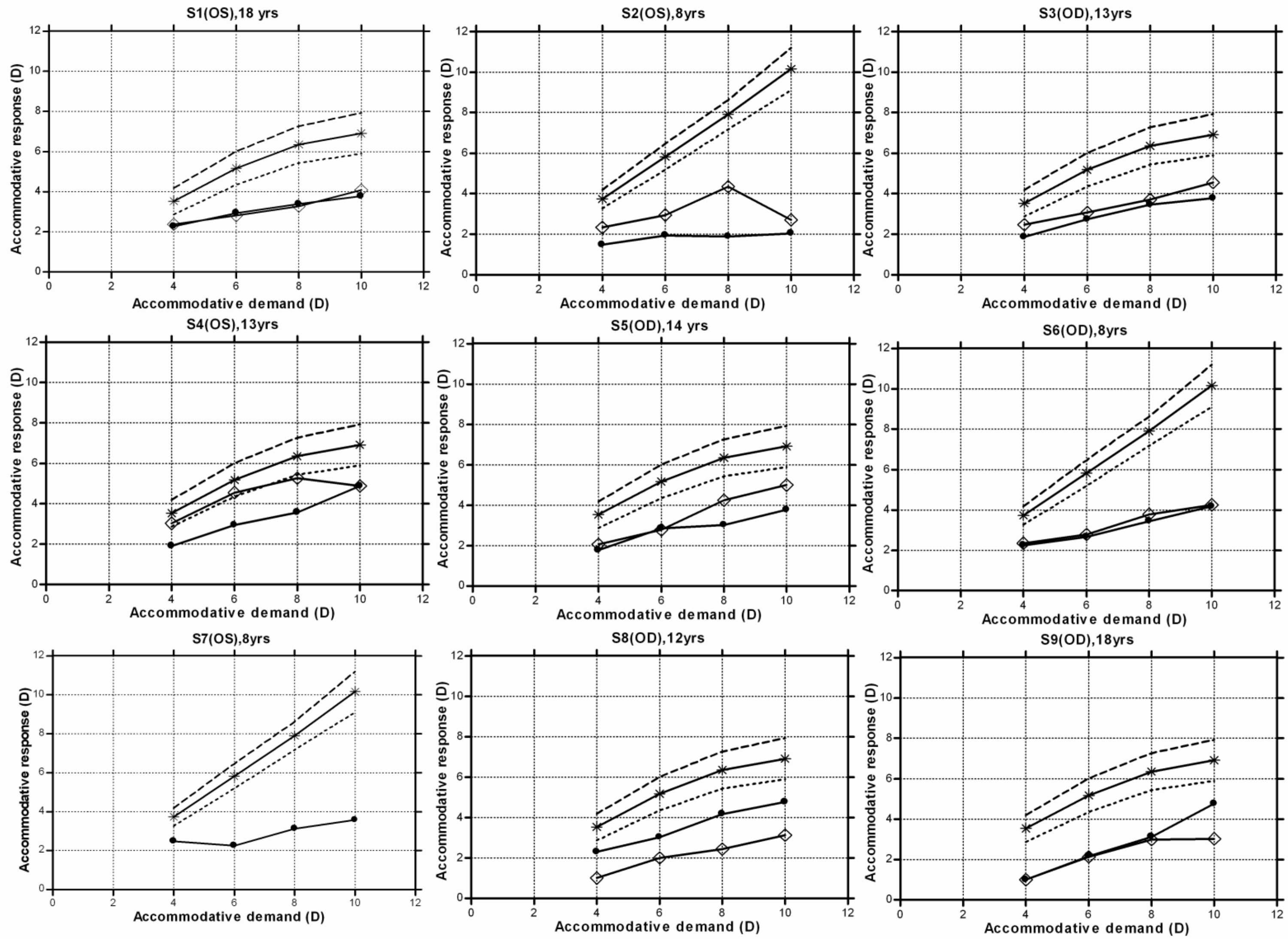
Table 3-2: Coefficients of agreement of cycloplegic minus non-cycloplegic refraction of all participants

	Most hyperopic meridian(D)		**Mean sphere (D)**		**Cylinder (D)**	
	OD	OS	OD	OS	OD	OS
Mean and SD of differences (D)	0.14 ±0.59	0.17 ±0.78	0.10 ±0.62	0.18 ±0.70	-0.09 ±0.77	0.02 ±0.41
Co-efficient of agreement (1.96 x SD)	1.15	1.53	1.22	1.37	1.51	0.81

Figure 3-4 shows the accommodative responses for all the subjects against the relevant age-related norms. The slope and the mean error of the accommodative response of each individual participant were compared with the normal 95% limits of the age-related controls for his/her age group, either 6-10 years or 11-26 years, from Leat and Mohr (Leat & Mohr. 2007). One hundred percent had a deficit in accommodation, either having reduced slope or increased mean error or both. This was true with both the habitual glasses and the new single vision glasses. For each of these age groups, a t-test showed that the difference between the DS subjects' accommodative response and that of the age-matched controls was significant in all cases (slope and mean error, 2 age groups, habitual and new glasses ($p<0.001$ allowing for Bonferroni correction) with the exception of the slope for the 11-26 year olds with new glasses.

These results include the data for accommodative responses for 8 and 10D, a working distance that is not used frequently in everyday life (although one subject had a reported habitual working distance of 15 cms). Young people with DS are more likely to use a 4D or perhaps a 6D distance. Therefore we also looked at the mean error of accommodation for 4 and 6D only (the slope cannot be reliably assessed for these 2 values, as a line would be drawn with only 2 data points). Again, all the subjects had a mean error that was outside of the normal age-related 95% range except for one subject whose error was normal with the new glasses.

Surprisingly, Cregg et al. (2001) noted that children with DS under-accommodated by a set amount, so that when changes of distance prescription were made, there was no change to the error of focus for close work. Therefore we examined our data to see if a similar pattern was found. Seven of the 14 subjects had a change in mean sphere of their prescription of 1D or more in the dominant eye or the eye which we chose for dynamic retinoscopy. Of these, all showed a change in accommodative response in the expected direction i.e. when more plus was added to the prescription, the lag decreased and when more minus was added the lag increased, although the change was not always of an amount equivalent to the change in prescription. Only one subject showed no change in lag, despite an increase in plus power in the new glasses.



S1(OS),18 yrs
S2(OS),8yrs
S3(OD),13yrs
S4(OS),13yrs
S5(OD),14 yrs
S6(OD),8yrs
S7(OS),8yrs
S8(OD),12yrs
S9(OD),18yrs
Accommodative response (D)
Accommodative demand (D)
0
2
4
6
8
10
12

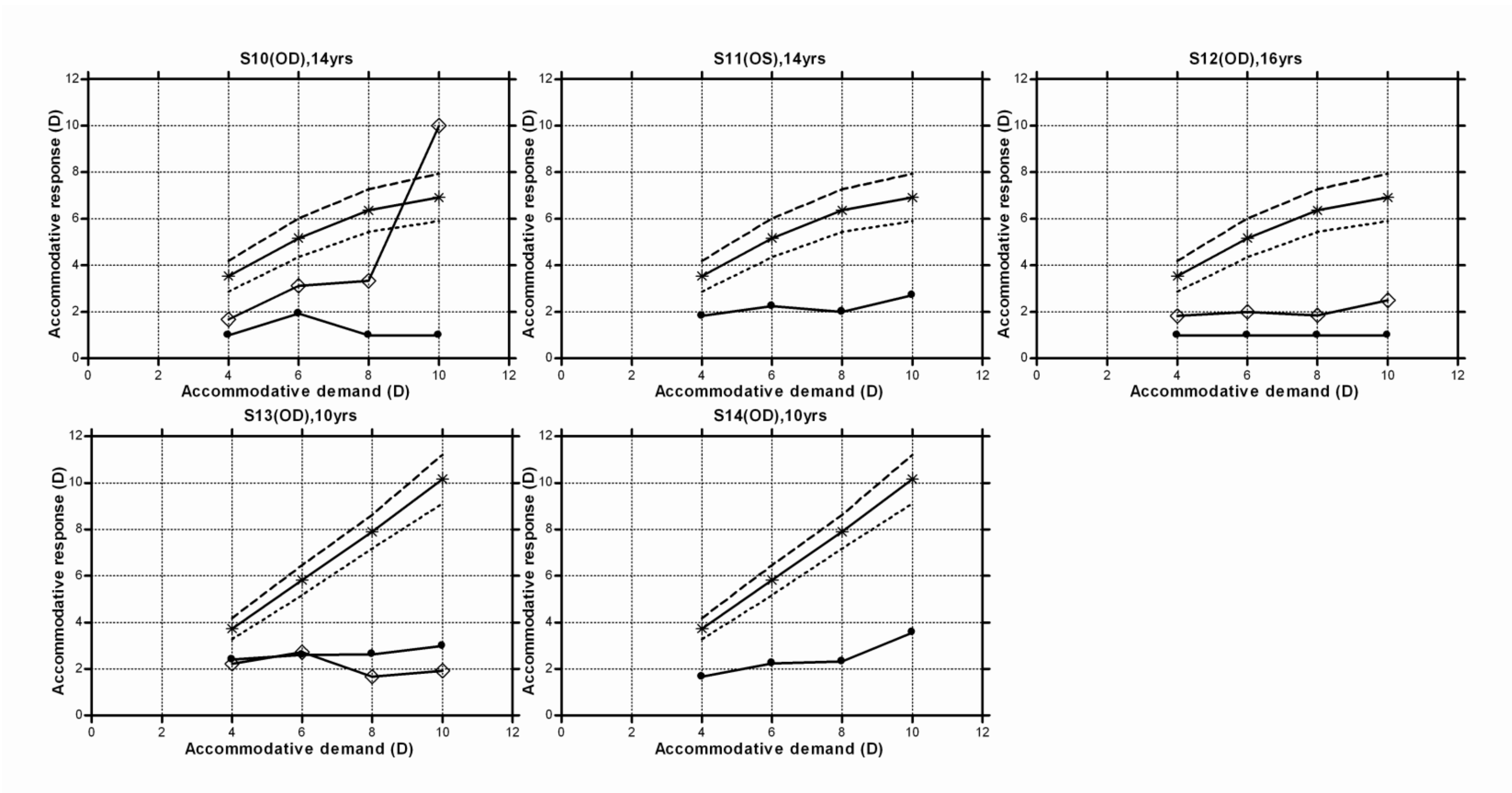


Figure 3-4: Accommodation response for each subject against the accommodative demand. Closed symbols indicate responses with habitual (either habitual glasses or without glasses) and open symbols indicate responses with new correction. The mean, upper and lower limits of the age-matched norms (Leat & Mohr. 2007) (6-10 years and 11-26 years) are plotted with an asterisk, small and large dashed lines respectively

Table 3-3: VA of all participants with change from habitual or no correction

			Change from habitual Rx					
Subject	**Distance VA (logMAR)**		**OD**		**OS**		**Near VA (logMAR)**	
	Old/no Rx	**New Rx**	**Sph (D)**	**Cyl (D)**	**Sph (D)**	**Cyl (D)**	**Old/no Rx**	**New Rx**
1	0.55	0.40	0	-1.75	-0.75	-0.25	0.375	0.6
2	0.50	0.475	+0.50	0	-0.5	-0.5	1	0.6
3	0.525	0.475	+0.75	0	+0.50	-0.50	0.6	0.475
4	0.30	0.40	+1.00	0	+1.00	-0.50	0.725	0.7
5	0.20	0.175	+0.5	-1.00	+0.75	-0.25	0.375	0.275
6	0.30	0.275	+2.00	-0.75	+1.75	-0.75	0.8	0.575
7	**0.25**	**0.25**	0	0	0	0	0.975	0.975
8	0.452	0.35	+1.50	-0.75	+1.00	-0.50	0.525	0.575
9	0.40	0.25	+0.25	-0.25	+0.25	-0.75	0.5	0.375
10	0.425	0.60	-1.00	-0.50	-0.75	0	0.875	0.65
11	**0.525**	**0.525**	0	0	0	0	0.325	0.325
12	0.375	0.275	+2.50	-1.00	+2.75	-1.00	0.375	0.2
13	0.525	0.60	-2.00	-3.50	+3.00	-3.00	0.975	0.675
14	**0.325**	**0.325**	0	0	0	0	0.6	0.6
Mean ± SD	*0.40 ± 0.11*	*0.42 ± 0.15*					*0.64 ± 0.25*	*0.54 ± 0.20*

*Bold numbers indicate those who did not receive a change in spectacles.

All of the subjects were able to perform a distance or near crowded Patti Pics matching VA test. The mean habitual near VA was poorer than distance VA (paired t-test, p=0.006), although this difference did not reach significance with the new glasses (paired t-test, p=0.07). There was no overall improvement of distance acuity with the new single vision glasses, but there was an improvement of near VA (paired t-test, p=0.015). The near VA with the old and new glasses is shown in Table 3-3 and the near visual acuities in Figure 3-5. The change in VA was generally as would be anticipated, assuming that the child exerts a constant amount of accommodation. This was particularly true for near VA, where most of those who were prescribed a net increase of plus power experienced improvement one or more lines of near VA and those who were prescribed more minus, a decrease. The corrected binocular distance VA (with new glasses) was 0.42+/-0.15 logMAR while binocular near VA was 0.54+/-0.20 and the distributions are shown in Figure 3-6. A paired t-test between corrected distance and near VA showed a borderline significance (p=0.067).

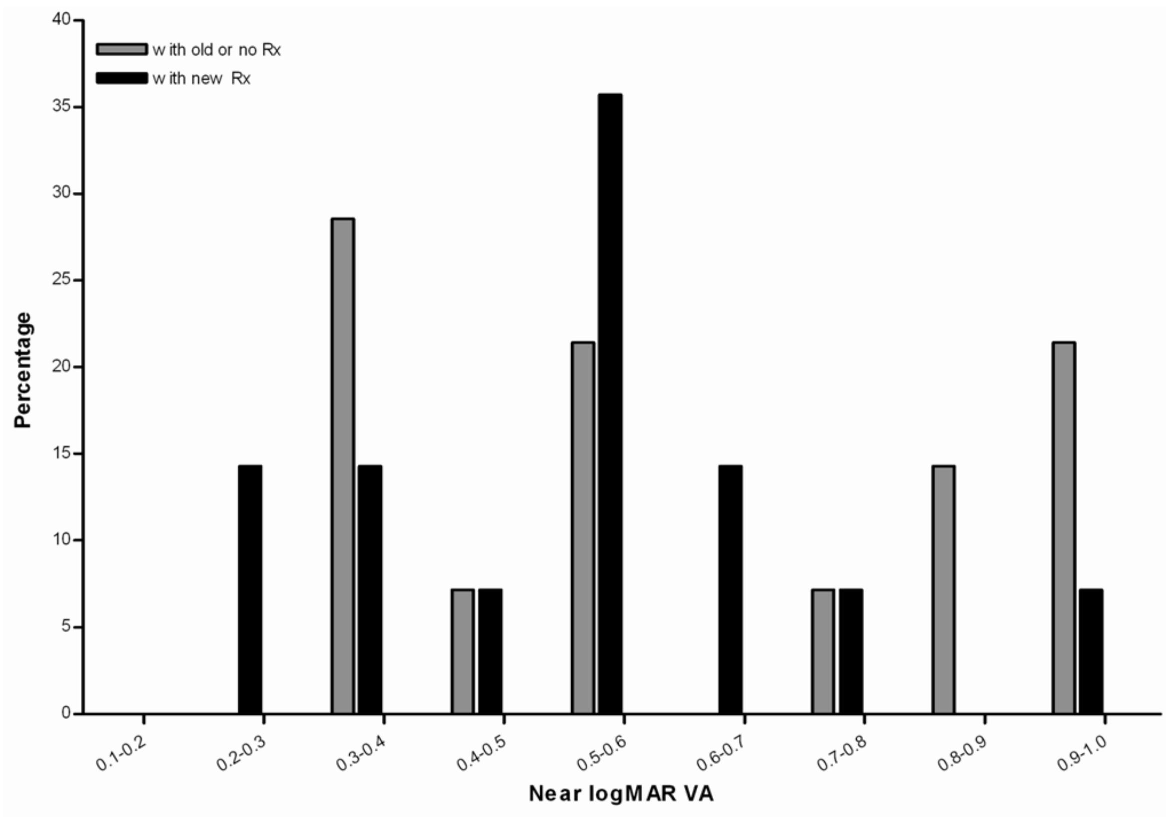


Figure 3-5: Distribution of near logMAR visual acuity with habitual (either habitual glasses or without glasses) and with new correction

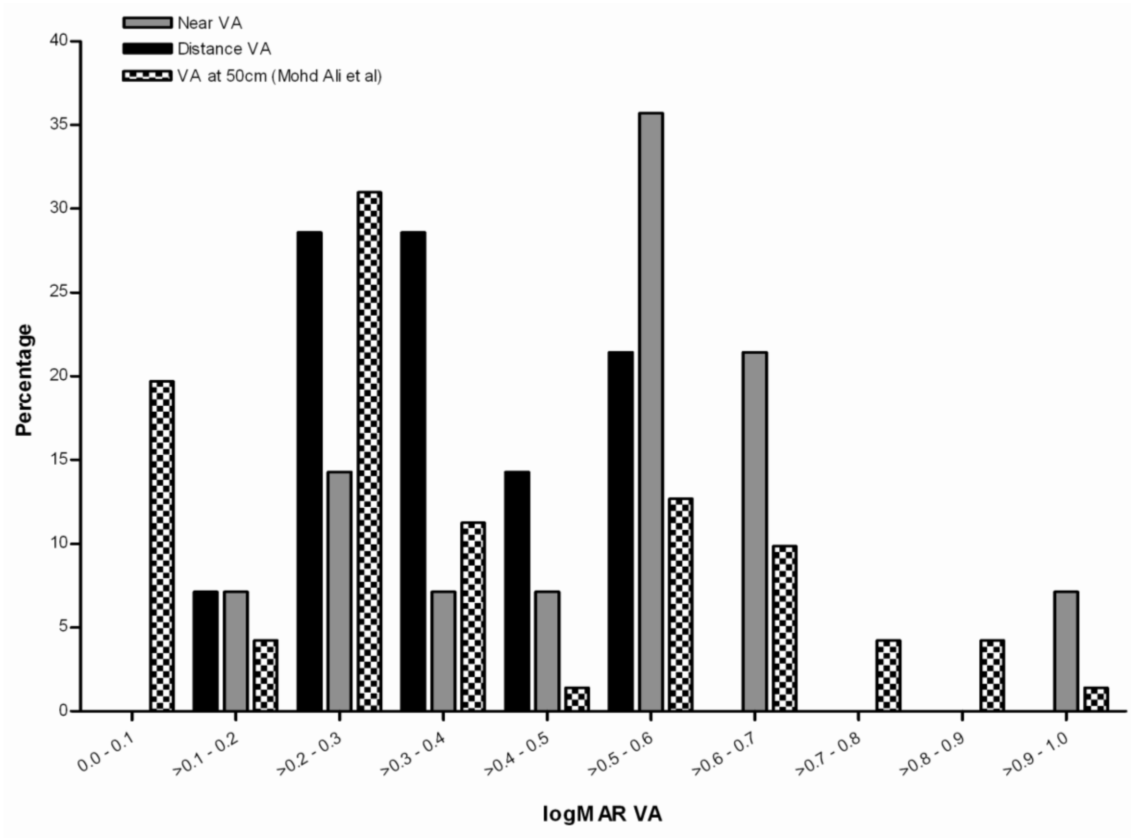


Figure 3-6 Distribution of distance and near binocular VAs compared to VA at 50cm in children with DS (Mohd-Ali et al. 2006)

3.5 Discussion

A greater prevalence of higher refractive error in people with DS is well established (Caputo et al. 1989, Cregg et al. 2003, Gardiner. 1967, Tsiaras et al. 1999, van Splunder et al. 2003, Woodhouse et al. 1997) although these studies differ on whether hyperopia or myopia is more common. Our data confirms the higher prevalence of hyperopia compared to van Splunder (2003) and the normal distribution of Gwiazda et al. (1993), as can be seen in Figure 3-2. Cregg et al. (2001) and Stewart et al. (2005)

also reported more hyperopia. There is also a tail in the high myopic end of the distribution. Thus we find a higher prevalence of both hyperopia and myopia. Higher levels of astigmatism have also been reported in people with DS ranging from 37-56.5 % (Gardiner. 1967, Stewart et al. 2005). In previous studies, high levels of anisometropia range from 11.6 % (Cregg et al. 2001) to 22% (Tsiaras et al. 1999) which is similar to the 14% found in the present study.

We found that 79% of the participants did not have an accurate refractive correction or had no glasses at all. It seems that this population is still not obtaining adequate basic eye care or that optometrists find it difficult to examine this population or are more reluctant to prescribe glasses. Lack of adequate eye care for patients with developmental delays has been discussed by several researchers and results in a high prevalence of uncorrected ocular disorders in individuals with intellectual disabilities (Liza-Sharmini et al. 2006, Mohd-Ali et al. 2006, Nagtzaam & Vink. 1998, Tsiaras et al. 1999, Woodhouse et al. 2000) . We have used a more stringent criteria for prescribing glasses, especially for hyperopes, than Woodhouse and colleagues. Applying our criteria for prescription of spectacles meant that all the participants except for one were prescribed distance glasses. Woodhouse and colleagues defined hyperopia as 3D or more (Cregg et al. 2001, Woodhouse et al. 1997) and suggest this criterion for prescribing glasses. We would argue that this is rather a high criterion for prescribing in clinical practice for children with DS and even for those without. The definition of hyperopia is variable, ranging from 0.5 to 3D. (Atkinson et al. 2002, Choi et al. 1995, Jones et al. 2005, Kleinstein et al. 2003, Laatikainen & Erkkila. 1980, Rosner & Rosner. 1997, Shankar et al. 2007, Zadnik et al. 2003). There is also lack of agreement and few guidelines on prescribing for hyperopia in children, especially low and moderate hyperopes (Cotter. 2007, Filips. 2008, Robaei et al. 2006) and it has often been assumed that they can “focus through” their hyperopia (Robaei et al. 2006). However, there is an increasing volume of information that indicates a link between even moderate levels of hyperopia and poorer reading or visual perceptual skills (Atkinson et al. 2002, Rosner & Rosner. 1997, Shankar et al. 2007). The question is what level of hyperopia should be prescribed?

Based on the literature, Leat et al.(Leat et al. 1999) have recommended that a prescription for hyperopia >2D from the age of 4 years should be considered and for lower amounts of hyperopia in the school years if there are symptoms. The Orinda study criteria for refractive correction were +1.00D or more of hyperopia and 0.75D or more of astigmatism and Dwyer and Wick (1995) showed improvements in accommodation and vergence in around 60% of school age patients after correcting refractive error according to the Orinda criteria. Cotter. (2007) suggests considering a prescription for 1.25D or more of hyperopia in the school years. The reduced accommodation in children with DS indicates prescribing at least for these degrees of hyperopia. Additionally, the fact that there was a net increase in near VA with the new glasses, for participants who were prescribed increased plus power (all of whom had deficits of accommodation), further emphasizes the need to prescribe for moderate or low levels of hyperopia, which we would define as >1D.

We observed no adverse reactions to Cyclopentolate. There is some suggestion in the literature of increase heart rate with injections of atropine (Harris & Goodman. 1968) and a greater degree and duration of dilation with topical instillation of atropine (Berg et al. 1959, Priest. 1960) and Tropicamide (Sacks & Smith. 1989), and therefore some authors have suggested that cycloplegics in general are contra-indicated in DS (Barnard & Edgar. 1996). However, there is no documented evidence of increased adverse reactions to topical Cyclopentolate (North & Kelly. 1987), although there may be longer or greater pupil dilation (Doughty. 2001). We found an average dilated pupil size of 10 mm in our study population. This compares to reports in Caucasians of 7mm pupil diameters after the instillation of 2 drops of 0.5% Cyclopentolate (Gordon & Ehrenberg. 1954). Thus we did obtain greater dilation than in children without DS, but there is no evidence of increased risk of adverse reactions. Woodhouse et al. (1997) also used Cyclopentolate on 14 children with DS and made no mention of adverse effects. We see no reason why Cyclopentolate should not be used in children with DS in the same manner and with the same routine precautions as for typically developing children.

However, because of the reduced accommodation it might be thought that cycloplegic refraction is less necessary in these children. We find that the increase of positive power with cycloplegic was not significant and was not as great as in children without DS, which ranges between 0.6D more plus power to 1.18D with Cyclopentolate (Egashira et al. 1993, Fotedar et al. 2007, Suryakumar & Bobier. 2003). Woodhouse et al. (1997) also found a smaller average difference between Mohindra and cycloplegic retinoscopy in children and infants with DS. However, our co-efficients of agreement ranged between 0.8 to 1.52D which are larger than in other studies. Repeatability of subjective and objective refraction has typically been found to be less than 0.5D in adults (Rosenfield et al. 1991, Goss & Zhai. 1994). Zadnik et al.(1992) found the repeatability of non-cycloplegic and cycloplegic retinoscopy in a group of pre-presbyopic adults to be 0.77 and 0.94D respectively. In children, Suryakumar & Bobier (2003) and Egashira et al.(1993) found a co-efficient of agreement of 0.96D and 0.99D respectively between non-cycloplegic and cycloplegic retinoscopy. We find higher values for the coefficient of agreement compared to all these studies, which may be because of generally poorer co-operation. Thus it seems that, although cycloplegic might not be needed to find latent hyperopia in children with DS, it may still be indicated for accurate results. Because of the greater dilation with Cyclopentolate in these children, it is important for the clinician to remember to judge his/her estimation of the retinoscopy reflex on the central zone.

This study also confirms the high prevalence of reduced accommodation found in other studies. In fact, we find that 100% of the subjects had deficits of accommodation. Woodhouse et al.(1993), in the original study, found 80%, based on the estimated amplitude of accommodation. This difference may be due to the way that we have analyzed the accommodative response in the current study (in terms of slope and mean error). Cregg et al.(2001) and Stewart et al.(2007) used the accommodative error index (AEI) as suggested first by Chauhan and Charman (1995), which describes the discrepancy between the measured accommodative response for stimuli at different distances and the accommodative demand. Stewart et al.(2007) found 76.3% had reduced accommodation, based on a definition of a lag ≥0.75D on at least two testing distances and on at least two occasions. Thus their criterion of normal accommodation was probably stricter than ours. If we apply their criteria, we still find that all our subjects exhibited abnormal accommodation. Cregg et al (2001) discuss whether reduced accommodation might be due to smaller pupils, resulting in a greater depth of focus. We do not find any evidence of smaller pupils in children with DS. In fact, they may tend to have larger pupils than children of the same age. We also do not find the same link with high hyperopia (they found more hyperopes had reduced accommodation). Eight of our subjects were not high hyperopes (≥3D spherical equivalent) yet all still exhibited reduced accommodation. These differences may be due to differences in the actual populations or due to sampling. We have a smaller number of subjects in the current study.

It was also reported by Cregg et al.(2001) that hyperopes tend to under-accommodate by a set amount, i.e. when hyperopia is corrected they still under-accommodate by the same amount. We did not observe this trend in our group. From Figure 3-4 we see the various patterns of accommodative responses when the child's correction was increased or decreased. Only one participant who had a larger change of prescription maintained the same lag with the old and the new spectacles. The others seemed to exert a more constant amount of accommodation. Also, the finding of improved near VA with an increase in plus power of the spectacles indicates that the child is not simply changing his/her accommodation to maintain a constant lag, but is exerting a more constant amount of accommodation and thus there is less defocus at near.

Our findings indicate that it is difficult to predict which children might benefit from a bifocal correction until the correct distance spectacles are worn. This is because some participants showed a change in their lag of accommodation once correct single vision glasses were prescribed. Clinically, we would currently recommend this management - prescribe the full distance prescription for a period of a few months and then re-measure the accommodative response before deciding on a bifocal prescription. The difficult case to deal with here would be the myope, who would be worse off for near work with a full correction and might reject the new glasses. In this case a reduced prescription might be wise. Our findings indicate that all the current participants will benefit from bifocals. We anticipate that bifocal prescription in this group of children will bring their focus to within the normal range for near work, further improve their near VA and thus facilitate their near work, which may translate into educational gains. The progress of this group will be assessed by following these children every month for 6 months before and after bifocal prescription, when basic reading and writing skills will be assessed.

3.6 Conclusion

These data confirm previous findings of a high prevalence of refractive errors, reduced visual acuity and reduced accommodation. It also indicates that young people with DS are still not being prescribed a full refractive correction and that near VA (with the habitual glasses) is reduced compared to distance VA. Prescribing for distance refractive error is expected to improve VA and correcting moderate hyperopia (>1D) improved near VA. The present results indicate that all subjects in this study may benefit from bifocal provision.

4 Early literacy and visual - perceptual skills in children with Down Syndrome before bifocal prescription

This chapter is in submission as follows:

Krithika Nandakumar,Mary Ann Evans , Susan J Leat. A Short-Term Longitudinal Study of Visual Perceptual Skills and Reading Ability in Children with Down Syndrome before Bifocal Prescription (Bifocals in Down Syndrome Study – BiDS) in *International Journal of Disability, Development and Education (Under review)*

Roles

	Concept / Design	Recruitment	Acquisition of data	Analysis	Write-up / publication
Nandakumar	Design of the number, writing tests, and Dolch test and compliance materials	Y	Y	Analysis of the data	Write-up
Evans	Suggestions in the choice of sub-tests and order of testing	-	-	Suggestions for analysis	Editing
Leat	Suggestions for administration of the tests and suggestion on written tests	-	-	Suggestions and direction	Editing

4.1 Overview

Disorders of the visual system, including reduced distant and near vision, are common in individuals with Down syndrome, and refractive errors frequently remain uncorrected. This study assessed the reading, printing, visual perceptual, and visual-motor integration skills of a sample of 14 children and young adults with Down syndrome aged 8 to 18 after they had been prescribed correct distance (single vision) glasses. A battery of tests was administered at baseline and selected reading subtests were administered in monthly probes for 6 months. This was in order to examine whether any natural progression occurred before providing bifocals to correct for the reduced near focussing ability (accommodation), constituting a second stage of the study. Overall the participants performed at an age-equivalent between 3-10 years. Chronological age and receptive vocabulary were significantly correlated with visual motor integration and word identification but not visual perception. Within the domain of visual perception, visual discrimination was a relative area of strength. The monthly subtests of reading and printing showed that for the majority of participants, there was no significant progression in the reading and the printing tasks over the 6 month period. The next phase of the study will discuss the impact of bifocals on the skills measured above and will be discussed elsewhere.

Keywords: Down syndrome, bifocals, literacy, writing, visual perceptual skills, reading

4.2 Introduction

Down syndrome (DS) is the most common cause of intellectual disability in individuals, according to the National Institute of Health, (2008) with a prevalence of 1 in 800 live births. Until the last few decades, it was less recognised that children with DS were capable of achieving primary reading and writing skills. In one of the earlier studies looking at academic achievement in this population, Hayden and Dmitriev (Hayden & Dmitriev. 1975) devised an educational program and showed that children with DS could be taught to read. A large number of studies in recent years have shown that children with DS are capable of learning to decode single words (Boudreau. 2002, Byrne et al. 2002, Lorenz et al. 1985) and many can achieve functional levels of reading (Casey et al. 1988, Laws. 2000, Sloper et al. 1990) and foundational skills such as writing and learning numbers (Duffen. 1976, Shepperdson. 1994, Turner & Alborz. 2003).

In a study by Lorenz et al. (1985), a series of questions was administered to teachers about reading-related skills, to understand the very early pre-reading and reading abilities of 58 children with Down syndrome aged 5 to 7 years. Their results showed that 19, 32 and 44 percent of the 5, 6 and 7 year olds respectively were able to read 5 to 10 words, and that the performance in each of the age groups depended on factors such as early instruction, parental attitudes, and the type of academic skills that were taught at different ages. It has been postulated that many children with DS are capable of "functional reading" by adulthood, which is interpreted as the ability to read at an 8-9 year old level (Bochner. 2001, Buckley. 2001). Bochner (2001) reported that

40-57% of young adults with DS can read books, magazines, menus, and recipes and that many will continue to develop reading abilities if given an opportunity to learn. Lorenz et al (1985) and Laws (2000) reported that placing children with DS in mainstream education resulted in important gains such as development of language and memory and development of higher reading abilities compared to children in special needs schools.

In comparison to the increasing volume of literature on academic/literacy skills in DS (Byrne et al. 2002, Fowler. 1995, Kay-Raining Bird et al. 2000, Kay-Raining Bird et al. 2008, Lorenz et al. 1985, Shepperdson. 1994, Sloper et al. 1990), there has been little focus on the development of writing skills in this population (Kay-Raining Bird et al. 2008). The few studies that describe the characteristics of reading and writing skills, and how they are related, are mostly in typically developing (TD) individuals (Berninger et al. 1992, Berninger et al. 1994, Berninger et al. 2002, Swanson & Berninger. 1996). In a recent study by Kay-Raining Bird et al. (2008) written narratives were studied for the first time in the DS population.

In addition to the school curriculum, several factors may at least weakly influence the literacy attainments of children with Down’s syndrome, including home experiences and phonological awareness. These factors are also relevant in the development of literacy skill in typically developing children (Snowling et al. 2008). One factor that might be especially relevant to reading and writing achievement in the DS population is vision. Most individuals with DS have a greater prevalence of disorders of the visual system and present with atypical ocular findings (Caputo et al. 1989, da Cunha & Moreira. 2004, Shapiro & France. 1985). These include reduced visual acuity (detail vision), strabismus (misaligned eyes), nystagmus (involuntary jerky movements of the eyes), keratoconus (steep corneas), cataracts and hyperplasia of the iris (Catalano. 1992, Pueschel & Gieswein. 1993). High refractive errors are common, yet persons with DS often are not often prescribed with glasses or do not have accurate glasses. Nandakumar and Leat (2009) observed that that 79% of participants with DS in the present cohort required new distance glasses, 55% requiring a change of current glasses and 45% requiring spectacles for the first time. Reduced accommodation (the ability to focus for near work), is another common manifestation in children with DS, being present in 55-100% of children with DS (Haugen et al. 2001, Haugen & Hovding. 2001, Lindstedt. 1983, Nandakumar & Leat. 2009, Woodhouse et al. 1993). Reduced accommodation will result in hyperopic defocus for close tasks, which is related to less well developed literacy skills in typically developing preschoolers (Shankar et al. 2007) and less well developed visual-motor skills (Atkinson et al. 2007). Reduced accommodation is correctable with lenses, such as reading glasses or bifocals, which would improve visual acuity and enable better focussing for close tasks.

The present study assessed the reading, printing, and visual perceptual skills of a sample of children ages 8 to 18 with DS after they had been prescribed correct distance (single vision) glasses. The description of the optometric baseline visual findings are given in Nandakumar and Leat (2009). The current paper describes the non-optometric baseline results for standardised tests of visual perceptual, visual motor and early literacy skills and an informal measure of printing ability. To our knowledge there are very few studies that have examined the progression of these skills in a population with DS over time. Hence we have monitored their progress using a subset of these tests of early literacy plus a test of printing over a 6 month period after the point of distance vision correction (where applicable). This was in order to examine their development before providing bifocals, constituting the second stage of the study to be described in a later report.

4.3 Methods

4.3.1 Participants:

Fourteen children (8 females, 6 males) aged 8-18 attending regular schooling were recruited into the study. Thirteen were previously diagnosed with Trisomy 21 and the fourteenth had a diagnosis of Robertsonian translocation. During recruitment, 2 subjects were excluded because of perceived inability to cooperate, judged from their optometric record (according to which they were not able to identify or match letters or shapes), or from parental reports. The average age of the final group of participants was 12.7±3.6. With the exception of 2 participants who were in a developmental education plan and 4 where no information on their educational plan was available, all other participants were enrolled in congregated classes in mainstream schools with educational assistance. Some may have had a portion of segregated teaching or withdrawal assistance. Exact data on this was known for a few participants, the number of hours of segregated/withdrawal teaching for word reading, phonological awareness, fine and gross motor skills ranging from 6.2 – 22.5 hours per week. One participant dropped out of the study during the follow up period and hence only the data for her baseline visit were included in the analysis.

The study participants were mostly recruited from the Paediatric and Special Needs Clinic, School of Optometry at the University of Waterloo, the local Down Syndrome Society, and advertisements in local newspapers and from Public and Catholic schools in the Kitchener-Waterloo region. The study was approved by the Office of Research Ethics at the University of Waterloo and all aspects of the study conformed to the requirements of the Declaration of Helsinki.

The inclusion criteria were as follows:

- Diagnosed with Down Syndrome
- Age between 5 years to 21 years, i.e. school age (note that children with Down syndrome often stay in school after the usual school leaving age - thus the higher age limit)
- Engaged in academic instruction, at least pre-literacy learning of letters
- Speaks English as first language
- Not already wearing bifocals
- Be verbal or able to understand instructions
- No significant eye conditions, such as keratoconus or cataract
- Not diagnosed with other neurological, sensory or behavioural disorders such as autism, microcephaly or significant hearing loss.

On recruitment into the study, all participants underwent a complete eye examination including accommodative ability and distance and near visual acuity. The distance and near visual acuity (level of detail vision) was measured with a standardized method, Patti pics symbols (Precision Vision. 2008) . Eleven of the 14 participants required new single vision distance lenses (either a change from a previous prescription or glasses for the first time). After 1-2 weeks with these lenses, the participants were seen for their baseline visit, when visual acuity through the new glasses was measured. Mean near logMAR visual acuity with the new glasses improved to 0.54±0.20 (equivalent to 6/21 or 20/70) from 0.64±0.25 (equivalent of 6/26 or 20/87). There was no significant improvement in the average distance visual acuity. The visual findings at the baseline and with the new distance vision glasses are reported in more detail in Nandakumar and Leat (Nandakumar & Leat. 2009). A copy of the school Individual Educational Plan (IEP) of participants was requested and kept in their files to be used later to compare against their progress. This helped us understand the current performance abilities of each child and the type and amount of instruction that the child was being given.

4.3.2 Tests administered:

A full battery of visual perceptual and literacy skills was administered at baseline. Each literacy or visual perception test was chosen based on the criteria that a) It should be a standardised and known test, b) Ideally more than 1 version of the test should be available, so that different versions could be used for the baseline and the monthly measures to reduce repetition effects, c) It should be a test for which there is previous data indicating that a majority of children with DS can perform the test.

The following tests were chosen and were administered at the baseline visit in the following order:

- The Beery™ Visual Motor Integration (BVMI) (Beery. 1997)
- Woodcock Reading Mastery Test (WRMT) *Subtests:* Letter Identification (LI), Word Identification (WI), Word attack (WA) (Form G) (Woodcock.1998)
- The Test For Visual Perceptual Skills (TVPS-R) *Subtests:* Visual Discrimination, Visual Form Constancy , Visual Closure (Gardiner. 1967)
- Dolch sight word list -1 (2008)
- For children unable to read any letters, a list of numbers was devised and used instead of the Dolch sight words
- Peabody Picture Vocabulary Test (PPVT) III (Dunn & Dunn. 1997)

The BVMI, WRMT, PPVT and TVPS are all standardised tests and the former three have been used previously in individuals with DS and quite reasonable performance has been shown (Bochner. 2001, Chapman. 2006, Cupples & Iacono. 2000, Fidler et al. 2005, Kay-Raining Bird et al. 2000, Kay-Raining Bird et al. 2008, Klein. 1999, Miolo et al. 2005, Spano et al. 1999, Wang et al. 1995). Unless otherwise stated, the standardised tests were administered according to the standard protocols, including the usual stopping rules. Variation from the standard procedures was generally in the starting point for the test. The BVMI, WRMT (LI, WI and WA) and TVPS-R were started on the first page for all participants.

Beery™ VMI. This test has a developmental sequence of geometric forms to be copied with a pencil on the test sheets. The short-form version was used and it has 21 items in increasing level of complexity. The examiner draws the first few simple shapes for the participants to copy and get familiarized with the test. The test identifies the individual's ability to integrate their visual and motor abilities (eye-hand coordination). Ages in the norming sample range from 2 to 18.

Letter Identification. The Letter Identification (LI) task of the WRMT measured the child's ability to name letters of the alphabet when presented in either upper or lower cases or in varying typefaces of increasing difficulty. Participants in the norming sample for the LI are in the range of 5 years-10 years 4 months.

Word Identification. This task measured the individual's ability to identify isolated English words ranging from high to low frequency of occurrence (Catts et al. 1999, Catts et al. 2002) and in increasing difficulty. Since this is a good indicator of reading ability, we used this test along with the Dolch sight words as an indicator of progress of reading ability. The norming sample range is from 5-33 years of age

Word Attack. The Word Attack subtest measured the individual's ability to read pronounceable nonsense words or pseudo words, i.e. non-words that follow English spelling patterns. This test measures the child's ability to use phonic and structural analysis to pronounce words with which they are unfamiliar. Participants in the norming sample ranged from 5 years – 18 years, 6 months.

For all the WRMT subtests, the test pages were enlarged by photocopying to allow for the reduced visual acuity for children with DS. Typical values of visual acuity for children with DS were taken from clinical records and the size of the letters and words in the stimulus pages were increased by a factor of 2.

Test of Visual Perceptual Skills-Revised subtests: Visual Discrimination. This subtest measured the individual's ability to match or determine exact characteristics of the form that is presented with one of the four similar forms. Visual Form Constancy measured the individual's ability to see a form and be able to find the form amongst the other forms presented even though they may be of a different size (larger, smaller), rotated, reversed or even hidden amongst other forms. Lastly, Visual Closure determined the individual's ability to determine from four incomplete forms presented, the one form that is the same as a completed form. The norming sample includes participants aged 4 years – 12 years 11 months.

Dolch sight words 1 and 2. This test consists of a set of high frequency words used in the English language and are therefore usually recognised by sight, grouped by reading level. This is not a standardized test but is commonly used as a teaching tool in schools for children at the elementary level. It has five levels: pre-primer, primer, first, second and third grade level. For each level of words, 8 words were randomly selected and printed in Arial 18 font size in two columns on each page, such that the difficulty of the test increased from page to page. Two sets of Dolch words were created, the first set being used for the full battery and the other for the monthly probes. The test was administered similarly to the WRMT. The test was stopped when 6 errors were made in a page and the page was completed. The score was calculated by the total number of words read correctly on each page and the total score was the sum of all page scores.

List of numbers. The number list had 9 numbers, arranged in random order in three columns on each page. Again, the difficulty on each page increased, by using numbers 1-9 on the first page, 10-19 on the second, 20-29 on the third and so on. An Arial font size 22 was used for the numbers. The test was stopped when 7 errors or more were made in a page and the page was completed. The score was calculated by taking the sum of correct scores on each page. Again, two versions were created, one used at the base line and the other for the monthly visits.

PPVT-III. This test measures receptive vocabulary in individuals over a wide age range. The participant is shown four pictures while the examiner says a single stimulus word. The participant verbally or non-verbally indicates the picture that best represents the stimulus word. It is positively correlated with verbal intelligence and was used to control for the mental age of the child. The parents were asked about their understanding of the approximate reading age of the child, according to their observation and school records. We used this to determine the starting point for the PPVT-III, for which the same criteria for basal and ceiling sets were used as in typically developing children.

4.3.3 Test Procedure

The order of testing at baseline was chosen such that the more interesting tests for the child were interleaved with the comparatively demanding tests. The BVMI was a good test for the child to start with and to develop rapport as most children like to trace or draw, and this would make them comfortable with the examiner and the testing. The WRMT was administered next so as not to leave it too late in the sequence, as it is more demanding. The TVPS-R was administered third to break up the reading tests, followed by the Dolch sight words or the list of numbers (depending on the ability of the child – those who could not perform the Dolch, were administered the numbers). The PPVT was done last (and only at baseline) since it was not one of our outcome measures and because it is more time consuming. Throughout the entire session(s), the children were given breaks when needed to minimise the effects of fatigue or boredom.

All the tests were administered by the first author in a quiet room designated for the study, under well lit conditions. The participant and the examiner were seated in comfortable chairs and a flat-topped desk of appropriate height for the participants was used. For younger children, a set of the child-sized tables and chairs were used for the child to be seated comfortably. The main light source in the room was from common white ceiling fluorescent lights, adequate enough to illuminate the test items and not cause any unwanted glare.

For the monthly visits (probes), a few subtests were chosen to be administered every month for 6 months. These probes acted as indicators of whether the child's performance improved with his/her correction. For each child, two reading sub-tests and two printing tests were chosen according to the child's ability to avoid the possibility of floor or ceiling effects. Thus not all children were given the same tests. The reduced battery also ensured that the monthly visits were short and not too tedious for the child and could be undertaken at the child's home if necessary. Retention of participants was a key factor as the total duration of the study, including the later phase of bifocal correction, would require almost a 14 month commitment from the parents and families. Although most of the monthly visits took place in the School of Optometry under the same circumstances as the full battery of tests, in some cases they were done at the child's home, when for various reasons, it was difficult for the child to attend the school. Similar testing conditions and surroundings were maintained, so that there were no distractions or interferences from other children or sources. Also, although it was intended that these probe visits should be monthly, due to illness, family circumstances etc it was not possible to maintain exact monthly intervals in all cases.

The monthly subtests were done in the following order:

- The child was asked to write his/her name (bold lined paper was provided). Note that we did not specify whether the child should print or use cursive writing, but all the participants printed, so we have referred to this task as printing throughout.
- The WRMT Form H ‑ Word Identification 2 (Woodcock.1998)
- The child was asked to print any ten letters and any ten numbers
- The Dolch sight words (2008) 2 or the number list 2

At the end of 6 months with single vision lenses, questionnaires were sent out to the mainstream classroom teacher of each child to gain information about the usual reading instruction provided to the child. The questions included the number of hours of regular class reading, number of hours with teaching assistant support, number of hours with indirect support, hours with withdrawal support, hours in a segregated class (if any). The teachers were also asked to quantify the number of hours of instruction per week that the child received for early literacy skills (letter recognition, alphabetics), reading instruction, writing, and arithmetic instruction and to estimate the child's glasses wearing time for each of the above mentioned activities. The children were given strong encouragement over the months to keep their glasses on with constant reminders, if required.

4.3.4 Analyses

The results for the norm-referenced tests were initially retained in the raw score form for analysis, but the age equivalents were also determined. Descriptive statistics were used for the full battery of tests. The probes were plotted against time (monthly visit) for each child against the age-matched expected scores and against scores for a typically developing child with the same starting score as each DS participant. The time scales were taken based on real time, i.e. if the participant missed a visit on a certain month, it was considered as a missed visit and would be seen as a gap in the data. Repeated measures ANOVA were used to determine any change from first to last testing. Correlation coefficients were computed against age for all literacy (except Word Attack and numbers), perceptual and writing measures. Paired t-tests or sign tests were used to analyze differences in age-equivalent scores between the tests administered at the baseline. The significance level was set to $p<0.05$.

The printing tasks were analysed by measuring the size and position of the written letters. The size was measured by the mean height of the middle zone of each letter (i.e. not including the ascenders and decenders, being the height of lower case "x") for each participant. The position was measured by the mean distance from the bottom of the middle zone of each letter (which should be placed on the base line) to the base line for each letter. This was given a negative score in millimetres if the letter position was below the line and a positive score if above the line, as shown in Figure 4-1. Means and standard deviation for the letter size and position measures were calculated for each partipant and plotted over time. The standard deviation gives a measure of the total amount of variation in letter position. These were plotted over time. Since negative and positive position scores would cancel when calculating the mean, the analysis of the mean letter positions was undertaken using the absolute values i.e. the removing the negative signs. Repeated measures ANOVA was used to analyze changes in letter size or position over the 6 visits.

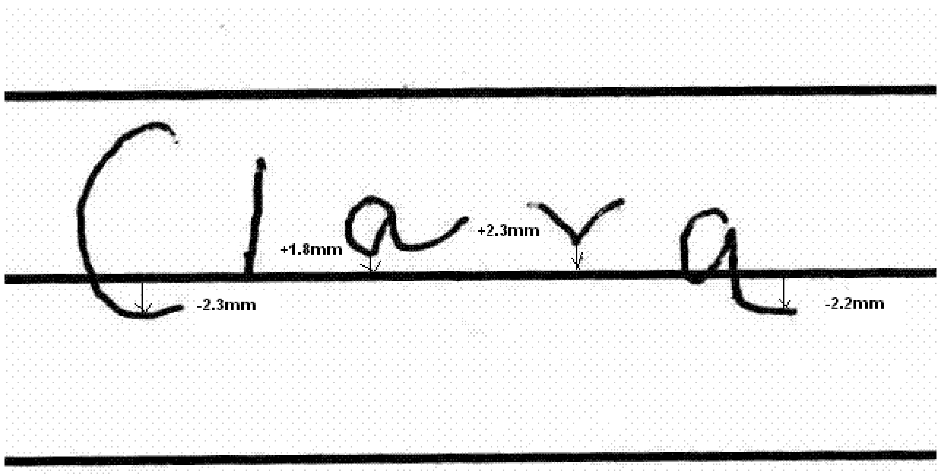


Figure 4-1: Analysis of the letter printing task

4.4 Results

4.4.1 Hours of instruction:

From the questionnaires sent to the teachers we received six responses regarding the number of hours of assisted teaching that the children received. For these participants we calculated the average number of hours per week. The averages are as follows for the respective activities: a) early literacy skills (e.g., letter recognition, alphabetics): 2.08 ± 2.33, b) reading instruction 2.22 ± 2.03, c) writing instruction: 2.18± 2.07 and d) arithmetic instruction 3.07± 1.77.

4.4.2 Baseline visit:

Histograms of the raw scores for all 14 subjects on the full battery of tests done at baseline are shown in Figure 4-2 a-i and descriptive statistics are provided in Table 4-1. These are scores obtained 1-2 weeks after receiving new single vision lenses (if prescribed). All subjects were able to score on the BVMI and the PPVT III. All but one were able to do the WRMT letter identification subtest. Nine and 8 participants respectively were able to identify some words in the WRMT Word Identification and Dolch sight word list. Five participants were unable to identify any words but were able to identify some numbers. Four participants were able to do some items of the WRMT Word Attack subtest. Twelve participants could score on the Visual Discrimination and Visual Form Constancy subset of the TVPS-R. Finally, 11 of them could obtain scores on the Visual Closure task. The mean raw scores for those who could obtain a score on each of the baseline tests are given in Table 4-1. These results indicate that the largest age-equivalent range was found for the PPVT-III (1-10.1 years) and that the highest age equivalent score was obtained on the Word Identification subtest.

Paired t-tests between the mean age-equivalent scores showed a significantly higher score on the BVMI than the PPVT-III ($p= 0.026$). Similarly age equivalent scores on the visual discrimination were seen to be higher than age-equivalents on the Visual Form Constancy ($p=0.05$) and Visual Closure ($p=0.016$). The Sign test was used to show differences between the Word Identification and Word Attack because some children did not obtain an actual score on the latter. Scores on Word Identification showed significantly better performance than for Word Attack (p=0.047).

Chronological age was significantly correlated with performance on the BVMI, WRMT-Word Identification, PPVT and the Dolch sight words (Table 4-2) but not with Letter Identification, or with any of the visual perceptual tests. Due to the small number of participants who could perform the Word Attack and the number test, correlations with age were not determined. PPVT-III scores were also positively correlated with the BVMI, Word Identification and Dolch sight words. In contrast, there was no significant correlation between either letter size or letter position on the writing measures and age or PPVT-III.

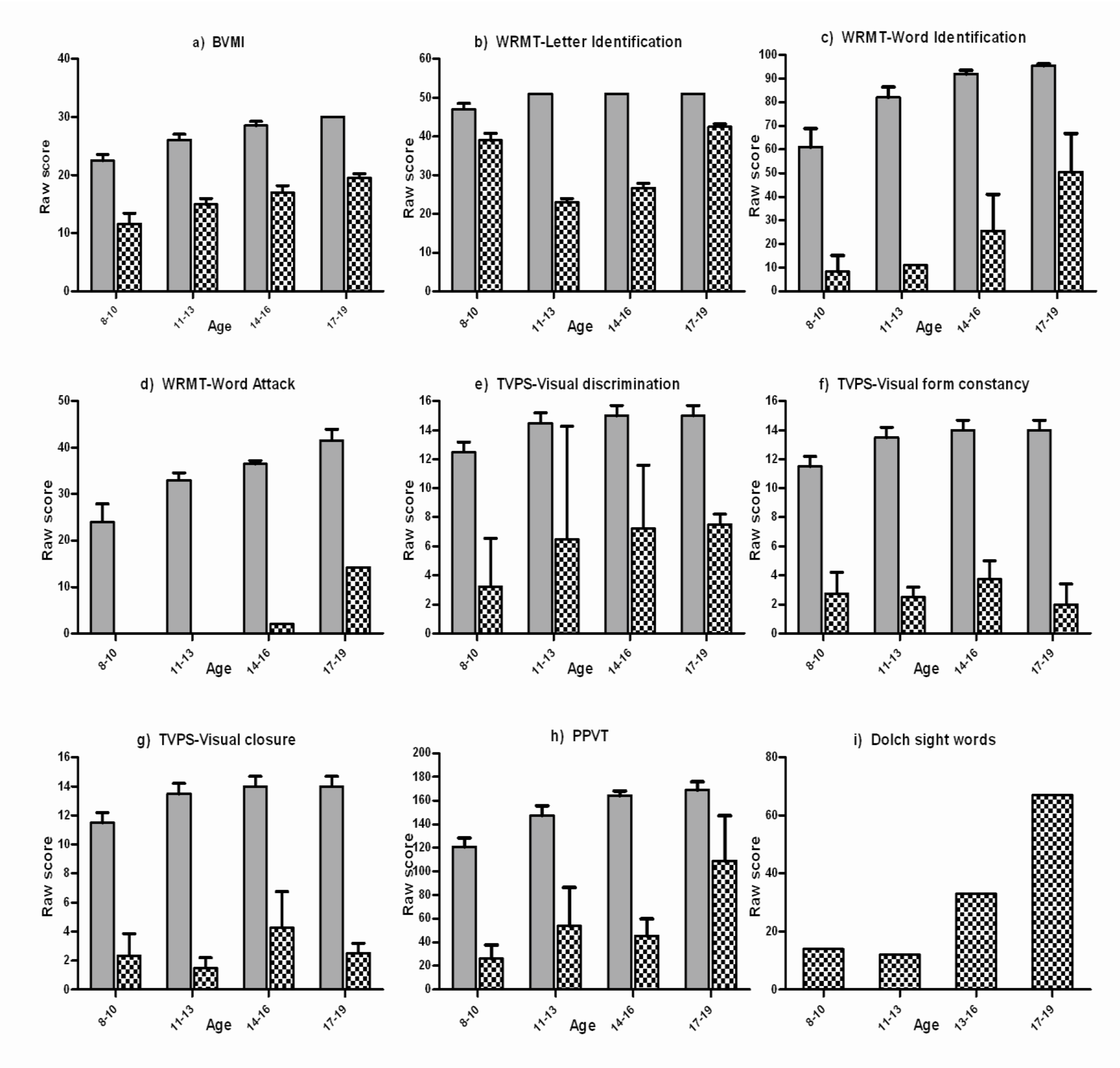


Figure 4-2: Bar charts of means of raw scores (chequered bars) of all participants grouped according to chronological age at the baseline. Mean age-expected raw scores (grey bars) (except Dolch sight words). Error bars show standard deviation of the mean.

Table 4-1 :The means and standard deviations of raw scores, range of scores obtained and corresponding age equivalents for all the tests at the baseline visit are given. The superscripts give the percentiles in that age group. Age equivalents are denoted by a-e. All figures are in decimals unless otherwise indicated. Column 2 and 3 are for all participants. Columns 4-6 are for those participants who could obtain a score on each test and the numbers are given in brackets (n).

Test	Range of raw scores	Range of a-e (years and months)	Mean +/- standard deviation (raw scores)	Mean +/- standard deviation for a-e	Range of a-e for all who obtained a score(years and months)	Actual age range of those who obtained a score
BVMI	10-20	3.11-7.6	15 ± 3.26	5.52 ± 1.22	3.11-7.6 *(14)*	8-18
Letter Identification	0-44	$5^{(9)}$-8.1	23.4 ± 15.2	6.16 ± 1.10	$5^{(17)}$-8.1 *(13)*	8-18
Word Identification	0-62	$5^{(13)}$-9.4	17.1 ± 19.7	7 ± 0.87	6.4-9.4 *(9)*	8-18
Word attack	0-20	$5^{(33)}$-8.2	3 ± 5.6	7.6 ± 0.57	7.4-8.2 *(4)*	14-18
* Visual discrimination	0-13	<4.3-8.9	5 ± 4.4	6.00 ± 1.73	<4.3-8.9 *(12)*	8-18
* Visual form constancy	0-5	<4-4.1	2.5 ± 1.7	4.1 ± 0.1	<4-4.1 *(12)*	8-18
* Visual closure	0-7	<4-5.1	2.3 ± 2.1	4.3 ± 0.4	<4-5.1 *(11)*	8-18
Dolch sight words	0-72	-	21.9 ± 27.7	-	- *(8)*	8-18
Number list	2	-	2	-	- *(5)*	8-18
PPVT-III	16-136	1-10.1	51.6 ± 34	3.7 ± 2.6	1-10.1 *(14)*	8-18

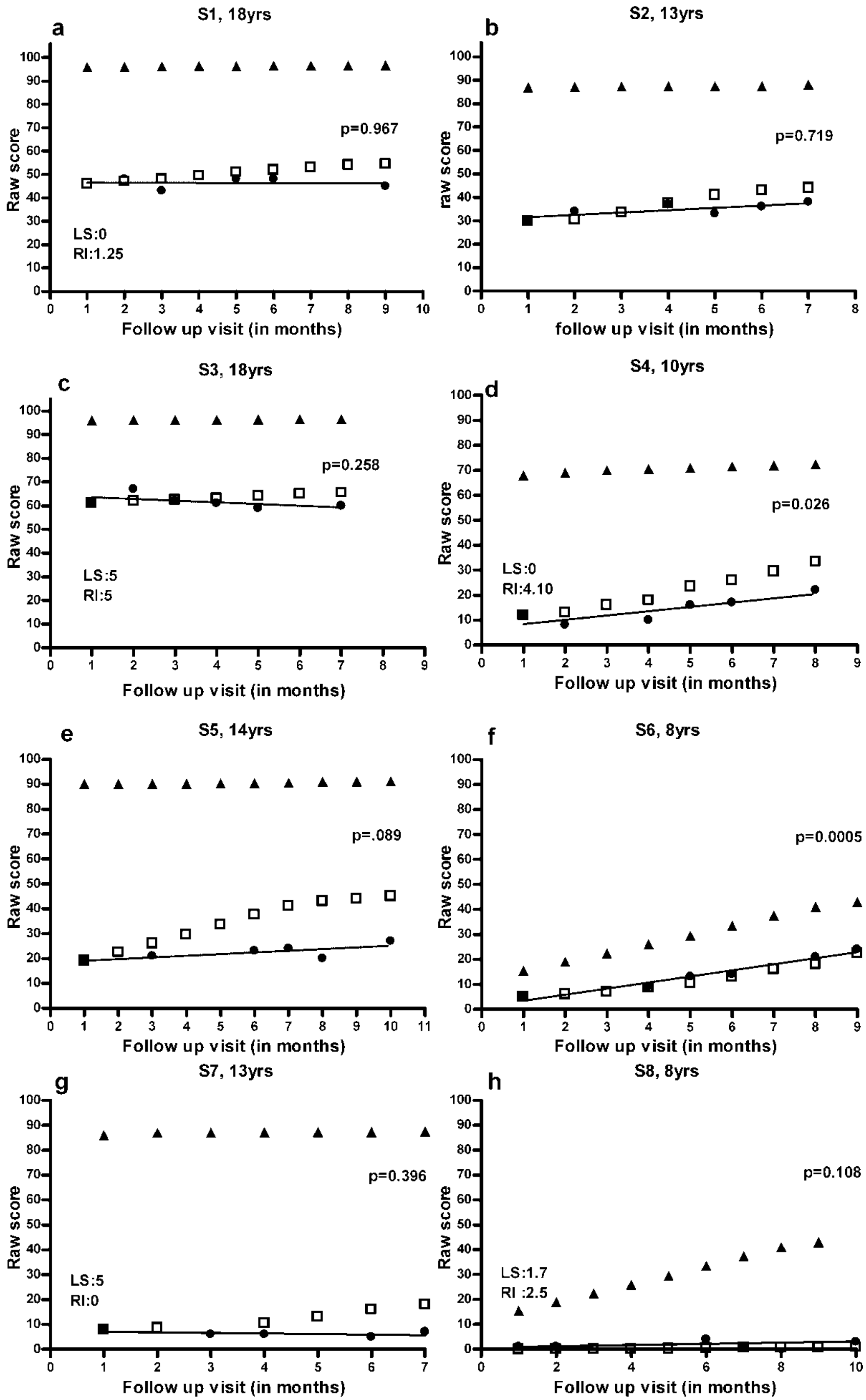
Note : The * symbol represents the tests where the age-equivalent scores were limited by the norming sample as it did not include young enough children i.e., the performance of some of the children with DS was lower than the lowest age of the norming sample

Table 4-2 Correlation coefficients with age and PPVT for all tests done at baseline, plus the initial probe values for the writing measures (r values with p values in brackets). The asterisk denotes p values that are significant (p<0.05). The p values remain significant even after applying the modified Bonferroni correction.

Tests	Correlations with age	Correlations with PPVT
BVMI	0.876 (p<0.001)*	0.725 (p=0.005)*
LI	0.442 (p=0.131)	0.550 (p=0.051)
WI	0.699 (p=0.008)*	0.621 (p=0.023)*
Dolch sight words	0.736 (p=0.004)*	0.785 (p=0.001)*
Vis Discrimination	0.474 (p=0.102)	0.544 (p=0.055)
Vis Closure	0.251 (p=0.407)	0.197 (p=0.520)
Vis Form Constancy	0.230 (p=0.450)	0.0275 (p=0.929)
PPVT	0.736 (p=0.004)*	-
Letter size	0.192 (p=0.503)	-0.274 (p=0.366)
Position of letters	0.403 (p=0.172)	-0.213 (p=0.485)

4.4.3 Monthly Probes:

WRMT- Word Identification. The monthly probes for each subject for the Word Identification task are shown in Figure 4-3a-f plotted against time, together with the expected raw scores for a typically developing child of the same age over a 6 month time period. The scores for a typically developing child matched to the initial score of each DS child are also shown. The age-matched and starting-matched scores were taken from the WRMT manual. The numbers of hours of reading instruction (RI) and literacy skills (LS) per week are shown for each participant, if the information was available. The slope of the regression line for each subject's raw scores is shown together with a *p* value (for the deviation from a slope of zero). It can be seen that six out of eight participants did not show slopes that were significantly different from zero. In addition, the trend lines for all the children are generally flatter than those for the typically-developing child with the same initial score. Repeated measures ANOVA on the Word Identification results does not show a significant effect of time on performance over the 6 month period, [$F\ (5.25) = 2.67,\ p: 0.10$)].



S1, 18yrs
a
p=0.967
LS:0
RI:1.25
Raw score
Follow up visit (in months)
S2, 13yrs
b
p=0.719
raw score
follow up visit (in months)
S3, 18yrs
c
p=0.258
LS:5
RI:5
Raw score
Follow up visit (in months)
S4, 10yrs
d
p=0.026
LS:0
RI:4.10
Raw score
Follow up visit (in months)
S5, 14yrs
e
p=.089
Raw score
Follow up visit (in months)
S6, 8yrs
f
p=0.0005
Raw score
Follow up visit (in months)
S7, 13yrs
g
p=0.396
LS:5
RI:0
Raw score
Follow up visit (in months)
S8, 8yrs
h
p=0.108
LS:1.7
RI :2.5
Raw score
Follow up visit (in months)

Figure 4-3: Scores of the monthly probes for each subject for the Word Identification task (circles). The triangles represent the expected raw scores for a typically developing child of the same age over a 6 month time period. The squares denote the scores for a typically developing child matched to the corresponding initial score of each participant . Hours of instruction if available are given for each participant for Literacy skills (LS), Reading instruction (RI) if this information was available.

*Other literacy and printing tests***.** The lack of improvement seen on the Word Identification subtest for most subjects was also apparent on the other tests. For the Dolch sight words, only one participant showed a pattern of improvement over the 6 months. In this participant, there was a slow but a steady increment in scores. Repeated measures ANOVA showed no significant effect of time on the performance of the group as a whole over 6 months [$F\ (5.25) = 1.08,\ p: 0.40$)]. None of the five participants showed a significant upwards slope in terms of improvement on their regression lines for the number test.

With regards to the various measures of writing, none of the participants showed a significant downward trend in letter size (i.e., smaller printing over time). Considering both the letter position means and standard deviations, there were no changes of letter positioning or its variability over the 6 months ($p>0.05$). All participants showed non-significant trends in the standard deviations for letter size and position. i.e. their writing did not become more consistent with time. Finally, repeated measures ANOVAs for letter size [$F(4,45)=2.181,\ p:0.073$] and position [$F(4,45)=1.849,\ p:0.122$], showed that there was no significant effect of time.

4.5 Discussion

The present study provided data on the literacy progression of a sample of Down syndrome children ages 8 to 18 years for a period of 6 months before bifocals were prescribed .

In the current study, we found an average raw score of 15 on the Beery Test of Visual Motor Integration which corresponds to an age-equivalent of 5.52. This is in agreement with other studies that have shown substantial delays on this test in participants with Down syndrome (Spano et al. 1999, Wang et al. 1995). Interestingly, Dykens et al.,(Dykens et al.2001) reported that participants with DS (average age 14) showed significant strengths on this test that exceeded their IQ expectations. Similarly in the current study, average age equivalent scores on the BVMI were higher than the age equivalent scores on the PPVT, which may be considered as a rough proxy for mental age. In this study we report higher raw scores in our group than Dykens et al., although the age range is comparable. This disparity could be attributed to the fact that the participants in our study were at a higher level of cognitive functioning because of the inclusion criteria.

As for the TVPS, no previous reports of TVPS performance in a DS population were located. The present study shows that the majority of the participants could obtain a result on this test and were performing at an age equivalent of between 4 and 9 years depending on the subtest, with the best performance on visual discrimination compared to visual form constancy or visual closure. These latter tests may entail a greater reasoning or problem solving component.

The mean age equivalent mean for Word Identification in the current study was 7 years which is not dissimilar to Fowler et al.(1995) and Boudreau (2002) who found a mean age equivalent of 8.5 (chronological age range 17-25) and 7.8 (chronological age range 5-17) respectively. The youngest participants who were aged 8 years old in this study could read between 0-6 words which is comparable to the reports of Lorenz et al (Lorenz et al. 1985). The current finding that children obtained higher scores in identifying real words than decoding unknown or pseudowords has also consistently been reported in other studies (Byrne et al. 2002, Cupples & Iacono. 2000, Fowler. 1995, Kay-Raining Bird et al. 2008). Decoding abilities have been shown to be poor in this population (Boudreau. 2002, Buckley & Bird. 1993, Kay-Raining Bird et al. 2000) mirroring the poor performance on the Word Attack subtest in our sample with an age equivalent of 7.6. It must be noted that only 4 of our 14 participants could do the test, and this could be the reason for the higher mean age equivalent in comparison to the Letter Indentification or Word Identification tasks.

Many of these children obtained a certain level of performance on the standardized reading tests ranging from age equivalents of 5 years, 6 years 4 month and 7 years 4 months on Letter Identification, Word Identification, and Word Attack respectively to highs of 8 years 1 month, 9 years 4 months, and 8 years 2 months, respectively, on the same tests, although, interestingly, it was not the oldest participants who attained these scores. This agrees with Buckley's suggestion that some children with DS can reach 8-9 year old levels of reading (Buckley. 2001). According to Buckley a 'reading age' of 8-9 years would be adequate to read books, newspapers and write letters and 60-70% of individuals with DS are able to achieve this functional level by their adult life. The two oldest participants in the current study, who were 18 years old, showed an age equivalent of 6-7 years and so are still not at this level of functional reading. There may yet be time for improvement for these individuals, as Buckley also states that many can still develop "functional levels" of literacy in their later years and continue to develop these skills with appropriate instruction as suggested by van Kraayenoord et al., (Van Kraayenoord et al. 2000).

When we consider the raw scores for the reading tests, there was considerable variation in baseline scores between our participants. This is in agreement with many other studies over the last decade showing a range of performance in raw scores for standardised tests in populations with DS (Boudreau. 2002, Cupples & Iacono. 2000, Dykens et al.2001 , Fidler et al. 2005, Kay-Raining Bird et al. 2008, Spano et al. 1999, Wang et al. 1995). However, to our knowledge ours is the first short-term longitudinal study to document reading and printing skills over a 6 month period with measures taken every month. Generally, there was little demonstrable improvement over the 6 month period. Bryne et al. (2002) showed a gradual improvement in reading single words over a 2 year period in children with DS using the British Ability Scales (Elliott. 1983). The score at the second year was better than at the first and there was better performance on both years compared to the baseline although their progress was much slower than the groups of averagely reading and reading-matched groups of children. Much akin to this, Laws and Gunn (2004), Cupples and Iacano (2000) and Kay-Raining Bird et al. (2000) reported that most of their participants improved in certain aspects of reading over the duration of the studies; ranging roughly across five years.

Although there was no demonstrable improvement for the group as a whole, two participants aged 8 and 14 years showed statistically significant positive slopes over time for the Word Identification, and for two other children, aged 8 and 14 years, the slope was marginally statistically significant. The invidivual (S4, 14 years) who showed a significant upward trend in the scores was given about 4.1 hours of reading instruction which was the second highest number of hours spent on reading instruction (the highest was 5.1 hours). No such information about reading instruction was available for the other participant (S6,8 years) who showed significant improvements with time. In the other participants, irrespective of the number of hours of reading instruction, no significant trend was observed. In the present study, we do not have any information on the commencement of literacy instruction in the participants.

Writing is a task involving fine motor skills and it is known that neuromotor development is delayed in children with DS (Cowie. 1970). Hypotonia or poor muscle tone has also been suggested to be the underlying cause for poor development of gross motor skills in DS (Cowie. 1970, Davis & Kelso. 1982). Performance on the printing task in which children were asked to print their name and a subset of numbers and letters showed no significant improvement over time in the quality with which the numbers and letters were printed. We are not aware of any previous studies that have used this kind of printing analysis in children with or without DS and therefore there are no norms against which we can compare our population. The closest study in children with DS is that of Kay-Raining Bird et al.(2008) wherein legibility of letters was measured but there was no description of these findings - only the written narratives are reported.

4.6 Limitations of the study

It is important to note that the study results should be considered as only descriptive for a number of reasons. The main reason is the small number of participants. Second, the necessary exclusion criteria meant that this study excluded lower functioning children with DS. Third, we do not have complete information about the children's schooling, types of instruction given, hours of reading instruction and interventions or the number hours of special instruction. This was because we did not receive the completed questionnaires from some teachers. Lastly there is the possibility of a practice or memory effect since the monthly tests were administered several times. However, since we found no signs of learning over the first 6 months, it would appear that memory or practice at the specific test is not a factor.

4.7 Conclusion

This paper describes our observations at baseline and over 6 months before the bifocal intervention. It is one of the few studies, looking at reading, printing and perceptual skills together with measures of literacy done every month. The data showed that the group of participants in our study were not at floor in the reading and perceptual skills. Participants were able to read a higher number of sight words and real words than sound out simple pseudo words. However there was no measureable development in the group's performance over the 6 month period, although a couple of the participants did show significant improvement for certain subtests. This indicates that overall the rate of progression is slow in this population.

5 Bifocals in children with Down syndrome (BiDS) – Visual acuity, accommodation and early literacy skills

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Roles

	Concept / Design	Recruitment	Acquisition of data	Analysis	Write-up / publication
Nandakumar	Design of the number, writing tests, and Dolch test and compliance materials	Y	(1) Refraction to determine SV correction (2) Dynamic retinoscopy for determining lag (3) All other data	Analysis of the data	Write-up
Leat	Suggestions for administration of the tests and suggestion on written tests	-	(1) Dynamic retinoscopy for determining lag (2) Rechecked refraction, add and signed prescription	Suggestions and direction	Editing and suggestions

5.1 Overview

Purpose: Reduced accommodation is seen in children and young adults with Down syndrome (DS), yet providing bifocals has not become a routine clinical management. This study investigates the impact of bifocals on visual function, visual perceptual and early literacy skills in a group of school children with DS.

Methods: In this longitudinal study each child was followed for 5 months with single vision lenses (SV) after which bifocals were prescribed if required, based on their accommodative response. Visual acuity (VA), accommodation, perceptual and literacy skills were measured after adaptation to bifocals and 5 months later. Educational progress and compliance with spectacle wear were assessed through school and parental reports.

Results: Fourteen children and young adults with DS participated in the study. Eighty five percent required bifocals with additions ranging from +1.00D to +3.50D. The mean near logMAR VA improved with bifocals (p=0.007) compared to SV lenses. Repeated measures ANOVA showed that there was more accurate focus (less accommodative lag) through the bifocals (p=0.002) but no change in the accommodation exerted through the distance portion compared to SV lenses (p=0.423). There was a main effect of time on sight words (p=0.013), word identification (p=0.047), visual closure (p=0.006) and visual form constancy (p=0.001).

Conclusion: Bifocals provide clearer near vision in DS children with reduced accommodation. This is shown by improved VA and decreased lag of accommodation. The results indicate that the improvement in VA results in improved scores in early literacy skills. Better compliance with bifocals over SV lenses was seen.

Key words: Down syndrome, accommodation, bifocals, literacy, visual perceptual skills, visual acuity, refractive error, longitudinal.

5.2 Introduction

Reduced accommodation has been shown to be present in the majority of pre-presbyopes with Down syndrome (DS) (Cregg et al. 2001, Haugen & Hovding. 2001, Nandakumar & Leat. 2009, Woodhouse et al. 1993). We have shown in an earlier report that 100% of participants had abnormal accommodative function across the stimulus distances tested both with their habitual correction or lack thereof and with newly prescribed single vision (SV) lenses (Nandakumar & Leat. 2009). Substantial amounts of under accommodation would result in near blur which could be a limiting factor for these children when doing prolonged near work. It is also possible that there is a subtle amblyopia due to this consistent blur for objects closer than infinity (reduced visual acuity with no associated physiological cause is another common finding in individuals with DS (Courage et al. 1994, Nandakumar & Leat. 2009). Additionally, a high prevalence of large refractive errors has been reported in individuals with DS (Cregg et al. 2003, van Splunder et al. 2003, Woodhouse et al. 1997). Hyperopia seems to be more prevalent than myopia (Cregg et al. 2001, Haugen et al. 2001, Mohd-Ali et al. 2006, Stewart et al. 2005), although equal percentages of both myopia and hyperopia have also been reported (Kim et al. 2002, van Splunder et al. 2003). It has been suggested that the prescription of positive lenses to correct any hypermetropia does not decrease the lag of accommodation as would be expected (Cregg et al. 2001), there may simply be a relaxation of accommodation, so that the lag remains constant. Later, however, it was shown that bifocals help to encourage more accurate accommodation in these children (Stewart et al. 2005). In our recent report, all the participants who were prescribed an increase in plus power (or a decrease in negative power) in their single vision glasses had an improvement in near visual acuity, indicating that they did not maintain a constant lag (Nandakumar & Leat. 2009).

Thus there is evidence that bifocals would decrease the blur caused by reduced accommodation in children with DS, but it remains to be seen how this would impact their everyday lives as readers. Uncorrected hyperopia in typically-developing children has been associated with reduced performance on early literacy tasks, receptive vocabulary and crowded VA (Shankar et al. 2007) and recently it has been reported that uncorrected hyperopes spend less time in near work than corrected hyperopes or emmetropes (French et al 2009). We would anticipate that hyperopic defocus would similarly impact children and young people with DS. Visual perceptual and literacy skills have been shown to be measureable in individuals with DS (Bochner. 2001, Cupples & Iacono. 2000, Kay-Raining Bird et al. 2008, Miolo et al. 2005) and it has been shown that children and young adults with DS can attain primary level literacy skills (Laws. 2000, Shepperdson. 1994, Sloper et al. 1990, Turner & Alborz. 2003).

The purpose of this study is to investigate near visual acuity, accommodation and visual perceptual skills before and after a bifocal prescription. Since this is the first longitudinal study of its kind, we undertook a descriptive, longitudinal, individual case-controlled study (each participant is his/her own control). Considering the preliminary nature of the study, a randomized study design was not undertaken at this stage as this would likely require a large multi-centre trial due to the variable performance of this population. We used frequent testing of a smaller number of participants in order to measure the progress before and after bifocal provision. Hence all of our participants who needed a correction at the initial visit were prescribed SV lenses and were followed up for five months before eligibility for bifocal prescription was determined.

We hypothesize that a) Prescribing bifocals in children with DS results in improved near visual acuity, b) The improved near acuity transforms into educational achievements at school c) The prescription of bifocals facilitates the development of improved corrected absolute levels of visual acuity. The results of the baseline visual function in this group of participants were reported in a previous manuscript (Nandakumar & Leat. 2009). Here we describe and compare the changes in accommodation, near visual acuity, visual perceptual skills and reading ability before and after the prescription of bifocals.

5.3 Methods

Fourteen participants aged 8-18 were recruited based on the inclusion criteria below:

- Diagnosed with Down Syndrome
- Age range between 5 years to 21 years and attending school
- Engaged in academic instruction and able to do at least pre-literacy or literacy tasks
- Speaks English as the first language
- Has not worn or currently wearing bifocals
- Must be verbal or able to understand instructions
- No other significant eye conditions, such as keratoconus or cataract
- No diagnosis of any neurological, sensory or behavioral disorders such as autism, microcephaly or significant hearing loss
- We did not exclude children with strabismus

All participants were subjected to a full optometric examination including:

- Distance visual acuity (VA) measured with crowded Patti-pics™ symbols (Precision Vision, Illinois) and near VA at the participant's habitual working distance measured with the Patti Pics MassVAT® version (Precision Vision, Illinois). The habitual reading distance was estimated by asking the parents for the approximate distance at which the child holds his/her books or reading material and by observing the child.
- The unilateral cover test (distance and near) or Hirschberg test
- Photopic pupil size measured with a pupil gauge
- Non-cycloplegic refraction (static retinoscopy)
- Accommodation measured with modified Nott retinoscopy, described by Leat and Mohr (Leat & Mohr. 2007) (by both observers KN and SL) at 4, 6, 8 and 10 dioptric distances
- Cycloplegic refraction using one or two drops of 0.5 or 1% Cyclopentolate hydrochloride
- Dilated fundus exam using direct ophthalmoscopy

All participants who required a change in the distance prescription were given new single vision (SV) lenses, free of cost to the participant. The participants were followed up after 1-2 weeks when visual acuity and accommodation were re-measured with the new SV lenses (SV baseline).

The following battery of visual perceptual and reading tests was administered in order at the SV baseline and then at the BF1 (after 5 months with single vision lenses) and BF2 (after 5 months with bifocals) visits:

- The Beery™ Visual Motor Integration (BVMI) (Beery. 1997)
- Woodcock Reading Mastery Test (WRMT-G) *Subtests:* Letter Identification (LI), Word Identification (WI), Word attack (WA) (Woodcock.1998)
- The Test For Visual Perceptual Skills (TVPS-R) *Subtests:* Visual Discrimination, Visual Form Constancy , Visual Closure (Gardner.1996)
- Dolch sight word list (2008)
- For children unable to read any letters, a list of numbers was made and was used instead of the Dolch sight words
- Peabody Picture Vocabulary Test (PPVT) III (Dunn & Dunn 1997) (This test was administered only at the SV baseline to determine the approximate functional age of the child)

For details of the initial optometric examination and the battery of tests, see Chapter 3, in which Figure 3-1 shows the whole protocol (Nandakumar & Leat. 2009). A summary of the findings from the baseline optometric examination is given in Table 5-1.

Table 5-1 Ocular findings from the baseline visual examination

Tests done at the baseline visual exam	**Findings**
Entering mean distance logMAR visual acuity (with old Rx or with no glasses) ± SD	OU: 0.40±0.11
Entering mean near logMAR visual acuity (with old Rx or with no glasses) ± SD	OU: 0.64±0.25
Cover test	4 esotropes (1 alternating & 3 constant), 1 esophore
Eye movements	2 jerky nystagmus, 1 latent nystagmus and 1 nystagmus at left gaze
Flash light examination of angles	14/14 normal open angles
Ocular motility	14/14 full and unrestricted
Average non-cycloplegic spherical equivalent ± SD	OD:0.78 ± 3.90 OS:1.12 ± 3.80
Average cycloplegic spherical equivalent ± SD	OD:0.90 ± 3.83 OS:1.12 ± 3.43
Direct opthalmoscopy	13/14 had healthy optic discs, good foveal reflexes, healthy maculae. The other child was uncooperative and the fundus examination was not possible

At the 6th month with SV lenses, accommodative function and near visual acuity were re-measured as at the SV baseline visit. Bifocals were prescribed based on the lag of accommodation measured through the SV lenses at 4D or 6D, whichever was closest to the participant's habitual working distance. Additions were prescribed when the lag was outside the 95% normal range at that distance based on norms from data from Leat and Mohr (Leat & Mohr. 2007). According to these norms, the accommodation lag should be <0.7D at 4D demand or <0.8D at 6D for the 6-10 year olds and <1.12D at 4D or <1.66 at 6D for the 11-19 year olds. Loose trial lenses were introduced over the habitual correction until the neutral point was brought within these norms. This enabled each child to receive a customized addition depending on their accommodative ability.

Airwear ™ (polycarbonate) Crizal™ coated straight topped D-segment bifocals, sponsored by Essilor Canada, were used for all participants. The bifocal segment top was placed at, or 1- 2 mm above, the pupillary centre in all participants (Stewart et al. 2005). Initially, the participants were asked to wear their bifocals for all tasks such as reading, writing, copying from the blackboard and working on the computer etc building up to full time wear. One to 2 (occasionally 3) weeks were given for adequate adaptation to the new lenses.

5.3.1 Initial bifocal visit (BF1) and 6th month bifocal visit (BF2):

The BF1 visit was after 2 weeks of bifocal wear (allowing the participant to adapt to the lenses) and the BF2 visit was 6 months later. At these visits, accommodation was re-measured through the bifocal segment and through the distance portion of the lenses with dynamic retinoscopy and near visual acuity was re-measured, again with the participant viewing through the bifocal segment. The battery of visual perceptual skills and reading tests was administered similarly to the SV baseline (except that the PPVT was omitted). All the participants used their bifocal segment during these tasks.

Two subtests of the battery of tests were performed monthly for 5 months before and after bifocal prescription to monitor for progress in skills. The specific subtests were chosen for each child such that the child could at least perform some level on the test and so that a ceiling would be avoided, i.e. so that changes could be measured. School reports and/or Individual Educational Plans (IEPs) were obtained for most participants throughout the study. This information was to be used to track the child's progress every term, before and after bifocals in addition to the monthly assessments. Parents were asked about compliance with spectacle wear at each main visit (SV baseline, BF1 and BF2).

All aspects of the study adhered to the tenets of the Declaration of Helsinki. Informed consent was obtained from the participants' parents and the research was approved by the Office of Research Ethics at the University of Waterloo, Canada.

5.3.2 Analysis:

Repeated measures ANOVA was used to analyze the accommodation responses between the SV baseline, BF1 and BF2 visits (4 accommodation demands [4, 6, 8 and 10D] x 3 visits). Accommodation was considered in terms of the measured lag of accommodation (neutral point with and without the add) and the accommodation exerted. These were followed by post-hoc tests (Bonferroni's correction). Repeated measures ANOVAs were also used for near visual acuities at the 3 visits and for the visual perceptual and reading test scores. The monthly sub-tests were used to determine test-retest reliability using Lin's concordance coefficient (ρc) (Lin.

1989). Partik's criteria (Partik et al. 2002) was used to grade the obtained ρc values according to the following: Values > 0.95 were regarded as excellent, > 0.90 as very good and >0.8 as fairly good. Within each group of tests (reading tests, visual perceptual skills, accommodation measures), the p level for significance was adjusted by the adjusted Bonferroni method (Jaccard & Wan. 1996), 0.05/n where n was the number of significant values obtained.

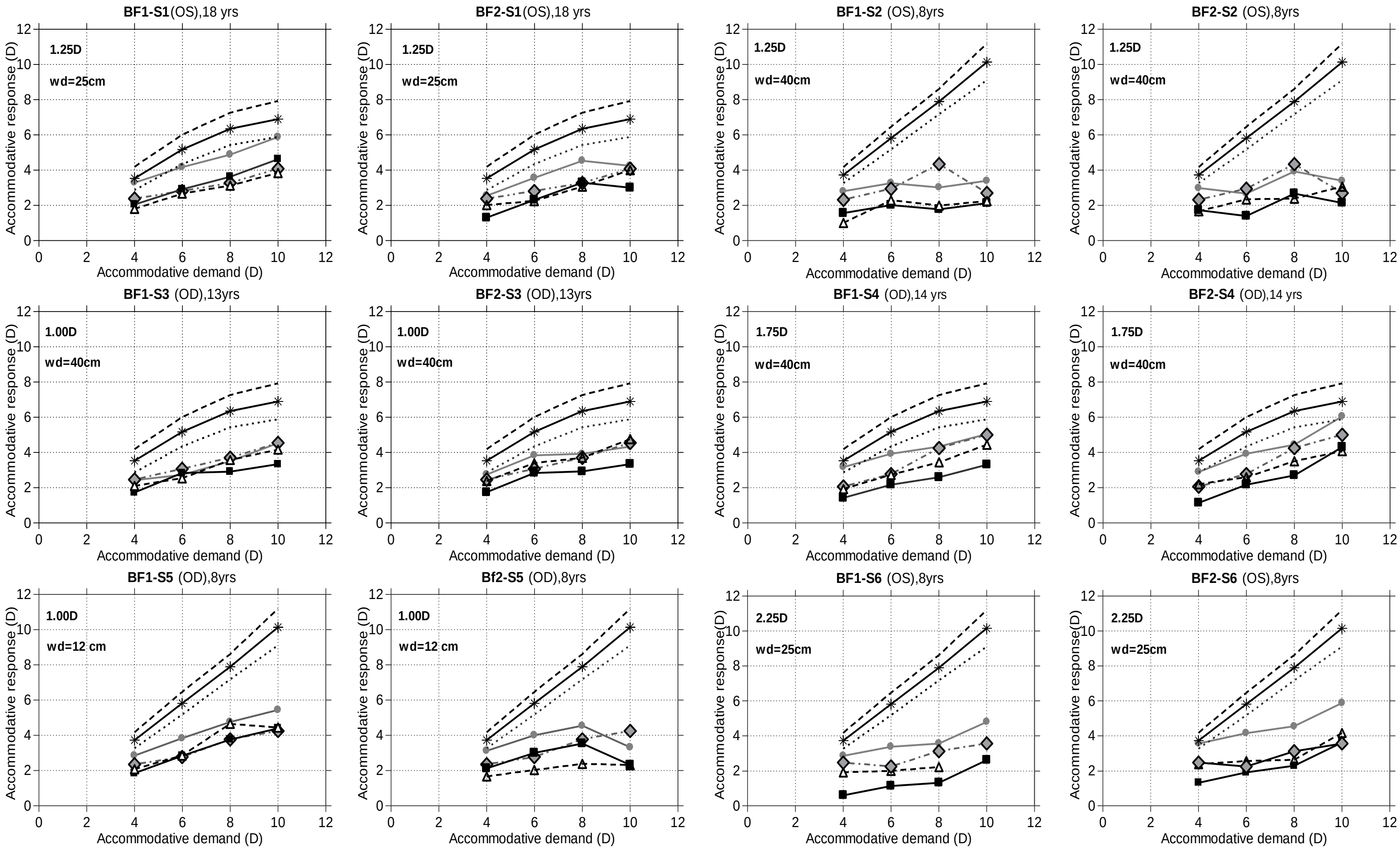
5.4 Results

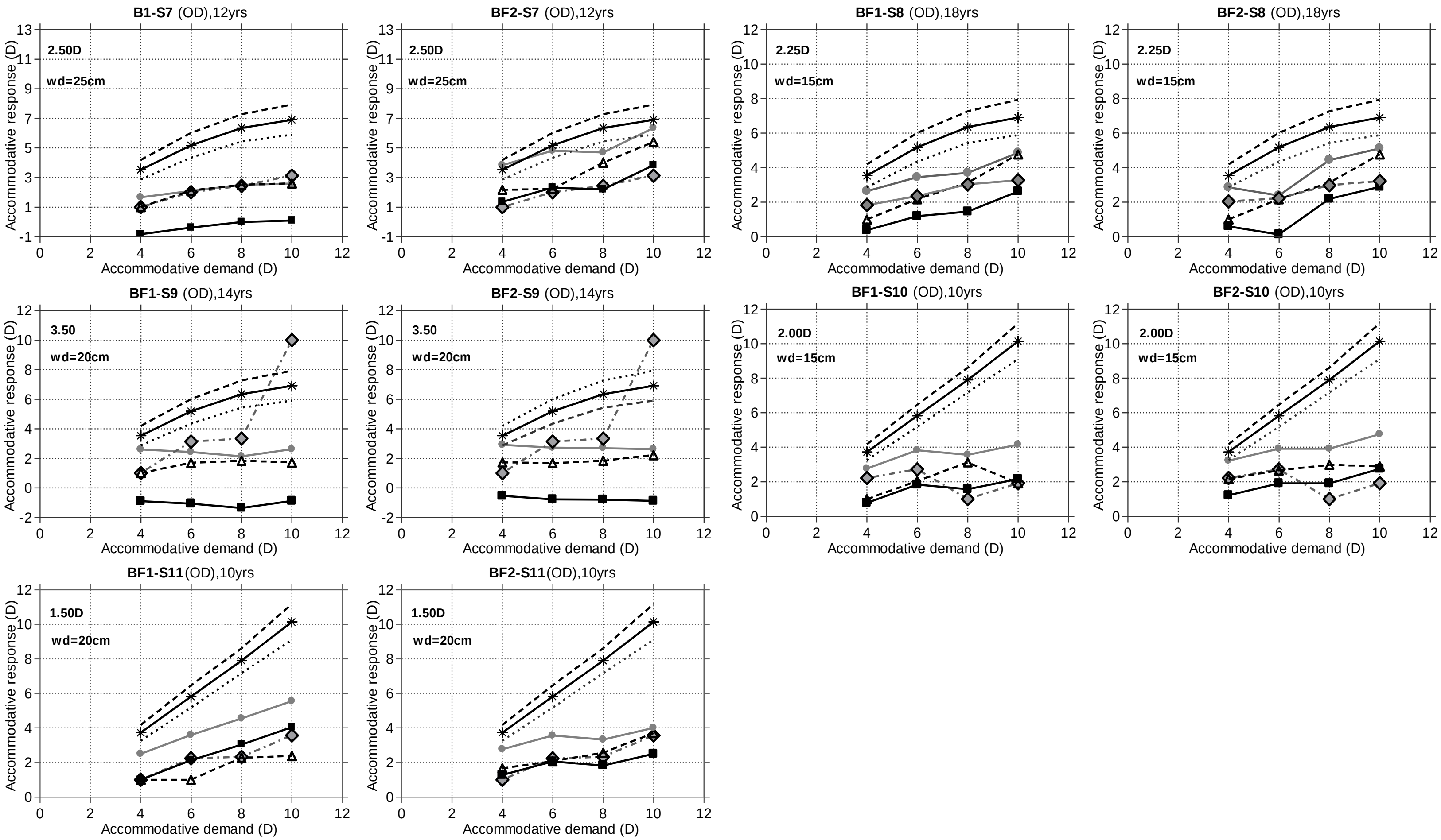
At the 6^{th} month visit with SV lenses, 11/13 participants met the criteria for prescription of bifocals. The additions prescribed ranged from +1.00D to +3.50D. Two participants, although having abnormal accommodative responses with their newly prescribed SV lenses for all the distances tested, had accommodative responses in the normal accommodative range at the 6^{th} month assessment at their habitual distance and so were not prescribed bifocals. One participant dropped out after the first few months and hence the data were not included in the current analysis.

We observed, during the monthly visits, that the children were more compliant in wearing their BF while doing the tasks, compared with SV lenses. Better compliance was reported by the parents for BF wear than for SV lenses in 6 participants and improved reading grade levels at school were reported in the school reports for 9 of them. A few examples are given : One participant improved 2 reading grade levels in the term following bifocal prescription, another from grade 2 to grade 5 reading levels, another participant who could identify numbers only from 1-10 improved to identifying 25 numbers in the term after bifocals were prescribed. In addition one participant, moved from grade 4 to grade 6 at school due to improved performance on reading skills.

5.4.1 Accommodation:

We describe the accommodation in terms of accommodative accuracy (neutral point of focus) and exerted accommodation (neutral point of focus – the power of the addition). The accommodative stimulus response plots for 4, 6, 8 and 10 Dioptric demands for all the participants through their SV lenses and through the bifocal segment and distance portion at BF1 and BF2 are shown in figure 5-1.


BF1-S1(OS),18 yrs
1.25D
wd=25cm
BF2-S1(OS),18 yrs
1.25D
wd=25cm
BF1-S2 (OS),8yrs
1.25D
wd=40cm
BF2-S2 (OS),8yrs
1.25D
wd=40cm
BF1-S3 (OD),13yrs
1.00D
wd=40cm
BF2-S3 (OD),13yrs
1.00D
wd=40cm
BF1-S4 (OD),14 yrs
1.75D
wd=40cm
BF2-S4 (OD),14 yrs
1.75D
wd=40cm
BF1-S5 (OD),8yrs
1.00D
wd=12 cm
Bf2-S5 (OD),8yrs
1.00D
wd=12 cm
BF1-S6 (OS),8yrs
2.25D
wd=25cm
BF2-S6 (OS),8yrs
2.25D
wd=25cm
Accommodative response (D)
Accommodative demand (D)



B1-S7 (OD),12yrs
2.50D
wd=25cm
BF2-S7 (OD),12yrs
2.50D
wd=25cm
BF1-S8 (OD),18yrs
2.25D
wd=15cm
BF2-S8 (OD),18yrs
2.25D
wd=15cm
BF1-S9 (OD),14yrs
3.50
wd=20cm
BF2-S9 (OD),14yrs
3.50
wd=20cm
BF1-S10 (OD),10yrs
2.00D
wd=15cm
BF2-S10 (OD),10yrs
2.00D
wd=15cm
BF1-S11(OD),10yrs
1.50D
wd=20cm
BF2-S11(OD),10yrs
1.50D
wd=20cm
Accommodative response (D)
Accommodative demand (D)

Figure 5-1: Accommodation response for each subject plotted against the accommodative demand (4, 6, 8 and 10D) at SV baseline (SV) and at the first visit with BF (BF1) shown on the left and last visit with bifocals (BF2) shown on right for each participant. Grey closed circles represent accommodative accuracy (through the bifocal), closed black squares represent the actual accommodation exerted through the bifocal, open triangles represent accommodation through the distance portion and grey diamonds represent accommodative accuracy through SV lenses at SV baseline. The mean, upper, and lower limits of the age-matched norms (Leat & Mohr. 2007) (6 to 10 years or 11 to 26 years) are plotted with an asterisk, short and long dashed lines, respectively. Add and working distance for each participant is shown.

Repeated measures ANOVA for accuracy of accommodation showed that there was a significant effect of time ($p = 0.002$) across the 3 main visits and no interactions (figure 5-2a). Post-hoc analysis showed that there was less accommodative lag (better accuracy of focus) at the BF1 visit than at the SV baseline ($p=0.019$) and at BF2 compared with SV baseline ($p=0.002$). No statistical significance was seen between BF1and BF2 ($p=0.56$). There was also the expected main effect of accommodative demand ($p<0.001$).

Apropos the exerted accommodation, repeated measures ANOVA showed a significant effect of time across the three main visits (p=0.014) (figure 5-2b), an effect of accommodative demand (p<0.001) and no interactions. Post-hoc analyses showed significant differences between BF1 and SV baseline (p=0.027), with the exerted accommodation being lower at BF1 compared to SV baseline and similarly between BF2 and SV baseline (p=0.05).There was no significant difference between BF1 and BF2 (p=0.862).

For the measures of accommodation that were taken through the distance portion, ANOVA showed that there was no effect of time (p=0.423), and again there was the expected effect of stimulus demand (p<0.001) (figure 5-2c).

For this set of ANOVAs, since there were two results that were significant at the p=0.05 level (accuracy of accommodation and accommodation exerted through the bifocal), the significance for the p value was adjusted to p=0.025 (0.05/2)for the accommodative accuracy and p=0.05 (0.05/1) for the exerted accommodation (adjusted Bonferroni correction) (Jaccard & Wan. 1996). Thus, these remain significant after the correction.

Figure 5-2a

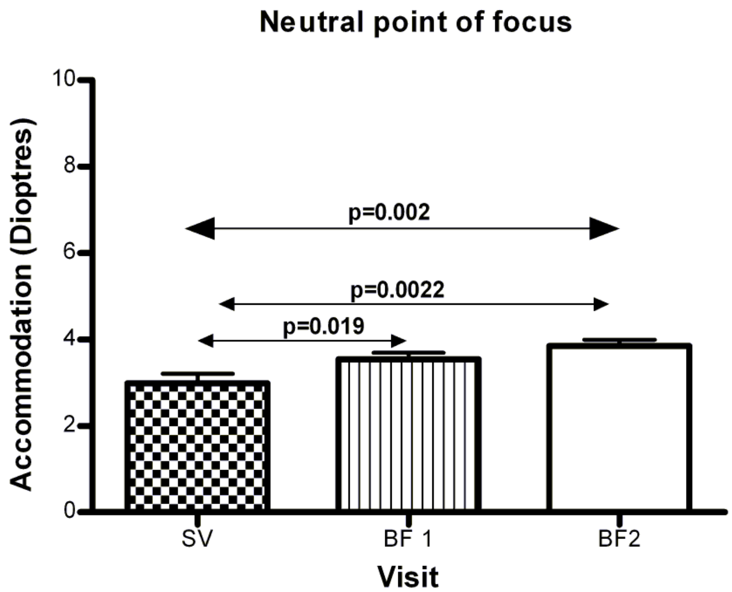


Figure 5-2b

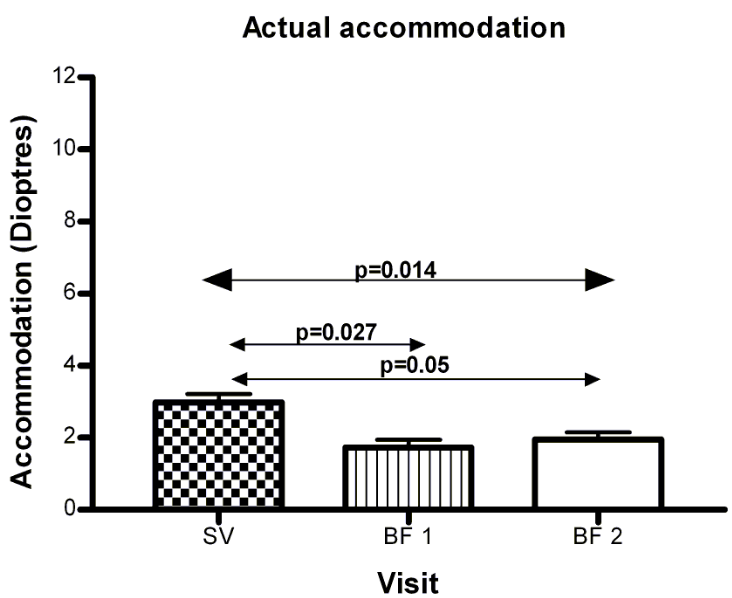


Figure 5-2c

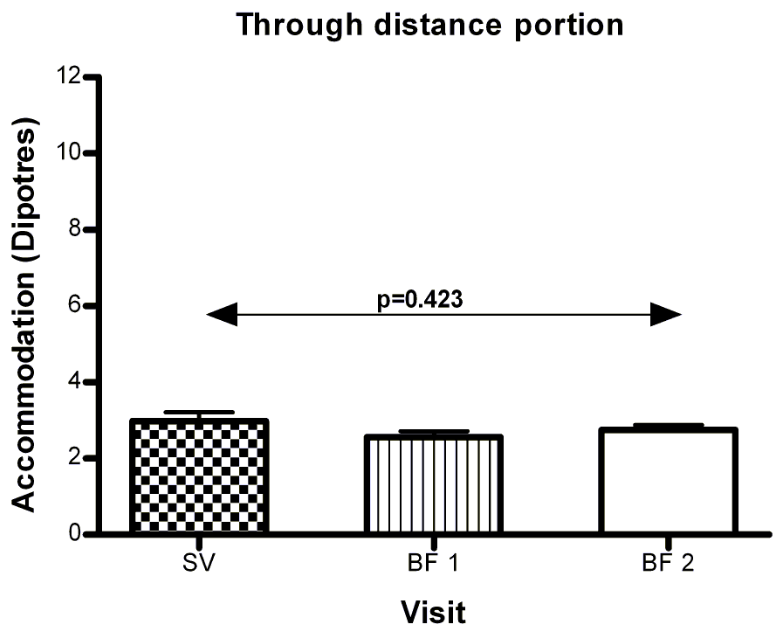


Figure 5-2 a, b and c: Bar charts of accommodation at 4, 6, 8 and 10D demands at three main visits : SV=SV baseline, BF1= initial visit with bifocals, BF2= final visit with bifocals. Figure 2a shows accuracy of accommodation (near point of focus), 2b shows the actual accommodation exerted through the bifocal and 3c shows accommodation through the distance portion. The significantly different scores (p<0.05) are indicated by arrows (large arrow heads indicating the main effects and small arrow heads indicating the post-hoc effects). The error bars show the standard error of the mean.

From figure 5-1, it can be seen that for all the subjects the accommodative responses were well outside the normal range with the SV lenses. The accuracy of accommodation improved significantly through bifocals in many of the participants (S1, S4, S5, S6, S8, S10, and S11) at BF1, particularly at the lower accommodation demands. Thus they demonstrate a reduced lag. Although these subjects showed improvement with the bifocals in place, only 1 participant had close to normal response curves through the bifocals (S1). In most cases, the exerted accommodation decreased with the bifocal in place (see S3, S4, S6, S7, S8, and S9). These subjects relaxed their accommodation through the bifocal. This is also indicated by the fact that the bifocal add was selected to bring the accommodative response into the normal range at 4 D or 6D, yet few showed an accommodation response in the normal range once they were wearing the bifocal. We also notice that in participants S1, S3, S4, S7, S9 there was fairly accurate focus through the bifocal for their habitual working distances, the lag being close to, or within, the normal range, but greater lags for the other distances. Similarly at BF2, a few participants (S3 and S7, and S4 only at 10D) showed an improvement in their accuracy of accommodation over the 5 months. With the exception of S1, who showed a small decrease in accuracy with time, all other participants showed no significant change over this period. None of the participants showed any obvious improvement in their accommodation response through the distance portion i.e. in no subject would removing the bifocal be clinically indicated.

5.4.2 Near Visual acuity:

The mean near logMAR VAs across the 3 main visits are shown in figure 5-3 and repeated measures ANOVA showed a significant effect of time (p=0.007). Post-hoc tests showed that there was a significant improvement in near VA with BF compared to SV lenses at the BF1 visit (p=0.014) and at the BF2 visit (p=0.020). There was no significant change between BF1 and BF2 (p=1.00).

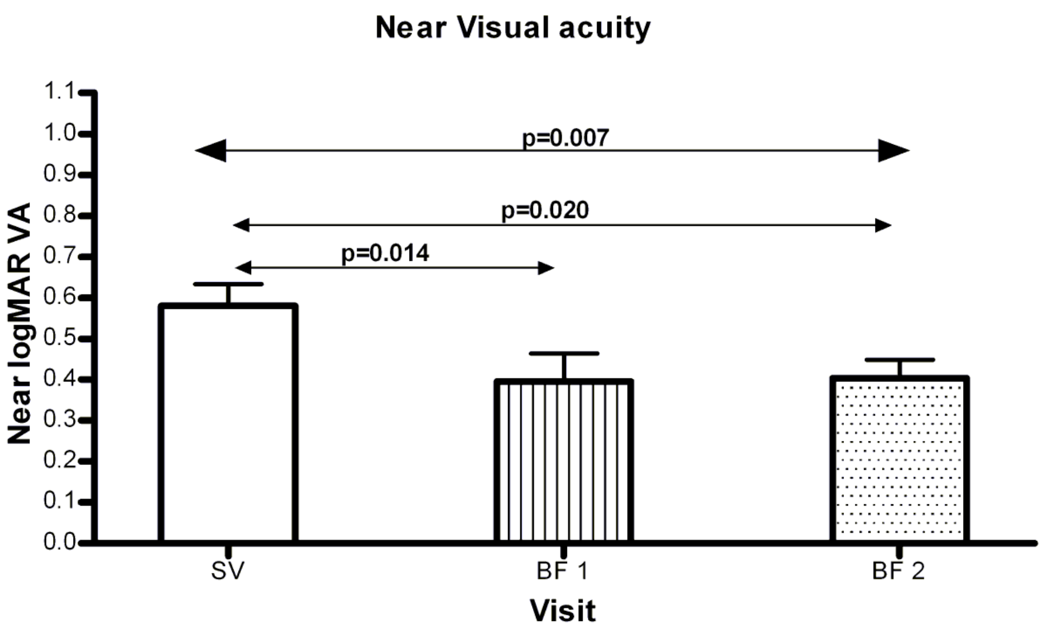


Figure 5-3: Bar charts of near logMAR VA against visits, SV=SV baseline, BF1= initial visit with bifocals, BF2= final visit with bifocals. The significantly different scores (p<0.05) are indicated by arrows (large arrow heads indicating the main effect and small arrow heads indicating the post-hoc significances). The error bars show the standard errors of mean.

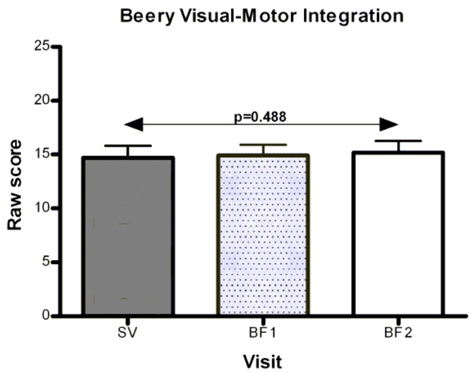
5.4.3 Visual perceptual and literacy skills :

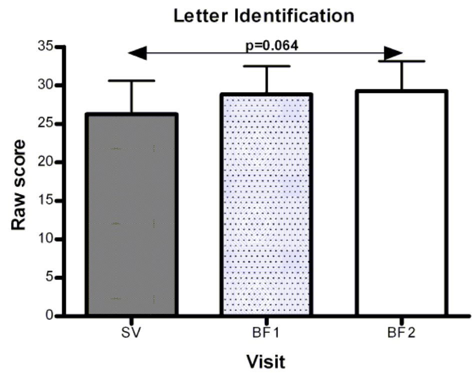
5.4.3.1 Test-retest reliability

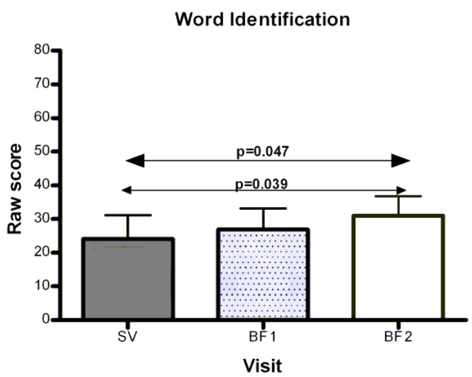
Reliability was calculated for the monthly sub-tests between the first two single vision lens visits, using Lin's concordance coefficient (ρc) with 95% confidence intervals. The Dolch had a ρc=0.95, WI, ρc =0.96 and the number test ρc=0.82 showing that the former two tests have very good/excellent reliability and the latter shows fairly good test-retest reliability.

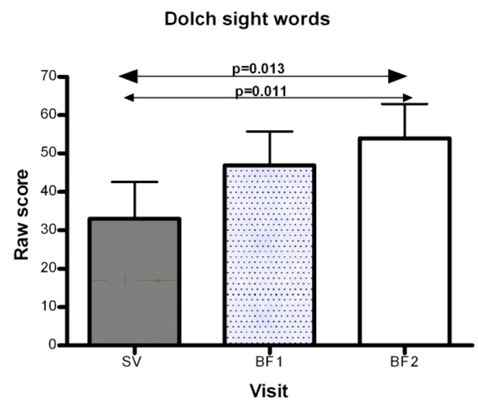
5.4.3.2 Repeated-Measures ANOVA

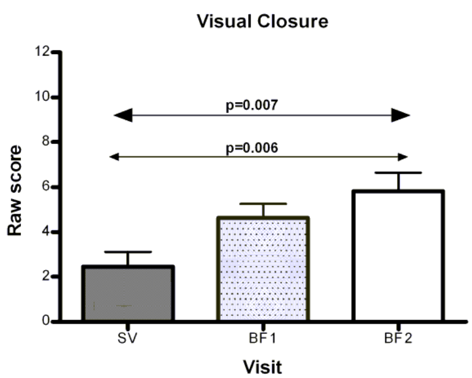
Repeated measures ANOVAs were used to compare the visual perceptual and reading scores between the three main visits (SV baseline, BF1 and BF2). There was a main effect of time on the Dolch sight words (p=0.013), WI (p=0.047), visual closure (p=0.007) and visual form constancy (p=0.001). The adjusted Bonferroni method was applied within the reading tests and the visual perceptual skills tests. The significance was adjusted to p=0.025 for the Dolch and p=0.05 for the WI and similarly p=0.025 for the visual form constancy and p=0.05 for the visual closure. Post hoc analysis (Bonferroni) showed that the performance at BF2 improved over the SV baseline for the Dolch (p=0.011), WI (p=0.039) and visual closure (p=0.006) and visual form constancy scores improved at BF1 visit over the SV baseline (p<0.001). Statistical significance was not seen for the other literacy and perceptual tests (p>0.05). The raw scores for all the perceptual and literacy skills administered at the 3 main visits are shown in figure 5-4. Since only a small number of participants (3 and 4 respectively), performed the word attack and the number test , the average scores are shown in figure 5-5.

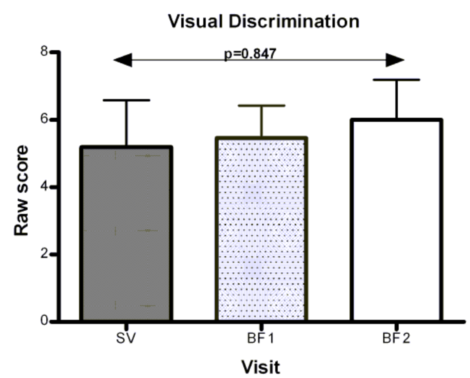

Beery Visual-Motor Integration
p=0.488
Raw score
0
5
10
15
20
25
SV
BF1
BF2
Visit


Letter Identification
p=0.064
Raw score
0
5
10
15
20
25
30
35
SV
BF1
BF2
Visit


Word Identification
p=0.047
p=0.039
Raw score
0
10
20
30
40
50
60
70
80
SV
BF1
BF2
Visit


Dolch sight words
p=0.013
p=0.011
Raw score
0
10
20
30
40
50
60
70
SV
BF1
BF2
Visit


Visual Closure
p=0.007
p=0.006
Raw score
0
2
4
6
8
10
12
SV
BF1
BF2
Visit


Visual Discrimination
p=0.847
Raw score
0
2
4
6
8
SV
BF1
BF2
Visit

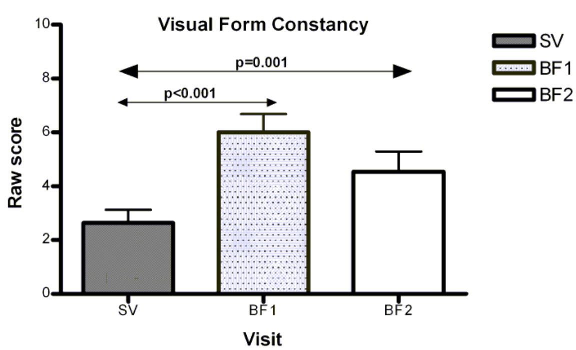

Visual Form Constancy
p=0.001
p<0.001
Raw score
0
2
4
6
8
10
SV
BF1
BF2
Visit
SV
BF1
BF2

Figure 5-4: Bar charts of raw scores obtained in all the perceptual and literacy tests at the three main visits (SV baseline, BF1 and BF2). The significantly different scores ($p<0.05$) are indicated by arrows (large arrow heads indicating the main effect and small arrow heads indicating the post-hoc significance).

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Figure 5-5a

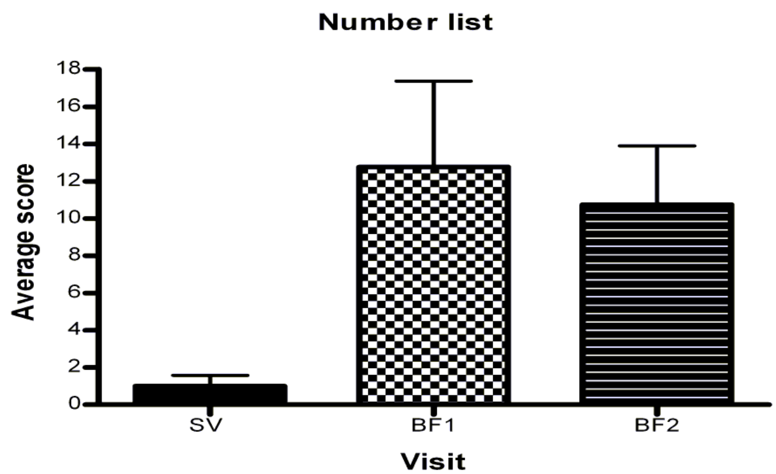

Figure 5-5b

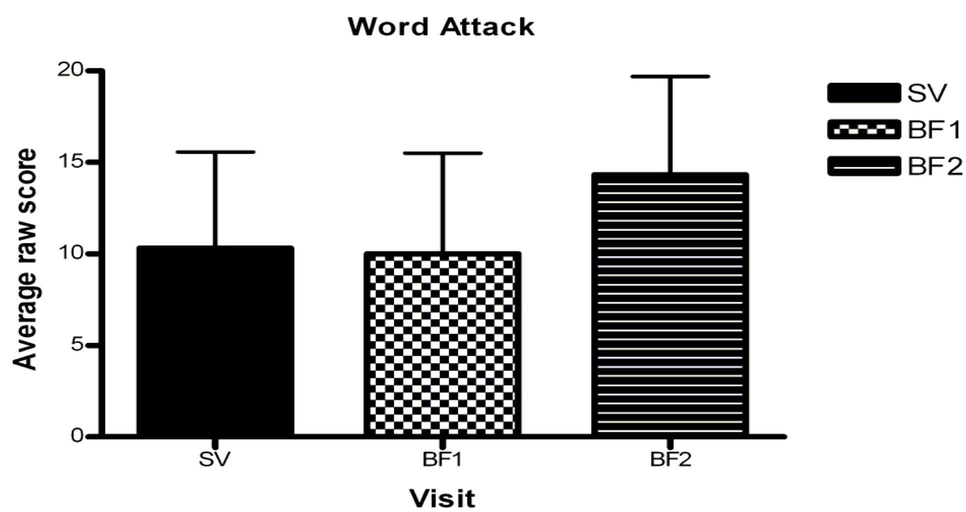

Figure 5-5: Bar charts of average raw scores for the Word attack test (n=3) (figure 5a) and number test (n=4) (figure 5b). The error bars show the standard error of mean

5.5 Discussion

To our knowledge this is the first study to investigate the impact of bifocals on reading, literacy and visual perceptual skills. In our previous report, we described the baseline optometric findings, including the fact that 100% of children with DS in our sample had reduced accommodative responses. Here we report the results of the impact of bifocals on the accommodative response, near acuity, reading and visual perceptual skills and reported school performance and compliance with wear.

Accommodation accuracy was found to improve with bifocals i.e. the lag of accommodation decreases and this would be commensurate with a decrease in near blur resulting in the improvements in visual acuity that were found. Additionally the results show that the participants do not maintain a consistent level of accommodation, but relax their accommodation to some extent when viewing through the bifocal segment. We find that there is neither a consistent level of accommodation maintained nor a consistent lag. They respond in an intermediate fashion. This is unlike the findings of Cregg et al.(2001) who suggested that their participants maintained a consistent lag when the power of the prescription lenses was changed. Our finding is interesting because the bifocal power was initially determined to cause the accommodation to fall within the age-matched normal range. It seems that when the children start wearing their bifocals they tend to relax their accommodation. There is improved accuracy of focus, although not quite sufficient to bring their response to within the age-matched norms.

The current study did not find that the accommodative response through the distance portion of the bifocals improved with time (at least after 5 months of wear) either as a group or individually. This is in variation with two recent reports from the same laboratory where the mean accommodative lag through the distance portion improved (Al-Bagdady et al. 2009, Stewart et al.2005). According to Stewart et al. (2005) bifocals encourage them to use their own accommodation unlike in presbyopic adults, where the lenses act as passive aids to overcome near defocus. It is not clear why our data are different from Al-Bagdady et al., and Stewart et al. The main difference seems to be that they prescribed the same add (2.5D) for all their children with reduced accommodation, while we prescribed a customized add based on the accommodative lag. In addition they categorized an improvement in accommodation response as being a response within the normal range in 2 out of the 3 distances tested (10, 6 and 4D) whereas in the current study the accommodation for statistical purposes was based on the numerical measures of accommodation (and lag) at all the 4 distances measured. So our analysis should have been more sensitive to showing changes. The studies also used different normal data sets, the current study using Leat and Mohr (2007) and the other two studies using McClelland and Saunders (2003). However, this difference does not explain the variation in findings either, as the McClelland and Saunders data has a narrower range of norms compared to those used here. Other differences could be the larger number of subjects and the duration of follow up. The former study had 40 children and teenagers aged 4.96-14.64 years at enrollment who were followed up anywhere between 1-7.8 years after bifocal prescription. In the current study there were 11 participants followed up over 5 months.

Our clinical experience at the School of Optometry in the University of Waterloo is also relevant here. Eight patients in the Pediatric and Special Needs clinic were prescribed bifocals prior to the current study, using the same prescribing protocol. Of these, 5 did not show an improvement in accommodation. One did show an improvement and so bifocals were removed, but a year later the accommodative response had reduced and bifocals were re-prescribed. One showed sufficient improvement for the bifocal power to be reduced (but not eliminated) and one was given an increase in add power after 1.5 years of wearing an add. Thus it seems that there is variability in response to bifocals, with many children continuing to require them. However, our current study and clinical findings do not support the concept that bifocals stimulate accommodation in any lasting fashion. We conclude that children with DS should be monitored closely and accommodation checked routinely, even after bifocals have been prescribed.

The current study demonstrates that bifocals do have an impact on the children's visual and school performance. Near visual acuity improved with bifocals, which is similar to the findings of Stewart et al. (2005). This confirmed our first hypothesis. Although there were a small number of participants in the current study, there was a statistically significant overall improvement in word recognition tests and in some visual perceptual skills. The Dolch sight words and Word identification task as well as 2 out of 3 visual perceptual subtests (visual form closure and visual constancy) showed significantly improved scores with bifocal lenses. This improvement in reading performance and visual perceptual tasks is expected to transfer into improvements in academic attainments in these children, thus supporting our second hypothesis. The children's school reports also support this conclusion, with improvements being noted in 9/11 children. It is important to note that the test font sizes were enlarged by a factor of 2 so that the print was within the acuity range of the children even with SV glasses, so that this improvement is not because the test was outside the acuity limit of the child before bifocals were prescribed. The improvement in acuity and focus may lead to greater ease, comfort and attention and a better acuity reserve (Lovie-Kitchin et al. 2001, Whittaker et al. 1993).

There is the possibility that these children would naturally improve in their reading performance with time, since they were all involved in classroom learning. There is also the possibility of a practice effect since the tests were administered several times (although different versions were used for the battery of tests and the monthly sub-tests). However, we find no signs of learning or practice over the first 5 months before the provision of bifocals (Nandakumar et al.,2010 *in submission*) i.e. these children have no significant natural progression of literacy skills over a 5 month period. It is also known that children with DS have poor short-term memory (Bower & Hayes. 1994). Thus we think that it is unlikely that the improvements after bifocal provision are due to natural progression or practice.

An important measure is compliance with spectacle wear, which indicates that the children were obtaining benefit. The reports from both the teachers and parents and from our observations in the laboratory show that the children were readily using the bifocals and adapted to them quicker than expected. In fact, they appeared to adapt to them more readily than the SV glasses, wearing them for the majority of their waking time.

5.6 Conclusion

Although preliminary and longitudinal in nature, the current study gives evidence of superior focusing, better near visual acuity, improved literacy skills over time and enhanced school performance as a result of bifocal prescription in children with DS. The children adapt well to bifocal wear. Bifocals should be considered in the clinical management of children and teenagers with Down syndrome. More long term studies are required to demonstrate whether long-term bifocal wear would improve absolute visual acuity.

6 Analysis of video recorded sessions of literacy and visual perceptual skills

This chapter is in submission as follows:

Nandakumar K, Briand K, Leat SJ. Bifocals in Down syndrome study (BiDS): Analysis of video recorded sessions of literacy and visual perceptual skills, in *Clinical and Experimental Optometry (Under review).*

Roles

	Concept / Design	Recruitment	Acquisition of data	Analysis	Write-up / publication
Nandakumar	Design of the number, writing tests, and Dolch test and compliance materials . Development of the measures to analyze the videos (time taken)	Y	(1) Refraction to determine SV correction (2) Dynamic retinoscopy for determining lag and add (3) All other data (4) Randomized and prepared videos for analysis	Analysis of the data	Write-up
Briand	-	-	-	Timing the videos	-
Leat	Suggestions for administration of the tests and suggestion on written tests	-	(1) Dynamic retinoscopy for determining lag (2) Rechecked refraction, add power and signed prescription	Suggestions and direction	Editing and suggestions

6.1 Overview

Purpose: In this longitudinal study a group of school children with Down syndrome (DS) who had reduced accommodation were prescribed with bifocals and followed in order to investigate the impact of bifocals on early literacy and visual perceptual skills. The changes with bifocals are described by monthly sub-test scores and the time taken to complete literacy and visual perceptual tasks.

Methods: Fourteen children with DS, aged 8-18 were followed for 5 months with single vision lenses and 11 were prescribed bifocals based on their accommodative ability and followed for another 5 months. A battery of reading and visual perceptual tests was administered before and after bifocal prescription. Monthly sub-tests of similar tasks were administered to measure progress. All the visits were videotaped to determine the time taken for the child to complete each task.

Results: There was significant improvement in the scores of the monthly subtests with bifocals (p=0.050, 0.025 and 0.023 for Word Identification (WI), Dolch sight words and number test respectively) and the rate of progress in monthly scores improved with bifocals for WI (p=0.008). RM ANOVA showed a significant decrease in the completion times with bifocals for the full battery of tests on the WI, *(p=0.0015).* There was significant correlation between the improvement in focus with bifocals and the completion time for the WI task *(p=0.004).*

Conclusions: This study demonstrates faster and improved performance on some literacy skills with bifocals. We recommend that bifocals be considered in children with DS presenting with inadequate accommodation to optimise their educational potential.

Key words: Down syndrome, bifocals, accommodation, literacy skills, visual perceptual skills, longitudinal.

6.2 Introduction

Down syndrome (DS) is the most common genetic cause of intellectual disability in individuals and is accompanied by mild to moderate mental challenge. There has been increased awareness on Down syndrome in the last few decades, with attention to intellectual, cognitive, physiological and anatomical aspects of the syndrome. In the context of intellectual functioning, there is a large body of literature on the educational abilities, literacy and functional skills in this population (Boudreau & Chapman. 2000, Buckley. 1995, Buckley et al. 1996, Byrne et al. 2002, Chapman. 2006, Chapman et al. 1998, Chapman & Hesketh. 2001, Fowler. 1995, Fowler et al. 1995, Kay-Raining Bird et al. 2000, Kay-Raining Bird et al. 2008, Kim et al. 2008, Klein. 1999, Morgan et al. 2004, Oelwein. 1995, Shapiro & France. 1985, Shepperdson. 1994). There is much less literature on the natural progression of skills. In a previous paper, we have described aspects of visual functioning and the progression of early literacy and visual perceptual skills in children and young adults with DS (Nandakumar et al. 2010, *in submission).*

A wide variety of ocular problems are present in this population including high refractive errors and these have been reported in many studies (Caputo et al. 1989, Catalano. 1992, Cregg et al. 2003, da Cunha & Moreira . 1996, Gardiner. 1967, Haugen et al. 2004, Nandakumar & Leat. 2009, Shapiro & France. 1985, Tsiaras et al. 1999, van Splunder et al. 2003, Woodhouse et al. 1997). Repeatedly, it has been shown that accommodation is reduced in this population (Cregg et al. 2001, Haugen & Hovding. 2001, Lindstedt. 1983, Nandakumar & Leat. 2009, Woodhouse et al. 1993, Woodhouse et al. 1996), yet correcting this with bifocals has generally not been incorporated in every day clinical practice. We were interested in the potential of bifocal prescription to impact visual function and academic performance in this population, as this has not been investigated before.

So we undertook a preliminary, descriptive, longitudinal, individual case-controlled study (each participant as his/her own control) of the impact of bifocal provision on visual acuity, accommodative function, early literacy, visual perceptual skills and printing skills. A battery of tests was given before and 5 months after bifocal prescription. We also decided to use frequent (monthly) testing of a small number of participants in order to measure the progress before and after bifocal provision. Our hypothesis is that bifocal provision, which improves near visual acuity and provides more accurate focus (Nandakumar & Leat, 2010), will result in educational gains at school as measured by:

a. Improved scores in standard early literacy and visual perceptual tests
b. Improved printing skills as measured by the average size and position of the letters on the line and the variability of these measures - a smaller and more uniform positioning of letters may indicate a more developed or mature writing.
c. Improvements in the completion times for the tasks i.e. faster performance (as measured from videotaped sessions) on these perceptual and literacy tests.

We have previously reported that there is less optical blur through the bifocals with commensurate improvements in near acuity (Nandakumar & Leat, 2010), and that there were significant improvements in the scores of some reading tests and visual perceptual tests used in the test battery. The current paper describes (1) The scores of the monthly tests of perceptual and early literacy skills before and after bifocal prescription; (2) The printing test results and (3) The time taken to complete the individual tests of the test battery and the monthly probes before and after bifocal prescription as observed from videotaped sessions.

The use of videotapes to transcribe or make observations during educational or interventional sessions in individuals with DS has been used before (Bird & Buckley. 1994, Cuskelly et al. 2001, Gallaher et al. 2002, Kim et al. 2008, Morgan et al. 2004, Mundy et al. 1988, O Toole & Chiat. 2006). Along with other measures, audio or video taping has typically been used as a secondary outcome (Chan & Iacono. 2001, Iverson et al. 2003, Kay-Raining Bird et al. 2008, van Bysterveldt et al. 2006). To our knowledge, the present study is the first of its kind which has measured the impact of bifocals on early literacy skills and in which each reading and perceptual tasks were videotaped at every visit.

6.3 Methods

A full detail of the study protocol is given in Nandakumar and Leat (Nandakumar & Leat. 2009). Fourteen children and teenagers with DS, aged 8-18 participated in the study (6 males and 8 females). At the initial visit, a basic eye examination was administered, which included measurement of distance and near visual acuities, binocular status and a cycloplegic refraction. Accommodative ability was measured using the modified Nott dynamic retinoscopy (Woodhouse et al. 1993). Participants were prescribed new single vision (SV) glasses if necessary. A battery of early literacy and visual perceptual tests was administered with new SV lenses (SV baseline). The results of these baseline visual function and early literacy and perceptual skills with single vision lenses are reported in Nandakumar & Leat (Nandakumar & Leat. 2009). The children were followed for 5 months, during which time they were seen monthly and a few sub-tests (probes) of the literacy skills, plus a printing task, were administered. After 5 months, the children were assessed for bifocals (based on their accommodative response) and eleven were prescribed with bifocals. The battery of tests was repeated with the bifocals (BF1 visit) and the probes were undertaken for another 5 months after which a final battery of tests was done (BF2 visit). All the sessions for the battery of tests and the monthly probes were video-taped. The results of accommodative function and early literacy and perceptual skills at the three main visits, SV baseline, BF1 and BF2 are given in Nandakumar and Leat (2010)*)*.

The battery of tests included the following (see Nandakumaret al., *in submission)* for more detail):

- The Beery™ Visual Motor Integration (BVMI) (Beery. 1997)
- Woodcock Reading Mastery Test (WRMT-G) *Subtests:* Letter Identification (LI), Word Identification (WI), Word attack (WA) (Woodcock.1998)
- The Test For Visual Perceptual Skills (TVPS-R) *Subtests:* Visual Discrimination, Visual Form Constancy , Visual Closure (Gardner.1996)
- Dolch sight word list (2008)
- For children unable to read any letters, a list of numbers was made and was used instead of the Dolch sight words
- Peabody Picture Vocabulary Test (PPVT)III (Dunn & Dunn. 1997) (This test was administered only at the SV baseline to determine the functional age of the child)

For the probes administered every month (with SV and BF lenses), the following tests were used in order:

- Printing his/her name
- The WRMT H (Word Identification 2)
- Printing any ten letters and any ten numbers
- The Dolch sight words (2nd list) or the number list depending on the child's ability

6.3.1 Videotaped sessions and analysis:

All the perceptual and literacy sessions were videotaped using a Sony Handy Cam (DCR-SR100) on a tripod. The recordings were re-named and randomised before analysis. The time taken to perform each page of every task was calculated from the video recordings by a naïve observer for all the battery visits and the monthly probes. The time taken for distractions and breaks, e.g., when the child needed water or a snack or dropped a pencil, was subtracted from the total time taken for each page of each task. Since it was possible that on some occasions a child might go further through the test before meeting the stopping criterion, the time taken up to the earliest page that the child completed on any visit was used in the analysis. For example, if the child progressed to page 5 on the SV visit, page 6 on the BF1 visit and page 7 on the BF2 visit, only the time taken up to page 5 was used for analysis, in order to make a fair comparison of time taken.

All facets of the study adhered to the Declaration of Helsinki and the study was approved by the Office of Research Ethics at the University of Waterloo, Canada.

6.3.2 Data analysis:

For each participant the probe raw scores were plotted against time and regression lines were determined to give a measure of the progression over time (indicated by the slope). Also the mean raw scores were calculated before and after bifocals. Paired t-tests were used to analyze the mean raw scores and slopes of the probes across subjects before and after bifocals.

The printing task was analyzed by measuring the size of each written letter and its position on the ruled line as shown in figure 6-1. The deviation from the line (in mm) was given a negative score if the letter was placed below the line and a positive score if it was above the line. Letter size and letter position means and standard deviations were calculated for each participant for each visit across all the monthly visits. The standard deviation gives a measure of the total amount of variation in letter position or size. Regarding letter position, since negative and positive position scores would cancel when calculating the mean, a secondary analysis was undertaken for the position scores in which the absolute value of the letter position was used. Paired-tests were used to analyze the mean letter size and letter position in all subjects before and after a bifocal provision. The time to perform the tests was analysed as follows. For the test batteries, a repeated measures ANOVA was used to analyze the time taken to complete each task at the SV baseline, BF1 and BF2. Paired t-tests were used to analyse the average time taken on the probes before and after bifocal correction. Also, the results were plotted over time and the slopes of the regression lines before and after bifocal prescription were analysed with paired t-tests.

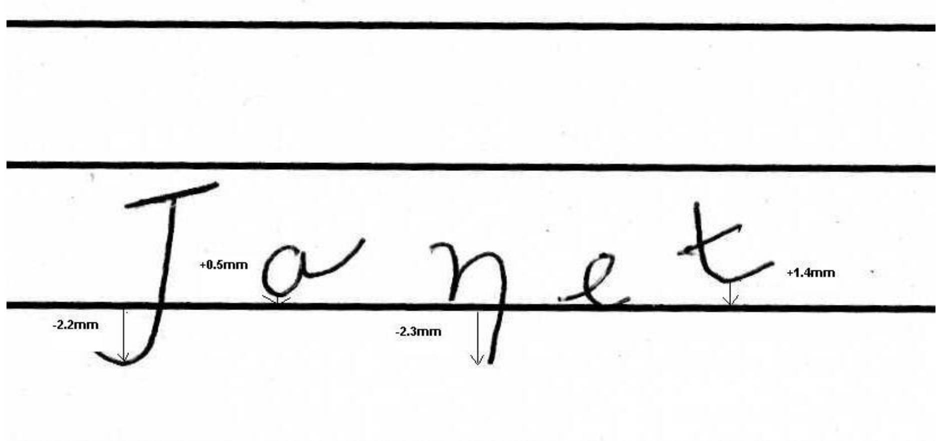

Figure 6-1: Analysis of the letter printing task. Positive and negative signs indicate if the letter was placed above or below the bold line.

In order to determine any factors which might predict improvements in reading scores or the times taken, Pearson's correlation coefficients were determined between the significant changes in the outcome measures before and after bifocal correction and; a) the difference in the near visual acuities at the BF1 and SV visits and b) the lag of accommodation with SV lenses and c) the difference in the accommodative response at the habitual working distances at the BF1 and SV visits.

6.4 Results

6.4.1 Probe scores (before and after bifocals):

6.4.1.1 *Early literacy and numeracy skills*

The raw scores obtained at the monthly probe visits over the 5 months before and after bifocals are shown in figure 6-2 (a), (b) and (c). One participant's values were excluded in this analysis, as s/he had missed too many visits and thus there were too few data points to calculate a meaningful slope. Paired t-tests were used to compare the slopes across all subjects before and after bifocals and showed a significant improvement in performance (an increase in the slope) for the Word Identification (p=0.008) but no significance for the Dolch sight words (p=0.346) or number list (p=0.62).

Figure 6-2a: Word Identification

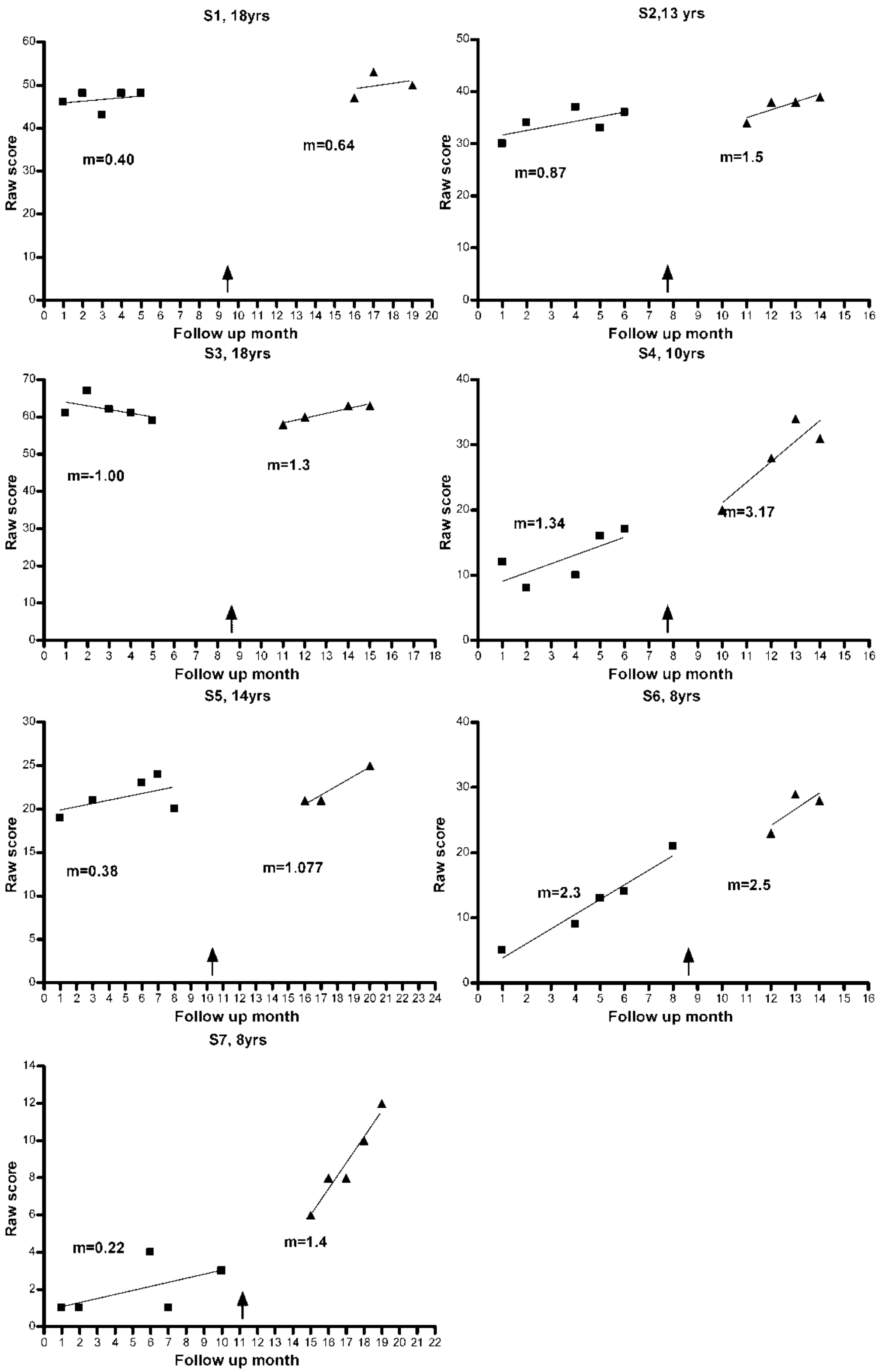

Figure 6-2b: Dolch sight words

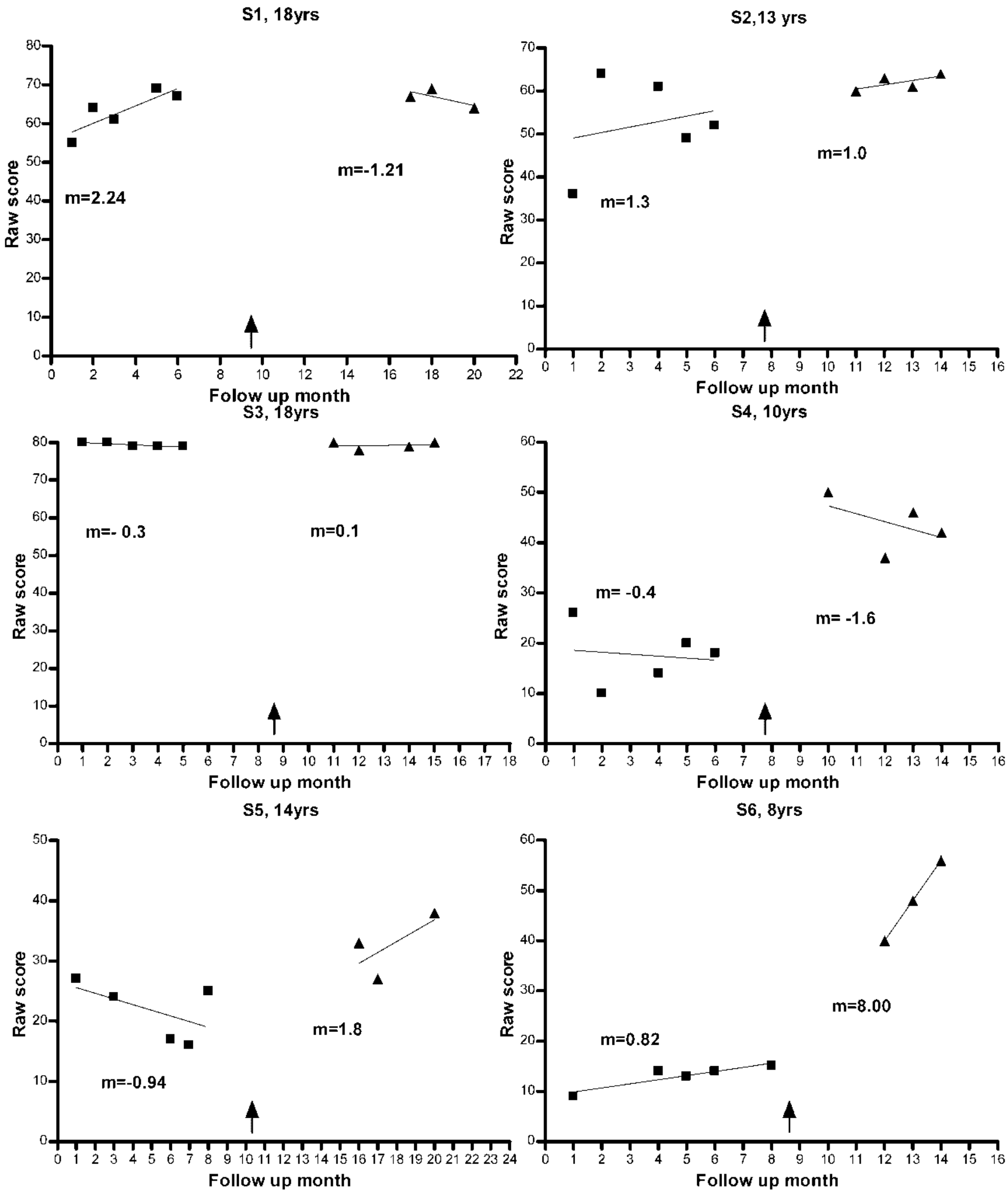

Figure 6-2c: Number list

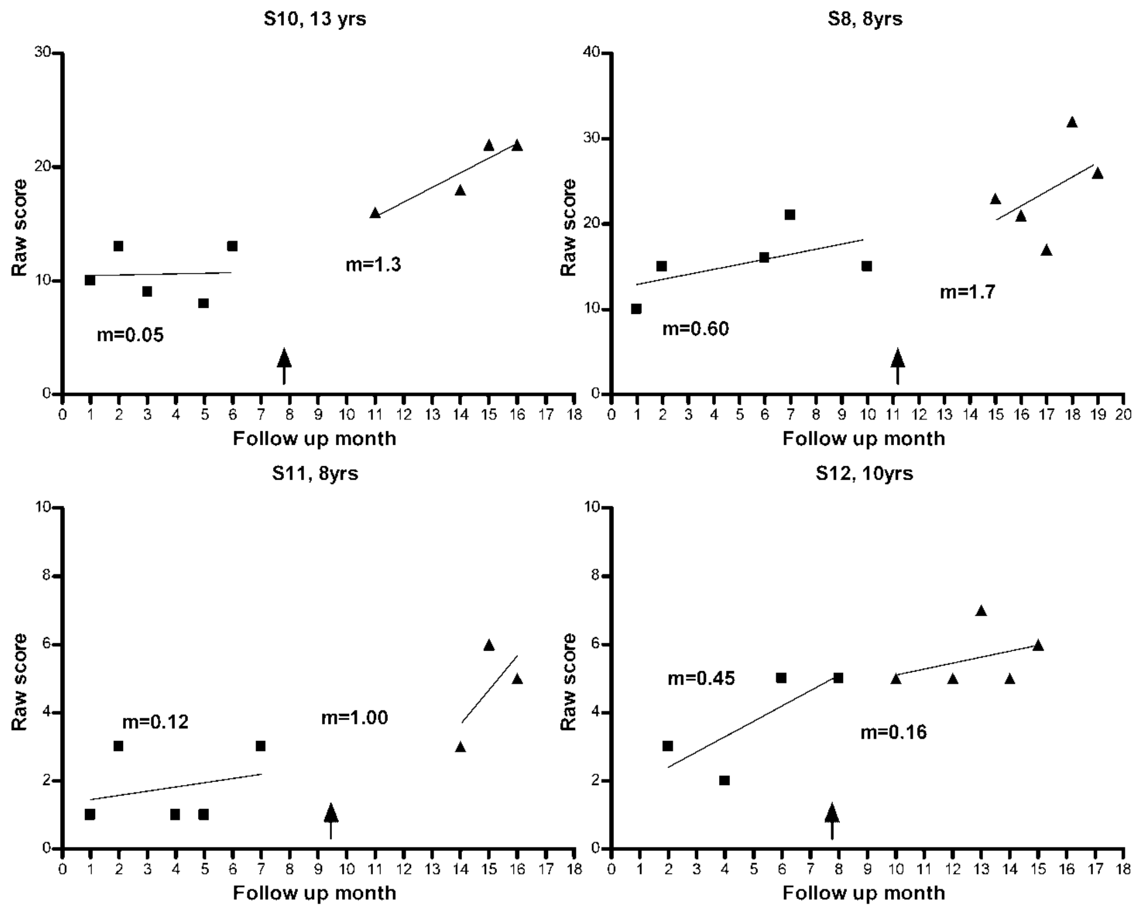

Figure 6-2: The raw scores obtained for each monthly probe visit by each participant are shown with respect to time, with time in months on the X-axis. The arrow indicates the month when bifocals were prescribed. The slopes (m) are shown on each figure.

When mean probe scores before and after bifocal provision were compared across subjects, there was a significant improvement in scores with bifocals for the Word Identification (WI), improving from 27.29±21.43 to 33.51±17.53, ($p=0.05$). The Dolch sight words improved from 41.2±27.42 to 55.5±16.86 ($p=0.025$) and the list of numbers from 7.89±6.27 to 13.63±9.57 ($p=0.023$).

Considering the data individually (Figure 6-2), it can be seen that two of the participants showed an obvious jump in either the slope of their scores or their average WI scores after bifocals were prescribed (S4 and S7, see Fig 6-2a). Similarly, 3 of the subjects showed such an improvement with the Dolch (S4, S5 and S6, Fig 6-2b) and 3 for the number test (S8, S10, S11, Fig 6-2c). The others (S1, S2, S3 and S12) showed little obvious change on any test. It is important to note that participant S3 experienced a ceiling effect on the Dolch sight words – she reached the upper limit of the test so that no further improvement could be measured. There were no significant correlations between the difference in the near visual acuity or accommodation exerted at the BF1and SV visits and the difference in mean probe scores before and after bifocals.

6.4.1.2 *Printing skills*

The means and standard deviations were calculated at each visit for letter size and position. Paired t-tests did not show any significant difference in the mean letter size or mean position of letters (letter size: $p=0.166$; letter position mean; $p=0.827$) and there was no change in the variability of letter size and position, i.e. there was no significant change in the standard deviations (letter size: $p=0.247$; letter position; $p=0.451$).

6.4.2 Video data:

6.4.2.1 *Main visits – Battery of tests*

Repeated measures ANOVAs across all subjects for the 3 main visits showed that there was a significant main effect of visit (figure 6-3) There was a decrease in the time taken to perform the task for the Word Identification *[F(2,14)=10.78,(p=0.0015)]*, Dolch sight words *[F(2,12)=3.94,(p=0.048)]* and there was a borderline significance for Visual closure *[F(2,20)=3.43,(p=0.052)]*. Post hoc (Bonferroni) tests showed that there was a significant decrease in the time taken to perform the WI task at the BF1 visit compared to SV (p=0.002) and at BF2 compared to SV (p=0.005) but no significant difference between BF1 and BF2 (p=1.00). The post hoc results for the Dolch sight words and Visual closure did not reach significance. There was also no significant difference in the time taken to perform all other literacy and visual perceptual skills across the three main visits. There were no significant correlations between the difference in the near visual acuity at the BF1and SV visits and the changes in time taken for the various battery sub-tests before and after bifocals. However there was a significant negative correlation between the time taken to complete the Word Identification task and both the lag of accommodation with SV lenses (r=-0.708, p=0.27) and the difference in the accommodative response before and after bifocal prescription (r= -0.813, *p*=0.004) i.e. those with poorer accommodative response and those who had a greater change in near focus benefitted more.

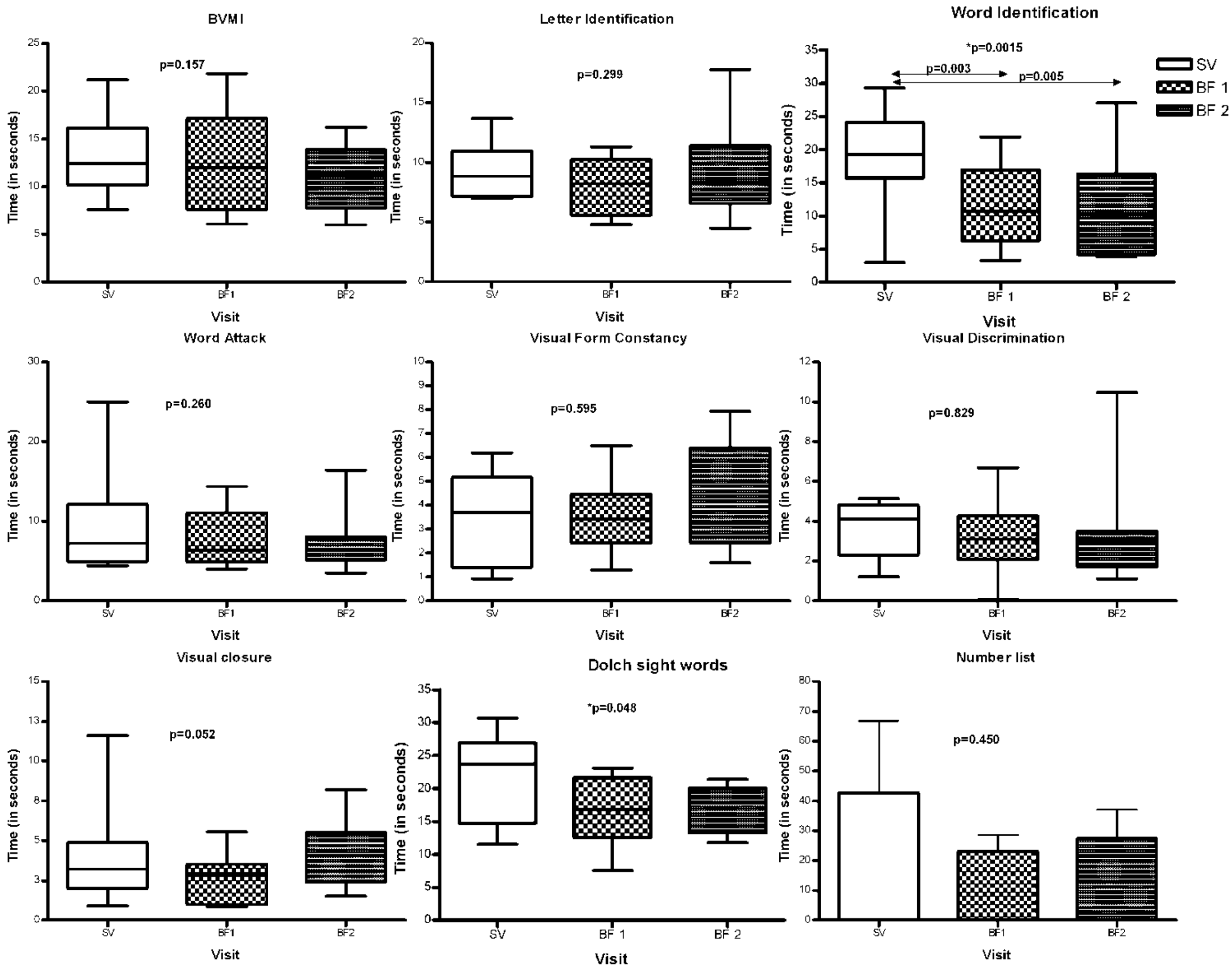

Figure 6-3: Box and whiskers plots of the average time (in seconds) taken to perform the battery of tests at the SV visit (white boxes), BF1 (chequered boxes) and BF2 (grey boxes). The significant p values are indicated by an asterisk. Significant post hoc results are indicated by double arrowhead lines. Boxes show the median, 25th and 75th percentile and whiskers show the lowest and highest values obtained, (except for number list where mean and SD is shown since only 3 participants underwent this test).

6.4.2.2 *Probes*

The results of the average time taken by each participant to complete each task in the probes before and after bifocals are shown in figure 6-4. Paired t-tests show that two out of the three literacy and numeracy tasks were completed faster with bifocals compared to the SV lenses namely, the number writing task ($p=0.001$) and Dolch sight words 2 ($p=0.025$), there being no significant difference for the WI test ($p>0.05$). It can be seen from Figure 4 that 6/10 children were faster in printing their names with the bifocals, 8/8 were faster at writing numbers, 7/7 were faster at the WI, 6/6 for the Dolch and 3/4 for the number test. There were no significant correlations between the difference in the near visual acuity or accommodative responses at the BF1and SV visits and the changes in time taken for the probes before and after bifocals.

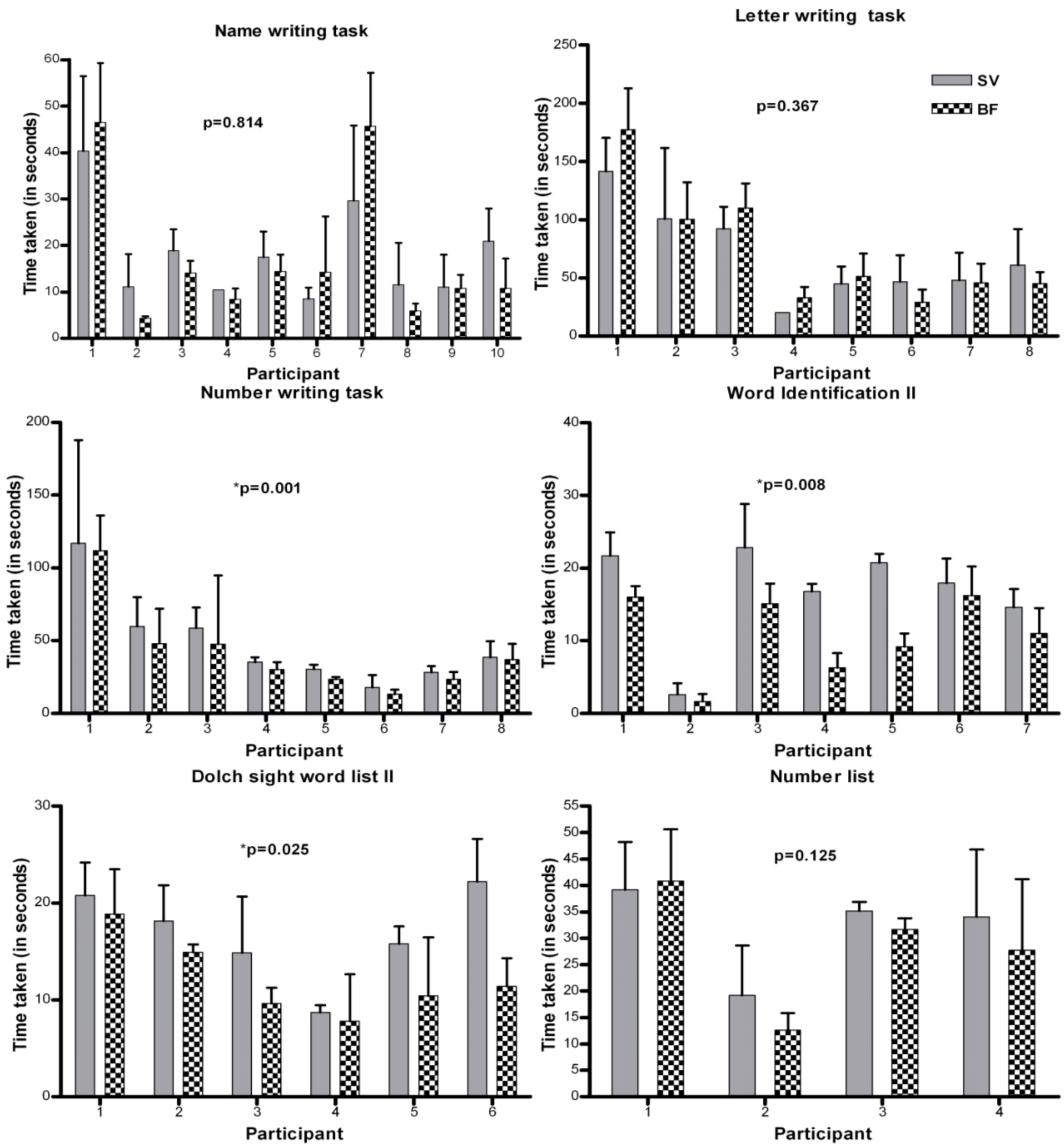

Figure 6-4 : Bar charts showing the average time taken to complete each task for each participant with SV lenses (shown by grey bars) and bifocals (shown by chequered bars). The p values are shown in each figure. Significant differences are indicated by an asterisk.

6.5 Discussion

In our understanding this is the first study of its kind to measure the impact of bifocals on early literacy, visual perceptual and printing skills. In addition, for the first time, reading and printing skills have been administered monthly and videotaped and timed before and after a bifocal prescription. We have previously reported that bifocals improve near visual acuity and performance on some literacy and visual perceptual tasks as measured in the battery of tests(Nandakumar & Leat, 2010),). In the present paper we have shown that there are also improvements in the monthly probe scores and the rate of progress of performance before and after bifocals as well as a decrease in the time taken to complete tasks after prescription of bifocals.

Our main outcome variable was the actual raw scores of the tests. In this report we show evidence of improvement with bifocals in the probe scores – the mean monthly probe scores improved with bifocals for the WI, Dolch and the number test across all subjects. This confirms our hypothesis that bifocals, which improve focus and give rise to improved near acuity, will lead to improvements of function in early literacy tests. Since these children were all receiving school instruction, it is possible that this progression may be due to natural learning over time. However, we have previously shown that there is little measurable improvement in skills in these children over a 5 month period (Nandakumar et al. *in submission*). It does seem likely, therefore that the changes we are observing in the current study are due to the bifocals and not natural progression. Another approach that was used to deal with the possibility of improvements due to natural progression was to consider the rate of learning, as measured by the slope of raw scores against time. We found that there was a significant increase in the slopes for the WI test across all participants and some observed improvements for individual subjects.

We did not find significant improvements for some of the measures that were used. This may be because of the variability of the performance of these children, which would mask small changes. However, it is interesting to observe that a number of individual participants demonstrated obvious changes in their rate of learning with bifocals (Figure 6-2) or a sudden jump in performance with bifocals (S6, S5, S4 on the Dolch; S7 on WI and S10 on the numbers). It is also noteworthy that no participant showed deterioration in his/her average performance. Those who showed these observable improvements were of a variety of ages (ranging from 8 to 13 years), which includes the majority of the participants' age range in this study. The two oldest children did not show improvements, but one of them (S3) was at the ceiling for the Dolch test. This variable performance could also be due to differences in when they were first introduced to formal learning, but this information was not available to us. Irrespective of their age, some younger children and teenagers are more skilled than their peers on reading and perceptual tasks. According to Buckley (Buckley. 2001) and Bochner (Bochner. 2001), children introduced to literacy in preschool years reach the highest level of achievement but individuals may make significant progress at any age into early adulthood if given an instruction (Van kraayenoord et al. 2000). This may indicate that introducing bifocals at any school age may be beneficial.

Our secondary outcome was the speed at which the children completed the tasks. On average the children performed the WI test faster with bifocals at both the main visits and after bifocal provision. Faster performance was also seen with the Dolch and the number writing test in the monthly probes. The visual perceptual tests administered in the study did not show quicker completion times with bifocals although a borderline significance was observed on the Visual closure subset.

For optometrists or ophthalmologists who are considering prescribing bifocals for such a child, it would be helpful to be able to predict which child would gain significant benefit. Our results show that it was not possible to predict this from improvements in VA. However a significant decrease in completion time with improvement in the accommodative response is seen with BF for the WI task. Similarly, those children with larger initial lags with the SV lenses, showed greater changes in time for completion of the WI task. This finding indicates that it is those children for whom there is greater initial lag and therefore greater improvement in focus who will gain the benefit of bifocals. However, since this was the only outcome parameter that correlated significantly with either VA or changes in accommodative lag, it appears that a bifocal correction should be considered for any child with DS with poor accommodation. Also there is potential for children of a variety of ages to obtain benefit.

We are not aware of any other literature on the progression of the formation of hand writing in children with DS or in typically developing individuals and hence we do not have a control group against which to compare our printing results. Individuals with DS have been reported to present with delayed motor skills which could result in poor finger coordination and poor formation of letters (Cowie. 1970, Davis & Kelso. 1982). It is possible that motor co-ordination is a greater determinant of handwriting than vision as we did not find any measurable improvement in letter formation of the printing tasks although the children wrote faster with the bifocals on the number writing task. Bergman & McLaughlin (Bergman & McLaughlin. 1988) report that even though conventional handwriting instruction would suffice for children to become good writers by 6-7 years of age, handwriting difficulties are very common in both typical and special education groups. We chose a task where there was no text to copy or trace but the participants had to write from memory as our interest pertained to observation of the formation of their printing for any improvement after a bifocal correction. Printing their names was still an attainable task as they would be taught to print their names and many would be required to print their names at school at various times.

6.6 Limitations of the study

The smaller number of participants in the study necessitates that the results be interpreted with caution. If anything, however, the small sample size would cause Type II errors – not finding differences that are present. In spite of the small sample, we were able to show changes after the bifocals. The small sample size was, in part, due to the commitment required for this study. It extended over 12-18 months and even 23 months in one participant and required monthly visits. This was demanding and some visits were inevitably missed due to bad weather or illness. Despite this, only one child missed many monthly visits and had to be excluded from the data analysis.

6.7 Conclusion

In this study, we have shown that bifocals, which improve near visual acuity and provide more accurate focus, enable children with DS to perform better for some reading tasks. There were increases in the rate of improvement for the whole group for one task (Word Identification) and improvements of the mean monthly scores for 3 tasks (WI, Dolch and numbers). There were observable improvements in some individual children. The children performed some tasks faster with bifocals (WI in the battery of tests, Dolch and printing for the monthly probes). Children with the greatest lags of accommodation before bifocal prescription showed the most benefit in time taken to complete the WI task, but this was the only outcome measure that could be predicted. So we conclude that a bifocal correction should be considered for all school-age children with DS who have reduced accommodation.

7 General discussion and conclusions

It has been previously reported that many children and teenagers with DS have high refractive errors. The results of Chapter 3 of this thesis confirm the presence of high refractive errors in Down syndrome. Generally there was a higher prevalence of hyperopia but there was a tail towards high myopia. Out of the 14 children who were recruited in the study 11 (79%) required a change in the prescription or were prescribed with new glasses. Six (43%) of the participants had incorrect prescriptions. Thus the finding in previous studies that many of these children still go uncorrected with or without an eye examination is confirmed. From a clinical perspective prescribing for hyperopic children has always been a matter of debate with respect to when and how much to prescribe. This is relevant to children with DS because of the high prevalence of high hyperopic errors. The Cardiff group corrected hyperopia of >3D in their participants with DS (Cregg et al. 2001, Woodhouse et al. 1997). However this is quite a high criterion for prescribing a correction both in individuals with or without DS. The current thesis used a more stringent criterion for changing the single vision spectacle correction, which was ≥1D of hyperopia or astigmatism and > 1D of myopia. In this thesis it is is recommended that even moderate or low amounts of hyperopia (>1D) should be corrected as there was evidence of improved near visual acuity with the increase in a hyperopic prescription. However there was no improvement in the distance visual acuity in this group of participants. The fact that the distance VA did not improve with a correction needs some explanation. It can be explained in terms of the distribution of refractive errors in the current study. There was a majority of hyperopes in the cohort which may explain why there was no significant improvement in the distance visual acuity albeit a clearer near acuity. Eight out of 11 participants who were given new glasses had a change in prescription in the positive direction as shown in Table 3-1 in Chapter 3. It may be that the children were able to accommodate for distance tasks for this small difference, but that they were not able to accommodate to overcome the extra demand for near. Hence the improvement at near was more notable. However, even though the distance VA was better than near VA before the prescription of new single vision glasses, this VA was still reduced compared to typically developing in the age group. The lower average distance VA after correction with no obvious associated physiological cause has been reported in this population (Courage et al. 1994, Tsiaras et al. 1999) with DS and is once again confirmed in this thesis.

There have been some suggestions about adverse reactions to some cycloplegics like atropine (Berg et al. 1959, Priest. 1960) and tropicamide (Sacks & Smith. 1989) and hence some authors have suggested that cycloplegics are generally contraindicated in individuals with DS (Barnard & Edgar. 1996). This suggestion is not confirmed for cyclopentolate , a cycloplegic used commonly in the typically developing population. In the present study, no adverse reactions occurred with either 2 drops of 0.5 or one drop of 1 % Cyclopentolate. Interestingly larger pupil sizes were observed in the present study. The average was 10mm with cyclopentolate compared to 7mm in a typically developing population (Gordon & Ehrenberg. 1954). The larger pupillary sizes with cyclopentolate, however, necessitates that the retinoscopic reflex should be judged in the central zone in order to avoid any errors in estimation. Thus there is evidence that cyclopentolate can be used in individuals with DS and may also produce accurate refraction results.

Reduced accommodation is another finding that has been reported repeatedly in this population and the correction of this was the prime focus of this study. The results described in Chapter 3 of this thesis yet again confirmed the presence of reduced accommodation which was found in 100% of the study participants. This reduced accommodative ability was present even with the newly prescribed SV lenses and was independent of refractive error.

In the next stage of the study bifocals were prescribed to correct for inaccurate accommodation. The limits of agreement between the measures of accommodation between the 2 observers in the current study are given in Appendix H. Bifocals were seen to significantly improve the accommodative accuracy showing that the bifocals helped in reducing the lag of accommodation especially at the lower accommodative demands. Unlike the initial report from the Cardiff group (Cregg et al. 2001), where the lag of accommodation remained constant after the single vision prescription was changed, in the current study the participants did not maintain a consistent level of accommodation. It was observed that they relaxed their accommodation to some extent when viewing through the bifocal segment and thus presented with neither a consistent level of accommodation nor a consistent lag. This finding was interesting as the bifocals were prescribed based on the lag of accommodation with SV lenses, i.e., the bifocal addition was prescribed such that the participant's accommodation was made to fall within the age-matched normal range for his/her habitual working distance. So it seems that these children may be relaxing their accommodation further, as soon as they start wearing their bifocals. This is, in fact, an opposite outcome to that found in the later studies from the Cardiff group (Al-Bagdady et al. 2009, Stewart et al. 2005), who found that bifocals appeared to stimulate the accommodative response in many individuals. Thus Chapter 5 of this thesis concludes that, even though a significant improvement in focus is seen, it is not sufficient to bring the response within the age-matched normal range of accommodative lag. This implies that the removal or discontinuation of bifocals was not clinically indicated in the current group of participants.

The cause of reduced accommodation in these individuals is not understood, but it does seem to be a different mechanism than presbyopia. In presbyopia, the amount of accommodation exerted remains the same at all times and this loss of accommodative ability is irreversible. In individuals with DS, however, the accommodative response is variable, that is, they modulate their accommodation (exert more or less accommodation) at different times and in different situations. Hence other possible explanations have to be considered. Woodhouse et al.,(Woodhouse et al. 2000) discuss the possibility of poorer levels of concentration exhibited by these children. It is possible that they produce normal accommodation according to their level of interest on the target but fail to maintain precision in their accommodative ability due to lack of motivation. They dismissed this explanation because increasing the cognitive load of the stimulus did not improve the accommodative accuracy in their study. Another possible explanation could be an abnormal interaction between accommodation and vergence in these individuals. This has not been studied in populations with DS. The interaction between the accommodation and vergence link has been studied in typically developing individuals and any changes in this link (a decrease or increase in the vergence) has been shown to cause a lag or lead of accommodation respectively (Schor. 1999). This is an area that could be investigated further. Abnormalities in the accommodation-convergence linkage may have possible effects on the accommodative ability of individuals with Down syndrome. Thus to date, it is still not known why accommodation is reduced in this population.

In contrast to previous reports from the Cardiff group of improved accommodative ability through the distance portion of the bifocals (Al-Bagdady et al. 2009, Stewart et al. 2005), none of the participants in the current study showed any obvious improvement in their accommodation response through the distance portion. Differences in protocol between the current study and that of the Cardiff group included: (1) Amount of the addition - the same add of 2.5D was prescribed for all the Cardiff study participants irrespective of the lag whereas customized adds based on the accommodative lag at the habitual working distance were used in the current study 2) The definition of improved or normal accommodation - the improvement in the accommodative response was considered to be normal in the Cardiff study if it was within the normal range in 2 out of the 3 distances tested (10, 6 and 4D). Alternatively, in the current study, the numerical measures of lags accommodation were considered at all the 4 distances tested (4, 6, 8 and 10 D) i.e. it was considered as a continuous variable, (3) The norms that were used for the age-matched accommodative response were different in the two studies and finally (4) The sample size and duration of the study - the Cardiff study had a larger sample size and greater duration of study.

In addition to improved accommodation, Chapter 5 also reported that the near visual acuity improved significantly with bifocals when compared with single vision lenses. It was observed that bifocals reduced the blur for near work and improved the near visual acuity thus confirming the 1st hypothesis of this thesis, which was that prescribing bifocals in children with DS will result in improved near visual acuity. It must be noted here that initially when SV glasses were prescribed in this group of participants, there was a significant difference in the distance and near VA (paired t-test, $p=0.008$). However it can be observed that after bifocal provision, the near visual acuity with bifocals is not significantly different from the distance visual acuity with SV lenses (t-test, $p=0.750$), that is, these 2 acuities are closer, showing that the bifocals definitely help in improving the near VA. Nevertheless this visual acuity is still reduced in comparison to the visual acuity of age-equivalent normals. Although no formal assessments of compliance were utilized in this thesis, improved compliance with bifocals was observed directly during the study visits in the laboratory as well as gathered from verbal reports from parents and teachers. This indicated that the children appreciated the improved clarity of near objects or near tasks, even though they did not verbally express this. There is a constant defocus at different distances other than infinity in these individuals during the period of development, which may cause reduced VA due to a subtle amblyopia. To understand the long term visual benefits of bifocals, studies of longer duration with participants of young age would be necessary and would show if bifocal wear would help in improving absolute levels of acuity.

Along with the measurement of accommodation and visual acuity, the natural progression of early literacy and printing skills has been studied and reported for the first time in this thesis. Chapter 4 describes the findings of the baseline battery of early literacy and visual perceptual skills and the natural progression of early literacy and basic printing skills after being prescribed SV lenses. Additional figures are shown in Appendix D-G. Appendices D and E show the natural progression of scores in the Dolch sight words and number list. The letter size and position mean and their standard deviations are shown in Appendices F-G, plotted across 6 months. Over the 6 month period with SV lenses, no significant improvements were seen on any of the early literacy or printing skills in the group as a whole although some progress was seen in a few individual participants (S6 in the Dolch sight words (shown in Appendix D), S4 and S6 in the Word Identification test (shown in Figure 4-3, Chapter 4)). The individual (S4) who showed a significant upward trend in the WI scores was given about 4.1 hours of reading instruction which was the second highest duration of reading instruction received in the group (the highest was 5.1 hours). Similar information about reading instruction was unavailable for the other participants who showed significant improvements with time. Hence it was not possible to study the influence of this factor on reading attainments in any systematic way. However in the other participants, there were no noteworthy improvements over time irrespective of the number of hours of reading instruction. Thus, there appeared to be no sign of learning or practice over the 6 months and hence the chances of a memory or practice effect affecting the scores (since the tests were repeated) were unlikely. This was important as any progress after bifocals could be attributed to the impact of bifocals.

Overall most participants were able to score on at least one test of early literacy. This was important as it allowed a measure of improvement to be made after bifocals were prescribed. For those who were unable to read any words, a number test, shown in Appendix B, was used. The raw scores obtained in the SV baseline showed considerable variability as a group but when adjusted for age, a more homogenous performance was observed (see Table 4-1 in Chapter 4). In other words, the group of participants perform more similarly to each other than would be anticipated by their age.

Against the understanding of little natural progression of early literacy and printing skills in this population, an improvement in performance was observed after bifocal prescription. The battery of tests administered with SV lenses was repeated again with bifocals both at the initial visit with bifocals as well as at 6^{th} months with bifocals and the findings are detailed in Chapter 5. The results of this chapter demonstrated that bifocals have a measureable, positive impact on the children's visual and school performance as shown by improved scores in some literacy tests. This confirmed the second hypothesis of the thesis – there was evidence of educational gains. This conclusion is also supported by the children's school reports - 9 out of 11 participants whose reports were available reported significant improvements at school (as shown by improved reading grade levels). The improvements in VA may have helped in improving the scores on reading and visual perceptual skills over time. Although it is not possible to measure if the improved performance was directly a result of the improved visual acuity, this could be deemed true as the improvement was observed just after VA improved. It must be noted here that the font sizes for all the reading tests were enlarged by a factor of 2 so that the print was within the acuity range of the children both with SV glasses and bifocals. This improvement in performance may be due to the improved visual acuity, decreased blur or less strain on the accommodation system (better focus). It may just be that the improved acuity and focus may have led to a greater ease, comfort and attention in the group of participants.

In order to test the 3rd hypothesis in the thesis, Chapter 6 determined the time taken to perform each task of reading, printing and visual perception from videotaped monthly and main visits with and without bifocals. This chapter described several unique approaches; 1) frequent testing of a few tests every month, 2) videotaping of all the sessions of literacy and visual perception (both monthly and main visits) in order to measure the time taken to complete the tasks and 3) assessment of printing skills, using a distinctive analysis of the quality of printing. There was evidence of improvement in the monthly probe scores as indicated by improved mean scores across all subjects on all the reading tests after prescribing bifocals. Yet again, this confirmed the hypothesis that bifocals resulted in improvements of function in early literacy tests. Although the factors of memory or practice were counted out by the results in Chapter 4, we still applied the original study design so that each participant would act as his/her own control. This was done by looking at changes in the slopes of the regression lines of their scores with respect to time. There was a significant increase in the slopes on the Word Identification subtest across all participants and some improvements in individual participants.

The printing task did not show any significant improvement in performance, i.e., there was no smaller or uniform sizing of letters indicating any development or maturity in writing with bifocals.

There was evidence of faster completion times for two tests of the test battery (WI and Dolch) and for 3 out the 6 monthly (WI, Dolch and the number writing task) probe tasks. When considering these participants individually as shown in figure 6-2 in Chapter 6, it was seen that a number of individual participants demonstrated obvious changes in their rate of learning with bifocals. This could once again be explained by the ease of performing near tasks with bifocals which helped to provide a better near acuity and better focusing ability.

From a clinical perspective, it is useful to know which child might benefit from a bifocal prescription. There was a significant correlation between the decrease in completion time and the improvement in the accommodative response with bifocals for the Word Identification task in Chapter 6. Additionally it was observed that children with larger initial lags with the SV lenses showed greater decreases in time taken for completion of the Word Identification task with their bifocals. This finding indicates that those children who present with greater initial lags of accommodation would be expected to gain a greater improvement in accommodation and would also be the ones who would gain most from a bifocal prescription. This is important as WI is a test of reading and it is noteworthy that an improved accommodative accuracy (less lag) has led to greater efficiency of performance in the scores on this reading test. In addition it was also observed that children of a variety of ages gained benefit from a bifocal prescription. Some might argue that all children with reduced accommodation should be offered a bifocal. This may be justified, as certainly it is true that none of the participants showed deterioration in performance with bifocals with any of the measures used in this study. It is important to remember that the current study did not include the lower functioning children and teenagers with DS and hence it is not possible to definitely extrapolate these findings to these individuals. Another question is the most appropriate way to determine the addition in this population. Clinically, it is recommended that the addition be prescribed based on the accommodative lag, considering the habitual working distance of each individual. This would ensure a more customized addition, in contrast to a same addition for all, irrespective of their levels of accommodative inaccuracy. This would be true, as in any clinical situation, a spectacle prescription is based on an individual's own error or condition, be it prescribing for refractive errors or strabismus. The prescription is never the same for everyone and is not the same lens for all. This should be the same when prescribing an addition in individuals with DS and it would be ideal if the add is customized to suit each individual's working distance and accommodative lag as described earlier. Thus, it is concluded that a bifocal prescription (or equivalent) is indicated in these individuals, but that the final prescription of any refractive correction is a decision based on clinical judgment and parental involvement.

From the findings in this thesis, it is suggested that clinicians should be at least as proactive in prescribing for hyperopia as for typically developing children, or in light of the reduced accommodation, be more proactive i.e., prescribe at a lower level of hyperopia. It is also recommended that a bifocal prescription should be considered in the clinical management of children and teenagers with Down syndrome who present with reduced accommodation. Both the full distance prescription and the bifocal may help to at least lower the risk of associated conditions such as strabismus and/or amblyopia in this population. This emphasizes the need to measure accommodative ability and correct for inaccurate accommodation in this population.

7.1 Future work

The current study was the first of its kind to measure the impact of bifocals on the educational attainments in a group of school children with Down syndrome, by observing a smaller number of participants and utilizing frequent measures of skill sets. From the findings in this initial study, there are possibilities of further studies and this section gives recommendations for future work.

7.2 Multifocals in children and young adults with DS

The current study could be replicated with the use of multifocal/ progressive addition lenses (PALs) instead of bifocals. PALs are used widely by many presbyopes as an alternative choice to bifocals. They have the advantage of providing a power for a range of intermediate working distances along with distance and near. In other words, clearer intermediate focus as provided by the PAL might be useful in many individuals who pursue interests such as painting or computing. This is the main functional advantage of PALs over bifocals which only provide focus for two discreet distances, usually distance and near. In addition, PALs are appealing cosmetically as there is no visible line on the lens, thus not showing that the person is wearing a presbyopic correction. Also in both the young and older individuals a better cosmetic appearance might be more helpful in motivating regular wearing of their correction. PALs could be prescribed for individuals with DS. The current thesis used bifocal lenses and we now know of their impact on academic attainments this population. Although the option of progressive addition lenses for correcting reduced accommodation has been suggested previously (Haugen et al. 2001), there have been no studies to measure the efficacy of PALs in individuals with DS. A future study, therefore, would be to replicate the current study with PALs instead of bifocals.

Being the first study of its kind, the present study included a wide range of tasks, to be certain to capture improvement. Future studies could be more focused on the outcome measures and use a few tasks to measure each aspect of literacy and visual perception. The same basic study design could be used with the following improvements or alternatives:

- Indirect measures from the current study show good compliance with bifocals. This outcome measure could be formalized with the use of a regularly administered questionnaire to parents and teachers.
- Choosing and utilizing a few tests from each task e.g., a reading task, a perceptual task, a printing task instead of multiple tasks of the same kind. This would help in reducing the time at each session and reduce fatigue. From the results of the current study we know that the almost all participants could do at least one reading task and many could not do the decoding task (Word Attack). Hence the Word Attack could be removed from the list of tests that would be administered. One reading task such as Word Identification, a test for visual motor integration such as the BVMI, two tests in the TVPS instead of three and a list of numbers for those who are unable to do any words could be used.
- For the printing task, a copying task instead of a task involving printing from memory could be used. This way, only the written aspect would be tested rather than including a cognitive demand while printing.
- For the follow up visits, one reading task, words or numbers, and one simple printing task could be utilized. Follow ups could be bimonthly instead of monthly. This would reduce the demands on the participants. The high demand of frequent visit was one reason why there were some missed visits in the current study

The suggested PAL study would be a good replication of the current study but would not show if PALS or bifocals are better or are similar to each other. Other study designs would have to be used to answer this questions, such as :

1) A randomized case controlled study with a cross over design, where one group of participants are prescribed bifocals and the other are prescribed with PALs and after a certain period of time (ideally 6 months or more) they would cross over and be prescribed with PALs or bifocals respectively.
2) A second design would be a full randomized clinical trial, either with two or a three-arms where the participants are prescribed with either PALs or bifocals (two arms) or including a single vision group as well as the control group (three arms).

7.3 Conclusion

This was also one of the few studies that assessed reading, printing and perceptual skills together with measures of literacy administered every month. To conclude, this thesis provides evidence of improved near visual acuity, accommodative accuracy and improved performance and rate of performance on tests of literacy with bifocals. Indirect measures of compliance also showed improved compliance with bifocal lenses compared to single vision lenses. Thus the clinical standard of care must change for individuals with DS. The clinical standard of care should include the measurement of accommodation and that clinicians be more ready to prescribe bifocals to children and young adults with DS who present with reduced accommodation. More studies are required to understand the long term impact of bifocals in individuals with DS and in similar populations.

References

References to Chapter 1&2

Al-Bagdady M, Stewart RE, Watts P, Murphy PJ & Woodhouse JM (2009): Bifocals and Down's syndrome: Correction or treatment? Ophthalmic Physiol Op **29**: 416-421.

Apell RJ. (1975): Clinical application of bell retinoscopy. J Am Optom Assoc **46** : 1023-1027.

Atkinson J. (1993): Infant vision screening: prediction and prevention of strabismus and amblyopia from refractive screening in the Cambridge Photorefraction Program. In Simons K. (ed). Early visual development, normal and abnormal. New York,Oxford University Press. 335-348.

Atkinson J, Anker S, Bobier W, Braddick O, Durden K, Nardini M & Watson P. (2000): Normal emmetropization in infants with spectacle correction for hyperopia. Invest Ophthalmol Vis Sci **41**: 3726.

Atkinson J, Anker S, Braddick O, Rutherford M, Edwards D & Cowan F. (2004): Deficits in selective visual attention and dorsal stream function: children with hyperopic refractive errors in infancy and children born prematurely. Invest Ophthalmol Vis Sci **45**: 3511.

Atkinson J, Anker S, Nardini M, Braddick O, Hughes C, Rae S, Wattam-Bell J & Atkinson S. (2002): Infant vision screening predicts failures on motor and cognitive tests up to school age. Strabismus **10**: 187-198.

Atkinson J, Braddick O, Nardini M & Anker S. (2007) Infant Hyperopia: Detection, Distribution, Changes and Correlates-Outcomes From the Cambridge Infant Screening Programs. Optom Vis Sci **84**: 84-96.

Atkinson J, Braddick O, Durden K, Watson P & Atkinson S. (1984): Screening for refractive errors in 6-9 month old infants by photorefraction. Br Med J **68**: 105.

Atkinson J, Braddick O, Robier B, Anker S, Ehrlich D, King J, Watson P & Moore A. (1996): Two infant vision screening programmes: prediction and prevention of strabismus and amblyopia from photo- and videorefractive screening. Eye (Lond) **10** : 189-198.

Baddeley A. (1998) Working memory. Comptes Rendus de lAcademie des Sciences Series III Sciences de la Vie **321**: 167-173.

Bailey N, Gunn P, Tovey C & Jobling A. (1989): Screening the vision of special school children with Down syndrome. J Intellect Dev Disabil **15**: 49-55.

Balkany TJ, Downs MP, Jafek BW & Krajicek MJ. (1979): Hearing loss in Down's syndrome. A treatable handicap more common than generally recognized. Clin Pediatr **18**: 116-118.

Banks MS. (1980): The development of visual accommodation during early infancy. Child Dev **51**: 646-666.

Becker L, Mito T, Takashima S & Onodera K. (1991) Growth and development of the brain in Down syndrome. Prog Clin Biol Res **373**: 133-152.

Berk AT, Saatci AO, Erçal MD, Tunç M & Ergin M. (1996): Ocular findings in 55 patients with Down's syndrome. Ophthalmic Genet **17**: 15-19.

Berninger V, Yates C, Cartwright A, Rutberg J, Remy E & Abbott R. (1992): Lower-level developmental skills in beginning writing. Read Writ **4**: 257-280.

Berninger VW, Abbott RD, Abbott SP, Graham S & Richards T. (2002): Writing and Reading: Connections Between Language by Hand and Language by Eye. J Learn Disabil **35**: 39.

Berninger VW, Cartwright AC, Yates CM, Swanson HL & Abbott RD. (1994): Developmental skills related to writing and reading acquisition in the intermediate grades. Read Writ **6**: 161-196.

Berninger V & Swanson H. (1994): Modifying Hayes and Flower's model of skilled writing to explain beginning and developing writing. Advances in cognition and educational practice **2**: 57-81.

Best TK, Siarey RJ & Galdzicki Z. (2007): Ts65Dn, a mouse model of Down syndrome, exhibits increased GABAB-induced potassium current. J Neurophysiol **97**: 892.

Best TK, Cho-Clark M, Siarey RJ & Galdzicki Z. (2008): Speeding of miniature excitatory post-synaptic currents in Ts65Dn cultured hippocampal neurons. Neurosci Lett **438**: 356-361.

Bharadwaj SR & Candy TR. (2008): Cues for the control of ocular accommodation and vergence during postnatal human development. J Vis **8**: 14.1.

Bieber J. (1974): Why nearpoint retinoscopy with children? Optom Wkly **173**: 78-82.

Blum HL,Peters HB & Bettman JW.(1968): Vision screening for elementary schools: The Orinda study. Berkeley: University of California Press.

Bobier W. (1990): Eccentric photorefraction: a method to measure accommodation of highly hypermetropic infants. Clin Vision Sci 5: 45–66.

Borish I. (1970): Clinical refraction, 3rd edition. Chicago: The Professional Press **2**: 172.

Borish I, Benjamin W & Benjamin W. (1998): Borish's Clinical Refraction. Philadelphia, WB Saunders.

Boudreau D. (2002): Literacy skills in children and adolescents with Down syndrome. Read Writ **15**: 497-525.

Braddick O, Atkinson J, French J & Howland HC. (1979): A photorefractive study of infant accommodation. Vision Res **19**: 1319-1330.

Brookman KE. (1983): Ocular accommodation in human infants. Am J Optom Physiol Opt **60**: 91-99.

Brooks D. (1972): Hearing Loss and Middle Ear Disorders in Patients with Down. J Ment Defic Res **16**: 21-29.

Browder DM & Xin YP. (1998): A meta-analysis and review of sight word research and its implications for teaching functional reading to individuals with moderate and severe disabilities J Spec Educ **32**: 130.

Brown N. (1973): The change in shape and internal form of the lens of the eye on accommodation. Exp Eye Res **15**: 441-459.

Buckley S (2001): Reading and Writing for Individuals with Down Syndrome: An Overview.The Down Syndrome Educational Trust.

Byrne A, MacDonald J & Buckley S. (2002): Reading, language and memory skills: a comparative longitudinal study of children with Down syndrome and their mainstream peers. Br J Educ Psychol **72**: 513-529.

Caputo AR, Wagner RS, Reynolds DR, Guo S & Goel AK. (1989): Down Syndrome: Clinical Review of Ocular Features. Clin Pediatr **28**: 355.

Carr J. (1970): Mental and motor development in young mongol children. J Intellect Disabil Res **14**: 205-220.

Casey W, Jones D, Kugler B & WatkinS B. (1988): Integration of Down's syndrome children in the primary school: a longitudinal study of cognitive development and academic attainments. Br J Educ Psychol **58**: 279-286.

Catalano RA. (1992): In Pueschel, S.M. & Pueschel, J.K. (Eds).Ophthalmologic concerns. Biomedical concerns in persons with Down syndrome.

Ciuffreda KJ. (1991): Accommodation and its anomalies. Visual Optics and Instrumentation : 231–279.

Ciuffreda K. (1998): Accommodation, the pupil, and presbyopia. Borish’s clinical refraction : 77–120.

Ciuffreda K & Kenyon R. (1983): Accommodation Vergence and Accommodation in Normals, Amblyopes and Strabismics. In: C. M. Schor, & K. J. Ciuffreda (eds). Vergence Eye Movements: Basic and Clinical Aspects. Boston: Butterworths: 101-173.

Cossu G, Rossini F & Marshall JC. (1993): When reading is acquired but phonemic awareness is not: A study of literacy in Down's syndrome. Cognition **46**: 129-138.

Cotter SA. (2007): Management of childhood hyperopia: a pediatric optometrist's perspective. Optom Vis Sci **84**: 103-109.

Courage ML, Adams RJ, Reyno S & Kwa PG. (1994): Visual acuity in infants and children with Down syndrome. Dev Med Child Neurol **36**: 586-593.

Cowie VA. (1970): A Study of the early development of Mongols.London, England : Pergamon.

Cregg M, Woodhouse JM, Pakeman VH, Saunders KJ, Gunter HL, Parker M, Fraser WI & Sastry P. (2001): Accommodation and refractive error in children with Down syndrome: cross-sectional and longitudinal studies. Invest Ophthalmol Vis Sci **42**: 55-63.

Cregg M, Woodhouse JM, Stewart RE, Pakeman VH, Bromham NR, Gunter HL, Trojanowska L, Parker M & Fraser WI. (2003): Development of refractive error and strabismus in children with Down syndrome. Invest Ophthalmol Vis Sci **44**: 1023-1030.

Cunningham C & McArthur K. (1981): Hearing loss and treatment in young Down's syndrome children. Child Care Health Dev **7**: 357-374.

Cupples L & Iacono T. (2000): Phonological awareness and oral reading skill in children with Down syndrome. J Speech Lang Hear Res **43**: 595-608.

Currie DC & Manny RE. (1997): The development of accommodation. Vision Res **37**: 1525-1533.

Czepita D, Lodygowska E & Czepita M. (2008): Are children with myopia more intelligent? A literature review. Ann Acad Med Stetin **54**: 13-16.

da Cunha RP, Moreira J. (Aug 1996): Ocular findings in Down's syndrome. Am J Ophthalmol **2**: 236-244.

Dahle AJ & McCollister FP. (1986): Hearing and Otologic Disorders in Children with Downs Syndrome. Am J Ment Defic **90**: 636-642.

Down JL. (1866): Observations on an ethnic classification of idiots. London Hospital Clinical Lectures and Reports **3**: 259-262.

Doyle SJ, Bullock J, Gray C, Spencer A & Cunningham C. (1998): Emmetropisation, axial length, and corneal topography in teenagers with Down's syndrome. Br Med J **82**: 793-796.

Duffen I. (1976): Teaching reading to children with little or no language. Remedial Educ **11**: 139-142.

Dwyer P & Wick B. (1995): The influence of refractive correction upon disorders of vergence and accommodation. Optom Vis Sci **72**: 224-232.

Dykens EM, Rosner BA & Ly TM. Drawings by Individuals With Williams Syndrome. (2001): Are People Different From Shapes? Am J Ment Retard **106**: 94-107.

Eames TH. (1955): The influence of hypermetropia and myopia on reading achievement. Am J Ophthalmol **39**: 375-377.

Ehrlich DL, Braddick OJ, Atkinson J, Anker S, Weeks F, Hartley T, Wade J & Rudenski A. (1997): Infant emmetropization: longitudinal changes in refraction components from nine to twenty months of age. Optom Vis Sci **74**: 822-843.

Elliott D. (2003): Clinical Procedures in Primary Eye Care : New York:Butterworth-Heinemann.

Fanning G. (1971): Vision in children with Down's syndrome. Aust J Optom 54: 74-82.

Fincham E. (1951): The accommodation reflex and its stimulus. Br J Ophthalmol **35**: 381-393.

Fisher R. (1973): Presbyopia and the changes with age in the human crystalline lens. J Physiol (Lond) **228**: 765.

Fowler A, Doherty B & Boynton L. (1995): The basis of reading skill in young adults with Down syndrome. Down syndrome: Living and learning in the community : 182–196.

French AN, Rose KA, Burlutsky G, Mitchell P, Sydney Childhood Eye Study. (2009): Does correction of hyperopia affect the pattern of children's activities, and does this differ from that in emmetropic children? Invest Ophthalmol Vis Sci **50.** E-abstract 3961.

Garcia A & Cacho P. (2002): MEM and Nott dynamic retinoscopy in patients with disorders of vergence and accommodation. Ophthalmic Physiol Opt **22**: 214-220.

Gardiner PA. (1967): Visual defects in cases of Down's syndrome and in other mentally handicapped children. Br J Ophthalmol **51**: 469-474.

Garner LF. (1983): Mechanisms of accommodation and refractive error. Ophthalmic Physiol Opt **3**: 287-293.

Garzia R & Nicholson S. (1990): Visual function and reading disability: an optometric viewpoint. J Am Optom Assoc **61**: 88-97.

Glasser A & Campbell C. (1999): Biometric, optical and physical changes in the isolated human crystalline lens with age in relation to presbyopia. Vision Res **39**: 1991-2015.

Glasser A & Campbell MCW. (1998): Presbyopia and the optical changes in the human crystalline lens with age. Vision Res **38**: 209-229.

Gray LS, Winn B, Gilmartin B & Eadie AS. (1993): Objective concurrent measures of open-loop accommodation and vergence under photopic conditions. Invest Ophthalmol Vis Sci **34**: 2996-3003.

Grosvenor T. (1970): Refractive State, Intelligence Test Scores, and Academic Ability. Am J Optom Arch Am Acad Optom **47**:355-361.

Gwiazda J, Thorn F, Bauer J & Held R. (1993): Emmetropization and the progression of manifest refraction in children followed from infancy to puberty. Clin Vis Sci **8**: 337-344.

Gwiazda J, Thorn F & Held R. (2005): Accommodation, accommodative convergence, and response AC/A ratios before and at the onset of myopia in children. Optom Vis Sci **82**: 273.

Haugen OH, Hovding G & Lundstrom I. (2001): Refractive development in children with Down's syndrome: a population based, longitudinal study. Br J Ophthalmol **85**: 714-719.

Haugen OH & Hovding G. (2001): Strabismus and binocular function in children with Down syndrome. A population-based, longitudinal study. Acta Ophthalmol Scand **79**: 133-139.

Haugen OH, Hovding G & Riise R. (2004): Ocular changes in Down syndrome. Tidsskr Nor Laegeforen **124**: 186-188.

Haugen OH, Høvding G & Eide GE. Biometric measurements of the eyes in teenagers and young adults with Down syndrome. Acta Ophthalmol Scand **79**: 616-625.

Hayden A & Dmitriev V. (1975): The multidisciplinary preschool program for Down’s syndrome children at the University of Washington Model Preschool Center. Except Infant **3**: 193–221.

Haynes H, White BL & Held R. (1965): Visual accommodation in human infants. Science **148**: 528-530.

Heath GG. (1956): Components of accommodation. Am J Optom Arch Am Acad Optom **33**: 569-579.

Henderson SE, Morris J & Frith U. (1981): The motor deficit in Down's syndrome children: A problem of timing. J Child Psychol Psychiatry **22**: 233-244.

Hosaka A. (1988): The growth of the eye and its components. Japanese studies. Acta Ophthalmol Suppl **185**: 65-68.

Howland H. Early refractive development. Early Visual Development, Normal and Abnormal, K.Simmons, ed.Oxford U.Press, New York, 1993 : 5–11.

Howland HC. (1985): Optics of photoretinoscopy: results from ray tracing. Am J Optom Physiol Opt **62**: 621-625.

Howland HC, Dobson V & Sayles N. (1987): Accommodation in infants as measured by photorefraction. Vision Res **27**: 2141-2152.

Hung GK, Ciuffreda KJ & Rosenfield M. (1996): Proximal contribution to a linear static model of accommodation and vergence. Ophthalmic Physiol Opt **16**: 31-41.

Ingram R, Arnold P, Dally S & Lucas J. (1991): Emmetropisation, squint, and reduced visual acuity after treatment. Br Med J **75**: 414.

Ingram R, Gill L & Lambert T. (2000): Effect of spectacles on changes of spherical hypermetropia in infants who did, and did not, have strabismus. Br J Ophthalmol 84(3): 324.

Jaeger E. (1980) Ocular findings in Down's syndrome. Trans Am Ophthalmol Soc **78**: 808.

Jarrold C & Baddeley AD. (1997): Short-term memory for verbal and visuo-spatial information in Down’s syndrome. Cogn Neuropsychiatry **2**: 101-122.

John FM, Bromham N, Candy T & Woodhouse J. (2002): Steady-state VEP and behavioural measures of visual acuity in infants and children with Down syndrome. Ophthalmic Physiol Opt **22**: 572-572.

John FM, Bromham NR, Woodhouse JM & Candy TR. (2004): Spatial vision deficits in infants and children with Down syndrome. Invest Ophthalmol Vis Sci **45**: 1566-1572.

Juel C. (1980): Comparison of word identification strategies with varying context, word type, and reader skill. Read Res Q **15**: 358-376.

Kay-Raining Bird E, Cleave P & McConnell L. (2000): Reading and phonological awareness in children with Down syndrome: A longitudinal study. Am J Speech Lang Pathol **9**: 319-330.

Kay-Raining Bird E, Cleave PL, White D, Pike H & Helmkay A. (2008): Written and oral narratives of children and adolescents with Down syndrome. J Speech Lang Hear Res **51**: 436-450.

Klein BP. (1999): Contrasting Patterns of Cognitive Abilities of 9- and 10-Year-Olds With Williams Syndrome or Down Syndrome. Dev Neuropsychol **16**: 177-196.

Kleinstein R. (1987): Epidemiology of Presbyopia. Presbyopia: Recent research and reviews from the third international symposium. New York: Professional Press Books : 12-18.

Korenberg J, Chen X, Schipper R, Sun Z, Gonsky R, Gerwehr S, Carpenter N, Daumer C, Dignan P & Disteche C. (1994): Down syndrome phenotypes: The consequences of chromosomal imbalance. Proc Natl Acad Sci U S A **91**: 4997-5001.

Koretz J, Cook C & Kaufman P. (1997): Accommodation and presbyopia in the human eye. Changes in the anterior segment and crystalline lens with focus. Invest Ophthalmol Vis Sci **38**: 569.

Kruger PB, Aggarwala KR, Bean S & Mathews S. (1997): Accommodation to stationary and moving targets. Optom Vis Sci **74**: 505-510.

Kruger PB & Pola J. (1985): Changing target size is a stimulus for accommodation. J Opt Soc Am A **2**: 1832-1835.

Laws G. (2000): Language and Memory Development in Children with Down Syndrome at Mainstream Schools and Special Schools: a comparison. Educ Psychol **20**: 447-457.

Laws G & Gunn D. (2002): Relationships between reading, phonological skills and language development in individuals with Down syndrome: A five year follow-up study. Read Writ **15**: 527-548.

Leat SJ, Shute RH & Westall CA. (1999): Assessing Children's Vision: A Handbook : Butterworth.

Leat SJ. (1996): Reduced accommodation in children with cerebral palsy. Ophthalmic Physiol Opt **16**: 385-390.

Leat SJ & Gargon JL. (1996): Accommodative response in children and young adults using dynamic retinoscopy. Ophthalmic Physiol Opt **16**: 375-384.

Leat SJ & Mohr A. (2007): Accommodative Response in pre-presbyopes with visual impairment and its clinical implications. Invest Ophthalmol Vis Sci **48**: 3888-96.

Lejeune, J.Gautier M., and Turpin, R. (1959): Étude des chromosomes somatiques de neuf enfants mongoliens. Compt Rend Acad Sci **248**: 1721-1722.

Lindstedt E. (1983): Failing accommodation in cases of Down's syndrome: A preliminary report. Ophthalmic Genet **3**: 191-192.

Little JA, Woodhouse JM, Lauritzen JS & Saunders KJ. (2007): The impact of optical factors on resolution acuity in children with Down syndrome. Invest Ophthalmol Vis Sci **48**: 3995-4001.

Locke LC & Somers W. (1989) A comparison study of dynamic retinoscopy techniques. Optom Vis Sci 66(8): 540-544.

Lorenz S, Sloper T & Cunningham C. (1985): Reading and Down's Syndrome. Br J Spec Educ**12**: 65-67.

Lowe RF. (1949): The eyes in mongolism. Br J Ophthalmol **33**: 131.

Maddock R, Millodot M, Leat S & Johnson C. (1981): Accommodation responses and refractive error. Invest Ophthalmol Vis Sci **20**: 387.

Marcell MM & Armstrong V. (1982): Auditory and visual sequential memory of Down syndrome and nonretarded children. Am J Ment Defic **87**: 86-95.

Marcell MM, Harvey CF & Cothran LP. (1988): An attempt to improve auditory short-term memory in Down's syndrome individuals through reducing distractions. Res Dev Disabil **9**: 405-417.

Mathews S & Kruger PB. (1994): Spatiotemporal transfer function of human accommodation. Vision Res **34**: 1965-1980.

McClelland JF & Saunders KJ. (2004): Accommodative lag using dynamic retinoscopy: age norms for school-age children. Optom Vis Sci **81**: 929.

McClelland JF & Saunders KJ. (2003): The repeatability and validity of dynamic retinoscopy in assessing the accommodative response. Ophthalmic Physiol Opt **23**: 243-250.

McLin LN,Jr, Schor CM & Kruger PB. (1988): Changing size (looming) as a stimulus to accommodation and vergence. Vision Res **28**: 883-898.

Mutti DO. (2007): To emmetropize or not to emmetropize? The question for hyperopic development. Optom Vis Sci **84**: 97.

Nott I. (1925): Dynamic skiametry, accommodation and convergence. Am J Optom Physiol Opt **6**: 490-503.

Pau H & Kranz J. (1991): The increasing sclerosis of the human lens with age and its relevance to accommodation and presbyopia. Graefes Arch Clin Exp Ophthalmol **229**: 294-296.

Paudel N, Leat SJ, Adhikari P, Woodhouse JM & Shrestha JB. (2010): Visual defects in Nepalese children with Down syndrome. Clin Exp Optom **93**: 83-90.

Phillips S & Stark L. (1977): Blur: a sufficient accommodative stimulus. Documenta Ophthalmol **43**: 65-89.

Pueschel S & Gieswein S (1993): Ocular disorders in children with Down syndrome. Downs Syndr Res and Pract **1**: 129-132.

Pueschel S. (1981): Ophthalmological aspects in Down syndrome. Down Syndrome: Papers and Abstracts for Professionals **4**: 4-7.

Rabin J, Van Sluyters RC & Malach R. (1981): Emmetropization: a vision-dependent phenomenon. Invest Ophthalmol Vis Sci **20**: 561-564.

London R & Wick B. (1998): Patients with amblyopia and strabismus. Borish’s clinical refraction : 1119–1133.

Robaei D, Kifley A, Rose KA & Mitchell P. (2006): Refractive error and patterns of spectacle use in 12-year-old Australian children. Ophthalmol **113**: 1567-1573.

Robert B, Johnson A & Tsuetaki TK. (1984): Comparison of laser and infrared techniques for measurement of the resting focus of accommodation: mean differences and long-term variability. Ophthal Physiol Opt **4**: 327-332.

Rohr A & Burr DB. (1978): Etiological differences in patterns of psycholinguistic development of children of IQ 30 to 60. Am J Ment Defic **82**: 549-553.

Roizen NJ, Mets MB & Blondis TA. (1994): Ophthalmic disorders in children with Down syndrome. Dev Med Child Neurol **36**: 594-600.

Roizen NJ & Patterson D. (2003): Down's syndrome. The Lancet **361**: 1281-1289.

Roizen NJ, Wolters C, Nicol T & Blondis TA. (1993): Hearing loss in children with Down syndrome. J Pediatr **123**: 9-12.

Rosenfield M, Ciuffreda K & Hung G. (1991): The linearity of proximally induced accommodation and vergence. Invest Ophthalmol Vis Sci **32**: 2985.

Rosenfield M & Logan N. (2009): Optometry: Science, Techniques and Clinical Management : Butterworth-Heinemann.

Rosenfield M & Cohen AS. (1995): Push-up amplitude of accommodation and target size. Ophthalmic Physiol Opt **15**: 231-232.

Rosenfield M, Portello JK, Blustein GH & Jang C. (1996): Comparison of clinical techniques to assess the near accommodative response. Optom Vis Sci **73**: 382-388.

Rosner J & Rosner J. (1997): The relationship between moderate hyperopia and academic achievement: how much plus is enough? J Am Optom Assoc **68**: 648-650.

Rosner J & Rosner J. (1987): Comparison of visual characteristics in children with and without learning difficulties. Am J Optom Physiol Opt **64**: 531-533.

Rosner J & Rosner J. (1989): Relation between tonic accommodation and visual perceptual skills development in 6- to 12-year-old children. Optom Vis Sci **66**: 526-529.

Sago H, Carlson EJ, Smith DJ, Kilbridge J, Rubin EM, Mobley WC, Epstein CJ & Huang TT. (1998): Ts1Cje, a partial trisomy 16 mouse model for Down syndrome, exhibits learning and behavioral abnormalities. Proc Natl Acad Sci U S A **95**: 6256-6261.

Saladin JJ. (1998) Phorometry and Stereopsis. Borish’s clinical refraction : 724-773.

Saunders KJ, Margaret Woodhouse J & Westall CA. (1995): Emmetropisation in human infancy: rate of change is related to initial refractive error. Vision Res **35**: 1325-1328.

Saw SM, Tan SB, Fung D, Chia KS, Koh D, Tan DTH & Stone RA. (2004): IQ and the association with myopia in children. Invest Ophthalmol Vis Sci **45**: 2943.

Schapero M, Cline D & Hofstetter HW. (1980): Dictionary of Visual Science, 3rd Ed. In: Anonymous : Radnor, PA: Chilton Bppk Co: **20**, 657.

Shankar S, Evans MA & Bobier WR. (2007): Hyperopia and emergent literacy of young children: pilot study. Optom Vis Sci **84**: 1031-1038.

Shapiro MB & France TD. (1985): The ocular features of Down's syndrome. Am J Ophthalmol **99**: 659-663.

Shepperdson B. (1994): Attainments in reading and number of teenagers and adults with Down syndrome. Down Syndr Res Pract **2**: 97-101.

Shott SR, Joseph A & Heithaus D. (2001): Hearing loss in children with Down syndrome. Int J Pediatr Otorhinolaryngol **61**: 199-205.

Siderov J & Johnston AW. (1990): The importance of the test parameters in the clinical assessment of accommodative facility. Optom Vis Sci **67**: 551-557.

Sivak JG & Bobier CW. (1978): Accommodation and chromatic aberration in young children. Invest Ophthalmol Vis Sci **17**: 705-709.

Sloper P, Cunningham C, Turner S & Knussen C. (1990): Factors related to the academic attainments of children with Down's syndrome. Br J Educ Psychol **60**: 284-298.

Smith G. (2006): Refraction and visual acuity measurements: What are their measurement uncertainties? Clin Exp Optom **89**: 66-72.

Smith-Bindman R, Hosmer W, Feldstein VA, Deeks JJ & Goldberg JD. (2001): Second-trimester ultrasound to detect fetuses with Down syndrome a meta-analysis. JAMA **285**: 1044-1055.

Spano M, Mercuri E, Rando T, Panto T, Gagliano A, Henderson S & Guzzetta F. (1999): Motor and perceptual-motor competence in children with Down syndrome: variation in performance with age. Eur J Paediatr Neurol **3**: 7-13.

Sriubienė M, Andriuškevičiūtė I & Sinkus A. Age-related dynamics of eye refraction in children with Down syndrome. Biologija **1:** 22-24.

Sterner B, Gellerstedt M & Sjostrom A. (2004) The amplitude of accommodation in 6-10-year-old children-not as good as expected. Ophthalmic Physiol Opt **24**: 246-251.

Stewart RE, Margaret Woodhouse J & Trojanowska LD. (2005): In focus: The use of bifocal spectacles with children with Down's syndrome. Ophthalmic Physiol Opt **25**: 514-522.

Stewart RE, Woodhouse JM, Cregg M & Pakeman VH. (2007): Association between accommodative accuracy, hypermetropia, and strabismus in children with Down's syndrome. Optom Vis Sci **84**: 149-155.

Suttle CM & Turner AM. (2004): Transient pattern visual evoked potentials in children with Down's syndrome Ophthalmic and Physiological Optics **24**: 91-99.

Swanson HL & Berninger VW. (1996): Individual Differences in Children's Working Memory and Writing Skill. J Exp Child Psychol **63**: 358-385.

Teasdale TW, Fuchs J & Goldschmidt E. (1988): Degree of myopia in relation to intelligence and educational level. Lancet **2**: 1351-1354.

Teller D (1997): First glances: the vision of infants. the Friedenwald lecture Invest Ophthalmol Vis Sci **38**: 2183.

Theodore Grosvenor. (1996): Primary Care Optometry: The Binocular VisionEamination. In: Anonymous : Butterworth-Heinemann, Boston, MA: 319-319.

Tsiaras WG, Pueschel S, Keller C, Curran R & Giesswein S. (1999): Amblyopia and visual acuity in children with Down's syndrome. Br Med J **83**: 1112.

Turner S & Alborz A. (2003): Academic attainments of children with Downs syndrome: A longitudinal study. Br J Educ Psychol **73**: 563-583.

Van Kraayenoord CE, Moni KB, Jobling A & Ziebarth K. (2000) Broadening approaches to literacy education for young adults with Down syndrome. In Monica Cuskelly, Anne Jobling, and Sue Buckley (Eds.),Down Syndrome Across the Life Span .

van Splunder J, Stilma JS, Bernsen RM, Arentz TG & Evenhuis HM. (2003): Refractive errors and visual impairment in 900 adults with intellectual disabilities in the Netherlands. Acta Ophthalmol Scand **81**: 123-129.

Varnhagen CK, Das J & Varnhagen S. (1987) Auditory and visual memory span: Cognitive processing by TMR individuals with Down syndrome or other etiologies. Am J Ment Defic **91**: 398-405.

Wallace DK, Chandler DL, Beck RW, Arnold RW, Bacal DA, Birch EE, Felius J, Frazier M, Holmes JM & Hoover D. (2007): Treatment of bilateral refractive amblyopia in children three to less than 10 years of age. Am J Ophthalmol **144**: 487-496.

Wang P, Doherty S, Rourke S & Bellugi U. (1995): Unique Profile of Visuo-Perceptual Skills in a Genetic Syndrome. Brain Cogn **29**: 54-65.

Wharry RE & Kirkpatrick SW. (1986): Vision and academic performance of learning disabled children. Percept Mot Skills **62**: 323-336.

Williams W, Latif A, Hannington L & Watkins D. (2005): Hyperopia and educational attainment in a primary school cohort. Br Med J **90**: 150.

Williams SM, Sanderson GF, Share DL & Silva PA. (1988): Refractive error, IQ and reading ability: a longitudinal study from age seven to 11. Dev Med Child Neurol **30**: 735-742.

Wiseman FK, Alford KA, Tybulewicz VLJ & Fisher E. (2009): Down syndrome-Recent progress and future prospects. Hum Mol Genet **18**: R75.

Woodhouse JM, Cregg M, Gunter HL, Sanders DP, Saunders KJ, Pakeman VH, Parker M, Fraser WI & Sastry P. (2000): The effect of age, size of target, and cognitive factors on accommodative responses of children with Down syndrome. Invest Ophthalmol Vis Sci **41**: 2479-2485.

Woodhouse JM, Meades JS, Leat SJ & Saunders KJ. (1993): Reduced accommodation in children with Down syndrome. Invest Ophthalmol Vis Sci **34**: 2382-2387.

Woodhouse JM, Pakeman VH, Cregg M, Saunders KJ, Parker M, Fraser WI, Sastry P & Lobo S. (1997): Refractive errors in young children with Down syndrome. Optom Vis Sci **74**: 844-851.

Woodhouse JM, Pakeman VH, Saunders KJ, Parker M, Fraser WI, Lobo S & Sastry P. (1996): Visual acuity and accommodation in infants and young children with Down's syndrome. J Intellect Disabil Res **40** : 49-55.

Young FA (1963): Reading, measures of intelligence and refractive errors. Am J Optom Arch Am Acad Optom **40**: 257-264.

References to Chapter 3

http://www.teachnology.com/worksheets/language_arts/dolch/list/. (Accessed 13 February 08).

http://www.precision-vision.com/index.cfm. (Accessed 13 February 2008).

Atkinson J, Anker S, Nardini M, Braddick O, Hughes C, Rae S, Wattam-Bell J & Atkinson S (2002): Infant vision screening predicts failures on motor and cognitive tests up to school age. Strabismus **10**: 187-198.

Barnard S & Edgar D (1996): Pediatric Eye Care: Blackwell Science.

Beery KE & Beery NA (2004): The Beery-Buktenica Developmental Test of Visual-Motor Integration (VMI) –5th edition, NCS Pearson, Minneapolis.

Berg JM, Brandon MW & Kirman BH (1959): Atropine in mongolism. Lancet **2**: 441-442.

Bochner S (2001): A study of functional literacy skills in young Adults with Down syndrome. Int J Dis Dev Educ **48**: 67-90.

Boudreau D (2002): Literacy skills in children and adolescents with Down syndrome. Reading and Writing **15**: 497-525.

Byrne A, MacDonald J & Buckley S.(2002): Reading, language and memory skills: A comparative longitudinal study of children with Down syndrome and their mainstream peers. Br J Educ Psychol **72**: 513-529.

Caputo AR, Wagner RS, Reynolds DR, Guo S & Goel AK (1989): Down syndrome: Clinical Review of Ocular Features. Clin Pediatr **28**: 355.

Catalano RA (1992) In Pueschel SM & Pueschel JK (Eds).Ophthalmologic concerns. Biomedical concerns in persons with Down syndrome.

Chauhan K & Charman WN (1995): Single figure indices for the steady-state accommodative response. Ophthalmic Physiol Opt **15**: 217-221.

Choi TB, Lee DA, Oelrich FO, Amponash D, Bateman JB & Christensen RE (1995): A retrospective study of eye disease among first grade children in Los Angeles. J Am Optom Assoc **66**: 484-488.

Cotter SA.(2007): Management of childhood hyperopia: a pediatric optometrist's perspective. Optom Vis Sci **84**: 103-109.

Courage ML, Adams RJ, Reyno S & Kwa PG (1994): Visual acuity in infants and children with Down syndrome. Dev Med Child Neurol **36**: 586-593.

Cregg M, Woodhouse JM, Pakeman VH, Saunders KJ, Gunter HL, Parker M, Fraser WI & Sastry P (2001): Accommodation and refractive error in children with Down syndrome: cross-sectional and longitudinal studies. Invest Ophthalmol Vis Sci **42**: 55-63.

Cregg M, Woodhouse JM, Stewart RE, Pakeman VH, Bromham NR, Gunter HL, Trojanowska L, Parker M & Fraser WI (2003): Development of refractive error and strabismus in children with Down syndrome. Invest Ophthalmol Vis Sci **44**: 1023-1030.

Cupples L & Iacono T (2000): Phonological Awareness and Oral Reading Skill in Children With Down Syndrome. Journal of Speech, Language and Hearing Research **43**: 595-608.

da Cunha RP & Moreira J (Aug 1996): Ocular findings in Down's syndrome. Am J Ophthalmol **2**: 236-244.

Deacon M, Woodhouse J & Watts P (2005): Investigation of ocular alignment and binocular single vision in children with Down's syndrome. Int Congr Ser **1282**: 88-92.

Doughty MJ (2001): Ocular Pharmacology and Therapeutics: A Primary Care Guide: Butterworth-Heinemann Medical.

Down JL (1866): Observations on an ethnic classification of idiots. London Hospital Clinical Lectures and Reports **3**: 259-262.

Dunn LM & Dunn LM (1997): Peabody Picture Vocabulary Test-(PPVT-III). Circle Pines, MN: American Guidance Service.

Dwyer P & Wick B (1995): The influence of refractive correction upon disorders of vergence and accommodation. Optom Vis Sci **72**: 224-232.

Egashira SM, Kish LL,Twelker JD, Mutti DO, Zadnik K & Adams AJ (1993): Comparison of cyclopentolate versus tropicamide cycloplegia in children. Optom Vis Sci **70**: 1019-1026.

Fidler DJ, Most DE & Guiberson MM (2005): Neuropsychological correlates of word identification in Down syndrome. Res Dev Disabil **26**: 487-501.

Filips R (2008): Low Plus Lenses for Children. Ophthalmology **115**: 222-223.

Fotedar R, Rochtchina E, Morgan I, Wang JJ, Mitchell P & Rose KA (2007): Necessity of cycloplegia for assessing refractive error in 12-year-old children: a population-based study. Am J Ophthalmol **144**: 307-309.

Gardiner PA (1967) Visual defects in cases of Down's syndrome and in other mentally handicapped children. Br J Ophthalmol 51(7): 469-474.

Gardner MF (1996): TVPS-R, Test of Visual-Perceptual Skills (Non-Motor)-Revised: Psychological & Educational Publications.

Gordon DM & Ehrenberg MH (1954): Cyclopentolate hydrochloride: A new mydriatic and cycloplegic agent; a pharmacologic and clinical evaluation. Am J Ophthalmol **38**: 831-838.

Goss DA & Zhai H (1994): Clinical and laboratory investigations of the relationship of accommodation and convergence function with refractive error. A literature review. Doc Ophthalmol **86**: 349-380.

Gwiazda J, Thorn F, Bauer J & Held R (1993): Emmetropization and the progression of manifest refraction in children followed from infancy to puberty. Clinical vision sciences **8**: 337-344.

Gwiazda J, Scheiman M, Mohindra I & Held R (1984): Astigmatism in children: changes in axis and amount from birth to six years. Invest Ophthalmol Vis Sci **25**: 88-92.

Harris WS & Goodman RM (1968): Hyper-reactivity to atropine in Down's syndrome. N Engl J Med **279**: 407-410.

Haugen OH, Hovding G & Lundstrom I (2001): Refractive development in children with Down's syndrome: a population based, longitudinal study. Br J Ophthalmol **85**: 714-719.

Haugen OH & Hovding G (2001): Strabismus and binocular function in children with Down syndrome. A population-based, longitudinal study. Acta Ophthalmol Scand **79**: 133-139.

Haugen OH, Hovding G & Riise R (2004): Ocular changes in Down syndrome. Tidsskr Nor Laegeforen **124**: 186-188.

Hayden A & Dmitriev V (1975): The multidisciplinary preschool program for Down’s syndrome children at the University of Washington Model Preschool Center. Exceptional infant **3**: 193–221.

John FM, Bromham NR, Woodhouse JM & Candy TR (2004): Spatial vision deficits in infants and children with Down syndrome. Invest Ophthalmol Vis Sci **45**: 1566-1572.

Jones LA, Mitchell GL, Mutti DO, Hayes JR, Moeschberger ML & Zadnik K (2005): Comparison of ocular component growth curves among refractive error groups in children. Invest Ophthalmol Vis Sci **46**: 2317-2327.

Kay-Raining Bird E, Cleave P & McConnell L (2000): Reading and phonological awareness in children with Down syndrome: A longitudinal study. American Journal of Speech-Language Pathology **9**: 319-330.

Kim JH, Hwang JM, Kim HJ & Yu YS (2002): Characteristic ocular findings in Asian children with Down syndrome. Eye **16**: 710-714.

Klein BP (1999): Contrasting Patterns of Cognitive Abilities of 9- and 10-Year-Olds with Williams Syndrome or Down Syndrome. Dev Neuropsychol **16**: 177-196.

Kleinstein RN, Jones LA, Hullett S, Kwon S, Lee RJ, Friedman NE, Manny RE, Mutti DO, Yu JA & Zadnik K (2003) Refractive error and ethnicity in Children. Arch Ophthalmol **121**: 1141.

Laatikainen L & Erkkila H (1980): Refractive errors and other ocular findings in school children. Acta Ophthalmol (Copenh) **58**: 129-136.

Laws G (2000): Language and Memory Development in Children with Down Syndrome at Mainstream Schools and Special Schools: a comparison. EducPsychol **20**: 447-457.

Leat SJ, Shute RH & Westall CA (1999): Assessing Children's Vision: A Handbook: Butterworth.

Leat SJ & Mohr A (2007): Accommodative Response in pre-presbyopes with visual impairment and its clinical implications. Invest Ophthalmol Vis Sci **48**: 3888-96.

Lindstedt E (1983): Failing accommodation in cases of Down's syndrome: A preliminary report. Ophthalmic Genet **3**: 191-192.

Liza-Sharmini AT, Azlan ZN & Zilfalil BA (2006): Ocular findings in Malaysian children with Down syndrome. Singapore Med J **47**: 14-19.

Maurer D, Lewis T & Brent H (1989): The effects of deprivation on human visual development: Studies of children treated for cataracts. Applied Developmental Psychology **3**: 139–227.

Mohd-Ali B, Mohammed Z, Norlaila M, Mohd-Fadzil N, Rohani CC & Mohidin N (2006): Visual and binocular status of Down syndrome children in Malaysia. Clin Exp Optom **89**: 150-154.

Nagtzaam L & Vink M (1998): Visual and hearing impairment in people with intellectual disability. Current Opinion in Psychiatry **11**: 527-530.

North RV & Kelly ME (1987): A review of the uses and adverse effects of topical administration of atropine. Ophthalmic Physiol Opt **7**: 109-114.

Priest JH (1960): Atropine response of the eyes in mongolism. Am J Dis Child **100**: 869-872.

Pueschel S & Gieswein S (1993): Ocular disorders in children with Down syndrome. Downs Syndr Res and Pract **1**: 129-132.

Robaei D, Kifley A, Rose KA & Mitchell P (2006): Refractive error and patterns of spectacle use in 12-year-old Australian children. Ophthalmology **113**: 1567-1573.

Rosenfield M, Ciuffreda K & Hung G (1991): The linearity of proximally induced accommodation and vergence. Invest Ophthalmol Vis Sci **32**: 2985.

Rosner J & Rosner J (1997): The relationship between moderate hyperopia and academic achievement: how much plus is enough? J Am Optom Assoc **68**: 648-650.

Sacks B & Smith S (1989): People with Down's syndrome can be distinguished on the basis of cholinergic dysfunction. J Neurol Neurosurg Psychiatry **52**: 1294-1295.

Shankar S, Evans MA & Bobier WR. (2007): Hyperopia and emergent literacy of young children: Pilot study. Optom Vis Sci **84**: 1031-1038.

Shapiro MB & France TD (1985): The ocular features of Down's syndrome. Am J Ophthalmol **99**: 659-663.

Spano M, Mercuri E, Rando T, Panto T, Gagliano A, Henderson S & Guzzetta F (1999): Motor and perceptual-motor competence in children with Down syndrome: Variation in performance with age. Eur J Paediatr Neurol **3**: 7-13.

Stephen E, Dickson J, Kindley AD, Scott CC & Charleton PM (2007): Surveillance of vision and ocular disorders in children with Down syndrome. Dev Med Child Neurol **49**: 513-515.

Stewart RE, Margaret Woodhouse J & Trojanowska LD. (2005): In focus: the use of bifocal spectacles with children with Down's syndrome. Ophthalmic Physiol Opt **25**: 514-522.

Stewart RE, Woodhouse JM, Cregg M & Pakeman VH (2007): Association between accommodative accuracy, hypermetropia, and strabismus in children with Down's syndrome. Optom Vis Sci **84**: 149-155.

Suryakumar R & Bobier WR (2003): The manifestation of noncycloplegic refractive state in pre-school children is dependent on autorefractor design. Optom Vis Sci **80**: 578-586.

Tsiaras WG, Pueschel S, Keller C, Curran R & Giesswein S (1999): Amblyopia and visual acuity in children with Down's syndrome. Br Med J **83**: 1112.

Turner S & Alborz A (2003) Academic attainments of children with Down’s syndrome: A longitudinal study. Br J Educ Psychol **73**: 563-583.

van Splunder J, Stilma JS, Bernsen RM, Arentz TG & Evenhuis HM (2003): Refractive errors and visual impairment in 900 adults with intellectual disabilities in the Netherlands. Acta Ophthalmol Scand **81**: 123-129.

Wallace DK, Chandler DL, Beck RW, Arnold RW, Bacal DA, Birch EE, Felius J, Frazier M, Holmes JM & Hoover D (2007): Treatment of Bilateral Refractive Amblyopia in Children Three to Less Than 10 Years of Age. Am J Ophthalmol **144**: 487-496.

Wang P, Doherty S, Rourke S & Bellugi U (1995): Unique Profile of Visuo-Perceptual Skills in a Genetic Syndrome. Brain Cogn **29**: 54-65.

Williams SM, Sanderson GF, Share DL & Silva PA (1988): Refractive error, IQ and reading ability: a longitudinal study from age seven to 11. Dev Med Child Neurol **30**: 735-742.

Woodcock R. Woodcock Reading Mastery Tests-Revised (WRMT-R/NU) (1998): Circle Pines, Minnesota: American Guidance Service.

Woodhouse JM, Griffiths C & Gedling A (2000): The prevalence of ocular defects and the provision of eye care in adults with learning disabilities living in the community. Ophthalmic Physiol Opt **20**: 79-89.

Woodhouse JM, Meades JS, Leat SJ & Saunders KJ (1993): Reduced accommodation in children with Down syndrome. Invest Ophthalmol Vis Sci **34**: 2382-2387.

Woodhouse JM, Pakeman VH, Cregg M, Saunders KJ, Parker M, Fraser WI, Sastry P & Lobo S (1997): Refractive errors in young children with Down syndrome. Optom Vis Sci **74**: 844-851.

Zadnik K, Manny RE, Yu JA, Mitchell GL, Cotter SA, Quiralte JC, Shipp MD, Friedman NE, Kleinstein RN & Walker TW (2003): Ocular component data in schoolchildren as a function of age and gender. Optom Vis Sci **80**: 226-236.

Zadnik K, Mutti DO & Adams AJ (1992): The repeatability of measurement of the ocular components. Invest Ophthalmol Vis Sci **33**: 2325-2333.

References to Chapter 4

http://www.teachnology.com/worksheets/language_arts/dolch/list/. (Accessed 13 February 08)

http://www.precision-vision.com/index.cfm (Accessed 13 February 08).

Atkinson J, Braddick O, Nardini M & Anker S (2007): Infant Hyperopia: Detection, Distribution, Changes and Correlates-Outcomes From the Cambridge Infant Screening Programs. Optom Vis Sci **84**: 84.

Beery K (1997): The Beery-Buktenica Test of Visual-Motor Integration with Supplemental Developmental Tests of Visual Perception and Motor Coordination.

Berninger V, Yates C, Cartwright A, Rutberg J, Remy E & Abbott R (1992): Lower-level developmental skills in beginning . Read Writ **4**: 257-280.

Berninger VW, Abbott RD, Abbott SP, Graham S & Richards T (2002): and Reading: Connections Between Language by Hand and Language by Eye. J Learn Disabil **35**: 39.

Berninger VW, Cartwright AC, Yates CM, Swanson HL & Abbott RD (1994): Developmental skills related to and reading acquisition in the intermediate grades. Read Writ **6**: 161-196.

Bochner S(2001): A Study of Functional Literacy Skills in Young Adults with Down Syndrome. Int J Dis Dev Educ **48**: 67-90.

Boudreau D (2002): Literacy skills in children and adolescents with Down syndrome. Read Writ 15(5): 497-525.

Buckley S & Bird G (1993): Teaching children with Down's syndrome to read. Downs Syndr Res Pract **1**: 34-41.

Buckley S (2001): Reading and for Individuals with Down syndrome: An Overview : *Down Syndrome Issues and Information.* doi:10.3104/9781903806098. The Down syndrome Educational Trust; 2001. (Accessed January 23, 2010).

Byrne A, MacDonald J & Buckley S (2002): Reading, language and memory skills: a comparative longitudinal study of children with Down syndrome and their mainstream peers. Br J Educ Psychol **72**: 513-529.

Caputo AR, Wagner RS, Reynolds DR, Guo S & Goel AK (1989): Down Syndrome: Clinical review of ocular features. Clin Pediatr **28**: 355.

Casey W, Jones D, Kugler B & Watkins B (1988): Integration of Down's syndrome children in the primary school: a longitudinal study of cognitive development and academic attainments. Br J Educ Psychol **58**: 279-286.

Catalano RA (1992): In Pueschel, SM & Pueschel, JK (Eds).Ophthalmologic concerns. Biomedical concerns in persons with Down syndrome.

Catts HW, Fey ME, Tomblin JB & Zhang X (2002): A Longitudinal Investigation of Reading Outcomes in Children With Language Impairments. J Speech Lang Hear Res **45**: 1142-1157.

Catts HW & Kamhi AG (1999): Language and Reading Disabilities : Allyn and Bacon. Boston, MA

Chapman R (2006): Language learning in Down syndrome: The speech and language profile compared to adolescents with cognitive impairment of unknown origin. Downs Syndr Res Pract **10**: 61-66.

Cowie VA. (1970) A Study of the Early Development of Mongols. London, England: Pergamon.

Cunningham CC, Glenn S, Lorenz S, Cuckle P & Shepperdson B (1998) :Trends and outcomes in educational placements for children with Down syndrome. Eur J Spec Needs Educ **13**: 225-237.

Cupples L & Iacono T. (2000) Phonological Awareness and Oral Reading Skill in Children With Down Syndrome. Journal of Speech, Language and Hearing Research 43(3): 595-608.

da Cunha RP, Moreira J. (Aug 1996) Ocular findings in Down's syndrome. Am J Ophthalmol 2(122): 236-244.

Davis WE & Kelso JA (1982):Analysis of "invariant characteristics" in the motor control of Down's syndrome and normal subjects. J Mot Behav **14**: 194-212.

Duffen I (1976): Teaching reading to children with little or no language. Remedial Educ (3): 139-142.

Dunn LM & Dunn LM (1997): Peabody Picture Vocabulary Test-(PPVT-III). Circle Pines, MN: American Guidance Service .

Dykens EM, Rosner BA & Ly TM. Drawings by Individuals With Williams Syndrome (2001): Are People Different From Shapes? Am J Ment Retard **106**: 94-107.

Elliott CD (1983): British Ability Scales: Spelling Scale.Windsor:NFER-Nelson Pub. Co.

Fidler DJ, Most DE & Guiberson MM (2005): Neuropsychological correlates of word identification in Down syndrome. Res Dev Disabil **26**: 487-501.

Fowler A (1995): Linguistic variability in persons with Down syndrome: Research and implications. Down syndrome: Living and learning in the community : 121–131.

Gardiner PA (1967): Visual defects in cases of Down's syndrome and in other mentally handicapped children. Br J Ophthalmol **51**: 469-474.

Haugen OH, Hovding G & Lundstrom I (2001): Refractive development in children with Down's syndrome: a population based, longitudinal study. Br J Ophthalmol **85**: 714-719.

Haugen OH & Hovding G (2001): Strabismus and binocular function in children with Down syndrome. A population-based, longitudinal study. Acta Ophthalmol Scand **79**: 133-139.

Haugen OH, Hovding G & Riise R (2004): Ocular changes in Down syndrome. Tidsskr Nor Laegeforen **124**: 186-188.

Hayden A & Dmitriev V(1975): The multidisciplinary preschool program for Down's syndrome children at the University of Washington Model Preschool Center. Except Infant **3**: 193-221.

Kay-Raining Bird E, Cleave P & McConnell L (2000): Reading and phonological awareness in children with Down syndrome: A longitudinal study. Am J Speech Lang Pathol **9**: 319-330.

Kay-Raining Bird E, Cleave PL, White D, Pike H & Helmkay A (2008): Written and oral narratives of children and adolescents with Down syndrome. J Speech Lang Hear Res **51**: 436-450.

Klein BP (1999): Contrasting Patterns of Cognitive Abilities of 9- and 10-Year-Olds With Williams Syndrome or Down Syndrome. Dev Neuropsychol **16**: 177-196.

Laws G (2000): Language and Memory Development in Children with Down Syndrome at Mainstream Schools and Special Schools: a comparison. Educ Psychol **20**: 447-457.

Laws G & Gunn D (2004): Phonological memory as a predictor of language comprehension in Down syndrome: a five-year follow-up study. J Child Psychol Psychiatry **45**: 326-337.

Lindstedt E (1983): Failing accommodation in cases of Down's syndrome: A preliminary report. Ophthalmic Genet **3**: 191-192.

Lorenz S, Sloper T & Cunningham C. (1985) Reading and Down's Syndrome. Br J Spec Educ **12**: 65-67.

Margaret J Snowling, Hannah M Nash and Lisa M Henderson (2008): The development of literacy skills in children with Down syndrome: Implications for intervention. Downs Syndr Res Pract : 62-67.

Miolo G, Chapman RS & Sindberg HA (2005): Sentence comprehension in adolescents with Down syndrome and typicallydeveloping children role of sentence voice, visual context, and auditory-verbal short-term memory. J Speech Lang Hear Res **48**: 172-188.

Nandakumar K & Leat SJ (2009): Bifocals in Down Syndrome Study (BiDS): Design and baseline visual function. Optom Vis Sci **86**: 196-207.

Pueschel S & Gieswein S. (1993): Ocular disorders in children with Down syndrome. Downs Syndr Res and Pract **1**: 129-132.

Shankar S, Evans MA & Bobier WR (2007): Hyperopia and emergent literacy of young children: pilot study. Optom Vis Sci **84**: 1031-1038.

Shapiro MB & France TD (1985): The ocular features of Down's syndrome. Am J Ophthalmol **99**: 659-663.

Shepperdson B (1994): Attainments in reading and number of teenagers and adults with Down syndrome. Downs Syndr Res Pract **2**: 97-101.

Sloper P, Cunningham C, Turner S & Knussen C (1990): Factors related to the academic attainments of children with Down's syndrome. Br J Educ Psychol **60**: 284-298.

Spano M, Mercuri E, Rando T, Panto T, Gagliano A, Henderson S & Guzzetta F (1999): Motor and perceptual-motor competence in children with Down syndrome: variation in performance with age. Eur J Paediatr Neurol **3**: 7-13.

Swanson HL & Berninger VW (1996): Individual differences in children's working memory and skill. J Exp Child Psychol **63**: 358-385.

Turner S & Alborz A (2003): Academic attainments of children with Downs syndrome: A longitudinal study. Br J Educ Psychol **73**: 563-583.

Van Kraayenoord CE, Moni KB, Jobling A & Ziebarth K (2000): Broadening approaches to literacy education for young adults with Down syndrome. In Monica Cuskelly, Anne Jobling, and Sue Buckley (Eds). Down syndrome across the life span.

Wang P, Doherty S, Rourke S & Bellugi U (1995): Unique Profile of Visuo-Perceptual Skills in a Genetic Syndrome. Brain Cogn **29**: 54-65.

Woodcock R. Woodcock Reading Mastery Tests-Revised (WRMT-R/NU) (1998): Circle Pines, Minnesota: American Guidance Service.

Woodhouse JM, Meades JS, Leat SJ & Saunders KJ (1993): Reduced accommodation in children with Down syndrome. Invest Ophthalmol Vis Sci **34**: 2382-2387.

References to Chapter 5

Al-Bagdady M, Stewart RE, Watts P, Murphy PJ & Woodhouse JM (2009): Bifocals and Down's syndrome: correction or treatment? Ophthalmic Physiol.Op **29**: 416-421.

Beery K.E. Beery &Beery N.A (2004): The Beery-Buktenica Developmental Test of Visual-Motor Integration (VMI)–5th edition, NCS Pearson, Minneapolis.

Bochner S (2001): A study of functional literacy skills in young adults with Down syndrome. Int J Dis Dev Educ **48**: 67-90.

Bower A & Hayes A (1994): Short-term memory deficits and Down syndrome: A comparative study. Downs Syndr Res Pract **2**: 47-50.

Courage ML, Adams RJ, Reyno S & Kwa PG (1994): Visual acuity in infants and children with Down syndrome. Dev Med Child Neurol **36**: 586-593.

Cregg M, Woodhouse JM, Pakeman VH, Saunders KJ, Gunter HL, Parker M, Fraser WI & Sastry P (2001): Accommodation and refractive error in children with Down syndrome: Cross-sectional and longitudinal studies. Invest Ophthalmol Vis Sci **42**: 55-63.

Cregg M, Woodhouse JM, Stewart RE, Pakeman VH, Bromham NR, Gunter HL, Trojanowska L, Parker M & Fraser WI (2003): Development of refractive error and strabismus in children with Down syndrome. Invest Ophthalmol Vis Sci **44**: 1023-1030.

Cupples L & Iacono T (2000): Phonological awareness and oral reading skill in children with Down syndrome. J Speech Lang Hear Res **43**: 595-608.

Dolch sighwordlist: http: //www.teachnology.com/worksheets/language_arts/dolch/list/. (Accessed February 2008)

Dunn LM & Dunn LM (1997): Peabody Picture Vocabulary Test-(PPVT-III). Circle Pines, MN: American Guidance Service.

Fidler DJ, Most DE & Guiberson MM (2005): Neuropsychological correlates of word identification in Down syndrome. Res Dev Disabil **26**: 487-501.

French AN, Rose KA, Burlutsky G & Mitchell P (2009): Sydney Childhood Eye Study: Does correction of hyperopia affect the pattern of children's activities, and does this differ from that in emmetropic children? Invest Ophthalmol Vis Sci **50**.E-Abstract 3961.

Gardner MF (1996): TVPS-R, Test of Visual-Perceptual Skills (Non-Motor)-Revised: Psychological & Educational Publications.

Haugen OH, Hovding G & Lundstrom I (2001): Refractive development in children with Down's syndrome: A population based longitudinal study. Br J Ophthalmol **85**: 714-719.

Haugen OH & Hovding G (2001): Strabismus and binocular function in children with Down syndrome: A population-based, longitudinal study. Acta Ophthalmol Scand **79**: 133-139.

Jaccard J & Wan CK (1996): Quantitative applications in social sciences. In: LISREL approaches to interaction effects in multiple regression. California: 114.

Kay-Raining Bird E, Cleave PL, White D, Pike H & Helmkay A (2008): Written and oral narratives of children and adolescents with Down syndrome. J Speech Lang Hear Res **51**: 436-450.

Kim JH, Hwang JM, Kim HJ & Yu YS (2002): Characteristic ocular findings in Asian children with Down syndrome. Eye **16**: 710-714.

Laws G (2000) Language and memory development in children with Down syndrome at mainstream schools and special schools: A comparison. Educ Psychol **20**: 447-457.

Leat SJ & Mohr A (2007): Accommodative Response in pre-presbyopes with visual impairment and its clinical implications. Invest Ophthalmol Vis Sci **48**: 3888-3896.

Lin LI (1989): A concordance correlation coefficient to evaluate reproducibility. Biometrics **45**: 255-268.

Lovie-Kitchin JE, Bevan JD & Hein B (2001): Reading performance in children with low vision. Clinin Exp Optom **84**: 148-154.

McClelland JF & Saunders KJ (2003): The repeatability and validity of dynamic retinoscopy in assessing the accommodative response. Ophthalmic Physiol Opt **23**: 243-250.

Miolo G, Chapman RS & Sindberg HA (2005): Sentence comprehension in adolescents with Down syndrome and typically developing children: Role of sentence voice, visual context, and auditory-verbal short-term memory. J Speech Lang Hear Res **48**: 172-188.

Mohd-Ali B, Mohammed Z, Norlaila M, Mohd-Fadzil N, Rohani CC & Mohidin N (2006): Visual and binocular status of Down syndrome children in Malaysia. Clin Exp Optom **89**: 150-154.

Nandakumar K & Leat SJ (2009): Bifocals in Down Syndrome Study (BiDS): Design and baseline visual function. Optom Vis Sci **86**: 196-207.

Partik BL, Stadler A, Schamp S, Koller A, Voracek M, Heinz G & Helbich TH (2002): 3D versus 2D ultrasound: Accuracy of volume measurement in human cadaver kidneys. Invest Radiol **37**: 489.

Shankar S, Evans MA & Bobier WR (2007): Hyperopia and emergent literacy of young children: pilot study. Optom Vis Sci **84**: 1031-1038.

Shepperdson B (1994): Attainments in reading and number of teenagers and adults with Down syndrome. Downs Syndr Res Pract **2**: 97-101.

Sloper P, Cunningham C, Turner S & Knussen C (1990): Factors related to the academic attainments of children with Down's syndrome. Br J Educ Psychol **60**: 284-298.

Turner S, Alborz A (2003): Academic attainment of children with Down's syndrome. Br J Educ Psychol. **73**: 563-583.

Stewart RE, Margaret Woodhouse J & Trojanowska LD (2005): In focus: The use of bifocal spectacles with children with Down's syndrome. Ophthalmic Physiol Opt **25**: 514-522.

van Splunder J, Stilma JS, Bernsen RM, Arentz TG & Evenhuis HM (2003): Refractive errors and visual impairment in 900 adults with intellectual disabilities in the Netherlands. Acta Ophthalmol Scand **81**: 123-129.

Whittaker SG, Lovie-Kitchin J & Optom MS (1993): Visual requirements for reading. Optometry Vision Sci **70**: 54.

Woodcock R. Woodcock Reading Mastery Tests-Revised (WRMT-R/NU) (1998): Circle Pines, Minnesota: American Guidance Service.

Woodhouse JM, Meades JS, Leat SJ & Saunders KJ (1993): Reduced accommodation in children with Down syndrome. Invest Ophthalmol Vis Sci **34**: 2382-2387.

Woodhouse JM, Pakeman VH, Cregg M, Saunders KJ, Parker M, Fraser WI, Sastry P & Lobo S (1997): Refractive errors in young children with Down syndrome. Optom Vis Sci **74**: 844-851.

References for Chapter 6

http://www.teachnology.com/worksheets/language_arts/dolch/list/. (Accessed 13 February 08)

Beery K. (1997): The Beery-Buktenica Test of Visual-Motor Integration with Supplemental Developmental Tests of Visual Perception and Motor Coordination.

Bergman K & McLaughlin T (1988): Remediating Handwriting Difficulties with Learning Disabled Students: A Review. BC J Spec Educ **12**: 101-120.

Bird G & Buckley S (1994): Meeting the Educational Needs of Children with Downs Syndrome. In: Portsmouth: University of Portsmouth.

Bochner S (2001): A Study of Functional Literacy Skills in Young Adults with Down Syndrome. Int J Dis Dev Educ **48**: 67-90.

Boudreau DM & Chapman RS. (2000): The relationship between event representation and linguistic skill in narratives of children and adolescents with Down syndrome. J Speech Lang Hear Res **43**: 1146.

Buckley S. (1995) In: Nadel L, Rosenthal D (Eds). Down syndrome: Living and learning in the community. New York: Wiley & sons: 158–69.

Buckley S, Bird G & Byrne A (1996): Reading acquisition by young children.I n: Stratford B, Gunn P (Eds). New approaches to Down syndrome. London: Cassell: 268-79.

Buckley S (2001): Reading and Writing for Individuals with Down Syndrome: An Overview : Down Syndrome Issues and Information. doi:10.3104/9781903806098. The Down syndrome Educational Trust; 2001. (Accessed January 23, 2010).

Byrne A, MacDonald J & Buckley S (2002): Reading, language and memory skills: a comparative longitudinal study of children with Down syndrome and their mainstream peers. Br J Educ Psychol **72**: 513-529.

Caputo AR, Wagner RS, Reynolds DR, Guo S & Goel AK (1989): Down syndrome: Clinical Review of Ocular Features. Clin Pediatr **28**: 355.

Catalano RA (1992): In Pueschel, SM & Pueschel, JK (Eds).Ophthalmologic concerns. Biomedical concerns in persons with Down syndrome.

Chan J & Iacono T (2001): Gesture and word production in children with Down syndrome Augment Altern Commun **17**: 73-87.

Chapman R. (2006): Language learning in Down syndrome: The speech and language profile compared to adolescents with cognitive impairment of unknown origin. Downs Syndr Res Pract **10**: 61-66.

Chapman RS (1997): Language development in children and adolescents with Down syndrome. Ment Retard Dev Disabil Res Rev **3**:307-12.

Chapman RS & Hesketh LJ (2001): Language, cognition, and short-term memory in individuals with Down syndrome. Downs Syndr Res Pract **7**: 1-7.

Chapman RS, Seung HK, Schwartz SE & Kay-Raining Bird E (1998): Language Skills of Children and Adolescents With Down Syndrome II. Production Deficits. J Speech Lang Hear Res **41**: 861-873.

Cowie VA. (1970) A study of the earlydevelopment of mongols. London, England: Pergamon.

Cregg M, Woodhouse JM, Pakeman VH, Saunders KJ, Gunter HL, Parker M, Fraser WI & Sastry P (2001): Accommodation and refractive error in children with Down syndrome: cross-sectional and longitudinal studies. Invest Ophthalmol Vis Sci **42**: 55-63.

Cregg M, Woodhouse JM, Stewart RE, Pakeman VH, Bromham NR, Gunter HL, Trojanowska L, Parker M & Fraser WI (2003): Development of refractive error and strabismus in children with Down syndrome. Invest Ophthalmol Vis Sci **44**: 1023-1030.

Cuskelly M, Einam M & Jobling A (2001): Delay of gratification in young adults with Down syndrome. Downs Syndr Res Pract **7**: 60-67.

da Cunha RP & Moreira J (1996) :Ocular findings in Down's syndrome. Am J Ophthalmol **2**: 236-244.

Davis WE & Kelso JA (1982): Analysis of "invariant characteristics" in the motor control of Down’s syndrome and normal subjects. J Mot Behav **14**: 194-212.

Dunn LM & Dunn LM (1997): Peabody Picture Vocabulary Test-(PPVT-III). Circle Pines, MN: American Guidance Service.

Fowler A (1995): Linguistic variability in persons with Down syndrome: Research and implications. In: Nadel L, Rosenthal D (Eds). Down syndrome: Living and learning in the community. New York: Wiley & sons: 121–31.

Fowler A, Doherty B & Boynton L (1995): The basis of reading skill in young adults with Down syndrome. In: Nadel L, Rosenthal D (Eds). Down syndrome: Living and learning in the community. New York: Wiley & sons: 182–96.

Gallaher K, van Kraayenoord C, Jobling A & Moni K (2002): Reading with Abby: A case study of individual tutoring with a young adult with Down syndrome. Downs Syndr Res Pract **8**: 59-66.

Gardiner PA (1967): Visual defects in cases of Down's syndrome and in other mentally handicapped children. Br J Ophthalmol **51**: 469-474.

Gardner MF (1996): TVPS-R, Test of Visual-Perceptual Skills (Non-Motor)-Revised : Psychological & Educational Publications.

Haugen OH & Hovding G (2001): Strabismus and binocular function in children with Down syndrome. A population-based, longitudinal study. Acta Ophthalmol Scand **79**: 133-139.

Haugen OH, Hovding G & Riise R (2004): Ocular changes in Down syndrome. Tidsskr Nor Laegeforen **124**: 186-188.

Iverson JM, Longobardi E & Caselli MC (2003): Relationship between gestures and words in children with Down's syndrome and typically developing children in the early stages of communicative development Int J Lang Commun Disord **38**: 179-197.

Kay-Raining Bird E, Cleave P & McConnell L (2000): Reading and phonological awareness in children with Down syndrome: A longitudinal study. Am J Speech Lang Pathol **9**: 319-330.

Kay-Raining Bird E, Cleave PL, White D, Pike H & Helmkay A (2008): Written and oral narratives of children and adolescents with Down syndrome. J Speech Lang Hear Res **51**: 436-450.

Kim O, Kendeou P, van den Broek P, White MJ & Kremer K (2008): Cat, Rat, and Rugrats: Narrative Comprehension in Young Children with Down Syndrome. J Dev Phys Disabil **20**: 337-351.

Klein BP (1999): Contrasting Patterns of Cognitive Abilities of 9- and 10-Year-Olds With Williams Syndrome or Down Syndrome. Dev Neuropsychol **16**: 177-196.

Lindstedt E (1983): Failing accommodation in cases of Down's syndrome: A preliminary report. Ophthalmic Genet **3**: 191-192.

Morgan M, Moni K & Jobling A (2004): What's it all about? Investigating reading comprehension strategies in young adults with Down syndrome. Downs Syndr Res Pract **9**: 37-44.

Mundy P, Sigman M, Kasari C & Yirmiya N (1988): Nonverbal Communication Skills in Down Syndrome Children. Child Dev **59**: 235-249.

Nandakumar K & Leat SJ (2009): Bifocals in Down Syndrome Study (BiDS): Design and baseline visual function. Optom Vis Sci **86**: 196-207.

Oelwein PL (1995): Teaching Reading to Children with Down Syndrome: A guide for parents and teachers. Maryland: Woodbine House.

O□Toole C & Chiat S (2006): Symbolic functioning and language development in children with Down syndrome. Int J Lang Commun Disord **41**: 155-171.

Shapiro MB & France TD (1985): The ocular features of Down's syndrome. Am J Ophthalmol **99**: 659-663.

Shepperdson B (1994): Attainments in reading and number of teenagers and adults with Down syndrome. Downs Syndr Res Pract **2**: 97-101.

Tsiaras WG, Pueschel S, Keller C, Curran R & Giesswein S (1999): Amblyopia and visual acuity in children with Down's syndrome. Br Med J **83**: 1112-1114.

van Bysterveldt AK, Gillon GT & Moran C (2006): Enhancing phonological awareness and letter knowledge in preschool children with Down syndrome. Int J Dis Dev Educ **53**: 301-329.

Van Kraayenoord CE, Moni KB, Jobling A & Ziebarth K (2000): Broadening approaches to literacy education for young adults with Down syndrome. In Monica Cuskelly, Anne Jobling, and Sue Buckley (Eds). Down syndrome across the life span.

van Splunder J, Stilma JS, Bernsen RM, Arentz TG & Evenhuis HM (2003): Refractive errors and visual impairment in 900 adults with intellectual disabilities in the Netherlands. Acta Ophthalmol Scand **81**: 123-129.

Woodcock R. Woodcock Reading Mastery Tests-Revised (WRMT-R/NU). 1998: Circle Pines, Minnesota: American Guidance Service.

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Woodhouse JM, Pakeman VH, Saunders KJ, Parker M, Fraser WI, Lobo S & Sastry P (1996): Visual acuity and accommodation in infants and young children with Down's syndrome. J Intellect Disabil Res **40**: 49-55.

References in Chapter 7

Al-Bagdady M, Stewart RE, Watts P, Murphy PJ & Woodhouse JM (2009): Bifocals and Down’s syndrome: correction or treatment? Ophthalmic Physiol.Op **29**: 416-421.

Barnard S & Edgar D. (1996) Pediatric Eye Care: Blackwell Science.

Berg JM, Brandon MW & Kirman BH. (1959): Atropine in mongolism. Lancet **2**: 441-442.

Courage ML, Adams RJ, Reyno S & Kwa PG. (1994) Visual acuity in infants and children with Down syndrome. Dev Med Child Neurol 36(7): 586-593.

Cregg M, Woodhouse JM, Pakeman VH, Saunders KJ, Gunter HL, Parker M, Fraser WI & Sastry P. (2001): Accommodation and refractive error in children with Down syndrome: cross-sectional and longitudinal studies. Invest Ophthalmol Vis Sci **42**: 55-63.

Gordon DM & Ehrenberg MH. (1954): Cyclopentolate hydrochloride: A new mydriatic and cycloplegic agent; a pharmacologic and clinical evaluation. Am J Ophthalmol **38**: 831-838.

Haugen OH, Hovding G & Lundstrom I. (2001): Refractive development in children with Down's syndrome: a population based, longitudinal study. Br J Ophthalmol **85**: 714-719.

Priest JH. (1960) Atropine response of the eyes in mongolism. Am J Dis Child 100: 869-872.

Sacks B & Smith S. (1989): People with Down's syndrome can be distinguished on the basis of cholinergic dysfunction. J Neurol Neurosurg Psychiatry **52**: 1294-1295.

Schor C. (1999): The influence of interactions between accommodation and convergence on the lag of accommodation. Ophthalmic Physiol Opt **19**: 134-150.

Stewart RE, Margaret Woodhouse J & Trojanowska LD. (2005): In focus: the use of bifocal spectacles with children with Down's syndrome. Ophthalmic Physiol Opt **25**: 514-522.

Tsiaras WG, Pueschel S, Keller C, Curran R & Giesswein S. (1999): Amblyopia and visual acuity in children with Down's syndrome. Br Med J **83**: 1112.

Woodhouse JM, Cregg M, Gunter HL, Sanders DP, Saunders KJ, Pakeman VH, Parker M, Fraser WI & Sastry P. (2000): The effect of age, size of target, and cognitive factors on accommodative responses of children with Down syndrome. Invest Ophthalmol Vis Sci **41**: 2479-2485.

Woodhouse JM, Pakeman VH, Cregg M, Saunders KJ, Parker M, Fraser WI, Sastry P & Lobo S. (1997): Refractive errors in young children with Down syndrome. Optom Vis Sci **74**: 844-851.

Appendix A

Example of a Dolch sight word test page

about	bring
clean	done
drink	fall
full	grow

Appendix B

<u>Example of a number test page</u>

32	34	39
35	31	38
33	37	36

Appendix C

Classroom questionnaire which was sent out to teachers at the end of each term

Bifocal Spectacle Lens Correction in Down syndrome (BiDS)

Classroom Questionnaire

Principal Investigator: Dr. Susan Leat

Co-investigators: Dr Mary Ann Evans, Department of Psychology, University of Guelph, and Dr Patricia Cleave, Dalhousie University.

Student investigator: Krithika Nandakumar

Questionnaire for teachers (to be completed regarding the prior 6 months period. If the student was not in your class for all of this period, please estimate as closely as possible)

Student's name (participant) ______________________

Please indicate the number of hours per week in the following settings, and the content/goals of the instruction in each setting (please circle all that apply)

Hours in regular class _________Content in regular class: word level reading (i.e. isolated words), spelling, phonological awareness, reading, writing, arithmetic, fine motor co-ordination, gross motor co-ordination, other

____

Hours with indirect support ________Content with indirect support: word level reading, spelling, phonological awareness, reading, writing, arithmetic; fine motor co-ordination, gross motor co-ordination, other ____

Hours with teaching assistant _________Content with TA: word level reading, spelling, phonological awareness, reading, writing, arithmetic, fine motor co-ordination, gross motor co-ordination, other

____

Hours with withdrawal assistance _________ Content in withdrawal: word level reading, spelling, phonological awareness, reading, writing, arithmetic, fine motor co-ordination, gross motor co-ordination other

____

Hours in a segregated class __________Content in segregated class: word level reading, spelling, phonological awareness, fine motor co-ordination, gross motor co-ordination other -

____

____

Please estimate the number of hours per week that the child receives

Hours of early literacy skills (e.g letter recognition, alphabet) _________________

Hours of reading instruction _________________

Hours of writing instruction __________________

Hours of arithmetic instruction ________________

For the following activities, please circle how much the child wears his/her glasses

Reading and reading related activities ………. Always/mostly/sometimes/rarely/never

Writing …………………………………………… Always/mostly/sometimes/rarely/never

Arithmetic ……………………………………… Always/mostly/somtimes/rarely/never

Computer work ………………………………… Always/mostly/sometimes/rarely/never

Playground ……………………………………… Always/mostly/sometimes/rarely/never

Gym ………………………………………………… Always/mostly/sometimes/rarely/never

Looking at the board/overheards...…………… Always/mostly/sometimes/rarely/never

Are there any general comments that you would like to add.

____

____

____

____

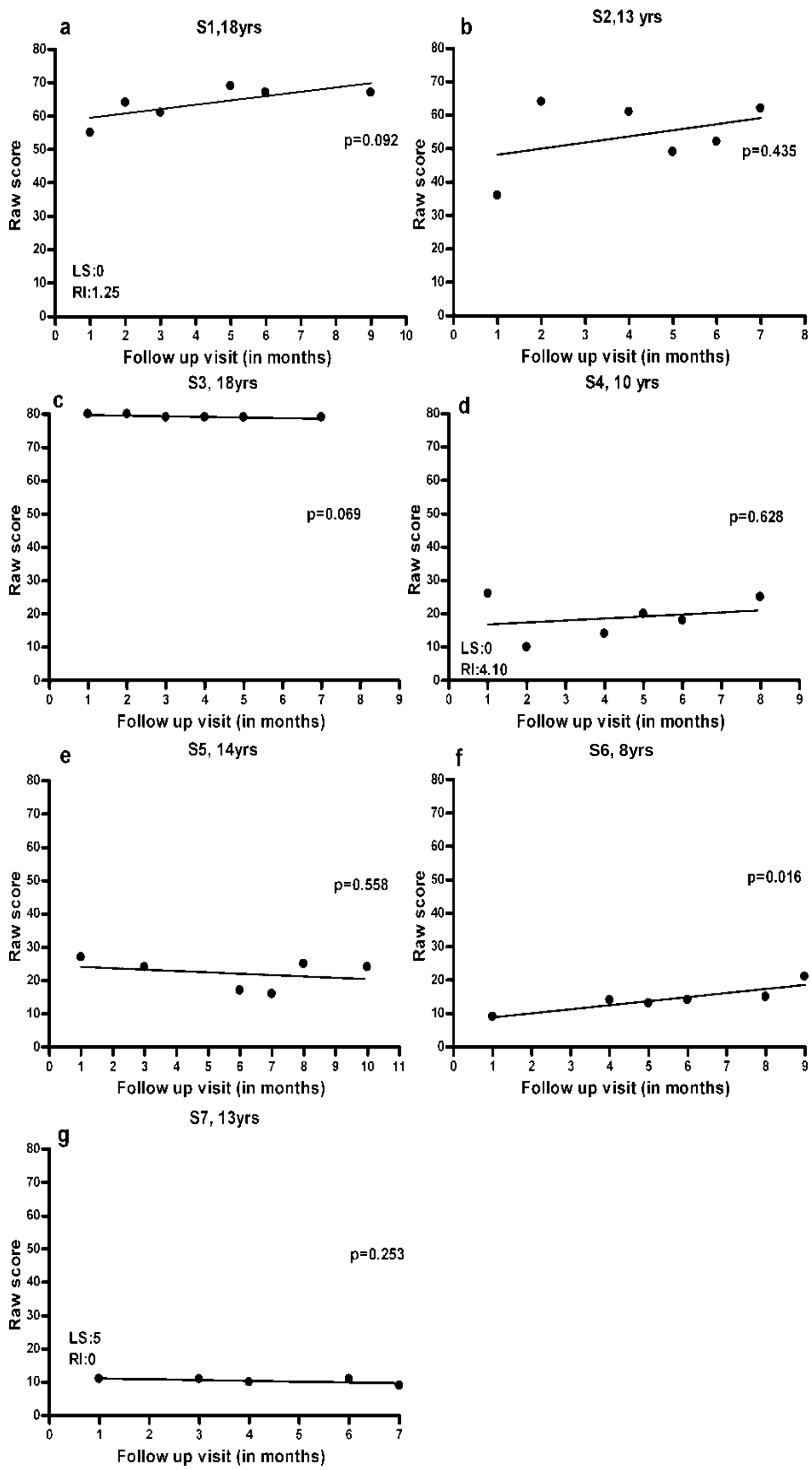
Form filled in by _____________________

Date _______________________________

Please return to Krithika Nandakumar, Graduate Student, School of Optometry, University of Waterloo, Waterloo, N2L 3G1.

Appendix D

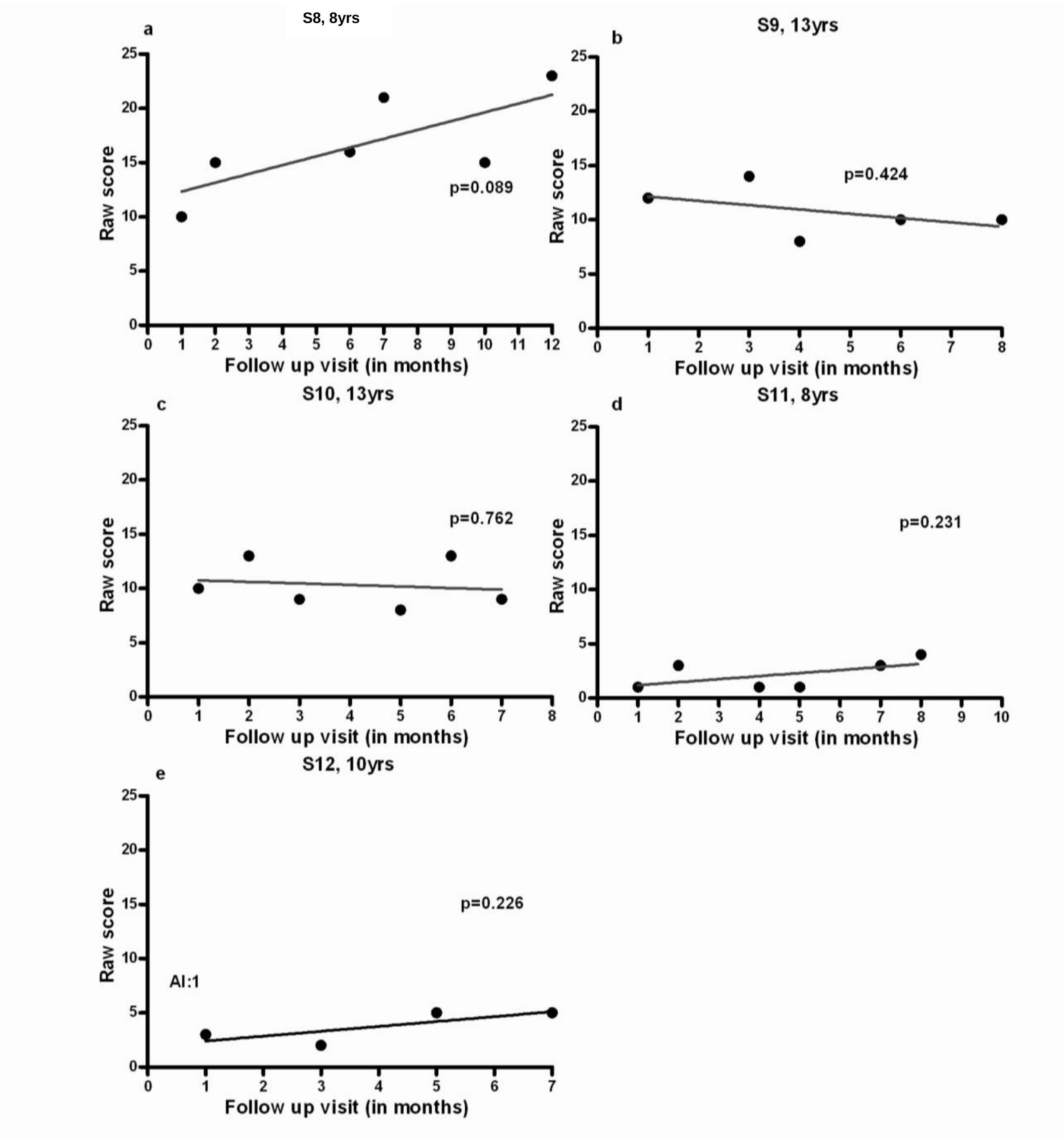
Natural progression of raw scores for the Dolch sight word list with single vision lenses



RI and LS show the numbers of hours of reading instruction and literacy skills instruction received respectively per week for the participants where information was available. Each p value shows if the slope of the regression lines over 6 months is significantly different from zero.

Appendix E

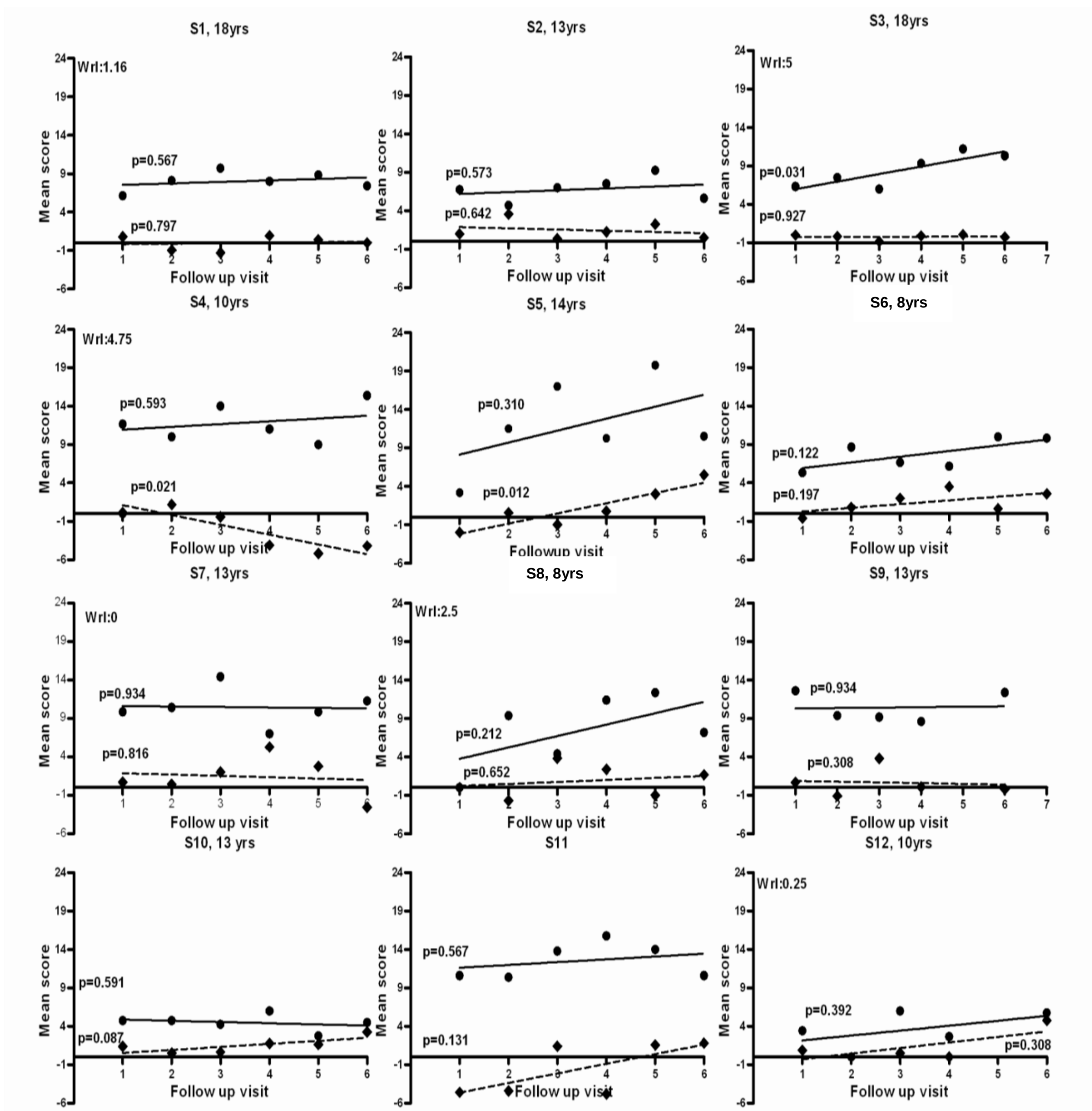
Natural progression of scores for the number list with single vision lenses



AI shows the numbers of hours of arithmetic instruction received per week for the participants where information was available. Each p value shows if the slope of the regression line over 6 months is significantly different from zero.

Appendix F

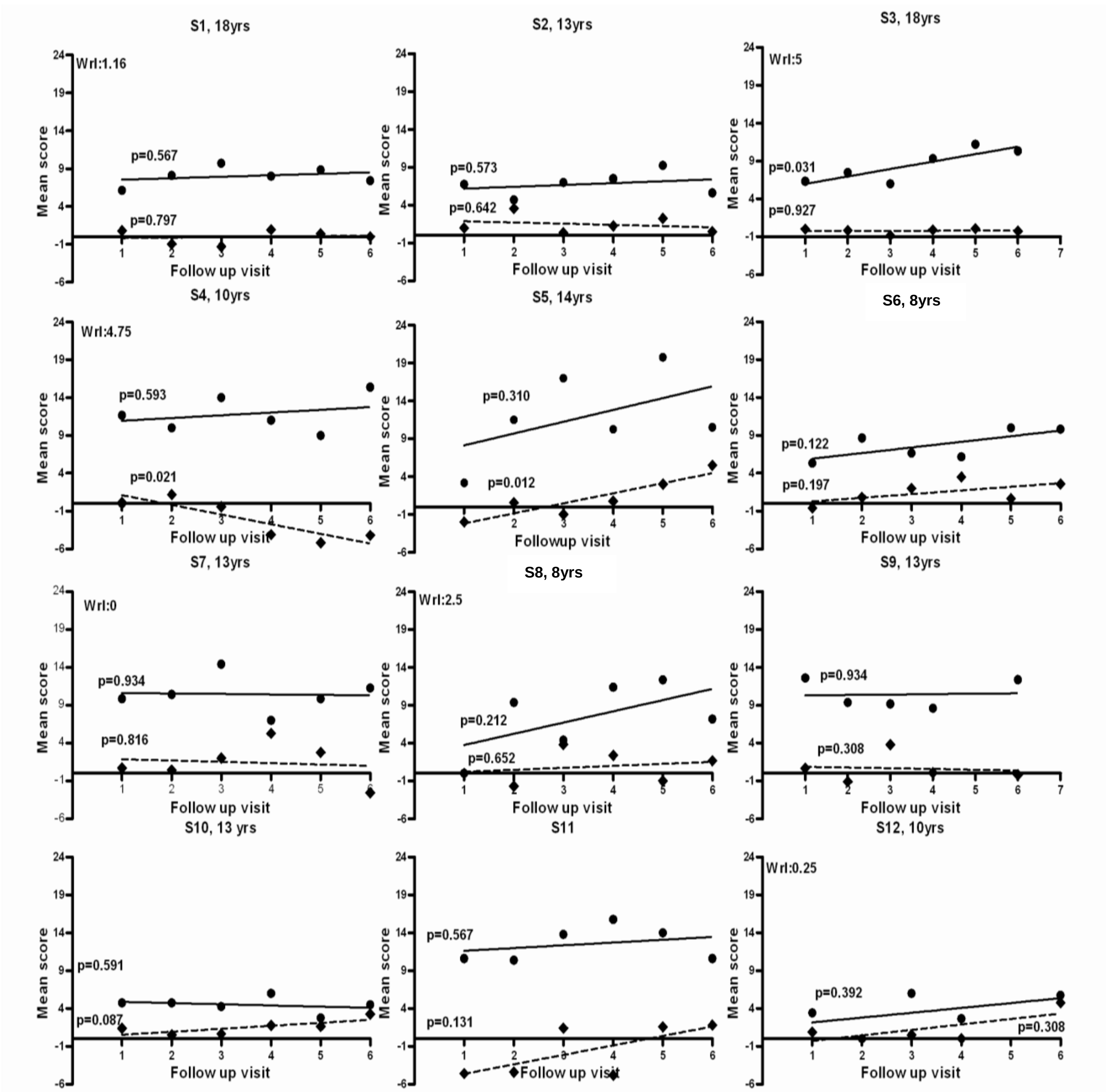
<u>Mean letter size and position of letters over 6 months with single vision lenses</u>



Mean letter size (shown by circles and solid lines) and position of letters (shown by diamonds and dashed lines) over 6 months with single vision lenses. WrI shows the hours of written instruction received per week. p values show if the slopes of the regression lines for each measure of printing are significantly different from zero

Appendix G

<u>Standard deviations of letter size and position over 6 months with single vision lenses</u>



Mean letter size (shown by circles and solid lines) and position of letters (shown by diamonds and dotted lines) over 6 months with single vision lenses. WrI shows the hours of written instruction received per week. p values show if the slopes of the regression lines for each measure of printing are significantly different from zero

Appendix H

Limits of agreement (LOA) between observers KN & SL for dynamic retinoscopy

Stimulus (D)	Bias (mean difference d)	SD of bias	Lower limit (d - 1.96 SD)	Upper limit (d + 1.96SD)
At SV baseline				
4	0.041	0.336	-0.619	0.701
6	-0.071	0.711	-1.464	1.322
8	-0.304	0.970	-2.206	1.597
10	0.016	1.044	-2.030	2.062
When determining bifocal addition				
4	0.142	0.259	-0.365	0.650
6	0.060	0.690	-1.108	1.223
8	-0.485	0.813	-2.080	1.108
10	-0.471	1.254	-2.929	1.986
Through bifocal add				
4	-0.024	0.388	-0.785	0.736
6	0.276	0.523	-0.748	1.301
8	0.225	0.783	-1.308	1.759
10	0.373	1.278	-2.133	2.877

Limits of agreement between measures of accommodative responses using dynamic retinoscopy at 4, 6, 8 and 10 D distances between observers KN and SL. Paired t-tests between both sets of observations were not significantly different ($p>0.05$) for all test distances.

Appendix I

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Susan J. Leat; Jennifer L. Gargon

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