An Evaluation of Vision Screening Protocols in Young Children

by Mythili Ilango

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Certificate of Original Authorship

I, Mythili Ilango declare that this thesis, is submitted in fulfilment of the requirements for the award of Doctor of Philosophy, in the Graduate School of Health at the University of Technology Sydney. This thesis is wholly my own work unless otherwise referenced or acknowledged. In addition, I certify that all information sources and literature used are indicated in the thesis. This document has not been submitted for qualifications at any other academic institution.

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Thesis Abstract

Background and Aims

There is a growing body of evidence supporting vision screening for preschool children and a recent evaluation of the New South Wales (NSW) Statewide Eyesight Preschooler Screening (StEPS) program found the program to be highly appropriate and cost-effective. However, there are no universally accepted protocols for vision screening, either nationally or internationally. In this context, this thesis aimed to address several questions related to ideal approaches to childhood vision screening. This included, to determine if visual acuity screening is more accurate at school age compared to preschool age, the comparability of referral rates and appropriateness of referral thresholds using the Sheridan Gardiner and HOTV logMAR charts, and whether including additional tests in screening protocols would improve detection of conditions. In addition, this thesis aimed to define the ocular conditions that may reduce vision at different ages and whether repeat screening may be required later in childhood. Finally, this thesis examined the impact of cycloplegia and refraction method for measurement of refractive errors in children and the natural history of hyperopic refractive errors to examine the need for detection and prescription of refractive correction.

Methodology

To answer the aims of this thesis, we have drawn on a number of relevant data sources. Existing datasets from the series of population-based studies of eye health in metropolitan Sydney children, the Sydney Childhood Eye Disease Studies that included, the Sydney Paediatric Eye Disease Study (SPEDS), the Sydney Myopia Study (SMS) and the 5-6 year follow-up Sydney Adolescent and Vascular Eye Study (SAVES) were utilised for analysis. These studies collectively examined a total of 7266 children between 6 months and 17 years of age. All children had a comprehensive ocular examination including, age-appropriate visual acuity testing, orthoptic assessment and cycloplegic autorefraction.

As part of the main project of this thesis, the Preschool Vision Screening Study (PVSS), 94 four year old children were recruited through StEPS. Vision screening was performed in preschool and childcare settings according to StEPS protocols using both the Sheridan

Gardiner visual acuity chart and the HOTV logMAR chart, followed by an additional orthoptic examination. The StEPS program referral criteria was used to refer children who did not pass screening. One month post-screening parents or guardians of children who were referred from screening were followed-up to determine the outcome of referral and barriers related to successful follow-up.

Results

This thesis has provided additional evidence that four years of age is ideal for vision screening, when amblyopia and early refractive errors are common causes of reduced vision and as intervention is time-sensitive to optimise treatment outcomes, and to address reduced vision prior to school entry. In addition, accuracy of vision screening was not compromised at preschool age in comparison to early school age screening, with visual acuity having a high sensitivity and specificity for amblyopia and myopic refractive errors. At 12 years of age, there was a substantial increase in the prevalence of myopia and this remained a significant cause of reduced vision in older children. Targeted school screening for those at risk of developing myopia or education for children and parents to increase detection and reporting of symptoms would be an appropriate and cost-effective approach to increasing myopia detection at this age.

Visual acuity testing had considerably lower sensitivity for the detection of hyperopic refractive errors in preschool children, suggesting that current vision screening protocols may not successfully detect this refractive error. There was a hyperopic mean refraction in the 6-12 month age group in Sydney that subsequently decreased through childhood. Interestingly, this analysis revealed a more myopic mean refraction in children with darker irides, likely related to lower efficacy of cycloplegia. This may result in under-detection of hyperopia in this population. There has been debate about the necessity of refractive correction for hyperopia in childhood and whether spectacle correction may interrupt normal emmetropisation to reduce hyperopia. However, data in this thesis has shown that children who are hyperopic, particularly those with high hyperopia, often remained significantly hyperopic into adolescence and that refractive correction did not impair

reduction of hyperopia through emmetropisation. Thus, there is likely benefit to both the detection and prescription of refractive correction for children with significant hyperopia.

The StEPS program has recently transitioned from the Sheridan Gardiner visual acuity chart to the gold-standard HOTV logMAR chart. Comparison of these two tests to determine the impact on referral rates and whether current referral criteria were likely appropriate was conducted. Visual acuity was considerably higher using the HOTV logMAR than found with the Sheridan Gardiner chart, indicating that referral rates in StEPS are likely to reduce after the transition to the logMAR chart. The current referral criteria of visual acuity worse than 6/9 is even more suitable now since the mean visual acuity of preschool children was 6/7.5 using the HOTV logMAR. However, it is recommended that an additional referral criterion of ≥ 2 line visual acuity difference between eyes be considered. The inclusion of additional screening tests did not significantly increase detection of childhood ocular conditions.

Conclusion

Overall, the series of chapters presented in this thesis have provided further evidence of the most effective protocols for vision screening in childhood including, that four years is the optimal target age for vision screening and that a referral threshold of 6/9 is appropriate when using HOTV logMAR for preschool children. The findings in this thesis further indicate that repeat screening in the early school years would not be valuable but, targeted screening or education in adolescence may support the detection and management of myopia. Finally, this thesis has shown the challenges of detecting hyperopia using visual acuity screening and as correction does not impact normal emmetropisation, detection of hyperopia and its correction is likely to have benefit for children.

Publications and Presentations

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Ilango M, French AN, Rose KA. Refractive Error in Preschool Children: Sydney Paediatric Eye Disease Study. 2017; Asia ARVO, The Association for Research in Vision and Ophthalmology Conference, Brisbane, Australia.

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Glossary of Abbreviations

Abbreviation	Full term
ABS	Australian Bureau of Statistics
AL	Axial Length
ANOVA	Analysis of Variance
ATS	Amblyopia Treatment Study
BCVA	Best Corrected Visual Acuity
BPEDS	Baltimore Paediatric Eye Disease Study
BSV	Binocular Single Vision
CCES	Community Children's Eye Service
CEHW	Community Eye-Health Workers
CI	Confidence Interval
CLEERE	Collaborative Longitudinal Evaluation of Ethnicity and Refractive Error Study
CR	Corneal Radius
D	Dioptres
DC	Dioptres Cylinder
DS	Dioptres Sphere
ETDRS	Early Treatment of Diabetic Retinopathy Study
EVA	Electronic Visual Acuity

GOS	General Optometry Service
IOD	Intraocular difference
IOL	Intraocular Lens
LE	Left Eye
Log MAR	Logarithm of the minimum angle of resolution
MEPEDS	Multi-ethnic Paediatric Eye Disease Study
NHMRC	National Health and Medical Research Council
NSW	New South Wales
OCT	Optical Coherence Tomography
OMs	Ocular Movements
OR	Odds Ratio
PEDIG	Paediatric Eye Disease Study Group
PVSS	Preschool Vision Screening Study
RAF	Royal Air Force
RAPD	Relative Afferent Pupillary Defect
RE	Right Eye
RESC	Refractive Error Study in Children Study
SAVES	Sydney Adolescent Vascular Eye Study
SCES	Sydney Childhood Eye Studies
SER	Spherical Equivalent Refraction

SES	Socioeconomic Status
SG	Sheridan Gardiner
SMS	Sydney Myopia Study
SPEDS	Sydney Paediatric Eye Disease Study
SPSS	Statistical Package for the Social Sciences
STARS	Strabismus, Amblyopia and Refractive Error Study
STEPS	Statewide Eyesight Preschooler Screening
UK	United Kingdom
USPSTF	United States Preventative Services Task Force
UTS	University of Technology Sydney
VA	Visual Acuity
VIP	Vision in Preschoolers study group
WHO	World Health Organisation

Chapter 1
Literature Review

1.1 Overview

This chapter examines current literature regarding the development of vision in children, with a focus on the importance of vision screening, including its primary purpose of screening for amblyopia and its risk factors. The evidence relating to the most suitable age for vision screening for the early detection and timely treatment of childhood eye disorders is also explored. The chapter further summarises the current evidence regarding international vision screening protocols, with a particular focus on the New South Wales Statewide Eyesight Preschooler Screening Program (StEPS). Current childhood vision tests, referral pathways and barriers to follow-up on referral after screening have also been surveyed. Finally, this chapter describes the justification for and aims of this thesis.

1.2 What is vision?

Vision is an important human sense that enables us to interact with the world around us. Good vision depends on four important anatomical factors:

1. Clear optical media (cornea, anterior chamber, crystalline lens and vitreous) for light to pass through to reach the retina (the neurosensory element) at the back of the eye
2. The sharpness of the image formed on the retina
3. Intact visual pathways to allow the image to be communicated centrally
4. Fully developed visual cortex and visual association areas that process and interpret the images present.

1.2.1 How does the eye work?

In an adult with a normal eye, light rays enter the eye through the cornea, which is the front surface of the eye. The cornea (as the primary lens of the eye) bends (converges) the light rays as they pass to the pupil and through the eye's crystalline lens which further converges the light rays. The light rays then passes through the vitreous and ideally come to sharp focus on the retina. This sharp focus on the retina, particularly in the centre of the macula (fovea) allows for good visual acuity (the ability of the eye to resolve fine detail) to be achieved. These light signals are then converted to electrical signals (action potentials) within the retina and conveyed along the neuronal fibres of the visual pathway. The pathway includes the optic nerve, chiasm and tract, and then the lateral geniculate nucleus and optic radiations and terminates in the visual cortex.

There are numerous ocular conditions that could cause reduced visual acuity. These include refractive errors that create a de-focused image and cataract that causes scattering of light, degrading the image formed on the retina. Retinal pathologies (e.g. retinal dystrophies, macular degeneration) and tumours of the optic pathway or demyelination of optic axons may prevent the formation of action potentials or their capacity to reach the visual cortex. These are just a few of the many pathologies that can hinder the generation of good vision.

1.3 Visual Development in Children

Newborn infants are in a state of early visual development. This development requires that all the structures of the eye are able to transmit light to the retina, that the visual pathway is able to convey light information to the visual cortex and that the cortex is intact.

1.3.1 Development of the Eye

A newborn baby's eye is not adult size, being about 70% of its adult size and about 50% of adult volume. The axial length of the neonatal eye is approximately 74% of an adult human eye.[1] When coupled with a relatively more mature anterior eye (cornea and crystalline lens) it means that a newborn is likely to have a hyperopic (long-sighted) refractive error, that is, the optical power of the eye does not match the shortened axial length of the eye. This was borne out by the work of Cook and Glassock who found that close to 75% of babies are hyperopic.[2] The remaining babies tended to have a myopic (short-sighted) refractive error, which appeared to arise from excessive lenticular power. This is known to occur in premature children and is known as myopia of prematurity.[3] While neonatal refractive errors do cause de-focused images, the pupils of a baby's eyes are relatively constricted, which has the effect of increasing depth of focus, overcoming some optical blur associated with the mismatch between the optical power of the eye it's axial length.[4] The greater determinant of visual acuity in neonates lies in their underdeveloped retinal structure and neural pathways.

The macula and more specifically the fovea, are central specialised neural areas of the retina. The fovea in particular is packed with a dense mosaic of cone photoreceptors that convert light into chemical signals, enabling high levels of visual acuity in daylight conditions.[5] The cone photoreceptors are responsible for detecting fine visual detail and are in high numbers specifically at the fovea, which allows greater sampling of visual objects than more widely scattered receptors would.

At the time of birth, the neonatal fovea is large in diameter (1100μm) but not fully developed due to incomplete migration of the ganglion cell and inner nuclear layers away from the fovea.[6-8] This means that at birth there is an incomplete foveal pit that affects the rate at which light rays reach the photoreceptors located in the outer retina. The inner and

outer segments of the cone photoreceptors are short and thick (diameter of 7.5μm compared to an adult diameter of 4μm) with a shorter outer segment length than found in adults and are considered immature. The shorter thicker cones create a limit to the density of the cone mosaic, therefore, at birth cone density is quite low at 36,000 cones/mm^2 compared to adult size.

At 15 months of age, the foveal pit has formed in the inner retinal layers, and the most central macula zone, known as the foveola, is exclusively occupied by cone photoreceptors and is now smaller in diameter (725μm) due to an inward movement of the cones.[8] This has the effect of creating an increased cone density of 52,000 cones/mm^2. The cones have also matured at this age, with an increase in outer segment length of the cone to 22μm.

At 45 months of age, the foveal pit has decreased in diameter to adult proportions (650-700μm) and foveolar cone diameter is similarly to adult size (2μm).[8] However, both cone density (108,000 cones/mm^2) and the cone outer segment length (30μm) while increased, are both still only 50% of adult proportions. Additionally, the number of cones at the foveola has not increased from birth and also remains at 50% of adult numbers. These under-developed foveolar anatomy parameters have a significant effect on visual acuity and further growth beyond this age is required to reach adult levels.

1.3.2 Development of the Visual Pathway

Myelination of the nerve fibres of the visual pathway allows for rapid transmission of action potentials from the retinal ganglion cells to the visual cortex. During the later gestation period, there is progressive myelination of the visual fibres, occurring first centrally in the optic tract, then gradually outwards along the optic nerve, with the orbital portion of the optic nerve being the last to be myelinated.[9] The majority of visual nerve fibres are at least thinly myelinated at birth. Thickening of the myelin sheaths continues to progress rapidly in the immediate post-natal period, however, at seven months of age the myelin sheath is still not yet thick enough to provide maximum rates of neural transmission. This means that the partial myelination of the visual fibres limits the optimal rate of conduction of neural impulses. At 2 years of age, all the myelinated nerve fibres in the orbital portion of the pathway are found to be moderately covered by myelin and further layers of myelin

continue to be laid down until around 5.5 years of age, when heavy myelination is present. It is to be noted that some optic nerve axons remain unmyelinated even in adulthood and retain a low conduction velocity. The variable speed and organization of neural signals communicated by the visual pathway from the retina to the visual cortex must be able to facilitate all aspects of visual experience and are largely determined by the characteristic of retinal ganglion cell types and the temporal frequency and latency of neural impulses.

1.3.3 Critical period for visual development

Following birth, evidence shows that exposure to external visual stimuli is crucial for visual development.[10] This enhances neural connections within the brain, facilitating the capacity for vision and visual perception. In 1970, Hubel and Wiesel conducted seminal experiments, imposing monocular visual deprivation via occlusion of one eye at different ages of development in cats and for different time periods.[11] Their results revealed that even short periods of visual deprivation in early life, resulted in loss of response of those cortical cells in the primary visual cortex (V1) deprived of visual stimuli. This loss remained even after occlusion of the eye was removed, revealing the permanent nature of the damage. With increasing age, longer periods of stimulus deprivation were required to affect the same level of damage to V1 cortical cells. They also determined that after a certain age, monocular occlusion did not have any effect on cortical cell development. This stage of early development, where cortical cells could be permanently affected, became known as the critical period for development for neural structures.

1.3.4 Neural Plasticity

Neural plasticity is the time frame in which the brain is able to reorganize and enhance neural connections, in response to changes in stimuli from the external environment.[12] This period of neural plasticity extends beyond the critical period of development, with some suggesting that it extends till approximately the first decade of life [13] and others suggesting it covers the first six years.[14] The differences between these conclusions are due to differences in methodology where Keech and Kutschke[14] looked into the time frame for the onset of conditions that resulted in hindered visual development (amblyopia), while others have explored the time frame for both the onset of these conditions and for their effective

treatment.[13] Based on these measures, it has been suggested that the period of neural plasticity could extend up to 12 years of age.[13,15] More recent studies have observed improvements in vision with amblyopia treatment in children up to 18 years of age, indicating that there may be residual neural plasticity in older children.[12,16,17]

1.3.5 Ocular Conditions that could hinder normal visual development

There are a wide range of conditions that can cause vision impairment and blindness in childhood. Some of these are congenital or develop during infancy, such as retinal dystrophies, congenital glaucoma, ocular albinism, nystagmus, coats disease, optic nerve glioma, optic nerve hypoplasia and cortical vision impairment.[18] These conditions, however, are not the focus of this thesis as they are not able to be currently remediated and are therefore, not the focus of vision screening, which aims to detect enmasse treatable conditions. It is to be noted however, that some of these more serious conditions can present at screening, particularly if they are uniocular, mild in severity or at an early stage, such as some retinal dystrophies. Conditions such as large strabismus (eye turn), ptosis, cataract and retinopathy of prematurity (ROP), can be detected (or in the case of ROP, screened for) in infancy. Refractive errors, amblyopia and less noticeable strabismus can be found at pre-school age and are considered treatable conditions. When any of these conditions occur at an early age, in particular during the critical and neural plasticity periods of development, this can hinder normal cortical visual development in the affected eye/s and if left untreated cause permanent visual deficit.

1.4 Screening for Childhood Ocular Conditions

1.4.1 Principles of Screening

The aim of any screening program is to enable early detection of a condition that may have already caused pathological change but will benefit from timely management. In 1968, under the auspices of the World Health Organisation (WHO), Wilson and Jungner put forward 10 principles of early disease detection that represent the starting point for screening decisions.[19] These 10 principles covered key elements of screening including, what should be known about the health problem, the natural progression of the condition, and also the availability of treatment, the characteristics of the screening tests and the cost-effectiveness of detecting positive cases via screening. These are then all compared to expenses for the medical system when the condition is not screened for and presents with later, more substantial costs.

Box 1.1 Principles of Screening[19]

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(1) The condition sought should be an important health problem.

(2) There should be an accepted treatment for patients with recognized disease.

(3) Facilities for diagnosis and treatment should be available.

(4) There should be a recognizable latent or early symptomatic stage.

SCREENING FOR DISEASE

(5) There should be a suitable test or examination.

(6) The test should be acceptable to the population.

(7) The natural history of the condition, including development from latent to declared disease, should be adequately understood.

(8) There should be an agreed policy on whom to treat as patients.

(9) The cost of case-finding (including diagnosis and treatment of patients diagnosed) should be economically balanced in relation to possible expenditure on medical care as a whole.

(10) Case-finding should be a continuing process and not a "once and for all" project.

In 2018, a systematic review and modified Delphi consensus process was conducted to refine Wilson and Jungner's original 10 principles for population-based screening.[20] The consolidated screening principles were re-organised and elaborated into 12, and included components of recruitment, testing, information access, follow-up, patient education and support, staff training and program management and evaluation. The refined screening principles placed a greater emphasis on evaluating aspects of the screening program itself.

Box 1.2 Refined Set of Consolidated Screening Principles[20]

Disease/condition principles

1. Epidemiology of the disease or condition

2. Natural history of disease or condition

3. Target population for screening

Test/intervention principles

4. Screening test performance characteristics

5. Interpretation of screening test results

6. Post-screening test options

Program/system principles

7. Screening program infrastructure

8. Screening program coordination and integration

9. Screening program acceptability and ethics

10. Screening program benefits and harms

11. Economic evaluation of screening program

12. Screening program quality and performance management

1.4.2 Principles applied to Childhood Vision Screening

The need to screen children for a variety of ocular conditions has had a long history and is generally endorsed by eye health care practitioners. However, with the release of a systematic review of preschool vision screening based on the principles stated by Wilson and Jungner by Snowdon and Stewart-Brown in 1997, the practice of preschool vision screening was called into question.[19,21] The authors based their recommendation of consideration of discontinuing existing programs and not implementing any new preschool vision screening programs, was based on the lack of quality evidence found for the natural history of the targeted conditions, namely amblyopia, refractive error and strabismus. They also cited the inadequate evidence for any disability associated with these conditions and for the effectiveness of treatments.

One noted that the evidence cited by Snowdon and Stewart-Brown to argue that the natural history of amblyopia was uncertain and that it might spontaneously resolve with age, was of poor quality in itself[22] while the negative impacts of spectacle wearing and patching for amblyopia were overstated. The lack of randomised clinical trials to provide evidence that amblyopia treatment was effective, was observed to be likely difficult to overcome.[22,23] Creating controls for such clinical trials, by leaving children with amblyopia untreated, could in the light of extensive clinical experience, be deemed unethical. The challenge remains about how to address the provision of robust evidence to support childhood vision screening, while there is no evidence to suggest that vision screening of children does harm and that in fact, may do good.

In Australia, the National Health and Medical Research Council (NHMRC) commissioned the Child Health Screening and Surveillance Review in 2002. This report also found little firm evidence to support or reject the value of childhood vision screening but recommended the continuation of neonatal screening in the context of an adequate early detection program.[24] This review questioned and recommended further research into the extent of disability and burden of disease for amblyopia and benefits and harms of its treatment. By contrast, a later systematic review of the available literature regarding vision screening from neonates

to age 16, concluded that the evidence now supported the vision screening of pre-school children.[25]

In the same year as the NHMRC report, Rahi and colleagues published evidence for a crucial point, finding in later birth cohort recruited in 1958, that untreated uniocular amblyopia puts amblyopic individuals at an increased risk of vision impairment and blindness later in life.[26] Ocular pathology and injuries in the non-amblyopic eye conferred a 1.2% lifetime risk of bilateral vision loss for these people. In fact for individuals who had unilateral amblyopia and faced vision loss in their non-amblyopic eye later in life, 65% of them were unable to continue with paid employment and were less likely to have completed a university degree.

It has been suggested however, that the psychological implications of treatment such as glasses for refractive error and patching for amblyopia and any other instances of unnecessary treatment, may pose a risk of harm.[27] The likelihood for potential harms appear low and likely outweighed by the potential benefits of improving visual acuity, but actual evidence is scarce. It has been reported that while amblyopia treatment may be associated with some level of distress, it had no impact on the child's well-being or behaviour during or after the treatment period[28] and that on average amblyopia treatment was well tolerated by the child and their family.[29] However, in order to minimise any potentially harmful impacts, a balance between managing amblyopia and ensuring psychosocial well-being should be considered and included within treatment guidelines.[30]

The United States Preventative Services Task Force (USPSTF) has further supported pre-school vision screening, stating that treatment of amblyopia was effective and that the harms of this treatment were minimal[27]. The USPSTF's recommendation was that vision screening must be conducted at least once between age three to five years, with the aim of early detection of amblyopia and its risk factors, rating this to have moderate benefit. Moreover, guidelines developed in 2018 by the American Academy of Pediatrics (AAP)[31] and the American Association for Paediatric Ophthalmology and Strabismus (AAPOS)[32] have supported routine childhood vision screening on the basis of available prospective cohort

research and expert consensus, both of which suggest that vision screening is beneficial in decreasing the incidence of vision loss in early childhood.[33]

The United Kingdom (UK) National Screening Council guidelines published in 2013, also suggest that an orthoptic-led screening program for vision defects (amblyopia, strabismus, refractive error) should be offered for children aged four to five years.[34,35] The 2019 Canadian guidelines are also in support of childhood vision screening and suggest that at least one comprehensive ocular examination is necessary prior to school-entry.[36] In the Netherlands, a seven year follow-up of childhood vision screening found a reduction in the prevalence of amblyopia within screened populations, suggesting a positive effect of childhood vision screening.[37] Overall, despite wide variation in the implementation of screening programs internationally, overall current guidelines suggest that there is a benefit of vision screening in children, with most recommendations for universal vision screening to occur at preschool age.[27,33-36]

While the need to screen children for amblyopia is widely endorsed, Snowdon and Stewart-Brown[21] called the practice into question, primarily based on the lack of evidence that treatment led to improvement and whether it caused disability. Even though it has been found that untreated amblyopia poses an increased risk of vision impairment later in life[26] it is to be remember, that amblyopia is not the only condition detected by vision screening. Uncorrected refractive error is both a risk factor for amblyopia and accounted for a large percentage of vision impairment in preschool children in Australia (69.7%).[38] There also is evidence that learning and education may be impacted when leaving refractive errors uncorrected.[39,40] This makes both amblyopia and refractive error conditions that should be targeted by pre-school vision screening programs, so these conditions can be treated before school entry.

1.5 Amblyopia

Amblyopia is a condition where "the patient sees nothing and the doctor sees nothing."[41] It translates to *dim-sightedness,* derived from the Greek words *amblys* meaning "blunt" and *ōps* meaning "eye" or "to see." It is defined by reduced visual acuity in one or both eyes in the absence of ocular pathology and in the presence of an amblyogenic risk factor. Typically removal of pathology and/or correction of refractive error will not improve visual acuity to age-normal values when amblyopia is present.

There are three basic causes of amblyopia: stimulus deprivation, strabismus and anisometropia. How amblyopia arises is through one or more of the following mechanisms:

<u>Stimulus deprivation</u> occurs when there is a de-focused or absent image in one or both eyes in childhood, leading to a poor or no visual stimulus in the eye, and therefore in the visual cortex, resulting in amblyopia. Stimulus deprivation amblyopia is usually caused by high refractive error (due to images being substantially de-focused) or ocular pathology such as congenital cataract or ptosis that covers the visual axis, thereby preventing light entering the eye and affecting visual development. These forms of amblyopia can be monocular or in the case of bilateral cataracts or high uncorrected refractive error in each eye, amblyopia can be bilateral.

<u>Cortical Suppression,</u> also described as 'signaling inhibition', can arise due to disruption of binocular function, usually caused either by the presence of strabismus (an eye turn) or anisometropia (difference in refraction between the two eyes of at least one diopter). Children with strabismus use suppression to avoid a diplopic image, related to the strabismic eye receiving an image 'off' the visual axis. This means that the image in the strabismic eye is not aligned with the fovea. The incompatible nature of the images received in each eye, leads to the visual information from the strabismic eye being suppressed at a cortical level.

In anisometropic amblyopia, the unequal refractive error between the two eyes will cause the images in each eye to also be dissimilar, both in image size (anisocoria) and degree of

blur. Suppression of the input from the eye with the greater refractive error may occur if the degree of dissimilarity of images causes an inability to fuse the two mismatched images. This then leads to the eye with the greater refractive error to develop anisometropic amblyopia,[42] with higher degrees of anisometropia being associated with greater severity of amblyopia.[43-45]

If these conditions persist, it will halt visual development causing monocular amblyopia. It is to be noted that the severity of amblyopia has been found to be correlated with the strength of suppression.[46] Suppression will also prevent the development of binocular vision.[47] In an animal model (infant monkeys aged 3 weeks – 9 months), it was found that only relatively short periods (one week) of disruption of visual input could induce neural suppression and decrease cortical binocular disparity sensitivity.[48] This suggests that the development of vision is exquisitely sensitive to visual input during early development.

1.5.1 Anatomical changes in relation to Amblyopia

Cortical changes in response to stimulus deprivation were first described by Hubel and Wiesel in seminal electrophysiological and morphological experiments that took place over 50 years ago. In response to depriving one eye of visual stimulus by lid suturing in kittens early in life, they reported morphological changes (atrophy) and lesser physiological changes of the responses of cells in the layers of the lateral geniculate nucleus (LGN) receiving input from the sutured eye.[11,49] Importantly, they noted that these changes did not occur in an older kitten and adult cat, laying the foundation for the understanding of the critical period for development. A second paper in the same year reported similar physiological deficits in the visual cortex, however here the physiological effects were greater than the impact on cortical morphology.[50] In animals reared without visual deprivation, they had also found that 80% of cortical cells respond identically to both eyes, with only 10% responding to one eye only.[51] Yet when Hubel and Wiesel's experiments were conducted on kittens with imposed strabismus[52] despite both eyes receiving light stimulus they still found that neural abnormalities were produced in area 17 of the visual cortex, demonstrating that the cortical

cells redistribute themselves to favour the non-strabismic eye when there is an alteration to visual information reaching the other eye.

This has led to the understanding that when amblyopia develops there is inhibition of development of appropriate binocular neural connections in the primary visual cortex.[53,54] Therefore, functionally there is reduced cortical activity, confirmed in response to visual stimulation of the amblyopic eye at the level of the primary visual cortex as well as other extra-striate areas.[53,55-58] A reduction in the number of cells in the primary visual cortex that receive input from the amblyopic eye has also been reported, along with a reduction in the number of cells that receive binocular input (i.e. binocular cortical cells) [53,58-61] causing an overall reduction in grey matter volume.[62] Studies on humans and animals with amblyopia have also been found to have an enlargement of cortical receptive field size, which reflects the loss of spatial resolution frequencies and contrast sensitivity.[53,63-66] Studies on humans with amblyopia using visual evoked potentials and neuroimaging techniques have similarly demonstrated structural changes and reduced activation in the primary visual cortex.[53-55,58,67-70]

While it has long been thought that amblyopia was primarily a cortical condition, anatomical changes in the lateral geniculate nucleus (LGN) including those morphological changes reported by Wiesel and Hubel,[49] have been further reported, including reduced growth of parvocellular cells that are known to subserve higher levels of visual acuity and other largely cone photoreceptor functions, such as colour vision. These cells with reduced growth in response to amblyopia are found in layers II, III and V in the ipsilateral lateral geniculate body, and layers I, IV and VI of the contralateral lateral geniculate body.[71,72] Functional deficits have also been reported in the responses of LGN cells in relation to amblyopia.[73]

More recently, with the use of optical coherence tomography (OCT) there is some evidence that the foveal structure in an amblyopic eye is also changed. Observations include increased retinal thickness at the fovea, reduced foveal pit depth along the horizontal meridian and flattening of the nasal and temporal sides of the foveal pit compared to those without amblyopia.[44,74] Other studies have, however, reported that there were no

significant differences in foveal structure between the amblyopic and non-amblyopic eye,[44,75] but one of these studies found that the peripheral retina in the amblyopic eye was significantly thicker than the fellow non-amblyopic eye.[75]

There is also some disagreement in regards to whether these foveal changes only occur in the presence of strabismus and not in those with anisometropic amblyopia.[76] Landa and colleagues, however found that the retina at the fovea was thicker in both strabismic and anisometropic amblyopia eyes but noted that the foveal retina in eyes with strabismic amblyopia were thicker than those with anisometropic amblyopia.[74] The authors also noted that typically amblyopia treatment was more successful in those with anisometropic amblyopia, which they concluded could be attributed to anisometropic amblyopic eyes having less structural change than strabismic amblyopic eyes.

The retinal involvement in the genesis of amblyopia is an area that requires further research in order to fully understand its mechanism. Importantly, there is a need to clarify whether any retinal structural changes are evident before the development of amblyopia, and therefore causal, or are a consequence of the lack of cortical feedback to the retina after amblyopia has developed. In addition, it may be important to know if successful amblyopia treatment alters the retinal structure as vision improves and whether intractable amblyopia is characterised by failure in the development of foveal structures.

1.5.2 Natural History of Amblyopia

Amblyopia develops during the critical period for visual development in response to amblyogenic risk factors, and thus rehabilitation of the affected eye has its greatest effect through treatment initiated (occlusion therapy via patching or atropine) within the period of neural plasticity. It is to be noted that amblyopia does not develop after neural plasticity even in the presence of amblyogenic risk factors.[77] It has also long been observed that in younger children, if treatment is not carefully monitored, reverse amblyopia can occur.[78] While this again indicates the malleability of the infantile visual system, these cases are rare and usually temporary in nature.[79]

The effectiveness, efficiency and sensitivity to traditional amblyopia treatment declines with age, with less or limited success in older children using traditional occlusion therapy and the most successful treatment with superior visual outcomes is carried out with the period of neural plasticity.[13,80,81] While amblyopia can be reversed via occlusion and/or atropine penalization treatment, it is to be noted that while visual acuity is recovered in the immediate, there is a rebound effect that does occur in some children. One study reported that 27% of occlusion trials resulted in recurrence of amblyopia after treatment was discontinued and that this was inversely correlated with patient age (up to 10 years).[82] The PEDIG studies reported that 24% of successfully treated amblyopic patients deteriorating by at least two visual acuity lines within one year of cessation of treatment.[83] Similarly, it was observed that 17% of treated children had a recurrence of amblyopia in a study by Nilsson and colleagues, with all cases associated with microstrabimus, although this study had a very small sample size.[84] There are suggestions however, that weaning occlusion therapy may be a useful method for reducing the risk of amblyopia recurrence and that children should be monitored in case of recurrence to just past 8 years of age.[85]

Studies have less commonly reported visual acuity gains in response to amblyopia treatment in children up to 18 years of age.[16,17,86] A meta-analysis of four PEDIG randomized control trials confirmed that amblyopia is more responsive to treatment in children younger than 7 years of age (within the period of neural plasticity) compared with children 7 to 12 years of age.[87] More recently, perceptual learning and binocular treatments have been trialed and have been found to have greater success in older children and adults, however, the long-term effects, and costs associated with this are yet to be evaluated before being widely available.[81,88-92] Furthermore, it is not well understood whether this would be successful in children with severe amblyopia and whether the patients trialed for these treatment methods had previously had traditional occlusion therapy. Therefore, despite the potential for perceptual learning enhancing vision in adolescents and young adults, it is still recommended that amblyopia and amblyogenic ocular conditions must be detected as early as possible to ensure maximum benefit in achieving the best possible visual acuity with treatment.[93]

1.5.3 Epidemiology of Amblyopia

Uniocular amblyopia in most epidemiological studies is defined as a difference of two or more lines in visual acuity between the person's two eyes in the presence of an amblyogenic risk factor such as strabismus and/or anisometropia.[94-97] Bilateral amblyopia is diagnosed as significantly reduced visual acuity for age in both eyes, usually associated with bilateral high refractive error or congenital cataract in both eyes. These conditions are diagnosed after exclusion of other possible conditions and ocular pathologies that can lead to reduced visual acuity and after best-corrected visual acuity is determined, to eliminate the effects on visual acuity of uncorrected or under-corrected refractive error.

Population based studies have reported amblyopia prevalences between 0.4-3.4%.[94-104] A recent meta-analysis of 60 studies (1,859,327 subjects) showed that the pooled prevalence rate of amblyopia was 1.44%.[105] The variation in reported prevalences is in part due to differences in amblyopia definition, as some studies did not always explicitly require amblyopia risk factors to be present[100,102] or a two line inter-ocular visual acuity difference for diagnosis of uniocular amblyopia.[98,100,102,103] In addition, some children may not be classified amblyopic if they were already under treatment with improved visual acuity at time of examination,[98,100] which would result in an underestimation of the population prevalence. There are also differences in methodology, particularly related to method of ascertainment, including one study that was based on a home screening test conducted by parents which is likely to contain significant inaccuracy.[100]

There are four 'sister' population-based studies that used a consistent protocol in pre-school children; two are based in the United States; in Los Angeles (Multi-ethnic Paediatric Eye Disease Study -MPEDS) and Baltimore (Baltimore Paediatric Eye Disease Study - BPEDS), another in Australia (Sydney Paediatric Eye Disease Study - SPEDS) and lastly, a study based in Singapore (Strabismus, Amblyopia, and Refractive Error Study - STARS). These studies have less inter-study variation and found similar prevalences of amblyopia (1.2% to 2.1%) in preschool aged children (Table 1.1).[94-97] However, MPEDS, BPEDS and STARS did not include those children with a previous history (by parental report) of amblyopia or amblyopia treatment, which may have underestimated their prevalence of amblyopia, whilst SPEDS

included these children and had obtained letters from treating ophthalmologists to confirm diagnosis and treatment.

Table 1.1 Prevalence of Amblyopia in Preschool Aged Children

Population-based Study	Amblyopia Prevalence percentage (number of amblyopia cases/sample population)	Age Range (months)	Of the children with amblyopia, the percentage of children with:		
			Anisometropic Amblyopia	*Strabismic Amblyopia*	*Bilateral Amblyopia**
STARS[96] *(2010) Singapore*	1.19% (20/1682)	30-72	55.00%	15.00%	30.00%
MEPEDS[95] *(2008) Los Angeles, USA*	2.06% (69/3350)	30-72	56.52%	18.84%	21.74%
BPEDS[97] *(2009) Baltimore, USA*	1.23% (19/1546)	30-71	31.58%	31.58%	5.26%
SPEDS[94] *(2012) Sydney, Australia*	1.90% (27/1422)	30-72	25.92%	18.51%	37.04%

*The majority of cases of bilateral amblyopia were caused by high refractive error

SPEDS found that of the ocular conditions associated with amblyopia in preschool children, anisometropia conferred the highest odds ratio for amblyopia (OR: 27.82) followed by hyperopia (OR: 15.33) then strabismus (OR: 13.10, all *p<.001*).[94] Strabismus has been found to affect 0.8-3.4% of children aged 6 months to 6 years.[95-97] In a similar age group, anisometropia has been found to affect 1-8.5% of children aged between 3 to 6 years of age.[106-108] These conditions are significant contributors to the prevalence of amblyopia.

1.6 Refractive Error

One of the major causes of reduced vision in preschool children is uncorrected refractive error.[38,109] Refractive errors occur when there is an imbalance between the refractive power of the optical components of the eye (cornea and crystalline lens) and the eye's axial length. This causes the light rays entering the eye to come to a focus either behind or in front of the fovea, creating a de-focused image and reduced vision.

There are four main types of refractive state; myopia, hyperopia, astigmatism and emmetropia. Myopia (short-sightedness) occurs when the axial length of the eye grows too long for its optical power, resulting in light rays coming to a focus in front of the retina causing blurred distance vision. This form of myopia is the most frequently occurring.[110] Myopia less commonly occurs due to increased lenticular power (increased crystalline lens thickness and/or refractive index). This is a known entity in children who are born prematurely and while this can be associated with retinopathy of prematurity (ROP),[111-113] myopia of prematurity can occur in isolation.[113-115] Myopia occurring in the presence of childhood keratoconus, increasing the curvature of the cornea, should also not be overlooked.[116]

Hyperopia (long-sightedness) is when the axial length of the eye is too short and results in light rays to come to a focus virtually behind the retina. This tends to cause vision to be blurred at near. This is the most common refractive state in children and being mildly hyperopic is not a disadvantage in terms of vision, as children have a robust level of accommodation (increased optical power of the crystalline lens) that is able to overcome mild to moderate levels of hyperopia to obtaining clear distant and near vision. However, it is to be noted that children with even high levels of hyperopia can report normal or near normal visual acuity.[39] This is likely to be due to the application of excessive degrees of reserve accommodation, thus increasing the optical power of the eye so that light rays entering the hyperopic eye can still be brought to a focus on the retina.

Astigmatism is characterised by a difference in refractive power along different ocular meridians of the cornea and/or lens.[117] Simple astigmatism occurs when there is a refractive error in one meridian and emmetropia in the other. Compound astigmatism is when there is

a difference in power between the two meridians of the same refractive type, for example, hyperopic in both meridians but with a difference in power. Complex astigmatism is when the patient is myopic in one meridian and hyperopic in the other. Astigmatism will cause blurred vision through two focal points being generated (one focal point is usually less blurred than the other). When astigmatism occurs in conjunction with myopia, it does not further decrease visual acuity, which appears to be determined by the spherical power of the eye. However, it does affect visual acuity if there are only very low levels of spherical refractive error or with significant hyperopia, in those with higher levels of astigmatism (<-1.50D).[39]

Emmetropia is the state where there is no refractive error; that is the axial length of the eye and its optical components are at balance, causing images to be focused on the retina without any accommodation, resulting in clear vision. This is not a common state of refraction in childhood and it has been argued that emmetropia is not the biological endpoint of refractive development in children, with mild hyperopia being the more common state.[118]

1.6.1 Natural history of refractive development in childhood

Refractive error changes over the span of life, with the majority of neonates starting life relatively hyperopic (Table 1.2). The long held understanding of neonatal hyperopic refractive error was supported by Cook and Glasscock, who found that 74.9% of newborns had hyperopia ranging +1 to +12DS.[2] This early hyperopia is related to the relatively small eye in neonates (70% of adult size). A high prevalence of hyperopia and low prevalence of myopia (3%) has also been found in infants aged 1-48 months.[119]

However, MEPEDS found that children younger than 12 months may be less hyperopic than previously understood[120]. The mean refraction for African American children aged 6-11 months (+0.60) in MEPEDS was much less hyperopic and to a lesser extent for Hispanic children (+1.29D) than studies conducted on children of European descent.[2,119] Additionally their prevalence of myopia for the African American (13.7%) and Hispanic (6.4%) infants in the 6-11 month age group was also much greater than that found by those of European descent.[2,119,120] However, it is to be noted that, infants under 12 months of age could only

be administered 0.5% cyclopentolate for medical reasons. This is a potential limitation in accurately determining refraction at this early age and could result in a number of infants exhibiting pseudo-myopia. The increased prevalence of myopia in the 6-11 months age group in MEPEDS, may be attributed to the reduced efficacy of cycloplegia and exertion of accommodation, causing a myopic shift.[120] This limitation is enhanced when children have darker coloured irides (common for African American and Hispanic populations), which are known to resist the effects of cycloplegia.

Table 1.2 Average Refraction up to 6 years of age

Age	Mayer et al. 2001[119] (Boston, USA)	Kuo et al. 2003[122] (Tennessee, USA)	Ojaimi et al. 2005[123] (Sydney, Australia)	French et al. 2013[124] (Sydney, Australia)
1 months	+2.20	+1.40		
2.5				
4				
6	+1.79			
1 year	+1.57			
2	+1.19			
2.5				
3	+1.00			
4	+1.13			
5				
6			+1.27	+1.3

Note: Spherical Equivalent is given in lens diopters (D)

Interestingly, a study by Gwiazda revealed that the pattern of refractive development in the first year of life can be different to later years; with some babies who were originally myopic having a reduction in their myopic refractive error towards mild myopia or even emmetropia.[121] However, these results must be treated with some caution as non-cycloplegic retinoscopy was used to determine refraction, which again leads to the question of the accuracy of these measures. Furthermore, as there were no accompanying biometric measures of the eye it is unknown if this possible change was attributed to loss of optical power in the crystalline lens and/or cornea or changes in axial elongation, or both.

Following the first year of life, changes in refraction slow, corneal power reduction caused by flattening of the corneal curvature slows by the end of the first year of life and largely ceases by 1-2 years of age.[125,126] However, the power of the crystalline lens consistently decreases until about 10 years of age, after which it slows down.[127-130] This decrease in lens power over this time period, balances out increases in axial length, minimising the negative shift in refraction (myopic shift) that would occur if axial elongation was the only change in ocular biometric parameters.

1.6.2 Epidemiology of Refractive Error

Clinically significant refractive error is usually defined as a spherical equivalent of ≤−1.00 diopters (D) for myopia; ≥+2.00 D for hyperopia; and ≥+1.00 D cylinder power for astigmatism.[39] However in epidemiological studies, myopia is usually defined as a refractive error ≤-0.50D, hyperopia is defined with greater variation (≥+1.00 to ≥+3.00 D) and is often age dependent, with lower refraction chosen for adolescents and adults.[131-134]

Based on the STARS, MEPEDS, BEPEDS and SPEDS studies, it was found that the prevalence of hyperopia was greatest in the younger children and tended to decrease with age (Table 1.3).[131-134] A proportion of the population will remain hyperopic throughout childhood and into adulthood. This could be seen as a failure of the eye to emmetropise and may be due to genetic determinants leading to an overall shorter axial length.[127] Leaving high levels of hyperopia (>+3.50) uncorrected is a known risk factor of amblyopia (OR: 15.33)[94] and has been found to have a 13 times greater risk of strabismus[135] in preschool children. Additionally, sustaining high levels of accommodation, particularly in high hyperopia, may induce headaches, eyestrain and other aesthenopic symptoms. These in turn may even have a negative effect on a child's learning, as they may avoid activities such as reading that cause them discomfort. Hyperopia has been found to be associated with lower educational attainment and possibly reading difficulties, providing a further argument for early childhood vision screening prior to school-entry.[39,40]

Table 1.3 Prevalence of Refractive Error (Hyperopia and Myopia) in Preschool Aged Children

Population-based Study	Infants and preschool: 6-72 months		Preschool age: 36-72 months	
	Hyperopia >+2.00D	*Myopia <-0.50D*	*Hyperopia >+2.00D*	*Myopia <-0.50D*
STARS[133]	7.8%	11.4%	3.94%	4.5%
MEPEDS[131]	22.3%	3.19%	20.8%	6.1%
BPEDS[132]	23.6%	4.6%	21.6%	3.8%
SPEDS[134]	15.4%	3.3%	14.8%	2.2%

In the preschool population, myopia prevalence tends to be low due to shorter axial lengths in childhood (Table 1.3).[131-134] However, in recent years the rising prevalence of myopia[136] associated with earlier onset[137-139] has also been recognised as a major international public health challenge because of the associated public health costs[140-142] and the likelihood that earlier onset of myopia may lead to high myopia (≤-5 dioptres) later in life.[140,143] High and moderate levels of myopia are associated with severe sight-threatening pathology related to excessive elongation of the eye, such as retinal detachment, myopic maculopathy, staphyloma and a number of other associated disorders, including glaucoma and cataracts.[140,143,144]

Australia has been an outlier in terms of the distribution of refractive error, with a relatively low prevalence of myopia[145] and the Sydney Myopia Study (SMS) and follow-up Sydney Adolescent and Vascular Eye Study (SAVES) have confirmed this outlier status in children. However, Australia is not immune to the worldwide trend of increasing myopia prevalence, it has almost doubled in European Caucasian children over a 5-6 year period (Age 12 years: 4.4% vs. 8.6%)[124] within the Sydney studies. In addition, high levels of myopia were detected in high school students in academically selective schools[123,124,146,147] and 3.92% of 17 year

olds in these schools had high myopia and therefore at high risk of vision-threatening pathology later in life.[148] Delaying the onset and progression of myopia is crucial, as this will decrease the likelihood of progression to high myopia as there is less time for excessive axial elongation during the period of growth in childhood. This emphasizes the importance of timely detection of myopia in order to implement interventions (such as low dose atropine,[149] orthokeratology[150] more time spent outdoors[151] or optical intervention with spectacles with novel lens design)[152,153] to slow its progression early in life .

The prevalence of astigmatism varies between populations due to multiple factors, one being its definition across studies, with some defining astigmatism as ≥0.50DC[154] and ≥0.75DC[154,155] of cylindrical power and others as≥1.00DC,[154,156] ≥1.50DC[131,133,156,157] and ≥2.00DC.[156]

Another important factor for variation is ethnicity, which may have a genetic basis. MEPEDS found that African American (OR: 1.47, 9%) and Hispanic children (OR: 2.38, 13%) had more astigmatism compared to White American children (6%) aged 6-72 months.[158] A Chinese study of young children aged 3-6 years reported a moderate prevalence of astigmatism (21.1%)[159] while a studies of Taiwanese children (mean age: 5 years) reported a lower prevalence of 13.3%,[160] suggesting that all variation is not accounted for by ethnicity alone. The Collaborative Longitudinal Evaluation of Ethnicity and Refractive Error Study Group (CLEERE) study conducted on a wider age group of children (5-17 years), found again that Hispanic children had the highest prevalence of 36.9%, than children of Asian ethnicity at 33.6%, whilst African American children had the lowest prevalence of astigmatism (20%), even lower than White American children in the same study (26.4%).[161] In the multi-country Refractive Error Studies in Children (RESC)[155] examining similar age groups (5-15 years) with the same protocols, the highest prevalence of astigmatism (27.2%) was reported for children in Chile who are genetically linked to Hispanic children.[162] A very low prevalence of astigmatism of 4.3% was found in Nepalese children[163] and is somewhat similar to the low prevalence found in rural India (9.7%)[164] though it is to be noted that the Nepalese population is of mixed ethnicity with a significant proportion closely related to Tibetan people who are East Asian.

Age is also another important factor in the variation in the prevalence of astigmatism. During infancy (<12 months) it has been found that there is an increased likelihood of having astigmatism,[165] three times more than children who were 5-6 years.[158] This may be related to the early change in the axis of the astigmatism from a neonatal against-the-rule astigmatism to with-the-rule as the cornea flattens with eye growth. Interestingly, while the Northern Ireland Childhood Errors of Refraction (NICER) Study Phase 1 reported the prevalence of astigmatism to not have significantly changed at follow-up (6-7 years: 24%, 12-13 years: 20%), it was not always the same children who had astigmatism at both time periods but that while some children developed astigmatism, others became non-astigmatic.[156] These results were further supported in the NICER study Phase 2, which reported that the prevalence of astigmatism was unchanged in both the younger cohort (6-7 years: 17.5% and 9-10 years: 22.9%) and older cohort (12-13 years: 18.4% and 15-16 years: 17.4%).[166]

Astigmatism is also frequently associated with myopia.[167,168] In a study of Chinese children, who had a high level of astigmatism, it was found that they had a greater myopic shift in refraction and increased axial elongation.[159] Similarly, MEPEDS also found that participants with myopia were 4.6 times likely to have astigmatism than those without refractive error, whilst those with hyperopia were 1.6 times as likely.[158]

Uncorrected astigmatism has a negative association with multiple domains of academic readiness in pre-school aged children.[169] SPEDS also reported astigmatism as the main refractive error causing vision impairment (51.3%) in the preschool age group.[38] High degrees of astigmatism (>1.50D) has also been associated with the development of amblyopia and progressive myopia all of which forms a solid basis for aiming to provide early detection of this condition.[160]

1.7 The Target Age for Childhood Vision Screening

Determining the optimum age for vision screening has been a topic of debate. While it would be optimal to screen children as often as possible during the first eight years of life, several factors should be considered including: the child's capacity to cooperate and their ability to perform a standardised test, the age at which a condition manifests and maximum age for optimal treatment outcomes. Also to be considered is the level of skill required for a screener to perform the vision test and costs involved for the health care system, among others.[21,25,170-173]

Childhood vision screening can be broken up into four age stages that target differing visual abnormalities.[1] The condition targeted in prematurely born babies is retinopathy of prematurity, where timely detection and treatment of this condition is essential for the prevention of vision impairment.[174] During the infantile period, strabismus, congenital cataract[175] and congenital glaucoma[176] are also targeted childhood ocular conditions but are not always part of screening programs. Where Bruckner's reflex test is used, strabismus and opacities of the eye can be detected[177] while congenital glaucoma can be suspected through a range of symptoms and signs. The recommendation is that in children suspected of these conditions, screening is performed twice, once when they are newborn and again prior to 6-8 weeks of age.[177]

A large congenital strabismus is usually observed by family, friends and medical professionals or can be detected with the Bruckner's test.[178] To ensure the child gains potential for binocular vision by developing binocular cortical cells, strabismus surgery should be performed during the first 6 months of age.[175] Also if successful, this may reduce the likelihood of amblyopia. Detection of such conditions are usually prior to preschool due to the child having a noticeable ocular condition or demonstrating signs of functionally poor vision requiring further investigation.

Vision screening in preschool children aims to detect amblyopia, refractive error and strabismus (including microtropia and intermittent strabismus).[103,179,180] The majority of these conditions are not always visible and may not noticeably functionally affect the child, so would not yet have been addressed. Other conditions that can be detected at this age

which are considered more sight and even life-threatening and that can develop after infancy include optic nerve glioma, retinoblastoma and optic nerve hypoplasia.[181,182] These conditions are rare but are, on occasion, detected at vision screening.

The final group of children targeted for screening are older school children for whom refractive error is the main targeted condition, particularly myopic refractive error that has later onset.[1] By this age, it would be hoped that amblyopia and ocular pathology would already be detected and managed and therefore should not be the primary conditions that are targeted.

Traditionally, in Australia after screening of newborns, vision screening was performed at the school age of five to six years. This was primarily to successfully gain universal access to children of this age. However, at that age, these children are nearing the end of the period of neural plasticity and correction of ocular disorders at this age is not as effective as correcting them earlier to maximise visual outcomes, especially for amblyopia.[25,183] Detecting and treating ocular conditions at preschool age of four years, prior to commencing school, will ensure early access to treatment, optimising treatment outcomes[183] and will also address any problems prior to the first year of school. This may encourage confident and keen learners with optimal vision and may pre-empt any psychosocial factors that may arise from patching treatment for amblyopia.[28,30,184]

1.7.1 Infancy vs. Pre-school Screening

A study of a prospective birth-cohort in the Netherlands, the Rotterdam Amblyopia Screening Effectiveness Study (RAMSES) examined the effectiveness of a vision screening program consisting of both pre-verbal (1-24 months) and preschool (36-72 months) vision screening.[101] A final school screening at age seven years was performed by orthoptists to determine outcomes of early vision screening (n=2964). Of the 100 children with amblyopia (prevalence: 3.4%), 15% were detected by pre-verbal screening (predominantly strabismic amblyopia) and 41% by preschool screening. Twenty six children were self-referred, the majority who had strabismic amblyopia, which may be related to the visible nature of the strabismus. This study concluded that while pre-verbal screening detected strabismic amblyopia earlier, noting that cases of strabismus were more likely to be self-referred,

preschool screening from age three years onwards, contributed most to the detection of amblyopia. It was concluded that omitting screening at 24 months had little impact on the effectiveness of screening programs.[185] A further report from the same cohort calculated the sensitivity and specificity of the early screening to be 73% and compare to 83% at pre-school age.[37] Also noted, was that at age seven years there was residual amblyopia confirmed for only 23 children (0.8%). These findings support the argument for preschool vision screening, where the majority of children with amblyopia would be detected and have the opportunity for timely treatment. It is also important to note that some children would have developed amblyopia after infancy.[186]

1.7.2 School vs. Pre-school Screening

Studies have shown that the rates of screening generally increase with age, particularly from the age four to six, capturing a wider coverage of children, since they would be attending school or preschool [171,187]. Screening children in these locations means they would be more accessible and at these ages they will have higher testability, with the use of age-appropriate tests enabling them to perform subjective ocular tests including visual acuity. This will provide a better indication of their functional vision rather than relying on objective testing methods. Some studies have concluded that the optimal time for successful vision screening is specifically at school-entry particularly due to the high take up rate.[171,188] However, this has to be balanced against pre-school vision screening which increases the probability of superior treatment outcomes compared to school screening.

A population mother and baby cohort study known as the Avon Longitudinal Study of Parents and Children (ALSPAC) conducted in the UK, compared the effect of orthoptic screening (visual acuity and cover testing) at preschool age in districts with no preschool screening programs to the more common vision screening program at school entry at four to five years of age.[183] All children were re-tested at age seven years by an orthoptist and it was found that the prevalence of amblyopia was 45% less in the children who had received preschool screening compared to those who had not, although this was only of borderline significance. In further suppport, children who had been treated for amblyopia following preschool screening, had better visual acuity in the amblyopic eye than those treated after

detection at school-entry vision screening. In terms of optimal outcomes for amblyopia treatment, the evidence suggests that pre-school screening is, on balance, the most appropriate age to conduct vision screening.

1.8 Determining Vision Screening Test Protocols

The key test in vision screening is the measurement of visual acuity. Visual acuity aims to quantify the minimum separable detail that the eye can see. While visual acuity testing is easy to administer, minimally invasive and results are well understood, there is a lack of consistency throughout the world on which visual acuity test should be used for vision screening of the pre-school population. There is also question as to whether additional tests are required to improve the accuracy of vision screening. Other screening measures have been suggested, to be used in combination with visual acuity testing or alone. These include stereopsis, cover test, photoscreening and use of autorefractors without cycloplegia.

1.8.1 Criteria for Vision Screening

Screening tools can be evaluated by Wilson and Jungner's criteria: acceptability, reliability, and validity.[19] A test that is minimally invasive with an outcome that is well understood would be considered to have high acceptability by the person screened, their family in the case of children, and by the community, including the screeners and health practitioners who interpret the results. Reliability refers to the consistency of test outcomes and is an important measure of accuracy. Validity refers to the sensitivity of the test, that is, how well it can identify those with the targeted condition (true positives). It also important to ensure low numbers of false positives in order to avoid unnecessary stress on the child and their family as well as on the medical system and associated costs. Specificity is the ability of the test to exclude those without the targeted condition (true negatives). It is also important to have low numbers of false negatives, as not detecting those with an ocular condition, can have implications for the health outcomes for the individual.

1.8.2 Visual acuity testing for preschool age

1.8.2.1 Construction of Optotypes and Snellen Chart Design

In 1862 Snellen created a visual acuity chart that was based on the construction of "optotypes" rather than focusing on printing letter sizes.[189,190] His principle was that each optotype should be drawn within a 5 by 5 unit square with the thickness of detail to be one

fifth the dimensions of the whole optotype. He also primarily chose letters to be used as the optotypes and that each letter should fill the 5 by 5 square as evenly as possible.

Figure 1.1 Snellen Optotype [190]

Snellen defined standard vision as the ability to recognise an optotype when it subtended 5 minutes of arc with its detail subtending 1 minute of arc at the visual angle of the eye (anterior nodal point of the eye)[191] when viewed at distance of 20 feet or 6 metres.[192] This visual angle was nominally chosen based on the observation of the astronomer Robert Hooke that two luminous points or stars could be discriminated by the naked eye if they were separated by a distance that would subtend 1 minute of arc at that point of observation. This observation has been used to determine the dimensions of all standard optotypes and this theoretical formulation is the foundation for the construction of optotypes even today.

In 1959, Louise Sloan introduced a restricted set of optotypes with a known approximate difficulty to perceive, equivalent to the Landolt C optotype.[193] The 10 chosen letters that were designed and conformed to Snellen's principles were "S, D, K, H, N, O, C, V, R and Z." Sloan planned to use all 10 letters on each line of the visual acuity chart, except at larger sizes where the 10 letters would not fit. This would mean that each line of a visual acuity chart was of almost equal difficulty, with the size of the optotypes progressively decreasing in size down the chart. The chart was designed to have an approximate logarithmic progression in the size of the optotypes.

Figure 1.2 Sloan Distance Visual Acuity [193]
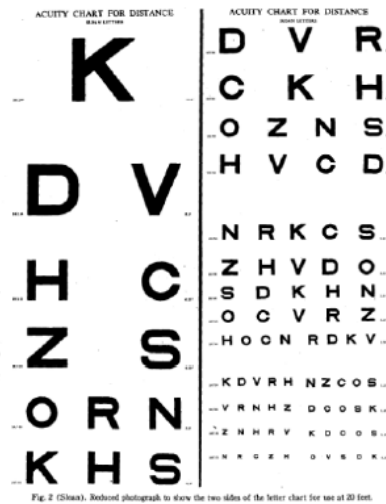
1.8.2.2 Crowding and Amblyopia Detection

It is well recognised that optotypes presented linearly (in lines of systematically varying optotype size)[194] or single optotypes surrounded by bars at a set distance, known as crowding, improves the detection of amblyopia (target condition of preschool screening).[195-197] A lack of crowding around the test optotypes, such as occurs in single letter booklets, typically overestimates visual acuity, which could have an impact on amblyopia detection and monitoring of treatment for amblyopia. Hilton and Stanley (1972) found there was a reduction in visual acuity ranging from one to six lines when comparing visual acuity achieved from a crowded visual acuity test (Sheridan Gardiner Linear) and a non-crowding visual acuity test (Sheridan Gardiner Singles) in patients with amblyopia.[198] Interestingly, when looking at the patient's non-amblyopic eye, such differences in visual acuity using the two methods was not observed, which is indicative that the eye with amblyopia is more affected by crowding than non-amblyopic eyes. Therefore, crowding is deemed essential to a visual acuity test to increase its sensitivity for detection of amblyopia. In children's visual acuity tests, crowding is achieved by either presenting the optotypes in linear format or with crowding bars around a single optotype. Another method is to have the test optotype ringed by a series of equally spaced optotypes to achieve uniform crowding.

1.8.2.3 Letter-based Visual Acuity Testing in Children

In 1930 Pugmire and Sheridan found a need for a visual acuity test designed for school aged children (five years and older) in order to increase the testability of visual acuity in these children.[199] A visual acuity chart was created with the chart having no more than three letters on each line and with the child tracing the shape of the letter in the air if they could not name it. The letters chosen were based on the idea of common letters that are used in everyday life and seen in books, such as 'O V X S U N T E A and L.' These letters were then revised based on feedback from their original use, to incorporate the optotypes now known as STYCAR letters ' V A T O H U X C and L.' The STYCAR visual acuity chart also had only 3 letters on each line and was designed to be performed at a six metre distance with a card containing the letters for the child to match instead of drawing the shape of the letter in the air.[200] However, it was noted that children demonstrated less interest in the visual acuity chart at a six metre distance, and the test was changed to remain a six metre test but using a mirror, so as to bring the tester closer to the child to improve testability. The only non-reversible letters (C and L) from the nine letter set were removed and a matching card continued to be used. The viewing of the letters through a mirror was not successful in this age-group and the testing distance was moved to three metres.[201] This test was called Sheridan Gardiner and used a flip booklet with single letters isolated in the centre of the page. This improved testability amongst younger children compared to a visual acuity chart presented linearly.[80,201] However, the single letters lacked sensitivity in detecting some ocular conditions, in particular amblyopia due to the lack of crowding.[80,197,198,202]

The single letters or optotypes from the flip book were also incorporated into a linear Sheridan Gardiner test with optotype size evolved from the Snellen visual acuity chart but using the reduced set of optoypes to improve testability in children, particularly those who were pre-literate.[200,203] The Sheridan Gardiner vision test initially used seven mirror-image letters A H O T V U and X. Further adaptations to the test were made for use in children younger than five years of age, using the letters H O T V and X. Sheridan found that there was confusion by younger children of the letters V and X reduced the set further to the four letters H O T V for use in this younger age group.[200,203]

A primary limitation of the Snellen visual acuity test, is the non-uniform spacing between letters and lines of letters in these visual acuity charts. This was addressed by the development of vision tests using logarithmic progression of optotype size (logMAR), with equal spacing between lines of optotypes and the optotypes themselves.[204] This method of progression of optotype size was subsequently introduced into visual acuity tests for children.[80,195,205,206] The Sheridan Gardiner HOTV letters have been incorporated into the gold-standard logMAR chart to provide a standardised vision chart design for use in young children, with the reduced letter set potentially increasing testability. No study to date has made a direct comparison of the Sheridan Gardiner vision test to a logMAR vision test using the HOTV letter set for testing vision in children. This comparison would be particularly useful for the Australian NSW Statewide Eyesight Preschooler Screening (StEPS) program since their recent change to HOTV logMAR from Sheridan Gardiner Linear vision charts could have an impact on the program and appropriateness of referral criteria for the StEPS program.[207]

The Amblyopia Treatment Study (ATS) developed an electronic visual acuity (EVA) testing method that has proven to have high testability in children aged 3 years and under, 80%[208] rising to 93% in those aged three to four years;[209] peaking at >94% in those aged four and over.[210,211] This chart has a logarithmic progression of optotype size. It also follows Snellen principles in the design of the limited HOTV letter set, making it suitable for the four year old age group.[212] This test is presented as a single optotype with four crowding bars surrounding it, spaced at half the optotype size distance from the central optotype. This creates sufficient crowding to ensure detection of amblyopia. This test is commercially available https://www.mstech-eyes.com/products/category/other-speciality-products.

In a prior study on Australian preschool children, it was found that the ATS EVA test resulted in visual acuity that was approximately one line better in comparison to the linear ETDRS or HOTV logMAR visual acuity tests.[208] Even when conducted on children older than five years, this one line difference was also evident. All these visual acuity tests are correctly calibrated. The one line difference may be due to cognitive ability rather than purely about visual capability, since the ATS EVA test design involves a single optotype with crowding bars (find

the letter in the box) may be less confusing for a child than a linearly presented test. However, in this study, ATS EVA was always tested first and the poorer visual acuity obtained on the ETDRS or HOTV logMAR charts may also be due to fatigue. However, the study findings would suggest that the ATS EVA is a highly appropriate visual acuity test to consider in the context of ensuring high testability. However, it required a computer and monitor which may not be ideal for transporting between screening sites and it may prove too expensive for a vision screening program.

Another vision screening visual acuity test that has been developed in Australia is the Melbourne Initial Screening Test that is used in Victoria, Australia.[213] The test consist of five letter Sheridan Gardiner Single Test (H,O,T,V,X) with a matching board at three metres for preschool children aged 42 to 54 months. This test was found to have a high level of testability in that age group (95%). The limitation of this test was the use of only a single optotype size (6/10) with a pass/fail criterion and therefore being unable to grade the severity of the reduced vision which can affect referral pathways. This also meant that a difference in visual acuity between the two eyes would not be recorded, thereby removing a crucial part of the definition of amblyopia. Furthermore, since this was a single-optotype test without the use of crowding bars, the detection of amblyopia would also be compromised as previously discussed.

The Keeler crowded logMAR chart used at three metres, is another letter-based chart used in the United Kingdom for vision screening of four to five year olds, with a reported sensitivity of 70.4% and specificity of 82.2% at the 6/9 cut-off.[214] In a comparison to the Snellen visual acuity test, it was found that the Keeler test had a higher false positive referral rate (Keeler: 17.95%, Snellen: 15.09%).[215] However, this was found to be due to the use of a 6/7.5 visual acuity cut-off which coincides with the age-normative VA value for 4-5 year old children. After lowering the cut-off to 6/9, it was found that the two tests had similar rates of false positives (Keeler: 7.69%, Snellen: 7.08%).

The tumbling E chart is commonly used in countries where the Roman (Latin) alphabet characters are not used [216-225] and has been shown to have high testability, as the optotypes

are directional and do not require the child to know their letters. It was reported that 90% of children aged three to six years were able to perform an ETDRS tumbling E chart.[225] Another study in Portuguese children aged three to four years also found high testability (95%).[226] Although Thomas and colleagues found the Tumbling E to have high sensitivity in detecting significant refractive errors (90.2%), the specificity was found to be lower at 69.8%.[227] However, Sanker and colleagues found the linear logMAR E chart to have a lower testability of 77.7% in a group of children aged three to six,[228] so there is some uncertainty about the value of this test in a preschool population. There also seems to be discrepancy in regards to the normative visual acuity achieved with the tumbling E chart compared to the HOTV logMAR chart. Children were not achieving an average of 6/7.5 on the tumbling E chart,[225] which would be the expected age norm visual acuity for a five to six year old child.[229] It was found that when a common cut-off of 6/9 was applied, nearly 90% of all children aged four would be referred, which would lead to a high number of false positives.[225] This raises the question of whether the reduced visual acuity found when using the Tumbling E is due to cognition limits and fatigue rather than an accurate representation of a preschool child's visual acuity.

Although the Tumbling E chart is a directional letter test which conforms to Snellen's principles, it is restricted in that young children have a limited comprehension of right and left.[230] It may also not be as accurate as using HOTV or the four Lea symbols, since there are only three reliable directions to test (down, up, and horizontal (left or right)[1] due to confusion of right vs. left directionality in children. Therefore chance can play a greater role in the determination of visual acuity in preschoolers. Further research is required to determine whether this chart is suitable for vision screening in preschool children and whether countries using this chart should consider changing to HOTV logMAR or Lea Symbols with a matching board instead. At this stage however, the American Academy of Paediatrics does not recommend Tumbling E for use in preschool vision screening, as such young children will not have developed the ability to express orientation of optotypes.[194]

1.8.2.4 Picture Visual Acuity Tests in Children

Although using letter optotypes are ideal in conforming to Snellen's principles, they are not always ideal for testing children's visual acuity, particularly for pre-literate children. Picture or shape optotypes are thought to be more acceptable and engaging for younger children.[231] As such, the children may be able to name or match a picture more easily than a letter, potentially increasing the testability in children aged two to four years who are classified more difficult to examine.[229,231-234] The use of such optotypes, particularly with matching cards, may enable more reliable visual acuity results to be obtained in children younger than pre-school age.

There is, however, a limitation in the majority of picture-based visual acuity charts. It is difficult to follow Snellen's principles for optotype design, such as constructing pictures with the defining detail being 1 unit of size within the 5 x 5 overall size, as well as ensuring standardisation between pictures. This means that the way to best conform to Snellen principles would be enlargement of the optotype itself. Despite the improvements that have been made, picture optotype visual acuity tests are not directly comparable to letter-based visual acuity tests.[231,233,234] However, picture charts are considered very valuable in testing the visual acuity of pre-literate children and are often the only way to test them.[231] However, wherever possible it is important to move to letter-based visual acuity tests, which is possible in children who are old enough to match letters and who are at an age where it has been shown that letter based vision charts have high testability.

There are numerous picture visual acuity charts that have been created to examine young children. Of the picture tests, Lea Symbols is highly favoured, while the Allens Figures that have previously been used in screening, are no longer recommended for use.[194] Allens Figures were not standardised and did not follow Snellen's principles. They also had the tendency to overestimate visual acuity in comparison to the Snellen chart when tested on adults.[235]

Kay Pictures

Another picture test that has been used in vision screening, particularly in the United Kingdom are Kays Pictures.[236,237] These pictures, although differing somewhat from Snellen's

principles, do have a form of standardisation. Instead of the five by five square for its optotypes, it used a 10 by 10 square, subtending overall 10 minutes of arc instead of five minutes of arc, but the detail still subtends 1 minute of arc.[238] It is this change in construction that may underlie the tendency for Kays pictures to overestimate visual acuity compared to a letter based visual acuity test.[234,239,240] Additionally, some of the optotypes in Kay's pictures were much easier to identify than others with some containing more detail within the optotype than others, making some of the sizes of optotypes more readily identified. Furthermore, some of the original Kays pictures required modifications to make them more familiar to children today; for example the telephone used in Kay's pictures is no longer identified commonly as a phone in this age of the mobile phone.

Figure 1.3 Optotype from the Kays Pictures test[238]
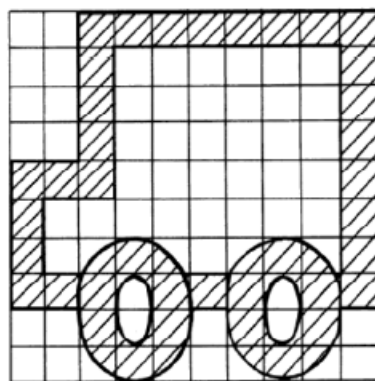
In recent years, Kays Picture underwent some modifications to address this issue.[241] The new test design involves a six-picture format with single picture presented with crowding bars. It was found that the latest optotypes were reliably recognised by the paediatric population (18 months to five years) and this new version has demonstrated good reliability and comparability to the gold standard logMAR visual acuity assessment.[241] While Kays Pictures has been shown to overestimate visual acuity in comparison to letter-based vision tests[236,237] by around +0.10 logMAR (one line)[237] the test was able to reliably identify interocular differences in visual acuity, demonstrating its capacity to identify amblyopia. Kay

Pictures is widely used in paediatric clinical practice, however, further research would be required to confirm its usefulness in vision screening.

Figure 1.4 A singly crowded optotype from the updated Kays Pictures[241]

Lea Symbols

Lea Symbols were developed with the purpose of ensuring standardisation in picture optotypes. The chart uses the same principles as the logMAR chart with logarithmic progression of optotype size and the same number of optotypes per line. It uses four readily identifiable symbols (house, square, circle and apple) that have been calibrated against the Landolt C vision test for standardisation and are 1.5 times larger than the equivalent Snellen optotypes.[242]

Figure 1.5 Picture of Lea Symbols[242]
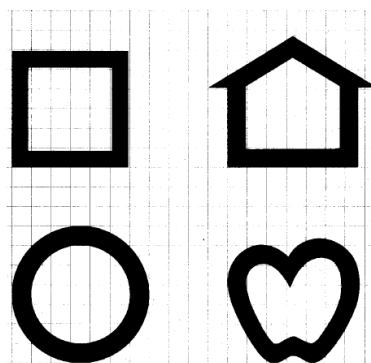
Comparison of Lea Symbols and Kays Picture Test

The testability of Lea symbols at three and five years of age (97.2%-99.4%, respectively) is somewhat higher than that found with the older version of Kays Pictures (three to 16 years: 86%).[239,241,243] However, the revised Kays Pictures has more comparable testability at >95% for children aged three to five years.[244] While Kays Pictures has been found to overestimate visual acuity, Lea Symbols do not, making it a more suitable test for vision screening.[236]

Comparison of Lea Symbols and HOTV Test

Numerous studies have compared the use of Lea Symbols to the HOTV test, which are both visual acuity tests currently recommended for preschool screening.[194] They have high testability, with improving testability with age in children aged two to six years.[245-248] For pre-school aged children, testability for Lea Symbols was greater than 90% and for the HOTV tests when presented in a single line format with crowding bars.[243] At age four, both these tests had greater than 95% testability, with a slightly greater testability with Lea Symbols (Lea: 97.2% and HOTV: 96%). The linear HOTV logMAR test also has high testability (≥90%) in children aged four to six years.[208,249,250]

Poor reliability was found for both tests (Lea Symbols r=0.63, HOTV r=0.71) when re-tested six weeks apart however, this was attributed to the majority of children improving their visual acuity due to a 'learning effect.'[251] Other studies have reported better reliability for these tests, with Holmes and colleagues reporting a high rate of reliability (r=0.82) for the HOTV test.[212] In younger children aged three, there was no difference in visual acuity measures on re-test for the Linear Lea Symbols but there was one line improvement for the Linear HOTV logMAR test.[247]

In terms of validity, Lea and HOTV have been found to not be significantly different. For children aged four, it was found that the sensitivity for detecting at least one ocular condition was 65% with the Lea Symbols and HOTV: 57% while specificity was high for both tests (Lea Symbols: 90%; HOTV: 87%) and not significantly different.[233] Therefore, both these tests seem equally valid for use in the detection of common childhood conditions: amblyopia, refractive error and strabismus. Lea symbols may have a slight advantage in

screening younger children (≤ three years) tending to record better visual acuity than HOTV optotypes,[233,243] possibly due to familiarity with pictures versus letters.[231] However, this difference was not statistically significant. Therefore, both tests can be recommended for vision screening purposes for the three to five age group, though the wider utilisation of HOTV may make it more acceptable.[194]

1.8.3 Other test protocols used in screening

1.8.3.1 Cover Test

Cover test is the gold standard test for the detection of strabismus. The inclusion of a cover test in the Vision in Preschooler's study screening program, improved the sensitivity for detection of strabismus by up to 25%.[209] However, cover testing is a very technical skill and can only be performed accurately by those who have undergone specialist training, such as orthoptists, optometrists and ophthalmologists and this may impose a cost that can't be justified if strabismic amblyopia is readily detected via vision testing. At screening, large angle strabismus can be readily observed by trained lay screeners and nurses. It is likely that intermittent strabismus and micro-strabismus may not detected at screening but these are mostly less problematic in terms of visual acuity. Cover tests may be more appropriate in the setting of secondary screening by orthoptists.

1.8.3.2 Stereopsis

While cover testing is gold standard for detection of strabismus, stereopsis tests are easier to administer and can be performed by lay screeners. Children with strabismus may have disruptions in their binocular vision, while poor vision in one eye or both can be reflected on testing stereopsis. Intermittent strabismus may go undetected if a reasonable level of binocularity is retained and children with a microtropia, may demonstrate a reduced level of stereopsis.[252] Despite the utility of stereopsis tests there is disagreement about their reliability as screening tools for amblyopia and strabismus.

In a small study of participants aged four to 78 years, examining the validity of four stereoacuity tests for the detection of strabismus, Lang I was found to have the best sensitivity (89.8%) and specificity (95.2%) while Lang II was slightly less (84.7%; 79.8%

respectively).[253] In contrast a large study of six year old children found that Lang II had sensitivity < 32% for anisometropia, strabismus and amblyopia but high specificity (>98%).[254] The age difference between these studies may have been a contributing factor.

Other studies, using a range of stereoacuity tests such as: Frisby, the Randot test, Titmus, TNO, Stereosmile, The Randot Preschool stereoacuity test and the Random dot E have been evaluated in children aged between two to 13 years and have concluded that their sensitivity for detection of amblyopia has been low (10-47%) when the specificity is set to a high level (85-98%).[243,255-257] This demonstrates that even in the presence of amblyopia, children are able to pass stereopsis tests.[258] These studies have demonstrated that stereopsis tests cannot be utilised as a screening tool on their own for the detection of amblyopia and strabismus and may not make any additional contribution to visual acuity testing.

Ohlsson et al.'s study, conducted in 12-13 year old children, concluded that not only were none of these stereopsis tests suitable for amblyopia or strabismus screening, but that the results obtained were variable for children without ocular conditions as well.[256] It is unlikely that this is an issue of cognition in this age group and indicates that there was no defined way in which a normal response could be differentiated from problematic response in those without any ocular condition. However it must be noted that all stereopsis tests in this study demonstrated higher sensitivity for the detection of strabismus than for amblyopia and as these are tests for binocularity, cases of anisometropic amblyopic may be less detectable. This may require further research in pre-school populations, using age appropriate tests, to determine if any additional cases of strabismus would be detected beyond those found by observation and vision screening.

1.8.3.3 Instrument-based screening for ocular conditions

There are primarily two instrument-based options for screening for ocular conditions in young children; they are photo-screening and non-cycloplegic autorefraction. While instrument-based screening is not a focus of this thesis, the merits of these methods of screening will be briefly discussed here, in the context of pre-school screening for ocular abnormalities. The main advantages of both these methods of screening is that they

produce relatively objective measures of ocular parameters, they are non-invasive, are relatively quick to perform, are reliable even in those children with limited literary or cognitive capacity and require minimal cooperation from the child. Their main disadvantage is cost and they have some limitations. Judgement of their effectiveness must be ultimately measured against whether they confer any benefit in screening a child who is able to co-operate with age-appropriate visual acuity testing.

Photo-screeners capture an image from a child's undilated pupil (non-cycloplegic) through a camera system coupled with off-axis (eccentric) flash of light causing a crescent shaped red reflex (crescent shaped). This appearance of the crescent-shaped reflex allows binocular measurement of refraction and eye alignment, made by an examiner looking at the photo images or by using a computer-based analysis system that may be incorporated in the device. This means that photo-screeners are designed to detect refractive errors and amblyogenic risk factors such as anisometropia or strabismus but are not able to directly determine amblyopia per se. The first commercial photo-screener became available in the late 1990s known as the MTI photo-screener.[259] There are now a variety of portable commercial photo-screeners available, the best known being the plusoptiX, also known as the Power Refractor and the Spot screener. While found to be highly testable in pre-school and younger populations, overall the reliability of these devices in identifying amblyogenic risk factors has been found to be variable.[243,260-264] More recently, an infrared photo-screener (2WIN)[265] and a smart phone app known as GoCheck Kids[266] have been developed. These, as yet, require testing in large pre-school populations to determine their utility and reliability.

A recent systematic review of the use of photoscreeners and auto-refractors in screening programs noted that these devices aim to detect amblyogenic risk factors rather than amblyopia itself[267] and that the most cost-effective approach is vision screening of children at an age when they are able to perform visual acuity tests. This agrees with the conclusion that the use of photo-screening appears to be most effective in children younger than pre-school age[268] and these devices are efficient and effective until an age when a child can reliably perform optotype-based vision screening effectively.[269] It is the recommendation of

the American Academy of Pediatrics [270] and the US Preventive Services Task Force[271] that photo-screeners are recommended for use in children three years old and younger.

Auto-refractors use a projected infrared light source and sensors to detect when the light reflected by the eye is correctly focused and they are a widely employed clinical and research tool, used both without and with cycloplegia. As their name suggests they are designed to measure refraction and are therefore able to detect significant refractive errors, including anisometropia. The best known of these are the hand-held SureSight Screener and Retinomax. The significant difference between auto-refractors and photo-screeners is their ability to detect strabismus. It should also be noted that all auto-refractors, when used without cycloplegia, have a high accuracy in measuring astigmatism, including determining its axis and may be useful as a screening tool in populations with a high prevalence of astigmatism.[272] The Retinomax has been widely used as a screening, clinical and research tool to measure refraction in paediatric populations, however it has been found to not be a valid tool even when used with cycloplegia, as overall the Retinomax appears to shift measured refractions in a myopic direction[273,274] and this may be of concern if trying to detect early-onset myopia and significant hyperopia. The Retinomax K-Plus and Retinomax K-Plus 2 without cycloplegia have been found to have a low sensitivity for detecting hyperopia (33-46%).[261,275]

1.8.3.4 Screening for significant hyperopia

Children in particular have a large amount of accommodative reserves and therefore have a high ability to accommodate, enabling them to overcome hyperopic refractive errors for the duration of a screening test. This is evident even when testing visual acuity in young adolescents, where even high levels of hyperopia could be overcome by accommodative reserves in some instances.[39] Similarly, using photo-screeners and auto-refractors without cycloplegia, hyperopia will frequently be underestimated and myopia overestimated at all ages,[276] but particularly in children.[277,278]

While children with significant hyperopia often perform well on visual acuity tests it has to be asked whether they can sustain the same level of accommodation over a period of time.[40] Are they able to sustain it throughout a school day for example, with poor academic performance having been linked with uncorrected hyperopia.[279] Uncorrected hyperopia has also been linked to deficits in attention, visual motor integration and visual perception[280] while correction of hyperopia may alleviate some of these issues.[281] If this is considered in conjunction with the link between moderate to high hyperopia and amblyopia, it is a refractive state that needs to be effectively screened for.

At this present time there is no reliable method to detect significant hyperopia in screening settings and cycloplegic refraction remains the only effective method. There is some possibility that ocular biometric measures, such are measured by non-invasive devices if translated into a portable cost-effective device may have some utility when used without cycloplegia. Such technology exists in the IOLMaster (Carl Ziess Pty Ltd) which is table mounted and has been shown to have moderate to high testability in children older than 3 years.[232,282,283] The axial length of the eye (AL) is known to be correlated with its refractive state, with a shorter axial length indicating hyperopia and a longer axial length, myopia[284] and the relationship between AL and refractive error overall has been shown to have a moderate to high correlation.[285]

It is also known that males tend to have longer axial lengths offset by flatter corneas than females.[284] Combining corneal radius (CR) with AL to calculate a ratio known as AL/CR may provide a good indication of refractive state of the eye. The emmetropic eye is theoretically said to have an AL/CR ratio of 3.[286] A lower AL/CR ratio would therefore could be indicative of hyperopia. Thus far AL/CR has been primarily used to predict myopia[286-288] and recently, Scheiman and colleagues described longitudinal changes in corneal curvature/radius and axial length in 6-12 year olds and found that increases in the AL/CR ratio were found as myopia progression occurred.[289] This study also found that AL/CR had a greater correlation to magnitude of myopia than axial length alone ($p<.001$). Therefore, AL/CR may be both an indicator of hyperopia as well as a useful predictor of myopic onset and progression level. If the usefulness of the AL/CR ratio could be further proven and its costs and practicality could

be assessed, the development of a non-invasive screening ocular biometric device may provide a solution to the vexed problem of screening children for significant hyperopia.

1.9 Referral Criteria and Normative Visual Acuity

Referral criteria are dependent on the test used and age of the child. Visual acuity develops with age, therefore normative visual acuity according to the age of the child needs to be considered (Table 1.4). At this point, there is no universal referral cut-off criteria considering the fact that differing age groups/ stages are vision screened along with various visual acuity tests used in screening programs (Table 1.5). Whilst the normal visual acuity is accepted as 6/6 in adults, children do not attain this level of visual acuity until early school years. In the Sydney Paediatric Eye Disease Study (SPEDS) for children aged three, the mean visual acuity was 6/11 using a logMAR chart (ETDRS or HOTV) and improved to 6/7.5 for children aged four to six years[229] (Table 1.4). In line with this, the StEPS program uses a cut-off visual acuity of 6/9 using either the HOTV logMAR or Sheridan Gardiner Linear vision test, which is ideal considering that the normative visual acuity for preschool children is 6/7.5.[184,207,290] Similarly, in the UK, a referral criteria of <6/9.5 in one or both eyes is used to determine whether a child has failed vision screening using the Keeler crowded logMAR chart for preschool children.[214,215]

A study conducted in the UK examined differing visual acuity referral criteria using the Sheridan Gardiner Singles test, found that when using a cut-off criteria of 6/6 (visual acuity worse than 6/6) there was a high rate of false positives compared to using a 6/9 cut-off. However, the downside was that at the 6/9 cut-off, sensitivity had declined to 70.6% from 97.2% at the 6/6 cut-off indicating that some children with ocular conditions may be missed.[291] Using an intermediary vision cut-off of 6/7.5 still led to a high false positive rate, with the 6/9 cut-off allowing for improved accuracy.[215] The Swedish country wide vision screening program for four year old children using HOTV logMAR prior to 1992 had a referral cut-off of less than 0.8 decimal (6/7.5) re-examined those children 18 months later who had vision between this cut-off and the 0.65 decimal (approximately 6/9.5) level and found that few required any treatment, suggesting that a cut-off of 6/9.5 was appropriate.[292] When later re-examining children aged 6,[293] it was again found that children who had had a vision screening result of 6/7.5 rarely needed treatment. However, in another study it was shown that that raising the cut-off to visual acuity ≤6/12 in children aged four to five years achieved a sensitivity for ocular conditions of 86.4%.[294]

Whilst these studies have demonstrated that the sensitivity at the 6/12 cut-off is high, there is the risk that children with ocular conditions may be missed at this level of visual acuity. A ≤6/9 criteria has not been demonstrated to result in over-referral and so would be the more appropriate criteria. Using a referral criteria of 6/7.5 or 6/6 would result in over-referral with a large number of false positives (high number of children with no ocular defect) considering that the normative visual acuity for preschool children is 6/7.5. Using a slightly higher cut-off of 6/9 means that the number of false positives can be kept to minimum whilst also ensuring that children with ocular conditions are not missed.

Table 1.4 Normative Visual Acuity up till 6 years of age

Age	Mayer et al. (1995)[295]	Leone et al. (2014)[229]	Pan et al. (2009)[296]	Drover et al. (2008)[297]
1 months	6/240			
2.5	6/95			
4	6/75			
6	6/38	6/30		
1 year	6/24			
2	6/19	6/24		
2.5	6/15	6/18	6/9.5	
3	6/9	6/11	6/9	6/7.5
4	6/7.5	6/7.5	6/7.5	
5			6/6	6/6
6				

Table 1.5 VA Charts and Referral Criteria for Childhood Vision Screening

Country	Target Age Division	VA Chart	Testing Distance	Referral Cut-off	Accuracy and Testability
Australia (NSW) [184,207]	Preschool (4 years)	Sheridan Gardiner, HOTV LogMAR	3 or 6m	6/9^{-2}	
Australia (Victoria)[213]	Preschool (3.5-4.5 years)	Sheridan Gardiner 5 single letter optotypes	3m	6/10	Testability: 95%
Canada[298,299]	Infants and preschool	HOTV LogMAR, Lea Symbols	-	-	
Croatia[250]	Preschool (4-6 years)	Lea Symbols	3m	6/9	Sensitivity: 100% Specificity: 96.68% Testability: 99.19%
Egypt[300]	School-aged (6-12 years)	Snellen, Tumbling E	-	6/12	Sensitivity: 92.80%
India[219,220]	School –aged (4-16 years)	Tumbling E	3m or 6m	6/12 or 6/9	**Teachers:** Sensitivity: 47.25%, 46.22% Specificity: 95.65%[219] **Community Eye-Health Workers:** Sensitivity: 83%, Specificity: 99.8% **Teachers:** Sensitivity: 72.3% Specificity: >99% [220]
Iran[217,224]	Preschool and school-aged (2-6 years)	Tumbling E	6m	6/12 or 6/9	Sensitivity: 74.5%, Specificity: 97.2% [217]
Italy[301]	Infants (7 months) and pre-school (3-5 years)	Lea Symbols	3m	6/9 (3 years old) 6/7.5 (5 years old)	
Israel[222]	School-aged (6-7 years and 13-14 years)	Tumbling E	6m	6/12	Agreement of Referral Recommendations: 85.8%
Japan[302]	Preschool (3.5 years)	Landolt C	2.5m	6/12	Sensitivity: ~50% (Nurses tested children who had not been tested at home as well)

Malaysia[303]	School-aged (7,12 and 15 years)	Snellen	6m	6/9	Sensitivity: 50.7%
Netherlands[37]	Infants (1 ,3, 6-9 and 14-24 months) and preschool (36, 45, 60-72 months)	Landolt C (45-72months) /Amsterdam Picture Chart (36 months)	5m	6/7.5	Sensitivity: 73% Specificity: 83%
New Zealand[304]	Preschool	Parr Chart	4m	6/12	Sensitivity: 89% Specificity: 47%
Oman[305]	School-aged (Year 4, 7 and 10)	Snellen, LogMAR	3m		
South Africa[223]	School-aged (12-18 years)	Snellen, Tumbling E		6/12	
South Korea[306]	Preschool (3-6 years)	Picture chart	3m	6/12 (3 years), 6/9.5 (4 and 5 years)	
Taiwan[221]	School-aged (11-12 years)	Tumbling E	6m	6/9	
United Kingdom[171,214,294,307]	Preschool (4-5 years)	Sonnksen chart, Keeler crowded LogMAR, Kays Pictures	-	6/9.5 (crowded logMAR), 6/7.5 (uncrowded logMAR), 6/7.5 (Kays pictures), 6/9.5 (Keeler)	Sensitivity: 86% [171] and 86.4% - 6/12 cut-off [294] Sensitivity: 70.4%[214] Specificity: 84.4%
United States[308-315]	Infants, preschool (3-5 years) and school-aged	Lea Symbols, Snellen, Kindergarten eye charts, ClearChart 2 digital acuity system, Allens figures	3m	6/12 (preschool), 6/9 (school age)	PPV: 64.5% [309]

1.9.1 Limitations of Screening using Visual Acuity Tests

Whilst visual acuity has demonstrated as being successful in the detection of amblyopia and myopia, the SMS showed that it has less sensitivity for detecting hyperopia and astigmatism[39] as well as abnormalities of binocular function such as strabismus without amblyopia. Myopia was found to be detected reliably (sensitivity 97.8%) at a 6/9.5 cut-off for children aged 12 year old children in the SMS and similarly in the Collaborative Longitudinal Evaluation of Ethnicity and Refractive Error (CLEERE) study for children aged 6-14 years at a sensitivity of 88%.[39,316] However, there was no reliable distance visual acuity cut-off for clinically significant hyperopia in either study, with the recommendation that cycloplegic autorefraction is required for an accurate diagnosis whenever hyperopia is suspected. Astigmatism (in the absence of myopia) was not reliably detected in the SMS by visual acuity, however, was reported to be successfully detected in the CLEERE study at a sensitivity of 97% but a lower specificity of 70% at the 6/9.5 cut-off. In agreement with SMS the Northern Ireland Childhood Errors of Refraction (NICER) study on school children also reported that while myopia had 92% sensitivity (12-13 year olds) for being detected at the 6/9.5 cut-off, hyperopia and astigmatism were not reliably detected on visual acuity testing.[317] Therefore, it must be noted that screening methods require improvement and it must be considered whether additional or alternative testing could be beneficial in improving the accuracy in detecting these childhood ocular conditions. It should also be noted that, because visual acuity is a subjective test, its use is limited to cooperative children with sufficient cognitive abilities.

1.9.2 Can tests additional to visual acuity measures be the solution?

The detection of refractive error and/or amblyopia in the majority of vision screening protocols relies on a reduction in visual acuity in one or both eyes in the absence of optical correction. In order to strengthen the detection of refractive errors, some groups have explored combining visual measure with photo-screening devices [243,261], or handheld-autorefractors.[243] However, as discussed the sensitivity of these devices for detecting particularly hyperopia is limited. There has been some suggestion that testing near visual acuity could provide additional evidence of hyperopia, with the clearer vision at distance

than at near. A study of near and distance visual acuity in a group of six to 12 year olds [318] found that the combination of the two tests was more accurate for detecting significant refractive error than each test alone, however as this study was conducted in China, the tumbling E chart was used which, as previously discussed, has some issues regarding reliability. Use of the combination of near and distance visual acuity to assist determination of refractive errors may not be applicable in younger children with larger reserves of accommodation [319] and therefore more research is needed in this area.

Another condition which may be missed by vision screening alone is strabismus. Whilst amblyopia is successfully detected with visual acuity testing,[39] strabismus without the presence of amblyopia such as intermittent strabismus, alternating strabismus and microtropia, may not be detected upon visual acuity testing. It must be noted that children with a large angle strabismus can be readily observed by family and friends of the child prior to screening, and therefore, have received a diagnosis from an eye health professional prior to the period of screening. However, small angle strabismus such as a microtropia and intermittent strabismus may not be as obvious and additional testing such as a cover test and/or stereopsis may need be considered in order to ensure these conditions are successfully detected. However, cover testing requires trained specialists such as orthoptists to be able to perform the skill. Stereopsis whilst easy to administer, has been found to have varying levels of accuracies over a number of studies.[123,243,253-257] Further investigation is required to determine the improvement in accuracy for detection of childhood ocular conditions by including these tests and to also determine their practicality.

1.10 International Models for Vision Screening

There are numerous childhood vision screening programs worldwide that have great diversity in protocols even within countries. Variations include testing procedures such as the age at which screening is conducted, the method of recruitment to the program, the visual acuity test used (Table 1.6 Validity for detection childhood ocular conditions using common Pediatric VA tests, screened by different personnel) and the testing distance, the qualifications and training of screeners, the referral criteria, and diagnostic pathways. In the UK, screening at age four to five has been recommended, however, the conduct of the vision screening programs have variability in terms of test procedures, type of screener and referral pathways.[34,294] While the US has set recommendations from the United States Prevenatative Services Task Force (USPSTF) for screening to occur at least once between the ages of three to five, with the aim of detecting amblyopia and its risk factors, variation continues to exist between states and communities using different test procedures and protocols.[34,320,321] Canada also presents with variation in its current practices in vision screening.[298] Of the European Union countries, 35 had vision screening programs, with seven of them regionally based and even for those countries with national screening programs, there is variation in their testing protocols.[322] Whilst the age of vision screening differed, the majority of programs had vision screening before age five and reported a high participation rate and population coverage by their program. Therefore, there are currently numerous vision screening programs throughout the world and within countries, with no single universal vision screening program to date.

1.10.1 Type of Vision Screener

There are different types of screeners or personnel utilised in vision screening programs throughout the world (Table 1.6). The screeners utilised include orthoptists, health professionals without formal training in visual assessment including; nurses, health visitors, general practitioners or medical officers or lay screeners with no health professional training are employed to conduct the vision screening. Orthoptic screening seems to be most accurate,[25,294] with numerous studies concluding that orthoptists as screeners have led to more effective vision screening programs than those delivered by those of non-ocular

background screeners.[21,323] High accuracy of screening in terms of sensitivity and specificity have been noted in screening programs delivered by orthoptists [171-173,291,324,325] and there was high agreement between orthoptists and paediatric ophthalmologists in diagnoses obtained from comprehensive paediatric eye examination; particularly for diagnosis of amblyopia, strabismus and retinal conditions.[326]

However, despite the evidence present for the accuracy and effectivity of vision screening delivered by orthoptists, it remains common in numerous countries for lay screeners or nurses to perform vision screening. This is due manpower and the higher cost of using orthoptists as screeners and in some countries a lack of available orthoptists to perform screening. Recommendations in the UK suggest that vision screening programs should be orthoptic-led, with orthoptists taking primary responsibility for training of screening staff and administration of the program to improve accuracy of screening performed by non-eye care professionals such as nurses or lay screeners.[35] The NSW StEPS program similarly has enrolled/registered nurses or lay screeners who complete a training module created by an orthoptist, followed by training by orthoptists and more experienced screeners to perform the actual vision screening.[207] Additionally, orthoptists provide secondary screening for children who are unable to be tested at preschools or who are high priority referrals (children with visual acuity ≤6/18). This enables some of the children who are provided high priority referrals to be reclassified as routine referrals, <6/9^{-2} (+1.5%) suggesting that the orthoptic secondary screening has greater accuracy.

Table 1.6 Validity for detection childhood ocular conditions using common Pediatric VA tests, screened by different personnel

Author	Study Type	Sample	Personnel		Accuracy		Type of VA test
					Sensitivity	Specificity	
Vision in Preschoolers (2005)[327]	Observational Study	3-5 years, n=1452	Nurse Screeners		Linear Lea Symbols 0.49	Similar sensitivities achieved by nurses and lay screeners when the specificity is set at 0.90	Linear Lea Symbols and Single Lea Symbols
			Lay Screeners		0.37	Single Lea Symbols 0.61		
Bolger et al. (1991)[328]	Cohort Study	3.5 years n=374	Orthoptists		0.54		Sheridan Gardiner
			Clinical Medical officer		0.23		
Garretty et al. (2017)[294]	Prospective study	4-5 years, n=7807	Orthoptist led, delivered by health care support workers		0.864		Keeler Crowded LogMAR test
Jarvis et al. (1991)[325]	Cross-sectional study	5-35 months, n=7000	Younger Cohort	Orthoptist 5 month screen	0.25	0.997	
				9 month health visitor check	0.17	0.997	
			Older Cohort	Orthoptist 35 month screen	1.00	0.983	Sheridan Gardiner letter matching or Kay picture tests
				30 month health visitor check	0.43	1.00	
Sabri et al. (2019)[329]	Prospective Observational Study	4-14 years, n-690	Non-eye care Trainee Screeners (final year undergraduate students)		0.80	0.75	Snellen crowded letters or Lea symbols; near VA with Rosenbaum chart or Lea symbols; Ischihara and Randot stereoacuity
Robinson et al. (1999)[330]	Longitudinal Study	3-5 years, n=3434	Public Health Nurses		Range: 0.604-0.709 (of the overall vision screening including visual acuity, Hirschberg and stereoacuity)	Range: 0.699-0.797 (of the overall vision screening including visual acuity, Hirschberg and	Cambridge crowding cards – single letters at 3 metres

					Average: of the 3 years: 0.64	stereoacuity) *Average*: of the 3 years: 0.75	
Kaur et al. (2016)[219]	Observational Study	5-16 years, n=30205	Teachers	Phase 1	0.47		6/9 Tumbling 'E'optotypes
				Phase 2	0.46	0.96	
Spowart et al. (1998)[331]	Observational Study	5 years, n=776	Nurses		0.83	0.95	Single optotype test, Glasgow acuity cards (3 metre linear)
Teerawattanon et al. (2014)[332]	Cross-sectional descriptive and analytical study	4-12 years, n=5885	Pre-primary school teachers		0.25	0.98	Lea symbols distance visual acuity chart (4-6 years old)
			Primary school teachers		0.59	0.98	E-chart (7 years old), Snellen chart (8-12 years old)
Toufeeq and Oram (2014)[171]	Observational Study	4-5 years, n=3721	Orthoptist-led vision screening		0.86		Sonsken Linear Crowded and single logarithm of the minimum angle of resolution
Sharma (2008)[218]	Observational Study	Year 1 and 2 (middle school), n=1892	Teachers	Detecting uncorrected presenting visual acuity ≤6/12	0.94	0.91	Non-illuminated Tumbling E charts at 6m
				Presenting visual acuity	0.85	0.85	
Wormald (1991)[324]	Retrospective study	Mean age 1980 cohort=4.3 years n=298, Mean age 1982 cohort=4.4 years n=598	Community Orthoptists		0.90	0.99	Snellen chart at 6m (Sheridan Gardiner or Kay Pictures when cooperation is poor)
Shukla et al. (2018)[216]	Cross-sectional study	n=6056	Teachers		0.923	0.726	6/12 Snellen Tumbling E optotype
Marmamula et al. (2018)[220]	Part of a large epidemiological study	4-15 years, n-6197	Community Eye-Health Workers (CEHW)		0.83	0.998	6/12 Tumbling E optotypes
			Teachers		0.723	>0.99	

1.10.2 Models of Referral Pathways post vision screening

Worldwide, vision screening programs also differ in their referral pathways and there is sometimes variation and a lack of clarity. Majority of vision screening programs from the US have referrals to either an optometrist or ophthalmologist with responsibility entirely up to the parent/guardian to ensure the follow-up appointment is made and attended[333-338] Furthermore, vision screening is frequently performed in paediatrician's offices during their annual general check-up and so referral decisions are also often influenced by the paediatrician. In schools where there are school nurses, it is up to the nurse to ensure that there are protocols or procedures in place for children to be followed up with an eye health professional.[339] In a pilot program, implementing vision screening in a few sites, using a protocol that followed the USA national guidelines,[32,340,341] children who failed the vision screening were provided referrals and parents advised to organize a follow-up comprehensive examination with an optometrist or ophthalmologist.[187] It was found that 56% of the referred children did not attend their follow-up appointment or their outcomes from examination were not communicated to the screening program.

The UK also has great variation in their vision screening models, differing with location, as well as structurally different referral pathways. There has been consideration regarding creating standardised models of screening in conjunction with clear referral pathways to ensure children who fail vision screening receive the appropriate treatment.[294,342,343] It has been suggested that a community-based model could provide secondary screening and provision of spectacles in order to reduce over-referral to hospital services[343] and this has been shown to be effective, with that the community service filtered unnecessary referrals to the Hospital Eye Service.[342] The community service model had an additional advantage in reducing the time between screening and a follow-up appointment being made and attended, therefore facilitating more timely treatment. Other models use orthoptists as primary screeners and then refer to either general optometric services or to the Hospital Eye Service if warranted.[294] In this model, those referred to the optometric services had visual acuity between 6/9.5-1 and 6/12+1 while those with poorer vision were referred to Community Children's Eye Service and onwards referral to Hospital Eye Services only

occurred if, in the option of the orthoptist/optometrist, it was needed, reducing the level of false positive referrals to the Hospital Eye Service.

Overall, there is limited literature determining the relative success of different models of referral pathways. In the USA, referrals are mostly to private optometrists and ophthalmologists with less than half of the children being successfully followed-up and reported as having had care.[187] There may be numerous barriers to follow-up care including access, finances and parental/guardian time, where a referral pathway to private clinics would be less successful in populations with poor health care access, lack of health insurance and socioeconomic disadvantage. The community- based screening clinics (optometric and orthoptic assessments) have the advantage of reduced loss to follow-up care, providing cost-effective care to children without generally overloading hospital services.[294,343]

In the StEPS program, high priority referrals (visual acuity ≤6/18) are referred to hospital-based Pediatric, Ophthalmic Outpatients Clinic (POOCs).[207] Children who failed screening at the 6/9^{-2} cut-off, have a similar referral pathway to the USA in that they would need to attend follow-up care at a private clinic, either optometric or ophthalmic and the success of this referral pathway depends on the parent to make sure the appointment is booked and attended. In such models of referral there is a need to look at potential barriers to follow-up after referral for such children and to recommend evidence-based strategies to ensure referral pathways are optimal, ensuring children's ocular conditions are addressed adequately and in a timely manner.

1.10.3 Barriers to parental action from vision screening

Failure to act on referrals from vision screening programs is a well-known problem and is an established limitation for the success of vision screening programs.[333] The recent StEPS evaluation found that overall 10% of NSW parents/guardians did not act on referrals for children who failed StEPS screening (children with visual acuity <6/9-2 in at least one eye).[207] Nearly 11% of high priority referrals (children who had visual acuity ≤6/18 in at least one eye) were not acted on in rural and regional areas and 4.9% were not acted on in metropolitan local health districts. It is currently not known why parents in rural and regional areas were less likely to act on referral, however it is hypothesised that this could be related to a lack of paediatric services in such areas or a need to travel far, particularly to paediatric ophthalmic outpatient's clinics (POOCs).

There have only been a handful of previous studies looking into barriers to parental action on referrals from vision screening. The majority of studies conducted in this area performed telephone surveys, with one other unsuccessfully utilising mailed questionnaires[335] and another utilising focus groups.[338] Whilst studies in this area have sampled a broad range of ethnic and socioeconomic groups, most have been focused on populations within the USA.[334-338,344] The USA does not have universal access to public healthcare, which potentially is a significant financial barrier that may not be relevant to other populations such as Australia, where generally healthcare can be accessed for free.[345] Therefore, specific barriers to action on referral may be location/jurisdiction specific, signifying the importance of identifying barriers within existing provision of health services and implementing customised action plans to overcome such barriers.

Studies with diverse population samples have found the overall important barriers to care to be: communication of test results, logistical challenges and parental knowledge of ocular conditions and the importance of timely care.[186,333,334,336-338,346] One study conducted in USA found that the addition of an appointed coordinator to arrange care was able to overcome certain logistic challenges and improved the percentage of action on referrals from 34% to 66%.[186] Financial barriers were not an identified issue in this particular study since all children are covered by 'Medicaid' in the state of Tennessee, USA where this study was

conducted. Similarly another study in Michigan, USA, found that a predominant issue was doubting the accuracy of vision screening which again, may be due to lack of understanding of ocular conditions, whilst other barriers such as finances and ethnicity were not significant in this study.[336]

A study in Philadelphia, USA that had a much lower proportion of parental action on referral, also found a major issue to be a parental lack of understanding of childhood ocular conditions.[344] This location, however, differed to the other USA locations, facing additional major barriers including financial and language that may have exacerbated lack of knowledge of ocular conditions and understanding of the importance of timely management. Findings from studies that focused on vulnerable populations with known socioeconomic hardship and in immigrants with cultural, language and financial barriers, cannot be generalised to other locations, as the extent and types of barriers within these populations could be unique.[335,344,347] Overall, key barriers that were identified in the previous literature include (Table 1.7):

1) Lack of parental awareness on understanding of childhood ocular conditions
2) Lack of perceived urgency
3) Parent's lack of time
4) Lack of available transportation
5) Lack of medical coverage
6) Socio-economic hardship
7) Cultural attitudes and language difficulty
8) Logistic challenges in scheduling appointments
9) Parents not being informed about the screening results

The applicability and significance of these barriers will depending on the design of the screening program, configuration of the local health care system and the attitude and knowledge of parents. Table 1.7 shows the common barriers to parental action on referral from vision screening identified in previous literature. Studies have estimated the percentages of children not attending follow-up care to be as low as 24%[336] and in other

programs as high as over half of the parents failing to act on recommendations to attend further assessment and treatment.[334,344] With such wide variation in rates of following the recommendations from the results of vision screening, it is important that the barriers to seeking further eye health care are sought for individual screening programs and the populations covered by such programs.

Limitations in Barriers Research

A limitation of previous research in this area has been that the parents may not be able to be contacted to identify any barriers they may face in scheduling and attending appointments following referral.[333,335,337] This limitation can be due to several reasons; including families moving to different cities or countries, not wanting to answer calls from unknown numbers or due to the researchers calling at inconvenient times, such as during work hours. Another barrier to research is families may have low motivation to participate in research or follow-up. Further, parents may feel embarrassed about their reasons for non-attendance and therefore avoid participation in follow-up activities.

A further limitation impacting all previous studies in this area, stems from the small sample size of respondents.[337] Mailed questionnaires, in addition to phone calls, have been trialled to improve response rates, but have been unsuccessful with one study reporting that no participants responded via this method.[335] A useful strategy for future research may be to contact the parents via email or text when calls are missed and to state the purpose of the call while asking for a suitable time to make contact. This may increase the possibility of successfully contacting families. Given, the limitations in generalisability and sample size in this area, further research is required to determine barriers according to specific locations and screening programs so that these may be addressed with follow-up procedures and strategies to assist families to attend follow-up care.

Table 1.7 Common Barriers to Acting on Referral from Vision Screening

Author (Year)	Country/City	Percentage of children with unscheduled appointments	Barriers						
			Parental unawareness of screening result	*Logistics: difficulty scheduling appointments, assuming they will be contacted with regards to scheduling appointments parental lack of time*	*Parental lack of understanding on childhood ocular conditions or need to act on referral*	*Accessibility /transport*	*Financial (e.g. lack of medical coverage)*	*Socio-economic hardship*	*Culture and Language*
Yawn et al. (1998)[338]	America/ Rochester	-	✓	✓	✓	N/A	✓	✓	✓
Kemper et al. (2004)[336]	America/ North Carolina	24%	✓	✓	✓	✓	x	x	x
Kemper et al. (2004)[337]	America/ North Carolina	24.4%	✓	✓	✓	✓	✓	N/A	✓
Tjiam et al. (2011)[346]	Netherlands/ Rotterdam	23.0%	✓	✓	✓	N/A	N/A	✓	✓
Wang et al. (2011)[347]	Canada	-	N/A	✓	N/A	✓	✓	✓	✓
PCCY (2008)[344]	America/ Philadelphia	63%	✓	N/A	✓	✓	✓	N/A	✓
Su et al. (2013)[334]	America	53.4%	✓	✓	✓	N/A	N/A	N/A	N/A
Williams et al. (2013)[335]	America/ Philadelphia	71%	✓	✓	N/A	✓	✓	✓	✓
Slingsby et al. (2017)[333]	America/ Western South Dakota	39.7%	✓	✓	✓	✓	✓	N/A	x

1.11 Benefits and Harms of Childhood Vision Screening Programs

Childhood vision screening aims to detect ocular conditions, primarily amblyopia and significant refractive error, with referral for comprehensive ocular assessment and treatment if required. However, the review by Snowdon and Stewart-Brown in 1997 had questioned the benefits of preschool vision screening [21]. The recommendation to not implement any new preschool vision screening programs and to consider discontinuation of existing programs was primarily based on a lack of evidence to support aspects of the programs and their benefit. However, it was noted that in some instances, such evidence would be impossible to provide, such as a comparative study of the benefits of amblyopia treatment and use of spectacles for refractive error with control groups who would receive no (placebo) treatment. Such a study design could be deemed unethical, as leaving children untreated can lead to lifelong visual and lifestyle consequences.[26]

The coverage of children in preschool vision screening has also been raised as a concern, as there is a high participation rate in school-based programs[171,219,348] compared to those that targeted pre-school aged children.[207,217,224,301,349,350] School-based programs had an apparent advantage of centralising the children into one place, providing minimal inconvenience to parents as the child was already at school and school age children being generally more cooperative with vision testing (Table 1.8). The study conducted in India on school screening had the highest participation rate at 99.7% which may be due to having a larger age range of children up to 16 years of age, with older children being more cooperative to screening. However, they also did not specify obtaining consent from parents which may have meant that they were able to access all available children.[219] Griffith and colleagues had a comparatively lower participation rate of 55.2% over a six year period at school screening and this was despite multiple visits to the school for screening and attempts to encourage parents to consent for screening.[308] This may be due to a high proportion of people in Cleveland, USA being of a low socioeconomic status and generally unable to act on referrals with a lack understanding the importance of eye health.[351]

Despite the advantage of universal access to children at schools, it should be noted that preschool screening programs have a reasonably high participation rate.[207,250,301,304] Some of

the reasons for lower proportions of children being screened can include the method used, for example a screening program in China utilised a home-based screening[349] with a participation rate of 69.9%. While this may be convenient in terms of having no requirement to travel anywhere for screening, the accuracy and confidence in the result may be questionable.

Additionally in some programs, screening was not conducted at preschools but rather required families to travel to a health care centres, which reduces the participation rate.[301,352] Of those who conducted screening at the preschool itself, most results were based on relatively smaller sample sizes that may not be representative of whole population participation rates in preschools.[172,349,350,353] An exception is the StEPS program in New South Wales, Australia, who have reported participation rates from a large sample of 719,686 children who had been offered screening making the participation rate of 75.6% a more reliable indication of the rates that could be achieved.[207]

While a study in the UK[353] reported that the participation rate was greater at five years (school-age) compared to 3-3.5 years (75%) (preschool-age), the researchers stated that since the study had taken place, the uptake of screening at preschool age was over 90%. It was further stated that a high proportion of referred children at preschool required orthoptic treatment and that half the children who had amblyopia at age 5, had not been screened at preschool.

It has been suggested however, that the psychological implications of treatment such as spectacles for refractive error and patching for amblyopia and any instances of unnecessary treatment may pose a risk of harm.[27] It has been reported that while amblyopia treatment may be associated with some level of distress, it had no impact on the child's well-being or behaviour during or after the treatment period [28] and that on average amblyopia treatment was well tolerated by the child and their family.[29] There is a concern for bullying related to patching treatment for amblyopia particularly at schools, however this treatment can be performed at home (avoiding patching at school) and preschool screening would mean the condition can be detected and addressed earlier, avoiding such psychosocial concerns by acknowledging visual concerns prior to school entry.[184,354]

A study conducted on the compliance of patching stated that there was a level of distress involved for both the parents and child, particularly at the early stages for treatment and it was suggested that interventions on compliance with patching should address difficulties faced by parents.[355] Psychosocial elements involved with patching treatment included self-efficacy, belief in treatment, presumed ideas on patching and social stigma, for example, the potential that there was an injury related to patching or that it was a harsh form of treatment. Such attitudes can influence the success of treatment and reduce levels of compliance.[356] While these elements do exist, it is important to note that perceptions of the parent and child are modifiable factors that can be addressed through the health professional's understanding of the family's psychology related to treatment. Therefore, while the likelihood of these harms are reported to be low, a balance between managing amblyopia and ensuring psychosocial well-being should be considered and included within treatment guidelines.[30]

Despite the potential negative impacts of treatment, it is to be noted that if amblyopia is untreated and persists into adulthood, there is a high risk of vision impairment later in life due to injury or disease to the non-amblyopic eye.[26] This increased risk can be attributed to the higher prevalence of ocular disorders that occur with age including cataract and age-related macula degeneration. At least 185 people in the UK with unilateral amblyopia have vision loss at a level that is detrimental to their quality of life.[26] A further study found that those with amblyopia have been found to have three times the risk of visual impairment compared with those without amblyopia.[357] Untreated amblyopia may also harm school performance and even adult self-image.[354,358] Therefore, the absence of childhood vision screening could mean psychological implications of living with amblyopia, as well as being at an increased risk of vision impairment.

Uncorrected refractive error may impact upon educational progression and even daily living.[40,358] Anisometropia is additionally an amblyogenic risk factor.[94] Whilst large angle strabismus should be easily observable and therefore will receive the medical attention required without the need for screening, small angle strabismus is not as easy to observe.[358] Although small angle strabismus is cosmetically acceptable, strabismus is an amblyogenic risk factor and at the time of screening, amblyopia may or may not have developed.[94] If

strabismic amblyopia develops, appropriate treatment regimens need to be put in place to ensure vision is restored. Other ocular motility issues including Brown's and Duane's syndrome and ocular pathologies such as cataract, retinoblastoma, optic nerve glioma and optic nerve hypoplasia may also be detected through childhood vision screening and are conditions that would require further assessment and treatment.[358]

Therefore, the benefits of childhood vision screening, in particular at preschool age, would ensure ocular conditions can be detected and that appropriate timely treatment can be put into place prior to school entry which would promote good prognosis (superior visual outcomes).[184] It would also mean that children would have adjusted with their treatment plan prior to entering the educational world, enabling keen learners to be encouraged.

Table 1.8 Childhood Vision Screening Programs' rate of participation, dependent on age and location of screening

Country	Screening Population and number offered screening	Target Age Division	Percentage Screened	Location of Screening
Australia[207]	Preschools and childcare facilities in New South Wales (n=719 686)	Preschool (4 years)	75.6% (n=564 825)	Preschool
Iran[217]	In all provinces of Iran (n=2 166 851)	Preschool and Kindergarten (3-6 years)	67% (n=1.4 million)	School (Kindergarten), Visual Assessment Centre
Iran[224]	Ardabil Province (n=75 137)	2-6 years	51.7% (n=38 844)	School (Kindergarten), Visual Assessment Centre
Croatia[250]	City of Zagreb County (n=16 896)	Preschool (4-4.5 years)	92.61% (n=15 648)	Kindergarten class
UK[172]	Walsall (n=3623)	Preschool (3-4 years)	78% (n=2830)	Preschool
UK[352]	Cambridge (n=8566)	Preschool (3.5 years)	79.3% (n=6794)	Local Health Centre, Designed Mobile Van
New Zealand[304]	South Auckland (n=5572)	Preschool (3-6 years)	88.22% (n=4916)	Preschool
United States[350]	Six preschools (n=283)	Preschool (3-5 years)	64% (n=181)	Preschool
China[349]	Guangzhou, 10 Kindergartens (n=3300)	Preschool (3-6 years)	69.9% (n=2308)	Home-based screening

Italy[301]	Bolzano District (n=7772)	Infants (7 months), Preschool (3 and 5 years)	78.82% (n=6126)	Health Care Centre
UK (North-east England)[348]	Geographical area covered by the Newcastle upon Tyne NHS Hospitals Trust for two primary vision screening programs at preschool (n=4567) and school (n=6082)	Preschool (3.5 years) School (4-5 years)	Preschool: 60% (n=2742) School: 96% (n=5824)	Preschool group: Local Clinic/ Nursery schools School group: School
UK (North-west England)[353]	Two samples of children in Warrington aged 3-3.5 (n=2736) and 5 years (n=2582)	Preschool (3-3.5 years) School (5 years)	3 years: 75% (n=2041) 5 years: 94% (n=2432)	Preschool School
United States[308]	Cleveland Public Schools	Pre-kindergarten, Kindergarten, First Grade (mean age: 6 years)	55.2% (over a 6 year period) n=63 841 (over a 12 year period)	Mobile Screening Van at Schools
India[219]	166 schools (government and semi-government) in the Ludhiana district (n=30 298)	School (4-16 years)	99.7% (n=30 205)	School
UK[171]	155 state and three private schools (n=4013)	School (4-5 years)	92.85% (n=3726)	School

1.12 Justification and Aims

Worldwide, vision screening programs vary significantly. Thus, there is no standard protocol used for vision screening in preschool aged children. This study aims to investigate some of the current gaps in the literature, with a focus on the protocols used in the Statewide Eyesight Preschooler Screening Program (StEPS).

1. To determine the prevalence of reduced vision in children and it's most common causes.

2. To evaluate the impact of refraction methods, iris colour and cycloplegia on refraction measurements.

3. To determine the natural history of children with hyperopia and if hyperopia should be corrected.

4. To determine the comparability of school vs. preschool screening.

5. To determine the comparability of two vision charts used in the StEPS program, Sheridan Gardiner Linear and HOTV LogMAR.

6. To determine whether visual acuity testing alone or vision screening with additional orthoptic testing would yield the most accurate referrals.

7. To determine the barriers to successful follow-up after referral

Chapter 2
Methods

2.1 Overview

The data and results presented in this thesis are derived from a suite of epidemiological studies. Chapters 3, 4, 5 and 6 present results from three large population-based studies collectively known as the Sydney Childhood Eye Disease Studies. The data from these studies was collected over an extended period of time, from 2003 to 2011. While I did not participate in the overall design of these studies or in data collection, the databases from these studies have been analysed to answer specific research questions which I posed related to childhood visual development and testing of ocular status. These questions directly impact the fundamental understanding of aspects of childhood vision screening. Further to these studies was the Preschool Vision Screening Study (PVSS) that was devised and conducted by me as the primary researcher. This study was embedded in the StEPS pre-school screening program and commenced in 2019. The data collection was curtailed in February 2020 due to health restrictions imposed by NSW Health related to the advent of COVID 19 and the StEPS program itself did not run in 2020. The data from this study is described in Chapter 7.

2.2 The Sydney Childhood Eye Disease Studies

The Sydney Childhood Eye Disease Study comprised of three population-based studies; the Sydney Paediatric Eye Disease Study (SPEDS) conducted from 2007-9, Sydney Myopia Study (SMS) conducted in 2003-5 and, its follow-up cohort, the Sydney Adolescent and Vascular Eye Disease Study (SAVES) in 2009 -11. SPEDS examined infants and children between 6 months to 72 months (6 years) of age. SMS sampled children in two school grades Year 1 and Year 7; with a mean age of 6 and 12 years at baseline, respectively and followed up these children and adolescents 5-6 years later in SAVES then aged 12 and 17 years. All children underwent an age appropriate comprehensive ocular examination which included visual acuity, stereoacuity, cover test, convergence, ocular motility and cycloplegic refraction (cyclopentolate 0.5% or 1%). Questionnaires were completed by parents to obtain demographic and health information and by the older children in SAVES.

2.2.1 The Sydney Paediatric Eye Disease Study (SPEDS)

The impetus for a paediatric study in Sydney came from a need to research more closely the natural history of development of vision and refraction in infants and pre-school aged children. The maximum age of 6 years was determined by the average age of the younger children examined earlier in the SMS study. At the same time the researchers were approached by the principal researcher from paediatric studies in the USA and it was agreed that SPEDS would follow a similar protocol to the USA studies,[211,359] one located in Los Angeles, known as MEPEDS and the other in Baltimore, known as BPEDS. The SPEDS examination protocol was essentially the same as in these studies, except for minor adjustments to the questionnaire to make it appropriate for an Australian population. There was also an adjustment to the re-testing of children with poor visual acuity, the USA studies always tested the right eye first, but in SPEDS when re-testing visual acuity, the worse eye was always tested first, which negated the finding from the USA studies, that overall left visual acuity was poorer than that of the right eye. These three studies were followed by a similar study conducted in Singapore, known as STARS.[360]

The protocol for SPEDS has been previously published[38] and the study received ethical approval from the University of Sydney Human Research Ethics Committee, and adhered to the tenets of the Declaration of Helsinki. SPEDS recruited a representative population-based sample for metropolitan Sydney using a stratified random cluster sampling strategy. The metropolitan area was stratified by socioeconomic status (SES), according to the Australian Bureau of Statistics (ABS) 2006 census data into low, middle and high SES, and suburbs within strata were grouped into three ABS geographical areas; inner city, middle suburban and outer suburban regions. Suburbs with moderate proportions of preschool-aged children were then randomly selected from these regions with a selection of four suburbs, one outer suburban, one inner city and two middle suburban suburbs. The inclusion of these four suburbs was considered to be sufficient for a representative sample of preschool-aged children residing in Sydney.

Information sheets about the study were delivered to each household identified using the 2006 ABS census map within the selected suburbs. Flyers were also placed at local health

care centres, preschools and day care centres. Recruitment staff then door-knocked to identify households that included children between 6 months to 6 years of age for inclusion in the study and invited them to participate in the study.

Households with eligible children were provided a package which included information about the study and two questionnaires as well as consent forms. Consenting families then had appointment times arranged at a study clinic located at either Quakers Hill (outer suburban) or Campsie (on the border of middle and inner city regions). The two questionnaires to be completed by parents included a total of 176 questions to obtain information on ethnicity, parental education and employment, the child's birth and medical history, as well as ocular information for the child and their family members.

SPEDS enumerated 3333 eligible children aged 6 months to 6 years and examined a total of 2461 children (73.8% participation rate). Ocular examinations were conducted by medical officers and orthoptists between 2007 and 2009. These included age appropriate assessments for visual acuity, ocular alignment and ocular pathology.

Visual Acuity was examined using, Teller Acuity Cards II (Stereo Optical Co. Inc., Chicago, IL) for children aged less than 24 months, the Electronic Visual Acuity (EVA) HOTV system at 3m (Jaeb Centre for Health Research, Tampa, Florida) with matching and retro-illuminated logMAR chart with EDTRS or HOTV optotypes at 2.44m (Vector Vision CSV-1000, Vector Vision, Inc., Dayton, OH) for children aged 30 months and older. Matching cards for HOTV were provided whenever needed. Lea Symbols were trialled for a short period but were discontinued as they were no more testable at younger ages than the EVA HOTV test (personal correspondence). Visual acuity was assessed monocularly by patching each eye and then using a staircase method until threshold visual acuity could be determined. Children with optical correction had their visual acuity tested with and without their correction.

Ocular alignment was examined using a cover test and prism bar cover test to detect strabismus or heterophoria at near and distance, with and without glasses. Ocular motility and convergence near point (RAF rule in older children) was also performed to assess

versions, ductions and vergence. Binocular single vision was assessed with Lang II stereo card (Lang stereotest, Forch, Switzerland) and Randot preschool stereoacuity test (Stereo Optical Company, Chicago, IL) was used for children aged ≥30 months and Stereo Smile Test II (Stereo Optical Inc., Chicago, IL) for children aged <30 months.

Iris colour was graded through the appearance of the undilated eye, against reference photographs that were classified standard as blue, green-hazel, tan-brown and dark-brown.

Figure 2.1 Iris Colour Reference Photographs[361]

Blue

Brown/Tan

Hazel

Dark Brown

Cycloplegic refraction was obtained using either Retinomax K-Plus 2 (Nikon Corporation, Tokyo, Japan), Canon RK-Fl table-mounted autorefraction (Canon, Tokyo, Japan) or streak retinoscopy. Cycloplegia was achieved with parental consent using; Amethocaine 0.5% followed by two cycles, 5 minutes apart of Cyclopentolate 0.5% for children younger than 12 months or Cyclopentolate 1% for children 12 months and older. For children who failed to dilate or with dark irides, an additional drop of Tropicamide 1% and/or Phenylephrine 2.5% was administered. The anterior eye was examined using a Haag-Streit slit-lamp (Koeniz, Switzerland) and fundus exam was by indirect ophthalmoscope. Retinal photographs were taken by a non-telecentric fundus camera (Canon CF-60UVi fundus camera, CF-DA camera

adapter, EOS-lOD digital camera; Canon Inc., USA) and ocular biometry using an IOLMaster (IOLMaster, Carl Zeiss Meditec, Jena, Germany) in children ≥30 months after cycloplegia.

2.2.2 The Sydney Myopia Study (SMS) and Sydney Adolescent Vascular and Eye Study (SAVES)

The SMS was the first of the Sydney Childhood Eye Studies, devised primarily to determine the prevalence of myopic refractive errors in light of the epidemic of myopic occurring in a number of countries and to examine possible risk factors that might assist to explain the rapid rise in its prevalence. It was also the first examination of childhood eye conditions in a representative sample of Australian school children. The methodology of the SMS has been previously published.[123] The Sydney metropolitan area was stratified by socioeconomic status (SES) using the 2001 ABS census data into nine strata. A total of 34 primary schools and 21 secondary schools across Sydney were randomly selected with preferential selection of schools from the highest strata. These include 5 primary schools and 2 high schools in the top SES and a random but proportionate mix of public, religious and private schools. The study was approved by the Human Ethics Committee of the University of Sydney, the Catholic Education Office and by the New South Wales Department of Education. The research adhered to the tenets of the Declaration of Helsinki. Informed written consent was obtained from at least one parent or guardian. Verbal consent was also obtained from each child prior to commencing the examination on the day.

Each school was approached by the lead researcher inviting participation and with the principal of the school's agreement, information sessions were held with teachers, parents and pupils. Information packages including information sheets, consent forms and questionnaires were sent to all eligible children. Questionnaires were completed by parents of participating children including questions regarding sociodemographic information such as ethnicity, parental education and employment; the child's birth and medical history and typical daily activities on weekdays and weekends.

Between 2003 and 2005, SMS examined 4093 children in two samples; Grade 1 children with mean age 6 and Grade 7 with a mean age 12 years. The study team included ophthalmologists, medical practitioners, orthoptists and optometrists. Assessments were conducted to examine visual acuity, ocular alignment, ocular pathology and cycloplegic refraction. Visual acuity was measured monocularly using a retro-illuminated EDTRS logMAR

chart (Vectorvision™ CSV-1000; Vectorvision,Inc., Arcanum, OH) at 2.44m and under controlled illumination, near vision was measured using the logMAR HOTV near vision chart at 33 cm. A matching card is available for children unable to read the letters. Children with optical correction had their visual acuity tested with and without their correction. Cover test, prism bar cover test and ocular movements were conducted by an orthoptist to establish ocular alignment. The presence of binocular single vision was tested using the Lang II stereo card (Lang-stereotest, Forch, Switzerland), TNO test for stereoscopic vision (Laméris Ootech BV Nieuwegian, The Netherlands) and 4 dioptre prism test to detect microtropia. After dilation (amethocaine 1%, cyclopentolate 1% and tropicamide 1% twice, 5 minutes apart and 2.5% phenylephrine if still poorly dilated), ocular biometry was measured using the IOLMaster™ (Carl Zeiss, Meditec AG Jena, Germany), a Haag-Streit slit-lamp (Koeniz, Switzerland) was used to examine the anterior segment of the eye. Refraction was measured on average 25 minutes after the last cycloplegic eye drop was administered using a Canon autorefractor (model RK-F1; Canon, Tokyo, Japan) and Optical Coherence Tomography: Stratus OCT3™ (Model 3000; Zeiss,Meditec Inc., CA, USA) was used for posterior segment examination with fundus photography using a Canon 60◦ Mydriatic Fundus Camera (model CF-60UVi, Canon Inc., Tokyo, Japan).

In the SMS five to six year follow-up study, known as SAVES, the examination procedures and questionnaires were the same as those used in the original study so that direct comparison could be made between the baseline and follow-up data. Of the original 34 primary schools included in SMS, children were still enrolled in 13 of these primary schools and re-examined at those schools. Of the 21 secondary schools included in SMS, 20 still had the same children enrolled at the school and were re-examined. Children unable to be examined at their original school were invited to attend an eye clinic or at one of the study schools if it was close to their home address. Again the same detailed questionnaires were administered to parents for demographic as well as the child's health and ocular health since SMS. Questionnaires were also administered to children to obtain information on daily activities, ocular health and ocular symptoms. A total of 2130 children from the original SMS were included in SAVES during 2009-2011 representing 52% of the SMS cohort and an

attrition rate of 10% per year for the five to six year follow-up. Included in SAVES were a further 941 children enrolled in the schools who participated in the study and consented to be included in SAVES.

<u>Ethics Approval</u>

Ethics approval for all three studies were obtained from the Human Research Ethics Committee of the University of Sydney, and the studies adhered to the tenets of the Declaration of Helsinki. In addition, SMS and SAVES obtained ethics approval from the New South Wales Department of Education and Training, and the Catholic Education Office. Informed written consent was obtained from at least one parent or participants who were over the legal age of consent (18 years) prior to examination. Verbal consent was also obtained from each child prior to commencing the examination on the day.

2.3 The Pre-school Vision Screening Study (PVSS)

The PVSS was conducted within the New South Wales (NSW) Statewide Eyesight Preschooler Screening (StEPS) Program and addressed some keys aspects of the program, in particular in relation to the protocol for vision testing. StEPS is a free universal vision screening program for all four year old children in NSW prior to school entry[184,207,290] and has been operating in the state since 2009. The aim of the StEPS program is to identify childhood ocular conditions early so that treatment outcomes can be maximised and to avoid any impact on learning at school. The screening involves a visual acuity test, performed by the StEPS screeners when they visit the child's preschool/early childhood centre or at a NSW Health Child & Family Health Service or a catch up StEPS clinic (if the child misses out on screening). The program is led by StEPS coordinators in each NSW Local Government Areas (LGA). The screeners themselves are lay or nurse screeners.

Recently the StEPS Program has transitioned from the Sheridan Gardiner Linear Chart to the HOTV logMAR chart. However, these visual acuity charts have not been systematically compared and, despite the change in charts, the StEPS program referral criteria has not been reviewed. In addition, the HOTV logMAR chart used in StEPS has an additional line (6/15) which is not available on the Sheridan Gardiner Linear Chart. StEPS refers children with 6/18 or worse vision as high priority referral which, provides the option for parents to attend a Paediatric Ophthalmic Outpatients Clinic (POOC) within the public hospital[184] (Appendix 2a: StEPS Protocol). However, the addition of the 6/15 line on the HOTV logMAR chart means that there is potential for some children who would have previously received a high priority referral, to now be a general referral, altering their referral pathway substantially. This project was devised in order to determine the comparability of the two charts, the most appropriate referral criteria and the potential impact of the change on referrals.

Furthermore, referring on the basis of visual acuity alone may not detect all ocular conditions that require treatment. Thus, it is important to determine whether a battery of tests would obtain more accurate referrals than vision screening alone. Finally, barriers to successful follow-up of children who are detected as having ocular abnormalities at vision

screening must be looked into in an Australian context to identify and address these issues on a national level. The PVSS was originally designed to address all these aspects in two phases. Unfortunately the second phase addressing barriers to acting on referral by parents and attending eye care practitioners was halted by the restrictions imposed by COVID 19 Health regulations in early 2020.

Recruitment: Of the 15 LGAs in NSW, the Sydney LGA was selected because of its broad cross section of suburbs from inner city to middle suburbia, capturing a range of socioeconomic status groupings (based on education, employment and income) and a wide diversity of languages and different ethnicities.[362] Preschools in the Sydney LGA were approached from April 2019 to February 2020 and invited via phone and/or email to participate in the current study in conjunction with the StEPS Program. Eleven preschools agreed to participate in the StEPS program and PVSS. After verbal consent by the preschool directors for their centres to participate, StEPS (Appendix 2a: StEPS Protocol, Appendix 2b: StEPS Brochure) and PVSS consent forms, information sheets (Appendix 1a: PVSS Participation Information and Consent Form and Appendix 1c: PVSS Preschool Information and Consent Form) and the PVSS demographics questionnaires as a pack (Appendix 1b: PVSS Demographics Questionnaire), were emailed or sent via post to the preschools after confirming the number of eligible children within each preschool. Children who were born before July 31st 2015 were eligible for participation in 2019 and children born before July 31st 2016 were eligible for participation in 2020. These forms were then sent out by the preschools to the parents/guardians of the children who would participate in the study. Consent was obtained separately for the StEPS program and the PVSS. Copies of the information sheets and consent letters are included in Appendix 1a, 2a and 2b.

Questionnaires: were completed by parents/guardians to obtain demographic information including date of birth, gender and ethnicity. Additionally, information on family history of ocular conditions and parental concerns and observations were also obtained. Contact details of parents/guardians were also collected to facilitate follow-up. A copy of the questionnaire is included in Appendix 1b: PVSS Demographics Questionnaire.

The StEPS Vision Testing Protocol: had originally used the Sheridan Gardiner Linear visual acuity chart with a matching letter card at a distance of 3 metres or 6 metres, depending on the available space for screening in the preschool. During 2018, the NSW LGAs commenced transitioning to the HOTV logMAR visual acuity chart, again using an appropriate matching card (Appendix 2a: StEPS Protocol) and testing distance was still adjusted to the available space. The visual acuity referral criteria (Box 2.1) remained unchanged after the introduction of the HOTV logMAR chart. Parents of children were notified and referred through the StEPS program according to their result using the HOTV logMAR chart. A pass was visual acuity better than $6/9^{-2}$ in both eyes, borderline pass was visual acuity equal to $6/9^{-1}$ or $6/9^{-2}$ in either eye, routine referral was visual acuity poorer than $6/9^{-2}$ in either eye and a high priority referral was when visual acuity was equal to or worse than 6/18 in either eye. Children with abnormalities detected on orthoptic testing (see Phase 1 of the PVSS) were also referred through the STEPs screening program for further ocular examination.

Prior to commencing the PVSS, I received training on the StEPS screening protocol for testing visual acuity and shadowed one of the StEPS screeners for a day screening in a preschool, who evaluated my performance according to the StEPS protocols.

Box 2.1 StEPS Referral Criteria

- ≥ 6/9 Pass, No Referral
- $6/9^{-1}$ to $6/9^{-2}$ Borderline Pass, Visual acuity recommended to be re-tested in 12 months
- < $6/9^{-2}$ Routine Referral to an eye health professional
- ≤ 6/18 High Priority Referral to an eye health professional. Your local StEPS coordinator can facilitate rapid referral to a NSW Health Paediatric Ophthalmic Outpatient's Clinic (POOC) within the public hospital.
- Unable to be tested/incomplete screen: Referral required
- Vision within normal limits but requires referral for other finding (e.g. strabismus, ptosis, head tilt, pathology): Referral required

PVSS Phase 1: Children had their vision screened at their respective preschools/childcares using the HOTV logMAR visual acuity chart (Good-lite, US, retro-illuminated 6 metre chart) and the Sheridan Gardiner Linear chart (BOC Instruments, Silverwater, NSW, Aus, retro-illuminated 6 metre chart) with matching cards provided. Each child was tested monocularly, using a patch occluder or occlusion glasses. Visual acuity was tested at distance of 6 metres using the staircase method. This involved commencing at the top line (6/60) of the vision chart, and progressively testing one letter from each line. When the child makes an error or becomes hesitant, the last line where they recognised the letter correctly, is then tested fully. The testing continues until a threshold visual acuity is obtained. This differed slightly from the StEPS protocol (Appendix 2a: StEPS Protocol) where the stair-casing of visual acuity was performed only to the 6/9 visual acuity line, the line on the chart used as the cut-off for referral.

The order of using the HOTV logMAR and Sheridan Gardiner vision tests was alternated between children to avoid confounding variables such as fatigue biasing results obtained. This was followed by an orthoptic examination including, HOTV crowded near visual acuity test (Good-Lite, Elgin, IL, US), cover test (near and distance), ocular movements, convergence near point using the RAF rule (Grafton Optical, UK), 15^BO test, 4^BO test, stereopsis testing including Lang II (Lang stereotest, Forch, Switzerland), TNO (Laméris Ootech BV Nieuwegian, The Netherlands) and the Randot Preschool stereotest (Stereo Optical Company, Chicago, IL), pupil assessment and ophthalmoscopy (red reflex) (Optimed, NSW, Aus).

Phase 2: Parents/guardians with children who were referred from StEPS for further eye care who had taken part in Phase 1 of the study, were contacted one month later via phone or emailed when they were not reachable by phone, to determine whether a follow-up appointment has been made and/or attended. When a follow-up appointment was not made or not attended, reasons were questioned. After the child attended their follow-up appointment and with consent from parents/guardians, eye care practitioners were contacted to obtain the child's diagnoses.

The impact of Covid-19: Whilst the PVSS data collection had planned to be continued through 2020, with the outbreak of covid-19 this was not possible given that NSW Health had posed restrictions on the StEPS program which itself did not run in this year. Whilst ten families had been recruited as part of Phase 2, this part of the project had been halted due to covid-19. Furthermore, additional projects had been proposed related to barriers to follow-up care in the StEPS program, which were also brought to a halt until further notice.

Ethics Approval and Consent: Informed written consent was obtained from all parents/guardians of participating children prior to vision screening. Wherever possible verbal consent was sought from children prior to the ocular examination. Children had the right to refuse to perform any test throughout the examination. The tests used in the examination were non-invasive and unlikely to cause any harm or distress to the children assessed. This study was approved by the Human Research Ethics Committee of the University of Technology Sydney and adhered to the tenets of Declaration of Helsinki.

Chapter 3
Reduced Vision in Childhood

3.1 Abstract

Aim: To determine the proportion of children aged 3-17 years with reduced vision in Sydney, Australia and the eye conditions that are likely to be detected by visual acuity screening. Additionally, to investigate whether there are changes in the prevalence and incidence of reduced vision with age and the necessity for additional screening in older children.

Method: Children aged 3-5 years from the Sydney Paediatric Eye Disease Study (SPEDS), 6 and 12 years from the Sydney Myopia Study (SMS) and its five year follow-up at 12 and 17 years from the Sydney Adolescent and Vascular Eye disease Study (SAVES) were included as part of the cross-sectional analysis. A longitudinal analysis of new cases of reduced vision between baseline (SMS) and follow-up examination (SAVES) included only children assessed at both time points. All children had a comprehensive ocular examination, including presenting (with habitual refractive correction, if present) and uncorrected visual acuity (VA). For the 3-5 year olds, the Electronic Visual Acuity HOTV test was used, while for the 6, 12 and 17 year olds, a retro-illuminated ETDRS logMAR chart was used. Reduced uncorrected VA was defined as <6/12 for either eye and was further classified according to severity as, mild (<6/12), moderate (<6/18) or severe (<6/60). Bilateral reduced vision was defined as reduced vision in both eyes with the classification of severity according to VA in the better-seeing eye, to be reflective of functional vision.

Results: The overall prevalence of reduced vision increased with age with a significant difference between all age groups (p<.001). The lowest proportion of reduced vision was in the 3-5 year old children (4.5%) and the highest in the 17 year olds (17.7%). Amblyopia accounted for 21% and 28% of reduced vision in the two youngest age cohorts (3-5 years and 6 years). There was a substantial decline in the relative proportion of reduced vision attributed to amblyopia as children increased in age, while other causes including refractive error increased, with > 80% of children with reduced vision in the older cohorts (12 and 17 years) being classified as myopic. Approximately 80% of children aged 12 and 17 years who had a refractive error and reduced vision, already had glasses, with the majority having good vision with their prescribed correction. Incident cases of reduced vision at 12 and 17 years

revealed that these were largely caused by myopia with 73% of 17 year olds already wearing glasses but the rate of glasses wear was comparatively lower in 12 year olds (52%).

Conclusion: Amblyopia as a cause of reduced vision was proportionally higher at a younger age and this supports current recommendations for screening to be conducted at preschool age in order to optimise treatment outcomes. Whilst refractive error was a significant cause of reduced vision at all ages, the majority of children with refractive error at older ages had been prescribed refractive correction. Community education and targeted myopia screening at 12 years of age may be more advisable than repeat population vision screening over school years in Sydney, Australia.

3.2 Introduction

Vision impairment is ranked 6th according to global burden of disease.[363] At least 2.2 billion people worldwide have a vision impairment and of these, at least 1 billion have an ocular condition that could have been prevented or treated.[364] Although the majority of those with vision impairment are over the age of 50, childhood vision impairment accounts for 4% of global visual impairment, yet one third of the costs associated with vision impairment and blindness is attributed to children's vision impairment.[365] A recent study found that uncorrected refractive error was one of the leading causes of blindness and moderate to severe vision impairment in children, with the authors recommending interventions at schools to improve effective coverage for those who require spectacles.[366]

A number of childhood ocular conditions, including refractive error and amblyopia are treatable. However, particularly for amblyopia, treatment is time sensitive to the period of neural plasticity and early treatment has been shown to be most effective for reversing visual impairment.[15,16,86,366,367] There is also a potential benefit of timely treatment to minimise the impact of reduced vision on a children's ability to learn, socialise and participate in daily activities, as well as the costs associated with vision impairment continuing into adulthood.[365,366] A focus group study conducted for 6-12 year old children with vision impairment, found a significant impact on quality of life. The most common areas of concern were ability to cope at school included: having access to appropriate print size, assistive devices and experiencing reading difficulties (21%). Also reported was future expectations and frustrations such as: lack of cure for the condition or worsening of the condition overtime (14%) and psychosocial wellbeing (13%).[368] In the circumstance that visual impairment cannot be treated, early detection remains valuable for the provision of low vision aides and support services.[369-372] Coupled with the need for early treatment, this adds a further argument for the importance of vision screening in childhood.

Current recommendations for vision screening indicate that a target age of 3-5 years is most appropriate for treatment of amblyopia and correction of refractive errors prior to school entry.[27,34,35,321] As the prevalence of eye conditions detected by vision screening, including amblyopia, strabismus and refractive error, differ with location, ethnicity, socioeconomic

status among other population specific factors, the referral rate from vision screening programs can vary with the community being screened.[94-97,124,132-134,136,139,146,366,373-377] The aim of this study is to determine the proportion of children with reduced vision in Sydney, Australia and the conditions likely to be detected by vision screening. An additional question related to vision screening is whether repeat screening at older ages may be necessary to detect new cases of reduced vision, particularly with the onset of myopia. As such, a further aim is to investigate the incidence of reduced vision in school-aged children enrolled in a longitudinal cohort and access the necessity for additional screening at older ages.

3.3 Method

Children aged 3-5 years from the Sydney Paediatric Eye Disease Study (SPEDS) during 2007-2009, 6 and 12 years at baseline in the Sydney Myopia Study (SMS) over 2003-2005 and 12 and 17 years, respectively at 5-6 year follow-up (2009-2011) in the Sydney Adolescent and Vascular Eye Disease Study (SAVES), were included in the cross-sectional data analysis. A longitudinal analysis of new cases of reduced vision between baseline (SMS) and follow-up examination (SAVES) included only children assessed at both time points.

All children had a comprehensive ocular examination, including presenting (with habitual refractive correction, if present) and uncorrected visual acuity (VA). For the 3-5 year olds, the Electronic Visual Acuity HOTV test was used, while for the 6, 12 and 17 year olds, a retro-illuminated ETDRS logMAR chart was used. Best corrected visual acuity was additionally performed in the 12 and 17 year old children when presenting VA was reduced. VA achieved was analysed in 'number of letters read correctly' for the analysis. Ocular pathology was diagnosed by slit lamp examination and/or a dilated fundus assessment by indirect ophthalmoscopy in younger children and retinal photography in older children. Refer to 2.2.1 and 2.2.2 for more detailed methods from SPEDS, SMS and SAVES.

Definitions:

Reduced vision (VA <6/12) was defined according to uncorrected VA in either eye. Severity was based on the World Health Organisation [364] definitions of vision impairment; mild (<6/12), moderate (<6/18) or severe (<6/60). Bilateral reduced vision was defined as reduced vision in both eyes with the classification of severity according to VA in the better-seeing eye to be reflective of functional vision. Corrected VA was also determined using a child's current spectacle correction.

Refractive error was defined for each participant according to spherical equivalent refraction (SER) calculated as sphere + 1/2 cylinder in the eye with reduced vision. Myopia defined as ≤-0.50 dioptres (D), hyperopia ≥+2.00 D and astigmatism ≥1.00 D. Unilateral amblyopia was defined as a difference of ≥2 lines between the eyes and bilateral amblyopia as VA <6/12 in both eyes, in the presence of an amblyogenic risk factor such as; manifest

strabismus as detected on cover test at near and/or distance fixation, anisometropia (≥1.00D difference between eyes) or high refractive error in one or both eyes (>+5.00D, <-5.00D) and in the absence of other pathology that may explain reduced vision.

Statistical Analysis: Data was analysed using SPSS (v22 IBM US). Mean VA (number of ETDRS letters) and SER was calculated for each age group cross section, with a t-test used to determine whether there was a significant difference in refraction between age groups. For this analysis the data from the right eye only was used due to a high correlation between right and left eye. The prevalence of reduced vision for each age group was calculated using the WHO definition and chi-square analysis was used to determine if differences in prevalence were statistically significant. Of those children who had reduced vision in each age group, the proportion attributed to different ocular conditions was also calculated and chi square was again used to determine statistical significance of differences in proportions. A participant was classified as having an ocular condition, including refractive error when it was present in either or both eyes. The impact of refractive correction on the prevalence of reduced vision was also determined. The incidence of new cases of reduced vision for children aged 12 and 17 years (SAVES) was based on change in VA from baseline at 6 and 12 years respectively with the attributing ocular condition also determined.

3.4 Results

There were a total of 986 children aged 3-5 years (SPEDS), 1739 children aged 6 (SMS), 3456 children aged 12 (SMS n=2345 and SAVES n=1111) and 1649 children aged 17 (SAVES) who were included in the current analysis.

Normative Visual Acuity and Refraction by Age

Mean uncorrected VA was 6/9.5+2 letters for children aged 3-5 years and 6/7.5 for children aged 6 (Table 3.1). By 12 years in both the SMS and SAVES cohorts, the mean VA had reached close to 6/6. At 17 years the mean VA was relatively lower at 6/7.5-1, but with corrected VA, the mean was 6/6+2.

The mean SER at 3-5 years was +1.29 and remained stable in the 6 year old cohort (+1.27D), with no significant shift in refraction between these two age groups (p=0.5). There was subsequently a myopic shift in mean SER with increasing age, reaching near to +0.50 at 12 years in both the SMS and SAVES cohort, with a further significant reduction in mean SER to +0.03 at 17 years of age (p <.0001). Differences in mean SER between age groups were significant between the 3-5 and 6 years groups and at 12 years (SMS and SAVES) and 17 years (all p <.0001).

Table 3.1 Mean visual acuity and mean refraction through childhood

Study	Age Group	Mean Uncorrected Visual Acuity *No. letters read (Standard Deviation), Snellen Fraction*	Mean Best Corrected Visual Acuity (BCRVA) *No. letters read (Standard Deviation), Snellen Fraction*	Mean Cycloplegic Refraction (SER) *Mean (Standard Deviation)*
SPEDS	3-5 years	47 (6.5), 6/9.5+2	-	+1.29 (1.13)
SMS	6 years	50 (4.7), 6/7.5	-	+1.27 (0.88)
	12 years	54 (12.9), 6/6-1	55 (4.9), 6/6	+0.49 (1.34)
SAVES	12 years	53 (12.0), 6/6-2	54 (5.5), 6/6-1	+0.47 (1.33)
	17 years	51 (15.1), 6/7.5+1	57 (4.0), 6/6+2	+0.03 (1.49)

Note: Data from right eye only

Prevalence of Uncorrected Reduced Visual Acuity in Childhood

The overall prevalence of uncorrected reduced VA increased with age with a significant difference between all age groups (p<.001). The lowest proportion of reduced vision was in the 3-5 year old children (4.5%) and the highest in the 17 year olds (17.7%) (p<.001) (Table 3.2). There was no statistically significant difference in prevalence of reduced VA between the two 12 year age groups (SMS and SAVES) (p=0.3). The severity of reduced vision also seemed to increase with age, with the majority in the younger age cohorts with reduced VA classified as having only mildly reduced vision (3-5 years: 77.3%, 6 years: 78.9%), compared to the older age groups (12 and 17 years) where most reduced vision was classified as moderate (46.7%-69.4%). Additionally, the proportion of those with bilateral reduced vision initially decreased from the 3-5 year age group (52.3%) to 6 years (32.4%) after which, it increased with age, with differences in the prevalence of bilateral reduced vision between all age groups considered statistically significant (all p<.001). This was including a significant difference between the two 12 year old age groups (p=0.019), with a 4% higher proportion of bilateral reduced VA in the SAVES cohort.

Table 3.2 Prevalence of reduced vision (<6/12) and severity by age throughout childhood

Age Group	Total number of Children	Overall Prevalence of Reduced Vision n (%)	Children with Reduced Vision			
			Mild Reduced Vision n (%)	Moderate Reduced Vision n (%)	Severe Reduced Vision n (%)	Bilateral Reduced Vision n (%)
3-5 years (SPEDS)	986	44 (4.5%)	34 (77.3%)	9 (20.5%)	1 (2.3%)	23 (52.3%)
6 years (SMS)	1739	71 (4.1%)	56 (78.9%)	14 (19.7%)	1 (1.4%)	23 (32.4%)
12 years (SMS)	2345	257 (10.9%)	103 (40.1%)	120 (46.7%)	34 (13.2%)	164 (63.8%)
12 years (SAVES)	1111	134 (12.1%)	35 (26.1%)	93 (69.4%)	6 (4.5%)	91 (67.9%)
17 years (SAVES)	1649	292 (17.7%)	111 (38.0%)	162 (55.5%)	19 (6.5%)	212 (72.9%)

Note: based on unaided visual acuity

Causes of Reduced Vision by Age

3-5 years

Of the 44 children with reduced vision in the group aged 3-5, the primary cause was refractive error (n = 30, 68.2%), with the most common being significant astigmatism (n=25) and hyperopia (n=17) (Table 3.3). However, on further analysis, only 3 children had hyperopia alone and all other children with hyperopia had an additional condition that could account for their reduced vision. In most cases (64.7%), this additional ocular condition was significant astigmatism. Overall, 18.1% had myopic refractive error at this age and 20.5% had amblyopia as a cause of reduced vision.

6 years

Of the 6 year old sample, 4.1% (n=71) were classified as having reduced uncorrected VA, with 78.9% (n=56) of those having mild reduced vision, 19.7% (n=14) with moderate reduced vision and 1.4% (n=1) having severely reduced vision (Table 3.2). The most frequent condition associated with reduced vision was refractive error (88.7%, n=63) including, myopia (18.3%), hyperopia (46.5%) and/or significant astigmatism (54.9%). Despite this, only 31 of the 63 children with reduced vision attributed to significant refractive error were found to use glasses. With refractive correction alone, 21 were no longer classified as having reduced vision (Table 3.4). Of the remaining 10 children, only 3 had an increase in their vision when wearing their glasses from moderate to mild reduced vision. The other 7 children still had the same severity of reduced vision while wearing their glasses. Out of these 10 children, 50% had amblyopia, 50% required an update in glasses and one child had ocular pathology. Other overall causes of reduced vision in this age group were amblyopia (28.2%) and strabismus (25.3%) (Table 3.3).

12 years

There were two cohorts of 12 year old children one from SMS (baseline) and the other from SAVES (follow-up of 6 year olds) who were separately analysed. For both 12 year old cohorts, the proportion of children with reduced vision was just over 10% (SMS 12 years: 10.9%, and SAVES 12 years: 12.1%), which was more than double the rate of reduced

uncorrected VA for the children aged 6 (all p<.001, Table 3.2). The majority of 12 year old children with reduced vision were classified as having moderately reduced vision (SMS 12 years: 46.7% and SAVES: 69.4%). The proportion of those with severely reduced vision was substantially higher in the children aged 12 in SMS (13.2%) compared to the younger sample aged 6 (1.4%). In contrast, the rate of severely reduced VA in the SAVES 12 year old cohort was considerably lower (4.5%) than those aged 12 in SMS. There was a greater proportion of children with bilateral reduced vision in both 12 year old cohorts (SMS: 63.8%, SAVES: 67.9%), than in the younger children aged 6.

This corresponded with a high number of children with refractive error in both cohorts (n=362), specifically myopia, which was the attributed cause for a high proportion of reduced vision at age 12 (SMS: 83.7%, SAVES: 82.8%, Table 3.3). Additionally, for the SMS cohort there was a higher proportion of anisometropia (50.6%), significantly greater (all p<.001) than other age groups, including the 12 year old children from SAVES (22.4%). Of those with reduced vision and significant refractive error at 12 years, 74.3% (n=269) over both cohorts owned a pair of glasses, although a quarter still had reduced vision with their refractive correction (Table 3.4). The prevalence of reduced vision declined with correction worn, after which, only five children still had reduced vision in the SMS 12 year old cohort, caused either by residual amblyopia or ocular pathology. No children remained with reduced vision in the SAVES 12 year old cohort. There were 14.8% of children with reduced vision attributed to amblyopia in the SMS cohort, and 3.7% with amblyopia in the SAVES cohort at age 12 years (Table 3.3).

17 years

A total of 17.7% (n=292) of children were classified as having reduced uncorrected VA at 17 years with 38.0% (n=111) classified as mild, 55.5% (n=162) as moderate and 6.5% (n=19) classified as having severely reduced vision (Table 3.2). This represents a further increase in reduced uncorrected VA in this age group compared to those aged 12. Again, in this age group, refractive error accounted for the largest proportion of reduced vision, specifically myopia (82.2%) (Table 3.3). Of those children with refractive error, most (n=215, 81.1%) already owned a pair of glasses, a higher proportion than in 12 year old children (75%). Only

nine of the 215 children with glasses in the 17 year age group had an incorrect or outdated script and were classified as having reduced vision even with their glasses (Table 3.4). With corrected visual acuity, all of these children were classified as no longer having reduced vision. There were also less cases of strabismus and no children with amblyopia in this age group, indicating that these conditions may have been successfully treated by this age (Table 3.3).

Table 3.3 Common causes of reduced vision in childhood

Causes of Reduced Vision	3-5 years (SPEDS) (n=44)	6 years (SMS) (n=71)	12 years (SMS) (n=257)	12 years (SAVES) (n=134)	17 years (SAVES) (n=292)
Amblyopia	20.5%	28.2%	14.8%	3.7%	0.0%
Strabismus	9.0%	25.3%	10.9%	8.2%	3.4%
Anisometropia	6.8%	22.5%	50.6%	22.4%	15.1%
Myopia	18.1%	18.3%	83.7%	82.8%	82.2%
Hyperopia	38.6%	46.5%	7.4%	6.7%	4.5%
Astigmatism	56.8%	54.9%	33.5%	29.9%	30.1%
Pathology	2.3%	8.5%	5.8%	6.7%	7.2%

Table 3.4 Impact of refractive correction on the proportion of children with reduced vision in those with significant refractive error

Age Group	Number with Refractive Error of those with Reduced Vision	Percentage with glasses	Percentage with reduced vision even with glasses	Reduced Vision after Best Corrected Visual Acuity
6 years (SMS)	63	49.2% (n=31)	32.3% (n=10)	-
12 years (SMS)	238	82.8% (n=197)	27.4% (n=54)	2.5% (n=5)
12 years (SAVES)	124	58.1% (n=72)	18.1% (n=13)	0
17 years (SAVES)	265	81.1% (n=215)	4.2% (n=9)	0

<u>Incidence of New Cases at Follow-up</u>

Table 3.5 shows the incidence of new cases of reduced vision in the 5-6 year follow-up of the SMS cohorts, as part of SAVES. A total of 978 children were examined at both 6 and again at 12 years and 1362 children were examined at 12 and again 17 years. These are the children included in this analysis. Between 6 years (SMS) and 12 years (SAVES) there were an additional 66 cases of reduced vision (7.2% incidence rate per 6 years), with the predominant cause being myopia and just over 50% already having been prescribed glasses. From 12 years (SMS) to 17 years (SAVES), there were 79 additional cases (6.4% incidence rate per 5 years) of reduced vision, with the most prevalent condition again being myopia and almost three quarters of the children with reduced vision already had been prescribed refractive correction.

Table 3.5 Incidence of New Cases at Follow-up (SMS to SAVES, 5-6 years apart)

SAVES Data	Incident cases of Reduced Vision <6/12	% with glasses at follow-up	Predominant cause of Reduced Vision at follow-up
Follow-up age 12	66	51.5%	Myopia – 88%
Follow-up age 17	79	73.4%	Myopia – 66.2%

3.5 Discussion

This chapter aimed to identify the prevalence and common causes of reduced vision through childhood and into adolescence, with the intention of understanding the proportion of reduced vision and ocular conditions that may be detected by visual acuity screening at different ages. While, visual impairment has been previously described in preschool and school-aged children,[369,378] the prevalence of reduced vision and attributed causes are likely to vary with age and location, as well as over time, given increases that have been seen in the prevalence of myopia in recent decades.[136] It is important to define the expected proportion of referrals and conditions likely to be detected for each screening population. The availability of population-based data for children from preschool age to 17 years from the collective Sydney Childhood Eye Disease Studies, has allowed a novel analysis comparing reduced vision and attributed causes across ages for Australian urbanised children. The availability of longitudinal data from SAVES further uniquely provides an opportunity to investigate incident cases of reduced vision through school years and whether repeat screening may be necessary at older ages.

The prevalence of uncorrected reduced vision remained steady at just over 4% in preschool (3-5 years) and early school age (6 years) children, subsequently there was an increase with advancing age to a high of 18% towards the end of schooling (17 years). Severity of reduced vision also increased with increasing age. This was in parallel to increases in prevalent myopia with age which, was present for more than 80% of children with reduced vision in the older cohorts. Age-normative mean visual acuity, initially increased for children between 3-5 and 6 years, and 6 and 12 years, at which time a mean of 6/6 was reached, coinciding with the visual system being fully developed.[379] Following this, there was a decline in uncorrected VA in the 17 year old age group. This was again related to increases in prevalent myopia, however, as reflected in the increase in mean corrected visual acuity to 6/6.

The recommended age for vision screening is preschool age,[27,183,298,321,354,380] corresponding to our 3-5 year old age group. Although, some screening programs still target early school years at approximately 5-6 years of age.[171,187,188,219,221,223,300,303,305] This examination of data

from large population-based studies found that the prevalence of reduced vision was 4.1% and 4.5% in these age groups, respectively, suggesting that at a minimum in terms of finding 'additional' ocular conditions at an school entry compared to pre-school age there is no advantage in this Australian urbanised population. It is to be noted a slightly higher cut off for reduced vision (<6/12) was used, than what is typically used in pre-school vision screening programs as a referral cut-off (<6/9).[184,250,301] So, direct comparison with referral rates from vision screening programs is not practical as these are likely to be higher than this analysis of significantly reduced vision in these populations. The current referral cut-offs used in vision screening programs represents a conservative approach to referral and shifting to a cut-off of <6/12 would increase the sensitivity but reduce specificity of a vision screening program. The lower referral cut off has been deemed appropriate for a pre-school vision screening program and is likely to reduce false negative screening. [215,292,293]

The cut-off of <6/18 used In this study for moderately reduced uncorrected vision more closely corresponds to the criteria for a 'high priority' referral (≤6/18) from screening in the NSW Statewide Eyesight in Preschooler Screening (StEPS) Program.[184] The current analysis based on VA testing by orthoptists with a comprehensive diagnostic assessment, estimates that the screening program should have approximately 1% of the total screened population referred as 'high priority' in preschool aged children. This somewhat less than the high priority rate (2.2%) found in the StEPS program.[184] This difference can in part be explained by the variation in cut-off VA used and that the data was collected by experienced orthoptists in the Sydney Childhood Eye Studies compared to nurse and lay screeners used in the StEPS program.

The most common eye conditions found in this study would indicate that vision screening at preschool age is most likely to find refractive errors, followed proportionally by amblyopia and strabismus. This corresponds with target conditions for childhood vision screening, although the emphasis tends to be given to amblyopia detection due to the need for early treatment.[87,93,183,367] The main goal of vision screening is to detect conditions that are 'invisible', or not readily noticed parents or reported by the child and as such, they are unlikely to seek the required medical attention. Such 'invisible' conditions include amblyopia, as it is most commonly unilateral and it may not be noticed due to the other eye

having good vision.[184] Correction of refractive errors that significantly reduce visual acuity, is beneficial prior to school entry [40,184,357,381] and for early onset myopia, detection at a young age provides the opportunity to commence optical and/or pharmacological intervention to reduce progression to high myopia.[149,382-385] Thus, there is considerable value in detecting refractive errors early and this should be considered a benefit and important aim of preschool vision screening alongside amblyopia detection. These findings support current recommendations for screening to be conducted at preschool age to support optimal amblyopia treatment outcomes and correction of vision prior to school commencement.

Amblyopia accounted for 21% and 28% of reduced vision in our two youngest age cohorts (3-5 years and 6 years). There was a substantial decline in the relative proportion of reduced vision attributed to amblyopia as children increased in age, whereas other causes including refractive error and to a lesser extent ocular pathology increased in prevalence. Interestingly, the percentage of children with amblyopia was lower in the SAVES 12 year old cohort compared to those from SMS at the same age. A possible reason for this is that the SAVES 12 year old children are a follow-up of the SMS 6 year olds, and may have been identified as amblyopic at 6 years, allowing for treatment of their amblyopia to be commenced. Although amblyopia treatment can still be undertaken at age 6, success has been reported to be lower and those with moderate or severe amblyopia may not reach a good level of visual acuity in their amblyopic eye.[386] Therefore, late detection of amblyopia leaves children at an increased risk of vision impairment and blindness in adulthood, in the situation that the better seeing eye suffers from disease or injury.[26]

Strabismus is not typically the focus of vision screening programs, as visual acuity may not be affected, unless strabismic amblyopia develops or there is a concurrent condition. There is also some evidence that constant manifest strabismus may be detected by family observation and care sought prior to preschool vision screening.[358,387] Nevertheless, children with strabismus and reduced vision due to refractive error or strabismic amblyopia can be identified on vision screening, providing an important pathway into care. In the 3-5 year old age group, strabismus was only present in 9% of children with reduced vision but, this increased to 25% in the 6 year old children before a steady decline to a low of 3% in 17 year olds. The increase at 6 years could be related to the onset of, particularly intermittent

strabismus. As strabismus carries psychosocial implications for children in their social settings such as at school [94,368,388] detection and treatment is likely to have a positive impact. There is also the potential for prevention of amblyopia and maintenance of vision. Some screening protocols include observation and stereopsis testing to increase the possibility of detecting strabismus that has not significantly impacted visual acuity,[184] however, it is unclear whether there is significant added benefit, and this may also require additional training for screeners.

The majority of reduced vision across all age groups was due to uncorrected refractive error, aligned with findings from other studies that indicate refractive error is the main cause of vision impairment in childhood.[366,380,389,390] In agreement with previous studies on preschool children, astigmatism was found to be a major cause of reduced vision in children aged 3-6 years.[131,157,272,391,392] A number of children with reduced vision at younger ages were also found to have hyperopia. However, it is known that children with hyperopia are able to accommodate, effectively compensating for their refractive error on visual acuity testing.[39] In the small proportion of children with hyperopia and reduced vision had an additional condition that accounted for their reduced vision. Thus, visual acuity screening alone is unlikely to detect the majority of significant hyperopia in children, but hyperopia may be incidentally detected when present concurrently with another ocular condition that causes reduced vision.

There was an increase in anisometropia as a cause of reduced vision in the 12 year olds from SMS only, which may be due to uneven axial elongation between the eyes with the onset of myopic refractive error at this age.[393] This trend however, was not observed in the 12 year old children from SAVES, possibly indicating this was a chance finding for this particular cohort. A high proportion of anisometropia as a cause of reduced vision at this age may have implications for binocularity due to a difference in clarity of the two images and may be an important condition to target in this age group.[394] Fortunately however, whilst anisometropia is a risk factor for amblyopia,[94] children at this age have passed the period of neural plasticity within which amblyopia develops.[13,41] By age 17 years, the proportion of anisometropia in those with reduced vision was again lower.

There were increases in the overall prevalence of reduced vision at the age of 12 years, and a further increase at 17 years of age. This was related to a higher myopia prevalence in these age groups, corresponding with the typical onset of school myopia, that is said to have its onset around age 10-12.[147] Visual acuity testing has been found to be highly sensitive for the detection of even low levels of myopia, since myopic refractive error cannot be masked by accommodation of the crystalline lens in the same way hyperopia may be.[39] The percentage of severe and bilaterally reduced vision also increased in the two older age groups as levels of myopia rose with ongoing myopic progression.

There is a viewpoint that repeat screening may be necessary at school age, to detect new cases of reduced vision caused by the onset of myopia. Approximately 80% of children aged 12 from SMS and 17 years from SAVES with a refractive error and reduced vision already had glasses, with the majority having good vision with their prescribed correction. There was, however, a lower rate of glasses wear in children who were aged 12 years in the SAVES sample (60%) who would have been unlikely to be myopic at baseline aged 6. The overall high rate of glasses wearing for those with myopia, indicates that symptoms of blur may be reported by children of this age and care sought. Our longitudinal analysis of incident cases of reduced vision revealed that these were largely caused by myopia and while, 73% of 17 year olds were already wearing glasses, this rate was comparatively lower in 12 year olds with incident myopia (52%) suggesting that children of this age have not yet reported their reduced vision. It is important to note that access to public health care is likely to have an impact on rates of correction of refractive errors. In Australia, the federal Medicare program provides free access to optometry care for all citizens[395] and this is likely to have contributed to the high rate of correction of refractive errors. However, prescription spectacles are an additional cost to families, although can be covered at least partially by private healthcare providers which may be a disincentive for some families.

Based on the current findings, if additional school screening were to be recommended, it would be most valuable at approximately age 12, which corresponds to the final year of primary schooling and the commencement of secondary schooling in Australia. There is, however, no indication from the data that repeat screening in early primary school years is likely to have any substantial benefit. An alternative and likely more cost-effective approach

to detecting new onset myopia in older children is to educate parents and children on eye health, with the aim of increasing prompt reporting of symptoms. Also, as we can profile risk of myopia based on demographics such as ethnic background and parental myopia, and risk factor exposure including, competitive schooling, low time outdoors and high near work demand,[147] there is further potential for targeted myopia screening for those at risk, rather than whole population screening.

The large sample of population-based data across a wide age range used in this analysis has uniquely placed allowed examination of the prevalence and causes of reduced vision across childhood. The availability of longitudinal population data from the SAVES study is another significant strength of the current analysis. However, longitudinal data was only available for analysis between 6 and 12 years and 12 and 17 years, respectively. Crucially, longitudinal analysis of new cases of reduced vision could not be conducted between preschool age and school commencement. Further research is warranted to investigate whether there is substantial new-onset reduced vision between these ages, though the cross sectional data presented does not support that this is a significant issue. An additional limitation was that we did not consider the impact of previous screening or eye examinations on the prevalence of reduced vision. Use of uncorrected visual acuity, should somewhat mitigate this impact within our analysis, as it more closely reflects the proportion of reduced vision in an untreated population. However, we cannot determine the impact of previous detection and treatment for amblyopia based on the current analysis and so, the proportion of vision loss caused by amblyopia particularly at older ages is likely to be higher in a completely unscreened and untreated population.

We have described the profile of children with reduced vision and the proportional ocular conditions that are present throughout childhood and that are likely to be detected by population vision screening. Our results demonstrate that amblyopia, which is a priority to detect and treat early, is proportionally a more common cause of reduced vision at a young age. This is supportive of current recommendations for screening to be conducted at preschool age to facilitate optimal treatment outcomes being achieved.[35,321] Refractive error is a significant cause of reduced vision at all ages, with early myopia and astigmatism a priority for detection in young children and school myopia becoming more prevalent from

the age of 12 years onwards. The majority of children with refractive error at older ages had already been prescribed refractive correction, including those with incident reduced vision in our longitudinal analysis. It is likely that community education and/or targeted myopia screening at approximately 12 years of age would be more appropriate than repeat population vision screening during school years. Understanding the profile of the population to be screened is vital for determining the referral burden likely to result from vision screening and for examining the success of preschool vision screening programs.

Chapter 4
The Detection of Refractive Errors in Young Children and Factors Impacting Accuracy

4.1 Abstract

Aim: Recent reports suggest that the mean refraction in infants may be less hyperopic than previously thought. We aimed to examine the distribution of refraction in a population-based study of Australian preschool children and to evaluate the impact of refraction method, iris colour and cycloplegia.

Method: 2462 children aged 6-78 months had a comprehensive eye examination including cycloplegic (cyclopentolate 0.5% for ≤12 months and 1% >12 months) refraction using retinoscopy (majority <30 months), retinomax (K-Plus 2 autorefractor; Nikon Corporation, Tokyo, Japan) (majority <30 months) or table-mounted (RK-F1 Auto Ref-Keratometer; Canon, Tokyo, Japan) (majority ≥30 months) according to the child's ability. Ocular biometry was measured using the IOLMaster (Carl Zeiss Meditec, Jena, Germany) in those ≥30 months of age. Ethnicity was ascertained by parental questionnaire and iris colour was graded prior to dilation using reference photographs. Spherical equivalent refraction (SER) of the right eye was used for analysis using SPSS (v22, IBM, NY).

Results: Mean SER varied between the age groups (6-12, 13-30, 31-48 and >48 months, $p<.001$) with infants 6-12 months (+1.49D) being the most hyperopic. The least hyperopic were the 13-30 months group (mean +1.05D) where the Retinomax was predominantly used, excluding Retinomax, the mean SER was +1.21D. Children with darker irides were less hyperopic than those with lighter irides, even in children of European Caucasian ethnicity ($p=0.008$). Axial length/corneal radius ratio was a good predictor of refraction ($r=-0.64$, $p<.001$) and unlike refraction did not differ significantly between any of the iris colour groups ($p=0.2$).

Conclusion: The Retinomax and darker irides negatively shifted refractive measures, inconsistent with ocular biometry. This suggests that where refractive measures and/or level of cycloplegia are uncertain, biometric measures may assist in accurately classifying refractive errors.

4.2 Introduction

Population-based studies of preschool children in the United States and Singapore suggest that children under 12 months of age may be less hyperopic than previously understood.[120,132,133] The Multi-Ethnic Paediatric Eye Disease Study Group (MEPEDS) found significant differences between ethnic groups in distribution of refraction, where African American children had a less hyperopic mean refraction (6-11 month age group +0.60 SE) than Hispanic children (+1.29).[120] Consistent with this, African American children had a significantly higher prevalence of myopia (almost double) in comparison to Hispanic children who had a significantly higher prevalence of hyperopia.[120] This ethnic difference was supported by findings from the Baltimore Paediatric Eye Disease Study (BEPEDS) where the mean spherical equivalent (SER) for White American children (+1.49DS) was more hyperopic than African American children (+0.71DS) and the prevalence of myopia was also lower in the White American children (0.7% vs. 5.5%).[132] The mean refraction for African American children from both BPEDS and MEPEDS was more myopic (6-11 month age group) and to a lesser extent for Hispanic children also (<23 months age group), compared to previous studies of children of European descent.[2,119] In contradiction, a large population-based study conducted on older participants (12-35 years) of African American and White American descent revealed that while myopia prevalence has increased over time for both ethnic groups, White American participants had a greater prevalence of myopia than African American participants.[396]

Like MEPEDS and BPEDS, the Strabismus, Amblyopia and Refractive Error in Young Singaporean Children (STARS) study also found the prevalence of myopia (11%) to be high in children younger than 6 years, whilst hyperopia was low (7.5%), consistent with the well-known high prevalence of myopia in older Singaporean children.[133] This was however, paralleled by a low prevalence (1.4%) of clinically significant hyperopia (≥ +3DS), considerably lower than reported in BEPEDS for White American children (8.9%).[132,133] It is to be noted that high levels of myopia are not found in all populations of East Asian children, with a study by Lan et al. (2013) of 3-6 year old Chinese children finding that their average refraction was similar to children of European descent at the same age.[397]

While Lan et al (2013)[397] included only participants older than 3 years of age, where the full concentration of 1% cyclopentolate could be administered, STARS, MEPEDS and BEPEDS included participants under 12 months old,[120,132,133] who could only be administered 0.5% cyclopentolate for medical reasons and, reflecting a limitation in determining refractions at this early age. The increased prevalence of myopia in the 6-11 months age group in BEPEDS,[132] MEPEDS[120] and STARS,[133] thus may be related to reduced efficacy of cycloplegia, causing a myopic shift. This limitation may be enhanced when children have darker coloured irides, which are known to resist the effects of cycloplegia. As such, care must be taken when interpreting variation in refraction between ethnic groups, where there is a difference in the prevalence of darker irides, particularly in very young children. Meng et al. (2012), hypothesised that iris colour may itself be a risk factor for myopia, where blue irides are able to reflect back the high-energy visible light, while it penetrates other iris colours making them more susceptible to myopia.[398] However, this hypothesis has not been thoroughly investigated.

In all children and young adults, ensuring adequate cycloplegia is important for accurate refraction to determine prevalence and natural history of refractive development.[278,399] The issue of reduced efficacy of cycloplegia in those with darker irides can be understood through measurement of axial length, which has a high correlation to the refractive state of the eye, and axial length to corneal radius ratio (AL/CR) which is an even better predictor of the refractive state of the eye.[286-288] Additionally, the Retinomax (predominantly used in MEPEDS and BPEDS)[120,132] is known to produce refractions that are more myopic compared to table-mounted auto-refractors and streak retinoscopy.[400] This may have also impacted the prevalence of myopia in these studies.

The impact of these factors and the development of refraction in the 6 months to 6 years age group is not well understood, particularly given these recent indications that myopic refractions of children under 12 months may occur more frequently than previously understood. Thus, the current study aims to identify the age distribution of refractive error in Australian preschool children whilst simultaneously examining the effect of iris colour, ethnicity and refraction method.

4.3 Method

The Sydney Paediatric Eye Disease Study (SPEDS) included a population-based sample of children aged 6 to 78 months (Refer to Chapter 2.2.1 The Sydney Paediatric Eye Disease Study (SPEDS) for a more detailed methods). The SPEDS examination protocols were based on MEPEDS and BEPEDS, allowing for direct comparison. [211,359] A questionnaire was administered to obtain demographic information including ethnicity. Ethnicity was ascertained by the self-identified ethnic origin of both parents using ethnic categories consistent with the Australian Standard Classification of Cultural and Ethnic Groups. All children had a comprehensive ocular examination including, cycloplegic refraction and ocular biometry. Iris colour was graded according to the appearance of the undilated eye, against reference photographs that were classified against four photographic standards as blue, green-hazel, tan-brown and dark-brown (Figure 2.1).[361] A stringent cycloplegic protocol was used with 1% cyclopentolate administered twice in children older than one year and 0.5% in children 12 months and younger. Refraction measurements were obtained following cycloplegia, using the table-mounted autorefractor (RK-F1 Auto Ref-Keratometer; Canon, Tokyo, Japan) in children 30 months and older. For the majority of children under 30 months, the hand-held retinomax (K-Plus 2 autorefractor; Nikon Corporation, Tokyo, Japan) and streak retinoscopy were utilised with the child fixating at 66cm. Preference of method of refraction in order according to child's ability was table-mounted autorefractor, hand-held streak retinoscopy then retinomax. For children aged 30 months and older, IOLMaster was performed for standard biometric measures and the Axial Length to Corneal Radius Ratio (AL/CR) was calculated.

Statistical Analysis: Data was analysed using SPSS (v22 IBM US). Impact of age, ethnicity and iris colour on AL/CR and refraction was analysed through one-way analysis of variance (ANOVA), post-hoc Tukey Honest Significant Difference (HSD) tests, Pearson's correlation coefficients (r) and independent samples t-tests. Participants were classified as having myopia (≤-0.50DS), emmetropia (-0.50 -+2.00), mild hyperopia (+2.00 to <+3.00DS) and high hyperopia (≥+3.00DS) according to the spherical equivalent refraction (SER) of their right eye. Stratification of refractive category by age, ethnicity and iris colour was performed and a Chi-square was used to compare between groups.

4.4 Results

Of the 2462 participants, 2292 had complete data which were included for this analysis (Table 4.1). The mean SER varied significantly between age groups (p<.001) with infants aged 6-12 months being the most hyperopic (+1.49D) (Table 4.2). There was no significant difference (p=0.4) in the mean SER for the two older age groups 31-48 (+1.26D) and >48 months (+1.21D), where the method of refraction was either obtained by table-mounted autorefraction (Canon) or retinoscopy. However, for those aged 13-30 months where the Retinomax was predominantly used, the mean SER was significantly less hyperopic (+1.05D) than all other ages (6-12 months: p<.001, 95% CI, 0.29 to 0.59, p=0.001, 31-48 months: 95% CI, -0.34 to -0.09, >48 months: p=0.014, 95% CI, -0.284 to -0.03). With Retinomax measures excluded, the mean SER was considerably more hyperopic +1.21D, while in this age group for Retinomax alone the mean SER measured +0.92D.

Table 4.1 Participant Demographics

Age	Total	European Caucasian	East Asian	South Asian	Other
6-12	296	131	68	40	57
13-30	510	240	95	66	109
31-48	570	271	112	77	110
>48	916	423	200	121	172
Total	2292	1003	465	292	448

Table 4.2 Mean Spherical Equivalent Refraction for Age and Ethnic groups

	European Caucasian	East Asian	South Asian	P-value	All Ethnicities
6-12 months	1.73	1.33	1.15	*.031*	1.49
13-30 months	1.24	0.69	0.74	*p<.001*	1.05
31-48 months	1.52	1.00	0.88	*p<.001*	1.26
>48 months	1.39	0.96	1.02	*p<.001*	1.21
P-value	*.002*	*.004*	*0.17*		*p<.001*
All Age Groups	1.43	0.97	0.94	*p<.001*	

A significant difference in mean SER was found between the three ethnic groups (European Caucasian: +1.43D, East Asian: +0.97, South Asian: +0.94, p<.001, Table 4.2), across all age groups. The difference in mean SER according to iris colour was also statistically significant across all age groups (blue: +1.55D, hazel: +1.44D, brown: +1.33, dark brown: +1.05, p<.001). Children with brown irides were found to be significantly less hyperopic than children with blue irides (p=0.021, 95% CI, 0.03 to 0.40). The effect of iris colour on mean refraction remained significant within children of European Caucasian origin who had the greatest mix of iris colours, when analysed separately (p=0.008, Figure 4.1 and Figure 4.2). Overall, children with dark brown irides had a significantly less hyperopic mean SER than those with blue (95% CI, 0.38 to 0.62), hazel (95% CI, 0.23 to 0.56) and brown irides (95%CI, 0.14 to 0.43, all p<.001,Figure 4.3). Children of East (p<.001, 95% CI, 0.33 to 0.59) and South Asian (p<.001, 95% CI, 0.34 to 0.64) ethnicity had a consistently less hyperopic mean SER than European Caucasian children (Table 4.2) and if only children with dark brown irides were considered, there remained a less hyperopic mean SER for East and South Asian children compared to European Caucasian children (all p<.001).

Figure 4.1 Number of children with each Iris Colour in each Ethnicity
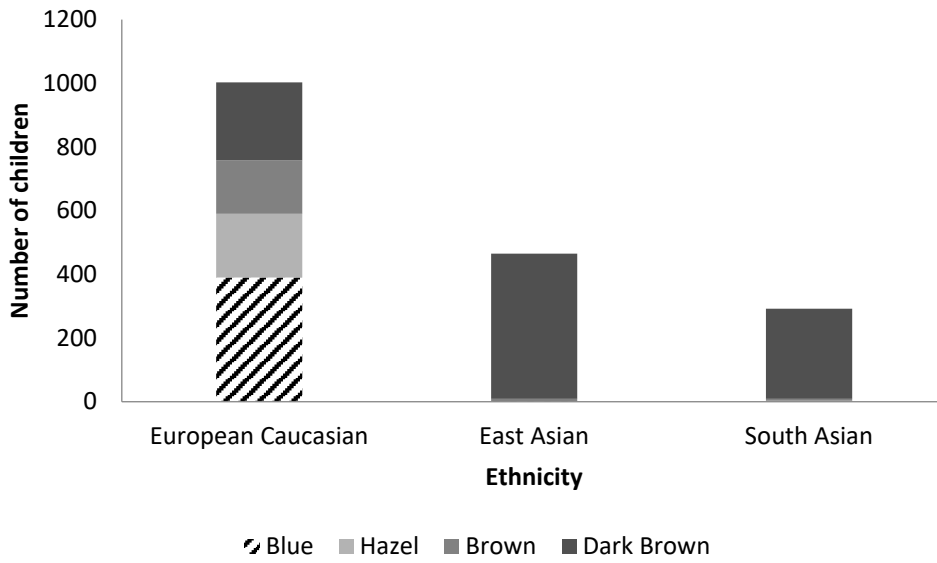
Figure 4.2 Mean Spherical Equivalent Refraction for different iris colours within European Caucasian children
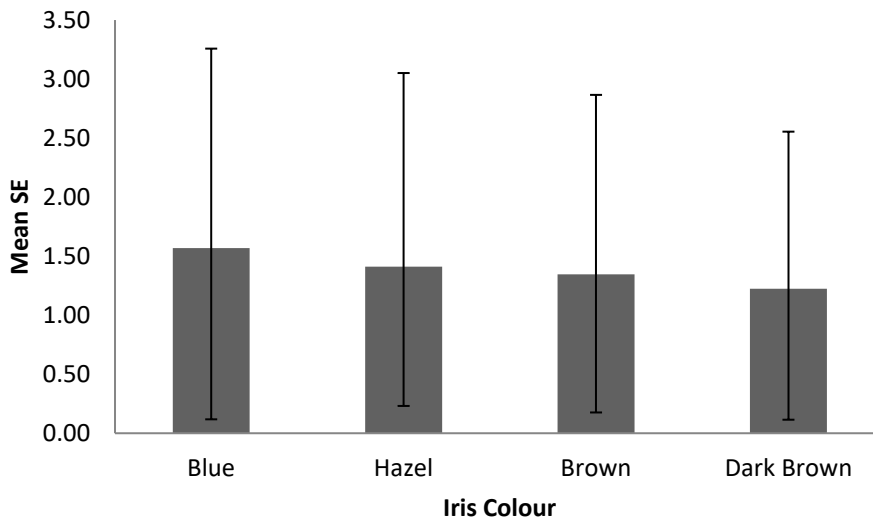
Figure 4.3 Mean Spherical Equivalent Refraction for different Iris Colours
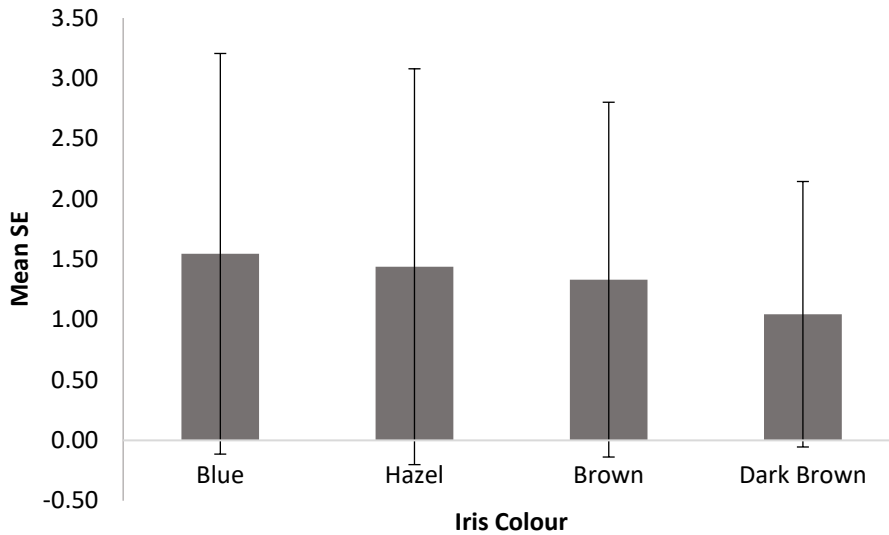
For the children who were 30 months and older with biometry data available, mean axial length (AL) was found to increase by 0.13mm between the children in the 30-48 month age group (22.12mm) and the older children (>48 months; 22.25mm).   The AL/CR, which takes into account changes in corneal radius that may be occurring at this age, was found to be a good predictor of refraction (r=-0.694, p<.001, 95% CI, -0.74 to -0.64, Figure 4.4). The mean AL/CR ratio slightly increased from 2.85 to 2.87 respectively between the groups aged 30-40 months and those >48 months. However, changes in mean AL/CR or axial length were not statistically significant by either ethnicity (Table 4.3) or iris colour (Table 4.4). Within the European Caucasian children only, AL/CR did not differ statistically significantly with iris colour (p=0.3), although AL was longer in those with darker irides (blue: 22.12mm, hazel: 22.08mm, brown: 22.29mm, dark brown 22.36mm), with the difference in AL with iris colour being statistically significant (p=0.028).

Figure 4.4 Mean Spherical Equivalent Refraction vs. AL/CR
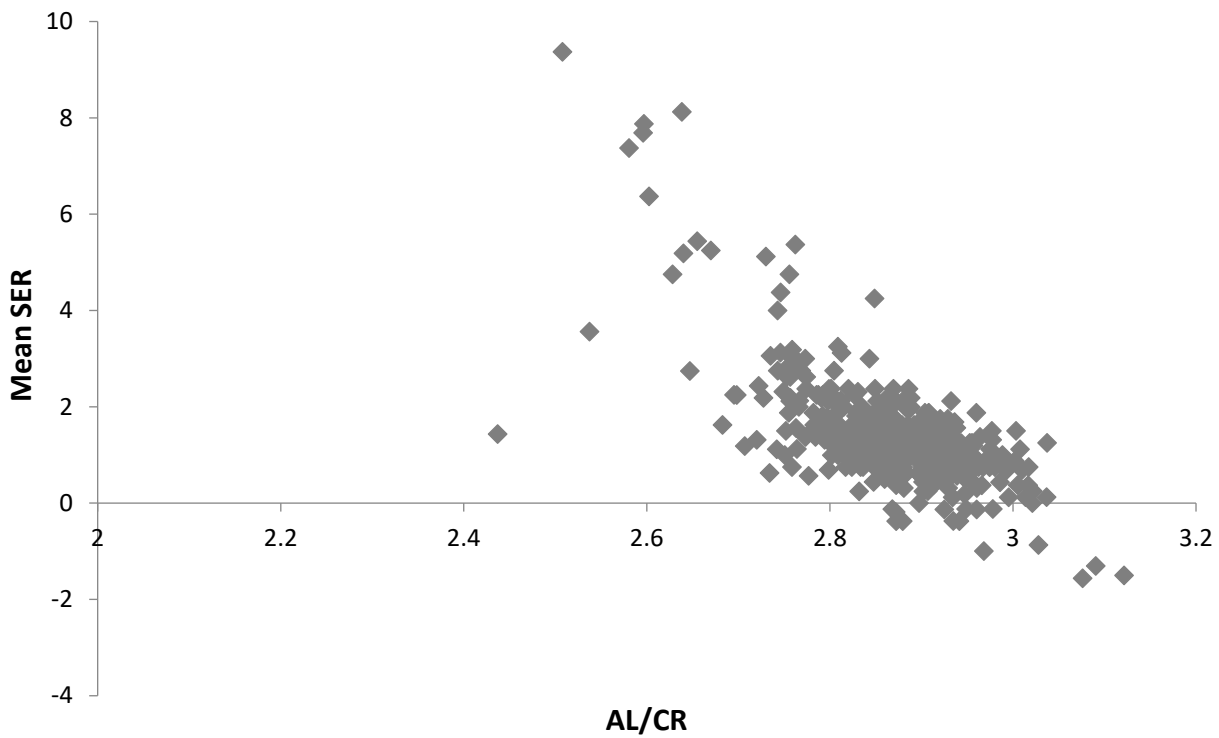
Table 4.3 Mean AL/CR, Axial Length and Spherical Equivalent Refraction for each Ethnicity (Age ≥ 30 months)

Ethnicity (Age ≥ 30 months)	Mean AL/CR	Axial Length	Mean SER
European Caucasian	2.86	22.19	1.58
East Asian	2.85	22.12	1.38
South Asian	2.88	22.35	1.13
P-value	0.38	0.69	.014

Table 4.4 Mean AL/CR, Axial Length and Spherical Equivalent Refraction for each Iris Colour (Age ≥ 30 months)

Iris Colour (Age ≥ 30 months)	Mean AL/CR	Axial Length	Mean SER
Blue	2.85	22.14	1.69
Hazel	2.86	22.08	1.57
Brown	2.86	22.23	1.51
Dark Brown	2.87	22.28	1.17
P-value	0.19	0.07	.001

The majority of children at all ages were classified as having no significant refractive error. Children with dark brown iris colours had the greatest prevalence of myopia (5.9%) which compared to those with brown (1.7%), blue (1.4%) and hazel (0.5%) iris colours, $p<.001$ (Table 4.5). This variation in prevalence of myopia according to iris colour was consistent across all age groups, but only differences in the 13-30 month ($p=0.033$) and 31-48 month ($p<.001$) age groups were statistically significant. In the 13-30 month age group, 8.3% of children with dark brown irides were classified as being myopic which was higher than the children with blue (3.1%), hazel (0%), and brown (1.7%) iris colours. Similarly in the 31-48 month age group, 5.6% of children with dark brown irides were classified as myopic compared to 0% in all other iris colours.

Table 4.5 Prevalence of refractive groups by iris colour across all age groups

Iris colour	Myopia	Emmetropia	Mild Hyperopia	High Hyperopia
Blue	1.4%	79.3%	11.5%	7.7%
Hazel	0.5%	80.3%	15.1%	4.0%
Brown	1.7%	83.4%	10.0%	4.8%
Dark Brown	5.9%	83.3%	7.6%	3.2%

When stratified by ethnicity, a statistically significant difference in prevalence of myopia by iris colour were only present in European Caucasian children (p<.001) (Table 4.6), who had the greatest mix of iris colours (Figure 4.1). European Caucasian children with blue and hazel irides had only 1.5% and 0.6% prevalent myopia, respectively. While, European Caucasian children with brown and dark brown irides, had a comparatively higher rate of myopia 2.5% and 2.4%, respectively. The majority of children of East Asian ethnicity who were not myopic, were classified as having no significant refractive error (87.5% brown and 82.4% dark brown irides) with very low levels of hyperopia in this ethnic group. Children in the South Asian ethnic group, predominantly had dark brown irides, with 7.3% of children with this iris colour being myopic.

Table 4.6 Prevalence of myopia by iris colour for each ethnic group

Iris colour	European Caucasian	East Asian	South Asian
Blue	1.5%	-	-
Hazel	0.6%	-	0%
Brown	2.5%	0%	0%
Dark Brown	2.4%	8.0%	7.3%
All	6.0%	8.0%	7.3%

4.5 Discussion

In this chapter, we aimed to investigate the refractive status of Australian preschool children, whilst examining the impact of refraction method and limitations of cycloplegia. The mean refractive error remained mildly hyperopic for all ages and ethnic groups, however, overall, there was a significant decline in the mean hyperopic SER with increasing age. Infants aged 6-12 months were the most hyperopic age group, with mean SER shifting negatively towards emmetropia with increasing age. There was a more negative shift in the children aged 13 months to 30 months than would be anticipated from the refractive measures seen for the children aged more than 30 months, however, this was the age group that was predominantly measured using the Retinomax. The findings regarding myopic shift in this study caused by use of Retinomax, replicates previous observations regarding its use.[273,274] When the Retinomax data is excluded, with the mean refractive error of +1.21D for the children in this age group substituted, it can be seen refractive error was relatively stable in children older than age one. This suggests that the process of emmetropisation may be most active in the first year of life compared to the older age groups analysed in this study. The trend of decline in SER was apparent for all three ethnic groups, particularly the decline from the mean SER in the 6-12 month age groups to later ages. However, the decline in SER with age was only statistically significant for those children of European Caucasian and East Asian ethnicity. The children of European Caucasian ethnicity had a more hyperopic mean SER at all ages and lower levels of myopia compared to children of both East Asian and South Asian ethnicity.

Recent studies in the United States and Singapore have found a less hyperopic mean SER in children aged 6-12 months than was found in this study using the same lower dose of cyclopentolate (0.5%).[120,132,133] The reduced dosage of cyclopentolate is generally recommended for infants under the age of one due to higher rates of adverse reactions seen with 1% cyclopentolate in this age group. Infants within this age range are thought to have adult-like accommodative responses in terms of accuracy and speed[401,402] with the suggestion that proximity of objects drives the accommodative response more strongly than blur.[403] This suggests that the proximity of the Retinomax and even streak retinoscopy may elicit accommodative responses to a greater extent in the presence of 0.5% cyclopentolate

than seen when using 1% cyclopentolate. While this may provide a partial explanation of the less hyperopic mean SER and higher prevalence of myopia seen in the SPEDS sister studies in the USA and Singapore in the infants under one year of age, it does not explain why a similar finding was not made in the Sydney study using the same protocol.

In MEPEDS, African American infants (6-11 months) had the least hyperopic mean SER (+0.60D) for any age group in the study,[120] while the Hispanic infants of the same age had a mean SER (+1.29D) which more closely approximates the level of mean SER found in SPEDS. Similarly, for the youngest African American age group in BEPEDS,[132] infants (6-11months) had a mean spherical error of +0.45D but in this study the low mean sphere persisted (mean sphere for all; +0.42DS) with little change. Conversely, the refractive measures for White American children in BPEDS more closely resembled that seen in the SPEDS study, with a mean sphere of +1.46DS at 6-11 months, declining in those aged 12-23 months and remaining relatively stable thereafter. The STARS study also found lower mean sphere in infants aged 6-11 months (+0.85DS)[133] that then somewhat persisted in the older age groups in pattern similar to the African American children in the BPEDS study. This pattern of minimal refractive change with age in the predominantly Chinese children in STARS and overall low mean hyperopic refraction did not parallel that of the East Asian children in the SPEDS study.

This variable pattern of refractive development seen across the four sister studies is not easily explained. Firstly, there may be an impact of the lower dose of cyclopentolate used in infants under 12 months of age causing less hyperopic mean refractive measures in this age group. This is most clearly demonstrated in the MEPEDS study where when the higher dose was administered in the older children, a more hyperopic mean refraction was found. Of course this could also be occurring in the other studies, for example the mean SER in the SPEDS European Caucasian infants (+1.73D) and the overall of mean of +1.49D, is still somewhat lower than the mean SER (approximately +2.00D) found in a Boston study of some 500 infants and young children using 1% cyclopentolate.[119] In turn their finding is not too dissimilar to the seminal findings made by Cook and Glasscock in 1950[2] using atropine cycloplegia in new born infants, with the mode of refraction being just greater than +2.00D.

This still leaves open a second possibility; that these later studies are indicating a comparatively less hyperopic mean refraction for the children in all studies and ethnic groups than was seen some decades ago, which may parallel the current myopia epidemic[136] though this can only be speculated at this time. The effect of the myopia epidemic may also be there in a comparison of the refractive measures in the Chinese children in STARS,[133] with them having a less hyperopic mean SE (overall; +0.69D) and more myopia (11.4%) than the children of East Asian ethnicity in SPEDS (overall mean SER +0.97D; myopia: 8.0%). This difference in myopia prevalence in children of the same ethnic group in differing locations, parallels comparative findings previously made between the SMS (Sydney) and SCORM (Singapore) in children aged 6-7 years.[404]

Finally, there is a third possibility, that these differences across studies reflects both the ethnic background of the respective samples and the clinically well-known decreased efficacy of cycloplegia in those with darker irides. As iris colour was recorded in SPEDS and we have a population of mixed ethic origins, the comparison of iris colour and ethnicity reveals that the children who predominantly had dark brown irides were of South Asian and East Asian ethnicity. Those with darker irides were found to have the lowest mean SER error and the highest prevalence of myopia. As has been speculated[398] this could be an effect of the level of light absorption by the higher concentration of melanin in darker irides that in turn may limit the amount of light entering the eye and limiting the protection from myopia development provided by sunlight.[151] If this were the case it would be expected that measures of axial length would vary with iris colour, being greatest in those with darker irides. However, as seen in Table 4.4, though there is a slight trend towards a longer axial length with increasing iris colour, this was not statistically significant. Similarly, change by iris colour was not accompanied by a difference in the AL/CR ratio, which further takes into account variation in corneal curvature. This suggests that the impact of iris colour on measured refraction is most likely to be due to reduced efficacy of cyclopentolate in those children with darker irides. Nor was this an effect of ethnicity. When the data on iris colour was examined in the SPEDS children of European Caucasian ethnicity alone, again there was an effect on SER, with the children with the darker irides having the least hyperopic SER.

It is also to be noted that while SER varied with ethnicity in SPEDS, again differences in AL and AL/CR were found to not be significant across the ethnic groups. Fotedar and colleagues emphasised the impact of lack of cycloplegia on refractive measures in children up to 12 years of age, indicating that clinically significant differences in refractions would be found between children who are cyclopleged versus children who are not cyclopleged adequately.[278] The findings from this study only emphasises the importance of full cycloplegia when trying to determine the refractive state in younger children. If accommodation remains, even only partially active, it may result a shift of refraction in a myopic direction. Biometric measures such as AL and AL/CR ratio with is high correlation to SER, could be used to interpret the efficacy of cycloplegia regimes.[286-288] As AL/CR did not have a correlate with iris colour and did not differ significantly between the different iris colour and ethnic groups, and can be measured non-invasively, it may yet prove to be a useful tool in the management of refractive error in young children.

The pattern of most reduction in the mean hyperopic refraction occurring in the first year of life, followed by a plateauing of refractive measures was seen in all the ethnic groups in SPEDS. This was also seen in the study by Mayer and colleagues[119] and in the majority of the SPEDS sister studies.[120,132,133] This stabilisation of refraction at this age despite axial length growing with age; is reflective of a lower dispersion of refractions with age, an increasingly kurtosis in distribution of refraction and process of emmetropisation.[2,121] Furthermore, the two older age groups 31-48 months and >48 months from SPEDS had no significant differences in refractions, reflective of plateauing and less spread in the distribution of refraction.[121]

However, there were two exceptions to this pattern of refractive changes with age. One was the African American children in BPEDS[132] and the other, the Singaporean Chinese children in STARS.[133] It is notable that the refraction for these ethnic groups at all ages in both studies was considerably and consistently lower than other studies, including the White American children also measured in BPEDS. Whether this is an indication of future early onset myopia or an artefact of measurement is unclear. This would have to be explored in

longitudinal studies to determine if this has is a true and lasting impact on refractive development, as these two ethnic groups to date, have had very different trajectories in terms of myopia prevalence, with the population of young adolescents in Singapore reporting myopia prevalences in the region of 80%.[405]

Further a study conducted on an older population (12-35 years) found that White American participants had a higher prevalence of myopia compared to those of African American origin, although both populations had increased in myopia prevalence overtime.[396] This increased prevalence of myopia in older children may be due to cultural, environmental and lifestyle factors [139,374,406,407] with issues of cycloplegia having a greater effect in younger children.[120,132,133] This again was demonstrated when comparing the STARS to Lan's study on 3-6 year olds in China[397] where the full cyclopentolate could be administered since the children were older, resulting in refractions that were similar to children of European Caucasian descent, suggesting that rather than effect of ethnicity, the STARS study may be due to reduced efficacy of cycloplegia to some extent.[133]

A key strength of this analysis was that SPEDS was a large population-based representative sample of Sydney children, encompassing significant numbers of European Caucasian, East Asian and South Asian children. The study also utilised a stringent cycloplegia protocol, systematically recorded iris colour and undertook ocular biometric measures as soon as the children were old enough. The use of 0.5% cyclopentolate in the youngest infants was not ideal but ethically correct given the possibility of adverse side effects in these infants. While the children of East Asian and South Asian ethnicity had one iris colour predominate, the European Caucasian children had a wide variation enabling analysis of iris effects on refraction in this ethnic group. The use of the Retinomax in the group aged 13 to 30 months was a significant limitation and it may be advisable to use streak retinoscopy in this age group to obtain more accurate measures.

The current study's findings indicate that mean refractions of young Australian children become less hyperopic with age and plateau around +1.2 dioptres after 12 months of age and that is consistent with the majority of studies worldwide. Further research is required to

determine the extent of the effect of iris colour on cycloplegia and to develop an appropriate cycloplegia individualised regime for young children. While it is hypothesised that the resistance of dark irides to cycloplegia had caused an apparent myopic shift in studying refractive development in these young children, there remains a question about the presence or extent of an ethnic influence on refractive development at this young age. It is still uncertain whether there is an interplay between iris colour and ethnicity and the development of myopia, which may be best answered with longitudinal studies. Ocular biometric measures in the future may assist in determining the refractive state of the eye with greater accuracy while assisting in determining which ocular parameters may play the greatest part in early refractive development.

Chapter 5
Longitudinal Change in Refraction and Axial Length in Children with Hyperopia

5.1 Abstract

Aim: To investigate the longitudinal change in refraction and axial length of children with hyperopia. To determine whether children with hyperopia reach emmetropia in childhood, and to understand whether refractive correction had an impact of emmetropisation for children with hyperopia.

Method: Children who participated in the Sydney Myopia Study (SMS), aged 6 and 12, were followed-up after 5 years in the Sydney Adolescent Vascular and Eye Study (SAVES) at ages 12 and 17. All children had a comprehensive eye examination including cycloplegic autorefraction. Spherical equivalent refraction (SER) was calculated and 92 younger (SMS: 6 years) and 31 older children (SMS: 12 years) with significant hyperopia ≥+2.00 diopters (D) were identified at baseline. Axial length (AL) was measured by IOL Master.

Results: For the children in the younger cohort with significant hyperopia (≥ +2.00D) at baseline, by follow-up only 40.2% were still significantly hyperopic. In the older cohort, with significant hyperopia at baseline, 74.2% remained significantly hyperopic. Overall, the younger cohort had significantly greater refractive change (mean -0.80D) and greater growth in AL (0.73mm) than the older cohort (mean SER change: -0.39D, p=0.005; mean AL change: 0.23mm, p<.001). None of the older cohort reached emmetropia while in the younger cohort, one became emmetropic and another myopic. The greatest change of mean SER (-0.81D) and AL growth (0.73mm) was seen in the younger children with moderate hyperopia at baseline (≥ +2.00 to <+3.50D) and was significantly more than the change in SER and AL in those with baseline high hyperopia (p=0.002). This difference in change of SER and AL with level of hyperopia at baseline was not significant in the older cohort (SER p=0.170; AL p=0.439). Correspondingly, the proportion of children with persistent hyperopia (change in SER <-0.50D) was greatest in the older cohort (53.3%) while only 22.1% of the younger cohort were persistently hyperopic. Higher baseline SER was associated with persistent hyperopia only in the younger cohort, while shorter AL was not associated in either cohort. For those whose baseline SER was ≥+3.50D, 55% in the younger and 91.7% in the older cohort remained highly hyperopic (r=0.737, p<.001 and r=0.736, p<.001, respectively). In both cohorts, children with refractive correction had a greater

mean change in refraction (younger with glasses: -1.15D, younger without glasses: -0.71D, older with glasses: -0.56D, older without glasses: -0.28), which was statistically significant in the younger cohort, (p=0.035) but not in the older cohort (p=0.1).

Conclusion: The majority of children with significant hyperopia remained hyperopic. This was particularly evident for those who had high hyperopia at baseline, with nearly all of children aged 12 with high hyperopia remaining highly hyperopic at age 17. This clearly demonstrated that eye growth at this age was not driven by blur, but parallels the slow of growth with age. Wearing of hyperopic correction did not appear to interfere with the process of emmetropisation in the younger children.

5.2 Introduction

It is clear that neonates have a wide distribution of refractive errors; however, this distribution narrows within the first year of life through the process of emmetropisation.[119,121,165,408] This initial emmetropisation that occurs is an active process and is visually driven.[119,165] At the end of the first year of life, the majority of infants have a mild hyperopic refraction, that subsequently slowly reduces throughout childhood. It has been argued that mild hyperopia is in fact the ideal state of refraction in childhood, with adequate reserves of accommodation able to easily overcome this level of refractive error and avoiding a shift into myopia.[118] This second phase of emmetropisation occurs with axial length increasing in parallel with general body growth and offset by a reduction in the power of the crystalline lens while changes in cornea curvature are minimal at this age.[409,410]

Studies have determined that the rate of emmetropisation is generally in proportion to the initial refractive error and tends to slow with increasing age.[165,411] Therefore, children with little refractive error, tend to have small shifts in refraction[165,408] and for those with larger refractive errors, both hyperopic and myopia,[121] there is a greater change in refractive state. While it has been demonstrated that during the first six months of life, children with higher levels of hyperopia demonstrated a more rapid decrease in refractive error than those with lower levels of hyperopia,[165] it is unclear as to whether this trend continues after the first year of life.

Jones and colleagues found that children with hyperopia at age six were likely to remain hyperopic at age 14.[127] They further reported that children with persistent hyperopia had a higher initial refraction, which remained highly hyperopic. However, they had a similar pattern of change in ocular biometric components as was seen in emmetropising hyperopic children. This suggests that that there is a reduction in hyperopic refractive error for children with mild hyperopia, paralleled by their ocular growth. However similar patterns of ocular growth in the children who were persistent hyperopes did not result in a reduction in hyperopia. This is in contrast with the pattern of refraction change and ocular growth that appears to occur in infancy[121]. As such, it could be suggested that persistent hyperopia is not the result of a failure to emmetropise, but is due to abnormal or halted eye development in

infancy that may or may not be related to genetic determinants. Therefore, emmetropia may not be reached for persistent hyperopes who due to a high level of hyperopia in infancy, may exhaust the emmetropisation processes before reaching mild hyperopia or emmetropia.

Given that early emmetropisation is a visually driven process, concerns have been raised that refractive correction for young children with hyperopia may disrupt the process of emmetropisation.[412] This has led to the suggestion that children with hyperopia should be under-corrected to ensure that the visual signals such as hyperopic defocus remain, so that emmetropisation continues normally.[412] Other studies suggest refractive correction is advantageous and that it does not impair emmetropisation.[413,414] Since the process of emmetropisation occurs most rapidly in the first year of life, any significant hyperopia present after this period may be more likely to remain. In addition, the second phase of emmetropisation may never happen or be slowed.[127,415] Despite inconsistent evidence, eye health professionals often leave hyperopic children uncorrected, particularly at a young age due to their good visual acuity[39] or they are routinely under-corrected.[416,417] It has also been suggested that although correction should be prescribed for children with moderate hyperopia when associated with strabismus or reduced visual acuity, there is insufficient evidence to recommend correction for all children with moderate hyperopia.[418] Although visual acuity may not be reduced in children with hyperopia due to their high accommodative reserves, there is the potential that sustained accommodative effort in children who are uncorrected or under-corrected could have detrimental effects on their education and learning, or lead to greater risk of developing amblyopia and strabismus.[40,94,415]

In order to determine appropriate prescribing protocols for children with hyperopia, we need to better understand the natural history of this condition including, both changes in refraction and biometry with growth in childhood as well as the effects of correction. There is currently limited evidence from population-based studies on the process of emmetropisation and the natural history of hyperopic refractive errors in older children. Therefore, this study aims to determine the longitudinal change in refraction and biometry for children with hyperopia and whether they effectively emmetropise through childhood.

Additionally, this study aims to determine the impact of refractive correction on the process of emmetropisation in older children.

5.3 Method

Data from the Sydney Myopia Study (SMS), children aged 6 years and the 5-6 year longitudinal follow-up Sydney Adolescent Vascular and Eye Study (SAVES), children aged 17 years was utilised for the analysis in this chapter. Refer to Chapter 2.2.2 for a more detailed method for the SMS and SAVES.

Spherical equivalent refraction (SER) was calculated and 92 children from the younger and 31 children from the older cohort with significant hyperopia ≥+2.00 diopters (D) were identified at baseline (SMS). The refractive and biometric outcomes for these children, now aged 12 years and 17 years in the younger and older cohort respectively, were determined at follow-up (SAVES). Cycloplegic (cyclopentolate 1%) autorefraction was measured using a table mounted autorefraction (model RK-F1; Canon, Tokyo, Japan) Ocular biometric data was measured using the IOLMaster™ (Carl Zeiss, Meditec AG Jena, Germany) and axial length (AL) calculated as the average of five valid measures.

At follow-up, myopia was classified as ≤-0.50D, emmetropia as >-0.50D to <+0.50, mild hyperopia as ≥+0.50D to <+2.00D, moderate hyperopia as ≥+2.00D to <+3.50D and high hyperopia as ≥+3.50D based on SER of the right eye. A change of ≥0.50D was regarded as being a true change in refraction over the 5 year period since anything less may be the result of instrumental error.

Statistical Analysis: Data was analysed using SPSS (v22 IBM US). Descriptive statistics were used to calculate the mean refraction at baseline and follow-up and change in refraction and axial length in both cohorts over the 5-6 year period. One-way ANOVA was used to determine whether the differences in baseline refraction had an impact on rate of change in refraction and axial length. Chi-square were used to calculate the proportion of children who had changes in refraction and axial length in both cohorts and to determine the impact of glasses on these changes. Logistic regression was used to calculate odds ratios of significant hyperopia (≥+2.00D) at follow-up, as the binary outcome variable, in the presence of moderate hyperopia and high hyperopia (≥+3.50D) at baseline. Linear regression was used to determine the amount of change in refraction that occurred over the 5-6 year follow-up period, according to baseline refraction.

5.4 Results

Of the 31 children in the older cohort with significant hyperopia at baseline, 74.2% remained significantly hyperopic at follow-up and of the 92 younger children with significant hyperopia at baseline, only 40.2% remained significantly hyperopic. Overall, the younger cohort had a greater mean refractive change over the five year period (mean SER change -0.80D) than the older cohort (mean SER change -0.39D, p=0.005), see Table 5.1. Correspondingly, at follow-up there was observed to have been a significantly greater increase in AL in the younger cohort (mean +0.73mm) than in the older cohort (mean +0.23mm, p<.001).

Table 5.1 Overall changes in mean refraction and axial length from baseline to follow-up for both cohorts

	Younger Cohort Mean *(Standard Deviation)*	Older Cohort Mean *(Standard Deviation)*	P-Value
Baseline SER	+2.93D (*1.09*)	+3.44D (*1.20*)	0.030
Follow-up SER	+2.13 (*1.34*)	+3.05D (*1.39*)	0.001
Change in SER	-0.80D (*0.74*)	-0.39D (*0.54*)	0.005
Baseline AL	22.02mm (*0.62*)	22.23mm (*0.72*)	0.120
Follow-up AL	22.75mm (*0.69*)	22.46mm (*0.73*)	0.048
Increase in AL	0.73mm (*0.34*)	0.23mm (*0.18*)	<.001

Of the children with hyperopia 77.9% of the younger cohort and 46.7% of the older cohort had a change in SER ≥-0.50D. In the younger cohort, those with persistent hyperopia were significantly more hyperopic at baseline (mean +3.33D, p=0.038) than those whose hyperopia decreased (mean +3.33D and +2.75D respectively, p=0.038). In the older cohort, baseline SER was not significantly different between those with persistent hyperopia and those whose hyperopia decreased (p=0.7). Baseline AL did not significantly differ between those with persistent hyperopia and those whose hyperopia decreased ≥0.50D for either cohort (younger cohort p=0.8, older cohort p=0.6).

When changes in mean SER and AL from baseline to follow-up were considered by the level of baseline hyperopia present, a more complex picture emerged. Those in the younger cohort with moderate baseline hyperopia had the greatest rate of change in both SER (-0.81D) and AL (0.73mm) of all hyperopic children (Table 5.2). These changes in SER and AL were significantly more than the changes observed for those younger children who had high hyperopia at baseline (SER: p=0.002, AL: p=0.015). In the older cohort, the rate of change of SER and AL in those with baseline moderate or high hyperopia was not significantly different. However, is to be noted that the mean axial elongation for the younger children with even baseline high hyperopia was more than double that measured for the older children with either high or moderate hyperopia, clearly demonstrating a slowing of axial elongation with age.

Table 5.2 Changes in mean refraction and axial length from baseline to follow-up for both cohorts by degree of hyperopia at baseline

Cohort	Moderate Hyperopia (≥+2.00 to <+3.50D)	High Hyperopia (≥+3.50D)	P-Value
Younger Cohort Change in SER	-0.81D	-0.38D	0.002
Younger Cohort Change in AL	0.73mm	0.56mm	0.015
Older Cohort Change in SER	-0.49D	-0.22D	0.170
Older Cohort Change in AL	0.25mm	0.20mm	0.439

Figure 5.1 Proportional change in refractive category from baseline moderate and high hyperopia to follow-up refractive status for both the younger and older cohorts.
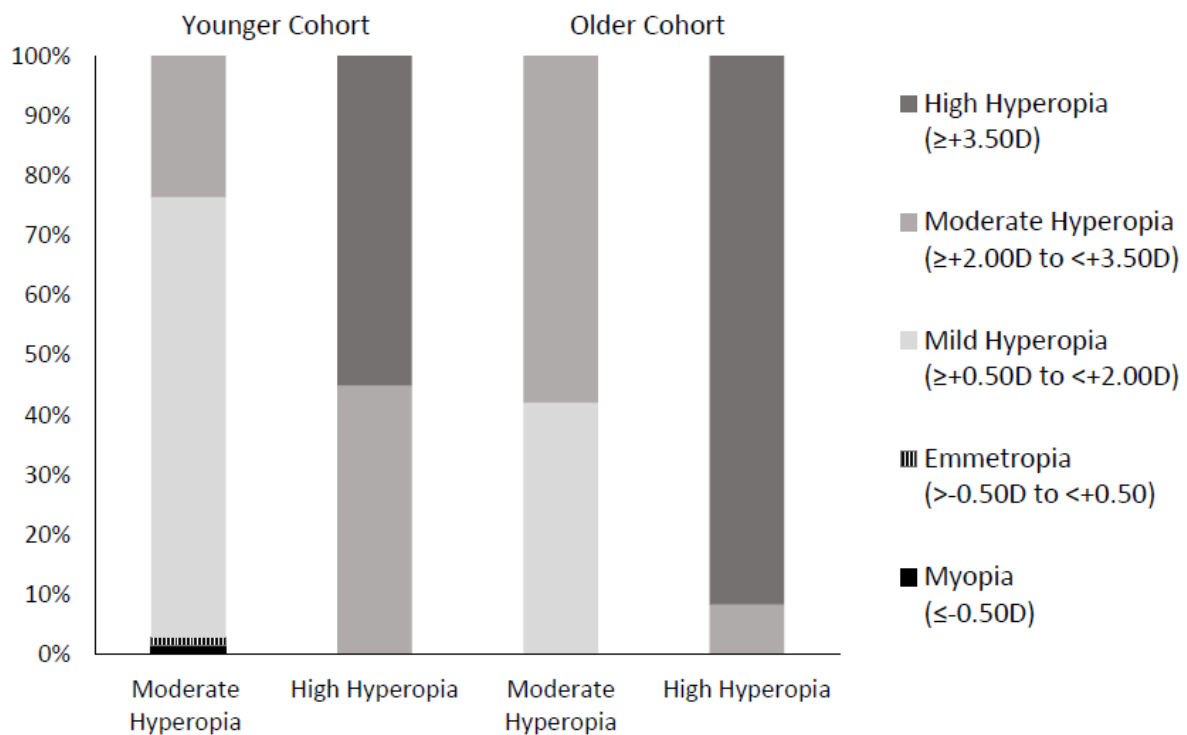
When considering only the children with high hyperopia at baseline, 55% of the younger and 91.7% of the older cohort remained highly hyperopic (≥3.50D) at follow-up (r=0.737, p<.001 and r=0.736, p<.001, respectively) (Figure 5.1). This differed to the children with moderate hyperopia at baseline in the younger cohort, where the majority (73.6%) had become mildly hyperopic at follow-up, while one became emmetropic and the other myopic (-1.31D). In the older cohort, of the children with moderate hyperopia at baseline, 42.1% had mild hyperopia at follow-up.

In both cohorts, children with SER <+2.00D at follow-up were found to have been significantly less hyperopic at baseline (younger cohort, p<.001, older cohort p=0.003) than those whose SER remained ≥+2.00D (Table 5.3). In the younger cohort, decreases in mean SER were significantly greater for those with the lower SER (<+2.00D) at follow-up, compared to those who remained significantly hyperopic (p <.0001). In the older cohort, only the decrease in SER was significantly more for those who had a SER <+2.00D at follow-up (-0.81mm) compared to those who remained significantly hyperopic (-0.24mm)

(p=0.007). In the younger cohort, both the mean baseline AL was significantly longer (p=0.006) and change AL over 5 years greater (p=0.001) in those whose SER was <+2.00D at follow-up, compared to those who remained significantly hyperopic. This pattern was seen to a lesser extent in the older cohort but was not significant.

Table 5.3 Mean change in refraction and axial length at follow-up for children with significant baseline hyperopia (≥+2.00 D) in the younger and older cohorts

| | At follow-up | | | | | |
| | Younger Cohort | | | Older Cohort | | |
	≥+2.00D	<+2.00D*	P-Value	≥+2.00D	<+2.00D*	P-Value
Mean Baseline SER	+3.79D	+2.35D	<.0001	+3.79D	+2.41D	0.003
Change in SER	-0.45D	-1.04D	<.0001	-0.24D	-0.81D	0.007
Mean Baseline AL	21.81mm	22.16mm	0.006	22.10mm	22.61mm	0.086
Change in AL	+0.59mm	+0.82mm	0.001	+0.19mm	+0.33mm	0.079

This includes those emmetropic or myopic at follow-up

Linear regression analysis found that baseline refraction was a poor predictor of change in refraction for both cohorts (younger: r=0.034, p=0.7; older: r=0.153, p=0.4), but was a good predictor of refraction at follow-up at 12 (r=0.831, p<.001) and 17 years (r=0.923, p<.001). A baseline refraction of high hyperopia (≥+3.50D) compared to moderate hyperopia (≥+2.00D-<+3.50D), significantly increased the odds of having significant hyperopia (≥+2.00D) at follow-up for both cohorts (younger: OR: 2.22, 95% CI 1.369-3.608; older: OR: 85.50, 95% CI 6.824-1071.268), particularly in the older cohort.

In the younger cohort, 83 children with significant hyperopia had data on glasses wear. Only 10 of these children with significant hyperopia had refractive correction at baseline. Of

these, eight remained significantly hyperopic at follow-up, whilst two became mildly hyperopic. Of the eight children who remained significantly hyperopic, all had high hyperopia at baseline. There were 73 children with significant hyperopia in the younger cohort without refractive correction at baseline. One became emmetropic and 46 were classified as mildly hyperopic (63%) at follow-up, all of whom were moderately hyperopic at baseline. The remaining 26 children (37%) persisted to be significantly hyperopic (≥+2.00D) at follow-up. Interestingly, the proportion of children with glasses was greater in the older cohort, with 15 of 29 children with significant hyperopia at baseline, presenting with habitual refractive correction. Of those with refractive correction at baseline, 60% were classified as having high hyperopia. Of those with glasses, 12 children remained significantly hyperopic and three had become mildly hyperopic. There were 14 children who had no refractive correction in this cohort, with six becoming mildly hyperopic and eight children remaining significantly hyperopic.

Table 5.4 Effect of refractive correction on mean spherical equivalent refraction in children with significant hyperopia, at baseline and follow-up for both cohorts.

| | Glasses worn at Baseline | | | | | |
| | Younger Cohort | | | Older Cohort | | |
	Glasses Mean SER D	No Glasses Mean SER D	P-Value	Glasses Mean SER D	No Glasses Mean SER D	P-Value
Baseline	+4.72	+2.71	<.0001	+3.87	+2.86	0.021
Follow-up	+3.57	+2.00	<.0001	+3.31	+2.58	0.047
Change in Refraction	-1.15	-0.71	0.035	-0.56	-0.28	0.145

The mean refraction for children with glasses at baseline in the younger cohort was +4.72D, compared to +2.71D for those without glasses (p<.001) (Table 5.4). This difference was also seen in the older cohort, with children with glasses having a mean refraction at baseline of +3.87D compared to +2.86D in those without glasses (p=0.021). In both cohorts, children with glasses had a greater change in refraction (younger cohort with glasses: -1.15D vs without glasses: -0.71D, older cohort with glasses: -0.56D vs without glasses: -0.28). However, this difference was statistically significant in the younger cohort (p=0.035) but not in the older cohort (p=0.1).

5.5 Discussion

While there has been extensive research in the development of childhood myopia, there is limited evidence available to demonstrate the natural history of hyperopia in childhood[127,415,419] and the effects of wearing of refractive correction on the process of emmetropisation.[413,415,420,421] This chapter aimed to understand the pattern of longitudinal refractive change and ocular axial elongation in children with significant hyperopia and whether these children effectively emmetropise with ocular growth through childhood and whether the wearing of optical correction influences emmetropisation.

Those children who effectively emmetropised and were no longer significantly hyperopic at follow-up were more likely to be in the younger age cohort and to have had lower levels of baseline hyperopia and greater refractive change and axial elongation over the follow-up period, than children who were persistently hyperopic. Thus, there was a greater proportion of children who effectively emmetropised in the baseline six year old cohort compared to the 12 year old cohort, particularly if they had moderate hyperopia at baseline. Consistent with this, the majority of children in both cohorts with high hyperopia at baseline remained highly hyperopic at follow-up, in particular those aged 12 at baseline. This suggests that there are two groups of hyperopic children; those with limited refractive change over time, and those who reduce their hyperopia over time, effectively dividing hyperopic children into persistent or emmetropising hyperopes, as similarly observed by Jones and colleagues.[127]

There was also an impact of age on the pattern of refractive change, with emmetropisation slowing with increasing age with a corresponding slowing of axial elongation. The slowing of ocular growth with increasing age is consistent with previous findings for longitudinal change in refraction for all children from this study,[422,423] as well as, progression of myopic refractive errors that also slow with age.[424,425] The impact of this on childhood hyperopia is that those who are significantly hyperopic at a young age, have a greater opportunity to clear their hyperopia through a greater and more effective period eye growth than older children, with the current analysis showing that 60% of younger children, compared to only 26% of older children, no longer had significant hyperopia at follow-up. As those children with hyperopia at 12 years of age, largely remained hyperopic at 17 years, we could assume

that children with hyperopia who have not emmetropised by age 12 years are likely to have persistent hyperopia throughout adolescence into adulthood. Children at both six and 12 years with high hyperopia rarely reduced their hyperopia to a lower level over the follow-up period and may continue to require refractive correction in the long-term.

These findings are similar to previous studies of refractive change in hyperopic children which, have also noted that hyperopic children often remain hyperopic through childhood.[127,129,426] A population-based longitudinal analysis of data from 6 to 14 year olds in the Orinda Longitudinal Study of Myopia, compared ocular component growth curves for four refractive error groups (persistent emmetropia, persistent hyperopia, emmetropising hyperopia and myopia).[127] They reported a similar growth pattern of ocular biometric components for both the children with emmetropising and persistent hyperopia, even though according to their definition of persistent hyperopia, the SER was not changing substantially. As found in the current study, those with little change in their hyperopic refraction, also had a greater level of hyperopia initially, and as such would be unlikely to emmetropise sufficiently to become non-hyperopic beyond early adolescence. Another longitudinal study of school children by Hirsch and colleagues, found that the refraction of children at age five to six was closely related to that found at age 13-14 and concluded that if greater than +1.50D of hyperopia was present at age five to six, children were likely remain hyperopic.[426] Given that the mean refraction at age six years from the Sydney Myopia Study was +1.27D, the results of the study by Hirsch and colleagues has likely been impacted by the myopic shifts in refraction seen with non-cycloplegic refraction and thus, their cut-offs may not be reliable, but the pattern of progression of hyperopia is similar.

The factors that determine whether hyperopia is persistent or emmetropising are currently unclear. Although, this appears to be influenced by something more than baseline refraction given the differing rates of refractive change for children with higher compared to lower levels of hyperopic refractive error, indicating earlier differences in ocular growth in infancy. Jones and colleagues came to a similar conclusion, suggesting that it is most likely that persistent hyperopia is due to a disruption of eye growth early in life, rather than being due to an error in growth through childhood. Whether this has a genetic basis or is influenced by environmental and visual experiences, as is the case for myopia,[144,148,427] warrants further

investigation. Previous research of refractive changes in infancy have indicated that those with greater neonatal refractive errors have a greater shift in refraction during the active phase of emmetropisation in the first months of life, than those with lower initial refractive errors.[121,165,408] These findings suggest a highly active process of emmetropisation in infancy that is visually stimulated, with greater refractive errors and resultant defocus, leading to significant and rapid refractive changes towards emmetropia.

It would also be of interest to examine children with significant hyperopia after initial emmetropisation in the first year of life, to determine if two groups emerge early in childhood, those who have lower rates of eye growth and those who emmetropise more rapidly, and continue to demonstrate this pattern of eye growth through childhood. Recent studies on the emmetropisation process after the initial years of life, have found that for children aged 3-6.5 years[419,428,429] that there is little change in refraction despite significant axial elongation. This is evidently due to lens thinning that is working towards stabilising refraction at a mildly hyperopic level which is advantageous in avoiding myopia. This significant axial elongation was not noted in our group of children with significant hyperopia, particularly in children with high hyperopia.

Iribarren and colleagues conducted their study on slightly older children (school aged children six to nine years)[129] and found there to be minimal change in refraction with the annual rate of change of SER to be -0.31D for persistent emmetropes and an even lower rate of -0.22D per year for emmetropising hyperopes. The rate of change of refraction for emmetropising hyperopes was found to be slightly greater than the children with moderate hyperopia in our study but almost three times the rate of children with high hyperopia. In the current study, children aged six at baseline with moderate hyperopia had a rate of change in refraction (-0.16D per year) that was double that of those with high hyperopia (-0.08D per year) over the 5 year period, reflective of the persistent nature of refraction in children who have high hyperopia.

There has been some speculation in the literature that spectacle correction may inhibit emmetropisation in children with hyperopia, although findings have been inconsistent.[411-414] Children with glasses at baseline tended to have higher levels of hyperopia at all ages,

possibly suggesting an increased likelihood of reduced visual acuity or reporting of symptoms in children with higher hyperopia.[430] Refractive correction did not appear to change whether hyperopic refractive errors reduced over time, with both the children who wore glasses and those who did not, either remaining hyperopic or emmetropising, with no indication that this was impacted by glasses wear itself. In fact, refractive correction may even help promote emmetropisation, with children who wore glasses demonstrating a greater change in refraction between baseline and follow-up, particularly for the younger cohort. This makes sense, since rapid emmetropisation that is visually-guided is largely complete after the first year of life,[415] and it appears that emmetropisation thereafter is more passively associated with ocular growth. Thus, the current investigation suggests that glasses wear does not impact emmetropisation and there should be no barrier to prescription of spectacles for children with significant hyperopia.

Although, many children with hyperopia do not have significantly reduced visual acuity on examination,[39] there may still be substantial benefits to correction of hyperopic refractive errors. Obtaining clear vision in the presence of a hyperopic refractive error requires substantial accommodative effort and thus may result in aesthenopic symptoms associated with eye strain and there is some evidence that lack of correction may result in avoidance of near work.[40,431,432] Hyperopia has previously been associated with reduced academic performance[279,433-436] and although it is unclear whether refractive correction improves the academic performance of children with hyperopia, provision of clear vision to optimise educational outcomes is considered important. Additionally, there is a greater risk of amblyopia and strabismus in the presence of uncorrected hyperopia in young children.[415] Given the potential benefits of correcting significant hyperopia and the lack of evidence that there is any harm, the reluctance of eye health professionals to prescribe optical correction for hyperopia seems unwarranted. The current analysis has not considered the level of refractive correction given and whether this has any impact on emmetropisation of hyperopia. This warrants further investigation to determine if full correction or partial correction would be more appropriate.

Although substantial research effort has been dedicated to investigating the natural history of myopic refractive errors, there has been comparatively limited attention given to

hyperopia. Longitudinal evidence in particular is scant. As the current analysis is based on a large population-based study of school aged children and longitudinally followed change in hyperopic refractive errors over time, this is a significant addition to the scholarly literature in this area. We were, however, limited by having only a small sample of children with significant hyperopia and fewer still who had refractive correction at baseline. Thus, further research on a larger sample size of children with hyperopia would be beneficial. Larger numbers of hyperopic children with refractive correction would also provide a further opportunity to determine if full or partial correction and age of first prescription is important.

In conclusion, this chapter has shown that children with hyperopia often remain significantly hyperopic throughout childhood. This was especially the case for older children and children with higher levels of hyperopia at baseline. In contrary to previous results, persistent hyperopes in this study tended to have less negative shift in refraction over time, in addition to a higher baseline refractive error. While active emmetropisation in early infancy appears to be visually guided with greater refractive shifts for children with higher levels of refractive error, the current results conversely showed greater refractive change towards emmetropia for children with lower levels of hyperopia. Crucially, refractive correction did not appear to interrupt normal emmetropisation in childhood, indicating that there should not be hesitancy to prescribe correction for children with significant hyperopia to reduce excessive accommodative demand and provide clear vision.

Chapter 6
The Efficacy of Vision Screening Protocols in Pre-school and School-aged Children

6.1 Abstract

Introduction: To determine the efficacy of visual acuity assessment for the detection of eye conditions in children aged four and six and the impact of additional testing on the accuracy of screening referrals.

Method: Data on refractive error, strabismus and amblyopia from the four year olds from the Sydney Paediatric Eye Disease Study (SPEDS) and six year olds from the Sydney Myopia Study (SMS) were used for this analysis. Visual acuity was measured using single-surround HOTV letters (EVA ATS; four years, n=215) and HOTV logMAR (EDTRS; six years, n=1741). A comprehensive ocular assessment was also conducted.

Results: Detection of childhood ocular conditions was slightly more accurate in the children aged six compared to those aged four, however, this difference was not statistically significant (four years: AUC 0.68 (95% CI: 0.58 to 0.78); six years: AUC 0.78 (95% CI: 0.74 to 0.81)). A visual acuity cut-off of <6/9.5 had a high sensitivity and specificity for detection of amblyopia in both cohorts. Visual acuity <6/7.5 yielded a high sensitivity or detecting amblyopia, but reduced specificity particularly at age six (four years: 81.8%, six years: 47%). Hyperopia was not satisfactorily detected using the <6/9.5 cut-off (sensitivity - four years: 45.5%, six years: 52.8%). Detection of hyperopia improved using the <6/7.5 cut-off, particularly for children aged six, however, the specificity was low (six years: 47.7%). Near visual acuity detected no new cases of hyperopia. Visual acuity was not reliable for detecting strabismus in either cohort. The addition of cover testing to a <6/9.5 cut-off, detected a further 29 cases of strabismus in both cohorts. Lang II detected eight additional cases of strabismus in the six year olds but no additional cases in four year old children, whilst the Randot Preschool Stereotest detected three additional cases of strabismus in four year olds.

Conclusion: The recommended age for screening is four years to ensure early detection and treatment of amblyopia. A visual acuity cut-off <6/9.5 is accurate for detection of amblyopia at age four and older. However, visual acuity did not reliably detect non-myopic refractive errors, particularly hyperopia, even at high levels. The Randot Preschool Stereotest, which

has a relatively high sensitivity for detection of strabismus and is also easily administered in preschool aged children, could be a recommended addition to vision screening, however, further investigation is required to determine the practicality of this in a screening context.

6.2 Introduction

There is no universal agreement on the ideal protocols for childhood vision screening, nor the optimal visual acuity cut-offs to determine whether a child has age-normal vision.[25] When determining an accurate visual acuity cut-off for screening, the sensitivity and specificity for detection of target conditions must be investigated. The emphasis of vision screening in childhood is the identification of amblyopia and amblyogenic risk factors including, refractive error and strabismus.[25,179,180,321] As visual acuity norms vary with age,[229] the appropriate visual acuity cut-off is likely to be age-specific, depending on the screening population. Consideration also needs to be given to the visual acuity chart used, which may also influence visual acuity referral thresholds, with current recommendations indicating that HOTV logMAR is the most appropriate visual acuity chart for vision screening.[194,321]

The StEPS program uses a cut-off visual acuity of 6/9-2 using either the HOTV logMAR or Sheridan Gardiner Linear vision test, which is ideal considering that the normative visual acuity for preschool children is 6/7.5.[184,229,290] Similarly in the United Kingdom (UK), a referral criteria of <6/9.5 in one or both eyes is used to determine whether a child has failed vision screening using the Keeler crowded logMAR chart for preschool children.[214] The majority of vision screening programs in the United States also use a 6/9 (20/30) cut-off.[308-310,312,314] Studies that have compared different referral thresholds using various visual acuity charts have generally found that in comparison to lower visual acuity cut-offs of <6/6[291] or <6/7.5,[215] the cut-off of <6/9 yields a higher specificity and thus a lower false positive referral rate. The Swedish country-wide vision screening program for four year old children using HOTV logMAR used a referral cut-off of less than 6/7.5 and re-examined children who had vision between 6/7.5 and 6/9.5 some 18 months later finding that very few required any treatment.[292,293] A higher referral threshold of ≤6/12 in screening for children aged four to five years in the UK, was reported to yield a high sensitivity of 86.4%.[294] However, there is the risk that specificity would be low at this threshold and children with significant ocular conditions may be missed.

Traditionally, vision screening was conducted in the early years of school and a number of programs still target children at this age.[21,171,188] Targeting school-age children has the

advantage of having a captive population for screening in schools and has been shown to lead to a high screening rate, increasing universality of screening programs.[171,219,300,348] However, a recent evaluation of the Statewide Eyesight for Preschoolers Screening (StEPS) program found that a high screening rate can be achieved by screening in preschool and community childcare settings.[207]

While children at school age can also be more cooperative, leading to high testability and accuracy of visual acuity testing,[437] more recently, there has been a shift in recommendations to screen vision at preschool age to ensure optimal outcomes for the treatment of amblyopia.[35,184,321] An observational study in the UK found that preschool vision screening provided a 45% reduction in amblyopia prevalence via improved treatment outcomes at age seven, compared to those who were screened at school entry.[183] In further support, a retrospective study concluded that there was no evidence to support that screening at preschool age led to an increase in incorrect referrals.[438]

Most screening programs use visual acuity testing, which has long been accepted as the gold-standard method of assessing vision and has high sensitivity for detecting amblyopia and myopic refractive error, but not hyperopia, which can be quite prevalent in young children.[39,184] Amblyogenic risk factors, including strabismus are clinically detected with specialised tests such as cover test and stereopsis. While cover testing is gold standard for detection of strabismus and is best conducted by those with specialist training, stereopsis tests are easier to administer and can be performed by lay screeners.[282,439] However, whether including additional tests as part of a screening protocol can improve the accuracy of vision screening referrals and detection rates for amblyogenic risk factors is uncertain.

Therefore this chapter aimed to determine the accuracy of visual acuity screening in two age groups, four year olds at pre-school and six year old school children; and to determine the ideal visual acuity cut-off for referral at these ages. The impact of additional orthoptic testing on the accuracy of vision screening in detecting childhood ocular conditions was also investigated.

6.3 Methods

Visual Acuity (VA) results and data on refractive error, strabismus and amblyopia from SPEDS and SMS were used for analysis in this chapter (see Chapter 2.2.1 and 2.2.2 for a more detailed methods for the SPEDS and SMS). Two age groups were chosen for this investigation, children aged four (SPEDS) and six (SMS). Visual acuity was measured electronic vision chart (single-surround HOTV) using the Amblyopia Treatment Study (ATS) protocol in 215 four year olds and a HOTV logMAR chart in 1741 six year olds. Two visual acuity cut-offs (visual acuity worse than 6/7.5 and visual acuity worse than 6/9.5) were assessed for their accuracy in detecting childhood ocular conditions. In the six year olds, near visual acuity was also assessed using a near HOTV logMAR chart at 40 centimeters and the 6/9 cut-off was utilised for the analysis. All children had a comprehensive ocular examination including cover test, Lang II (absent stereopsis considered fail), Randot Preschool Stereotest (≥400" considered fail, available for four year olds only), ocular health check using either fundus photography or indirect ophthalmoscopy and a cycloplegic auto-refraction.

Definitions: Spherical equivalent refraction (sphere + 1/2 cylinder) was calculated and clinically significant refractive errors were classified as myopia ≤-1.00D, hyperopia ≥+3.00D and astigmatism ≥1.00D in either eye. Anisometropia was defined as a ≥1.00D difference between the eyes. Based on the Baltimore Pediatric Eye Disease Study (BPEDS), bilateral amblyopia was defined as best corrected visual acuity <6/12 in both eyes and unilateral amblyopia as a difference of at least 2 lines in visual acuity between the eyes with an amblyogenic risk factor present (anisometropia, manifest strabismus, ≥+4.00D hyperopia, ≤-6.00D myopia, ≥2.50D astigmatism, previous ocular history of patching, strabismus or strabismus surgery) in the absence of ocular pathology [132].

Statistical Analysis: Data was analysed using SPSS (v22 IBM US). Positive predictive values (PPV), negative predictive values (NPV), sensitivity and specificity of visual acuity thresholds for the detection of different childhood ocular conditions were calculated by cross-tabulation across the two age groups. Receiver Operating Characteristic (ROC) curve analysis was performed for the right eye to determine the area under the curve (AUC) which

determined the usefulness of screening at age four compared to age six. The difference AUC between the two age groups was determined by comparison of the confidence intervals. The impact of additional tests to the accuracy of detection was determined for each condition.

6.4 Results

There were 33 children (15.3%) in the sample aged four, and 199 children aged six (11.4%), who were classified as having an ocular condition. When achieving a VA of at least 6/7.5 was required to pass (referral cut-off), the sensitivity for detection of ocular conditions was lower for children aged four (sensitivity: 48.5%) compared to children aged six (sensitivity: 86.4%). There was a higher specificity at age four for this visual acuity threshold (specificity: 84.6%) than at age six (specificity: 50.9%). The positive predictive values (PPV) for this referral cut-off at both these age groups were low but slightly higher for the children aged four (PPV 4 years: 36.5%, 6 years: 18.9%) whilst the negative predictive values (NPV) were high for both age groups (NPV 4 years: 90.1%, 6 years: 96.6%). For both cohorts, the sensitivity for detection of ocular conditions dropped when required to achieve a VA of at least 6/9.5 to pass (4 years: 24.2%, 6 years: 45.7%). However, there was an increase in specificity (4 years: 97.3%, 6 years: 94.7%) and PPV (4 years: 61.5%, 6 years: 53.2%) with a slight drop in NPV (4 years: 87.6%, 6 years: 93.0%).

ROC analysis showed visual acuity was a significant measure for detection of ocular conditions in both cohorts but, stronger in the six year old sample with an area under the curve (AUC) of 0.78 (95% CI: 0.74 to 0.81); than in the four year olds, who had an AUC 0.68 (95% CI: 0.58 to 0.78) (Figure 6.1). However, this difference in AUC between age groups was not statistically significant. When strabismus was removed as an ocular condition, the AUC only improved slightly but not significantly for both cohorts at four years: AUC 0.70 (95% CI: 0.60 to 0.80); and six years: AUC 0.80 (95% CI: 0.76 to 0.83).

Figure 6.1 Receiver-operating characteristic curves (ROC) of right eye visual acuity values for the detection of all targeted childhood ocular conditions for (A) four year olds and (B) six year olds.

A)   Four years   B)   Six years

AUC 0.68, 95% CI=0.58- 0.78

AUC 0.78, 95% CI=0.74 – 0.81
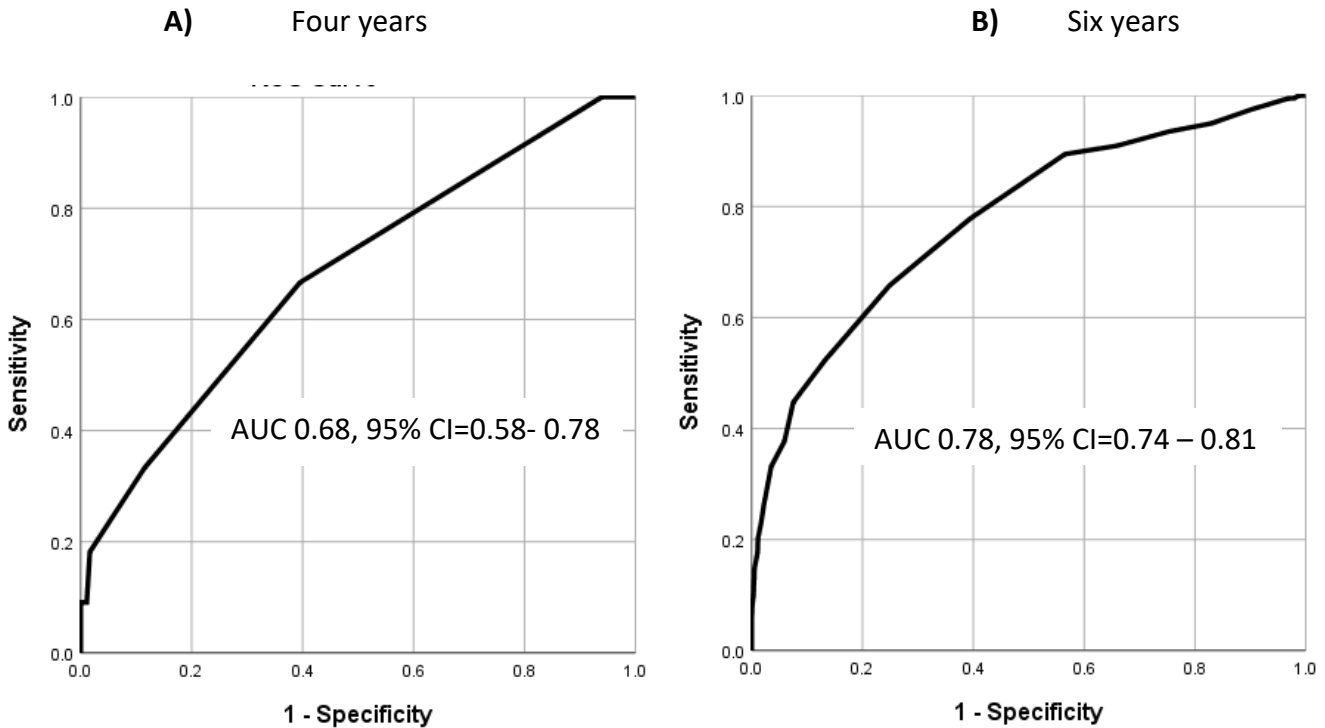
There were six cases of amblyopia in the children aged four, and 17 cases in the children aged six. Whilst there was a sensitivity of 100% for detection of amblyopia at the <6/7.5 cut-off among the four year old children, there was one child who went undetected at the 6/9.5 cut-off, leading to a lower sensitivity but improved specificity compared to 6/7.5 (Table 6.1). There was a sensitivity of 100% for detection of amblyopia at both the <6/7.5 and <6/9.5 visual acuity cut-offs for children aged six, although there was improved specificity at the <6/9.5 visual acuity cut-off.

Table 6.1 The sensitivity and specificity value of different visual acuity cut-offs for the detection of childhood ocular conditions at age four and six.

Age	Condition *Percentage(n=)*	VA <6/7.5 Sensitivity	Specificity	*95% Confidence Interval (CI)*	VA <6/9.5 Sensitivity	Specificity	*95% Confidence Interval (CI)*
4 years	Amblyopia	100%(6)	81.8%(171)	0.467-0.670	83.3%(5)	96.2% (201)	0.508-0.872
	Strabismus	50.0% (4)	80.7%(167)	0.435-0.633	37.5%(3)	95.2%(197)	0.423-0.783
	Anisometropia	60.0%(3)	80.7%(167)	0.429-0.629	60.0%(3)	95.2%(197)	0.429-0.791
	Myopia	50.0%(2)	80.3%(167)	0.353-0.949	25.0%(1)	94.2%(196)	0.280-0.912
	Hyperopia	72.7%(8)	82.6%(166)	0.481-0.687	45.5%(5)	96.0%(193)	0.497-0.858
	Astigmatism	44.4%(8)	77.5%(100)	0.451-0.674	22.2%(4)	94.6%(122)	0.438-0.823
6 years	Amblyopia	100%(17)	47.0%(801)	0.482-0.537	100%(17)	90.9%(1550)	0.501-0.598
	Strabismus	86.5%(32)	47.3%(796)	0.487-0.542	35.1%(13)	90.7%(1526)	0.483-0.578
	Anisometropia	95.5%(21)	47.1%(794)	0.483-0.538	63.6%(14)	90.7%(1528)	0.490-0.587
	Myopia	100%(7)	46.9%(795)	0.476-0.531	100%(7)	90.5%(1536)	0.473-0.568
	Hyperopia	84.9%(45)	47.7%(787)	0.492-0.547	52.8%(28)	91.5%(1511)	0.525-0.625
	Astigmatism	89.3%(108)	49.4%(782)	0.524-0.578	51.2%(62)	93.3%(1477)	0.615-0.716

There were 8 children with a manifest strabismus in the four year old sample and 37 in the six year old sample. This condition was not accurately detected in the four year olds at either visual acuity cut-off (Table 6.1). The sensitivity for detection of strabismus was much higher at the 6/7.5 cut-off for the children aged six however, the specificity was lower compared to the 6/9.5 cut-off (Figure 6.2). The addition of Lang II to a 6/9.5 cut-off detected no new cases of strabismus for children aged four, but detected an additional eight cases in the six year old children, thereby improving the sensitivity for strabismus detection from 35.1% to 56.8% with only a slight drop in the specificity. The use of the Randot Preschool Stereoacuity test, in addition to the 6/9.5 cut-off for the four year old children, detected three additional cases of strabismus, improving the sensitivity for detection of strabismus

from 37.5% to 75.0% with no significant change in specificity (Figure 6.2). Interestingly, the Randot Preschool Stereoacuity test also detected an additional case of amblyopia and astigmatism that was not detected by visual acuity at this threshold. The addition of a cover test to the 6/9.5 cut-off, which is the gold standard diagnostic test for strabismus, detected a total of 29 additional cases across both cohorts. The overall impact in terms of detecting additional cases of strabismus with cover testing would be 2.3% (n=five) for the preschool children and 1.4% (n= 24) for the school aged children.

Figure 6.2 Sensitivity and specificity of strabismus detection at the <6/9.5 visual acuity threshold in four- and six-year-old children and the impact of including stereopsis tests and cover test in increasing detection rates
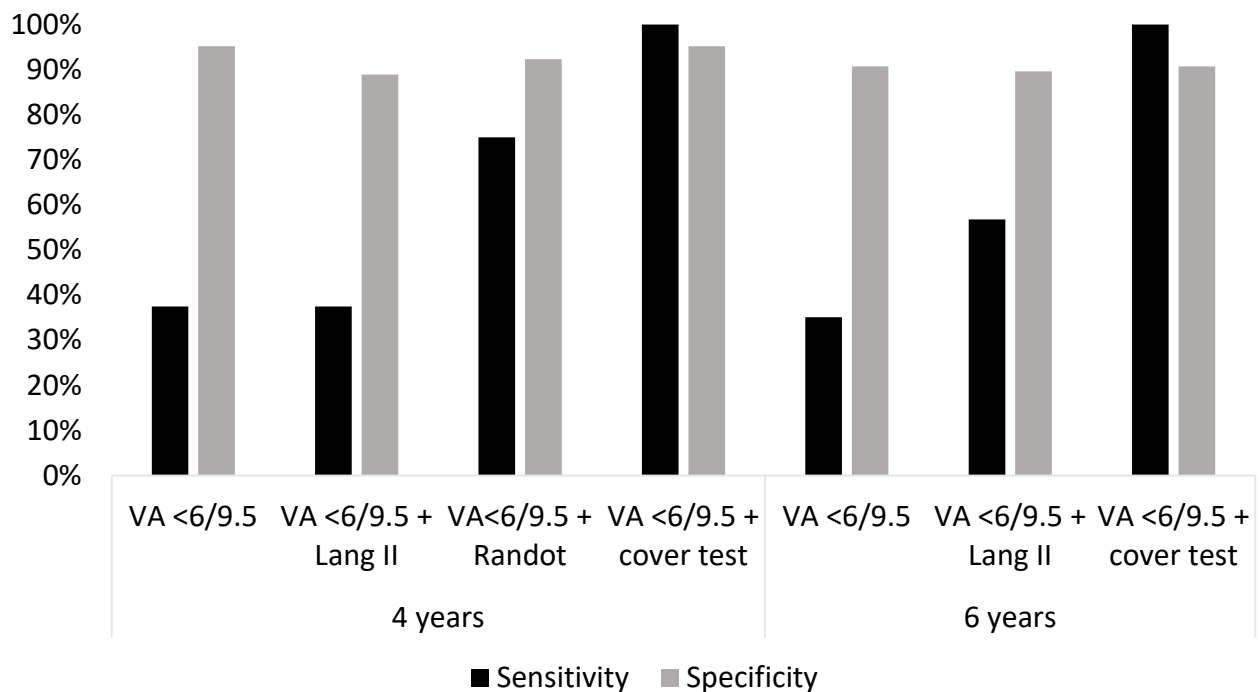
A visual acuity threshold of 6/7.5 successfully detected 72.7% of the 11 children in the four year old sample with clinically significant hyperopia and two of the four children with myopia, with specificity of 82.6% and 80.3%, respectively. Specificity for the children aged four increased at the 6/9.5 cut-off but the sensitivity was reduced for the detection of these refractive errors (Table 6.1). The one four year old child with high myopia (right eye: -4.00, left eye: -5.63), was successfully detected at both visual acuity cut-offs. In the six year old cohort, seven children had myopia and 53 had hyperopia. There was a sensitivity of 100%

for the detection of myopia at both visual acuity cut-offs, with a much higher specificity at the 6/9.5 cut-off compared to 6/7.5. The sensitivity for detection of hyperopia was lower at six years at the 6/9.5 visual acuity cut-off, but with a higher specificity compared to the 6/7.5 visual acuity cut-off. Overall, there was a higher sensitivity but lower specificity for the detection of hyperopia at the 6/9.5 cut-off at six years compared to four years. The addition of near visual acuity testing in six year old children, failed to detect any new cases of hyperopia.

Astigmatism was poorly detected in the four year old sample at both visual acuity cut-offs (Table 6.1), with only cases of astigmatism ≥2.50D reliably detected. Astigmatism was better detected in six year old children at the 6/7.5 visual acuity cut-off but with a poor specificity. The specificity improved at the 6/9.5 cut-off for the six year olds, but conversely the sensitivity was poor. Of the 59, six year old children who went undetected at the 6/9.5 visual acuity cut-off, three of them had astigmatism at a level ≥2.50D and were successfully detected at the 6/7.5 visual acuity cut-off.

Anisometropia was present in five children aged four and 22 children aged six. Of those with anisometropia, 60% were detected at both visual acuity cut-offs at four years, with an improved specificity at the 6/9.5 visual acuity cut-off (Table 6.1). Of the children aged six, 95.5% were detected at the 6/7.5 visual acuity cut-off but, with a low specificity. There was improved specificity at this age with the 6/9.5 visual acuity cut-off but with a lower sensitivity.

6.5 Discussion

The current study aimed to determine the value of childhood vision screening at preschool compared to school-age; to determine a suitable visual acuity threshold for referral and whether additional tests including stereopsis, cover test and near visual acuity would improve the accuracy of referrals. Overall, distance visual acuity was a good test for childhood ocular conditions at both ages, with no significant loss of accuracy when screening at preschool (four years) compared to school age (six years). However, there was considerable variation in sensitivity and specificity by condition, with amblyopia and myopic refractive errors being detected more readily than other refractive errors and strabismus. A threshold visual acuity for referral of <6/9.5 was more appropriate than <6/7.5, as with the <6/7.5 visual acuity cut-off, specificity for ocular conditions was lower, which could lead to substantial over-referral and additional burden on eye care services.

There has been ongoing debate regarding whether screening should be conducted at preschool or school age, with screening at school age often considered favourable due to high testability and accuracy of visual acuity testing and high screening rates.[171,188,219,300,348] In our two population-based samples at four years and six years, the current analysis has demonstrated that there is no indication that screening at school age offers significantly improved accuracy compared to preschool screening. This is in agreeance with a previous retrospective study that similarly found no evidence that earlier screening would compromise accuracy.[438] Thus, screening at four years of age provides good testability and accuracy[208] and early screening has the benefit of maximising amblyopia treatment outcomes, as well as, addressing eye conditions prior to school entry, potentially avoiding detrimental impacts on educational outcomes.[87,184] As such, our results strongly support current recommendations to target four year old preschool children for vision screening.[179,321]

Specificity for the detection of a variety of ocular conditions was very high at the visual acuity threshold of <6/9.5 in both cohorts. This aligns with current recommendations from both the United States[308-310,312,314,321] and United Kingdom[179,214,215] and is consistent with the threshold for referral used in the NSW StEPS program.[184,290] When comparing the accuracy

of referral thresholds at both ages, sensitivity was higher at <6/7.5 but, specificity was higher at <6/9.5. In determining appropriate referral criteria, a balance between sensitivity (low false negative referral) and specificity (low false positive referral) must be identified. Although the sensitivity for detection of ocular conditions was good at the 6/7.5 cut-off for children aged six, the associated poorer specificity is not ideal. This poor specificity arises because the mean visual acuity for children of this age is 6/7.5. Similarly, in four year old children specificity was higher at a cut off of <6/9.5 and as such, this threshold is more appropriate for screening referral.

Amblyopia is the primary target of childhood vision screening programs and our findings are in agreeance with previous literature, where amblyopia was successfully detected at both visual acuity cut-offs and in both age-groups.[317] There was a greater specificity for amblyopia detection at four years of age, particularly at the 6/9.5 cut-off, again suggesting this is an appropriate referral threshold. Myopia was also successfully detected at both cut-offs for the six year old children, consistent with previous studies that have found visual acuity to be accurate for myopia detection.[39,170,317] In the four year old sample only, there was low sensitivity for detection of myopia although specificity was very high. However, there was only a small number of children with myopia in this age group and as such, this finding requires further investigation in a population with higher levels of myopia at a young age. In utilising visual acuity for myopia detection, close attention should be paid to ensuring children do not use eyelid squinting to improve visual acuity through a pinhole effect.

The <6/9.5 cut-off did not successfully detect hyperopia or astigmatism in either age group, in agreement with previous studies.[39,170,317] Although hyperopia was successfully detected at the <6/7.5 cut-off in the six year old cohort, the low specificity aligns with normative visual acuity for this age group,[229] and suggests that this threshold would not be feasible to utilise in screening protocols. Interestingly, near visual acuity did not add diagnostic value to the assessment and detection of children with even clinically significant hyperopia. This is inconsistent with a previous study by Jin et al. of 4416 children aged 6-12 years that reported that near visual acuity was better for detecting high hyperopia and that a combination of distance and near visual acuity had the biggest area under the curve for

detecting high hyperopia.[318] The difference between the current study and that reported by Jin and colleagues is perhaps due to the current study being conducted on younger children who have larger reserves of accommodation to overcome hyperopic blur even at high levels of hyperopia. [39] Whilst the detection of astigmatism was also poor, no child with high levels of astigmatism was missed at either cut-off for the four year old children; however, three children aged six, with high levels of astigmatism were missed at the 6/9.5 cut-off but detected at the 6/7.5 cut-off.

To determine if the addition of further testing in vision screening protocols could improve the accuracy of screening, particularly for the detection of strabismus, we examined the impact of cover test, Lang II and the Randot Preschool Stereotest. Cover testing is the gold-standard for strabismus detection and unsurprisingly detected all cases of strabismus. However, a disadvantage of using cover test in a vision screening context is that it is a skill that requires specialist orthoptic training and vision screening is usually conducted by lay screeners and nurses. Although studies have suggested that orthoptic screening would be preferable as it would involve more comprehensive investigation and thereby greater accuracy of referrals,[172,173,291,324,325] the feasibility and cost-effectiveness of this has not yet been determined in an Australian context. The addition of Lang II and/or the Randot Preschool Stereotest did detect additional cases of strabismus, although the feasibility and time constraints of including either of these tests in a screening protocol would also need to be evaluated. Lang II, although quick and easy to use, only provides a gross measure of stereopsis and lacks accuracy in younger age groups.[254,256] The Randot Preschool Stereotest may be particularly useful in supporting the detection of strabismus in four year old preschool children, with our findings demonstrating an appropriate sensitivity, but again its overall utility in a screening protocol would have to be further assessed. The justification for detecting smaller angle or intermittent strabismus, that are not readily detected by families and observers, is the potential for developing future amblyopia or their possible association with substantial refractive errors, particularly hyperopia, which may not be readily detected by visual acuity tests.

This is the first study to directly examine the effectiveness of screening referral cut-offs in a population-based sample of preschool, compared to school-aged children. However, the

current analysis had several limitations that need to be considered when interpreting these findings. Firstly, the sample size for the four year old sample, was considerably smaller than the six year old sample. However, both samples were population-based, and therefore this comparison is likely to be valid. In addition, different visual acuity tests were used between the two cohorts, which may impact the comparability of visual acuity results, since the type of test can influence the accuracy achieved. Despite this, both tests have optimal crowding for the detection of amblyopia and utilise the same standardised optotypes and sizing. Thus, the different tests used in this analysis are likely to have had limited impact on the results obtained.

In conclusion, the detection of amblyopia had an excellent sensitivity at both age groups at both cut-offs, demonstrating that visual acuity testing is highly useful to detect this condition. Our findings support current recommendations that screening at preschool age is accurate and most appropriate to implement early treatment, and that a threshold visual acuity of <6/9.5 for referral is optimal for the detection of ocular conditions. While, visual acuity screening is accurate in detection of myopic refractive errors, hyperopia is less accurately detected. Near visual acuity did not improve hyperopia detection and investigation of non-invasive methods of screening for hyperopia is warranted. The Randot Preschool Stereotest improved strabismus detection, an important risk factor for amblyopia, and could be considered as an addition to visual acuity screening however, the practicality of including this in a screening context needs to be evaluated.

Chapter 7
Preschool Vision Screening Study – The Statewide Eyesight Preschooler Screening (StEPS) Program

7.1 Abstract

Aim: The StEPS program recently transitioned from the Sheridan Gardiner linear chart to using the HOTV logMAR chart. This study aimed to determine the comparability of these two visual acuity tests, the impact on referral thresholds and whether additional orthoptic tests improved the accuracy of referrals. The barriers to acting on referral from preschool vision screening in relation to the StEPS program was also investigated.

Method: Ninety four, four year-old children were recruited through the StEPS program. Children had their vision screened at their respective preschools, using the Sheridan Gardiner chart and the HOTV logMAR chart, followed by an orthoptic examination that included a cover test at near and distance fixation and stereopsis tests. Children who did not pass screening were referred for further examination through the STEPs program. In the second part of this study, parents/guardians of children who were referred from vision screening were followed up one month after screening to determine whether a follow-up appointment had been made and attended and if not, what barriers had impacted successful follow up.

Results: An additional 19.1% of children qualified for routine referral ($<6/9^{-2}$) with the Sheridan Gardiner chart (n=24) compared to the HOTV logMAR chart (n=6). There were two high priority referrals ($\leq6/18$) using the Sheridan Gardiner visual acuity chart and none using the HOTV logMAR. The difference in mean visual acuity between the HOTV logMAR chart and the Sheridan Gardiner chart was statistically significantly (logMAR: 0.0665, approximately 3.5 letters, p<.001). Of the two children who had an inter-ocular difference of greater than two visual acuity lines using HOTV logMAR, one was provided a routine referral and the other classified a borderline pass. Only three additional children were detected with more comprehensive orthoptic assessment, with one already under the care of an eye health professional. There were 10 children referred, of which two were referred due to inability to screen and were later diagnosed with developmental/behavioural problems. Of the eight children referred for failing vision screening, 50% did not attend follow-up appointments. Reasons for non-attendance included family issues, forgetting appointments and not perceiving there to be a problem.

Conclusion: Visual acuity measures and testability rates were significantly higher using HOTV logMAR than Sheridan Gardiner chart. Screening using HOTV logMAR, currently considered the gold-standard test, resulted in a substantially reduced referral rate overall. The median visual acuity achieved with HOTV logMAR was approximately 6/7.5 and therefore, the current visual acuity cut-off of <6/9 is suitable for referral when using this chart. The current StEPS referral criteria does not consider inter-ocular difference and these results suggest this may be a beneficial inclusion. Additional testing did not detect a significant number of additional referrals and would not be recommended for screening protocols. Loss to follow up is a significant problem for vision screening programs and approaches to reducing barriers to acting on referrals are important for ongoing success of vision screening programs.

7.2 Introduction

The New South Wales Statewide Eyesight Preschooler Screening (StEPS) Program is a free universal vision screening program for four year old children in NSW.[184] The program has been shown to be effective, with 96.4% of four year old preschool children in NSW being offered screening and 75.6% of them being screened.[207] The StEPS screening program aligns with current recommendations for preschool vision screening at age four,[27,179,440-442] when testability with letter-based visual acuity charts is acceptably high.[443,444] This is also an appropriate age for effective implementation of amblyopia treatment and to ensure vision problems are addressed prior to school entry.[25,298,445]

There are a number of different vision test designs available for preschool children. Previously, no particular test has been recommended universally for this age group, however, recent evidence and recommendations suggest that HOTV logMAR is the best designed and most standardised vision chart and appropriate for vision screening.[194] The StEPS program initially used the Sheridan Gardiner (SG) Linear chart but has recently transitioned to HOTV logMAR to align with these recommendations.[207] The SG Linear chart utilises seven mirror image letters (X, O, T, U, H, A and V), whilst the HOTV logMAR chart uses only four of these letters (H, O, T and V). It is well recognised that optotypes presented linearly (in lines of systematically varying optotype size) or single optotypes surrounded by bars at a set distance, known as crowding, are crucial for the detection of amblyopia.[195-197]

A primary limitation of the Snellen and therefore the Sheridan Gardiner visual acuity test is the non-uniform spacing between letters and lines of letters in these tests. This was addressed by the development of vision tests using logarithmic progression of optotype size (logMAR), with equal spacing between lines of letters and the letters themselves, which has progressively been introduced in visual acuity tests for children.[80,195,205,206] No study to date has made a direct comparison of the SG vision test to a logMAR vision test using the HOTV letter set, for testing vision in children. This comparison would be particularly useful for the StEPS screening program, since their recent change to HOTV logMAR from SG Linear could have an impact on the testability and appropriateness of referral criteria for the StEPS program.

In addition, the HOTV logMAR chart used in StEPS has an additional line (6/15) that is not available on the previously used SG Linear Chart. StEPS refers children with 6/18 vision or worse, as a high priority referral which, provides the option for parents/guardians to attend a Paediatric Ophthalmic Outpatients Clinics (POOC) within the public hospital system in NSW.[184] However, the addition of the 6/15 line on the HOTV logMAR chart means that there is potential for some children who would have previously received a high priority referral, to now be a routine referral, altering their referral pathway substantially.

While, visual acuity testing has long been accepted as the gold-standard method of assessing vision and is particularly useful in detecting amblyopia and myopic refractive error,[39] other conditions such as strabismus are clinically detected with other specialised tests such as cover test and stereopsis. Thus, a further area of inquiry is whether the incorporation of additional testing is feasible and whether it would increase the accuracy of vision screening within the StEPS screening program.

Failure to act on referrals from vision screening programs is a well-known problem and is recognised as an established limitation for the success of vision screening programs. Previous studies conducted predominantly in the United States (US) have reported loss to follow-up rates as high as over half of the parents/guardians with referred children failing to act on recommendations to attend for further assessment and treatment.[333-335,344] A major barrier identified from vision screening programs in the US has been lack of insurance and concerns about financial burden of follow-up care. In Australia, free ophthalmic care through public hospitals is available, particularly in metropolitan locations and free optometric consults can be accessed through the Australian Government Medicare program.[395] However, a recent evaluation of the StEPS program found that overall 10% of parents/guardians did not act on routine referrals for children who failed StEPS screening (children with visual acuity <6/9^{-2} in at least one eye).[207] Nearly 11% of high priority referrals (children who had visual acuity ≤6/18 in at least one eye) were not acted on in rural/regional areas and 4.9% were not acted on in metropolitan local health districts. Barriers to accessing appropriate follow-up care are likely to vary in Australia compared to other locations, although this has not been investigated.

Therefore, this chapter has a number of aims:

1. To assess the comparability of the Sheridan Gardiner Linear and the HOTV logMAR visual acuity charts and determine how the chart used may vary referral rates in the StEPS program.

2. To examine whether additional orthoptic testing can improve accuracy of referrals from vision screening.

3. To investigate barriers to follow-up care after referral from the StEPS program.

7.3 Method

Refer to Chapter 2.3 for a more detailed explanation of methods for the Preschool Vision Screening Study.

Ninety-four 3.5-4 year-old preschool children were recruited through the StEPS Program from 2019 to 2020. Children were recruited from eleven different preschool and childcare centres within the Sydney Local Health District (LHD) in metropolitan Sydney. It is to be noted that further recruitment was curtailed by the advent of COVID 19 health restrictions in 2020.

Phase 1: Vision screening was conducted using StEPS screening protocols (refer to Appendix 2a) at the preschool or childcare centre using the HOTV logMAR chart with matching card (Good-lite, US, retro-illuminated 6 metre chart) and Sheridan Gardiner Linear (BOC Instruments, Silverwater, NSW, Aus, retro-illuminated 6 metre chart) also with a matching card. The order of using the HOTV logMAR and Sheridan Gardiner vision tests was alternated between children to avoid confounding variables such as fatigue, biasing results obtained. This was followed by an orthoptic examination that included, cover test at near and distance fixation, stereopsis using Lang II (Lang stereotest, Forch, Switzerland), the TNO stereoacuity test (Laméris Ootech BV Nieuwegian, The Netherlands) and the Randot Preschool Stereotest (Stereo Optical Company, Chicago, IL). Referral to eye care professional services was based on the results of the HOTV logMAR vision test, according to the StEPS referral criteria (Box 2.1). Children with abnormalities detected on orthoptic testing were also referred through the STEPS screening program for further assessment.

Phase 2: Parents/guardians with children who were referred from the Preschool Vision Screening Study were contacted one month after screening via phone to determine the outcome of referral and whether a follow-up appointment had been made and/or attended. When not reachable by phone, an email was sent to obtain contact with parents/ guardians. Where parents/ guardians indicated that a follow-up appointment was not made or not attended, the barriers to accessing follow-up care were discussed. After the child attended

their follow-up appointment and with parental consent, eye care practitioners were contacted to obtain the child's diagnoses.

Consent and Ethics: Informed written consent was obtained from all parents/guardians of participating children prior to vision screening. This study was approved by the Human Research Ethics Committee of the University of Technology Sydney and adhered to the tenets of Declaration of Helsinki.

Statistical Analysis: Data was analysed using SPSS (v22 IBM US). The distribution of visual acuity within the sample was skewed when using both charts, in particular when using the Sheridan Gardiner (skewness - Sheridan Gardiner: 2.510, HOTV: -0.244). Therefore we selected non-parametric statistical tests to compare the two visual acuity charts for this analysis. To compare the two visual acuity charts, Spearman's correlation coefficients were calculated and Wilcoxon signed-rank test was used to compare medians and interquartile ranges between the HOTV logMAR and Sheridan Gardiner visual acuity tests. A Bland-Altman plot was also created to visually assess agreement between the two visual acuity tests. Frequency tables and descriptive statistical analyses were utilised to determine differences in number of referrals between the charts at various thresholds consistent with the StEPS referral criteria. The additional orthoptic tests were examined descriptively. Phone interview data was analysed descriptively to identify emerging themes.

7.4 Results

<u>Comparability of HOTV LogMAR and SG Linear Visual Acuity charts:</u>

Of the 94 children screened, 73 passed screening with a visual acuity better than 6/9 in both eyes using HOTV (77.7%) and an additional 13 were classified as a borderline pass. In comparison, only 28 passed vision screening using the SG chart (44.7%), with 23 classified as borderline pass. This reflected a 19.1% higher rate of routine referral when using the SG chart (n=24) compared to the HOTV chart (n=6). There were no high priority referrals using the HOTV chart, but two children were considered high priority referrals using SG. One of these children passed screening when performed with the HOTV chart, with a number of letter confusions demonstrated when they tested with the SG chart with its more extensive optotype set and the other child achieved a borderline pass when the HOTV chart was used (Figure 7.1, Table 7.1). Two children were unable to be tested by either the HOTV or SG Linear test and were referred on the basis of inability to be screened. Another child was unable to be tested using the SG chart, yet was a routine referral using the HOTV logMAR.

Figure 7.1 Referral status using the Sheridan Gardiner Linear chart compared to the HOTV logMAR chart.
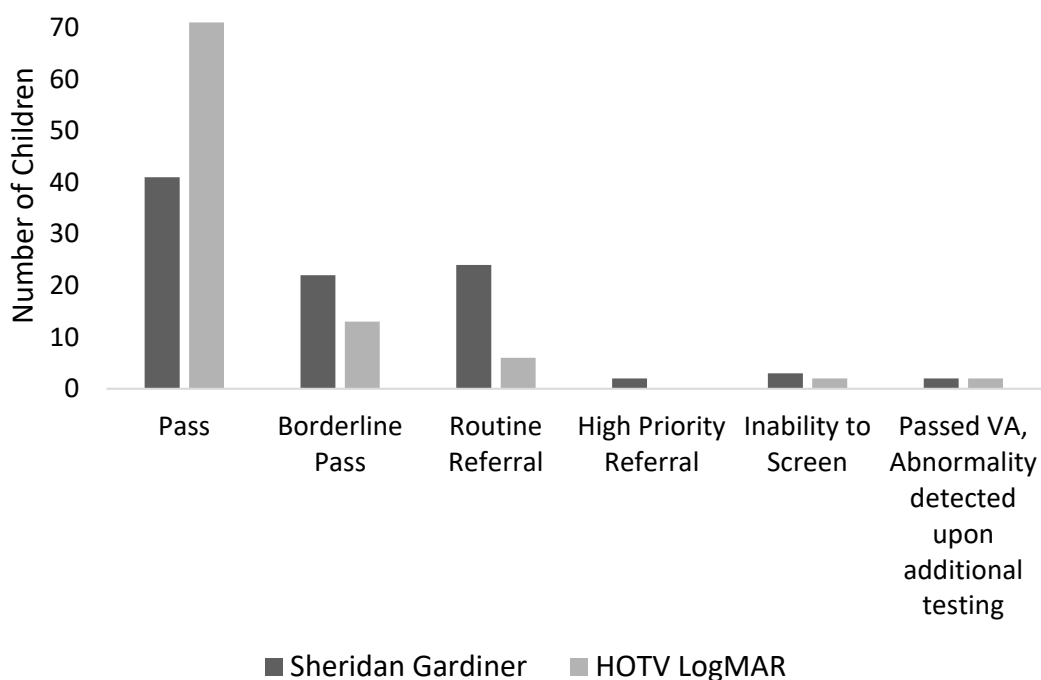
Table 7.1 Agreement in referral status between HOTV logMAR and Sheridan Gardiner

		HOTV LogMAR					
	Referral Status	*Passed*	*Borderline Pass*	*Routine Referral*	*High Priority Referral*	*Inability to screen*	*Abnormality on additional testing*
Sheridan Gardiner	*Passed*	40	1	0	0	0	0
	Borderline Pass	20	2	0	0	0	0
	Routine Referral	10	9	5	0	0	0
	High Priority Referral	1	1	0	0	0	0
	Inability to screen	0	0	1	0	2	0
	Abnormality on additional testing	0	0	0	0	0	2

There was no statistically significant difference in median visual acuity between the right and left eyes using either the HOTV (Z=0.537, p=0.591) or SG (Z=-1.246, p=0.213) vision tests. Therefore, to compare the difference in median visual acuity between the two charts, only the data for the right eye was used. A Wilcoxon Signed-Ranks Test indicated that the median outcome achieved with the HOTV test (median logMAR: 0.120, IQR:0.040-0.176 (median Snellen equivalent: 6/7.5[+1]))was statistically significantly higher than the SG test (median logMAR: 0.176 IQR:0.136-0.216 (median Snellen equivalent: 6/9)) (Z=-6.374, p<.001).There was a positive moderate-high correlation (Spearman's correlation r=0.665, p<.001, 95% CI: 0.528-0.769) between visual acuity results obtained with the two charts, but this relationship was not considered very strong since children on average performed better when tested with the HOTV logMAR chart (Figure 7.2). Additionally, on the Bland-Altman plot showed a clinically significant mean difference between the two charts of 3.5 letters (mean logMAR=-0.0665, limits of agreement: -0.291-0.158). For the majority of participants

the points were plotted below zero, showing that mean visual acuity was higher when measured with HOTV in comparison to Sheridan Gardiner (Figure 7.3).

Figure 7.2 Comparison of the logMAR VA values using the right eye for the HOTV logMAR and Sheridan Gardiner Linear charts.
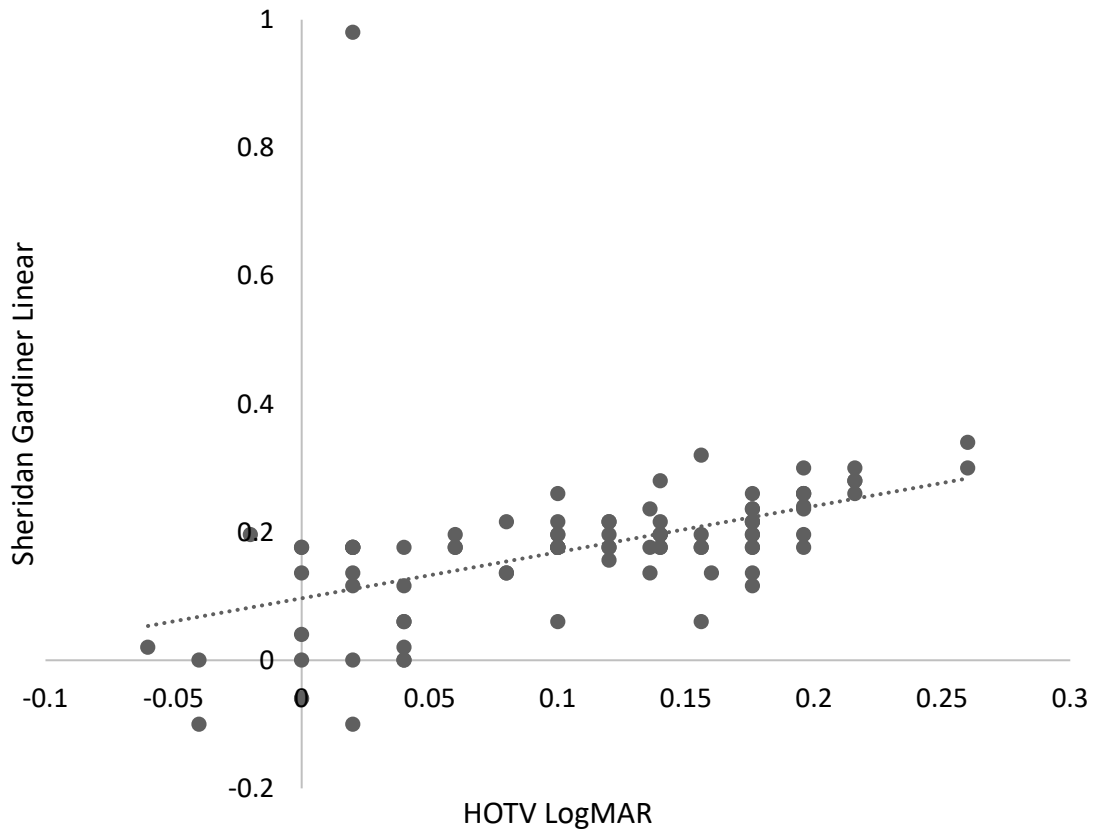
Figure 7.3 Bland-Altman Plot: An assessment of agreement between the HOTV logMAR and Sheridan Gardiner Linear chart
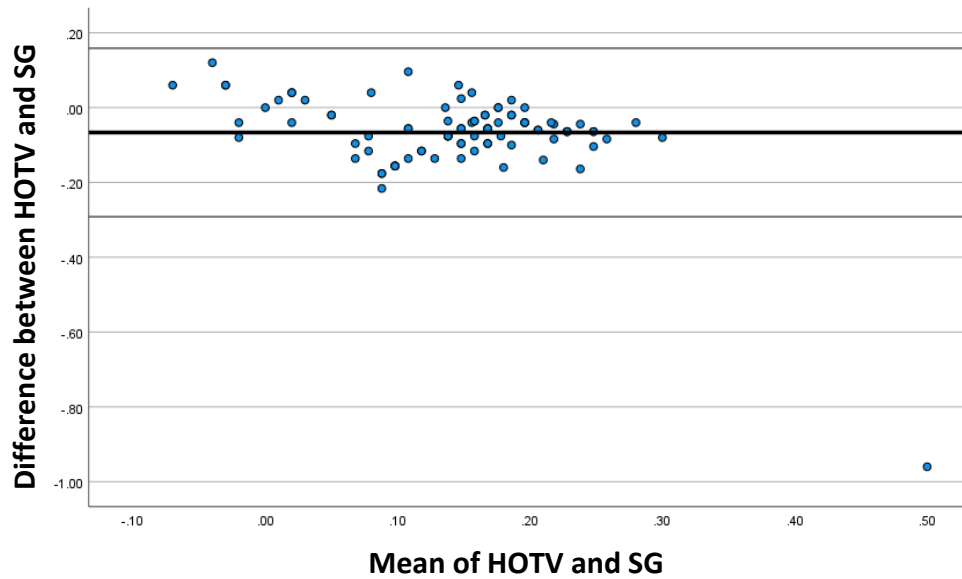
There were two children with an inter-ocular difference of greater than two lines (≥10 letters) using the HOTV logMAR chart. Based on the current StEPS referral criteria, one child was classified as a routine referral after failing to attain the 6/9-2 threshold and the other was a borderline pass. There were six children who had an inter-ocular difference of ≥2 lines of visual acuity when using SG, two of whom also had a significant inter-ocular difference found when tested with the HOTV logMAR chart. Of the additional four children detected as having an inter-ocular difference ≥2 lines on testing with the SG, two had a pass result on both tests, meaning the eye with the worse visual acuity was still better than the threshold for referral. The remaining two children were both classified as high priority referrals on SG, with one becoming classified a borderline pass and the other a pass when tested using the HOTV logMAR chart.

Additional orthoptic testing detected abnormalities in three children who were not detected on the basis of visual acuity testing using the HOTV logMAR chart. One child had gaze-evoked nystagmus on ocular motility testing and interestingly, also had an inter-ocular difference of ≥2 lines with SG but not when tested using the HOTV logMAR chart. Another child who had an intermittent exotropia and equal visual acuity between eyes, passed screening using both visual acuity tests. This child was not provided a referral as they were already under the care of an eye health professional. Observation of pupil size detected

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another child who had anisocoria and was classified as a borderline pass according to their visual acuity when tested using SG but passed visual acuity testing with the HOTV logMAR chart (Figure 1).

Outcomes of referral from screening:

Overall, 10 children were referred from screening in this study. Two children were referred due to inability to be screened, six were routine referrals based on their visual acuity being <6/9-2 but better than 6/18 in both eyes and two were referred based on the orthoptic assessment. Of the eight children referred for failing vision and/or orthoptic screening, 50% (n=4) attended follow-up appointments, three of whom had been routine referrals and one a referral based on the orthoptic assessment. Of the two children who were referred for further assessment due to inability to screen, both were found to have developmental /behavioural problems and were under the care of a child psychologist following screening. One was additionally being managed by a speech pathologist.

Two of the children referred based on their vision/orthoptic assessment, received follow-up ophthalmology care, one was attended an optometrist and one received secondary screening by the StEPS orthoptist. The parent of the child reviewed by the StEPS orthoptist reported that this was due to rapid availability of this consultation, appreciated after experiencing a high level of concern following the screening report and also because they did not have access to Medicare services, being neither a citizen nor permanent resident of Australia. Of the further two children referred who attended follow-up care this was with an ophthalmologist. Both were prescribed glasses for myopia and astigmatism, and one was also prescribed patching for amblyopia. The one child who attended the optometrist was prescribed glasses for hyperopia.

Three children did not attend a follow-up appointment. Two of these children were siblings (twins) and had been classified as routine referrals. The reason given for non-attendance was because the appointment with the optometrist was inadvertently missed and the research team was informed that the family intended to reschedule. On a further follow-up call, the appointment had not been rescheduled and this was attributed to family problems and a lost referral letter. The third child had not had a follow-up appointment scheduled

with no decision made whether an appointment would be made in future. The parents/guardians reported being happy that their child's vision was within normal limits, as they were referred on the basis of additional orthoptic assessment. The final child who was referred was a routine referral and their parents or guardians were not able to be contacted to determine the follow-up outcome.

7.5 Discussion

This is the first study to directly compare the utility of the HOTV logMAR and Sheridan Gardiner chart in preschool aged children, particularly in relation to a screening setting. We aimed to determine the comparability of visual acuity results and referral rates based on the StEPS referral criteria obtained when the two charts were used. This is an important and timely analysis to determine the impact of the change in vision charts used in the StEPS program on expected referral rates and the appropriateness of pre-determined referral thresholds. We further sought to understand whether additional orthoptic testing would improve the accuracy of screening referrals. Finally, we investigated referral outcomes and barriers to acting on referral to access eye care services.

We have demonstrated a significant difference in the visual acuity results based on the two vision charts, with consistently better visual acuities obtained using HOTV logMAR compared to SG chart. This resulted in an almost 20% higher rate of referral using SG based on the current study. A significant contributor to this would be that the median visual acuity for this sample of children using SG chart was 6/9, closely corresponding to the referral threshold used in the StEPS program. In comparison, the median visual acuity using HOTV logMAR was approximately 6/7.5, making the 6/9-2 referral threshold significantly more appropriate for the use of this chart. HOTV logMAR has several advantages compared to SG, including standard letter progression and uniform crowding and as such, is considered the gold-standard for visual acuity screening.[194] Thus, we expect that the visual acuity results obtained using the HOTV logMAR are accurate. This has two implications for the StEPS program; firstly, that the referral rate using HOTV logMAR is likely to be significantly lower than previously found when the SG chart was used and secondly, that the accuracy of referrals is likely to increase, following the transition to the logMAR chart.

An additional impact on referral was observed at the level of high priority referral, which according to the StEPS referral criteria, corresponded to a visual acuity of ≤6/18. As visual acuity was, on average, one line better using HOTV logMAR compared to SG chart, there is potential for a number of children to no longer be classified as being a high priority referral using the new visual acuity chart. While two children were classified as high priority based on SG visual acuity test, no children were classified as such when using HOTV logMAR. To

further compound this effect, the HOTV logMAR chart has an additional line of letters corresponding to 6/15, that lies between the 6/18 and 6/12 lines available on the SG chart. This means that children with visual acuity of >6/18 but <6/12 would have previously been considered a high priority referral, but would not be, based on HOTV logMAR. As high priority referral results in greater access to public ophthalmology services, consideration should be given to whether ≤6/18 remains a reasonable threshold for this level of referral, or whether it would be appropriate to reduce the threshold to <6/15 to capture the children who would have been classified as a high priority referral previously. It is to be noted that only a small number of children were considered a high priority referral using either chart in the current study and further evaluation of the impact on high priority referrals on a larger scale would be valuable to determine if visual acuity <6/15 captures cases that require routine care or specialised care in POOCs.

There were less children who passed, and more children who were classified as routine referrals and borderline passes, when the Sheridan Gardiner chart was used compared to using the HOTV logMAR chart. There are only four letters necessary for children to accurately match using HOTV optotypes compared to seven SG optotypes, increasing the difficultly of this test. Thus, the higher rate of routine referral and borderline pass could be the result of lower testability using SG optotypes for preschool aged children. It was observed that the children often confused the letters A, V, X, and U during testing, supporting this hypothesis and confirming the appropriateness of the subset of letters H, O, T and V used in the logMAR chart. There are other fundamental differences between the two charts, for example the difference in the number of letters on each line and uniform crowding and spacing between letters, which could have also contributed to improved testability for HOTV logMAR. Higher testability also meant that the HOTV logMAR chart could be completed in a shorter amount of time compared to the SG chart. Two children who received a very poor visual acuity result and another who was unable to be tested using SG, two of whom subsequently passed and the other obtained a borderline pass when tested with HOTV logMAR, while all children were testable using HOTV logMAR, confirming its high rate of testability and accuracy in this age group. As vision screening is often conducted by nurse and lay screeners, the increased testability of HOTV logMAR makes this a very appropriate test for vision screening purposes and may reduce unnecessary referrals

for children who were unable to be screened using the SG chart. This observation is in agreement with other evidence that the HOTV logMAR test has high testability (≥90%) in children aged four to six years.[208,249,250].

Interestingly, the two children who were unable to be screened using either visual acuity chart in the present study were found at follow-up to have previously undiagnosed behavioural/developmental problems. This was consistent with an anecdotal findings reported in the recent evaluation of the StEPS program that suggested children referred as unable to be screened were often later diagnosed with developmental problems such as autism.[207] This may represent an unexpected but valuable additional benefit of vision screening for preschool children.

Definitions of amblyopia in the literature focus on reduced age norm visual acuity or a difference in visual acuity between the two eyes of at least two lines.[95-97] As such, it has previously been suggested that children who have an inter-ocular difference of at least two lines of visual acuity are of clinical importance, even if their visual acuity is good in both eyes compared to the norm for age.[446] This implies that they should be referred from vision screening programs as a potential diagnosis of amblyopia or with an amblyogenic risk factor such as anisometropia or strabismus. However, the StEPS program referral criteria does not currently include interocular difference. One of the two children who had an inter-ocular visual acuity difference of at least two lines in the current study, was classified a borderline pass according to the StEPS criteria for referral, meaning that they would not routinely be referred. As this could potentially be a false negative result, it is recommended that consideration be given to a referral criteria of a difference of ≥ 2 lines of visual acuity between eyes being adopted. However, in order to implement this, a further protocol change would be required in the StEPS program, ensuring that visual acuity is tested to threshold, rather than only to 6/9 and that a staircase technique of testing be used to rapidly establish threshold acuity.

The more comprehensive orthoptic assessment only detected three additional children, who were not identified by visual acuity testing, one of whom was already under the care of an eye health professional for strabismus. This suggests that only a small proportion of additional children would be detected with more comprehensive testing. Considering this,

as well as the extensive time required to perform these tests, and that this would require either employment of orthoptists as vision screeners or additional specialised training for nurse and lay screeners, it would be difficult to recommend this as a further course of action. Whilst using orthoptic screening has been proven to be successful in a previous study,[437] the costs required to implement this may outweigh its benefits and it has yet to be evaluated in an Australian context. Extending the vision screening time to conduct further tests also lacks feasibility within a preschool or childcare environment.

The StEPS protocol encourages observation by screeners, in addition to visual acuity examination, to detect further conditions that may not have reduced visual acuity alone. This is likely to be sufficient in a screening scenario to detect large-angle strabismus and visible ocular pathology. This too requires some training of screeners, but is more likely to not take a great deal of time and be performed with reasonable accuracy within a vision screening program. It is recommended that an observation protocol is incorporated into vision screening programs that ensures a screener performs thorough observation of the child's eyes to ensure easily observable conditions are not missed. It is also possible that children with strabismus and ocular pathology may already be receiving care by an eye health professional. This was demonstrated in the current study where a child who passed visual acuity screening and had equal visual acuity in both eyes, had an intermittent exotropia but was already under the care of an eye health professional. The role of orthoptists as secondary screeners, is likely to be more feasible and has been used to good effect in the StEPS program.[207]

The current analysis found that only half of those referred attended follow-up care, consistent with the low rates of action on referral reported from a number of previous studies of vision screening programs.[333,334,336,337,346] This follow-up rate in this small sample was much lower than the 10.9% rate reported in the StEPS evaluation, however, the rate reported from over nine years of data from StEPS is more likely to be an accurate reflection of the outcomes from preschool vision screening in Australia.[207] Poor follow-up, subsequent to vision screening, is a significant problem and impacts the effectiveness of the screening programs overall. Reasons for non-attendance found in this study were family issues, forgetting appointments and not perceiving there to be a problem. This is fairly similar to

findings from previous studies, potentially indicating that similar issues may impact follow-up in Australia as found elsewhere.[333-338,344,346,347] However, Australia has the advantage of Medicare-covered optometry services[395] which, does reduce some of the issues related to cost that can be present, particularly in the United States.[336,337] Education about eye conditions and the importance of early intervention may also be beneficial to improvement of rates of follow-up care.[344] A larger detailed investigation of barriers to follow-up is warranted to determine how best to encourage and facilitate action by parents to attend eye-care professionals in a timely manner.

It is to be noted that in this small sample, one parent was unable to be contacted to determine follow-up status. This appears to be a frequent problem when trying to follow-up results of vision screening as seen in previous studies.[335] The StEPS evaluation also found inconsistency in returning reports from eye care professionals.[207] To improve monitoring and reminders for children who are referred, automated and electronic reporting methods could be investigated. Parental reports of referral outcome may also not be accurate and it is likely to be better to obtain information directly from the treating eye care professionals, who would be able to provide a more detailed version of any ocular conditions and treatment the children may require.

This study is novel in its comparison of two visual acuity charts previously used for preschool vision screening in the StEPS program and evaluation of the impact of the shift to HOTV logMAR on referral rates from the program. However, there are some limitations that need to be considered. Firstly, as not all children were referred for a comprehensive ocular assessment, the sensitivity and specificity rates for different childhood ocular conditions cannot be determined from this study. Although, this would be a valuable direction for further research, our aim was focused on comparability of visual acuity measures and impact on referral rates, as opposed to a comparison of the accuracy of each test.

Barriers to follow-up from vision screening has not been previously investigated in an Australian population and as such, despite our small sample size for this phase of the study, the interview data has provided some insight into barriers that may play a role in the Australian context. Further research should be conducted on a larger sample of children following referral and ideally, would also examine the impact of rural and metropolitan

location. This would enable the development of strategies to mitigate barriers to follow-up and potentially reduce loss to follow-up from the StEPS program.

In conclusion, there was a significant difference in referral rate between the HOTV logMAR and SG vision charts, which may be attributed to greater testability of the HOTV logMAR and perhaps reflects that this test is more cognitively compatible for the developmental stage of a four year old preschool child. Transition to the HOTV logMAR chart from the SG chart in the NSW StEPS program is likely to have reduced false positive over-referral using the existing referral criteria of <6/9, and therefore is a positive change. Consideration should be given to whether the 6/18 threshold for high priority referral remains sufficient to capture all children with high-risk ocular conditions and whether a significant inter-ocular difference in visual acuity should be included as an additional referral criterion.

Chapter 8
Conclusions and
Future Directions

8.1 Conclusions

Current recommendations indicate that vision screening should be conducted for preschool aged children.[27,35,321] Vision screening at this age ensures early detection and timely management of ocular conditions, particularly amblyopia, whose treatment is time-sensitive to the period of neural plasticity and prompt treatment as early as possible is most effective for optimal treatment outcomes.[13,80,81,183] Detection of amblyogenic risk factors including, refractive error and strabismus is also appropriate to prevent the development of amblyopia. Untreated childhood visual impairment has the potential to impact educational outcomes and untreated amblyopia poses a risk of permanent vision loss later in life.[26] The NSW StEPS program has been shown to be an effective model for obtaining a high screening rate in the preschool population as well as a cost-effective screening program.[207] However, there are no universally accepted protocols for vision screening and there is substantial variation in screening programs both within Australia and internationally. This thesis has addressed a number of research questions related to optimal vision screening approaches, including greater understanding of the natural history of hyperopia that could be utilised to develop or refine current protocols.

Chapter 3 "Reduced Vision in Childhood" demonstrated that vision screening in childhood is necessary to detect and manage the significant proportion of children with prevalent reduced vision. In agreement with the currently targeted ocular conditions by preschool vision screening programs, causes of reduced vision in preschool and at early school age in a population-based sample where shown to be refractive errors, followed proportionally by amblyopia and strabismus. This provides further support for targeting preschool aged children to ensure effective treatment of amblyopia, correction of refractive errors prior to school entry and for early onset myopia, early intervention to slow progression to high myopia with age.

The prevalence of reduced vision increased with age in our analysis, with a significant proportion of later-onset reduced vision, aligning with increases in the occurrence of school myopia through adolescence. However, most children with refractive errors in the older age-groups had previously been prescribed refractive correction, which when worn, enabled good vision. This suggests that older children may report symptoms of myopia more readily

than younger children, leading to self-directed treatment for this condition. To promote the detection and correction of myopia through school years, it may be beneficial to perform targeted screening at 12 years of age for high risk populations and to educate children and their parents on eye health to improve reporting of symptoms and access of community eye health services.

Chapter 4 "The Detection of Refractive Errors in Young Children and Factors Impacting Accuracy" examined the distribution of refractive errors in infants and young children and the impact of refraction methods on the accuracy of refractive measures. Two factors were identified that negatively shifted refractive measures towards myopia, the use of the Retinomax and reduced efficacy of cycloplegia in children with dark coloured irides. Our analysis suggested that there is a true iris colour effect on refraction, as there was no significant difference in ocular biometric measures with iris colour. A trend towards more myopic refractive errors in children with darker irides was also demonstrated in the European Caucasian children, suggesting this was not an effect of ethnicity, as has been previously suggested.[398] Thus, a rigorous protocol for cycloplegia is necessary for accurate diagnosis of refractive errors, to prevent overestimation of myopia and underestimation of hyperopia.

The mean refraction through childhood remained hyperopic, with an initial reduction after 6-12 months of age, followed by a plateauing of refractive status, while further gradual reduction in hyperopia can be expected with passive axial elongation through childhood. Significant hyperopia in children is poorly detected by visual acuity measures and cycloplegic refraction is often necessary for diagnosis. This has led to some debate as to whether detection of hyperopia is a priority for vision screening although, even though hyperopia is a risk factor for amblyopia[94] and strabismus[415] suggesting it is an important condition to detect. In addition, some concern has been raised that refractive correction may interrupt normal emmetropisation for children with hyperopia. In **Chapter 5** "Longitudinal Change in Refraction and Axial Length in Children with Hyperopia" it was found that most children with significant hyperopia remained hyperopic through adolescence. This was particularly the case for children with high hyperopia at baseline. Refractive correction of hyperopia did not appear to interfere with the normal process of emmetropisation. As such, our findings

suggest that provision of refractive correction for children with significant hyperopia is not harmful and is likely to be beneficial to reduce strain on accommodative systems, impacts on learning and lessen the risk of amblyopia and strabismus. This means the detection of hyperopia should be a consideration for vision screening and non-invasive and screening-appropriate methods warrant investigation.

Chapter 6 "The Efficacy of Vision Screening Protocols in Preschool and School-aged Children" aimed to examine whether accuracy of vision screening was compromised at preschool age compared to school-age screening and further, whether additional orthoptic testing could improve detection of hyperopia and other amblyogenic risk factors that may be missed on visual acuity testing. This chapter found that vision screening at age four did not compromise the accuracy of visual acuity test results obtained compared to screening at six years of age. This provides further evidence that there is no need to delay vision screening to school age, particularly with the consequential sacrifice to amblyopia treatment effectiveness and loss of other benefits from optimising visual outcomes through early intervention.

There was good sensitivity and specificity for the detection of amblyopia and myopic refractive errors with a visual acuity threshold of 6/9.5 in the four year old children in this analysis, consistent with the StEPS program referral criteria.[290] A very small percentage of children with ocular conditions were identified through additional tests, with the majority being captured by visual acuity testing alone. Although, near visual acuity is often suggested as a screening test for hyperopia,[318] there were no additional cases of hyperopia detected by near visual acuity, indicating that children are able to clear their vision through accommodation even at this close distance in the presence of significant degrees of hyperopia. As only a small number of additional cases of strabismus were detected by stereopsis testing, there is unlikely to be sufficient benefit of the inclusion of stereopsis in vision screening programs, and a protocol of ocular observation for signs of obvious ocular pathology and strabismus would be more appropriate. It is also important to note that obvious conditions such as large angle constant strabismus and overt ocular pathologies are often detected through family observation and may be addressed prior to vision screening.[358]

Chapter 7 "The Preschool Vision Screening Study (PVSS)" investigated the comparability of referral rates using two visual acuity charts, the Sheridan Gardiner Linear and the HOTV LogMAR charts in 94 preschool children recruited through the NSW StEPS program. These visual acuity charts had not been previously compared and as the StEPS program had recently transitioned to using HOTV logMAR it became pertinent to examine the potential impact on the appropriateness of referral criteria. Findings from the PVSS indicated that current StEPS referral criteria remained appropriate for screening using the HOTV logMAR. However, a higher median visual acuity using the HOTV logMAR is likely to reduce referrals from the StEPS program. The HOTV logMAR visual acuity test had the additional advantage of being more testable, likely due to the logarithmic progression of optotype size and the use of only four letters compared to the seven used in the original Sheridan Gardiner and this has the potential to reduce false positive outcomes on vision screening. The analysis in this chapter also suggests that a two line or greater intraocular difference in visual acuity should be included in the StEPS referral criteria to ensure mild cases of amblyopia do not go undetected. Although, this would require an adjustment to screening protocols to ensure visual acuity is tested to minimum threshold rather than ceasing at the 6/9 threshold for referral.

The final aim of chapter 7 was to identify common barriers to acting on referral from preschool vision screening. While only a small number of children were referred from the PVSS, there was a substantial loss to follow up in this group. Interviews with those who did not act on referral revealed that family problems, forgetting appointments and not perceiving there to be a problem were the most common reasons for non-attendance of follow-up appointments after referral from StEPS. These findings were consistent with previous barriers reported in the literature,[333-338,344,346,347] however, further investigation in a larger population of referred children would be appropriate. Implementing measures to overcome barriers to increase the number of children who do attend follow-up appointments would substantially increase the success of vision screening programs. Possible measures that could be taken include education on the importance of vision and treatment of ocular conditions, improving convenience of access and reducing financial barriers to seeking eye health care.

In summary, the main findings of this thesis are that:

1. Screening at preschool age is ideal for detecting amblyopia and early refractive errors, particularly myopia and accuracy of visual acuity testing at this age is comparable with young schoolchildren.

2. Inclusion of additional tests in screening protocols did not significantly improve detection rates for ocular conditions including for hyperopia and strabismus.

3. Myopia onset in adolescence increases the prevalence of reduced vision and targeted repeat screening at 12 years of age along with community education may be appropriate to increase detection and treatment.

4. A stringent cycloplegia protocol is necessary for accurate measures of refractive error, particularly for children with dark coloured irides.

5. The Retinomax shifts refractive measures in a myopic direction and may not be appropriate for accurate diagnosis of refractive error.

6. Children who are significantly hyperopic in childhood commonly remain hyperopic into adolescence and would benefit from refractive correction, as this did not interfere with normal emmetropisation.

7. The HOTV logMAR visual acuity chart was more testable and produced higher overall visual acuities and lower referral rates compared to the Sheridan Gardiner chart.

8. The current StEPS referral thresholds remain appropriate for use with the HOTV logMAR. Although, consideration should be given to reducing the 'high priority' threshold as children who were previously captured within this category are less likely to be using the HOTV logMAR.

9. A revision to the StEPS referral criteria to include an interocular difference of two or more visual acuity lines should be included to avoid missed cases of amblyopia.

10. Barriers to follow-up after referral from the StEPS program were similar to previous studies in our small sample and automated electronic reminders for appointments, improving access to cost-neutral tertiary care and education on the importance of eye health may assist in improving follow-up rates from screening.

8.2 Future Directions

There are several future directions for research, extending from the findings of this thesis. The comparability of the HOTV logMAR and Sheridan Gardiner vision chart was undertaken with examination of mean visual acuity and proportion of referrals based on the StEPS thresholds. However, as a comprehensive ocular examination was not conducted as part of the PVSS, we were unable to compare sensitivity and specificity of these two tests and this would be an appropriate avenue for further investigation. Further, as we had a low rate of children with poor vision in our sample, we were unable to form conclusions about the impact of the additional 6/15 visual acuity line between 6/18 and 6/12 on the HOTV logMAR chart on the high priority referral rate in StEPS. This is important to determine, as it would be unfortunate if children with poor vision and severe ocular conditions missed out on the high priority referral pathway that prioritises access to paediatric ophthalmology though public hospitals. As a result of the transition to HOTV logMAR chart an evaluation of high priority referral outcomes is warranted, particularly as these preliminary results in this study suggest that there will be some impact on high priority referrals from our comparison, given the significantly better mean visual acuity using the HOTV logMAR chart.

Our findings showed that visual acuity testing was accurate for the detection of amblyopia and myopia but, less so for hyperopia and that there is some impetus to improve detection and correction of significant hyperopia. The gold standard method of diagnosis is through cycloplegic refraction, although this is not appropriate in a screening context as it is invasive and requires technical expertise and safe use of medications. As the additional tests utilised in this thesis were unable to increase detection of hyperopia, further research should investigate alternative options. One possibility is use of ocular biometry to determine axial length to corneal radius, which is non-invasive and has been shown to correlate highly with cycloplegic refraction in previous studies.[286-289] Currently equipment to measure ocular biometry such as the IOLMaster and Lenstar, is not portable, restricting it's applicability to a community screening program like StEPS. However, the IOLMaster has been found to have moderate to high testability in children older than three years, making it appropriate for use in four year olds. [232,282,283] If a portable non-invasive ocular biometer could be designed specifically for screening purposes, this would go some way in addressing the issue of hyperopia detection. For diagnostic refractive error measures it would be beneficial to

determine appropriate cycloplegia regime, specifically for children with dark coloured irides to ensure they are adequately cyclopleged and cannot mask any hyperopia through use of residual accommodation.

Failure to act after referral from screening can compromise the efficacy of screening programs. The evaluation of the StEPS program showed a significant proportion of loss to follow-up that was particularly of concern in rural and regional areas of NSW. We have identified some barriers to follow-up but, our sample was not of a sufficient size to draw definitive conclusions, having been curtailed by COVID 19 health restrictions. Although, there have been other reports on barriers to follow-up from screening, these have been predominantly from the United States[334-338,344] and it is important to note that Australia has the advantage of public hospital services and Medicare-covered optometry services.[395] In this context, it would be beneficial to conduct larger scale research on the barriers to acting on referrals from the StEPS program and to examine differences between metropolitan, regional and rural areas, enabling implementation of solutions tailored to the requirements of different local health districts and Australia. There is likely to have been further disruption due to the COVID-19 pandemic of the StEPS program which ceased during most of 2020 and examining access to care in this context and effective methods of improving access through telehealth may have ongoing benefit particularly within rural and regional areas.

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Appendices

Appendix 1a: PVSS Participation Information and Consent Form

The study is being conducted within this institution by:

Professor Kathryn Rose
Kathryn.Rose@uts.edu.au
+61 2 9514 9222

Dr Amanda French
Amanda.French@uts.edu.au
+61 2 9514 7238

Miss Mythili Ilango
Mythili.Ilango@student.uts.edu.au
+61 2 9514 4124

**Discipline of Orthoptics,
Graduate School of Health,
University of Technology Sydney**

Vision Screening Methods in Preschool Children
[UTS HREC ETH18-2642]

INFORMATION FOR PARENTS/GUARDIANS
This information sheet is for you to keep.

What is this study about?

Your child has been invited to participate in a study that involves comparing different vision screening methods. Vision screening is important for preschool children as the early detection and management of eye disorders (pathology, strabismus/squint eye, refractive error, amblyopia/reduced vision without a cause) are crucial in order to develop and maintain a good level of vision. Many eye disorders go undetected as they cannot be detected by behavioural changes and observation alone. Monocular vision testing (one eye at a time) is important in detecting such conditions. If children with or at risk of eye disorders are not detected within a certain age period, their underlying condition may worsen or affect them to a greater extent and their visual development may be compromised.

What will the study involve?

If you agree to participate in this study, you will first be asked to complete a questionnaire about demographic information and family history of any childhood eye conditions. Contact details of parents/guardians will also be requested in this questionnaire.

Following this, a vision screening test will be conducted on your child at their preschool. This will involve a vision test as well as a check for any eye turns (squint eye). No eye drops or direct contact with the eyes will be made during this process. If any abnormalities are detected, your child will be referred to an ophthalmologist through the New South Wales Statewide Eyesight Preschooler Screening (StEPS) program for further ocular examination.

After one month of testing, if your child was detected as having or at a risk of a particular eye condition, you will be contacted via email or phone to obtain information about your appointment referral status. Following your child's appointment, their practitioners will be contacted to obtain information on their diagnosis with your consent and the practitioner's consent. Practitioners will be provided details of the research prior to obtaining their consent.

Are there any risks/ inconvenience?

There are no particular risks for this study. The vision screening assessment will involve no contact with the eyes. However, vision will be tested one eye at a time and this involves wearing occluder glasses frames with one eye covered during the process.

What will happen if I say no?

Participation in this study is entirely voluntary. It is completely up to you whether or not you decide to take part. If you decide not to participate in the research, your child will still have access to vision screening as part of the StEPS program. If you decide not to participate, it will not affect your relationship with the researchers or the University of Technology Sydney.

If you choose to participate, you can withdraw at any time without having to give a reason. If you wish to withdraw from the study you can do so by contacting Mythili Ilango or Dr Amanda French (contact details below). However, it may not be possible to withdraw your data from the study results if these have already had your identifying details removed.

Confidentiality

All the information collected from you for the study will be treated confidentially, and only the researchers named above will have access to it. For the study, your child will be assigned an ID number for the purpose of deidentification. The study results may be presented at a conference or in a scientific publication. In any publication, information will be provided in such a way that you cannot be individually identified.

What if I have concerns or a complaint?

When you have read this information, we will discuss the study with you and answer any questions you may have. If you would like to know more information at any stage or you have any concerns that you think we can help you with, please feel free to contact Mythili Ilango on 9514 4124 or Mythili.Ilango@student.uts.edu.au or Dr Amanda French on 9514 7238 or Amanda.French@uts.edu.au.

You will be given a copy of this form to keep.

NOTE:
This study has been approved by the University of Technology Sydney Human Research Ethics Committee [**UTS HREC ETH18-2642**]. If you have any concerns or complaints about any aspect of the conduct of this research, please contact the Ethics Secretariat on ph: +61 2 9514 2478 or email: Research.Ethics@uts.edu.au, and quote the UTS HREC reference number. Any matter raised will be treated confidentially, investigated and you will be informed of the outcome.

UTS UNIVERSITY
OF TECHNOLOGY
SYDNEY

CONSENT FORM
Vision Screening Methods in Preschool Children
[UTS HREC ETH18-2642]

I _____ agree to allow my child _____ to participate in the research project Vision Screening Methods for Preschool Children [UTS HREC ETH18-2642] being conducted by researchers from the Discipline of Orthoptics, Graduate School of Health.

I have read the Participant Information Sheet or someone has read it to me in a language that I understand.

I understand the purposes, procedures and risks of the research as described in the Participant Information Sheet.

I have had an opportunity to ask questions and I am satisfied with the answers I have received.

I freely agree to participate in this research project as described and understand that I am free to withdraw at any time without affecting my relationship with the researchers or the University of Technology Sydney.

I understand that I will be given a signed copy of this document to keep.

I agree:
☐ for my child to have a vision screen and for their results to be used in this study
☐ to be contacted by the researchers for follow-up information
☐ for my child's practitioner to be contacted for follow-up information

I agree that the research data gathered from this project may be published in a form that:
☐ Does not identify me in any way
☐ May be used for future research purposes

I am aware that I can contact Mythili Ilango on 9514 4124 or Mythili.Ilango@student.uts.edu.au or Dr Amanda French on 9514 7238 or Amanda.French@uts.edu.au if I have any concerns about the research.

_____   ____/____/____
Name and Signature [participant]   Date

_____   ____/____/____
Name and Signature [researcher or delegate] Date

Appendix 1b: PVSS Demographics Questionnaire

Study Research ID:

UTS UNIVERSITY OF TECHNOLOGY SYDNEY

Name of Parent/ Guardian _____

Relationship to child _____

Mobile Number _____

Home Phone Number _____

Address _____

Postcode _____

Email Address _____

Another Contact _____

Relationship to child _____

Mobile Number _____

Home Phone Number _____

Address _____

Postcode _____

Email Address _____

Name of Child _____

Gender ☐ Male
☐ Female
☐ Other

Date of Birth _____/_____/_____

UTS UNIVERSITY OF TECHNOLOGY SYDNEY

Child's Ethnic Background
☐ European Caucasian
☐ East Asian
☐ South Asian
☐ Middle Eastern
☐ Aboriginal/Torres Strait Islander
☐ Other _____

Do you have any concerns regarding your child's eyes?   ☐ Yes ☐ No

If yes, what are your concerns?

_____

_____

_____

Is your child under the care of an eye health professional?   ☐ Yes ☐ No

Contact Details for your child's eye health professional

_____

_____

_____

If yes, for what condition?

_____

Does your child currently wear glasses?   ☐ Yes ☐ No

Has your child been prescribed patching treatment?   ☐ Yes ☐ No

Has anyone in the child's immediate family been prescribed glasses during childhood?
☐ Yes ☐ No

Has anyone in the child's immediate family been prescribed patching treatment during childhood?
☐ Yes ☐ No

Do you have a family history of any other childhood eye problems?
☐ Yes ☐ No
If yes, who and what childhood eye problems?

_____

_____

_____

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Appendix 1c: PVSS Preschool Information and Consent Form

The study is being conducted within this institution by:

Professor Kathryn Rose
Kathryn.Rose@uts.edu.au
+61 2 9514 9222

Dr Amanda French
Amanda.French@uts.edu.au
+61 2 9514 7238

Miss Mythili Ilango
Mythili.Ilango@student.uts.edu.au
+61 2 9514 4124

**Discipline of Orthoptics,
Graduate School of Health,
University of Technology Sydney**

Vision Screening Methods in Preschool Children
[UTS HREC ETH18-2642]

INFORMATION FOR PRESCHOOLS
This information sheet is for you to keep.

What is this study about?

Your preschool has been invited to participate in a study that involves comparing different vision screening methods. Vision screening is important for preschool children as the early detection and management of eye disorders (pathology, strabismus/squint eye, refractive error, amblyopia/reduced vision without a cause) are crucial in order to develop and maintain a good level of vision. Many eye disorders go undetected as they cannot be detected by behavioural changes and observation alone. Monocular vision testing (one eye at a time) is important in detecting such conditions. If children with or at risk of eye disorders are not detected within a certain age period, their underlying condition may worsen or affect them to a greater extent and their visual development may be compromised.

What will the study involve?

If you agree to participate in this study, parents of children at your preschool will first be asked to complete a questionnaire about demographic information and family history of any childhood eye conditions. Contact details of parents/guardians will also be requested in this questionnaire.

Following this, four year old children at your preschool will receive a vision screening test. This will require a 3 metre room at your preschool, free from distractions. A vision test as well as a check for any eye turns (squint eye) will be conducted. No eye drops or direct contact with the eyes will be made during this process. If any abnormalities are detected, the child will be referred to an ophthalmologist through the New South Wales Statewide Eyesight Preschooler Screening (StEPS) program for further ocular examination.

After one month of testing, if a child at your preschool was detected as having or at a risk of a particular eye condition, their parents will be contacted via email or phone to obtain information about your appointment referral status. Following the child's appointment and with parental consent their practitioners will be contacted to obtain information on their diagnosis.

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Are there any risks/ inconvenience?

There are no particular risks for this study. The vision screening assessment will involve no contact with the eyes. However, vision will be tested one eye at a time and this involves wearing occluder glasses frames with one eye covered during the process.

What will happen if I say no?

Participation in this study is entirely voluntary. It is completely up to you whether or not you decide to take part. If you decide not to participate, it will not affect your relationship with the researchers or the University of Technology Sydney and they will still be screened through the StEPS screening program.

If you choose to participate, you can withdraw at any time without having to give a reason. If you wish to withdraw from the study you can do so by contacting Mythili Ilango or Dr Amanda French (contact details below). However, it may not be possible to withdraw your data from the study results if these have already had your identifying details removed.

Confidentiality

All the information collected from your preschool for the study will be treated confidentially, and only the researchers named above will have access to it. For the study, each child will be assigned an ID number for the purpose of deidentification. The study results may be presented at a conference or in a scientific publication. In any publication, information will be provided in such a way that the child and the preschool cannot be individually identified.

What if I have concerns or a complaint?

When you have read this information, we will discuss the study with you and answer any questions you may have. If you would like to know more information at any stage or you have any concerns that you think we can help you with, please feel free to contact Mythili Ilango on 9514 4124 or Mythili.Ilango@student.uts.edu.au or Dr Amanda French on 9514 7238 or Amanda.French@uts.edu.au.

You will be given a copy of this form to keep.

NOTE:
This study has been approved by the University of Technology Sydney Human Research Ethics Committee [**UTS HREC ETH18-2642**]. If you have any concerns or complaints about any aspect of the conduct of this research, please contact the Ethics Secretariat on ph: +61 2 9514 2478 or email: Research.Ethics@uts.edu.au, and quote the UTS HREC reference number. Any matter raised will be treated confidentially, investigated and you will be informed of the outcome.

CONSENT FORM FOR PRESCHOOLS
Vision Screening Methods in Preschool Children
[UTS HREC ETH18-2642]

Blackfriar's Children's Centre agrees to participate in the research project Vision Screening Methods for Preschool Children [UTS HREC ETH18-2642] being conducted by researchers from the Discipline of Orthoptics, Graduate School of Health.

I have read the Information Sheet or someone has read it to me in a language that I understand.

I understand the purposes, procedures and risks of the research as described in the Information Sheet.

I have had an opportunity to ask questions and I am satisfied with the answers I have received.

I freely agree to participate in this research project as described and understand that I am free to withdraw at any time without affecting my relationship with the researchers or the University of Technology Sydney.

I understand that I will be given a signed copy of this document to keep.

I am aware that I can contact Mythili Ilango on 9514 4124 or Mythili.Ilango@student.uts.edu.au or Dr Amanda French on 9514 7238 or Amanda.French@uts.edu.au if I have any concerns about the research.

_____   ____/____/____
Name and Signature [director]   Date

_____   ____/____/____
Name and Signature [researcher or delegate]   Date

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Appendix 2a: StEPS Protocol

Policy Directive

NSW GOVERNMENT | Health

Statewide Eyesight Preschooler Screening (StEPS) Program

Summary	Detailed requirements for the consistent management and implementation of the StEPS program in LHDs. The policy contains procedures associated with the roles and responsibilities of StEPS personnel including training requirements, identifying four year old children for vision screening, vision screening protocols, referral pathways and reporting requirements.
Document type	Policy Directive
Document number	PD2018_015
Publication date	22 May 2018
Author branch	Health and Social Policy
Branch contact	(02) 9424 5944
Replaces	PD2012_001
Review date	22 May 2023
Policy manual	Patient Matters Manual for Public Health Organisations
File number	16/3507
Status	Active
Functional group	Clinical/Patient Services - Baby and Child, Governance and Service Delivery
Applies to	Community Health Centres, Local Health Districts, Specialty Network Governed Statutory Health Corporations
Distributed to	Divisions of General Practice, Ministry of Health, Public Health System
Audience	Administration, StEPS Coordinators, LHDs, Specialty Network Governed Statutory Health Corporation, Child and Family Health Nurses, GPs

STATEWIDE EYESIGHT PRESCHOOLER SCREENING (StEPS) PROGRAM

PURPOSE

The purpose of this policy directive is to guide StEPS coordinators in the consistent implementation and management of the Statewide Eyesight Preschooler Screening (StEPS) program at the Local Health District (LHD) level.

This policy directive describes the roles and responsibilities of StEPS personnel and training requirements, identifying four year old children for vision screening, vision screening protocols, referral processes and reporting requirements so that childhood vision problems can be detected early and treatment outcomes maximised.

MANDATORY REQUIREMENTS

LHDs must ensure compliance with the requirements set out in this policy directive as the basis for administering the StEPS program in LHDs. Mandatory requirements for the StEPS program are:

- Vision screening protocols relating to consent, vision screening, assessment, referrals, referral follow up, and reporting and data management (Section 2).
- All four year old children in LHDs, including disadvantaged groups and children with special needs, should be offered the StEPS program, to meet StEPS performance benchmarks (Section 3).
- StEPS vision screening staff must be suitably trained and provided with the necessary equipment and resources to conduct vision screening (Sections 4 and 5).
- All standardised templates attached to this policy are used by LHDs when administering the StEPS program (Section 7).
- LHDs must develop operating processes consistent with this policy directive, to maximise screening and meet local needs in each LHD.

IMPLEMENTATION

The Ministry of Health provides funding to assist LHDs in the implementation of the StEPS program in NSW. This policy directive applies to all staff and relevant managers involved in delivering the StEPS program in LHDs across NSW.

Roles and Responsibilities

Ministry:

- Provide mandatory requirements and guidelines for the implementation and management of the StEPS program.

- Evaluate the overall efficiency and performance management of the StEPS program in LHDs across NSW.

- Meet regularly with all LHDs through the StEPS Coordinators Meetings to review overall progress and implementation of the StEPS program in LHDs.

- Ensure the content of this StEPS policy directive is effectively communicated to all staff involved in coordinating the StEPS program in NSW.

LHDs:

- Actively identify all four year old children in their LHDs to offer them a free StEPS vision screen.

- Assign responsibility and personnel to implement the StEPS program in line with this policy directive.

- Ensure appropriate vision screening staff are employed, that vision screening staff are trained to undertake the StEPS vision screen, and staff are provided with appropriate equipment and resources to carry out the functions of the StEPS program.

- Ensure compliance and full implementation of this policy directive in their LHD.

- Ensure that the budget provided for the StEPS program is expended on implementing the StEPS program.

- Provide all required reports to the Ministry of Health relating to screening activity, referrals, assessments, follow ups, monitoring and reporting.

- ensure that StEPS performance benchmarks are achieved and maintained (Section 3.3)

- Ensure the content of this StEPS policy directive is effectively communicated to all staff involved in implementing the StEPS program in the LHD.

REVISION HISTORY

Version	Approved by	Amendment notes
May 2018 (PD2018_015)	Deputy Secretary, Strategy and Resources	Revision and update of StEPS policy directive, including updates to procedures, privacy information and correspondence to parents
January 2012 (PD2012_001)	Deputy Director-General Population Health	New document outlining the policy directive for the StEPS program

ATTACHMENTS

1. Statewide Eyesight Preschooler Screening (StEPS) Program: Procedures.

Statewide Eyesight Preschooler Screening (StEPS) Program

NSW GOVERNMENT | **Health**
PROCEDURES

CONTENTS

1 BACKGROUND

The Statewide Eyesight Preschooler Screening (StEPS) program is a universal, evidence based, free vision screening program for all four year old children in NSW.

The program actively identifies all four year old children in NSW to offer them a free StEPS vision screen and is designed to identify childhood vision problems early, prior to school entry, so that treatment outcomes can be maximised.

The StEPS program is an important component of the NSW Child Health Screening and Surveillance Program, as documented in the NSW Personal Health Record (PHR), the 'Blue Book'. The NSW PHR recommends a vision examination at the newborn health check, vision surveillance at the 1-4 week, 6-8 week, 6 month, 12 month, 18 month, 2 year and 3 year child health checks, and a monocular visual acuity screen at the 4 year child health check.

Vision develops from birth to approximately eight years of age, and is fully mature by the mid-teenage years. Early identification and treatment of eye and vision problems aims to optimise vision prior to starting school and reduces the likelihood of permanent vision loss. The StEPS program targets children at four years of age, the first opportunity for a child's visual acuity to be reliably screened at a population level.

While eye health surveillance can monitor a child for outward signs of eye or vision problems, the two most common childhood vision problems, amblyopia and refractive error, cannot be detected by family history, vision surveillance or observing a child's behaviour or appearance. These vision disorders can only be detected if a monocular visual acuity screen is conducted by a trained vision screener.

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2 VISION SCREENING PROTOCOLS

2.1 StEPS Referral Pathway Flowchart

The figure below outlines the StEPS Referral Pathway:

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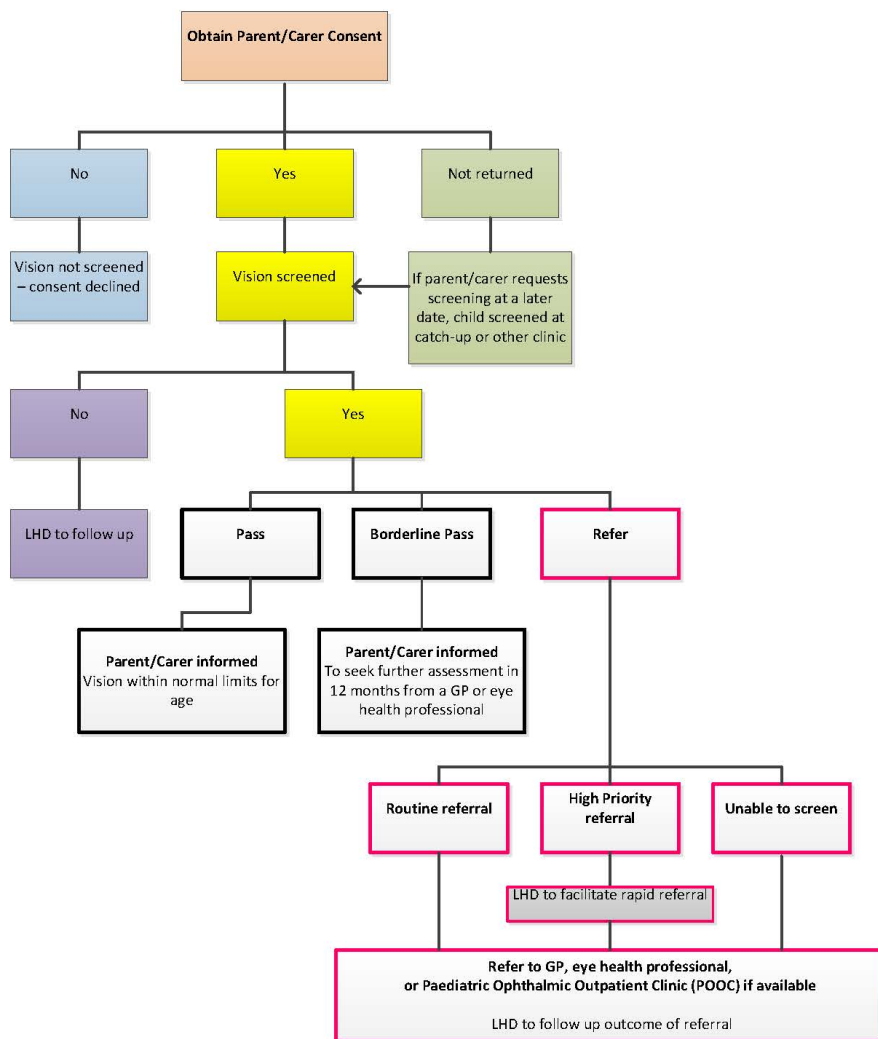
2.2 Pathway for screening, referral, assessment and follow up

LHDs must have clearly documented protocols, consistent with this StEPS Policy Directive for approaching services to offer the StEPS vision screening program, offering parents/carers the StEPS vision screen for their child, the provision of the vision screening service, documenting the outcome of the vision screen, informing parent/carers of the outcome of the vision screen, and for referral and follow up of referrals.

Standardised templates attached to this policy (Section 7) must be used to implement and administer the StEPS program in NSW.

2.3 Consent

Consent from parent/carer for child to participate in the StEPS program at a preschool, child care centre or other service must be obtained prior to undertaking the StEPS vision screen. The following standardised information letter and consent forms are to be used to obtain signed consent:

- *StEPS Important Notice for all Parents/Carers* (Attachment 2).
- *StEPS Consent and Results Form* (Attachment 3).

Consent forms, information letters and flyers about the StEPS program and LHD privacy information should be provided to the preschool/child care centre where the screening will occur at least two weeks prior to the screening date. Completed and signed consent forms must be collected prior to the screening date or on the day of screening. Consent may be accepted by the StEPS vision screener up to and including the day of screening. If verbal consent is provided, this must be documented on the consent form by the vision screener.

If a consent form is returned and the parent/carer has consented to screening, but the child is absent on the day of screening, a follow-up screening offer should be made. At least two vision screening follow up offers should be made (and documented) where consent is obtained but screening is not conducted.

If the consent form is not returned, LHDs should have screening options available for parents/carers who request screening for their child at a later date, such as catch-up and other clinics.

If consent is not provided, that is, the consent form is returned but consent is declined, this must be recorded appropriately.

To monitor and assist in accurately recording the number of StEPS vision screenings offered, it is recommended that LHDs enquire about the number of children at each centre who:

- Are four years of age, or who will be turning four years of age.
- Are eligible to attend school in the following calendar year.

The target group of children for StEPS screening are those aged four years who are starting school the following year. Children who are five years of age and have not previously received a StEPS vision screen are also eligible to be offered the StEPS program. Three year old children who are eligible to start school the following year may be screened at the StEPS coordinator's discretion.

2.4   Vision Screening

LHDs must coordinate and organise the StEPS vision screening with relevant parties at a suitable screening location. Consideration should be given to preschools/childcare centres with specific attendance patterns such as split week attendance to ensure a high uptake of screening and the number of screening days required to screen all children appropriately. StEPS vision screening staff should arrange an appropriate area to conduct the StEPS vision screening in consultation with the preschool/child care centre.

Wherever possible, StEPS vision screening staff must conduct a monocular visual acuity screening test using the approved 6 metre HOTV logMAR chart or Sheridan Gardiner Linear Chart. If the screening location does not have the required space available for the 6 metre chart, the approved 3 metre HOTV logMAR or Sheridan Gardiner Linear Chart can be used. The matching board corresponding to the chart used is to be provided to all children to enable children to match the letter indicated to by the vision screener with the letter on the matching board.

Vision screeners should also review the consent form carefully noting any parental/carer concerns, perform a visual inspection of the eyes and observe the child carefully (for example, does the child constantly close one eye in sunlight, do both eyes move together equally in all direction of gaze, does the child consistently tilt their head or turn their face to one side) to determine if any abnormalities may be present which could affect either the vision or the child's general eye comfort. If there are concerns following visual inspection of the eyes, for example, red eyes, red lid margins, or excessive watering, the child should be referred to their General Practitioner. Vision screeners should carefully observe and refer any possible eye or vision abnormalities even if the visual acuity result is within normal range.

To conduct a monocular visual acuity test, the screener must occlude the left eye first using the recommended occlusion glasses. A folded tissue is placed between the occluded eye and the glasses. If the child already wears glasses, use a single-use eye patch with a tissue between their glasses and the eye patch. LHD infection control procedures must be followed.

The test results for each eye must be accurately recorded by the vision screener on the *StEPS Consent and Results Form* (Attachment 3) and *Notification of StEPS Vision Screening Results Letter* (Attachment 4) as appropriate.

2.5 Documenting Results of the Vision Screening

2.5.1 Consent and Results Form

The vision screener must complete the results section of the *StEPS Consent and Results Form* (Attachment 3) to document the vision screening results.

All sections of the *StEPS Consent and Results Form* must be completed, signed and dated. Relevant actions relating to completing a *StEPS Results Notification Letter* and *StEPS Referral Letter* must be identified on the form. All *StEPS Consent and Results Forms* must be promptly forwarded to the StEPS Coordinator as per LHD procedures.

2.5.2 Notification of StEPS Vision Screening Results Letter

The *Notification of StEPS Vision Screening Results Letter* (Attachment 4) is used to inform parents of the outcome of vision screening and must be completed and forwarded to all parents/carers of children who participated in the StEPS program.

Notification of the vision screening result should be provided as soon as practical, preferably on the day of the screening.

If the parent/carer has indicated on the *StEPS Consent and Results Form* (Attachment 3) that the child is under the care of an eye health professional, the vision screener must advise on the *Notification of StEPS Vision Screening Results Letter* (Attachment 4) for parent/carer to continue care. If there are any concerns about the child's current treatment, vision screeners must discuss this with their StEPS Coordinator. LHDs should encourage parents/carers to add the *Notification of StEPS Vision Screening Results Letter* to their child's Personal Health Record.

All parents are encouraged to ensure their child attends a Before School Health Assessment at 4 years of age, as per the NSW Personal Health Record (Blue Book).

2.5.3 Inclusion of forms and letters in the electronic medical record

If the child's electronic medical record is available, the *StEPS Consent and Results Form* (Attachment 3) is scanned and forms part of the child's electronic medical record. The *StEPS Referral Letter* (Attachment 5) may be scanned and included as correspondence accompanying the child's medical record.

2.5.4 Confidentiality

All information collected and results are confidential and must not be provided to or discussed with others, including staff at the preschool or child care centre, without parent/carer consent. To ensure privacy, all *Notification of StEPS Vision*

Screening Results letters (Attachment 4) are to be placed in a sealed envelope with the child's name on the outside of the envelope. Vision screeners must liaise with relevant parties (e.g. preschool/child care director) at the screening location to determine the most appropriate mechanism for providing the results of the StEPS vision screen to parents/carers.

2.5.5 StEPS Referral Letter

All parents/carers of children who require a referral must be provided with a *StEPS Referral Letter* (Attachment 5). The referral letter may be completed by the vision screener, StEPS Coordinator or Administration Officer as per LHD procedures.

2.6 Referral Criteria

The StEPS program uses pass/refer criteria that correlate to specific, evidence-based visual acuity results. Following the StEPS vision screen, the criteria for making a referral based on the vision screening result are as follows:

a) Pass - visual acuity of 6/9 (3/4.5) or above

- A child with visual acuity of 6/9 (3/4.5) or above in both eyes is considered to have passed the StEPS visual acuity screen.
- Referral is not required.

b) Borderline Pass - visual acuity of 6/9-1 (3/4.5-1) or 6/9-2 (3/4.5-2)

- A child with visual acuity of 6/9-1 (3/4.5-1) or 6/9-2 (3/4.5-2) in one or both eyes is considered a borderline pass.
- Parents/carers are advised to re-test in 12 months by an Eye Health Professional.

c) Refer - visual acuity of less than 6/9-2 (3/4.5-2) in one or both eyes

- A child with visual acuity of less than 6/9-2 (3/4.5-2) in one or both eyes is considered to have not passed the StEPS visual acuity screen.
- Parents/carers are advised to have their child's eyes tested by a General Practitioner or Eye Health Professional.

d) Refer - obvious pathology

- A child with obvious pathology on observation of external eye and adnexa that is currently untreated should be referred for review.
- Parents/carers are advised to have their child's eyes reviewed by a General Practitioner.

e) High Priority Referral - visual acuity of 6/18 (3/9) or less in one or both eyes

- A child with visual acuity of 6/18 (3/9) or less in one or both eyes is considered a high priority referral.

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- Parents/carers are advised to have their child's eyes tested by a General Practitioner or Eye Health Professional as a matter of urgency.

- Referral may be made to Paediatric Ophthalmic Outpatient Clinics (POOCs) according to *StEPS Referral Protocols for POOCs* (Attachment 6).

f) Refer - unable to be screened

- A child who has a valid consent but is unable to be screened, for example if they are uncooperative or unable to perform the test, should be referred.

- Parents/carers are advised to follow up with an Eye Health Professional.

2.7 Follow-up of referrals

All referrals from the StEPS Program must be actively followed up by the StEPS Coordinator as per this Policy Directive and LHD procedures. Wherever possible, StEPS Coordinators should ensure that High Priority Referrals receive a diagnostic vision assessment within one month, and other referrals receive an assessment within six months.

StEPS Coordinators are to offer assistance to families to ensure the child receives a diagnostic eye assessment within the appropriate timeframe. This may include, but is not limited to, offering secondary screening Orthoptic services and/or referral to the StEPS Paediatric Ophthalmic Outpatient Clinics (POOCs). StEPS Coordinators should consider any barriers to receiving a diagnostic assessment and subsequent treatment and assist families wherever possible to access appropriate services.

StEPS Coordinators must monitor all follow up referrals and report on the outcomes. If no eye health professional report is received and the outcome is unknown, the parent/carer must be contacted to determine the outcome and the result recorded. If possible, the name of the eye health professional who provided the assessment/treatment should be sought from the parents and the eye health professional then contacted to confirm the outcome.

The *StEPS Referral Outcomes Report* (Attachment 10) must be completed to record the outcome of the referral as a result of the StEPS vision screening. These reports can be used to demonstrate the accuracy of vision screening undertaken and the effectiveness of the StEPS program.

2.8 Mandatory Reporting for the StEPS program

StEPS Coordinators must complete and submit StEPS Screening Activity and StEPS Referral Outcomes reports for the StEPS program to the Ministry. Where an electronic medical record system is available in the LHD, electronic reporting and data extraction files should be submitted as reports to the Ministry. If

electronic medical records are not available, these may be submitted as manual reports using the following templates:

- Quarterly *StEPS Screening Activity Report* (Attachment 9)
- Quarterly *StEPS Referral Outcomes Report* (Attachment 10)

2.9 Data Management

LHDs are responsible for developing and maintaining a database to record all children who have participated in the StEPS program. This will enable ease of scheduling, screening, tracking referrals, follow up referrals, reporting on referral outcomes and responding to enquires from parents/carers on vision screening.

All children who have participated in the StEPS program must be recorded on a database developed and maintained by LHDs. This database must include client/patient identifying details and parents contact details in accordance with PD2007_094 Client Registration Policy', as well as screening location, date of screening, result of screening, follow up of referrals, and the outcome and diagnosis following referral where applicable. It is recommended that terminology used to record the outcomes of referrals is consistent with language used in the referral outcomes report.

Where an electronic medical record system is available in the LHD, the appropriate electronic documentation for StEPS should be completed and data extraction files submitted as reports to the Ministry of Health.

2.9.1 Retention and Disposal of StEPS patient/client records

For all children who receive a StEPS vision screen, the *StEPS Consent and Results Form* (Attachment 3) must be incorporated into the main Community Health client record system and retained until the child attains or would have attained the age of 25 years. This applies to children who are found to have no abnormality on screening, as well as those children who receive a borderline pass or are referred for any reason.

Where the StEPS Consent and Result Form is in paper format and is not imaged or scanned, the original paper form must be retained for 25 years. It can then be disposed of according to LHD procedures.

Where the StEPS Consent and Result Form is imaged or scanned, the original Form should be retained until it has been verified that the scanned copy clearly displays all elements of the original record, as per NSW State Records 'General Retention and Disposal Authority – Public Health Services: Patient/Client Records' (GDA 17). Once verified, the paper Form can then be disposed of according to LHD procedures. The imaged Form must be retained for 25 years.

3   IDENTIFYING FOUR YEAR OLD CHILDREN

3.1   Identifying Four Year Old Children

All four year old children in NSW are to be actively identified to be offered a free StEPS monocular visual acuity screen by StEPS Coordinators within their designated LHDs. Strategies to identify four year old children may include, but are not limited to, contacting the following services to offer the StEPS program:

- preschools
- child care centres
- family day care services
- early intervention services
- refugee services
- Child and Family Health Services
- playgroups
- immunisation clinics
- Department of Education and Communities, Schools for Specific Purposes
- Community vision screening days
- School Orientation programs (this strategy should only be used where the eligible child was not able to be identified through alternative strategies).

3.2   Disadvantaged groups of children and children with special needs

Disadvantaged groups of children and children with special needs are to be actively identified to ensure they are offered StEPS screening. For the purposes of the StEPS program, the following groups of children are classified as 'disadvantaged groups':

- Aboriginal and Torres Strait Islander children.
- Children attending 'Early Intervention Services'.
- Children attending 'Schools as Community Centres (SACCs) Playgroups'.
- Children whose parents attend Mental Health Services.
- Children in 'Out of Home Care'.
- Refugee children.
- Socioeconomically disadvantaged children.

Children with special needs are children who have been identified with developmental delay and/or neurological deficits.

StEPS Coordinators are to develop local strategies that meet the needs of their LHD in order to ensure maximum vision screening and equity of access to the StEPS program for all four year old children.

3.3 Service Level Agreement

The Service Level Agreement of the StEPS program is:

- A minimum of 80% of eligible four year old children have screening conducted

Wherever a parent/carer completes a StEPS consent form and agrees to their child participating in the StEPS program the LHD must make every effort to ensure that the child's vision is screened according to StEPS protocols.

LHDs are to ensure that the StEPS Service Level Agreement is maintained according to the estimated target population numbers of four year olds in their LHD provided by the Ministry of Health.

4 StEPS PERSONNEL

4.1 Vision Screening Staff

StEPS vision screening staff are employed by LHDs, under the supervision of LHD StEPS Coordinators to conduct monocular visual acuity screening assessments for four year old children.

StEPS vision screening must be conducted by suitably trained staff competent in using the StEPS vision screening equipment to undertake vision screening for four year old children. Screening assessments are undertaken in locations deemed appropriate by LHDs and can include settings such as preschools, child care centres, community settings and Child and Family Health Services.

StEPS vision screening staff are responsible for:

- liaising effectively with preschool and child care centre staff, parents, team members and other health care professionals in a professional and caring manner
- conducting vision screening according to vision screening protocols consistent with this StEPS policy directive relating to obtaining consent, referral processes, appropriate testing set up, vision screening equipment gathering and utilising information as required for effective vision screening
- ensuring the vision screening process creates minimal disruption to the location where screening is undertaken

- ensuring the confidentiality and privacy of the child is maintained at all times and all relevant information about the screening process and vision screening results is provided to parents/carers

- ensuring all mandatory requirements and reporting mechanisms relating to vision screening, consent, referrals processes, notification of results and LHD protocols are undertaken

- adhering to all LHD Work Health and Safety and Infection Control protocols

- maintaining vision screening equipment and reporting malfunctioning equipment to the StEPS Coordinator

- advising the StEPS Coordinator of any issues, incidents, problems or concerns that arise during a vision screening session.

4.2 StEPS Coordinator

StEPS Coordinators are employed by LHDs to implement, coordinate and manage the day to day operations of the StEPS program.

StEPS Coordinators develop and maintain strong links with all relevant stakeholders in their LHD, such as child health services, parents and carers, early childhood education and care providers, eye health professionals, general practitioners, medical specialists, Aboriginal Community Controlled Health Services, early intervention and coordination programs and other government and non-government agencies, to promote the StEPS program and to ensure the StEPS program is delivered effectively in their respective LHDs.

StEPS Coordinators are responsible for:

- ensuring all four year old children in their LHD are actively identified and offered a StEPS vision screen, including providing screening services as required

- recruiting vision screening staff as required, training and/or arranging the training to be provided to StEPS vision screeners by a suitably qualified health professional

- supervision and professional development of StEPS vision screeners to ensure that competency in vision screening is achieved and maintained, and that all applicable LHD protocols are followed

- ensuring transportation is available for StEPS vision screeners to travel to screening locations, according to resources available in the LHD. This may include access to a motor vehicle or approval to use private vehicles with the provision of a mileage allowance according to LHD protocols

- ensuring all appropriate supplies and maintenance of equipment, relevant forms and promotional material is available to conduct StEPS vision screening

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- maintaining the confidentiality and privacy of the children screened and providing support to parents as appropriate in the period between vision screening and diagnostic assessment

- developing vision screening protocols for screening, referral, assessment and follow up consistent with the StEPS Policy Directive

- developing local processes to ensure disadvantaged groups of children and children with special needs are actively identified for the StEPS program

- data management and monitoring of key performance indicators, vision screening referral rates, referral outcomes, follow up referrals and submitting relevant reports to the NSW Ministry of Health as required.

- setting up and maintaining a database to record information on all four year old children who participated in the StEPS program for quality management

- effectively managing the LHD StEPS budget to ensure the program is implemented efficiently in the LHD including all printing costs relating to information flyers, brochures, letters and forms on the StEPS program

- attending NSW Ministry of Health StEPS Coordinators meetings as required and being the main point of contact for the StEPS program in their LHDs

4.3 StEPS Administration Officer

StEPS Administration Officers are employed by LHDs to provide administrative duties as deemed appropriate by the StEPS Co-ordinator. Duties may include, but are not limited to, arranging and confirming vision screening bookings, organising consent form packages, StEPS data entry and general office tasks.

4.4 Orthoptist

Orthoptists may be employed to provide comprehensive secondary vision screening for children referred via the StEPS program. Orthoptists may also provide vision screening services for children identified with 'special needs' and undertake additional vision screening tests considered appropriate to a child's individual developmental level. Orthoptists may also investigate and diagnose ocular motility disorders and assist in transitioning the family to timely diagnostic assessment services where appropriate.

Orthoptists may also assist in the training of vision screening staff.

4.5 StEPS Outpatient Clinics

Dedicated StEPS tertiary Paediatric Ophthalmic Outpatient Clinics (POOCs) have been established for children identified with potentially significant vision loss and referred as a 'High Priority Referral'. POOCs will ensure that such children receive a diagnostic vision assessment in a timely manner so that treatment outcomes can be maximised. Ongoing management and treatment of a child diagnosed with a vision

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problem via POOCs should be at the discretion of the eye health professional in consultation with the parent/carer.

Referrals to POOCs are available from anywhere in NSW. Children can be referred according to *StEPS Referral Protocols for Paediatric Ophthalmology Outpatient Clinics (POOCs)* (Attachment 6). The *StEPS Referral Form for POOCs* is at Attachment 7.

5   TRAINING

5.1   StEPS Training Package

To be certified as competent, vision screening staff must:
- satisfactorily complete modules one and two of the *StEPS Training Package* through the NSW Health Education and Training Institute (HETI).

- complete a minimum of four hours practical experience at a screening location (with the StEPS Coordinator or an Orthoptist if possible).

- be assessed as competent after three months of screening using the *StEPS Competency Checklist for Vision Screeners* (Attachment 8) and annually thereafter.

5.2   Supervision and Professional Development

Following completion of modules one and two, in addition to supervised practical experience, ongoing professional development and mentoring opportunities for vision screeners should be locally arranged by LHDs as appropriate. This may involve opportunities to work with an experienced vision screener for the first three months of vision screening wherever possible; participation in Orthoptic clinics; and/or other professional development opportunities identified by the LHD.

It is the responsibility of the StEPS Coordinator to ensure that all dedicated StEPS vision screening staff, and all LHD staff who undertake StEPS vision screening, are proficient in undertaking a StEPS vision screen prior to being deemed qualified to undertake a StEPS vision screen unsupervised.

Ongoing supervision and performance management of vision screening staff, and other health staff who provide StEPS vision screening, is to be undertaken by LHDs according to LHD protocols. This should include performance reviews of vision screening staff referral rates and where appropriate, actions undertaken to address performance factors and skill development.

6   GLOSSARY OF TERMS

Adnexa
For the purposes of this document, adnexa refers to the appendages of the eye. These include but are not limited to the eyelids, conjunctiva, lacrimal apparatus and orbit.

Amblyopia
Amblyopia is reduced or 'dim' vision in an eye which appears to be normal. It is sometimes called 'Lazy Eye'. This is a serious eye defect which often goes undetected in childhood. If amblyopia is not diagnosed and treated early, the vision in the affected eye may be permanent and cannot be corrected with glasses or surgery.

Eye Health Professional
For the purposes of this document, an *Eye Health Professional* refers to registered ophthalmologists, orthoptists and optometrists.

Refractive Error
A refractive error occurs when the shape of an eye is abnormal or does not bend (or refract) light properly, which results in blurred vision. The three most common refractive errors are myopia (short sightedness), hyperopia (long-sightedness) and astigmatism.

HOTV logMAR chart
A visual acuity screening chart used in the StEPS program. LogMAR charts feature the same number of letters on each line, which progressively reduce in size according to a geometrical progression.

Sheridan Gardiner Linear Chart
A visual acuity screening chart used in the StEPS program. Linear charts feature an increasing number of letters on each line, which linearly reduce in size.

Visual Acuity
Visual acuity refers to the measurement of the eye's capacity to see an object, for example a letter on a vision chart, at a certain distance. This measurement is taken one eye at a time with the child wearing their correcting glasses or contact lenses (when needed). It is usually recorded in a format that compares the child's vision results to a certain standard.

Visual Acuity Screening
Also referred to as vision screening, this is the testing of visual acuity using pass/fail criteria to a specific standard correlated to an age appropriate level of acceptable vision.

Vision Surveillance
Vision surveillance is defined as the monitoring of vision development for signs of eye or vision problems and includes observation, family history, reported visual behaviours and some vision tests, e.g. corneal reflections, ocular movements and response to occlusion.

7   LIST OF ATTACHMENTS

1. Implementation Checklist
2. StEPS Important Notice for all Parents/Carers
3. StEPS Consent and Results Form
4. Notification of StEPS Vision Screening Results letter
5. StEPS Referral Letter
6. StEPS Referral Protocols for Paediatric Ophthalmology Outpatient Clinics
7. StEPS Referral Form for Paediatric Ophthalmology Outpatient Clinics
8. Competency Checklist for Vision Screeners
9. StEPS Screening Activity report
10. StEPS Referral Outcomes report

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7.1 Attachment 1: Implementation checklist

LHD/Facility:				
Assessed by:			**Date of Assessment:**	
IMPLEMENTATION REQUIREMENTS	**Not commenced**	**Partial compliance**	**Full compliance**	
1. All StEPS, Child and Family Health, and other relevant NSW Health staff are informed about this policy.	❏	❏	❏	
	Notes:			
2. LHDs develop and implement local policies, guidelines and procedures to support this policy.	❏	❏	❏	
	Notes:			
3. LHDs ensure that vision screeners working within StEPS services and any other services delivering StEPS screening have completed the StEPS HETI eLearning Modules and other StEPS training requirements, as outlined in this policy.	❏	❏	❏	
	Notes:			

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7.2   Attachment 2

StEPS Important Notice for all Parents/Carers[1]

Title of Service Provider

Within Local Health District

Local Health District Logo

IMPORTANT NOTICE FOR ALL PARENTS/CARERS OF
4 YEAR OLD CHILDREN

Dear Parent/Carer,

RE: STATEWIDE EYESIGHT PRESCHOOLER SCREENING (StEPS)

The Statewide Eyesight Preschooler Screening (StEPS) program is an initiative of the NSW Ministry of Health and offers all 4 year old children a free vision screening assessment.

- It is **highly recommended** all 4 year old children participate in screening as vision problems may not be detected unless a child's vision is screened by a trained vision screener.

- Your child's vision will be screened one eye at a time and **no drops** will be used.

- All parents/carers of children who have their vision screened through the StEPS program will be informed of the results of their child's vision screening assessment.

- If a possible vision problem is found, parents/carers will receive a letter asking them to have their child's vision fully tested by an eye health professional.

- Please complete and sign the attached consent form and return it to *(name of centre)* as soon as possible so that a trained vision screener can test your child's eyes.

- If your child has already had a StEPS screen, there is no requirement for a second screening.

The StEPS program is for screening purposes only. Screening tests may not always be accurate and sometimes a screening may cause a false alarm or miss a problem. Occasionally a new problem may occur after a child has had their vision screened. For this reason, if you have concerns about your child's eyes, either now or at any time in the future, please have your child's eyes fully tested by an eye health professional.

Any personal health information collected as part of the StEPS program will only be used in accordance with privacy law. If you have any questions regarding the StEPS program, please contact *(contact person)* on *(telephone number)*.

Thank you for participating in the StEPS program.

> **Important**: As well as a StEPS vision screen, all children should have a general health check before they start school. Please make an appointment with your local child and family health nurse or GP. Take your child's NSW Personal Health Record (Blue Book), if they have one, to your health check appointment.

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[1] Suggested wording only. LHDs should maintain basic content but may modify according to local needs.

7.3   Attachment 3

NSW GOVERNMENT | Health

OFFICE USE ONLY		
FAMILY NAME	MRN	
GIVEN NAME	☐ MALE ☐ FEMALE	
D.O.B. _____ / _____ / _____	M.O.	
ADDRESS		

StEPS CONSENT AND RESULT

LOCATION / WARD

COMPLETE ALL DETAILS OR AFFIX PATIENT LABEL HERE

SMR020004

Holes Punched as per AS2828.1: 2012
BINDING MARGIN - NO WRITING

✍ **PARENT / GUARDIAN TO COMPLETE** *(please use black or blue pen)*

Parent / Guardian (relationship to child) Name: _____

Mobile: _____ Home Phone: _____

Address: _____

_____ Postcode: _____

Email: _____

CONSENT FOR VISION SCREENING

I understand that the Statewide Eyesight Preschooler Screening (StEPS) Program is for screening purposes only. Screening tests, checks and examinations can never be 100% accurate. Sometimes a screening may cause a false alarm or miss a problem. Occasionally a new problem may occur after your child has had a screening test. For this reason, if you have concerns about your child's eyes now or at any time in the future, please see an eye health professional.

☐ Yes, I **consent** to have my child's vision screened   Signed: _____ Date / /

☐ No, I **decline** to have my child's vision screened because (please tick below)

☐ already received a screen ☐ already under care ☐ other Signed: _____ Date / /

Verbal consent: Yes ☐ No ☐

Name _____ Designation _____ Signed _____ Date / /

Reason for verbal consent _____

✍ **CHILD'S DETAILS** *(please use black or blue pen)*

Child's Name: _____ Gender: M ☐ F ☐

Date of Birth: _____ Medicare Number: ☐☐☐☐☐☐☐☐☐☐ / ☐

Name of Preschool / Child Care Centre: _____

Days child attends centre (please tick all that apply):   Mo ☐ Tu ☐ We ☐ Th ☐ Fr ☐

If your child attends another centre, please state _____

Days child attends other centre (please tick all that apply):   Mo ☐ Tu ☐ We ☐ Th ☐ Fr ☐

Indigenous Status

Is your child of Aboriginal or Torres Strait Islander origin?

☐ Yes - Aboriginal   ☐ Yes - Torres Strait Islander   ☐ Yes - Both   ☐ Neither   ☐ Unknown

Pre-Screening Questions – *please answer all of the following questions:*

- Are you concerned about your child's vision?   Yes ☐ No ☐

If yes, what are your concerns? _____

- Is your child currently under care for their vision? Details _____ Yes ☐ No ☐
- Does your child have a turned or lazy eye (squint or strabismus)?   Yes ☐ No ☐
- Did anyone in the family have eye problems in childhood?   Yes ☐ No ☐

If yes, please provide details: _____

_____

NH606646 080616

NO WRITING

Page 1 of 2

StEPS CONSENT AND RESULT

SMR020.004

Attachment 3 (Continued)

NSW GOVERNMENT | Health

StEPS CONSENT AND RESULT

	OFFICE USE ONLY	
FAMILY NAME		MRN
GIVEN NAME		☐ MALE ☐ FEMALE
D.O.B. _____ / _____ / _____	M.O.	
ADDRESS		
LOCATION / WARD		
COMPLETE ALL DETAILS OR AFFIX PATIENT LABEL HERE		

VISION SCREENER TO COMPLETE

Location of Screening:

Vision screening chart used:	HOTV ☐	Sheridan Gardiner	☐
Vision screening distance:	6 metres ☐	3 metres	☐
Vision was tested:	Without glasses ☐	With glasses	☐

Visual acuity result: RVA _____   LVA _____

COMMENTS / OBSERVATIONS

_____
_____
_____
_____
_____
_____
_____
_____
_____
_____
_____

RESULT

☐ .....Pass (vision within normal limits for age)

☐ .....Borderline Pass (follow-up by parent/guardian in one year)

☐ .....Referred for further assessment (for general referrals and not other referral types listed below)

☐ .....Referred – High priority referral

☐ .....Referred due to unable to screen/incomplete screen

☐ .....Referred – vision within normal limits but requires referral for other finding

☐ ...Absent on the day of screening

☐ ...Currently under care for vision

NOTIFICATION FORM COMPLETED?   Yes ☐   No ☐   **REFERRAL LETTER COMPLETED?**   Yes ☐   No ☐

Screener's Name:	Signature:	Date:

Consent and Result Form to be forwarded to the StEPS Co-ordinator

FOLLOW-UP BY LHD WITHIN 1 MONTH?   Yes ☐   No ☐   Date of contact   /   /

FOLLOW-UP BY LHD WITHIN 3 MONTHS?   Yes ☐   No ☐   Date of contact   /   /

OUTCOME:

StEPS Co-ordinator:	Signature:

Page 2 of 2   NO WRITING

7.4 Attachment 4

Notification of StEPS Vision Screening Results

> **Important**: All children should have a health check before they start school with their local child and family health nurse or doctor. Please keep this letter in your child's NSW Personal Health Record (Blue Book) and take it to your health check appointment.

Your child_____, DOB _____
had his/her vision screened using a vision chart approved in the StEPS program to conduct visual acuity screening in children. The result of screening is below:

☐ **Pass – vision is within normal limits for age**
   If you have any concerns about your child's eyes at a later date please have their eyes fully tested by an eye health professional

☐ **Vision is borderline upon screening**
   Please ensure your child has his/her eyes retested by an eye health professional in one year. Vision in children continues to develop as the child grows. Younger children may have their vision within normal limits for their age however require additional testing in one year to ensure their vision fully develops to the normal adult level

☐ **Vision requires further assessment**. You will be contacted by letter or telephone

☐ **Your child is under the care of an eye health professional** and this *Local Health District* advises you to continue this care

☐ **Your child's vision was unable to be screened** due to illness or distraction on the day. Please have your child's vision fully assessed by an eye health professional

☐ **Your child was absent on the day of screening**. Please contact _____ on _____ to arrange a time for your child to have their eyesight screened.

Comments:	
If you wish to discuss the above results further, please contact:	
Name:	Telephone number:
Vision Screener:	Signature:

NOTE: The Statewide Eyesight Preschooler Screening (StEPS) program is for screening purposes only. Screening tests, checks and examinations can never be 100% accurate. Sometimes a screening may cause a false alarm or miss a problem. Occasionally a new problem may occur after a child has had their vision screened. For this reason, if you have concerns about your child's eyes, either now or at any time in the future, please have your child's eyes fully tested by an eye health professional.

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7.5 Attachment 5

StEPS Referral Letter

Date:

Dear

Re: _____ DOB:_____ MRN:_____ M / F

Centre:

Following the recent vision screening of your son or daughter by the StEPS program, it is recommended that you have his/her eyes fully tested by an eye health professional as your child may require glasses and/or treatment for reduced vision or an eye muscle imbalance.

You are advised to carry out this recommendation as soon as possible.

Vision screening chart used:	HOTV	☐	Sheridan Gardiner	☐
Vision screening distance:	6 metres	☐	3 metres	☐
Vision was tested:	Without glasses	☐	With glasses	☐

Vision screening result: Right Eye _____ Left Eye _____

Comment:

Further information may be obtained by telephoning or writing to the address below.

Medicare rebates are available for children's vision assessments; however, costs may vary between eye health professionals and eye health services. Your eye health professional or eye health service will be able to provide further information on the costs of their service.

Please take this letter with you when you have your child's eyes fully tested.

A report from your eye health professional would be greatly appreciated, sent to the address below. Alternatively, please ask your eye health professional to complete the attached tear off slip and forward it to the address below.

- -

Results Notification

Child's Name: _____ Date of birth: _____ MRN (if applicable):

Preschool: _____ Date of assessment: _____ Clinic/Provider:

Outcome: Please select **all** relevant categories:

Refractive error	☐	Anisometropia	☐	Emmetropia	☐
Amblyopia	☐	Strabismus	☐	Other (please specify)	☐
Monitor/review	☐	Discharge (no treatment or review required)	☐		

Diagnosis and treatment plan: _____

Send to:_____

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7.6 Attachment 6: Referral Protocols for Paediatric Ophthalmology Outpatient Clinics

Referral Process	Contact
Sydney Children's Hospital at Westmead (SCHW)	
• Only *High Priority* referrals accepted • Clinics are held at the SCHW • Satellite clinics are also held at Mt Druitt and Campbelltown • Parents/carers are to be advised to take a copy of their child's StEPS vision screening results with them when attending the clinic appointment • Parents/ carers are requested to obtain a referral from their GP also • Referral forms are to be completed and forwarded via email to both nominated contacts • SCHW will then contact the parent/guardian via post or phone with appointment details	Referrals for all clinics are to be forwarded to: Lindley Leonard, Senior Orthoptist lindley.leonard@health.nsw.gov.au **and** Louise Brennan, Senior Orthoptist louise.brennan@health.nsw.gov.au **Tel:** (02) 9845 2877
Mt Druitt Satellite Clinic	
• Only *High Priority* referrals accepted • Parents/carers are to be advised to take a copy of their child's StEPS vision screening results with them when attending the clinic appointment • Parents/ carers are requested to obtain a referral from their GP also • Referral forms are to be completed and forwarded via email to both nominated contacts • Mt Druitt Hospital will then contact the parent/guardian via post or phone with appointment details	Referrals for all appointments are to be forwarded to: Jacqueline Gow Administration Supervisor, Ambulatory Care Department jacqueline.gow@helath.nsw.gov.au **and** Lisa Fuller, Administration Officer lisa.fuller@health.nsw.gov.au **Tel:** (02) 9881 1552

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Campbelltown Satellite Clinic	
• Only *High Priority* referrals accepted • Parents/carers are to be advised to take a copy of their child's StEPS vision screening results with them when attending the clinic appointment • Parents/ carers are requested to obtain a referral from their GP also • Referral forms are to be completed and forwarded via **fax to 46344600** • Campbelltown Hospital will then contact the parent/guardian via post or phone with appointment details	**Tel:** (02) 4634 4162

St George Eye Clinic	
• All children can be referred directly to StEPS Paediatric Ophthalmic Outpatient Clinics • Children who did not receive a secondary vision screen or are 'unable to be screened' are requested to book an *'Orthoptic Only'* appointment • All referrals require a referral form to be completed which is to be BOTH emailed and faxed to the nominated contact. • Forms are to be emailed and faxed in bulk once per week to assist staff to collate referrals. • Referral forms should note the type of service required e.g. *High Priority Referral, General Referral* or *Orthoptic Only* • St George Eye Clinic will contact the child's parent/guardian to book an appointment	Kim Marchant, Orthoptist Kim.Marchant@health.nsw.gov.au Tel: (02) 9113 2153 Fax: (02) 9113 2113

Sydney / Sydney Eye Hospital	
• All children can be referred directly to StEPS Paediatric Ophthalmic Outpatient Clinics • Children who did not receive a secondary vision screen or are 'unable to be screened' are advised to book an *'Orthoptic Only'* appointment • All referrals require a referral form to be completed and emailed to the nominated contact • Referral forms should note the type of service required e.g. *High Priority Referral, General Referral* or *Orthoptic Only* • Referrals are requested via email once per week only • Sydney Eye Hospital will contact the child's parent/guardian via letter to book an appointment	Susan Downing, Orthoptist Susan.downing@health.nsw.gov.au Tel: (02) 9382 7060 Fax: (02) 9382 7354

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Statewide Eyesight Preschooler Screening (StEPS) Program

NSW GOVERNMENT | **Health**

PROCEDURES

Sydney Children's Hospital Randwick	
• <u>All</u> children will be reviewed by an Orthoptist <u>and</u> an ophthalmologist or ophthalmic registrar for the first visit • Follow up appointment is with an Orthoptist only • Where possible refer children who have received a secondary vision screen unless a *High Priority* referral • Referral forms are to be completed and forwarded via email to the nominated contact • Sydney Children's Hospital Randwick will then contact the parent/guardian to book an appointment	Danielle Morgan Orthoptist, StEPS Danielle.morgan@health.nsw.gov.au 9382 0657
Bankstown Hospital	
• <u>All</u> children will be reviewed by an Orthoptist <u>and</u> an ophthalmologist or ophthalmic registrar • All referrals require a referral form to be completed and emailed to the nominated contact • Referral forms should note the type of service required i.e. *High Priority Referral, General Referral* • Bankstown Hospital will contact the child's parent/guardian to book an appointment	Karen Pedemont Area Director Orthoptics Service Manager Orthoptics Bankstown Karen.Pedemont@sswahs.nsw.gov.au **Tel:** (02) 9722 7873

7.7 Attachment 7

StEPS Referral Form to Dedicated Paediatric Ophthalmic Outpatient Clinics

To: **From:**

_____

Phone: **Date:**

_____

☐ **High Priority Eye Referral**   ☐ **General Eye Referral**   ☐ **Orthoptic Only**

Screens conducted:

☐ **Primary screen – Date:**_____   ☐ **Secondary screen - Date:** _____

Interpreter Required: ☐ No   ☐ Yes   **Language**: _____

Child's first name: _____   **Child's surname:** _____

MRN:_____

Male ☐   **Female** ☐

Date of Birth: _____

Address: _____

Parent/Carer's Name: _____

Parent/Carer's Contact Number: _____

Medicare Number: _____

Referred by: _____   **Date:** _____

Comments:

_____

_____

Please forward a report on the outcome of the referral to:

StEPS Coordinator

Address:

Email address:

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NSW GOVERNMENT Health

PROCEDURES

7.8 Attachment 8

Statewide Eyesight Preschooler Screening (StEPS) Program

Competency Checklist for Vision Screeners

Screener Name:
Location:
Assessor Name & Position:

Criteria	Outcome – *please tick*	
Liaises effectively with service providers in a professional manner	☐ Competent	☐ Not yet Competent
Liaises with parents/carers in an appropriate manner	☐ Competent	☐ Not yet Competent
Effectively communicates with children using age appropriate language and positive re-enforcement	☐ Competent	☐ Not yet Competent
Equipment set up:		
Understands of use of 3m and 6m chart	☐ Competent	☐ Not yet Competent
6 metre test used if space permits	☐ Competent	☐ Not yet Competent
Small chair for child	☐ Competent	☐ Not yet Competent
Table for screeners equipment	☐ Competent	☐ Not yet Competent
Adult size chair for screener	☐ Competent	☐ Not yet Competent
Appropriately lit room with no glare on chart	☐ Competent	☐ Not yet Competent
Accurate measurement of test distance marked out on the floor	☐ Competent	☐ Not yet Competent
Adequate supply of consumables	☐ Competent	☐ Not yet Competent
Consent and Explanation:		
Ensures consent is gained and appropriately completed before screening each child	☐ Competent	☐ Not yet Competent
Reviews all pre-screening questions on consent form	☐ Competent	☐ Not yet Competent
Provides a clear explanation of the procedure to the child in a kind and friendly manner	☐ Competent	☐ Not yet Competent
Confirms the child understands what is required and practices use of matching board	☐ Competent	☐ Not yet Competent
Screening:		
Screens visual acuity using the HOTV and/or Sheridan Gardiner Linear Chart (6m or 3m)	☐ Competent	☐ Not yet Competent
Screens vision in the right eye by covering the left eye using the approved occlusion glasses with a fresh tissue folded appropriately beneath	☐ Competent	☐ Not yet Competent
Records visual acuity for right eye correctly	☐ Competent	☐ Not yet Competent

Screens vision in the left eye by covering the right using the approved occlusion glasses and a tissue	☐ Competent	☐ Not yet Competent
Records visual acuity for the left eye correctly	☐ Competent	☐ Not yet Competent
Starts pointing from the top of the chart and works down in a random fashion	☐ Competent	☐ Not yet Competent
Allows the child enough time to respond	☐ Competent	☐ Not yet Competent
Clearly points to individual letters from below	☐ Competent	☐ Not yet Competent
Avoids isolating letters	☐ Competent	☐ Not yet Competent
Ensures other children waiting to be screened cannot see the chart	☐ Competent	☐ Not yet Competent
Performs a visual inspection of the eyes to rule out any other pathology	☐ Competent	☐ Not yet Competent
Follows LHD infection control and hand hygiene protocols	☐ Competent	☐ Not yet Competent
Documentation:		
Ensures results of the vision testing and follow-up required are documented on: • *StEPS Consent and Results Form* • *Notification of StEPS Vision Screening Result letter* • *StEPS Referral Letter* (as per LHD procedure)	☐ Competent ☐ Competent ☐ Competent ☐ N/A	☐ Not yet Competent ☐ Not yet Competent ☐ Not yet Competent
Ensures *Notification of StEPS Vision Screening Results* letters are placed in a sealed envelope for parent/carer	☐ Competent	☐ Not yet Competent
Follow-up:		
Follows appropriate referral protocol as per LHD procedures	☐ Competent	☐ Not yet Competent
Procedures & Guidelines:		
Demonstrates sound understanding of LHD StEPS Program protocols	☐ Competent	☐ Not yet Competent
Displays knowledge of equipment care, maintenance and storage	☐ Competent	☐ Not yet Competent
Demonstrates compliance with LHD infection control and Work Health and Safety procedures	☐ Competent	☐ Not yet Competent
Ensures correct disposals of consumables	☐ Competent	☐ Not yet Competent
Troubleshooting:		
Appropriately manages and adapts to children with challenging behaviour	☐ Competent	☐ Not yet Competent
Monitors referral rate whilst screening	☐ Competent	☐ Not yet Competent
Rechecks screening distance for accuracy as required	☐ Competent	☐ Not yet Competent
Data Collection:		
Follows LHD protocols for data entry	☐ Competent	☐ Not yet Competent
Understands and uses StEPS electronic medical records	☐ Competent ☐ N/A	☐ Not yet Competent
Ensures *StEPS Consent and Results Form* is sent to StEPS Co-ordinator	☐ Competent	☐ Not yet Competent

Comments:	
Assessor Signature:	**Date:**

7.9 Attachment 9: StEPS Screening Activity Quarterly Report

StEPS
Statewide Eyesight
Preschooler Screening

	LHD:	**Select LHD**
	Period:	**Jul - Sep 2016**

		Data Item	Number	Comments (optional)
OFFERS	A	**No. of StEPS screens offered this period**		
	B	*Of the children offered StEPS screens this period, how many:*		
	C	Accepted offer		
	D	Declined offer due to vision screen previously conducted		
	E	Declined offer due to other reasons		
	F	No response (i.e., consent forms not received)		
	G	Total	0	
	H	VALIDATION CHECK: (G) MUST EQUAL (A)	OK	
ACCEPTED	I	*Of the accepted offers this period, how many children were:*		
	J	Screened (visual acuity tested)	0	
	K	Not Screened - Absent on day of screening	0	
	L	Not screened - Other reasons	0	
	M	Total	0	
	N	VALIDATION CHECK: (M) MUST EQUAL (C)	OK	
SCREENED	O	Children screened this period where parental/carer consent to conduct screening was received in *previous period(s)*	0	
	P	Children screened this period where parental/carer consent to conduct screening was received *this period*	0	
	Q	Total StEPS screens conducted this period	0	
PATHWAYS	R	*Of the children screened this period, how many:*		
	S	Passed Visual Acuity Screen	0	
	T	Borderline Pass (follow up in 1 year)	0	
	U	Referred for further assessment (excludes High Priority referrals and children referred due to unable to screen/incomplete screen)	0	
	V	Referred - High Priority Referral	0	
	W	Referred due to unable to screen/incomplete screen	0	
	X	Total	0	
	Y	VALIDATION CHECK: (X) MUST EQUAL (Q)	OK	
ABORIGINALITY	Z	*Of the children screened this period, how many were:*		
	AA	Aboriginal	0	
	AB	Torres Strait Islander	0	
	AC	Aboriginal and Torres Strait Islander	0	
	AD	Non-Aboriginal	0	
	AE	Aboriginal status Not Stated	0	
	AF	Total	0	
	AG	VALIDATION CHECK: (AF) MUST EQUAL (Q)	OK	

7.10 Attachment 10: StEPS Referral Outcomes Quarterly Report

LHD	Year	Quarter
Please select LHD	2016	April-June

TABLE 1: OUTCOMES				
	Referral Type	Number of High Priority Referrals	Number of Further Assessment Referrals	Number of 'Unable to be screened/ incomplete screen' referrals
A	**Total referrals**			
B	Ocular pathology or condition diagnosed			
C	Monitor/Review			
D	Currently under care for vision			
E	Further investigation required			
F	No abnormality detected			
G	Referral not followed up by parent			
H	Lost to follow up (unable to be contacted			
I	Total Outcomes	0	0	0
J	Validation A=I	ok	ok	ok

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TABLE 2: DIAGNOSIS				
StEPS Statewide Eyesight Preschooler Screening	Referral Type	Number of High Priority Referrals	Number of Further Assessment Referrals	Number of 'Unable to be screened/ incomplete screen' referrals
K	Ocular pathology or condition	0	0	0
L	Refractive Error			
M	Emmetropia			
N	Anisometropia			
O	Amblyopia			
P	Strabismus			
Q	Mixed- Anisometropia & Amblyopia			
R	Mixed-Strabismus & Amblyopia			
S	Mixed- Refractive Error & Strabismus			
T	Mixed- Refractive error, Strabismus & Amblyopia			
U	Mixed- Refractive Error & ambylopia			
V	Mixed- anisometropia & strabismus			
W	Mixed- refractive error and anisometropia			
X	Other mixed condition			
Y	Other mixed condition comment			
AA	Cataract			
AB	Conjunctivitis			
AC	Chalazion			
AD	Optic Nerve disorder			
AE	Glaucoma			
AF	Ptosis			
AG	Binocular vision disorders (excluding strabismus)			
AH	Nystagmus			
AI	Corneal Pathology			
AJ	Colour vision deficiency			
AK	Miscellaneous			
AL	Miscellaneous Comment			
AM	Total Diagnosis	0	0	0
AN	Validation cell K=AM	ok	ok	ok

(left margin label: DIAGNOSIS)

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Appendix 2b: StEPS Brochure

StEPs

Statewide Eyesight Preschooler Screening

Please contact:
StEPS Coordinator

Ph: 9562 5400

E: SLHD.StEPS@sswahs.nsw.gov.au

NSW GOVERNMENT | **Health**
Sydney
Local Health District

StEPs
Statewide Eyesight Preschooler Screening

What you need to know
about the StEPS program

NSW GOVERNMENT | **Health**
Sydney
Local Health District

What is StEPS?

The StEPS program is an initiative of NSW Health and offers all 4 year old children free vision screening.

Why would my child need their vision screened?

- Children rarely complain of eye problems
- Children may not realise they can't see well
- Some children can see well with one eye but have very poor vision in the other eye
- Children's eyes may look OK and parents/carers might think that their child can see well but some children might still have a vision problem
- The only way to tell if a child has a vision problem is to have the child's vision tested one eye at a time.

Did you know?

- If a child has a 'lazy eye' it may lead to severe vision loss or blindness in that eye if not treated
- If a child has a vision problem, the earlier the problem is detected and treated the better the vision outcome
- If parents wear glasses or had vision problems as a child their children are more likely to have vision problems too
- After a certain age some childhood vision problems cannot be treated and the child will have poor vision for the rest of their life - glasses won't help
- Low birth weight babies and children with neurological problems are at a greater risk of developing eye problems

Does my child need their vision screened?

NSW Health advises all children to have their vision screened before they start school and strongly recommends that all 4 year old children participate in the vision screening program.

How can my child access the StEPS program?

Your Local Health District will target preschools and child care centres to offer all four year old children a free vision screening. To have your child's vision screened you will need to complete a consent form and return it to your child's preschool/childcare centre.

You can also have your four year old child's vision screened for free through your local Child & Family Health Service. Contact details for your local Child & Family Health Service are on the back of this pamphlet.

How will I know if my child has a vision problem?

Every parent/carer of children who have a vision screening will be informed of the results of their child's vision screening assessment. Should a vision problem be detected you will be contacted by your Local Health District and asked to have your child's eyes fully tested by an eye health professional.

It is important to ensure every child's vision develops normally throughout childhood.

The StEPS program is a vision screening program and does not offer a full diagnostic assessment. If you have any concerns about your child's vision you are recommended to have your child's vision tested fully by an eye health professional.