VISION PROBLEMS AMONG CHILDREN WITH OCULO-CUTANEOUS ALBINISM ATTENDING SPECIAL EDUCATION SCHOOLS IN THE NORTHERN PROVINCE OF SOUTH AFRICA

by

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DEDICATION

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DECLARATION

I, the undersigned declare that the dissertation hereby submitted to the University of the North, for the degree of Master of Optometry has not previously been submitted by me for a degree at this or any other university; and that it is my own work in design and in execution.

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LIST OF ABBREVIATIONS

OCA	=	Oculocutaneous albinism
OCA 1	=	Oculocutaneous albinism (Tyrosinase negative type)
OCA 2	=	Oculocutaneous albinism (Tyrosinase positive type)
OCA 3	=	Rufous or red albinism
OA	=	Ocular albinism
DHICA	=	5,6- dihydroxyindole-2-carboxylic acid
OD	=	Right eye
OS	=	Left eye
OU	=	Both eyes
SPSS	=	Statistical Packages for Social Sciences
VA	=	Visual acuity
CSF	=	Contrast sensitivity function
m	=	Metres

OCA	=	Oculocutaneous albinism
cm	=	Centimetres
cpd	=	Cycles per degree of visual arc
Sec/arc	=	Seconds of arc

DEFINITION OF TERMS

Words	Definitions
Amblyopia	Reduced visual acuity which is not correctable by refractive means and is not attributable to obvious structural or pathological anomalies.
Chediak-Higashi syndrome	An autosomal-recessive disorder characterised by partial albinism, photophobia, massive leukocytic inclusions, psychomotor abnormalities and recurrent infections.
Astigmatism	Refractive condition in which the eye's optical system is incapable of forming a point image for a point object on the retina.
Congenital nystagmus	A repetitive, rhythmic oscillation of one or both eyes in any or all fields of gaze, with jerky movements which are present at birth or shortly thereafter.
Corpus collosum	This is the largest commissure (connecting fibres) in the brain connecting the cerebral hemisphere.
Dyschromatopsia	Imperfect discrimination of colours or partial colour blindness.
Foveal hypoplasia	underdevelopment of the fovea

Hermansky-Pudlak syndrome	A rare autosomal-recessive disease characterized by oculocutaneous albinism, the absence of platelet dense-bodies and ceroid storage disease.
Heterophoria	A latent deviation of the visual axes of the two eyes; a deviation brought about by eliminating all stimuli to fusion.
Hypermetropia (Hyperopia)	(Farsightedness) refractive state of the eye in which the accommodated eye would focus the image behind the retina.
Lentigens	Pigmented flat spots (macules) in the skin, associated with increased melanin synthesis in melanocytes and often found in sun-exposed areas. Freckles (ephelides) are small lentigens.
Light fogging	Blurring of vision due to excessive light entering the eye.
Melanin	A dark, sulfur-containing pigment normally found in the hair, skin, ciliary body, choroid of the eye, pigment layer of the retina and certain nerve cells.
Melanosome	The organelle in the melanocyte that contains the melanin pigment. It has an internal protein matrix in which melanin in synthesized and stored.
Myopia	(Nearsightedness) A refractive anomaly in which, with the accommodation relaxed, parallel rays of light converge to focus in front of the retina.

Mutation	An alteration in the nucleotide sequence of a DNA molecule.
Oculocutaneous albinism	Hereditary defect in the metabolism of melanin resulting in the decrease or absence of this pigment in the skin, mucosa, hair and eyes.
Photophobia	Abnormal sensitivity of the eye to light.
Polymorphism	Frequent hereditary variations at a genetic locus.
Retino-geniculo-cortical fibres	Retinal fibres passing through the lateral geniculate body en route to the visual cortex.
Strabismus (Tropia)	A manifest deviation of the visual axes of one or both eyes.
Tyrosine	A naturally occurring non-essential amino acid present in most proteins. It is a precursor of melanin, catecholamines and thyroid hormones.
Visual acuity	Acuteness of vision; usually the finest detail that the eye can detect.

ABSTRACT

Albinism is a congenitally inherited hypomelanosis that may involve the normally pigmented structures of the body such as the eyes, skin and hair (oculocutaneous albinism) or be limited to the eyes (ocular albinism). Albinism is associated with several visual problems which include: congenital nystagmus, reduced visual acuity (VA), strabismus, astigmatism, high spherical errors, photophobia, poor depth perception, and poor contrast sensitivity. The severity of each of these conditions varies from one type of albinism to another and from one subject to another. Several factors have been implicated in causing the vision problems in individuals with albinism. The major ones include: absence of melanin in the normally pigmented structures of the eye (iris, retinal pigment epithelium and choroid), poor foveal differentiation (foveal hypoplasia) and misrouting of optic nerve fibres at the optic chiasma.

The existing information on the visual status of people with albinism is based on findings from small populations of individuals with albinism. No study on vision problems has been carried out specifically on children with albinism in any part of the world, findings of which are expected to differ from adults or mixed populations. The purpose of this study, therefore, was to determine the magnitude and prevalence of visual problems (visual acuity, refractive errors, contrast sensitivity, depth perception and colour vision) among children with oculocutaneous albinism. Subjects were children attending the special education schools in the Northern Province of South Africa. This study was the first visual performance study to be conducted on a large population with oculocutaneous albinism. It was also probably the first of such study conducted in Africa.

A battery of optometric procedures which included visual acuity measurement, cover tests, retinoscopy, subjective refraction, contrast sensitivity, ophthalmoscopy, stereo-test and colour vision tests were used to study the visual status.

A total of 153 black South African children with oculo-cutaneous albinism was included in this study (147 OCA2, 3 OCA1 and 3 brown albinism). They included 77 (50.3%)

males and 76 (49.7%) females and their ages ranged from 7 to 17 years (mean =10.76 +/- 2.5 years).

Nearly thirty five percent (34.6%) of subjects had strabismus ranging from 2 to 25 prism diopters (pd) (mean= 6.96 +/- 5.63 pd) for exotropia and from 2 to 30 prism diopters (mean = 8.96 +/- 7.50 pd) for esotropia.

The uncorrected distance visual acuities were: right eye (OD): finger counting (FC) to $6/7.5^{-2}$ ($20/25^{-2}$) (0.1^{-2} logMAR); left eye (OS): $6/75^{+2}$ ($20/250^{+2}$) (1.1^{+2} logMAR) to $6/7.5^{-3}$ ($20/25^{-3}$) (0.1^{-3} logMAR); both eyes (OU): $6/60^{-1}$ ($20/200^{-1}$) (0.1^{-1} logMAR) to 6/7.5 (20/25) (1.00 logMAR). The uncorrected near VA was: OD: Finger counting to 0.63^{-2} M (Snellen equivalent of $6/9.5^{-2}$) (0.2^{-2} LogMAR); OS: 5.0 M (6/75) (1.1 logMAR) to 0.8^{+2} M ($6/12^{+2}$) (0.3^{+2} logMAR) OU: 4.0^{2} M ($6/60^{2}$) (1.0^{2} logMAR) to 0.63^{2} M ($6/9.5^{2}$) (0.2^{-2} LogMAR).

Myopia (67.6%) was more common than hyperopia (30.8%) or emmetropia (1.7%). The nearest equivalent spherical powers observed ranged from -12.00 D to +2.75 D (mean= -1.48 +/-2.28 D) for the right eye and from -8.25 D to +5.75 D (mean = -1.36 +/-2.18 D) for the left eye. Astigmatism was present in 92.2% of all the eyes. Myopic astigmatism (62.1%) was more prevalent than hyperopic (30.4%) and simple (7.5%) types. With-the-rule (WTR) astigmatism was more common (47.0%) than oblique (OBL) (28.0%) and against-the-rule (ATR) (25.0%) astigmatism.

The corrected distance visual acuities were: right eye (OD): FC to $6/6^{-1}$ ($20/20^{-1}$) (0.0^{-1} logMAR); left eye (OS): $6/75^{+2}$ ($20/250^{+2}$) (1.1^{+2} logMAR) to $6/6^{-1}$ ($20/20^{-1}$) (0.0^{-1} logMAR); both eyes (OU): 6/60 (20/200) (1.0 logMAR) to 6/6 (20/20) (0.0 logMAR). The corrected near VA were OD: Finger counting to 0.4^{+2} M ($6/6^{+2}$) (0.0^{+2}logMAR); OS: 3.2^{+1} M ($6/48^{+1}$) (0.9^{+1} logMAR) to 0.4^{+1} M ($6/6^{+1}$) (0.0^{+1} logMAR) and OU: 2.5 M (6/38) (0.8 logMAR) to 0.4^{+1} M ($6/6^{+1}$) (0.0^{+1} logMAR). A large proportion (71.2%) of the subjects showed significant improvement in distance VA with the best optical corrections. The VA improvement ranged from one letter to 3 acuity lines (15 letters). The improvement in the near VA (monocular and binocular) ranged from 1.0 to 5.0 logMAR (one to 5 acuity lines) (5 to 25 letters). One hundred and twenty nine (84.3%)

children were classified as partially sighted and 24 (15.7%) were classified as normally sighted based on the distance visual acuity values.

Contrast sensitivity function (CSF) test revealed that, a maximum spatial frequency of 18 cpd was perceived binocularly and the peak sensitivity was at 3 cpd for all age groups. The mean sensitivity for each spatial frequency were: 2.6 for 1.5 cpd, 2.0 for 3.0 cpd, 1.0 for 6 cpd, 0.4 for 12 cpd and 0.1 for 18 cpd. Contrast sensitivity function resolution acuity and peak sensitivity increased only by about 1 octave following optical corrections.

A large proportion (89.5 %) of the subjects demonstrated stereopsis, although poor. Few subjects (10.5%) could not perceive the minimum stereoacuity of 500 seconds of arc at 40 cm on Randot stereo test. Stereoacuities values at 40 cm ranged from 500 to 70 seconds of arc (mean= 294.0 +/- 112.6 sec arc).

One hundred and twenty seven (83.0%) of the children had normal colour vision, while others failed the colour vision tests. Those who failed included: 7 deutanopes, 8 protanopes, 6 tritanopes and 2 were achromats. Among those who failed the tests, 58.0% were males and 42.0% were females.

It was concluded that children with oculocutaneous albinism exhibit poor visual acuity which can be improved significantly in most individuals. High refractive errors: astigmatism, myopia and hyperopia were also observed. Although most of the children demonstrated stereoacuity, the values were significantly lower than expected in the normally pigmented individuals of similar age groups. Also, the subjects exhibited poor contrast sensitivity which remained poor after correction. Colour vision, however, was normal in most children.

Findings in this study will contribute to the knowledge of vision problems in individuals with albinism. It is recommended that the vision problems in children with albinism should be corrected at an early age so that maximum visual function can be maintained and amblyopia avoided.

CHAPTER 1

1. INTRODUCTION

1.1 ALBINISM

Albinism is a congenitally inherited hypomelanosis that may involve the normally pigmented structures of the body such as the eyes, skin and hair. The condition is generally classified into ocular albinism (OA) and oculocutaneous (OCA) types. When only the eyes are affected, the condition is called ocular albinism (OA), whereas when both the eyes and skin are affected; the condition is called oculocutaneous albinism (Witkop, 1970; Oetting et al., 1996). Ocular albinism is less commonly noted than OCA, and the affected individuals will have reduced melanin pigment primarily in the eyes (King and Summers, 1988). The main subdivisions of OCA are: tyrosinase negative (OCA1), tyrosinase positive (OCA2) and rufous or red (OCA3) types. OCA type 1 (OCA1) refers to a clinical picture of hypopigmentation resulting from mutations of the tyrosinase gene on chromosome 11q14-21 (King et al., 1995). Individuals with OCA1 are born with a complete absence of pigment in the skin, eyes, and hair. For many individuals, this complete lack of pigment persists throughout life (OCA1A) because their tyrosinase gene mutations are responsible for an inactive enzyme (Oetting and King, 1999). Other individuals with OCA1 have mutations that result in residual tyrosinase activity, producing a spectrum in the amount of pigment in the skin, hair, and eyes (OCA1B) (Oetting and King, 1999). The defective gene in the tyrosinase-positive albinism (OCA2) codes for a transmembrane protein that has been mapped to chromosome 15q11.2-12 (Summers, 1996). These individuals are typically born with melanin pigment in their hair, in contrast to the white hair that is present at birth in OCA1. The gene in OCA2, coding for a membrane protein, has been referred to as the "P gene" because similar clinical and genetic features are found in the pink-eyed-dilution (p) mouse. The function of the protein in human remains uncertain (Rinchik et al., 1993). Those with OCA2 in Africa have a distinctive phenotype, with

sandy to yellow coloured hair, blue to hazel eyes and pale chalky coloured skin (Lund, 1997). Another type of OCA, Rufous or red (OCA3) are recognized by their reddish skin and hair colour (Manga et al., 1997). In this type of OCA, nystagmus and foveal hypoplasia were uncommon and misrouting of the optic fibres measured by visual evoked potentials was not found (Kromberg et al., 1990). There is very little clinical research data available on OCA3 and the phenotype is incompletely documented (Oetting and King, 1999). Oculocutaneous albinism may also be a secondary association occurring with systemic disorders such as Hermansky-Pudlak syndrome (mutation on chromosome 10) and Chediak-Higashi syndrome. In all cases of albinism, the inherited deficiency is in the production of melanin pigment (Summers, 1996). Other less common types of albinism are brown albinism and albinoidism. The former type results in tan skin, brown hair and blue or brown eyes (Kromberg et al. 1990) and has been reported in Africans, African Americans and Caucasian populations. In the latter, although a form of hypopigmentation is exhibited, it is not accompanied by nystagmus, photophobia and visual impairment (Abadi and Pascal, 1989; Lyle et al., 1997).

1.1.1 Prevalence

Albinism affects people all over the world in varying degrees. It rarely affects Europeans, frequently Africans, only a minority of American Indians, who nevertheless, when an ethnic group is concerned, presents one of the highest incidence in the world (Lyle et al., 1997). It affects nearly 1 in 39,000 whites and is more common among black Americans (Kinnear et al., 1985; Fulcher et al., 1995). Various types of albinism is particularly prevalent in sub-Saharan Africa. Albinism affects 1 in 1634 school children in Swaziland (Ewusie, 1988) and 1 in 2,833 school children in Harare, Zimbabwe, (Kagore and Lund, 1995). The condition affects 1 in 3900 in Soweto, South Africa. The high incidence of albinism among the black South African population has been attributed to the practice of consanguineous marriage among certain tribes (Kromberg and Jenkins, 1982).

The oculocutaneous (OCA2) type is the most common type of albinism in the United

States of America, occurring in 1 in 36,000 among Caucasians and 1 in 10,000 in African Americans (Witkop et al., 1989). It is one of the most common recessively inherited conditions in sub-Saharan Africa. There is a high incidence of OCA2 among specific African populations: 1 in 7900 population among Bamileke tribe of Cameroon (Aquaron, 1990), 1 in 1100 among the Ibos of Nigeria (Okoro, 1975; Witkop et al., 1989), an overall occurrence of 1 in 4182 in Zimbabwe (Lund, 1996) with a higher prevalence in the capital city, Harare (1 in 2833) (Kagore and Lund, 1995) and 1 in 3,900 in South Africans (Ramsay et al., 1992). The frequency of OCA2 in the Northern Province of South Africa is particularly high, at 1 in 1500 neonates (Venter et al., 1995).

Brown albinism initially was defined in the Nigerian population (King et al., 1980) and has been found to occur only in the Black, or Negroid, populations of Africa and America (King et al., 1995). Rufous (Red) OCA principally has been reported in the Black populations of Africa and Papua New Guinea (Stannus, 1913) (cited by Manga et al., 1997); Walsh, 1971; Kromberg et al., 1990). In Southern Africa, brown and rufous oculocutaneous abinism (BOCA and ROCA) have been reported mainly in South Africa, where BOCA affects 1 in 8,580 black school children (Kromberg et al., 1990) and ROCA affects 1 in 8,500 South Africans (Kromberg et al., 1990). Although issues relating to ocular albinism have been studied by several authors (O'Donnell et al., 1978; Charles et al., 1993; Shiono et al., 1995), the prevalence has not been properly documented. According to Kinnear et al., (1985), almost 1:50000 people has autosomal recessive ocular albinism.

1.1.2 Molecular Basis

In albinism, the amino acid tyrosine which is necessary for melanin production is either not produced or produced in an inactive form. This is because the enzyme, tyrosinase is either absent or inactive in their body. This results in lack of pigmentation of part or all of the normally pigmented structures of the body (Oetting and King, 1996).

The synthesis of melanin pigment is complex, requiring the products of many genes. Two types of melanin are produced: black-brown eumelanin, and yellow-red

pheomelanin; synthesis of these types of melanin is localized in the melanosomes within the melanocytes in the skin, hair follicle and eye (Oetting and King, 1996). More than 65 separate loci have been identified in mice that affect either the production of melanin pigment or melanocyte distribution and pattern development, or both (Silvers, 1979). The human homoloques of several of these loci have been isolated and mutations in at least six of these genes are associated with albinism (Oetting and King, 1999). Different mutations of the same gene can produce a spectrum of pigmentation associated with albinism, from a complete absence to near normal levels of melanin.

Mutations in the five different genes responsible for different types of oculocutaneous albinism and mutation in one gene are responsible for the ocular albinism. The mutations of the most common types of albinism include: (i) the tyrosinase gene (TYR) and OCA1 (MIM#203100); (ii) the OCA2 gene and OCA2 (MIM#203290). A number of other genes are being characterized for other types of albinism and related conditions, e.g the HPS gene and Hermansky-Pudlak syndrome (MIM# 203300); the CHS gene (CHS1), and Chidiak-Higashi syndrome (MIM# 214500) and X-linked ocular albinism gene and OA1 (MIM# 300500) (Oettings and King, 1999). The key enzyme in the melanin biosynthetic pathway is tyrosinase, which is inactive in OCA1 cases. The function of only two of the gene products is known: tyrosinase and tyrosinase-related protein-1, both of which are enzymes in the melanin biosynthetic pathway. Some of these genes that are responsible for the various types of albinism are shown in Table 1.

Table 1*: Some types of albinism, gene products and their chromosomal location.

Type of albinism	Gene product	Chromosomal Location
OCA I of several subtypes	Tyrosinase enzyme	11q14-q21
Tyrosinase-related protein, OCA IV or brown OCA	TRP-1	9p23
OCA 2 or brown albinism	P protein	15q11.2-q12

* Adapted from a table presented by Lyle et al.,1997.

Tyrosinase, associated with OCA1, is a copper-containing enzyme that is responsible

for the rate-limiting step in the melanin pathway: conversion of tyrosine to dopaquinone (Cooksey et al., 1997). A total of 88 mutations of the tyrosinase gene have been reported in the tyrosinase gene associated with OCA1 (Oetting and King, 1999). There are also six polymorphisms in the tyrosinase gene, two of which result in amino acid substitutions and many affects the biology of the enzyme (Tripathi et al., 1991).

The OCA2 gene encodes a transmembrane protein with unknown function. The most prevalent mutation of the OCA2 gene associated with OCA2 is a deletion of exon 7 (Durham-Pierre et al., 1994, 1996; Spritz et al.,1995). An intragenic deletion mutation has been identified as the most common cause of OCA2 in the South African black population (114/146 (0.78) OCA2 chromosomes) (Stevens et al., 1995) as well as in neighbouring countries such as Zimbabwe (Puri et al., 1997) and other parts of Sub-Saharan Africa (Durham-Pierre et al., 1994; Spritz et al., 1995).

The tyrosinase-related protein-1 (TYRP1) associated with OCA3, has been shown to have DHICA oxidase activity, placing it distal to tyrosinase in the eumelanin pathway (Jimenez-Cervantes et al., 1994).

1.1.3 Skin Cancer

People with albinism have an increased risk of skin cancer, particularly squamous and basal cell carcinomas, but may have a lower risk of malignant melanomas (Kinnear, et al., 1985; Kromberg et al., 1989). The high risk of skin cancer is due to very little melanin pigment produced in the skin resulting in an increased sensitivity to ultraviolet radiation. About 40% of people with tyrosinase positive (OCA2) develop dark pigmented patches such as: dendritic freckles, ephilides, lentigens, goose foot lentigens and actinic lentigens (Findlay et al., 1962; Hall et al., 1976; King et al., 1980; Witkop, 1985; Kromberg et al., 1989; Lookingbill et al., 1995) on sun exposed parts of their body, reflecting a capacity to form melanin.

Those with dendritic freckles are less likely to develop solar keratoses and skin

cancers than those without freckles (Bothwell, 1997). Skin cancer among individuals with albinism in the equatorial regions of the world has a higher incidence and a more ominous course, perhaps due to neglect and many individuals with albinism in these areas die because of skin cancer (Okoro, 1975; Luande et al.,1985)

1.1.4 Intellectual Capacity

There are conflicting reports about the intellectual capacity of individuals with albinism A child with albinism has been considered to be intellectually retarded (Segers, 1929; cited by Fulcher et al., 1995) but there are recent reports that the intelligence of an individual with albinism does not differ from that of their non-albino siblings (Beckham 1946; Manganyi et al., 1974; Fulcher et al., 1995). Although the children with the condition had normal intelligence, they seem to perform more poorly in tests of reading, spelling and arithmetic ability (Fulcher et al., 1995). The reason for the poor performance in those areas of their study might be due to the vision problems such as poor visual acuity, lack of binocular vision and poor contrast sensitivity (Fulcher et al., 1995).

1.1.5 Myths and Beliefs

Although albinism is a common condition in Africa, accurate knowledge and information about the condition in some countries on the continent is inadequate. As a result of the unusual appearance and symptoms which are particularly striking in the black population, albinism is surrounded by many myths and beliefs which has led to discrimination and lack of acceptance by the various communities (Kromberg and Jenkins, 1984). In Nigeria for instance, nowhere in the traditional views on albinism is it believed to be associated with inbreeding, rather, weird aetiological theories ranging from punishment from the gods, to conception during menstruation, or to seeing frightening sights during pregnancy (Okoro, 1975). Social life is generally difficult for individuals with albinism and neither their relatives nor anyone else would agree that

most people with albinism are normal in every aspect except for the absence of pigment in their skin, hair and eyes (Okoro, 1975). In Zimbabwe, a common belief is that someone who had been unkind or unpleasant to an individual with albinism would bear a child with albinism (Lund, 1997). Others regarded albinism as a curse from God for the faults of the parents and people with albinism are regarded as outcast by the family and the community at large (Lund, 1997). In South Africa, it is believed that a child with albinism is born as a result of various foods taken in excess during pregnancy or due to the mother conceiving during menstruation or positively as a sign of good luck (Sacharowitz, 1999). In the Venda area of the Northern province of South Africa, it is a common belief that individuals with albinism do not die, but disappear. The Albinism Society of South Africa (ASSA) was established in 1992 in an attempt to educate communities on the various aspects of albinism and provide a support base for its members (Sacharowitz, 1999). These activities of the ASSA will help to deal with the superstitions associated with albinism and result in individuals with the condition being accepted into their communities.

1.2 VISION PROBLEMS ASSOCIATED WITH ALBINISM.

Several vision problems have been associated with albinism. These include: high refractive errors, strabismus, reduced visual acuity, poor contrast sensitivity, congenital nystagmus, photophobia, poor depth perception and colour vision deficiency. (Taylor, 1978; O'Donnel and Green., 1981; Abadi and Dickinson., 1983; Kinnear et al., 1985; Guo et al., 1989; Perez-Carpinell, 1992; Summers, 1996; Sacharowitz, 1999). These vision problems have been associated with neural anomalies which include foveal hypoplasia (O' Donnel et al., 1976; Fulton et al., 1978) and aberrant optic pathway projections (Guillery et al., 1973; Guillery et al., 1975; Creel et al., 1974; 1978; 1981; Guillery et al., 1984).

Anatomical evidence in the cat, monkey and human has shown that albino central retina contains a ganglion cell layer, rods and cones with outer segment morphology

similar to those found in the parafovea at 5^0 (Fulton et al., 1978; Guillery et al., 1984; Stone et al., 1978). This has led to the hypothesis that the albino central retina is underdeveloped (Wilson et al., 1988 a; 1988 b). Also, foveal cone spacing has been estimated to be 3-4 times greater than in the normal fovea (Fulton et al., 1978) suggesting foveal underdevelopment which will reduce the functional capability of the fovea.

The optic projection anomaly is in the form of a reduction in ipsilaterally destined retinofugal projections. In albinism, certain temporal retinal ganglion fibres which should normally project ipsilaterally, decussate erroneously at the optic chiasma leading to abnormal arrangement of fibres in the lateral geniculate body and altered representation of the eye in the visual cortex (Witkop et al., 1973). Comparable anomalous retinofugal projections have been demonstrated in other species of mammals with albinism (Witkop et al., 1982). The origin of the abnormal decussation of the retinostriate fibres has been attributed to the reduction or absence of melanin in the eyes during embryogenesis (Witkop et al., 1973; La Vail et al., 1978; Silver, 1984; Webster et al., 1988).

1.2.1 Refractive Status

There are many reports on the refractive status of individuals with normal vision. The prevalence and distribution of ametropia vary greatly with age in a normal population. According to Grosvenor (1989) refractive status distributes normally at birth, but early in infancy, the majority of children are found to be somewhat hyperopic with a tendency towards myopia up to 20% or more in the 20-year-old ones. A drift toward hyperopia with advancing age has also been reported (Hyams et al., 1977; Kragha, 1987; Chung et al., 1996; Raliavhegwa and Oduntan, 2000) indicating that more older people are hyperopic than myopic in contrast with the younger generations. There are reports, also of greater incidence of myopia in females than in males (Richler and Bear, 1980; Johnson et al., 1979). There are contrary reports, however, that myopia is more

prevalent in males than in females (Hyams et al., 1977; Raliavhegwa and Oduntan, 2000). Astigmatism is also influenced by certain factors such as age, sex and race. Infants showed less astigmatism, the extent increasing through childhood with those showing a higher spherical error revealing the greater increases (Borish, 1970). It has been reported that the majority of children and adults have with-the-rule (WTR) astigmatism (Grosvenor, 1989; Gordon, 1990; Chan et.al., 1993) but during the later adult years, there is a tendency for WTRA to decrease and ATRA to increase (Grosvenor, 1989). Greater incidence of ATR has been reported by others (Kragha, 1987; Raliavhegwa and Oduntan, 2000). These differences may be due to differences in race, age and gender of the subjects studied. There are observations also, that WTRA is relatively more common in males than in females and that this type increases in males but decreases in females with age (Grosvenor, 1989).

There are not many reports on the refractive status of individuals with albinism. There is a general consensus, however, that they have high spherical refractive errors combined with astigmatism (Fonda et al., 1971; Perez-Carpinell et al.,1992; Sacharowitz, 1999). Myopia has been reported to be common in individuals with albinism (Fonda et al., 1971; Perez-Carpinell et al.; Sacharowitz, 1999). There are contrary reports, however, that hyperopia was more prevalent in these individuals (Taylor, 1976; Fulcher et al., 1995). The difference may be attributed to the types of albinism and the age of the subjects studied. In a previous study, 12.5% of all people with albinism (47 OCA1, 18 OCA2 and 14 OA types) had spherical errors (myopia and hyperopia) over 10.50 diopters and another 33% had spherical errors with magnitude between 4.50 and 10.00 D (Fonda et al., 1971). In another study in which 7 OCA1 and 9 OCA2 subjects of various ages were studied (Taylor, 1976), it was found that refractive errors ranged from -5.00 D to +9.50 diopters. Spherical equivalent errors ranging from -8.00 D to +6.00 D were reported by Guo et al., (1989) in 19 subjects with oculocutaneous albinism, age ranging from 15 to 20 years. Perez-Carpinell et al. (1992) studied the vision defects of 9 subjects with oculocutaneous albinism (3 of whom were OCA1 and 6 were OCA2 types) aged between 7 and 39 years. They found refractive errors ranging from -10.00 D to +7.00 D. A more recent retrospective

study, (Sacharowitz, 1999) in which over 6 years clinical records of South Africans with OCA2 type and of various ages were reviewed, the spherical equivalent refractive errors ranged from -20.00 D to +5.50 D.

Astigmatic errors are also common in individuals with albinism. Perez-Carpinell et al. (1992) reported that 50% of the eyes examined had astigmatism, whereas, Sacharowitz (1999) reported that 76% of the eyes had astigmatism. There are reports that hyperopic astigmatism (83.0%) was more common than other types of astigmatism (Fulcher et al.,1995). According to Taylor (1976), 78.1% of the subjects studied had hyperopic astigmatism. There are contrary reports that myopic astigmatism (46.3%) was more common in individuals with albinism (Sacharowitz, 1999). Regarding the axes of the astigmatism, it is generally agreed that with-the-rule astigmatism (WTRA) is more prevalent among people with albinism (Loshin and Browning, 1983; Dickinson and Abadi, 1984; Davis et al, 1997; Lyle et al.,1997). The various aspects of refractive status in individuals with albinism have not been sufficiently studied as many of the reports were based on small population samples. Also, no study has been specifically conducted on children population. A large population sample studies are considered necessary in order to document the refractive status of individuals with various types of albinism and to establish what improvement in vision could be achieved following refraction. Also, there is a need for studies on large children population. This will help to determine what eye care assistance should be directed towards children with albinism as many children with albinism has been reported to drop out from school partly because of uncorrected refractive errors and partly because of discouragement (Okoro, 1975).

1.2.2 Visual Acuity

Experience has shown that visual acuity of 20/20 can be achieved by children with normal visual development at the age of 5 to 6 years. Visual acuity is variably reduced in albinism, depending in part on the type of albinism and amount of ocular melanin

pigment (Fonda et al., 1971; King and Summers, 1988; Kinnear et al., 1985; Charles et al., 1993). The reduction in VA may be due, in part, to the foveal hypoplasia (O' Donnel et al., 1976; Fulton et al., 1978) or structural and anatomical alterations of the foveal cone photoreceptors (Fulton et al., 1978; Guillery et al., 1984; Stone et al., 1978). The primary source of subnormal acuity is hypoplasia of the macula, which generally includes absence of a foveal pit, absence of normal macula pigment, absence of a rod-free area, the presence of extrafoveal rods and cones, and an abnormal vasculature pattern in the macula area (O' Donnell and Green, 1981). Other contributory factors are nystagmoid movements, intraocular scatter of light due to excessive light transmission, degradation of the retinal image and possible amblyopia that may result from high ametropia and strabismus. Previous reports have shown that some individuals with albinism have almost normal distance vision (20/20) (Castronuovo et al., 1991; Summers et al., 1991; Summers et al., 1996). In others, VA can be as poor as 20/400 (Witkop et al., 1973; O'Donnell et al., 1976; O'Donnell et al., 1978; Cheong et al., 1992). Distance visual acuity of individuals with tyrosinase negative OCA (OCA1) has been reported to be 6/60 to 6/120 (20/200 to 20/400) (Kinnear et al, 1985; Gross et al., 1995). It has been reported to be 6/18 to 6/120 (20/60 to 20/400) in those with OCA2 (Kinnear et al., 1985). In brown albinism, VA has been reported to be 20/50 to 20/150, while in ocular albinism (X-linked and Autosomal OA), it has been reported to be 20/50 to 20/400 (Summers et al., 1996).

Near vision has been reported to be generally better in albinism as a result of dampening of nystagmus on convergence (Biswas, 1999). The author, however, did not report the values of the visual acuities. As the macula is not fully developed in people with albinism, the expected significant improvement in visual acuity is not achieved when the refractive error is corrected (Perez-Carpinell et al., 1992). According to this author, an improvement in VA with the best optical corrections (far and near) of less than 6/30 (20/100) in five subjects was observed and there was an improvement in visual acuity of 6/9 and 6/10.5 (20/30 and 20/35) respectively in two subjects. No improvement was observed in two subjects. Sacharowitz (1999) reported a significant improvement in distance visual acuity with optical correction in 60% of the

patients with OCA2 but in 40% of the subjects, there was no significant improvement in distance VA. The author, however, did not present the VA values of the subjects. Refraction combined with nystagmus surgery has also been considered to improve VA in individuals with albinism. Improvement in VA of two or more Snellen lines was observed following large recession nystagmus surgery and refraction in individuals with albinism (Davis et al., 1997). Visual acuity has also been reported to improve during the nystagmus cycle, where the eye movement velocity is < 10° per second (Biswas, 1999). Visual acuity in children with albinism has not been specifically studied and this needs to be documented.

1.2.3 Contrast Sensitivity

Contrast sensitivity function (CSF) has been considered to be a valuable tool for assessing qualitative aspects of vision, especially in abnormal visual conditions such as amblyopia (Levi et al., 1977) and ocular pathology (Arden, 1978; Comeford, 1979). Even though only sinusoidal targets are used in contrast sensitivity assessment, the contrast sensitivity function (CSF) provides a description of the visual system's ability to detect and identify spatial complex objects (Loshin and Browning, 1983). When contrast sensitivity (CS) is plotted against spatial frequency the resulting graph is termed the contrast sensitivity function (CSF).

There are only a few reports on the contrast sensitivity in people with albinism. There is a consensus, however, that CS is poor in individuals with the condition (Loshin and Browning, 1983; Abadi and Pascal, 1989; Perez-Carpinell et al.,1992). According to Abadi and Pascal (1989) , the overall shape of the CSF of people with albinism is similar to that of a normal observer, however, the peak of maximum sensitivity is shifted to a lower spatial frequency of about 1 cycle/ degree. While the high frequency cut-off of a normal subject was about 40 cpd, for those with albinism, the maximum frequency was 8 cpd (Abadi and Pascal, 1989). Perez-Carpinell et al. (1992), also reported a maximum frequency perceived by subjects with albinism binocularly being 6 cpd, with

an average of 3 cpd. Through the telescopic aid, the CSF resolution and peak sensitivity was reported to increased for all the subjects (6 OCA2 and 2 OA) examined, the increased in resolution was, however, less than would be predicted based on magnification (Loshin and Browning, 1983). The increase in sensitivity was attributed to stimulation of more sensitive peripheral retina (Loshin and Browning, 1983). People with albinism have also been reported to show sensitivity difference between vertical and horizontal CSF gratings; being more sensitive to the latter than the former. According to Abadi and Pascal (1989), for the normal observer, the CSFs for horizontal and vertical periodic targets were identical but in albinism there were distinct differences. Individuals with albinism show greater sensitivity to horizontal than vertical CSF grating; this meridional difference was attributed to high astigmatic error, nystagmus and possible neural modifications in individuals with albinism (Abadi and King-Smith, 1979; Abadi and Pascal, 1989; Loshin and Browning, 1983; Dickinson and Abadi, 1985). With lower spatial frequencies there is often a 'cross-over' such that sensitivity to vertical contours is equal to or even greater than that to horizontal contours (Abadi and Pascal, 1989; Loshin and Browning, 1983). None of the reports that have examined contrast sensitivity in albinism has specifically been designed to study this visual function in children. Such data will help to establish the level of contrast sensitivity in children with albinism, or whether CSF values differ in children from those of adults, and what improvement is expected following refraction.

1.2.4 Colour vision

Several reports have been presented on colour vision in albinism. While about 7% to 8% of normal males and 0,5% of normal females have congenital X-linked colour vision defects (Pokorny et al. 1979), there are several reports that individuals with albinism might have a certain degree of colour vision deficiency (Pickford, 1958; Pickford and Taylor, 1976; Perez-Carpinell et al., 1992). Other reports, however, noted that colour vision in albinic individuals is generally normal (O' Donnel and Green, 1981; Lyle et al., 1997; Oduntan 1998). It has, however, been reported that minor anomalies of colour

vision could be detected in albinic individuals using anomaloscope, electro-retinography and electro-oculography tests (Taylor, 1976). According to Taylor (1976), the type of colour vision defect which appears to be peculiar to albinism is quite a minor one, but of great theoretical significance as a possible indicator of colour receptor development and action. The nature of the defect is a red deviation (protonomalous) which will explain the failure by many observers to detect it, since pseudo-isochromatic tests are too insensitive (Taylor, 1976). In a study of vision defects in albinism which included colour vision evaluation, Perez-Carpinell et al. (1992), using Ishihara's plates, the Roth 28-Hue test and Davico's anomaloscope, found only two cases of colour deficiency (one simple deutan and one red deviant) out of the six subjects (OCA2) examined, whereas, those belonging to OCA1 type were all chromatically anomalous (two red deviants and one simple deutan). Others (four subjects) had normal colour vision. Colour vision deficiency in individuals with albinism is to be expected since individuals with albinism have a poorly developed fovea, which is the region of the retina used for colour vision. There is a need for more studies of colour vision in individuals with albinism especially in a large population in order to document the prevalence of the various types of colour vision deficiency in these individuals. If it is confirmed that not all individuals with albinism have colour vision deficiency, this may suggest that the fovea is more developed in some individuals with albinism than others. It may be of interest to find out if those with good colour vision have better VA as the more developed the macula, the better the VA. Also, It may be of interest to find out if any particular group of individuals with albinism has better colour vision than other groups. This is, however, beyond the scope of the present study, therefore, awaits a future study.

1.2.5 Depth Perception.

The ability to perceive depth or relative distance on the basis of retinal disparity that gives rise to a sensation of depth is termed stereoacuity (Romano et al., 1975). It is expressed in visual angle of disparity in seconds of arc. A major importance of

stereoacuity is in the clinical assessment of binocular vision. A reduction or absence of stereoacuity may be caused by anisometropia, strabismus or amblyopia (Saunders et. al., 1996). Also, stereoacuity is highly sensitive to refractive condition especially when there is a difference in visual acuity between the two eyes (Lang, 1983; Manny et. al., 1991). This agrees with the notion that all optical, neural and motor components in both eyes must be working normally for normal stereoacuity to be achieved. It has led to stereopsis being proposed as an ideal test for overall visual function (Simons and Reinecke, 1974). Stereopsis is not present at birth but usually appears during the third or fourth postnatal month (Shea et al., 1980; Held et al., 1980) and the time course for development in infants is extremely rapid (Held et al., 1980). It is generally agreed that stereoacuity improves with age (Shea et al., 1980; Held et al., 1980; Al-Mubrad and Oduntan, 1996). It has been suggested that the maturation of stereoscopic capacity is nearly complete in children three to five years old (Fox et al., 1986). Using the Titmus stereotest, Al-Mubrad and Oduntan (1996) found that a stereoacuity of 20 seconds of arc can be achieved by children with normal binocular vision aged six years while others could not achieve that level even at 12 years. A recent study (Oduntan et al., 1998) using the Randot stereoacuity test reported a range of stereoacuity for age groups 6 to 12 years to be 70 to 20 seconds of arc.

Depth perception (stereoacuity) has been reported to be absent or poor in individuals with oculocutaneous albinism (Creel et al., 1981; Kinnear et al., 1985 ; Apakarian, and Reits, 1989; Guo et al., 1989; Lyle et al.,1997). The poor stereopsis in individuals with albinism has been attributed to misrouting of retino-geniculo cortical fibres. In normal humans, approximately one half of the fibres project to the ipsilateral hemisphere but in human with albinism, more than one half of the fibres cross to the contralateral geniculate body (Guillery et al., 1975). The paucity of ipsilateral optic nerve fibres projections severely disrupts binocular activation of neurons in the visual cortex (Distafano et al., 1984). The misrouting also might be expected to disrupt binocular function in people with albinism, thereby affecting depth perception. Anatomic misrouting of retino-geniculo-cortical fibres is dramatic enough to have led some investigators to suggest that stereopsis in albinism might depend not on the

convergence of direct ipsilateral and contralateral retino-geniculo-cortical innervation but rather on information transfer via the corpus callosum (Apakarian and Reits, 1989). The poor depth perception observed in individuals with albinism may also be due to high refractive errors since poor depth perception is associated with poor refractive status (Donzis et al.,1983; Larson and Lanchance, 1983) and most individuals with albinism are known to have significant refractive errors. Also, the foveal hypoplasia may degrade depth perception capability. Although there are reports of poor depth perception in individuals with albinism, the reports were based on findings from general studies of albinism and none has been specifically designed to study this visual function in children with albinism. It is, therefore, necessary to study depth perception in the children with albinism in greater detail.

1.2.6 Other Visual Problems: Nystagmus, Photophobia and Strabismus.

Nystagmus is a repetitive, rhythmic, involuntary movement of the eyes, either having equal velocity and amplitude in both direction or having a slow movement in one direction and a fast movement in the other (Grosvenor, 1989). It can be congenital and can occur in many CNS disorders or caused by bilaterally reduced vision in early childhood (Buckingham, 1993) . It occurs in a wide range of eye disorders of childhood such as cataract, glaucoma and some disorders of the retina. It may also be found in children who have multiple disabilities such as Down's Syndrome. The prevalence of nystagmus in the general population is not known accurately but it is believed to affect around 1 in 1,000 individuals (RNIB and RCO, 1999).

Nystagmus is a consistent finding in albinism although occasionally, individuals exist with albinism but without nystagmus (Kriss et al., 1992). The nystagmus usually appears in the first 2 or 3 months of life and often reduced in magnitude with age (Biswas, 1999). In infancy the eye movements are typically pendular and of large amplitude, especially if the ambient light levels are high (Biswas, 1999). Like those with congenital idiopathic nystagmus, some individuals display a 'null zone' (a position

of gaze in which the nystagmic intensity is at a minimum) and many possess a 'neutral zone' (a gaze area in which the nystagmus fast phase reverses direction) (Abadi and Pascal, 1989). An abnormal head posture may be adopted by certain individuals to maintain the eyes in a position of relative stability (where visual acuity is likely to be maximal) whilst looking straight ahead. This happens if the null zone is located in eccentric gaze (Abadi and Pascal, 1989).

A rare form of nystagmus, periodic alternating nystagmus, is a conjugate horizontal jerk nystagmus that periodically reverses its direction (Abadi and Pascal, 1994). According to these authors, this form has a higher prevalence among individuals with albinism, and the null zone in this type of nystagmus may shift periodically and unpredictably. Nystagmus in individuals with albinism might be a result of anomalous visual pathways and foveal hypoplasia (Collewijn et al., 1985; Cucchiaro, 1985). Excessive transmission of light into the retina can lead to light fogging and reduction in visual capability, which may also result in nystagmus (Abadi and Pascal, 1989). Nystagmus can be influenced by many factors, such as gaze positions, vergence and fixational eye movements (Abadi and Dickson, 1986). The level of stress or fatigue experienced by the subject also has a significant effect on nystagmus intensity and hence on visual acuity (Abadi and Pascal, 1989).

Photophobia (an undue sensitivity to light) may be caused by several factors such as hypopigmentation of ocular tissues which are normally pigmented. It is a common problem in individuals with albinism and caused by the lack of uveal and retinal pigment resulting in high transmission of light to the retina. Light scatter by the iris and multiple internal reflections produce both discomfort and disability glare, resulting in photophobia (Abadi and Pascal, 1989).

A high incidence of strabismus has been associated with albinism (Perez-Carpinell et al., 1992; Fulcher et al., 1995; Lyle et al., 1997). Typically 90% of people with albinism are strabismic (Lyle et al, 1997). According to Fulcher et al. (1995), 50% of the individuals with albinism were strabismic. Esotropia (80%) has been reported to be

more prevalent than exotropia (Lyle et al., 1997). An abnormal head posture has been reported to be adopted by approximately 50% of individuals with albinism, especially when reading (Perez-Carpinell et al., 1992; Fulcher et al., 1995). The abnormal head posture is considered to be a compensatory mechanism for the strabismus. No structural feature has, however, been implicated in the presence of strabismus in individuals with albinism. This needs to be investigated but it is not within the scope of the present study.

Although the above literature review shows that various vision problems have been reported in individuals with albinism, some of these reports were part of general studies of albinism while others were based on the study of a small number of subjects showing genetically different types of albinism. This, therefore, dictates the need for a study designed specifically to study the vision of individuals with albinism using a large subject population with only one type or where one type of OCA, such as OCA2 predominates. Such study will allow statistically significant conclusions to be drawn on the particular type of albinism.

1.3 MOTIVATION FOR THE PRESENT STUDY.

The following is the motivations for this study:

i. There are few records of previous study on vision problems in African individuals with albinism. Since genetic and environmental factors influence refractive status and other visual functions, it is expected that African individuals with albinism may have different refractive status from those from other continents due to different genetic and environmental influences. This study will verify this claim and shed light on the degree of differences in visual status between individuals with albinism in Africa and those from other continents. Also, the existing information on vision problems in individuals with albinism is either from information obtained from a general study of albinism or from vision studies conducted on a very small population, usually under 20 subjects. There is a need to conduct a study on the vision problems of a large number of subjects, in order to be able to draw statistically reliable conclusions.

ii. Although vision problems among children have been studied all over the world, no study has been specifically devoted to vision problems in children with albinism. This may be due to the low prevalence of albinism in many parts of the world. Since there are many children with albinism in South Africa, and education brings these children together in few locations, this country is ideal for this study.

iii. A large percentage of what a child learns is via the visual system, therefore it is important to provide vision care for this category of children and give the necessary assistance, so that their learning process is not disturbed. Many children all over the world, including South Africa, however, do not have the opportunity of an eye examination. Those with significant refractive errors may develop amblyopia (uncorrectable vision) due to uncorrected refractive error. There is no cure for albinism nor for the visual problems associated with the

condition. The vision can, however, be improved with appropriate vision devices (Abadi and Pascal, 1989). Our experience has shown that children with albinism attending the special education schools in the Northern Province have significant errors which need to be corrected, one of the motivation for this project is to provide the children with an opportunity for an eye examination.

iv. Individuals with albinism are likely to have colour vision deficiency. This is quite understandable because it is well documented that the macula in this category of people is not well developed. Our experience in colour vision screening in children with albinism, however, showed that not many of the children with the condition have colour vision deficiency, thus suggesting the need for a comprehensive colour vision test for individuals with albinism in order to determine the prevalence of deficiency among these people.

1.4 AIM AND OBJECTIVES

The aim of the study is:

i. To study the magnitude and prevalence of vision problems (visual acuity, refractive errors, astigmatism, contrast sensitivity, colour vision and stereopsis) among children with oculocutaneous albinism attending the special education schools in the Northern Province of South Africa.

The objectives of the study will include the following:

i To provide statistical data on vision problems studied for children with albinism in special education schools in the Northern Province of South Africa.

ii To establish if there were differences in the vision problems among these

children compared with those reported in the literature.

iii To recommend standard visual examination procedures necessary for children with albinism.

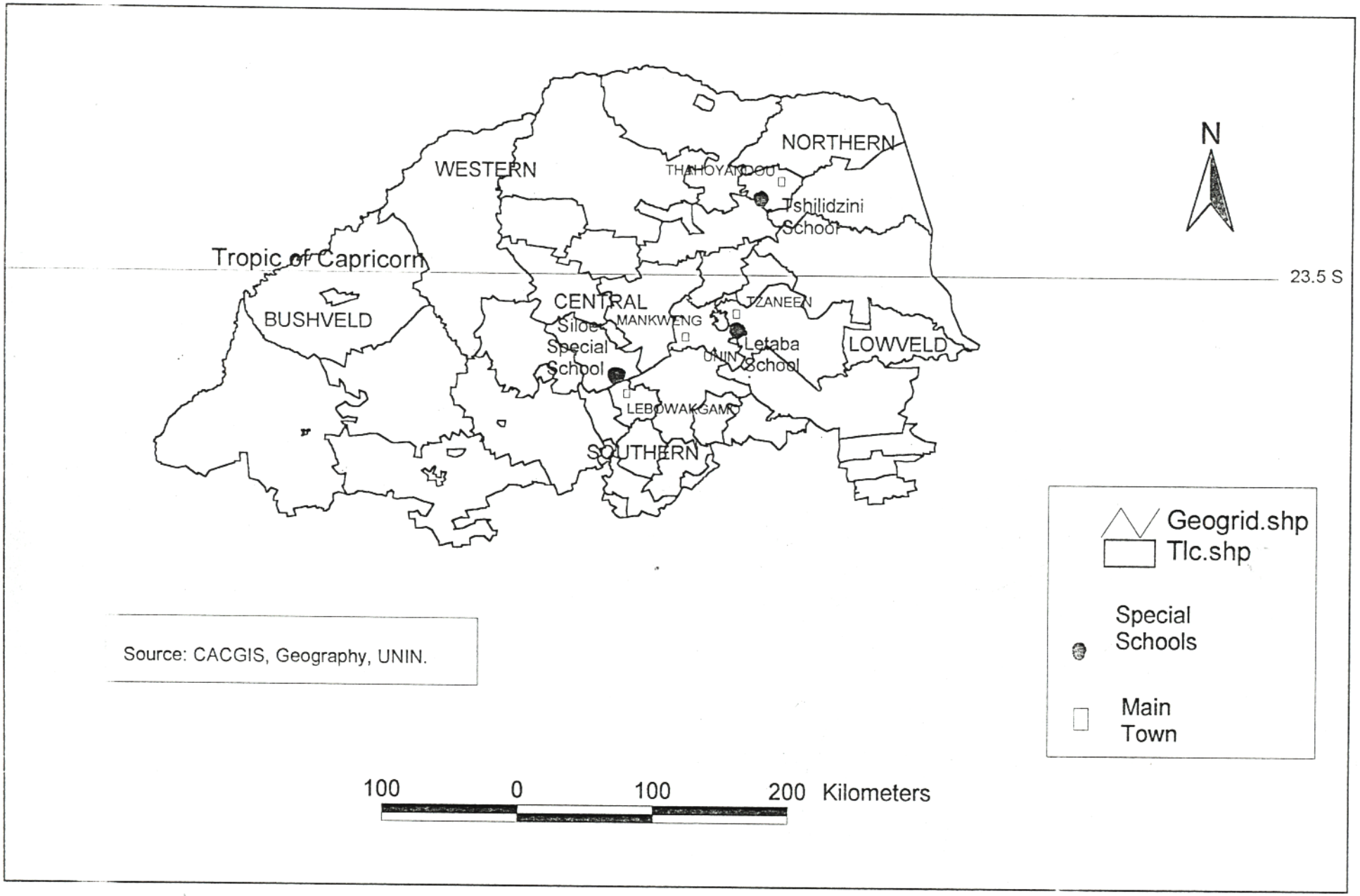
CHAPTER 2

2. SUBJECTS, MATERIALS AND METHODS

2.1 Subject and study population

It was planned to study the vision problems of between 120 and 150 children of both sexes with oculocutaneous albinism attending special education schools in the Northern Province of South Africa. This number was considered appropriate because it will be more than half of the total number of children with albinism in those schools. There were about 200 children with albinism in the schools at the time of commencement of the study. The ages of the children ranged from 7 to 17 years. The children were from Siloe school for the blind, Lebowagomo; Tshilidzini special school, (Thohoyandou) Venda; and Letaba school, Tzaneen. The locations of the schools in the Northern province are shown in Figure 1.

Fig. 1 The location of Special Education Schools in The Northern Province



2.2 Material

Table 2 below shows the tests performed, the purpose and the equipment used in each test. Certain tests (cover tests and ophthalmoscopy) were included in the procedures in order to provide comments on general ocular status of the children.

Table 2: Tests performed, purpose and equipment used for each test.

Tests	purpose	Equipment
1.visual acuity measurement (distance and near)	To measure the visual acuity of the subjects	Illiterate distance LogMAR chart (Oduntan and Briggs, 1999) (see Figure 2) and Numerical LogMAR near chart (Hyvarinen LEA) (unpublished) (Figure 3)
2. Cover tests	To determine if the subjects had tropia.	Cover paddle and measuring prism bars.
3.Retinoscopy	To measure the refractive error of the subjects objectively.	Streak retinoscopes (Welch Allyn)
4. Subjective sight testing	To determine the refractive error of the subjects subjectively.	Trial lenses (spherical and cylindrical), trial frame and Jackson cross cylinders.
5. Depth perception	To determine the ability of the subjects to perceive depth.	Randot Stereo test (Stereo Optical Company, Chicago, USA).

6. Contrast sensitivity function.	To measure the contrast sensitivity of the subjects	Distance Vistech Vision Contrast test system (Vistech Consultant Inc., 1983).
7. Colour vision test	To study the colour vision status of the subjects.	Ishihara pseudo - isochromatic plates, Farnsworth panel D-15 (Saturated and de-saturated tests)
8. Direct ophthalmoscopy	To examine the external and internal structures of the eye.	Direct ophthalmoscope (Welch Allyn)

2.3 Method

The method (procedures) employed for each test is as described below. Where considered necessary, a brief description of the test is presented for the purpose of clarity.

2.3.1. Distance visual acuity (VA) measurement

The Distance visual acuity was measured with a LogMAR illiterate chart (Oduntan and Briggs, 1999). This chart was design for use at 4 or 6 metres (see Figure 2). The subject was seated at a distance of 4 m from the chart. The room illumination was not measured because a light metre was not available. When considered necessary the lights were switched off and the amount of light in the room varied by adjusting the area of the window covered by the blind . The test procedure was explained to the subject and was instructed to cover the left eye with a cover paddle while the right eye was being tested. He or she was instructed to tell where the strokes of each optotypes on the chart were facing (up, down, left or right) beginning from the largest to the smallest ones. In cases when the subject could not see the letters at the distance of 4 m the distance was reduced to 2 m or 1 m depending on how poor the VA was. He or she

was encouraged to go on reading until a whole line was read out incorrectly. The subject was allowed but not forced to guess at threshold. The best VA line read was recorded in logMAR form with a plus or minus numerical value indicating the number of additional letters read or missed respectively in the previous lines. When the VA for the right eye had been obtained, the right eye was occluded and the left eye of the subject was tested the same way as the right. Then the subject was asked to keep both eyes open to obtain the binocular VA using the same procedure. Where test distances other than 4 m was employed, the VA values were adjusted to 4 m equivalents. For the purpose of analysis, each optotype read correctly by the subject was scored as 0.02 LogMAR unit because change in letter size between one line and the next is 0.1 log MAR, and there were five letters in every line.

2.3.2 Near visual acuity measurement

The subject was presented with the numerical LogMAR near acuity chart (see Figure 3) at a distance of 40 cm. The room illumination was not measured because the light metre was not available. Extra light , however, was not provided for near to prevent photophobia. The procedure for the measurement and recording of the near VA was similar to that used to obtain the distance VA.

Figure 2: Showing a picture of the Arabic letter distance logMAR chart used to measure the distance visual acuity (Oduntan and Briggs, 1999)

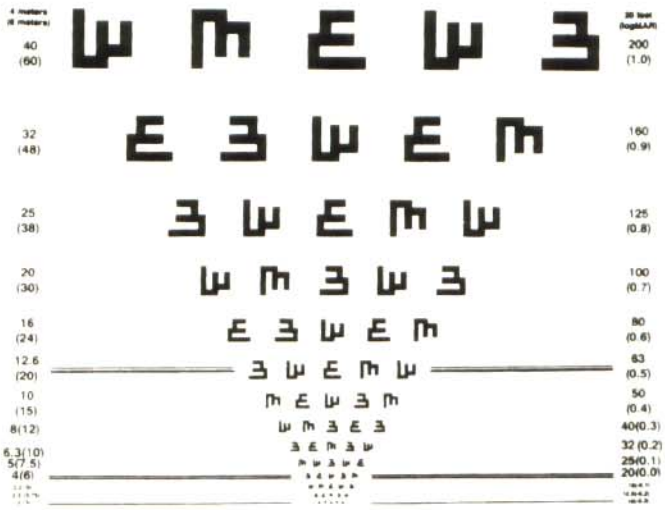
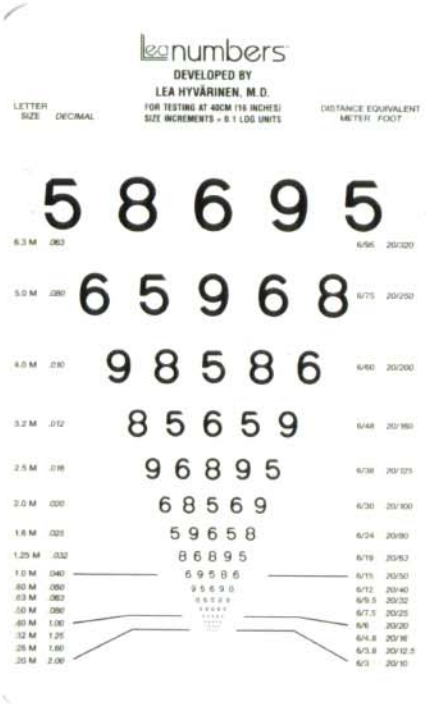


Figure 3: Showing a picture of the numerical logMAR (Lea numbers) chart used to measure near visual acuity.



2.3.3 Cover tests

The tests were: unilateral and alternating cover tests. These tests were used to determine if the subject was strabismic or heterophoric and the amount of the tropia was measured. The amount of phoria was not measured because it was difficult due to the nystagmus movement in the eyes of the children. For the same reason, vertical tropias were not measured. Also, the cover tests were performed only at near only because, nystagmus was minimal at near.

2.3.3.1 Unilateral cover test at 40 cm

Many of the subjects had no previous prescription, and those who had did not have them during the test. The test was done without the prescription. The subject was instructed to look at a large number on the numerical LogMAR near chart placed 40 cm. Illumination was sufficient enough so that the movement of the eye being examined could be observed but always at a level low enough to avoid photophobia.

The occluder was placed in front of the right eye, held there for some few seconds and then removed. During this time the left eye was observed for any movement. When no tropia existed, the left eye made no movement, maintaining steady fixation both when the right eye was covered or uncovered. If the right eye was strabismic (deviating either inward, outward, up or down), the left eye would still maintain fixation when the right eye was covered and when it was uncovered, just as if there were no strabismus. However, if the left eye was strabismic, covering the right eye caused it (left eye) to turn outward (in esotropia) or inward (in exotropia) to take up fixation. Removal of the cover caused the eye to move back to its strabismic position. The left eye was then covered for some few seconds and opened. The right eye was observed for any movement. Absence of any movement of the right eye indicated either that there was no tropia in that eye or that the left eye might be strabismic. A movement of the right eye on covering the left eye, however, indicated strabismus. An outward movement indicated esotropia and an inward movement indicated exotropia. The type of tropia (exotropia or esotropia) was recorded on the record form.

2.3.3.2 Alternating cover test at 40 cm

This test provides a method of measuring the magnitude of the ocular deviation in both strabismic and non-strabismic conditions. It does not determine whether the patient is strabismic or not. The subject was instructed to fixate a best acuity number at 40 cm to maintain accommodation and keep the small number in focus. The amount of ocular deviation, strabismus (tropia) was measured (estimated) using a prism bar. A low power from the prism bar was placed in front of one eye. The occluder was placed in front of one eye for few seconds, and then quickly removed and placed in front of the other eye. As the occluder was moved from one eye to the other, the eye without the prism in front was observed for any movement. Prism power was gradually increased until a reversal movement was noted. If exo (eye turned inwards) movement was observed prior to the use of prisms, base in prism was increased until no movement other than the usual nystagmus movement was noted, and it was then further increased until eso (eye turned outwards) movement was noted. If eso movement was observed prior to the use of prism, base out prism was increased until no movement other than the usual nystagmus movement was noted, and it was then further increased until exo movement was noted. The prism that neutralized the tropia was the one before reversal movement was observed. The magnitude of tropia for each subject was recorded.

2.3.4 Retinoscopy

The subject seated comfortably in a dimly lit room. The trial frame was properly adjusted on her or his face. Neither eye of the subject was occluded. The subject was instructed to fixate a large letter on a distance chart at 6 metres. The test was done at a distance of 67 cm. The subjects's eyes were fogged with +1.50 diopter lenses (to compensate for the working distance). The retinoscopic light was driven across the pupil along the 0-180 and then 90 degrees meridians. The movement of the retinoscopic reflex inside the eye relative to the retinoscopic light movement on the iris was observed. If movement was seen when scoping, spherical lenses (and cylindrical lenses if necessary) were added until movement was neutralized (no relative movement) as explained below. Plus spherical lenses were used to neutralize "with"

movement (the retinoscopic reflex and light on the iris moving in the same direction) and minus spherical lenses were added to neutralize "against" movement (retinoscopic reflex and the light on the iris moving in opposite directions). If there was a "with" movement in the two meridians, the meridian with the slowest "with" movement was neutralized first using plus spherical lenses. When the meridian was neutralized, then the movement in the other meridian was checked to determine if it was "against" or "neutral" (no relative movement). If "neutral", no lenses were needed, if "against", it was neutralized with minus cylindrical lenses with appropriate axis. If there was an "against" movement in both meridians, the meridian with the fastest "against" movement was neutralized first with minus spheres and the other meridian was neutralized with negative cylindrical lenses with appropriate axis . When the "against" movements in the two meridians had the same magnitude, after neutralizing one meridian with minus spheres, the other meridian was found to be neutral and no cylindrical lenses were added. If there was a "with" movement in one meridian and an "against movement" in the other, the meridian with the "with" movement was neutralized first with plus spherical lenses and the other meridian with an "against" movement was neutralized with minus cylindrical lenses with appropriate axis. The two meridians were subsequently checked for neutrality. The findings (spherical, cylinder and axis) were recorded as the objective refraction value. The visual acuity through the lenses was also measured and recorded.

2.3.5 Subjective monocular sight testing and binocular balancing

Following retinoscopic procedure, the subjective sight testing was done. One eye, usually the left was occluded while testing the other eye. The subject's attention was directed to the best VA line obtained from the retinoscopic procedure. A spherical lens chosen from retinoscopic value was placed in a trial frame. He or she was asked to read smaller prints on the VA chart while appropriate lenses were added in the trial frame. The lens was adjusted (power increased or decreased) until the best spherical lens was obtained. The cylindrical lens obtained from the retinoscopic procedure was then placed on top of the spherical lens with the determined cylindrical axis. The subject was then asked to read the next smaller prints. The lens power was changed

according to his or her response until the best cylindrical power was obtained. The Jackson cross cylinder was used to refine the axis and power of the cylinder. The spherical and the cylindrical power obtained constitute the corrective lenses for that eye. The other eye was then tested the same way to get the best lens or lenses for that eye. With the best refractive corrections on both eyes, the eyes were opened and the subject was asked to read smaller prints binocularly on the VA chart. Binocular balancing of the lenses (when considered possible and necessary) was done by fogging both eyes with +1.00 lens. The patient was asked to fixate the letters in the best VA line. The eyes were occluded alternately, and the patient was asked to tell which eye has better vision. The eye with better vision was fogged (vision reduced with plus lenses) in steps of +0.50 lens until vision in both eyes was of equal clarity. The fogging lens was then reduced binocularly in steps of 0.25 or 0.50 diopters depending on the magnitude of the refractive error, until best VA was achieved. The lenses providing best VA in each case were recorded as the corrective lenses for that subject.

2.3.6 Contrast sensitivity measurement

The Vistech contrast sensitivity chart has 1.4 degrees circular patches of sinusoidal gratings. The chart has 5 rows of patches, each containing a different spatial frequency (1.5, 3, 6, 12, and 18 cpd). Each row contains eight contrast levels; highest on the left, lowest on the right. There is a ninth column on the far right containing zero contrast circular patches. The subject sat comfortably at a distance of 3 metres from the chart. The room illumination was high enough for the patient to see the gratings. The test was done binocularly because it was observed that the contrast sensitivity of the children was very poor. The CS was measured without and with the best optical correction for those subjects with a prescription. The subject was instructed to look on the chart and tell grating orientations. He or she was allowed to guess at his or her threshold, based upon appearance. The contrast sensitivity of the patient was determined by the lowest contrast grating patch orientation that he or she identified correctly. The lowest contrast grating patch that the subject could identify on each cpd was recorded on the record form.

2.3.7 Stereo acuity test

The Randot Stereo test was used to measured the stereo acuity of each subject at a distance of 40 cm. The test consists of three subsets of vectographs which include: six shapes used for testing gross stereopsis (600 seconds of arc disparity); 3 rows of 5 cartoon animals per row, the figures in each row having the same disparity of 400, 200 and 100 seconds of arc respectively. The last subset consisting of 10 circular disparity areas, the disparity ranging from 400 to 20 seconds of arc. The room illumination (was not measured for reason given earlier) but high enough to observe the test targets but not at a level that may cause photophobia. Stereoacuity was measured with the best optical correction. The subject wore a pair of polarized glasses over his or her corrective lenses given after the subjective refraction. The subject was asked to identify each of the shapes used in the gross stereopsis test. After identifying all the shapes, he or she was asked to point to which of the cartoon animals seemed to stand out from the page. If the subject could identify all the animals, he or she was instructed to point out the circle that seemed to stand out from the others. Each subject was allowed to guess at thresholds (where he or she found the test difficult) during the test. Any target missed by the subject was presented again. If he or she got it, the test was continued. If he or she failed to get it at a second attempt, the test was stopped. The level of stereopsis of the last target chosen correctly was recorded in seconds of arc.

2.3.8 Colour vision testing: Ishihara plates (with subjective prescription).

The subject was seated comfortably on a chair. He or she was instructed to hold the Ishihara test plates at a distance of 75 cm or closer when necessary, and such that the plane of the paper was at right angle to his or her line of vision. The subject was asked to read the numbers on the plates. An assessment of readings of the plates determined the normality or defectiveness of colour vision. If the subject could read numbers on 13 or more plates the colour vision was regarded as normal, if numbers on only 9 or less plates were read, the colour vision was regarded as deficient. The type of colour vision deficiency was recorded as indicated in the manual.

2.3.9 Colour vision testing: The Farnsworth Panel D-15 test (with subjective prescription).

The Farnsworth panel D-15 test is a shortened version of the well-known Farnsworth-Munsell 100 Hue Test. It is intended for screening purpose rather than for in-depth study of a colour vision defect. There are two test types: saturated (standard test) and desaturated. The Munsell Hue (type of colour) are the same for these two tests. The desaturated is more demanding because the colour samples are less saturated (by 2 units of Munsell chroma) and lighter (by 3 units of Munsell value) (Atchison et al., 1991). The desaturated test is chiefly useful for detecting slight anomalies in colour perception, as well as for confirming that a subject has good colour grading ability. Each set of discs contain a reference or pilot cap and fifteen numbered discs. The samples are chosen to represent approximately equal hue steps for the natural colour circle. The movable caps are numbered on the back according to the ideal colour circle.

The tests were conducted at the patient's working distance under daylight illumination (near a window). The tests were administered in the same sequence for all patients, i.e. first the standard D-15 and then the desaturated D-15. The caps were pre-arranged in random order on a white surface. The subject was instructed to arrange the caps in sequential order of colours starting with the cap closest in colour to the fixed reference cap. The order of the caps was plotted directly on the score sheet on a diagram that showed correct cap positions extending in a circle from the reference cap. A line was drawn connecting the points representing the caps as arranged by the subject. In the correct order the line retraced the hue circle for normals. In this study we adopted a pass-fail criterion such that "test failure resulted when a subject made more than two simple errors or more than one major crossing. According to Fletcher and Voke (1985) a simple error constituted skipping a single number in sequence and major error occurs when distance caps are placed next to each other; the line connecting these caps crosses the hue circle. All subjects who failed the test at the first attempt were retested to minimise experimental errors in the form of confusion, misunderstanding of instruction and compliance. The type of colour vision deficiency

was recorded on the record form.

2.3.10 Ophthalmoscopy

The patient was seated comfortably looking at a small fixation object 4 metres away. Illumination level in the room was reduced to a low level to allow pupil dilation. The ophthalmoscope was used to examine the external and the internal part of the eye. The structures examined for normality included: eyelids, conjunctiva, cornea, iris, aqueous and vitreous humour, the crystalline lens, fundus, disc margins, cup disc ratio, blood vessels and foveal reflex.

2.4 Ethical and Legal Considerations.

i. All tests performed during this project were within the scope of optometric practice in South Africa. All tests were non-invasive and none presented discomfort to the subject.

ii. The proposal was previously approved by the University of the North Senior degrees and Ethics Committees.

iii. Permission was obtained from the parent of every child, principals of the schools and the Department of Education, Northern Province before the commencement of the project.

iv. The consent form (appendix A) was sent to the parent of each child for completion through the children. Only children whose parents gave consent for the child to participate in the study was examined.

V. The procedure of each test was explained to each subject before the test was carried out.

Vi. The results of the vision examination was made available to the school authorities to be forwarded to the parents of the children.

Vii. Individual results will be kept confidential.

2.5 DATA ANALYSIS, REPORTING OF DATA AND IMPLEMENTATION OF RESULTS.

Data collected during the test procedures was recorded in a specially designed form (appendix B) . Descriptive statistics was applied to analyse results using the SPSS and Microsoft Excel statistical packages.
Most of the results were reported in the form of frequency tables and graphs generated by the statistical packages.

The outputs of the study will include the following:

1. M.Sc dissertation.
2. Presentation to Professional colleagues and the staff of the special schools.
3. Publications in peer reviewed journals.

CHAPTER 3

3. RESULTS

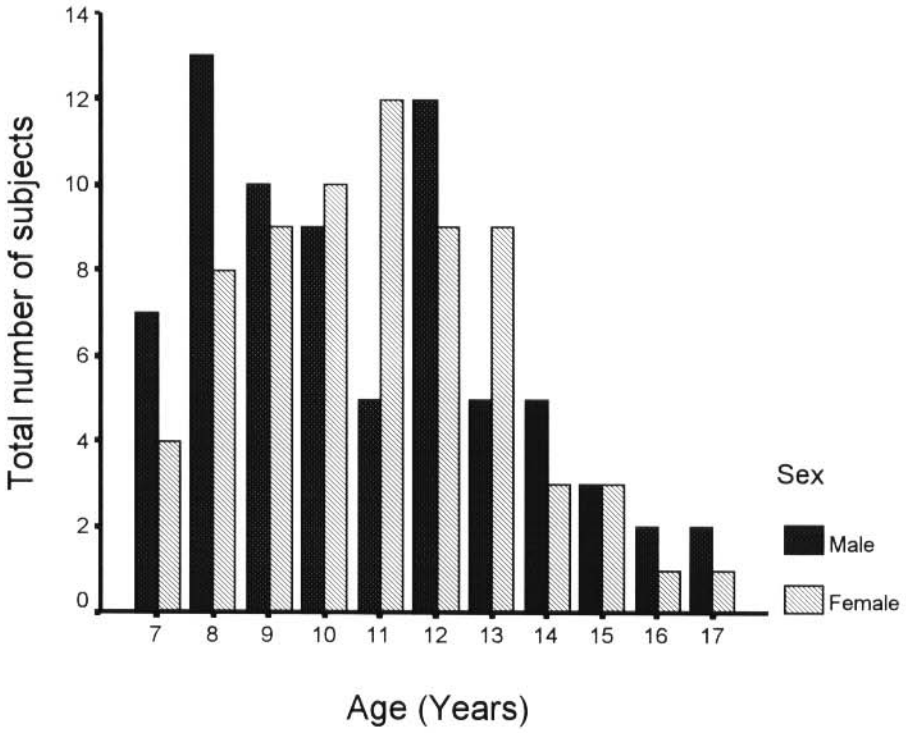
3.1 SUBJECTS, EXTERNAL EXAMINATION AND OCULAR ALIGNMENT.

The subjects included in the study were 153 black South African children with oculocutaneous albinism. There were 77 (50.3%) males and 76(49.7%) females. Their ages ranged from 7 to 17 years with a mean of 10.76 +/-2.50 years. A frequency distribution of their ages is shown in Table 3. The age and sex distributions are shown in Figure 4. Most of the subjects, 41(92.2%), were 7-14 years of age. There were 6 subjects aged 15 and only 3 subjects each in the 16 and 17 years age groups.

Table 3: Frequency distribution of ages of the subjects and the percentages. A large proportion of the subjects were aged between 7 and 14 years.

Age groups (Years)	Number of subjects	Percentages (%)
7	13	8.5
8	22	14.4
9	19	12.4
10	21	13.7
11	19	12.4
12	22	14.4
13	14	9.2
14	11	7.2
15	6	3.9
16	3	2.0
17	3	2.0
TOTAL	153	100.0

Figure 4: The graph showing the age and sex distribution of the subjects. There were more males between the ages of 7 and 9, thereafter, the number of females increased up to the age of 13, then the number of males slightly increased.



External observation showed that 147 children had wheat-straw hair, 3 had white hair and 3 had brown hair. On the basis of these phenotypes 147 (96.1%) children were considered to have tyrosinase - positive oculocutaneous (OCA2) albinism, 3 (2.0%) tyrosinase- negative (OCA1) and 3 (2.0%) had brown albinism. The colour of the iris ranged from blue, hazel to grey. Thirty one (20.3%) of the subjects had pigmented macules, whereas, 122 (79.7%) did not have. No child had significant ocular pathology. Plates 1 to 6 show the children with albinism.

Plate 1: A group of boys with oculocutaneous albinism at Siloe school.

Plate 2: A Group of girls with oculocutaneous albinism at Siloe school

Plate 3: Three boys with tyrosinase positive oculocutaneous albinism (OCA 2).

Plate 4: A boy at Tshilidzini school (Venda) with tyrosinase positive oculocutaneous albinism (OCA 2) and pigmented spots on the face and other parts of the body exposed to the sun. This shows the ability of his skin to produce some melanin pigment.

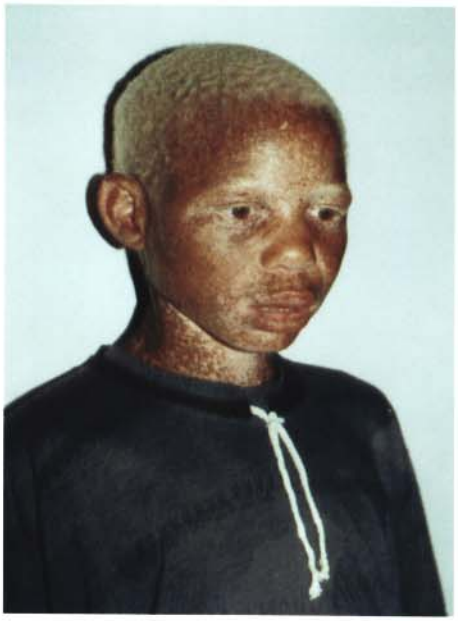


Plate 5: A girl at Siloe school with brown albinism.

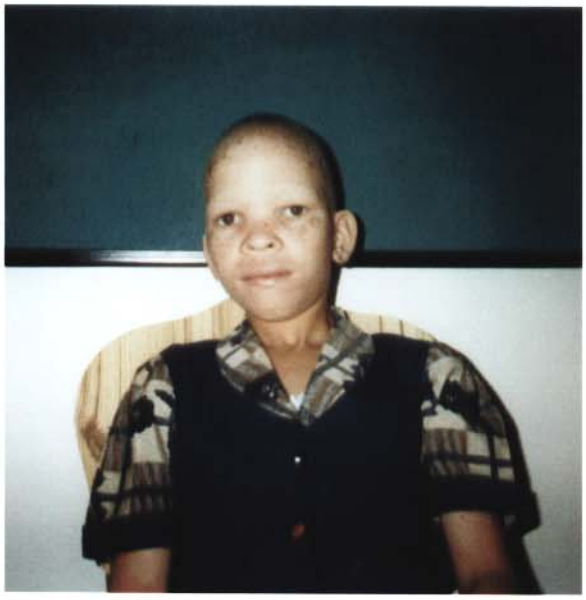


Plate 6: The examiner examining a learner with oculocutaneous albinism.

Family history revealed that the number of children per family ranged from 1 to 10; and the number of children with albinism per family ranged from 1 to 5. The percentage of albinic children per family ranged from 10 to 100 percentage. A large proportion (150) (98.0%) of the children reported that their parents were without any form of albinism. Only the mothers of 3 (2.0 %) children, two of these were twin sisters were albinic.

The main visual complaint of the subjects (78, (51.0%)was reduced vision at near and far. Fifty nine subjects (38.6%) reported reduced vision at distance only whereas, 3 (2.0%) reported reduced vision at near only. Thirteen (8.5%) subjects reported good vision at near and distance. Photophobia was a common complaint among all the children.

Estimation of ocular deviation by cover tests revealed that 53 (34.6%) subjects had tropia. These included 25 (47.2 %) exotropia and 28 (52.8 %) esotropia. The magnitude of exotropia ranged from 2 to 25 prism diopters with a mean of 6.96 +/- 5.63 prism diopters, whereas, esotropia ranged from 2 to 30 prism diopters with a mean of 8.96 +/- 7.50 prism diopters. The summary of ocular alignment findings are shown in table 4. The ocular alignment data for the children are presented in appendix D1. All the subjects had nystagmus, horizontal with occasional oscillatory or mixed nystagmus.

Table 4. Showing the ages, number of eyes with exotropia or esotropia and the mean values of the tropias. Both exo and esotropias were common among the children. The highest mean values were 19 pd for esotropia and 15.5 pd for exotropia and these occurred among the children aged 9 years.

AGE	EXOTROPIA		ESOTROPIA	
	Number of eyes	Means (Δ)	Number of eyes	Means (Δ)
7	1	4.00	0	0.00
8	1	6.00	1	4.00
9	2	15.50	2	19.00
10	5	7.0	5	15.50
11	3	7.33	4	12.50
12	4	6.00	4	6.50
13	0.00	0.00	5	5.60
14	3	3.33	3	13.33
15	4	7.50	2	5.00
16	0.00	0.00	1	4.00
17	2	6.00	1	4.00
TOTAL	25	6.96	28	8.96

3.2 VISUAL ACUITY

The uncorrected and corrected distance visual acuities are presented in Table 5.

The uncorrected and corrected near visual acuities are presented in Table 6.

The improvement in the near VA (monocular and binocular) ranged from 1.0 to 5.0 logmAR (one to 5 acuity lines) (5 to 25 letters).

The distance visual acuities (OD, OS, OU) (uncorrected and corrected) and the

improvement in vision following optical correction are shown in Table 7. The table shows that the maximum improvement in VA recorded monocularly was better than binocular visual acuity. The mean uncorrected and corrected distance VA values for OD, OS and OU for each age group are illustrated in Figures 5,6 and 7. As revealed by the t-tests, there was no significant difference in uncorrected visual acuity between the right and left eyes ($t= -0.456$, $p= 0.647$). Similarly, there was no significant difference between the corrected visual acuities of the right and the left eyes ($t= 0.07$, $p= 0.944$). With optical correction, there was no significant difference in the improvement in visual acuity between the right and the left eyes ($t= 1.06$, $p= 0.291$). Comparison of uncorrected and corrected visual acuities showed that the corrected values were significantly better than the uncorrected values. The corrected OD was significantly better than uncorrected ($t= -17.36$, $p< 0.0001$). Also, the corrected visual acuities for OS was significantly better than the uncorrected ($t= -17.38$, $p < 0.0001$). The corrected OU visual acuities was significantly better than uncorrected ($t= -17.92$, $p < 0.0001$).

A large proportion, 71.2%, of the children showed a significant improvement in VA of one to three logMAR acuity lines binocularly with the best optical correction (see Figure 8 showing visual acuity improvement versus age). However, 17% had VA less than one line (less than 5 letters). There was no improvement in visual acuity in 11.8% subjects with the best optical correction. One hundred and twenty nine (84.3%) subjects were classified as low vision patients (VA worse than 6/18 for the better eye with the best optical correction). Twenty four (15.7%) were classified as having normal vision.

Table 5: Showing the ranges of uncorrected and corrected distance visual acuities for the right, left and both eyes in Snellen acuity (Metres and Feet) and LogMAR forms.

Eye	(VA) SNELLEN ACUITY				(VA) LogMAR	
	Uncorrected		corrected		Uncorrected	Corrected
	Metres	Feet	Metres	Feet		
OD	FC to $6/7.5^{-2}$	FC to $20/25^{-2}$	FC to $6/6^{-1}$	FC to $20/20^{-1}$	FC to 0.1^{-2}	FC to 0.0^{-1}
OS	$6/75^{+2}$ to $6/7.5^{-3}$	$20/250^{+2}$ to $20/25^{-3}$	$6/75^{+2}$ to $6/6^{-1}$	$20/250^{+2}$ to $20/20^{-1}$	1.1^{+2} to 0.1^{-3}	1.1^{+2} to 0.0^{-1}
OU	$6/60^{-1}$ to 6/7.5	$20/200^{-1}$ to 20/25	6/60 to 6/6	20/200 to (20/20)	0.1^{-1} to 1.0	1.0 to 0.0

Table 6: Showing the ranges of uncorrected and corrected near visual acuity values for the right, left and both eyes in Snellen acuity (Metre equivalent and M notation) and LogMAR forms.

Eye	(VA) SNELLEN ACUITY				(VA)LogMAR	
	Uncorrected		corrected		Uncorrected	Corrected
	Metres Equivalent	M notation	Metres Equivalent	M notation		
OD	FC to $6/9.5^{-2}$	FC to 0.63^{-2}	FC to $6/6^{+2}$	FC to 0.4^{+2}	FC to 0.2^{-2}	FC to 0.0^{+2}
OS	6/75 to $6/12^{+2}$	5.0 to 0.8^{+2}	$6/48^{+1}$ to $6/6^{+1}$	3.2^{+1} to 0.4^{+1}	1.1 to 0.3^{+2}	0.9^{+1} to 0.0^{+1}
OU	$6/60^{-2}$ to $6/12^{+2}$	4.0^{-2} to 0.8^{+2}	6/38 to $6/6^{+1}$	2.5 to 0.4^{+1}	1.0^{-2} to 0.3^{+2}	0.8 to 0.0^{+1}

Table 7 : Showing the descriptive statistics of the visual acuities (VA) (in terms of the number of letters read) (uncorrected and corrected) for the right (OD), left (OS) and both (OU) eyes (N= 153). The minimum (Min), maximum (Max), mean and (SD) standard deviation are shown. Finger counting (FC) was scored as zero logMAR for the purpose of analysis. (Please note: 1 letter read = 0.02 logMAR).

Note: The VA improvement was not derived from the min. and max. values on the table. This is because, the min. and max values belonged to different subjects, and the most improved VA belonged to another subject.

EYE	VISUAL ACUITY (logMAR)								
	Without RX				With RX				Max. VA Improvement
	Min	Max	Mean	SD	Min	Max	Mean	SD	
OD	FC (Zero)	0.96	0.37	0.17	FC (Zero)	1.08	0.50	0.18	0.40
OS	0.04	0.94	0.37	0.17	0.0 4	1.08	0.49	0.18	0.44
OU	0.08	1.00	0.40	0.16	0.10	1.10	0.52	0.18	0.34

Figure 5: Showing the mean uncorrected and corrected visual acuity scores for the right eye for all age groups. The corrected VA score s was better in each case for all age groups. The mean VA score among the 15-17 years old may not be representative enough for the age group as there were 6 children in the 15 years and 3 children each among the 16 and 17 years age group.

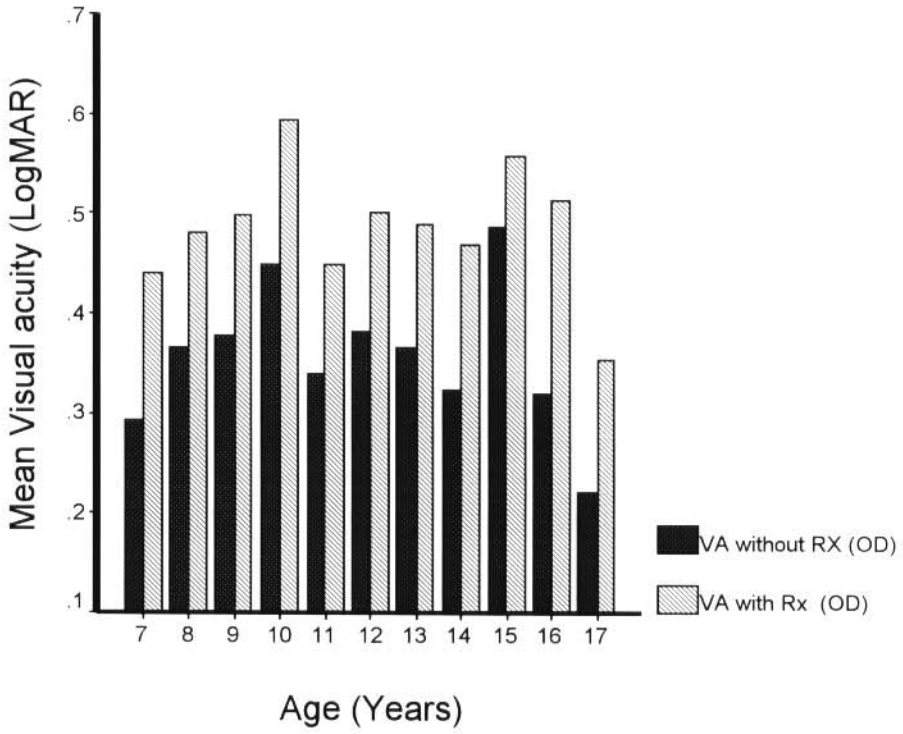


Figure 6: Showing the mean uncorrected and corrected visual acuity for the left eye for each age group. The corrected VA was better in each age group. The mean improvement in most cases was greater than 0.1 logMAR. The mean VA improvement between the ages of 15 and 17 may not be representative enough because of the reason given earlier.

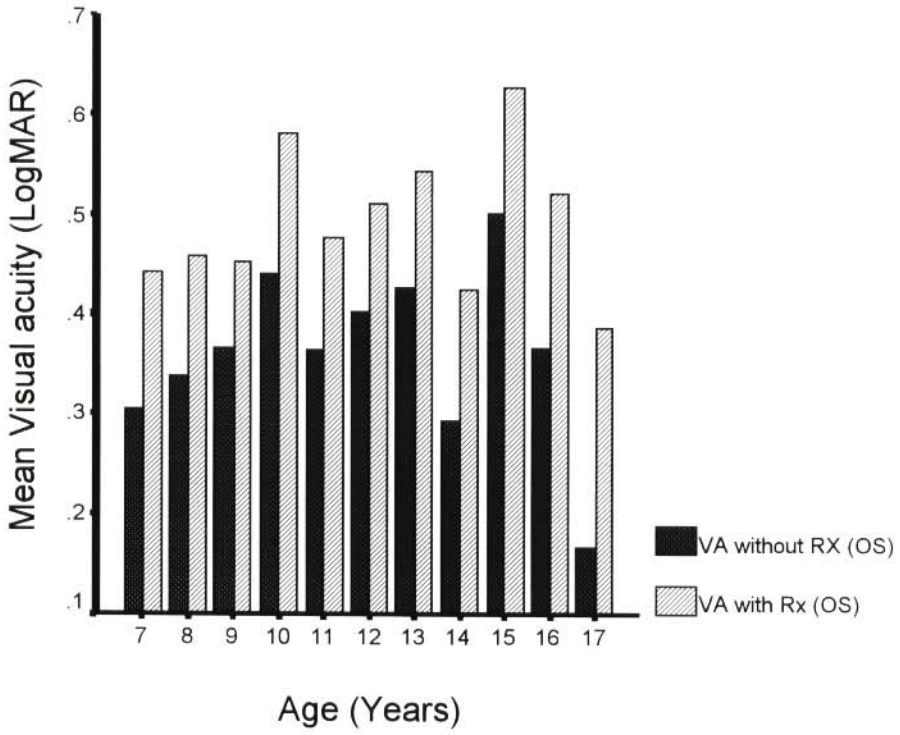


Figure 7: The graph showing the mean binocular uncorrected and corrected visual acuity of the subjects. As expected, the corrected VA was better than uncorrected for all age groups.

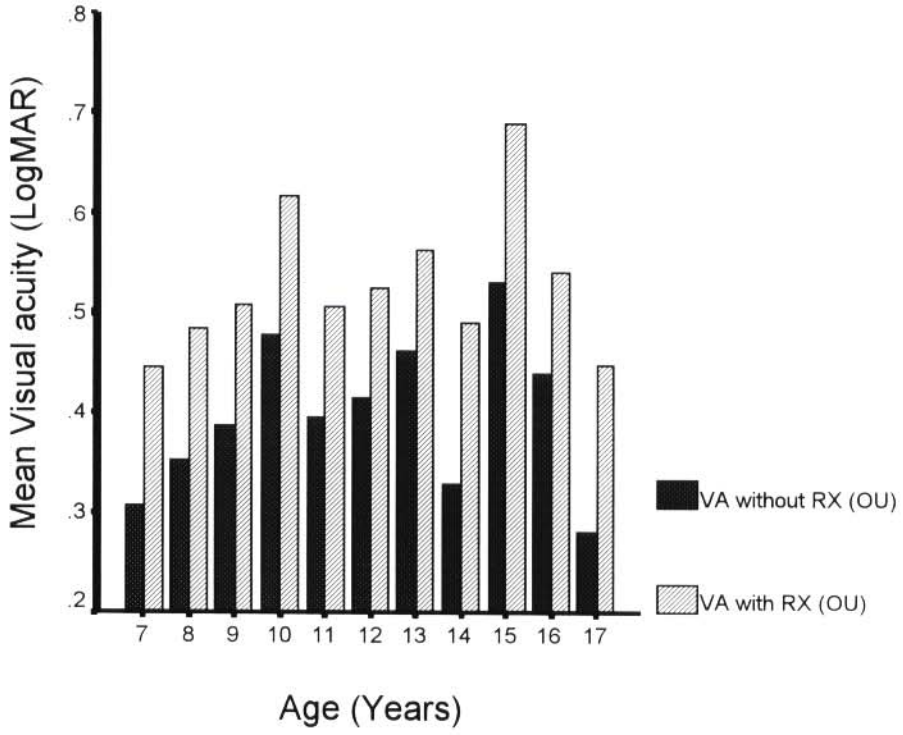
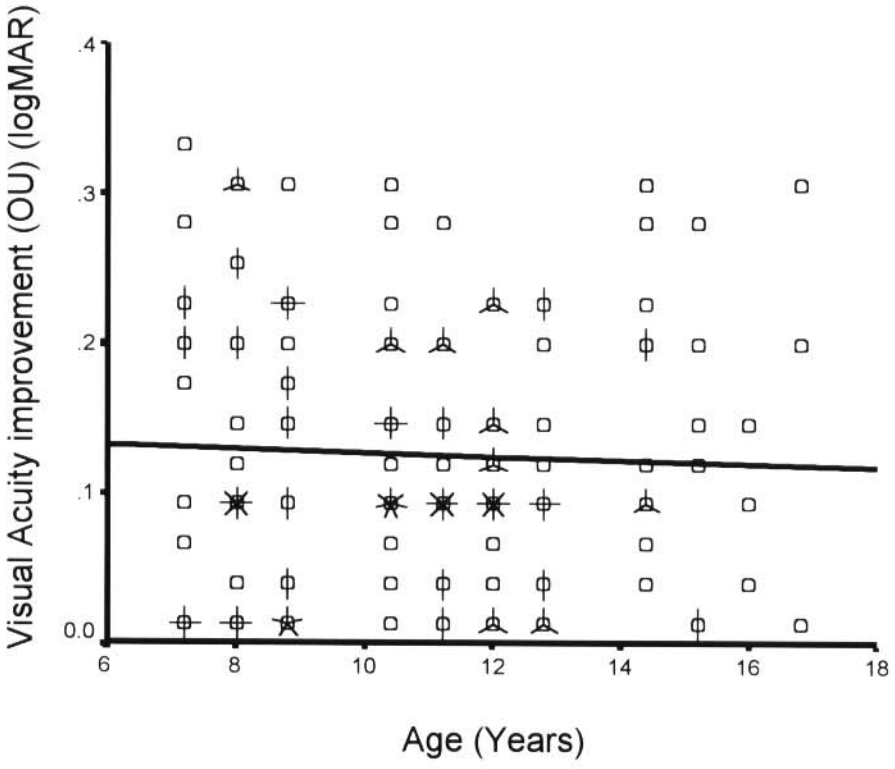


Figure 8: A scatter plot to illustrate the improvement in binocular visual acuity scores (logMAR) for both eyes with the best optical correction. Many of the subjects showed an improvement in VA of one to three lines (0.1- 0.3 logMAR) (5 to 15 letter improvement). Each square represents a plot for one subject and each petal represents an additional subject. The regression equation is y = - 0.0018 x + 0.139 where y represents the visual acuity improvement and x is the age of the patients in years. The regression line shows that there is a slight decrease in visual acuity with increase in age.



3.3 REFRACTIVE ERRORS

Many (72.6%) of the children had significant refractive errors (≥ 1.00 D). The nearest equivalent spherical powers (F_{NE} = sphere + 1/2 cylinder) for the right eye ranged from -12.00 D to +2.75 D (mean = -1.48 +/- 2.28 D) and from -8.25 to +5.75 D (mean= -1.36 +/- 2.18 D) for the left eye. Comparison of nearest equivalent spherical powers between the right and the left eyes using t-test showed no significance difference (t = -1.074, p= 0.284). Myopia (67.6%) (nearest equivalent) was more prevalent among the children than hyperopia (30.7%) or emmetropia (1.7%) (see Tables 8 and 9 and Figure 9). For those with myopia, the nearest spherical equivalent powers ranged from -0.25 D to -12.00 D for the right eye and from -0.25 D to -8.25 D for the left eye with means and standard deviation of -2.59 +/- 1.99 D and -2.38 +/- 1.79 D respectively. Among those with hyperopia, the nearest equivalent spherical powers ranged from +0.25 D to +2.75 D with a mean and standard deviation of +0.78 +/- 0.50 D for the right eye and +0.25 D to +5.75 D (mean = 0.90 +/- 0.89 D) for the left eye. A large proportion (24%) of the negative powers ($F_{NE)}$ were between -0.25 and -2.25 (see Table 9 and Figure 9). A large proportion (46.9%) of hyperopia were between +0.25 and +0.75 Diopters (see Table 9 and figure 9). The incidence of hyperopia was highest (60.5%) among those aged 9 years and lowest (0.00%) among those aged 17 years (see Table 8). There were only 3 subjects under this age group. The distributions of nearest equivalent spherical powers among males and females are shown in Table 9. The distribution of each type of nearest equivalent powers is illustrated in Figure 9. Low hyperopia (+ 0.25 to +2.00 Diopters) and low myopia (- 0.25 to -3.00 Diopters) were more common than other magnitudes of spherical errors. The distribution of the F_{NE} according to magnitude (low, medium and high) is shown in Table 10.

TABLE 8. Showing the age groups, number of eyes, number of eyes with myopia, and hyperopia (F_{NE}) and the percentage incidence of the spherical errors among the different age groups. The incidence of myopia was higher than hyperopia among all the age groups. It ranged from 39.5% among 9 years old to 100% among the 17 years olds. There were, however, only 3 subjects in the latter age group. Hyperopia was absent among the 17 years old but had highest incidence (60.5 %) among the 9 years olds.

Age groups (Years)	Number of eyes	Number of eyes with myopia			Incidence of myopia (%)	Number of eyes with hyperopia			Incidence of hyperopia (%)
		OD	OS	Total		OD	OS	Total	
7	26	9	8	17	65.4	4	5	9	34.6
8	44	14	14	28	63.6	8	7	15	34.1
9	38	7	8	15	39.5	12	11	23	60.5
10	42	11	13	24	57.1	9	8	17	40.5
11	38	14	15	29	76.3	5	4	9	33.7
12	44	18	20	38	88.4	3	2	5	11.6
13	28	10	9	19	67.9	3	4	7	25.0
14	22	9	8	17	77.3	2	3	5	22.7
15	12	5	5	10	83.3	1	1	2	16.7
16	6	2	2	4	66.6	1	1	2	33.4
17	6	3	3	6	100	0	0	0	0.00
TOTAL	306	102	105	207	67.6	48	46	94	30.7

TABLE 9. Distribution of magnitude of nearest equivalent spherical powers and the percentages (in bracket) among the males and females in the population. A large proportion of the errors were between +0.25 to +1.50 for hyperopia and between -1.00 to -2.25 for myopia. Hyperopia was more prevalent in males (17.6%) than in females (13.1%), whereas, myopia was more prevalent in females(35.9%) than in males (31.7%).

SPHERICAL POWERS	MALES		FEMALES	
	Hyperopia	Myopia	Hyperopia	Myopia
0.25-0.75	34 (62.9)	7 (7.2)	30 (75.0)	8 (7.3)
1.00-1.50	16 (29.6)	29 (29.9)	8 (20.0)	36 (32.7)
1.75-2.25	1 (1.9)	33 (34.0)	2 (5.0)	33 (30.0)
2.50-3.00	1 (1.9)	9 (9.3)	0 (0.0)	9 (8.2)
3.25-3.75	1 (1.9)	3 (3.1)	0 (0.0)	7 (6.4)
4.00-4.50	0 (0.0)	2 (2.1)	0 (0.0)	2 (1.8)
4.75-5.25	0 (0.0)	1 (1.0)	0 (0.0)	4 (3.6)
5.50-6.00	1 (1.9)	2 (2.1)	0 (0.0)	6 (5.5)
6.25-6.75	0 (0.0)	4 (4.1)	0 (0.0)	2 (1.8)
7.00-7.50	0 (0.0)	3 (3.1)	0 (0.0)	3 (2.7)
7.75 and above	0 (0.0)	4 (4.1)	0 (0.0)	0 (0.0)
TOTAL	54 (17.6%)	97 (31.7%)	40 (13.1%)	110 (35.9%)

Figure 9: The graph showing the distribution of nearest spherical equivalent powers among the subjects. Low hyperopia (+ 0.25 to + 2.00 Diopters) and low myopia (- 0.25 to - 3.00 Diopters) were more common than other magnitudes (high and medium) of spherical errors. Myopia was more prevalent except between 0.25 and 0.75 diopters.

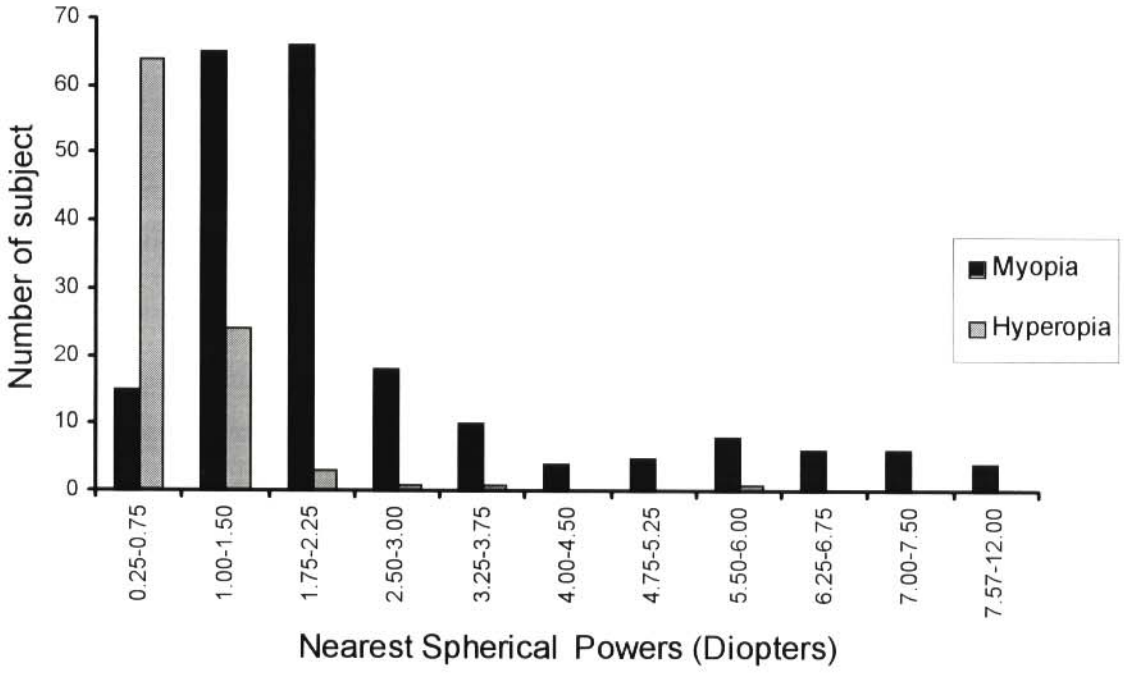


Table 10: showing the classification of refractive errors according to magnitude among males and females. Low positive and negative F_{NE} astigmatism were more common among both sexes than other magnitudes. No subjects among the females had medium positive F_{NE} and none among males and females had high positive F_{NE}. Few subjects (4.0% males and 2.0% females) had high F_{NE}.

Subjects (Sex)	Positive spherical equivalent (hyperopia)			Negative spherical equivalent (Myopia)		
	Low 0.25-2.00 D	Medium 2.25-5.00 D	High > 5.00 D	Low 0.25-3.00 D	Medium 3.25- 6.00 D	High > 6.00 D
Males	51 16.7%	3 1.0%	0	78 25.4%	8 2.6 %	11 4.0%
Females	40 13.1%	0	0	86 28.1%	19 6.2%	5 1.6 %
Total	91 (29.7%)	3 (1.0%)	0	164 (53.6%)	27 (8.8 %)	16 (5.2 %)

Astigmatism occurred in 282 (92.2%) of the eyes. One hundred and forty (140) right eyes (91.5 %) had astigmatism with the correcting cylinder values ranging from -0.25 D to -3.50 D (mean = -1.19 +/- 0.77 D). One hundred and forty two (142) left eye (92.8%) had astigmatism with correcting cylinders values ranging from -0.25 D to -4.50 D (mean = -1.22 +/- 0.77 D). T-test revealed no significant difference between the correcting cylinder powers between the right (OD) and the left eyes (OS) (t= -1.05, p= 0.284). The distribution of the corrective cylindrical powers for right and left eyes of males and females is shown in Table 11 and the frequency distribution are illustrated in Figure 10. A large proportion of the correcting cylinders (42.2%) were between -0.25 and -0.75 D. Only 2 (0.7%) subjects had correcting cylinder powers ranging from -4.00 to -4.50 D (see Table 11). Astigmatism was classified according to magnitude and the distribution according to this classification is shown in Table 12. The incidence of astigmatism was high in all age groups (72.7% to 100%) (see Table 13).

The types of astigmatism included 30.4 % hyperopic, 62.1% myopic and 7.5% simple type (see Figure 11). With-the-rule (WTR) astigmatism (axis of the corrective cylinder within 30 degrees of the horizontal meridian) (0^0 to 30^0 or 150^0 to 180^0) was more common than other types. It occurred in 131 (47.0%) eyes (see Table 14) and 55.7% occurred in females and 44.3% males. The next common type was oblique astigmatism (OBLA) (axis of the correcting cylinder located at more than 30 degrees from the horizontal or vertical meridian but outside the location of the "with-the-rule" and "against-the-rule" the rule astigmatism, from). It occurred in 80 (28.0%) eyes and (51.3% males and 48.7% in females) (see Table 14). The against-the-rule astigmatism (ATRA), (Axis of the corrective cylinder within 30 degrees of the vertical) (60^0 to 90^0 or 90^0 to 120^0) occurred in 71 (25.0%) of the eyes and 60.6% of subjects with this type of astigmatism were males and 39.4% were females (see Table 14). The incidence of types of astigmatism in relation to age in shown in Table 15 and illustrated in figure 12.

Table 11: Showing the distribution of correcting cylinder powers in the right and left eyes of males and females and percentages. A large proportion of the errors were between - 0.25 and -1.50 D.

Cylindrical powers	Number of right eyes (OD) with astigmatism			Number of left eyes (OS) with astigmatism			Total number of subjects with astigmatism
	Males	Females	Total	Males	Females	Total	
-0.25 to -0.75	27 19.3%	35 25.0%	62 44.3%	30 21.1%	27 19.0%	57 40.1 %	119 42.2%
-1.00 to -1.50	22 15.7%	20 14.3%	42 30.0%	26 18.3%	26 18.3%	52 36.6%	94 33.3%
-1.75 to -2.25	12 8.6%	10 7.10%	22 15.7%	9 6.4%	10 7.0%	19 13.4%	41 14.5%
-2.50 to -3.00	6 4.3%	6 4.30%	12 8.5%	6 4.23%	5 3.52%	11 7.8%	23 8.2%
-3.25 to -3.75	2 1.4%	0	2 1.42%	1 0.7%	0	1 0.7%	3 1.1%
-4.00 to -4.50	0	0	0	1 0.7%	1 0.7%	2 1.4%	2 0.7%
Total	69 49.3%	71 50.7%	140 100%	73 51.4%	69 48.6%	142 100%	282 100%

Table 12: Showing the classification of astigmatism errors according to magnitude and their distribution among both sexes. A large percentage (52.5%) of the children had moderate astigmatism error. Only 1.3% males and 0.3% females had high astigmatic errors.

Subjects	Astigmatism		
	Low 0.25-0.75 D	Medium 1.00- 3.00 D	High > 3.00 D
Males	57 18.6%	81 26.5%	4 1.3%
Females	62 20.2%	79 26.0%	1 0.3%
Total	119 (38.8%)	160 (52.5%)	5 (1.6%)

Figure 10: Showing the distribution of cylindrical powers among the subjects. Medium astigmatism error (1.00 - 3.00 D) was more prevalent than other magnitudes of astigmatic errors.

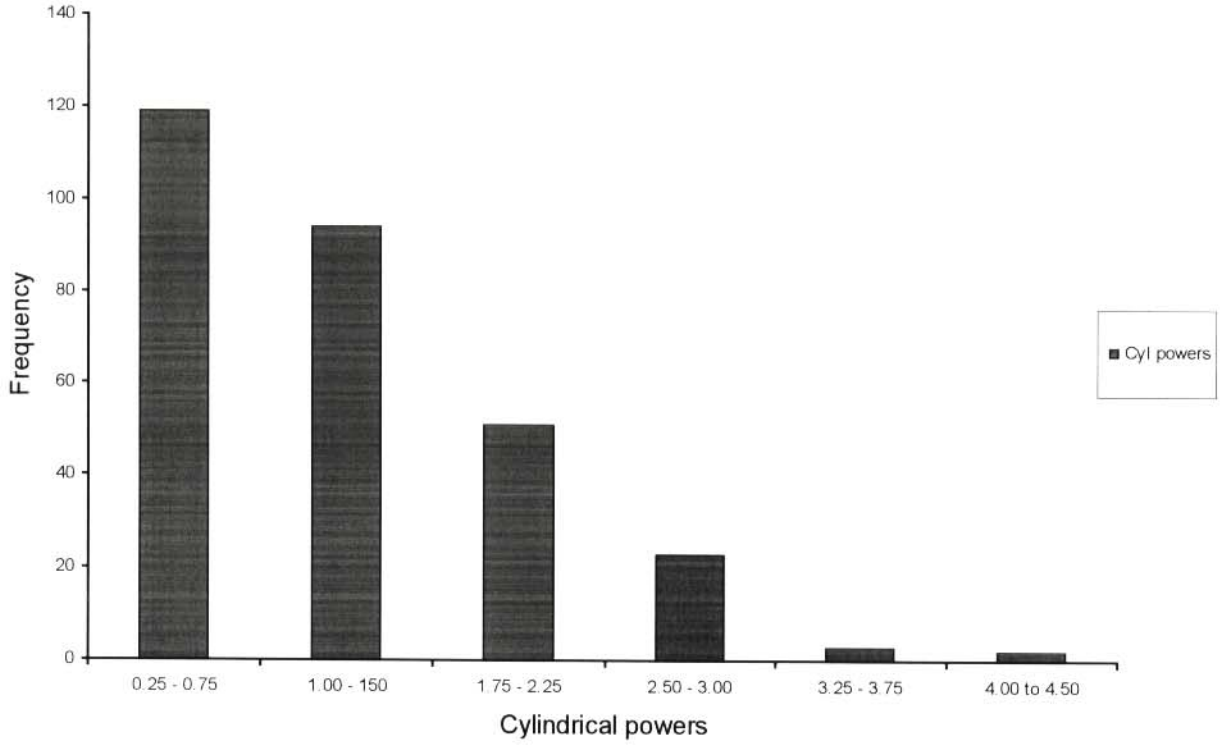


Table 13. Distribution of astigmatism and their percentages among the different age groups. The table shows that the incidence of astigmatism was high in the population, ranging from 72.7% among the 14 year to 100% among the 9, 16 and 17 age groups. The 100% incidence among the 16-17 age groups is considered to be due to the small number of subjects among this age groups.

AGE	NUMBER OF EYES	NUMBER OF EYES WITH ASTIGMATISM	INCIDENCE (%)
7	26	23	88.5
8	44	42	95.5
9	38	38	100
10	42	39	93.0
11	38	34	89.5
12	44	39	88.6
13	28	28	100
14	22	16	72.7
15	12	11	92.0
16	6	6	100
17	6	6	100
Total	306	282	

Figure 11: Showing the prevalence of types of astigmatism. With-the-rule astigmatism was more common than other types.

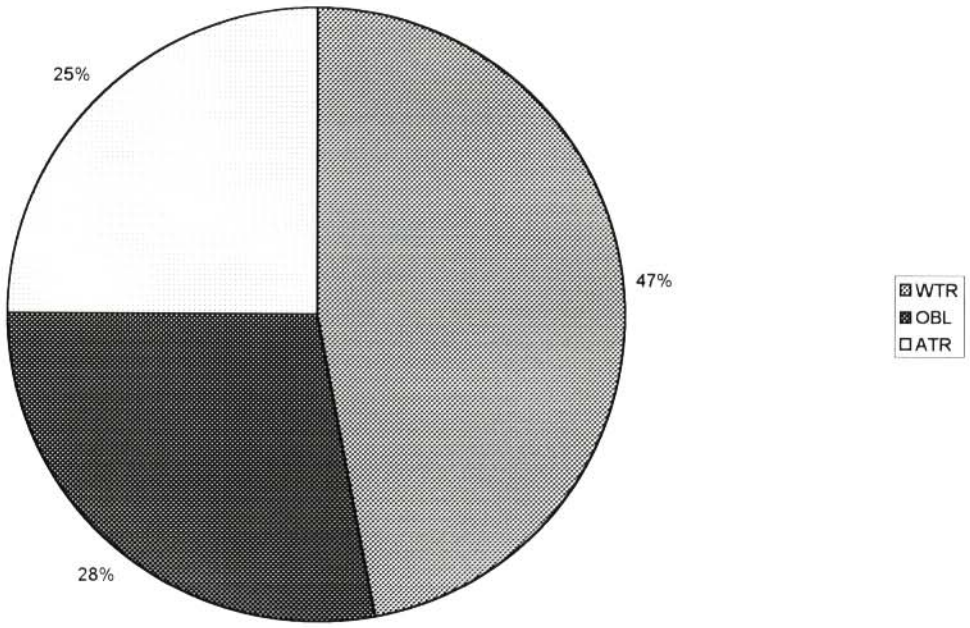


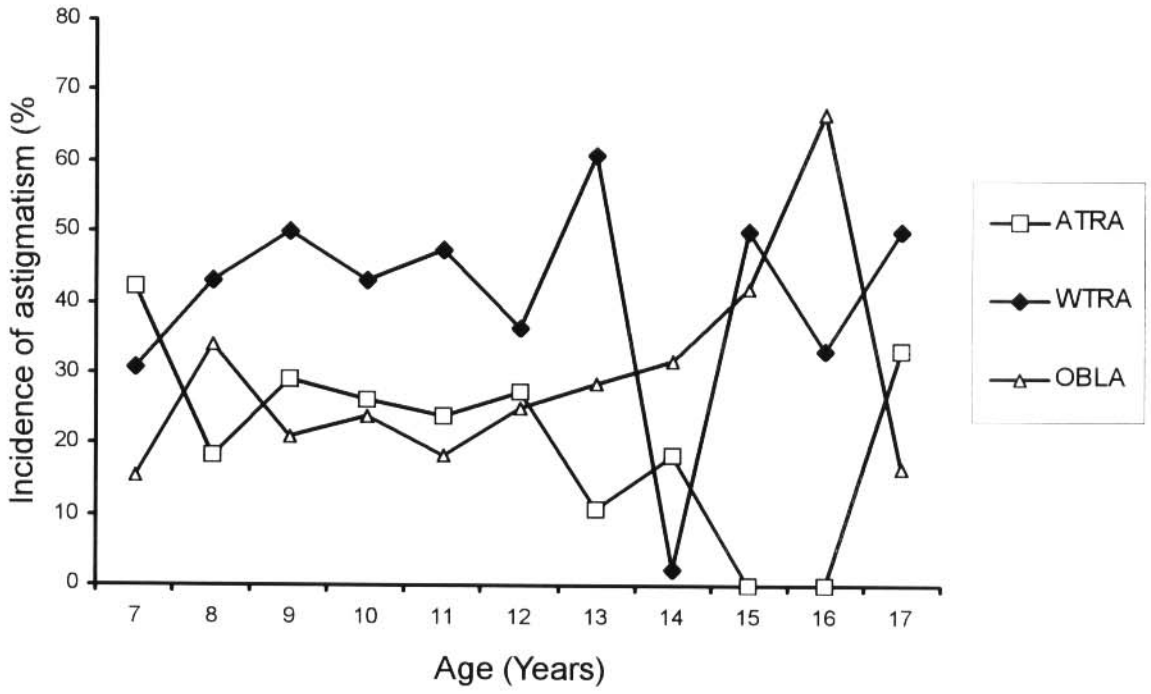
TABLE 14. Showing the distribution of the various types of astigmatism in relation to sex and the percentages (in brackets). With-the-rule astigmatism (WTRA) was more common and against- the- rule (ATRA) was the least common. WTRA was more common in females than in males and ATRA was more common in males than in females. OBLA was slightly more common in males than in females. (M is male, F is female and T is total).

	WTRA			ATRA			OBLA		
	M	F	T	M	F	T	M	F	T
OD	25 (45.5)	30 (54.5)	55 (42.0)	22 (53.7)	19 (46.3)	41 (57.7)	22 (50.0)	22 (50.0)	44 (55.0)
OS	33 (43.4)	43 (56.6)	76 (58.0)	21 (70)	9 (30)	30 (42.3)	19 (52.8)	17 (47.2)	36 (45.0)
ALL	58 (44.3)	73 (55.7)	131 (47.0)	43 (60.6)	28 (39.4)	71 (25.0)	41 (51.3)	39 (48.7)	80 (28.0)

Table 15: Showing the distribution of types of astigmatism among the subjects in each age group and the incidence (in brackets). The trend of incidence of types of astigmatism with age is not quite consistent but WTRA and OBLA appeared to increase with age whereas ATRA appeared to decrease with increasing age.

Age	No. of eyes	Number of eyes with ATRA (Incidence of ATRA %)	Number of eyes with WTRA (Incidence of WTRA %)	Number of eyes with OBLA (Incidence of OBLA %)
7	26	11 (42.3)	8 (30.8)	4 (15.4)
8	44	8 (18.2)	19 (43.2)	15 (34.1)
9	38	11 (29.0)	19 (50.0)	8 (21.0)
10	42	11 (26.2)	18 (43.0)	10 (23.8)
11	38	9 (24.0)	18 (47.3)	7 (18.4)
12	44	12 (27.3)	16 (36.4)	11 (25.0)
13	28	3 (10.7)	17 (60.7)	8 (28.6)
14	22	4 (18.2)	5 (2.40)	7 (31.8)
15	12	0 (0.00)	6 (50.0)	5 (42.0)
16	6	0 (0.00)	2 (33.3)	4 (66.7)
17	6	2 (33.3)	3 (50.0)	1 (16.7)

Figure 12: The graph showing the distribution of types of astigmatism in relation to age. The trend of the incidence of the types of astigmatism is not quite consistent.



3.4. CONTRAST SENSITIVITY

Contrast sensitivity function measurement indicated that the maximum frequency of 18 cpd can be perceived without or with correction by certain individuals but not by others. The peak sensitivity was 3 cycles/degree (cpd) without or with optical correction. The range and mean binocular contrast thresholds without and with the optical correction for all the subjects are presented in figures 13, 14 and 15 respectively. The mean CSF resolution acuity, and peak sensitivity increased slightly when measured through the optical correction (see Figure 15 and Table 17). The mean binocular sensitivity threshold for each age group does not fall within the norms published for the VCTS 6500 stippled area (See Figures 13, 14 and 15). Tables 16 and 17 show the mean values for the last target (CS)

correctly seen between 1.5 and 18 cpd spatial frequencies measured without and with the best optical correction for different age groups. Tables 14 and 15 show that there was only a slight increase in contrast sensitivity with age.

Figure 13: Graph showing the normal range of contrast sensitivity in the distance Vistech contrast sensitivity test and the range of contrast sensitivity without optical correction for the children with albinism.

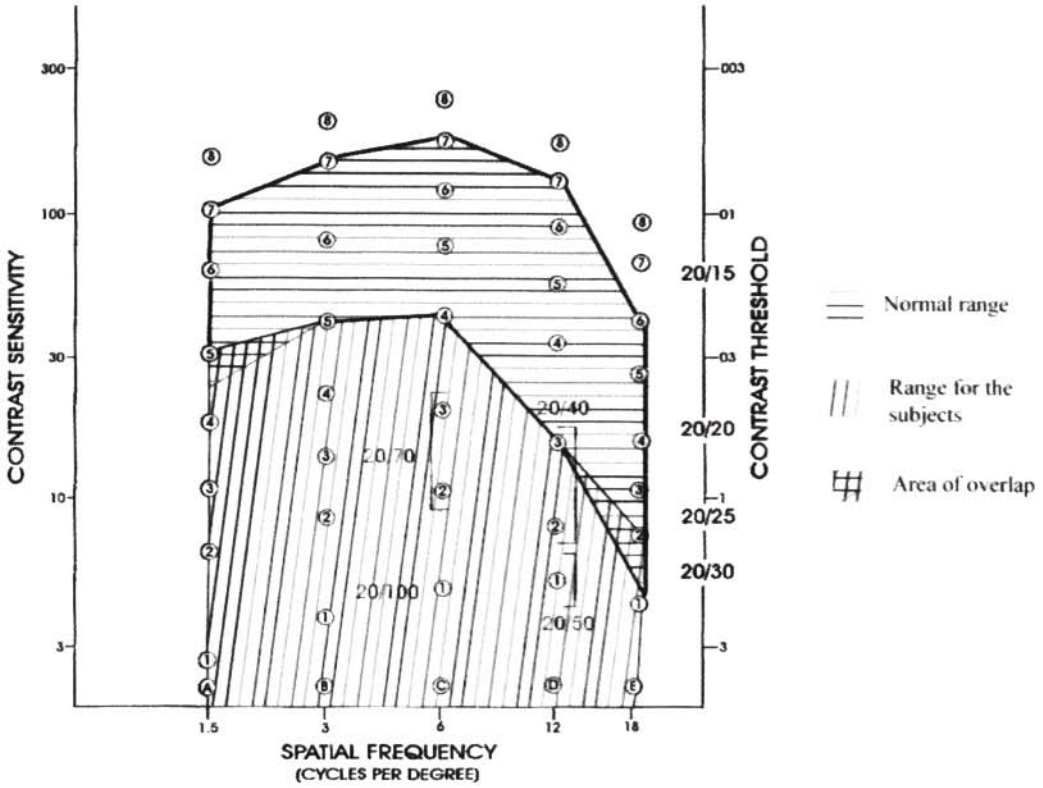


Figure 14: Graph showing the normal range of contrast sensitivity as indicated in the distance Vistech contrast sensitivity test and the range of contrast sensitivity with optical correction for children with albinism.

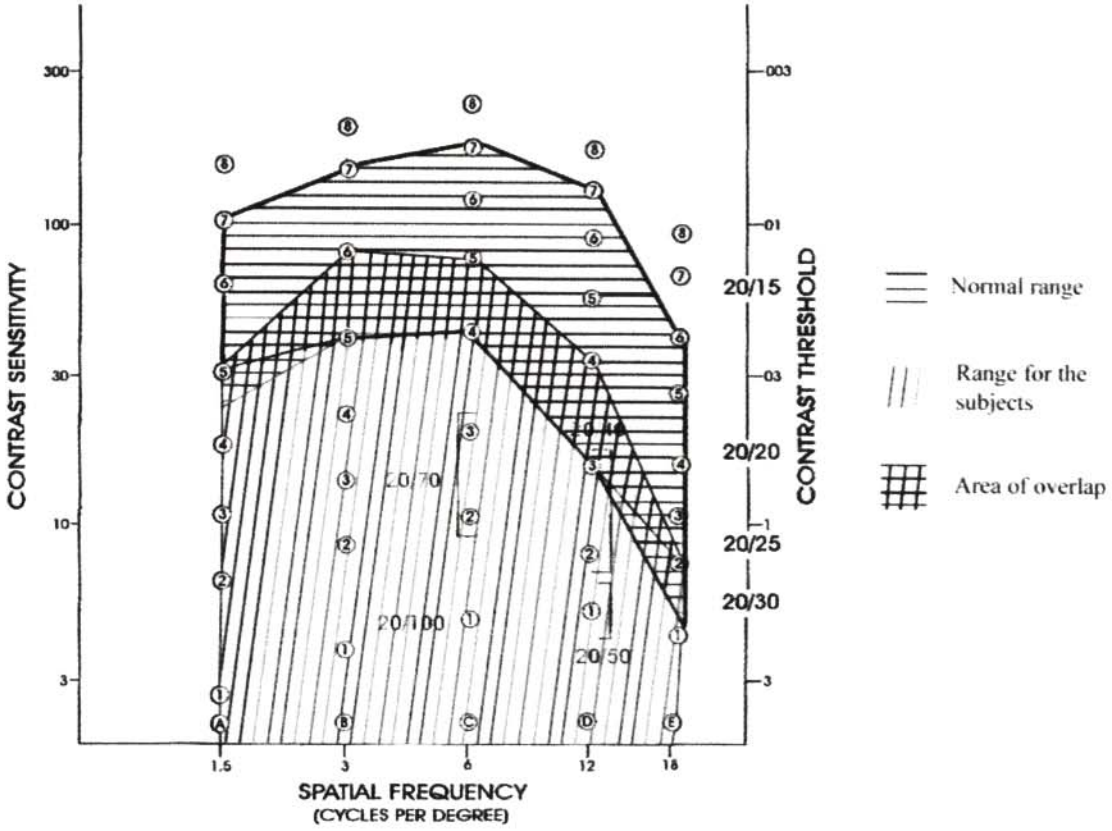


Figure 15: showing the mean binocular contrast sensitivity of all the subjects without and with the prescription. The contrast sensitivity and peak sensitivity increase slightly when measured with the optical correction. The mean values (without and with optical corrections) were quite below the average means for normal individuals.

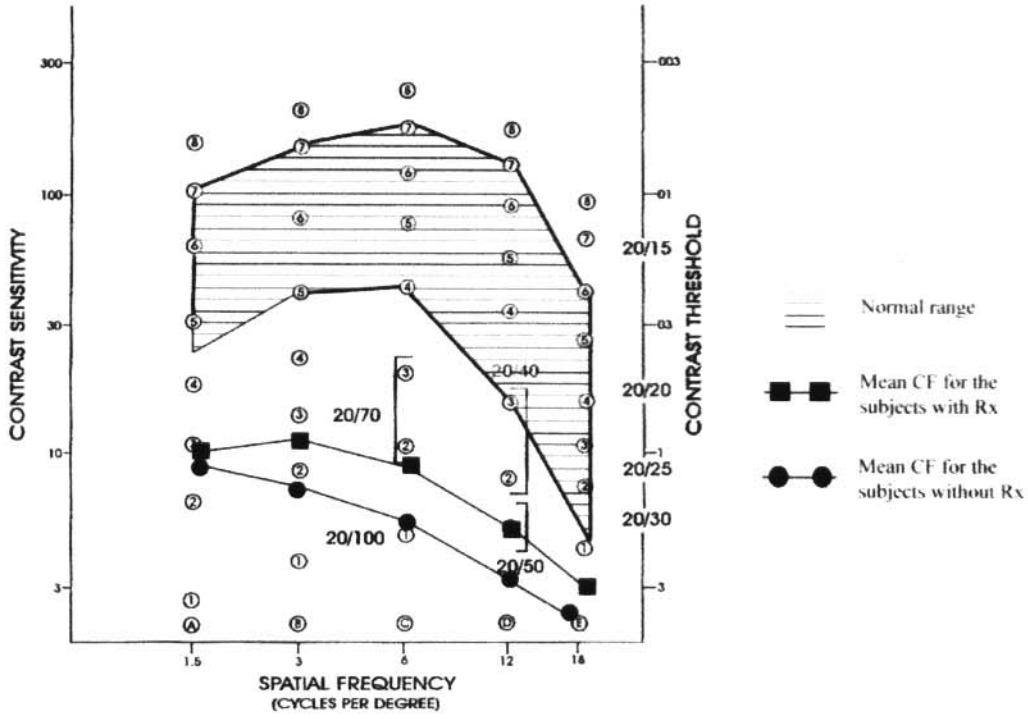


Table 16: Showing the last target seen without the prescription in the Vistech distance test in different spatial frequencies 1.5 to 18 (cpd) among all the age groups. The mean values of the last target correctly seen (CS) decreased with increase in spatial frequencies in all the age groups. The mean contrast sensitivity, however, increased with increase age in each spatial frequencies. All the test range were lower than the expected normal range for each age group.

The range, mean and SD of contrast sensitivity (without Rx) in the VCTS 6500 (Vistech Distance test) in different spatial frequencies A to E (cpd) in different age groups			
Cpd	AGE GROUPS (Years)		
	7-10 years	11-14 years	15- 17 years
1.5 (A) (Expected normal range 4.5-7) Test range mean +/- SD	 0-5 2.6 +/- 1.5	 0-5 2.4 +/- 1.5	 0-5 3.0 +/- 1.5
3 (B) (Expected normal range 5-7) Test range mean +/- SD	 0-5 2.0 +/- 1.6	 0-5 2.0 +/- 1.44	 0-5 2.3 +/- 1.5
6 (C) (Expected normal range 4.3-7) Test range mean +/- SD	 0-5 1.0 +/- 1.0	 0-4 1.2 +/- 1.1	 0-3 1.1 +/- 1.1
12 (D) (Expected normal range 3-7) Test range mean +/- SD	 0-3 0.1 +/- 0.1	 0-3 0.1 +/- 0.7	 0-2 1.1 +/- 0.8
18 (E) (Expected normal range 1-6.5) Test range mean +/- SD	 0-2 0.4 +/- 0.5	 0-2 0.5 +/- 0.7	 0-1 0.3 +/- 0.45

Table 17: Showing the expected normal range of contrast sensitivity (CS) in each spatial frequencies, the range of CS with the prescription for each age group and the mean values of the CS in each age group in all spatial frequencies 1.5 to 18 (cpd). The ranges of the CS do not fall within the normal value for all the spatial frequencies. The mean of the last target correctly seen (CS) decreased with increase in spatial frequencies and increased with increase in age in all the spatial frequencies (1.5 - 18 cpd).

The range, mean and SD of contrast sensitivity (with Rx) in the VCTS 6500 (Vistech Distance test) in different spatial frequencies A to E (cpd) in different age groups			
Cpd	AGE (Years)		
	7-10 years	11-14 years	15- 17 years
1.5 (A) (Expected normal range 4.5-7) Test range mean +/- SD	 0-5 2.9 +/- 1.3	 0-5 2.8 +/- 1.4	 1-5 3.3 +/- 1.3
3 (B) (Expected normal range 5-7) Test range mean +/- SD	 0-6 2.2 +/- 1.48	 0-5 2.7 +/- 1.6	 1-5 3.0 +/- 1.4
6 (C) (Expected normal range 4.3-7) Test range mean +/- SD	 0-5 1.6 +/- 1.1	 0-5 1.7 +/- 1.2	 0-3 1.3 +/- 1.3
12 (D) (Expected normal range 3-7) Test range mean +/- SD	 0-4 1.2 +/- 0.95	 0-4 1.0 +/- 0.92	 0-3 1.3 +/- 1.3
18 (E) (Expected normal range 1-6.5) Test range mean +/- SD	 0-2 1.0 +/- 0.6	 0-2 0.6 +/- 0.6	 0-2 0.7 +/- 0.1

3.5. STEREOACUITY

A large proportion 137 (89.5%) of the subjects demonstrated certain level of stereopsis, while sixteen (10.5%) subjects failed to demonstrate stereopsis on the Randot test. For all the children who demonstrated stereoacuity, the values were very poor compared with expected values for their ages. The maximum stereoacuity demonstrated was 70 seconds of arc and the minimum was 500 seconds of arc with a mean of 294.0 +/- 112.6 seconds of arc (500 seconds of arc is the minimum measurable stereoacuity with this test). A large proportion of the subjects (69) (45.1%) achieved stereoacuity of 200 seconds of arc. Three subjects perceived stereoacuity of 140 seconds of arc and one subject achieved stereoacuity of 70 seconds of arc (see Table 18).

The total number of subjects who achieved each level of stereoacuity is illustrated in Figure 16 and the number of subjects in each age group who achieved each level of stereoacuity is illustrated in Figure 17. A scatter plot of the individual subject threshold points plotted against age is shown in Figure 18 (0 seconds of arc indicates that the subject was not able to perceive the minimum (500 seconds of arc) measurable stereoacuity with Randot test. The mean stereoacuity values for each age group in relation to sex is presented in Table 19. The mean stereoacuity does not appear to show any trend with sex (see Table 19). The mean stereoacuity among each age group is illustrated in Figure 19. The mean stereoacuity appeared to increase slightly with age (see Figures 18 and 19).

Table 18: Showing the number of subjects who achieved each level of stereoacuity at the distance of 40 cm and the percentage. A large proportion of subjects achieved stereoacuity of 200 seconds of arc.

STEREOACUITY (seconds of arc)	NUMBER OF SUBJECTS	PERCENTAGE (%)
0	16	10.5
70	1	0.7
140	3	2.0
200	69	45.1
250	4	2.6
400	50	32.7
500	10	6.5

Figure 16: The total number of subjects who achieved each level of stereoacuity in seconds of arc at 40 cm. A large proportion of the subjects achieved 200 seconds of arc. Only one subject achieved 70 seconds of arc.

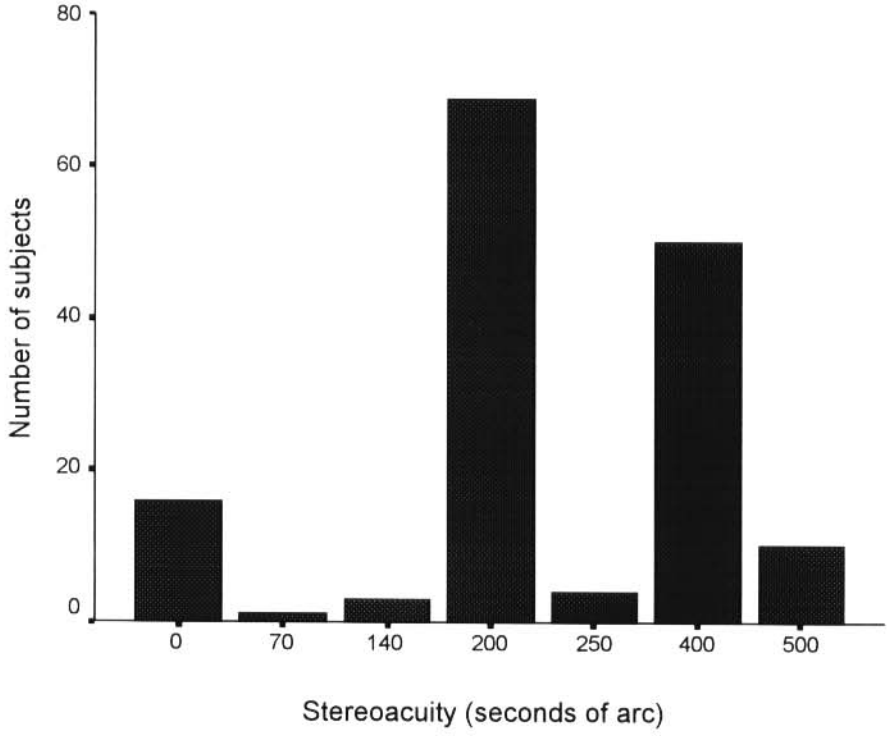


Figure 17: Showing the number of subjects in each age group who achieved each level of stereoacuity. The graph shows that more children achieved 200 seconds of arc between the ages of 7 and 13 years. For most of the children 200 seconds of arc was the maximum level of stereoacuity that could be achieved .

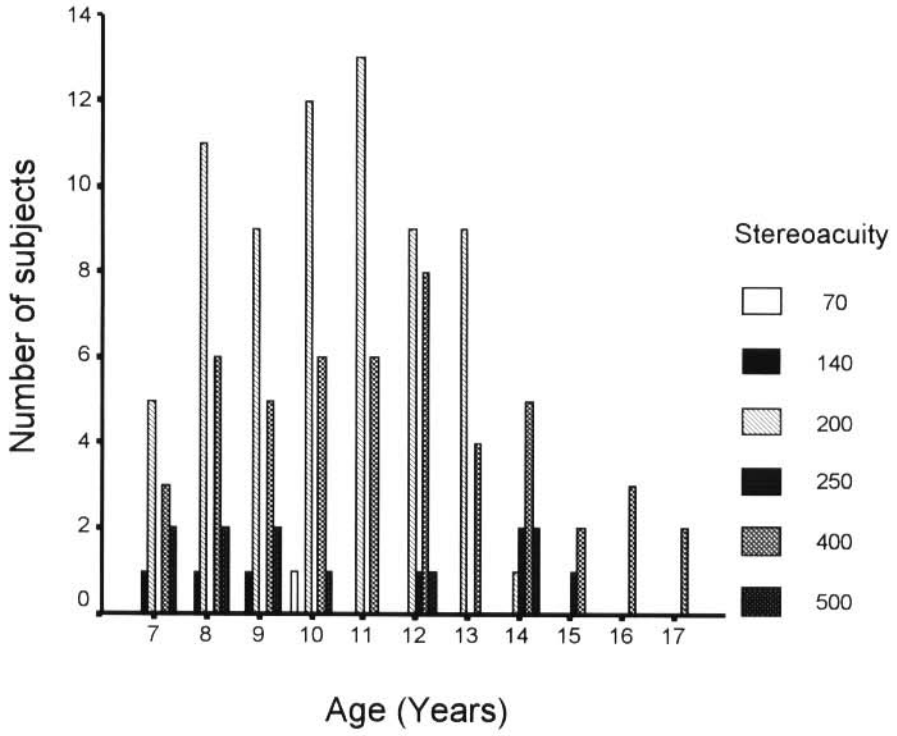


Figure 18: The scatter plot showing the individual subject data points. Each square represents one subject and each petal represents an additional point. The regression equation is y = 2.698 x + 234.3 where y represents the stereoacuity and x is the age of the subject in years. There was a slight increase in the amount of stereoacuity value with age. Zero value shows that the patient could not achieve any stereoacuity in the Randot test.

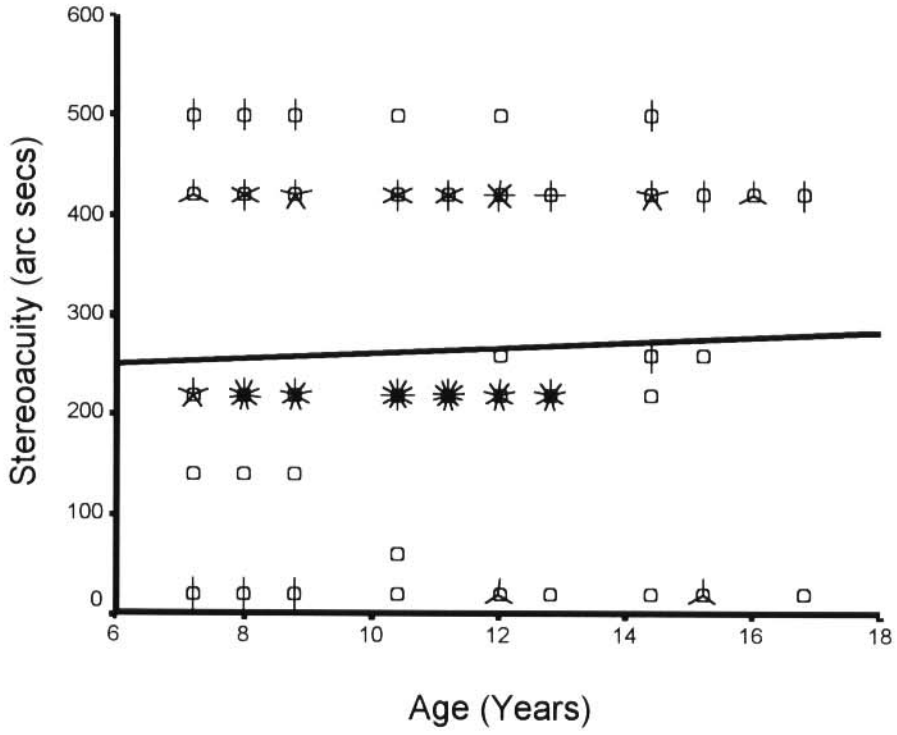
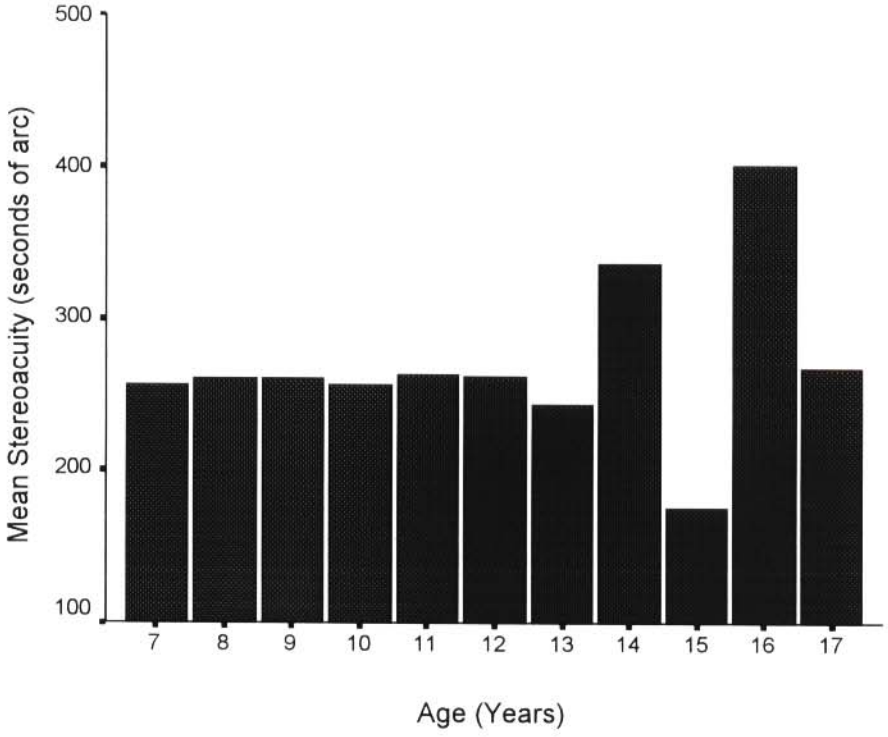


Table 19: Showing the range, means and standard deviations of stereoacuity among male and female subjects of different age groups. Females appeared to have slightly higher mean stereoacuity values between the ages of 7 and 9 and between 13 and 17 years. Males, however, appeared to have higher mean stereoacuity values among the 10 to 12 years old age groups. Therefore, no sexual trend was established.

Range, mean and standard deviation values of stereoacuity						
	Male			Female		
Age	Range	Mean	Std. Deviation	Range	Mean	Std. Deviation
7	400	217.50	169.52	300	320.0	164.32
8	500	233.85	128.68	500	300.00	158.11
9	300	310.00	119.72	500	204.44	163.33
10	430	263.33	137.30	400	250.00	124.32
11	200	300.00	1.09.54	200	246.15	87.71
12	300	325.00	113.82	400	185.00	149.16
13	400	200.00	141.42	200	266.67	100.0
14	500	335.75	165.11	300	337.50	137.69
15	0	0.00	0.00	150	350.00	86.60
16	0	400	0.00	0	400.00	0.00
17	400	200	282.84	0	400.00	0.00

Figure 19: The mean stereoacuity (seconds of arc) at 40 cm for each age group of subjects. The mean stereoacuity values appeared to increase slightly but irregularly with age. There were few (12) subjects between the ages of 15 and 17 years, which may account for the inconsistent shape of the graph at that area.



3.6 COLOUR VISION TESTS

One hundred and twenty seven (83.0%) children passed all the three colour vision tests, hence were considered to have normal colour vision. Others (7) (4.6%) failed all the three tests or failed one or two tests (19) (12.4%). Among those who passed, 137 passed the Ishihara, 141 passed the saturated D-15 and 134 subjects passed the desaturated panel D-15 test. Sixteen subjects failed the Ishihara test. Among those who failed, 9 were deutanopes (6 mild and 3 strong deutanopes) and 6 were protanopes (4 mild and 2 strong protanope). One of the children had total colour blindness. Twelve subjects failed the saturated D-15 test (3 mild protanopes, 1 mild deutanopes, 3 mild tritanope, 2 achromats and 3 with mixed variables), whereas, 19 failed the desaturated (2 mild deutanopes, 4 mild protanopes, 6 mild tritanopes, 2 achromats and 5 subjects with variable results). Among those with colour vision deficiency (those who failed all tests or failed one or two tests), 15 (58.0%) were males and 11 (42.0%) were females. Seven (27%) were classified as deutanopes , 8 (30.8%) protanopes , 6 (23.0%) tritanopes, 2 (7.7%) achromats and 3 (11.5%) subjects had mixed variables. Table 20 shows the number of subjects who failed one or more of the colour vision tests and the types of colour vision deficiency.

Among those who passed Ishihara, five failed both the unsaturated and desaturated D-15. Out of the 12 who passed the saturated panel D-15 test, 9 failed the Ishihara test. Among the 134 subjects who passed the desaturated, 7 failed the Ishihara test. None of those who failed the saturated passed the desaturated D- 15 test (Table 20). Among the 12 subjects who failed the Farnsworth panel D-15, seven subjects failed the desaturated D-15 test while all of them failed the desaturated D-15 test. The colour vision data is presented in Appendix D 1, and the summary of the findings is presented as appendix D 2.

Table 20: Showing the number of subjects who failed one or more of the colour vision tests. D= Deutan, P= Protan, A= Achromat, T= Tritan and MV= Mixed variables

Sex	Ishihara			Saturated panel D-15					Desaturated panel D-15				
	D	P	A	D	P	T	A	MV	D	P	T	A	MV
Male	7	3	1	1	1	1	1	1	2	1	3	1	2
Female	2	3	0	0	2	2	1	2	0	3	3	1	3
Total	9	6	1	1	3	3	2	3	2	4	6	2	5

Figure 20: A chart Showing the percentage of subjects with colour vision defects and the percentage of those without colour vision defects. This graph shows that a large proportion of children have normal colour vision.

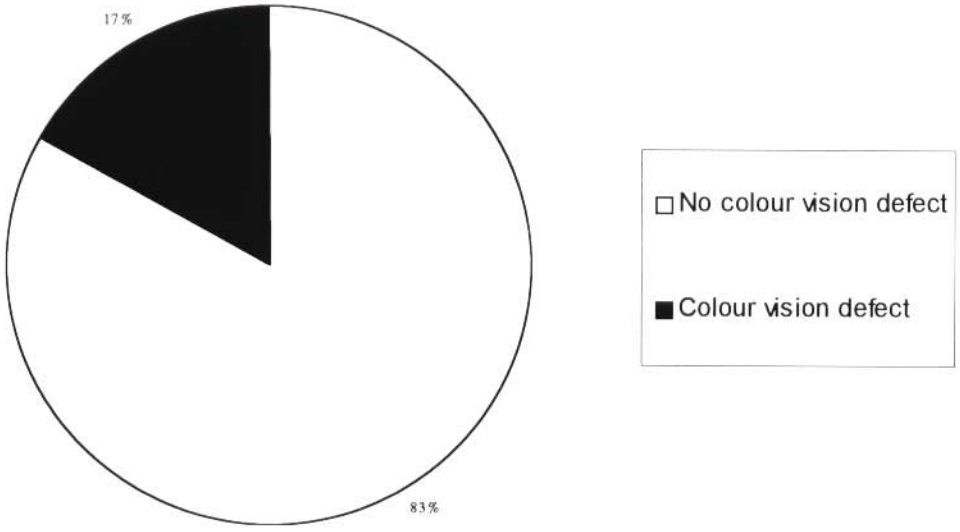


Figure 21: Showing the percentages of different colour vision defects found using the Ishihara test. A large proportion of subjects were Deutanopes. Only one subject had total colour blindness.

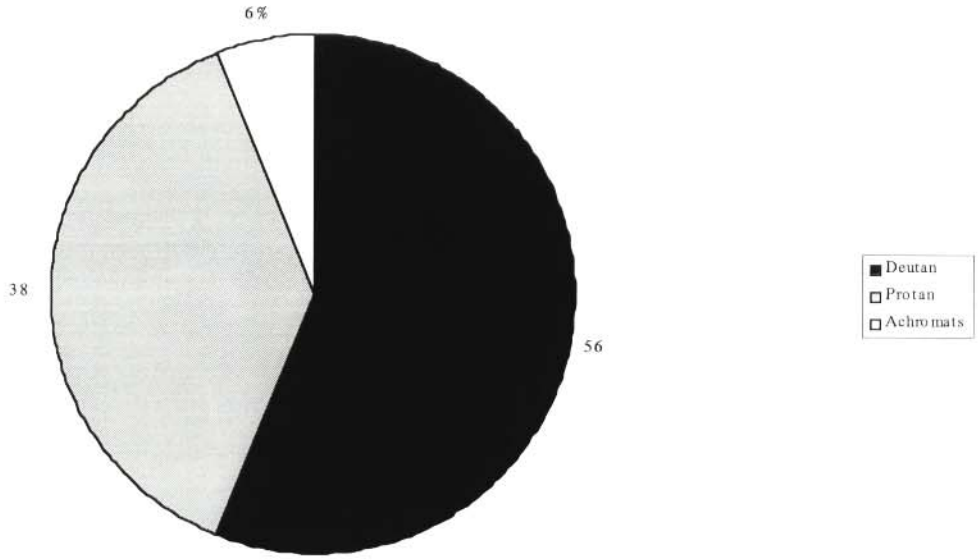
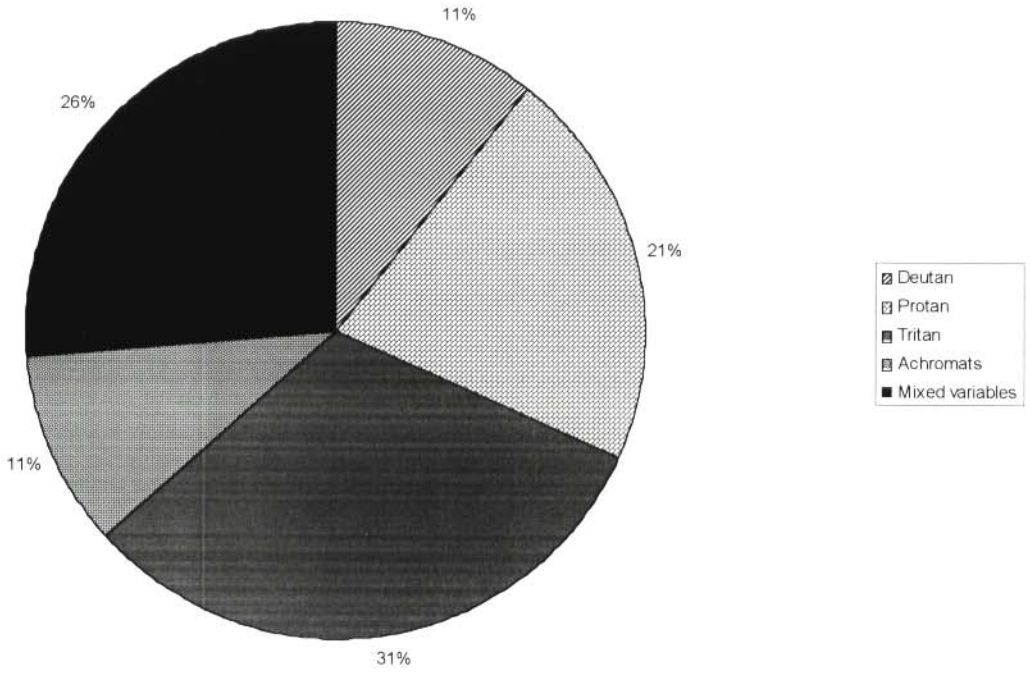


Figure 22: showing the percentages of the different colour defects found with the Farnsworth panel D-15 tests. A large proportion (31%) of subjects were tritanopes. Twenty six percent of the subjects showed mixed variables.



CHAPTER 4

4. DISCUSSION

4.1 STUDY SITES AND POPULATION

The special education schools in the Northern Province of South Africa accommodate visually and physically disabled children. One of these (Siloe school), however, admits only visually disabled children. Children with albinism are often sent to these special schools due to their poor vision and because their parents feel that they will be happy there with their peers. The schools, therefore, have many children with albinism mainly OCA (OCA2). A large percentage (72%) of children included in this study were from the Siloe school for the blind as this school has more children with albinism than the others. Most children in these schools were from the Northern province, with a few from Gauteng and Mpumalanga provinces. Only children aged 7 to 17 years were included in this study. Those below 7 years were not included because the pilot study showed that they could not respond efficiently to subjective eye examination. The 17 years cut off was made because the aim was to study the vision problems of children in South Africa. Individuals who are 18 years old are regarded as adults. The total number of children examined was 153, which is far more than half of the children (200) with albinism in the schools, therefore, considered enough to draw firm conclusion on the vision problems of this group of children.

4.1.1 Classification

All forms of oculocutaneous albinism are inherited as autosomal recessive traits, but can be distinguished from each other on the basis of their clinical, biochemical, ultrastructural and genetic characteristics (Witkop, 1971; Jay et.al, 1976).

As new genes involved in the pigment production are identified and mutational analysis undertaken, the classification of the different types of albinism relies more and more on sophisticated molecular analysis. The clinical method of diagnosis, involves

classification of types of oculocutaneous albinism based on the colour of hair, skin and eyes. Those with oculocutaneous albinism of the tyrosinase negative subgroup have no clinically visible pigment on the skin and have white hair, whereas, the larger tyrosinase positive subgroup produce some pigment, therefore, have pale wheat straw to yellow hair (Lyle et al., 1997). Those with brown albinism have tan skin and brown hair (Kromberg et al., 1990). This clinical method of classification has been used by several authors (Taylor, 1976; Perez-Carpinell et al., 1992; Sacharowitz, 1999). A disadvantage of this method is that, whereas the OCA1 individuals of any race are totally amelanotic, the degree of hypopigmentation seen in OCA2 individuals is modified by the racial and genetic background of the patient and also by the gradual increase of pigmentation with age (Abadi and Dickinson, 1983). Although this method is considered unreliable, it has been used in this study because the methodology involved in the biochemical, ultrastructural and molecular methods of diagnosis is beyond the scope of this study. The 3 types of albinism described in this study, OCA1, OCA2 and brown albinism, are relatively easy to distinguish on the basis of clinical observation. The vast majority of the children had OCA2, which has a classic phenotype among black people. Individuals with white hair and blue eyes were classified as tyrosinase-negative (OCA1) type, those with wheat-straw hair and grey or hazel eyes were classified as tyrosinase-positive (OCA2) type and those with brown hair, skin and eyes were classified as having brown albinism (OCA3). This classification was made to present a general picture of the types of children with albinism included in this study. No conclusion, however, will be drawn in this study on the basis of the types of oculocutaneous albinism because of the unreliability of the clinical method of classification. This study showed that 96% of children show the classic OCA2 phenotype seen in this type of albinism in Africa. A minority (20.3%) of the subjects have pigmented patches indicating that these subjects are able to produce more melanin pigment than the other subjects (79.7%).

4.2 VISION PROBLEMS

As indicated in the introduction, people with albinism have several vision problems which include reduced visual acuity, high refractive errors, high astigmatism (Fonda et al., 1971; Taylor, 1978; O'Donnell and Green, 1981, Kinnear et al., 1985; Abadi and Pascal, 1989; Perez-Carpinell et al., 1992; Sacharowitz, 1999), poor contrast sensitivity (Loshin and Browning ,1983; Abadi and Pascal, 1989; Perez-Carpinell et al., 1992) poor depth perception (Apakarian and Reits, 1989; Guo et al., 1989) strabismus (Guo et al., 1989; Fulcher et al., 1995; Lyle et al., 1997), photophobia and nystagmus (Fonda et al., 1971; Taylor, 1978; O'Donnell and Green, 1981; Abadi and Pascal, 1989; Kinnear et al., 1985; Lyle et al., 1997; Perez-Carpinell et al., 1992; Sacharowitz, 1999). Although there is no cure for albinism, the poor vision problems can be partially corrected by optical means.

Blurry vision is the main visual complaint in individuals with albinism. This has been attributed to the absence or decrease of melanin pigment in the eyes and presence of refractive errors. Absence or lack of melanin in the iris and retinal pigmented epithelium results in light fogging thereby degrading visual acuity. Findings in this study showed that more than half (52.1%) the children reported reduced vision at near and far. Although some of the children (7.1%) reported that they can see well at near and far without correction, optometric findings revealed that they had poor VA at both near and far.

4.2.1 Strabismus (Tropia)

All the subjects in this study had nystagmus (mostly horizontal) which made it very difficult to obtain an accurate magnitude of tropia and to determine the direction of deviations. Some of the subjects could have had a small angle of tropia which was not diagnosed due to the presence of nystagmus. Similar difficulty was reported by Guo et al., (1989) who reported that an accurate measurement of ocular alignment was

difficult in the presence of nystagmus.

Strabismus (Tropia) is a common occurrence in individuals with albinism. No structural feature has, however, been implicated as causative factor of strabismus in these individuals. It is possible that the decreased visual acuity monocularly (disuse resulting in exotropia) and high refractive errors (hyperopia resulting in esotropia) are responsible for the manifest ocular deviation.

The incidence (35%) of strabismus found in this study is lower than 50% reported by Fulcher et al.(1995) and 80% presented by Lyle et al., (1997). It, is, however, higher than (26.7%) presented by Guo et al., (1989). The differences in these reports may be due to difference in the age of subjects, types of albinism studied and the methods used in measurement of tropia (Fulcher used Bruckner method, prism and alternate cover test and Guo et al. (1989) did not mention the method used in his study). It may also be due to difference in sample size.

The slightly higher occurrence of esotropia (18.3%) than exotropia (16.3%) in the present study is in agreement with 16% of esotropia reported by Guo et al. (1989). It, also agreed with the study by Guo et al., 1989 and the review by Lyle et al., (1997) that esotropia is more prevalent than exotropia in individuals with albinism. This difference is not considered significant and no reason is given for the slight preponderance of esotropia.

High refractive error such as hyperopia in children can result in strabismus. The latter in turn can cause amblyopia in the deviating eye as a results of long-continued suppression (Grosvenor, 1989). Therefore, there is a need for the refractive errors and strabismus in the children with albinism to be corrected at an early stage before amblyopia sets in.

4.2.2 Visual acuity

Visual acuity is reduced in albinism as a result of several factors such as foveal hypoplasia, misrouting of optic fibres at chiasma, presence of deprivational amblyopia, astigmatic refractive errors and possibly high level of retinal irradiances since birth (Davis et al., 1997). While certain aspects of causative factors such as macular hypoplasia cannot be alleviated, others, such as refractive status and excessive transillumination can be addressed, therefore, resulting in improvement in vision.

The logMAR charts (Figures 1 and 2) were used to measure VA in the present study because they are devoid of the several deficiencies which have been identified in the Snellen charts and their variations (Bailey and Lovie, 1976; Westheimer, 1979; Oduntan and Briggs, 1999). Also, they can be used to measure and score low acuities accurately, which can then be included in statistical analysis (Bailey and Lovie, 1976). The scoring method used with logMAR, also allows arithmetic procedures, including regression analysis and parametric statistics to be applied legitimately to visual acuity scores (Wild and Hussey, 1985; Lovie and Kitchin, 1988). Another advantage of the letter by letter scoring technique associated with the logMAR principle is that by scoring each letter, the clinical grading scale is effectively made five times finer (for a chart with five letters per line) than scoring by line, which consequently improves the clinician's ability to detect damage (Lovie and Kitchin, 1988). In addition, the logMAR charts make it ease to convert non-standard test distance values to standard test distance values. The VA of certain subjects in this study were poorer than can be measured at 4 metres. It, therefore, became necessary to reduce the testing distance. The logMAR facilitated this process because, for each 0.1 log unit reduction in viewing distance, there is 0.1 log unit increase in the angular subtense of all the letters in every row of the chart. In order to facilitate the conversion to the standard testing distance of 4 metres, the testing distance was reduced by dividing by a ratio of 0.1 log unit or 0.12589. For the purpose of analysis, each letter read by the subject was scored as 0.02 logMAR.

Findings in this study show that there were improvements in both monocular and binocular visual acuities with optical correction. The corrected binocular distance visual acuity of children with albinism ranged from 6/60 (20/200) (1.0 logMAR) to 6/6 (20/20) (0.0 logMAR). These values are better than corrected binocular visual acuity (6/120 to 6/18) presented by Kinnear et al. (1995). Also, it was observed that distance VA can be improved by up to 3 (15 letters) acuity lines in most (71.2%) children with best optical correction. This improvement is considered significant. This is consistent with the reports presented by Perez-Carpinell et al. (1992) that a significant improvement in visual acuity was observed in many of the subjects studied. It is also in agreement with the conclusion drawn by Sacharowitz (1999) who reported significant improvement in distance VA in 60% of the subjects following a review of refractive data of South African individuals with albinism. It is also noteworthy, however, that insignificant improvement (less than one acuity line) was achieved by a small percentage (28.8%) of the children. This is also in agreement with existing reports (Perez-Carpinell et al., 1992; Sacharowitz, 1999) that improvement in VA was very limited in certain individuals. Although, no significant improvement in VA was measured in these children, an improvement in the quality of their vision was reported by many of them with best optical correction. The lack of improvement in VA following refraction and correction with best optical correction may be related to the severity of one or more factors causing poor vision in albinism.

The significant improvement in near visual acuity (monocular and binocular) following optical correction shows that children with albinism can benefit from refractive correction not only at far but also at near. The slightly better VA at near compared with the far (corrected and uncorrected) is believed to be due to the dampening of nystagmus on convergence as previously suggested by Biswas, 1994. The near visual acuities could not be compared with other data because previous authors did not present near VA data in individuals with albinism, although there are general comments that near VA is generally better than VA at far. All previous reports that we have examined reported and discussed only the distance visual acuities.

Even with the best optical correction, a large percentage (84.3%) of the children in this study would still be classified as partially sighted (distance VA of less than 6/18) (WHO, 1980). As vision of a large proportion of the subjects can be improved significantly with best optical correction; if children with albinism can be provided with optical corrections such as tinted spectacles or contact lenses early in life, before amblyopia sets in, the chances are that they will retain this good level or even further improve their level of visual acuity. It is, therefore, recommended that the children with albinism should be examined optometrically within the first or second year of life and provided with optical correction.

4.2.3. Refractive errors

The correct way of quantifying and analysing refractive powers in research has been controversial (Harris, 2000). It is, however, generally agreed that meaningful statistical analysis cannot be performed with sphere, cylinder and axis considered separately (Saunders, 1980; Harris, 1988a; Harris 1988b; Harris 1989; Harris, 1997; Thibos, 1992; Deal 1993; Harvey, 1995; Harris, 2000). Saunders (1980) suggested that a necessary and sufficient condition for obtaining a mean value for the various components of the power requires the summation of all parameters of the variates while maintaining the integrity of their physical values. According to Harris (2000), although, sphere, cylinder and axis are unambiguous and meaningful together as a representation of power when separated, they are not invariant under spherocylindrical transposition. An appropriate method, therefore, is that which represents power as a single entity rather than separate components (sphere, cylinder and axis). Nevertheless, certain authors (Karabatsas et al., 1998; Goggin and Pesudovs, 1998a, Goggin and Pesudovs, 1998b) still attempt the analysis of separated refractive components.

The multivariate statistical methods developed by Harris (Harris, 1988; Harris, 1989; Harris, 1991) and Malan (1990) have the advantages of representing dioptric power as a single entity, and therefore, are appropriate for analysis of refractive data. A current

problem with that approach, however, is that most practitioners and researchers are not conversant with the mathematical statistical methodology involved in this approach. In view of the above, the method of using the nearest equivalent spherical power (F_{NE} = sphere +1/2 cylinder) which has been previously referred to as spherical equivalent and employed by several authors (Kragha, 1987; Lam et al., 1994; Lavery et al., 1988; Parssinen, 1987; Fledelius, 1984) is employed in this study. This concept has, however, been criticized by several authors. According to Lavery et al. (1988), when this procedure is used, the detail of the cylinder power and axis is lost, and if the means of the individual components are found, the results do not give an accurate picture of the most representative prescription. The method also completely ignores the direction of the cylindrical axis, it, therefore, has neither real physical nor physiological meaning (Saunders, 1980). Also, because both the hyperopia and astigmatism (in negative cylinder form) diminish with increasing age in certain populations, there is a tendency for the changes to cancel each other out in the calculation of spherical equivalent power. The changes in the refractive errors, therefore, are not well represented by the use of spherical equivalent values (Chung et al., 1996). In spite of these various criticisms, the nearest equivalent spherical power methods is employed in the present presentation because, currently, many practitioners and ophthalmic researchers are more conversant with it compared to the multivariate method of analysis. It also satisfies the invariance concept under spherocylindrical transposition (Harris, 2000). According to magnitude, refractive errors have been classified as low (+0.25 to +2.00 D), moderate (+2.25 to +5.00 D) or high (>5.00 D) for hyperopia; low (-0.25 to -3.00 D); moderate -3.25 to -6.00 D, or high (> 6.00 D myopia) and for astigmatism, low (-0.25 to -0.75 D), moderate(-1.00 to -3.00 D) or high (> -3.00 D) (Borish, 1970). These classifications were used in this study. In the present study 83.3% of the subjects had low refractive errors (myopia and hyperopia), 9.8 % had moderate and only 5.6% of the subjects had high refractive errors (see Table 10). This finding disagrees with several reports (Fonda et al., 1971; Perez-Carpinell et al., 1992; Lyle et al., 1997; Sacharowitz, 1999) that individuals with albinism have high refractive errors. The differences may be due to the ages of the subjects studied. Others have studied mixed ages or adults whereas only children were included in this study.

That the majority of children were myopic (67.6%) and a smaller proportion were hyperopic (30.8%) in the present study is in agreement with previous studies (Fonda et al., 1971; Sacharowitz, 1999). The maximum positive and negative spherical powers observed in this study (+5.75 D and -8.25 D respectively) were, however, lower than +/-10.0 D reported by Fonda et al., (1971) and -20.0 D (for myopia) reported by Sacharowitz (1999). The maximum positive spherical powers in this study was however higher than +5.50 D (hyperopia) reported by Sacharowitz (1999).

The differences in these reports may be attributed to differences in the age distribution of the subjects studied and differences in criteria used in determining and tabulating results. The high occurrence of myopia in females being higher than in males observed in this study is in agreement with the report of Richler and Bear (1980) and Johnson (1979) that myopia was more prevalent in females than in males in normally pigmented subjects. It is, however, contrary with the report by Hyams et al., (1977) and Raliavhegwa and Oduntan (2000) that myopia was more common in males than in females. It is possible that there are no real sex differences in spherical refractive errors. More data are needed to establish any sex-related trends in both the normally pigmented and those with albinism.

The high occurrence of astigmatism (92.2%) (cylindrical powers $\geq$ -0.25 D) agrees with previous reports (Perez-Carpinell et al., 1992; Fulcher et al., 1995; Sachrowitz, 1999). The prevalence in the present study is, however, higher than 76% reported by Sacharowitz (1999) and 50% reported by Perez-Carpinell (1992). It is possible that there is a higher prevalence of astigmatism in the children with albinism.

The occurrence of myopic astigmatism (62.1%) in this population agrees with the findings of Sacharowitz (1999) who reported that this type of astigmatism was more common (46.3%) among the individuals with albinism studied. The findings, however, disagree with those of Fulcher et al.,(1995) (studied 18 subjects) who reported that hyperopic astigmatism was more common than other types of astigmatism in a study of 18 subjects with albinism. Perez-Carpinell et al., (1992) (9 subjects), however, did

not observe any dominant type in his study involving 9 subjects. These differences may be due to differences in the number and ages of subjects studied. Further studies, using large population is needed to establish which type of astigmatism is more common in individuals with albinism.

In the literature, astigmatism are often grouped together as WTR, ATR, and OBL (Loshin and Browning, 1983; Dickinson and Abadi, 1984; Lavery et al., 1988; Chan et al., 1993; Lam et al., 1994; Davis et al., 1997; Raliavhegwa and Oduntan, 2000). This mode of grouping has some shortcomings, for instance the cut off values are arbitrary and inconsistent. While some authors used 30 degrees cut off (Chan et al., 1993; Lam et al., 1994; Raliavhegwa and Oduntan, 2000), others used 15 degrees (Lavery et al.,1988; Fledelius, 1984). The fifteen degrees difference can be significant if the cylindrical power is moderate or high, thereby making comparison of data difficult. Also, that method does not take account of the cylindrical powers, therefore, lacks meaning. In view of these and other shortcomings of this type of grouping, this mode of grouping has been considered inappropriate (Thibos, 1992; Holladay et al., 1998; Harris, 2000). According to Harris (2000), grouping by axes introduces discontinuity into what in reality is continuous; the net effect is distortion and loss of information and results in diminished value. This approach is now giving way to other methods such as those based on vectors in what are sometimes called double angle plots (Thibos, 1992; Holladay et al., 1998). The current thinking in optometry, however, remains in terms of WTRA, ATRA and OBLA, also previous reports on astigmatism in individuals with albinism used this type of classification, therefore, this mode of grouping has been employed in presenting that aspect of the data in this study in spite of the various criticisms. This classification allows us to compare our findings with the existing data. In the present study WTRA astigmatism was more prevalent (47.0%) than the oblique (28.0%) and ATR (25.0%) astigmatism. This agreed with the reports of various authors: Loshin and Browning, 1983; Dickinson and Abadi, 1984; Davis et al., 1997; that WTRA is more common in individuals with albinism. The WTRA type was more prevalent (55.7%) than other types in females than in males in the present study. The relation of axis of astigmatism to sex of subjects has not previously been examined in

individuals with albinism. Further studies on a large number of individuals with albinism are necessary to establish the trend of axis of astigmatism with sex.

4.2.4 Contrast sensitivity

The contrast sensitivity function (CSF) has proven clinically to be a useful tool for detection of neurological or visual pathway problems (Virsus, 1981; Kupersmith, 1984). Further, the Vistech Contrast Sensitivity System has been reported to be useful and understandable by children as young as 4 years of age and enables the clinician to perform an accurate, reliable and quick examination of the contrast sensitivity function (Rogers et al., 1987; Scharre et al., 1990). In view of these advantages, the Vistech Contrast Sensitivity system was used to measure the CSF of the children in this study.

Reduced contrast sensitivity in normally pigmented children has been associated with visual disorder such as amblyopia, strabismus, cataract and glaucoma (Scharre et al., 1990). As indicated in the introduction, individuals with albinism are known to perform poorly on the contrast sensitivity tests. The poor performance has been attributed to increased spacing of the central cones, high astigmatism errors and nystagmus (Abadi and Pascal, 1989; Loshin and Browning, 1983; Kelly and Burbeck, 1984). The poor contrast sensitivity values observed in this study is therefore, consistent with existing reports. The maximum frequency of 18 cycles per degree (cpd) perceived by the subjects in the present study, however, was higher than 16 cpd reported by Perez-Carpinell et al. (1992). The variance may be due to differences in the numbers and ages of subjects in the studies. The average CS curve obtained in this study was considered to be significantly low; with a maximum sensitivity of 3 cpd (see Figure 15). This low value is consistent with the reports of Perez-Carpinell et al. (1992) and Loshin and Browning (1983) that individuals with albinism have lower contrast sensitivity threshold compared to the normally pigmented population. The low improvement in contrast sensitivity observed in the subjects following optical correction is attributable

to such factors as nystagmus, macula hypoplasia and excessive trans-illumination (resulting in light fogging), and possibly the misrouting of the optic nerve fibres.

The reduced contrast sensitivity in children with albinism indicates that these children will have difficulties in detecting border contrast. The slight increase in the mean CSF resolution acuity and peak sensitivity with the optical corrections, may, however, be of advantage to the subjects. If, children with albinism can be provided with appropriate optical correction at an early age, it is possible that the correction can make a difference in detecting border contrasts in their everyday life. This further dictates the need for early vision correction for children with albinism.

4.2.5 Depth perception

Stereoacuity can be measured using apparent depth in two-dimensional tests or real depth in three-dimensional situations. Tests based on two-dimensional objects are used clinically and include the Titmus, Random dot E, Frisby, Lang and Randot stereo tests. The Titmus stereotest is very popular among clinicians world wide due to its simplicity in administration with very young children. However, a major disadvantage of the test is that patients may respond positively to it using lateral displacement cues (Cooper and Warshowsky, 1997). The Randot stereotest is regarded as superior to the Titmus test because it has a wide range of disparity presentation and contains no monocular cues (Cooper et al., 1979). Because of these advantages, the test was used in this study. Individuals with albinism are generally known to exhibit poor depth perception (Apakarian and Reits, 1989; Guo et.al, 1989; Lyle et al., 1997). Findings in this study agree with this notion. A large proportion (89.5%) of children in this study demonstrated stereopsis ranging between 500 and 70 seconds of arc, which is considered to be very poor for their age group. Others (10.5%) could not achieve any level of stereoacuity in the Randot stereotest. It is possible that the latter group of children had stereopsis which was less than 500 seconds of arc, therefore, stereopsis could not be measured with the Randot stereo test at 40 cm. The present findings, however, disagree with the report by Creel et al.(1981) that individuals with albinism

appear to have no stereopsis.

Although many (45.1%) of the children in this study achieved 200 secs arc, this value is significantly lower than 20 sec arc that had been reported for children of the same age group without albinism (Oduntan et al. 1998). Only one subject who achieved 70 sec arc can be considered to have fairly normal depth perception. The poor depth perception found here may be due to misrouting of the optic nerve fibres. This opinion has previously been expressed by certain authors (Apakarian and Reits, 1989; Guo et al, 1989 and Lyle et al., 1997) that misrouting of the optic nerve fibres in individuals with albinism adversely affects binocular vision, hence stereopsis. Also, It may be a consequence of the strabismus observed in the subjects, as these conditions have been reported by several authors to result in degraded depth perception (Donzis et al., 1983; Larson, 1983). Poor stereopsis will present difficulty to these children in their everyday vocational and avocational activities where a need to judge depth is involved. The poor depth perception means that the children will have difficulties in activities such as sports; for instance, in following a ball in soccer. Also, they will have problems in other activities such as seeing the edge of steps. It is possible that, if the strabismus and refractive status are corrected early in life, the depth perception may be enhanced.

4.2.6 Colour Vision

Congenital colour vision deficiencies are a relatively common occurrence and many ocular diseases are associated with changes in colour vision (Pokorny et al., 1979; Adams, 1982; Kalloniatis et al., 1989). Both congenital and acquired dyscromatopsias would affect partially sighted children in everyday classroom situations. Also, colour vision defect could provide a barrier to the child in acquiring visual information, both with respect to colour confusion created by the colour vision defect and the reduced ability to see colours by the subjects (Porkony et al, 1979). Therefore, early colour vision examination could be of benefit to many of the young children with colour vision

defects as they will became aware of the problem earlier and get advise on how to adapt to the defect.

There are many tests used for colour vision evaluation. These include the anomaloscope, Ishihara, and saturated and de-saturated Farnsworth panel D-15. Anomaloscopes provides a definitive diagnosis of type and severity of congenital colour vision defects but are expensive instruments and few practitioners have access to them (Atchison et al., 1991) . We had no access to an anomaloscope, hence, the Ishihara and Farnsworth panel D-15 (Standard D-15 and Desaturated D-15) were used in this study.

The Ishihara pseudoisochromatic test is the most widely used screening test for red-green colour deficiency and clinical trials have for a long time showed that it is the most effective for detecting colour vision defect (Sloan and Hebel, 1956; Belcher et al., 1958; Pokorny et al., 1979). The disadvantage being that it only detects red-green defect (Fletcher and Voke, 1985). It, has, therefore been suggested that it should be used with the other tests such as the Farnsworth panel D15 (Porkony et al., 1979). In agreement with this suggestion, the Ishihara test was used with the Farnsworth panel D-15 tests in this study.

The Farnsworth panel D-15 has been recommended for studying colour vision in the partially sighted subjects (Fletcher and Voke, 1985, Kalloniatis and Johnston, 1990). Farnsworth's Dichotomous panel D-15 (standard test) is designed to identify those observers with moderate to severe losses of colour discrimination (Farnsworth, 1947). The disadvantage being that its sensitivity is limited until a significant congenital colour defect is present (Helve, 1972) or until a definite visual acuity loss is present in acquired defects (Bowman, 1980). It has been suggested that this limited sensitivity of the D-15 can be compensated for by supplementing its use with the desaturated D-15 (Dubois-Poulsen and Lanthony, 1973) for those subjects manifesting only slight loss of colour discrimination. The desaturated D-15 panel is a more demanding test

because the colour samples are less saturated (by 2 units of Munssel Chroma) and lighter (by 3 units of Munssel value) than the standard D-15 test caps (Atchison et al., 1991). Some people with colour defects who pass the standard D-15 test will therefore fail the desaturated test because it needs better colour discrimination to pass (Atchison et al., 1991). Because of the positive features of these tests (Ishihara, Farnsworth panel D-15 tests (saturated and desaturated), they were used for this study.

As indicated in the introduction, earlier reports have associated albinism with colour vision deficiency (Pickford, 1958; Pickford and Taylor, 1968; Taylor, 1976, Perez-Carpinnel et al., 1992). More recent reports, however, suggested that most individuals with albinism have normal colour vision (O'Donnel and Green, 1981; Lyle et al., 1997; Oduntan, 1998). The findings (83% pass rate) of the present study agree with the latter opinion that many children with albinism have normal colour vision.

The 17% of children who had colour vision deficiency in this study was lower than 58.8% presented by Perez-Carpinell et al., (1992) and 95% reported by Taylor (1976) whose findings suggested that individuals with albinism were likely to have colour vision deficiency. The disagreement in findings may be attributed to differences in the sensitivity of the tests used by various authors in detecting the colour vision defects and the types of albinism studied. Perez-Carpinell et al., (1992) for example used the Ishihara, the Roth 28-Hue test and the Davi's anomaloscope, while Taylor used an anomaloscope. Anomaloscopes are quite sensitive, therefore may be able to identify children with slight colour deficiencies, hence the large number of failures in the previous studies than the present one.

Also, the number and type of subjects studied could account for the differences in the percentage of failures. Perez-Carpinell studied only 9 subjects which is considered to be small (6 of whom had OCA2 type of albinism). Taylor (1976) also studied 16 only (with various types of albinism), whereas, the present study had a large number of subjects consisting of mainly of OCA2 individuals.

This study showed that a slightly higher percentage (58.9%) of males than females (42.0%) had colour vision defects. This agrees with previous reports (Thuline, 1972; Pokorny et al., 1979; Swanson and Everett, 1992) and suggests that males are more likely to have colour vision defects than females. This is expected since genetically, both deutan and protan defect are X-linked and therefore, more common in males, while tritan defects are inherited as an autosomal dominant trait, therefore, are of equal prevalence among males and females (Birch, 1999). Also, that 30.8% of the subjects with colour vision deficiency were red deviants was in agreement with the previous reports that most of the subjects with albinism examined were red deviants (Taylor, 1976, Perez-Carpinell et al., (1992).

Some of the subjects (11.5%) showed mixed variables (there was no clear confusion axis to show the type of colour vision defect) with the Farnsworth-panel D-15 tests. Subjects who exhibit mixed variables have been reported to confuse colours such as blue with green , yellow with green but could identify orange (Fletcher and Voke, 1985). This may be the case in the present study.

The three tests employed in the study gave slightly different results (see Table 20). This is related to the varied sensitivities of the tests.

Children with colour defects will experience difficulties when colour coded materials are used for teaching in the classroom. Also, they are sometimes misjudged by teachers or peers as being either unintelligent or uncooperative (Steward and Cole, 1989). Diagnosis of the colour defects can help alleviate these problems, as the child, parents, and teacher can become aware of the colour vision defect and the types of mistakes it produces. In addition, although, there is no cure for colour vision deficiency, early diagnosis can improve the child's adaptation to the dysfunction and help colour defective individuals avoid planning on vocations which require normal colour vision (Swanson and Everett, 1992). There is therefore, a need for the eye care practitioners to detect and correctly diagnose these colour vision defect and convey their findings to the parents and teachers.

4.3 CONCLUSION AND RECOMMENDATIONS

The objectives of this study include: Firstly, provision of statistical data on vision problems of South African children with oculocutaneous albinism, secondly, establishment of whether or not, there were differences in the vision problems among these children compared with those reported in the literature and thirdly, recommendation of standard visual examination procedure necessary for children with albinism.

This study showed that children with albinism have several significant vision problems. A large percentage (84.3%) had low vision (corrected VA less than 6/18), significant refractive errors ($\geq$ 1.00 D) (62.7% myopia and 29.4 % hyperopia) and strabismus (34.6%). Contrast sensitivity and depth perception were poor in all subjects. A large proportion (83.0%), however, had good colour vision whereas, 17.0% had poor colour perception. Findings in this study, which included by far the largest group of children with albinism studied to date, agree with most of the previous reports on vision problems in individuals with albinism. The few differences have been highlighted and discussed.

It was found that best optical corrections significantly improved the visual acuity in many of the children. It follows, therefore, that if the children can be provided with the best optical correction early in life, the chances are they will maintain this or achieve better level of visual acuity later in life. If, however, the refractive errors are not corrected in time, amblyopia may set in and prevent them from achieving their maximum level of vision. Also, best optical correction will slightly improve their contrast sensitivity function and depth perception. It is, therefore, recommended that:

1. Children with albinism should be given a comprehensive eye examination early in life.

2. They should be provided with appropriate optometric or ophthalmological

assistance as early as possible. As photophobia is a common occurrence in individuals with albinism, spectacles and contact lenses should be tinted or provided in photochromic form. The absorptive lenses will also provide protection against ultraviolet and shortwavelength visible light.

3. The parents and the teachers should be given necessary information and advice regarding the child's vision following an eye examination. They should also be given the task of monitoring compliance in the use of whatever optical aid provided, as children are careless regarding compliance in the use of optical devices.

4. The children should be provided with regular annual eye care services which will include an eye examination and the provision of necessary optical devices or other appropriate interventions.

5. As many of the children can be classified as partially sighted, the optical services may include magnifiers for reading at near, as these are cost effective options.

With implementation of these recommendations, improvement in visual functioning will be achieved in South African children with albinism.

CHAPTER 5

5. REFERENCES

Abadi, R.V. and Dickinson, C.M. (1983). Monochromatic fundus photography of human albinos. *Arch. Ophthalmol.* **101,** 1706 - 1711.

Abadi, R.V. and Dickinson, C.M. (1986). Waveform characteristics in congenital nystagmus. *Doc. Ophthalmol.* **64,** 153-167.

Abadi, R.V. and Pascal, E. (1989). The recognition and management of albinism. *Ophthal Physiol. Opt.* **9,** 3 -15.

Abadi , R.V. and Pascal, E. (1994). Periodic alternating nystagmus in humans with albinism. *Invest. Ophthalmol. Vis. Sci.* **35,** 4080-4086.

Abadi, R.V. and King-Smith, P.E. (1979). Congenital nystagmus modifies oriental detection. *Vision Res.* **19,** 1409-1411.

Adams, A.J. (1982). Chromatic and luminosity processing in retinal diseases. Am. J. Optom. *Physiol. Opt.* **59,** 954-960.

Al-Mubrad, T.M. and Oduntan, A.O. (1996). Titmus stereoacuity norms in Saudi Primary School children. *Saudi J. Ophthalmol.* **10,** 27 -30.

Apakarian, P. and Reits, D. (1989). Global stereopsis in human albinos. *Vision Res.* **71,** 355 - 367.

Aquaron, R. (1990). Oculocutaneous albinism in Cameroon, a 15-year follow-up study. *Ophthal. Paediatr. Genet.* **4,** 255 - 263.

Arden, G.B. (1978). The importance of measuring contrast sensitivity in cases of visual disturbance. *Br. J. Ophthalmol.* **62,** 198 - 209.

Atchison, D.A., Bowman, J.K. and Vingrys, A.J. (1991). Quantitative scoring methods for D15 panel tests in the diagnosis of congenital colour vision deficiencies. *Optom. Vis. Sci.* **68,** 41-48.

Bailey, I.L. and Lovie, J.E. (1976). New design principle for visual acuity letter chart. *Am J. Optom. Physiol. Opt.* **53,** 740-745.

Beckham, A.S. (1946). Albinism in Negro children. *J. Genet. psychol.* **69,** 199 - 215.

Belcher, S.J., Greenshields, K.W. and Wright W.D. (1958). A colour vision survey. *Br. J. Ophthalmol.* **42,** 355-359.

Birch, J. (1999). Colour vision deficiency-An introduction. *Optician.* **21,** 22-32.

Biswas, S. and Lloyd, I.C. (1999). Oculocutaneous albinism. *Arch. Dis. child.* **80,** 565 - 569.

Borish, I. (1970). Clinical refraction. 3rd ed. 127 -131. Professional press, Chicago.

Bothwell, J.E. (1997). Pigmented skin lesions in tyrosinase - positive oculocutaneous albinos: a study in black South Africans. *Internat. J. Dermatol.* **36,** 831 - 836.

Bowman, K.J. (1980). The clinical assessment of colour discrimination in senile macular degeneration. *Acta Ophthalmol.* (Openh.) **58,** 337-346.

Buckingham, T. (1993). Visual problems in childhood. 98-100. Butterworth-Heinemann Ltd, Oxford.

Castronuovo, S., Simon, J.W.and Kandel, G.L. (1991). Variable expression of albinism within a single kindred. *Am. J. Ophthalmol.* **111,** 419-426.

Chan, O. Y. and Edwards, M. (1993). Refractive errors in Hong Kong Chinese pre-school children. *Optom. Vis. Sci.* **70,** 501-505.

Charles, S.J., Green, J.S., Grant, J.W., Yates, J.R.N. and Moore, A.T. (1993). Clinical features of affected males with X-linked ocular albinism. *Br. J. Ophthalmol.* **77,** 222-227.

Cheong, P.Y., King, R. A. and Bateman, J. B. (1992). Oculocutaneous albinism: variable expressivity of nystagmus in a sibship. *J. Pediatr. Ophthalmol. Strab.* **29,** 185-188.

Chung, K.M., Mohidin, N., Yeow, P.T. and Tan, L. L. (1996). Prevalence of visual disorder in Chinese School children. *Optom. Vis. Sci.* **73,** 695 - 700.

Collewijn, H., Apakarian, P. and Spekreijse, H. (1985). The ocular motor behavior of human albinos. *Brain* **108,** 1-28.

Comeford, J.P. (1979). Contrast sensitivity functions for clinical optometry. *Am. Optom. Assoc.* **50,** 683 - 686.

Cooksey, C.J., Garratt, P.J., Land, E.J., Pavel, S., Ramsden, C. A., Railey, P. A. and Smit N. P.M. (1997). Evidence of the indirect formation of the catecholic intermediate substrate responsible for the autoactivation kinetics of tyrosinase. *J. Biol. Chem.* **222,** 26226-26235

Cooper, J., Feldman, J. and Meldin, D. (1979). Comparing stereoscopic performance of

children using the Titmus, TNO and Randot stereotests. *J. Am. Optom. Assoc.* **50,** 281-285.

Cooper, J. and Warshowsky, J. (1997). Lateral displacement as a response cue in the Titmus stereotest. *Am. J. Optom. Physiol. Optics* **74,** 537-541.

Creel, D.J., Summers, C.G. and King, R.A. (1974). Asymmetry visually evoked potentials in human albinos: evidence for visual system anomalies. *Invest. Ophthalmol.* **13,** 430-440.

Creel, D.J., O'Donnell, F. E. Jr and Witkop, C.J. Jr (1978). Visual system anomalies in human ocular albinos. *Science* **201,** 931-933.

Creel, D., Spekreijse, H. and Reits, D. (1981). Evoked potentials in albinos: efficacy stimuli in detecting misrouted optic fibers. *Electroencephalogr. Clin. Neurophysiol.* **52,** 595-603.

Cucchiaro, J. (1985). Visual abnormalities in albino mammals. In: cell and developmental biology of the eye, heredity and visual development. Sheffield, J.B. and Hilfer, S.R. (eds). 63-83. Springer Verlag, New York.

Davis, P. L., Baker, R.S. and Piccione, R.J. (1997). Large recessive nystagmus surgery in albinos: effect on acuity. *J. Pediatr. Ophthalmol. Strab.* **34,** 279 - 285.

Deal, F. C. (1993). Magnitude, distance and orthogonality for thin sphero-cylindrical lenses. *Optom. Vis. Sci.* **70,** 454-461.

Dickinson, C.M. and Abadi, R.V. (1984). Corneal topography of humans with congenital nystagmus. *Ophthal. Physiol. Opt.* **4,** 3 -13.

Dickinson, C.M. and Abadi, R.V. (1985). The influence of nystagmoid oscillation on contrast sensitivity in normal observers. *Vision Res.* **25,** 1089-1096.

Di Stefano, M., Bedard, S., Marzi, C. A. and Lepore, F. (1984). Lack of binocular activation of cells in area 19 of the Siamese cat. *Brain Res.* **303,** 391-395.

Donzis, P.B., Pappazzoo, J.A., Burde, R.M. and Gordon, M. (1983). Effect of binocular variation of Snellen visual acuity on Titmus stereoacuity. *Arch. Ophthalmol.* **10,** 903-932.

Dubois-Poulsen, A. and L'Anthony, P. (1973). Le Farnsworth-15 désatuŕe. *Bull. Soc. Opthalmol.* **73,** 861-866.

Durham-Pierre, D., Gardner, J.M., Nakatsu, Y., King, R. A., Franke, U., Ching, A. and Aquaron, R. (1994). African origin of an intragenic deletion of the human P gene in tyrosinase positive Oculocutaneous albinism. *Nat. Genet.* **7,** 176 - 179.

Durham-Pierre, D., King, R.A., Naber, J.M., Laken. S. and Brilliant, M.H. (1996). Estimation of carrier frequency of a 2.7 kb deletion allele of the P gene associated with OCA2 in African Americans. *Hum. mut.* **7,** 370 - 373.

Ewusie, J. Y. (1988). Characterization of the genetic profile of Swaziland, the ABO blood groups and albinism. *Swazi Sci. Technol.* **9,** 45 - 55.

Fansworth, D. (1947). The Farnsworth Dichotomous Test for colour blindness panel D-15 manual. Psychol Corporation, New York.

Findlay, G.H. (1962). On giant dendritic freckles and melanocyte reactions in the skin of an albino patient. *S. Afr. Lab. Clin. Med.* **8,** 68 -72.

Fledelius, H.C. (1984). Prevalence of astigmatism and anisometropia in adult Danes. *Acta. Ophthalmol.* **62,** 391-400.

Fletcher, R. and Voke, J. (1985). Defective colour vision: fundamentals, diagnosis and management. Adam Hilger Ltd, Bristol.

Fonda, G., Thomas, H. and Gore, G.V. (1971). Educational and vocational placement and low vision corrections in albinism. *Sight Sav. Rev.* **4,** 29 -35.

Fox, R., Patterson, R. and Francis, E.L. (1986). Stereoacuity in young children. *Invest.* Ophthalmol. *Vis. Sci.* **27,** 598 - 600.

Fulcher, T., O'keefe, M., Bowell, R., Lanigan, B., Burke, T., Carr, A., O'Rourke, M. and Bolge, M. (1995). Intellectual and Educational Attainment in albinism. *J. Pediatr. Ophthalmol. Strab.* **32,** 368-372.

Fulton, A.B., Albert, D.M. and Craft, J.L. (1978). Human albinism: light and electron microscopy study. *Arch. Ophthalmol.* **96,** 305-310.

Goggin, M. and Pesudovs, K. (1998a) Assessment of surgically induced astigmatism: toward an international standard. J. Cataract. Refract. Surg. **24,** 1548-1550.

Goggin, M. and Pesudovs, K (1998b). Assessment of surgically induced astigmatism: toward an international standard II. *J. Cataract. Refract. Surg.* **24,** 1552-1553.

Gordon, A. (1990). Refractive error in a Puerto Rican rural population. *J. Am. Optom. Assoc.* **61,** 870-874.

Gross, A., Kunze, J. and Maire, R.F. (1995). Autosomal recessive neural crest syndrome with albinism, black lock, cell migration disorder of the neurocytes of the gut and deafness:

ABCD syndrome. *Am. J. Genet.* **56,** 322-326.

Grosvenor, T.P. (1989). Primary Care Optometry, 2nd edn.,30 - 44. Professional press, Chicago.

Guillery, R.W. and Kass, J.H. (1973). Genetic abnormality of visual pathways in a "white" tiger. *Science* **180,** 1287-1289.

Guillery, R.W., Okoro, A.N. and Witkop, C.J. Jr. (1975). Abnormal visual pathway in the brain of a human albino. *Brain Res.* **96,** 373-377.

Guillery, R.W., Hickey, T.I., Kass, J.H., Felleman, D.J., Debruyn, E.J. and Sparks, D.L. (1984). Abnormal central visual pathways in the brain of an albino green monkey (*Cercopithecus aethiops*). *J. Comp. Neurol.* **226,** 165-183.

Guo, S., Reinecke, R.D., Fendick, M. and Calhoun, J.H. (1989). Visual pathway Abnormalities in albinism and infantile nystagmus: VECPs and stereoacuity measurements. *J. Pediatr. Ophthalmol. Strab.* **26,** 97 - 104.

Hall, M., Eusebi, B. and D' Angelo, I. (1976). Multiple pigmented naevi in an albino. *Br. J. Dermatol.* **98,** 685- 692.

Harris, W.F. (1988a). The matrix representation of dioptric power. Part 1: An introduction. *S. Afr. Optom.* **47,** 19-23.

Harris, W.F. (1988b). Squaring the sphero-cylinder, the equivalent of squaring the refractive power matrix. *Ophthal. Physiol. Opt.* **8,** 458-459.

Harris, W.F (1989). The matrix representation of dioptric power. Part 2: Adding obliquely-crossed sphero-cylinders. *S. Afr. Optom.* **48,** 22-24.

Harris, W.F. (1991). Representation of dioptic power in Euclidean 3-space. *Ophthal. Physiol. Opt.* **11,** 130-136.

Harris, W.F. (1997). Dioptric power : Its nature and its representation in three and four dimensional space. *Optom. Vis. Sci.* **74,** 349-366.

Harris, W.F. (2000). Astigmatism. *Ophthal. Physiol. Opt.* **20,** 11-30.

Harvey, E.M., Miller, J.M. and Dobson, V. (1995). Reproducibility of corneal astigmatism measurements with a hand held keratometer in preschool children. *Br. J. Ophthalmol.* **79,** 983-990.

Held, R., Birch, E. and Gwiazda, J. (1980). Stereoacuity in human infants. *Proc. natl. Acad. Sci. USA.* **77,** 5572-5574 .

Helve, J. (1972). A comparative study of several diagnostic tests of colour vision used for measuring types and degrees of congenital red-green defects. *Acta. Ophthalmol. Suppl* 115.

Holladay, J.T., Dudeja, D.R. and Koch, D.D. (1998). Evaluating and reporting astigmatism for individual and aggregate data. *J. Cataract. Refract. Surg.* **24,** 57-65.

Hyams, S.W., Pokotilo, E. and Shkurko, G. (1977). Prevalence of refractive errors in adults over 40 years: a survey of 8102 eyes. *Br. J. Ophthalmol.* **61,** 428 - 432.

Jay, B., Carruthers, B. and Treplin, M.C.W. (1976). Human albinism. *Birth defects* **12,** 415-426.

Jimene-Cervantes, C., Solana, F., Kobawashi, T., Urabe, K., Hearing, V.J., Lozano, J.A.

and Garcia, B. (1994). A new enzymatic function in the melanogenic pathway: the DHICA oxidase activity of tyrosinase related protein -1 (typ1). *J. Biol. Chem.* **269,** 17993 - 1800.

Johnson, G.J., Matthews, A. and Perkins, E.S. (1979). Survey of ophthalmic conditions in a Labrador community. I: refractive errors. *Br. J. Ophthalmol.* **63,** 440 -448.

Kagore, F. and Lund, P.M. (1995). Oculocutaneaous albinism among school children in Harare, Zimbabwe. *J. Med. Genet.* **32,** 859 - 861.

Kalloniatis, M., Harwerth, R.S., Smith III, E.L. and De Santis, L. (1989). Colour vision anomalies in glacoma. *Optom. Vis. Sci.* **66,** (suppl) 203.

Kalloniatis, M. and Johnston, A.W. (1990). Colour vision characteristics of visually impaired children. *Optom. Vis. Sci.* **67,** 166-168.

Karabatsas, C.H., Cook, S.D., Papaefthymiou, J., Turner, P. and Sparrow, J.M. (1998) . Clinical evaluation of keratometry and computerized videokeratography : Intraobserver variability on normal and astigmatic corneas. *Br. J. Opthalmol.* **82,** 637-642.

Kelly, D.H. and Burbeck, C.A. (1984). Critical problems in spatial vision CRC. *Crit. Rev. Biomed. Eng.* **10,** 125 - 177.

King, R.A., Creel, D., Cervenka, J., Okoro, A. N. and Witkop, C.J. (1980). Albinism in Nigeria with delienation of new oculocutaneous type. *Clin. Genet.* **17,** 259-270.

King, R.A. and Summers, C.G. (1988). Albinism. *Dermat. Clin.* **6,** 217 - 228.

King, R. A., Hearing, V.J. and Creel, D.J. (1995). Albinism. In the metabolic and molecular basis of inherited disease (eds. Scriver, C.R., Beaudet, A.L., Sly, W.S. and Valle, D.)

4353-4392. McGraw-Hill, New York .

Kinnear, P.E., Jay, B. and Witkop, C.J. (1985). Albinism. *Surv. Ophthalmol.* **30,** 75 - 101.

Kragha, I.K.O. (1987). The distribution of refractive errors in Nigeria. *Ophthal. Physiol. Opt.* **7,** 241 - 244.

Kriss, A., Russell- Eggit, I. Harris, C.M., Lloyd, I.C. and Taylor, D. (1992). Aspects of albinism. *Ophthalmol. Pediatr .Genet.* **13,** 89-100.

Kromberg, J.G.R. and Jenkins, T. (1982) Prevalence of albinism in the South African Negros. S. Afr. Med. J. **61,** 383 - 386.

Kromberg, J.G.R. and Jenkins, T. (1984). Albinism in South African Negros: III. Genetic counseling issues. *J. Biosoc. Sci.* **16,** 99-108.

Kromberg, J.G.R., Castle, D., Zwane, E.M. and Jenkins, T. (1989). Albinism and skin cancer in Southern Africa. *Clin. Genet.* **36,** 43 -52.

Kromberg, J.G.R., Castle, D.J., Zwane, E.M., Bothwell, J., Kidson, S., Bartel, P., Phillips, V.L. and Jenkins, T. (1990). Red Rufous albinism in Southern Africa. *Ophthalmol. Pediatr. Genet.* **11,** 229 - 235.

Kupersmith, J.J., Seiple, W.H. and Nelson, J.I. (1984). Contrast sensitivity loss in multiple sclerosis. *Invest. Ophthalmol. Vis. Sci.* **25,** 632-639.

Lam, C.S., Goh, W.S., Tang, Y.K., Tsui, K.K., Wong, W.C. and Man, T.C. (1994). Changes in refractive trends and optical components of Hog Kong Chinese aged over forty. *Ophthal. Physiol. Opt.* **14,** 383-388.

Lang, J. (1983). A new stereotest. *J. Pediatr. Ophthalmol. Strab.* **20,** 72 -74.

Larson, W.I. and Lanchance, A. (1983). Stereoscopic acuity with induced refractive errors. *Am. J. Optom. Physiol. Opt.* **60,** 509-513.

La Vail, J.H., Nixon, R.A. and Sidman, R.L. (1978). Genetic control of retinal ganglion cell projections. *J. Comp. Neurol.* **182,** 399-422.

Lavery, J.R., Gibson, J.M., Shaw, D.E. and Rosenthal, A.R. (1988). Refraction and refractive errors in an elderly population. *Ophthal. Physiol. Opt.* **8,** 394-396.

Levi, D.M. and Harwerth, R.S. (1977). Spatio-temporal interactions in anisometropic and strabismic amblyopia. *Invest. Ophthalmol. Vis. Sci.* **16,** 90 -95.

Lookingbill, D.P., Lookingbill, G.L. and Leppard, B. (1995). Actinic lentigines versus skin cancer risk in albinos in Northern Tanzania. *J. Am. Acad. Dermatol.* **33,** 299 - 300.

Loshin, P.S. and Browning, R.A. (1983). Contrast sensitivity in albinotic patients. *Am. J. Optom. Physiol. Opt.* **6,** 158 - 166.

Lovie, J.E. and Kitchin, J.E. (1988). Validity and variability of visual acuity measurements. *Ophthal. Physiol. Opt.* **8,** 363-370.

Luande, J., Henschke, C.I. and Mohammad, N. (1985). The Tanzanian human albino skin. *Cancer* **55,** 1823 - 1828.

Lund, P.M. (1996). Distribution of oculocutaneous albinism in Zimbabwe. *J. Med. Genet.* **33,** 641 - 644.

Lund, P.M. (1997). The distribution and education of pupils with albinism in Zimbabwe. *Trans. Zim. Assoc.* **71,** 9-13.

Lyle, M.W., Sangster, J.O. and Williams, T.D. (1997). Albinism: an update and review of the literature. *J. Am. Optom. Assoc.* **68,** 623-645.

Malan, D.J. (1990). Computer program for calculating mean refractive error. *S. Afr. Optom.* **49,** 83-85.

Manga, P., Kromberg, J.G.R., Box, N.F., Sturm, R.A., Jenkins, T. and Ramsay, M. (1997). *Am. Hum. Genet.* **61,** 1095-1101.

Manganyi, N.K., Kromberg, G.R. and Jenkins, T. (1974). Studies on albinism in the South African Negros. 1: intellectual maturity and body image differentiation. *J. Biosoc. Sci.* **6,** 107 - 112.

Manny, R.E., Martinez, A.T. and Fern, K.D. (1991). Testing stereoacuity in the preschool children : Is it clinically useful? *J. Pediatr. Ophthalmol. Strab.* **28,** 223 - 231.

O'Donnell, F.E., Hambrick, G.W., Iliff, W.J. and Stone, D.I. (1976). X-linked ocular albinism. An oculocutaneous macromelanosomal disorder. *Arch. Ophthalmol.* **94,** 1883-1892.

O'Donnell, F.E., King, R.A. and Green, W.R. (1978). Autosomal recessively inherited ocular albinism: a new form of ocular albinism affecting females are severely as males. *Arch. Ophthalmol.* **96,** 1621-1625.

O'Donnell, F.E. and Green, W.R. (1981). The eye in albinism. In Clinical Ophthalmol (ed. Duane, T.D.) Vol 4, Chapt. 38, 1-10. Harper and Row, New York.

Oduntan, A.O. (1998). Colour vision in albinism. Unpublished data.

Oduntan, A.O., Al-Ghamdi, M. and Al-Dosari, H. (1998). Randot Stereoacuity norms in a population of Saudi Arabian children. *Clin. Exp. Optom.* **81,** 193 - 197.

Oduntan, A.O. and Briggs, S.T. (1999). An arabic letter distance visual acuity test chart for young children and illiterate adults. *Ophthal. Physiol. Opt.* **19,** 431-437.

Oetting, W.S., Brilliant, M.H. and King, R.A. (1996). The clinical spectrum of albinism. *Mol. Med. Today*. 330 - 335.

Oetting, W.S. and king, R.A. (1999). Molecular basis of albinism. Mutations and polymorphisms of pigmentation genes associated with albinism. *Human. Mut.* **13,** 99- 115.

Okoro, A.N. (1975). Albinism in Nigeria. *Br. J. Dermatol.* **92,** 485 - 492.

Parssinen, T.O. (1987). Relation between refraction, education, occupation and age among 26-and 46-year-old Finns. *Am. J. Optom. Physiol. Opt.* **64,** 136-143.

Perez-Carpinell, J. Capilla, P., Illueca, C. and Morales, J. (1992). Vision defects in albinism. *Am. Acad. Optom.* **69,** 623-628.

Pickford, R.W. (1958). Colour vision of three albinos. *Nature* **181,** 361 - 362.

Pickford, R.W. and Taylor, W.O.G. (1968). Colour vision of two albinos. *Br. J. Ophthalmol.* **52,** 640 - 641.

Pokorny, J., Smith, V.C., Verriest, G. and Pinckers, A.L.G. (1979). Congenital and acquired colour vision defects. Grune and Straton, New York.

Puri, N., Durham-Pierre, D., Aquaron, R., Lund, P.M., king ,R.A. and Brilliant, M.H. (1997). Type 2 oculocutoneous albinism (OCA 2) in Zimbabwe and Cameroon: deletion of the 2.7 kb deletion allele of the P gene. *Human. Genet* .**100,** 651-656.

Raliavhegwa, M. and Oduntan, A. O. (2000). A retrospective study of the refractive status of a South African black population. *S. Afr. Optom.* **59,** 72- 77.

Ramsay, M., Colman, M.A., Stevens, G., Zwane, E., Kromberg, J., Farrell, M. and Jenkins, T. (1992). The tyrosinase positive oculocutaneous albinism locus maps to chromosome 15q 11.2 -q12. *Am. J. Hum. Genet.* **51,** 879 - 884.

Richler, A. and Bear, J.C. (1980). The distribution of refraction in three isolated communities in Western Newfoundland. *Am. J. Optom. Physiol. Opt.* **57,** 861-871.

Rinchik, E.M., Bultman, S.J., Horsthemke, B., Lee, S.T., Strunk, K.M., Avidano, K.M., Jong, M.I.C. and Nicholls, R.D. (1993). A gene for the mouse pink-eyed dilution locus and for human type II oculocutaneous albinism. *Nature* **361,** 72- 76.

Rogers, G.L,. Bremer, D.L. and Leguire, L.E. (1987). Contrast sensitivity functions in normal children with the Vistech method. *J. Pediatr. Ophthalmol. Strab.* **24,** 216-219.

Romano, P., Romano, I. and Puklin, J. (1975). Stereoacuity development in children with normal binocular vision. *Am. J. Ophthalmol.* **79,** 966 -971.

Royal National Institute for the blind (RNIB) and Royal College of Ophthalmologists (RCO) (1999). Understanding nystagmus. page 5. RNIB, RCO and nystagmus network, London.

Sacharowitz, H.S. (1999). An overview of oculocutaneous albinism in South Africa. *S. Afr. Optom.* **58,** 105-109.

Saunders, H. (1980). A method for determining the mean value of refractive errors. *Br. J. Physiol. Opt.* **34,** 1-11.

Saunders, K.J., woodhouse, J.M. and Westhall, C.A. (1996) The modified Frisby stereotest. J. Pediatr. Ophthalmol. Strab. **33,** 323 - 327.

Scharre, J.E., Cotter, S.A., Block, S.S. and Kelly, S.A. (1990) Normative contrast sensitivity for young children. Optom. Vis. Sci. **67,** 826-832.

Segers, J.E. (1929) Examen psychologique d'unenfant albinol. J. Neurol. Psych. **29,** 613-622.

Shea, S.L., Fox, R., Aslin, R.N. and Dumain, S.T. (1980). Assessment of Stereopsis in human infants. *Invest. Ophthalmol. Vis. Sci.* **19,** 1400 - 1404.

Shiono, T., Tsunoda ,M., Chida, Y., Makazawa, M. and Tamai, M. (1995). X-linked ocular albinism in Japanese patients. *Br. J. Ophthalmol.* **79,** 139-143.

Silvers, W.K. (1979). The coat colour of mice - A model for mammalian gene action and interaction. Springer - Verlag, New - York.

Silvers, J. (1984). Studies on the factors that govern directionality of axon growth in the embryonic optic nerve and at the chiasma of mice. *J. Comp. Neurol.* **223,** 238-251.

Simons, K. and Reinecke, R.D. (1974). A reconstruction of amblyopia screening and stereopsis. *Am. J. Ophthalmol.* **78,** 707 - 713.

Sloan, L.L. and Habel, A.B. (1956). Tests for colour deficiency based on the pseudoisochromatic principle. *Am. Arch. Ophthalmol.* **55,** 229-239.

Spritz, R.A., Fukai, K., Holmes, S.A. and Luande, J. Frequent intragenic deletion of the P Gene in Tanzanian patients with type 11 Oculocutaneous albinism (OCA2) (1995). *Am. J. Hum. Genet.* **56,** 1320 - 1323.

Stannus, H. S. (1913). Anomalies of pigmentation among natives of Nyasaland. *Boimetrika* **9,** 333-365.

Sterward, J.M. and Cole, B.L. (1989). What do colour vision defectiveness say about everyday tasks? *Optom. Vis. Sci.* **66,** 288-295.

Stevens, G., Van Beukering, J. Jenkins, T. and Ramsay, W. (1995). An intragenic deletion of the P gene is the common mutation causing tyrosinase-positive oculocutaneous albinism in Southern African Negroids. *Am. J. Hum. Genet* **56,** 586 - 591.

Stone, J., Rowe, M.H. and Campion, J.E. (1978). Retinal abnormalities in the Siamese cat. *J. Comp. Neurol.* **202,** 521-538.

Summers, C.G., Creel, D. and Townsend, D. (1991). Variable expression of vision in sibs with albinism. *Am. J. Med. Genet.* **40,** 327-331.

Summers, C.G. (1996). Vision in albinism. *Trans. Am. Ophthalmol. Soc.* **94,** 1095-1155.

Summers, C.G. Oetting, W.S. and King, R.A. (1996). Diagnosis of oculocutaneous albinism with molecular analysis. *Am. J. Ophthalmol.* **121,** 724-726.

Swanson, W.H. and Everett, M. (1992). Colour vision screening of young children. *J. Pediatr. Ophthalmol. Strab.* **29,** 49-54.

Taylor, W.O.G. (1976). Albinism and colour defects. *Mod. Probl. Ophthalmol.* **17,** 292 - 298.

Taylor, W.O.G. (1978). Visual disabilities of oculocutaneous albinism and their alleviation. *Trans. Opthalmol. Soc. UK.* **98,** 423-445.

Thibos, L.N. (1992). Application of Fourier analysis to the description of optical lens power and to the statistical analysis of refractive error. *Optom. Vis. Sci.* **69,** (suppl) 114.

Thulin, H.C. (1972). Colour blindness in children. The importance and feasibility of early recognition. *Clin. Pediatr.* **11,** 295-299

Tripathi, R.K., Giebel, L.B., Strunk, K.M. and Spitz RA (1991). A polymorphism of the human tyrosinase gene is associated with temperature - Sensitive enzymatic activity. *Genet. Exp.* **1,** 103-110.

Venter, P.A., Christianson, A.L., Hutamo, C.M., Makhura, M.P. and Gericke, G.S. (1995). Congenital anomalies in rural black South African neonates- a silent epidemic. *S.A. M. J.* **85,** 15-20.

Virsus, V,. Lehtro, P. and Rovamo, J. (1981). Contrast sensitivity in normal and pathological vision. *Doc. Opthalmol. Proc. Series.* **30,** 263-272.

Walsh, R.J. (1971). A distinctive pigment of the skin in New Guinea Indigenes: *Ann. Hum. Genet.* **34,** 379-388.

Webster, M.J., Shatz, C.J. and Silver, J. (1988). Abnormal pigmentation and unusual morphogenesis of the optic stalk may be correlated with retinal exon misguidance in embryonic Siamese cats. *J. Comp. Neurol.* **269,** 592-611.

Westheimer, G .W. (1979). Scaling visual acuity measurement . *Arch. Ophthalmol.* **97,** 327-330.

Wild, J.M. and Hussey, M.K. (1985). Some statistical concepts in the analysis of vision and visual acuity. *Ophthal. Physiol. Opt.* **5,** 63-71.

Who offset Publ (1980). Methods of assessment of avoidable blindness. **54,** 1-42.

Wilson, H.R., Mets, M.B., Nagy, S.E. and Kressel, A.B. (1988a). Is the albino visual system a useful model for infant spatiotemporal vision? *Vis Res.* **28,** 979-990.

Wilson, H.R., Mets, M.B., Nagy S.E. and Ferreira, V.P. (1988 b). Spatial frequency and orientation tuning of visual mechanisms in the human albino. *Vis Res.* **28,** 991-999.

Witkop, C.J., Nance, W.E., Fawls, R.F. and White, J.G. (1970). Autosomal recessive oculocutaneous albinism in man: Evidence for genetic heterogeneity. *Am. J. Hum. Genet.* **22,** 55-74.

Witkop, C.J. (1971). Albinism: In Advances in human genetics (Harris H, Hischonrn K eds) vol 2, 61-142. Plenum Press, New York,

Witkop, C.J. Jr., Hill, C.W. and Desnick, S. (1973). Ophthalmologic, biochemical platelet and ultrastructural defects in the various types of oculocutaneous albinism. *J. Invest. Dermatol.* **60,** 443-456.

Witkop, C.J., Jay, G., Creel, D. and Quillery, R.W. (1982). Optic and otic neurologic abnormalities in oculocutaneous and ocular albinism. *Birth Defects* **18,** 299-318.

Witkop, C.J. (1985). Inherited disorders of pigmentation. *Clin. Dermatol.* **94,** 70 - 134.

Witkop, C.J., Quevedo, W.C., Fitz Patrick, T.B. and King, R.A. Albinism (1989). The metabolic basis of inherited disease. In Scriver, C.r., Beaudet, A.L., Sly, W.S., Valle, D. eds, 6th edn., 2905 - 2947. Mc GrawHill, New York.

APPENDIX A: UNIVERSITY OF THE NORTH CONSENT FORM.

UNIVERSITY OF THE NORTH
ETHICS COMMITTEE

PROJECT TITLE:

PROJECT LEADER:

CONSENT FORM

I, ---------- hereby voluntarily consent to participate in the following project: **A study of vision problems among children with oculocutaneous albinism attending special education schools in the Northern Province of South Africa.**

I realise that:

1. The study deals with **vision problems among children with oculocutaneous albinism attending special education schools in the Northern Province of South Africa.**

2. The procedure or treatment envisage may hold some risk for me that cannot be foreseen at this stage;

3. The Ethics Committee has approved that individuals may be approached to participate in the study.

4. The experimental protocol, ie. The extent, aims and methods of the research, has been explained to me.

5. The protocol sets out the risk that can be reasonably expected as well as possible discomfort for persons participating in the research, an explanation of the anticipated advantages for myself or others that are reasonably expected from the research and alternative procedures that may be to my advantage;

6. I will be informed of any new information that may become available during the research that may influence my willingness to continue my participation;

7. Access to the records that pertain to my participation in the study will be restricted to persons directly involved in the research;

8. Any questions that I may have regarding the research, or related matters, will be answered by the researchers;

9. If I have any questions about, or problems regarding the study, or experience any undesirable effects, I may contact a member of the research team;

10. Participation in this research is voluntary and I can withdraw my participation at any stage;

11. If any medical problem is identified at any stage during the research, or when I am vetted for participation, such condition will be discussed with me in confidence by a qualified person and/ or I will be referred to my doctor;

12. I indemnify the University of the North and all persons involved with the above project from any liability that may arise from my participation in the above projects

or that may be related to it, for whatever reasons, including negligence on the part of the mentioned persons.

_____	_____
Signature of researched person	Signature of witness
_____	_____
Signature of person that informed the researched person	Signature of parent/guardian

signed at_____this _____ day of _____200-

APPENDIX B: Record Form

ALBINO VISION PROJECT

Department of Optometry,

University of the North.

RECORD FORM

Name..... **Age**..... **Sex**.....

School..... **Home address**.....

.....

Case History

Visual acuity	Distance	Near
OD		
OS		
OU		

Cover tests

Strabismus	Heterophoria

Retinoscopy

OD

OS

Monocular subjective

OD..... OS..... VA: OD..... OS.....OU.....

Binocular balance

OD..... OS..... VA: OD.....OS.....OU.....

Near: OD.....OS.....OU.....

Contrast sensitivity (VISTEC)

Stereoacuity (Randot)

Colour vision tests

Ishihara

Farnsworth panel D15.

Ophthalmoscopy

APPENDIX C: REFRACTIVE STATUS AND DISTANCE VISUAL ACUITY DATA FOR THE CHILDREN WITH OCULOCUTANEOUS ALBINISM N= 154.

REFRACTIVE STATUS AND VISUAL ACUITY DATA																		
S/N	AGE	SEX	RX (RIGHT EYE)				RX (LEFT EYE)				VA (RIGHT EYE)			VA (LEFT EYE)			VA (BOTH EYE)		
			Sph	Cyl	Axis	N/E	Sph	Cyl	Axis	N/E	V1	V2	V3	V1	V2	V3	V1	V2	V3
1	13	M	+1.25	-2.00	15	+0.25	+1.00	-2.00	155	0.00	0.56	0.62	0.06	0.56	0.62	0.06	0.54	0.64	0.1
2	12	M	-1.00	0.00	-	-1.00	-1.25	-0.25	80	-1.25	0.44	0.50	0.06	0.40	0.5	0.10	0.44	0.5	0.06
3	14	M	+1.00	0.00	-	+1.00	+1.00	-	0.00	+1.00	0.34	0.40	0.06	0.32	0.36	0.04	0.34	0.4	0.06
4	13	F	+1.00	-0.75	170	+0.75	+1.00	-0.75	30	+0.75	0.38	0.62	0.24	0.44	0.62	0.18	0.44	0.54	0.1
5	14	F	-0.75	-0.50	145	-1.00	-1.25	-	0.00	-1.25	0.50	0.56	0.06	0.40	0.58	0.18	0.48	0.58	0.1
6	13	M	-0.75	-0.25	30	-1.00	-0.75	-0.75	140	-1.00	0.46	0.52	0.06	0.48	0.52	0.04	0.5	0.52	0.02
7	13	F	-1.00	-0.50	75	-1.25	-1.25	-1.00	73	-1.75	0.34	0.44	0.10	0.34	0.34	0.00	0.4	0.42	0.02
8	13	F	-1.00	-0.75	130	-1.25	-1.75	-0.50	135	-2.00	0.32	0.50	0.12	0.30	0.50	0.20	0.34	0.54	0.2
9	12	F	+1.50	-1.50	10	+0.75	+2.50	-3.00	160	+1.00	0.26	0.60	0.24	0.36	0.50	0.14	0.26	0.50	0.24
10	12	F	-1.00	-0.75	150	-1.25	-0.75	-1.75	165	-1.50	0.42	0.54	0.18	0.48	0.60	0.12	0.54	0.54	0.10
11	12	F	-1.25	-0.50	65	-1.50	-1.50	-0.50	150	-1.50	0.44	0.5	0.10	0.44	0.44	0.00	0.48	0.52	0.04
12	12	M	-1.00	0.00	-	-1.00	-0.75	-1.25	140	1.25	0.44	0.44	0.06	0.44	0.44	0.00	0.44	0.48	0.1
13	12	F	+1.25	0.00	-	+1.25	-1.50	-	0	0	0.34	0.50	0.10	0.44	0.54	0.10	0.46	0.54	0.02

REFRACTIVE STATUS AND VISUAL ACUITY DATA																		
S/N	AGE	SEX	RX (RIGHT EYE)				RX (LEFT EYE)				VA (RIGHT EYE)			VA (LEFT EYE)			VA (BOTH EYE)		
			Sph	Cyl	Axis	N/E	Sph	Cyl	Axis	N/E	V1	V2	V3	V1	V2	V3	V1	V2	V3
14	12	F	-2.25	-1.00	45	-2.75	-3.00	-1.75	165	-3.75	0.42	0.50	0.08	0.34	0.34	0	0.46	0.48	0.02
15	13	M	+1.00	-0.50	15	+0.75	+1.00	-0.50	180	+0.75	0.22	0.4	0.18	0.50	0.50	0	0.44	0.40	0.08
16	13	F	-1.25	-0.50	110	-1.50	-1.00	-1.50	180	-1.75	0.30	0.40	0.10	0.28	0.40	0.12	0.26	0.04	0.14
17	14	M	-8.00	0.00	-	-8.00	-6.50	-1.00	15	-7.00	0.12	0.20	0.08	0.12	0.20	0.08	0.10	0.50	0.30
18	13	M	-1.00	-1.00	35	-1.50	+0.75	-0.25	65	+0.50	0.50	0.50	0	0.50	0.56	0.06	0.50	0.50	0.00
19	11	F	-1.25	-0.50	35	-1.50	-1.50	-0.50	15	-1.75	0.42	0.52	0.10	0.38	0.50	0.12	0.40	0.50	0.10
20	15	M	-1.75	-0.75	135	-2.00	-0.75	-1.50	25	-1.50	0.06	0.22	0.12	0.28	0.50	0.22	0.32	0.60	0.28
21	13	F	-0.75	-0.50	45	-1.00	-0.75	-0.50	165	-1.00	0.44	0.50	0.06	0.46	0.60	0.14	0.50	0.60	0.10
22	11	F	+1.25	-1.50	175	+0.75	+0.50	-1.25	180	-0.25	0.58	0.60	0.02	0.38	0.46	0.10	0.56	0.66	0.10
23	14	M	-5.00	-1.75	115	-5.75	-5.00	-1.50	165	-5.25	0.05	0.36	0.31	0.05	0.12	0.07	0.08	0.12	0.03
24	12	M	-1.00	-0.50	65	-1.25	-1.75	-0.50	115	-1.50	0.30	0.40	0.1	0.30	0.40	0.10	0.30	0.40	0.10
25	11	F	-1.25	-0.25	64	-1.25	-0.75	-0.75	65	-1.00	0.24	0.30	0.06	0.40	0.50	0.10	0.40	0.50	0.10
26	11	F	-5.00	0.00	-	-5.00	-3.00	0.00	-	-3.00	0.16	0.20	0.04	0.12	0.22	0.10	0.20	0.20	0.00

REFRACTIVE STATUS AND VISUAL ACUITY DATA																			
S/N	AGE	SEX	RX (RIGHT EYE)				RX (LEFT EYE)				VA (RIGHT EYE)			VA (LEFT EYE)			VA (BOTH EYE)		
			Sph	Cyl	Axis	N/E	Sph	Cyl	Axis	N/E	V1	V2	V3	V1	V2	V3	V1	V2	V3
27	12	F	+1.50	-0.75	15	+0.25	+1.00	-2.50	145	-0.25	0.56	0.68	0.12	0.60	0.68	0.08	0.40	0.60	0.20
28	11	F	-1.00	-1.25	45	-1.50	-1.00	-1.00	15	-1.50	0.42	0.52	0.1	0.54	0.64	0.10	0.42	0.54	0.12
29	12	F	-1.50	-0.50	53	-1.75	-1.75	-0.50	15	-2.00	0.48	0.66	0.18	0.40	0.50	0.10	0.46	0.50	0.04
30	14	M	-1.25	-3.00	25	-0.25	-100	-0.75	140	-1.25	0.44	0.52	0.08	0.60	0.52	0.08	0.44	0.56	0.12
31	12	M	+0.75	-3.00	85	-075	+0.75	-3.00	105	-0.75	0.50	0.54	0.04	0.54	0.56	0	0.20	0.30	0.10
32	11	M	-1.00	-0.75	15	-1.25	-1.00	-0.75	15	-1.25	0.48	0.58	0.10	0.40	0.58	0.1	0.52	0.66	0.14
33	12	M	-1.00	-0.50	25	-1.25	-0.75	-1.00	75	-1.25	0.46	0.46	0	0.44	0.56	0.1	0.46	0.62	0.16
34	13	F	-1.25	-0.50	15	-1.50	-0.75	-1.75	165	-1.50	0.34	0.44	0.1	0.56	0.52	0.18	0.48	0.70	0.22
35	10	F	-1.50	-0.50	45	-1.75	-1.50	-0.25	20	-1.50	0.34	0.54	0.20	0.48	0.56	0.20	0.50	0.62	0.12
36	14	M	+1.00	-0.50	130	+0.75	+1.00	0.00	-	+1.00	0.1	0.30	0.20	0.46	0.3	0.20	0.40	0.60	0.20
37	10	F	+1.50	-1.75	25	+0.75	+1.50	-1.50	15	+0.75	0.52	0.52	0	0.34	0.46	0.16	0.32	0.46	0.14
38	10	F	-1.25	-1.75	25	-2.25	-1.25	-2.75	165	-2.50	0.42	0.54	0.12	0.36	0.56	0.10	0.26	0.30	0.04
39	12	M	-1.00	-1.75	65	-1.75	-1.25	-0.75	130	-1.50	0.48	0.70	0.22	0.1	0.66	0.1	0.42	0.68	0.24

REFRACTIVE STATUS AND VISUAL ACUITY DATA																		
S/N	AGE	SEX	RX (RIGHT EYE)				RX (LEFT EYE)				VA (RIGHT EYE)			VA (LEFT EYE)			VA (BOTH EYE)		
			Sph	Cyl	Axis	N/E	Sph	Cyl	Axis	N/E	V1	V2	V3	V1	V2	V3	V1	V2	V3
40	13	F	-1.75	-0.75	145	-2.00	-0.75	-0.75	175	-1.00	0.42	0.62	0.2	0.54	0.64	0.10	0.50	0.62	0.12
41	11	F	-1.25	-1.00	25	-2.25	-1.00	-1.75	140	-1.75	0.40	0.56	0.16	0.36	0.58	0.22	0.40	0.60	0.20
42	11	F	-2.75	-0.75	45	-3.00	-3.25	-0.75	3	-3.50	0.30	0.50	0.20	0.24	0.38	0.14	0.32	0.46	0.14
43	11	F	-2.00	-1.50	145	+1.25	+1.25	-1.00	115	+0.75	0.22	0.30	0.08	0.28	0.30	0.02	0.26	0.30	0.04
44	10	F	-2.25	-0.25	90	-2.25	-1.50	-0.25	100	-1.50	0.30	0.46	0.16	0.46	0.58	0.12	0.42	0.68	0.24
45	10	F	-1.50	-1.00	70	-2.00	-1.50	-0.25	160	-1.50	0.42	0.70	0.28	0.54	0.66	0.12	0.60	0.70	0.1
46	10	F	+1.00	-1.75	25	+0.25	+1.00	-1.75	5	+0.25	0.70	0.80	0.1	0.62	0.86	0.24	0.70	0.90	0.2
47	10	F	+1.75	-1.00	45	+1.25	+1.75	-1.25	145	+1.25	0.64	0.70	0.06	0.64	0.70	0.06	0.60	0.70	0.2
48	11	M	-1.75	-0.50	170	-2.00	-1.25	-1.00	5	-1.75	0.42	0.42	0	0.44	0.54	0.10	0.46	0.50	0.04
49	10	M	-2.75	-0.25	135	-2.75	-2.50	-0.75	95	-2.75	0.42	0.54	0.12	0.38	0.50	0.12	0.40	0.50	0.10
50	14	M	-2.25	-0.25	45	-2.25	-1.25	-2.50	45	-2.50	0.30	0.60	0.30	0.40	0.62	0.22	0.62	0.28	0.34
51	10	F	-1.00	-0.50	140	-1.25	-1.25	-0.75	130	-1.50	0.44	0.64	0.20	0.50	0.64	0.14	0.44	0.64	0.20
52	9	F	+1.75	-1.75	5	+1.00	+1.50	-1.75	175	+0.75	0.46	0.50	0.02	0.48	0.50	0.02	0.46	0.50	0.04

REFRACTIVE STATUS AND VISUAL ACUITY DATA																			
S/N	AGE	SEX	RX (RIGHT EYE)				RX (LEFT EYE)				VA (RIGHT EYE)			VA (LEFT EYE)			VA (BOTH EYE)		
			Sph	Cyl	Axis	N/E	Sph	Cyl	Axis	N/E	V1	V2	V3	V1	V2	V3	V1	V2	V3
53	11	F	+0.75	-1.00	28	+0.25	+1.00	-1.75	180	+0.25	0.46	0.56	0.10	0.44	0.56	0.12	0.50	0.66	0.16
54	8	F	-0.75	-1.75	130	-1.50	-0.75	-1.75	10	-1.50	0.28	0.30	0.02	0.28	0.30	0.02	0.3	0.3	0.00
55	11	F	-4.50	-2.25	30	-5.50	-5.00	-2.25	30	-6.00	0.18	0.44	0.26	0.18	0.44	0.26	0.16	0.44	0.28
56	11	F	-3.50	-1.75	65	-4.25	-3.75	-1.00	125	-4.25	0.24	0.32	0.08	0.26	0.34	0.17	0.26	0.34	0.08
57	11	M	+0.75	-0.25	15	+0.75	+0.75	-0.50	130	+0.50	0.60	0.66	0.06	0.54	0.66	0.12	0.48	0.60	0.12
58	10	M	-1.50	-1.00	15	-2.00	-1.50	-1.25	140	-2.00	0.36	0.60	0.20	0.50	0.60	0.1	0.50	0.60	0.1
59	11	M	-1.75	-1.00	130	-2.00	-2.00	-1.75	155	-2.75	0.40	0.48	0.02	0.26	0.48	0.22	0.40	0.46	0.06
60	14	M	-1.00	0.00	-	-1.00	-1.00	-0.50	75	-1.25	0.56	0.48	0.02	0.30	0.36	0.06	0.48	0.60	0.12
61	9	M	+1.25	-1.25	5	+0.75	+1.25	-1.25	5	+0.75	0.58	0.70	0.12	0.05	0.05	0	0.66	0.76	0.10
62	8	M	-1.25	-1.75	45	-2.00	-2.25	-1.50	145	-3.00	0.50	0.58	0.08	0.4	0.52	0.00	0.48	0.58	0.10
63	11	F	+1.00	2.25	45	+0.75	+1.00	-0.50	120	+0.75	0.58	0.76	0.18	0.58	0.76	0.12	0.66	0.76	0.10
64	9	M	-0.75	-1.00	20	-1.75	-0.50	-1.50	145	-1.50	0.54	0.70	0.16	0.54	0.64	0.18	0.50	0.64	0.10
65	9	M	+0.75	-2.75	50	+0.25	+1.00	-0.50	115	+0.75	0.56	0.66	0.10	0.60	0.70	0.10	0.48	0.66	0.14

REFRACTIVE STATUS AND VISUAL ACUITY DATA																		
S/N	AGE	SEX	RX (RIGHT EYE)				RX (LEFT EYE)				VA (RIGHT EYE)			VA (LEFT EYE)			VA (BOTH EYE)		
			Sph	Cyl	Axis	N/E	Sph	Cyl	Axis	N/E	V1	V2	V3	V1	V2	V3	V1	V2	V3
66	9	M	+2.00	-2.75	30	+0.50	+1.75	-2.75	150	+0.50	0.34	0.60	0.26	0.38	0.60	0.22	0.38	0.70	0.32
67	8	M	-0.75	-2.50	55	-2.00	-0.75	-2.25	160	-2.00	0.40	0.70	0.30	0.30	0.70	0.30	0.34	0.66	0.32
68	9	M	-6.00	-0.5	95	-6.25	-8.00	-0.50	30	-8.25	0.20	0.20	0	0.14	0.20	0.06	0.26	0.26	0.00
69	10	M	+1.25	-2.75	85	0	+1.25	-1.75	30	+0.50	0.60	0.78	0.18	0.52	0.76	0.24	0.66	0.76	0.10
70	9	M	+1.00	-0.50	120	+0.75	+1.00	-0.50	65	+0.75	0.40	0.76	0.36	0.50	0.62	0.12	0.44	0.66	0.22
71	9	F	+1.00	-0.75	65	+0.75	+0.75	-0.75	140	+0.50	0.40	0.66	0.24	0.36	0.50	0.14	0.44	0.66	0.22
72	9	F	+1.00	-0.50	150	+0.75	+1.00	-0.50	165	+0.75	0.47	0.66	0	0.47	0.66	0.24	0.42	0.66	0.24
73	9	F	-6.00	-1.75	35	-6.75	-5.00	-1.25	135	-5.50	0.18	0.18	0	0.20	0.2	0	0.20	0.20	0
74	9	M	+1.75	-1.25	180	+1.25	+2.00	-1.00	85	+1.50	0.32	0.32	0.16	0.32	0.32	0	0.36	0.36	0
75	7	M	+1.50	-1.50	50	+0.75	+1.25	-2.00	10	+0.50	0.44	0.60	0.10	0.44	0.66	0.22	0.46	0.64	0.18
76	11	M	-6.50	-0.75	175	-6.75	-8.00	-0.50	155	-8.25	0.20	0.30	0.10	0.04	0.20	0.16	0.20	0.30	0.10
77	9	M	+1.00	-1.00	60	+0.50	+2.25	-2.25	160	+1.00	0.22	0.30	0.26	0.22	0.32	0.10	0.30	0.32	0.02
78	8	M	+1.50	-1.75	25	+0.75	+1.25	-2.25	180	+0.25	0.54	0.80	0.20	0.54	0.72	0.18	0.48	0.78	0.30

REFRACTIVE STATUS AND VISUAL ACUITY DATA																			
S/N	AGE	SEX	RX (RIGHT EYE)				RX (LEFT EYE)				VA (RIGHT EYE)			VA (LEFT EYE)			VA (BOTH EYE)		
			Sph	Cyl	Axis	N/E	Sph	Cyl	Axis	N/E	V1	V2	V3	V1	V2	V3	V1	V2	V3
79	8	F	-0.75	-1.25	30	-1.25	+1.25	-0.75	160	+1.00	0.44	0.64	0.20	0.46	0.64	0.18	0.44	0.70	0.26
80	7	F	-1.00	-3.00	5	-2.50	-1.50	-1.50	150	-2.25	0.40	0.64	0.24	0.40	0.66	0.26	0.40	0.64	0.24
81	8	M	+1.75	-1.50	35	+1.0	+1.75	-1.75	145	+1.00	0.26	0.42	0.16	0.46	0.56	0.10	0.44	0.56	0.12
82	8	F	-2.50	-2.50	25	-3.75	-2.50	-2.50	85	-3.25	0.24	0.34	0.10	0.24	0.36	0.14	0.26	0.34	0.08
83	9	F	+3.00	-2.50	29	+1.75	+3.00	+3.00	18	+1.75	0.44	0.52	0.18	0.50	0.60	0.10	0.50	0.68	0.18
84	8	F	+0.75	-1.25	80	+0.25	+0.75	+0.75	130	+0.50	0.44	0.62	0.22	0.52	0.62	0.10	0.54	0.68	0.14
85	8	F	-0.50	-0.75	45	-0.75	-0.50	-1.00	160	-1.00	0.60	0.64	0.04	0.66	0.68	0.08	0.60	0.68	0.08
86	10	M	+2.50	-2.25	67	+1.50	+1.75	-1.50	160	+1.00	0.34	0.44	0.10	0.30	0.40	0.10	0.36	0.40	0.04
87	11	M	-12.0	0.00	-	-12.0	-1.75	-1.25	165	-2.25	0.05	0.19	0.01	0.40	0.30	0	0.40	0.40	0.00
88	8	M	-1.75	-1.25	45	-2.25	-1.50	-1.25	15	-2.00	0.36	0.40	0.04	0.30	0.52	0.14	0.34	0.44	0.10
89	9	F	-1.75	-0.75	75	-2.25	-1.75	-1.25	35	-2.25	0.50	0.7	0.20	0.52	0.04	0.10	0.42	0.66	0.24
90	9	M	-6.00	-3.50	175	-6.75	-5.50	-4.00	25	-7.50	0.08	0.12	0.02	0.04	0.60	0.08	0.08	0.12	0.04
91	8	F	-1.25	-0.25	35	-1.25	-1.00	-0.50	145	-1.25	0.62	0.72	0.10	0.60	0.32	0.12	0.60	0.70	0.10

REFRACTIVE STATUS AND VISUAL ACUITY DATA																			
S/N	AGE	SEX	RX (RIGHT EYE)				RX (LEFT EYE)				VA (RIGHT EYE)			VA (LEFT EYE)			VA (BOTH EYE)		
			Sph	Cyl	Axis	N/E	Sph	Cyl	Axis	N/E	V1	V2	V3	V1	V2	V3	V1	V2	V3
92	8	M	+2.00	-1.25	150	+1.50	+1.50	-1.00	150	+1.50	0.48	0.40	0.00	0.32	0.32	0.00	0.46	0.50	0.04
93	8	M	+1.50	-3.50	35	+0.25	+1.50	-3.50	140	+0.25	0.20	0.36	0.16	0.14	0.24	0.10	0.22	0.32	0.10
94	8	F	-6.50	-1.50	60	-7.25	-6.50	-1.50	160	-7.25	0.09	0.20	0.14	0.08	0.20	0.12	0.10	0.20	0.10
95	7	M	-6.50	0.00	-	-6.50	-7.00	0.00	-	-7.00	0.08	0.30	0.28	0.01	0.30	0.20	0.08	0.28	0.20
96	7	F	-2.00	-2.25	60	-3.00	-2.00	-1.25	80	-2.50	0.30	0.30	0.00	0.30	0.30	0.00	0.30	0.30	0.00
97	8	M	-1.25	-1.50	25	-2.00	-1.25	-1.50	30	-2.00	0.48	0.60	0.12	0.48	0.60	0.12	0.30	0.60	0.30
98	7	F	-6.50	-1.00	135	-7.00	-6.00	0.00	-	-6.00	0.09	0.16	0.00	0.08	0.20	0.12	0.08	0.30	0.20
99	7	F	-1.75	-1.00	110	-2.25	-1.50	-1.00	135	-2.00	0.30	0.40	0.10	0.32	0.40	0.10	0.34	0.40	0.06
100	7	M	+1.25	-1.50	25	+0.50	+1.75	-2.00	165	+0.75	0.48	0.76	0.28	0.52	0.70	0.18	0.54	0.76	0.24
101	7	M	-3.00	-2.00	110	-4.00	-3.50	-1.25	60	-4.00	0.20	0.34	0.14	0.22	0.32	0.10	0.20	0.30	0.10
102	7	F	-1.00	-1.00	180	-1.50	+0.75	-0.50	-	+0.50	0.40	0.60	0.20	0.32	0.62	0.30	0.36	0.64	0.28
103	8	M	+1.25	-	-	+1.25	+1.00	0.00	-	+1.00	0.32	0.32	0.00	0.32	0.32	0.00	0.40	0.40	0.00
104	7	F	+1.50	-1.25	45	+1.00	+1.75	-1.25	155	+1.25	0.30	0.70	0.40	0.34	0.66	0.32	0.30	0.64	0.34

REFRACTIVE STATUS AND VISUAL ACUITY DATA																		
S/N	AGE	SEX	RX (RIGHT EYE)				RX (LEFT EYE)				VA (RIGHT EYE)			VA (LEFT EYE)			VA (BOTH EYE)		
			Sph	Cyl	Axis	N/E	Sph	Cyl	Axis	N/E	V1	V2	V3	V1	V2	V3	V1	V2	V3
105	8	M	+1.00	-1.00	70	+0.50	+0.75	-1.25	165	+0.25	0.38	0.68	0.30	0.36	0.22	0.24	0.40	0.66	0.26
106	8	M	+0.75	-1.25	180	+0.25	+0.75	-1.75	10	-0.25	0.46	0.60	0.14	0.46	0.60	0.14	0.40	0.60	0.20
107	7	M	+1.25	-1.75	10	+0.50	+1.00	-1.25	175	+0.50	0.30	0.30	0.00	0.30	0.30	0.00	0.30	0.30	0.00
108	16	M	+3.75	-2.25	45	+2.75	+3.50	-0.75	45	+3.25	0.16	0.40	0.24	0.32	0.46	0.14	0.32	0.42	0.10
109	8	M	-1.75	-0.50	155	-2.00	-1.75	-1.00	120	-2.25	0.46	0.46	0.00	0.10	0.10	0.00	0.10	0.10	0.00
110	9	M	-5.00	-1.00	45	-5.50	-5.00	-0.75	90	-5.25	0.16	0.26	0.10	0.14	0.24	0.10	0.16	0.26	0.10
111	7	F	-1.50	-0.75	90	-1.75	-0.75	-0.75	90	-1.00	0.40	0.46	0.06	0.42	0.42	0.00	0.42	0.42	0.00
112	9	F	+0.75	-0.50	180	+0.50	+0.75	-0.50	170	+0.50	0.86	0.94	0.10	0.90	0.10	0.10	0.76	0.96	0.20
113	13	M	-2.75	-0.50	50	-3.00	-2.00	-0.75	135	-2.25	0.40	0.64	0.24	0.30	0.64	0.34	0.40	0.64	0.24
114	8	M	-2.75	-1.00	90	-3.25	-3.00	-1.00	180	-3.50	0.26	0.30	0.04	0.30	0.30	0.04	0.26	0.26	0.00
115	14	F	-3.50	-0.50	110	-3.75	-2.75	-0.75	110	-3.25	0.22	0.44	0.22	0.26	0.34	0.10	0.22	0.42	0.20
116	9	F	-1.75	-1.00	75	-2.25	-2.00	-1.75	135	-2.75	0.40	0.40	0.00	0.24	0.40	0.00	0.40	0.40	0.00
117	8	F	-1.25	-1.50	95	-2.00	-1.25	-1.25	130	-1.75	0.20	0.30	0.10	0.20	0.30	0.10	0.20	0.72	0.20

REFRACTIVE STATUS AND VISUAL ACUITY DATA																			
S/N	AGE	SEX	RX (RIGHT EYE)				RX (LEFT EYE)				VA (RIGHT EYE)			VA (LEFT EYE)			VA (BOTH EYE)		
			Sph	Cyl	Axis	N/E	Sph	Cyl	Axis	N/E	V1	V2	V3	V1	V2	V3	V1	V2	V3
118	14	F	-1.50	-0.75	45	-1.75	-1.50	-1.00	45	-2.00	0.54	0.70	0.16	0.50	0.70	0.20	0.50	0.72	0.22
119	11	M	-1.25	-0.75	110	-1.50	-1.50	-0.50	110	-1.75	0.30	0.50	0.20	0.44	0.60	0.16	0.40	0.60	0.20
120	12	M	-1.25	-1.75	60	-2.00	-1.75	-0.75	55	-2.00	0.30	0.40	0.10	0.32	0.46	0.14	0.36	0.44	0.12
121	17	M	-3.00	-0.50	110	-3.25	-2.50	-0.50	70	-2.75	0.26	0.40	0.14	0.22	0.40	0.18	0.32	0.44	0.24
122	12	F	+1.00	-1.00	30	+0.50	+0.75	-1.50	170	+0.25	0.60	0.70	0.10	0.60	0.70	0.10	0.60	0.70	0.10
123	12	M	+0.25	-1.75	40	-0.50	+0.25	-1.50	140	-0.50	0.52	0.72	0.20	0.54	0.72	0.20	0.62	0.76	0.14
124	12	M	0	-	-	0.00	-1.00	-2.25	170	-2.00	FC	FC	0.00	0.30	0.70	0.40	0.30	0.40	0.10
125	10	M	+0.50	-0.75	30	+0.25	-0.50	-0.50	95	-0.75	0.48	0.68	0.20	0.20	0.50	0.10	0.40	0.50	0.10
126	10	F	-7.75	-0.50	35	-2.00	-1.50	-1.50	130	-2.25	0.26	0.40	0.14	0.14	0.40	0.10	0.3	0.40	0.10
127	9	F	-2.50	-0.50	170	-2.75	-2.75	-1.50	45	-3.50	0.08	0.30	0.21	0.21	0.30	0.10	0.12	0.26	0.14
128	10	M	+0.50	-0.25	110	+0.50	+0.75	-1.50	65	+0.50	0.46	0.66	0.20	0.20	0.76	0.10	0.56	0.76	0.20
129	12	F	-1.25	-0.75	140	-1.75	-1.25	-1.25	150	-1.75	0.38	0.60	0.22	0.22	0.46	0.16	0.26	0.42	0.16

REFRACTIVE STATUS AND VISUAL ACUITY DATA																		
S/N	AGE	SEX	RX (RIGHT EYE)				RX (LEFT EYE)				VA (RIGHT EYE)			VA (LEFT EYE)			VA (BOTH EYE)		
			Sph	Cyl	Axis	N/E	Sph	Cyl	Axis	N/E	V1	V2	V3	V1	V2	V3	V1	V2	V3
130	12	F	-2.75	-3.00	160	-5.25	-4.50	-1.00	45	-5.00	0.20	0.40	0.20	0.22	0.40	0.18	0.28	0.50	0.22
131	10	F	+0.75	-0.50	170	+0.50	+1.00	-1.25	155	+0.50	0.46	0.66	0.20	0.50	0.70	0.20	0.42	0.74	0.32
132	10	F	+1.25	-1.50	30	+0.50	+0.50	-2.00	150	-0.50	0.40	0.70	0.30	0.40	0.60	0.20	0.38	0.66	0.28
133	10	F	+2.75	-3.00	110	+1.25	+2.75	-3.00	20	+1.25	0.24	0.30	0.06	0.28	0.30	0.02	0.30	0.30	0.00
134	13	F	-1.00	-0.50	170	-1.75	-1.00	-1.00	175	-1.50	0.36	0.60	0.24	0.42	0.64	0.22	0.44	0.66	0.22
135	12	M	-0.50	-2.75	35	-2.00	-0.50	-2.75	145	-2.00	0.40	0.50	0.10	0.40	0.50	0.10	0.40	0.50	0.10
136	8	M	-1.00	-1.50	110	-1.75	-1.25	-1.50	20	-2.00	0.10	0.20	0.10	0.10	0.24	0.14	0.10	0.20	0.10
137	12	M	-1.50	-0.50	90	-1.75	-1.00	-0.50	90	-1.25	0.36	0.46	0.10	0.34	0.46	0.12	0.34	0.48	0.14
138	7	M	-1.50	-0.50	90	-1.75	-1.50	-0.50	90	-1.75	0.14	0.16	0.02	0.20	0.20	0.00	0.20	0.20	0.00
139	10	F	-0.50	0.00	-	-0.50	-0.75	0.00	-	-0.75	0.56	0.60	0.00	0.30	0.50	0.20	0.56	0.64	0.08
140	11	M	-1.50	-0.50	90	-1.75	-2.00	0.00	-	-2.00	0.20	0.3	0.10	0.40	0.40	0.00	0.50	0.58	0.10
141	17	F	-5.00	-0.50	15	-5.25	-5.50	-0.50	175	-5.75	0.10	0.22	0.12	0.10	0.30	0.20	0.20	0.50	0.30
142	15	F	+0.75	-1.00	15	+0.25	+1.00	-0.75	165	+0.50	0.44	0.54	0.10	0.34	0.38	0.04	0.38	0.50	0.12

REFRACTIVE STATUS AND VISUAL ACUITY DATA																		
S/N	AGE	SEX	RX (RIGHT EYE)				RX (LEFT EYE)				VA (RIGHT EYE)			VA (LEFT EYE)			VA (BOTH EYE)		
			Sph	Cyl	Axis	N/E	Sph	Cyl	Axis	N/E	V1	V2	V3	V1	V2	V3	V1	V2	V3
143	15	F	-6.00	0.00	-	-6.00	-6.00	-1.00	145	-6.50	0.08	0.10	0.02	0.04	0.16	0.12	0.08	0.10	0.02
144	15	M	-1.25	-0.25	45	-1.25	-0.75	-1.00	45	-1.25	0.78	0.80	0.02	0.80	0.94	0.14	0.80	0.94	0.14
145	13	F	+0.75	-1.50	150	0	+0.75	-0.50	150	+0.50	0.09	0.10	0.10	0.50	0.50	0.00	0.74	0.8	0.04
146	10	F	-1.00	-1.00	45	-1.50	-1.00	0.00	-	-1.00	0.60	0.74	0.14	0.58	0.68	0.10	0.60	0.74	0.14
147	12	M	-2.50	-0.50	180	-2.75	-2.00	-0.50	180	-2.25	0.10	0.20	0.10	0.05	0.10	0.10	0.10	0.20	0.10
148	15	F	-0.25	-0.50	40	-0.50	-0.50	-0.50	170	-0.75	0.96	1.08	0.12	0.94	1.08	0.14	1.00	1.20	0.02
149	17	M	-1.25	-0.75	50	-1.50	-0.75	-1.00	150	-1.25	0.30	0.44	0.14	0.18	0.46	0.28	0.50	0.50	0.00
150	16	M	-0.75	-1.00	150	-1.25	-1.50	-0.50	35	-1.75	0.30	0.54	0.24	0.30	0.54	0.24	0.42	0.58	0.16
151	16	F	-1.00	-0.50	175	-1.25	-1.50	-0.75	40	-1.75	0.50	0.60	0.10	0.48	0.56	0.08	0.58	0.62	0.04
152	14	F	-1.75	-0.75	5	-2.00	-1.00	-0.50	170	-1.25	0.38	0.60	0.22	0.36	0.56	0.20	0.44	0.64	0.20
153	15	M	-1.50	-0.75	180	-2.00	-1.00	-0.75	180	-1.25	0.60	0.60	0.00	0.60	0.7	0.10	0.60	0.80	0.20

V1 = Uncorrected visual acuity.
V2 = corrected visual acuity.
V3= V1-V2: visual acuity improvement.

APPENDIX D 1: OCULAR ALIGNMENT , CONTRAST SENSITIVITY AND COLOUR VISION FOR CHILDREN WITH OCULOCUTANEOUS ALBINISM N= 153.

OCULAR ALIGNMENT, CONTRAST SENSITIVITY, STEREOACUITY AND COLOUR VISION DATA																
S/N	AGE	SEX	STRAB/PHORIA (Prism diopters)	CONTRAST SENSITIVITY (CPD/CONTRAST) WITHOUT Rx					WITH Rx					STEREO ACUITY (Sec/arc)	COLOUR VISION		
				1.5	3	6	12	18	1.5	3	6	12	18		ISHIHARA	PANEL D-15 (Saturated)	PANEL D-15 (Desaturated)
1	13	M	3 EsoT	3	1	0	1	0	3	2	2	1	1	200	P	P	P
2	12	M	4 ExoT	1	1	0	0	0	3	2	0	0	0	400	P	P	P
3	14	M	3 ExoT	1	1	0	0	0	1	1	1	0	0	400	P	P	P
4	13	F		2	1	1	0	0	4	1	1	0	0	200	P	P	P
5	14	F		1	1	0	0	0	3	1	1	0	0	200	P	P	P
6	13	M	6 EsoT	3	1	2	0	0	3	1	1	2	1	200	P	P	P
7	13	F	10 EsoT	1	0	1	0	0	1	2	3	0	0	200	P	P	P
8	13	F		2	1	0	0	0	4	5	1	0	0	200	P	P	P
9	12	F	3 ExoT	0	0	0	0	0	3	1	1	0	0	200	P	P	P
10	12	F		3	1	1	2	1	5	5	3	1	1	200	P	P	P
11	12	F		3	1	3	1	1	3	1	3	1	1	400	P	P	P
12	12	F		4	4	2	0	0	4	4	2	0	0	400	P	P	P
13	14	F	6 EsoP	2	1	1	0	0	3	4	1	1	0	400	P	P	P

OCULAR ALIGNMENT, CONTRAST SENSITIVITY, STEREOACUITY AND COLOUR VISION DATA																
S/N	AGE	SEX	STRAB/PHORIA (Prism diopters)	CONTRAST SENSITIVITY (CPD/CONTRAST) WITHOUT Rx					WITH Rx					STEREO ACUITY (Sec/arc)	COLOUR VISION		
				1.5	3	6	12	18	1.5	3	6	12	18		ISHIHARA	PANEL D-15 (Saturated)	PANEL D-15 (Desaturated)
14	12	F	6 EsoT	1	1	1	2	0	1	1	1	3	0	*	P	P	F
15	13	M		1	0	1	0	1	2	4	2	0	2	*	P	P	P
16	13	F		2	1	1	1	1	2	2	2	1	1	400	P	P	P
17	14	M	4 ExoT	3	3	3	2	0	3	3	3	2	0	*	P	P	P
18	13	M		3	3	2	0	0	3	3	2	0	0	200	P	P	F
19	11	F	6 ExoT	1	1	1	1	0	3	4	3	1	1	200	P	P	P
20	15	M	4 ExoT	2	1	0	0	0	3	1	0	1	0	*	P	P	P
21	13	F	4 EsoT	1	1	1	0	0	1	4	4	2	1	400	P	P	P
22	11	F		5	5	1	1	1	5	5	2	1	1	200	P	P	P
23	14	M		1	2	3	2	0	1	2	3	1	1	400	P	P	F
24	12	M		0	0	1	0	0	0	0	1	0	0	200	P	P	P
25	11	F		5	1	0	1	1	5	1	2	1	1	200	P	P	P
26	11	F		1	1	0	0	0	1	1	0	0	0	200	P	P	P

OCULAR ALIGNMENT, CONTRAST SENSITIVITY, STEREOACUITY AND COLOUR VISION DATA																
S/N	AGE	SEX	STRAB/PHORIA (Prism diopters)	CONTRAST SENSITIVITY (CPD/CONTRAST) WITHOUT Rx					WITH Rx					STEREO ACUITY (Sec/arc)	COLOUR VISION		
				1.5	3	6	12	18	1.5	3	6	12	18		ISHIHARA	PANEL D-15 (Saturated)	PANEL D-15 (Desaturated)
27	12	F		2	2	1	1	0	5	5	3	1	1	200	P	P	P
28	11	F		4	1	1	1	0	4	4	1	1	0	200	P	P	P
29	12	F		3	1	2	1	0	5	5	2	1	1	200	P	P	P
30	14	M		1	4	4	1	1	1	4	4	2	1	400	P	P	P
31	12	M		5	4	0	0	0	5	4	1	0	0	500	P	P	P
32	11	M		3	1	1	1	1	4	4	3	3	2	200	P	P	P
33	12	M		3	1	1	1	1	3	4	2	1	0	400	P	P	P
34	13	F		1	1	1	1	1	3	2	2	1	1	200	P	P	P
35	10	F	6 EsoT	1	1	1	1	1	1	2	2	1	1	200	P	P	P
36	14	M	18 EsoT	0	0	0	0	0	4	2	0	0	0	500	F	F	F
37	10	F		5	3	1	3	1	5	2	3	3	2	200	P	P	P
38	12	F		1	4	1	1	0	4	3	4	3	3	200	P	P	P
39	13	M	2 EsoT	1	1	1	1	1	5	5	3	4	1	200	P	P	P

OCULAR ALIGNMENT, CONTRAST SENSITIVITY, STEREOACUITY AND COLOUR VISION DATA																
S/N	AGE	SEX	STRAB/PHORIA (Prism diopters)	CONTRAST SENSITIVITY (CPD/CONTRAST) WITHOUT Rx					WITH Rx					STEREO ACUITY (Sec/arc)	COLOUR VISION		
				1.5	3	6	12	18	1.5	3	6	12	18		ISHIHARA	PANEL D-15 (Saturated)	PANEL D-15 (Desaturated)
40	13	F	6 EsoT	1	1	1	1	1	3	1	2	1	1	200	P	P	F
41	11	F	14 EsoT	1	1	1	1	1	1	4	4	1	1	200	P	P	P
42	11	F		5	2	1	1	1	2	4	3	2	1	200	P	P	P
43	11	F		1	1	1	1	1	1	3	3	1	1	200	P	P	P
44	10	F	11 EsoT	3	2	1	1	1	5	5	5	4	4	200	P	P	P
45	10	F	30 EsoT	3	1	1	1	1	5	5	4	4	1	200	P	P	P
46	10	F	3 ExoT	1	1	1	1	1	3	4	3	2	1	200	P	P	P
47	10	F	20 ExoT	5	5	2	1	1	3	3	1	3	1	200	P	P	P
48	11	M		5	5	3	1	1	3	5	3	3	1	200	P	P	P
49	10	M	25 EsoT	3	3	1	1	1	4	5	3	1	0	200	P	P	P
50	14	M		1	3	1	1	0	5	1	1	1	0	400	P	P	P
51	10	M		5	5	5	1	1	5	5	3	3	1	200	P	P	P
52	9	F	18 EsoT	3	1	1	1	1	3	1	3	1	1	200	P	P	P

OCULAR ALIGNMENT, CONTRAST SENSITIVITY, STEREOACUITY AND COLOUR VISION DATA																
S/N	AGE	SEX	STRAB/PHORIA (Prism diopters)	CONTRAST SENSITIVITY (CPD/CONTRAST) WITHOUT Rx					WITH Rx					STEREO ACUITY (Sec/arc)	COLOUR VISION		
				1.5	3	6	12	18	1.5	3	6	12	18		ISHIHARA	PANEL D-15 (Saturated)	PANEL D-15 (Desaturated)
53	11	F		5	5	2	1	1	5	5	1	1	1	400	P	P	P
54	8	F		4	1	1	1	0	3	1	1	1	0	400	P	P	P
55	11	F		1	1	1	1	0	2	2	1	1	1	400	P	P	P
56	11	F	4 EsoT	1	1	1	0	0	4	5	2	1	1	400			
57	11	M		3	1	1	1	1	3	5	1	1	1	400	P	P	P
58	10	M		3	4	1	1	1	4	5	4	1	1	400	P	P	P
59	10	M	14 EsoT	5	5	1	1	1	3	5	5	1	2	70	P	P	P
60	14	M	14 EsoT	4	3	1	2	0	4	5	1	2	0	400	F	P	P
61	9	M		3	1		1	1	1	1	1	1	1	200	P	P	P
62	8	M	12 EsoT	5	1		1	1	3	3	1	1	1	200	P	P	P
63	11	F		3	5		1	1	3	3	5	1	1	200	P	P	P
64	9	M		3	1		2	1	3	3	1	1	1	400	P	P	P
65	9	M	6 ExoT	3	1		1	1	1	3	1	1	1	200	P	P	P

OCULAR ALIGNMENT, CONTRAST SENSITIVITY, STEREOACUITY AND COLOUR VISION DATA																
S/N	AGE	SEX	STRAB/PHORIA (Prism diopters)	CONTRAST SENSITIVITY (CPD/CONTRAST) WITHOUT Rx					WITH Rx					STEREO ACUITY (Sec/arc)	COLOUR VISION		
				1.5	3	6	12	18	1.5	3	6	12	18		ISHIHARA	PANEL D-15 (Saturated)	PANEL D-15 (Desaturated)
67	8	M		3	1	2	1	1	3	1	1	3	1	200	P	P	P
68	9	M		1	0	0	0	0	1	1	2	1	1	200	F	P	P
69	10	M		3	1	1	1	1	4	3	3	3	2	200	P	P	P
70	9	M		3	3	2	1	1	3	3	1	2	2	200	P	P	P
71	9	F		3	2	1	1	1	3	1	1	2	1	140	P	P	P
72	9	F		1	1	0	0	0	1	1	1	1	0	*			
73	9	F		1	1	0	0	0	1	1	1	1	0	400	F	F	F
74	9	M	10 EsoT	1	0	0	0	0	3	1	1	1	0	400	F	F	F
75	7	M	4 EsoT	1	1	0	0	0	3	1	1	1	1	200	F	P	P
76	11	M	20 EsoT	3	1	1	0	0	3	1	1	1	1	400	F	P	P
77	9	M	4 EsoT	0	0	0	0	0	3	1	1	1	0	400	F	P	P
78	8	M		3	2	1	1	1	3	1	1	1	0	200	P	P	P
79	8	F		3	4	0	0	0	2	3	1	2	1	200	F	P	P

OCULAR ALIGNMENT, CONTRAST SENSITIVITY, STEREOACUITY AND COLOUR VISION DATA																
S/N	AGE	SEX	STRAB/PHORIA (Prism diopters)	CONTRAST SENSITIVITY (CPD/CONTRAST) WITHOUT Rx					WITH Rx					STEREO ACUITY (Sec/arc)	COLOUR VISION		
				1.5	3	6	12	18	1.5	3	6	12	18		ISHIHARA	PANEL D-15 (Saturated)	PANEL D-15 (Desaturated)
80	7	F		1	0	0	0	0	1	1	0	0	0	500	P	P	P
81	8	M		3	1	1	1	0	5	3	3	2	0	200	F	F	P
82	8	F		3	1	1	0	0	3	1	1	1	0	400	F	F	F
83	9	F	8 ExoT	5	1	1	1	0	3	1	1	1	1	200	P	P	P
84	8	F	4 EsoT	4	3	0	0	0	4	3	2	1	1	200	P	P	P
85	8	F	12 EsoT	0	0	0	0	0	1	1	0	0	0	*	F	F	P
86	10	M	4 ExoT	3	1	0	0	0	3	2	1	0	0	200	P	P	P
87	11	M	8 ExoT	0	0	0	0	0	3	1	1	1	0	200	F	P	F
88	8	M		0	0	0	0	0	1	1	1	0	0	140	F	F	F
89	9	F		1	1	0	0	0	3	1	1	1	0	200	P	P	P
90	9	M	10 EsoT	1	1	0	0	0	3	1	0	0	0	200	P	P	P
91	8	F		5	4	1	1	0	5	5	3	2	1	200	P	P	P
92	8	M	13 EsoT	1	2	2	1	0	2	3	3	1	0	200	F	F	F

OCULAR ALIGNMENT, CONTRAST SENSITIVITY, STEREOACUITY AND COLOUR VISION DATA																
S/N	AGE	SEX	STRAB/PHORIA (Prism diopters)	CONTRAST SENSITIVITY (CPD/CONTRAST) WITHOUT Rx					WITH Rx					STEREO ACUITY (Sec/arc)	COLOUR VISION		
				1.5	3	6	12	18	1.5	3	6	12	18		ISHIHARA	PANEL D-15 (Saturated)	PANEL D-15 (Desaturated)
93	8	M	2 ExoT	0	0	0	0	0	0	0	0	0	0	200	P	P	F
94	8	F		0	0	0	0	0	1	1	1	0	0	400	P	P	P
95	7	M	4 ExoT	3	3	2	0	0	5	3	3	1	1	*	P	P	P
96	7	F	8 ExoT	4	1	1	1	1	5	2	2	1	1	500	P	P	P
97	8	M		2	1	1	1	1	2	1	1	1	1	400	P	P	P
98	7	F		2	1	1	1	2	2	1	1	2	2	200	P	P	P
99	7	F		5	2	2	0	0	1	4	2	3	1	200	F	P	P
100	7	M	3 EsoT	5	3	0	0	0	4	1	1	1	1	400	F	P	P
101	7	M		3	1	1	0	0	3	3	0	0	0	*	P	P	P
102	7	F		5	4	2	2	0	3	5	1	1	0	200	P	P	P
103	8	M	4 ExoT	1	2	2	1	0	5	6	3	3	1	400	F	P	P
104	7	F		1	2	1	1	1	3	4	1	1	1	140	P	P	P
105	8	M		5	1	2	1	1	3	1	1	1	1	200	P	P	P

OCULAR ALIGNMENT, CONTRAST SENSITIVITY, STEREOACUITY AND COLOUR VISION DATA																
S/N	AGE	SEX	STRAB/PHORIA (Prism diopters)	CONTRAST SENSITIVITY (CPD/CONTRAST) WITHOUT Rx					WITH Rx					STEREO ACUITY (Sec/arc)	COLOUR VISION		
				1.5	3	6	12	18	1.5	3	6	12	18		ISHIHARA	PANEL D-15 (Saturated)	PANEL D-15 (Desaturated)
106	8	M		1	0	0	0	0	3	1	1	1	1	500	F	P	P
107	7	M	16 EsoT	3	2	2	1	1	2	1	1	1	2	400	P	P	P
108	16	M	4 ExoT	2	2	2	1	1	4	4	1	3	2	400	P	P	P
109	8	M		1	1	1	1	1	1	1	1	0	0	200	P	P	P
110	9	M		2	1	1	1	0	2	1	1	1	0	500	P	P	P
111	7	F		2	4	1	0	0	3	2	1	1	0	200	F	P	F
112	9	F		4	4	3	2	1	5	5	3	2	1	500	F	P	P
113	13	M		4	4	2	1	1	4	3	3	2	1	400	F	P	F
114	8	M		3	4	2	0	0	2	2	1	1	0	*	F	P	P
115	14	F		3	3	0	0	0	1	1	1	1	1	500	P	P	P
116	9	F		3	2	2	2	1	2	2	2	1	1	200	P	P	P
117	8	F		3	2	3	0	0	2	2	1	1	1	500	P	P	P
118	14	F	8 EsoT	3	5	3	0	0	2	2	2	2	1	400	P	P	P

OCULAR ALIGNMENT, CONTRAST SENSITIVITY, STEREOACUITY AND COLOUR VISION DATA																
S/N	AGE	SEX	STRAB/PHORIA (Prism diopters)	CONTRAST SENSITIVITY (CPD/CONTRAST) WITHOUT Rx					WITH Rx					STEREO ACUITY (Sec/arc)	COLOUR VISION		
				1.5	3	6	12	18	1.5	3	6	12	18		ISHIHARA	PANEL D-15 (Saturated)	PANEL D-15 (Desaturated)
119	14	F	8 EsoT	3	5	3	0	0	2	2	2	2	1	400	P	P	P
120	12	M		3	2	3	1	1	4	2	2	2	1	400	P	P	F
121	17	M	6 ExoT	1	1	0	0	0	1	1	1	0	0	400	P	P	P
122	12	F		4	4	0	0	0	3	1	1	0	0	*	P	P	P
123	12	M		4	4	3	3	4	5	5	4	2	0	200	P	P	P
124	12	M		1	1	1	1	0	1	1	1	1	1	200	P	P	P
125	10	M		5	4	3	0	0	4	3	1	0	0	500	P	F	F
126	10	F		1	1	0	0	0	2	2	2	1	0	400	P	P	P
127	9	F		1	1	1	0	0	2	3	2	2	0	*	P	F	F
128	10	M		5	5	3	2	1	4	3	1	1	0	400	P	P	P
129	12	F		2	2	0	0	0	2	2	1	0	0	*	P	F	F
130	12	F		1	1	1	0	0	1	1	1	0	0	250	F	F	F

OCULAR ALIGNMENT, CONTRAST SENSITIVITY, STEREOACUITY AND COLOUR VISION DATA																
S/N	AGE	SEX	STRAB/PHORIA (Prism diopters)	CONTRAST SENSITIVITY (CPD/CONTRAST) WITHOUT Rx					WITH Rx					STEREO ACUITY (Sec/arc)	COLOUR VISION		
				1.5	3	6	12	18	1.5	3	6	12	18		ISHIHARA	PANEL D-15 (Saturated)	PANEL D-15 (Desaturated)
131	10	F		4	4	1	1	1	2	2	2	2	1	200	F	P	P
132	10	F		4	3	0	0	0	3	1	2	0	0	400	F	F	F
133	10	F		0	0	0	0	0	1	1	1	0	0	400	P	F	F
134	13	F		2	1	0	0	0	2	3	1	0	0	400	P	P	P
135	12	M		3	2	1	0	0	3	1	1	1	1	200	P	P	P
136	8	M		2	2	1	1	1	2	2	1	1	1	400	P	P	P
137	12	M		4	3	4	0	0	4	4	3	0	0	400	P	P	F
138	7	M		1	1	0	0	0	1	1	1	0	0	400	P	P	P
139	10	F		4	1	0	0	0	4	4	2	2	1	*	P	P	P
140	11	M		2	1	0	0	0	2	2	1	1	1	400	P	P	P
141	17	F		3	2	1	1	1	2	2	1	1	1	400	P	F	F
142	15	F		4	3	2	1	0	4	4	3	2	1	400	P	P	P
143	15	F		0	0	0	0	0	1	1	0	0	0	400	P	F	F

OCULAR ALIGNMENT, CONTRAST SENSITIVITY, STEREOACUITY AND COLOUR VISION DATA																
S/N	AGE	SEX	STRAB/ PHORIA (Prism diopters)	CONTRAST SENSITIVITY (CPD/CONTRAST) WITHOUT Rx					WITH Rx					STEREO ACUITY (Sec/arc)	COLOUR VISION		
				1.5	3	6	12	18	1.5	3	6	12	18		ISHIHARA	PANEL D-15 (Saturated)	PANEL D-15 (Desaturated)
144	15	M		4	4	2	2	0	4	4	3	3	2	*	P	P	P
145	13	F		2	1	1	0	0	2	2	1	0	0	200	F	P	F
146	10	F		4	3	1	0	0	4	3	1	0	0	400	P	F	F
147	12	M		1	3	2	2	1	3	3	2	2	0	400	P	P	P
148	15	F		5	5	2	1	0	5	5	3	3	1	250	P	P	P
149	17	M		1	1	1	0	0	4	3	1	0	0	*	P	P	P
150	16	M		3	2	0	0	0	4	4	0	0	0	400	P	P	P
151	16	F	4 EsoP	4	4	3	2	1	4	4	3	2	1	400	P	P	P
152	14	F		4	4	2	1	1	3	3	3	2	1	250	P	P	F
153	15	M	4 ExoT	2	2	0	0	0	2	2	0	0	0	*	P	P	P

M= Male
F = Female
*** Indicates that a minimum stereoacuity of 500 seconds of arc could not be perceived (the subject did not demonstrated stereoacuity with the Randot stereotest).**

APPENDIX D 2. SHOWING THE SUBJECTS WHO FAILED ONE OR MORE OF THE COLOUR VISION TESTS

SN	INITIALS/ (SEX)	TESTS		
		ISHIHARA PLATES	FARNSWORTH D- 15 SATURATED	FARNSWORTH D-15 DESATURATED
1	IB (F)	Pass	Pass	Fail mild tritanope
2	VM (F)	Pass	Pass	Fail Mixed variables
3	MM (M)	Fail mild deutanope	Pass	Fail mild deutanope
4	MA (M)	Fail strong protanope	Fail mild protanope	Fail mild protanope
5	RM (M)	Fail mild protanope	Pass	Pass
6	MM (F)	Fail strong deutanope	Fail achromat	Fail achromat
7	MP (M)	Fail achromat	Fail achromat	Fail achromat
8	MR (M)	Fail mild deutanope	Pass	Pass
9	NC (M)	Fail strong deutanope	Pass	Pass
10	FM (F)	Fail mild deutanope	Fail mixed variable	Fail mixed variables
11	ML (F)	Fail mild protanope	Pass	Pass
12	MD (M)	Pass	Pass	Fail mild tritanope
13	MM (M)	Fail mild deutanope	Pass	Pass
14	MT (M)	Fail strong deutanope	Pass	Pass

15	NT (M)	Fail mild protanope	Pass	Pass
16	MN (M)	Fail mild deutanope	Pass	Fail mixed variables
17	RM (M)	Pass	Pass	Fail mild tritanope
18	DR (M)	Fail mild deutanope	Fail mild deutanope	Fail mild deutanope
19	MK (M)	Pass	Fail mild tritanope	Fail mild tritanope
20	NM (F)	Pass	Fail mild tritanope	Fail mild tritanope
21	MM (F)	Fail mild protanope	Fail mild protanope	Fail mild protanope
22	MM (F)	Fail mild protanope	Fail mild protanope	Fail mild protanope
23	ML (F)	Pass	Fail mild tritanope	Fail mild tritanope
24	EM (M)	Pass	Fail mixed variables	Fail mixed variables
25	KS (F)	Pass	Fail Mixed variables	Fail mild protanope
26	BK (F)	Pass	Pass	Fail Mixed variables