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Research Article

Oculo-visual Functions in Patients Living with Sickle Cell Disease in Kano

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Abstract

Sickle Cell Disease (SCD) is one of the most common life-threatening, single-gene inherited blood disorders in the world that leads to higher risk of early mortality. The increase in life expectancy of the SCD patients in recent years has led to the emergence of more complications of the disease, e.g. ocular complications. This increasing complication has prompted this study in order to discover the possible effects of SCD on oculo-visual functions. This study assessed the oculo-visual functions of patients living with sickle cell disease within Kano, in order to determine the possible effects of SCD on their oculo-visual functions. All patients aged from 7 years and above who reported for routine follow up in the adult and paediatric sickle cell clinics in Murtala Muhammad Specialist Hospital (MMSH) and Muhammad Abdullahi Wase Hospital (MAWH) during the period of this study were examined. A verbal interview was done to obtain the demographic data of the subjects. Visual functions testings and ocular examination were performed by the use of Snellen's distance and near acuity charts, Amsler grid, Ishihara chart, MARS contrast sensitivity chart, direct ophthalmoscope and air-puff non-contact tonometer. The mean age and standard deviation of the participants were 20 years (cl: 17-26) and SD (10.02) respectively. Decreased visual acuity was found in 43.5%, visual impairment in 5.5% of 200 eyes of 100 participants. Contrast sensitivity loss in 84.5%. Visual field defects in 9.5%. Cataract in 2.5%, non-proliferative and proliferative sickle cell retinopathy findings in 11.0% and 0.5% respectively, whereas intraocular pressure and colour vision sensitivity were normal in all the 200 eyes. Ocular findings and oculo-visual function disorders occur in patients living with sickle cell disease in Kano with the exception of colour sensitivity and intraocular pressure that show normal results. These disorders occur in both males and females and also across all ages.

Keywords: Sickle Cell Disease (SCD); Non-Proliferative Sickle Cell Retinopathy; Proliferative Sickle Cell Retinopathy; Visual Impairment; Oculo-Visual Functions; Single-Gene Inherited Blood Disorders

Acronyms and Abbreviations

AHW: Abdullahi Wase Hospital; AKTH: Aminu Kano Teaching Hospital; BCVA: Best Corrected Visual Acuity; BTM: Beta-thalassemia Major; CS: Contrast Sensitivity; Hb: Hemoglobin; HbSC: Heterozygous; HbSS: Homozygous; IOP: Intraocular Pressure; NO: Nitric Oxide; MMSH: Murtala Muhammad Speacialist Hospital; OCT: Optical Coherance Tomography; PH: Potent Hydrogen; PSR: Proliferative Sickle Cell Retinipathy; SCA: Sickle Cell Anemia; SCR: Sickle Cell Retinopathy; SD: Standard Deviation; SPSS: Statistical Package for the Social Science; VA: Visual Acuity; WHO: World Health Organization

Introduction

Sickle Cell Disease (SCD) is one of the most common life-threatening, single-gene inherited blood disorders in the world that leads to higher risk of early mortality. The term 'sickle cell disease' is used to refer to all entities in which there is polymerization of hemoglobin (Hb) within red cells with basic clinico-pathological features, i.e. presence of sickle red cells in the blood and clinical illness caused by abnormal erythrocytes. These abnormal erythrocytes adhere to the blood vessel wall (endothelium) and produce the manifestations of the disease through vital interactions with leucocytes, platelets, and various plasma factors such as cytokines

and coagulation proteins. This condition predisposes individuals to multi-organ acute and chronic complications linked with significant morbidity and mortality. Some ocular complications of sickle cell disease are retinal changes resulting from occlusion of retinal vessels especially in temporal periphery (SCD retinopathy), refractive errors, vitreous hemorrhage due to vaso-occlusion which can result to transient visual impairment or retinal detachment with permanent blindness, and corneal abnormalities.

The average life-expectancy for people with SCD is estimated to be between 42 and 48 years among which approximately 85% survives for at least 20 years of age.

Background of the study

Sickle cell disease (SCD) is an autosomal recessive haemoglobinopathy occurring due to a defect in the beta chain of the haemoglobin molecule. Its effects on the eye have been reported in the adnexa, anterior and posterior segments.

Sickling haemoglobinopathies are caused by one or more abnormal haemoglobins that induce red blood cells to adopt an anomalous shape under conditions of physiological stress such as hypoxia and acidosis, with resultant vascular occlusion. Sickle cell anemia, the prototypical (and most prevalent) hemoglobinopathy is one of a group of hemoglobinopathies which are hereditary disorders caused by inherited mutations that lead to structural abnormalities in hemoglobin, stems from a mutation in the β-globin gene that creates sickle shaped hemoglobin. (Bruce Muchnick P334, 2008) [15].

Sickle cell disease (SCD) was first described in Africa under a variety of names that related to cultural and spiritual beliefs. James B. Herrick was an American physician who made the first clinical discovery and reported in 1910 'peculiar elongated and sickle-shaped red blood corpuscles in a case of severe anemia' affecting a medical student from Grenada, initially the disease was known as Herrick's syndrome and it was not until 1922 when a fourth case reported by a medical student, Verne R. Mason, from Johns Hopkins Hospital that the term 'sickle cell anemia (SCA)' was first used (Tamer Hassan., *et al*. Medicine (Baltimore) 2021. 13/01/2022) [27].

Normal hemoglobins are tetramers composed of two pairs of similar chains. On average, the normal adult red cell contains 96% HbA (α2β2), 3% HbA2 (α2δ2), and 1% fetal Hb (HbF, α2γ2). HbS is produced by the substitution of valine for glutamic acid at the sixth amino acid residue of β-globin. In homozygotes (HbSS), all HbA is replaced by HbS, whereas in heterozygotes (HbSC), only about half is replaced. This sickle cell type hemoglobin causes deformation of erythrocyte under conditions of decreased oxygen tension (Bruce Muchnick P334, 2008) [15].

Sickle cell disease causes red blood cells to become sticky and rigid. Sickle cells can block blood flow in small blood vessels depriving the eye of oxygen and cause damage. This is called sickle retinopathy that can progress to severe proliferative sickle cell retinopathy, bleeding into the eye, detachment of the retina or even loss of vision (Tamer Hassan., *et al*. Medicine (Baltimore). 2021. 13/01/2022) [27].

The increase in life expectancy of the SCD patients in recent years has led to the emergence of more complications of the disease, e.g. ocular, which in the past were uncommon. Sickle cell disease can affect virtually every vascular bed in the eye and can cause blindness in the advanced stages. (A. O Fadugbagbe., *et al*. Ann Top paediatr. 2010. 13/01/2022) [6].

Anatomic alterations in sickle cell anemia stem from (1) the severe chronic hemolytic anemia, (2) the increased breakdown of heme to bilirubin, and (3) microvascular obstructions, which provoke tissue ischemia and infarction.

Clinical course

Homozygous sickle cell disease usually is asymptomatic until 6 months of age when the shift from HbF to HbS is complete. The anemia is moderate to severe; most patients have hematocrits 18% to 30% (normal range, 36% to 48%). The chronic hemolysis is associated with hyperbilirubinemia and compensatory reticulocytosis. The most serious of these are the vaso-occlusive, or pain crises, acute chest syndrome and stroke, which sometimes occurs in the setting of the acute chest syndrome. Although virtually any organ can be damaged by ischemic injury, the acute chest syndrome and stroke are the two leading causes of ischemia-related death. A second acute event, aplastic crisis, is caused by a sudden decrease in red cell production. As in hereditary spherocytosis, this usually is triggered by the infection of erythroblasts by parvovirus B19.

Clinical manifestations of SCD are also related to complex metabolic pathways that include endothelial actions, inflammation, nitric oxide (NO) bioavailability, oxidative stress, and regulation of the adhesiveness of several types of blood cells.

Incidence/epidemiology

SCD occurs in regions of the world where malaria is or was previously common. Individuals in these areas have a fitness benefit in carrying only a single sickle-cell gene (sickle cell trait). These areas include: Africa; Mediterranean Europe; Middle East; Saudi Arabia; some regions of India; the Caribbean; South and Central America.

In United State of America, SCD affects > 70,000 African-American and 1 in 375 new-born. In the United Kingdom, it affects 1 in 2,400 live births across ethnic groups, and >12,000 individuals are living with SCD.

In Africa, the HbS gene frequency is 10-30% and about 200,000 new cases of sickle cell disease occur each year. The incidence of SCD in other parts of the world is increasing due to population migration.

An overall sickling prevalence of 10.4% was found in over 4,000 selected subjects in a study done in Lusaka, Zambia. However, this varied from 4.5% to 16.8% in different tribal groups. Nigeria being the epicenter zone of SCD has about 4-6 million people living with the disease (1 in every 4 Nigerians has sickle cell trait). Annually, about 300,000 newly diagnosed SCD children are born worldwide. Sub-Saharan Africa contributes about 75% of the number, and accounts for 100,000-150,000 newborns living with SCD annually (33% of the global burden of SCD). Therefore, Nigeria occupies a strategic position in the epidemiology of SCD from the global perspective. Specifically, the HbS gene has a prevalence of 24-25% in Nigerians and only the homozygous SCD (Hb SS) and Hb SC are known to cause significant clinical problems in Nigeria.

The city of Kano has a large cohort of SCD patients. The SCD patients' population is about 1,570 at the outpatient clinics of AKTH, of which 1,300 are paediatric patients. AKTH receives regular SCD referrals from Murtala Muhammad Specialist Hospital, Kano, which is a secondary health facility with about 11,000 registered SCD patients. The prevalence of SCD in Kano is as high as 40.3%.

Earlier done in a 1969 survey of some 10,000 blood samples found 187 cases diagnosed with HbSS, most of them in infants. However, no haemoglobin C and no beta-thalassaemia were found. Haemoglobin H disease and other evidence for alpha-thalassaemia were found. Patients with SCD are now living much longer than in the past. Deaths occurring due to SCD globally were previously very high. From the 1960s, multiple researchers have reported an almost total absence of HbSS among samples of African adults.

A study done by Barckley in a Zambian mining town between 1969-1971 showed excess mortality rates of 60% by age 12 years. However in the recent past, some studies are suggesting that survival might be improving. For example, in a study conducted in Lusaka, Zambia, Athale and Chintu reported that the case-fatality rate among children with HbSS who were admitted to the University Teaching Hospital decreased from 18.6% in 1970 to 6.6% during 1987-1989. A WHO progress report noted a decline in under-5 mortality in sub-Saharan Africa by 28% since 1990.

Homozygotes have sickle cell anemia (all hemoglobin is HbS), and heterozygotes have sickle cell trait (only 50% of hemoglobin is HbS); 8% of Africans have sickle cell trait because of the protective effect conferred by the mutation versus malaria infection, and 0.2% of Africans have sickle cell disease (Principles Of Pathology P196).

Oculo-visual functions

The functions of the eye and its visual system to maintain the integrity and rigidity of the ocular structures as well as the shape and alignment to provide information and or perception (vision) of objects within the environment or surrounding and the real world, including perception of shape, colour, form and size helping individuals to perform well while interacting with the visual environment (functional vision).

Visual acuity

Visual acuity is defined as the "spatial resolving capacity of the visual system" (Benjamin, 2006) and refers to the sharpness of vision or the patient's ability to recognize a minimum size target. Visual acuity is customarily abbreviated as 'VA'. The measurement of visual acuity should be performed at every visit on completion of a case history. It is one of the most informative tests conducted. Visual acuity provides information on; Refractive status of the eye, indication of macula function, and indication of neural integrity.

Intrao cular pressure

Tonometry/Intraocular pressure measurement is a clinical technique that provides a measurement of the internal pressure of the eye (also known as intraocular pressure, IOP, ocular tension). Technically speaking, the value for a given IOP represents the combined resistance of the different layers of the eye and the intraocular pressure itself. Sickling of HbS cells is favoured by the relatively deoxygenated conditions in the anterior chamber. The deformed, sickled and pliable cells are unable to negotiate the tra-

becular meshwork leading to increased intraocular pressure. SCD patients with hyphaema present an important ocular emergency, which usually occurs as a result of trauma or surgery. Red blood cells tend to sickle and obstruct the flow of aqueous humour as a result of low PH and partial pressure of oxygen.

Individuals with sickle cell disease are prone to neovascular glaucoma. Ischaermia occurring due to blood vessels obstruction by sickled red cells may lead to neovascularization in trabecular meshwork and the iris. This can eventually lead to anterior chamber outflow tract obstruction causing raised intraocular pressure.

Colour vision

Is the visual function that allows one to perceive variations among physical wavelengths of light that compile the visible spectrum, Persons with normal colour vision are termed trichromats. Trichromats have three divisions of cone photoreceptors within the retina which contain photopigments to absorb the wavelengths of light within the visible spectrum.

Erythrolabe refers to the photopigment that absorbs red wavelengths.

Chlorolabe refers to the photopigment that absorbs green wavelengths.

Cyanolabe refers to the photopigment that absorbs blue wavelengths

Contrast sensitivity

While the more familiar visual acuity test assesses resolution of the eye and visual system and the processing of high retinal image spatial frequencies, it measures an extensity threshold, i.e. the minimum size target that can be resolved or recognized while contrast sensitivity test instead assesses processing of relatively low retinal image spatial frequencies, it measures intensity threshold i.e. the minimum luminance difference that can be seen.

Contrast sensitivity can be diminished by a host of retinal disorders such as glaucoma, age related macular degeneration, by ocular media opacities and other optical disorders (refractive error) or visual pathway diseases like optic neuritis and retrobulbar optic neuritis and others eg amblyopia often with minimal or no diminution of visual acuity.

Contrast sensitivity is the patient's ability to differentiate between light and dark stimulus by assessing the perception of black on white.

Contrast Sensitivity (CS) can also be defined as the ability to perceive sharp and clear outlines of very small objects and its use to identify minute differences in the shadings and patterns. CS helps detect objects without a clear outline and distinguish them from their background contrast. Contrast sensitivity function is usually performed when the integrity of the visual pathway is questionable or when the media opacity is present. Contrast sensitivity function provides useful information about functional or real-world vision that is not provided by visual acuity, including the likelihood of falling, control of balance, driving, motor vehicle crash involvement, reading, activities of daily living and perceived visual disability and therefore contrast sensitivity should be included with visual acuity and visual fields in the definitions of visual impairment and visual disability and for legal definitions of blindness. In addition, it can provide more sensitive measurements of subtle vision loss than visual acuity. Contrast Sensitivity test can be used for establishing baseline contrast sensitivity prior to an intervention (such as cataract extraction), for identifying functional losses in low contrast perception (often associated with glare sensitivity), or for functionally monitoring disease progression.

Visual field

According to Benjamin (1998) in Borish's Clinical Refraction, the visual field (VF) is that" area of space that a person can see at one time". Even though we function binocularly in most circumstances, the clinical testing of the visual field is rarely conducted binocularly. The monocular visual field is 3-dimensional and known as the "Hill of Vision". The outer edges of which represent the outermost limits of the area in space, termed the visual field, that can be seen at any one time, any target irrespective of its size or intensity, cannot be seen beyond this area. The outer or absolute limits of the monocular visual field are: superior: 55-60 degrees, inferior 70 degrees, temporal 100 degrees and nasal 60 degrees. The shape of the visual field is therefore that of horizontal oval.

Statement of the problem

Sickle cell disease is the most common genetic disease worldwide (A O Fadugbagbe., et al. Ann Top Paediatr. 2010. 13/01/2022) [6]. The increase in life expectancy of the SCD patients in recent years has led to the emergence of more complications of the disease, e.g. ocular, which in the past were uncommon. Sickle cell disease can affect virtually every vascular bed in the eye and can cause blindness in the advanced stages. This increasing complications has prompted this study in order to discover the possible effects of SCD on oculo-visual functions.

Justification

Although prevalence of sickle cell disease is high in Kano state, there is no study done to determine ocular manifestations and oculo-visual functions The World Health Organization (WHO) at its 56th session in 2006 recommended that SCD be acknowledge as a public health issue and emphasized the urgent need to establish simple cost effective strategies to reduce the morbidity and mortality associated with SCD in countries most affected [16].

A number of interventions have thus been promoted, such as public health programs including: new-born screening, health education, and immunization, this has resulted in increased life –expectancy. Hence, more patients with SCD are living into adulthood. Mortality attributable to SCD has decreased and the mean age of death is increasing.

The increase in life–expectancy may lead to increased number of patients with visual impairment. As a result of this, eye care providers will be faced with a burden of a unique challenge of managing these patients.

The information obtained will thus be useful in highlighting oculo-visual function features of sickle cell disease patients in Kano, Nigeria. Therefore, this will assist in setting up of an appropriate screening protocol and follow up. It will also foster a multidisciplinary approach in the management of sickle cell disease patients involving pediatricians, physicians, ophthalmic practitioners and other allied health workers.

Scope of the study

This study was conducted at Murtala Muhammad Specialist Hospital (MMSH), and Muhammad Abdullahi Wase (Nassarawa) Hospital in Kano, Kano is a city in northern Nigeria and the capital of Kano State. Kano State is the second largest city in the country (Nigeria) and has the population close to 15,076,892 million people and occupies a total area of 20,131 kilometer square, and has the latitude and longitude coordinates of 12.000000, 8.516667. Kano State divided into three zones/ districts namely Kano Sourth, Kano Central and Kano North; it consists of 44 local governments and is bounded by Katsina State to the northwest, Jigawa State to the northeast, Bauchi State to the southeast, and Kaduna State to the southwest.

Aim

The study aims at assessing the oculo-visual functions of patients living with sickle cell disease within Kano, in order to determine the possible effects of SCD on their oculo-visual functions.

Objectives

To assess the ocular health and test visual functions of patients living with SCD.

To determine the common oculo-visual function disorders in people living with SCD.

To ascertain the effects of SCD on oculo-visual function.

Significance

The data obtained will be a very useful tool for the eye care practitioners in the management of people living with SCD.

The result will let the general physicians to know about the ocular problems SCD patients may have and therefore refer them as early as possible whenever they report to them with any ocular symptoms.

It will also help the SCD patients and their parents or gurdians know the effect, the condition has on their eyes and try to visit eye care professionals for routine checkup.

Methodology

Study design

This study is a hospital based prospective cross sectional study.

Ethical clearance

Ethical clearance was collected from Kano State Ministry for health.

Approval

Approval was obtained from the ethics committee of each of the study locations and, oral and written consent of the primary care givers of subjects were obtained and patient informed consent forms were given and signed by either the subjects or their parents or gurdians.

Study population

100 Sickle cell disease patients attending the adult and paediatric clinics, in the selected hospitals were examined.

Using sample size calculation for a single cross-sectional survey:

$$n = \frac{1.96^2 * P_{exp}(1-P_{exp})}{d^2}$$

Where: n = required sample size
P_{exp} = expected prevalence
d = desired absolute precision

Given that P exp = 0.34 (George IO et al), it therefore follows that

(1-Pexp) = (1 – 0.34) = 0.66.

We let the desired absolute precision (d) to be 10%. The estimated sample size is given by

$$n = \frac{1.962 \times 0.34 \times 0.66}{0.12} = 86$$

The estimated sample size of 86 was expanded to 100 to allow for 10% possible non respondent. The prevalence for ocular manifestations of 34% used to calculate the sample size was obtained from a Nigerian study.

Study area/location

Study areas were Murtala Muhammad Specialists Hospital (MMSH), and Muhammad Abdullahi Wase (Nassarawa) Hospital in Kano.

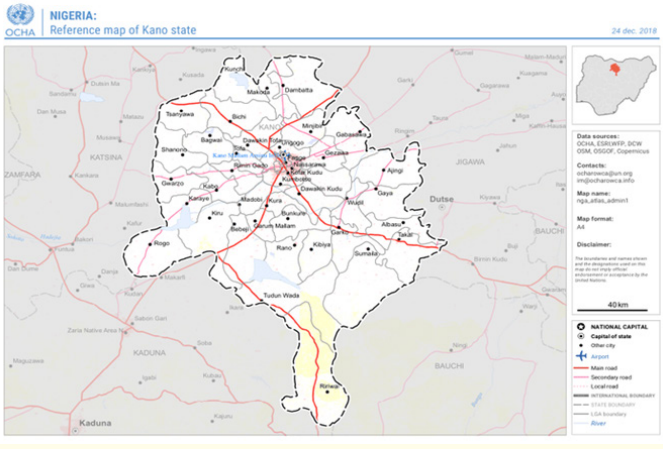

Figure a

Sampling technique

All eligible patients who reported for routine follow-up at the adult and paediatric sickle cell clinics at MMSH and AWH were included.

Materials

- Monocular Direct Ophthalmoscope (wech allyn).
- Distance and near Snellen Visual Acuity Charts.
- Colour Vision chart (Ishihara).
- Trial Frame, Pin hole and Occluder.
- Retinoscope (keeler).
- MARS Contrast Sensitivity Chart.
- Amsler Grid. And
- Pulse Air Tonometer.

Inclusion criteria

All Patients from seven and above years who are diagnosed with sickle cell disease at the selected hospitals and who are residents of Kano metropolis were included.

Exclusion criteria

Patients below 7 years of age were excluded because of special resources for examination and the expected difficulties in examining this age group.

Any sickle cell patient with another systemic and or manifest ocular diseases thought to have the same ocular presentations (comorbidities) with SCD was counted out.

Data collection procedure

The study was explained to the subjects, they were then given the information form detailing the study, those who agreed to be part of the study were given the consent form to sign which would also be signed by the principal investigator.

Once the study participant consented, the procedure of ocular examination then began with obtaining the demographic data after which the examinations followed. The demographic data was obtained by interviewing the patients and or their parents or guardians.

Visual acuity measurement

Distant Snellen letter V.A chart or illiterate E chart was hung at the distance of six (6) meters from the subjects' sitting position at about the subjects' eye level, and the subjects were then asked to read the letters on the chart to determine their distant V.A. Subjects were then made to read the sentences/letters of the near Snellen chart at about arm's length distance, normally in order to determine their near V.A. pin hole acuity and refraction (i.e. objective and subjective) were done to get the best corrected visual acuity (BCVA) in optometry clinic, AKTH, for patients with poor vision.

Contrast sensitivity

The chart was placed at a distance of 50cm from the subject's sitting position at eye level with proper lightning of the environment, subjects were instructed to read the chart with either eye while the other was occluded, it was measured monocularly and then binocularly and the result was recorded using the score sheet.

Visual field

Amsler grid chart was placed on a table and subjects sitted on examination chair comfortably fixating the central dot of the chart from a distance of 30cm, subjects were first presented with chart 1, and asked to report if any of the lines were distorted, dim, broken or disappeared and then followed by chart 2 and all relevant charts depending on the response received. Subjects that reported any abnormally were given a recording form and pencil to draw what they saw.

Colour vision

The book containing the plates was held at 75cm from the subject and tilted so that the plane of the paper was at right angles to the line of vision in a room that was lit adequately by daylight. Plates were presented to the subjects and asked to identify the numbers printed on each plate and a total of 9 plates were u to make the procedure easier and faster, the number of plates identified would then be recorded.

Ophthalmoscopy

Subjects were seated comfortably on the examination chair and were instructed to be fixating at a distant target eg wall of the semi-darkened examination room, Wech allyn monocular direct ophthalmoscope was used to examine the subjects' eyes by holding it at a certain degree eg 15 degree, starting from distance, observing the external structures of the eye, and moving closer until a clear view of the internal structures is obtained, maintaining both the patient and examiner's settings, subjects were examined by using right eye and right hand for the subjects' right eye and using left hand and left eye for their left eye.

Tonometry

Tonometry was carried out with a potable Pulsair intelliPuff Non-Contact Tonometer (Keeler Instruments, Inc.) to measure IOP while the subjects were sitted comfortably on a chair.

Data analysis

The collected data were cleaned and entered in a computer's Microsoft excel data base. It was then exported to IBM SPSS Statistics for analysis.

Results

The response rate was 100% with the full sample size being examined and none of the participants withdrawn from the study.

Characteristics n = 100	n (%)
Age—groups	
7_____16	45 (45.0)
17_____26	35 (35.0)
27_____36	11 (11.0)
37_____46	7 (7.0)
47_____56	2 (2.0)
Gender	
Female	56 (56.0)
Male	44 (44.0)

Table 1: Characteristics of the study participants.

Table 1 Above shows that ages of the participants ranged from 7 years to 56 years old. Majority were in the age group 7 –16 years (45.0%). The mean age and standard deviation of the respondents is 20 years (cl: 17 –26) and SD (10.02) respectively.

The above bar chart shows that, the majority of the respondents came from Nassarawa, participants amounting to 21 (21.0%), followed by Kumbotso having 17 (17.0%) participants, and the Ungogo with 15 (15.0%) then Tarauni with 13 (13.0%), Dala with 10

(10.0%), Gwale having 9 (9.0%), Fagge having 8 (8.0%) and and finally Kano Municipal having the least participants with just 7 (7.0%)

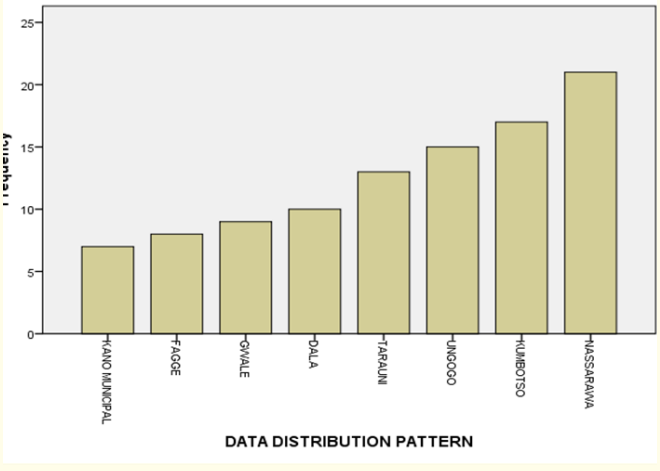

Figure 1: Local government area of participants.

Visual function tests findings

Visual acuity

Table 2 Shows that the majority of the participants 102 (51.0%) had normal vision whereas decreased vision had a prevalence of 87 (43.5%), visual impairment was 11 (5.5%) and there was no blindness recorded.

Table 2: Visual acuity in the affected eyes.

Visual acuity n = 200 eyes of 100 participants	n (%)
Normal vision	102 (51.0)
Decreased vision (VA)	87 (43.5)
Visual impairment	11 (5.5)
Severe visual impairment	0 (0.0)
Blindness	0 (0.0)

Contrast sensitivity findings

The above pie chart shows the result of CS findings where the test shows that 169 (84.5%) eyes had abnormal CS findings whereas only 31 (15%) eyes had normal CS scores out 200 eyes.

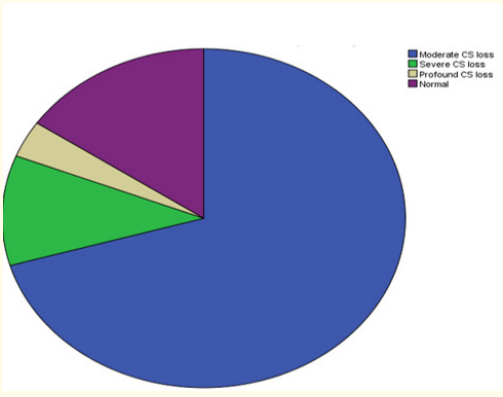

Figure 2: Contrast sensitivity findings.

Visual field

The above chart shows the amsler grid visual field test results with 19 (9.5%) of eyes having abnormal findings and 181 (90.5%) eyes that were normal.

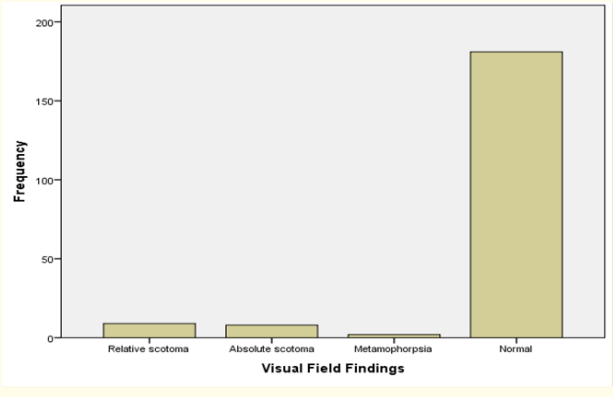

Figure 3: Visual field findings.

Clinical examination findings

As shown in figure 4 above, ocular media opacity was present in 2.5% of the eyes. Most eyes of the participants 97.5% had normal or clear media.

Posterior segment findings

Table 3 Shows internal examination findings where 11% eyes had non-proliferative findings, 0.5% of eyes had proliferative findings and 86% of eyes had normal fundal background.

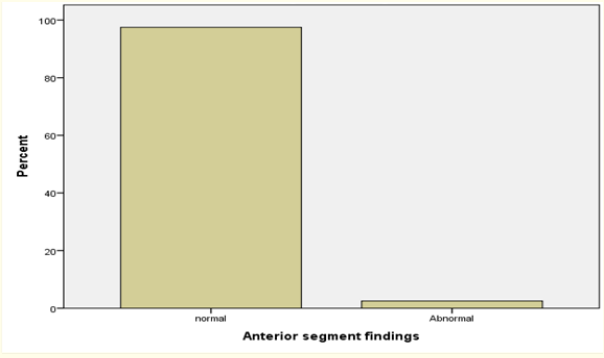

Figure 4: Anterior Segment Findings.

Posterior segment findings n = 95 eyes of 100 participants	n (%)
Non-proliferative Findings	22 (11)
Venous tortuosity	14 (7.0)
Artero-venous tortuosity	5 (2.5)
Optic disc sign of sickling	1 (0.5)
Artero-venous nipping	2 (1.0)
Proliferative findings	1 (0.5)
Normal findings	172 (86)

Table 3: Posterior segment findings.

Discussion

In this study, the number of the subjects declined with increasing age, with 45 subjects (45%) in the age group 7—16 years and only 2 subjects (2%) in the age group 47—56 years. This may reflect the lower life expectancy of sickle cell patients when compared to the general population in Kano. However some other factors might contribute to this findings such as reduced mobility of older patients. The study also revealed that ocular manifestations and oculo-visual functions loss and or defects or impairment were common among patients living with SCD at Murtala Muhammad and Muhammad Abdullahi Wase (Nassarawa) Hospitals in Kano.

Decreased visual acuity (visual loss) was noted in this study to be 87 (43.5%) eyes, visual impairment 11 (5.5%) eyes while no eye had legal blindness and causes were astigmatic refractive error, cataract and non-proliferative sickle cell retinopathy and proliferative sickle cell retinopathy. Those with decreased visual acuity had normal best corrected visual acuity (BCVA) on refraction or pin hole acuity. This study is similar to many previous studies such as that of Condon and Serjeant (1976) at the University of West Indies, Kingston, Jamaica [10]. Which this study is more closely related to, as it had documented causes of visual loss that are same but had higher prevalence. Other studies that were similar to this are: Comparative study by Moriarty., et al. (1988) that found the incidence of visual loss to be 31 per 1000 eye-years observation among eyes with proliferative disease compared to 1.4 per 1000 eye-years observation among eyes with non-proliferative disease [4]. And that of Fadugbagbe., et al. (2013) who documented that visual impairment was present secondary to proliferative sickle retinopathy [6]. This study is contrary to some previous studies also, these include:

Talbot., et al. (1982) prospective study carried out in Jamaica who found visual acuity to be 6/9 or better in all children he examined and Spyridon., et al. (2010) comparative study that confirmed that contrast sensitivity can be significantly impaired but Snellen visual acuity seems unaffected [8,9].

This study found high prevalence of contrast sensitivity loss/impairment amounting to as high as 169 (84.5%) eyes with 141 (70.5%) eyes having moderate CS loss, 21 (10.5%) eyes having severe CS loss and 7 (3.5%) eyes having profound CS loss whereas only 31 (15.5%) having normal score. This finding goes hand in hand with that of Spyridon., et al. (2010) comparative study to evaluate and compare contrast sensitivity in patients with Beta-Thalassemia Major and Sickle Cell Disease Undergoing Regular Transfusions and Chelation Therapy with Desferrioxamine in Epirus Vision Centre, Ioannina, Greece. Where he also found impairment of contrast sensitivity in both groups [8] and also with that conducted by Gilles., et al. (2017) in a single referral centre for sickle cell disease in 2016 and also found same result [22].

The visual field test findings in this study also shows visual field defects in some subjects. Where only 19 (9.5%) eyes had visual defects and these include 9 (4.5%) relative scotoma, 8 (4.0%) absolute scotoma and only 2 (1.0%) eyes had metamorphopsia whereas 181 (90.5%) eyes had normal visual field. This finding is almost similar to the ones conducted earlier by Gilles., et al. (2017) in a single referral centre for sickle cell disease in 2016 in a retrospective study [22]. And that of Kwok., et al. (2022) that found binasal visual field defects due to sickle cell maculopathy in a case report to assess the visual field in a 26-year-old African American man with

sickle cell disease who noticed blurry vision in both eyes [28] and the difference between this study and those by Gilles., et al. and Kwok., et al. is that this used amsler grid chart and therefore, tests for central 10-20 degree visual field while standared automated perimetry was used for their own and therefore, tested for central 25 to 30 degree.

Out of the 200 eyes of 100 participants examined only 5 (2.5%) eyes found to have media opacity (cataract) indicating that the prevalence is low in this study compared to the previous ones, studies that had reported same result include that of Condon and Serjeant (1976) at the University of West Indies, Kingston, Jamaica [10] and Osafo-Kwaako., et al. (2011) in a Hospital-based cross-sectional study at Korle-bu Hospital, Accra, Ghana [2]. This study does not report any other anterior segment sign of sickling like conjunctival signs including coma shaped vessels, injections and iris signs i.e. iris depigmentation, iris atrophy and rubeosis as reported by previous studies.

The prevalence of posterior segment manifestations of SCD (i.e. sickle cell retinopathy) in this study is 23 (11.5%) eyes. This includes non-proliferative fundus signs 22 (11.0%) eyes and they are, tortuosity of the major retinal vessels i.e. venous tortuosity 14 (7.0%) and artero-venous tortuosity 5 (2.5%), optic disc sign of sickling 1 (0.5%) and artero-venous nipping 2 (1.0%) eyes and then proliferative sickle cell retinopathy which is only 1 (0.5%). These findings are supported by many preceded studies like that of Condon and Serjeant (1976) at the University of West Indies, Kingston, Jamaica. Where they found retinal detachment in two, vitreous detachment in one, long standing optic atrophy, tortuosity of the major retinal vessels present in 18 (30 percent) patients affecting the veins alone in 13 and the arteries and veins in five [10]. Fadugbagbe., et al. (2013) who also found proliferative sickle cell retinopathy as a cause of visual impairment [6]. Osafo-Kwaako., et al. (2011) in a Hospital-based cross-sectional study at Korle-bu Hospital, Accra, Ghana, he found PSR in 12.9% of subjects he examined [2]. Moriarty., et al. (1988) in a comparative study also reported the prevalence of both proliferative and non-proliferative disease [24].

Colour vision testing revealed normal findings in all 100 subjects examined. In the previous studies like that of Roy., et al. (1987) where they used Farnsworth-Munshell 100-hue test to compare colour vision in patients with sickle cell anaemia and normal controls, found that SCD patients had significantly more blue-yellow and mixed colour vision defects. ROY., et al. (1988) in another comparative study using The Lanthony D15 desaturated test reported same result as the first one [23,24]. No wonder this study did not catch a single colour vision defect because Ishihara test was used, As Ishihara chart is only efficient in screening for red-green colour defects (that is mostly congenital) and cannot detect for blue-yellow defect (which is always acquired) this shows that SCD patients might not have congenital colour defect.

Finally intraocular pressure is found to be less than 21mmHg in all 200 eyes of the subjects; this is similar to what Osafo-Kwaako., et al. (2011) in a Hospital-based cross-sectional study at Korle-bu Hospital, Accra, Ghana also reported [2], so also Abdi Daher., et al. (2005) found normal IOPs in all SCD patients he examined [1]. But in a study done by George (2012), in Nigeria he found out that 1.1% of the cases studied had glaucoma [3].

Conclusion, Recommendations, and Limitations

Conclusion

From the results of this study, it can be concluded that:

- SCD patients have oculo-visual function disorders.
- Oculo-visual function disorders in SCD occurred in 2.5% anterior segment and 11.5% posterior segment in Kano.
- The commonest ocular disorder among SCD patients is vascular tortuosity.
- There is oculo-visual function impairment in SCD patients in Kano.
- There is decrease in visual acuity in 43.5% and visual impairment in 5.5% while there was no blindness documented.
- Contrast sensitivity impairment occurred in 84.5% and majority fall in moderate CS loss category accounting for 70.5%.
- Visual field defect occurred in 9.5% with relative scotoma having the highest occurrence.
- Colour vision sensitivity was normal using the Ishihara test.
- And finally intraocular pressure (IOP) was bellow 21mmHg in SCD patients in Kano.

Recommendations

Recommendations are as follow:

- All patients with sickle cell disease should at least have two to three ophthalmic examinations in childhood and a more regular frequent follow up in adulthood.

- Further study should be carried out in Kano and to include OCT and visual field test using standard automated perimetry for better diagnosis of SCD maculopathy and retinopahy.
- Primary care givers/physicians should refer any SCD patient who complain of ocular symptoms to eye care professionals for checkup.

Limitations

- Lack of ocular coherence tomography (OCT). With OCT, Fundus findings such as macular hole, macular oedema and other retinal abnormalities due to SCD might have been reported.
- Lack of other colour vision testing tools like Farnsworth-Munsell D-15, Lenthony D15 desaturated test, Farnsworth-Munshell 100 hue and City University test charts resulted in reduction of reported colour vision defects especially blue-green deficiency.
- Due to lack of standard automated perimetry more visual field defects beyond 10 degree of central visual field like hemianopias might have been reported.
- Failure to meet agreement with the general physicians and or care givers of the subjects to perform dilated fundus examination which could have allowed for more fundus signs like angiod streak, salmon patch haemorrhage and schisis cavity to be revealed.

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