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Editorial: Cataract auditing as a tool for improving effective cataract surgical coverage

Globally, there is an estimated 216.6 million people with visual impairment (80% uncertainty interval [UI] 98.5-395.1 million)[1]. Of these, 39 million people are blind (80% UI 12.9-65.4 million)[1]. Cataract is the leading cause of blindness globally, affecting approximately 12.6 million people in 2015 (80% UI 3.4–28.7)[1]. Cataract is also the cause of one quarter of moderate and severe vision impairment, affecting an estimated 52.6 million people (80% UI 18.2–109.6)[1]. Cataract surgery is the only definitive management for cataract with an estimated 25 million surgical cases per year. It is a highly effective sight restoration intervention.

There are several measures used to monitor the effectiveness of cataract surgery such as cataract surgical rate, cataract surgical coverage, sight restoration rate and others. In 2017, the concept of effective cataract surgical coverage started to take shape. This refers to proportion of people aged 50 years or older with operated cataract or operable cataract who have a good postoperative presenting visual acuity of 6/18 or better. According to the Lancet Global Health Commission on Global Eye Health: beyond vision 2020, the median effective cataract surgical coverage for 48 countries was less than 50% for an outcome of 6/18 or better and an operable cataract threshold of worse than 6/60 between 2000-2019. This would be much less than the WHO recommended target of 80%. Much of the skewing towards the worse outcomes was in Sub Saharan Africa (SSA)[2].

Several things are needed to improve effective cataract surgical coverage in many parts of the world. One mechanism for which this can be done is continuous quality improvement through cataract surgical outcomes monitoring. Monitoring cataract outcomes is an important activity to ensure quality and safety of the cataract surgical services. Routine monitoring of pre-operative vision, operative and post-operative data of each operated patient calculates the visual outcome and assesses the quality of cataract surgery. It is assumed that encouraging eye surgeons to monitor their own results, over time, will lead to better outcomes of cataract surgery.

In Uganda, we have piloted the "Sustained Routine Cataract Auditing (SROCA)" intervention to improve cataract outcomes. The overall purpose of this project is to test the feasibility of introducing a cataract outcome monitoring practice in Uganda and the impact of this intervention on the quality and safety of cataract surgery. This work is part of a larger 3-year project "Increasing Equitable Access to Effective Cataract Services in West, and Southwest Uganda."funded by Fred Hollows Foundation in partnership with CBM and implemented by Ruharo Mission Hospital and Mbarara University and Referral Hospital Eye Centre (MURHEC).

SROCA draws lessons from the maternal mortality audits in Uganda to provide a safe, positive, and non-judgmental environment of objectively reviewing, discussing and sharing cataract surgery outcomes. It is currently being piloted at the major high volume cataract centers in Southwestern Uganda. The platform includes routine collection of data of all adult patients who undergo cataract surgery at the participating hospitals. Customized monthly audit reports are presented to the clinical teams based on the WHO classifications of outcome. In addition, the worst 5th percentile cases are discussed to explore reasons for their poor outcomes and the teams are facilitated to discuss a continuous quality improvement strategy. The participating hospitals have an opportunity to meet at quarterly audit network meetings to share lessons and best practices. The trend of outcomes will be monitored annually to monitor improvement over time.

The individual hospitals will be able to have candid review meetings and utilize their data to improve their clinical practice into better patient outcomes. At the hospital level, this will result into peer-to-peer adoption of improved surgical technique and medical management, improved case selection, revised protocols for intraoperative and postoperative management. As a network, the participating hospitals will discuss overall results and share lessons and challenges to support each other through this shared learning process.

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Clinical and optical coherence tomography features of adult-onset foveo-macular vitelliform dystrophy mimicking age-related macular degeneration

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ABSTRACT

Objective: To describe the clinical and Optical Coherence Tomography (OCT) findings of Adult-Onset Foveo-Macular Vitelliform Dystrophy (AFVD) mimicking age-related macular degeneration.
Method: A retrospective study design was employed which reviewed clinical and optical coherence tomography images of patients with AFVD from September 2019 to March 2021.
Results: There were 12 (9 male and 3 female) study subjects with a mean age of 62.75 years. The presenting visual acuity ranged from 20/100 to 20/20. Of 19 eyes, 10 eyes showed vitelliform lesions; 5 eyes had pseudo-hypopyons, and 4 eyes were in the vitelliruptive stage. OCT showed disruption of the IS/OS interface in 8/19 eyes (3 eyes with stage I, 1 in stage II, and 4 eyes with stage III disease). Optically clear (non-reflective) subretinal lesions were present in OCT images from 6 eyes (5 in pseudo-hypopyon and 1 in vitelliruptive stages). Four of five eyes in the pseudo-hypopyon stage had intact IS/OS photoreceptor interfaces.
Conclusions: An optically clear space is a distinct feature of AFVD that is different from the pattern of fluid in neovascular AMD that may have a protective role against the disruption of the photoreceptor interface. It is important to be aware of the stages of AFVD and to differentiate them from neovascular AMD, as AFVD does not require treatment with injections of anti-vascular endothelial growth factor.

Key words: Adult-onset foveo-macular vitelliform dystrophy, Age-related macular degeneration, Optical coherence tomography

INTRODUCTION

Adult-Onset Foveo-Macular Vitelliform Dystrophy (AFVD) was first described by Gass in 1974[1]. In its original description, this disease was called peculiar foveomacular dystrophy. It generally presents in the fourth to sixth decades of life in patients who are visually asymptomatic, who have mild blurring of vision, or who have mild metamorphopsia. The typical patient has bilateral, asymmetric, foveal or perifoveal, yellow, solitary, round to oval elevated sub-retinal lesions of less than 1/3 disc diameter, often with central pigmentation[2, 3].

There is a four-stage classification for AFVD based on the one that was established for Best vitelliform macular dystrophy. These are vitelliform, pseudo-hypopyon, vitelliruptive, and atrophic, in progressive order[4]. Many patients show a different stage in each eye, multiple characteristics of different stages within a single lesion, and sometimes even multifocal vitelliform lesions[5]. The vitelliform stage is the classic dome-like lesion with sub-retinal vitelliform material. In the pseudo-hypopyon stage, the vitelliform material settles inferiorly within the sub-retinal space, creating an optically empty zone superiorly, with hyper-reflective material inferiorly and a horizontal demarcation at the interface. The vitelliruptive (scrambled egg) stage is characterized by the collapse of the dome-like shape into a lesion with heterogeneous vitelliform material and hyper-reflective clumps within the inner retina. The atrophic stage is characterized by a homogenous plaque of atrophy centered on the macula.

Based on spectral-domain OCT findings in patients with AFVD, it was hypothesized that early changes involve the layer between the RPE and the ellipsoid zone, first with the accumulation of material beneath the sensory retina[6,7].

Clinico-pathological studies confirmed the sub-retinal location of the vitelliform material, demonstrating pigment-laden cells, lipofuscin granules, photoreceptor debris, and RPE cells in varying stages of disintegration in the sub-retinal space[8-10].

Due to this clinical variability of AFVD, there is considerable overlap with age-related macular degeneration. Choroidal neovascularization (CNV) can also occur in macular dystrophies, although it is rare and appears to have a relatively favorable prognosis as compared to CNV in AMD[11]. Most often, AFVD is misdiagnosed as AMD. It is therefore important to carefully define the ophthalmoscopy and imaging

characteristics of this maculopathy to improve diagnostic accuracy. The purpose of our study was to describe the clinical and OCT morphologic features in patients with AFVD that help differentiate from nv-AMD.

MATERIALS AND METHODS

A retrospective study design was employed which reviewed the clinical and imaging studies of patients who have had an evaluation at WGGA Eye Center and Roha Specialized Eye Clinic, in Addis Ababa, Ethiopia from September 2019 to March 2021.

Inclusion criteria were all patients with a clinical diagnosis of AFVD, confirmed by the presence of a sub-foveal, round, yellowish lesion and with the corresponding presence of hyper-reflective sub-retinal material on Spectral-Domain Optical Coherence Tomography (SD-OCT). Five yellow eyes with no apparent macular lesions were excluded.

All study subjects had a comprehensive ophthalmology examination including measurement of Best-Corrected Visual Acuity (BCVA) at 3M with Snellen's visual acuity chart, intraocular pressure with Topcon non-contact tonometry, dilated bio-microscopic examination, fundus photography, and optical coherence tomography using 3D OCT-1, Maestro-I/and II, Topcon, Tokyo, Japan. The field of view of 3D OCT-1 used was a 6mm by 6mm area centered on the fovea.

Nineteen eyes had macular scans, and both 3D and 2D cross-sectional images were assessed with particular attention to the Inner Segment/Outer Segment (IS/OS) of photoreceptor cells, the retinal pigment epithelium, the morphology, and the stage and location of vitelliform lesions.

The procedure of this investigation conformed to the tenets of the Declaration of Helsinki, and the Ethics committees of WGGA and Roha Specialized Eye Clinic approved the retrospective revision of the clinical data. The data were kept anonymous after extraction from both the clinical and imaging findings.

RESULTS

There were 12 (9 male and 3 female) study subjects with a mean age of 62.75 years. Cases 1, 2, 3, 4, 6, 7, 9, 10, 11 and 12 noticed a gradual blurring of vision, while case 5 presented with sudden onset of distortion of vision in the right eye. Case 8 had no complaint of reduced vision, and the lesions were detected during a routine fundus evaluation (Table 1).

Table 1: Patient characteristics

Age/gender	Study eye	VOD	VOS	Integrity of IS/OS interface OD	Integrity of IS/OS Interface OS	Stage OD	Stage OS
1. 67/M	OU	20/50	20/40	Diffuse disruption	Focal disruption	III	II
2. 63/M	OU	20/70	20/70	Focal disruption	Focal disruption	III	III
3. 82/M	OU	20/60	20/100	Disrupted	Disrupted	I	I
4. 70/M	OS	NA	20/60	NA	Disrupted	NA	I
5. 58/F	OD	20/20	NA	Intact	NA	II	NA
6. 59/M	OD	20/30	NA	Intact	NA	II	NA
7. 64/F	OS	NA	20/25	NA	Intact	NA	I
8. 55/M	OS	NA	20/70	NA	Segmental disruption	NA	III
9. 40/M	OU	20/60	20/20	Intact	Intact	II	I
10. 54/M	OU	20/20	20/30	Intact	Intact	I	I
11. 70/M	OU	20/60	20/60	Intact	Intact	I	I
12. 71/F	OU	20/100	20/30	Intact	Intact	I	II

Note: NA is to indicate eyes that were excluded as they did not have AFVD lesions

The presenting visual acuity ranged from 20/100 to 20/20. Seven study subjects had bilateral disease, while five were unilateral. Their five fellow eyes were excluded from analysis because they did not have yellow macular deposits or other signs of AFVD.

Nineteen eyes fulfilled clinical and OCT inclusion criteria for AFVD. There were 10 eyes with vitelliform lesions (Stage I); 5 pseudo-hypopyon eyes (stage II); 4 vitelliruptive eyes (stage III) and no atrophic stage eyes (stage IV).

OCT through the macular lesion demonstrated optically clear spaces in all 5 eyes with pseudo-hypopyon stage and in 1 eye with vitelliruptive stage (case 2 right eye). OCT showed disruption of IS/OS of photoreceptors in 8/19 (42%) of study eyes (3 eyes with stage I, 1 eye with stage II, and 4 eyes with stage III disease). It was evident that both the Inner Segment/Outer Segment (IS/OS) interface and the External Limiting Membrane (ELM) were disrupted in two eyes (Case 1, OS and case 2, OD), and as a result of this, hyper-reflective clumps were seen in the inner retina (Figures 1, 2).

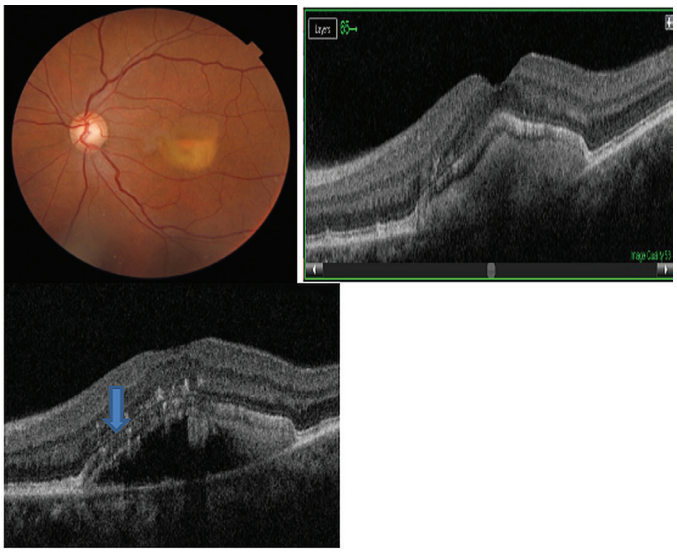

Figure 1: Colour fundus photo and optical coherence tomography of a 67-year-old male: Fundus photo OD: Central, layered vitelliform lesion. Optical coherence tomography B- scan slice with hyper-reflective dome-shaped lesion beneath a demarcation line and a non-reflective (optically clear) space above the demarcation line. Note the irregular and focally disrupted IS/OS layer and external limiting membrane (ELM) and hyper-reflective foci in the outer nuclear layer

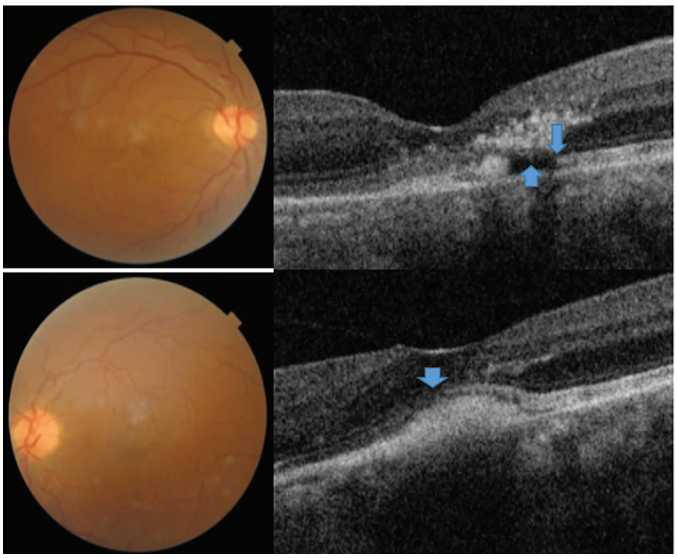

Figure 2: Colour fundus photo and optical coherence tomography of a 63-year- old male: Fading central yellow deposits OU, OCT scan OD: heterogeneous reflectivity beneath the neurosensory retina with the optically clear zone (up arrow), Note the segmental disruption of IS/OS layer and focal disruption of the external limiting membrane and hyper-reflective clumps in the outer nuclear layer and outer plexiform layer (down arrow). Note the dome-shaped sub-retinal hyper-reflective deposit with disrupted IS/OS interface (downward arrow)

In three eyes (cases 3 & 4), fundus photography showed multiple round yellowish macular deposits, and their corresponding SD-OCT revealed discrete hyper-reflective sub-retinal deposits with distortion of the contour of the IS/OS layer that appears to be reticular-pseudodrusen-like lesions (Figures 3, 4).

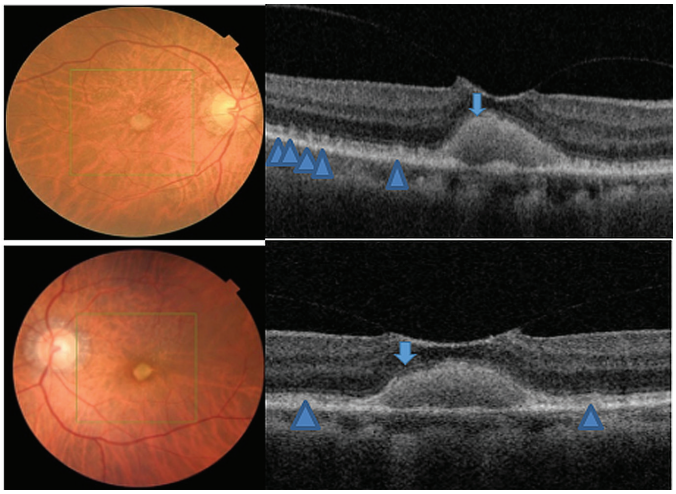

Figure 3: Colour fundus photo and Optical Coherence Tomography (OCT) images of an 82-year-old male. Fundus photo: circumscribed, raised yellow macular lesion associated with multiple, round yellowish-white discrete deposits OU. Optical coherence tomography: sub-foveal dome-shaped hyper-reflective lesion, with early VMT OU. Note the disruption of IS/OS layer (down arrows) and mounds of granular deposits representing reticular-pseudo-drusen-like lesions lying in the sub-retinal space and altering the contour of IS/OS interface (triangles) OU

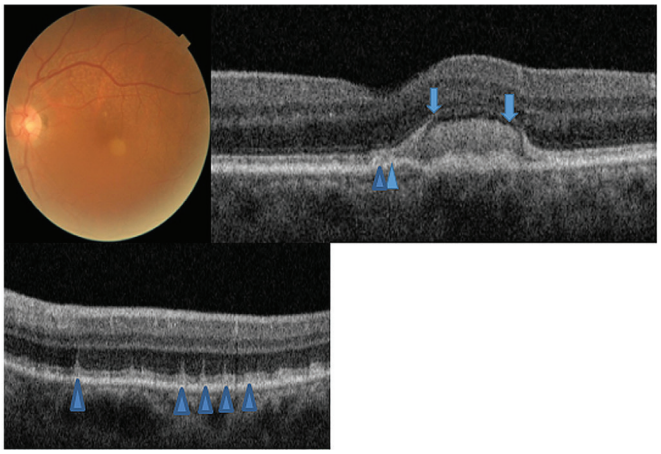

Figure 4: Colour fundus photo and optical coherence tomography images of a 70-year-old male: Fundus photo: vitelliform macular lesion and associated multiple round oval yellowish deposits mostly in the superior perifovea OS. OCT: Juxtafoveal sub-retinal hyper-reflective dome-shaped lesion. Note the disrupted and attenuated IS/OS zone (down arrows) and conical shaped hyper-reflective sub-retinal lesions protruding to the outer nuclear layer seen mostly in the superior perifovea (triangles)

DISCUSSION

The term adult-onset foveomacular vitelliform dystrophy currently used in the literature likely represents a group of disorders with overlapping phenotypes[12]. Some cases have an autosomal dominant trait while similar phenotypes are explained by a variety of genetic, toxic, immune-mediated, degenerative, and mechanical mechanisms[12].

Optically clear (non-reflective) subretinal lesions were present in OCT images from 6/19 eyes (5 in pseudo-hypopyon and 1 in vitelliruptive stages).The OCT image of case 1, OS (Figure 1), was of interest with a hyper-reflective dome beneath the demarcation line and an overlying optically clear space. This is a pseudo-hypopyon stage representing partial liquefaction of vitelliform material.

This optically clear space is a distinct feature of AFVD different from the pattern of fluid seen in neo-vascular AMD. The sub-retinal fluid in neo-vascular-AMD assumes a more horizontal position as there is a generalized macular disease and less resistance for the fluid to spread in the interstitial space of the retina. It is also associated with neovascular membranes seen by SD-OCT as a hyper-reflective band/or patch beneath the retinal pigment epithelium, in the subretinal space, or bridging the RPE and retina. Intra-retinal fluid can be seen based on the stage of the disease and the type of choroidalneovascular membrane.

In AFVD, optically clear cystic spaces lying between the retinal pigment epithelium and the neurosensory retina conform to the dome-shaped lesion. This optically clear region was also considered to represent a loss of the photoreceptors and/or accumulation of eosinophilic fluid, as was reported by Jaffe and Schatz[9]. In addition, the accumulation of sub-retinal fluid may be due to the decreased ability of RPE cells to pump fluid from the sub-retinal space. In vitelliform macular dystrophy, the sub-retinal fluid appears to be caused by an underlying defect of VMD2, which encodes bestrophin, a calcium-sensitive chloride channel protein found in the basolateral membrane of RPE cells[6,13].

It is important to note the SD-OCT findings of case 1, OS (Figure 1) and case 2, OD (Figure 2) with pseudo-hypopyon and vitelliruptive stages where hyper-reflective clumps were seen in the outer nuclear and outer plexiform layers. These lesions in the inner retinal layer may correspond to migrated pigment-laden cells that gained access through the disrupted IS/OS and external limiting membrane layers, which is in agreement with previous OCT and clinicopathologic reports[7,8]. Such hyper-reflective dots may reflect part of an absorption process of the vitelliform lesion and may not represent lesions of AMD.

In contrast to AMD in which histopathological abnormalities commence as basal laminar deposits beneath the RPE, the earliest changes of AFVD involve the photoreceptors[10]. The SD-OCT findings in our series also confirm the location of the vitelliform material in the sub-retinal space lying between the photoreceptors and retinal pigment epithelium.

Another interesting observation was that 4/5 of eyes with the pseudo-hypopyon stage had intact IS/OS interfaces. Cone photoreceptors can function relatively well despite the physical separation from the RPE because of an alternative, intraretinal visual pigment regeneration route[12,14,15].The classic pigment regeneration route involves photo isomerization of 11-cis- retinal to all-trans-retinal in the photoreceptors and the shuttling of all-trans-retinol from the photoreceptors to the RPE and 11-cis- retinal from the RPE to the photoreceptors. Recent evidence suggests the existence of a second retinoid visual cycle within the retina that is independent of RPE to specifically support rapid cone pigment regeneration[14,15]. The sedimentation of vitelliform material inferiorly, and the resulting optically clear space between the material and the IS/OS interface of photoreceptors may shield the IS/OS interface from the effect of the vitelliform material. Therefore, we hypothesize that the presence of fluid in the sub-retinal space could have a protective role against the disruption of the IS/OS of photoreceptors at least until the disease progresses to the next stage. We understand this has to be proven through a long-term, prospective study with enough study subjects.

In our series, SD-OCT images showed disruption of the IS/OS interface in 8/19 (42%) of study eyes. Interestingly, 3/19 eyes with disrupted IS/OS layers had vitelliform lesions, and associated reticular-pseudo-drusen-like lesions (Figures 3, 4). Although most eyes in the vitelliform stage have intact IS/OS and preserved vision, some eyes could develop disruption of the IS/OS interface in the early stage of AFVD. One or any combination of the descriptions could explain the mechanisms how some eyes in the early stage of AFVD develop the disruption of the ellipsoid zone.

(i) Although we could not make any assumptions on how long the vitelliform material had been in the sub-retinal space, we think that even in the early stage of AFVD, there could be local phagocytic dysfunction of RPE. And as a result of this functional failure, pigment-laden cells and other components of vitelliform material could cause damage to the photoreceptors.

(ii) The presence of reticular-pseudo-drusen-like deposits could have contributed to mechanisms that led to the disruption of IS/OS of photoreceptors described in 3 eyes.

(iii) Moreover, previous studies explained the challenge of delineating the IS/OS interface, due to the elevated dome-shaped lesions. Alternatively, it has been reported in a clinicopathological study of AFVD eyes that inner segments may be stunted, which may represent a sign of photoreceptor cell dysfunction[7,8].

AFVD was associated with different types of drusen that include subretinaldrusenoid deposits (reticular pseudodrusen), basal laminar deposits (cuticulardrusen), or drusen that are typical for AMD[1,12,16]. The subretinal location of both psuedodrusen and vitelliform lesions might reflect a common pathogenic mechanism associated with the altered function of the RPE-photoreceptor complex[12].

The sudden onset of metamorphopsia noticed in the right eye of case 5 could be the result of an abrupt collapse of the vitelliform lesion that produced morphologic changes in the neurosensory retinal layer. Careful monitoring of this patient showed no sign of choroidal neovascularization, and there was complete resolution of complaints in subsequent weeks.

CONCLUSIONS

Based on the finding of intact IS/OS interface in eyes with most pseudo-hypopyon stages, we hypothesize that the presence of fluid that shields the photoreceptor cells from the effect of vitelliform material could have a protective role from disruption of IS/OS interface. It is important to be aware of pseudo-hypopyon and vitelliruptive stages of AFVD that have sub-retinal fluid and hyper-reflective intra-retinal changes and to differentiate this condition from neo-vascular AMD. Making this distinction can help avoid unnecessary interventions and risks associated with intravitreal treatment.

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Clinical presentation and management of retinoblastoma at Queen Elizabeth Central Hospital, Blantyre, Malawi

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ABSTRACT

Objective: Retinoblastoma is the commonest intraocular malignancy in childhood worldwide commonly affecting children in the first 5 years of life. The primary goal of retinoblastoma treatment is to improve survival. This is achievable through early detection. We conducted this study to report the clinical presentation and management of retinoblastoma in Blantyre, Malawi.

Methodology: This was a retrospective case series in which all files of patients who presented with a clinical diagnosis of retinoblastoma from 1st January to 31st December 2017 were reviewed. Data extracted included clinical presentation, lag time, examination findings, investigations and treatment provided. Categorical variables were summarized as percentages and frequencies whilst continuous variables were analysed as medians.

Results: A total of 43 patient records were retrieved. The median age was 31 months with a median lag time of 3 months (IQR 0.1 – 48 months) with most patients (n =37, 86%) presenting to the retinoblastoma treatment unit within two weeks of being referred. There was no gender preponderance with a male to female ratio of 1:1 Most patients presented with leukocoria (n = 29, 67.4%) and almost a third had proptosis (n = 13, 30.2%).Thirty six patients (83.7%) had a unilateral presentation. Patients were clinically staged using the ICRB staging. There was limited availability of facilities for thorough patient evaluation with 21 (48.8%) patients being examined under anaesthesia, B-scan being performed in 14 (32.6%) patients and only 6 (13.9%) patients undergoing Magnetic Resonance Imaging (MRI). Enucleation and chemotherapy were the only treatment options available with chemotherapy being given to 40 (93%) of the patients and 28 (65.1%) children underwent enucleation apart from chemotherapy over the study period.

Conclusions: The median lag time to presentation was 3 months and most of the delay in presentation was at the level of the patient before the first contact with the health system. There is a need to improve treatment options for retinoblastoma at Queen Elizabeth Central Hospital whilst also improving community awareness and early case detection at the primary level of health care.

Key words: Presentation, Lag time, Examination, Investigations, Treatment

INTRODUCTION

Retinoblastoma is an intra-ocular cancer that occurs due to a mutation of the RB1 gene[1]. It is the most common intraocular malignancy in children, with an incidence of 1 in 15,000 to 1 in 18,000 live births worldwide[1,2].This tumour arises from the primitive photoreceptor cells in the developing retina that contain predisposing mutations in both copies of the RB1 gene. It can be unilateral or bilateral, growing as a solitary or multifocal tumour[3].The mean age at diagnosis is 12 months for bilateral tumours and 24 months for unilateral tumours[4].

Leukocoria is the most common presenting sign of retinoblastoma[4]. Abramson *et al*[5] reported that some children with retinoblastoma develop a squint[4]. This was further noted to be the second most common presentation in the developed world. Advanced intraocular tumours present with pain, glaucoma or buphthalmos[5]. A study done by Goddard *et al*[6] indicated that patients may present with orbital or metastatic disease as the tumour progressed and metastases occur most commonly in the central nervous system, bones, bone marrow and liver. This shows that most children with retinoblastoma are likely to be systemically well. Poor visual tracking, glaucoma, and inflammation were also seen as other presenting signs[6]. A study conducted in the Democratic Republic of Congo (DRC) showed similar results with leukocoria being the most common presenting factor followed by strabismus[7].

Clinical examination includes imaging and examination under anaesthesia to stage the tumour and

rule out other causes of leukocoria[4]. Delay in diagnosis can be divided into parental associated delay and health care associated delay[8]. The duration between when a parent recognized a symptom and when they presented to the hospital is known as lag time. Lag time is a prognostic indicator that often determines the treatment that a patient will receive. Those with a long lag time are likely to undergo eye removal surgery compared with patients with a low lag time who would likely receive globe-salvaging or sight-salvaging treatments.

Lag time is used as an indicator that is associated with possible poor outcomes and mortality[8]. A high lag time is reported to be associated with high-risk retinoblastoma that includes choroidal and optic nerve invasion.

Classification of retinoblastoma has gone through many changes as treatment strategies have evolved. In the 1960s, Reese and Ellsworth developed the Reese-Ellsworth classification which showed to predict globe-salvage[9]. This was a time where external beam radiation was the primary form of treatment[9]. However, the Reese-Ellsworth classification failed to incorporate vitreous and subretinal seeding, and a more modified classification was developed to predict success. Eventually, the International Classification of Retinoblastoma (ICRB) was developed, placing an emphasis on focal and diffuse vitreous and subretinal seed[10]. This is the classification that is widely used to date.

Community awareness of retinoblastoma is important in early diagnosis of retinoblastoma. This consists of regular teaching programs for paediatricians and general physicians, including the key symptoms of retinoblastoma. The timely diagnosis of retinoblastoma increases the chance of treatment success[11].

Currently, the retinoblastoma therapy aims to prevent blindness and serious side effects that can reduce life expectancy and quality life of the patients. Conservative therapy includes photocoagulation, cryotherapy, chemotherapy and radiotherapy[12]. Surgical practice includes enucleation and exenteration[11]. However, exenterations are now highly discouraged. In Indonesia, about 20% of children that come with intraocular retinoblastoma are treated with enucleation[14]. This study also showed that mortality rates were high if parents refused the indicated surgical therapy. Meel *et al*[13] also reported that death was caused by tumour metastasis. This shows us that there is a need of community action to promoting early diagnosis and treatment for child with retinoblastoma, especially in a developing country.

Malawi's child eye health tertiary facility is located in the Southern region of the country and an average of 45 patients with retinoblastoma are managed annually at the facility. We conducted a descriptive case series at the hospital to report the clinical presentation and management of patients with retinoblastoma.

MATERIALS AND METHODS

Setting: This descriptive case series study was undertaken in the Ophthalmology Department at Queen Elizabeth Central Hospital (QECH) in Blantyre, Malawi, which is the country's paediatric ophthalmology referral facility. The center treats at least four new patients with a diagnosis of retinoblastoma each month. The treatment is provided in collaboration with the Paediatric Oncology Department at QECH.

Study participants: All files of children who presented with a clinical diagnosis of retinoblastoma from 1st January to 31st December 2017 were included in the study.

Data collection: All data collected was paper-based and documented onto a structured data collection tool. This included socio demographic features, pre-referral pathway, and family history. The lag time was defined as the duration between when the parent first noticed ophthalmic symptoms and the first presentation to QECH.

Data was collected on whether there was use of investigations such as B-scan and MRI and also on how the retinoblastoma was graded (International Classification for Retinoblastoma). Those with proptosis were documented as extra-ocular retinoblastoma. The findings of examination were documented whether they underwent examination under general anaesthesia or not. Data was also collected on the treatment modality given to patients. No pathological results were documented as this service was not readily available at the time of the study.

Data analysis: Data was coded, entered and managed using Microsoft Word Excel and analysed using STATA version 15. Categorical variables were summarized as percentages and frequencies whilst continuous variables were analysed as medians.

Ethical clearance: The College of Medicine Research and Ethics Committee (COMREC) approved the study protocol (P.07/18/2445). To maintain the confidentiality and anonymity of patients, the data was extracted without identifiers and names were replaced with unique identification numbers.

RESULTS

The study population (n=43) had a median age of 31 months (inter-quartile range 3 – 82 months) with a male-to-female ratio of 1:1 and most (n = 20, 46.5%) were referred from a secondary level health facility followed by tertiary hospital referrals (n = 18, 41.9%) with only 11.6% (n = 5) being referred from the primary level of health care. Most patients (n = 37, 86%) arrived to QECH within two weeks of being referred.

Table 1: Patient characteristics and social demographics of those with retinoblastoma at QECH (n=43)

	No.	(%)
Age (months)	31 (3-82)*	
0-6	9	20.9
7-12	9	20.9
13-18	4	9.3
Above 18	21	48.9
Sex		
Male	21	48.8
Female	22	51.2
Referral facility		
Primary	5	11.6
Secondary	20	46.5
Tertiary	18	41.9
Pre-referral treatment		
Yes	8	18.4
No	35	81.4
Arrival time following referral (weeks)		
Less than 2	37	86
More than 2	6	14

*Median (inter-quartile range)

Presenting features: The majority of the population noticed the symptoms after 18 months of age (n=22, 51.1%) and eye involvement was mostly unilateral (n=36, 83.7%). Most of the patients presented with a white lesion (n=29, 67.4%) in the eye followed by eye swelling (n= 19, 44.2%). There were no children that presented with strabismus or buphthalmos. None of the children presented with metastatic disease. The median lag time to presentation was 3 months (inter-quartile range 0.1 – 48 months) with the majority of the children (n=24, 55.8%) presenting within the first 3 months of noticing the symptoms (Table 2).

Table 2: Presenting features of children with retinoblastoma at QECH (n=43)

	No.	(%)
Age symptoms were first noticed (months)		
0-6	7	16.3
7-12	11	25.6
13-18	3	7
>18	22	51.1
Involvement		
Unilateral	36	83.7
Bilateral	7	16.3
White lesion		
Yes	29	67.4
No	14	32.6
Swelling		
Yes	19	44.2
No	24	55.8
Poor vision		
Yes	7	16.3
Not documented	36	83.7
Pain		
Yes	2	4.7
No	41	95.3
Redness		
Yes	4	9.5
No	39	90.5
Lag time (months)	3*(0.1-48)	
0-3	24	55.8
3-6	4	9.3
6-12	9	20.9
>12	6	13.9

*Median

Examination findings and investigations done: The most common examination feature noted was leukocoria (n=32, 74.4%) whilst pseudohypopyon was the least common examination finding (n = 2, 4.6%). A third (32.6%) of the patients had a B-scan done as part of their assessment whilst Magnetic Resonance Imaging (MRI) was performed in only 6 children. Just over half of the study population (n=22, 51.2%) were examined without utilizing anaesthesia (Table 3).

Table 3: Patient evaluation of children with retinoblastoma at QECH (n=43)

	No.	(%)
Leukocoria		
Yes	32	74.4
No	11	25.6
Proptosis		
Yes	13	30.2
Not documented	30	69.8
Pseudohypopyon		
Yes	2	4.6
No	41	95.4
Staging (ICRB)		
Group D	30	69.8
Group E	13	30.2
B-Scan		
Yes	14	32.6
No	29	67.4
MRI		
Yes	6	13.9
No	37	86.1
EUA		
Yes	21	48.8
No	22	51.2

Treatment modalities: Almost two-thirds of the patients had undergone an enucleation surgery as treatment for retinoblastoma (n=28, 65.1%) and a third (n=15, 24.9%) were awaiting surgery. The majority of the population had received pre-surgical or post-surgical chemotherapy (n=40, 93%). Radiotherapy treatment was not available at the facility (Table 4).

Table 4: Treatment given to children with retinoblastoma at QECH (n=43)

	No.	(%)
Enucleation		
Done	28	65.1
Pending	15	34.9
Chemotherapy		
Administered	40	93
Pending	3	7

DISCUSSION

This study reports a median age of 31 months at the time of diagnosis of retinoblastoma. This is similar to findings from other African countries such as Zambia and Kenya [14,15].The male to female ratio of our study population is also similar to the findings from the majority of studies on retinoblastoma where the sex ratio has been reported as 1:1[1,2,6,14]. The majority of the patients were referred from secondary health facilities followed by from tertiary health facilities. 41.9% of the patients being referred from tertiary facilities highlights the need to equip other tertiary facilities in Malawi with human resources and facilities for treating retinoblastoma patients. At the same time, a small percentage of referrals from the primary level (11.6%) points to the need for strengthening

primary eye care to facilitate early detection and referral of retinoblastoma cases from the primary level.

The median lag time of 3 months before presentation was long compared to reports from high income countries where lag times are as short as three days[5,15,16]. However, the lag time was shorter compared to previous findings from DRC, Cote d'Ivoire, Zambia and Kenya where the median lag time ranged from 6.8 months to 9.27 months[6,14,16]. It is important to note that it took less than two weeks for 86% of the patients to present for treatment at QECH Blantyre following referral. This implies that the delay in presentation can be attributed to delays by patients in making contact with the health system and not to delays with referrals within the health system. Some of the patient-related barriers to seeking eye health care in general in low income countries include lack of awareness, long distances to the nearest health facility and high transportation costs[18]. It is thus important to conduct studies to investigate the main barriers to accessing eye care for retinoblastoma patients in Malawi to guide community-level interventions to improve timely detection.

Almost three-quarters of the patients from our study had presented with leukocoria. This is in line with most reports on the commonest presentation of retinoblastoma from Western countries such as United States of America and France where at least half of the cases present with leukocoria[19-21]. This is also comparable to reports from other countries in Sub-Saharan Africa such as Republic of Cote d'Ivoire, the Democratic Republic of the Congo and Kenya, where 70 to 90% of patients with retinoblastoma present with leukocoria.

Leukocoria was followed by orbital cellulitis (37.2%). This is over double (13.2%) what Nyaka reported in 2010[3]. Orbital cellulitis is a rare presenting feature in the developed world. Mullaney *et al*[22] reported 4.8% in Saudi Arabia. In the United States and other developed nations cellulitis is noted in 10% of patients that are diagnosed with retinoblastoma[23].

In our study 44% of patients presented with swelling of one eye. Proptosis is a feature of extra-ocular retinoblastoma and was found in this study to be 30% with an additional 10% reported to have an orbital mass. These findings are similar to Nyaka[3] who reported 44.1% of the patients with swelling of the eye in 2010. It is known, that proptosis is seen frequently in low and middle income countries. A study done in Zambia noted that proptosis occurred in 47% of their patients and was the most common presenting sign[15]. Other developing countries such as Nigeria and Zimbabwe reported 85% and 65% respectively[16]. This was in sharp contrast to what was seen in the developed countries such as USA (0.5%) and Korea (1.4%)[22].

According to the International Classification of Retinoblastoma (ICRB), proptosis is a feature of advanced retinoblastoma which signifies extraocular involvement of the tumour and in our study, about a third of the patients had proptosis. A high percentage of retinoblastoma patients with proptosis has also been reported from other countries in Sub-Saharan Africa such as Zambia, Nigeria and Zimbabwe where 47% to 85% of the patients had proptosis[22]. First-time presentation with extraocular retinoblastoma in low income countries is attributed to delayed presentation[22]. This is in contrast to reports from developed countries such as USA and Korea where only 0.5% to 1.4% of patients with retinoblastoma present with proptosis[22].

The patients that were staged in our study had group D & group E tumours. Due to the advancement of the tumours at presentation, other treatment modalities such as brachytherapy or eye salvaging interventions such as intraocular chemotherapy cannot be offered. Besides lack of their availability.

Due to limited access to facilities for anaesthesia for small children at the time of the study, most patients (51.2%) underwent dilated fundoscopy without anaesthesia. The B-scan ultrasound machine was also not functional at the time and only 32.6% underwent ultrasonography as part of evaluation. In addition, the MRI machine was not functional for most of the study period and only 13.9% underwent MRI imaging. All these three facilities that were of limited availability are important for accurate patient evaluation, tumour grading and monitoring response to treatment[2]. It is important for retinoblastoma centres to have these facilities available to optimise patient management.

Enucleation was performed in 65.1% of the patients. All patients with retinoblastoma at QECH receive the same standard treatment regardless of lag time and staging. This constitutes three cycles of chemotherapy followed by enucleation which is subsequently followed by another three cycles of post-operative chemotherapy (vincristine, etoposide and cyclophosphamide). Although cryotherapy is available, the treatment is not normally given due to advanced disease at the time of presentation. Other treatment modalities such as brachytherapy, radiotherapy and intra-ocular chemotherapy are not available. This is in contrast to western countries where patients have access to more treatment options and that enable globe-salvaging therapeutic approaches[23]. Pathological staging was not done due to the unavailability of a pathologist at the public hospital at the time of the study. A collaboration with the paediatric oncology team has enabled tissue samples to be sent to a paying facility for histological confirmation and staging. Those that present to hospital with metastatic disease and given palliative chemotherapy which consists of 6 cycles of chemotherapy followed by enucleation. Then another 3 cycles of chemotherapy is given. Radiation therapy is still not available in the country.

As Malawi plans to develop retinoblastoma services in the country, collaboration with local experts in other disciplines such as paediatric oncology, radiology and pathology is paramount. At the same time, international multidisciplinary collaborations such as with the

retinoblastoma network offer an opportunity for training in newly introduced treatment options, strengthening referral pathways and research on interventions that can improve treatment outcomes[21].

From our study findings, it is however not enough to improve the diagnostic and therapeutic facilities for retinoblastoma at tertiary level without effective interventions to promote early detection at primary level. There is thus a need for studies on the most effective models for health promotion at community level to increase community awareness on the signs of retinoblastoma and promote earlier presentation. At the same time, there is need for research into the primary eye care interventions that would improve detection at primary level in Malawi.

A major limitation of our study is the retrospective nature of the study where some medical records had incomplete details to enable accurate reporting on clinical features such as tumour staging. However, we were able to demonstrate the long lag time to presentation for retinoblastoma patients and areas that can be improved in the health system to improve retinoblastoma outcomes.

CONCLUSIONS

The median lag time between detection of the first symptom and visiting the retinoblastoma treatment center was 3 months. Most of the delay in presentation was at the level of the patient before the first contact with the health system. There is a need to improve treatment options for retinoblastoma whilst also improving community awareness and early case detection at the primary level of health care.

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Competing interests: The authors declare that they have no competing interests.

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Outcomes of autologous blood conjunctival graft for pterygium surgery at Mbarara University and Referral Hospital Eye Centre and Ruharo Eye Centre

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ABSTRACT

Background: Pterygium management in its advanced stages needs surgery and conjunctival grafting to minimize recurrences. Traditionally, conjunctival grafts were being sutured. Newer techniques such as attachment of the graft with autologous blood are becoming prevalent and outcomes are comparable to other methods of grafting. A few autologous blood coagulum grafts had been performed in Mbarara University and Referral Hospital Eye Centre (MURHEC) and the outcomes were not well known.
Objective: The study determined the average duration of surgery using autologous blood conjunctival grafts, stability of the graft and associated discomfort post operatively.
Methods: A case series study with 19 eyes that received sutureless autologous blood coagulum grafts on the bare sclera was conducted. Surgery duration was timed from time of speculum insertion to speculum removal. Patients were assessed on days 1 and 14 for discomfort and graft stability.
Results: Nineteen eyes received autologous blood coagulum grafts following their pterygium excision. The mean age of participants was 37 (20-55) years. The average duration for the surgery was 31 (25-40) minutes. Thirteen (68.4%) eyes had their grafts adherent on all corners on their first post-operative day. By the 14th day post operatively, 94.4% had their grafts well secured. One (5.3%) eye had graft displaced on 3 sides and it was reattached with sutures. Five participants (27.8%) had minimal graft dehiscence nasally which later closed.
Conclusion: Autologous blood coagulum takes a shorter duration, has stable grafts and with minimal post-operative discomfort. The procedure is equally effective in securing the graft however some patients may have medial dehiscence.

Key words: Autologous Blood Conjunctival grafts (ABC), Pterygium, Graft stability, Outcomes

INTRODUCTION

Pterygium prevalence increases geographically towards the equator between Latitude 40^0N and 40^0S. The association is even greater in people exposed to outdoor environments[1]. Pterygium is initially asymptomatic and later as it advances, it is associated with cosmetic disfigurement, itching, recurrent inflammation, visual impairment, diplopia from motility restriction, and difficult to wear contact lens. Lesions larger than 3.5mm onto the cornea are likely to be associated with greater than one Diopter astigmatism, tear film instability, and can grow to obliterate the pupil. These effects warrant the excision of the pterygium to eliminate the induced symptoms. Conjunctival growth excision with grafting minimizes this risk of recurrence[2,3].

Use of sutures which is commonly used is technically difficult with a long learning curve, prolonged surgery duration, pronounced post-operative discomfort, expensive sutures, increased hospital visits most associated with suture irritation. One Vicryl suture costs around 34 US$ and it is used on a single person. Fibrin glue is costly, approximately 14 US$ per patient and not readily availability in most centers[4]. Autologous Blood Coagulum (ABC) grafting is not commonly used despite its low cost, simple learning curve, short surgery duration, better post-operative comfort and there is insufficient data on its outcomes to support its extensive use as a preferred method.

Autologous blood coagulum grafting is a simple procedure that can be done by any ophthalmic surgeon including ophthalmic clinical officers. It is easy to learn and there is no experience and technical skills needed unlike use of sutures. This study sought to introduce the use of autologous blood coagulum to secure a conjunctival autograft for routine pterygium operations and we wanted to describe the surgical outcomes at MURHEC and Ruharo Eye Centre.

MATERIALS AND METHODS

Ethical considerations

Approvals were sought from the Departments of Ophthalmology (MUST), Faculty Research and Ethics Committee (MUST), Institutional Ethical Review Committee of Mbarara University of Science and Technology (Ref MUREC 11/12-20). All participants consented. All data generated from both hard copy consents and participants' results were stored in key and lock.

Study design and setting

This was a hospital-based case-series study conducted in two specialized ophthalmology hospitals. Mbarara University and Referral Hospital Eye Centre (MURHEC) and Ruharo Eye Centre (REC) are specialized eye hospitals in South-Western Uganda. They are both located in Mbarara Municipality about 5km apart.

Data collection

The study enrolled 19 eyes from 17 adult participants who were eligible that underwent pterygium surgery and received autologous blood conjunctival grafting at MURHEC and Ruharo Eye Centre in the months of December 2020 to May 2021. Pregnant women, suspicious lesions of malignancy, infected eyes, temporal pterygium and those with recurrent pterygia were not included in the study.

A questionnaire was administered to collect data and full eye examinations performed on all participants. A pre-tested structured questionnaire was used to collect data.

Examinations included visual acuity using a Snellen`s projection chart, the anterior and posterior segment examination was performed using a slit lamp and 90 D lenses. Pterygium was graded depending on the extent of corneal involvement. Grade I - crossing the limbus, Grade II - midway between limbus and pupil, Grade III - reaching up to pupillary margin, Grade IV - crossing pupillary margin[5]. Sections of the questionnaire were filled both preoperatively, intraoperatively for surgery duration and any intraoperative complications were documented. Postoperatively, the participants were reviewed in the clinic on day 1 and at 2 weeks later, following a routine follow-up procedure for extraocular surgery. At each visit, the VA was taken, full eye examination done. Stability of the graft and post-operative discomfort were assessed. Graft stability diagnosis was based on attachment of the graft onto scleral bed and conjunctival edges.

Surgical technique

The procedures were done by the same investigator under an operating microscope using the same technique. Sterile conditions following standard operating procedures were followed on all cases. All surgeries were performed under local anaesthesia using a 2% lignocaine given either retrobulbar or peribulbar. All procedures were done under the supervision of a specialist ophthalmologist. The surgeon (investigator) had undergone training and supervision from the ophthalmologists with experience in the method and had the required competence and experience to perform the procedure. A speculum was inserted and maintained in place to enhance exposure for the entire length of the surgery.

Sub-conjunctival and sub-pterygial 0.25ml lignocaine solution was injected. The neck of the pterygium was then lifted up with the help of fine-toothed forceps. The body of the pterygium was dissected 4mm from the limbus, down to the bare sclera, and reflected over the cornea. The pterygium head and cap were avulsed using dissection forceps by maintaining consistent fine traction followed by careful crescent blade excision of corneal remnants. Only the thickened portions of the conjunctiva and the immediate adjacent and subjacent Tenon's capsule showing tortuous vasculature were excised. Care was taken to avoid conjunctival plica excision and extensive dissection of tenons. Haemostasis was allowed to occur spontaneously without the use of cautery. In case of excessive bleeding by a vessel, fine glass rod cautery was applied carefully to avoid extensive burnt area. Saline flushes were not used. Cotton buds were used to remove the excess haemorrhage. The cornea was kept moist by applying saline-soaked cotton buds throughout the procedure. If enough blood was not available to provide autologous fibrin, small perforating veins and capillaries were purposely fractured (though seldom required) to encourage a thin layer of fresh blood to cover the bare sclera. The size of the bare sclera defect (mm) was measured with Castroviejo calipers.

0.25ml of lignocaine was injected subconjunctival on the donor site. Careful dissection between donor graft conjunctiva and Tenon's layer was done while fashioning the 0.1mm oversized conjunctivo-limbal graft from the superotemporal bulbar conjunctiva. The limbal edge of the graft was carefully positioned at the host limbal tissue edge. No attempt was made to directly close the full extent of the excision wound, allowed natural graft positioning without tension.

The scleral bed was being viewed through the transparent conjunctiva and to ensure residual bleeding does not relift the graft, small central haemorrhages were given tamponade with direct compression using non-

toothed forceps muscle hook or non-irrigating Vectis until haemostasis was achieved, usually within 8–10 minutes. The stabilisation of the graft was tested with a cotton bud centrally and on each free edge to ensure firm adherence to the sclera. This was similar to a method used by other studies[1,6-8].

Ocular chloramphenicol ointment was applied in minimal amounts to cover the cornea on the half away from the graft before the eyes were covered with a cotton gauze pad and secured with plaster. The dressing was removed the following day after 24hrs without replacement for initial examination.

Surgical time was noted from the time of speculum insertion up to the speculum removal. Time was recorded in minutes. No assistant was used to eliminate a confounder on duration of surgery.

Post operatively, all eyes were treated with dexamethasone/gentamicin combined eyedrops 4 hourly a day for 2 weeks and prednisolone acetate 1% drops continued for a further 4 weeks. Patients also applied HPMC 3 times a day in the operated eyes. Patients were reviewed on day1 and day 14. For the one participant whose graft did not adhere to the recipient site intra operatively, the graft was sutured and this participant was dropped from the study.

For grafts that were displaced post-operatively, they were taken back for suturing. Amethocaine drops were used for anaesthesia, and patients were allowed home to continue care. No eye patching was necessary. They received dexamethasone/gentamicin combined eye drops 6 times a day for 2 weeks and prednisolone acetate 1% eye drops for a further 4 weeks. Sutures were removed after 7 days or 14 days depending on the patient's convenience to come earlier.

Data analysis

Data from 19 eyes was analysed manually for frequencies. Surgery duration was analysed for the mean.

RESULTS

Objective one: Duration of surgery

A total of 19 eyes from 17 patients were enrolled in the study, 18 from MURHEC and 1 from Ruharo eye centre.

Table 1: Time taken in minutes to perform entire procedure

Time taken (minutes)	Frequency
<30	6
≥30	13
Total	19

The average duration for the surgery was 31 (25-40) minutes from the time of speculum insertion until speculum removal.

Objective two. Stability of the graft

Table 2: Status of the graft on day 1 and day 14 post operatively

Status of graft	Day 1	Day 14
Adherent all 4 corners	13 (68.4%)	17 (94.4%)
Displaced 1 side	5 (26.3%)	1 (5.6%)
Displaced 2 sides	0 (0.0%)	0 (0.0%)
Displaced 3 sides	1 (5.3%)	0 (0.0%)
Graft lost	0 (0.0%)	0 (0.0%)
Total	19 (100%)	18 (100%)

Thirteen patients (68.4%) had their grafts adherent on all corners on their first post-operative day. Up to 94.4% had their grafts well secured by 14th day post operatively and beyond.

Objective three: Post-operative discomfort

Table 3: The presenting symptoms on day 1 and day 14 post operatively

Nature of abnormal sensation	Day 1	Day 14
Foreign Body (FB) sensation	13 (41.9)	6 (19.4%)
Itching	3 (9.7%)	3 (9.7%)
None	3 (9.7%)	20 (64.5)
Pain	6 (19.4%)	1 (3.2%)
Tearing	5 (16.1)	0 (0.0%)
Red	1 (3.2%)	0 (0.0%)
Photosensitivity	0 (0.0%)	1 (3.2%)
Total	31 (100%)	31 (100%)

Foreign body sensation was experienced by 13 (41.9%) patients and was the commonest symptom on day1 and had halved by day14. Six (19.4%) patients reported pain as their symptom on day 1.

Table 4: Level of pain perceived by participants

Pain scale	Day 1	Day 14
None = no pain at all	0 (0.0%)	14 (77.8%)
Very mild= has pain but easily tolerated	12 (63.2%)	3 (16.7%)
Mild = has pain causing some discomfort	5 (26.3%)	0 (0.0%)
Moderate = has pain causing discomfort that interferes with usual activity or sleep	2 (10.5%)	1 (5.6%)
Severe = has pain that completely interferes with usual activity or sleep	0 (0.0%)	0 (0.0%)
Total	19 (100%)	18 (100%)

Twelve (63.2%) patients had very mild pain on day 1 post-operatively and of these, only 3 (16.7%) still had mild pain at the 14th post-operatively.

DISCUSSION

To prevent the recurrence of pterygium, many surgical techniques have been developed. The "ideal" pterygium management is still an ongoing debate because there is no technique regarded as a gold standard and no single method of surgery has proved as superior. In this study, 19 eyes were enrolled for autologous blood coagulum conjunctival grafts with primary nasal pterygia ranging from grades 1 to 3.

Surgical time

In this study, the average duration for the surgery timed from speculum insertion to speculum removal upon completion of the procedure was 31 (25-40) minutes. Autologous blood grafting takes a shorter duration compared to suturing since it avoids the tedious process of suturing under a microscope with instruments. On the other hand, suturing needs skill of good hand coordination and it has long learning curve, complex to learn and one has poor outcomes initially. Suturing and autologous blood coagulum grafting share similar procedural steps initially but suturing takes additional time performing the skillful procedure using instruments under a narrow field of view of the microscope. This effect of extended time with suturing is even felt more when the person suturing is not experienced where it may take more than 90 minutes. Autologous blood grafting saves time further and costs in the long run because there will be no repeated visits to the clinic and time spent removing sutures. This study duration was comparable to 28 minutes observed in another study[9]. It was significantly shorter than 44.8 minutes observed by Kumar and Singh, 2019[20]. Pterygium surgery with the use of sutures took an averagely longer than 40 minutes as reported in some studies. In different studies in different years, there was a significant difference between sutured group and the autologous blood group in terms of surgical time[10,11]. A report done in the year 2018 showed suture surgery took an average of 33.73 min[12]. Although it was averaging our time, it is associated with extra costs and longer learning curves.

Stability of graft

The majority of the patients had their grafts adherent on all corners on their first post-operative day. Up to 94.4% had their grafts well secured by the 14th day post operatively

and beyond. This showed that autologous blood facilitates adherence of the graft on the scleral bed.

Using autologous blood coagulum conjunctival grafts creates even tension across the whole graft interface and there is no direct tension on the free graft edges as with sutures. This minimizes the stimulus for subconjunctival scar tissue formation[6]. A few millimeters of dehiscence was initially seen in 26.3% that later was covered by conjunctival growth by 14th day. Owing to the fact that the graft is not tethered at its edges and when contraction ensues during the healing process, there may be small graft dehiscence of about half a millimeter in some individuals. This is usually well tolerated and does not need further intervention. It is assumed that harvesting a graft that is slightly larger than the area to be grafted by about half a millimeter will cater for this. This was thought to be due to under-perfusion of the scleral bed[14]. Similar studies had noted dehiscence[5,15]. This study observed closer rates to those observed by Ghoz *et al.*, 2018[14], who noted a 33% gap between the graft and conjunctiva. In this study, patients did not lose grafts possibly because we instructed and encouraged our participants not to rub their eyes and also we used a smaller sample size. This finding is in contrast to what was observed in other studies[3,16,17] who observed graft loss. The graft loss and dehisce was attributed to vigorous eye rubbing.

Post-operative discomfort

This study observed foreign body sensation as the commonest symptom on day 1 which had halved by day 14. The initial feeling could be attributed to the edematous graft, raw cut surfaces both on the donor site and recipient site and unevenness of the different conjunctival surfaces. The fast resolution of this symptom is due to resolution of the edema, growth of the graft to cover the edges and healing of the donor site. It could also be due to the effect of medication which favours resolution of the inflammation. By day 14 post operatively, most (73.7%) participants had no symptoms. Such similar observations were seen in studies by Bhatia *et al.*, 2017[2]. In studies that compared suturing to autologous blood grafts, there was a statistically significant difference in the two groups about scores of the postoperative foreign body sensation, pain, epiphora[18]. Many other studies that compared autologous grafts and sutures showed that sutures were associated with prolonged post-operative patient discomfort[10,19].

Limitations of the study

(i) The study population and follow-up time were relatively small in our study.
(ii) No blinding technique was used in the analysis of postoperative discomfort and pain, this was subjectively assessed by the operating surgeon.

CONCLUSIONS

(i) The average surgery duration using autologous blood grafts was 31 minutes, shorter than other studies have shown with suturing.
(ii) The grafts in this study were stable, there were slight dehiscences in some grafts which were well tolerated and eventually covered the defect. The cosmesis was better than the initial presentation.
(iii) Patients experienced less post-operative discomfort related to autologous *in situ* blood coagulum grafts. There were no adverse events observed with the use of autologous blood grafts, the procedure was relatively safe.

RECOMMENDATIONS

(i) This technique should be used for grafting after pterygium surgery because it saves time and it is cost effective. It is suitable for grafts which have a reduced surface area. Large areas for grafting were not studied.
(ii) Care should be taken to harvest slightly larger grafts (1mm) to cater for graft contraction that happens post operatively.
(iii) An adequate explanation should be given to the patient post-operatively to avoid rubbing the eyes while using these techniques. We did not lose grafts because we encouraged our patients not to rub eyes.

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Competing interests: The authors declare that they have no competing interests

Ethical approval: This study was approved by the Mbarara University Research Ethics Committee (MUST REC).

Informed consent: We confirm that all eligible participants or their caregivers provided consent to participate in this study and that the study obtained approval from the Mbarara University Research Ethics Committee

Consent for publication: All authors consented to have this work published including photos.

Availability of data: The datasets used during the current study are available from the corresponding author on reasonable request.

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The clinical profile and operative outcomes of adult patients undergoing glaucoma surgery at Queen Elizabeth Central Hospital, Blantyre, Malawi

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ABSTRACT

Objective: This study was conducted to describe the characteristics and operative outcomes of adult patients undergoing glaucoma surgery at the Eye Department at Queen Elizabeth Central Hospital, Blantyre, Malawi.
Methods: This was a retrospective case series review of medical records. Case files of adult patients who underwent glaucoma surgeries between March 2019 and February 2020 were reviewed. The procedures of interest were trabeculectomy, Ahmed glaucoma valve surgery, and combined trabeculectomy and small incision cataract surgery. The primary outcome measure was the percentage post-Operative Intraocular Pressure (IOP) drop with a reduction of 30% or greater classified as a successful surgery. The pre-operative and post-operative median IOPs were compared using Wilcoxon's sign-ranked test. In addition, the Kruskal-Wallis test was used to compare the median IOPs of the three procedures. A multivariate binary logistic regression model was used to analyse possible factors associated with the surgical outcome.
Results: Sixty-two patient records fitting the inclusion criteria were found, reviewed, and analysed. There was a male to female ratio of 2:1. Two-thirds of the study population presented with blindness in one eye, and three quarters had advanced glaucoma. Successful IOP reduction was achieved in 70.9% and 91.9% of eyes on days one and seven post-operation, respectively. There were no statistically significant pre-operative or post-operative factors associated with the outcome identified in this study.
Conclusions: This study shows advanced disease, advanced age and male preponderance among the patients undergoing glaucoma surgery at a tertiary level eye unit in Malawi. Furthermore, the glaucoma surgeries achieved high rates of successful IOP lowering effects comparable to rates from similar studies in other parts of Africa.

Key words: Glaucoma surgery outcomes, Trabeculectomy, Glaucoma drainage device, Intraocular pressure

INTRODUCTION

Glaucoma is the second leading cause of blindness in the world[1]. The number of people with glaucoma is projected to increase to 111.8 million by 2040[1]. In 2020, a global analysis estimated the number of people aged 50 years and above who are blind from glaucoma to be 3.6 million[2].

Africa has the highest prevalence of blindness globally, with 15% attributed to glaucoma[3]. Similarly, local data from a Rapid Assessment of Avoidable Blindness (RAAB) survey in the Southern Region of Malawi reported that glaucoma accounts for 15.8% of blindness[4]. The burden of glaucoma is disproportionately higher in developing countries[5]. This problem is further exacerbated by a lack of disease awareness, poor access to eye care, sub-optimal diagnostic and management tools, as well as socio-economic deprivation.

Patients who suffer from glaucoma experience profound changes in their everyday lives due to functional visual loss, inconvenience, cost of treatment, as well as the side effects of treatment. The absence of symptoms during the early stages is challenging for those affected because they tend to be unaware of any visual impairment until the disease is advanced[1]. This has led to glaucoma being described as the "silent thief of sight"[1].

The main goal of glaucoma management is lowering intraocular pressure (IOP), as this is recognised as the only modifiable risk factor for the development and progression of the condition[6]. Early detection and intervention are crucial in preserving vision[6]. Surgery is usually the preferred form of treatment in Africa, as maintaining lifelong medical therapy is often unachievable[6]. The treatment's success depends on the sustained lowering of IOP and a decrease in the progression of visual loss.

We conducted a retrospective case series review of medical records to describe the clinical profile and operative outcomes of adults undergoing glaucoma surgeries at Queen Elizabeth Central Hospital (QECH) in Blantyre, Malawi. The outcome of interest that determined the success of the procedure was post-operative IOP.

Currently, the success rate of adult glaucoma surgery in Malawi is unknown; hence filling this knowledge gap is crucial in optimising the approach to glaucoma management.

MATERIALS AND METHODS

Setting: This was a retrospective case series review of medical records at the Eye Department of QECH in Blantyre, Malawi, a major referral eye care centre in the southern region of Malawi and one of five units conducting glaucoma surgeries in the country.

Study population: The operating theatre register was used to identify adult patients who underwent surgical treatment for glaucoma from March 2019 to February 2020. The procedures of interest were trabeculectomy, combined manual small incision cataract surgery and trabeculectomy and Ahmed Glaucoma Valve (AGV) implantation. The AGV surgeries were performed without an anti-metabolite agent, while the other procedures of trabeculectomy and combined cataract and trabeculectomy were done using Mitomycin-C (MMC) and 5-Flourauracil (5FU).

Data collection: A total of 76 adult patient case files were identified from the operating theatre register. Of these, 62 (81.6%) paper patient record files were retrieved from the patient records office and analysed in this study. The patient data were extracted using a structured data collection tool from the patient record files. All records belonged to Malawian residents of African origin.

Demographic information was collected from the patient records files. Details on pre-operative assessment such as visual acuity, IOP, CDR, type of glaucoma, previous incisional surgery in the eye to undergo surgery, which eye was operated on and whether the contralateral eye was blind were also recorded. In addition, details on intraoperative findings and post-operative clinical features were recorded. Visual acuity was converted to and graded according to the WHO classification of visual impairment (Appendix 1). The glaucoma was categorised as advanced of the CDR was 0.8 or greater, or they had one eye already blind from glaucoma. The visual acuity had three categories: unchanged, better, or worse. The visual acuity was graded as unchanged if it remained in the same WHO visual impairment category before and after surgery. The visual acuity was graded as worse if it changed to a category with a higher degree of visual impairment after surgery.

Data analysis: Patient data collected was coded, entered, and managed using Microsoft excel. The cleaned data was exported to STATA version 14 for analysis. The primary outcome measure was post-operative IOP reduction at one week. The post-operative IOP reduction was categorised into 'successful' and 'not successful', where success was defined as a percentage IOP reduction of 30% or more. In addition, intra-operative and post-operative complications were recorded and evaluated for association with the success or failure of the glaucoma surgeries. We carried out a bivariate analysis of each predictor variable against the surgical outcome and entered all variables with a p-value <0.25 into the multivariable logistic regression model. We presented results as crude adjusted Odds Ratios (OR), with corresponding 95% Confidence Intervals (CI) and p-values. We used a two-tailed $p<0.05$ to indicate statistical significance. All statistical analyses were conducted using STATA version 14.

Ethical clearance: The study protocol adhered to the tenets of the Declaration of Helsinki and was granted ethical approval by the College of Medicine Research and Ethics Committee (ethics approval certificate number P.11/20/3189). Confidentiality and anonymity of participants were maintained by removing patient identifying markers and collecting data anonymously by providing each participant with a unique study identification number.

RESULTS

Characteristics of the study population: The mean age of the patients was 59.1 years (95% CI: 54.8-63.4) SD ±2.2. The ages of the study patients ranged from 19 to 91 years, and 43 (69.3%) were male.

In the study population, 38 (61.3%) of the 62 eyes had some level of visual impairment prior to surgery. Furthermore, 41 (66.1% 95% CI: 54.0- 78.2) patients had unilateral blindness, that is, having a visual acuity less than 3/60 in the fellow eye, which is graded as blindness according to the WHO criteria for visual impairment (Appendix 1). Three-quarters of the study population had advanced glaucoma, with a CDR of 0.8 or greater. The median (IQR) of the CDR was 0.9 (0.8-1.0). The rest of the population characteristics are shown in Table 1.

Appendix 1: WHO classification of visual impairment

Category	Presenting distance Visual acuity	
	Worse than	beter or equal to
0- mild or no impairment		6/18
1- moderate visual impairment	6/18	6/60
2- severe visual impairment	6/60	3/30
3- blindness	3/60	1/60 or CF at 1m
4- blindness	1/60	PL
5- blindness	NPL	

Copied from the WHO International Statistical Classification of Diseases and Related Health Problems. 10th revision 2016 Chapter VII H54 Blindness and low vision.
CF: Counting fingers
WHO: World Health Organisation
PL: Perception of Light
NPL: No Perception of Light

Table 1: Characteristics of the study population

Variable	Mean age	Median age	IQR
Age	59.1	60.5	50-71
Variable	Frequency	Percentage	95% CI:
Sex			
Male	43	69.4	(57.6- 81.2)
Female	19	30.6	(18.8- 42.4)
Comorbidities			
Diabetes	3	4.8	(0.1- 10.3)
Pre-op vision impairment in operated eye			
Normal to mild visual impairment (6/6 -6/18)	24	38.7	(26.2- 51.2)
Moderate visual impairment (6/24 -6/60)	18	29	(17.4- 40.6)
Severe visual impairment (5/60 -3/60)	10	16.1	(6.7- 25.5)
Blindness (2/60 -1/60)	8	12.9	(4.3- 21.5)
Blindness (HM-LP)	2	3.3	(1.2- 7.7)
Blindness (NLP)	0	0	
Laterality of vision			
Blindness in the fellow eye	41	66.1	(54.0- 78.2)
No blindness in the fellow eye	21	33.9	(21.8- 45.9)
CDR in operated eye (N=55)			
1.0	23	41.8	(28.4- 55.3)
0.8-0.9	19	34.6	(21.6- 47.5)
0.6-0.7	8	14.5	(4.9- 24.2)
<0.6	5	9.1	(1.2- 16.9)
Prior incisional surgery in operated eye			
Yes	14	22.3	(11.9- 32.6)
No	48	77.7	(67.3- 88.1)
Type of glaucoma			
POAG	53	87.1	(78.5- 95.7)
JOAG	7	11.3	(3.2- 19.4)
PACG	1	1.6	(0.6- 4.8)
Operated eye			
Right eye	41	66.1%	(54.0- 78.2)
Left eye	21	33.9%	(21.8- 45.9)

Surgical procedures, complications, and post-operative interventions: All the trabeculectomy and combined cataract and trabeculectomy procedures were done with an anti-metabolite, the majority with MMC. There were seven (11.3%) eyes that experienced nine intra-operative complications.

There were 18 post-operative complications in 11 (17.7%) eyes. Hyphaema was the commonest post-operative complication. Three eyes experienced bleb leaks which required bleb resuturing in the operating theatre. Despite experiencing bleb leaks, these patients maintained a good IOP in the range of 6-10 mmHg. No eye developed post-operative endophthalmitis. The rest of the complications and interventions are shown in Table 2.

Comparison of pre-operative and post-operative vision: The WHO visual acuity grading of 29 (47.5%) of the 61 eyes remained unchanged, while 21 (34.4%) eyes experienced worsening after the glaucoma surgery. Of the 11 patients who experienced improved vision after surgery, six (54.5%) of them belonged to the combined cataract and trabeculectomy group. Figure 1 shows the comparison of pre-operative and post-operative visual outcomes.

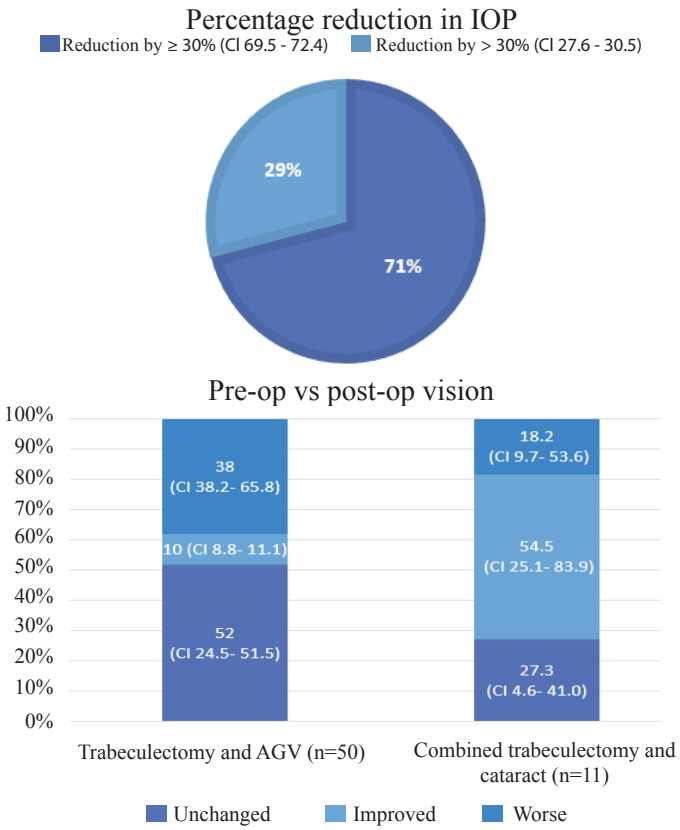

Figure 1: Comparison of pre-operative and post-operative vision andI OP in the study population

Table 2: Surgical procedures, complications and post-operative intervention

Variable	Frequency	Percentage
Procedure performed		
Trabeculectomy with MMC	36	58.1
Trabeculectomy with 5-FU	5	8.1
Ahmed glaucoma valve	10	16.1
Combined cataract and trabeculectomy	11	17.7
Intra-operative complications (n=10)		
Conjunctival buttonhole	3	30.0
Vitreous loss	1	10.0
Iris trauma	3	30.0
PI extension	3	30.0
Post-operative complications (n=18)		
Hyphaema	5	27.7
Hypotony	4	22.2
Flat anterior chamber	3	16.7
Bleb leak	3	16.7
Post-op interventions (n=37)		
Massage	16	43.3
Suture release	8	21.6
Double eye padding	4	10.8
Bleb resuturing	3	8.1
Oral acetazolamide	2	5.4
Intracameral OVD injection	1	2.7
OVD washout	1	2.7
Foreign body removal	1	2.7
Scleral flap revision	1	2.7

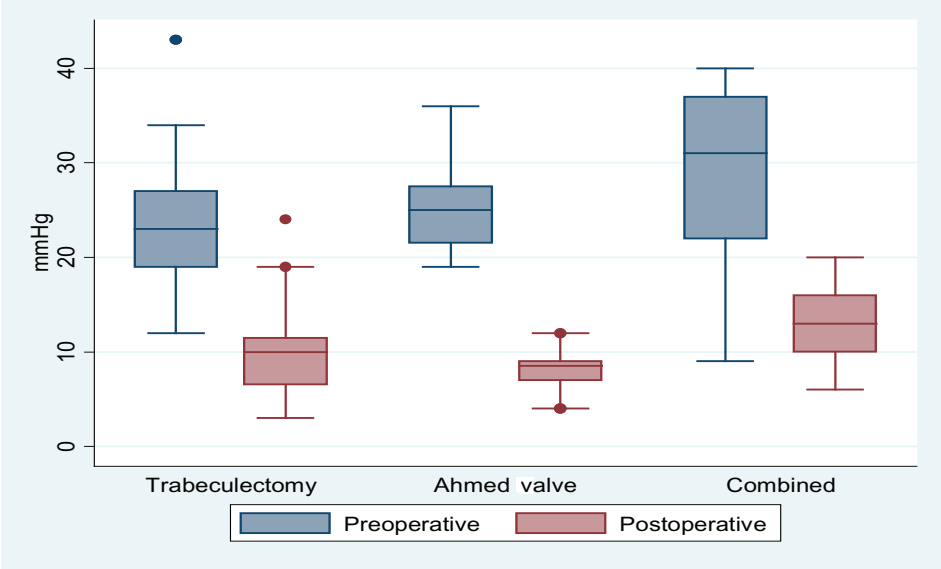

Figure 2: Comparison of pre-operative and post-operative IOP by surgical procedure

Comparison of median IOP: The three surgical procedures resulted in a 60% mean IOP reduction post-operatively. The median pre-operative IOP was 24 (IQR 20-29). The post-operative median IOP was ten on both day one (IQR 8-18) and week one (IQR 7-12.5) post-operatively. The collective pre-operative and post-operative median IOPs were compared using Wilcoxon's sign ranked test. The pre-operative IOP was 25 (CI 23.0-27.0) and the post-operative IOP was 15 (CI 12.0-18.0). A statistically significant IOP reduction was detected between pre-operative and post-operative measurements (mean difference 10.0 CI 6.8-13.3), with a P-value of <0.001.

Comparison of IOP by procedure: Of the three procedures, AGV implantation resulted in the lowest median IOP. Eyes that underwent combined trabeculectomy and cataract surgery had the highest pre-operative IOP with the widest distribution compared to AGV or trabeculectomy surgeries. In addition, AGV produced the narrowest range of post-operative IOPs. This is shown in Figure 2.

A Kruskal-Wallis test was used to compare the median IOPs of the three procedures. The median pre-operative IOP for the trabeculectomy, Ahmed valve and combined cataract and trabeculectomy were 23.9 (21.6- 26.2), 25.4 (20.9- 29.9) and 28.9 (21.9- 35.9) while the post-operative IOP were 10.0 (8.4- 11.6), 7.9 (6.1- 9.7) and 12.5 (9.9- 15.2) respectively. There was a statistically significant difference in the population median IOPs between the three procedures with a P-value of 0.045.

Factors affecting the surgical outcome: A multivariate binary logistic regression model was used to identify possible factors associated with the outcome of the glaucoma surgeries. There were no statistically significant factors that affected the surgical outcome identified in this study.

DISCUSSION

The mean age of the study population was 59.1 years, with the majority (88.7%) of patients being above 40 years of age. Other African studies that analysed glaucoma surgery outcomes have reported mean age in the range of 56-67 years[7–10].

There was a gender disparity among the patients who underwent surgery in our study with 69.4% of the population being male. This is similar to a previous study done at the same study location[11], and is also similar to studies from Nigeria, Ghana, and Tanzania where men comprised 74%, 65% and 72%, respectively[12–14] of POAG patients presenting at eye care facilities. One possible reason for the lack of equity is the interplay of cultural factors in low-income countries like Malawi[15]. For example, women are less likely to be educated and access information regarding health care services. The males are usually the breadwinner and women do not have control over the household income or any influence over decision making in the home[16].

In our study, three-quarters of the patients had advanced glaucoma, with two-thirds of the operated eyes having some degree of visual impairment prior to surgery and 16.2% being blind. Most patients with glaucoma in low-income countries present with advanced disease due to late presentation[12,13]. One reason for late presentation with glaucoma is a low socio-economic status[17]. Other barriers to accessing eye care services in Africa in general include lack of awareness about glaucoma, cultural belief and misconceptions about the cause of blindness, shortage of ophthalmologists, lack of functional referral systems, and inadequate access to eye care facilities[1,3,18,19]. All these factors culminate in an increased burden of avoidable blindness.

Regarding surgical outcomes, 70.9% of eyes that underwent glaucoma surgery achieved a successful IOP reduction one day post-operatively, rising to 91.9% at one week. In comparison, a review article on the success rates of glaucoma surgeries in Africa reported a range from 61.8% to 90%[20]. It is worth noting that these studies had varying follow-up times, with the final follow-up time ranging from 6 to 60 months. Although a longer follow-up time allows for a comprehensive understanding of the success rate of glaucoma surgeries, it is essential to study immediate post-operative outcomes as they are a proxy indicator of the long-term success of the surgical treatment [21,22]. Literature shows that early post-operative IOP reduction is linked to the long-term success of glaucoma surgery[21,22].

The IOP was lower in the trabeculectomy group compared to the combined cataract surgery and trabeculectomy group, similar to other research on combined surgery in managing eyes with cataracts and POAG[23]. However, in the latter, the procedure performed was the phacotrabeculectomy which is considered the "treatment of choice" in glaucoma patients with coexisting cataracts[24]. In contrast, the combined procedure performed in this study population was trabeculectomy and manual small-incision extracapsular cataract extraction.

The AGV surgery produced the lowest mean post-operative IOP. This finding is similar to that reported in the landmark Tube Versus Trabeculectomy (TVT) study. It was found that both trabeculectomy with anti-metabolite and tube surgery resulted in a significant and sustained reduction of IOP, with the latter having a higher success rate. However, at the end of the follow-up period of 5 years, both procedures had similar IOP reduction[25].

In addition, the AGV resulted in a lower range of post-operative IOPs compared to the trabeculectomy and combined cataract and trabeculectomy procedures. While the efficacy of a trabeculectomy can be influenced by a surgeon's experience[26], the AGV contains a standard filtration route with a valve mechanism primed to open and close so that aqueous humour flow is maintained in the range of 8–12 mmHg[27]. This suggests that the AGV may result in more predictable post-operative IOP levels compared to trabeculectomy and combined cataract and trabeculectomy. The AGV devices are not readily available at our hospital, and as such, their use is limited to patients with failed trabeculectomy or secondary glaucomas.

In the trabeculectomy and AGV group 38% of eyes had worsening of visual acuity. This occurrence in the early post-operative period has been reported in other studies[28,29], with some improvement to baseline vision seen on follow up. In addition, 52.0% of the operated eyes had the same level of visual acuity before and after surgery which is a favourable outcome since the primary goal of surgery is to stabilise visual acuity and disease progression by lowering IOP. It is imperative that the patient, through careful counselling understands expected outcome, minor risk of post-operative visual decline and the high risk of blindness without surgery[6].

An improvement in visual acuity was seen in six (54.5%) of eyes that underwent the combined cataract and trabeculectomy procedure. Similar findings were reported in a study done in Eastern Africa that assessed the visual and IOP outcomes in combined cataract and trabeculectomy surgery[7]. Visual outcomes were encouraging, considering the advanced glaucomatous optic neuropathy. At discharge, there was an improvement in visual acuity in 58 (41%) of 142 eyes compared with the pre-operative measurement; on the other hand, the vision was unchanged in 43 (30%) eyes and was worse in 20 (29%) of eyes.

One limitation of the study was that due to the Covid-19 pandemic, the number of patients that presented at QECH with glaucoma during the last quarter of the study time period was very low. This resulted in having a small sample of patients with this condition, which may have influenced the non-significant findings obtained in the study. Another limitation is the retrospective nature of the study. QECH uses paper-based system of records, which only contains post-operative data up to 10 days. Our recommendation is that a large-scale prospective study should be carried out to provide a complete understanding of the post-operative outcomes.

CONCLUSIONS

This study found that most patients undergoing glaucoma surgeries at QECH were males and had advanced disease. In addition, this study showed that approximately two-thirds of the glaucoma surgeries performed at the Eye Department of QECH achieve a successful IOP reduction comparable to rates seen in studies published in other parts of Africa. The Ahmed valve glaucoma surgery was the most successful surgical procedure for lowering IOP.

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Availability of data and materials: The datasets used during the current study are available from the corresponding author on reasonable request.

Competing interests: The authors declare that they have no competing interests.

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Juvenile retinoschisis in a 4 year old boy: case report

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ABSTRACT

X-Linked Juvenile Retinoschisis (XLJR) is a rare congenital disease of the retina in children with a prevalence of between 15,000 to 30,000. It almost always occurs in males and is characterized by poor vision due to resultant macular degeneration. We present a case report of this condition in a 4 year old boy whose only presenting symptom was poor vision. He had concurrent mixed astigmatism which initially masked this diagnosis. Failure of the poor vision to improve despite spectacle correction prompted further evaluation. The definitive diagnosis was made through ocular imaging using retinal photography and Optical Coherence Tomography (OCT). High index of suspicion and close follow-up is critical to point to rare cases that need further investigation in unexplained visual symptoms in children; which can be masked by common conditions such as refractive errors and amblyopia.

Key words: Juvenile retinoschisis, X- linked retinoschisis, Juvenile foveoschisis, Juvenile macula degeneration, Poor vision in children

INTRODUCTION

Juvenile retinoschisisis caused by mutations in the RS1 gene, which encodes retinoschisin, a protein involved in intercellular adhesion and likely retinal cellular organization[1]. It is present at birth but usually present at school age due to increased visual demands. The purpose of the case report is to highlight this rare retinal pathology in the paediatric population, the challenges of making the diagnosis in our set-up and to emphasize the importance of patient follow up.

CASE REPORT

This is a case report of a 4 year old male child who presented at a paediatric eye centre in Nairobi with poor vision at both near and far distance for unspecified long duration. He had difficulties in reading and writing at school. There was no variability between day and night vision nor any associated ocular or systemic symptoms.

Initial evaluation of the child revealed visual acuity of 6/36 in both eyes. Cycloplegic refraction confirmed significant mixed astigmatism of +3.00/ -3.00 x180 in both eyes. He had normal anterior segment and unremarkable fundus findings on indirect ophthalmoscopy in both eyes. Spectacle prescription was given. On the first follow-up review after 3 months, the child was still struggling to read at both near and far distance and the visual acuity had only improved by one line to 6/24, despite the spectacle prescription and reported compliance. We queried amblyopia from the uncorrected astigmatism but still ordered further ophthalmological work up because of the marked straining that was noted on attempt to read near work. Corneal topography to assess for keratoconus was normal. Positive findings were found in the retina in both eyes, with fundus photography showing characteristic spoke wheel pattern radiating from the fovea, typical of juvenile retinoschisis (Figure 1a and 1b). Optical coherence tomography also revealed characteristic foveal schisis in juvenile retinischisis involving the superficial neural retina and thinning of the retina (Figure 2a and 2b). The child was immediately put on topical dorzolamide and referred for low vision management. He was advised on environment modification at home and school in addition to the spectacle correction in order to optimize the visual function. He is also on close follow up with the paediatric ophthalmologist and the retina specialist for close to two years since the diagnosis. The vision has remained the same and the serial fundus and optical tomography images have remained stable as per the latest review. The authors have received consent from the parents of the child and permission to publish the case report from Eagle Eye Diagnostic & Laser Centre, Nairobi, Kenya where the patient is being managed.

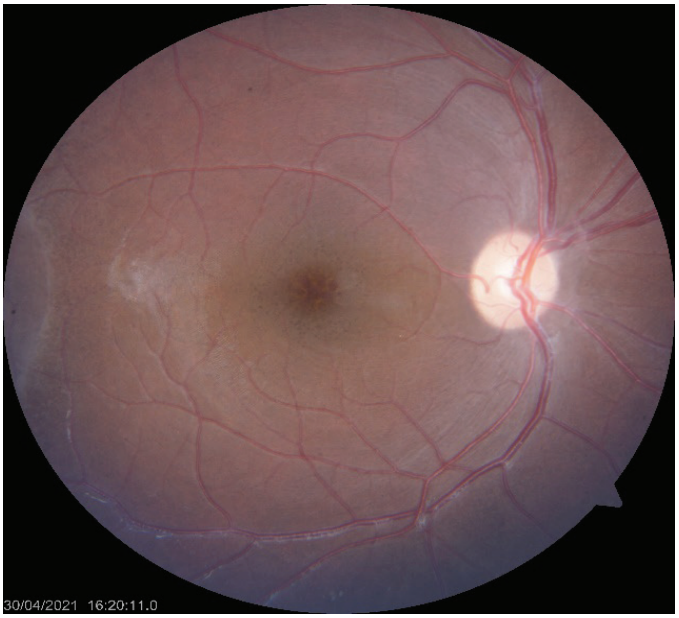

Figure 1a: Fundus photo right eye

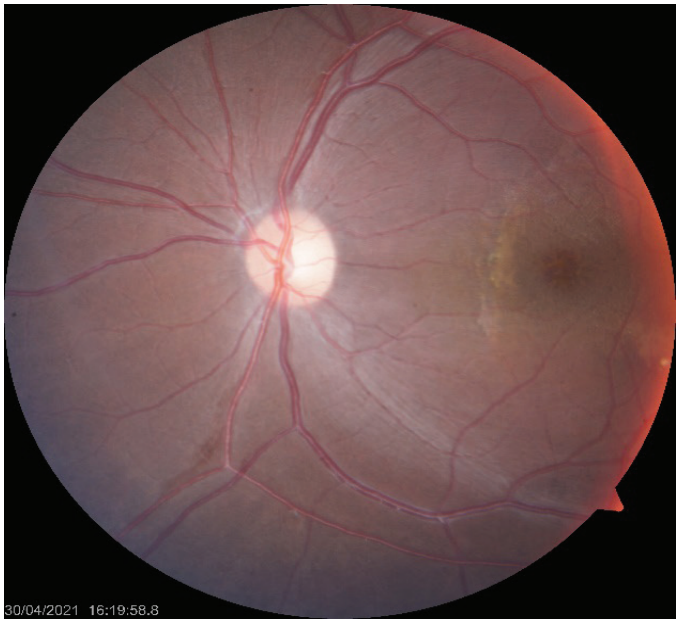

Figure 1b: Fundus photo left eye

Figure 1a and 1b: Characteristic spoke wheel pattern radiating from the fovea for the right and left eye respectively

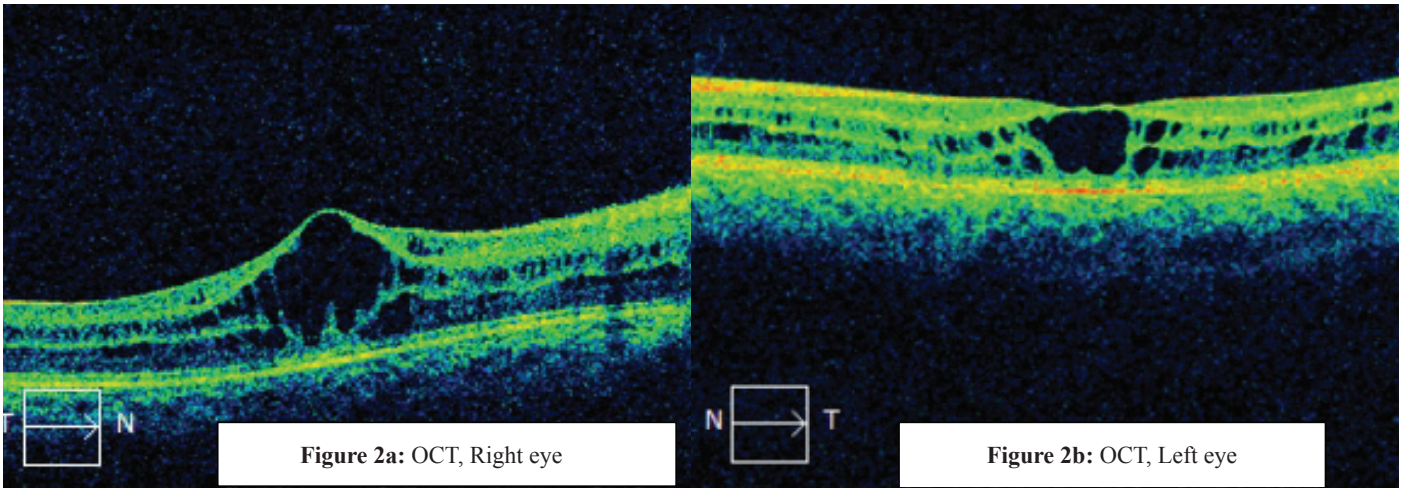

Figure 2a: OCT, Right eye

Figure 2b: OCT, Left eye

Figure 2a and 2b: OCT: Cystic spaces primarily in the inner nuclear and outer plexiform layers in the right and left eye respectively

DISCUSSION

Juvenile retinoschisis is one of the main causes of juvenile macular degeneration in males. It mostly presents in school going children with poor vision, as was the case in our patient. Rarely, may it occur in younger children causing nystagmus and strabismus[2].

In childhood the findings can be very subtle, and the diagnosis can easily be missed or confused with amblyopia. The best way to view the characteristic features is by use of a slit- lamp, contact lens and the red free filter[1]. This could explain why it was not possible to pick the findings during the initial clinical fundus examination which was performed using indirect ophthalmoscope and a 20 dioptre lens. In addition, picking the fine retinal changes in a child can be challenging. The characteristic symmetric spoke wheel pattern radiating from the fovea is seen on fundus examination and photography[3].

The criteria used for the clinical diagnosis in absence of genetic testing in our patient were; male patient, presentation in the first decade, characteristic symmetric macular involvement on digital photography and hypo reflective cavities on Optical Coherent Tomography (OCT). OCT is particularly useful in detection of the lamellar schisis which are normally not visible on clinical examination. It also enables detection of splitting of other layers of the retina in addition to the nerve fibre layer. Peripheral retinoschisis occurs in 50% of the patients[4,5]. This had not been observed in this patient as yet. Other useful tests that can be performed to support the diagnosis include; Fundus autoflorescence which reveals increased autoflorescence that highlight the areas of foveal schisis. Fluorescein angiography shows non- petaloid leakage unlike that seen in cystoid macula oedema, with pooling of dye in schisis cavities at late phase. On standard full-field electroretinogram (ffERG) there is reduced b-wave

with preserved a-wave (negative waveform). This ERG finding is not specific to XLJR and can also be observed in other retinal diseases. Genetic testing to confirm the RS1 mutation can be done[1]. The genetic testing was not feasible in our patient.

Management of this condition is by a retinal specialist and/ or paediatric ophthalmologist. Oral or topical carbonic anhydrase inhibitors such as acetazolamide and dorzolamide are effective in flattening the cysts[6-8]. Eventually the cysts gradually reduce by adolescence in the absence of any intervention[3]. Laser photocoagulation to prevent retinal detachment is recommended, though it has a low risk of inducing the same. Low vision devices and environment adjustments such as appropriate sitting position in class, increasing contrast in the text are useful in enhancing the visual functioning. With the new advances on gene therapy, definitive treatment of the XLRS is possible through gene replacement therapy[9]. Genetic counselling is thus paramount for the patient and the relatives.

Complications that can occur include rhegmatogenous retinal detachment, vitreous haemorrhage and foveal ectopia in approximately 5% of the patients[1,8]. Vision often declines in the first two second decades, then stabilises until the fifth or sixth decade. Further deterioration occurs after this period due to macula atrophy, hence lifelong visual monitoring and rehabilitation is essential[3]. XLRS diagnosed in children is often severe with poor prognosis regardless of the age of the patient at presentation[5].

CONCLUSION

High index of suspicion is needed to diagnose such rare retinal diseases in children, which may be masked by other diagnoses such as amblyopia and refractive errors. Where possible further evaluation should be carried out to investigate unexplained visual symptoms in children.

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Bilateral congenital eyelids eversion: case series and review of literature

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ABSTRACT

Congenital Upper Eyelid Eversion (CUEE) is a rare and benign congenital condition characterized by everted upper eyelids with prominent chemosis. We report two cases of bilateral congenital upper eyelid ectropion; an ophthalmic examination of one of the cases showed asymmetry in the extent of upper eyelid eversion and chemosis. The further paediatric evaluation showed no revealing in the two cases. For the two cases, treatment of the eyelids eversion was conservative, combining topical steroids, antibiotics, and lubricants. Chemosis was reduced progressively. One of the cases reported by the authors had an asymmetric severity and variation in time to respond to conservative treatment, which makes it peculiar to other reported cases of congenital upper eyelid ectropion. In this patient, the upper eyelid with severe chemosis and eversion took extended time. Congenital upper eyelid ectropion is common among black infants, and most infants show excellent anatomic and functional results with conservative treatment if managed timely and promptly.

Key words: Congenital ectropion, Upper eyelid eversions, Asymmetric presentation, Black infants, Conservative management

INTRODUCTION

Congenital Upper Eyelid Eversion (CUEE) is a rare and benign congenital disorder that usually presents at birth and is described as a complete eversion of the upper eyelid associated with swelling, conjunctival prolapse, and severe conjunctival chemosis[1]. In addition, CUEE is associated with blepharophimosis syndrome, colloidal skin disease, and Down syndrome. Moreover, a higher preponderance of the disease is seen in black infants[2-4].

We report two rare cases of bilateral upper eyelid eversion presented to hospital of which one had an asymmetric clinical feature at presentation. Our case report aims to describe a previously unreported asymmetric feature of upper eyelid ectropion. It also highlights the treatment response to conservative management.

CASE REPORTS

Case 1

A 4-hour-old male neonate, the first child to a 26-year-old mother, was referred to our centre from a private clinic where he was delivered on account of protrusions of red mass of tissue from both eyes since birth. Both the pregnancy and delivery were uneventful. The mother had an ante natal follow-up at the same clinic. The baby was born at full term via vaginal delivery without using any instruments. Ophthalmic examination revealed complete eversion of both upper eyelids with severe conjunctival chemosis and hyperemia (Figure 1). The eyeballs were examined by carefully opening the eyelids with a cotton-tipped applicator under topical anaesthesia. Both globes were normal size, and the anterior segment evaluation was unremarkable in both eyes. A further detailed systemic examination was unremarkable.

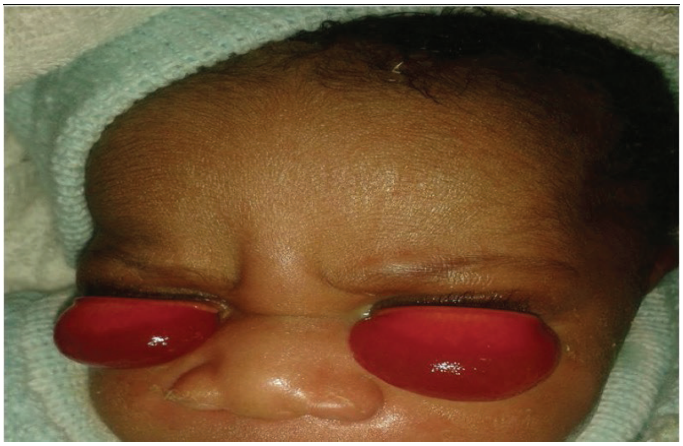

Figure 1: A 4-hour-old child born with complete bilateral eversion of the upper eyelids with severe conjunctival chemosis and hyperemia

Manual repositioning of the eyelids was not successful. Moist dressings with hypertonic saline, a topical antibiotic-corticosteroid ophthalmic ointment, Terra-cortril ointment® (10 mg/mL Oxytetracycline, 5 mg/mL Hydrocortisone Acetate and 10000 u/mL Polymyxin B Sulphate) three times daily and lubricants with Tear Naturale® (dextran 701 mg / ml and hypromellose mg / ml) applied hourly for lubrication, infection prophylaxis, and chemosis reduction as part of a conservative treatment under close supervision. The eyes were patched between

drops. After four weeks of conservative treatment, the upper eyelid's chemosis had sufficiently subsided, the lid naturally flipped (Figure 2), and there was no ocular complication.

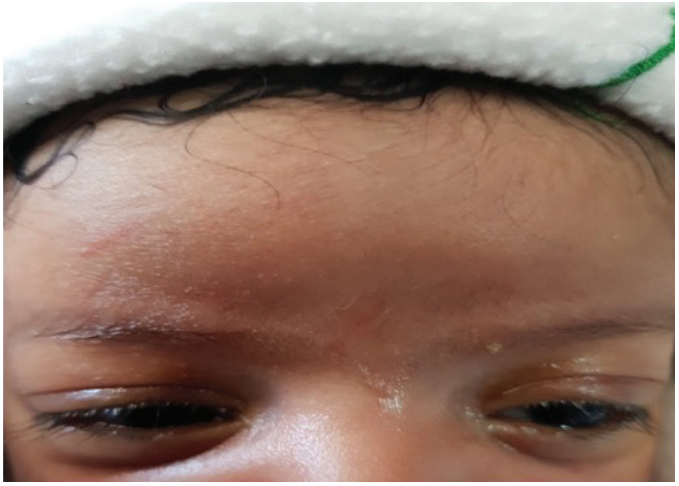

Figure 2: Four weeks after the presentation, the same patient had complete resolution of bilateral upper eyelid eversion and chemosis.

Case 2

A 2-day-old female neonate presented to the emergency ophthalmology clinic with a chief complaint of outward eversion of both eyelids since birth, with the left eyelid intermittently reverting back by itself. She was the fourth child of a 25-year-old woman. Pregnancy was uneventful with regular antenatal follow-ups. The child was full term and was born by normal vaginal delivery, with no instrumentation.

Ophthalmic examination revealed complete of the right upper eyelid and partial eversion of the left upper eyelid with prominent conjunctival hyperemia and chemosis ((Figure 3A).

Figure 3A: A two-day-old infant with fully bilateral everted upper eyelids and chemosis, worse on the right upper eyelid

The eyeballs were examined by carefully opening the eyelids using a cotton tip applicator under topical anaesthesia. Both globes were normal-sized, and anterior segment evaluation was unremarkable in both eyes. Further detailed systemic examination was unremarkable.

Manual repositioning of the eyelids was not successful. Similar management to the first patient was given. After three days, the left upper eyelid's chemosis had sufficiently subsided, the lid naturally flipped, and the right upper eyelid chemosis markedly reduced (Figure 3B).

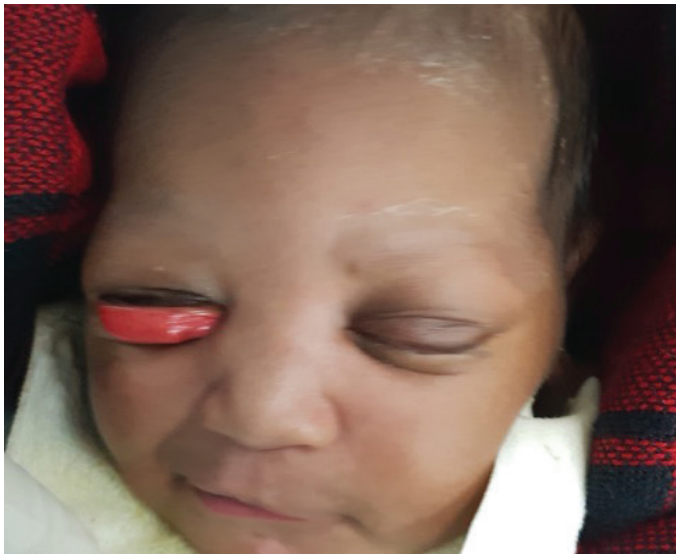

Figure 3B: After three days of topical therapy, resulting in resolution of eyelid edema in the left eye and naturally flipped left upper eyelid and no longer taped.

The same conservative treatment with eye patches was continued to the right eye, which did not improve, until day 18, when the right upper lid began to realign naturally (Figure 4). Two months later, both eyelids were normal, and there was no ocular complication.

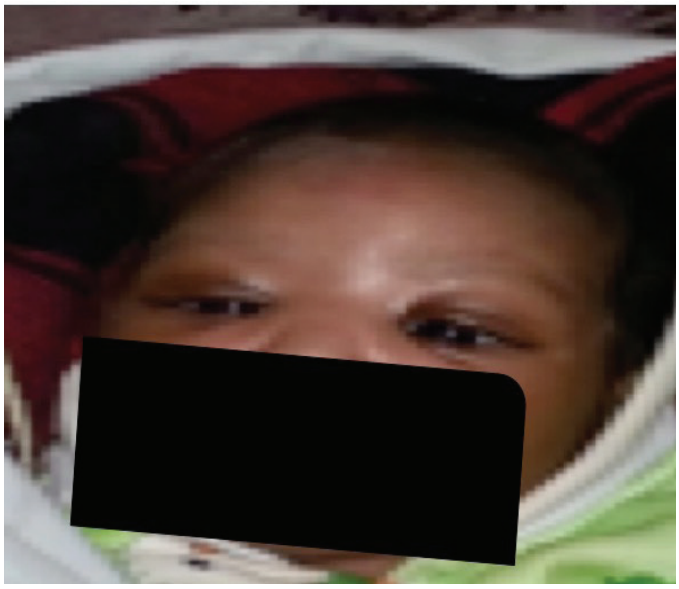

Figure 4: The same patient 5 with complete resolution of bilateral upper eyelid eversion and chemosis

DISCUSSION

Congenital Upper Eyelid Eversion (CUEE) is a rare abnormality, and Adams initially documented it as a case of "double congenital ectropion" in 1896[1]. Congenital Eyelid Imbrication Syndrome (CEIS) and Congenital Floppy Eyelid Syndrome (CFES) are important differential diagnoses for CUEE. CEIS is characterized by upper eyelids overriding the lower eyelids, usually seen in patients with floppy eyelid syndrome[5]. Floppy eyelid syndrome is frequently associated with Down syndrome[6]. In the present cases, no overriding of the upper lid over the lower eyelid and no features of Down syndrome were noted on systemic evaluation.

Bentsi-Ebchill from Ghana reported the greatest number of CUEE cases, 14 in total[7]. The exact aetiology of CUEE is unknown. The absence of a tarsal plate, vertical shortening of the anterior lamella, levator disinsertion, and overlapping of the lower eyelid edge under the upper lid are some of the hypothesized aetiologies for CUEE[7,8]. In addition, the orbicularis spasm may serve as a sphincter once it has everted, creating a cycle of conjunctival strangulation and edema due to venous stasis[4]. However, a histopathologic investigation of a deceased 9-day-old neonate with CUEE failed to reveal anatomic lid anomalies to explain the illness[9].

Rarely, the disorder may be linked to collodion skin disease or Down syndrome[2,10-12]. Otherwise, as seen in Table 1, most cases reported in healthy children were in Africans[3,4]. Birth trauma has been suggested as a potential mechanism for CUEE, even if the actual aetiology is unknown[4,13]. Similar to our case, the majority of the reported cases have been following both normal vaginal delivery[7,14-17].

Table 1: Clinical presentation of reported cases of congenital upper eyelid ectropion

Features	Sayadi *et al*[13]	Ibraheem *et al*[17]	Bentsi-Enchill [7]	Adeoti *et al*[14]	Present
No. of cases	1	1	14	3	2
Age at presentation	4 h	6 days	1h-1 case 6h- 1 case 14h- 1 case 1day-6 cases 2 days-2 case 3 days- 3 case	4 h 3days 4 days	4 h 2 days
Sex	Male	Female	10-male 4-Female	3 Male	1 Male 2 Female
Ethnicity	White	Black	Black	Black	Black
Method of delivery	Vaginal	Vaginal	13 vaginal 1 caesarean	Vaginal-all	Vaginal
Laterality of eversion	Bilateral	Bilateral RE>LE	Unilateral RE-3 Unilateral LE-3 Bilateral -6 Bilateral LE>RE-1 Bilateral RE>LE-1	Bilatearl-2 Bilateral LE>RE	Bilateral Bilateral RE>LE-1
Country	Tunisia	Nigeria	Ghana	Nigeria	Ethiopia
Systemic diseases	Umbilical hernia	None	None	None	None
Treatment	Topical Lubrication, Antibiotics and Application Hypertonic Saline	Topical lubrication, antibiotics and application hypertonic saline	Lid suture Subconjunctival hyaluronidase injection Antibiotic treatment	Topical lubrication, antibiotics and application hypertonic saline	Topical lubrication, antibiotics and application hypertonic saline
Recovery	21 days	5 Days	1-2 days	8th day-1case 10th day-2 cases	2 weeks

Like in our cases, the disorder is frequently present from birth[1,17-20]; however, there have been recorded cases of later presentation[10]. It is clinically defined by the protrusion of the edematous upper lid conjunctival fornix leading to upper eyelid eversion[1,18,19].

Although unilateral involvement has been noted[7,15-19], most publications describe bilateral involvement of the upper lids. Bilateral asymmetric presentation is rare and these cases were seen in black Africans like our cases (Table 1).

The role of surgical interventions and injections was only limited in studies published earlier[7] or in cases of CUEE with Down syndrome[12]. Conservative management with local antibiotic ointment, hypertonic saline, lubricants, and moist pressure with manual repositioning of the eyelids has been reported to resolve CUEE in a few days to weeks [2,16,17,19,20-22]. In the current cases, similar to the above reports, the patient responded to conservative management. The left upper eyelid, which had partial eversion, responded in 2 days, whereas the right upper eyelid, which had total eversion, took three weeks to respond fully.

CONCLUSIONS

CUEE is a rare eyelid disorder of uncertain cause. As seen in our cases, black infants with CUEE have no other associated systemic disorders. Black infants with CUEE rarely present with asymmetric CUEE, and most infants with CUEE usually show excellent anatomic and functional results with conservative treatment. These case series may expand the current recognition and understanding of this rare entity.

Consent: Written informed consent was attained from the patient's parents to publish this case report. This report does not contain any personal identifying information.

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Conflicts of Interest: The authors have no conflicts of interest.

Authorship: Both authors attest that they meet the current ICMJE criteria for authorship.

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Perspective

Social entrepreneurship eye care model in Africa: A case of Kisii Eye Hospital, Kenya

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Many definitions have emerged to describe what social entrepreneurship stands for. A generic one goes 'Social entrepreneurship is the process by which individuals, startups and entrepreneurs develop and fund solutions that directly address social issues'[1]. Mohammed Yunus in his book - *Building a Social Business*, described it as a new kind of capitalism that serves humanity's most pressing needs. Therefore, the motivation for starting a social enterprise is not, so that you can be your own boss or to build an empire as it is traditionally, but rather to address a social problem that is bothering your conscience. Social entrepreneurs are individuals who identify a social problem and apply entrepreneurial spirit, business insight, and leadership to solve the problem. These are people with new ideas to address major problems, who are relentless in the pursuit of their visions, people who simply will not take "no" for an answer, who will not give up until they have spread their ideas as far as they possibly can[2-4]. Bill Drayton, the founder of Ashoka described social entrepreneurs as people who cannot just give fish or teach how to fish but rather the ones who cannot rest until they have revolutionized the fishing industry.

Getting closer home, as eye care providers we have in our hands the big burden of blindness in Sub-Saharan Africa (SSA) that has eluded the eye care providers over the years. It is a big social problem and as Prof. Allen Foster puts it – we need to be emotional. Emotions that will lead to action. The government eye care services have been in existence for several years and with support from development NGOs have made great attempts to address this problem. The universities and colleges have produced eye care workers, done research and outreach programs. Through these a lot of advances have been made in various fields of eye care in SSA. However, using parameters like Cataract Surgical Rates (CSR), and Cataract Surgery Outcome Monitoring (CSOM) it is evident that Africa is performing way below what is expected in terms quantity and quality of eye care service delivery[5-7]. It is in this backdrop that a new thinking is imperative. Social entrepreneurship has revolutionized eye care in other regions like India[2,3]. Can it do it for Africa?

The author founded Kisii Eye Hospital in 2013 as a comprehensive eye hospital that runs as a social enterprise with a mission to offer high quality, affordable and high-volume eye care services in the rural population of Southwestern Kenya. Prior to this he was an assistant professor and head of ophthalmology in Aga Khan University Hospital, Nairobi. The main motivation to start this model of hospital is the perpetual backlog of blindness especially attributed to easily and completely treatable conditions like cataracts and refractive errors despite all the efforts that were seemingly being made. The notion is that an African ophthalmologist who lives and works in Africa with firsthand experience of the blindness problem is best placed to come up with a solution and offer leadership to a program that will tackle the problem.

Cost is one of the leading barriers to access of eye care services in Africa[8]. The social enterprise model is designed to offer quality eye to all where the rich can pay the normal cost and the poor will have their fees subsidized or even no fees for a similar level of care. The strategy was to start a facility where the founder who understands the problem and the solution well, has full control of the Manpower, Money, and Materials [3M] in the facility.

Starting a high-quality eye hospital in the rural area in Africa is full of challenges. These will range from financial resources, source of equipment, supply chain of consumables, skilled manpower for this specialized service in a rural area etc. The hospital was started on a very low budget by leasing an apartment building and renovating section by section as funds became available.

The main sources of funding were initially from selling of family property, local bank loans and friends' donations. Later some international grants and zero interest loans started trickling in. The hospital was not designed to rely on grants and donations but by being self-sustaining. Up to the present period, grants whenever available have only contributed to about 7% or less of the revenue. Regarding skilled manpower, it was realized from the beginning that the mid-level ophthalmic personnel are the backbone of such a practice. These include ophthalmic nurses and technicians, counsellors, outreach staff, refractionists among others. The only way to obtain such staff in this region was to train them. The hospital therefore embarked on an intensive training program that leveraged on best practice training programs elsewhere which had shown success.

As the hospital celebrates its 10th Anniversary, great strides have been made to demonstrate that it has achieved great successes and looking forward for much more. Most important to note is that the hospital is financially self-reliant, a key factor to prove that the social enterprise model can be a success in Africa. In terms of numbers the hospital started by attending to 6,469 patients in 2013 and that increased steadily to 31,914 in 2019 before the effects of Covid-19 pandemic affected the operation of the hospital and now it is catching up. A total of 188,875 patients have been treated in the facility since inception (Figure 1). In 2013 the hospital performed 404 cataract surgeries. This number also increased greatly to 2,356 cataract surgeries in 2019 before the decline in 2020. By 2022 the numbers have bounced back to 2,016 cataract surgeries (Figure 2). Cataract surgeries serve a dual purpose of having the most significant impact in reduction of blindness in the community as well as being key to sustainability. It is recognized that the poverty index in the rural areas in Kenya is higher than 50% in several counties[9]. To break the greatest barrier to access eye care in the poor communities, a total of 48% of cataract surgeries have been offered free of charge. This cost is met by either supporters or the cross-subsidization model. To manage the cost recovery the hospital performs a minimum of 40% phacoemulsification of the walk-in patients. There is great attention to the quality of cataract surgery outcome monitoring. The hospital made a concerted effort to

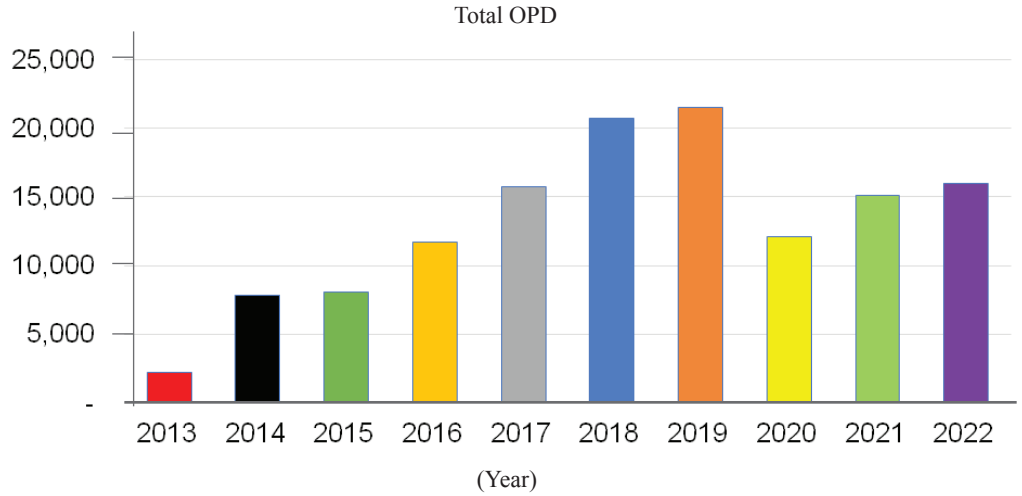

Figure 1: Patients attended per year

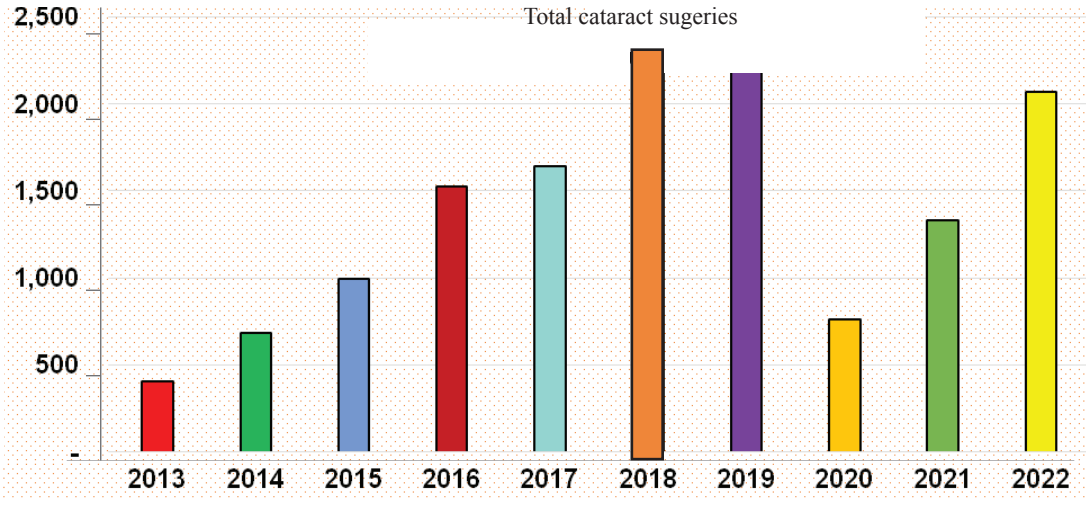

Figure 2: Cataract surgeries done per year

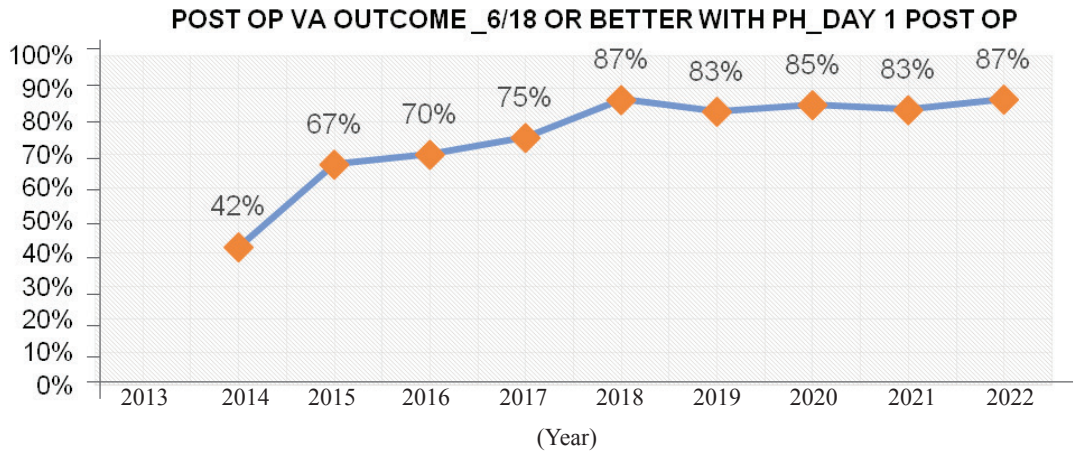

Figure 3: Cataract surgery outcome

meet the WHO recommended target of > 80% success in cataract surgery outcome from the beginning. The target was achieved in 2018 and consistently maintained and surpassed to date (Figure 3). And these figures include the outcome of surgeries for residents from the neighbouring universities who regularly come for training in the facility.

The optical department also similarly serves the dual function of reduction of reversible poor vision as well as sustainability. Significant emphasis is made in dispensing spectacles in a cost effective and timely manner. The hospital has so far dispensed 25,836 spectacles with the yearly breakdown as shown in Figure 4. As a

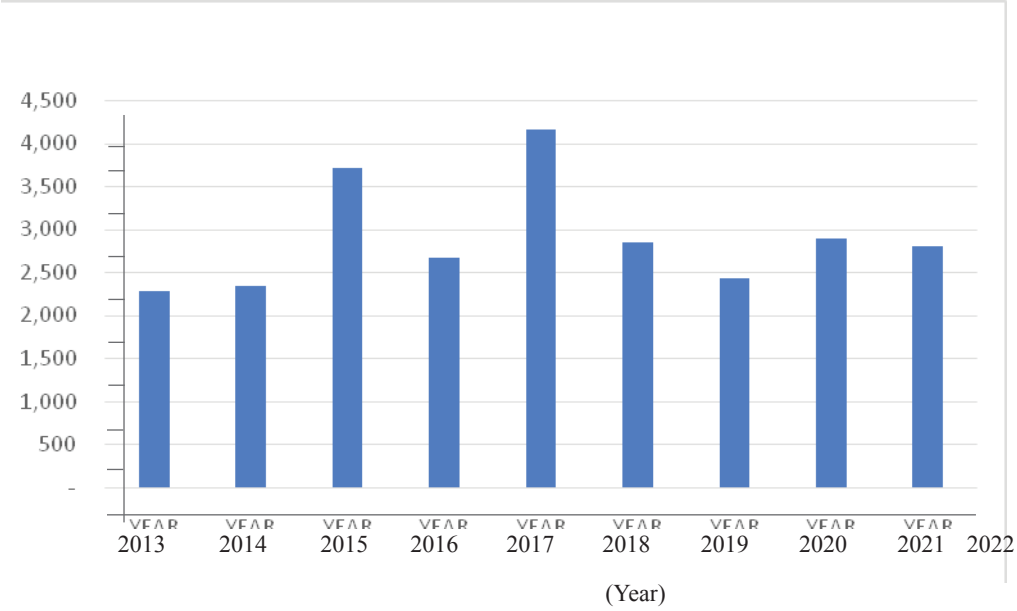

Figure 4: Spectacles dispensed

comprehensive eye hospital, skills are developed to serve other specialties like glaucoma, cornea, medical retina, and paediatric ophthalmology.

Traditionally the hospital has been running base hospital and outreach services. It has however recognized that outreach program, as much as they help reach to the non-customers [those who need the services but would ordinarily not be able to access them], they are not effective in giving a sustainable eye care in the community. The outreach is planned in the community at the convenience of the hospital and the clients may not be having the need then, and when the need occurs, there is a tendency that they will wait until the outreach happens again, often too late for timely intervention. The hospital embarked in a program of vision centres – these are small permanent facilities set up to provide primary eye care services to semi-rural and rural communities. Ophthalmic nurses or assistants operate the centres and will counsel, treat, dispense spectacles, and refer complicated and patients needing surgeries to a main hospital. It is envisaged that a vision centre will greatly improve the eye health seeking behavior of the community as one can travel a short distance and get quality care whenever there is need to. The hospital has established vision centres with the standard best practice of setting up and running one. It is in the process of establishing many more in the coverage area to replace outreach camps.

Looking forward, a stage is now set for further growth by strengthening the existing programs of training and standard processes as well as establishing other hospitals

in the region for wider coverage. It has been observed through the establishment and other similar ones in SSA that this model is a viable option of providing eye care in this region with a potential to offer the solution to the elimination of avoidable blindness. Setting up of more such models by others in the region is encouraged and the Kisii Eye Hospital is ready to share their experience.

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THE COECSA EXAMINATIONS

The College of Ophthalmology of Eastern, Central & Southern Africa (COECSA) aims to address the chronic shortage of ophthalmologists in Eastern, Central & Southern Africa (ECSA) region, as well as improve the quality of eye care services in the region. One of the ways the College does this is to continually assess the quality, safety and efficiency of the patient care provided by ophthalmologists through administering of examinations.

The main purpose of COECSA examinations is to examine candidates who are in training or have completed their post-graduate training in ophthalmology and wish to become fellows of COECSA, with all attendant benefits. The examination tests the knowledge, understanding and skills acquired by trainees or practicing ophthalmologists. Our international standard exams are designed to be as relevant, objective and stringent as possible.

Our exams include both Written and Clinical Fellowship Exams which ultimately lead to the award of Fellow of COECSA (FCOphth -ECSA). Further, the COECSA Exams form a complete stand-alone pathway to qualification as an Ophthalmologist, when complemented by training in an accredited facility that delivers and tracks learning as described in the COECSA Residency curriculum.

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The complete examination pathway for one to become a Fellow of The College of Ophthalmology of Eastern, Central Southern Africa, is as follows;

1. Written Part 1A - Basic Sciences
2. Written Part 1B – Optics, Refraction, and Instruments
3. Written Part 2- Clinical Ophthalmology
4. Part 3 -Clinical Exam (Viva & OSCE)

The College endeavors to make the Written Exams affordable to the candidates, thus allow more access the exam. The fees applicable on our exams are as follows;

a. Written Part 1A: Basic Ophthalmic Sciences = **$120**
b. Written Part 1B: Optics and Refraction = **$80**
c. Written Part 2: Clinical Ophthalmology = **$150**
d. Part 3: Clinical Fellowship Exam (Exit) = **$ 250**
e. Examination Centre Admin Fee (Compulsory): **$30** per sitting

Exam update

Friday, 23rd June, 2023, was a historical day for the College of Ophthalmology of Eastern, Central and Southern Africa (COECSA), as the College administered its first ever Online Written Examinations. 14 Accredited Exam Centers that underwent rigorous preparations, from 7 countries across the Eastern Central and Southern Africa (ECSA) region hosted the exams. The 7 countries include Ethiopia, Kenya, Rwanda and Somalia. Others are Tanzania, Uganda and Zambia. A total of 132 candidates were admitted to sit for the three (3) exams on offer, as follows:

✓ Written Part 1A - Basic Sciences: 46 Candidates
✓ Written Part 1B - Optics and Refraction: 54 Candidates
✓ Written Part 2- Clinical Ophthalmology: 32 Candidates

COECSA would like to thank all the Centers and Exam Coordinators who have volunteered to deliver the exam. In the words of COECSA Exams Advisory Board Chairperson, Prof. Ciku Mathenge: "*To our Coordinators, you are all volunteers which is what makes this even more amazing. Some*

of you have nothing to do with ophthalmology yet you offered your centres. It is a privilege for us to work with you. Together let us deliver an exam that makes all of us proud. We will all be there to support one another".

The College further extends its gratitude to all our partners who have supported this journey, especially PYOTT Foundation and Ophthalmology Foundation. We further thank all those that contributed to the entire exam development process. COECSA remains committed to making this annual exam sustainable, and accessible across the African continent and beyond.

For further information about COECSA Exams, please write to: exams@coecsa.org

NOTES

THE JOURNAL OF OPHTHALMOLOGY OF EASTERN CENTRAL & SOUTHERN AFRICA

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