

# Juvenile-onset Open-Angle Glaucoma at the University Teaching Hospitals - Eye Hospital, Lusaka Zambia

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## ABSTRACT

**Purpose:** To demonstrate the socioeconomic, demographic and clinical characteristics associated with patients of juvenile-onset open-angle glaucoma (JOAG) at the University Teaching Hospitals Eye Hospital (UTHs - EH).

**Methods:** This was a cross-sectional survey for Juvenile Open Angle Glaucoma (JOAG) conducted at the UTHs - EH in, Lusaka, Zambia from January to December 2013. All participants aged between 18 and 39 years had a full ocular examination after capturing demographic and socioeconomic information. The ocular examination included visual acuity, intraocular pressure (IOP) and cup

disc ratio (CDR) and tests performed were central corneal thickness (CCT) and visual fields. Severity was graded based visual field (VF) in the worse eye using the advanced glaucoma intervention study score. Univariate and multivariate logistic regression, stratified by age group and gender, was used to determine the association between demographic factors and JOAG and between clinical factors and JOAG.

**Results:** Of the 1625 patients recruited for the study, 309 were POAG patients. Of the 309 POAG patients, 140 aged 20 to 39 years old had bilateral JOAG. The distribution of the 140 participants was 98 (70.9%) females and 42 (29.4%) males. Thirteen (9.3 %) were aged 20 – 24 years, 29 (20.7%) 25 – 29 years, 44 (31.4%) 30 – 34 years, and 54 (38.6%) 35 – 39 years. The mean age of the patients was 25.1 ± SD 8.7 years. The prevalence of JOAG was 8.6%

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(140/1625) distributed as 2.6% (95% CI 1.3, 3.9) males and 6.0% (95% CI 4.7, 9.2) females. There was a female preponderance of (71.2% vs. 28.8%; OR 2.98, 95% CI 2.3, 6.7,  $p=0.021$ ). Eighty-five (60.7%) had complained of poor vision and 24 (17.1%) of eye pain. However, 24 (17.1%) presented with no definite symptoms. Patients with a positive family history presented 3.7 years earlier ( $P=0.034$ , CI: 1.37-7.9) compared to those without a family history. Lower socioeconomic status (Odds ratio [OR] 3.5,  $P=0.013$ , CI: 1.2-17.2), and higher IOP (OR 6.7,  $P=0.002$ , CI: 2.6-21.8) were associated with severe glaucomatous visual field defects. High myopia ( $-6.47 \pm 5.00$  Diopters) was present in 70.9% of patients. The patients with myopia also had a severe elevation of IOP of ( $35.8 \pm 18.5$  mmHg).

**Conclusions:** The study found a high prevalence of JOAG at 8.6%. The patients with JOAG presented late with advanced disease and high IOP. Clinical, socioeconomic, and demographic factors are contributory to the severity of JOAG among JOAG patients.

**Recommendation:** Early detection of cases during eye health care outreach programmes such as school and community screening of children and adults could be of great benefit in creating awareness, demand, early detection and prompt commencement of treatment. Glaucoma should no longer be considered a condition of the people aged 40 years and above.

## INTRODUCTION

Glaucoma is a heterogeneous group of optic nerve diseases divided into congenital, juvenile-onset and adult-onset categories.<sup>1</sup> JOAG is a rare subset of POAG characterized by an autosomal dominant pattern of inheritance.<sup>2,3</sup> The affected age range is between three and 39 years and commonly present with myopia and severely elevated IOP with large fluctuations.<sup>4</sup> Advanced glaucomatous optic neuropathy and severe VFD are other clinical features associated with JOAG.<sup>5</sup> It also tends to progress faster.<sup>6</sup> Some studies have reported that a

considerable number of patients with JOAG tend to present late with advanced disease.<sup>7,8</sup> The difference between JOAG and the late congenital glaucoma is that the JOAG would not have clinical features such as buphthalmos, Haab's striae, anterior segment dysgenesis and ocular or other systemic developmental anomalies.<sup>4</sup>

Various studies have reported different ages of JOAG onset. Some studies have observed that the onset age varies from country to country, region to region and continent to continent and may vary from race to race.<sup>9,10</sup> The onset of JOAG is usually associated with an early age (3 years), and in most instances, it affects individuals in their childhood and early adulthood.<sup>6</sup> In Africa, the reports from Cameroon and Nigeria indicated 12 years as the age of onset of JOAG.<sup>9,11</sup> These reports were contrary to the findings in the USA, and Asia where the onset age of JOAG was reported as 18 and 21.3 years, respectively.<sup>2,12</sup> These findings demonstrated that JOAG occurred early in black Africans compared to other races.

The prevalence of JOAG varies from country to country, region to region and continent to continent and may vary from race to race. An epidemiological study from the Dallas Glaucoma registry reported that JOAG comprised about 4% of the cases of childhood glaucoma with JOAG defined as idiopathic glaucoma arising in children older than three years of age.<sup>10</sup> In another report from the USA, 1 in 50,000 individuals had JOAG while among Cameroonians, the prevalence was 0.4%.<sup>4,9</sup> A population-based study in Olmstead County, Minnesota, USA reported the incidence of JOAG at 0.38 per 100,000 residents between 4 and 20 years of age.<sup>13</sup> Alliot et al. (1998) and Komolafe et al. (2011), reported the prevalence of JOAG of 3.4% and 7.0%, respectively, of all glaucoma patients attending ophthalmic consultant clinics in two West African countries.<sup>11,14</sup>

The information on the clinical profile of patients with JOAG in African sub-region is limited.<sup>4,15</sup> The reason for this could be the presumed rarity of the

condition and the general thinking that glaucoma only occurs in people aged 40 years and above. As JOAG patients are young with an expected more extended life expectancy than older adult glaucoma patients, their morbidity duration is likely to be very long. Thus, early diagnosis is fundamental so that sight can be retained for a long time by the patients. However, most diagnoses are made late due to patients' late presentation. This situation entails severe disease at first contact, often associated with visual impairment and blindness which can significantly impair the patient's quality of life and limit daily living activities for the entire lifespan.<sup>16</sup> The presumed rarity of JOAG could probably contribute to the deficient description of its clinical characteristics and prognosis. There is no information on the prevalence and characteristics of JOAG among the Zambian population.

This study attempted to demonstrate the demographic features, clinical characteristics, socioeconomic factors and the VF picture in patients with JOAG seen in an out-patient eye clinic of a tertiary eye hospital in Lusaka, Zambia.

## **MATERIALS AND METHODS**

A cross-sectional survey of 1,625 participants aged 18 to 98 years old was conducted on POAG at the UTHs Eye Hospital in Lusaka, Zambia. The UTHs Eye Hospital is the national referral hospital which provides ophthalmological surgical and clinical services. The UTHs' Eye Hospital is estimated to cater for more than 21,000 clients annually for both routine and morbidity driven health care. The clients that attend this clinic come from across the country and include both self- and system-referrals, representing all age groups and all ethnic groups.

A systematic random sampling using 50%-time sampling was employed which meant that of the 220 (on average) eye patients seen in the outpatient eye clinic every month, 110 were to be picked to participate in the study. This selection translated to a minimum of 1320 participants to be recruited into the study for twelve months. To cater for attrition and assuming a response rate of 80%, the sample

size of the study pegged at 1,714 participants. The JOAG patients were to be obtained from the 1,714 participants. Only 1625 eye patients participated were recruited, giving a response rate of 94.8%. Of the 309 POAG patients, 140 patients aged 20 to 39 years had JOAG.

The subjects' inclusion criteria were age of 18 years or more, clear ocular media, glaucomatous optic neuropathy in at least one eye and visual field loss consistent with optic nerve damage in at least one eye. The lower age limit of 18 years was considered because of the study design which stipulated to include adults only. Exclusion criteria included evidence of secondary causes of elevated IOP, history of intraocular surgery, pigmentation of the angle greater than grade 3 or peripheral anterior synechia, conditions other than glaucoma affecting the VF, presence of any other retinal or neurological pathology and no light perception. A positive family history of glaucoma was defined as the presence of one or more relatives (first or second degree) of the patient who reportedly had been diagnosed with glaucoma by an ophthalmologist. The participants were classified into different socio-economic classes depending upon their urban or rural background, respectively.

### *Diagnosis of JOAG*

The diagnosis of JOAG was based on

- The presence of characteristic glaucomatous optic neuropathy (neural rim thinning, focal notching or a vertical cup-to-disc ratio >0.5)
- Visual field defects not attributable to other causes;
- Open anterior chamber angle configuration on gonioscopy in the affected eye
- Age ranging between 18 and 39 years of age as confirmed through the national identification document called green national registration card (NRC).
- IOP in mmHg whether raised or not by Goldmann applanation tonometry.<sup>17</sup>

The indicators of JOAG, therefore were:

(a) *Optic disc status*: The optic disc was examined using a 78D Volks lens (Volks Optical, Inc., Mentor, OH) at X16 magnification after adequate pupillary dilation. When there was evidence of glaucomatous optic nerve damage, that is, cupping of  $>0.5$  with or without notching supported by visual field changes, it was referred to as glaucomatous. When there was no such evidence of glaucoma, it was referred to as non-glaucomatous.

(b) *IOP*: IOPs were considered normal if it was  $\leq 21$ mmHg. Values  $>21$ mmHg or a difference of 4mmHg or more between the two eyes were considered abnormal. The IOP was measured on two separate occasions with a Haag Streit Goldmann applanation tonometer under topical anaesthesia, and the average value was considered for the study.

(c) *Gonioscopy*: This was performed with a Volk 3-Mirror Gonio Lens. Grade three (3) and four 4 were considered as open angles.

(d) *Visual fields*: Visual fields were plotted for all participants declared as suspects of glaucoma using Humphrey Field Analyzer I - I series 24-2 array (Zeiss, Oberkochen, Germany). Subjects with visual field defects suggestive of glaucoma were confirmed as glaucoma if there were either glaucomatous optic disc changes or high IOP. The presence of characteristic glaucomatous field defects affecting the area within  $10^\circ$  from fixation was categorized as severe visual field loss, while characteristic glaucomatous field defects not affecting areas within  $10^\circ$  from fixation were categorized as mild/moderate visual field loss. Unreliable test results were excluded if the fixation loss was more than 30% or a false-positive or false-negative was greater than 33%. Those with no visual field defects that could be attributed to glaucoma were labelled as non-glaucomatous field changes.

Detailed medical history, full ocular examination findings including slit-lamp biomicroscopy, best-corrected visual acuity, refractive error, and central corneal thickness (measured using a Kacon Ophthalmic Ultrasound System, China), were also

recorded. The documented variables were demographic data such as age, sex, education status, socioeconomic status, occupation and the ocular clinical characteristics including the presenting visual acuity, IOP, the cup disc ratio and the type of treatment given. *The diagnosis of JOAG was made by ophthalmologists.*

#### *Analysis*

Collected data was entered in Microsoft Excel version 2007 and transferred to Stata version 12.0 for further storage and analysis. Univariate and multivariate logistic regression analyses were used to assess and estimate the association of sex, age and gender with JOAG. The variables in the model were age, residence, and stratification were done by sex and age group. Chi-square test and an independent 't'-test were applied to compare categorical and continuous data, respectively. The odds ratio was calculated with a 95% confidence interval. A univariate and then multivariate binomial logistic regression analysis of the factors associated with severity at presentation was performed. The disease severity at presentation using a worse eye VFD and with binocular field defects at presentation was analysed separately. A *p*-value less than or equal to 0.05 was considered statistically significant.

#### *Ethical considerations*

This study was performed in accordance with the tenets of the Helsinki declaration. The University of Zambia Biomedical Research Ethics Committee approved the study (reference number 013-08-12). Further approval was obtained from the Ministry of Health of Zambia through the UTH.

## **RESULTS**

Of the 309 POAG participants, 140 participants aged between 20 and 39 years, hence classified as having JOAG. The mean age of the patients was  $25.1 \pm$  SD 8.7 years (range 20–39 years). The mean age among the females was  $26.1 \pm 9.5$  compared to the males  $24.1 \pm 7.9$ . Thirteen (9.3 %) were aged 20 – 24 years, 29 (20.7%) 25 – 29 years, 44 (31.4%) 30 – 34 years, and 54 (38.6%) 35 – 39 years.

The prevalence of JOAG was found to be 8.6% (140/1625); [95% CI, 5.1%, 11.3%), Table 1.

**Table 1: Gender Distribution Vs Prevalence of POAG Among Participants by Age Group in a Hospital Survey in POAG at the UTHs – EH (n = 1,625)**

Age group	N (%)	Gender	POAG patients	Prevalence		
				By gender in each age group (%)	By age group (%)	By type of POAG
20 – 24	60 (3.69)	male	4	0.20	0.80	<b>JOAG</b>
		female	9	0.60		
25 – 29	125 (7.69)	male	6	0.40	1.79	<b>140/1,625</b> <b>(8.6)</b>
		female	23	1.40		
30 – 34	200 (12.31)	male	13	0.80	2.71	
		female	31	1.80		
35 – 39	253 (15.57)	male	19	1.20	3.32	
		female	35	2.10		
40 – 44	82 (5.05)	male	3	0.20	0.55	<b>POAG</b>
		female	6	0.40		
45 – 49	94 (5.78)	male	1	0.10	0.55	<b>169/1,625</b> <b>(10.4)</b>
		female	8	0.50		
50 – 54	201 (13.37)	male	3	0.20	2.03	
		female	30	1.80		
55 – 59	132 (8.12)	male	14	0.90	2.58	
		female	28	1.70		
60 – 64	37 (2.28)	male	9	0.60	1.35	
		female	13	0.80		
= 65	441 (27.14)	male	21	1.30	3.32	
		female	33	2.00		
<b>TOTAL</b>	<b>1625</b> <b>(100)</b>		<b>309</b>	<b>19.0</b>	<b>19.0</b>	<b>309/1,625</b> <b>(19.0)</b>

There were 98 (70.9%) females and 42 (30.1%) males; Table 2.

**Table 2: Socio-Demographic Profile of JOAG Participants at the UTHs EH (n = 140)**

<b>Variable</b>	<b>Number of JOAG Participants (n)</b>	<b>Proportion (%)</b>	<b>p-value</b>
<b>Age group (years)</b>			0.022
20 – 24	13	9.3	
25 – 29	29	20.7	
30 – 34	44	31.4	
35 – 39	54	38.6	
<b>Sex</b>			0.013
Male	42	30.1	
Female	98	69.9	
<b>Occupation</b>			0.012
Student	16	11.4	
Informal Employment	35	25.0	
Formal Employment	22	15.7	
Subsistence Farming	23	16.4	
Unemployed	44	31.4	

The clinical and socioeconomic profile was statistically significant for female gender, older age group, positive family history, poor VA, severe visual field changes, cup disc ratio of more than 0.7 and occupation as shown in Table 3.

**Table 3: Clinical and Socioeconomic Profile of JOAG Study Participants, n = 140**

Variable	Characteristic	Univariate Analysis				Multivariate Analysis			
		Odds Ratio (OR)	95%, CI		p-value	Odds Ratio (OR)	95%, CI		p-value
			Lower	Upper			Lower	Upper	
Sex	Male (Ref)	1.00							
	Female	3.99	1.98	4.53	0.001	3.12	2.15	5.08	0.027
Age group (years)	20 - 24 (Ref)	1.00							
	25 – 29	1.31	1.04	1.56	0.025				
	30 – 34	1.21	1.07	1.45	0.012				
	35 – 39	1.71	1.26	2.38	0.001	3.33	2.55	5.78	0.030
Reason of presenting to the hospital	Poor vision	6.67	2.44	10.67	0.009	4.51	1.11	6.55	0.031
	Routine eye examination	0.89	0.99	3.22	0.789				
	Ocular pain	0.71	0.89	2.71	0.944				
	Refractive error (Ref)	1.00							
Family history of glaucoma	Yes	12.833	1.45	14.12	<0.001	17.88	4.78	16.89	0.001
	No (Ref)	1.00							
Presenting visual acuity (in the better eye)	>6/18 (Ref)	1.00							
	6/18-6/60	1.43	1.04	1.61	0.490				
	<6/60-3/60	2.33	1.55	5.56	0.001	2.01	1.31	4.99	0.017
	<3/60-NPL	1.82	1.99	4.22	0.012				
Cup disc ratio (in the better eye)	<0.5 (Ref)	1.00							
	0.5 – 0.7	2.55	1.88	5.90	0.017				
	>0.7	5.99	2.88	11.34	0.001	5.09	2.17	11.01	0.033
Visual field changes (in the better eye)	Mild (Ref)	1.00							
	Moderate	0.87	0.97	3.19	0.786				
	Severe	2.33	1.55	5.56	0.001	2.12	1.03	5	0.040
Intraocular pressure (IOP)	Male (Ref)	1.00							
	Female	3.55	2.89	10.66	0.032				
Duration (more than three years)	No (Ref)	1.00							
	Yes	1.64	1.23	2.62	0.0323				
Occupation	Student (Ref)	1.00							
	Informal Employment	3.61	2.09	5.99	0.007	2.99	2.09	4.32	0.025
	Formal Employment	3.55	2.89	10.66	0.012				
	Subsistence Farming	1.33	1.07	1.44	0.003	1.19	1.97	3.31	0.031
	Unemployed	1.71	1.26	2.38	0.001	2.55	1.36	4.89	0.008

Note: IOP = intraocular pressure; NPL = No perception of light; CI = Confidence interval

Participants with a positive family history presented at least three years earlier,  $p = 0.323$ . Those with a positive family history were ten times more likely to present with severe visual field defects. Most of the JOAG participants, 60.7%, presented to the eye hospital due to poor vision and 24 (17.1%) had JOAG detected during routine ocular examination. While 63 (45.0%) had a positive family history of glaucoma, 77 (55.0%) participants did not.

The majority had visual acuity of worse than 6/60, while 65.1% of the participants had significant severe visual field changes. The mean IOP of all the JOAG participants at presentation was  $35.8 \pm \text{SD } 18.5$  mmHg (range 10.0–57.8 mmHg). The mean IOP for male participants was  $31.2 \pm \text{SD } 14.5$  mmHg (range 10.0–50.0 mmHg) and for females  $40.4 \pm \text{SD } 15.4$  mmHg (range 10.0–58.0 mmHg), Table 4. The mean IOP increased with the age group. The age group 35 – 39 years had the highest mean IOP;  $43.5 \pm 12.5$  mmHg; Table 4.

**Table 4: Pre-treatment mean intraocular pressure (IOP) profile of JOAG study participants, n = 140**

Variable	Pre-treatment Mean IOP (mmHg) $\pm$ SD	p value
All patients (N = 140)	35.8 $\pm$ SD 18.5 mmHg	
Male group (n = 42)	31.2 $\pm$ SD 14.5 mmHg	0.012
Female group (n = 98)	40.4 $\pm$ SD 15.4 mmHg	
20 – 24 group (n = 13)	27.3 $\pm$ SD 7.0 mmHg	
25 – 29 group (n = 29)	33.6 $\pm$ SD 9.7 mmHg	0.022
30 – 34 group (n = 44)	38.8 $\pm$ SD 11.4 mmHg	
35 – 39 group (n = 54)	43.5 $\pm$ SD 12.5 mmHg	

Note: SD = Standard deviation; IOP = Intraocular pressure

Table 5 shows a comparison of the clinical parameters between 91 patients with advanced glaucoma and 49 non-advanced glaucoma in 140 participants with JOAG. The IOP was significantly higher in the advanced group ( $44.9 \pm 9.7$  mmHg) than in the unadvanced group ( $26.7 \pm 7.3$  mmHg,  $p = 0.009$ ).

**Table 5: Clinical profile comparisons between eyes with advanced glaucoma and non-advanced glaucoma, n = 140.**

Variable	With Advanced Glaucoma (n = 91)		With Non-advanced Glaucoma (n = 49)		p-value*
	Mean reading	SD	Mean reading	SD	
Mean age at diagnosis (years)	26.3	$\pm 6.9$	23.9	$\pm 10.5$	0.033
Refractive error (dioptre)	-7.0	$\pm 0.75$	-5.92	$\pm 0.67$	0.011
Vertical cup-to-disc ratio	0.79	$\pm 0.06$	0.74	$\pm 0.05$	0.011
IOP at enrolment (mmHg)	44.9	$\pm 9.7$	26.7	$\pm 7.3$	0.009
MD at enrolment (dB)	-14.11	$\pm 2.67$	-6.55	$\pm 1.25$	0.003

Note: Values are expressed as mean  $\pm$  SD unless otherwise indicated. IOP = intraocular pressure; MD = mean deviation of the visual field test. \* Independent T-tests analysed the visual field parameters between the eyes with advanced glaucoma and non-advanced glaucoma. The advanced group had a slightly significantly higher proportion, 60.3%, of family history compared to the unadvanced group (39.7%,  $p = 0.038$ ).

The bivariate logistic regression analyses shown in Table 6 demonstrates that IOP was significantly associated with advanced glaucoma (odds ratio (OR), 1.33;  $p = 0.009$ ). The multivariate analysis maintained this huge significant association ( $p < 0.001$ ) between the two variables. The multivariate analysis equally demonstrated a significant deep association between advanced glaucoma with refractive error (odds ratio (OR), 1.188;  $p = 0.001$ ). In multivariate analyses, advanced glaucoma was also very significantly associated with the IOP (OR, 1.399;  $p < 0.001$ ).

Table 6: Association Between Advanced Glaucoma and Clinical Factors, n = 140

Variable	Characteristic	Bivariate analysis				Multivariate analysis			
		OR	95%, CI		p-value	OR	95%, CI		p-value
			Lower	Upper			Lower	Upper	
Family history <sup>†</sup>	Yes	2.83	1.45	14.12	0.003	2.78	0.94	11.91	0.043
	No (Ref)								
Refractive error (dioptr)	Yes	1.26	0.99	1.91	<0.001	1.18	1.55	2.11	0.011
	No (Ref)								
IOP (mmHg)	Yes	1.33	1.18	1.96	0.009	1.40	1.17	1.91	<0.001
	No (Ref)								

Note: In bivariate and multivariate analyses, generalized estimating equations were conducted.

OR = odds ratio; CI = confidence interval; IOP = intraocular pressure;

<sup>†</sup>These data of family history occurred in 63 of 140 participants eyes included in the analysis, of which there were 91 participants in the advanced glaucoma group and 49 participants in the unadvanced glaucoma group.

The socio-economic evaluation revealed that 60.22% of the participants were rural dwellers. Rural dwellers comprising mostly subsistence farmers had 31.21% while unemployment stood at 23.5%. The JOAG problem seemed to be more among the participants who were not in formal employment (72.41%).

The univariate logistic regression analyses shown in Table 7 demonstrates that IOP was significantly associated with advanced glaucoma (odds ratio (OR), 1.333;  $p = 0.009$ ). The multivariate analysis maintained this huge significant association ( $p < 0.000$ ) between the two variables. The multivariate analysis equally demonstrated a significant deep association between advanced glaucoma with refractive error (odds ratio (OR), 1.188;  $p = 0.001$ ). In multivariate analyses, advanced glaucoma was also very significantly associated with the IOP (OR, 1.399;  $p < 0.000$ ). The socio-economic evaluation revealed that 60.22% of the participants were rural dwellers. Rural dwellers comprised of subsistence farmers standing at 71.21% while unemployment stood at 73.5%. The JOAG problem seemed to be more among

the participants of the lower socioeconomic strata (72.41%).

## DISCUSSION

To our knowledge, this is the first cross-sectional survey conducted in an African setting that looked at the prevalence, demographics, socioeconomic and clinical characteristics of JOAG. JOAG is a rare subset of POAG.<sup>6</sup> In this study, 140 (45.31%) of the 309 POAG patients had JOAG in the black Zambian population, suggesting that JOAG is a common condition in the black Zambian people. This study showed that JOAG had preponderance for the female gender, myopic refractive state and severe elevation of IOP; just as reported in other studies.<sup>5,18</sup> The older JOAG patients had severe, bilateral disease and made up a higher proportion of those with a positive family history of glaucoma and late presentation. This finding is similar to that by Kwun et al. (2016).<sup>6</sup> There are no studies among young patients with early-onset glaucoma that have investigated the factors associated with severity at presentation<sup>19</sup>, despite Brandt et al. (2001), reporting that POAG is significantly more common, develops earlier, and is more severe in blacks than white populations.<sup>19,20</sup> This

study observed that JOAG participants had a severe presentation. The study also observed that poor socio-economic status was associated with the greater disease severity at presentation among JOAG patients just like reported by Gupta et al. (2013).<sup>19</sup> The explanation could probably be that participants of low social status delayed seeking medical advice due to either ignorance or cost constraints.

In this study, despite the challenges in the severity of the disease, all the participants received the appropriate treatment. One hundred and two (72.90%) patients were commenced on medical treatment, while 38 (27.11%) underwent surgical treatment. All the cases requiring surgical treatment had trabeculectomy surgery performed on them.

Patients of JOAG are known to have high IOP and severe glaucomatous damage at presentation compared to adult POAG patients.<sup>14,21</sup> In this study, the mean IOP in JOAG participants was ranging from  $27.30 \pm 11.25$  mmHg to  $47.50 \pm 17.45$  mmHg. The mean IOP between males and females was significantly statistically different. The participants aged 20 – 24 years had a lower mean IOP compared to those aged 35 – 39 years and there was a statistically significant difference among the four age groups. The validation of this suggestion could only be through another research focussed on IOP in POAG patients. The mean IOP among the study patients was also higher than what was reported among the Cameroonians and Nigerians.<sup>9,11</sup> The finding of this study compares with mean IOP of 38.5 mmHg reported by Kwun et al. (2016) and Wiggs et al. (1995).<sup>6,21</sup> Of interest is the fact that 3.5% of the patients had IOP, which was less than 21 mmHg at presentation. This finding conforms with the reporting of most researchers that patients with JOAG usually have high IOP (>30 mmHg). Among the Cameroonians, only 5.3% had IOP that was less than 21 mmHg.<sup>9</sup>

Among the 140 JOAG patients, the mean age at diagnosis was similar to that of the Koreans, Cameroonians and Nigerians.<sup>6,9,11</sup> The minimum age of 20 years recorded in this study was not the onset

age, but merely the age of the youngest patient recruited because the study focused POAG in adults hence the enrolment was for all those aged 18 years and above. From the study, it was apparent that most of the participants had JOAG for years before they presented to the facility as evidenced through the late presentation and disease severity. The suggestion that screening for glaucoma must only be for people aged 40 years and above could have compounded the late presentation because the clinicians did not screen patients younger than 40 years for POAG. Furthermore, the belief has been that POAG occurs in adults aged 40 years and above.<sup>20,22</sup> The reflection that the belief and practice do not favour early screening of young adults is in the high number of JOAG participants (60.7%) who presented with poor vision to the facility. The other surprising aspect was that more than 50% of the participants had refractions done on them over and over with spectacles prescribed for myopia several times when their diagnosis was not myopia, but glaucoma.

In all these participants, the examiners at local facilities conducted no fundoscopy despite them being eye health personnel. The fundoscopy performed at the UTHs EH in more than 50% of the JOAG patients was their first.

Late presentation among glaucoma patients is a problem highlighted in many previous studies.<sup>6,7,8,11</sup> In this study, 65.0% of the participants presented late, of which 77.8% had some form of disease manifestation such as ocular pain which was the case in 17.1%. The proportion of the patients that had a positive family history suggestive of glaucoma was rather low, 45.0% compared to 88.3% in Cameroon.<sup>5</sup> This study from Cameroon demonstrates the importance of adequate sensitisation and education of the public on glaucoma to prevent blindness from JOAG.

Ninety-one (65.1%) patients in this study presented with a severe VFD. The binocular VFD for grading severity of glaucoma, 63.5% of the JOAG patients, had severe VFD, 12.3% had moderate field defects, and 37% had a mild VFD. There was an association between the age at diagnosis with the severity of VFD. The younger patients with JOAG were as much

likely to have a severe disease as those presenting later. This study showed that VF progression was associated with a higher IOP and high myopia. There was an association between family history and VF progression. On the contrary, in the case of POAG in older patients, an evidence-based review showed that there was no significant relationship between the patient's family history of glaucoma and VF progression.<sup>23</sup>

The asymptomatic nature of glaucoma also makes people present very late to the hospital. This presentation could also be attributed to the poor health-seeking behaviour of the people especially those from lower socioeconomic status and rural settings.<sup>6,19,24-27</sup> Among patients with stroke, Kitko et al. (2008), reported that lack of knowledge, fear of hospital, and denial were the factors affecting health-seeking behaviour.<sup>28</sup> These factors could pose the same challenge for glaucoma. Blinding eye diseases such as glaucoma are usually associated with many myths, especially when they present early in life. As such, there is a tendency to seek improper treatment options.<sup>29</sup> In Nigeria, the study by Ashaye et al. (2006), showed several misconceptions about the causes of eye diseases among the Yoruba people, with fatalistic beliefs such as witchcraft ranking highest in the cause of blindness.<sup>29</sup> Probably all such factors could have contributed to the late presentation among the patients in this study.

In this study, the JOAG participants with a positive family history presented 3.7 years earlier than those without a family history. The study by Wu et al. (2006) observed that patients with familial POAG had a more significant disease severity and an earlier onset age at diagnosis compared to patients with sporadic disease.<sup>30</sup> Regarding the severity of JOAG based on the optic nerve evaluation, 77.9% of the patients presented with CDR of more than 0.7, which was statistically significant. The situation was worse with patients who were subsistence farmers and the unemployed who mostly constituted the group of poor socioeconomic status in this study. Elsewhere, factors were associated with severity at

presentation among adult glaucoma patients include the lower socio-economic status, presence of family history and poor health literacy.<sup>31-33</sup>

## CONCLUSION

This study observed that the prevalence of JOAG in this population is much higher than previously thought and that the levels of glaucoma qualify to consider glaucoma as a significant public health problem in Zambia with a significant genetic association and an early onset. The risk factors of JOAG in this population included sex, age, race and family history. JOAG was severe and bilateral in older JOAG patients who also made up a higher proportion of those with a family history of glaucoma and late presentation, suggests a need for early-onset primary screening strategies as part of the overall response to control this problem.

## RECOMMENDATION

Early detection of cases during eye health care outreach programmes such as school and community screening of children and adults could be of great benefit in creating awareness, demand, early detection and prompt commencement of treatment. Glaucoma should no longer be considered a condition of the people aged 40 years and above. More cross-sectional studies must be performed to characterise the JOAG further.

## Conflict of Interest

The authors report no conflict of interest. The authors alone are responsible for the content and writing of this article.

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