Refractive Errors in Children with Down syndrome in Lagos State, Nigeria.

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Abstract

Down syndrome is the most common chromosomal disorder and many with the condition tend to be more at risk of several ocular disorders than those without Down syndrome. The study was aimed at assessing the distribution of refractive errors in children with Down syndrome between 5-18 years of age. The cross-sectional study used data from 104 children with Down syndrome from selected exclusive special needs schools in Lagos State, Nigeria. Visual acuity was measured using the Lea symbol chart and non-cycloplegic refraction was carried out using both static retinoscopy and autorefraction. Significant refractive error was defined as myopia, hyperopia and astigmatism of 0.75D and above respectively. Data was analysed using IBM SPSS statistical software version 20.1. Chi-square and independent t test were used to test the hypotheses. A total of 91 children with Down syndrome were examined; 52 (57.1%) were males with a mean age of 13.6 +3.8 years. The study showed that refractive errors was present in 82 (95.3%) of the participants, with astigmatism being the commonest form of refractive error occurring in 61(67%) followed by hyperopia 12(13.2%) and myopia 7(7.7%) of the 91 participants studied. The study also supported the null hypothesis that there is no significant difference between the types of refractive error and gender (p=0.8331). Recorded visual acuities also revealed a generalised reduced visual acuity which significantly improved with best optical correction (p <0.001). This study indicates the need for people with Down syndrome to be provided with prompt eye care services.

Keywords: Refractive error, Down syndrome, visual acuity, prevalence, distribution

Introduction

Down syndrome (DS) or Down's syndrome, also known as trisomy 21, is a genetic disorder caused by the presence of all or part of a third copy of chromosome 21. It is the most common chromosomal abnormality in humans[1], occurring in about 1 per 1000 babies born each year. The incidence is estimated to be about 1 in 600 live births[2]. It occurs in people of all races and economic levels, though older women have an increased risk of having a child with Down syndrome but the cause of non-disconjuction is still unknown[3]. This condition is typically associated with physical growth delays, characteristic facial features and mild to moderate intellectual disabilities[4]. In recent history, advances in medicine and science have enabled researchers to investigate the characteristics of people with Down syndrome. About 60% of people

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with Down syndrome have ocular manifestations. Ocular findings in Down syndrome include a wide range of visual acuities due to refractive errors and amblyopia, strabismus, nystagmus, lid abnormalities including prominent, upwards slanting of the palpebral fissure, epicanthal folds, lid infections including blepharitis, blepharoconjunctivitis, chalazion and hordeola. Furthermore, they may have nasolacrimal duct obstruction, corneal ectasia, iris brushfield spots seen in about 90% cases especially those with lightly pigmented irides, presenile cataracts, glaucoma, and retinovascular anomalies[5-7].

The distribution of refractive errors in school aged children with Down syndrome and cerebral palsy are different from that of typical children. Investigations have all revealed very distinct, complex and aberrant visual development in children with Down syndrome from early months of life until school age[8-10]. Studies have shown that in children with Down syndrome, the distribution of refractive errors in the first years of life mirrors that of typical children but widens over time rather than narrows, also emmetropisation is believed to fail in most of these children with down syndrome and cerebral Palsy[11-13].

People with Down syndrome have been reported to have a higher incidence of refractive errors[8,14]. Reports on the prevalence of children with refractive errors vary in literature but it is generally agreed to exceed 40%[15] and this high prevalence occurs amongst school children with Down syndrome as well as adults[11,16]. Refractive errors and squint maybe present from an early age and persist into childhood[8,9,17]. The most common refractive error is hypermetropia which often reduces spontaneously in other children, is likely to persist beyond infancy in DS subjects[9].

Despite the high prevalence of large refractive errors in children with Down syndrome, longitudinal data show that these are not always present in early infancy[17]. The prevalence of astigmatism among infants (0–12 months) has been reported to be 45–53% (defining astigmatism ≥ 1.00 D) in studies using non-cycloplegic techniques18,19 and as 65% (astigmatism of ≥ 0.75 D) using photorefraction[20].

Although the incidence of ocular anomalies in children with Down syndrome varies in different studies, they have shown that children with Down syndrome were more at risk for several ocular disorders than typical children. Whilst studies in Port Harcourt[21] and Benin[22] respectively have reported on ocular manifestations in Down syndrome, there are no documentations in Lagos state. This is in contrast with comprehensive studies carried out on refractive errors in children including infants and school aged children with Down syndrome in Europe, Americas and Asia[8,23,24]. Furthermore, whereas the distribution and prevalence of refractive

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error amongst school children in different population settings is known[25-30], little is known about the prevalence of refractive error among children with Down syndrome in Nigeria.

Children with Down syndrome are often neglected when national eye health and rehabilitation programmes are planned therefore the need for this study. The rationale behind this study is that refractive errors in children and people generally with Down syndrome is largely under reported compared to children without Down syndrome. There is need to investigate this problem in other to know the distribution of refractive errors among children with Down syndrome and to be able to compare its prevalence with that of children without Down syndrome. The findings will assist eye care practitioners in managing the visual problems associated with Down Syndrome as well as improving the quality of life in this special population.

Method

The study was a cross sectional analytic study design using quantitative method of data collection. The study area was Lagos, Lagos State, Nigeria. Lagos State is located in the south west geopolitical zone of Nigeria. It has six educational districts and there are two categories of special schools in Lagos State, five exclusive schools and 38 inclusive schools. Only the schools exclusively for special children were used for the study. Surulere and Yaba in districts two and three were purposively selected for this study because most of the special needs schools in Lagos state are located in Surulere and Yaba. All the three State owned exclusive special needs schools in Surulere and Yaba were selected for the study, also it was necessary to include the Down Syndrome Foundation, a non-profit, non-governmental foundation which is recognised by Lagos State and exclusively dedicated only to persons with Down Syndrome, also located in Surulere, Lagos State. The reason was to increase the sample size for the study as this foundation has the largest population of children with Down syndrome in Lagos State. These schools already have confirmed cases of children with Down Syndrome. The desired sample size was 104, and sampling was proportionately done according to the size of the four schools selected. Simple random sampling was used to select children who met the inclusion criteria (children 5-18 years old with Down syndrome who has written informed consent from parents were selected). Selected children who were uncooperative, even with the assistance of the school teacher were excluded, also those who were absent from school were not included in the study.

A total of 91 children out of the 104 selected children were examined. This was because some children were uncooperative while some were absent from school due to ill health and other reasons on the day of eye examination. The following eye test was carried out for all children recruited for the study: Visual Acuity assessment at distance and near using the Lea symbol for distant/near testing (Brand, copm, country) depending on their abilities; External eye examination using pen light; internal eye exam using the ophthalmoscope; ocular alignment using alternate cover/uncover test without Prisms; Hirsberg tests also for ocular alignment (corneal reflex test); Non-cycloplegic retinoscopy using streak retinoscope; Auto-Refraction using Auto Ref-keratometer PRK-5000 Potec co Ltd.; Subjective refraction when possible using trial frame and lenses.

All examinations were done over the same period between 9am-1pm. Due to poor attention and concentration

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skills in this population, attention was motivated using fixation target, toys, clapping of hands and calling of names. These were used as strategies for improving attention and concentration. Stickers were given to each participating child for a job well done. Only the Lea symbols were used for testing the visual acuity. The criteria used for classification of refractive errors were; myopia ≥ -0.75DS, hyperopia ≥ + 0.75DS, astigmatism ≥ - 0.75DC.

Ethical approval for this study was obtained from the Ethics Committee of the Lagos State Ministry of Health and permission to enter the schools was obtained from the Lagos State Ministry of Education using the approval from the Ministry of Health. Consent was sought from the State Universal Basic Education Board (SUBEB) and the school authorities. Informed consent was obtained from parents of the children who participated in the study through the schools. Uncooperative children who were unable to be examined and whose parents or guardians did not give consent to the eye examination were excluded.

Statistical analysis of the data obtained was performed using the IBM Statistical Package for Social Sciences (SPSS) version 20.1. Chi-square and the independent t test statistics were used to test for association between variables. The results were represented using appropriate tables and figures, showing frequencies and percentages.

Results

A total of 91 participants were examined in this study. This comprised of 52 (57.1%) males and 39 (42.9%) females (Figure 1). The mean age of the 91 subjects was 13.6 ± 3.7 years with a median of 14.0 years. There was no statistical significant difference in the mean age of females (M=13.1, SD =3.9) and males (M=14, SD =3.6), (P = 0.280)

The distribution of presenting visual acuity is shown in Table 1. Visual acuity could not be determined in 17 right eyes and 16 lefts of the patients. A total of 48 (52.7%) subjects had normal or mild to moderated visual impairment (VA between 6/6 and 6/18) in each eye, 26 (28.6%) and 27 (29.7%) had low vision (VA <6/18 to 3/60) in the right and left eye respectively using the International Classification of Disease (ICD). There was improvement in visual acuity of the subjects after subjective refraction. The number of the participants with vision of 6/18 or better increased from 48 to 72 after best correction.

Of the total 91 participants, the refractive status could not be determined in four subjects (all male), while five subjects had emmetropia. The prevalence rate of refractive errors among children with Down's syndrome in the present study was 95.3% (95% CI = 88.9 – 97.4%). The prevalence of refractive errors in male and female participants was 93.8% (95% CI = 85.8 – 97.9%) and 94.9% (95% CI = 86.1 – 98.7%)respectively. There was no significant difference in the prevalence of refractive errors in male and female participants (p = 0.8331) (Table 3), In terms of the magnitude, the refractive errors ranged from – 11.00 DS to + 4.25 DS (spherical equivalent) while the astigmatism ranged from – 0.50 DC to – 4.00 DC Astigmatism was the most common refractive error being present in 61 (67.0%) of the subjects examined followed by hyperopia, 12 (13.2%) and myopia, 7 (7.7%).

The prevalence of astigmatism was further analysed by types. Out of the 61 subjects with astigmatism, 27 (44.3%) had hyperopic astigmatism followed by myopic astigmatism, 19 (31.1%); simple astigmatism, 12 (19.7%) and mixed astigmatism, 3 (4.9%). More male participants had hyperopic astigmatism (30.8%) while more female participants had myopic astigmatism (30.8%). However, there was no statistically significant relationship between the type of astigmatism and the gender of children with Down syndrome ($\chi^2 = 2.559$, p = 0.465).

Discussion

Refractive anomalies in patients with Down syndrome are very common and their incidence vary from 65% to 100%[31,32]. The present study is a further confirmation of refractive

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anomalies in patients with Down syndrome. In this study, refractive errors occurred in over half of the population of the children with Down syndrome examined. Out of the 91 children studied, 82 had one form of refractive error or the other. This result varies with the prevalence of refractive error among children without Down syndrome in Nigeria which ranges from 5% to 15% [25,27-30]. The distribution of refractive error in this study revealed that the most common form was astigmatism, followed by hyperopia and myopia. This supports the study by Adio et al.[21], where more than half (76.2%)of the 42 children with Down syndrome screened had refractive errors, also further supporting this finding are results in similar studies conducted within and outside Nigeria[8,9,31,32].

Of the 61 cases of astigmatism; hyperopic astigmatism was the highest, followed by myopic astigmatism, simple astigmatism and mixed astigmatism. The percentage was higher than that found in a population of children and young adults without Down Syndrome in various other studies[8,14,23]. The distribution in this study also supports studies carried out by Haugen et al.[9] where they reported that astigmatism was found to be the highest occurring refractive error, followed by hyperopia and myopia. The study however did not distinguish between the types of astigmatism present. In some other similar studies, by Kim and Hwang[24], Ljubic and Trajkovski[23] astigmatism was also found to have the highest occurrence followed by hyperopia and myopia.

In a study by Cregg et al.[17] of 123 children with Down syndrome, the most prevalent refractive error was hyperopic astigmatism which is in line with the result of this study. This trend contradicts that found in school children without Down syndrome of comparable age group where myopia was found to have the highest prevalence with environmental, hereditary, increased near work and recently computer or visual display suggested to play a role [25-28].

The high incidence of refractive errors in children with Down syndrome is believed to be caused by failure of emmetropisation process in Down syndrome. The change in spherical refractive error was found to be minimal throughout childhood and early adulthood. According to another study, the children had a specific development of astigmatism of oblique nature which was also found in this study and is suggested to be due to mechanical induction through the eyelids[12]. The thinning of corneal stroma may account for the steeper cornea and the high frequencies of astigmatism seen in Down Syndrome due to lower corneal rigidity. The cornea in Down syndrome is thought to be thinner and steeper and corneal shape plays a significant role in the development of astigmatism in Down syndrome. This may also predispose this population to keratoconus. Oblique astigmatism is commonly found in this group of patient population[9], Statistical test also found that there is no significant relationship between the type of astigmatism and the gender of participants in this study.

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In the analysis to determine if the refractive errors found in this population were gender dependent, it was found that the type of refractive error present was independent of the sex of the subjects and there was no significant difference in the prevalence of refractive error in male and female studied (p=0.8231). This corresponded to a study by Murthy[33] in which the distribution of refractive errors between boys and girls did not differ.

It is worth noting that esotropia was the most occurring ocular deviation seen among children with Down syndrome, only two cases of exotropia was seen. This is similar to findings in other studies[22,34]. The high occurrence of strabismus poses a high risk of amblyopia in this population and as such further buttressing the need for early intervention. Due to the magnitude of refractive errors found in this population, ranging between -11.00DS to +4.25DS and astigmatism from -0.50DC to- 4.00DC in this study, the need for correction to avoid amblyopia cannot be over emphasized because refractive errors has a significant impact on a child's education, development and life generally. It is important that effective strategies are developed to eliminate the barrier to refractive error correction in this population of intellectually disabled patients. The high prevalence of refractive error found in this study signifies a need for spectacle correction in Down syndrome than in a general population.

Comprehensive eye examination, regular vision screening, follow ups and appropriate remedial action must be taken by eye care practitioners. This calls for the need of appropriate spectacles for the correction of refractive errors and accommodation lag present as well as measures to care for the myriad of other ocular anomalies which may be present in this group.

Conclusion

Refractive error was found to be a significant cause of visual impairment among children with Down syndrome. Astigmatism was the most common type of refractive error, followed by hyperopia and myopia, while hyperopic astigmatism was the highest occurring type of astigmatism seen. The implication of uncorrected refractive error in causing amblyopia further emphasizes the need for early intervention in this population that is often neglected. The study revealed that refractive error in children with Down syndrome was not gender dependent. There was also no significant difference in the type of astigmatism found and the gender of the subjects. Provision of spectacles, necessary ophthalmic correction and management will invariably improve the visual efficiency, education, general wellbeing and the quality of life for these groups of children, a step closer to acceptance, inclusion and integration into the society.

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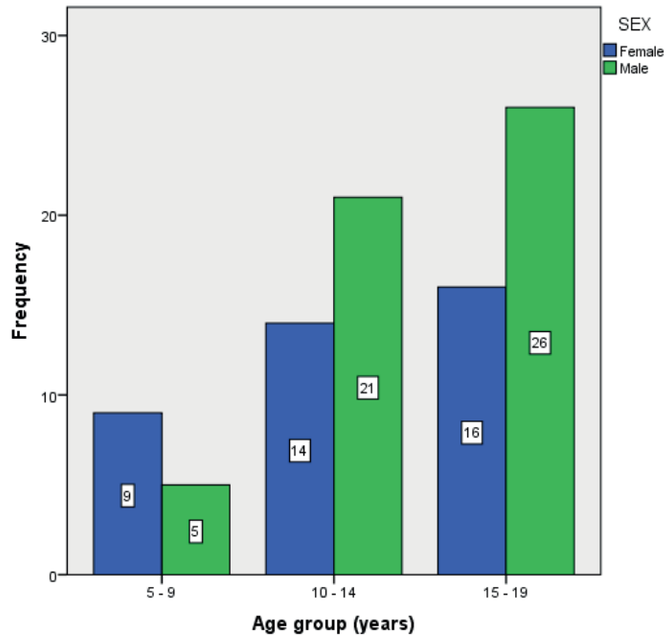

Fig 1: Age and gender distribution of the study subjects

Table 1
Distribution of presenting visual acuity for the right and left eye of subjects

Visual acuity*	Right Eye (%)	Left Eye (%)
Better than 6/6 to 6/12	42 (46.1)	42 (46.1)
Worse than 6/12 to 6/18	6 (6.6)	6 (6.6)
Worse than 6/18 to 6/36	11 (12.1)	13 (14.3)
Worse than 6/36 to3/60	15 (16.5)	14 (15.4)
Undeterminable	17 (18.7)	16 (17.6)
Total	**91(100.0)**	**91(100.0)**

*Normal or mild to moderated visual impairment (VA between 6/6 and 6/18), low vision (VA <6/18 to 3/60)

Table 2
Distribution of best corrected visual acuity in the right and left eye of subjects

Visual acuity*	Right Eye (%)	Left Eye (%)
Better than 6/6 to 6/12	68 (74.7)	69 (75.8)
Worse than 6/12 to 6/18	3 (3.3)	3 (3.3)
Worse than 6/18 to 6/36	2 (2.2)	1 (1.1)
Worse than 6/36 to 3/60	0 (0.0)	1 (1.1)
Undeterminable	18 (19.8)	17 (18.7)
Total	**91(100.0)**	**91(100.0)**

*Normal or mild to moderated visual impairment (VA between 6/6 and 6/18) low vision (VA <6/18 to 3/60)

Table 3
Distribution of refractive error by gender among the study subjects

REFRACTIVE ERRORS	MALE n (%)	FEMALE n (%)	TOTAL n (%)	p-value
Emmetropia	3 (3.3)	2 (2.2)	5 (5.5)	0.8331
Hyperopia	7 (7.7)	5 (5.5)	12 (13.2)	
Myopia	5 (5.5)	2 (2.2)	7 (7.7)	
Astigmatism	31 (34.0)	30 (32.9)	61 (67.0)	
Antimetropia	2 (2.2)	0 (0.0)	2 (2.2)	
Undeterminable	4 (4.4)	0 (0.0)	4 (4.4)	
Total	**52 (57.1)**	**39 (42.9)**	**91 (100.0)**	

p>0.05 Test not significant @95% confidence interval using Chi-square test statistics

Table 4
Absolute frequency and relative frequency of refractive error in children (n=86) with down syndrome

Prevalence	Absolute Frequency (n)	Relative Frequency (%)
Refractive error.	82	95.3 (95%CI 88.9 – 97.4%)
No Refractive error	4	4.7
Total	**86**	**100**

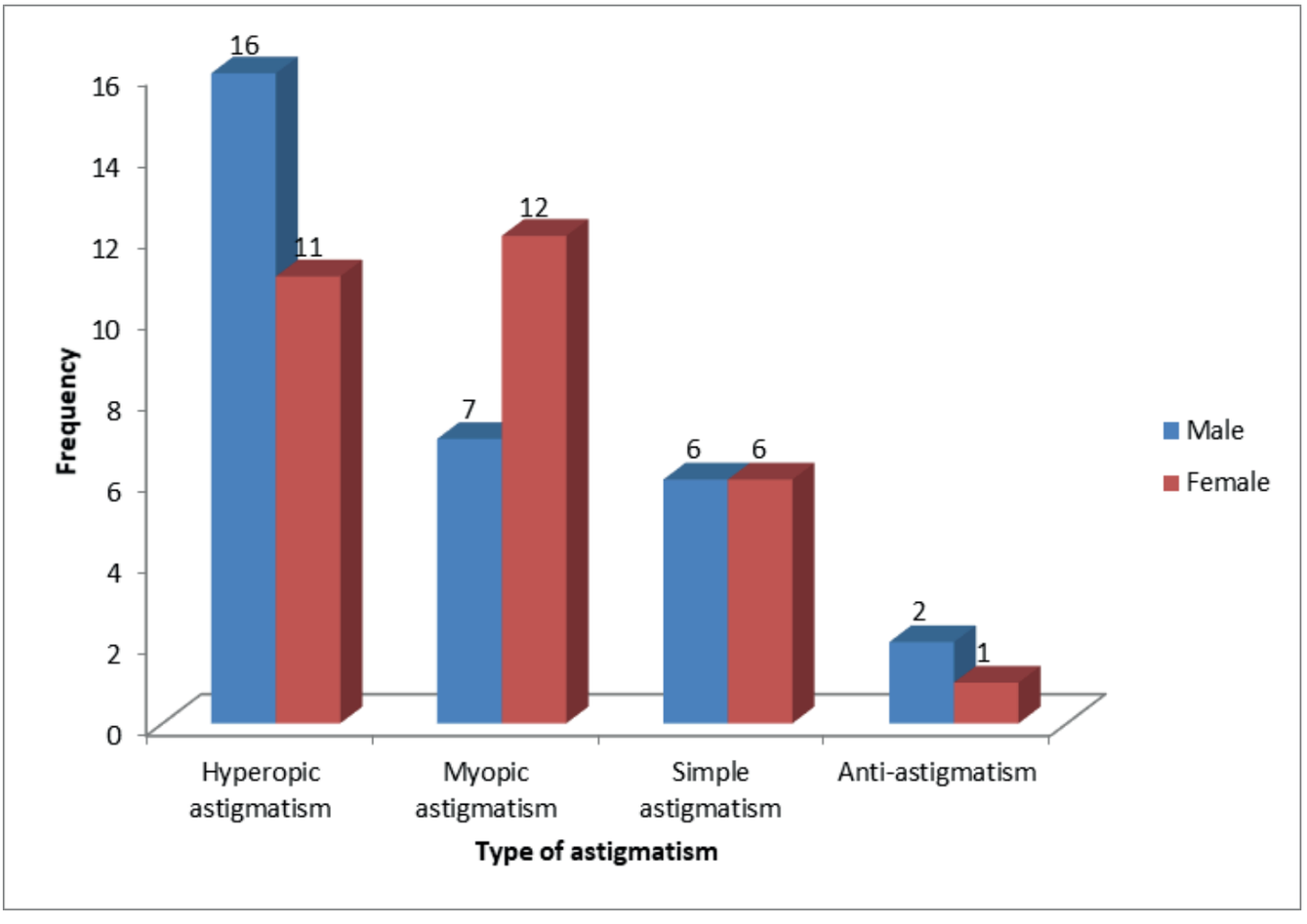

Figure 2: Distribution of the type of Astigmatism by gender among children with Down syndrome