

Ophthalmic Disorders among Children with Down Syndrome: An Insight into Parents' Knowledge, Awareness Level and Attitude

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ABSTRACT

Introduction: Down syndrome is a genetic disorder caused by the presence of an extra copy of chromosome 21. Ophthalmic disorders affecting children are even more frequent in those with DS and sometimes tend to be more severe.

Objectives: This study attempts to probe into parents' knowledge, awareness and attitude regarding management of ophthalmic disorders among children with Down Syndrome (DS).

Methods: This cross-sectional study was conducted during the World Down Syndrome Day event on March 21, 2013 in Jeddah, Saudi Arabia. Parents of children with DS who attended the event were requested to participate in the survey that was conducted in the form of a questionnaire. The questionnaire was designed to assess parents' knowledge, awareness level and attitude toward common ophthalmic disorders affecting children with DS. Descriptive and analytical statistical methods were used to analyze the collected data written between Mar 21, 2013 and February 2014.

Results: A total of 72 parents participated in the questionnaire-based survey. Among these, 49 (74.2%) reported taking their children for ophthalmology visits, and 27 (37.5%) for routine visits at varying intervals. Among the enumerated eye conditions, the most frequently reported one was up-slanting eyes (52.8%), followed by epicanthal folds (38.9%) and myopia (33.3%). However, up to 62.5% of the parents were not aware

about other eye conditions such as nystagmus, brush field spots, and keratoconus.

Conclusions: Most parents are not aware of the ophthalmologic disorders affecting children with DS, suggesting a need to embark upon large scale awareness campaigns to educate the parents of children with DS. Parent's awareness is crucial for early diagnosis and proper follow-up of ocular disorders in children with DS.

Keywords: Down Syndrome, Eye Disorders, Ocular Diseases, Parents, Knowledge.

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INTRODUCTION

Down syndrome (DS), also known as trisomy 21, is a genetic disorder caused by the presence of an extra copy of chromosome 21. It is the most common chromosomal abnormality in humans. In Saudi Arabia, the incidence of DS has been reported in 1 of 554 live births;¹ while in the United States, 1 in every 691 of births each year, according to the Centres for Disease Control (CDC).² Common ophthalmic disorders (eye problems) that occur in any group of children are even more frequent in those with DS and sometimes tend to be more severe.^{3,4} Morphologically, children with DS have obliquely placed eyes, more than normally distant internal canthi and very narrow palpebral fissure. In addition, several ocular abnormalities such as strabismus, nystagmus, keratoconus, cataract, hyperplasia of the iris, and refractive errors

occur more frequently in children with DS than in their healthy counterparts, with prevalence ranging from 7.6% to 82%, depending on the abnormality.³⁻⁷ For example, the prevalence of refractive errors (hyperopia, myopia, and astigmatism) reported in the medical literature ranged between 3% and 62.3% among DS children,⁶⁻¹¹ while prevalence of strabismus, more frequently esotropia than exotropia, ranged between 5% and 35%.^{7,11-13} Other ophthalmic disorders reported in patients with DS include high incidence of conjunctivitis (5.6%–6.7%) and glaucoma (6.7%).^{6,14}

A study conducted at the University Eye Hospital of Ljubljana, Slovenia, reported nystagmus (29.2%), esotropia (26.1%), epiphora (21.5%), brush field spots (16.9%), lens opacities

(12.3%), abnormalities of the retinal vessels, foveal hypoplasia or retinal pigment epithelium hyperplasia (32.2%), and optic disc pallor (7.6%) in children with DS. Hyperopia (36.9%) was the most frequent refractive error in the group, followed by astigmatism (29.2%) and myopia (24.6%).¹⁵ Down Syndrome is almost always diagnosed at birth and requires lifelong parenting and supervision. Therefore, parents' awareness is crucial in the prevention and treatment of ophthalmic disorders in children with DS. However, there seems to be a serious dearth of medical evidence highlighting the awareness and knowledge level of parents of children with DS about ophthalmic disorders and their attitude towards this frequently reported complication of DS.

The current study was carried out among parents of children with DS to investigate their levels of awareness, knowledge and attitudes toward the most frequently reported ophthalmic disorders in DS.

MATERIALS AND METHODS

A cross-sectional design, questionnaire-based survey was conducted among parents of children with DS who attended the World Down Syndrome Day event, a public awareness campaign organized on March 21, 2013 at King Abdulaziz University Hospital in Jeddah, Saudi Arabia. The questionnaire was presented to all attendees and explained by the investigators, to be auto-filled by volunteering participants. The informed consent was orally obtained from all participants prior to inclusion and the

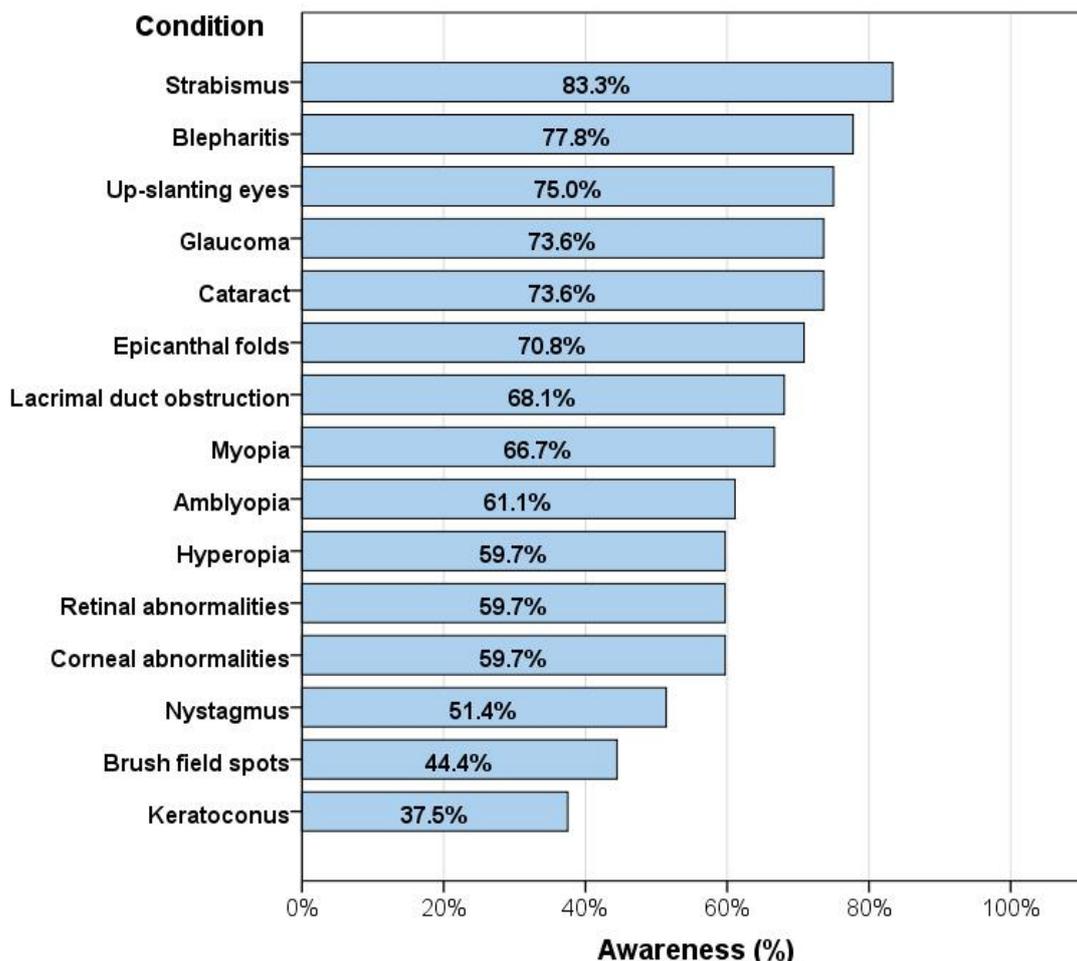
study was approved by the Biomedical Ethics Research Committee of King Abdulaziz University.

In an attempt to increase the response rate, the questionnaire was designed in a simple fashion including the following items: 1) child's age and gender; 2) history of visits to an ophthalmologist; 3) frequency of routine visits to a physician; 4) parent's source of information on ophthalmic diseases; 5) parent's awareness about 15 listed eye conditions (e.g. up-slanting eyes, epicanthal folds, myopia, hyperopia, strabismus, etc.) and; 6) parent's knowledge about the past or present existence of each of these conditions in their children. The questionnaire underwent face and content validation prior to study.

Statistical Analysis

Data was analyzed using the Statistical Package for the Social Sciences, version 20.0 (SPSS Inc., Chicago, IL, USA). Descriptive statistics was performed to analyze all variables including response patterns of the participants. According to the number of eye conditions acknowledged, participants were divided into three knowledge-level categories: A) weak (0 to 4 conditions); B) fair (5 to 9 conditions); and C) good (10 to 15 conditions). A cross-tabulation was performed to correlate the level of knowledge with the frequency of ophthalmology clinic visits, as well as with the existence of ophthalmologic history in the affected child. Chi-square test was used to analyze the correlation between categorical variables. Results were considered statistically significant ($P < 0.05$).

Figure 1: Awareness about different eye conditions in Down Syndrome among parents of afflicted children. (Bars present the percentage of parents who declared being aware of the given condition.)



RESULTS

Demographic characteristics of diseased children

All the parents attending the awareness campaign agreed to complete the questionnaire (n=72; response rate, RR=100%). Children demographics showed mean±SD age= 9.2±6.9 [1, 36] years, 31 (43.1%) males and 36 (50.0%) females; while child's gender was not indicated in 5 (6.9%) of cases.

Awareness about ophthalmic disorders

Figure 1 shows the percentage of parents who declared being aware about the given condition, which has been explained by the investigator. Awareness was highest for strabismus (83.3), followed by blepharitis (77.8%) and up-slanting eyes (75.0%); whereas, nystagmus (51.4%), brush field spots (44.4%) and keratoconus (37.5%) had the lowest awareness rates.

Table 1: Parent-reported ophthalmic disorder in children with Down Syndrome.

Condition	Present in or experienced by child					
	Yes		No		Do not know	
	Freq.	%	Freq.	%	Freq.	%
Strabismus	18	25.0	42	58.3	12	16.7
Blepharitis	18	25.0	38	52.8	16	22.2
Up-slanting eyes	38	52.8	16	22.2	18	25.0
Cataract	4	5.6	49	68.1	19	26.4
Glaucoma	1	1.4	52	72.2	19	26.4
Epicanthal folds	28	38.9	23	31.9	21	29.2
Lacrimal duct obstruction	13	18.1	36	50.0	23	31.9
Myopia	24	33.3	24	33.3	24	33.3
Amblyopia	7	9.7	37	51.4	28	38.9
Hyperopia	12	16.7	31	43.1	29	40.3
Corneal abnormalities	2	2.8	41	56.9	29	40.3
Retinal abnormalities	1	1.4	42	58.3	29	40.3
Nystagmus	7	9.7	30	41.7	35	48.6
Brush field spots	0	0.0	32	44.4	40	55.6
Keratoconus	0	0.0	27	37.5	45	62.5

More than one condition may be experienced by a single child; awareness rate is calculated as the percentage of parents knowing whether their child has or has not the given condition.

Table 2: Parents-reported ophthalmic care among children with Down Syndrome.

Variables	Frequency	Percentage
Number of eye conditions in diseased child		
0	6	8.3
1-2	36	50.5
≥3	30	41.7
Diagnosis site of eye disorder		
Ophthalmology clinic	41	56.9
Pediatric clinic	5	6.9
Family medicine	1	1.4
Other	11	15.3
No answer	14	19.4
History of eye surgery	59	81.9
Blindness	0	0.0
Ever took child for a visit to an ophthalmologist (n=66)		
Yes	49	74.2
No	17	25.8
Ever took child for follow up visits (n=66)		
Yes	27	40.9
No	39	59.1
Frequency of follow up visits (n=49)		
Only when needed	22	33.3
Every 6 months	19	28.8
Once a year or more	8	12.1

Parent’s awareness and knowledge about their children’s eye condition

Among the enumerated eye conditions, the most frequently reported one was up-slanting eyes (52.8%), followed by epicanthal folds (38.9%) and myopia (33.3%). However, some parents were not aware whether their children had keratoconus (62.5%), brush field spots (55.6%) and nystagmus (48.6%) (Table 1). On the other hand, half of the parents (n=36; 50.0%) reported that their children had or have experienced 1 or 2 of the enumerated eye conditions, 30 (41.7%) reported ≥3 eye conditions; and 6 (8.3%) reported none of the enumerated eye conditions among their children. Other parent-reported ophthalmological conditions showed a history of eye surgery in 59 (81.9%) cases and no case of blindness. Regarding the site of diagnosis, 41 (65.1%) of the participants reported that their child’s ophthalmologic disorder was diagnosed at an ophthalmology clinic, 5 (7.9%) at a paediatric clinic, and only one case was diagnosed at a family medicine clinic, while the remaining 11 (17.5%) cases were diagnosed at other locations. Among all the participating parents, 11 (17.5%) reported that the eye disorder was diagnosed at child’s birth (Table 2).

Assessment of attitude towards ophthalmic conditions showed that 49 (74.2%) parents reported that their children had never attended an ophthalmologist clinic; among these, 27 (55.1%) had regular follow-up visits every 6-12 months (Table 2).

Sources of knowledge about ophthalmological disorders

Analysis of sources of knowledge about ophthalmological disorders showed that the most frequently reported source was the internet (n=41; 56.9%), followed by ophthalmology clinics (n=28; 38.9%) and books (n=22; 30.6%). Other sources reported by parents included television, media publications and “word-of-mouth” information (Figure 2).

Impact of knowledge on care attitude

There was no correlation between parents’ knowledge level (number of eye conditions correctly identified) and attitude towards ophthalmological care (frequency of ophthalmology clinic visits). Analysis showed that 68.7% of parents with good knowledge declared taking their children for regular ophthalmology visits versus 80.0% and 78.6% with fair and weak knowledge, respectively (p=0.610) (Table 3).

Figure 2: Sources of knowledge about ophthalmological disorders among parents of children with Down syndrome.

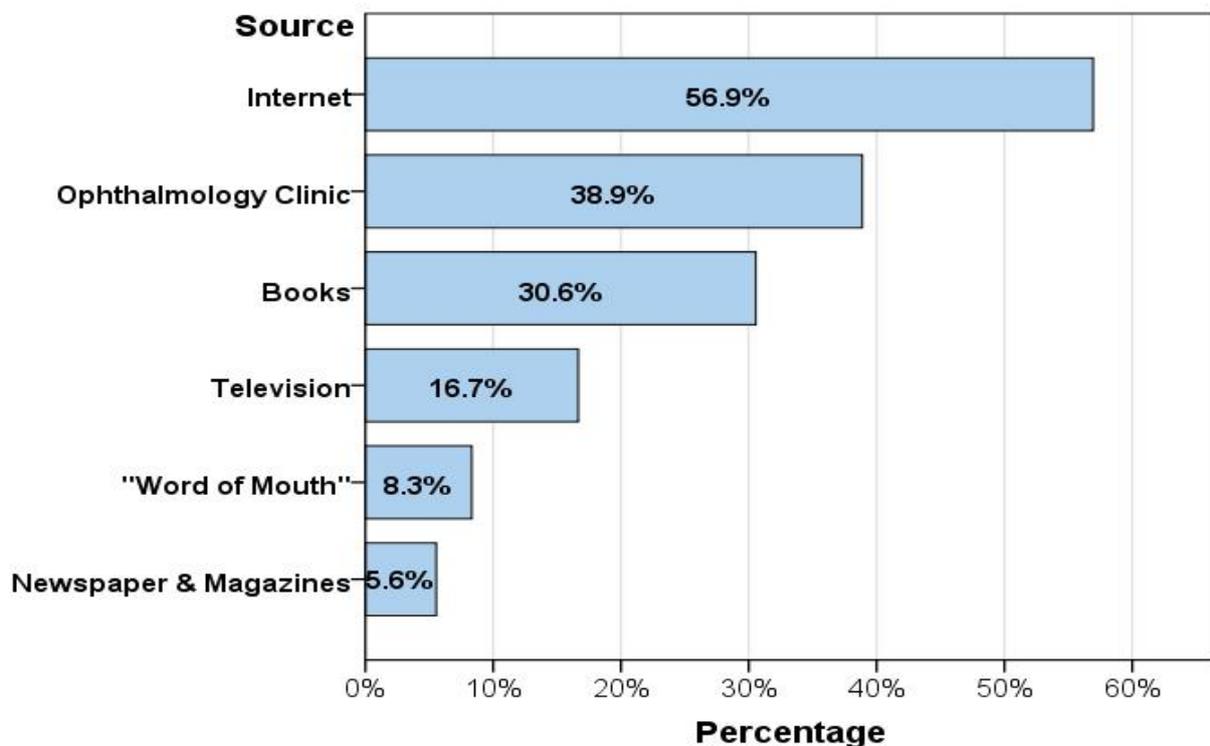


Table 3: Relationship of ophthalmological care attitude and parents’ knowledge.

Attitude	Knowledge level			Total	P-value
	Weak (n=14)	Fair (n=20)	Good (n=32)		
Never visited a clinic	3 (21.4)	4 (20.0)	10 (31.3)	17	.610
Visited a clinic	11 (78.6)	16 (80.0)	22 (68.7)	49	

Data are presented as frequency (percent); weak knowledge: knowledge about ≤3 conditions; fair knowledge: knowledge about 4-7 conditions; good knowledge: knowledge about ≥ 8 conditions; because of some missing data, the total number of participants does not sum up to the total (N=72); test used: Chi-square test.

DISCUSSION

Being the first attempt ever to assess parents' awareness levels and perception of common eye disorders among their children with DS, this is a novel study in its objective. Therefore, the findings of this study could not be contrasted with other previous data. However, findings emanating from studies performed among the general population revealed that parents generally fail to recognize the commencement of visual disturbances in their children. A recent study by the Vision Council of America reported that 27% of parents and guardians who had not taken their children for an eye check-up believed that their children were too young to develop vision problems.¹⁶ Furthermore, the previous study highlighted that most people including parents, believe that eye check-ups are only necessary when a vision problem appears or when a pair of spectacles is needed.¹⁶

In this study, 49 of 66 parents (74.2%) reported taking their children for a visit to an ophthalmologist and only 27 (55.1%) admitted taking their children for regular follow-up visit. A low rate of regular follow-up may suggest that a high proportion of DS-related ophthalmologic diseases remain undiagnosed. In other cases, high proportion of noncompliance may be associated with diagnosed cases being lost to follow-up. Underdiagnoses, under-reporting and delayed diagnosis of visual disorders is likely to be the tip of the iceberg among all the medical concerns in DS. This results in a considerable number of cases diagnosed in adulthood, with an increased risk of end-stage eye complications like impaired vision.¹⁷

Despite of having ran the questionnaire in the local language, in simple words and with limited use of medical terminology, we found that ocular conditions were unfamiliar to a significant proportion of parents, thereby increasing the hypothetic proportion of children being undiagnosed. On the other hand, with regards to the main objective of this study, varying levels of awareness and knowledge between the specific eye disorders was observed. Common ophthalmic disorders such as myopia, amblyopia and hyperopia were insufficiently known by parents of children with DS, as evidenced by the relatively high proportion (33%-40%) of parents being unaware about such conditions.

Regarding knowledge sources, parents mainly obtained information about ophthalmic disorders from the internet and ophthalmology clinics. Because eye disorders are more common in children with DS,^{5,7} it is crucial for parents of children with DS to be more aware of ophthalmic diseases that might affect their children. Health care providers should organize public awareness campaigns to promote routine regular follow-up visits in children with DS, including ophthalmologic visits. Education programs could be organized at ophthalmology clinics, using visual and audio material focusing on eye health in children with DS. Parents' participation is crucial to the care of patients with DS. Therefore, improving parents' knowledge and raising their awareness about the increased ophthalmologic risk in their children is a critical determinant in the primary and secondary prevention of ophthalmic disorders, ensuring early diagnosis and regular follow-up.

This study has some limitations that may warrant a discussion. First, the small sample size (n=72) is not representative of the population of children with DS in Jeddah. Second, participants were recruited during an awareness campaign, which may constitute an important selection bias as the voluntary attendance

of such events suggests the prior existence of a basic level of awareness. Third, we did not collect participants' demographic data, mainly for the sake of simplification. However, findings of a study conducted in families of children with DS showed that family demographics were likely to be important factors for healthy family function and consequently for the quality of care of children with DS.¹⁸ Further large scale multi-centric evaluations are warranted to reach more robust results and conclusions regarding this issue, which may provide decisive and constructive information to improve the management of patients with DS in the future. With regards to findings from the current study, it is highly recommended to educate parents of children with DS regarding the common eye diseases, besides other health concerns of this category of people. For that purpose, Down Syndrome Medical Interest Group guidelines can be used, which were developed for people with DS with an objective to outline the basic medical essentials of follow-up of their disease.¹⁹

CONCLUSION

Major lacunae exist in knowledge and awareness about the ophthalmic risk associated with DS among parents of afflicted children. Epicanthal folds, up-slanting eyes and myopia were the best known among the investigated ophthalmologic disorders; probably because they were the most frequently experienced by the participant's children. On the other hand, more common ophthalmologic conditions such as glaucoma, cataract, strabismus, nystagmus and lacrimal duct obstruction were associated with lower awareness levels.

Given that parents' participation is crucial to the care of children with DS, parents' education about the increased ophthalmologic risk in their children is a determinant factor for early diagnosis and regular follow-up of ophthalmologic disorders in these patients. We recommend the use of Down Syndrome Medical Interest Group guidelines, developed for people with DS with an objective to outline the basic medical essentials of follow-up of their disease.

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