Ophthalmologic Screening in A School for the Hearing Impaired in Turkey

İşitme Engelliler Okulundaki Çocuklarda Göz Sağlığı Taraması

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ABSTRACT Objective: The aim of this study was to screen the ophthalmic disorders in a school for the hearing impaired and to draw the attention to the necessity of early educational and career modifications of children with Usher syndrome. **Material and Methods:** The study population was 208 of 214 students in a school for hearing impaired in Ankara; ages ranged from 7 to 19 years. All students underwent an ophthalmologic examination, including uncorrected and corrected visual acuity measurement, assessment of manifest refraction, slit lamp biomicroscopy and fundoscopy. Cover/uncover test was done to detect a tropia of more than 10 prism diopters. Students with an ophthalmic pathology were invited for a detailed eye examination. Chi square test and binary logistic regression analysis were used for statistical analysis. **Results:** Of the 208 students, 72 (34.6%) had an ocular abnormality. One hundred eighty four (88.5%) children presented with normal fundus findings. Of the 24 patients with an ophthalmoscopic finding, twelve (5.8%) were diagnosed with retinitis pigmentosa, three with Waardenburg syndrome and four with atypical retinal pigmentary changes. Female gender ($p< 0.05$) and the presence of a refractive error ($p< 0.0001$) were significantly correlated with the existence of ophthalmoscopic findings. **Conclusion:** Early referral to an ophthalmic examination is mandatory for the hearing impaired children. In previous studies, higher frequency of ophthalmic disorders in the hearing impaired population had been reported; present study also emphasizes the necessity of educational and career modifications in hearing impaired children with retina pathologies. Detection and early intervention of ophthalmologic abnormalities is important for the normal development of cognitive and social skills and also for education and career modifications.

Key Words: Hearing loss; vision screening; visually impaired persons; Usher syndromes; retinitis pigmentosa; education, special

ÖZET Amaç: Çalışmanın amacı, işitme engelliler okulu öğrencilerinde göz patolojilerinin belirlenmesi ve Usher sendromu olan çocukların erken dönemde eğitim ve meslek düzenlemelerinin gerekliliğine dikkat çekmektir. **Gereç ve Yöntemler:** Çalışma grubu Ankara'daki bir işitme engelliler okulundaki, 7-19 yaşları arasındaki 214 öğrencinin 208'ini kapsamaktadır. Tüm öğrencilere göz muayenesi uygulanmış; düzeltmeli ve düzeltmesiz görme keskinliği, manifest refraksiyon, biyomikroskopi ve fundoskopik muayene yapılmıştır. Açma kapama testi ile 10 prizm diyoptri üzerindeki göz kaymaları saptanmıştır. Herhangi bir göz patolojisi olan çocuklar ayrıntılı bir göz muayenesine davet edilmiştir. İstatistik değerlendirme için Ki-kare ve "binary logistic regression" analizi kullanılmıştır. **Bulgular:** İki yüz sekiz öğrencinin 72 (%34.6)'sinde bir göz patolojisi gözlenmiştir. Yüz seksen dört (%88.5) öğrencide normal fundus bulguları saptanmıştır. Oftalmoskopik bulguları olan 24 hastanın 12 (%5.8)'sinde retinitis pigmentoza, üçünde Waardenburg sendromu, dördünde ise atipik retinal pigmenter değişiklikler görülmüştür. Kadın cinsiyet ($p< 0.05$) ve kırma kusuru varlığı ($p< 0.0001$) ile oftalmoskopik bulguların varlığı arasında önemli korelasyon saptanmıştır. **Sonuç:** İşitme engelli çocuklarda göz patolojilerinin oranı yüksektir ve erken dönemde göz muayeneleri yapılmalıdır. Ülkemizde ve yurtdışında yapılan önceki çalışmalarda da benzer oranlarda göz patolojisi varlığı saptanmış; fakat uzun vadede eğitim ve meslek düzenlemesi gereği yeterince vurgulanmamıştır. Göz patolojilerinin saptanması, bilişsel ve sosyal yeteneklerin geliştirilmesi ve eğitim ile meslek seçiminde düzenlemelerin yapılması açısından gereklidir.

Anahtar Kelimeler: İşitme kaybı; görme taraması; görme özürlü bireyler; Usher sendromları; retinitis pigmentoza; eğitim, özel

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The prevalence of ophthalmologic abnormalities in the hearing impaired and deaf children is reported approximated 50 percent.[1-3] Vision is the principal way of learning and interacting with the family and the environment. Children with sensorineural hearing impairment and deafness have more dependence on their visual function to gather information and improve social skills. The need for early detection of ophthalmologic abnormalities is important for hearing impaired children in order to develop normal educational, social and cognitive skills.

The ophthalmologic screening of the hearing impaired is also important in some other aspects. The ophthalmologic examination may give additional information about the exact diagnosis of the cause of hearing loss. Another important aspect is the detection of retinal pigmentary changes in the hearing impaired children. Approximately 8.6-22% of hearing impaired and deaf students has some kind of pigmentary retinopathy, which has the potential to end with deaf-blindness.[1] For this patient group, educational modifications and career planning are important, because some of these patients will additionally have visual impairment beyond early adulthood.[4-6] Another concern for these patients is the need for professional support to the family and children for adapting to this challenging situation.

Besides ophthalmologists, the information in this article is also important for Ear, Nose and Throat specialists, pediatricians, primary health care practitioners and special educators for their early encounter with hearing impaired and deaf children.

MATERIAL AND METHODS

The study population was the students of Yahya Özsoy Primary School for Hearing Impaired in Ankara. This screening was performed by the written request of the school administration, aiming for the early-detection of visual disturbances which might interfere with the academic performance of some students. The school is the largest primary school for hearing impaired in Turkey and one of the three schools for the hearing impaired in Ankara. According to the legal requirements, these children should be educated in classrooms composed of fewer than 10 students and some part of the educational period should be in mainstream educational institutes. Accommodations and educational activities are supported by the Turkish Ministry of Education and children from remote geographic areas can stay in school on a 24 hour basis.

A level of 70 dB hearing loss was a prerequisite to be accepted in these schools in Turkey at the time of the study. This level is accepted as severe hearing loss and these students do not hear conversations. They are visually oriented and have serious problems of speech and language development.[1]

Socio-demographic and medical history was recorded by two authors (DÇ, OÖ) using face to face interview method and also by using the health and education files of the students. A questionnaire was enrolled to the students, which included questions about the presence of consanguinity between parents, presence of visual problems during night and day, history of hearing impairment and visual disturbances of any member within the family, prenatal diseases and age of the parents during pregnancy. Three special educators experienced in sign language assisted the research team with each student, directing the questions and communicating with students using sign language. Ankara University Faculty of Medicine Department of Public Health Administration approved this screening project. The school administration had taken consents of the families of the students for the ophthalmic examination and the study was performed in concordance with the tenets of Declaration of Helsinki.

Of 214 students, 208 were evaluated at their school; the participation rate of study was 97.1%. All students underwent an ophthalmologic examination which included uncorrected and corrected visual acuity measurement with a tumbling E Snellen chart, assessment of manifest refraction, slit lamp biomicroscopic examination and fundoscopic examination with a 90 diopter (D) lens. Cover/uncover test was done to detect the presence of a tro-

pia more than 10 prism diopters. Ophthalmologic assessments were performed by two ophthalmologists (Aİ, DC). Children who present with visual acuities between 0.8 and 1.0 with a Snellen acuity chart (Snellen equivalents converted to decimal ratios), with no ocular complaints and a manifest refraction between -1.00 and + 1.00 D; no anterior segment, retinal and oculomotor abnormality were considered as ophthalmologically "normal". Any student who presented with a corrected monocular visual acuity of 0.6 or less; visual disturbances during night and day, manifest refraction of more than +1.0 D and -1.00 D; tropia of more than 10 prism diopters and suspected pathology with biomicroscopy and ophthalmoscopy were invited to ophthalmology office for a detailed eye examination.

Astigmatism of less than 1 D with manifest refraction was accepted as spherical error or emmetropia. Refractive errors were measured using the spherical equivalents of hyperopes and myopes of the eye with the higher refractive error. Anisometropia is defined as 1 diopter (spherical equivalent) difference of manifest refractive error between the eyes. If best corrected visual acuity was less than 0.6 and if there was more than two lines of Snellen visual acuity difference between the eyes, as well as absence of an organic pathology, amblyopia was considered. Children with, and suspected of, retinal pigmentary changes and who stated to have night vision disturbances with a family history of night blindness were also invited for a detailed eye examination.

These selected patients underwent a complete ophthalmologic examination including determination of best corrected visual acuity, a slit lamp examination of the anterior segment and retinal examination with a 90 D lens with full pupillary dilation. These examinations have been performed by two authors (Aİ, DC) at Ankara University Faculty of Medicine, Department of Public Health, Visual Impairment Rehabilitation Center.

For statistical analysis, Chi square test and binary logistic regression analysis were used; a "p value" less than 0.05 was considered as significant.

RESULTS

The students' age range was between 7 to 19 years with a mean of 12.83 ± 3.0. One hundred eleven (53.4%) were boys and ninety seven (46.6%) were girls. Of the 208 students examined, 72 (34.6%) had an ocular abnormality. One hundred forty five (69.7%) did not have a manifest refraction error equal to or more than ±1.0 D in the eye with higher refractive error. A hyperopia between + 1.00 and + 3.00 diopters was detected in 19 (9.1%) students and a hyperopia more than +3.00 was detected in three (1.4%) eyes. In the eyes with a higher refractive error, myopia between 1.00 and 5.00 diopters was found in six (2.9%) and myopia more than 5.0 D in one (0.5%) of the eyes. Astigmatism between 1.00-1.75 D was found in 25 (12.0%) children and astigmatism equal to or more than 2.0 D was measured in eight (3.8%) eyes. No astigmatism was found in 173 children, 16 (7.7%) children presented with myopia and 14 (6.7%) had a hyperopic astigmatism. No anisometropia was detected in 195 (93.8%) children, four (1.9%) children had mixed or irregular astigmatism. An anisometropia of more than 1 D is detected in 11 (5.3%) patients. The refractive data of eyes with a higher refractive error are listed in the table (Table 1).

A manifest strabismus was observed in five (2.4%) children; keratoconus and cataract interfering with vision were detected in two children. Seventeen (8.2%) of the children had amblyopia. Three students were mentally retarded and none of them could be evaluated for visual acuity measurement.

As for ophthalmoscopic findings (Table 1), 184 (88.5%) children were classified as having normal fundus findings. Twelve (5.8%) children were ddiagnosed with retinitis pigmentosa by typical retinal appearance, and these children were classified as Usher Syndrome. One child had traumatic optic atrophy, and two children had myopic degeneration. Three patients had (1.4%) Waardenburg syndrome, which is characterized by white forelock, heterochromia iridis, bilateral deafness, telecanthus and patchy fundus depigmentation. Macular abnormalities were observed in two children. Four patients detected to have retinal pigmentary chan-

TABLE 1: Ophthalmologic findings in hearing impaired students.

Ophthalmic Findings	n	%
Refractive error*		
No refractive error	145	69.7
Hyperopes (1-3 D)	19	9.1
Hyperopes (3.25 D and higher)	3	1.4
Myopes (1-5 D)	6	2.9
Myopes (5.25 D and higher)	1	0.5
Astigmatism (1-1.75 D)	25	12.0
Astigmatism (2 D and higher)	8	3.8
Unable to evaluate	1	0.5
Fundus findings		
Normal	184	88.5
Retinitis pigmentosa	12	5.8
Pale optic nerve (traumatic)	1	0.5
Myopic degeneration	2	0.9
Waardenburg syndrome	3	1.4
Retinal pigmentary changes	4	2.0
Macular abnormalities	2	0.9

* Refractive error data of the eye with higher refractive error.
No visual acuity data of 3 mentally retarded children.

ges; a differential diagnosis could not be carried out with fundus findings only. Rubella retinopathy or other syndromes which cause retinal pigmentary changes associated with hearing impairment could be the etiologic factor in these patients.

Factors associated with ophthalmoscopic pathologies are shown at the table (Table 2). Gender ($p< 0.05$) and refraction errors ($p< 0.0001$), especially a manifest astigmatism equal to or more than 1D were significantly correlated with the existence of fundus pathologies. Consanguinity of parents, family history of visual or hearing impairment and the age of siblings were not significantly correlated with the existence of fundus pathologies.

DISCUSSION

Of the 208 deaf students screened, 72 (34.6%) had an important ocular pathology. Sixty two (29.8%) had a manifest refraction of myopia, hyperopia or astigmatism, more than 1 D. One hundred forty five (69.7%) had a refractive error less than 1 D. The prevalence of each type of refractive error is presented in Table 1.

The frequency of refractive errors in a normal school population in Turkey was reported around 10%.[7] This frequency is less than the half of the frequency of refractive errors in the hearing impaired population in this study. Kvarnström et al. reported the prevalence of ametropia as 7.7% in a visual screening of Swedish children, and Donnelly et al. also reported 5.82% ametropia in a population based study in Ireland.[8,9] Although there are discrepancies for defining the "abnormal" values for refractive errors in these different studies; the high prevalence of refractive errors in the hearing impaired students can easily be noticed. This relative increase in the frequency of refractive errors strongly emphasizes the need for an ophthalmologic screening in the hearing impaired and deaf population.

In a similar study in Turkey, almost the same frequency (29.8%) was reported for refractive errors in deaf children.[2] Although different refraction thresholds were used and cycloplegic refraction was applied to all students in that study, the results were quite similar with present study. We chose not to perform a cycloplegic refraction to students who had no visual complaints, who presented with a visual acuity of more than 0.6 and a manifest refraction less than ± 1.0 D for myopia, hyperopia and astigmatism. According to our findings, a manifest refraction examination is sufficient and a cycloplegic refraction is not needed in this patient group for screening purposes.

Increased prevalence of astigmatism in the hearing impaired and deaf population has been reported and the reported prevalence in this study (11.1%) was greater than six-fold compared to normal eyes.[1] Using the same threshold (1 D), we found 15.8% of astigmatism in the hearing impaired population. This amount of astigmatism may cause amblyopia and should be corrected as early as possible. Leguire et al. performed refraction examination with paralysis of accommodation, however, in the present study manifest refraction was performed.[1] We believe, if we had performed cycloplegic refraction, the prevalence for refractive errors would have been slightly higher.

TABLE 2: Variables that may be associated with fundus findings.

	Fundus Pathology					
	No		Yes			
Variable	**n**	**%**	**n**	**%**	**Odds Ratio**	**(95%) Confidence Interval**
Gender*						
Male	103	92.8	8	7.2		
Female	81	83.5	16	16.5	**2.543**	1.037-6.237
Age (NS)						
7-10	45	84.9	8	15.1	1.955	0.635-6.019
11-14	72	87.8	10	12.2	1.528	0.526-4.435
15+	66	91.7	6	8.3		
Age of mother at birth (NS)						
34 and lower	151	90.4	16	9.6		
35 and higher	9	81.8	21	18.2	2.097	0.416-10.560
Age of father at birth (NS)						
34 and lower	139	89.7	16	10.3		
35 and higher	21	87.5	3	12.5	1.241	0.333-4.626
Consanguinity of parents (NS)						
No	84	89.4	10	10.6		
Yes	62	87.3	9	12.7	1.219	0.468-3.180
Hearing, visual impairment in parents (NS)						
No	164	88.6	21	11.4		
Yes	20	87.0	3	13.0	1.171	0.321-4.280
Hearing, visual impairment in siblings (NS)						
No	108	90.8	11	9.2		
Yes	70	86.4	11	13.6	1.543	0.635-3.750
Refractive error**						
No	140	96.6	5	3.4		
Hyperopes	19	86.4	3	13.6	4.421	0.977-20.001
Myopes	5	71.4	2	28.6	**11.200**	1.732-72.430
Astigmatism	19	57.6	14	42.4	**20.631**	6.678-63.735

NS: Not Significant,

* $p< 0.05$, ** $p< 0.0001$.

It is also found that, if a deaf child had a manifest astigmatism more than 1 D, the odds that he might have an important ophthalmoscopic pathology increases 20.631 times (6.678-63.735, 95% CI) (Table 2). The increase in associated ophthalmoscopic pathologies is 11.2 times (1.732-72.430, 95% CI) for myopia and 4.421 times (0.977-20.001, 95% CI) for hyperopia. It is hard to propose an explanation for this significant increase in fundus pathologies with our study design. The importance of this finding is that, astigmatism more than 1 D with associated myopia in a deaf child should alert the ophthalmologist about the existence of possible ophthalmologic pathologies.

The incidence of anisometropia was 5.3% in our study group. The reported rates 4.8% in Turkey and 7.3% in Leguire et al's study are similar to our findings.[1,2] Amblyopia frequency was found as 8.2% in our study. The 2.9% difference between the incidences of amblyopia (8.2%) and anisometropia (5.3%) in our study might arise from the presence of tropias less than 10 prism diopters, which may be the cause of amblyopia without anisometropia. The reported incidence of amblyopia in normal school children in Turkey is around 5%.[7,10] For hearing impaired and deaf population, amblyopia incidence was found as 15.3% in a study in Turkey, 4.4% in Leguire et al.'s and 3.6% in Guy et al.'s stu-

dies.[1-3] Former two studies used the same criteria for amblyopia; best corrected visual acuity of less than 20/30 in either eye. Although for frequencies reported amblyopia are not very consistent; the lowest frequency reported is approximately two times higher than the amblyopia frequency in normal hearing population.[3] This shows that ophthalmologists should also pay particular attention for higher incidence and the need for early detection of anisometropic amblyopia in the hearing impaired and deaf patients.

An important retinal abnormality (Table 1) was detected in 11.5% in our study group and retinal pigmentary changes in 12 (5.8%) children were found to be consistent with retinitis pigmentosa. Hanioğlu-Kargı et al.'s reported one case (0.9%) of retinitis pigmentosa and seven (6.8%) cases of retinal pigmentary changes.[2] The reported rate of retinitis pigmentosa in their study (0.9%) is very low compared to present study's findings (5.8%). However, the rate of all retinal pigmentary changes in study of Hanioğlu-Kargı et al. (7.7%) and (7.8%) the present study have been found to be similar.[2] Two children (1.0%) presented with macular pathologies, and four children with (1.9%) atypical pigmentary changes. Atypical pigmentary changes in these four patients may be related to congenital rubella syndrome, which is a cause of hearing impairment associated with retinal pigmentary changes.

In our opinion, the most important ophthalmologic aspect for the hearing impaired and deaf children is the possible existence of Usher syndrome.[11] This syndrome is an autosomal recessive disorder, characterized by congenital sensorineural deafness and progressive pigmentary retinopathy and is responsible for approximately half of the acquired deaf-blindness in the population.[5] The age of onset of the symptoms in this syndrome is variable and the exact prognosis is not known for every individual. The unfavorable prognosis of this clinical condition poses a necessity of a detailed fundus examination in the hearing impaired children for early educational and vocational modifications.

Vision and hearing are the two senses which enhance each others' efficiency. When deaf-blindness occurs, communication becomes extremely difficult without proper education. The most important aim of the rehabilitation of deaf-blindness is reducing the isolation of the patient. Cochlear implants are revolutionary and promising aids to help deaf blind people to hear and communicate. Identification of the mutations in genetic disorders may enlighten the path to inventions of newer medical and genetic treatments or alleviation of deaf-blindness. The aim of the psychological rehabilitation should at least be fostering realistic expectations and also aim to endure the patient and the family's hope of treatment in some near future.

The importance of this study should be assessed in two aspects for pediatricians, primary care physicians, special educators and Ear, Nose and Throat specialists and ophthalmologists. Due to an approximate chance of 50% in a hearing impaired child to present with an ocular abnormality (Table 3), it is essential to refer these children to an ophthalmologist as early as possible. The second important aspect of these findings is the need for a career planning and educational modification for children with co-morbidities, like Usher syndrome. Children who have a hearing abnormality as-

TABLE 3: The distribution of findings of hearing impaired and deaf children from different studies.

First Author of Study	Sample size	Population Characteristics of Study Group	Frequency of any Eye pathology	Frequency of Retinal Pathology
Hanioglu-Kargı[2]	104	7-20 y/o, deaf	42/104 **(40.4%)**	7/104 **(7%)**
Leguire[1]	357	6-22 y/o, deaf	225/357 **(63%)**	88/357 **(24.6%)**
Ceyhan	208	7-19 y/o, deaf	72/208 **(34.6%)**	20/208 **(9.6%)**
Guy[3]	110	5-16 y/o hearing impaired, deaf	48/110 **(43.6%)**	(16/110 **14.5%**)

sociated with retinal pigmentary changes have a higher chance of deteriorating into a deaf-blind status in early adulthood. The families and educators of these patients should be informed for a possible visual loss. A special educational scheme that provides earlier educational degrees for these children should also be considered by education policy-makers.

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