Ocular profile among hearing impaired children

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ABSTRACT

Objective: To report ocular abnormalities among identified hearing impaired children and their prevalence in relation to normal hearing children.

Methods: A sample of 302 identified children with bilateral sensorineural hearing loss at King Abdul-Aziz University Hospital and Rehabilitation Medical Center of the Ministry of Health, Riyadh, Kingdom of Saudi Arabia, after full audiological assessment, aged below 15 years, were screened ophthalmologically during the period from December 1997 to December 2002. Visual testing and cycloplegic refraction using 1% Cyclogyl was performed followed by retinal examination using direct and indirect ophthalmoscopy. Electroretinography was recorded for those with retinal pigmentary changes. One hundred control normal hearing children were examined, and chi square test applied.

Results: A total of 302 hearing impaired children were examined, 199 (66%) were boys and 103 (34%) were girls, age ranged between 2-15 years. Hearing loss was bilateral severe to profound. One hundred and eighty-four (61%) have one or more ocular abnormalities, while 147 (48.7%) had one or more significant error of refraction. In the control cases, 23% had error of refraction with a chi square test of 20.2 ($p<0.001$), which is significant.

Conclusions: Ocular anomalies are more commonly found among deaf children than those with normal hearing. Early ophthalmological assessment of hearing impairment is advisable to detect any ocular anomalies, followed by correction to help in the academic performance of these children.

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Hearing impaired children depend on other senses to acquire knowledge, and mainly the sensation of vision. They are more dependent on visual cues for communication. The degree which persons with hearing impairment need visual information when conversing is proportional to the amount of information that is lost due to hearing impairment.[1] The person with severe to profound hearing loss is likely to be more dependent on visual information to communicate than is an individual with a mild auditory impairment.[2] Visual information is transmitted by a manual or oral communication system. This complex form of communication allows for transfer of information via the visual channel when both the sender and the receiver are familiar with the same system of symbols.[3] Therefore, vision assessment in this population is required as the presence of visual impairment will add to the suffering of these hearing impaired children and to avoid the educational difficulties posed by hearing loss in the presence of unrecognized visual problems. This survey reports the ocular abnormalities among 302 deaf children in Riyadh and its rate in relation to normally hearing children, and to stress the importance of ophthalmological assessment of the hearing impaired.

Methods. This is a prospective study carried out during the period December 1997 to December

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2002 at King Abdul-Aziz University Hospital and the Rehabilitation Medical Center of the Ministry of Health, Riyadh, Kingdom of Saudi Arabia. A sample of 302 hearing impaired children were subjected to full ear, nose and throat (ENT), audiological and ophthalmological clinical examinations. History including parent relationship, pregnancy especially acquiring infection such as Rubella, and postnatal medical history was recorded. Full ocular examination by an ophthalmologist, and ENT by an otorhinolaryngologist was carried out. An orthoptist and an audiologist also participated in the assessment. Pure tone audiometry using Madson O.B. 8-22 and tympanometry using GSI-31 was carried out. Brain evoked response audiometry was performed for all children. Visual testing, modified according to age and cooperation of the children was carried out, uncorrected, best-corrected visual acuity, cycloplegic refraction using 1% Cyclogyl was performed followed by retinal examination using direct and indirect ophthalmoscopy. The anterior segments of each eye were assessed using biomicroscope and papillary reaction was noted. Children with pigmentary change with the congenital cataract children were sent for electroretinography (ERG). The ERG is the record of electrical changes of the retina that is caused by transient light stimulation. This was recorded using electrode with FZ reference. Flash visual evoked potential (FVEP) and pattern shift visual evoked potential (PSVEP) were also studied using OZ-FZ derivation. Flash stimuli were given from a distance of 30 cm from the eye for ERG and FVEP recording. The PSVEP were carried out using checkboard screen at a distance of 100 cm from the eye.[4]

Results. A total of 302 hearing impaired children, aged 2-15 years (mean age 8.7 years) were examined, 199 (66%) were boys and 103 (34%) were girls. Consanguinity was found among 27%. All the children had sensorineural hearing loss (SNHL) bilaterally, 127 (42%) with profound SNHL and 175 (58%) with moderate to severe SNHL, average 40-75 dB HL. Thirteen hearing impaired children were too young to be examined ophthalmologically. One hundred and eighty-four (61%) have one or more ocular abnormalities, 147 of which (48.7% of the total) had one or more significant errors of refraction. **Table 1** shows the ocular findings among the hearing impaired children. **Table 2** shows the comparison of ocular finding in the control normal hearing group and the hearing impaired group.

Discussion. Hearing impairment, especially severe to profound when bilateral, is a handicap that interferes with speech and language development especially when the hearing loss occurs during early childhood or is congenital. Children therefore use other senses to compensate for the hearing handicap. Vision is the most important in this regard. Visual skills or speech reading is a visual activity, and the acuity of vision is critical in the decoding process. Hardick et al[3] postulated that they could rank-order successful and unsuccessful speech readers on the basis of visual acuity. Furthermore, these authors observed a significant relationship between eye blink rate and speech reading ability, with poorer speech readers demonstrating higher eye blink rates. Recently Johnson and Snell,[5] showed that distance has a significant effect on visual acuity, children with visual acuity 20/80 or better should be able to speech read at 5 feet with an adequate degree of accuracy. When the speech reader is positioned 22 feet from the talker it is necessary that the speech reader has visual acuity no poorer than 20/30. If; however, the speech reader has one eye of 20/30 or better he should be able to speech read at a level comparable to individuals with normal binocular vision under similar viewing conditions. Gibson,[6] stated that our eyes receive

Table 1 - Ocular findings in 302 hearing impaired children.

Finding	N	(%)
Normal findings	118	(39)
Myopia	63	(21)
Hypermetropia	15	(4.9)
Astigmatism	36	(12)
Anisometropia	9	(3)
Manifest strabismus	11	(3.6)
Pigmentary changes	18	(5.9)
Congenital cataract	7	(2.3)
Pale fundi	6	(2)
Visual field defect	6	(2)
Not examined	13	(4.3)
Total	**302**	**(100)**

Table 2 - The overall prevalence of refractive errors among the hearing impaired and normal children.

Finding	Hearing children %	Hearing impaired children N	(%)	Difference %
Normal findings	77	118	(39)	38
Myopia	12	63	(20.9)	8.9
Hypermetropia	2	15	(4.9)	2.9
Astigmatism	6	36	(12)	10.9
Amblyopia	2	15	(4.9)	2.9
Manifest strabismus	1	11	(3.6)	2.6
Anisometropia	-	7	(2.3)	-
Total	**100**	**265**	-	-
Overall prevalence	**23**		**(48.7)**	**28.3**

visual stimuli that are interpreted at a cortical level and provide us with visual information. This information, in turn enables us to make a selective response to the original stimuli. Thus, when interpreting speech visually, the speech reader first sees the movement of the lips, which the cortex classifies as speech. The accuracy of the speech readers response to these stimuli is partially a function of how well the peripheral to central visual process enables him to discriminate among the speaker's articulatory movements.

Congenital deafness unfortunately may be associated with visual abnormalities, since these children have no hearing experiences from early life, therefore, visual impairment further reduces their ability to learn and to adjust themselves to their environment, thus increasing the suffering of already handicapped children. In this survey, ocular abnormalities were found to occur in the hearing impaired children examined in comparison to normal hearing children. In this study 5 children were found to have retinal pigment changes, 4 with profound sensorineural hearing loss and reduced vestibular response and one with moderately severe hearing loss and normal vestibular response. These cases were considered as Usher syndrome. The association between deafness and pigmentary retinopathy is well known, and several syndromes have been described, Usher syndrome is the best known. Retinitis pigmentosa (RP) is a heredoretinal degenerative disease. In Usher syndrome it is of an autosomal recessive inheritance, characterized by sensorineural hearing loss and RP. It is heterogenous and occurs in 2 forms: type I with profound nonprogressive congenital hearing loss associated with absent vestibular response to caloric stimulation, and type II with nonprogressive moderate to moderately severe congenital hearing loss.[7] According to the study of Schukrecht,[8] the deafness in Usher syndrome is due to atrophy of the cochlea and loss of the non motile ciliated cells. The common neuroectodermal embryological origin of the retinal pigment epithelium, and the organ of corti explains the association between deafness and retinal pigmentary changes. The deafness is believed to be present at birth, while the initial symptoms of RP do not usually occur until the second or third decade of life. Physiological changes in the retina, as measured by ERG occurs early in the disease, before anatomical changes and before the patient becomes aware of symptoms such as night blindness and tunnel vision.

Those children with congenital cataracts underwent cataract surgery. The low incidence of rubella in this country may be attributed to the compulsory vaccination of all girls from the age of 10 while at school, and may be due to the proper antenatal care. The 6 children with pale fundi have no signs of albinism, but their parents were cousins and with positive family history of deafness. This hypopigmentation of the optic fundi is a phenotype expression of underlying and, as yet poorly understood, genetic defects affecting hearing.

In our study, 38% of the hearing impaired children were found to have ocular abnormalities. Nicoll and House,[9] in a study of 78 hearing impaired children found 33% with ocular abnormalities. Woodruff,[10] in his study of vision anomalies in deaf children found a high rate of 55% with ocular problems, compared to 12-32% found in sample of children in elementary school.

In conclusion, ocular anomalies are more commonly found among congenitally deaf children, than those with normal hearing. Early ophthalmological assessment in these children is strongly advisable to detect any ocular anomalies and correct these as early as possible to minimize the effect on the performance of these already handicapped children. Otolaryngologists, pediatricians and general physicians should be aware of the site problems associated with deafness and to refer affected children for ophthalmological assessment before supplying hearing aids, and institute speech and hearing training.

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