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Original Article

Ophthalmic Abnormalities among Children Treated with Cochlear Implants

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OBJECTIVES: To review the ocular abnormalities in children treated with cochlear implant.

MATERIALS and METHODS: A total of 51 children (29 boys, 22 girls) who were under 18 years old, presented previously with severe to profound hearing loss, and underwent cochlear implantation surgery were included in this study prospectively. A detailed ophthalmic examination, including refraction, best corrected visual acuity, ocular motility, slit-lamp biomicroscopy, and dilated fundus examination, was performed for each patient.

RESULTS: Mean age of the patients was 80.10±38.64 (range, 18-168) months. A total of 13 (25.4%) children had at least 1 ophthalmic abnormality. The majority of the detected ophthalmic abnormalities were hyperopia and astigmatism (6 patients had hyperopia, 5 had astigmatism, and 2 had hyperopia plus astigmatism). Strabismus (esotropia) was found in 2 patients, 2 patients had refractive amblyopia, and 2 patients had nystagmus. Moreover, 3 patients had microcornea, 2 patients had cataract, and 1 patient had epiblepharon. Optic disc coloboma (3 patients), choroidal coloboma (1 patient), and pigmentary abnormality (1 patient) were noticed on fundus examination. Congenital rubella syndrome (2 patients), Waardenburg's syndrome (1 patient), and CHARGE syndrome (coloboma, heart defects, choanal atresia, growth retardation, genital abnormalities, ear abnormalities) (1 patient) were also present.

CONCLUSION: Children treated with cochlear implant should be consulted with an ophthalmologist to identify any treatable ocular abnormality.

KEYWORDS: Cochlear implants, hearing loss, eye diseases

INTRODUCTION

Cochlear implants are the electronic devices that were developed to restore hearing loss by directly stimulating the auditory nerve. Cochlear implantation was approved for the management of children and adults with severe to profound sensorineural hearing loss that showed no or limited benefit from conventional hearing aids [1-3].

Visual and auditory systems are responsible for more than 95% of information acquisition [3, 4]. Hearing impairment during infancy and early childhood severely retards the development of speech and language skills of the children. In these children, the visual system is the major source for gaining information from the outside. Thus, additional visual problems in a deaf child may profoundly impair the cognitive functions and the educational and social development of the child and highlights emerge the importance of ocular assessment in deaf children. In previous studies, the prevalence of ophthalmic abnormalities was found to be higher in children with hearing disability, ranging from 32% to 60% [4-9]. A wide variety of conditions can be the etiologic factor for both hearing and visual impairment in a newborn or during the early childhood period [3-15].

In this study, we aimed at evaluating the rate and nature of the ophthalmic abnormalities in a cohort of pediatric population with cochlear implant.

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MATERIALS AND METHODS

Children (age<18 years) who previously presented with severe to profound hearing loss and underwent cochlear implantation surgery at the Otorhinolaryngology Department of Dokuz Eylul University School of Medicine were included in this study prospectively. A comprehensive eye exam was performed by one of the 2 authors (Z.A. and G.A.) at the Ophthalmology Department of Dokuz Eylul University School of Medicine between October 2016 and December 2017.

The study was approved by the ethics committee of our faculty and was performed in adherence to the guidelines of the Declaration of Helsinki (2016/16-16). Informed consent form was taken from the patients' parents.

The children were evaluated with a detailed ophthalmic examination, including refraction, best corrected visual acuity (BCVA), ocular motility, slit-lamp biomicroscopy, and dilated fundus examination.

The Allen figures or Snellen visual acuity chart was used for evaluating BCVA according to the adaptation of the child. Amblyopia was defined as an interocular difference of 2 or more lines in BCVA resulting from refractive, strabismic, or deprivational sources in the absence of any organic pathology. Ocular motility was assessed in 9 directions of gaze, evaluating both ductions and versions. Strabismus was evaluated with Hirschberg and cover-uncover tests. Cycloplegic refraction was evaluated with retinoscopy. Significant cycloplegic refractive error was defined as hyperopia if there was a refractive error of 1.5 D or more with manifest esotropia and if there was a refractive error of more than 3.0 D in the absence of strabismus; it was defined as myopia if there was a myopic refractive error of more than 1.0 D and astigmatism and if there was at least 1.5 D difference in refractive error between the 2 principal meridians. Anisometropia was defined as a difference of 1.0 D between the 2 eyes.

Statistical Analysis

Statistical analysis was performed using the Statistical Packages for Social Sciences (SPSS) version 15 (SPSS Inc., Chicago, IL, USA).

RESULTS

A total of 51 children (29 boys, 22 girls) were included in this study. Mean age of the patients was 80.10±38.64 (range, 18–168) months. The etiology of deafness and ophthalmic findings of the patients are summarized in Table 1 and Table 2, respectively. A total of 13 (25.4%) children had at least 1 ophthalmic abnormality. The majority of the detected ophthalmic abnormalities were hyperopia and astigmatism (6 patients had hyperopia, 5 had astigmatism, and 2 had hyperopia plus astigmatism). Strabismus was found in 2 patients, and both of them had esotropia; 2 patients had refractive amblyopia; and 2 patients had nystagmus. Moreover, 3 patients had microcornea, 2 patients had cataract, and 1 patient had epiblepharon. Optic disc coloboma (3 patients), choroidal coloboma (1 patient), and pigmentary abnormality (1 patient) were noticed on fundus examination. Congenital rubella syndrome (2 patients), Waardenburg's syndrome (1 patient), and CHARGE syndrome (coloboma, heart defects, choanal atresia, growth retardation, genital abnormalities, ear abnormalities) (1 patient) were also present.

Table 1. Etiology of deafness in all patients

Etiology	Number of Patients
Idiopathic	33 (64.7%)
Familial	8 (15.6%)
Enlarged vestibular aqueduct	4 (7.8%)
Congenital rubella syndrome	2 (4%)
Auditory neuropathy	2 (4%)
CHARGE syndrome	1 (2%)
Waardenburg's syndrome	1 (2%)

CHARGE: coloboma, heart defects, choanal atresia, growth retardation, genital abnormalities, ear abnormalities.

DISCUSSION

Ophthalmic pathologies in the patients treated with cochlear implants are variable, and anterior and posterior parts of the eye could be affected by abnormal development of the cornea, lens, optic nerve, choroid, and retina. The retina, the retinal pigment epithelium, and the optic nerve develop from the neural ectoderm. This development starts during the fourth embryonic week. Moreover, the inner ear develops from the surface ectoderm during the same embryonic period. Thus, environmental factors may cause developmental abnormalities both in the eye and the ear. In the literature, many studies state that there is a very high prevalence of ocular abnormalities in deaf children because of this close link of embryonic development [4-15]. In contrast, chromosomal abnormalities and gene mutations may also cause the unusual development of both the eye and the ear [15-19].

In the literature, the prevalence of ophthalmic abnormalities was found to be higher in children with hearing disability, ranging from

Table 2. Ophthalmic abnormalities of the patients

Ophthalmic Abnormality	Etiology of Deafness	Number of Patients
Pure astigmatism	Idiopathic	2
Esotropia+hyperopia	Familial	2
Hyperopia+astigmatism+ refractive amblyopia	Idiopathic	2
Nystagmus+hyperopia	Auditory neuropathy	1
Microcornea+cataract+ optic disc coloboma+ hyperopia+nystagmus	Congenital rubella syndrome	1
Microcornea+cataract+ optic disc coloboma+ hyperopia	Congenital rubella syndrome	1
Microcornea+choroidal coloboma+hyperopia	CHARGE syndrome	1
Epiblepharon+astigmatism	Idiopathic	1
Optic disc coloboma+ astigmatism	Idiopathic	1
Retinal pigmentary abnormality+astigmatism	Waardenburg's syndrome	1

CHARGE: coloboma, heart defects, choanal atresia, growth retardation, genital abnormalities, ear abnormalities.

32% to 60% [4-9]. However, in normal-hearing children, the prevalence of ophthalmic problems was found to be between 2.4% and 30% and prone to increase with age [15, 20-30]. Correlation between the cause of deafness and the prevalence of ophthalmic problems in deaf children is not clearly known. Armitage et al. [31] did not find any significant difference in visual impairment between children with congenital and acquired deafness. Woodruff [32] studied the prevalence of ophthalmic problems in 460 deaf children according to the cause of deafness. It was shown that inherited deafness appears to be associated with the fewest visual abnormalities. Congenital rubella had the highest prevalence and was associated with the broadest spectrum of ocular and visual problems. They found that children who had congenital rubella, neonatal sepsis, and Rh incompatibility showed higher rates of strabismus and amblyopia. In our study, patients with idiopathic and familial deafness were associated with less severe ocular abnormalities. Furthermore, 13 of the 51 children (25.4%) had at least 1 ophthalmic abnormality, the majority of which were refractive errors. Leguire et al. [33] found that children and young adults (age range, 6-22 years) with deafness greater than 80 dB had a higher prevalence of refractive errors than children with deafness less than 80 dB. Besides, there was a statistically significant increase in the prevalence of retinal abnormality in children with deafness greater than 80 dB compared to those with deafness less than 80 dB. They found that rubella retinopathy was 13.4% in children with deafness greater than 80 dB, whereas it was 6.1% in children with deafness less than 80 dB. Congenital rubella, syphilis, cytomegalovirus, and toxoplasmosis are well-known clinical conditions associated with both deafness and ophthalmic disorders [32,34]. In our study, only 2 patients were diagnosed with congenital rubella syndrome and had both anterior and posterior ocular abnormalities, such as microcornea, cataract, optic disc coloboma, and nystagmus. In addition to congenital infections, numerous syndromic, genetic, and other conditions (i.e., cerebral palsy, CHARGE syndrome, Cogan's disease, Cornelia de Lange syndrome, Aicardi syndrome, albinism, Alport's syndrome, Alström syndrome, Batten disease, meningitis, Stickler's syndrome, Usher syndrome, trisomy 21 and trisomy 18, Waardenburg's syndrome, metachromatic leukodystrophy, mitochondrial cytopathy, Moebius syndrome, mucopolysaccharidosis, prematurity, and spinocerebellar degeneration) can affect both the eyes and the ears. [15]. There were no patients with postnatal infectious conditions causing hearing loss in our study, but there were 2 other patients with syndromic conditions (1 patient had Waardenburg's syndrome, and other patient had CHARGE syndrome).

In children with severe to profound deafness, cochlear implantation provides development of spoken language skills, but most of these children continue to be at risk for significant difficulties in reading and writing skills and executive functioning [35]. Additional visual impairments may further deteriorate these functions [36]. Thus, early detection and treatment of visual problems of children with cochlear implant is critical for their neurocognitive development. Ophthalmic pathology in a patient with cochlear implant may be correctable (i.e., refractive errors) or treatable (i.e., amblyopia, cataract, strabismus). In some situations, ophthalmic pathology may be untreatable (i.e., optic atrophy or pigmentary retinopathy) but helps the physician to identify the cause of deafness.

CONCLUSION

Children treated with cochlear implant should be screened for ophthalmic disorders to identify any treatable ocular abnormality limiting the visual acuity, such as refractive errors, strabismus, amblyopia, and cataract. Furthermore, patients diagnosed with untreatable ocular pathologies such as pigmentary retinopathy, optic nerve abnormality, or cortical dysfunction should be referred for the appropriate educational, psychological, and genetic counseling.

Ethics Committee Approval: Ethics committee approval was received for this study from the Ethics Committee of Dokuz Eylul University School of Medicine (2016/16 -16).

Informed Consent: Written informed consent was received from all the patients' parents who participated in this study.

Peer-review: Externally peer-reviewed.

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