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CLINICAL STUDY

Ophthalmological screening of a paediatric cochlear implant population: a retrospective analysis and 12-year follow-up

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Abstract

Aims **To determine the nature and prevalence of ophthalmological findings for a cohort of children in a paediatric cochlear implant program and to assist the clinician in devising an investigative plan for this population.**

Methods **Retrospective medical record review of children who underwent multichannel cochlear implantation at a tertiary care hospital between February 1996 and July 2008.**

Results **In all, 141 children (mean age 28 months, range 16 months to 9 years) had complete medical record documentation consisting of orthoptic and opthalmological examination, including cycloplegic refraction. A total of 59 children (41.8%) had ocular abnormalities with refractive errors being the most common abnormality. Hypermetropia was the most common refractive error and was found in 21 children (14.8%). Strabismus was found in six patients, with constant esotropia being the most common. Ocular pathology (excluding refractive or muscle abnormalities) were found in nine patients (6.3%). Three patients had syndromes associated with ocular findings including Waardenburg and Usher syndrome. During the follow-up period, 14 children were fitted with prescription lenses, 3 had strabismus surgery, and 2 underwent ptosis.**

Conclusions **Routine orthoptic and ophthalmologic examination can be beneficial in the initial evaluation of children assessed for cochlear implants. Electroretinography is useful in evaluating children with unexplained congenital sensorineural hearing loss, suggestive symptoms including night blindness, unexplained reduction in visual acuity, or delayed motor milestones. Routine yearly follow-up may aid in the detection of changing refractive errors and the possibility of later-onset retinal degeneration.**

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Introduction

Cochlear implants have steadily improved over the last decades, dramatically changing the prospects for profoundly deaf children.[1] A majority of cases of congenital and infantile hearing loss are sensorineual in origin. These devices are thus successful in restoring some hearing by electrical stimulation of proximal neural elements. It is now accepted that early cochlear implantation may facilitate rapid transformation of a deaf child to a fully functioning member of the community.[2]

Ophthalmic problems in deaf children have a serious effect on development of their communication skills. As visual and auditory channels are responsible for more than 95% of information acquisition,[3] any ophthalmic disorder may thus negatively affect on this process. It is well documented that the incidence of visual problems is higher among the deaf than a corresponding population of hearing individuals.[4–6]

At referral for cochlear implantation, the specialist service should arrange for the child to

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be seen by an orthoptist and an ophthalmologist as soon as possible. This assessment may need to be repeated at intervals after the diagnosis.[7] The aims of screening for ophthalmic disorders are to identify any ocular abnormality limiting visual acuity, any pigmentary retinopathy, or optic nerve abnormality. Some disorders may be correctable (eg, refractive) or treatable (eg, cataracts).[8] It is crucial to establish normal visual acuity, as hearing-impaired children are extremely dependent on vision to compensate for the loss of auditory input.[4] Those children with non-correctable and non-treatable visual disorders, such as retinitis pigmentosa in Usher's syndrome, are especially good candidates for early cochlear implants, which will improve listening and spoken language skills.[9]

Deaf blindness is more than the sum of deafness and blindness on their own. It is a unique and entirely different condition requiring a multi-disciplinary approach.[10] Deaf children are heavily dependent on their sense of vision to develop efficient communication skills. Hearing and visually impaired children are significantly affected, being less able to lip read, less cooperative, and less capable of manual tasks then hearing-impaired children with normal vision.[11] If unrecognized, especially in the early years of life, children may fail to develop the ability to develop linguistic and communicative skills, with a significant effect on education, socioemotional development, and future professional prospects.

The objective of this study was to determine the nature and prevalence of ophthalmological findings for a cohort of children in a paediatric cochlear implant program and to assist the clinician in devising an investigative plan for this population.

Materials and methods

Study population

The study was a retrospective review of the medical records of 196 children who underwent multichannel cochlear implantation at Beaumont University Hospital, Dublin, Ireland between February 1996 and July 2008. Beaumont University Hospital is the National Irish Referral Centre for Cochlear Implants and is a member of the British Cochlear Implant Group representing all the Cochlear Implant Centres throughout the United Kingdom and Ireland. Cases were identified through the electronic database of paediatric patients who received a cochlear implant.

Data collection

Data collected from the medical records included patient gender, age, cause of hearing loss (if known), relevant prenatal, perinatal and postnatal factors, family history of hearing loss, genetic testing results, and orthoptic and ophthalmological assessment.

Visual assessment included visual acuity (Snellen letters, Sheridan Gardner singles, Kay pictures, Cardiff Acuity Cards, or fixation/following evaluation), cover test, complete motility examination, and binocular function evaluation (Frisby stereoacuity) by an orthoptist. Convergence and optokinetic nystagmus were recorded. All children underwent slitlamp examination, direct and indirect ophthalmoscopy, and cycloplegic retinoscopy by an ophthalmologist. Electrodiagnostic tests were not performed routinely. Patients underwent yearly control evaluations after the initial diagnosis unless otherwise indicated.

Results

Records of 141 ophthalmological examinations carried out in our department during the study period were available for review. All 141 children (mean age 28 months, range 16 months to 9 years) had complete medical record documentation consisting of orthoptic and opthalmological examination, including cycloplegic refraction. The ophthalmological findings and aetiology of deafness are summarized in Tables 1 and 2, respectively. The criteria for defining a vision disorder as significant were obtained from the study on children with severe and profound sensorineural deafness by Armitage *et al*.[5]

Visual function was remarkably good in our patients with only 34 children (23.6%) having visual acuity worse than six out of nine patients. A total of 59 children (41.8%) had ocular abnormalities with refractive errors being the most common abnormality. Hypermetropia was the most common refractive error and was found in 21 children (14.8%). Strabismus was found in six patients, with constant esotropia being the most common. Ocular pathology (excluding refractive or muscle abnormalities) were found in nine patients (6.3%). In one patient, visual loss occurred in early childhood because of congenital horizontal pendular nystagmus.

One child was diagnosed with Waardenburg syndrome after the finding of iris heterochromia and telecanthus. Two patients with an unidentified cause of deafness and positive history for late walking (mean age 25 months), were diagnosed with Usher syndrome after abnormal electroretinography. During follow-up, reduced visual acuity, optic disc pallor, and attenuated retinal vessels occurred at the age of 5 years; however, retinitis pigmentosa occurred only later than the age of 8 years, comparable to that in children described earlier. During the follow-up period, 14 children were fitted with prescription lenses, two underwent ptosis surgery and three children had strabismus surgery.

Table 1 Ophthalmological findings in the paediatric cochlear implant population

	Number	*Percentage of total (%)*
Visual acuity <6/9	34	23.6
Refractive error	37	26.4
Hypermetropia >3.0 D	21	16.8
Myopia >1.0 D	5	4.2
Astigmatism ⩾1.5 D	4	2.8
Anisometropia ⩾1.0 D	3	2.8
Strabismus	11	8.4
Constant esotropia	5	4.2
Constant exotropia	3	2.8
Intermittent esotropia	2	1.4
Stereopsis		
Normal	95	67.3
Reduced	14	9.9
Unidentified	32	22.6
Ocular pathology	9	6.3
Anterior segment		
Iris heterochromia	1	0.7
Anterior polar cataract	2	1.4
Retinal anomalies		
Pigmentary retinopathy	2	1.4
Optic nerve anomalies		
Optic disc pallor	2	1.4
External eyelid anomalies		
Epicanthal folds	2	1.4
Telecanthus	1	0.7
Ptosis	2	1.4
Nystagmus	1	0.7

Table 2 Aetiology of deafness in this study

Aetiology	N
Unknown	75
Congenital	68
Familial	19
Connexin 26 anomaly	31
Jervell and Lange-Nielsen syndrome	3
Pendred syndrome	4
Waardenburg syndrome	1
Usher's syndrome	2
Maternal diabetes	3
CMV	2
Rh disease	3
Acquired	17
Prematurity	5
Meningitis	9
Gentamicin ototoxicity	3

Discussion

Earlier studies have reported a high incidence of visual impairment among the hearing-impaired population. (Table 3) Most of them assessed deaf children in residential or day schools and included children of all ages and causes. Only one other study assessed children who underwent cochlear implantation.[25] This retrospective study with a 12-year follow-up, provides data on the prevalence and characteristics of ophthalmological findings after screening of a paediatric cochlear implant population. Follow-up for many of our patients was long enough to show objective benefit to our patients.

Consistent findings show that 12–60% of deaf children have significant vision impairment (Table 3). These studies vary in patient population, the types of visual problems they have assessed, the tests used, and the criteria to define and classify visual impairment. Thus, the incidence rates for specific visual problems are often not directly comparable across studies. In a comparative analysis,[4] the incidence of visual impairment in the hearing-impaired group is more than two and a half times higher than that found in a normal school population.[6]

The role of screening is to determine visual acuity and identify abnormalities amenable to specific treatment and better overall management. Second, this on-going commitment aids in identification of hereditary hearing loss syndromes that are associated with ocular findings. Several authors concluded that deaf students had a high frequency of eye disease,[7,22,27] and that early diagnosis and treatment could improve their quality of life and that the potential benefits of ophthalmologic screening in deaf children is enormous.[22] Armitage *et al*[5] found that in 48% of deaf children, visual impairment had remained undetected before screening. The British National Screening committee recommended that children with deafness should have routine expert eye examination performed by an orthoptist and ophthalmologist.[7] Our findings suggest that all children assessed for cochlear implants would benefit from routine ophthalmic assessment.

Ocular findings are quite variable and could affect the anterior and posterior segment of the eye with abnormal development of the lens, optic nerve, and retina. These higher rates of ocular abnormality have been attributed to the retina and cochlea maturing during the same embryological period (fourth embryonic week) from the same embryonic layer (ectoderm), which may be susceptible to genetic and/or environmental factors.[8] The most frequent ophthalmic finding is refractive errors (26.4%), similar to the earlier studies. Fourteen of our patients with refractive errors were fitted with corrective spectacles. Correction of refractive errors has been found to bring significant improvement even in motor behaviours.[32]

Table 3 Studies of ophthalmological findings in patients with hearing impairment

Year	*Author*	N	*Ocular abnormality (%)*	*Refractive error (%)*	*Strabismus (%)*	*Retinopathy (%)*
1967	Suchman[11]	104	58	25	—	—
1970	Lawson[12]	80	54	47	3	9
1971	Dayton[13]	237	60	58	2	25
1973	Alexander[14]	572	50	35	12	22
1974	Pollard[15]	511	33	25	9	7
1976	Mohindra[16]	77	60	51	19	10
1978	English[17]	698	—	—	—	12
1978	Quinsland[18]	186	—	54	—	10
1982	Johnson[19]	620	—	49	9	17
1982	Walters[20]	1951	—	17	7	11
1984	Regenbogen[4]	150	45	27	11	5
1985	Woodruff[21]	460	55	30	9	9
1987	Fillman[3]	210	—	48	8	3
1988	Rogers[22]	360	43	25	—	—
1989	Ma QY[23]	279	32	18	7	29
1992	Leguire[24]	505	49	23	8	22
1994	Siatowski[25]	54	61	44	4	4
1994	Elango[26]	165	58	14	11	35
1995	Armitage[5]	83	46	29	20	10
2001	Brinks[27]	217	48	16	5	26
2002	Mafong[28]	49	31	66	7	7
2003	Guy[29]	110	44	39	6	11
2003	Hanioğlu[30]	104	40	30	18	9
2008	Bakshaee[31]	50	32	28	6	24

In a prospective study of the prevalence of vision disorders, other than refractive anomalies, the most prevalent conditions in the clinical paediatric population are binocular and accommodative disorders.[33] At initial assessment, binocular vision was reduced and absent in 9.7 and 4.2%, respectively, similar to previously reported.[5] The incidence of manifest strabismus in our study (8.4%) was significantly higher than that of 4.6% found in a normal population.[6] During the follow-up period, 11 children (7.8%) underwent conservative or surgical correction.

The likelihood of visual impairment is independent of the aetiology of the hearing loss. Armitage *et al*[5] did not find any statistically significant difference in the incidence of visual impairment among the congenital and acquired hearing-impaired groups. However, children with deafness >80 dB had a statistically significant increase in the prevalence of retinal abnormality compared with children with deafness <80 dB.[24]

Studies of hearing-impaired populations indicate a genetic basis in approximately 50% of all hearing loss.[34] Cochlear implant assessment includes imaging studies and genetic testing in addition to ophthalmologic referral. Despite the enormous heterogeneity of genetic hearing loss, variants in one locus, Gap Junction Beta 2 (GJB2) or connexin 26 (CX26), account for up to 50% of cases of nonsyndromic sensorineural hearing loss.[35] We did not find any ophthalmological abnormalities in the subgroup of 15 patients with CX26 mutations. Rarely, autosomal dominant CX26 mutations may result in Keratitis-Ichthyosis-Deafness (KID) syndrome characterized by congenital hearing loss associated with skin lesions and vascularizing keratitis.[36]

Approximately 15–30% of hereditary hearing impairment involves other organ systems and occurs as a syndrome. In addition, abnormal ocular findings are associated with certain common deafness syndromes. In 8% of congenitally deaf children examined, eye examination confirmed a previously suspected cause of deafness, and in another 8%, a diagnosis that had not been suspected was made.[37] Thus, ophthalmic examination may prove invaluable to identify or clarify the cause of deafness such as CHARGE association, Usher syndrome or congenital CMV or Rubella.[7]

Some studies recommend electrodiagnostic testing as part of routine ophthalmologic evaluation of children with sensorineural hearing loss to identify Usher syndrome.[9,25] Electroretinography is a noninvasive test that identifies retinitis pigmentosa before the onset of funduscopic and visual abnormalities. Mets *et al*[9] found a 10% prevalence of Usher's syndrome in children with severe to profound sensorineural hearing loss. Two patients in our group, with an unidentified cause of deafness and positive history for late walking, were diagnosed with Usher syndrome after abnormal

electroretinography. Retinitis pigmentosa occurred only later with long-term follow-up after the age of 8 years. As early retinal changes are uncommon, electroretinography should be considered in children with unexplained profound congenital sensorineural hearing loss and delayed motor milestones.[38]

The limitations of this study should be noted. First, a major limitation of our study was its retrospective nature. Second, the results of ophthalmologic examinations were unavailable for many of our patients, introducing the possibility of selection bias. Many of our patients were referred to their local ophthalmologists for assessment before the introduction of mandatory ophthalmology screening, the results of which were unavailable. Despite these limitations, we believe our data support routine ophthalmological screening of children in the cochlear implant population.

The high prevalence of visual abnormalities in the paediatric cochlear implant population emphasizes the importance of thorough ophthalmological screening. The first aim is to determine visual acuity. Some of the symptoms can be successfully corrected surgically, ensuring better visual development and improving the quality of life after cochlear implantation. Second, uncommon ophthalmological manifestations can be used in identification of hereditary hearing loss syndromes that are associated with ocular findings.

It remains to be agreed which tests should be performed. Routine orthoptic and ophthalmologic examination can be beneficial in the initial evaluation of children assessed for cochlear implants. Electroretinography is useful in evaluating children with unexplained congenital sensorineural hearing loss, suggestive symptoms including night blindness, unexplained reduction in visual acuity, or delayed motor milestones. Routine yearly follow-up may aid in the detection of changing refractive errors and the possibility of later-onset retinal degeneration.

Conflict of interest

The authors declare no conflict of interest.

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