Coll. Antropol. **32** (2008) 4: 1111–1113
Original scientific paper

Etiology of Deafness in Children Cochlear Implant Candidates in Croatia

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A B S T R A C T

The aim of this study was to provide more information on the causes of sensorineural hearing loss (SNHL) in children cochlear implant candidates in Croatia. The retrospective study included 270 children candidates for cochlear implantation between January 1997 and January 2005 at our institution. The medical assessment of the candidates included the history, physical examination, radiologic evaluation of the temporal bone and audiologic assessment. A family history of SNHL had 82 (30.4 %) candidates. The prematurity and/or complicated perinatal course was found in 35 (12.9%) of candidates. Computerized tomography (CT) scan analysis identified 44 (16.3%) candidates presenting with an inner ear malformation. Overall, a definite or probable cause of SNHL was identified in 58.9% of candidates and 41.1% had no obvious cause. The results of the study might give us better insight into the potential causes of SNHL and allow more timely intervention, allowing children with SNHL to reach their potential.

Key words: *sensorineural hearing loss, children, cochlear implantation, CT, temporal bone, Croatia*

Introduction

The incidence of severe to profound sensorineural hearing loss (SNHL) in children is approximately 1:2000 at birth and 6:1000 by 18 years of age[1]. The severe to profound unilateral losses are often not recognized until kindergarten. The causes of SNHL are many, ranging from genetic to infective, both congenitally and postnatally acquired, to pharmacologic. The relative importance of these factors has not been fully assessed, and in about one-third of the cases no cause is found[2]. It is estimated that 50% of cases of childhood hearing impairment are due to genetic factors, about 20% to 25% of cases are due to identifiable environmental causes, occurring prenatally or perinatally, during the neonatal period, or later in life, and 25% to 30% are sporadic cases of unknown etiology. Most authors attribute 75% to 80% of genetic deafness to autosomal recessive genes and 18% to 20% to autosomal dominant genes; the rest are classified as X-linked or chromosomal disorders[3]. Genetic forms of hearing loss may be congenital or of delayed onset, progressive or nonprogressive, unilateral or bilateral, and they may be syndromic or nonsyndromic. In the literature, 20% of children with sensorineural hearing loss have had associated radiological anomalies of the temporal bone. The use of high-resolution imaging techniques resulted in the detection of a large percentage of children with radiographically detectable anomalous cochleovestibular anatomy. Cochleovestibular anomalies including those of the internal auditory canal can be identified with current high-resolution scans. Jacker et al. classified four types of osseous and membranous dysplasia: *complete aplasia* (Michel's aplasia), common cochleovestibular chamber *(CC) deformity*, hypoplastic cochlea *(HC)* and incomplete partition *(IP)*. IP includes the classic Mondini malformation in which the basal turn of the cochlea is normal, hearing may be present, and no anatomical basis exists on which to expect a perilymph fistula[4–8]. Most countries do not have mandatory hearing screening at birth for all children, regardless of the risk factors, so many children are missed. 50% of the losses occur after the newborn period, so only ongoing surveillance will identify losses in these children. In an effort to provide more information on the causes of SNHL we performed this retrospective study. The study was carried out at tertiary institution, Department of Otolaryngology – Head and Neck Surgery, University Hospital Sestre Milosrdnice, Zagreb, one of two cochlear implant centers in Croatia. 65% of all cochlear implantations in Croatia were performed at our department.

Received for publication February 13, 2007

Patients and Methods

The study was carried out to review the etiology of deafness in the group of congenital and prelingually deaf children, candidates for cochlear implantation (CI), less than 18 years of age at the time of clinical evaluation for the cochlear implantation, between January 1997 and January 2005. All children had severe to profound sensorineural hearing loss and were considered audiological candidates based on the documented hearing loss. The medical assessment of cochlear implant candidates included the otologic history, physical examination, radiologic evaluation of the temporal bone and audiologic assessment. The auricle was inspected for signs of congenital deformity of the external ear and a detailed head and neck examination was performed to rule out congenital deformities. All candidates underwent a standardized protocol of temporal bone imaging consisting of preoperative high-resolution thin-section CT scanning to rule out a congenital deformity of the inner ear and following changes were detected: malformations of the inner ear, the inner auditory canal, meningititis-induced cochlear obliteration, post-traumatic temporal bone changes, local auditory ossicle malformation, inflammatory middle ear changes, the high position of the jugular vein bulb. The CT images were assessed by neuroradiologists. A retrospective study was made of the CI candidates history and the preoperative CT findings. The medical charts of 270 children were reviewed in our study.

Results

The retrospective study included 270 children candidates for cochlear implantation at our institution. The distribution of male and female patients was approximately equal. The age of candidates at time of clinical preoperative evaluation for cochlear implantion ranged 5 months to 14 years (mean age, 3.9 years). All patients undergoing evaluation had bilateral severe to profound SNHL. The hearing loss was identified before 1 year of age in 161 (59.6%) of these patients. A family history of SNHL had 82 (30.4 %) candidates. The meningitis and/or prenatal maternal factors were present in 18 (6.7%) candidates. The prematurity and/or complicated perinatal course was found in 35 (12.9%) of candidates (Table 1). The radiologic evaluation of the temporal bone (Table 2) showed normal cochleovestibular anatomy in 226 (83.7%) candidates. CT-scan analysis identified 44 (16.3%) patients presenting with an inner ear malformation in the study. The inner ear malformations included 18 (6.7%) patients with dilated vestibular aqueducts, 12 (4.4%) with vestibulocochlear dysplasia, 10 (3.7%) with Mondini malformation, and 4 (1.5%) with ossified cochlea. The inflammatory middle ear changes were present in 15 (5.6%) and the high position of the jugular vein bulb in 32 (11.8%) candidates, which are important because it cause higher risk of intraoperative or postoperative complications. Overall, a definite or probable cause of SNHL was identified in 58.9% of patients and 41.1% had no obvious cause of SNHL.

TABLE 1
EPIDEMIOLOGIC FEATURES OF CHILDREN COCHLEAR IMPLANT CANDIDATES IN CROATIA

Etiology	No. of patients
family history of SNHL	82 (30.4%)
meningitis and/or prenatal maternal factors	18 (6.7%)
prematurity and/or complicated perinatal course	35 (12.9%)

TABLE 2
CT TEMPORAL BONE EVALUATION OF CHILDREN COCHLEAR IMPLANT CANDIDATES IN CROATIA

CT findings	No. of patients
normal cochleovestibular anatomy	226 (83.7%)
inner ear malformation	44 (16.3%)
dilated vestibular aqueduct syndrome	18 (6.7%)
vestibulocochlear dysplasia	12 (4.4%)
Mondini malformation	10 (3.7%)
ossified cochlea	4 (1.5%)
inflammatory middle ear changes	15 (5.6%)
high position of the jugular vein bulb	32 (11.8%)

Discussion and Conclusion

Eighty-five percent of children with SNHL are thought to suffer from congenital or early acquired hearing loss[9]. Our data show that 30.4% of children undergoing evaluation had a hereditary factor that may have contributed to their hearing loss. These data have not changed significantly compared with previous studies[2]. The most common risk factors potentially contributing to SNHL are birth factors (premature birth and prolonged neonatal intensive care unit stay), maternal factors (prenatal substance abuse, placental abruption, and toxemia) and acquired factors (meningitis, chemotherapy, ototoxic antibiotics and extracorporeal membrane oxygenation)[10]. The recent genetic studies have identified connexin 26 mutations in a significant proportion (50–80%) of patients with sporadic deafness[11]. Such information will give us better insight into the potential causes of SNHL.

Cochlear implantation (CI) involves the insertion of electrode systems in the inner ear in order to restore hearing in patients with sensorineural deafness. Cochlear implantation has been used to restore the sense of audition to deaf children. Positive CI results are closely related with careful selection of candidates. Pediatric cochlear implant selection criteria are age, bilateral profound sensorineural hearing loss, no benefit from appropriate amplification and willingness to participate in extensive rehabilitation program. The difficulty in determining with certainty that a small child is totally deaf and cannot benefit from a hearing aid is well known. For

this reason, a prolonged hearing-aid trial under close observation with appropriate aural rehabilitation is desirable for most children. Age at implantation is significant predictor of outcome in pediatric cochlear implantation. Prelingually deaf children should receive implants as early as possible to facilitate the later development of speech perception skills and speech intelligibility and thus maximize the health gain from the CI. Patient selection has emerged as one of the most important determinants of successful outcome after pediatric cochlear implantation. Preoperative temporal bone computed tomography (CT) and its results play an important role in selecting candidates for CI and circumvents potential difficulties and complications at CI. Detecting inner ear malformation in the pediatric population requires high-resolution axial and coronal CT scans[12]. High-resolution computed tomography has a high sensitivity for bony abnormalities. The children with anomalous cochleovestibular anatomy would have poorer outcomes and therefore would be poorer candidates as a result of their diminished ability to interpolate and use auditory information delivered through a cochlear implant[13]. These temporal bone anomalies are associated with a wide range of hearing loss, and presence or absence of related non-otological anomalies. The more severe the temporal bone deformity, the poorer is the hearing[14]. Perceptive and linguistic results depend on the type of the deafness. In progressive deafness, the perceptive and linguistic result are expected to be good. In congenital deafness, the results are more variable. Inner ear malformation in a child with profound hearing loss can no longer be considered to be a contraindication to cochlear implantation[15].

The patients included in this study represent 270 children with bilateral severe to profound sensorineural hearing loss – candidates for cochlear implantion at our department. This group of patients represent an increasing population considering mandatory newborn hearing screening in Croatia. The hearing loss was identified before 1 year of age in 59,6% of candidates and definite or probable cause of SNHL was identified in 58.9% of candidates. The results of the study might give us better insight into the potential causes of SNHL and allow more timely intervention, allowing children with SNHL to reach their potential.

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ETIOLOGIJA GLUHOĆE DJECE KANDIDATA ZA UMJETNU PUŽNICU U HRVATSKOJ

SAŽETAK

Cilj ove studije je bio prikupiti podatake o zamjedbenom oštećenju sluha kod djece kandidata za ugradnju umjetne pužnice u Hrvatskoj. Retrospektivna studija uključivala je 270 djece kandidata za ugradnju umjetne pužnice u razdoblju od siječnja 1997. do siječnja 2005. u našoj ustanovi. Klinička obrada kandidata uključivala je anamnezu, pregled, radiološku obradu temporalne kosti i audiološku obradu. Obiteljska anamneza na zamjedbeno oštećenje sluha bila je pozitivna kod 82 (30,4 %) kandidata. Prematuritet i/ili perinatalne komplikacije bili su prisutni kod 35 (12,9%) kandidata. CT obrada pokazala je malformacije unutarnjeg uha kod 44 (16,3%) kandidata. Siguran ili vjerojatni uzrok oštećenja sluha nađen je kod 58,9% kandidata, dok je kod 41,1% kandidata uzrok oštećenja sluha ostao nepoznat. Rezultati ove studije mogli bi nam dati bolji uvid u uzroke zamjedbenog oštećenja sluha i omogućiti nam pravovremeno djelovanje, omogućujući djeci sa zamjedbenim oštećenjem sluha da razviju svoj puni potencijal.