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After a first prelingually deaf child, does the family learn a lesson?

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BACKGROUND: Congenital sensorineural hearing loss (SNHL) is a common disability in children. It can affect normal language development and educational achievement. Today, the time to cochlear implant is delayed for many children, which in turn delays intervention and impacts outcomes. Lack of knowledge and experience with congenital SNHL in the family are critical factors that can delay identification and intervention.

OBJECTIVES: Compare treatment seeking behavior in families for a first and second congenitally deaf child. Design: Analytical, cross-sectional using medical record data.

SETTING: Ear specialist hospital in Riyadh.

SUBJECTS AND METHODS: All patients who presented to the cochlear implant committee from March 2016 to March 2018 and met criteria were included in the study. Data on when the subjects presented to hospital and were approved for cochlear implant were retrieved from the patient files and through phone calls to the family. The age of first suspicion, audiological testing, diagnosis, hearing aid fitting, and the decision for cochlear implant were compared between the first and second child in families with multiple children with congenital SNHL.

MAIN OUTCOME MEASURES: The timing difference between the first and second deaf child in seeking treatment.

SAMPLE SIZE: 116 (58 pairs).

RESULTS: The second child was suspected to have hearing loss 13.6 months earlier than the first child and presented to the cochlear implant committee for final decision 16.7 months earlier than his\her sibling. Differences in the mean ages at suspicion of hearing loss, presentation to the hospital for audiological evaluation, hearing aid fitting, diagnosis, and decision for cochlear implant by cochlear implant committee were statistically significant ($P<.001$).

CONCLUSION: Experience and knowledge has a major effect on early identification. We need to implement educational programs for the public to increase awareness of how to recognize a deaf child and what steps to take.

LIMITATIONS: Single-centered.

CONFLICT OF INTEREST: None.

In prelingually deaf children, sound deprivation can result in disruption or arrest of normal central auditory system development and cortical reorganization.[1] Cochlear implantation (CI) is efficacious in providing auditory stimulation by activating the central auditory pathway in deaf children.[1] However, a successful outcome is highly dependent on the age at implantation.[1] Earlier hearing loss identification and a shorter duration of deafness correspond to better outcomes.[1,2] In 2007, the Joint Committee on Infant Hearing (JCIH) stated that all infants with hearing loss should be diagnosed before 3 months of age, and should receive appropriate intervention by 6 months.[3] A study by Yoshinaga-Itano et al[4] found that children who were identified before 6 months of age had better language skills than those who identified after that age.

Application of universal neonatal hearing screening has a major role in the early identification of hearing loss progression. After implementation of hospital newborn hearing screening programs in the United States, the median age of diagnosis of hearing loss decreased dramatically from 11 months of age to 2 months, between 2006 and 2009.[5] However, not all newborns receive hearing screening, and passing the screening does not guarantee that progressive or late-onset hearing loss will not develop.[6]

Despite the emphasis on early identification and intervention, a study conducted in Saudi Arabia reported that the median age of hearing loss suspicion was 9.3 months. Moreover, the average delay between suspicion of hearing loss and diagnosis was 4 months, and the average time between diagnosis and fitting of amplification was 7 months.[7] In a study by Kitteral and Arjmand,[8] children with sensorineural hearing loss (SNHL) were grouped into three groups based on the etiology of hearing loss: genetic, at-risk, and not-at-risk groups. Children in the genetic group were suspected earlier than the children in the other groups. However, there was no difference in the age at diagnosis between the three groups.

Many children with SNHL experience delays between suspicion and the diagnosis of hearing loss. The most common reasons according to parents are the physicians' failure to corroborate the parents' suspicion of hearing loss.[9] In another study, knowledge deficit was a factor that delayed the choosing of cochlear implant as treatment.[10] In a study by Fitzpatrick, 9.3% of parents reported that the reason for the delay between diagnosis of hearing loss and cochlear implantation was family indecision.[11] Delay in identification of hearing loss and receiving appropriate intervention in children can have severe effects on speech, language, and cognitive development and can result in educational, social, and emotional difficulties.[12] Kirk et al studied the effects of age of implantation on 73 prelingually deaf children by analyzing the rate of growth of word recognition and language skills. Results showed that children younger than 3 years at implantation had significantly faster rates of language development than did the children with later implantation.[13] An article published in 2014 by Sarant et al reported that a family history of hearing loss predicted better outcomes (the study compared the language abilities of children with unilateral and bilateral CIs and analyzed the predictors of language development in this population). This finding emphasizes the importance of family experience and knowledge.[14] However, there is little in the literature on the effect of family experience and knowledge on early identification and presentation of their deaf child. To our knowledge, there are no studies comparing the first deaf child in the family with the second child on either time of presentation to health care centers or performance after cochlear implant. Given the impact of the timing of treatment seeking on patient outcomes, this study compared the families' approaches with their first deaf child and their second, and highlighted the impact of the family's experience with their first child on the timing of treatment initiation for the second affected child.

SUBJECTS AND METHODS

This analytical cross-sectional study was carried out in a tertiary-care hospital, the King Abdullah Ear Specialist Center (KAESC) at King Abdulaziz University Hospital in Riyadh, Saudi Arabia. The study included all pediatric patients with bilateral severe to profound hearing loss who were prelingually deaf, and presented to the cochlear implant committee for the first time, and had a brother or sister who was prelingually deaf as well. Excluded were adults, patients who presented for a second ear CI, patients with unilateral SNHL, patients who had developed a language before a CI, any patient with psychological or neurological conditions, and any patient with no brother or sister who was prelingually deaf.

This research was approved by the institutional review board at King Saud University Medical City. Confidentiality was maintained. The research was fully explained to all patient relatives, and verbal informed consent was obtained during the data collection phone call. Patient's parents were told that they were free to not participate and would not be included in the study if they wished.

We included all patients who presented to the cochlear implant committee at the center for the first time, between March 2016 and March 2018. Each pa-

tient with SNHL who was referred had presented to the cochlear implant committee at KAESC, registered and had undergone a process of evaluation and a series of investigations that were kept in a numbered electronic file in the center's registry. Subjects were selected from the CIC registry by applying inclusion and exclusion criteria by using a data collection sheet designed for this study. The data collection sheet consisted of two parts: The first part for recording the socio-demographic characteristics of the participants (i.e. age, gender, status of other ear, language status, medical status) and the second part for recording the ages at suspicion of hearing loss, diagnosis, hearing aid fitting, and presentation to the cochlear implant committee. Data were collected by phone calls with parents or direct caregiver (for those whose parents were deceased) and from patient files. The electronic patient record and the paper folders of the cochlear implant committee were both used in the data collection. The data were entered in an Excel sheet. The first child (control) was compared with the second child in terms of age (months) at suspicion, first audiological testing, confirmed diagnosis, hearing aids fitting, and date of presentation at cochlear implant committee.

Statistical tests were carried out using IBM SPSS software Version 22.0 (Armonk, NY: IBM Corp.). A paired sample *t* test was used to compare the study groups. A *P* value less than .05 was considered statistically significant.

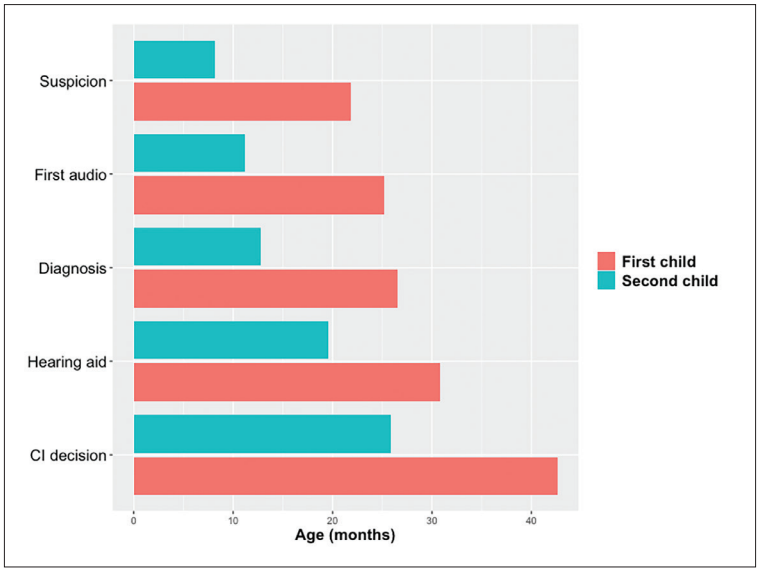

Figure 1. Ages of events related to hearing loss for first and second children.

RESULTS

One hundred and sixteen patients (58 pairs of brothers and/or sisters) fulfilled our inclusion criteria. Thirty (51.7%) of fathers and 22 (37.9%) mothers had a bachelors degree or higher. Forty-six (79.3%) fathers and 20 (34.5%) mothers were working at the time of the study. Most families (n=41, 70.5%) lived within 200 km of a cochlear implant center. The total monthly income of 31 (52.8%) families in the sample was 10000 Saudi Riyals or less. The mean ages for events related to hearing loss (suspicion, first audiological testing, confirmed diagnosis, hearing aid fitting, and presentation at cochlear implant committee) for each child are shown in **Figure 1**.

We found a statistically significant difference between the first and second child on the mean ages at suspicion of hearing loss, presentation to the hospital for audiological evaluation, hearing aid fitting, diagnosis, and decision for cochlear implant by the cochlear implant committee (paired sample *t* test, *P*<.001 (**Table 1**).

Table 1. The mean differences between the first and second child at each event.

	Mean difference	Standard. error of the mean	95% confidence interval of the difference		*P*
			Lower	Upper	
Age at suspicion	13.6	3.1	7.4	19.7	<.001
Age at first audio testing	13.9	3.1	7.6	20.2	<.001
Age at diagnosis	13.7	3.1	7.5	19.9	<.001
Age at HA fitting	11.3	3.3	4.7	17.8	.001
Age at cochlear implant committee	16.7	4.1	8.4	24.9	<.001

Values are mean age (months). Paired sample *t* test.

DISCUSSION

Cochlear implantation is the penultimate treatment for SNHL, but its effectiveness is time related. Better outcomes are expected if the child is presented early for investigation and intervention. As lack of knowledge and experience was one of the factors that delayed choosing cochlear implant as treatment,[10] the influence of family experience needed to be studied. The results of this study affirm the value of family knowledge on the early identification of hearing loss as well as on the process of cochlear implantation. The second child reached all tested checkpoints sooner and the journey to cochlear implant was completed by the mean age of 25.9 months, 16.7 months earlier than the older sibling. The most important gain in time was at the age of suspicion as the second child was suspected 13.6 months earlier than the first. After that, for both children it took about 3 months between suspicion and audiological testing and 1.5 months until a confirmed diagnosis. However, after having a confirmed diagnosis there was an unnecessary delay before being fitted with hearing aids for both groups (4.3 months for first child and 6.7 months for second child) which is contrary to the JCIH 2007 recommendation that they should be fitted withhearing aids within 1 month of diagnosis.[3] This delay might be explained by some patients needing a referral to tertiary care hospitals for hearing aids. Occasionally, they might be on a waiting list in governmental hospitals for hearing aids as hearing aids might not be available all the time.

The experience and knowledge of the family of congenital hearing loss has a major effect on the timing of presentation to the hospital. Based on the results of this study, the most dramatic reduction in time in the journey to seek treatment was at the initial suspicion of congenital hearing loss. A reduction in time at this point led to earlier treatment by cochlear implant. Knowing the importance of early intervention in congenital SNHL, we need to implement educational programs for the public to increase awareness of how to recognize a deaf child and what steps to follow. Additionally, parents of a deaf child need to be informed about the chances of having another child with the same condition so that they are alert for early signs of hearing loss. Although neonatal hearing screening has been the standard of care for all newborns in Saudi Arabia since 2016, those families with a hearing impaired child should be getting special treatment as a high-risk group by providing easier access to available services.

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