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Childhood hearing loss and risk profile in a South African population

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ABSTRACT

Objective: To describe the nature of hearing loss and associated risk profile in a South African population of infants and children diagnosed at a pediatric referral clinic.
Methods: A retrospective review of patient files for a pediatric auditory evoked potential clinic in Pretoria was conducted (January 2007–December 2011). Collected data included demographical information, risk factors from case history questionnaire, diagnosis (type and degree of hearing loss), documented age of caregiver suspicion and age of first diagnosis.
Results: Hearing loss was present in 73% (73/100) of cases evaluated. Permanent hearing losses (SNHL, ANSD and mixed) constituted 76% of losses. Unilateral hearing losses constituted 8% of SNHL and 20% of conductive hearing loss. ANSD was diagnosed in 21.4% and SNHL in 78.6% of permanent non-conductive hearing loss cases. The most prevalent SNHL risk was family history of hearing loss and for ANSD it was admittance to the NICU for more than 5 days. The majority of the sample was diagnosed with a permanent bilateral SNHL and ANSD after 36 months of age (47%) despite 40% already suspected of having a hearing loss before 12 months of age.
Conclusions: A high prevalence of ANSD was found with preventable risk factors often indicated. Age of diagnosis was significantly delayed, evidencing the lack of early hearing detection services in South Africa. The majority of children were diagnosed at ages precluding optimal benefits from early detection and subsequent intervention.

1. Introduction

More than 1 million babies are born annually in South Africa (1,059,417 in 2011) of which very few will be afforded the opportunity to have their hearing screened [1–3]. Despite the proven benefits of early detection of hearing loss and early intervention it is still uncommon practice in South Africa [3]. A survey of early detection services in the public health care system, which serves approximately 85% of the South African population [4], indicate that less than 7.5% of hospitals offer any infant hearing screening services [5]. The private health care system provides slightly better coverage with 53% of obstetric units offering some form of screening but only 14% offering universal newborn hearing screening [2]. Existing programs are also not sufficiently systematic and are plagued by suboptimal and variable protocols for early detection, follow-up and data management [2]. As a result it is estimated that less than 10% of South African newborns are likely to have their hearing screened [2,5]. Unsurprisingly the average age of hearing loss diagnosis has been reported to be between 23 and 31 months of age as opposed to the recommended 3 months of age [3,6,7].

Due to limited newborn hearing screening programs, resultant late identification, and insufficient data management [2,3,5] there is very limited systematic data on the nature and causes of permanent congenital and early onset hearing loss (PCEHL) in South Africa. The only reports date back to the 1970s and early 80s when a series of retrospective reviews of children in schools for the deaf were conducted across the country [8]. Within the diverse sample of 3064 school-aged children 25% presented with an acquired hearing loss, 7% with syndromic hearing loss, 11% with non-syndromic (familial) hearing loss and 57% with unknown causes (11% with other anomalies and 46% without other anomalies). The main risks associated with acquired deafness were maternal rubella, meningoencephalitis, "severe illness", jaundice, birth trauma and prematurity [8]. Since these results were reported much has changed in terms of the specification of risk factors and even the diagnostic categories

Abbreviations: ANSD, auditory neuropathy spectrum disorder; PCEHL, permanent congenital and early onset hearing loss; SNHL, sensorineural hearing loss.

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of hearing loss (i.e. auditory neuropathy spectrum disorder (ANSD)).

Recent reports have highlighted that developing regions such as sub-Saharan Africa may have risk factors for PCEHL that vary significantly from those established for developed world regions [9] and subsequently may present with a larger incidence of ANSD [10]. Unique developing world risk factors such as undernutrition, maternal high blood pressure and unskilled birth attendants has been associated with congenital and early onset hearing loss in Nigeria [9,11]. Alongside such unique risk factors is a higher incidence of existing risk factors (e.g. birth trauma, asphyxia, neonatal jaundice and ototoxicity) associated with poor maternal and child health services typical of many developing world regions [10,11,13,14]. The prevalence of ANSD has been reported to be as high as 16% in a population of Nigerian children with sensorineural hearing loss (SNHL) born outside hospital as opposed to a prevalence of 10% in a similar group born within hospital [10].

South Africa is characterized by diversity in culture, language and economic development, being classified as an upper middle-income country, largely developing with pockets of developed contexts [15]. To date there has been no description of the nature of PCEHL in South Africa. Apart from studies conducted in deaf schools three to four decades ago [8], no associated context-specific risk factors have been documented for childhood hearing loss. The current study therefore describes the nature of hearing loss and associated risk profiles in a South African population of infants and children diagnosed with hearing loss at a referral clinic.

2. Methods

Approval from the institutional ethics committee was obtained before any data collection commenced.

2.1. Study population

A retrospective review of patient files for the pediatric auditory evoked potential clinic at the University of Pretoria was conducted from January 2007 to December 2011. The clinic is scheduled once a week during university terms serving as a referral source in Pretoria and surrounding areas. Few public healthcare hospitals offer this type of diagnostic service in Pretoria and surrounding areas. If services are available waiting lists typically exceed 6 months. The current study clinic waiting period varies between 2 and 4 months. Evaluations were conducted by experienced pediatric audiologists employed at the University of Pretoria with support from final year audiology students. Test batteries comprise mostly objective test procedures including acoustic immittance measurements, otoacoustic emissions, auditory brainstem responses, auditory steady-state responses supplemented by behavioral audiometric procedures where possible.

2.2. Procedures

Records of all the patients who attended the pediatric auditory evoked potential clinic between January 2007 and December 2011 were reviewed in order to locate and obtain the patient files kept in the Department of Communication Pathology at the University of Pretoria. Files were drawn from the filing cabinet and relevant information documented onto a data collection sheet developed to assist the researchers in sorting, analyzing and organizing the data. Data collected included demographical information, risk factors documented on the case history questionnaire, test procedures conducted, diagnosis (type and degree of hearing loss), documented age of caregiver suspicion and age of first diagnosis. The captured data was subsequently transferred from the data collection sheet to an electronic database.

2.3. Data analysis

Data was analyzed on a statistical software package (IBM SPSS version 19). Descriptive measures were employed to describe the central tendency and normal distribution of recorded variables. A non-parametric test, the Mann–Whitney test, was employed to compare means between independent sub-samples using a significance level of 5%.

3. Results

Comprehensive diagnostic assessment information was obtained from the files of 100 children attending the pediatric hearing clinic at the University of Pretoria between January 2007 and November 2011. The rest of the files were incomplete and in some cases could not be sourced. More than half (53%) of the patients' caregivers were first language speakers of an African language as opposed to English (12%) and Afrikaans (35%). Less than half (46%) had access to some form of private medical aid. Referral sources for pediatric audiological assessments were from public health care hospitals (52%), speech–language therapists (14%), audiologists (11%), schools (10%), ENT specialists (4%) and other sources (9%).

3.1. Childhood hearing loss

Of the 100 children 73% (73/100) presented with hearing loss. Table 1 provides a description of the types of hearing losses across the sample. Permanent hearing losses (SNHL, ANSD and mixed) constituted 76% of hearing losses of which 4% included an additional conductive component (mixed). One in four hearing losses was purely conductive (23%) in nature. Except for one case (n = 1/17) of bilateral atresia all the other conductive losses (n = 16/17), including the mixed losses (n = 3), were due to middle ear effusion related to otitis media. Unilateral hearing losses constituted 8% of SNHL and 20% of conductive hearing loss. ANSD constituted 21.4% and SNHL (including mixed hearing losses) constituted 78.6% of permanent hearing losses (SNHL, ANSD and mixed). The minimum diagnostic criteria for diagnosing auditory neuropathy were the presence of otoacoustic emissions and/or a cochlear microphonic response between 80 and 90 dB with absent or severely abnormal ABR waves [16].

As illustrated in Fig. 1 approximately 50% of SNHL was of a profound degree (including moderate to severe, severe to profound and profound). All mixed hearing losses (n = 6) presented as a profound hearing loss. In 42% (n = 10/24) of ears diagnosed with ANSD the degree of hearing could not be specified. The remaining distribution of ears diagnosed with ANSD presented with a profound (57%; n = 8/14), severe (29%; n = 4/14) or mild (14%; n = 2/14) degree of hearing loss. Two thirds of ears with conductive

Table 1
Distribution of hearing losses (n = 73). n = number of cases; mixed hearing loss = SNHL and conductive.

Types of hearing loss	Bilateral		Unilateral		Uni- and bilateral	
	n	%	n	%	n	%
Sensorineural hearing loss	37	51%	4	5%	41	56%
Auditory neuropathy	12	16%	–	–	12	16%
Mixed hearing loss	3	4%	–	–	3	4%
Conductive hearing loss	13	18%	4	5%	17	23%
Total	65	89%	8	11%	73	100%