

CHILDHOOD HEARING LOSS AND RISK PROFILE IN A SOUTH AFRICAN POPULATION

De Wet Swanepoel^{1,2,3}

Lorné Johl¹

Danelle Pienaar¹

1. *Department of Communication Pathology, University of Pretoria, South Africa*
2. *Ear Sciences Centre, School of Surgery, the University of Western Australia, Nedlands, Australia*
3. *Ear Science Institute Australia, Subiaco, Australia*

Corresponding author:

Prof. De Wet Swanepoel

Department of Communication Pathology

University of Pretoria

C/o Lynnwood & University Roads

Hatfield, 0002

South Africa

Dewet.swanepoel@up.ac.za

ABSTRACT

Objective: To describe the nature of hearing loss and associated risk profile in a representative 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 to 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 & 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.

Keywords

Auditory neuropathy; hearing loss; infant; children; hyperbilirubinemia, risk factors; sensorineural, conductive.

Abbreviations

ANSD – Auditory neuropathy spectrum disorder

PCEHL – Permanent congenital and early-onset hearing loss

SNHL – Sensorineural hearing loss

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 [1,2,3]. 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 to 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 1970's and early 80's 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 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, 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.

METHODS

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

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. 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.

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.

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%.

RESULTS

Comprehensive diagnostic assessment information was obtained from the files of 100 children attending the paediatric 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 paediatric audiological assessments were from public health care hospitals (52%), speech-language therapists (14%), audiologists (11%), schools (10%), ENT specialists (4%) and other sources (9%).

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 & 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

Table 1. Distribution of hearing losses (n=73). *n* = number of cases; *Mixed hearing loss* = *SNHL & Conductive*

Types of hearing loss	Bilateral		Unilateral		Uni- & 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%

21.4% and SNHL (including mixed hearing losses) constituted 78.6% of permanent hearing losses (SNHL, ANSD & Mixed).

As illustrated in figure 1 approximately 50% of SNHL were of a profound degree (including moderate to profound, 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 hearing loss had a mild degree of hearing loss (70%; n=21/30) with a small number presenting with moderate (20%; n=6/30) and severe (7%; n=2/30) degrees of hearing loss (3% had no degree specified).

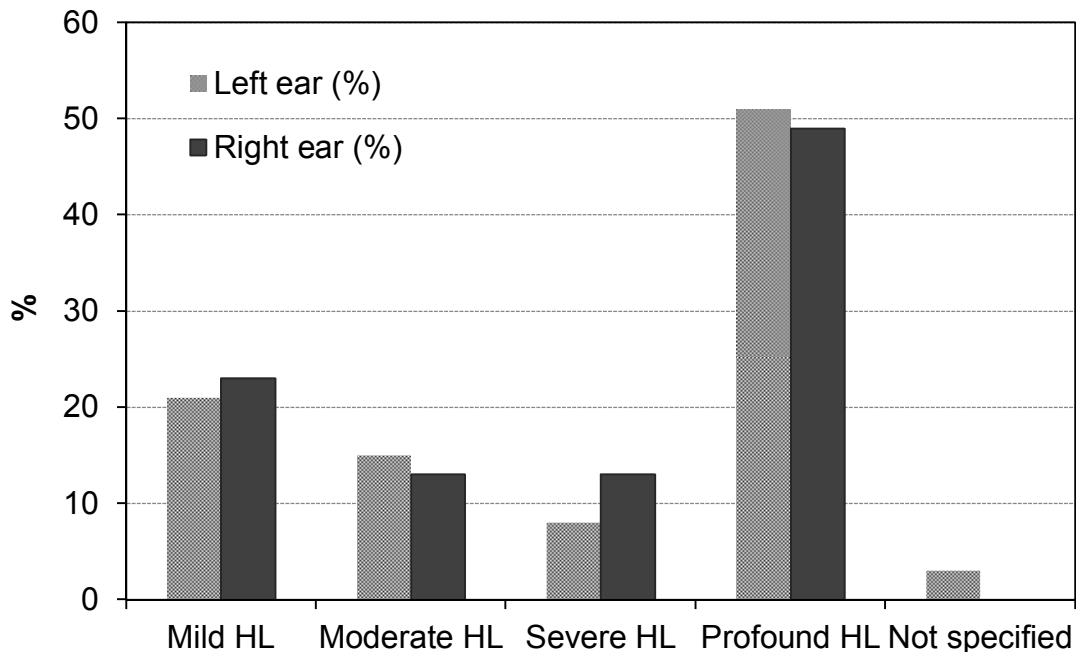


Figure 1. Distribution of sensorineural hearing loss degree (excluding ANSD) across ears (n=78)

Risk profiles

Table 2 provides a summary of the risk factors documented for the subjects with hearing loss. The syndromes included in the sample were Goldenhar, Cri-du-Chat and Prader Willi syndrome. The Cri-du-Chat subject was diagnosed with ANSD and the other with SNHL. The most prevalent SNHL risk was family history of hearing loss (27%) and for ANSD it was admittance to the NICU for more than 5 days (70%).

Table 2. Documented risk factors (*n*= number of children affected with completed risk profiles)

Risk Factors	No HL (n=19) % (n)	SNHL (n=29) % (n)	ANSD (n=10) % (n)	SNHL & ANSD (n=39) % (n)
Admittance to NICU > 5 days	16 (3)	14 (4)	70 (7)	27.5 (11)
Hyperbilirubinemia	-	3.5 (1)	50 (5)	15 (6)
Asphyxia	10.5 (2)	10 (3)	50 (5)	20 (8)
Extremely LBW < 1500g	21 (4)	3.5 (1)	40 (4)	12.5 (5)
Family history of hearing loss	16 (3)	27.5 (8)	20 (2)	25 (10)
Syndrome present	16 (3)	-	10 (1)	2.5 (1)
Congenital infections	-	-	10 (1)	2.5 (1)
Craniofacial defects	-	3.5 (1)	-	2.5 (1)
Bacterial meningitis	-	3.5 (1)	-	2.5 (1)

Age of hearing loss suspicion and diagnosis

Within the group diagnosed with bilateral permanent hearing loss (SNHL, ANSD & Mixed hearing loss; n=52), only 36 caregivers reported the age at which they suspected the presence of a hearing loss for the first time (Table 3). The majority of the sample population was diagnosed with a permanent bilateral SNHL and ANSD after 36 months of age (47%) despite 40% of this group already suspected of having a hearing loss before 12 months of age. A delay of 9 months or less between suspicion and diagnosis was evident for 25% of bilateral permanent losses. Only 20.4% of children with bilateral SNHL and ANSD were diagnosed before 18 months

of age and less than one in three (30.6%) before 24 months of age. Unilateral losses were suspected only after 36 months of age in 75% (n=3/4) of cases.

Table 3. Age of bilateral hearing loss suspicion and diagnosis (*n* = number of individuals affected)

	Age at suspicion (months)	Age at diagnosis (months)	Delay from suspicion to diagnosis (months)
	<i>n</i> =36	<i>n</i> =49	<i>n</i> =36
Mean	23.7	42.1	22.0
SD	18.9	27.6	16.3
Max	72.0	128.2	68.2
Min	2.0	2.2	0

There was no significant difference between the age of hearing loss suspicion and age of diagnosis for families with or without medical insurance ($p>0.05$; Mann-Whitney Test). No significant difference was also evident between the age of hearing loss suspicion for African first language compared to English and Afrikaans first language speaking families ($p>0.05$; Mann-Whitney Test).

Average age of HL suspicion in cases of ANSD was 32 months (28.7 SD; 6 cases) compared to 22 months (16.2 SD; 30 cases) for bilateral SNHL but was not statistically significant however (Mann-Whitney; $p>0.05$). The average age of HL

diagnosis in cases of ANSD was 45.1 months (41.1 SD; 12 cases) compared to bilateral SNHL of 41.1 months (22.6 SD; 37 cases) but was also not statistically significant (Mann-Whitney; $p>0.05$).

DISCUSSION

The cultural diversity and economic inequality of the South African population was represented in the research sample with the majority being African first language speakers (53%), more than half (54%) without private medical insurance and the majority (52%) referred from public health care facilities [1,4,15].

Three in every four children assessed at the clinic presented with a hearing loss of which 69% had a bilateral, and 7% had a unilateral permanent hearing loss (SNHL, ANSD & mixed). ANSD cases constituted a larger proportion (21.4%) of permanent hearing losses (SNHL, ANSD & mixed) than previously reported. Previous reports have indicated prevalence for ANSD of between 5 to 17% of SNHL in children [16,17,18,19]. This prevalence has shown to increase when only severe to profound degrees of SNHL are considered [20]. The only other report from sub-Saharan Africa was from Nigeria, which reported ANSD prevalence of 15.9% in SNHL cases for a cohort of babies born outside of hospital and 10.3% for a cohort born in hospital [10]. The current study ANSD prevalence is still slightly higher and may be attributed in part to the nature of the research cohort, which comprised a group of children referred for diagnostic testing at a specialized university clinic. A complex case, typical of ANSD, is more likely to be referred for diagnosis at a specialized university clinic. Furthermore however it may also reflect the increased environmental and

maternal and child health related risk factors (hyperbilirubinemia) predisposing ANSD in populations from developing contexts such as sub-Saharan Africa [10,12].

Within the sample population diagnosed with ANSD the most prevalent associated risk factor was admittance to the NICU for longer than five days. Olusanya et al.,[21] indicated that more than half (54.5%) of the children diagnosed with ANSD were admitted to hospital during the neonatal period for serious illness. NICU graduates have a significantly higher risk of ANSD as opposed to low-risk well baby populations [7,22]. Hyperbilirubinemia and asphyxia were the second most prevalent risk factors (50%) associated with ANSD. This is in agreement with the multi-centre study by Berlin and colleagues [17] wherein 48% of ANSD cases presented with hyperbilirubinemia as risk factor. Hyperbilirubinemia is more prevalent in African countries due to a higher incidence of glucose-6-phosphate dehydrogenase deficiency and lack of adequate medical treatment [13,23,24]. The Berlin and colleagues [17] study also reported a high prevalence for asphyxia-related risks including anoxia (17%), respiratory distress (15%) and artificial ventilation (23%).

The risks associated with SNHL were significantly less prevalent in comparison with the ANSD cases. The primary risk factor associated with SNHL was family history (27.5%), which was significantly higher than the 11% previously reported for South African children with hearing loss [8]. There is the possibility of this risk factor being over-reported in the current study since the case history form, completed by caregivers, specifies "*any family history of childhood hearing loss*", which may include reports of transient conditions. Considering the increase in prevalence between those children with SNHL and those without, reveal the prevalence (11.5%)

to be similar to that of Sellars & Beighton [8]. The other risks associated with SNHL included perinatal factors such as NICU admittance and asphyxia.

The degree of hearing loss was profound for 50% of SNHL ears and 57% of ANSD ears. A previous report on degree of hearing loss in a South African group of children indicated severe and profound degrees of hearing loss (61%) similar to that of the current study [6]. In a multicentre study of ANSD cases, profound hearing loss was reported in 37% of ears [17].

There was a substantial delay between the age of hearing loss suspicion by caregivers and the age of eventual diagnosis in the study population. Caregivers generally suspected hearing loss by 23.7 months of age, although the average age of diagnosis of a permanent SNHL was only at 42.1 months indicating an average delay of 22 months. These long delays evidence the limited newborn hearing screening programs available in public and private health care sectors [2,5]. The result is passive detection relying on caregivers who usually suspect hearing loss only after critical language development milestones have already passed [3]. Average age of suspicion for ANSD (32 months) was later and more variable compared to SNHL (22 months). This may partly be related to the inconsistent auditory responses that often characterize this population [17].

The mean age of parental suspicion in a Nigerian sample of school-aged children was reported to be between 12 to 24 months of age with an average 18 month delay until diagnosis of a hearing loss [25]. Previous studies in South Africa of children surveyed in schools for the deaf [6,26] reported average age of diagnosis to be 23

months in the Western Cape and 31 months in the Gauteng provinces of South Africa[6,26]. The current study, also situated in the Gauteng province, presented an average age of diagnosis significantly later (42 months) than the previous reports. Most likely reasons for the difference between studies relate to the difference in sample populations. As opposed to an unselected population referred for diagnostic hearing assessment the previous report for Gauteng province [26] included a population of children already enrolled in early intervention programmes. As a result they are predisposed to be from more developed socio-economic settings with infant hearing screening being more likely and access to services being better [2,3].

CONCLUSION

Hearing loss across this paediatric South African sample were typically permanent with a high prevalence of ANSD. Risks for ANSD were mostly preventable perinatal factors related to maternal and child health care. Age of hearing loss diagnosis was significantly delayed evidencing the lack of early detection services in the public and private health sectors of South Africa. The majority of children are diagnosed at ages which preclude optimal benefits from early detection and subsequent intervention.

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