

**PROFOUND CHILDHOOD HEARING LOSS IN A SOUTH AFRICA COHORT: RISK
PROFILE, DIAGNOSIS AND AGE OF INTERVENTION**

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ABSTRACT

Objective: To describe profound childhood hearing loss in a South African population of pediatric cochlear implant recipients in terms of risk profile, and age of diagnosis and intervention.

Methods: A retrospective review of patient files for 264 pediatric cochlear implant recipients from five cochlear implant programs was conducted. Data was captured from 264 eligible subjects, of which all were implanted between 1996 and 2013 and PCEHL was confirmed under the age of five years old. Data collected included demographical information, risk factors from case histories, diagnostic test procedures conducted, diagnosis (type, onset and degree of hearing loss) and documented ages of caregiver suspicion, initial diagnosis and intervention.

Results: Risk factors for permanent childhood hearing loss were present in 51.1% of cases, with the most prevalent risks being NICU admittance (28.1%), family history of childhood hearing loss (19.6%) and prematurity (15.1%). An associated syndrome was diagnosed in 10% of children and 23.5% presented with at least one additional developmental condition. Hearing loss for most (77.6%) children was confirmed as congenital/early onset, while 20.3% presented with postnatal onset of hearing loss. ANSD was diagnosed in 5% of children, with admittance to NICU (80%) and hyperbilirubinemia (50%) being the most prevalent risk factors for these cases. Hearing loss was typically diagnosed late (15.3 months), resulting in delayed initial hearing aid fitting (18.8 months), enrollment in early intervention services (19.5 months) and eventual cochlear implantation (43.6 months).

Conclusion: Most prevalent risk factors in profound childhood hearing loss were admittance to NICU, family history and prematurity. Diagnosis and intervention was typically delayed predisposing this population to poorer outcomes.

Keywords: profound hearing loss, risk factors, children, intervention, hyperbilirubinemia, auditory neuropathy

Abbreviations: ANSD, auditory neuropathy spectrum disorder; EHDI, early hearing detection and intervention; NICU, neonatal intensive care unit; NHS, newborn hearing screening; PCEHL, permanent congenital and early onset hearing loss; SNHL, sensorineural hearing loss.

INTRODUCTION

Congenital or early onset permanent bilateral hearing loss affect an estimated 798 000 newborns annually [1]. At least 90% of these reside in developing countries around the world, implying that almost 2000 infants with hearing loss are born daily in developing world regions [2]. Based on an estimated incidence of six per 1000 live births, 180 000 infants with permanent hearing loss are born annually in sub-Saharan Africa alone [3,4]. Profound hearing loss prevalence in developing regions is largely unknown with only a few previously reported estimates [5,6,7,8]. Although comprehensive population studies for Africa do not exist, available reports suggest that the prevalence of profound hearing loss is higher than the estimated 20-30% of children with permanent childhood hearing loss in the developed world [3,9,10,11].

The lifelong consequences of permanent congenital and early onset hearing loss (PCEHL) are well documented [12-15]; however, these consequences are exacerbated for children and their families when a profound degree of hearing loss is diagnosed. These include the lack of development of spoken language which results in restricted learning, literacy and educational achievements, as well as later employment opportunities [10,16]. Profound hearing loss also results in a considerable cost for both the child and society [16] with the costs expected to be even higher in developing countries [11]. Early auditory stimulation during periods of maximal receptiveness is therefore critical for this population, since congenital/early onset profound hearing loss alters the functional properties of the auditory system and impairs cortical development [10, 16-18].

Unfortunately it is estimated that less than 10% of the more than 1 million babies born annually in South Africa will have their hearing screened, implying that children with hearing loss will most likely miss out on necessary early auditory stimulation [19-21]. Within the public health care system, which serves approximately 85% of the South African population [22], less than 7.5% of hospitals offered any infant hearing screening services when surveyed in 2008 [19]. Slightly better coverage is provided in the private health care system, with 53% of obstetric units offering some form of screening, but only 14% offering universal newborn screening [20]. As a result, the average age of hearing loss diagnosis in South Africa has been reported to be between 23 to 44.5 months [23-25], in contrast to the recommended age of 3 months [26].

Despite recent reports on early hearing detection services in the public and private health care sectors of South Africa [19,20], information on the status of intervention in terms of amplification and enrollment into early intervention programs is limited [2]. Contextual data on profound childhood hearing loss, in particular, is non-existing. A report from the Western Cape

province on a representative sample of 54 children with hearing loss, most (61%) with severe to profound hearing loss, indicated the average age of initial hearing aid fitting and enrolment in early intervention to be 28 and 31 months respectively [23]. A survey conducted amongst speech therapy and audiology departments within public sector hospitals in South Africa reported that within a sample of 76 children aged 18 months or younger that were fitted with hearing aids 12 months prior to the survey, less than 7% received hearing aids by the age of 6 months, as recommended [19,26,27].

As a result of limited early hearing detection and intervention (EHDI) programs and poor data capturing and management amongst existing programs [20,24] the prevalence and nature of PCEHL in South Africa is largely unknown along with the associated risk profiles. Except for a series of etiological surveys of children in schools for the deaf dating back to the 1970s and early 80s [28], no data has been available to describe the risk profile of PCEHL in South Africa. At the time of these early etiological reports [28-30], diagnostic categories of hearing loss did not include auditory neuropathy spectrum disorder (ANSD). Also, with the advent of newborn hearing screening (NHS) the risk profiles for PCEHL were expanded and described more accurately [31]. This was not accounted for in these early South African reports [28-30]. Only in a recent report was the nature of hearing loss and associated risk profile described with consideration of ANSD for a population of infants and children diagnosed at a pediatric referral clinic in South Africa [24]. More than half of the diagnosed children (56%) presented with sensorineural hearing loss, with 50% being of a profound degree. ANSD was diagnosed in 21% of the cases, suggesting a larger prevalence for populations from developing contexts such as sub-Saharan Africa, as has previously been reported. This is attributed to an increased incidence of environmental, maternal and child health related risk factors predisposing ANSD [32,33].

Children with profound hearing losses are known to be identified at earlier ages and are predisposed to enter early intervention services earlier than children with less severe degrees of hearing loss [34]. However, the initiation of early intervention services are often delayed in the resource limited settings such as sub-Saharan Africa, where poor healthcare infrastructure, the lack of audiological services and widespread poverty impede the attainment of developed world benchmarks for intervention [4,23,26,35].

It can be expected that the risk profile for children with profound hearing loss may show marked distinctions from children with less severe degrees of hearing loss. Profound childhood hearing loss is more than just a sensory loss, since central nervous system consequences of congenital deafness are aggravated with an increase in degree of hearing loss [10]. Also, approximately 30% of children with a profound hearing loss are reported to have an additional disability, with cognitive impairment and neurodevelopmental disabilities being the most common [36,37].

Since the epidemiological profile of PCEHL differs across various regions of the world and since risk factors have been reported mostly for school-aged children [31], profiling the risk factors for profound hearing loss in younger children is an important epidemiological endeavor, especially in developing countries [31].

Recently reported findings from Swanepoel et al. [24] provide preliminary data on the nature of hearing loss and associated risk profiles for a small sample of infants with hearing loss in South Africa. However, data pertaining to additional developmental conditions and intervention was not available for this sample population. The current study therefore investigates profound childhood hearing loss in a South African population of pediatric cochlear implant recipients considering associated risk profiles, the diagnosis of hearing loss and age of intervention.

METHOD

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

Study population

There are currently eight independent cochlear implant programs throughout South Africa. All eight programs were approached to participate in this multicentre study, from which five programs committed to participation. Four programs are situated in the Gauteng Province, while the remaining program is in the Free State Province. A retrospective review of the patient files of pediatric cochlear implant recipients at these participating five programs was conducted. Data captured within a 8 month period resulted in a dataset of 264 eligible pediatric cochlear implant recipients, of which all were implanted between 1996 and 2013 and PCEHL was confirmed under the age of five years old. The children included in this study sample were diagnosed with PCEHL at various diagnostic audiology clinics throughout South Africa. When candidacy for cochlear implantation was confirmed, the children were referred to the nearest cochlear implant program for assessment. Once approved and implanted, a comprehensive patient file was opened for each child, containing records of their pre-operative case history and diagnostic audiological assessment data.

Procedures

Patient registers were reviewed at each of the five participating cochlear implant programs in order to locate pediatric cochlear implant recipients who were South African residents, and for whom PCEHL was confirmed under the age of five years old. The clinical files of the children who complied with these criteria were drawn from the filing cabinets at each participating

cochlear implant program and then reviewed retrospectively. Data captureurs were identified and trained for each participating cochlear implant program. An electronic database was developed to organize and capture the data in a consistent format amongst the participating programs. Data collected included demographical information, case history questionnaires containing documented risk factors, diagnostic test procedures conducted, diagnosis (type, onset and degree of hearing loss), as well as the age of caregiver suspicion, initial diagnosis and intervention.

Data processing and analysis

A commercially available statistical software package (IBM SPSS version 21) was utilized to analyze the obtained data. The central tendency parameters and the degree of variation of the captured variables were calculated using descriptive statistics. For the capturing of pre-operative behavioral pure tone and Auditory Steady State Response (ASSR) threshold data, when a “no response” was indicated in the clinical file at a respective frequency, the threshold was captured as 120 dBHL. The minimum diagnostic criteria considered for ANSD were the presence of oto-acoustic emissions and/or a response for a cochlear microphonic between 80 and 90 dB nHL, with the absence of or severely abnormal Auditory Brainstem Response (ABR) waves [38].

RESULTS

Demographics

The study population of 264 children were all approved for cochlear implantation and showed an even gender distribution (50.4% male and 49.6% female). Only 7.6% of children were public health care patients compared to 92.4% private health care patients. Ethnical category was distributed as 66.2% White, 20.5% Black, 7.6% Indian/Asian and 5.7% Colored. Just more than half (50.8%) of the children’s home languages was Afrikaans, 40.7% was English and 5.6% was

an African language (n=248). Most children (95.4%) had normal hearing parents, while both parents had hearing loss in 2.5% of the cases and one parent had hearing loss in 0.8% of the cases (n=241). The majority of children (73.8%) were communicating orally and received either auditory/oral or auditory/verbal style education. The current communication mode for 6% of the children was South African Sign Language, while 13.1% used Total Communication and the remaining 7.2% alternative manual modes of communication (n=252).

Risk profile

A total of 23 children (10%) were diagnosed with a syndrome (Table 1), with Waardenberg syndrome being the most prevalent (5.2%; n=12/229). Of the total sample, 23.5% (n=55/234) presented with at least one additional developmental condition (Table 1). The most frequently occurring conditions for this population were visual impairment (8.5%; n=20/234).

Table 1: Presence of syndromes and additional developmental conditions

Syndrome	% (n)
Waardenberg Syndrome	5.2 (12/229)
Ushers Syndrome	1.3 (3/229)
Pierre Robin Syndrome	0.9 (2/229)
Leopard Syndrome	0.9 (2/229)
Additional developmental conditions	% (n)
Visual impairment	8.5 (20/234)
Mobility impaired	3 (7/234)
Cerebral palsy	5.6 (13/234)
Autism	2.1 (5/234)
Apraxia	1.7 (4/234)
Cleft lip and/or palate	1.3 (3/234)

Within the sample of children diagnosed with ANSD specifically, 58.3% (n=7/12) presented with at least one additional developmental condition. Most occurring conditions for this

population was documented as visual impairment (16.7%; n=2/12) and cerebral palsy (16.7%; n=2/12).

Birth type was specified to be normal delivery in 40% of the cases and caesarian section in 60% of the cases (n=80). Average birth weight was 2543.2g (range: 710 - 4100, 902.6 SD; n=91) and average gestation age was 37 weeks (range: 24 - 42, 4.7 SD; n=128). Prenatal, natal and postnatal risk factors are listed in Table 2.

Table 2: Risk factor prevalence for profound childhood hearing loss (n=264)

	Total population n=264	ANSD n=12
Risk factors	% (n)	% (n)
Pre-natal risk factor		
Family history of permanent childhood hearing loss	19.6 (40/204)	10 (1/10)
Rubella	5.5 (12/219)	20 (2/10)
Cytomegalovirus	3.2 (7/219)	-
Twin/ triplet	3.2 (7/219)	10 (1/10)
Syphilis	0.5 (1/219)	-
Toxoplasmosis	0.5 (1/219)	-
Natal risk factor		
Admittance to NICU	28.1 (43/153)	80 (8/10)
Prematurity (<34 weeks gestation)	15.1 (33/219)	40 (4/10)
Low birth weight (<2500g)	8.7 (19/219)	10 (1/10)
Extremely low birth weight (<1500g)	5.5 (12/219)	10 (1/10)
Birth asphyxia	1.8(4/219)	-
Maternal hypertensive disorder in pregnancy	1.8 (4/219)	10 (1/10)
Rupture of membranes	0.9 (2/219)	10 (1/10)
Rh incompatibility	0.9 (2/219)	10 (1/10)
Birth trauma	0.5 (1/219)	-
Post natal risk factor		
Neonatal jaundice/ hyperbilirubinemia	10.5 (23/219)	50 (5/10)
Blood transfusion	2.3 (5/219)	20 (2/10)
Meningitis	10 (22/219)	-
Viral infection (unspecified)	5 (11/219)	10 (1/10)
Mumps	0.9 (2/219)	-
Measles	0.5 (1/219)	-
Tuberculosis	0.5 (1/219)	-

Admittance to neonatal intensive care unit (NICU) was recorded as the most prevalent risk factor for 28.1% of the total population (n=43/153). However, only a smaller subset of data was available pertaining to duration of NICU stay, indicating that 90.2% (n=37/41) of children were admitted to NICU for longer than 5 days. More than half (51.1%; n=112/219) of the study sample presented with one or more risk factors, while 48.9% (n=107/219) presented with no associated risk factors for hearing loss (Table 3). The occurrence of natal (23.3%; n=51/219) and postnatal (34.7%; n=76/219) risk factors were more evident than pre-natal (12.3%; n=27/219) risk factors.

Table 3: Occurrence of risk factors in children with profound hearing loss (n=264)

	PRE-NATAL risk factors % (n)	NATAL risk factors % (n)	POST-NATAL risk factors % (n)	TOTAL % (n)
Available data	83 (219/264)	83 (219/264)	83 (219/264)	83 (219/264)
No risk factors	87.7 (192/219)	76.7 (168/219)	65.3 (143/219)	48.9 (107/219)
Risk factors present	12.3 (27/219)	23.3 (51/219)	34.7 (76/219)	51.1 (112/219)
1 risk factor present	11.9 (26/219)	13.7 (30/219)	29.2 (64/219)	28.8 (63/219)
2 risk factors present	0.5 (1/219)	7.3 (16/219)	4.1 (9/219)	11 (24/219)
3 or more risk factors	-	2.3 (5/219)	1.4 (3/219)	11.4 (25/219)

Diagnosis and degree of permanent childhood hearing loss

Hearing loss for most (77.6%) children was confirmed as congenital/early onset, while 8.5% presented with progressive hearing loss, 11.8% with sudden hearing loss and 2% with unknown onset of hearing loss (n=246). The vast majority of children (95%) presented with sensorineural hearing loss (SNHL) (n=228/240) and 5% presented with ANSD (n=12/240). From the 264 clinical files that were reviewed for this study, no pre-operative hearing data was available for 60 (22.7%) children. The remaining 204 files contained pre-operative pure tone thresholds for 144

children (70.6%). Only the ears with comprehensive threshold data available at all required frequencies (0.5; 1; 2 and 4 kHz) were included. The mean pure tone average (PTA) air-conduction threshold (average of 0.5; 1; 2 and 4 kHz) for 197 ears was 108.6 dBHL (range: 90.0 - 120 dBHL; 9.2 SD). Available pre-operative behavioral audiological data confirmed a profound (>90 dBHL) degree of hearing loss in all ears with available data.

In line with the national cochlear implant evaluation protocol [39] all children (n=264) underwent diagnostic electrophysiological assessment pre-operatively and these original diagnostic test results were only available for 95 (36%) children. The files of these children were reviewed for ABR and ASSR test results specifically. The files of fifty (52,6%) children contained both ABR and ASSR test results, while 24 files (25.3%) contained only ABR test results and 21 files (22.1%) only ASSR test results (n=95). Pre-operative click-ABR threshold data was available for a total of 141 ears. A “no response” was obtained at the maximum output of the equipment (90 dBnHL) for 138 ears (97.9%), while for the remaining 3 ears (2.1%), an average click ABR threshold of 86.7 dBnHL (range: 80 - 90 dBnHL; 5.8 SD) was obtained (n=141). ASSR thresholds were available for 71 children and only the ears with comprehensive threshold data available at all required frequencies (0.5; 1; 2 and 4 kHz) were included. In a total of 119 ears the average ASSR threshold (determined for 0.5; 1; 2 and 4 kHz) was 112.0 dBHL (range: 90 – 120 dBnHL; 9.0 SD). Available pre-operative electrophysiological audiological data confirmed on average a profound (>90 dBHL) degree of hearing loss in all ears with available data.

Age of hearing loss suspicion and diagnosis

Data on whether newborn hearing screening (NHS) was conducted was available for only 85 children, of whom it was indicated that NHS was not done for 72.9% of them. Within the group of children with a confirmed congenital/early onset hearing loss (n= 191), only 64 caregivers reported the age at which they suspected the presence of the hearing loss for the first time (Table 4). The average age of diagnosis was 15.3 months (9.3 SD), with a delay of 5.3 months between suspicion and diagnosis of the hearing loss. The majority of children (94%) presented with a prelingual hearing loss (n=249).

Table 4: Age of congenital/early onset hearing loss suspicion and diagnosis

	Age at suspicion (months) <i>n=64</i>	Age at diagnosis (months) <i>n=121</i>	Delay from suspicion to diagnosis (months) <i>n=61</i>
Mean	11.3	15.3	5.3
S.D.	7.8	9.3	5.5
Max	36.0	45.0	27
Min	1.0	0.5	0

Age of intervention

Age of intervention was determined for children with confirmed congenital/early onset hearing loss (n=191/246). For this group the average age at implantation was 43.6 months (31.2 SD), with a delay of more than two years (24.7 months; 27.1 SD) between diagnosis and implantation (Table 5). Ages of initial hearing aid fitting and initial enrollment in early intervention services were available for 108 and 36 children, respectively (Table 5). Early intervention services, being either home- or centre-based, refer to any type of habilitative, rehabilitative or educational program provided to children with hearing loss and their parents [26]. On average, initial hearing aid fitting occurred at the age of 18.8 months (10.7 SD), 2.6 months (4.9 SD) after diagnosis.

Table 5: Ages at initial hearing aid fitting, cochlear implantation and initial enrollment in early intervention services (HA = hearing aid; CI = cochlear implantation; EI = early intervention)

	Age at initial HA fitting (months) <i>n=108</i>	Delay from diagnosis to fitting (months) <i>n=101</i>	Age at CI (months) <i>n=186</i>	Delay from diagnosis to CI (months) <i>n=116</i>	Age at initial enrollment in EI (months) <i>n=36</i>	Delay from initial HA fitting to EI services (months) <i>n=31</i>
Mean	18.8	2.6	43.6	24.7	19.5	3.2
S.D.	10.7	4.9	31.2	27.1	12.6	6.7
Max	51.0	33.0	188.0	164.1	60	27.0
Min	1.0	0	6.0	3.0	2	0

The average age of initial enrollment in early intervention services was 19.5 months (27.1 SD), with a delay of 3.2 months (6.7 SD) between initial hearing aid fitting and enrollment in early intervention services.

DISCUSSION

The risk profile of South African children with profound hearing loss in this study showed the most prevalent risks to be NICU admittance (28.1%), family history of childhood hearing loss (19.6%) and prematurity (15.1%). Occurrence of natal (23.3%) and postnatal (34.7%) risk factors were more common than pre-natal (12.3%) risk factors. NICU stay for longer than five days is considered as one of the most common risk factors for childhood hearing loss [24,40]. Data on the duration of NICU stay for this study was only available for 41 children. Of this smaller subset, admittance to NICU for longer than 5 days was indicated for 90.2% of children (n=37/41). For the cohort of children diagnosed with ANSD, admittance to NICU was also indicated as the most prevalent (80%) risk factor. NICU admittance is an established risk factor for ANSD [26,41] as indicated in a Nigerian study where over half (54.5%) the children diagnosed with ANSD were admitted to hospital for serious illness within the neonatal period [1].

Caesarian section constituted more than half (60%) of the births in the study sample. Whilst unusually high South Africa is known for high rates of caesarian births. Caesarian births in South Africa has been reported to constitute 21% of births en general and 43.1% of births amongst White women [42]. With 66% of the current sample being White this is likely to have increased the number of represented caesarian sections. Furthermore, the influence of malpractice litigation and a higher prevalence of high-risk pregnancies in this sample are also likely to have led to the high caesarian section rate for the study sample [43].

The second most common risk factor for the total sample was family history of permanent childhood hearing loss, revealing a higher prevalence (19.6%) than previously reported for developing contexts. In a Nigerian sample of children from schools for the deaf, family history of permanent childhood hearing loss was indicated as an associated perinatal factor for 6.9% children [44].

Neonatal jaundice/hyperbilirubinemia was indicated as the most common postnatal risk factor (10.5%) for the sample population, with a further 2.3% of children requiring a subsequent blood transfusion. Two separate Nigerian studies indicated a prevalence of 4.9% in a sample of children diagnosed with SNHL [45] and 13.5% in a cohort of children with severe-profound hearing loss [44]. Hyperbilirubinemia has been associated as the most commonly reported risk factor for ANSD, varying between 30 to 70% [38,46-48]. The current study had 50% of ANSD cases presenting with hyperbilirubinemia as a risk factor similar to a recent report from South Africa in which half of the ANSD cases (n=5/10) also presented with hyperbilirubinemia [24].

Meningitis occurred in 10% of the total study sample and was recorded as the second most prevalent postnatal risk factor. Bacterial meningitis is the most common cause of acquired SNHL

in infants and children and accounts for about 6% of all cases of SNHL in the pediatric population [49,50]. Among developing countries the incidence of meningitis is the highest in Africa, mainly in sub-Saharan Africa, often referred to as the sub-Saharan meningitis belt [51]. Meningitis prevalence ranging from 7.8% to 22% was reported for children with permanent hearing loss from Nigeria [45,52]. In Angola, severe-profound permanent hearing loss was diagnosed in 30% of children with acute bacterial meningitis [51].

One in every ten children in the current study also presented with an associated syndrome, with Waardenburg syndrome constituting 52% of these cases. Within a sample of pediatric cochlear implant recipients, Young et al. [40] also reported 9.5% of children presenting with a syndrome. Waardenburg syndrome however, constituted only 10.8% of these cases. A previous South African etiological survey dating back three decades [28] reported 6.6% of children with confirmed severe-profound hearing loss presenting with a syndrome, with Waardenburg syndrome being represented by 43.8% of cases. For the present study the most common additional developmental conditions were indicated as visual impairment (8.5%) and cerebral palsy (5.6%). This corresponds to a UK epidemiological study [53] where visual impairment was reported in 5.4 to 7.5% and cerebral palsy in 2.5 to 3.6% of children with profound hearing loss. Additional developmental conditions were particularly common in cases of ANSD for whom more than half (58%) had at least one additional developmental condition. This increased incidence of additional disabilities is typically reported for children with ANSD [46].

Up to 25% of bilateral childhood hearing losses reportedly have a postnatal onset [54]. In the current study population 20.3% of children presented with a postnatal onset with 8.5% having a late onset progressive hearing loss and 11.8% having sudden hearing loss. ANSD was diagnosed in 5% of the cases which compares to previous reports where the prevalence of ANSD in

children ranged from 4.1 to 14% [40,55,56]. For developing countries such as India and Egypt, the reported prevalence ranges are 13.4 to 14% [57,58] and reports available for sub-Saharan Africa suggest an even higher prevalence. In Nigeria the reported prevalence ranges between 10.3 to 15.9% [32] and in South Africa an even higher prevalence of 21.4% was recently reported [24]. In the current cohort of children with PCEHL the prevalence of ANSD is lower than expected and may be attributed in part to the majority of the research sample (92.4%) being private health care patients. In South Africa, perinatal risk factors are more likely to occur in public health care patients, since adequate health care are known to be more accessible for private patients [59]. Therefore, children in this study were less exposed to environmental, maternal and child health related risk factors that predispose ANSD in populations from the developing world [32,33]. Also, since this cohort include children being diagnosed with profound PCEHL over the past 18 years, it is possible that the diagnosis of ANSD was not differentiated from SNHL in earlier years, given that the first report on auditory neuropathy dates back to 1996 [60] and accurate diagnosis of the disorder only followed in subsequent years. The first documented diagnosis of ANSD in the current study cohort dates back to 2007 which corresponds to the first ANSD diagnosis in a recently reported retrospective study from South Africa [24].

In the current study, PCEHL was typically diagnosed late (15.3 months, 9.3 SD) resulting in delayed initiation of intervention. On average, caregivers suspected hearing loss by 11.3 months of age, with a delay of 5.3 months until eventual diagnosis. In spite of the age of parental suspicion corresponding with a Nigerian sample [61], this study's results indicate ages of suspicion and diagnosis much earlier than recent South African reports [23-25]. Children with profound hearing losses are however known to be identified at earlier ages than children with

less severe degrees of hearing loss, since the symptoms of profound hearing loss are more apparent and may prompt parents to seek audiological evaluation sooner [34]. This could be a reason for earlier suspicion, diagnosis and intervention within this study sample, in contrast to former South African reports that included children with degrees of hearing loss ranging from mild to profound. In spite of earlier diagnosis, NHS was not done for 72.9% of children (n=62/85), reflecting the current EHDI status in South Africa where NHS services are offered in only a few hospitals in both the public and private health care sectors [19,20].

With recent South African reports on infant hearing loss mainly focusing on screening and diagnosis, limited information is available on ages of intervention [2]. In the current study, earlier ages of intervention in terms of initial hearing aid fitting and initiation of early intervention services was reported in comparison to a study sample from the Western Cape [23]. For children not benefitting from acoustic amplification, early access to sound through early cochlear implantation has been widely advocated [10,62,63]. Nonetheless, the average age of implantation for children with a congenital/early onset hearing loss in this study exceeded three and a half years of age, indicating a delay of more than two years from diagnosis to implantation. Cochlear implants are not provided by the South African National Department of Health and caregivers of children requiring cochlear implantation need to have adequate finances or access to funding from a private medical aid to be able to acquire this technology [64]. In 2010 the total average costs for a child for the first five and ten years post implantation was determined to be 298 961 ZAR and 455 225 ZAR respectively [64], with the current USD/ZAR conversion rate being 1 USD to 11.21 ZAR. Since implant systems are manufactured outside of South Africa and imported for use, these costs may fluctuate as a result of exchange rate changes [64]. Also, as a result of the weakening ZAR against the USD the past few years, implantable devices have

become more expensive. With the average monthly income level of South African citizens in 2010 being only 2 800 ZAR [65], it is clear that funding constraints as well as a lack of prompt referral to specialized cochlear implantation services are likely contributing factors to late implantation. Since age of implantation rather than age of diagnosis is considered as the primary predictor of language outcomes in implanted children [66], effort should be made to identify factors contributing to delayed cochlear implantation. The current study population, consisting of predominantly White (66.2%) Afrikaans speaking (50.8%) children from the private health care system (92.4%) is a representative sample of pediatric cochlear implant recipients in South Africa. This sample is not however, representative of the larger South African population reflecting the general disparities in health care access across ethnicities. Current population estimates indicate that 79.8% of the population is of African ethnicity with 74.9% speaking an African first language [21, 67]. Also, only 15% of the population are covered by private health care financing, while the majority (75%) of the population rely on public health care for health services [22]. The demographic distribution of the study sample highlights the persistent health care inequalities for advanced interventions such as cochlear implants in South Africa with previously disadvantaged people groups still marginalized. Despite the selective nature of this South Africa study sample (i.e. more privileged) critical periods for intervention, prior to 12 months of age, are not realized for children with profound hearing loss [26,31,68,69].

CONCLUSION

The most prevalent associated risks for profound PCEHL in South Africa included NICU admittance, family history of childhood hearing loss, prematurity, hyperbilirubinemia and meningitis. Profound hearing loss was typically sensorineural with a congenital onset and a 5% prevalence of ANSD. Diagnosis of PCEHL was delayed, resulting in deferred ages for initial

hearing aid fitting, enrollment in early intervention services, and eventual cochlear implantation. Even though average ages for intervention were earlier than previously reported in South Africa, necessary early auditory stimulation required for optimal outcomes for children with profound PCEHL, is not typically realized.

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