Profile of childhood hearing loss in the Western Cape, South Africa

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Abstract

Objectives: To describe the nature, associated risk factors and age of diagnosis for childhood hearing loss in a South African cohort from the Western Cape Province.

Methods: A retrospective review of clinical data from children under six years of age with confirmed hearing loss at Red Cross War Memorial Children's Hospital (RCWMCH) was conducted between 1 January 2019 and 31 July 2019. Data collected included demographic information, type and degree of hearing loss, documented risk factors associated with hearing loss, and age of suspicion and diagnosis of hearing loss.

Results: The study sample included 240 children with hearing loss, with a mean age of 42 months (21.8 SD; range 2-72). More than two thirds (68.3%) of the children presented with bilateral hearing loss. The majority presented with conductive hearing loss (64.6%), followed by sensorineural (28.7%) and mixed hearing loss (3.3%) or auditory neuropathy spectrum disorder (3.3%). More than half (51.8%) of the bilateral sensorineural hearing losses were of a profound degree. The most prominent risk factor for conductive hearing loss was otitis media, for sensorineural hearing loss it was a family history of childhood hearing loss, and for auditory neuropathy spectrum disorder it was hyperbilirubinaemia. Approximately one third of patients (27.1%) with sensorineural hearing loss did not have any associated risk factors. The mean age of diagnosis of permanent congenital or early-onset hearing loss was 31.4 months (22.8 SD; range 2-72), with a mean delay of nine months (13.2 SD; range 0-60) between age of suspicion and diagnosis of hearing loss (n=93).

Conclusions: The large proportion of preventable hearing losses in this sample highlights the importance of maximising primary health care efforts to treat preventable causes timeously. Age of diagnosis of permanent congenital or early-onset hearing loss was severely delayed undermining prospects of positive outcomes through early intervention. Infant hearing screening services in the public health sector of South Africa should be prioritised alongside primary health care efforts to reduce preventable risks for hearing loss.

Key words: childhood hearing loss; risk factors; age of diagnosis

1. Introduction

An estimated 466 million people globally suffer from disabling hearing loss of more than 30dBHL in the better hearing ear, which equates to nearly 6% of the world's population [1]. Of these, 34 million are children [1]. Hearing loss is the second most prevalent developmental disability, affecting approximately 15.5 million children under the age of 5 years world-wide [2]. Sub-Saharan Africa is one of the regions where the prevalence of disabling hearing loss in children under the age of 14 years is greatest [3]. The World Health Organisation (WHO) estimates the prevalence of hearing loss for children aged between 5 and 14 years at 1.9% in sub-Saharan Africa as opposed to 0.4% in high-income countries [3].

Most cases of disabling childhood hearing loss have preventable causes that are common in low-to-middle-income countries (LMICs), and make up 48.9% of the aetiology of hearing loss in children [4,5]. Children born into lower socioeconomic contexts have a higher incidence of middle ear pathology and subsequent preventable hearing loss, as well as considerably less access to non-emergency health resources [6]. Adverse pre-, peri- and post-natal conditions are prominent risk factors for childhood hearing loss, especially in LMICs [7]. Higher rates of low birth weight and severe hyperbilirubinaemia, which are associated risk factors for childhood hearing loss, have been reported in LMICs [8-10]. Additionally, vaccine-preventable infections like rubella and meningitis, which are associated with sensorineural hearing loss in children, occur more commonly in LMICs [1,8].

Possible reasons for the difference in prevalence of hearing loss in high-income countries and LMICs include the absence of well-managed hearing screening programmes, the impact of poverty and malnutrition on hearing, lack of awareness of hearing loss and its devastating effects in children, and limited access to hearing healthcare in LMICs [11]. Furthermore, the proportion of hearing loss attributed to post-natal causes such as infectious diseases and middle ear disease is typically higher in LMICs [7].

Due to the limited availability of hearing screening programmes, as well as poor data capturing and management within existing programmes [12], the nature and associated risk profile of childhood hearing loss in South Africa is largely unknown. Apart from studies from nearly four decades ago conducted in schools for the deaf [13,14], only preliminary data on the nature and associated risk profile of childhood hearing loss in South Africa are available [15,16]. This study aims to provide one of the first reports on the profile of childhood hearing loss in the Western Cape Province of South Africa, by describing the nature, associated risk factors and age of diagnosis for childhood hearing loss in a cohort from Red Cross War Memorial Children’s Hospital (RCWMCH).

2. Methods

The study was approved by the University of Pretoria Research Ethics Committee of the Faculty of Humanities (HUM024/0419), the University of Cape Town Human Research Ethics Committee (365/2019), RCWMCH Ethics Committee (RCC203) and the Western Cape Health Research sub-directorate (WC_201906_023).

2.1 Study population

RCWMCH is one of only two dedicated paediatric academic hospitals in sub-Saharan Africa. It provides specialist diagnostic audiology and intervention services to children from birth to 13 years from the public health care sector. Only children under the age of six were included in this study due to the paucity of information on the hearing profile in very young children who are not yet of school-going age in South Africa.

2.2 Procedures

An electronic database, updated daily with patient data in the Department of Audiology at RCWMCH, was used to conduct a retrospective review of clinical data from children under the age of six years who were diagnosed with confirmed hearing loss between January 2019 and July 2019. Some data that were not routinely included in the electronic database were captured from clinical records in patient hospital files.

Data collected included demographic information, type and degree of hearing loss, documented risk factors associated with hearing loss, and age of suspicion and diagnosis of hearing loss. Only children under six years of age with confirmed hearing loss were included in the sample. The audiological test battery typically included tympanometry, acoustic reflex-testing, oto-acoustic emissions (OAEs), and frequency-specific air- and bone-conduction auditory brainstem response (ABR) testing where indicated. Behavioural audiometry (air- and bone conduction pure tone testing) was used where age-appropriate, to determine the type and degree of hearing loss. Normal peripheral hearing was defined as air-conduction thresholds ≤15 dBHL [17]. Hearing loss was indicated when the pure tone average was >15dBHL across three frequencies (500, 1000 and 2000 Hz). The minimum diagnostic criteria for auditory neuropathy spectrum disorder (ANSD) was the presence of OAEs or a clear cochlear microphonic response at 85 dBnHL and 95 dBnHL with absent or abnormal ABR waves [18].

2.3 Statistical analysis

Data were captured on Microsoft Excel 2016 (Microsoft Corp, Redmond, WA), and analysed using SPSS 24 (Version 24.0.IBM Corp., Armonk, NY). Descriptive statistical methods were used.

3. Results

A total of 1154 paediatric patients under the age of six years were seen at RCWMCH Department of Audiology during the study period (January 2019-July 2019). Approximately one in five (20.8%) of these patients were diagnosed with hearing loss.

3.1 Demographics

The mean age of the 240 patients younger than six years of age who were diagnosed with hearing loss was 42 months (21.8 SD; range 2-72) with slightly more males (55.0%). The majority of patients were of coloured background (53.7%). Foreign patients from neighbouring sub-Saharan African countries constituted 11.7% of the sample. English was recorded as home language by the majority of persons (40.4%), followed by isiXhosa (27.5%) and Afrikaans (20.4%). Most referrals (53.8%) were received from Ear-Nose-Throat (ENT) specialists, followed by medical out-patients (22.9%), district referrals (10%), genetics (5.8%), Cerebral Palsy Clinic (2.9%) and others (3.5%).

3.2 Type and degree of hearing loss

The majority of patients presented with bilateral hearing loss (68.3%) (Table 1). Conductive hearing loss (CHL) was the most prevalent type of hearing loss (65.0%). The degree of CHL for the worse ear was predominantly mild [17] (64.7%). Approximately one third of CHLs were of a moderate [17] degree (31.4%). Bilateral permanent hearing losses (SNHL, ANSD and mixed) made up 27.9% of hearing losses. Bilateral hearing losses made up 22.5% of SNHL and 40.4% of CHL. SNHL (including mixed hearing loss) constituted 91.7%, and ANSD constituted 8.3% of permanent hearing losses. SNHL was predominantly profound in nature. Figure 1 presents a profile of the degree of bilateral SNHL for the worse ear (n=54) and unilateral SNHL (n=16).

Table 1. Profile of hearing losses type and laterality (n=240)

Types of hearing loss	Bilateral % (n)	Unilateral % (n)	Combined % (n)
CHL	40.4 (97)	24.6 (59)	65.0 (156)
SNHL	22.5 (54)	6.6 (16)	29.1 (70)
Mixed hearing loss	2.5 (6)	0.4 (1)	2.9 (7)
ANSD	2.9 (7)	-	2.9 (7)
Total	68.3 (164)	31.6 (76)	100 (240)

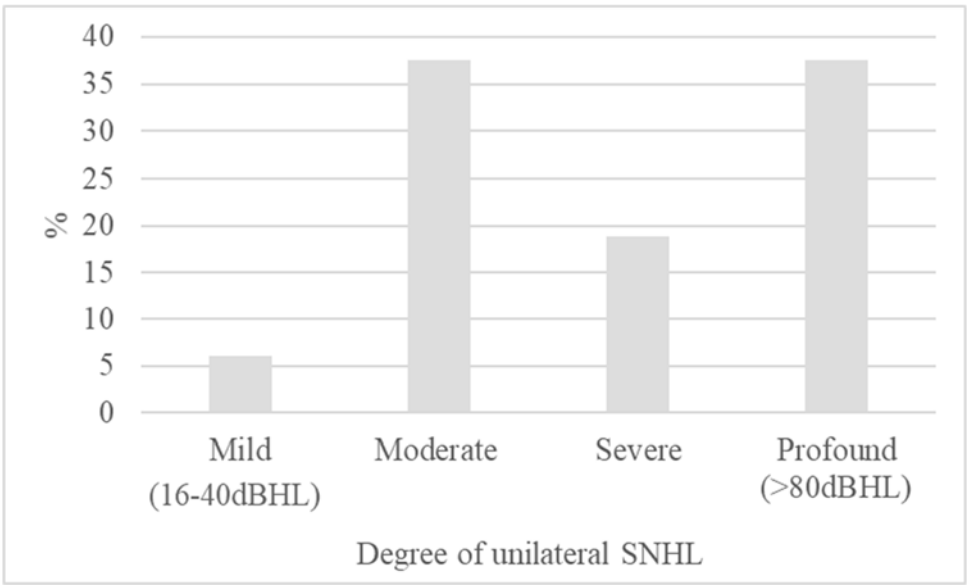

Figure 1: *Degrees of SNHL for pure tone average threshold across 500, 1000 and 2000 Hz (n=70)*

Table 2. Documented risk factors for childhood hearing loss (n=240)

	CHL (n=156) % (n)	SNHL (n=70) % (n)	ANSD (n=7) % (n)	Mixed HL (n=7) % (n)
Middle ear pathology	73.1 (114)	-	-	28.5 (2)
Tympanic membrane perforations	12.2 (19)	-	-	-
Syndromic	3.2 (5)	11.4 (8)	-	71.4 (5)
Hyperbilirubinaemia	1.9 (3)	7.1 (5)	85.7 (6)	-
Family history of childhood hearing loss	1.2 (2)	18.6 (13)	14.2 (1)	42.8 (3)
Microtia	5.7 (9)	-	-	28.5 (2)
Hypoxia	-	10 (7)	28.5 (2)	-
Cytomegalovirus	0.6 (1)	10.0 (7)	-	-
Bacterial meningitis	-	7.1 (5)	-	-
VLBW* < 1500g	-	1.4 (1)	57.1 (4)	-
Ototoxicity	-	4.2 (3)	-	-
TB** Mastoiditis	1.2 (2)	-	-	14.2 (1)
Rubella	-	2.8 (2)	-	-

**VLBW – Very low birthweight*
***TB - Tuberculosis*

3.3 Risk factors associated with hearing loss

The most prominent risk factor for conductive hearing loss was middle ear pathology (73.1%), for SNHL was a family history of childhood hearing loss (18.6%), and for ANSD it was hyperbilirubinaemia (85.7%). Middle ear pathology included otitis media (OM), acute otitis media (AOM), and chronic suppurative otitis media (CSOM). Syndromes included in this sample were Goldenhar, Trisomy 21, CHARGE, Pierre Robin and KID syndrome (Table

2).Approximately 70% of children with ANSD had two or more risk factors. Nearly one third (27.1%) of children with SNHL had no risk factors (Table 3).

Table 3. Number of risk factors for childhood hearing loss (n=240)

	1 Risk % (n)	2 Risks % (n)	3 Risks % (n)	None % (n)
CHL (n=156)	85.2 (133)	13.4 (21)	0.6 (1)	0.6 (1)
SNHL (n=70)	65.7 (46)	7.1 (5)	-	27.1 (19)
Mixed (n=7)	71.4 (5)	2.8 (2)	-	-
ANSD (n=7)	-	71.4 (5)	2.8 (2)	-

3.4 Age of hearing loss suspicion and diagnosis

Age of hearing loss suspicion and diagnosis were recorded for 93 patients with permanent congenital or early-onset hearing loss, and included SNHL (n=70), ANSD (n=7) and permanent congenital CHL secondary to structural and genetic aetiologies (n=13) (Table 4). Half of the participants in this sample (50%) were diagnosed with permanent hearing loss only after 36 months of age. Approximately only one third (29%) of children were diagnosed with permanent congenital or early-onset hearing loss before 12 months of age. On average, there is a delay of nine months (13.2 SD; range 0-60) between age of suspicion and age at diagnosis of hearing loss.

Table 4. Age of bilateral congenital or early-onset hearing loss suspicion and diagnosis (n=93)

	Age at suspicion (months)	Age at diagnosis (months)	Suspicion-to-diagnosis delay
Mean (SD)	22.4 (20.6)	31.4 (22.8)	9.1 (13.2)
Range	1-69	2-72	0-60

4. Discussion

Approximately two thirds of children diagnosed with hearing loss in this South African sample from the Western Cape Province presented with CHL. This is in line with recent reports from the WHO, which postulates that the leading causes of childhood hearing loss in LMICs are conductive and treatable [1]. The large number of children diagnosed with conductive hearing loss secondary to middle ear pathology could be attributed to the fact that the main referral source was from ENT specialists, and that RCWMCH is a tertiary referral facility with a combined ENT and Audiology service. It is evident that awareness and training for primary-level healthcare doctors and nurses in LMICs is important to provide effective first-line treatment for middle ear pathology such as AOM, OM and CSOM, so that hearing loss and subsequent adverse effects on hearing, speech – and language development can be minimised [4].

More than half (51.8%) of bilateral SNHL cases were of a profound degree. A previous South African study also indicated a profound degree of hearing loss in 50% of all SNHL cases [15].

Estimates from high-income countries suggest that profound hearing loss make up 20-30% of permanent childhood hearing loss [19]. The higher incidence of profound SNHL in this sample could be attributed to the fact that children with profound hearing losses tend to be identified sooner than children with less severe hearing losses, since the signs of profound hearing loss are more readily identified and may prompt parents to seek audiological assessment earlier [20]. Milder and even moderate losses, especially in the absence of newborn hearing screening programmes, may remain undetected until school failures or other behavioural patterns arise in school [20]. The profound nature in nearly half of SNHL cases implies that these children will not necessarily benefit optimally from hearing aids, and highlights the importance of early diagnosis of hearing loss, in order to refer timeously for cochlear implant assessment. Early auditory stimulation is essential for optimal speech- and language outcomes in children with severe-profound hearing loss [21]. Within the South African context, limited funding for cochlear implants within the public sector is available [22]. The financial implications associated with cochlear implantation has been identified by parents as the most prominent challenge regarding the paediatric cochlear implantation process in South Africa [22]. The high proportion of profound SNHL indicates that these children are audiological candidates for cochlear implantation. However, children in South Africa do not have equal access to cochlear implants, especially in the public healthcare sector, and therefore cochlear implantation is considered as a privileged intervention [16,22].

ANSD as a proportion of all SNHL (including mixed hearing losses) constituted 8.3%, which is in line with previous reports of 5-17% [16,23]. All of the children with ANSD diagnosis in this sample had hyperbilirubinaemia, which required phototherapy and blood transfusion. Hyperbilirubinaemia is more prevalent in African countries due to a higher incidence of G6PDD and limited treatment facilities [24,25]. More than half of the children diagnosed with ANSD in this sample were also born prematurely (<34 weeks gestation) with a very low birth weight of <1500g. Higher rates of very low birth weight have been reported in LMICs [8-10]. A previous South African study on the risk profile of children with profound hearing loss also included prematurity (<34 weeks gestation) as a risk factor in 15.1% of all cases, and 40% of ANSD cases [16].

More than two thirds (65.7%) of children with SNHL presented with at least one risk factor for hearing loss. The most prominent risk factor for SNHL was a family history of childhood hearing loss present in 18.6% of cases. This finding is in line with two previous South African studies on the risk profiles of children with SNHL [15,16]. A multi-centre study across cochlear implant programmes in South Africa reported on family history of permanent childhood hearing loss as a risk factor for SNHL (19.6%) [16]. A study conducted at a paediatric referral centre in Pretoria reported on any family history of childhood hearing loss as a risk factor for SNHL (27%) [15]. The higher incidence of family history reported in the Pretoria-study could have been due to the fact that parents were able to report on any family history of childhood hearing loss, including transient episodes of childhood hearing loss due to middle ear pathology [15]. The high incidence of syndromic risks (11.4%) for children with SNHL in this study could be attributed to the specialised tertiary institution where the data in the current study was collected. Nearly one third (27.1%) of patients with SNHL did not present with any risk factors for hearing loss,

highlighting the need for universal newborn hearing screening, and not only targeted high-risk screening in South Africa.

The average age of hearing loss diagnosis for children with permanent congenital or early-onset hearing loss (including SNHL, ANSD and permanent congenital CHL) was 31.4 months (n=93), surpassing two and a half years of age. This finding highlights the consequences of the lack of newborn hearing screening programmes and appropriate follow-up in the public sector of South Africa [12,26]. Delayed diagnosis of hearing loss results in delayed initiation of intervention, and predisposes this population to poorer speech- language and academic outcomes [16]. RCWMCH is a referral facility for many foreign patients from sub-Saharan Africa, including Zimbabwe, the Democratic Republic of the Congo and Malawi. Twenty-eight children in this sample were foreign patients (11.7%), and more than two thirds (64.8%) of them were diagnosed with severe-profound hearing loss for the first time at RCWMCH at ages well beyond recommended guidelines due to a lack of audiology services in their native countries.

The mean delay of nine months between age of suspicion and diagnosis of hearing loss in this study is less than the 22 month mean delay reported in a previous South African study [15]. The shorter time between suspicion and diagnosis of hearing loss in this study could be due to the high incidence of a family history of childhood hearing loss as a risk factor for SNHL in this sample, or due to many children (12.1%) in the sample having complex co-morbidities, resulting in more timeous referral to audiology at a tertiary institution.

5. Conclusions

The nature of childhood hearing loss at the RCWMCH tertiary health care facility was predominantly bilateral and conductive. The burden of preventable hearing loss in this sample was high, supporting the case for primary level healthcare facilities to treat preventable causes of hearing loss timeously. Age of diagnosis for permanent congenital or early-onset hearing loss was significantly delayed beyond recommended ages for optimal early intervention outcomes. Universal newborn hearing screening services in the public health sector of South Africa should be prioritised along with identification and early treatment of preventable risks for hearing loss.

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