International Journal of Pediatric Otorhinolaryngology 79 (2015) 8–14

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: www.elsevier.com/locate/ijporl

Profound childhood hearing loss in a South Africa cohort: Risk profile, diagnosis and age of intervention

Talita le Roux [a], De Wet Swanepoel [a,b,c,*], Anel Louw [a], Bart Vinck [a,d], Mashudu Tshifularo [e]

[a] Department of Speech-Language Pathology and Audiology, University of Pretoria, South Africa
[b] Ear Sciences Centre, School of Surgery, University of Western Australia, Nedlands, Australia
[c] Ear Science Institute Australia, Subiaco, Australia
[d] Speech-Language Audiology Department, Ghent University, Belgium
[e] Department of Otorhinolaryngology, University of Pretoria, South Africa

ARTICLE INFO

Article history:
Received 30 May 2014
Received in revised form 26 September 2014
Accepted 30 September 2014
Available online 28 October 2014

Keywords:
Profound hearing loss
Risk factors
Children
Intervention
Hyperbilirubinemia
Auditory neuropathy

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 5 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.

1. 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 [5–8]. 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 [3,9–11].

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

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.

* Corresponding author at: Department of Speech-Language Pathology and Audiology, University of Pretoria, c/o Lynnwood and University Road, Hatfield 0002, South Africa.
E-mail address: dewet.swanepoel@up.ac.za (D.W. Swanepoel).

http://dx.doi.org/10.1016/j.ijporl.2014.09.033
0165-5876/

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 and 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.

2. Method

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

2.1. 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 5 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.

2.2. 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 5 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 capturers were identified and trained for each participating cochlear implant program. An electronic database was developed to