

# Age of diagnosis for congenital hearing loss at Universitas Hospital, Bloemfontein

I R T Butler,<sup>1</sup> MB ChB, MMed (ORL); S Basson,<sup>2</sup> 2nd year MB ChB student; E Britz,<sup>2</sup> 2nd year MB ChB student; R de Wet,<sup>2</sup> 2nd year MB ChB student; G B Korsten,<sup>2</sup> 2nd year MB ChB student; G Joubert,<sup>3</sup> BA, MSc

<sup>1</sup> Department of Otorhinolaryngology, Faculty of Health Sciences, University of the Free State, Bloemfontein

<sup>2</sup> Faculty of Health Sciences, University of the Free State

<sup>3</sup> Department of Biostatistics, University of the Free State

Corresponding author: I R T Butler (butlerirt@ufs.ac.za)

**Background.** Congenital hearing loss affects 3 - 6/1 000 children worldwide. The benefits of early identification of hearing loss and early intervention have been clearly established. There are no previous studies reporting on the age of diagnosis of congenital hearing loss in the Free State province.

**Objectives.** To determine the age of diagnosis of congenital hearing loss in the Otorhinolaryngology Clinic at Universitas Hospital. Secondary aims included determining age at first visit, as well as the time delay between first visit and diagnosis, and documenting any interventions which took place.

**Methods.** A retrospective, descriptive study was undertaken, analysing data from 2001 to 2010.

**Results.** A total of 260 cases of congenital hearing loss were analysed. The median age of diagnosis of hearing loss was 44.5 months. The median age of first visit was 40.9 months, and the median delay between first visit and diagnosis was 49 days.

**Conclusions.** The median age of diagnosis far exceeds national and international benchmarks. This has a profoundly negative impact on the development and outcomes of children with hearing loss. These results have been used to motivate for the expansion of hearing screening and diagnostic services in the province.

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Congenital hearing loss is reported to affect 3 - 6/1 000 live births worldwide.<sup>[1]</sup> According to Statistics South Africa, approximately 900 000 children are born in South Africa annually, including 50 000 in the Free State province.<sup>[2,3]</sup> Therefore an estimated 150 - 350

children are born with permanent sensorineural hearing loss annually in the Free State, out of an estimated 6 000 in the whole of South Africa;<sup>[1]</sup> 90% of these children have no access to newborn hearing screening.<sup>[4]</sup>

It is well established that a lack of infant and early childhood auditory stimulation will result in a permanent functional communication handicap, with associated learning difficulties, impaired cognitive development, and emotional/psychological issues.<sup>[5,6]</sup> Furthermore, there is a proven negative impact on future vocational and socio-economic outcomes.<sup>[5,6]</sup> Conversely, infants who are identified with hearing loss before 6 months of age and receive appropriate intervention before the age of 1 year can develop communication skills on par with those of their peers with normal hearing.<sup>[1,7-9]</sup>

Universal newborn hearing screening (UNHS) is now a well-established norm in most developed countries, and has been endorsed by the Health Professions Council of South Africa (HPCSA) Professional Board for Speech, Language and Hearing Professions.<sup>[6]</sup> International benchmarks have been modified for local conditions and call for diagnostic testing to be performed by 4 months of age, with implementation of an appropriate intervention strategy by 8 months of age.<sup>[6]</sup>

In the Free State, targeted hearing screening services are available at 2 hospitals and 1 clinic in the public health service, while UNHS is available at 3 private hospitals.

A study conducted at a school for hearing-impaired children in the Western Cape found a mean age of diagnosis of 23 months, while an unpublished study from Gauteng found a mean age of diagnosis of 31 months.<sup>[1,10]</sup> To our knowledge, there are no data available on

the age of diagnosis of congenital hearing loss in the Free State. This study was undertaken to validate our perception that the current age of diagnosis of congenital hearing loss in our hospital was unacceptably high and to provide a benchmark against which improvements in healthcare delivery could be measured.

## Objectives

The primary aim of the study was to determine the age of diagnosis of congenital hearing loss in children seen in the Otorhinolaryngology Clinic at Universitas Hospital (a tertiary level public healthcare facility). Our secondary aims were to determine the age of first visit, as well as the time delay between first visit to our clinic and diagnosis of hearing loss, and to document any subsequent interventions.

## Methods

A retrospective, descriptive study was undertaken, which analysed clinical records of patients who underwent hearing evaluations by the Department of Audiology and Speech Therapy in the Otorhinolaryngology Clinic at Universitas Hospital. Our inclusion criteria were: diagnosis of hearing loss between 1 January 2001 and 31 December 2010; diagnosis before the age of 6 years; and a pure tone average (500 Hz, 1 KHz, 2 KHz, 4 KHz) worse than 30 dB in the better-hearing ear. We excluded any case with an acquired cause of hearing loss. The study was approved by the Ethics Committee of the Faculty of Health Sciences at the University of the Free State; permission to perform the study was obtained from the Clinical Head of Universitas Hospital.

## Results

A total of 2 383 patient records were analysed. Of these, 1 001 demonstrated a hearing loss (congenital or acquired) but only 260

fulfilled all the inclusion criteria. The median age of diagnosis of hearing loss for this group of 260 children was 44.5 months or 3.7 years (1 month - 5.9 years). The median age of first visit to our clinic was 40.9 months or 3.4 years (1 month - 5.9 years). The median delay between first visit to our clinic and diagnosis was 49 days. Data on management of the deafness were available in 169 cases: 26% ( $n=46$ ) of the children were referred directly to schools for the deaf (sign language medium), 50% ( $n=85$ ) were fitted with hearing aids or cochlear implants and enrolled in speech and language therapy (SLT) programmes, and 23% ( $n=38$ ) were referred back to district level to be fitted with hearing aids and enrolled in SLT programmes.

In analysing the age of diagnosis the 10-year period was divided in 2, and this revealed no difference in the age of diagnosis over time. We found that the median age of children referred to sign language-medium schools (54.8 months) was significantly older ( $p<0.0001$ ) than those fitted with hearing aids/cochlear implants and enrolled in SLT programmes (38.5 months).

## Discussion

Congenital hearing loss has long been seen as one of the most common birth defects.<sup>[6,9]</sup> Before the availability of otoacoustic emission (OAE) testing, hearing screening was targeted at those children with known risk factors. It has been shown that such targeted screening misses 50% of infants affected by congenital hearing loss.<sup>[6]</sup> The introduction of UNHS programmes in developed countries has dramatically reduced the age of diagnosis of congenital hearing loss and allowed the implementation of early intervention strategies during the critical first year of life.<sup>[1,5-12]</sup> The profound benefits to the child with congenital hearing loss, as well as the economic benefit to society in general, have been well established and underpin the motivation for all countries to provide universal newborn hearing screening services.<sup>[1,4-13]</sup> Studies have shown that the earlier the intervention, the better the outcomes, especially if the interventions begin before the age of 1 year.<sup>[9,12]</sup>

The HPCSA's Professional Board for Speech, Language and Hearing Professions has affirmed the need for such services in SA.<sup>[6]</sup> A number of adaptations to the international guidelines have been proposed in the light of challenges facing the country's public healthcare sector,<sup>[6]</sup> the most important of which is the change in screening context from birthing unit to primary healthcare clinic. Also included is the coupling of hearing screening to the immunisation schedule. The age at which diagnostic testing should be completed has been shifted from 3 to 4 months, while the age by which an appropriate intervention should be initiated has been shifted from 6 to 8 months. Despite these more lenient benchmarks, our age of diagnosis of 44 months (3.7 years) far exceeds the local benchmark of 4 months of age. It also exceeds by 13 months and 21 months respectively the findings of the two previous SA studies, which were smaller (with a combined total of 74 children) and studied children already enrolled in auditory-oral rehabilitation programmes.

Our data reveal a median 49-day delay between first presentation at our clinic and diagnosis of congenital hearing loss. This appears to be commensurate with international guidelines, but may be misleading as a large number of our children were diagnosed by subjective testing owing to their relatively older age. Subjective testing relies on observation of conditioned responses to sound stimuli; the older the child, the easier it becomes to condition them to respond. Reliable results may be obtained from approximately 6 months of age, but are generally complementary to objective test results in children under 2 years of age.

Although these subjective tests are time-consuming, they are less resource-intensive in terms of time, personnel, equipment, medication and monitoring costs than auditory brainstem response/auditory steady state response (ABR/ASSR) testing. The latter is an

objective form of testing, using surface recordings of auditory evoked potentials, and can be performed from birth. Universitas Hospital, as a tertiary referral hospital, is the only state facility in the province with the equipment to perform ABR/ASSR testing. The younger the child referred, the greater the reliance on ABR/ASSR testing for diagnosis, and the greater the strain on this special resource. This is reflected in our current waiting period for ABR/ASSR testing, which over the past year has ballooned from 3 months to 9 months. While a small percentage of parents chose sign language as a primary mode of communication for their children, our results suggest that those children referred to sign language-medium schools were past the age where auditory-oral communication was deemed possible.

It is clear that diagnostic services need to be expanded in the province to cope with current and future demand. The data generated by this study have been used to motivate for the procurement of ABR/ASSR diagnostic equipment by the 5 regional hospitals in the Free State, as well as for the establishment of a UNHS programme in the province's public sector, according to the HPCSA guidelines.

Our study has a number of shortcomings. We did not access information from other audiology services in the province's public sector, which meant we had no data on the age of diagnosis or on interventions provided at these centres. As none of these centres has ABR/ASSR equipment, we can only assume that children would be diagnosed by subjective means, which would usually take place at an older age. We feel that including data on these children would increase the age of diagnosis. We also did not collect data on the interval between diagnosis and the initiation of the interventions.

## Conclusion

The median age of diagnosis of hearing loss at the Otorhinolaryngology Clinic at Universitas Hospital was 44.5 months (3.7 years). This is far older than the benchmark suggested by the HPCSA, of 4 months of age. This late age of diagnosis has a profoundly negative impact on these children's communication, cognitive, educational and vocational outcomes. These data have been used to motivate for the expansion of hearing screening and diagnostic services in the province. The results will also be used as a baseline against which any improvements in service delivery can be measured.

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