# AUDITORY BRAINSTEM RESPONSE FINDINGS IN A GROUP OF NEUROLOGICALLY COMPROMISED CHILDREN: A RETROSPECTIVE STUDY.

Karen Mary Baillieu

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A Research Report submitted to the Faculty of Health Sciences, University of the Witwatersrand, Johannesburg, in partial fulfilment of the requirements for the degree of Master of Science in Medicine in Child Health and Neurodevelopment.

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# DECLARATION

I, Karen Mary Baillieu, declare that this Research Report is my own, unaided work. It is being submitted for the Degree of Master of Science in Medicine at the University of the Witwatersrand, Johannesburg. It has not been submitted before for any degree or examination at any other University.

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Karen Baillieu

_____day of _____20_____in_____

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## List of Abbreviations

| | |
|---|---|
| AABR | Automated Auditory Brainstem Response |
| ABR | Auditory Brainstem Response |
| AIDS | Acquired Immunodeficiency Syndrome |
| ASD | Autism Spectrum Disorder |
| ASHA | American Speech and Hearing Association |
| CHBAH | Chris Hani Baragwanath Academic Hospital |
| CP | Cerebral Palsy |
| CPA | Conditioned Play Audiometry |
| CSOM | Chronic Suppurative Otitis Media |
| dB | Decibel |
| dBeHL | Decibel Estimated Hearing Level |
| dBHL | Decibel Hearing Level |
| dBnHL | Decibel Normal Hearing Level |
| HIE | Hypoxic Ischemic Encephalopathy |
| HIV | Human Immunodeficiency Virus |
| HIV/AIDS | Human Immunodeficiency Virus/ Acquired Immunodeficiency Syndrome |
| HPCSA | Health Professions Council of South Africa |
| Hz | Hertz |
| kHz | Kilohertz |
| ms | Milliseconds |
| NICU | Neonatal Intensive Care Unit |
| OAE | Otoacoustic Emmission |
| Ω | Ohm |
| RVD | Retroviral Disease |
| SNHL | Sensorineural Hearing Loss |
| TBM | Tuberculosis Meningitis |
| UNAIDS | Joint United Nations Programme on HIV/AIDS |
| VRA | Visual Reinforcement Audiometry |
| WHO | World Health Organization |
| WMA | World Medical Association |

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## Abstract

**Objective:** There is a higher prevalence of hearing loss in children with diagnosed neurological disorders than the general paediatric population. It is therefore essential that these children have their hearing assessed. Conventional behavioural audiometry requires participation from the child, and in a majority of this population with neurological pathology this is not always possible owing to their neurocompromised state. These children will have to undergo objective testing, such as the Auditory Brainstem Response (ABR) in order to obtain estimated hearing thresholds, as this requires no active involvement from the patient. This study therefore aims to describe the audiological ABR findings in order to determine hearing function in this group and to establish a relationship between audiological ABR findings to behavioural audiometry findings where these exist in a group of neurologically disordered children in a tertiary hospital in South Africa.

**Methods:** A retrospective review was conducted on 40 ABR patient records of children between the ages of 5 months and 10 years diagnosed with a neurological disorder. Behavioural audiometry results were then sought for these children, where these existed. Hearing status was described for each child per ear for both objective and behavioural results, and descriptive statistics were conducted.

**Results:** 56.25 % (n=45) of ears in this study presented with normal hearing on ABR testing. No behavioural audiometry results were obtained in 72.5 % (n=58) of ears in this study. Results correlated between ABR and behavioural testing for only 7.5% (n=8) of ears tested and in all eight of these ears the hearing result was within normal hearing limits. Twelve and a half percent (n=10) of ears were misdiagnosed on behavioural testing. More premature infants were able to be tested behaviourally when compared to other pathologies. Cerebral palsy, Down's Syndrome, prematurity and RVD were the pathologies in which the most hearing losses were diagnosed.

**Conclusions:** Behavioural audiometry appears a largely unreliable method of hearing testing in children diagnosed with neurological disorders as results were obtained in only 27.5 % of the study sample; however it remains the gold standard in paediatric hearing testing in order to evaluate the entire auditory system and provides information on how a child processes sound, unlike ABR testing which only provides hearing information up to the auditory brainstem. This study highlights the high prevalence of hearing problems in children with neurological disorders and therefore the importance of hearing testing in this population. Hearing thresholds should be established for subsequent remediation via objective testing. Conditioning should continue simultaneously for a behavioural audiological test battery with adaptations for the child's developmental ability.

**Key Words**: *Neurological disorder; paediatric; hearing loss; ABR; behavioural audiometry;*

*South Africa*

# 1. Introduction

## Literature Review

### Early Detection of Hearing Loss

Hearing loss is a common sensory disability in children, with reports indicating this to be the most common sensory disability in the United States of America (Topol, Girard, St. Pierre, Tucker & Vohr, 2011). One to three per 1000 newborns are diagnosed with a congenital hearing loss in the United States (Topol *et al*., 2011), and one per 840 live births in the United Kingdom (Proops & Acharya, 2009). A reported 0.76 per 1000 children are diagnosed with a delayed-onset hearing loss between the age of 3 and 6 years in China (Lu *et al.*, 2011). In a study conducted in the United Kingdom, congenital hearing loss was diagnosed in 1.06 per 1000 births, and this increased to 1.65 per 1000 children by the age of 9 years (Fortnum, Summerfield, Marshall, Davis & Bamford, 2001).

Hearing loss has been reported to be more common in developing countries. Of the 800 000 children born with or acquiring a bilateral permanent hearing loss of 40 dB or worse each year, 90 % reside in developing countries such as South Africa (Friderichs, Swanepoel & Hall, 2012). A previous study found a prevalence rate of three per 1000 for the private healthcare setting and six per 1000 in the public health setting in South Africa (Swanepoel, Storbeck & Friedland, 2009).

Many studies have shown the benefits of identifying a hearing loss as early as possible (Topol *et al.*, 2011; Swanepoel *et al*., 2012; Swanepoel *et al*., 2009; Meyer, Swanepoel, le Roux & van der Linde, 2012; Fortnum *et al*., 2001). Late detection of hearing loss has adverse effects on the child's linguistic, "psychosocial, emotional and cognitive development" (Meyer *et al.*, 2012, p. 699). Late detection of hearing loss also decreases the child's quality of life (Fortnum *et al*., 2001).

Conversely, the earlier the detection of the hearing loss and the intervention is provided, the better the child's expressive speech and language will be (Friedrich *et al*., 2012). Early detection and intervention will decrease the burden of permanent hearing loss, and result in better educational opportunities (Meyer *et al*., 2012).

The aim of screening programmes is the early identification of hearing loss in order to improve long-term outcomes (Rai & Thakur, 2013). Cost-effectiveness studies of screening programmes identifying congenital hearing loss are in their infancy (Kemper & Downs, 2012). Preliminary data in the United States of America have shown that hearing screening programmes have proven cost-effective and that these programmes are beneficial (Kemper & Downs, 2012).

In South Africa, The Health Professions Council of South Africa (HPCSA) identified populations at risk of developing a hearing loss (Swanepoel, 2007). These children should, according to the HPCSA (Swanepoel, 2007, p. 28), have their hearing screened at birth if they present with any of these identifying factors:

- An illness or condition requiring admission of 48 hours or greater to a Neonatal Intensive Care Unit (NICU)
- Stigmata or other findings associated with a syndrome known to include a sensorineural and/or conductive hearing loss
- Family history of permanent childhood sensorineural hearing loss
- Craniofacial abnormalities, including those with morphological abnormalities of the pinna and ear canal
- In-utero infection such as cytomegalovirus, herpes, toxoplasmosis, rubella, HIV, or malaria

In addition, children with postnatal infections associated with sensorineural hearing loss including bacterial meningitis should be screened after a diagnosis of the infection (Swanepoel, 2007).

In South Africa, 85% of the population access public health care, which offers medical care either free of charge or at a reduced rate (Swanepoel *et al*., 2009). According to a recent survey-based study, only 7.5% of public hospitals in South Africa offer infant hearing screening, and only one hospital offered universal screening (Swanepoel *et al*., 2009). It can therefore be said that 90% of babies born in the public health setting in South Africa are not afforded the opportunity of newborn hearing screening (Swanepoel *et al*., 2009).

## Early Intervention for Neurologically Disordered Populations

A large number of children in developing countries present with neurological disorders, which cause high rates of mortality and long-term morbidity (Newton & Neville, 2009). The World Health Organization (WHO) Guidelines, as cited in the *Paediatric Neurology and Child Development Services in South Africa* Report (Department of Health, 2003), states that of the estimated 18 million children in South Africa, 10 % (or 1.8 million) will have a significant neurological disorder. This is in keeping with the global Burden of Disease, which has found that "mental and neurological disorders account for 9.8 % of the total burden of disease in low- and middle-income countries" (Patel, Goel & Desai, 2009, p. 37).

Neurological disorders in developing countries face twin difficulties. These pathologies are not only diagnosed less in these countries, a study in Zambia found that patients with these diseases receive less treatment for their condition, largely as a result of the lack of neurological expertise in these countries (Siddiqi, Atadzhanov, Birbeck & Koralnik, 2010). According to Siddiqi *et al*. (2010), the ratio of neurologists to the general population in African nations is 1:3.4 million. Eleven countries in the African continent report no neurologist at all (Siddiqi *et al*., 2010). The ratio of neurologists to the population is 1:26 000 in the United States of America (Siddiqi *et al*., 2010).

Many different medical conditions fall within the category of "neurodisability." Certain of these conditions are known to place affected individuals at higher risk of hearing impairment than the general population, for example Down's Syndrome (Abou-Elhamd, ElToukhy & Al-Wadaani, 2013). Other neurological disorders may result in a speech and language delay. In order for speech therapy to begin for the remediation of this delay, the child's hearing status should first be established (Spivak, 2007). Even a mild hearing loss will affect speech and language development, and any hearing loss should be amplified to the best aided condition before speech therapy begins (Spivak, 2007).

It has been established above that the earlier a hearing loss is detected and the faster appropriate intervention is provided therefore, the fewer adverse effects the hearing loss will have on the child. Indeed early intervention in hearing loss can improve the quality of life for the hearing-impaired child (Islami *et al.,* 2013). It is therefore vital that the hearing status of these children with neurological disorders be established, with as little delay as possible. These children, however, owing to their neurological disorders, struggle with conventional hearing testing and thus require objective testing such as the Auditory Brainstem Response (ABR) test (Swanepoel & Ebrahim, 2009). Objective testing in audiology is defined as those tests which supplement information obtained in the case history and behavioural tests, and make it possible to assess those patients unable to respond appropriately in behavioural testing (Gelfand, 2009).

## The Burden of Disease

The term "neurological disease" comprises many heterogeneous conditions with different causes (Valente, Ferraris & Dallapiccola, 2008). Hearing loss is associated with certain of these conditions (Northern & Downs, 2002). It is for this reason, and the fact that the disorders chosen place a large

burden on the healthcare system in South Africa that the following neurological disorders were identified for the focus of this study.

South Africa has the largest paediatric epidemic of Human Immunodeficiency Virus/ Acquired Immunodeficiency Syndrome (HIV/AIDS), or Retroviral Disease (RVD) in the world (Fatti, Bock, Grimwood & Eley, 2010). Of the 33 million HIV-infected individuals worldwide, 35 % reside in Southern Africa (Shisana *et al.,* 2009). In 2007, 2.7 million people became infected with HIV worldwide, 1.9 million of which reside in Southern Africa (Shisana *et al*., 2009). The Joint United Nations Programme on HIV/AIDS (UNAIDS) has defined the HIV/AIDS epidemic in South Africa as a "hyper-endemic epidemic" owing to the fact that 15 % of the population aged between 15 and 49 years were infected with the virus (Shisana *et al*., 2009).

According to Statistics South Africa, an estimated 5.26 million people are living with HIV/AIDS in South Africa in 2013 (Statistics South Africa, 2013). In May 2013 it was estimated that 10 383 746 people residing in South Africa are under 9 years of age (Statistics South Africa, 2013). An HIV Survey conducted in 2008 found that 2.5 % of children in South Africa aged 2 to 14 years were HIV-infected (Shisana *et al*., 2009). Therefore, applying this statistic to the population estimate of 2013, it can be estimated that there are 259 594 children under the age of 9 years in South Africa living with HIV/AIDS.

According to the guidelines in *Paediatric Neurology and Child Development Services in South Africa* (Department of Health*,* 2003), conditions such as HIV/AIDS and Tuberculosis place an increased burden of disease in the South African public health care system. All children with HIV/AIDS have neurodisability (Statistics South Africa, 2003). Therefore it can be surmised that approximately 260 000 children in South Africa present with a significant neurodisability resulting from HIV/AIDS.

Cerebral Palsy (CP) is a disorder arising from a permanent, non-progressive insult to the brain that results in disturbances in "sensation, perception, cognition, communication and behaviour" (Eunson,

2012, p. 361). This group of neurological deficits is the most common cause of motor deficits in childhood (Campbell, Hoon & Johnston, 2008) and affects an average of two to two and a half per 1000 live births worldwide (Eunson, 2012). However, the incidence was found to be higher in South Africa, where 10 in 1000 children had cerebral palsy in rural KwaZulu-Natal (Couper, 2002). The largest risk factor in causing CP is prematurity. Among preterm infants born before 28 weeks gestation the incidence of CP is 100 per 1000 live births, however this decreases to one per 1000 live births in children born full term (O'Shea, 2008).

Autism Spectrum Disorder (ASD) is thought to be a set of neurological disorders resulting in deficits of socialization, communication and the presence of rituals and stereotypies (Matson, Beighley & Turygin, 2012). Autism is present in some cases as early as birth and infancy (Arieff, Kaur, Gameeldien & van der Merwe, 2010). These children will then be unable to form normal social relationships, as well as be unable to communicate effectively (Arieff *et al*., 2010).

Globally, ASD prevalence has been reported as 11.3 per 1000, or one in 88 children (Autism and Developmental Disabilities Monitoring Network Principle Investigators, 2012). This study in the United States of America found that the incidence of ASD is higher in the white population (12.0 per 1000) and more common in males (18.4 per 1000) than in females (four per 1000) (Autism and Developmental Disabilities Monitoring Network Principle Investigators, 2012).

It is predicted by Autism South Africa that one in 150 children born in South Africa are affected with Autism, leading to a potential 270 000 children in South Africa living with the disease (Arieff *et al*., 2010). This discrepancy in the two values between the two countries could be due in part to cultural differences between South Africa and America leading to differences in the number of children diagnosed with ASD (Grinker *et al.,* 2012).

Down's Syndrome is the most common chromosomal abnormality, occurring in one in 800 births globally (Moore, 2008). This syndrome, also known as Trisomy 21, is caused in a vast majority of

cases by each cell in the body containing an extra 21st chromosome (Pitetti, Baynard & Agiovlasitis, 2013). Children with this syndrome have abnormal brain development and function, presenting with intellectual impairment (Moore, 2008).

The prevalence of Down's Syndrome has increased from nine to 11.8 per 10 000 births in the United States of America (Pitetti *et al*., 2013). This can in part be attributed to the increase in the number of mothers bearing children after the age of 35 years (Petetti *et al*., 2013), as more than half of the babies born with Down's Syndrome are born to mothers above this age (Lampret & Christianson 2007). The overall prevalence of Down's Syndrome increases with maternal age, from one per 1000 births in mothers under the age of 25 to 10 per 1000 births in mothers of 40 years and 70 per 1000 births in mothers aged 48 years (Lampret & Christianson, 2007).

In South Africa the prevalence of Down's Syndrome is slightly higher in rural populations (2.01 per 1000 live births) than in urban populations (1.8 per 1000 live births) (Lampret & Christianson, 2007). This syndrome is therefore as prevalent in South Africa as it is globally, owing to the cause being of a genetic nature.

15 million children are born prematurely (as classified as those children born before 37 weeks gestation) across the world each year (Nour, 2012). Sixty percent of these premature births occur in Sub-Saharan Africa and South Asia (Nour, 2012). According to Nour (2012), 12.3 % of all births in Sub-Saharan Africa are premature. These children are then at risk of developing visual impairment and neurodevelopmental effects ranging from learning difficulties to severe cognitive impairments to cerebral palsy (Nour, 2012).

Birth asphyxia, or the impairment of the exchange of oxygen and carbon dioxide (Kurinczuk, White-Koning & Badawi, 2010), results in approximately 10 % of infants born in America requiring some assistance to breathe at birth, 1 % of which require extensive resuscitation (Kattwinkel *et al.,* 2010). Of these, three per 1000 survivors in South West London and Western Australia were diagnosed

with neonatal encephalopathy, which is the clinical manifestation of disordered brain function in neonates (Kurinczuk, *et al.*, 2010). A further one and a half per 1000 infants have hypoxic-ischaemic encephalopathy (HIE), or a lack of oxygen in the blood supply, as stated in three studies conducted in the United Kingdom, Western Australia and Sweden (Kurinczuk *et al*., 2010).

In South African Public Sector Hospitals 35.3 % of neonatal deaths were attributed to hypoxia, and was the most common cause of neonatal mortality (Pattinson, Woods, Greenfield & Velaphi, 2005). This is higher than the global average of 23 %, which was the third most common cause of neonatal death after infections and preterm birth (Buchmann & Velaphi, 2009). Owing to the fact that asphyxia leads to neurological disorders, these infants can develop severe consequences such as cerebral palsy, epilepsy and other "significant cognitive, developmental and behavioural problems" (Kurinczuk *et al*., 2010, p. 329).

Hydrocephalus is a blanket term encompassing a variety of "etiological, pathological and age-dependent conditions" of children with impaired flow of cerebrospinal fluid (Stagno, Navarrete, Mirone & Esposito, 2013, p. 17). Children with hydrocephalus present with clinical symptoms, enlarged ventricles on neuroimaging, and alterations in cerebrospinal fluid dynamics (García *et al.,* 2013). Hydrocephalus can be a primary condition, or secondary to conditions such as head trauma, meningitis or brain tumours, to name a few (García *et al.,* 2013). In developed countries the prevalence rate of hydrocephalus ranges between 0.9 and 1.2 per 1000 (Stagno *et al*., 2013).

There is a lack of research on statistics of the number of children born with hydrocephalus in South Africa. According to Stagno *et al*. (2013) the prevalence of hydrocephalus in developing countries will be higher than that stated above owing to nutritional deficits, poor treatment of perinatal infections and inadequate antenatal diagnosis of the condition. A paper written by Qureshi, Piquer and Young (2013) attempted to estimate the burden of disease for hydrocephalus in Africa. These authors estimate that of the 250 million people residing in Central, Southern and East Africa, more than 14 000 will develop hydrocephalus annually within their first year of life (Qureshi *et al*., 2013).

The prevalence of meningitis in children varies depending on the age of the child and on the type of meningitis. A study by Levy, de la Rocque and Cohen (2009) estimated an overall incidence of 2.23 per 100 000 people in France for all ages and strains of meningitis. However, when age is taken into account it can be seen that meningitis is far more common in children than in adults (Levy *et al*., 2009). The incidence rate in France was found to be 44 per 100 000 for children under the age of 1 year and 6.9 per 100 000 children aged 1 to 4 years (Levy *et al*., 2009).

Tuberculosis Meningitis (TBM) is more prevalent in developing countries owing to the higher prevalence of tuberculosis in these nations (Principi & Esposito, 2012). The neurological disability resulting from TBM will depend on the level of obstruction of the cerebrospinal fluid from the reaction to the tuberculosis bacteria (Dekker *et al.,* 2011). More than half of this population will present with severe neurological sequelae (Principi & Esposito, 2012). In the Western Cape province of South Africa, the incidence of TBM is seemingly age-dependent, as there are approximately 31.5 children per 100 000 under one year of age affected with TBM, and 0.7 per 100 000 of children between the ages of 10 years and 14 years (Dekker *et al.,* 2011). TBM is the most common bacterial meningitis in children in the Western Cape (Dekker *et al*., 2011).

In view of pneumococcal meningitis, the incidence appears to remain stable across age groups below 17 years of age in HIV uninfected children (Nunes *et al.,* 2011). Pneumococcal meningitis is defined as the identification of pneumococcus in a sterile site, such as cerebrospinal fluid (Nunes *et al.,* 2011). The incidence of pneumococcal meningitis was reported to range between 28.9 and 39.6 cases per 100 000 children under the age of 18 years in South Africa (Nunes *et al*., 2011).

It can thus be seen in the above review that prevalence and incidence rates of neurological disorders in the paediatric population vary widely between studies, and that there is a general paucity of literature on these conditions, especially in South Africa. It is especially difficult to find information on the number of children with these conditions, and how this changes during childhood. This

dearth of published evidence within this context includes that of published research on hearing function in this population.

## Hearing Function and Neurological Disorders

Studies have shown that hearing loss is higher in the HIV-infected paediatric population than in the general paediatric population. Laughton, Cornell, Boivin and Van Rie (2013) state that 20 % of HIV-infected children in high income countries and 38 % in lower income countries presented with a hearing loss. This is higher than the incidence of three per 1000 births in the general paediatric population stated above.

In poorer resourced areas these hearing deficits were found to be largely conductive hearing loss as a result of chronic suppurative otitis media (CSOM) (Laughton *et al*., 2013). This differed from higher resourced areas, where the hearing losses found were mainly sensorineural in nature (Laughton *et al*., 2013). Hearing impairment for this HIV positive population has been linked to both a low CD4 count and a history of AIDS-defining illness (Laughton *et al*., 2013). However, this may be compounded by the fact that the global burden of disease for CSOM lies in Africa, South-East Asia and the Western Pacific (Mahadevan *et al*., 2012), thus more children in these countries will present with a conductive hearing loss, including those children encompassing the HIV positive population.

Children with Down's Syndrome present a special case in terms of audiology. Hearing loss is more common in children with Down's Syndrome than in those affected by other developmental disabilities and in healthy children (Abou-Elhamd *et al*., 2013). Age-related sensorineural hearing loss (SNHL) occurs approximately 30 to 40 years earlier in individuals with Down's Syndrome (Malt, *et al.,* 2013). This population was found to be 75 % more at risk of developing a hearing loss in America (Pitetti *et al*., 2013).

Children with Down's Syndrome have narrow ear canals (Malt *et al*., 2013), and Eustachian tube dysfunction (Austeng *et al.,* 2013). This most likely stems from muscle hypertrophy and hypertrophy of the mid-face that are characteristic of the syndrome (Austeng *et al*., 2013). As a result of these abnormalities, there is insufficient ventilation in the middle ear and stagnation of fluid, leading to an increase in the number of cases of CSOM seen in children with Down's Syndrome (Austeng *et al*., 2013). CSOM is significantly related with hearing loss, thus hearing loss is more common in children with Down's Syndrome than those without (Austeng *et al*., 2013).

In Utah, in the United States of America, 46.1 % of children with Down's Syndrome presented with a hearing loss in a study by Park, Wilson, Stevens, Harward and Hohler (2011). Of these children with a hearing loss, 88.2 % were diagnosed with a conductive hearing loss, 3.9. % had a SNHL and 2 % had a mixed hearing loss (Park *et al*., 2011).

Studies conducted in Erie in America have shown that 100 % of children with Autistic Spectrum Disorder will manifest auditory-related dysfunction of some level (Kulesza, Lukose & Stevens, 2010). These dysfunctions can range from hyperacusis (increased sensitivity to sound), delayed cortical responses to low frequency sounds, disrupted encoding of simple sounds, difficulty listening in noisy environments, to deafness (Kulesza *et al*., 2010).

Children with CP can have accompanying hearing loss (Rosenbaum, Paneth, Leviton, Goldstein & Bax, 2007). This can be any amount of decibel loss. It is therefore recommended that the hearing of every child with CP be assessed and remediated so as to provide the child with the best access to sound (Rosenbaum *et al*., 2007).

Eleven percent of children with CP in Europe have concomitant visual impairments (O'Shea, 2008), thus assessment and intervention of hearing loss becomes more important for these children, who will rely on information from the hearing modality to compensate for visual deficits. Identification of

concomitant problems in children with CP improves the quality of life and helps to structure the individualized management plan more effectively (Campbell *et al*., 2008).

Of the infants who have birth asphyxia, approximately 3 % present with a mild hearing loss, and a further 3 % present with a severe hearing loss worldwide (Berke, 2010). Therefore, these children will require auditory testing in order to diagnose and provide intervention for the hearing loss.

Similarly, between 5 to 10 % of premature infants globally are diagnosed with a hearing loss of some degree (Nour, 2012). In a study by Moore *et al.* (2012), 0.2 % of infants born before 27 weeks gestation in England were diagnosed with a profound hearing loss. Twenty five percent of this worldwide premature population will also develop a visual impairment (Nour, 2012), thus hearing will be very important in this population.

Hydrocephalus is often associated with hearing loss. According to Spirakis & Hurley (2003), 83 % of the children in their study conducted in Tampa, Florida, had a unilateral high-frequency cochlear hearing loss on the side of the shunt. Therefore, these children require hearing testing.

Dixon and Jones (2012) more recently found in Kentucky in America that increased cranial pressure can result in a bilateral SNHL as a result of the pressure limiting the transmission of sound through the cochlear aqueduct. They found that this hearing loss improved with surgery to decrease cranial pressure, and thus recommend that every child with hydrocephalus has a hearing test (Dixon & Jones, 2012).

Karanja, Oburra, Masinde and Wamalwa (2013) found that among a cohort of meningitic children in Kenya, 43.4 % of the sample presented with a SNHL post meningitis, of which 26.5 % presented with a mild to moderate hearing loss, and 16.8 % with a severe to profound hearing loss.

It can therefore be seen that hearing loss is more prevalent in children with the above neurological disorders than it is in the typical paediatric population. This therefore emphasizes the need for hearing testing in neurologically-compromised children.

## Assessing Hearing Function in Children

When testing hearing in children, it is preferable to utilize the cross-check principle whereby a variety of tests are utilized and the results compared in order to make a final diagnosis (Madell & Flexer, 2008). According to the HPCSA (2012), all students of Audiology should be competent in testing the hearing function of children in order to graduate the Degree.

For children less than 6 months it is recommended to rely mainly on objective measures of hearing, such as the ABR test (Madell & Flexer, 2008). Behavioural observation audiometry in this population can be attempted, for example observing if the child's sucking becomes faster in response to sound, however this is considered to be overall unreliable (Delaroche, Gavilan-Cellie, Maurice-Tison, Kpozehouen & Dauman, 2011).

Behavioural testing of hearing is the gold-standard of obtaining hearing thresholds (Delaroche *et al*., 2011). Behavioural audiometry allows threshold estimation across the entire frequency range for both air and bone conduction, which is essential information for speech and language development (Delaroche *et al*., 2011). Behavioural audiometry also provides an estimate of the entire auditory system from the outer ear to the auditory cortex, thus providing information on cognitive development (Delaroche *et al*., 2011). Even for children with complex needs, behavioural evaluations of hearing are preferable (Hall & Bondurant, 2009).

Once the child has achieved head control and has reached the developmental age to search for the origin of the sound, hearing can be tested behaviourally through visual reinforcement audiometry (VRA) (Proops & Acharya, 2009). This involves presenting a sound to the child through headphones, insert earphones or speakers and reinforcing positive responses by illuminating toys in a glass-fronted box (Proops & Acharya, 2009). Children able to follow simple commands, usually from 2 or 3 years of age can have their hearing tested through conditioned play audiometry (CPA) (Proops &

Acharya, 2009). In this test children are trained through repetition to perform a task each time they hear a sound, for example throwing a block into a bucket (Proops & Acharya, 2009).

However, according to Jerger and Hayes (1976), behavioural audiometry alone is not enough as the results obtained can be misleading and lead to misdiagnosis. Therefore a test battery approach is necessary (Madell & Flexer, 2008). This involves selecting multiple tests that both corroborate the results of previous testing, as well as provide information on different parts of the auditory pathway. By utilizing the test battery approach conclusions are not drawn from a single test and multiple pathologies can be identified (Madell & Flexer, 2008). This test battery approach is reportedly 20% better at diagnosing hearing loss than using a single test (Diefendorf & Wynne, 2004).

## Assessing Hearing in Special Needs Populations

The developmental age of the child will have an impact on the success of the behavioural audiometry attempt (Diefendorf & Gravel, 2000). For children with special needs, certain adaptations can be made to the test battery and the tasks to try and accommodate for their needs (Madell, 2008; Shoup & Roeser, 2007). Positioning, test stimuli and the required response can be altered, based on estimations of the motor and play skills of the child (Madell, 2008).

Children with neurological disorders are a population for which behavioural responses are difficult to ascertain (Madell, 2008). The responses may be much slower, they may fatigue quickly, or they may habituate to test stimuli and reinforcers faster than those children without neurological disorders (Madell, 2008). These children may also become fearful of the reinforcer or refuse headphones (Madell, 2008).

The behaviour of this population in the test may be so erratic that standard techniques cannot be utilized to test them (Northern & Downs, 2002). According to Proops & Acharya (2009), children

such as those diagnosed with ASD may not respond reliably in behavioural testing, and would thus require objective testing to make a definitive diagnosis. The ABR, as found in Portugal, is often the only test that offers information about the hearing status of these children as a result of their "lack of cooperation, attention deficit and cognitive dysfunction" (Coutinho, Rocha & Carmo Santos, 2002, p. 84).

According to the American Speech and Hearing Association (ASHA), for those children with developmental disabilities, the diagnosis of hearing loss should rely primarily on objective testing, such as the ABR, to evaluate hearing function (Purdy & Kelly, 2008). This should be done through obtaining frequency-specific information so that the audiogram can be estimated (Purdy & Kelly, 2008). However, immittance audiometry, OAEs, a comprehensive case history, behavioural observation, and functional hearing measures should also all be conducted or at least attempted in order to supplement the ABR results (Purdy & Kelly, 2008; Lancioni *et al.,* 1985).

## The ABR Test

The ABR is a brainstem auditory evoked potential that allows "neurophysiologic analysis of the auditory pathways between the inner ear and the upper brainstem" (Sleifer *et al.*, 2007, p. 2). An ABR is an electrophysiological test representing the "summed auditory neural activity" (Purdy & Kelly, 2008, p. 132). It tests the audiological pathway from the eighth cranial nerve to the midbrain in the brainstem (Newman & Sandridge, 2007).

Surface electrodes are placed onto the forehead and on the mastoid behind each ear so that electrical responses to the acoustic signal placed into the patient's ear can be measured (Newman & Sandridge, 2007). The response is a summation of the neural activity along the pathway and is described in terms of latency (time) and amplitude (voltage) (Purdy & Kelly, 2008).

The ABR response is measured within the first 10 msec after the presentation of the stimulus (Arnold, 2007). Through using both clicks and tone-burst stimuli the neural integrity of the auditory pathway from the vestibulocochlear (VIIIth cranial) nerve to the midbrain can be examined, and the hearing threshold can be estimated (Arnold, 2007).

The click ABR is an ABR tested using a brief duration and rapid onset click stimulus (Purdy & Kelly, 2008). The click is comprised of many frequencies, and is thus considered a broadband stimulus (Crumley, 2011). This makes it possible to record an ABR on a patient with a significant hearing loss, as long as there are areas of hearing ability in the frequency range of 2 kHz – 4 kHz (Crumley, 2011).

However, this can give a false impression of the patient's hearing, as there is a possibility of a child with a significant hearing loss in the higher frequencies showing responses in the ABR with a broadband stimulus if there is useable hearing in the low frequencies (Ferm, Lightfoot & Stevens, 2013). It is for this reason that there has been a move away from broadband stimuli in testing the hearing status of newborns (Ferm *et al.,* 2013).

Tone-burst ABR, alternatively, utilizes tone-burst stimuli that allow for frequency-specific assessment (Crumley, 2011). Through utilizing tone-burst ABR, a clinician is able to isolate low-frequency versus high-frequency hearing loss (Crumley, 2011), and reliably predict the pure-tone audiogram (Purdy & Kelly, 2008). However, a disadvantage is that the amplitude of the tone-burst stimuli is typically 70 % smaller compared to that of click stimuli, thus the testing time is twice as long in order to overcome this problem and allow for a more robust response (Ferm *et al.*, 2013).

Both of these stimuli are important in audiological practice. Currently, the click ABR has an important role to play in the diagnosis of ANSD, thus the accepted practice is to utilize broadband stimuli for this testing, and tone-burst stimuli to acquire information about the child's audiological abilities (Purdy & Kelly, 2008).

Similarly, the latency of the ABR response has been shown to be age-dependent (Coenraad, Immerzeel, Hoeve & Goedegebure, 2010). The latency of waves III and V decreases with age until two years, however the latency of wave I is seemingly stable from birth (Coenraad *et al.,* 2010). This has been attributed to a central maturation effect as a result of the process of myelination in the first two years of life (Coenraad *et al.,* 2010).

Although tone-burst ABR can predict a child's hearing ability, the relationship between the results of tone-burst ABR and pure tone audiometry for the same child is not exact (Purdy & Kelly, 2008). It is for this reason that correction factors are necessary that allow for the creation of an estimated audiogram based on the ABR results (Purdy & Kelly, 2008). Tables based on regression equations of a study in 1995 are commonly used for this purpose (Purdy & Kelly, 2008); however each clinic is encouraged to establish their own clinical norms based on the testing system utilized and the patient population utilizing the services (Arnold, 2007).

Bocskai, Németh, Bogár and Pytel (2013) studied the effects of sedation on the ABR. Although the ABR tracings for children under sedation contained less artefacts than awake tracings, this study found that sedatives can have negative side-effects in certain children (Bocskai *et al.,* 2013). These side-effects can include agitation, nausea, vomiting, coughing, low blood pressure, breathing difficulties and oxygen desaturation (Bocskai *et al.,* 2013).

The ABR has been shown to be a reliable and accurate manner with which to assess the hearing status of the paediatric population (Dornan, Fligor, Whittemore & Zhou, 2011). In order to optimize the efficiency of the test, different testing protocols can be used so that the maximum information regarding the child's hearing status is obtained in the shortest possible time (Purdy & Kelly, 2008). In this manner, should testing have to be aborted owing to the child awaking, a clinical decision should still be possible with the information available (Purdy & Kelly, 2008). Many different suggested protocols exist in order to achieve this aim (Purdy & Kelly, 2008).

## ABR in Neurologically Compromised Children

For children with special needs, the primary goal is to rule out bilateral hearing loss of a mild degree or worse (Diefendorf & Wynne, 2004). This, as shown above, is generally done through ABR testing. Studies have shown that children affected with neurological disorders have ABRs that differ from those with a normal neurological status (Kwon, Kim, Choe, Ko & Park, 2007). These differences therefore need to be taken into account when conducting hearing testing in these children.

According to the Journal of Korean Medical Science, the ABR for children in South Korea on the Autistic Spectrum typically presents with prolonged latency of wave V, as well as the Interpeak Latencies I-V and III-V (Kwon *et al*., 2007). This can indicate that children with Autistic Spectrum Disorder have an immaturity or dysfunction in the Central Nervous System (Kwon *et al*., 2007).

Kulesza *et al*. (2010) found that children on the Autistic Spectrum in Erie in the United States of America have immature brainstem anatomy, namely the Superior Olivary Complex. This dysmorphology in auditory brainstem nuclei results in auditory dysfunction mentioned above ranging from hyperacusis to deafness (Kulesza *et al*., 2010).

This study goes further to state that these structural deficits are present at birth, thus ASD could be diagnosed earlier through the use of ABR (Kulesza *et al*., 2010). However, these structural abnormalities vary within individuals diagnosed with ASD, resulting in diversity and challenges in diagnosing ASD (Kulesza *et al*., 2010). They purport that as ASD is a continuum depending on the extent of neurological dysfunction, auditory deficits observed will depend on the extent of dysmorphology in the auditory brainstem with deafness being the most severe (Kulesza *et al*., 2010). ABR therefore cannot be the sole manner of diagnosis (Kulesza *et al*., 2010).

In a population of children with Down's Syndrome in Poland, the ABR presented with short latencies and lower amplitudes, possibly owing to the accelerated maturation of the nervous system or

disturbances in the nervous system (Kręcicki, Zalesska-Kręcicka, Kubiak & Gawron, 2005). Peaks II, III, IV and V were found to have shorter absolute latencies, which leads to short I-II and I-III Interpeak Intervals (Kręcicki *et al*., 2005). This was found for intensities of at least 45 dB above the patient's threshold. The shorter latencies and amplitudes could also be attributed to the deficient cerebral organization and information processing common of these children (Kręcicki *et al*., 2005).

However, when Kuhn *et al*. (2013) conducted ABR testing on mice with Trisomy 21 in the United Kingdom, they found that waveform amplitudes and latencies and hearing levels for these mice compared to normal controls, and no abnormalities were exhibited in the ABR. This indicates that clarity on this population is needed, as definitive results on how Down's Syndrome affects the ABR are not available at present.

Jiang, Brosi, Shao and Wilkinson (2008) studied ABR tracings in infants in Shanghai, China and the United Kingdom with severe perinatal asphyxia. They found that during the first month of life the amplitudes of all waves, particularly wave V, were reduced (Jiang *et al*., 2008). This reflects the decreased number of cells in the response, resulting from neuronal impairment or death in the brainstem following asphyxia (Jiang *et al*., 2008).

The reduction in amplitudes continued well into the first month of life (Jiang *et al*., 2008). This prolonged decrease indicates that the neuronal impairment is therefore not likely to recover quickly (Jiang *et al*., 2008). Some cells may not only die at the time of the asphyxia, but continue to die hours and days after the event (Jiang *et al*., 2008).

Development of the auditory system is postulated, based on a cohort of Brazilian children, to occur in two phases (Casali & dos Santos, 2010). The first phase occurs in utero whereby the peripheral auditory system is developed by the 6$^{th}$ month of gestation (Casali & dos Santos, 2010). The second phase occurs after birth until 18 months of age and comprises of the maturity of the central auditory

pathways up to the brainstem (Casali & dos Santos, 2010). This is important to take into account in testing premature infants.

Absolute latencies of all waves are increased in neonates in comparison to adult values, possibly as a result of the immature auditory system not having undergone myelination, thereby increasing the conduction time (Casali & dos Santos, 2010). The absolute latency of Wave I decreases with an increase in gestational age and reaches adult values within the first few weeks of life, whereas the values of Waves III and V only reach adult latencies by two years of age (Casali & dos Santos, 2010). Therefore taking into account the child's gestational age is important in testing premature infants with ABR (Casali & dos Santos, 2010).

The only study conducted on ABR results in patients with hydrocephalus dates back to 1984. No recent studies could be found on the subject. In this study, 88 % of the hydrocephalic cohort in Chicago, America, indicated some abnormalities on ABR, ranging from delayed wave latencies to poor waveform morphology to raised hearing thresholds (Kraus, Özdamar, Heydemann, Stein & Reed, 1984).

Children in Guangxi Zhuang National Autonomous Region in China with CP presented with prolonged latencies when compared to neurologically uncompromised peers (Zhu, Wang & Liang, 2006). However there is a dearth of studies in this population, possibly owing to the vast differences in the hearing function of these children.

In a study by Topcu *et al.* (2002) in Diyarbakir, Turkey, 24 % of children with meningitis presented with abnormal ABR results. Interpeak latencies were increased for I – V and I – III, and 14.8 % presented with no visible waveforms at the maximum output of the equipment (Topcu *et al*., 2002). These differences are important to keep in mind in order to decrease the chance of misdiagnosis in this population.

The above information on how neurological disability affects the ABR was extremely difficult to locate. Most studies were based on a small sample size, and are out-dated. Worldwide, there is a severe lack of literature on how the ABR is affected by certain medical (especially neurological) conditions in children. For some of these conditions, such as Down's Syndrome and RVD, no information is available on how these diagnoses affect ABR results. Therefore, this is an area requiring further research; hence the importance of this study.

# 2. Methodology

## 2.1. Aims and Objectives

**Primary Aim of the Study:**

To describe the hearing test results in a group of neurologically compromised children.

**Study Objectives:**

1. To describe the audiological ABR findings in order to determine hearing function in this group.

2. To establish a relationship between audiological ABR findings and behavioural audiometry findings where these exist.

## 2.2. Research Design

A descriptive Retrospective Review of patient records was conducted on 40 audiological records of children between the ages of 5 months to 10 years from the Speech Therapy and Audiology Department at Chris Hani Baragwanath Academic Hospital.

In qualitative research a naturalistic approach is used whereby the aim is to understand phenomena in context-specific settings (Golafshani, 2003). This study aimed to describe the hearing status of a neurologically disordered cohort of children whose test battery included the ABR at the Audiology

Department of Chris Hani Baragwanath Academic Hospital. Traditionally, qualitative research aims for “illumination, understanding, and extrapolation to similar situations” (Golafshani, 2003, p. 600).

Audiological records of patients fitting the inclusion criteria were drawn from the Departmental Archive. These records were of children assessed between the months of September 2010 and February 2012. Results of Neurologically Typical children were not analyzed. Results of children diagnosed with one or more of the following disorders were included in this study:

- Cerebral Palsy
- Developmental Delay
- HIV/AIDS
- Hydrocephalus
- Down’s Syndrome
- Birth Asphyxia
- Autism Spectrum Disorder
- Meningitis

These pathologies were chosen, as according to the earlier described Burden of Disease, neurological disorders account for 10 % of children in low-resourced areas (Patel, Goel & Desai, 2009). These conditions are the most prevalently seen in children attending health care institutions in Soweto. For the purposes of hydrocephalus and CP, these blanket terms were utilized as this is what was recorded on the patient record. The subcategories of these pathologies were not recorded, thus it remains unknown as to with which type of hydrocephalus and CP the patient was diagnosed by the medical practitioner.

Testing results for behavioural testing attempts (if available) were also included in the data collection. The most recent test results were utilized for comparison to those results obtained objectively via Tone-Burst ABR.

**Equipment:** The records of the patients' ABR tests were drawn from the Resource Room of the Audiology Department. A laptop containing an Excel spreadsheet was utilized for data capturing and data management.

## 2.3. Participants

**Participants:** Forty audiological files and records of infants and children between the ages of 5 months and 10 years diagnosed with one of the above neurological disorders were included in this study. These records were of patients tested at the Audiology Department at Chris Hani Baragwanath Academic Hospital using the Eclipse Auditory Evoked Potential Machine during the months of September 2010 and February 2012. All children in this study were tested under natural sleep, i.e. without sedation. Owing to the side-effects outlined above, sedation is only used in rare cases in CHBAH, and thus this population would be too small in number to analyze for the purpose of this study. The patients had been diagnosed by an appropriate medical practitioner as presenting with one or more of these neurological disorders.

Results of behavioural testing attempted for these children were drawn. The most recent test results were utilized for those with multiple attempts. If the results of the behavioural attempts were inconclusive, this was documented. For those children for whom no results were available, "*no results*" was documented for the behavioural section.

**Recruitment:** Patient records were recruited from the archive at Chris Hani Baragwanath Academic Hospital. The first 40 records of patients meeting both the age and diagnostic criteria were included in the sample.

## 2.4. Test Protocol

All patients in this study were tested using the CHBAH ABR protocol, as indicated in the patient records. The following describes the manner in which all patients were tested. The ABR results consisted of Tone Burst thresholds obtained at one or more of the following frequencies: 500 Hz, 1 kHz, 2 kHz and 4 kHz. Threshold for at least one frequency in each ear was documented during testing so as to obtain ear-specific information regarding both ears.

Patients will initially have been scrubbed using Webcol alcohol swabs and NuPrep Skin Preparation Paste on the electrode sites. Electrodes were placed on the mastoid processes behind both the left and the right ears, high on the forehead in the midline (vertex) and directly below this between the eyes (ground). Impedance was less than 5Ω across all electrode sites for testing to commence. The EEG was set to 20 µV, with minimal rejects. All wave tracings with a large number of rejects and low wave reproducibility were deleted, and another tracing was conducted at that intensity.

The Tone Burst ABR was conducted at 44.1 cps, on an alternating polarity. Testing began at 60 dBnHL at 4kHz, and Wave V was traced until its disappearance. If Wave V could be clearly seen, the intensity was reduced by 10 dBnHL. If Wave V could not be seen, the intensity was increased by 5 dBnHL until the waveform was visible. A second tracing was conducted at the lowest intensity at which wave V could be seen to ensure that the downward deflection is Wave V.

Correction factors based on the transducer, patient age and frequency (see Appendix 2) were then applied in order to convert this result to a hearing threshold estimate in dBeHL. Should no waveform

be visible at the maximum intensity of the system, "NR" (or "*No Response"*) would be documented, followed by the maximum intensity at which the testing was conducted.

The behavioural test battery began with otoscopy and immittance testing (tympanometry and acoustic reflexes) in order to gain information about the child's outer and middle ear status. Depending on the child's age and developmental and physical capabilities, either Visual Reinforcement Audiometry (VRA) or Conditioned Play Audiometry (CPA) was then attempted under headphones. Testing began at 1 kHz at 60 dBHL where the child was familiarized with the task, and the threshold for each frequency was documented as the lowest level at which the patient gave two reliable conditioned responses to the sound (either a head turn or placing a toy in a bucket)

If the patient was averse to the headphones, testing in free field was attempted. Sound was presented through one of two speakers, and the same conditioned response would have been expected. Thresholds for this test are not ear-specific as the patient is listening with both ears simultaneously (Delaroche, Thiebaut & Dauman, 2004). Results of free-field testing pertain to at least one cochlea, but results cannot be documented per ear (DeConde Johnson & Seaton, 2011), therefore results were not recorded per ear for free-field testing. The better hearing ear will respond; therefore if the hearing loss is asymmetrical the worse ear may be misdiagnosed.

## 2.5. Data Analysis

Each patient's hearing status was described per ear based on the documented tone-burst ABR thresholds. The classification by Gelfand (2009) was utilized to describe the degree of hearing loss for each ear tested (see Appendix 1). This description was also done for those patients for whom behavioural results under headphones were available. For those patients tested behaviourally in free-field the overall hearing status was described, and one ear was marked as "*no results.*"

Descriptive statistics were conducted by the researcher for each set of objective and behavioural results, per ear, for all 40 subjects in terms of incidence and degree of hearing loss, as well as comparing ABR and behavioural results within the same ear. The pathologies were then documented in terms of occurrence, as well as the hearing results for each of the six main pathologies. Patient age was expressed in terms of the average and the standard deviation (in months).

## 2.6. Reliability and Validity

### 2.6.1. Reliability

Reliability refers to the stability of the test (Carter, Godoy, Marakovitz and Briggs-Gowan, 2009). In order for an audiological test to be reliable the test should consist of controlled stimuli in an acoustically-controlled environment (Frank & Rosen, 2007). All patients in this study were tested using the Interacoustics Eclipse System for ABR testing and a GSI 61 audiometer for behavioural testing, both calibrated and serviced annually, thus technical inconsistencies were minimized. Daily biologic calibration was conducted by the audiologist before using all equipment as per Departmental Protocol, which ensures the accuracy of the equipment (Frank & Rosen, 2007). All testing rooms were also sound-treated to reduce noise.

Owing to the fact that this study is a retrospective record review, controlling patient variables to a high degree of reliability could not be ensured. The testing conditions remained the same in each test to the best of the tester's ability. According to Carter *et al*. (2009), a detailed training manual and assessment protocol increases inter-rater reliability. All patients were tested with the same protocol, written by the researcher, thus maximizing reliability. Patient and test variables were

thought to also be managed by the fact that the behavioural audiometry and ABR testing procedures form part of standard audiology practice for qualified audiologists (Kanji, 2010).

The ABR test has established test-retest reliability (Song, Nicol & Kraus, 2011). Similarly, intra-subject reliability is small in that variations in the absolute latency of the click ABR differ very little between tests (Song *et al*., 2011). Therefore, results from the ABR test will give a reliable indication of the patient's hearing abilities.

### 2.6.2. Validity

Validity refers to the ability of a test to measure what it purports to measure (Carter *et al.,* 2009). Validity was enhanced by taking into consideration the effect of patient and environmental factors on ABR tracings. According to the ABR protocol utilized at CHBAH, "noisy tracings" (or those tracings for which the signal-to-noise ratio is worse than 3:1) are deleted, and another tracing is conducted. In this way, the response is replicable, and clearer deductions can be made.

Panacek (2007) recommends that in order to increase the validity of a retrospective review, variables should be defined before data is abstracted. Each patient had been diagnosed with a neurological disorder by a relevant professional before their ABR test, and this diagnosis was copied from the patient's hospital file by the tester. Degree of hearing loss was classified for each patient according to the classification by Gelfand (2009) (see Appendix 1). In this way the abstraction from the data records was consistent (Panacek, 2007).

Similarly, Panacek (2007) states that a uniform data collection form should be utilized in order to enhance the validity of the study. All patient data was captured onto a summary form (see Appendix 6), thus all data were captured uniformly for each patient.

### 2.6.3. Bias

ABR interpretation is subjective, as the tester has to identify waveforms in order to make a diagnosis. In order to reduce bias, the interpretation of the ABR waveforms was taken as written on the patient report. The waveforms for each child were interpreted by the tester at the time of the test. The diagnosis written on the page was taken, thus results were analyzed by one of five audiologists and not just the author.

## 2.7. Ethical Considerations

Permission to review records was obtained from CHBAH hospital management and the Head of Department of Speech Therapy and Audiology at CHBAH (see Appendices 3 and 4). Data collection only began following permission from the Postgraduate Review Committee, and the Human Research Ethics Committee (Medical) at the University of the Witwatersrand. The Ethical Clearance Certificate Number M120217 (see Appendix 5) was obtained on 24.02.2012. All of this was in accordance with the World Medical Association (WMA) *Declaration of Helsinki* (2008), which calls for all medical research to appear before an Ethics Committee.

Owing to the fact that it is a retrospective study of patient records, patient consent was not obtained. However, these patients attended a teaching and research hospital where records are utilized for research purposes. According to the WMA *Declaration of Helsinki* (2008), no harm should come to the patient. All subjects in this study sample attended CHBAH for a hearing test during which the patient was not harmed in any way. No additional intervention occurred for the purpose of this study.

Similarly, the WMA *Declaration of Helsinki* (2008) states that patient privacy should be ensured, and that no personal information should be divulged. A coding system was created whereby patients were allocated a number so that no identifying information was utilized. This ensured confidentiality and anonymity of the study sample. Therefore, this study abides by the ethical regulations of confidentiality and non-maleficence.

The WMA *Declaration of Helsinki* (2008) additionally states that testing should only be conducted by appropriately trained personnel. All testing conducted for the patient records was performed by audiologists working at CHBAH and registered with the HPCSA to practice audiology.

Justice is a major consideration in medical ethics (Pratt & Loff, 2011). Justice is defined differently in various sources, and consists of ensuring that the research conducted reacts to the health needs and priorities of the community or country in which it was conducted, that all benefits are fairly and freely available to participants during and after clinical trials, and result in the strengthening of ethical research in the community or country of the research participants (Pratt & Loff, 2011).

This research project adhered to the principle of justice in that, as evident in the literature review above, neurological disorders are rife in the paediatric population of South Africa, and thus comprise the burden of disease in this population.

Similarly, all patients had undergone ABR testing, thus no treatment options were withheld and all patients received equal opportunities in terms of hearing testing options. By adhering to these aspects of justice, this research contributes to an ethically-sound body of medical studies.

# 3. Results

## 3.1. Demographic Profile

A total of 40 patient files were drawn from the Departmental Archive. All patients resided in the catchment area for Chris Hani Baragwanath Academic Hospital in Soweto, Johannesburg. The sample consisted of 21 males and 19 females. The average age was 28 months, with a standard deviation of 24 months. The median age was 22 months, and the age range was 5 months to 115 months (9 years 7 months).

Results are described per ear for audiological analyses (N = 80), and per participant in analyzing the pathologies (N = 40). Certain patients had multiple neurological disabilities. For these patients all relevant diagnoses were taken into account in analysis if the disability appeared on the above list of neurological disabilities for this study. Eleven children presented with two neurological disabilities and five further children had been diagnosed with three of the neurological disabilities included in this study.

## 3.2. Results of the ABR Test

Of the 40 records drawn from the CHBAH Archive, 22 patients (55 %) presented with normal hearing bilaterally, ten (25 %) presented with bilateral symmetrical hearing loss, seven (17.5 %) presented with an asymmetrical hearing loss, and one patient (2.5 %) was diagnosed with a unilateral hearing loss.

It can be seen in Table 1 below that 45 of the ears tested (56.2 %) presented with normal hearing. Of these 45 ears, 44 (97.7 %) were bilateral and 1 child (2.5 %) presented with a unilateral hearing loss with normal hearing in the right ear. Eight ears (10 %) were diagnosed with a profound hearing loss in Electrophysiological testing. These results were obtained in five children, three with bilateral profound hearing losses and two with unilateral asymmetrical profound hearing losses diagnosed. All three children with bilateral profound hearing losses had been diagnosed with cerebral palsy.

| DESCRIPTION OF HEARING | NUMBER OF EARS |
|---|---|
| Normal Hearing | 45 |
| Profound Hearing Loss | 8 |
| Mild Hearing Loss | 7 |
| Moderately-Severe Rising to Mild Hearing Loss | 4 |
| Moderately-Severe Hearing Loss | 3 |
| Mild Sloping to Moderate Hearing Loss | 2 |
| Moderate Sloping to Severe Hearing Loss | 2 |
| Severe Sloping to Profound Hearing Loss | 2 |
| Moderate Hearing Loss | 1 |
| Moderate Rising to Mild Hearing Loss | 1 |
| Moderate Sloping to Moderately-Severe Hearing Loss | 1 |
| Severe Rising to Normal Hearing Loss | 1 |
| Severe Rising to Moderate Hearing Loss | 1 |
| Profound Rising to Moderately-Severe Hearing Loss | 1 |
| Profound Rising to Severe Hearing Loss | 1 |
| **Total** | **80** |

*Table 1: Table to Show the Description of Hearing Status and the Number of Ears Presenting with Each*

## 3.3. Results of Behavioural Testing

Figure 1 below depicts that for 58 out of a total 80 ears (72.5 % of the study sample) no results could be obtained with behavioural testing. No results could be obtained bilaterally in 24 children (60 %), and unilaterally in 10 children (25 %). Fifty percent (n=11) of the ears for whom behavioural results

were available presented with normal hearing. Three of these were children presenting with normal hearing in both ears under headphones. Five of these patients were tested in free field, and thus have normal hearing in at least one cochlea. Four ears (5 %) presented with a moderate hearing loss. A profound hearing loss was diagnosed in three ears (one unilateral and two bilaterally in 1 child), or 3.75 % of ears tested. Sloping hearing losses were diagnosed in three ears (3.75 %), and a severe-to-profound hearing loss was diagnosed in free field in at least one cochlea for one patient (1.25 %).

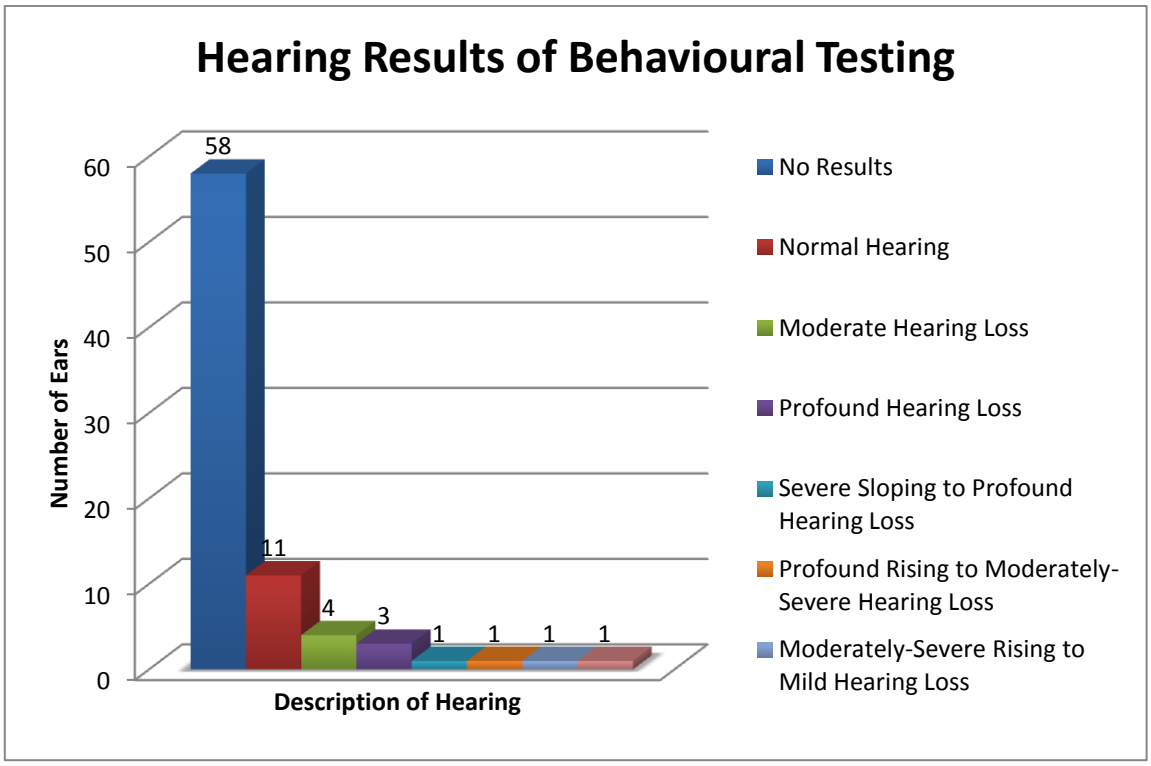


*Figure 1: Bar Graph to Show the Audiological Results of Behavioural Testing*

## 3.4. ABR Compared to Behavioural Testing Results

Of the sample of 80 ears 7.5 % (n=6) were diagnosed with normal hearing in both behavioural and objective testing, as depicted in Table 2 below. For 58 ears (72.5 %) no behavioural results were obtained. For the remaining 16 ears (20 %) there was a discrepancy between behavioural and objective testing. Therefore 92.5 % of the results differed in behavioural and ABR testing, and those results that correlated were only in normal hearing ears.

| BEHAVIOURAL TEST | ABR TEST | NUMBER OF EARS |
|---|---|---|
| No Results | Normal Hearing | 30 |
| No Results | Profound Hearing Loss | 8 |
| No Results | Moderately-severe Rising to Mild Hearing Loss | 4 |
| No Results | Moderate Sloping to Moderately-Severe Hearing Loss | 1 |
| No Results | Moderately-Severe Hearing Loss | 3 |
| No Results | Mild Hearing Loss | 5 |
| No Results | Moderate Sloping to Severe Hearing Loss | 1 |
| No Results | Profound Rising to Severe Hearing Loss | 1 |
| No Results | Mild to Moderate Hearing Loss | 2 |
| No Results | Severe Rising to Normal Hearing Loss | 1 |
| No Results | Severe to Profound Hearing Loss | 2 |
| **Normal Hearing** | **Normal Hearing** | **6** |
| Normal Hearing | Moderate Hearing Loss | 1 |
| Normal Hearing | Moderate Sloping to Severe Hearing Loss | 1 |
| Normal Hearing | Profound Rising to Moderately-Severe Hearing Loss | 1 |
| Normal Hearing | Mild Hearing Loss | 1 |
| Normal Hearing | Moderate Rising to Mild Hearing Loss | 1 |
| Moderate Hearing Loss | Normal Hearing | 4 |
| Moderately-Severe Rising to Mild Hearing Loss | Normal Hearing | 1 |
| Severe Rising to Moderately-Severe Hearing Loss | Normal Hearing | 1 |
| Severe Sloping to Profound Hearing Loss | Normal Hearing | 1 |
| Profound Rising to Moderately-Severe Hearing Loss | Normal Hearing | 1 |
| Profound Hearing Loss | Mild Hearing Loss | 1 |
| Profound Hearing Loss | Normal Hearing | 1 |
| Profound Hearing Loss | Severe Rising to Moderate Hearing Loss | 1 |
| **Total** | | **80** |

*Table 2: Table to Show the Results of Behavioural Compared to ABR Testing in the Same Ear*

It can be seen from Figure 2 below that for a majority of ears (72.5 %) no results were obtained behaviourally. 37.5 % (n = 30) of these ears, when later tested via ABR, had normal hearing, and 35 % (n = 28) were diagnosed with a hearing loss on electrophysiological testing.

For those ears with behavioural results, 7.5 % of ears with a hearing loss diagnosed objectively were falsely diagnosed with normal hearing based on behavioural testing. Similarly, 10 % of children with normal hearing in ABR were diagnosed with a hearing loss behaviourally. Two and a half percent of the sample who had a hearing loss were diagnosed with a hearing loss in behavioural testing, however the degree of the loss was worse in behavioural testing than the results obtained objectively.

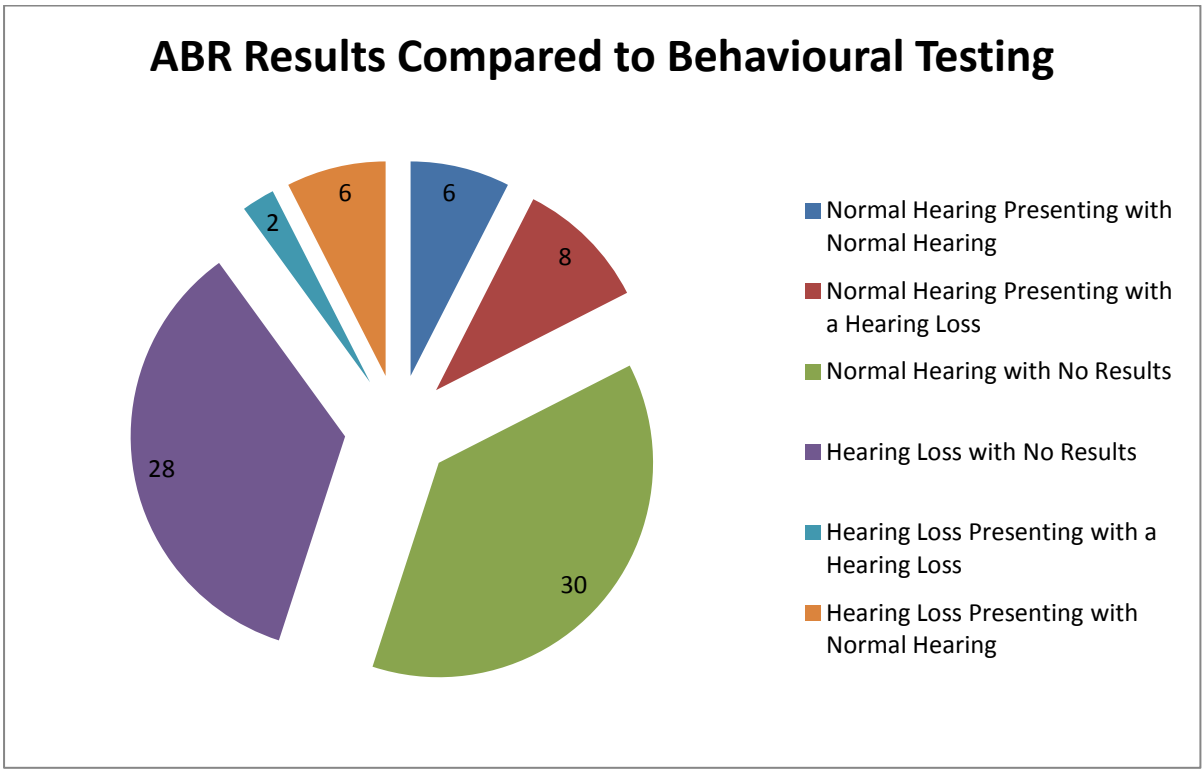


*Figure 2: Pie Chart to Show to Compare ABR Audiological Results with those obtained Behaviourally*

## 3.5. Results per Pathology

Figure 3 below indicates the number of children presenting with each diagnosis. The two most common diagnoses in the sample were prematurity (27.5 % of sample) and cerebral palsy (25 % of sample). Hydrocephalus (17.5 %), Down's Syndrome (12.5 %), meningitis (12.5 %) and birth asphyxia (10 %) were also common in the sample, yielding a combined 52.5 % of the sample. The rest of the pathologies were less common, only diagnosed in 1 or 2 patients each. Only two of the study sample (5 %) presented as RVD positive.

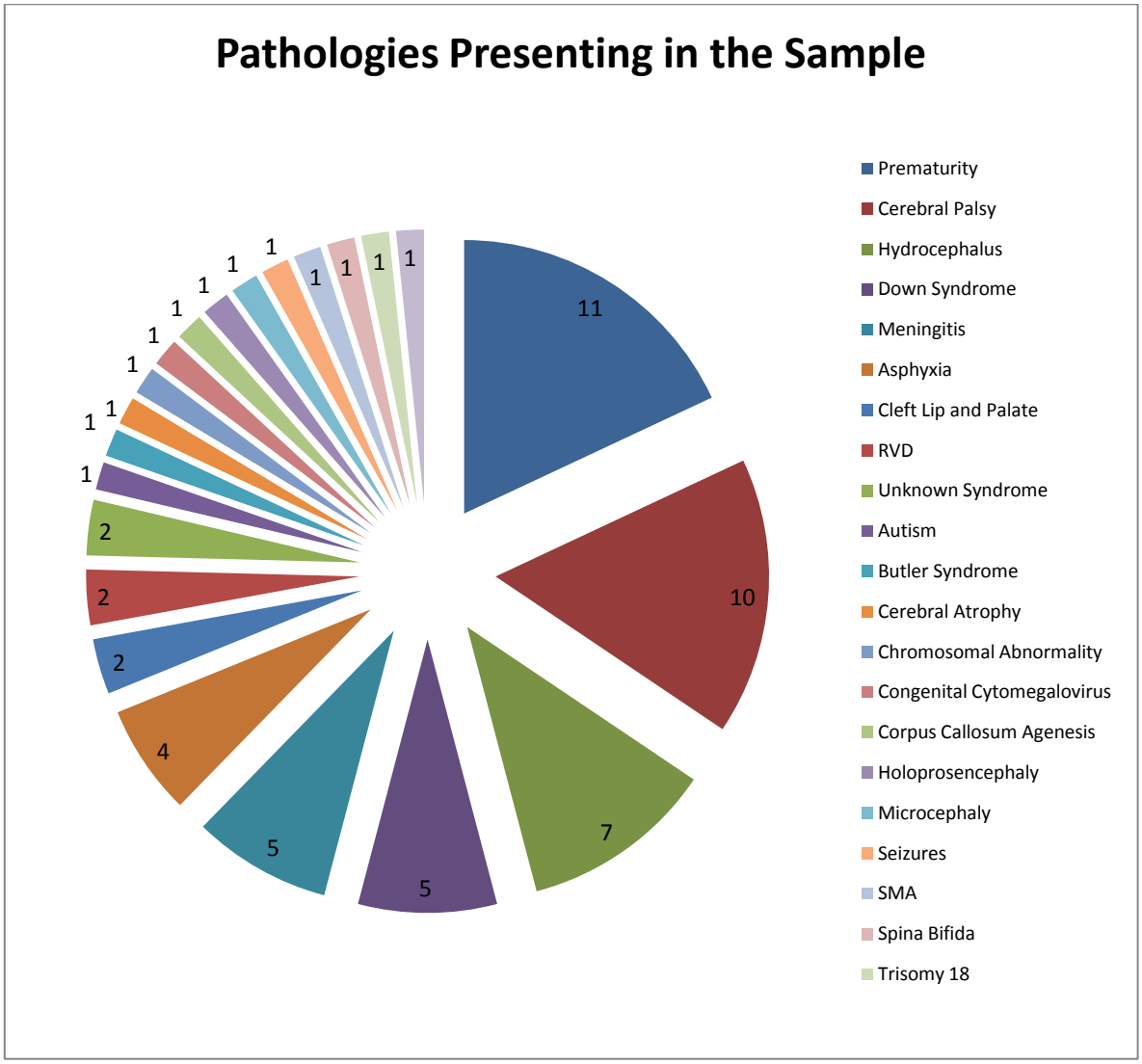


*Figure 3: Pie Chart to Show the Pathologies Represented in the Sample*

Figure 4 below indicates the number of hearing losses diagnosed per pathology. More hearing losses were diagnosed in ABR testing, with a total of 22 losses (55 %) diagnosed on ABR testing for these pathologies, and only six (15 %) on behavioural testing for the same subset of the sample. A majority of patients diagnosed with Cerebral Palsy, Down's Syndrome, prematurity and RVD presented with a hearing loss, as 19 patients out of a combined 28 patients with these conditions (67.8 %) were diagnosed with a hearing loss objectively.

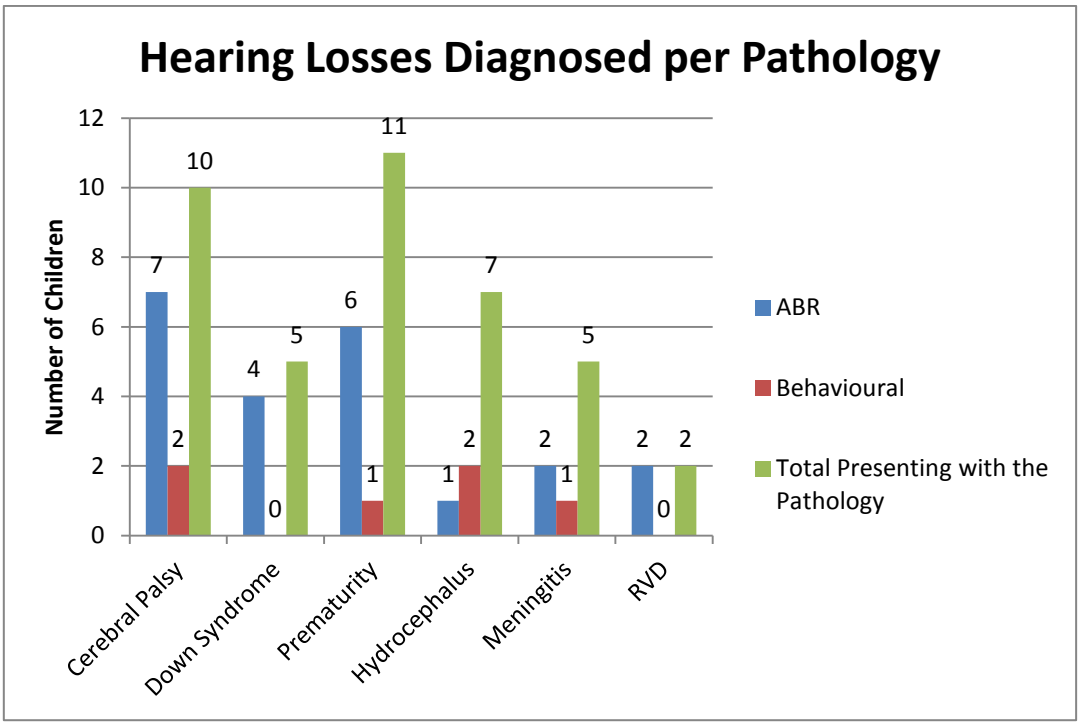


*Figure 4: Bar Chart to Show the Number of Hearing Losses Diagnosed on ABR and Behavioural Testing for the 6 Main Pathologies.*

Table 3 below indicates that, similarly, more patients presented with normal hearing in ABR testing than in behavioural testing for these main pathologies. A total of 18 children (45 %) were diagnosed with normal hearing in ABR testing, as opposed to seven (17.5 %) diagnosed with normal hearing behaviourally for the same subsample.

| | ABR | BEHAVIOURAL | TOTAL WITH PATHOLOGY |
|---|---|---|---|
| Cerebral Palsy with Normal Hearing | 3 | 1 | 10 |
| Down's Syndrome with Normal Hearing | 1 | 0 | 5 |
| Prematurity with Normal Hearing | 5 | 4 | 11 |
| Hydrocephalus with Normal Hearing | 6 | 1 | 7 |
| Meningitis with Normal Hearing | 3 | 1 | 5 |
| RVD with Normal Hearing | 0 | 0 | 2 |

*Table 3: Table to Show the Number of Children Presenting with Normal Hearing in Behavioural and ABR Testing per Pathology*

Table 3 also depicts that a majority of children with hydrocephalus (85.7 %) and with meningitis (60 %) were diagnosed with normal hearing on ABR testing. However, as seen in Figure 4 above, most of the children with CP, Down's Syndrome, prematurity and RVD were diagnosed with a hearing loss on ABR testing.

| | NUMBER WITH NO RESULTS | TOTAL | PERCENTAGE of SAMPLE (%) |
|---|---|---|---|
| Cerebral Palsy | 7 | 10 | 70 |
| Down's Syndrome | 5 | 5 | 100 |
| Prematurity | 6 | 11 | 54.6 |
| Hydrocephalus | 4 | 7 | 57.1 |
| Meningitis | 3 | 5 | 60 |
| RVD | 2 | 2 | 100 |

*Table 4: Table to Show the Number and Percentage of Patients for Whom No Results were Obtained Behaviourally*

Table 4 above indicates that for these six pathologies, a majority of the sample could not be tested behaviourally. Results could not be obtained for all the patients in the sample diagnosed with Down's Syndrome and with RVD. Seventy percent of children with CP could not undergo behavioural testing reliably, as well as 60 % of those with meningitis, 57.1 % with hydrocephalus and 54.6 % with prematurity.

No statistical hypothesis testing could be conducted on the data owing to the lack of complete behavioural results in this population and the fact that within the subset of patients with normal hearing, some levels were screened and some levels were definite thresholds below which the patient will not hear. These inconsistencies led to too many extraneous variables for statistical analysis.

# 4. Discussion

## Diagnosis of Hearing Status

An objective test is described as a test of hearing "without the patient's active participation in the test" (Northern & Downs, 2002, p. 209). According to Diefendorf and Gravel (2000), the responsiveness of the developmentally delayed child is associated with their developmental level, leading to both high false positive and false negative results in behavioural testing. In this study, of the ears with behavioural results, half were diagnosed with a hearing loss, whereas only 44 % of the entire sample presented with a hearing loss on ABR testing (see Table 1). This indicates higher false positive rates for behavioural testing. Similarly, as seen in Table 2, 11 ears presented with a hearing loss of worse degree on behavioural testing than on ABR testing, emphasizing the high false positive rate for behavioural testing. Five ears were diagnosed with normal hearing on behavioural testing; however these ears were diagnosed with a hearing loss on ABR testing (see Figure 2). This indicates a false negative rate in behavioural testing, and is in agreement with the finding of Diefendorf and Gravel (2000) of both high false negative and false positive results stated previously.

In this study population a total of 72.5% (n = 58) of the sample could not be tested behaviourally (as seen in Figure 1). More hearing losses were diagnosed in ABR than on behavioural testing. Thirty five ears were diagnosed with a hearing loss objectively versus 11 behaviourally. This indicates that most hearing losses are detected in a test where a response is not expected from the patient, and is in fitting with the finding of Swanepoel and Ebrahim (2009) that electrophysiological testing, via ABR or ASSR, would be necessary for those populations who are unable to cooperate or respond in behavioural testing.

Two and a half percent of ears (n=2) presented with a hearing loss on both behavioural testing and objective testing (see Figure 2 above). However the degree of hearing loss differed in the two tests

in that both of these children presented with a profound hearing loss on behavioural testing, however the hearing was found to be better on ABR testing (one with a mild hearing loss, and the other with a severe rising to moderate hearing loss) in both children. The two children seemingly did not show any response at high sound intensities tested, thus leading the tester to deduce that the child has a profound hearing loss. The child may have heard the sound, but may have not understood the task, or the conditioned response expected from them may have been too difficult for their developmental level, causing the child to not respond. These children, should the behavioural results indicating a profound hearing loss have been accepted, would have been overamplified in hearing aid fitting, causing further damage to the auditory system.

This discrepancy between behavioural and objective findings supports the study by Proops and Acharya (2009), who found that the reliability of behavioural tests depends on the skill of the tester as well as the state of the child; thus hearing may appear to be of a certain degree in behavioural testing owing to tester interpretation or a fatigued child, but may be of another degree in objective testing. Therefore, objective testing is particularly important in diagnosing hearing loss in a population with neurological disorders.

Likewise, more patients were diagnosed with normal hearing on ABR testing. Objectively 45 ears were diagnosed with normal hearing, whereas only 8 ears presented with this degree of hearing when tested behaviourally. Only six out of the 80 ears tested correlated between behavioural and ABR results and all of these ears were normal-hearing.

The above results obtained in this study suggest that behavioural testing was less sensitive at diagnosing both hearing loss and normal hearing than ABR testing for this study population, and confirms that some children can be misdiagnosed owing to lack of responses in behavioural audiometry, thus highlighting the need for objective testing in assessing auditory function in this population. This is consistent with the finding by Delaroche *et al*. (2004), who assert that some

multiply-handicapped children may never be able to be tested behaviourally and that objective testing will thus be vital in ascertaining their hearing status.

## Behavioural Testing in Neurologically Compromised Populations

This study aimed to describe the results of behavioural testing in a sample of neurologically disordered children and those results obtained with objective ABR testing. In the study behavioural results were obtained in only 22 of the 80 ears (27.5% of the sample). No behavioural results could be obtained for 24 of 40 children. Four of these children were aged 5 or 6 months, thus behavioural testing was not attempted owing to their developmental capabilities. For six children behavioural testing was attempted but was aborted as the child could not be tested. For a further six children testing occurred in free-field, thus no ear-specific results were obtained.

Aside from the four children aged 5 or 6 months, the sample were aged 7 months to 10 years, and are thus at the correct chronological age for behavioural testing. However, most of the children in this sample could not undergo behavioural testing owing to their developmental stage resulting from the neurological disorder. This is contrary to the findings of Madell (2008), who purports that by conducting estimates of the child's developmental age, auditory information can be obtained in the same manner as normal-developing children with the same developmental age. This disagreement may be a result of some children in the sample having multiple neurological disorders, and others being severely developmentally delayed. The child's developmental age is not estimated at CHBAH, as this is not part of the protocol for either behavioural or objective testing, which could also contribute to the discrepancy.

However, as stated previously, it has been well established that behavioural audiometry remains the gold standard in hearing testing as it provides an estimate of the entire auditory system from the

outer ear to the auditory cortex (Delaroche *et al*., 2011), and should be utilized as a supplement to objective testing for neurologically disordered children (Lancioni *et al.,* 1985). It is therefore vital that behavioural results be sought in all children, as objective testing does not give an overview of the entire auditory system. In this study population, however, most children were not able to undergo behavioural audiometry. In 20 children (half the study population) behavioural results were not even attempted. Behavioural audiometry needs to be included this into the test battery for every child in order to comply with the gold standard mentioned above. Therefore, based on the number of children in this population unable to cope in behavioural testing, this should be attempted later in the child's development than in normally developing children. Preliminary habilitation should be based on objective results and altered as behavioural results are obtained after time. Behavioural results are important in holistic treatment of hearing impaired children, as stated above.

In order to give the child with a neurological disorder the best chance of completing the task required for behavioural testing, Madell (2008) suggests making some adaptations to the test battery such as changing the stimulus, positioning the child appropriately, altering the expected response and presenting the stimulus differently. Behavioural testing should be attempted on this population with these adaptations; however interpretation by the tester should still be holistic, based on the case history, and cautious, ensuring that all results are cross-checked. The child may also need a number of sessions over which they are conditioned before they are able to understand the required response (Shoup & Roeser, 2007).With these adaptations and careful interpretation, behavioural testing may yield more results in this paediatric neurologically-disordered population than the current protocol if attempted at CHBAH. In this way the audiologist will gain information into the entire auditory system, and not just up to the auditory brainstem if only objective results are available.

All of the above indicates that results are difficult to obtain behaviourally for neurologically-disordered children. A majority of this cohort did not yield any behavioural results, and thus hearing testing relied completely on objective measures. Objective testing, however, only provides information as far as the auditory brainstem and not higher. Behavioural testing offers an idea of how the sound is perceived and processed by the child up to the auditory cortex. It is therefore important and should not be overlooked in order to assess how the child processes sound, and not just how it is perceived. The children in this population should therefore undergo behavioural audiometry in addition to objective testing in order to evaluate the entire auditory system, and this behavioural test should be adapted for the child by obtaining multiple responses at the same intensities, using a test battery to cross-check results and altering the conditioned response according to the child's developmental level. Interpretation of results should be holistic based on the entire testing battery.

## Difficulty Obtaining Behavioural Responses in Neurocompromised Populations

In this study for those children for whom behavioural information was obtained, 72.7 % (16 out of 22 ears) differed from the objective results. Of these 16 ears, 11 (68.8 %) presented with a worse degree of hearing than was diagnosed on objective testing. This is in fitting with the finding that children at lower developmental ages respond less to low-intensity sounds, thus hearing will appear worse than the true threshold (Diefendorf & Gravel, 2000). In order to respond the child must attend to the stimulus, and the nature of this response will vary between children, both of which are dependent on the level of neuromaturation (Delaroche *et al*., 2004). Low-functioning children may not even show an observable orientation response required for VRA such as a head turn (Lancioni *et al.,* 1985). This difficulty in the child responding and the tester not recognizing the response would lead to hearing thresholds appearing worse than the child's actual hearing abilities. In this way,

results obtained in behavioural testing are associated to the developmental level of the child (Diefendorf & Gravel, 2000), and is emphasized by the findings in this study that behavioural thresholds differ from objective results.

More ears were diagnosed with a profound hearing loss on ABR testing than on behavioural testing in this sample (8 ears versus 3 ears respectively). None of these children diagnosed with a profound hearing loss objectively had any behavioural results. This could be a result of incorrect conditioning of the child behaviourally, leading to the tester aborting the test on the premise that the child "could not be conditioned", and is consistent with the finding that should a child with a profound hearing loss be conditioned below the ear threshold, this child will not learn the conditioned response (Madell, 2008). Therefore, if a child does not appear to condition under headphones at loud intensities, vibrotactile conditioning with the bone oscillator should be attempted to ascertain whether this lack of response is a potential hearing loss or a result of developmental delay (Madell, 2008). The fact that no child in this study diagnosed with a profound hearing loss objectively was able to be tested behaviourally emphasizes the importance of correct conditioning, as suggested by Madell (2008), and should be highlighted in the CHBAH behavioural protocol. All of these children with profound hearing loss barring one were diagnosed with CP, with the last child having a multiple diagnosis of meningitis and hydrocephalus. It is therefore possible that the more developmentally delayed children presented with the worst hearing abilities; however more investigation into this matter would be necessary to draw conclusive results.

The Audiology Department at CHBAH services a large population, thus time pressure may lead to audiologists aborting the test and sending the child for an ABR in order to save time. The audiologist may therefore not include the adaptations and correct conditioning techniques that allow a neurologically-disordered child the best opportunity to respond in a behavioural hearing test, rather choosing to cease behavioural attempts.

The amount of clinical experience of the tester is negligible in this study as, according to the HPCSA regulations of undergraduate curriculum for Audiology, all students should be proficient in testing neurocompromised children in order to graduate. The Government Gazette states that a requirement of graduation for a Degree in Audiology is the assessment of central and peripheral hearing status in persons of any age in view of their communication needs (HPCSA, 2012).

## Hearing Loss with Regard to Diagnosis

It is important to know the diagnosis of the child, as this can assist the audiologist in knowing if a hearing loss should be expected and what type of hearing loss to anticipate should this be synonymous with the diagnosis (Madell, 2008). The audiologist will also be able to gauge, by knowing the diagnosis, what to expect with speech, language and auditory behaviours, and to estimate the expected cognitive and developmental age of the child (Madell, 2008). Based on this knowledge, the test can be altered to account for the child's needs, allowing the child the best opportunity to undergo reliable behavioural testing. Objective results can also be analyzed taking these factors into account.

In this study sample, 43.75 % (n=35) of the ears tested presented with a hearing loss on objective testing. This indicates that hearing loss is more common in this neurologically-disordered population than in the general paediatric population where the incidence of hearing loss is three per 1000 live births, and emphasizes the necessity to conduct hearing testing on these patients.

In terms of CP, of the 10 subjects diagnosed with this condition, four patients presented with bilateral profound hearing losses on ABR testing, and one patient presented with a unilateral profound hearing loss. Three presented with normal hearing bilaterally, and two presented with a moderate sloping to severe bilateral hearing loss. Therefore 40 % of this population was diagnosed

with a bilateral profound hearing loss, and half of the patients tested had a profound loss in at least one ear. There is a dearth of information on the trends in hearing loss in children with CP, however as stated previously, Rosenbaum *et al*. (2007) found that these children can have any amount of decibel loss. Further investigation with a larger sample could examine if, as this study indicates, profound hearing loss is the most common degree of hearing loss in children with CP. Owing to the fact that the subtype of CP was not documented on the patient record no conclusion can be drawn as to whether different hearing losses are more common in different types of CP. Further investigation into this matter would be necessary to obtain this information.

Similarly, only one out of the five children (20 %) diagnosed with Down's Syndrome presented with normal hearing in this study. Hearing loss was expected in this population based on studies mentioned previously that have found that these children present with fluctuating conductive hearing loss owing to the frequent otitis media present in a majority of this population as a result of the craniofacial structural abnormalities pertinent in this syndrome. Muscle hypertrophy common of the syndrome and hypertrophy of the mid-face lead to dysfunction of the eustachian tube, which contributes to the higher prevalence of otitis media and eventual hearing loss in this population as stated previously (Austeng *et al*., 2013). In this study sample, therefore, 80 % of children with Down's Syndrome presented with a hearing loss. This percentage is higher than the 46.1% quoted in the study by Park *et al*. (2011) in Utah in America. This could be a result of some patients being diagnosed with normal hearing on screening measures in a diagnostic hearing evaluation. When VRA or CPA is not possible, screening is attempted with a screening Automated ABR and Otoacoustic Emmisions (OAEs). Patients passing this screening are discharged with a diagnosis of normal hearing and given a date for an annual hearing recheck. Patients referring the screening are booked for an ABR, therefore the population of children with Down's Syndrome presenting in the ABR clinic at CHBAH will have a higher chance of presenting with a hearing loss as only those referring initial hearing screening will be seen for an ABR.

This higher incidence of hearing loss in the Down's Syndrome subpopulation found in this study may also be in part attributed to the higher prevalence of CSOM in lower-resourced countries such as South Africa than in higher income countries where treatment and medical staff are more available (Mahadevan *et al.*, 2012). The global burden of disease for CSOM lies in Africa, South-East Asia and the Western Pacific (Mahadevan *et al*., 2012); therefore more children will present with CSOM in South Africa and will thus be at higher risk of developing a hearing loss.

Of the patients born prematurely in this sample, 54.5 % were diagnosed with a hearing loss on ABR testing. This is higher than the global prevalence of 5-10 % stated previously (Nour, 2012). However, this study by Nour (2012) included infants with prematurity only. Of the premature infants in this study only 3 had no concomitant conditions. The other 8 patients had other conditions that put them at risk of hearing loss, such as cerebral palsy, asphyxia, RVD and holoprosencephaly. Of these three patients with no concomitant conditions, two had a hearing loss. This could also be a result of the higher prevalence of CSOM mentioned above, as both these children had rising audiograms often associated with CSOM.

This higher prevalence of hearing loss in the premature population in this study emphasizes the need for Newborn Hearing Screening. Universal newborn screening, whereby every infant is screened after birth, is the gold standard; however a lack of resources has prevented the instigation of this screening in South Africa (Friderichs *et al*., 2012). High-risk or targeted screening should, based on the results of this study, include at least all infants born prematurely.

Prematurity, as indicated in Table 4, yielded the least amount of "no responses" in behavioural testing of any of the five other common pathologies. This indicates that these children are generally able to undergo behavioural testing, given extra time to allow for brain maturation. This finding is in agreement with Madell (2008), who states that premature infants should be tested in the same manner as term infants. However, given the higher prevalence of hearing loss in this premature population than that of the general population as found in this study, hearing losses would then be

diagnosed at a later stage if more time was allowed for maturation. Early intervention for those infants later diagnosed with hearing losses would then be less effective.

Although both patients diagnosed with HIV/AIDS presented with a hearing loss in this study, both of these patients had concomitant diagnoses that could have caused hearing loss. One patient had Cytomegalovirus and CP, both of which have been shown to put children at risk of a hearing loss (Swanepoel, 2007). The other patient was born prematurely, which is also a risk factor in hearing loss (Swanepoel, 2007).

Similarly, 43.4% of children with meningitis were found to have a sensorineural hearing loss of some degree post meningitis in Kenya, as reported by Karanja *et al* (2013). However, in this study three out of the five children with meningitis (or 60 %) were diagnosed with a hearing loss on ABR testing. One child had a bilateral profound hearing loss, however this child had hydrocephalus as well. The other two children had a mild bilateral hearing loss. This is higher than the prevalence stated by Karanja *et al*. (2013), thus further investigation would be necessary to establish the type of hearing loss (sensorineural or conductive) by bone conduction testing, and thus to establish if the loss is permanent or may resolve with medical intervention. The fact that 60 % of the study population with meningitis presented with hearing loss reiterates the need to test this population once a diagnosis of meningitis is received.

In terms of hydrocephalus, the number of children in this study with hydrocephalus presenting with a hearing loss (one out of seven, or 14 %) is far lower than the average of 83 % stated above by Spirakis and Hurley (2003) in Tampa in America. However, this study by Spirakis and Hurley (2003) found high frequency cochlear losses on the side of the shunt. In this study high frequencies were not tested as tone-burst ABR was conducted up to 4kHz. Further testing such as OAEs should therefore be conducted on these children in order to rule out a high frequency hearing loss. Dixon and Jones (2012) found at the University of Kentucky in the United States of America that relieving intracranial pressure helped the hearing loss to resolve in most cases. All of the children in this study

had a ventriculoperitoneal shunt, thus pressure-related hearing loss may have resolved in some of these children. These patients should therefore be tested after the shunt is removed to check for remediation of hearing loss.

It can be seen in Table 4 above that the three pathologies yielding the least behavioural results are those for which the highest percentage of hearing loss was diagnosed. These are Cerebral Palsy, RVD and Down's Syndrome. This therefore emphasizes the importance of these children having their hearing tested, and the use of objective testing in obtaining results.

## HPCSA Risk Factors of Hearing Loss

As mentioned previously, the HPCSA have identified children with the following conditions to be at risk of developing a hearing loss, and should therefore receive a hearing screen (Swanepoel, 2007):

- An illness or condition requiring admission of 48 hours or greater to a NICU
- Craniofacial abnormalities
- In-utero infection such as cytomegalovirus, HIV
- Postnatal infections associated with sensorineural hearing loss including bacterial meningitis

For this study sample, out of the 40 patients selected, only 14 would have been at-risk if the above criteria were utilized. Very few of the patients born prematurely and those with birth asphyxia would fit the criteria of a 48 hour NICU admission. Most of these patients would have been moved out of the NICU as soon as they are stabilized and transferred to a high-care ward, thus not receiving hearing screening. Similarly, a minority of those patients diagnosed with cerebral palsy would have been screened based on the HPCSA High Risk Register, as this condition would only manifest once the child is older and begins to fall behind motor milestones.

Nine of the 26 patients with conditions not recognized by the at-risk register were found to have a hearing loss in this sample, thus 22.5% of children with a hearing loss diagnosed in this study sample would not have received a hearing screen. These were children with CP, prematurity and birth asphyxia. This indicates the need to revise the criteria for targeted screening, including all patients with CP and/or asphyxia as well as those patients born prematurely.

## Early Intervention and Neurological Disorder

It has been shown above that hearing loss is more prevalent in populations with neurological disorder than in the general paediatric population. It has similarly been proven that most of these patients are diagnosed with objective measures, as behavioural testing is unreliable in these children. According to the HPCSA in their position statement (Swanepoel, 2007), the guidelines for early intervention of hearing loss services in South Africa for hospital-based care are as follows:

- Patients at risk should receive hearing screening before 28 days of age
- Those referring on screening should receive diagnostic testing and a confirmation of hearing loss should be made before 3 months of age
- The child should be enrolled in an early intervention programme before 6 months of age

In this study, not one patient met the criteria above. For many of these children behavioural testing was attempted two or three times before a referral to the ABR clinic. This would delay the confirmation of hearing loss by a number of months.

It is therefore suggested that all patients with neurological disorders are screened as soon as a diagnosis is made. Those children that do not pass hearing screening should be referred directly for objective testing in order for habilitation to begin. Behavioural testing should then be attempted, as

this is the only measure that gives an indication of the functioning of the entire auditory system from the outer ear to the auditory cortex (Delaroche *et al*., 2011). The child should be conditioned over a number of appointments with the above-mentioned adaptations to the conventional behavioural test in order to meet their individual developmental needs. However, this should be supplementary to enrollment in an early intervention programme in order to afford the child the best possible language and communication development resulting from early habilitation. Once behavioural results are obtained, amplification devices should be reprogrammed based on these results.

# 5. Conclusion

Hearing loss is more prevalent in the paediatric population diagnosed with neurological disorders than the in general paediatric population. These neurologically-disordered children are more difficult to test with behavioural audiometry owing to their diagnoses. Behavioural thresholds are not possible in many members of this population, and those results obtained may be unreliable and lead to an incorrect diagnosis of the child's hearing status. Therefore, objective testing via tone-burst ABR is very important in this population, and should be included in the initial hearing test battery for any children diagnosed with a neurological disorder. Aural habilitation should begin based on the results of objective testing. All children with a neurological disorder should receive hearing screening, and those children who do not pass the screen should immediately receive electrophysiological testing. Behavioural audiometry should not be ignored, but continually attempted with adaptations for the child's developmental level in order to assess the entire auditory pathway up to and including the auditory cortex. Habilitation programmes should be based on objective measures initially, however once behavioural results are obtained the programme should be based on these, as this remains the gold standard and gives information of the entire auditory pathway.

# 6. Recommendations and Implications for the Study

It can be seen above that neurological disorders are more common in developing countries such as South Africa than in higher-resourced, developed countries. Moreover, hearing loss is more common in neurologically-disordered children than in the general paediatric population. This study has shown that, owing to the fact that a majority of children with the above-mentioned pathologies presented with hearing loss, all children should be referred for a hearing test once a diagnosis of a neurological disorder is received.

In order to test hearing, the results of these children should be based on both objective and behavioural test battery results. This study has shown that behavioural testing in isolation is of limited value, and will only be able to be conducted once the child is of a sufficient developmental age to understand the test. Objective testing should therefore be conducted in order to ascertain the status of the child's peripheral hearing system from the outer ear up to and including the eighth cranial nerve. Enrollment in a habilitation programme should be based on these results. The child should continually be conditioned for a developmentally-appropriate behavioural test battery, with alterations to encompass the child's unique abilities, in order to assess the entire auditory system up to the auditory cortex.

All children with a neurological disorder should have access, therefore, to both behavioural and objective testing. In District and Rural hospitals in South Africa there is currently a dearth of both, with a majority of the caseload falling on Tertiary hospitals such as CHBAH. Therefore, more appropriately-trained audiologists and relevant equipment should be allocated to District and Rural hospitals in order to afford children in these areas the opportunity for hearing and communication habilitation.

It is thus recommended that Universal Newborn Hearing Screening be implemented. Failing this, due to a lack of resources, all children diagnosed with a neurological disorder should immediately be

referred for a hearing test. This should comprise of both behavioural and objective testing, thus all equipment for these tests (namely diagnostic audiometers, VRA conditioning toy boxes, speakers for freefield testing, immittance equipment and an electrophysiological testing unit) should be available to the audiology department. Audiologists and the above-mentioned equipment should be allocated to hospitals of all levels ensuring that no patient is without the option of a hearing test.

Further research should be conducted into the typical responses in behavioural audiometry by children with neurological disorders. By knowing what responses to expect, the clinician will be able to garner as much behavioural information as possible, thereby gaining more insight into the child's auditory processing capabilities.

Research should also be conducted into exact alterations that could be made to the behavioural test battery according to the child's diagnosis. These alterations would make it more likely that the child would respond, and lead to more behavioural information being obtained.

# 7. Limitations of the Study

This study comprised of only 40 subjects. Some of the pathologies studied were present in only one or two of the subjects, thus it cannot be generalized to the population. Hydrocephalus and CP were utilized as “blanket terms” for these conditions, and children in the different sub-groups of these pathologies were not separated. Similarly, the nature of the data and the small number of subjects resulted in the ABR records not being viable for statistical analysis.

Bone conduction testing results were not available for many children. Many of the subjects were diagnosed with a mild hearing loss in the presence of abnormal Type B tympanometry, indicating middle ear effusion. According to the CHBAH protocol, these children would have been sent for ENT management and booked for a recheck assessment in 8 – 10 weeks, thus bone conduction testing was rarely conducted. Tympanometric results were not included in the study, so it cannot be stated whether subjects in this sample present with a permanent or a temporary hearing loss. Bone conduction cannot be tested on the Eclipse equipment at intensities exceeding 40 – 50 dBnHL, owing to stimulus artifact. Therefore, for those children presenting with profound hearing losses, bone conduction testing would have most likely yielded no identifiable waveforms at maximum stimulus levels. For these children it is assumed that the type of hearing loss is sensorineural, however this would not be able to be confirmed objectively.

The cohort of children in this study was mostly affected by more than one neurological disorder. For these children with multiple disabilities, it is difficult to report on the prevalence of hearing loss per pathology as found in this study as it is confounded by the multiple handicaps.

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# 9. Appendices

## Appendix 1: Severity of Hearing Loss

| **Degree of Hearing Loss** | **Pure Tone Average (dBHL)** |
|---|---|
| Normal Hearing | < 15 |
| Slight Hearing Loss | 16 – 25 |
| Mild Hearing Loss | 26 – 40 |
| Moderate Hearing Loss | 41 - 55 |
| Moderately-severe Hearing Loss | 56 – 70 |
| Severe Hearing Loss | 71 – 90 |
| Profound Hearing Loss | > 90 |

Reproduced from *Typically Used Categories to Describe the Degree of Hearing Loss based on the Pure-Tone Average* (p. 144) by Gelfand (2009).

## Appendix 2: Correction Factors for ABR Testing

The following correction factors were utilized in ABR testing in order to convert the threshold obtained in the ABR (which is in dBnHL) to an estimated hearing level in dBeHL.

| | **Click** | **500 Hz** | **1000 Hz** | **2000 Hz** | **4000 Hz** |
|---|---|---|---|---|---|
| Under 12 weeks corrected age | -5 | -15 | -10 | -5 | 0 |
| 12 - 24 weeks corrected age (AC & BC) | 0 | -20 | -15 | -10 | -5 |
| Over 24 weeks corrected age (AC & BC) | - 5 | -20 | -15 | -10 | -10 |

*Correction Values for Air Conduction ABR under Insert Earphones*

Adapted from:

*Guidelines for early audiological assessment and management of babies referred from the newborn hearing screening programme (Version2.5)* by Stevens *et al* (2011).

## Appendix 3: Letter to the Ethics Committee at Chris Hani Baragwanath Academic Hospital Requesting Permission to Use Patient Data

**CHRIS HANI BARAGWANATH ACADEMIC HOSPITAL**
**SPEECH THERAPY AND AUDIOLOGY DEPARTMENT**
**(011) 933 9263/4/5**
**P O Bertsham 2013**

1 February 2012

To: Ethics Committee
Chris Hani Baragwanath Academic Hospital

CC: Mrs. Naik

To Whom it May Concern,

My name is Karen Baillieu. I am a senior audiologist in the Speech Therapy and Audiology Department at Chris Hani Baragwanath Academic Hospital (PERSAL number 8298 7831). I am currently enrolled in an MSc Med degree in Child Health and Neurodevelopment at the University of the Witwatersrand (Student number 516 855). For this degree a research study must be done. I have chosen to conduct my study on the findings on the ABR test of neurologically impaired children. The title of the study is:

**Auditory Brainstem Response (ABR) Findings in a Group of Neurologically Compromised Children: A Retrospective Study.**

The study aims to describe the results of this test for children with diagnosed neurological and/or neurdevelopmental impairments. The ABR test is an objective test of hearing conducted on newborn and difficult-to-test patients in order to ascertain their hearing status, as no other option of hearing testing is available to these populations. The waveform morphology, replicability, absolute latencies and interwave latencies will be described for each patient. The audiological findings will also be described for each patient, which will also be correlated to behavioural audiological information (where available).

Each patient will be allocated a number, thus the results will be anonymous. The study is a retrospective literature review, whereby results of ABR tests conducted between September 2010 and March 2012 in the Audiology Department of Chris Hani Baragwanath Academic Hospital will be analyzed. The patient will have undergone the ABR test for assessment of hearing abilities (not for research purposes), therefore no extra procedures will be necessary for the study and no harm will be caused to the patient.

In order to conduct this study, I require access to patient records of testing conducted in the Speech Therapy and Audiology Department, specifically the ABR test data and where available the behavioural results.

I am requesting permission to conduct the study in the Audiology Department of Chris Hani Baragwanath Academic Hospital, and to have access to the results of the tests and to utilize them anonymously for the study.

Please find attached the proposal submitted and accepted by the Postgraduate Review Committee of the University of the Witwatersrand. Ethical approval has been submitted to the Ethics Committee of the University, and is still pending.

Karen Baillieu
Senior Audiologist

## Appendix 4: Letter of Permission from the Head of Department of Audiology at Chris Hani Baragwanath Academic Hospital

**GAUTENG PROVINCE**
HEALTH
REPUBLIC OF SOUTH AFRICA

Chris Hani Baragwanath hospital
Department of Speech Therapy/Audiology
PO Bertsham
2013
22 May 2012
011 933 9269
mwsadna@mweb.co.za

Ms Karen Baillieu

**Re Study : Auditory Brainstem Response Findings in a Group of Neurologically Compromised Children: A retrospective study**

Thank you for your application to conduct your research in our department, after discussion with the Senior Clinical Executive Ms Naik permission is granted for you to conduct your study at the Audiology Department at Chris Hani Baragwanath Academic Hospital. Please inform me about your starting date as soon as this confirmed.

Kind Regards

Balta

Dr Sadna Balton (Head of Department-Speech Therapy/Audiology)

## Appendix 5: Medical Ethics Committee Clearance Certificate

Ethical clearance was obtained from the Human Research Ethics Committee (Medical) at the University of the Witwatersrand on 24.02.2012.

The Ethical Clearance Certificate for Protocol Number M120217 was obtained.

**UNIVERSITY OF THE WITWATERSRAND, JOHANNESBURG**
Division of the Deputy Registrar (Research)

**HUMAN RESEARCH ETHICS COMMITTEE (MEDICAL)**
R14/49 Ms Karen Baillieu

| | |
|---|---|
| **CLEARANCE CERTIFICATE** | **M120217** |
| **PROJECT** | Auditory Brainstem Response Findings in a Group of Neurologically Compromised Children: A Retrospective Study |
| **INVESTIGATORS** | Ms Karen Baillieu. |
| **DEPARTMENT** | Department of Paediatrics & Child Health |
| **DATE CONSIDERED** | 24/02/2012 |
| **DECISION OF THE COMMITTEE*** | Approved unconditionally |

**Unless otherwise specified this ethical clearance is valid for 5 years and may be renewed upon application.**

**DATE** 24/02/2012 **CHAIRPERSON** .....
(Professor PE Cleaton-Jones)

*Guidelines for written 'informed consent' attached where applicable
cc: Supervisor : Prof Lorna Jacklin

---

**DECLARATION OF INVESTIGATOR(S)**
To be completed in duplicate and **ONE COPY** returned to the Secretary at Room 10004, 10th Floor, Senate House, University.
I/We fully understand the conditions under which I am/we are authorized to carry out the abovementioned research and I/we guarantee to ensure compliance with these conditions. Should any departure to be contemplated from the research procedure as approved I/we undertake to resubmit the protocol to the Committee. **I agree to a completion of a yearly progress report.**

***PLEASE QUOTE THE PROTOCOL NUMBER IN ALL ENQUIRIES...***

## Appendix 6: Spreadsheet Containing a Summary of All Data, Per Participant

| Patient No | D.O.B | Age (months) | Diagnosis |
|---|---|---|---|
| 1 | 02.01.2011 | 11 | Down Syndrome |
| 2 | 22.05.2007 | 53 | CP |
| 3 | 25.03.2011 | 7 | Chromosomal Abnormality |
| 4 | 01.11.2010 | 11 | SMA, RVD, Prematurity |
| 5 | 01.03.2011 | 5 | Prematurity, Asphyxia |
| 6 | 28.05.2009 | 31 | CP |
| 7 | 08.05.2010 | 11 | Prematurity, Asphyxia, CP |
| 8 | 14.03.2011 | 7 | Asphyxia |
| 9 | 17.02.2010 | 22 | Unknown Syndrome |
| 10 | 08.03.2011 | 6 | Prematurity, Asphyxia |
| 11 | 29.08.2007 | 52 | CP |
| 12 | 22.01.2007 | 54 | Unknown Syndrome, TB |
| 13 | 31.12.2007 | 47 | Butler Syndrome |
| 14 | 14.06.2007 | 47 | CP, cerebral atrophy, asphyxia |
| 15 | 21.01.2009 | 28 | Trisomy 18, cleft lip and palate |
| 16 | 30.12.2009 | 25 | Hydrocephalus, developmental delay |
| 17 | 08.05.2010 | 22 | Hydrocephalus, blindness |
| 18 | 17.06.2011 | 8 | Meningitis |
| 19 | 01.08.2011 | 7 | Prematurity, asphyxia |
| 20 | 29.01.2005 | 84 | CP, microcephaly |
| 21 | 28.07.2011 | 5 | Prematurity |
| 22 | 18.07.2010 | 19 | Down Syndrome |
| 23 | 19.12.2008 | 39 | Autism |
| 24 | 15.04.2011 | 7 | Prematurity |
| 25 | 12.10.2009 | 29 | Prematurity, holoprosencencephaly |
| 26 | 19.08.2011 | 5 | Down Syndrome |
| 27 | 04.08.2010 | 22 | Corpus callosum agenesis, cleft lip and palate |
| 28 | 11.07.2011 | 5 | Prematurity |
| 29 | 06.03.2008 | 48 | CP, seizures |
| 30 | 04.05.2010 | 22 | CP, CMV, RVD |
| 31 | 07.07.2002 | 115 | Meningitis, hydrocephalus |
| 32 | 01.12.2008 | 39 | Hydrocephalus |
| 33 | 27.08.2011 | 6 | Meningitis |
| 34 | 27.12.2010 | 13 | Hydrocephalus, spina bifida, meningitis, prem |
| 35 | 25.12.2006 | 62 | Down Syndrome |
| 36 | 12.05.2011 | 7 | Down Syndrome |
| 37 | 14.03.2007 | 60 | Hydrocephalus, meningitis |
| 38 | 01.07.2009 | 31 | CP |
| 39 | 09.07.2009 | 31 | Hydrocephalus |
| 40 | 03.03.2010 | 22 | CP, prematurity |

| | *Left Ear ABR* | | | | *Right Ear ABR* | | | |
|---|---|---|---|---|---|---|---|---|
| **Patient No** | **500 Hz** | **1 000 Hz** | **2 000 Hz** | **4 000 Hz** | **500 Hz** | **1 000 Hz** | **2 000 Hz** | **4 000 Hz** |
| 1 | | 55 | | 30 | | 55 | | 30 |
| 2 | NR 80 | NR 85 | NR 90 | NR 80 | NR 80 | NR 85 | NR 90 | NR 80 |
| 3 | | | | 0 | | | | <10 |
| 4 | 70 | | | 5 | 50 | | | 10 |
| 5 | | 25 | | 20 | | <15 | | <15 |
| 6 | | 35 | | 25 | | 70 | | 40 |
| 7 | | 25 | | 50 | 50 | | | 85 |
| 8 | | | | 0 | | | | -5 |
| 9 | <25 | <25 | <25 | <25 | | 30 | | <20 |
| 10 | 25 | <15 | <20 | <20 | 25 | <15 | <20 | <20 |
| 11 | | | | <20 | | | | <20 |
| 12 | | <25 | | <25 | | <25 | | 35 |
| 13 | | | | <10 | | | | <10 |
| 14 | <20 | <20 | <15 | <20 | | | | |
| 15 | | | | <30 | | | | |
| 16 | | <25 | | <25 | | | | <25 |
| 17 | <30 | 25 | | <20 | <30 | <15 | | <25 |
| 18 | | <20 | | <20 | | <25 | | <25 |
| 19 | | | 50 | 65 | | | 25 | 60 |
| 20 | NR 75 | NR 85 | NR 90 | NR 85 | NR 75 | NR 85 | | NR 85 |
| 21 | NR 80 | 85 | 80 | 45 | 70 | 60 | 40 | <25 |
| 22 | | | | 50 | | | | |
| 23 | | <25 | | <25 | | <25 | | <20 |
| 24 | <25 | <25 | | <25 | <25 | <25 | | <25 |
| 25 | | | | <20 | | | | <20 |
| 26 | 35 | <25 | | <25 | <25 | <25 | | <25 |
| 27 | | <25 | | <25 | | <25 | <25 | <20 |
| 28 | 40 | 75 | 65 | 65 | | <15 | <30 | 25 |
| 29 | | <15 | | <20 | | <30 | | <30 |
| 30 | | | | 35 | | NR 85 | NR 90 | NR 85 |
| 31 | | NR 85 | 90 | NR 85 | | NR 85 | 80 | 70 |
| 32 | | | | <30 | | | | |
| 33 | | 35 | | 30 | | 50 | 40 | 35 |
| 34 | | <30 | | <30 | | <35 | | <40 |
| 35 | <60 | 40 | 40 | <25 | | 30 | 30 | <25 |
| 36 | <20 | <25 | | <25 | | <25 | | <25 |
| 37 | | <35 | | <20 | | | | <30 |
| 38 | 80 | NR 85 | NR 90 | NR 85 | 100 | NR 100 | NR 100 | NR 95 |
| 39 | 25 | <25 | 30 | <25 | <25 | <25 | 30 | <25 |
| 40 | NR 80 | NR 85 | NR 90 | NR 85 | NR 80 | NR 85 | 90 | NR 85 |

| | *Left Ear Behavioural* | | | | *Right Ear Behavioural* | | | |
|---|---|---|---|---|---|---|---|---|
| **Patient No** | **500 Hz** | **1 000 Hz** | **2 000 Hz** | **4 000 Hz** | **500 Hz** | **1 000 Hz** | **2 000 Hz** | **4 000 Hz** |
| 1 | | | | | | | | |
| 2 | | | | | | | | |
| 3 | | | | | | | | |
| 4 | | | | | | | | |
| 5 | | | | | | | | |
| 6 | NR 115 | NR 115 | NR 115 | NR 110 | NR 110 | NR 115 | NR 110 | NR 105 |
| 7 | 35 | | | 15 | 10 | 10 | 10 | |
| 8 | | | | | | | | |
| 9 | | | | | 80 | 85 | 85 | 85 |
| 10 | | | | | | | | |
| 11 | | | | | | | | |
| 12 | | | | | | | | |
| 13 | 40 | 40 | 40 | 40 | 40 | 40 | 40 | 40 |
| 14 | | | | | | | | |
| 15 | | | | | 100 | | 55 | |
| 16 | | | | | | | | |
| 17 | | | | | | | | |
| 18 | | | | | | 40 | 40 | |
| 19 | | | | | | | | |
| 20 | | | | | | | | |
| 21 | | | | | | | | |
| 22 | CNT | CNT | CNT | CNT | CNT | CNT | CNT | CNT |
| 23 | | 25 | | | 25 | 30 | 25 | 25 |
| 24 | | | | | | | | |
| 25 | | | | | | | | |
| 26 | CNT | CNT | CNT | CNT | CNT | CNT | CNT | CNT |
| 27 | | | | | | | | |
| 28 | | | | | | | | |
| 29 | CNT | CNT | CNT | CNT | | 50 | 40 | 50 |
| 30 | CNT | CNT | CNT | CNT | CNT | CNT | CNT | CNT |
| 31 | | | | | | | | |
| 32 | 70 | 60 | 55 | | 60 | 60 | 50 | 30 |
| 33 | 20 | | 20 | 20 | 30 | 20 | | 20 |
| 34 | | | | | | | | |
| 35 | CNT | CNT | CNT | CNT | CNT | CNT | CNT | CNT |
| 36 | | | | | | | | |
| 37 | CNT | CNT | CNT | CNT | CNT | CNT | CNT | CNT |
| 38 | | | | | | | | |
| 39 | | | | | | | | |
| 40 | | | | | | | | |

| | *Free Field* | | | |
|---|---|---|---|---|
| **Patient No** | **500 Hz** | **1000 Hz** | **2000 Hz** | **4000 Hz** |
| 1 | | | | |
| 2 | | | | |
| 3 | | | | |
| 4 | | | | |
| 5 | | 25 | 25 | |
| 6 | | | | |
| 7 | | | | |
| 8 | | | | |
| 9 | | | | |
| 10 | | | | |
| 11 | | | | |
| 12 | | <25 | | <25 |
| 13 | | | | |
| 14 | | | | |
| 15 | | | | |
| 16 | 20 | 20 | 20 | 20 |
| 17 | | | | |
| 18 | | | | |
| 19 | | | | |
| 20 | | | | |
| 21 | 30 | 25 | 25 | 25 |
| 22 | | | | |
| 23 | | | | |
| 24 | 30 | 30 | 30 | 30 |
| 25 | | | | |
| 26 | | | | |
| 27 | | | | |
| 28 | | | | |
| 29 | | | | |
| 30 | | | | |
| 31 | | | | |
| 32 | | | | |
| 33 | | | | |
| 34 | | | | |
| 35 | | | | |
| 36 | | | | |
| 37 | | | | |
| 38 | | | | |
| 39 | | 85 | 90 | |
| 40 | | | | |

| Patient No | D.O.B | Age (months) | Diagnosis |
|---|---|---|---|
| 1 | 02.01.2011 | 11 | Down Syndrome |
| 2 | 22.05.2007 | 53 | CP |
| 3 | 25.03.2011 | 7 | Chromosomal Abnormality |
| 4 | 01.11.2010 | 11 | SMA, RVD, Prematurity |
| 5 | 01.03.2011 | 5 | Prematurity, Asphyxia |
| 6 | 28.05.2009 | 31 | CP |
| 7 | 08.05.2010 | 11 | Prematurity, Asphyxia, CP |
| 8 | 14.03.2011 | 7 | Asphyxia |
| 9 | 17.02.2010 | 22 | Unknown Syndrome |
| 10 | 08.03.2011 | 6 | Prematurity, Asphyxia |
| 11 | 29.08.2007 | 52 | CP |
| 12 | 22.01.2007 | 54 | Unknown Syndrome, TB |
| 13 | 31.12.2007 | 47 | Butler Syndrome |
| 14 | 14.06.2007 | 47 | CP, cerebral atrophy, asphyxia |
| 15 | 21.01.2009 | 28 | Trisomy 18, cleft lip and palate |
| 16 | 30.12.2009 | 25 | Hydrocephalus, developmental delay |
| 17 | 08.05.2010 | 22 | Hydrocephalus, blindness |
| 18 | 17.06.2011 | 8 | Meningitis |
| 19 | 01.08.2011 | 7 | Prematurity, asphyxia |
| 20 | 29.01.2005 | 84 | CP, microcephaly |
| 21 | 28.07.2011 | 5 | Prematurity |
| 22 | 18.07.2010 | 19 | Down Syndrome |
| 23 | 19.12.2008 | 39 | Autism |
| 24 | 15.04.2011 | 7 | Prematurity |
| 25 | 12.10.2009 | 29 | Prematurity, holoprosencencephaly |
| 26 | 19.08.2011 | 5 | Down Syndrome |
| 27 | 04.08.2010 | 22 | Corpus callosum agenesis, cleft lip and palate |
| 28 | 11.07.2011 | 5 | Prematurity |
| 29 | 06.03.2008 | 48 | CP, seizures |
| 30 | 04.05.2010 | 22 | CP, CMV, RVD |
| 31 | 07.07.2002 | 115 | Meningitis, hydrocephalus |
| 32 | 01.12.2008 | 39 | Hydrocephalus |
| 33 | 27.08.2011 | 6 | Meningitis |
| 34 | 27.12.2010 | 13 | Hydrocephalus, spina bifida, meningitis, prem |
| 35 | 25.12.2006 | 62 | Down Syndrome |
| 36 | 12.05.2011 | 7 | Down Syndrome |
| 37 | 14.03.2007 | 60 | Hydrocephalus, meningitis |
| 38 | 01.07.2009 | 31 | CP |
| 39 | 09.07.2009 | 31 | Hydrocephalus |
| 40 | 03.03.2010 | 22 | CP, prematurity |

| Patient No | *Hearing Result Objective RE* | *Hearing Result Objective LE* |
|---|---|---|
| 1 | Moderately-severe to mild HL | Moderately-severe to mild HL |
| 2 | Profound HL | Profound HL |
| 3 | Normal hearing | Normal hearing |
| 4 | Moderately-severe to mild HL | Moderately-severe to mild HL |
| 5 | Normal hearing | Normal hearing |
| 6 | Severe rising to moderate HL | Mild HL |
| 7 | Moderate sloping to severe HL | Moderate high frequency HL |
| 8 | Normal hearing | Normal hearing |
| 9 | Normal hearing | Normal hearing |
| 10 | Normal hearing | Normal hearing |
| 11 | Normal hearing | Normal hearing |
| 12 | Normal hearing | Normal hearing |
| 13 | Normal hearing | Normal hearing |
| 14 | Normal hearing | Normal hearing |
| 15 | Normal hearing | Normal hearing |
| 16 | Normal hearing | Normal hearing |
| 17 | Normal hearing | Normal hearing |
| 18 | Normal hearing | Normal hearing |
| 19 | Moderate sloping to severe high freq HL | Moderate sloping to moderately-severe HL |
| 20 | Profound HL | Profound HL |
| 21 | Severe rising to normal | Profound rising to moderately-severe HL |
| 22 | Moderately-severe HL | Moderately-severe HL |
| 23 | Normal hearing | Normal hearing |
| 24 | Normal hearing | Normal hearing |
| 25 | Normal hearing | Normal hearing |
| 26 | Mild low frequency HL | Mild low frequency HL |
| 27 | Normal hearing | Normal hearing |
| 28 | Normal hearing | Moderate sloping to severe HL |
| 29 | Normal hearing | Normal hearing |
| 30 | Profound HL | Mild HL |
| 31 | Profound rising to severe HL | Profound HL |
| 32 | Normal hearing | Normal hearing |
| 33 | Moderate rising to mild HL | Mild HL |
| 34 | Mild HL | Mild HL |
| 35 | Mild to moderate HL | Mild to moderate HL |
| 36 | Normal hearing | Normal hearing |
| 37 | Normal hearing | Normal hearing |
| 38 | Severe to Profound HL | Severe to Profound HL |
| 39 | Normal hearing | Normal hearing |
| 40 | Profound HL | Profound HL |

| **Patient No** | ***Hearing Result Behavioural RE*** | ***Hearing Result Behavioural LE*** |
|---|---|---|
| 1 | None | None |
| 2 | None | None |
| 3 | None | None |
| 4 | None | None |
| 5 | Normal hearing | None |
| 6 | Profound HL | Profound HL |
| 7 | Normal hearing | Normal hearing |
| 8 | None | None |
| 9 | Profound HL | None |
| 10 | None | None |
| 11 | None | None |
| 12 | Normal hearing | None |
| 13 | Moderate hearing loss | Moderate hearing loss |
| 14 | None | None |
| 15 | Profound rising to moderately-severe HL | None |
| 16 | Normal hearing | None |
| 17 | None | None |
| 18 | Moderate hearing loss | None |
| 19 | None | None |
| 20 | None | None |
| 21 | Normal hearing | None |
| 22 | None | None |
| 23 | Normal hearing | Normal hearing |
| 24 | Normal hearing | None |
| 25 | None | None |
| 26 | None | None |
| 27 | None | None |
| 28 | None | None |
| 29 | Moderate hearing loss | None |
| 30 | None | None |
| 31 | None | None |
| 32 | Moderately-severe rising to mild HL | Severe rising to moderately-severe HL |
| 33 | Normal hearing | Normal hearing |
| 34 | None | None |
| 35 | None | None |
| 36 | None | None |
| 37 | None | None |
| 38 | None | None |
| 39 | Severe to profound HL | None |
| 40 | None | None |