The Prevalence and Perceptions of Hearing Loss in Individuals Diagnosed with Adult Onset Motor Neuron Disease (MND)

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Declaration

I, Elena Philippou, hereby declare that this research report is my own work except as indicated in the references and acknowledgements. I am responsible for the content of this study and the conclusions presented. No part of this research report has been previously submitted for a degree at any other University/Institution.

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Abstract

Although it is well-known that motor neuron disease (MND) primarily affects motor neurons, the involvement of sensory pathways in the disease is currently receiving more attention. There is a dearth of information regarding the atypical effects of MND, resulting in limited understanding of the vulnerability of for example the auditory system.

The presence of hearing loss negatively impacts on participation across all communicative contexts, stripping individuals of autonomy and self-worth, ultimately resulting in withdrawal and isolation. These factors form the foundation for individual desire to pursue life-prolonging measures. Hearing loss, combined with dysarthria and the use of augmentative and alternative communicative strategies, implies that individuals with MND require additional support to meet their daily communicative needs.

This descriptive, exploratory study aimed to identify the prevalence of hearing loss in eight individuals with adult onset MND. In addition, perceptions relating to the implications of auditory impairment and value of auditory diagnosis were explored.

An evaluation of auditory function was performed on eight individuals with a neurologist confirmed diagnosis of MND. Auditory function was assessed using a comprehensive audiological test battery including both objective and subjective measures. Perceptions related to auditory impairment were determined using the Hearing Handicap Inventory for Adults (HHIA) and the Hearing Experience Questionnaire. Both individuals with MND and their primary caregivers completed the Hearing Experience Questionnaire.

The results of the study indicate that a high frequency sensorineural hearing loss was identified in six participants. Auditory handicap, as measured by the Hearing Handicap Inventory for Adults, was reported in four participants, with social handicaps reported more than emotional handicaps. Individuals with MND and their caregivers identified communication as the most important functional skill. Interestingly, the caregivers related more to the threats auditory impairment than individuals with MND.

The nature of hearing loss identified in this study mimics the pattern of a presbycustic (age-related) hearing loss. It is postulated that hearing loss may arise during disease course. Participants' limited understanding of the devastating consequences of hearing loss on quality of life highlights the need for inclusion of an audiologist as part of the multidisciplinary management team in MND. Audiological assessment, management, counseling and education will serve to guide the process of sensory regulation and limit psychosocial threats posed by MND. This will in turn promote enhanced quality of life and maintenance of individual autonomy.

Key Words: atypical symptoms; hearing loss; motor neuron disease; perceptions; sensory regulation; socio-emotional consequences

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Definition of Terms

Amyotrophic Lateral Sclerosis.

Amyotrophic Lateral Sclerosis (ALS) is the most common form of adult onset motor neuron degeneration, typically presenting in adults during the fourth to sixth decade of life. ALS commonly referred to as Lou Gerich's disease leads to upper and lower motor neuron signs. It is often used synonymously with MND (McLeod & Clarke, 2007). Clinical markers include spasticity and hyperflexia as well as progressive muscle weakness and muscle wasting as a consequence of upper and lower motor neuron degeneration respectively (Van Damme & Robberecht, 2009).

Note: The terms ALS and MND will be used interchangeably throughout this research report.

Augmentative and Alternative Communication.

Augmentative and alternative communication (AAC) is the term used to describe communication that is not speech based (Smith, 2005). It may be classified as aided, relying on an external device such as alphabet boards, computers; or unaided, relying on the individual's body e.g. eye tracking, gestures. AAC serves to replace or complement natural speech and/or writing (Lloyd, Fuller & Arvidson, 1997; Smith, 2005). AAC is used to enhance the communicative abilities of individuals presenting with expressive communicative difficulties as a result of diseases, such as stroke and MND.

Communication.

Communication refers to the process in which individuals are able to exchange, share and request information regarding individual needs, desires, perceptions and knowledge. Communication is a shared process between individuals that may be verbal (such as speech) and/or nonverbal (such as gesture and facial expression) (Duffy, 2005; Ross &Deverell, 2004). Successful communication is integral to quality health care and successful maintenance of social inclusion (Baladin et al., 2001).

Note: For the purpose of this study, 'communication' relating to the Hearing Experience Questionnaire will refer to expressive communicative abilities alone.

Conductive Hearing Loss.

A conductive hearing loss (CHL) is impairment in hearing that arises as a result of obstruction or damage along the outer and/or middle ear pathways. This prevents the sound signal from being effectively transmitted at any one of the following structural regions of the outer or middle ear: the pinna, the external auditory canal, the tympanic membrane, the middle ear ossicles (malleus, incus and stapes) (Marieb, 2001). A CHL may arise as a result of obstruction (e.g. cerumen, foreign body), infection (e.g. otitis media), structural damage (e.g. tympanic membrane perforation) or head trauma (e.g. ossicular discontinuity). A CHL is frequently reversed through medical and surgical procedures (Musiek & Baran, 2007).

Dysarthria.

Dysarthria is a neurologically-based motor speech disorder characterized by impaired movement of the articulators, phonation, resonance and control of respiration (Duffy, 2005). In progressive neurological disease such as MND a gradual loss of speech ability occurs with increasing severity alongside disease progression. This results in diminished expressive communicative abilities and ultimately a loss of verbal expressive abilities (Duffy, 2005).

Dysphagia.

Dysphagia is a disorder of swallowing, which occurs mainly in the elderly however can arise as a consequence of muscular weakness and/or in-coordination linked to stroke and/ or neurodegenerative disease (Marieb, 2001). Dysphagia presents with a series of physical, social and psychological consequences. Dysphagia is a high risk factor for food aspiration, which often leads to pneumonia and death (Ekberg, Hamdy, Woisard, Wuttge-Hanning, & Ortega., 2002).

Deglutition.

Deglutition refers to the action or the process of swallowing. This includes the oral stage whereby food is essentially prepared for the swallow (oral preparatory and oral transport), pharyngeal phases where food passes through the oropharynx leading towards the oesophagus and laryngeal musculature constrict to prevent aspiration and the oesophageal phases where food is passed into the stomach region (Cherney, 1994). A disruption of any of these phases places an individual at risk for aspiration, posing the risk of pneumonia and possible death (Ekberg et al., 2002).

Hyperacusis.

Hyperacusis defines a significant intolerance to environmental sounds and is a subjective experience that cannot be quantified by objective measures (Khalfa, Veuillet, Grima, Bazin, & Collet, 1999). It involves a decreased threshold of discomfort from sound and refers to individual discomfort caused by sounds that were previously tolerable to the listener and are tolerable to other listeners with normal hearing (Hesse, Hasri, Nelting, & Brehmer, 1999).

International Classification of Functioning, Disability and Health.

The International Classification of Functioning, Disability and Health (ICF) is a biopsychosocial approach to health, viewing disability and functioning as the outcomes of interactions between health conditions, environmental- and personal factors (Gagne, Jennings, & Southall, 2009). This is an interactive framework classifying disease according to body (structure and function), activities and participation and contextual classifications (environmental and personal) (Gagne et al., 2009).

Motor Neuron Disease.

Motor Neuron Disease (MND) is a progressive, neurological disease involving the degeneration of upper and lower motor neurons in the motor cortex, brainstem and spinal cord (Kuhnlein et al., 2008; McLeod & Clarke, 2007). It is a term often used interchangeably with ALS – the umbrella term classifying motor neuron diseases (Leigh et al., 2003, McLeod & Clarke, 2007). MND to date remains an incurable disease with palliative care and symptom control being of prominent interest (Kuhnlein et al., 2008).

MND is categorized according to the site at which symptoms present initially. This may occur in the form of spinal MND (lower motor neuron damage), bulbar MND (upper motor neuron and brainstem damage) or mixed MND (upper and lower motor neuron damage). Clinical markers include limb weakness of a progressive nature, respiratory insufficiency, spasticity and hyperflexia. Individuals with bulbar onset MND develop dysarthria and dysphagia. Limb symptoms inevitably present simultaneously or shortly after the presentation of initial symptoms (Wijesekera & Leigh, 2009). Bulbar symptoms are evident in approximately 30% of individuals during the initial stages of the disease; however these symptoms are evident in almost all individuals during the later stages of MND.

Presbycusis.

Presbycusis is a high frequency sensorineural hearing loss (SNHL) arising as a consequence of the degenerative changes brought about by aging. This degenerative process is isolated to the impairment or death of the outer hair cells of the cochlea, which results in a decline in hearing ability (Rappaport & Provencal, 2002). A presbycutic hearing loss is frequently managed through amplification in the form of hearing aids.

Prevalence.

The term prevalence is used to define the population percentage presenting with a disease or disorder during a specified time period. This includes both newly diagnosed and previously diagnosed cases (Weiten, 2001).

Quality of Life.

Quality of life (QoL) is a vast concept encompassing all aspects of daily living. This includes physical, social, emotional, spiritual and existential domains of daily life (Hardiman, Hickey & O'Donerty, 2004). QoL refers to the extent to which individual hopes and ambitions are fulfilled and met in reality (Mitsumoto & Del Bene, 2000).

Recruitment.

Recruitment refers to an abnormal growth of perceived loudness occurring at suprathreshold intensities and arises as a result of the lack of outer hair cell modulation common in diseases of the inner ear (Hesse et al., 1999).

Sensorineural Hearing Loss.

A sensorineural hearing loss (SNHL) is an impairment in hearing that arises as a result of damage to the inner ear structures of the auditory system (Marieb, 2001). This may include damage at the region of the cochlea (cochlear hearing loss) and/or along the auditory nerve (retrocochlear hearing loss) responsible for transmitting the sound signal to the auditory cortex of the brain (Musiek & Baran, 2007). SNHL may arise as a result of aging, noise exposure, head trauma and/ or viral infection (Musiek & Baran, 2007). A SNHL is typically irreversible; however hearing ability may be enhanced through the use of assistive devices such as hearing aids and/or surgical procedures such as cochlear implantations (Musiek & Baran, 2007).

Tinnitus.

Tinnitus refers to a sound sensation perceived by an individual without an extrinsically evident stimulus (Vesterager, 1997). Tinnitus is typically experienced as a subjective condition and is frequently paired with psychological and perceptual components, proving to be distressing to the individual. Tinnitus may be triggered along any site of the auditory pathway; however it is typically associated with hearing impairment of sensorineural nature. Individuals with tinnitus, typically experience impaired ability to listen in the presence of noise. Tinnitus is frequently documented amongst individuals with a history of noise exposure and/ or as part of the effects of the aging process on the auditory system (Vesterager, 1997). Tinnitus sufferers associate this with negative changes in QoL giving rise to concentration difficulties, irritability, difficulty understanding speech (Khalfa et al., 2009).

Abbreviations

AAC	Augmentative and Alternative Communication
ABR	Auditory Brainstem Response
ALS	Amyotrophic Lateral Sclerosis
CHL	Conductive Hearing Loss
CP*x*	Caregiver of Participant Number (CP1 – CP2 etc.)
dB	Decibel
DPOAE	Distortion Production Otoacoustic Emission
HEQ	Hearing Experience Questionnaire
HHIA	Hearing Handicap Inventory for Adults
HL	Hearing Level
ICF	International Classification of Functioning, Disability and Health
L	Left
MCL	Most Comfortable Level
MDT	Multi-Disciplinary Team
MND	Motor Neuron Disease
MMND	Madras Motor Neuron Disease
OAE	Otoacoustic Emission
P*x*	Participant Number (P1 – P2 etc.)
PT	Pure Tone
PTA	Pure Tone Average
QoL	Quality of Life
R	Right

SD	Standard Deviation
Sd	Speech Discrimination
SL	Sensation Level
SNHL	Sensorineural Hearing Loss
SNR	Signal to Noise Ratio
SRT	Speech Reception Threshold
TD	Threshold of Discomfort
WHO	World Health Organization

Chapter One

Orientation

Introduction

The following chapter provides an orientation to the study. This chapter comprises the rationale for the study by describing the background information that led to its development as well as the relevance of this area of research. Definitions of terminology used within the context of the research and an explanation of the various abbreviations used throughout this report are provided. Finally, an outline of each of the chapters in the study is provided.

Background

Motor neuron disease (MND) is a relatively rare neurodegenerative disorder that is characterized by progressive motor cell injury and death (Mandriolli, Fagliani, Nichelli & Sola, 2006; Shaw, 2005). Lower and upper motor neuron signs arise as a direct consequence of motor neuron degeneration (Mandriolli et al., 2006). MND presents as a multi-systemic disease affecting bulbar, limb and respiratory muscles. This impairs speech function and mobility, with death ultimately occurring as an outcome of respiratory failure (Mandriolli et al., 2006).

The International Framework for Health, Disability and Functioning (ICF) aims to define the impact of disease at varying levels, including physical structure and function, activities, participation and environmental factors (World Health Organization [WHO], 2001). Within the ICF framework, the effects of MND-related impairments extend across the spectrum of physical disability, activity limitations and participation restrictions. At a physical level individuals

experience a loss of limb function as well as progressive weakness of the speech musculature, muscles for deglutition (necessary for the process of swallowing) and respiratory muscles. Activity limitations arise in the form of reduced mobility and self-care ability, which in turn evolve into restriction of participation.

Participation restrictions in the form of withdrawal and isolation from social and community life as well as the deterioration of interpersonal relationships, prevent reintegration of the affected individual (Ng & Khan, 2011). This proves to have significantly damaging effects on individual quality of life (QoL) and autonomy (Ng & Khan, 2011), both of which are identified as key elements to sustaining individual desire to live with terminal illness (Kelly et al., 2003).

The current scope of research remains directed towards clinical trials of disease patterns and symptoms that are directly related to prolonging lifespan and aim to yield outcomes that may modify or alter the course of this disease (McLeod & Clarke, 2007). Current research in MND is directed towards clinical trials of various agents that may theoretically modify disease progression. A vast range of trials evaluating the role of agents such as insulin-like growth factor I, nimodipine, creatine and riluzole have failed to modify disease progression, while investigation of the effects of previously unexplored agents continue to date (Leigh et al., 2003). In the exploration of multi-facetted nature of MND, the involvement of possible sensory neuropathy has consequently long since failed to be recognized as a related clinical feature (Isaacs et al., 2007). Despite this, evidence for non-motor neuron involvement has been suggested, although beyond these initial suggestions, limited insight into atypical symptoms has

been documented (Maier et al., 2009). The emergence of research into sensory symptom presentation may be attributed to prolonged lifespan. This affords researchers the opportunity to investigate disease components emerging at later stages of the disease, which may previously have been masked by the extreme physical and physiological consequences (Logroscino et al., 2008).

While the importance of understanding and pursuing knowledge relating to those life threatening symptoms and course of progression remain fundamental to the current scope of MND research, the ICF framework highlights the relevance of a holistic management approach, whereby aspects of QoL form key factors shaping disease experience (WHO, 2001). The impact of hearing loss on social, emotional and communicative wellbeing is significant (Dalton, Cruickshanks, Klein, Klein, Wiley & Nondahl, 2003). As a result, the exploration of the involvement of auditory impairment on the affected individual in this multi-systemic disease serves to provide valuable insight into the implications of hearing impairment as a factor influencing QoL and individual experience of living with neurodegenerative disease. While the value of pursing such diagnosis is linked to a number of positive outcomes, it is also essential to consider the negative emotional consequences in an already vulnerable population and the risks related to this. The latter may serve as another factor influencing the range of available research in this field of exploration.

Hearing loss in MND adults was first reported by Maier et al. (2009), where indications of sensorineural hearing impairment linked to auditory neuropathy or conductive hearing impairment linked to paralysis of the tensor and veli palatini muscles was suggested (Gourie-

Devi & Suresh, 1988; Maier et al., 2009). No further research related to hearing impairment and adult-onset MND is available in the literature to date, despite the devastating effects this superimposes on individual wellbeing and communicative success.

The most socially and psychologically damaging consequences of hearing loss include difficulty with speech perception and a false sense of understanding (Dewane, 2010; Nachtegaal et al., 2009). These characteristics of hearing loss inevitably manifest in isolation, withdrawal and overreliance on others, leading to a loss of identity and dignity as well as a loss of autonomy and feelings of being burdensome (Dewane, 2010). In a disease such as MND, where individuals are repeatedly required to redefine themselves, lower personal expectations and redirect focus on other aspects of life in order to maintain a sense of self-worth, the additional burden of hearing impairment poses significant threats (Foley, O'Mahony, & Hardiman, 2007). Further to this, the documented shifts in the dynamics between the individual with MND and his/her caregiver (Foley, O'Mahony & Hardiman, 2007), additionally highlight the negative impact arising as a direct result of MND and its' associated primary consequences. The added involvement of hearing impairment is then likely to further hinder individual-caregiver dynamics, increasing caregiver burdens above the burdens faced by MND caregivers at a level of accommodating physical loss of function (Garstecki & Erler, 1999).

Individuals with MND continually undergo changes at both a physical and physiological level of functioning. Alongside such readjustments are emotional and psychological shifts, which frequently dictate the coping abilities of the individual in the face of progressive redefinition of function and identity (Hallberg, Hallberg & Kramer, 2008). It is acknowledged that adjustment

to hearing loss in individuals with MND does not have the same life threatening consequences as respiratory or swallowing difficulties. However, at a psychosocial level of functioning, hearing loss is well known to prevent individuals from achieving optimal social integration outcomes. This consequently directly impact on their psychological wellbeing (Foley, O'Mahony & Hardiman, 2007; Ng & Khan, 2011). Therefore, in line with the management of primary symptoms, the value of early auditory diagnosis on the psychological and social wellbeing of the individual with MND and the caregiver is significant. This is therefore critical to maintaining and enhancing the prolonged life afforded to the affected individual through the effective management of primary, life threatening physical and physiological symptoms.

MND individuals in their seventh decade of life are on the increase, suggesting a longer lifespan linked to the positive effects of multi-disciplinary management (Logroscino et al., 2010). While knowledge surrounding the effects of motor neuron degeneration on the auditory system is scarce, the parallel and corresponding age range for MND presentation and progression when reviewed against age-related hearing loss requires consideration. This suggests that hearing impairment related to natural aging may arise in the course of MND. This requires individuals to adjust to this loss of function as a means of maintaining autonomy and desirable QoL. Furthermore, it is through interactive and socialization opportunities that improvements in QoL are reported, consequently enhancing emotional and mental wellbeing and reportedly limiting the presentation of depressive symptoms (Averill, Kasarskis & Segerstrom, 2003; Boi et al., 2011).

With no cure available the current challenge remains to establish measures that facilitate prolonged independence and optimal QoL (Ng & Khan, 2011). Facilitative devices

(augmentative and alternative communication [AAC] systems for expressive communication, wheelchairs for mobility) serve to promote positive enhancements to QoL. This affords individuals with MND the opportunity to maintain a level of autonomy and dignity. In line with this, hearing aids and communicative strategies facilitate integration, inclusion and communicative success in interactive situations. QoL forms the foundation to all individuals irrespective of medical circumstance, and the detrimental effects of hearing loss on QoL. However, in the instance of a population dealing with a progressive loss of physical ability, the psychosocial value of maintaining a sense of normalcy.is imperative. Furthermore, in a population where motor function is significantly reduced stripping an individual of written or spoken abilities, the impairment of auditory function essentially closes down all forms of communication available to the individual. Without alternative approaches to rehabilitating and/ or guiding communication, the superimposed decline of auditory function forms the final element leading to communicative breakdown (Dalton et al., 2003)

It is apparent that the physical consequences of motor neuron degeneration (e.g. respiratory difficulties and aspiration), pose significant life threatening risks for individuals, and therefore remain the primary focal point in MND management. However, it is also necessary to consider QoL as an essential element that makes life-prolonging and/or sustaining measures worthwhile. Through addressing life prolongation, the individual is afforded an extended period of life, but without consideration for secondary and concomitant symptoms, it does not guarantee QoL. It is therefore of upmost importance that *all* symptoms influencing the functioning of the individual with MND be considered and effectively managed to ensure optimal outcomes from patient management at a physical level as well as an emotional and social level (Cedarbaum, 1996). The

relevance of acknowledging and understanding secondary symptoms serves to enhance the nature of multi-disciplinary team involvement aimed at symptom specific management. Facilitating individual self-empowerment and guiding the process of adaptation to progressive loss of function are the foundations to enhancing individual desire to live and engage in the surrounding world (Kelly et al., 2003). The relevance of understanding the involvement and associated effects of hearing loss in MND thus become profoundly apparent as a prominent factor dictating the QoL and autonomy of an individual facing neurodegenerative losses. .

This study therefore aims to explore the prevalence of hearing loss within the MND population. Further to this, the study serves to explore the history of auditory referrals amongst the sample, the perceived value of functional abilities including hearing and the relevance of auditory diagnosis as perceived by individuals with MND and their caregivers in the face of living with a fatal disease.

Chapter Outlines

This research report will be presented in six chapters. Chapter 1 provides a basic orientation to the study as well as the rationale for the study. This chapter includes definitions of terminology used throughout the research report, an explanation of the abbreviations used and an outline of the chapters in this report.

Chapter 2 provides an outline of the study's conceptual framework relating to the ICF. A description of MND reviewing the reported etiology and the classification of types and stages of MND progression follows. This chapter proceeds to address both motor and sensory symptoms of MND in relation to the ICF's framework of body; activities and participation. Particular emphasis is directed towards the involvement of the auditory system at a functional and structural level in MND. With the support of the available literature, the impact of auditory impairment on activity limitations and the effects this has on participation in daily life is highlighted. Brief mention of the ICF contextual factors as they relate to environmental and personal components is included alongside a review of the rehabilitative services involved in the MND management team. The level of involvement of the audiological discipline as part of the MND multi-disciplinary team is addressed in this chapter.

Chapter 3 provides a detailed review of the methodological paradigm applied to this study. This chapter includes a description of the primary and secondary objectives outlined in this study, followed by a detailed review of the selected research design. This follows with an outline of the research phases applied to this study and a review of the execution, outcomes and modifications that arose as a result of the pilot study; followed by the main study. A description

of participants is followed with a review of the equipment, measuring instruments and test battery. This chapter concludes with a review of data collection procedures, ethical considerations and statistical procedures.

Chapter 4 provides a thorough description of the results yielded as an outcome of this study. This includes a report on the prevalence of hearing loss in participants in this study in accordance with the primary objective of the study. In fulfillment of the secondary objectives of this study, this chapter provides a detailed description of the audiometric patterns identified within this sample. A review of the perceived social and emotional consequences of hearing loss in line with the Hearing Handicap Inventory for Adults (HHIA) follows. This chapter concludes with a review of the qualitative data acquired through the use of the Hearing Experience Questionnaire (HEQ). This includes a review of participant contact with the audiological discipline and breakdown of participant and caregiver perceptions related to the rating of functional skills. This chapter closes with an exploration of participant and caregiver perspectives relating to the value of auditory testing and diagnosis in MND.

Chapter 5 comprises a detailed discussion integrating the results of this study with relevant literature. This is discussed in accordance with each of the outlined objectives of this study and highlights significant outcomes of this study.

Chapter 6 provides concluding statements relating to the current study. This chapter includes a summary of the rationale and findings from this study. This is followed by a review of the limitations identified throughout the execution of this research. This chapter concludes

with the implications for future research and a set of recommendations for future research studies of a similar nature.

The appendices supply important information for the understanding of the data collection and analysis procedures, and thus the replication of the study.

Chapter Two

Literature Review

Introduction

The quality of life in adults with progressive, neurodegenerative disorders is severely affected at multiple levels of functioning, namely physical body structure and function, participation and activities of daily living. The following chapter serves as a review of the current literature related to MND, hearing loss and the interrelated implications of these. These factors will be discussed within the ICF framework. It further aims to provide evidence for the limited availability of literature related to suspected sensory involvements, such as hearing impairment in MND. Documented incidences of hearing impairment related to MND will be highlighted alongside hypotheses supporting these diagnoses. The importance of auditory function for maintenance of QoL is also presented in this chapter. Lastly, the information presented in this chapter serves to provide further insight into the current nature of multi-disciplinary management of MND in relation to the needs outlined in the ICF framework.

International Classification of Functioning, Health and Disability

The ICF is a scientific tool introduced by the World Health Organization (WHO) in a bid to establish an international framework serving to enhance functioning of health systems (WHO, 2001). In 2001, a total of 191 Member States of the WHO (including South Africa), adopted the ICF principles as the basis for the scientific standardization of data on health and disability on a worldwide scale (WHO, 2001). The ICF provides comparable information about the experience of health and disability, based on a biopsychosocial school of thought, where disease is no longer

defined in isolation by its physical consequences (Gagne et al., 2009). This framework provides a coherent view of varying health perspectives (including biological, social and individual perspectives) without making the error of converging the complex notion of disability into only one of the named components (Gagne et al., 2009). The ICF hence views disability and functioning as the outcomes of interactions between health conditions, environmental- and personal factors (See Figure 1).

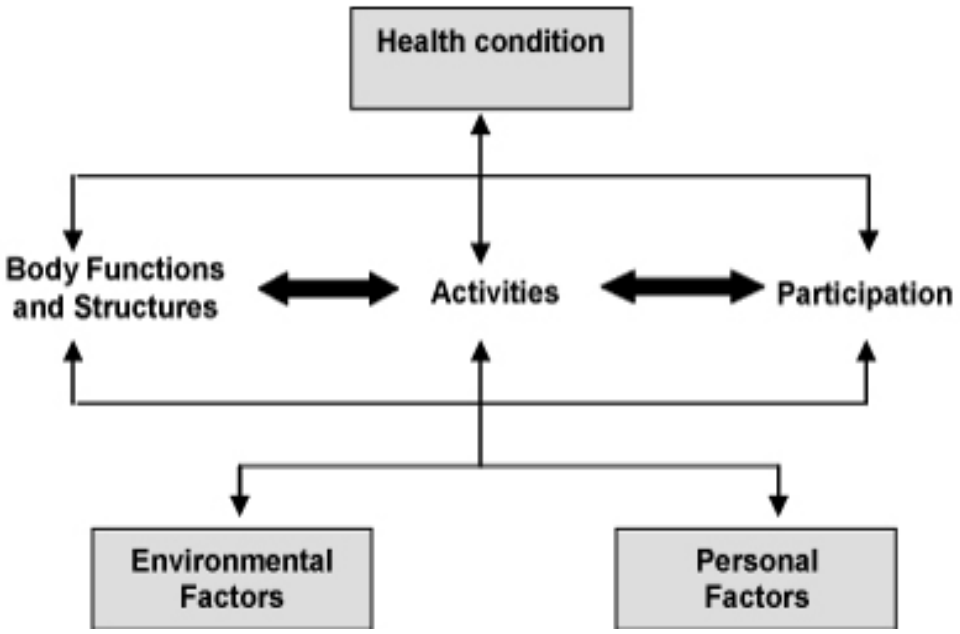

Figure 1. Outline of ICF Classification System (WHO & United Nations Economic and Social Commission for Asia and the Pacific [UNESCAP], 2008, p. 23)

This flexible framework comprises classifications according to body structure and function and activities and participation, as well as contextual classifications linked to environmental and personal factors (UNESCAP, 2008). Each classification is further subdivided into categories providing an in-depth discussion of the various components considered within

this framework (Table 1). The first category, body functions and structures, serves to describe the physical capabilities and physiological functions of the body in relation to a specific health condition. This may include functioning at the level of the respiratory system and/ or sensory and cardiovascular structure and function to name a few examples. The second category, activities, refers to the execution of tasks by the individual such as communicating, performing self-care tasks and domestic life (e.g. grocery shopping). Participation, then relates to the individuals' involvement in life situations (e.g. domestic, community, social and civic life, interpersonal interactions and relationships).

Table 1

ICF Categories and Classification

Body	
Function:	**Structure:**
Mental function	Structure of the nervous system
Sensory functions and pain	***The eye, ear and related structures***
Voice and Speech function	***Structures involved in voice and speech***
Functions of the cardiovascular, Haemotological, Immunological and Respiratory Systems	***Structure related to movement***
	Structure related to reproductive systems
Functions of the ***digestive***, metabolic, endocrine systems	Skin and related structures
Reproduction functions	Structures related to the cardiovascular, Haemotological, Immunological and ***Respiratory Systems***
Neuromusculoskeletal and movement-related functions	
Functions of the skin and related structures	Structures related to ***digestive***, metabolic, endocrine systems

Activities and Participation	
Learning and applying knowledge	***General tasks and demands***
Communication	***Mobility***
Self care	***Domestic life***
Interpersonal interactions and relationships	***Major life areas***
Community, social and civic life	

Environmental Factors	
Products and technology	***Support and relationships***
Services, systems and policies	***Attitudes***
Natural environment and human-made changes to environment	

(WHO & UNESCAP, 2008, p. 30)

The ICF is further divided into two contextual categories, namely environmental and personal factors. Environmental factors are defined as the factors that *'make up the physical, social and attitudinal environment in which people live and conduct their lives'* (UNESCAP, 2004, p.1) and form the external factors of the ICF (Royal College of Speech Language Therapists [RCSLT], 2009). These may include families, labour and employment services and systems available to the individual. The products and technology available to the individual, alongside the natural environmental and human made changes that facilitate maintained psychosocial wellbeing, support and relationships additionally comprise environmental factors. These include wheelchairs for mobility, adaptive equipment for eating and home modifications to accommodate these environmental assistors (Ball, Beukelman, & Pattee, 2004). Personal factors comprise those internal factors that are specific to the individual and vary amongst all individuals. These include age, gender, education, character, as well as past and current experiences (RCSLT, 2009). Personal factors are an important indicator of individual ability to cope with diagnoses and largely influence the manner in which different individuals experience disability (RCSLT, 2009).

The ICF framework has many advantages, the foremost of which is that it allows for a description of *all* difficulties experienced by an individual. This naturally leads to an intervention approach that is family-centered and committed to applying specific solutions that allow individuals to overcome activity and/or participation restrictions for enhanced QoL (Gagne et al., 2009). The ICF additionally allows for an integration of both medical and social aspects of disease rather than focusing entirely on the diagnosis itself. This further ensures that goals and solutions specific to the client are formulated promoting alleviation of individual difficulties and

optimal participation in activities considered to be limiting to the affected individual (Gagne et al., 2009).

Erdman (2009) reports that management approaches that are efficacious in addressing activity limitations and participation restrictions show improvements in psychological and psychosocial wellbeing. This approach to disease management therefore is positively associated with improved QoL for the individual with disease as well as the primary caregiver and support structure. The term QoL is widely described in the literature, however a universal definition of this is yet to be established. QoL relates to the extent to which an individual is able to pursue and realize their life goals (Mitsumoto & Bene, 2000), while others refer to QoL as those factors contributing to an individual's overall happiness and harmony (McLeod & Clarke, 2007). A common thread amongst all definitions of QoL, although a slightly more limiting perspective, is that it refers to general psychological wellbeing of the individual and the requirements necessary to realize this (Hallberg, Hallberg & Kramer, 2008). Poor QoL has been identified to be closely related to individual end-life decisions (Kubler, Winter, Ludolph, Hautzinger & Birbaumer, 2005). It is reported that individuals with lower QoL are more likely to decline life-sustaining treatments, further emphasizing the importance of a framework that considers all dimensions of individual functioning and involvement (Goldstein, Atkins, Landau, Brown & Leigh, 2006; Kubler et al., 2005).

Motor Neuron Disease

MND is one of the most common adult onset disorders of the motor neurons. MND is often referred to interchangeably as amyotrophic lateral sclerosis (ALS) or Lou Gerich's disease

(McLeod & Clarke, 2007; Van Damme & Robberecht, 2009). A consistent global incidence of 1,2 per 100,000 of the population per year is reported, whilst the prevalence is indicated at 5,2 per 100,000 (Wijesekera & Leigh, 2007). MND is a progressive, neurological disorder resulting in a gradual loss of function in the upper and lower limbs, speech, deglutition and respiratory abilities. MND typically presents during the fourth and sixth decades of life (Leigh et al., 2003; Shaw, 2005; Van Damme & Robberecht, 2009). Life expectancy is reported within 3-5 years from symptom onset (Hardiman et al., 2004; Logroscino et al., 2010; Van Damme & Robberecht, 2009). A 5-year survival is documented in 18-40% of individuals from symptom onset and in 5-30% of individuals from diagnosis (Mandriolli, Faglioni, Nichelli & Sola, 2006). Survival in some instances has been extended up to 10 years by respiratory support (Bach, 2002). In rare instances, juvenile forms of MND have been noted to present during the second decade of life (Shaw, 2005). These juvenile forms tend to present with a slower rate of progression and thereby afford individuals up to a few decades of life from symptom onset (Swash & Desai, 2000). The exact etiology of selective neuron degeneration has not yet been exclusively identified, however multi-factorial causes associated with oxidative stress, environmental factors and genetics are under investigation (Chancellor & Warlow, 1992; Shaw, 2005).

MND is confined to the upper and lower motor neurons in the motor cortex, brainstem and spinal cord (Kuhnlein et al., 2008), but in some instances cranial nerve nuclei may also be implicated (Shaw, 2005; Snell, 2001). MND, at a clinical and electrophysiological level, is described as a disorder purely of the motor system, with a salient pathological feature of progressive motor cell injury and death (Donaghy, 1999). However, recent studies suggest that motor neuron groups are primarily but not exclusively involved in MND; and are variable in

vulnerability to degenerative processes (Isaacs et al., 2007; Shaw, 2005).

Sensory neuropathy has long since failed to be recognized as a clinical feature of the MND spectrum, the presence of which has remained a cause of diagnostic uncertainty (Isaacs et al., 2007). It is postulated that a wider spectrum of MND related signs and symptoms are anticipated to become clinically apparent as a result of increased life expectancy due to medical and supportive care developments (Isaacs et al., 2007; Shaw, 2005). With no cure available and a growing range of symptoms, the challenge lies with prolonging independence and optimizing patient QoL (Ng & Khan, 2011).

Prolonging independence and optimizing QoL is best addressed with multi-disciplinary management that encompasses the symptomatic management, but also the physical, social and environmental factors spanning the spectrum of MND (Ng & Khan, 2011). Investigation surrounding the atypical features of MND is therefore essential for refining the standard clinical description of the disease. Further exploration, in support of the diagnostic process, also serves in reshaping the management of individuals with MND. This will ensure that disease features at both a primary and secondary level are accounted for and accommodated (Isaacs et al., 2007).

Classification and Progression of MND.

Classification of MND is determined according to the site at which the symptoms of MND initially presented and the course of degeneration noted from the initial symptom presentation (Doyle & Phillips, 2001).

MND symptoms may be characterized by a loss of function in the spinal motor neurons or bulbar musculature – the former presenting with a slower rate of deterioration than the latter (Ball et al., 2004; Turner, Parton, Shaw, Leigh, & Al-Chalabi, 2003). Spinal motor neuron degeneration may present as upper, lower or mixed loss of motor neuron function. A loss of lower motor neuron function is associated with muscular atrophy, weakness, and fasciculation. The loss of upper motor neuron function is marked with supranuclear features including brisk jaw jerk, emotional lability and bulbar muscle spasticity (Kuhnlein et al., 2008). Features of spasticity tend to present in the weakened atrophic limbs impacting gait and dexterity (Wijesekera & Leigh, 2009). Upper and lower motor neuron degeneration may present in isolation, however it is a combination of these that is most commonly noted. This is referred to as mixed MND. Mixed MND is noted in approximately two thirds of individuals where symptoms such as wasting and focal muscle weakness present in the upper and lower limbs either distally or proximally (Wijesekera & Leigh, 2009).

Bulbar symptoms in MND are observed in approximately 30% of patients during the initial stages, while almost all individuals present with these symptoms as the disease progresses (Kuhnlein et al., 2008; Van Damme & Robberecht, 2009). The degeneration of lower motor neurons is responsible for the presentation of bulbar palsy symptoms such as dysarthria and dysphagia. Dysarthria, a speech disorder resulting from weakness, paralysis or in-coordination of the speech musculature, is the most common initial symptom in bulbar onset MND (Ball et al., 2004). Dysphagia, the partial or total difficulty swallowing as a result of damage to the brain, muscles and/or nerves responsible for controlling swallowing, is the second most common symptom (Kuhnlein et al., 2000; Traynor et al., 2000).

Functional changes in various bodily regions progress at different rates in MND (Ball et al., 2004; Doyle & Phillips, 2001). In light of this variability, the MND classification system documents the progression of neuron degeneration in stages according to functional use of the upper extremities, mobility and speech throughout the course of the disease (Appendix A) (Riviere, Meininger, Zeisser & Munsat, 1998). This begins at the earliest stages where functional independence is maintained in two of the three specified modalities (speech, arm or leg movement) and a mild deficit exists in the remaining one region (Stage I). With progression of the disease and deterioration of function, the classification system concludes at Stage IV where the individual with MND has no functional use of two of the above named modalities and moderate or no functional use of the third region as well.

ICF and MND

Body Structure and Function: Symptoms of MND.

The ICF framework for classifying functioning, disability and health defines body functions as *'physiological functions of body systems (including psychological functions)'* (WHO & UNESCAP, 2008, p. 27). One such psychological function relates to the presence of dementia, a recently exposed MND-related symptom. Although frontotemporal dementia is estimated to occur in 5% of individuals diagnosed with MND (Van Damme & Robberecht, 2009) exact figures have yet to be established. The ICF's body structures are then defined as *'the anatomical parts of the body such as organs, limbs and their components'* (WHO & UNESCAP, 2008, p.27). As motor neurons gradually cease to function, muscle deterioration results in severe and extensive decline in an individuals' functional status (Doyle & Phillips, 2001). A loss of motor

ability is primarily reported as the core symptom of MND, with minimal report and/or documentation of the sensory effects of the disease.

The section to follow will provide a description of the motor systems involved in MND in line with the ICF definitions of bodily function and structure. This will then be followed by a discussion surrounding documented sensory involvement in MND on a broad scale spectrum and in relation to the auditory system, highlighting the gaps in research in this domain.

Motor Symptoms.

Individuals with MND are faced with a gradual loss of function. According to the categories described in the ICF (Table 1), these include decline in muscular function (mobility), mental functions (dementia), respiratory (breathing), digestive function and structure (dysphagia), voice and speech functions (dysarthria), as well as investigations into the effects of MND on eye (vision) structure (WHO & UNESCAP, 2008).

The deterioration of physical skills such as movement, functional use of the legs and arms for gross motor movements and fingers for fine motor manipulation remain key markers of the physical threats faced by an individual with MND (Ball et al., 2004).

Dysphagia is a key bulbar symptom. This, when paired in combination with progressive weakness of respiratory muscles, prevents reflexive cough action to enable clearance of the aspirated food (Kuhnlein et al., 2008), consequently leading to aspiration pneumonia and possible death (Ekberg et al., 2002). Dysohagia is documented in 86% of individuals with MND

(Kuhnlein et al., 2008). Muscular fasciculation of the tongue is reported in 64% of individuals. Fasciculation's involve the simultaneous contraction of groups of muscle fibers resulting in muscular twitching (Snell, 2001). Fasciculations at the level of the oral structures further compound difficulties in swallowing at the preparatory phase of the swallow (Kuhnlein et al., 2008).

The effects of MND on the communicative system can be devastating to the individual. The gradual deterioration of motor neuron function impacts the respiratory system in the form of respiratory muscle weakness, threatening both communicative competence and long-term survival (Ball et al., 2004). Respiratory muscle strength is necessary for fulfilling its role in the process of phonation, without which weakness in vocal intensity and strength becomes evident. The communicative system additionally relies on muscular function for movement of the oral motor structures for articulate expression. As a direct consequence, a decline in speech intelligibility is well documented in the MND population (Freed, 2000; Yorkston, Beukelman, Strand, & Bell, 1999). The decline is marked by impaired muscle tone affecting power and range of voice, breathing and oral movements as well as in-coordination of musculature for speech production (Duffy, 1995). Bulbar symptoms gradually become more pronounced as the disease progresses, with 93% of individuals developing dysarthria (Leigh et al., 2003; Kuhnlein et al., 2008). Dysarthria initially presents at varying levels of severity and this gradually evolves into a total loss of expressive verbal function. A total loss of expressive verbal function is reported in 80% of MND individuals as motor neuron deterioration progresses and motor neurons cease to function (Leigh et al., 2003).

Traditional curative interventions are currently inapplicable in the management of MND. This, as well as the rapidly progressive nature of this disease, emphasizes the importance of acknowledging systems that are preserved while a steady decline of bodily function occurs. It is suggested that certain motor neuron groups are less vulnerable to the pathological process of degeneration (Shaw, 2005). For example, the motor neurons in the upper brain stem nuclei responsible for the control of eye movements, the ocular nuclei, remain functional. The reduced vulnerability of the ocular nuclei therefore allow non-verbal, expressive communicative abilities to be somewhat maintained through the use of visual gaze and tracking. With maintained visual function, the importance of augmentative and alternative communication (AAC) strategies become central to maintaining communicative competence amidst deterioration of verbal expressive abilities (Ball et al., 2004). The reduced vulnerability of these visual motor neuron groups further support the need to determine the susceptibility of other systems, such as the auditory system, to the MND degenerative process. The current study thus placed emphasis on determining the prevalence of auditory impairment as a sensory symptom of MND.

Sensory Symptoms.

In recent years various MND studies have explored the involvement of sensory neuropathy in a disease dominated largely by motor dysfunction (Isaacs et al., 2007; Mondelli, Rossi, Passero, & Guazzi, 1993). Isaacs et al. (2007) reported that sensory symptoms were identified in 50% of participants diagnosed with MND (N = 111). These symptoms were confirmed with electrophysiological investigations, confirming the high incidence of sub-clinical, progressive sensory dysfunction (Isaacs et al., 2007). The involvement of the autonomic system, basal ganglia, dorsal columns, spinocerebellar tracts and extra-motor cortex were documented in 2-

10% of these individuals. Furthermore, a 30% reduction in the total number of myelinated nerve fibers was reported in participants with MND in contrast to a control group (Isaacs et al., 2007). Earlier studies support the presence of sensory dysfunction in MND to a lesser extent in 13-22% of participants (Mondelli et al., 1993; Theys, Peeters & Robberecht, 1999). These differences may be accounted for by the diagnostic limitations of electrophysiological investigations used in these earlier studies. An additional study by Pugdhal et al. (2007) explored sensory involvement in MND revealed a larger number of individuals presenting with normal sensory functions than otherwise. In this study (*N = 88),* a total of 20 participants presented with abnormalities within sensory parameters, while the remaining 68 participants presented without sensory abnormalities (Pugdhal et al., 2007). The study concluded that these findings were suggestive of unaffected sensory nerve function in MND. It was however later acknowledged by the authors of this study, that a high probability exists that sensory nerve dysfunction is a representation of a variant of the disease and hence could not be conclusively discarded based on the participants from that study alone (Pugdahl et al., 2007). It was furthermore suggested that sensory involvement may have been underestimated since electrodiagnostic tests were not included in the test protocol (Pugdahl et al., 2007).

One of the sensory symptoms infrequently documented in individuals with MND, is hearing loss (Gourie-Devi & Suresh, 1988; Maier et al., 2009). The auditory system comprises the outer, middle and inner ear. The outer ear region comprises the pinna, the ear canal and the tympanic membrane (Musiek & Baran, 2007). The pinna collects sound and directs it into the ear canal reaching the tympanic membrane, which is set into vibratory motion. This motion is transmitted along the ossicles of the middle ear namely, the incus, malleus and the stapes

(Musiek & Baran, 2007). The ossicles serve to stimulate vibratory motion from the air medium of the outer ear to the liquid medium of the inner ear stimulating the organ of hearing, the cochlea. This vibratory motion stimulates the transmission of the sound signal along the auditory nerve to the auditory cortex of the brain translating these signals into audible sound (Musiek & Baran, 2007). Uninterrupted transmission of the sound signal from the outer ear through to the level of the auditory cortex results in normal hearing abilities. In instances where sound is not efficiently conducted through the outer and/or middle ear region, a conductive hearing loss (CHL) arises, although this can frequently be medically or surgically corrected. Damage to the auditory system at the level of the cochlea and/or auditory nerve may result in a sensorineural hearing loss (SNHL). This can be further classified to be either a cochlear or retrocochlear loss, depending on the level of the auditory pathway at which damage occurs (Musiek & Baran, 2007). Sensorineural hearing loss generally cannot be reversed, however rehabilitation methods and assistive devices are available to assist in achieving improved sensory regulation.

Madras Pattern of Motor Neuron Disease (MMND), a form of MND, was reported by Gourie-Devi and Suresh (1988). MMND individuals present with an age of disease onset that is at least a decade earlier than typical adult-onset MND. Bilateral SNHL and bilateral facial nerve involvement are noted to present in individuals with MMND. This supports indications of sensory involvement in the MND spectrum of disease.

Audiological testing of individuals with MMND provided evidence for hearing impairment (Gourie-Devi & Suresh, 1988). Auditory testing revealed a bilateral SNHL in ten out of eleven

participants, whilst one participant presented with a unilateral SNHL. The presence of sensorineural impairment suggests damage either within the region of the inner ear (cochlea) and/or along the auditory nerve (retrocochlear) leading to the auditory cortex. This study did not report site of lesion testing to further isolate the specific location of auditory impairment. Severity varied amongst participants with 50% presenting with a severe loss and the remainder with a mild-moderate loss (Gourie-Devi & Suresh, 1988). While bilateral sensorineural hearing impairment remains a main feature of MMND, the presence of hearing impairment in individuals with typical adult onset MND has yet to be conclusively reported.

A single retrospective study provided the first hint of evidence linked to auditory involvement in adult onset MND and included investigation of reports of hyperacusis (Maier et al., 2009). The latter has yet to be reported in MND. All three participants in this study had undergone tracheostomy (35 months, 26 months and 25 months respectively since disease onset) and received long-term ventilation. Following retrospective review of these three cases, Maier et al. (2009) reported bilateral hearing impairment for all three participants (See Table 2). The test battery applied to the above study included pure tone audiometry and auditory evoked potentials, while structural lesions in the middle ear were clinically and pathoanatomically excluded (Maier et al., 2009). Speech audiometry measures were not included in this test battery.

Table 2

Retrospective Study: Hypoacusis in ALS Patients (Maier et al., 2009)

	Age	Gender	Time since onset of bilateral hearing loss	SNHL	CHL
P1	32	Female	78 months	**	
P2	47	Male	33 months	**	
P3	48	Male	31 months		**

Note: SNHL - Sensorineural hearing loss; CHL – Conductive hearing loss

The presence of SNHL is etiologically unknown however its presentation in two of the three participants suggested the presence of a neurodegenerative process with involvement of the auditory system (Maier et al., 2009). Sensorineural hearing impairment suggests damage within the inner ear region and/or along the auditory nerve leading to the auditory cortex. The cochlear organ of the inner ear is comprised of two types of neurons called the Type I and Type II afferent neurons (Spoendlin, 1971; Spoendlin & Schrott, 1989). These afferent neurons or sensory receptors are activated by the inner hair cells of the organ of Corti and are responsible for the innervations of the cochlear inner hair cells. This occurs at synapses whereby glucomate neurotransmitters communicate signals to the primary auditory neurons. The dendrites of these neurons belong to the auditory nerve, later uniting with the vestibular nerve to form the eighth cranial nerve – the vestibularcochlear or auditory nerve (Marieb, 2001). The auditory nerve is responsible for carrying the sound signal to the auditory cortex of the brain, beginning at the spinal ganglia (Marieb, 2001). Type I spinal ganglion neurons are large and myelinated whilst Type II neurons are smaller and unmyelinated (Spoendlin & Schrott, 1989). Type I afferent neurons comprise 95% of the neurons exclusively connected to the inner hair cells. Type II afferent neurons comprise 5% of the spinal ganglion cells and supply the outer hair cells

(Spoendlin, 1979; Spoendlin & Schrott, 1989). The findings of the study conducted by Isaacs et al. (2007) indicated a 30% reduction of myelinated fibers occurs as a sensory consequence of MND. The presence of a high percentage of myelinated neurons along the auditory pathway suggests that the effect of a 30% decline in myelinated fibers may threaten auditory ability. This further supports the hypothesis that a hearing loss of a sensorineural nature in individuals with MND is a viable consideration in need of more thorough exploration and research.

In contrast to sensorineural hearing impairment is the hypothesis that paralysis of the tensor tympani muscle, as a result of bulbar palsy symptoms, could result in a CHL (Ghadiali, Swarts & Doyle, 2003). The auditory system is protected from loud stimuli by the tensor tympani muscle, which pulls the malleus medially resulting in the damping of vibrations in the ossicles. This results in reduced sound amplitude (Marieb, 2001). This damping leads to in smaller amplitude of vibratory motion along the remaining components of the auditory pathway and consequently a reduction in the loudness of the sound signal (Marieb, 2001). An alternative hypothesis may be linked to the paralysis of the tensor and levetor veli palatini muscles, which pose a threat to the mechanical properties of the Eustachian tube. The impaired ability to actively dilate and increase the lumen in the cross-sectional region of the Eustachian tube arises as a result of bulbar palsy symptoms (Ghadiali, Swarts & Doyle, 2003). This leads to insufficient forces being applied to the membranous wall of the Eustachian tube during swallowing. The latter subsequently results in the development of negative middle ear pressures and places individuals at risk for developing otitis media with effusion (Ghadiali et al., 2003). The presence of otitis media would further support the hypothesis of conductive hearing impairment (Maier et al., 2009). Findings from the Maier et al. (2009) support the previously

undocumented presence of hearing loss in MND as one of the possible sensory symptoms of MND.

Sensory involvement may not be a key marker in the current MND diagnostic process, but may rather extend the phenotype of the disease to include a clinical or subclinical axonal sensory neuropathy (Isaacs et al., 2007). It is acknowledged that the low incidence of idiopathic neuropathy in the general population may account for its occasional presentation alongside MND (Isaacs et al., 2007). Other studies have reached parallel conclusions suggesting that sensory involvement, as part of multi-systemic degeneration is a valid claim deserving of further investigation (Pugdahl et al., 2007). Further investigation is of particular importance in light of advances in the medical management of MND that are resulting in the extended lifespan of individuals with MND (Isaacs et al., 2007; Pugdahl et al., 2007). This provides greater opportunities for exposing atypical symptoms, such as hearing impairment, that possibly present later in the disease and threaten individual wellbeing. This provides greater opportunities for exposing atypical symptoms, such as hearing impairment, that possibly present later in the disease and threaten individual wellbeing.

Activity Limitations and Participation Restrictions: MND.

The WHO views functionality, disability and health as an interactive process (WHO & UNESCAP, 2008). In instances of impaired body structures and functions that cannot be returned to normalcy, provision of solutions to alleviate limitations at a personal and a social level become paramount to the disease management approach (WHO, 2001). It is important to view individual functioning as the interaction between an individuals' general physical health

status alongside contextual factors (WHO, 2001). The ICF view activity limitations as *'difficulties in executing activities'* (WHO & UNESCAP, 2008, p. 27*)*, while participation restrictions are viewed as '*problems in involvement in life situations'* (WHO & UNESCAP, 2008, p.27*)*. This section will focus on the various activity limitations and participation restrictions faced by an individual with MND. These will be discussed with particular emphasis on the effects on communication and more specifically the threats brought about by hearing impairment.

Activities and participation as outlined by Table 1 include the following categories as they relate to MND: general tasks and demands linked to mobility, self-care, interpersonal relationships, social and community life and communication. Progressive physical weakness and loss of upper and/or lower extremity function threatens individual ability to perform activities of daily living and fulfill daily demands, such as writing, dressing, driving, walking and independent management of basic hygiene (bathing, brushing teeth).

Further to this, dementia is named as a possible symptom of MND and negatively affects an individuals' ability to perform common daily activities such as eating, bathing and continence. Restrictions in socialization and participation within group situations arise as a consequence of these limitations (UNESCAP, 2004).

Similarly, dysphagia commonly presents as a consequence of motor neuron degeneration. The difficulties in controlling liquids for fluid intake, chewing abilities and the process of deglutition further impacts daily activities and participation. It has been found that over 50% of participants with dysphagia no longer experience eating as a pleasurable activity as a direct result

of the associated limitations (Ekberg et al., 2002). Dysphagia directly affects the ability to eat and drink independent of risk. This in turn threatens individual participation in all social situations centered on dining experiences (e.g. restaurants, dinner invitations). Feelings of exclusion, isolation and distress during mealtimes were reported by participants in this study consequently resulting in increased reluctance and avoidance of eating in social settings that extend beyond the privacy of the home environment and primary caregiver (Ekberg et al., 2002). This not only threatens individual wellbeing, but places strain on the maintenance of relationships.

Respiratory muscle weakness results in individuals with MND experiencing respiratory insufficiency (Ball et al., 2004). These symptoms include disturbed sleep, fatigue, morning headaches, increased respiratory rate, as well as poor concentration and memory (Ball et al., 2004). While assisted ventilation of a non-invasive nature has been positively associated with improving patient QoL, invasive ventilation measures (e.g. tracheostomy) have negative effects on remaining communicative ability (Van Damme & Robberecht, 2009). Such respiratory symptoms further limit individual ability to perform activities of self-care, desire to engage in community and/or social activities and threaten the maintenance of established interpersonal relationships and/or establishing new relationships (Van Damme & Robberecht, 2009).

Dysarthria is identified as one of the most debilitating deficits experienced by individuals with MND (Leigh et al., 2003; Kuhnlein et al., 2008). The communication process relies on a two-way exchange whereby individuals receive and provide information. For this exchange to be effective, intact verbal expression and auditory reception is necessary. A breakdown in one or

both of these processes, results in a breakdown in interactive exchange (Glennen & DeCoste, 1997). Dysarthria impairs verbal communicative abilities and negatively affects communicative success in activities of speech. This includes making requests, expressing concerns and/or sharing information for functional or social purposes (Aronson, Ramig, Winholtz & Silber, 1992; Kuhnlein et al., 2008). Communication is a critical component in promoting psychological independence, establishing social closeness and maintaining independence in activities of daily living. Deterioration or loss of communicative abilities increases dependence on others, strips individuals of autonomy and in time provides evidence of a decline in social and emotional wellbeing (Ball et al., 2004). Activity limitations as a result of dysarthria further include reduced speech intelligibility and communicative ability. This has serious implications on participation and creates restrictions to an individual's participation at a level of social functioning i.e. church gatherings, family functions, and community gatherings. Furthermore, clinical/medical autonomy i.e. expressing thoughts, concerns or requests, directly related to the management of MND, may become restricted further damaging individual perception of control and desire to engage in the decision-making process.

A breakdown in auditory reception amongst this population would largely be brought about by deterioration or loss of auditory function. Hearing loss is often unrecognized and undertreated as a health disorder, despite the marked implications this has on individual QoL (Dalton et al., 2003). Hearing impairment in individuals with MND is further exacerbated by the additional symptoms and loss of function experienced by the affected individuals.

Activity Limitations and Participation Restrictions: Hearing loss and MND.

The prevalence of hearing impairment is reported to occur in 10 – 20 % of the population worldwide as a consequence of various auditory pathologies (Mathers, Smith & Concha, 2003). Age-related hearing loss, or presbycusis, occurs in approximately 46% of the population aged 48-65 years (Yueh, Shapiro, MacLean, & Shekelle, 2003). It is important to note that the age range in which MND diagnosis and progression occurs typically correlates with the range in which presbycutic hearing loss presents. It is therefore postulated that individuals with MND may at some stage experience hearing impairment, regardless of whether this is directly or indirectly related to the MND itself.

The impact of hearing loss on activities and participation in daily life does not vary according to the cause of onset, but is rather based on severity and the functional obstacles presented (Nachtegaal et al., 2009). This provides further support for the relevance of auditory testing within this population group. The consequent findings may then be instrumental in guiding sensory regulation of auditory function, for an unobstructed process of communicative interaction.

Hearing impairment inevitably affects health-related QoL at a psychosocial level (Chia et al., 2007). It introduces a range of activity limitations and restrictions in successful participation in daily life. Hearing loss in later adulthood may trigger an identity crisis and reactive depression may occur (Dewane, 2010). Associated psychosocial effects of hearing impairment include anxiety, distress, loneliness, somatization and poor social functioning (Nachtegaal et al., 2009). Additional consequences of impaired hearing include difficulties localizing sounds

and/or the threat of being unable to detect warning signals (Hallberg et al., 2008). The effects of hearing impairment may extend to activities such as watching television, understanding whispering, being unaware someone is talking and/or listening to the radio - all of which form important contributions to maintaining a sense of involvement in the world around. This is particularly relevant when dealing with a loss of multiple other functions (Dewane, 2010). Hearing loss furthermore introduces a false sense of understanding due to misinterpretations of speech sounds, creating feelings of disillusionment (Nachtegaal et al., 2009).

Since a sense of belonging is largely associated with mental health, a loss of hearing creates a psychological sensory confinement with harmful negative effects on individual belonging and mental wellbeing (Dewane, 2010). This has serious implications on an individual's ability to maintain identity, potentially resulting in communicative limitations. This leads to difficulty thinking, concentration and boredom. The resultant inattentiveness, distraction, isolation, depression and in some instances dementia leads to the most serious consequence of hearing loss - abandoning participation (Dewane, 2010; Gates & Mills, 2005).

The social stigmas associated with hearing impairment in adults include labels such as troublesomeness, slow-wittedness and tiresomeness (Dewane, 2010). The internalization of such prejudices result in a further blow to an individuals' self-esteem leading to depressive exhaustion and marked changes in individual personality (Dewane, 2010). When considering dementia and hearing loss it is also worthwhile to consider the related changes in character and personality. Where changes in character and personality occur, subjective detection of altered cognitive abilities may be impeded by the possibility of impaired hearing. This subsequently precludes

conclusive decision-making and recurrent communicative breakdown continues (Leigh et al., 2003). In instances where this is a direct result of impaired hearing, damaging consequences for psychosocial wellbeing and interpersonal relationships continue to be exacerbated in the midst of blurred diagnosis (Erber & Scherer, 1999).

Restrictions on an individual's execution of activities involving an exchange of information (e.g. expressing needs and wants, medical enquiries, legal interactions such as drafting of wills) impose barriers to independent functioning. Dysarthria in combination with hearing impairment results in voluntary or involuntary exclusion from social situations for fear of humiliation, isolation and loss of dignity (Foley et al, 2007; Kelly et al., 2003). Individuals typically report experiencing a lack of authenticity and feelings of alienation from people and activities that previously may have created a sense of belonging. This gradually leads to social exclusion, manifesting in the form of voluntary or involuntary withdrawal or avoidance of social contact. Social exclusion occurs when individuals lack a sense of integrated involvement and are deprived of participation in organized activities, meaningful roles or interactive successes (Mathers, Smith & Concha, 2003). Social exclusion and the consequences thereof will be further exacerbated by co-occurring physical limitations. Themes including a loss of autonomy and lack of dignity that arise as a result of social exclusion are closely related to individual desire to hasten death when living with a terminal illness (Kelly et al., 2003). The effects of hearing loss can therefore not be underestimated as a secondary feature separate from the disease itself.

It has been found that female participants with hearing impairment experience feelings of annoyance, irritation and anger (Hallberg et al., 2008). Male participants, who generally failed

to recognize the extent of their hearing difficulties, reported these feelings to a lesser extent. The application of non-verbal communication strategies, serving as a solution to facilitate the enhancement of participation in daily life, was also reported. However, these behaviours were noted more readily in female participants in contrast to male participants who generally showed more denial associated with hearing loss (Garstecki & Erler, 1999; Hallberg et al., 2008). The denial of hearing loss hinders success in addressing activity limitations and amplifies the restrictions on participation.

Despite the negative consequences brought about by hearing impairment, individuals with hearing impairment underestimate the consequences and the extent of withdrawal from interactions and/or avoidance of listening situations (Hallberg et al., 2008; Dalton et al., 2003). It is postulated that the avoidance of listening situations occurs as a result of the individuals' failure to accept their difficulties and pursue audiological rehabilitation (Dalton et al., 2003). It is further suggested that certain individuals show preference for self-deception as a means of reducing the discrepancies between reality and their perceived self-image (Hallberg & Barrenas, 1995). In a disease such as MND where individuals deal with the constant need to redefine their perceived image according to the gradual loss of functional abilities, secondary impairments, such as hearing loss, may not be readily acknowledged, and are consequently addressed with less urgency, if at all.

The effects of hearing impairment on the social and psychological wellbeing of individuals are hence profound (Ross & Deverell, 2004). The resultant social withdrawal, avoidance of previously positively associated hobbies and/or activities, exclusion from social

exchange and loss of empowerment pose obvious risks, further increasing the demands and stress of living with MND. Essentially, the absence of communicative success results in the patient becoming locked in; physically present, although immobile; and cognitively aware, amidst social exclusion (Ross & Deverell, 2004).

Contextual Factors: MND and Hearing Loss.

The contextual factors outlined by the ICF framework comprise external environmental factors and internal personal factors (WHO, 2001). Environmental factors that have an impact on the functioning of individuals with disability include the products and technology available to the individual. Alongside this are the natural environmental and human made changes that serve to facilitate psychosocial wellbeing, support and relationships. Personal factors such as individual attitudes and coping mechanisms for acknowledging and accepting diagnosis, alongside environmental factors are integral to the maintenance of individual QoL extending beyond the physical definition of the disease (WHO, 2001). For an individual with MND, emphasis needs to be directed at maintaining and improving QoL (McLeod & Clarke, 2007). Factors within the environment can hence be instrumental in either facilitating or limiting activities or participation restrictions faced by individuals with MND.

Individual attitudes and the attitudes of those around them form a key component in dealing with disease. The diagnosis of MND evokes various strong psychological reactions to the constant adjustments to the loss of functional abilities, loss of autonomy and self and the reality of death as the disease progresses (Averill, Kasarskis & Segerstrom, 2007). These experiences of loss arouse signs of depression, including withdrawal and self-isolation, often

resulting in a decline in individual willingness to prolong and sustain life (Erdman, 2009; Kelly et al., 2003). Beck's cognitive theory of depression suggests that the cognitive profile of depression is set off and revolves around themes of loss (Erdman, 2009). The ICF framework supports the notion that promoting QoL extends beyond the alleviation of physical burden and is rather more closely correlated to suffering, social support and sense of autonomy (McLeod & Clarke, 2007). This is supported by Simmons, Bremer, Robbins, Walsh and Fisher (2000) who found that the compensation for physical symptoms in MND does not markedly correlate to the alleviation of emotional distress. The latter is however more closely related to the existential and psychological aspects of life.

In a study conducted to understand the personal perceptions of QoL of individuals living with a progressive disease, participants expressed a strong desire to maintain their sense of self and to be viewed independently from their disability (Foley et al., 2007). It was further found that the ability of an individual to maintain a level of independence is largely determined by the interactions he/she engages in (Foley et al., 2007). This emphasizes the importance of functional auditory ability, without which these interactive processes are jeopardized.

Dignity was raised as an important theme linked to the individual's need to be respected and viewed independently of the disability. The lack of this was closely related to individual desires to hasten death (Foley et al., 2007; Kelly et al., 2003). Control, in terms of individual need to be *in* control and acknowledgement of the uncertainty paired with MND, creates feelings of vulnerability and dependency. Control is also linked to the sharing of thoughts and being respected in the decision-making process, which additionally add to the psychological effects of

MND (Foley et al., 2007).

Studies have indicated that individuals with progressive neurological diseases inevitably adapt to the disease and the progressive loss of physical function through a process of acknowledgement (Hardiman et al., 2004; Ross & Deverell, 2004). These individuals acknowledge the challenge of coping with feelings of loss and the strain of being physically dependent on others. This, when paired with feelings of vulnerability, results in a shift of focus being placed on maintaining the locus of control at an interactive level, thereby empowering themselves as autonomous in daily interactive experiences (Foley et al., 2007; Hallberg, 1999). Essentially, individuals are able to lower their personal expectations surrounding physical activity levels. They redirect this attention to other aspects of their lives in order to maintain a sense of self-worth and view themselves as purposeful contributors to the world around them (Hallberg et al., 2008; Ng & Khan, 2011). It is therefore through the control of environmental factors (such as the alleviation of certain symptoms, support and individual attitudes) that individual participation is maintained.

It is postulated that the obstacles to effective communication faced by individuals with MND are brought about by dysarthria, hearing loss or a combination of the two. This threatens autonomy and engagement in activities of daily living. Individual attitude towards MND, living with the various physical effects and the associated emotional and social consequences plays a critical role in the manner an individual deals with MND (Ball et al., 2004). The use of products and technology such as wheelchairs to aid mobility, adaptive eating equipment to accommodate eating difficulties and changes to the environment such as ramps and home modifications to

accommodate the changes in mobility can promote an improved attitude towards life (Ball et al., 2004).

Similarly, environmental factors related to communication difficulties (e.g. speech and hearing) can also enhance QoL for individuals with MND. Various communication-based strategies have been developed to facilitate the communicative success of individuals with dysarthria and subsequently reduce the activity limitations they face (Kuhnlein et al., 2008; Murphy, 2004; Leigh et al., 2003; Grauman, Betke, Gips, & Bradski, 2001; Lasker & Bedrosian, 2000). These strategies may include vocal strategies and/ or environmental alterations such as ensuring listeners have a full view of the speaker and/or AAC strategies. AAC strategies assist where deterioration in hand function and functional speech prevents alternate forms of communication (Leigh et al., 2003; Lasker & Bedrosian, 2000). These may include unaided strategies (such as facial expressions, gestural communication and eye movement) or low technology systems (such as alphabet cues and boards, and communication boards). Often eye gaze, a form of direct selection where individuals perform pre-learned eye movements directed to a communication board, may be utilized (Grauman et al., 2001). High technology AAC systems (such as speech generation systems or specialized eye tracking technology) may also be used to facilitate expressive communicative abilities (Murphy, 2004).

The use of AAC strategies and aids accommodate loss of expressive language abilities in MND, however facilitation of auditory deficits amongst this population is not met with equal urgency and importance (Dalton et al., 2003). This occurs despite the extent to which psychological and psychosocial factors related to hearing impairment restrict daily activities and

participation (Gates & Mills, 2005). Solutions to auditory reception breakdown should consequently not be excluded from the management of individuals with MND.

The benefits of achieving sensory regulation, often through the introduction of a hearing amplification device (e.g. a hearing aid) or communication training, positively contribute to QoL (Paolo et al., 2008). It has been found that the use of hearing aids demonstrate positive effects on an individuals' self-perceived hearing handicap, further supporting the benefit of identifying impairment and promoting the urgency for achieving successful sensory regulation (Tesch-Romer, 1997). Communication training, offered by audiologists, offers paired communication partners strategies to alleviate the extent of communicative breakdown. These strategies include non-verbal communication training (such as lip reading or sitting closer to a speaker), environmental modifications (furniture rearrangement, lighting changes) and psychosocial support (counseling) (Hallberg et al., 2008; Kramer, 2008; Michaud, Burnand, & Stiefel, 2004). These rehabilitative approaches serve to enhance communicative effectiveness and in doing so promote social inclusion (Gates & Mills, 2005; Michaud et al., 2004).

These rehabilitation or facilitation-based solutions form the foundation of the ICF framework, and serve to implement solutions that will overcome, alleviate or minimize an individuals' difficulties. This is particularly relevant in instances where disease is chronic and bodily structural and/or functional damage cannot be returned to normalcy (Gagne et al., 2009). Equipping the individual with the skills to communicate, assists in keeping him actively involved in the world around and lends the individual a degree of independence and autonomy in the face of irreversible physical decline and loss of physical function.

In addition to the personal effects of hearing impairment, hearing loss also threatens the functional success of relationships within the family structure (Hallam, Ashton, Sherbourne & Gailey, 2008). The value of support systems is highlighted by the ICF as an environmental factor contributing to the overall wellbeing of the individual (WHO, 2001). Numerous studies have reported that hearing loss does place considerable strain on family structures and enhance their vulnerability to failure (Hallam et al., 2008; Armero, 2001). However, the effects on the family/support structure appear to vary based on the extent and severity of the hearing loss. Family members of individuals with hearing impairment report significant increase in frustration and irritation and the effects are documented to reverberate throughout the family system (Dewane, 2010; Armero, 2001). The negative impact hearing impairment poses on intimate relationships, emphasizes the maladaptive behaviour and coping strategies implemented by a couple in crisis due to hearing impairment (Armero, 2001). Caregivers of hearing impaired individuals identified emotional consequences including deep resentment, anger, reclusiveness and depression. The caregivers were reported to be twice as likely to complain about reduced social activities than the individuals with hearing impairment. Affected individuals in turn begin to experience isolation from immediate family structures and a sense of guilt for their role in the breakdown of successful communication (Armero, 2001). This is supported by Dewane (2010) who reported that individuals with hearing impairment experience feelings of remorse for the constant need to request repetition. They further reported distrust, nervousness, irritability and reduced self-image associated with the effects of hearing loss on a family structure.

The devastating effects of hearing impairment heavily threaten the maintenance of caregiver-patient relationships and family structures. This is particularly true when hearing

impairment is superimposed on the additional challenges faced by individuals with MND and their families. These effects of hearing loss increase the psychological burden posed on individual perspectives of autonomy and QoL, in addition to having a marked impact on caregiver dynamics.

The process of providing an individual with the options available to alleviate or minimize the effects of a hearing impairment, allows for an inclusive management approach where the individual is able to prioritize rehabilitation goals. This promotes the likelihood of individual motivation to apply the outlined strategies, which have shown to have a positive effect on individual attitudes and behaviours as they encourage a sense of autonomy, empowerment and personal growth (Gagne et al., 2009). This in turn will promote positive dynamics within an individuals' support system.

The ICF framework therefore views the individual holistically and promotes enhanced activity and participation in daily living, through manipulating environmental factors to limit the extent of the physical, social and emotional effects of MND. To achieve success, a multi-disciplinary approach to patient management is paramount.

Multi-Disciplinary Management in MND

The management of progressive degenerative diseases should be directed towards managing the primary presenting symptoms and rehabilitating secondary symptoms (either directly or indirectly related to the disease itself). It is the role of the multi-disciplinary team (MDT) to manage all symptoms in a manner that affords the individual the time and ability to

dissociate worth from physical capability and reassign value to autonomy and maintenance of dignity at a psychological, social and existential level (Foley et al., 2007). The primary members of the MND management team include the neurologist, respiratory physician and gastroenterologist. These team members are responsible for the management of physical symptoms associated with breathing and swallowing, without which prolonged survival would not be possible. The secondary members of the MND MDT team are typically reported to comprise of the care coordinator, caregiver, general practitioner, occupational therapist, physiotherapist, speech therapist, dietician, social worker, palliative care teams and psychologist (Leigh et al., 2003; Mitsumoto & Del Bene, 2000). Although hearing impairment has been identified as one of the significant risk factors associated with a reduced QoL (Hallberg et al., 2008), the audiologist is not included in the MDT for individuals diagnosed with MND. This highlights an important gap in the process of promoting and enhancing QoL amongst the target population. Allowing an untrained professional to deal with the effects of hearing loss may result in an underestimation of the extent of frustration or misattribution the level of frustration (Dewane, 2010). The misattribution of the changes in personality associated with hearing impairment lead to unintentional mismanagement of the patient, further exacerbating the consequences of hearing loss (Dewane, 2010).

Ward, Phillips, Smith and Moran (2003) highlighted the importance of multi-disciplinary teamwork in patient management as an essential cornerstone to effectively managing individual needs and wants. An evaluation of the benefits of attending clinics catering specifically to the needs of individuals with MND revealed that clinic attendance was associated with a 47.3% decrease in one-year mortality risk (Traynor et al., 2003). A 7.5 month extension for median

survival was documented, while an extended 9.6 month prognostic improvement was documented for individuals with bulbar onset MND. This positive prognostic data was attributed to be a result of individuals being exposed to the coordinated symptomatic and disease-modifying therapies and treatments available as well as a consistent support structure (Traynor et al., 2003). Coordinated services are believed to eliminate the disjointed, overwhelming plethora of support services available to individuals with MND and their families (Ward et al., 2003). In contrast, the use of fragmented services and referrals result in delays in referral time frames, follow up and inter-professional communication, inevitably leading to misguided decision-making (Ward et al., 2003).

While the outcomes of coordinated services support the benefits of a multi-disciplinary approach to individual management, social, emotional and economic factors also play a role in attending such clinics (Ward et al., 2003). The implications of hearing impairment on social and emotional wellbeing, threaten individual willingness and motivation to attend such clinics, further bringing to light the value and relevance of inclusion of the auditory discipline within the MND management team.

Conclusion

MND is now referred to as a multi-system disease (Shaw, 2005; Isaacs et al., 2007; Pugdahl et al., 2007). The likelihood of MND-related sensory degeneration, such as hearing loss, is suggested as a possible presentation of disease variants. The use of invasive ventilation promotes the prolongation of disease course allowing neuronal categories that are susceptible to degeneration to become clinically apparent (Isaacs et al., 2007; Shaw, 2005). Furthermore, the

use of long-term ventilation as a life prolonging measures increases the clinical relevance of diagnosing atypical symptoms such as hearing loss (Maier et al., 2009). This is especially relevant in the older age groups where the associated natural decline in hearing ability is prevalent. The importance of clinically relevant research related to the wider spectrum of MND-associated features is essential and will assist in designing appropriate management plans for the individuals diagnosed with MND. This is critically important for the purpose of complex decision making as well as the modification of electronic communication systems and palliative care (Maier et al., 2009). Despite the tangential shifts from health-related symptoms to the social effects of a disease, as supported by the ICF, the effects of MND related to aspects of daily survival are often not viewed to be directly disease-related and hence are less readily addressed (Hardiman et al., 2004). The interaction of the physical limitations brought about by motor neuron degeneration paired with a loss of speech and verbal communicative abilities lead to limitations in communicative effectiveness and may result in withdrawal and avoidance of communicative interactions by the affected individual. These factors, in addition to the secondary symptoms that arise as a direct or indirect result of MND are likely to significantly impact on QoL and threaten participation in activities of daily living. Failing to acknowledge these symptoms ultimately impacts individual ability to take control of the disease and in contrast allows the disease to control the individual (Hardiman et al., 2004). Acknowledging the presence of hearing loss therefore ensures necessary modifications to facilitate receptive communication abilities. This in turn facilitates communication and promotes individual participation in daily living to proceed in a manner that is conducive to the maintenance of social inclusion and autonomy.

Due to the dearth of information on the effects of sensory degeneration in MND on the auditory system, the question of whether individuals with MND present with hearing loss was posed. It is postulated that individuals with MND may present with either a CHL or SNHL.

Chapter Three

Methodology

Introduction

This chapter provides a detailed review of the research methodology used in this study. The primary and secondary objectives of this study are identified, followed by a discussion of the research design. The research phases are presented, followed by the pilot study, its' findings and the recommendations that were formulated as a consequence. Lastly, a detailed review of the main study is described, including participant description, measuring instrumentation, data collection and data analysis procedures.

Objectives of the Study

Primary Objective.

The primary objective of the study was to determine the prevalence of hearing loss in South African individuals with adult onset MND.

Secondary Objectives.

The following objectives delineate the means by which the study was realized:

- To describe the audiometric findings in terms of the type, degree and configuration of hearing loss.
- To describe the perceived implications a hearing loss would impose on the psychosocial wellbeing of individuals with MND.
- To describe the perceived functional abilities in order of importance as reported by

individuals with MND and their caregivers.

- To describe perceptions of individuals with MND and their caregivers related to the value of auditory testing and diagnosis.

Research Design

A research design is defined as a plan of procedures to be used for the collection and analysis of data in order to evaluate a particular theoretical perspective (Denzin & Lincoln, 1998).

A mixed research method was employed for the current study. This study was executed as an exploratory, descriptive, cross-sectional research design, adopting qualitative and quantitative approaches to the different aspects of data collection and analysis. Triangulation, the method of combining a minimum of two theoretical perspectives or methodological approaches in a single study served to strengthen confidence in the research data by increasing the reliability and validity of qualitative findings (Thurmond, 2001). It was acknowledged that a dual qualitative-quantitative method such as the one applied to this study needed to be approached with caution, ensuring that the quality of data analysis and capturing was not sacrificed for data quantity (Silverman, 2000). Ultimately however this method of data capture is particularly well supported in instances where a limited and restricted sample population, such as the MND population highlighted in this study, is utilized. This was hence identified as a key design to promote optimal realization of the outlined research aims (Thurmond, 2001).

The **qualitative** component served to aid in exposing meaningful information, which may otherwise have remained unknown with the use of only quantitative data collection (Cresswell,

2003). This was applied in the acquisition of data and analysis of participant responses to the Hearing Experience Questionnaire (HEQ). Due to the limitations in sample size, qualitative review of audiological presentation and auditory related symptoms were analyzed using a descriptive approach, since quantitative numerical analysis of these findings alone would raise questionable strength of generalization. Qualitative analysis was applied to this study through the use of thematic content analysis. This design allows for categories and themes to emerge from the acquired data rather than preconceived categories (Kondracki & Wellman, 2002). Furthermore, the theoretical freedom of this design offers a flexible research tool, potentially exposing a rich, yet complex account of the acquired data (Braun & Clarke, 2006). Thematic content analysis is also an appropriate design in instances where existing theory or research literature on a phenomenon is limited (Hsieh & Shannon, 2005). The latter is true in the instance of MND and subjective auditory experiences and perceptions pertaining to hearing and communicative handicap levels as perceived by individuals with MND.

The use of a **quantitative** platform served to enhance the understanding and identification of common patterns amongst research participants allowing for broader comparisons to be made (Durrheim, 1999; Thurmond, 2001). This being said, the generalizability of information acquired on a limited sample population and size, such as the eight participants included in the current study, was acknowledged as a limitation and had to be approached with caution. Where the purpose of this research was to test unstudied domains associated with auditory ability that are atypical to those known MND symptoms, the acquired information also served to facilitate future studies where a larger sample size allowing for greater generalization may be possible (Creswell, 1994). Further benefit from quantitative research designs lie with the knowledge that

subjective opinion has minimal interference in the data analysis process, thereby encouraging greater consistency and reliability of results (Keller & Warrack, 2000).

The quantitative approach to of the current research followed a descriptive, cross-sectional, exploratory research paradigm. Furthermore, a multiple single case design was selected to support the qualitative component of this study.

A **descriptive** research design places emphasis on examining group differences and developmental trends amongst variables through the use of laboratory measures and various kinds of tests (Schiavetti & Metz, 2002). **Descriptive research designs** place emphasis on arranging, summarizing and presenting a set of data in a way that ensures that only meaningful elements of the data is extracted and interpreted accordingly (Keller & Warrack, 2000). Furthermore, a descriptive research design is useful when there is no theory to work from and/or when exploring a new area which has not been researched before on prior occasions (Mark, 1996). For the purpose of this study, a descriptive research design was used to satisfy the description of auditory test results based on the type, degree and configuration of auditory thresholds and objective test findings. This study did not aim to establish cause-effect relationships as in experimental research, but rather aimed to address whether there may be a correlation between the presence of MND and a hearing loss, deeming a descriptive design appropriate.

While descriptive research designs are advantageous due to their relatively non-invasive nature, a number of risks linked to observer and sample bias exist (Schiavetti & Metz, 2002).

Interactional bias occurs when a participants' behaviour is influenced by interaction with the researcher, while non-interactional bias occurs when the researcher affects the recording of the participants' performance. This can be done either through misinterpretation of results and/or dishonesty associated with recording of results and data (Schiavetti & Metz, 2002). This study posed a greater risk for non-interactional bias, which was addressed by abiding to specific test protocols and inter-rater result interpretation. Sample bias posed a risk to the external validity of this study (Schiavetti & Metz, 2002), although a series of pre-established criteria for inclusion were applied. This ensured that any willing participants who did not fall within the realm of these predetermined criteria were not included in the study.

A **cross-sectional design** was utilized since this study did not serve to provide evidence for the changes in hearing ability over the course of the participants' disease progression. Instead the current study served to obtain evidence for the presence or absence of hearing loss in MND individuals at a single time, with a single period of data acquisition (Leedy & Ormrod, 2001).

An **exploratory research** design was applied for the purpose of obtaining additional knowledge surrounding an area of limited available research (Sekaran, 2003). Exploratory research relies on extreme caution in making definitive conclusions. It however serves as an initial reference for insight into domains that have not yet been clearly defined (Babbi & Mouton, 2001). Due to the low prevalence of MND, further narrowed by the limited geographical region (Gauteng) being included in this study, the opportunities for experimental investigations on large sample sizes were not possible in this instance. An exploratory study therefore served to

facilitate the process of exploration, analysis and description of hearing loss in this sample population, offering insight and information that may be used as rationalization for studies where a larger sample size is obtainable (Pannbacker, Middleton, & Lass, 1994). It is therefore evident that although this approach is not always useful for definitive conclusions, it offers an opportunity for secondary research to branch out and acquire more comprehensive understanding into a condition, such as MND and its atypical symptoms.

A **multiple single case study method** was further utilized in this exploratory study. Case studies are typically designed to examine and analyze information on specific individuals in more depth and in the specific context of the research (Lewis, 2003). This is beneficial as a means of limiting the loss of essential information, which is typically at risk of occurring when examining group data (Lewis, 2003). One of the key benefits of engaging in case study research is associated with its' powerful ability to draw attention to phenomena that may have been overlooked in clinical practice (Grbich, 2003). The investigation of auditory presentation remains one such investigative domain that to date is undocumented as an atypical symptom of the disease. This lends itself to question whether hearing loss is not a symptom or concern among the MND population or whether auditory involvement and its' impact on QoL is overlooked in the intense management process of those typical MND-related symptoms.

Case study research has however also been criticized for its expansionistic nature as it does not allow for generalizations, nor does it allow for causal relationships to be established (Lewis, 2003). In essence, while case study research allows for a comprehensive, holistic review of individual perceptions and experiences (Grbich, 2003), a problem presents itself in applying the

acquired information to society in general. It is important to acknowledge that based on the limited number of persons with MND and the heterogeneity of this population a large experimental design was deemed inappropriate. A case study method thus allowed for more information to be acquired from fewer participants. This was further addressed through the use of *multiple* single case studies. Since no single participant study would be suitable in representing the complete view of auditory experiences and symptoms, each case was reviewed at an individual level. Following this, results were reviewed at a comparative level across cases serving to enhance the generalization of data across the limited sample size. A case study design is further limited by inter-subjectivity (Gummesson, 2007). This refers to the content analysis of a case, based on the interpretations and perceptions of the researcher. The latter typically tends to be a representation of judgments based on the individual world-view of the interpreter and therefore may vary from one person to another (Gummesson, 2007). The use of qualitative and quantitative measures, data coding and inter-rater analysis served to limit the impact of subjective interpretations and redirect the focus to reporting the content provided by each individual/caregiver and emergent themes rather than the researchers' judgments alone.

The application of a mixed research method essentially allowed for the limitations of quantitative analysis on a small sample size to be compensated by the strengths of a qualitative design (Creswell, 2003). This yielded a design that acknowledged its' strengths, compensated for its limitations and ensured a holistic, relevant and integrated use of the collected data.

Research Phases

The research comprised three major phases: Phase 1, the development phase, Phase 2, the

pilot study and Phase 3, the main study. The phases are outlined in Table 3.

Table 3

Research phases

Objectives	Description of activities
Phase 1: Developmental phase (April - June 2010)	
To determine the viability of locating participants within the geographical area	Contact was established with the Motor Neuron Disease Association of South Africa (MNDSA) (Appendix B and C), Gauteng Department of Health (GDoH)) and four neurologists in private practice in Gauteng.
To identify, select and modify measuring instruments be used in the study	A test protocol was designed for audiological testing, a case history interview (Appendix D) and HEQ (Appendix E & F) were developed and the HHIA (Appendix G) was selected.
To train the research assistant to ensure the reliability and validity of the results (Schiavetti & Metz, 2002).	A specific protocol detailing test measures and result interpretation was consolidated with a research assistant. All result interpretation was done in accordance with standardized protocols and published literature.
Phase 2: Pilot Study Phase (July- September 2010)	
This identifies practical problems in the research procedure and problem-solves procedural aspects and/ or flaws in instrumentation, which pose a threat to the execution of the main study (Teijlingen, Rennie, Hundley, & Graham, 2001).	
Execution of full test protocol to finalize the data collection measures, procedures and protocols to be used in the main study	Individuals provided informed consent (Appendix H). Two individuals complying with inclusionary criteria for participation underwent the full test battery. They further provided a review of the experience to aid identification of methodological obstacles and modification of these obstacles for correction.
Determine the face validity of HEQ	The researcher paired with an audiologist with five years clinical experience in the audiological field reviewed the face validity of the information acquired from pilot study participant responses to the questionnaire.
Phase 3: Main Phase (September 2010– April 2011)	
Identification and selection of participants	This was done in accordance with the inclusionary and exclusionary criteria established during the developmental study phase. A total of eight individuals met these criteria and were selected for inclusion in this study.
Data collection	Execution of audiometric testing and completion of individual and caregiver questionnaires marked this step in the main study.
Data analysis	Organization and logical arrangement of acquired data as well as statistical analysis and review of acquired information for descriptive review.

Pilot Study

The objectives of the pilot study were to (i) finalize the measuring instruments; (ii) determine the equipment to be used in the study; (iii) determine the need for adaptation of test

equipment; and (iv) determine the total testing time per participant. The results of the pilot study were then used to refine the methodology and to reduce the threats to internal and external validity (Teijlingen et al., 2001).

Participants.

Individuals provided informed consent (Appendix H). Two individuals with MND and their caregivers were included in the pilot study. These individuals were both members of the MNDSA and met the selection criteria as outlined for the main study. Both individuals were diagnosed with mixed onset MND and were classified as 'severe' according the MND Classification (Riviere et al., 1998). Participant one was a male aged 65.6 years, diagnosed 54 months prior to the current study with initial symptom presentation 66 months prior to the current study. Participant two was a female aged 49.6 years, diagnosed 24 months prior to the study, with initial symptom presentation 33 months prior to the study. Participant one reported occasional difficulty in noisy environments where participant two did not report experiencing decreased hearing sensitivity, but occasional instances of hypersensitivity. Data collected for both participants was included in the main study due to the limited sample size.

Procedures.

Once written permission was obtained from the MNDSA (Appendix C), the care worker was requested to identify potential participants and distribute informed consent forms. Two participants, who met the selection criteria, completed and returned the informed consent forms. Appointments were made at times most convenient to the participants and their caregivers.

Objectives.

The objectives, materials, equipment, procedures, results and recommendations made after the completion of the pilot study are outlined in Table 4.

Table 4

Objectives, materials, equipment, procedures, results and recommendation from pilot study

Objectives	Material/Equipment	Procedures	Results	Recommendations
To determine whether the required information would be obtained with the selected measuring instruments	Case history questionnaire	Reviewed by an independent research assistant prior to participant issue. The same procedures were used as for the main study.	Limited information regarding the hearing experiences, functional priorities and perceptions was obtained.	An additional set of questions (HEQ) was formulated for persons with MND detailing concerns regarding hearing loss – the symptoms, the need for diagnosis and the impact this would have on the social and functional abilities of the MND participant. Once developed this was issued to pilot study participants to allow for face validity review.
	HHIA	Reviewed by an independent research assistant prior to participant issue. The same procedures were used as for the main study. Data was encoded and analyzed using basic descriptive statistical procedures	The inventory offered valued differentiation between social and emotional involvement in participant's daily listening experiences as reported by the persons with MND.	No changes were recommended
	HEQ	Reviewed by an independent research assistant prior to participant issue. The same procedures were used as for the main study. Data was descriptively analyzed following a qualitative approach.	The HEQ provided data that fulfilled the outlined objectives of this study related to participant perceptions of hearing loss and the importance of auditory diagnosis. The HEQ neglected caregiver perspectives related to these themes.	An additional section of replicate questions were added to this questionnaire. The added set of questions was directed entirely at participant caregivers.

Objectives	Material/Equipment	Procedures	Results	Recommendations
To determine the equipment to be used in the study (refer to page 64)	Portable versus Conventional Equipment (AC 40 Audiometer versus Kuduwave 500)	The care worker and potential participants were consulted regarding the accessibility of the USHC. Result comparisons using portable versus conventional audiometers were obtained and reviewed for consistency	Based on feedback from the care worker, five potential participants reported difficulties in acquiring transport to the USHC for participation in this study. Discrepancies in test results were noted for portable versus convention audiometry (refer to page 63)	Testing at the USHC was deemed more appropriate and reliable when reviewed against the possibility of testing within the individuals home environment. Transport was offered to facilitate participation of individuals who were restricted in terms of transport to the test site.
To determine the total testing time per individual	Timer	The timer was set when data collection began (including rest intervals and general discussion)	The total time required for testing was approximately 178 minutes.	Participants need to be informed that about the estimated length of the appointment prior to appointment date. Principles of non-maleficence needed to be strictly adhered to and participant state carefully monitored by tester and caregiver

Selection of Equipment.

Testing procedures were executed in two different ways to determine the manner in which the most efficient and accurate test results were to be obtained:

University of the Witwatersrand Speech and Hearing Clinic.

The University of the Witwatersrand has a fully equipped audiology clinic with sound-proof test booths. Testing booths are separated by one-way glass allowing audiologist-patient contact to be maintained throughout testing. A fourth sound-treated testing room is equipped with the required materials and equipment for OAE and ABR testing. All rooms are within a close proximity to one another making it easy for participants to maneuver from one test room to another. Two of the booths are equipped with ramps, making wheelchair access into and out of the test rooms a relatively unchallenging task. Testing at the university clinic revealed optimal test conditions, allowing for testing to be executed in a manner that was efficient and consistent across individual test sessions. The benefit of fully calibrated equipment and a sound treated environment allowed for optimal test conditions and test results that were believed to be representative of the participants' auditory abilities.

Portable Testing.

The alternative option was that of portable equipment. The benefits of portable testing included the fact that the researcher would have been able to travel to participants' homes and conduct testing in an environment that was both comfortable and equipped for participants' needs. It was also initially suspected that this option would invite a higher positive response rate for participation in this study. This assumption however did not prove to be accurate, with only

one additional agreement for participation being obtained following the suggestion of portable testing. Portable testing also required a change in test equipment and implied the maneuvering of bulky equipment threatening the calibration of the equipment (Table 5).

Table 5

Test Equipment for both test conditions

	Otoscopic evaluation	**Immitance audiometry**	**Pure tone and speech audiometry**	**Otoacoustic emissions**	**Neurodiagnostic (ABR)**
Audiology clinic equipment	Otoscope	GSI 33	AC 40 Audiometer	Biological Diagnostic OAE	Interacoustics Eclipse
Portable testing equipment	Otoscope	GSI 33	Kuduwave 5000 *only pure tone	Interacoustics Eclipse	Interacoustics Eclipse

The KUDUwave 5000 is an automated computer-based audiometer with an option for manual use. The KUDUwave is a device that has met national (South African National Standards [SANS] 10182) an international standards for audiometer manufacturing and operation as well as United Kingdom and global organization certification (Mkwanazi, Mngemane, Molemong, & Tutshini, 2009). The KUDUwave 5000 utilizes circumaural headphones paired with built-in 3A insert earphones used as transducers. Additionally, the KUDUwave was developed with, a built-in sound blocking ability (ambidome) equivalent to that of a single wall soundproof booth. The benefit of this built in feature allows for hearing testing to occur with equal reliability both within or external to a sound booth. This device is further supported by a feature that allows for ambient background noise to be consistently monitored during testing, thereby alerting the tester should noise levels exceed those of compliant testing levels. A comparison of pure tone air conduction results for participant two using a conventional, non-portable audiometer (AC40) and a portable audiometer (KUDUwave 5000) is documented in Table 6. Testing with the conventional audiometer was performed in a sound-proof test booth,

while testing using the Kuduwave was performed in sound controlled settings in the natural environment of the individual i.e. bedroom/living room.

Table 6

Air Conduction Results, Participant 2 - Conventional versus Portable Audiometer

Audiometer	Ear	250Hz	500Hz	1000Hz	2000Hz	4000Hz	8000Hz
Conventional audiometer (AC40)	Right	10dB	10dB	10dB	5dB	10dB	10dB
Portable audiometer (Kuduwave 5000)	Right	30dB	25dB	15dB	10dB	10dB	10dB
Conventional audiometer (AC40)	Left	10dB	10dB	5dB	5dB	15dB	15dB
Portable audiometer (Kuduwave 5000)	Left	20dB	25dB	10dB	5dB	15dB	20dB

These results demonstrate inconsistency in findings particularly for the low frequency hearing range, where differences of up to 20dB were documented. Mkwanazi et al. (2009), reported test-retest reliability between a conventional audiometer and the Kuduwave 5000 to be exactly on par with exception to the low frequency range. An increase of 3dB across all frequencies, with exception to the low frequencies was documented between the Kuduwave and conventional audiometers (Mkwanzi et al., 2009). The latter is problematic as the introduction of a false positive conductive pathology diagnosis results in a management plan being devised in accordance with this pathological indication. Cautious cross-checking of a complete test battery would therefore be indicated and would threaten the exposure of poor cross-test correlation. While this unit of equipment reports the ability to eliminate of external noise interference through the built in ambidome paired with the sound elimination benefits of built-in insert earphones, the specificity of the pure tone thresholds recorded in contrast to conventional testing

posed a problem to the reliability of recordings. Furthermore the functionality of speech audiometry measures, in reference to speech reception threshold testing, provided questionable test reliability through pure tone average (PTA) and speech reception threshold (SRT) correlation scores. At the time of data collection, the Kuduwave was not yet designed to be used for executing most comfortable level thresholds, thresholds of discomfort and speech discrimination scores, all of which formed essential components of the outlined test battery and offer valued correlative information during the cross checking procedure.

In addition to the evident discrepancies in pure tone reliability, auditory brainstem response testing – a test of high sensitivity to external noise, internal noise and patient state proved problematic when used during home visits. While the researcher was to a certain degree able to control immediate external noise interference such as television sets, nearby conversations, telephonic noise and electrical noise sources from lighting or nearby electronic devices, these modifications could not replace the high quality noise control offered by a sound treated audiological test booth. Ambient noise, beyond the researchers' control, such as passing traffic resulted in a high recording rejection rate suggesting the need for increases in gain levels exceeding the recommended test parameters. This resulted in unfavourable trace morphology and poor repeatability of recordings. Further to this, each individual tested presented with differences in surrounding ambient and electrical sound interference, which could not be controlled outside a sound treated environment. For valid comparisons of test findings all participants are required to be tested under the same test conditions using the same test parameters. Pursuing home visits for data collection would have violated these conditions as set out by the equipment manufacturers. It thus became evident that these conditions were not

capable of yielding results worthy of diagnostic interpretation nor would they be truly valid for comparative purposes.

In summary, although home testing may have appeared to be a preferable method for participant recruitment and data acquisition, the actual execution of this was not feasible. Home visits proved effective in recruiting only one additional participant and the number of external, uncontrollable variables did not support the acquisition of audiological recordings of an acceptable quality and standard of reliability. The decision was therefore made to perform testing at the university clinic, where calibration of equipment and test conditions were consistent across all participants.

Summary

After completion of the pilot study minor modifications to the instruments and procedures were required. Amongst these modifications included the development of a qualitative questionnaire (HEQ) serving to explore individual perceptions relating to auditory abilities, impairment and the perceived value of auditory diagnosis. Following this, the pilot study further suggested a need for extension of the HEQ to incorporate a set of caregiver questions. The latter served to comparatively explore the views between individuals with MND and their caregivers in relation to the values of auditory abilities. The pilot study lead to changes in the initial intension of conducting testing at locations comfortable to the individuals e.g. their houses. Portable test equipment was deemed undesirable for the purpose of obtaining reliable, consistent results across the entire test battery. Consequently, a complete test battery comprising both behavioural and electrophysiological measures using equipment that was calibrated and consistent across all

individuals tested was viewed to be more beneficial to the acquisition of reliable and valid results across the entire sample. As a result of the relocation of testing to the USHC the pilot study allowed for accommodations for transport to be made, thereby aiming to facilitate individuals who chose to participant in the study but where restricted by issues of mobility and transport. Lastly, the pilot study allowed for the test duration to be taken into account and allowed the researcher to fully disclose details pertaining to the length of testing to individuals prior to their test date. This assisted in limiting individual anxiety on the test date and ensuring full disclosure prior to testing, allowing for individual withdrawal from the study prior to the test date should this information have been deemed undesirable.

Main Study

Participant Selection and Description.

Sampling strategy.

A non-probability, purposive sampling strategy was used to select participants who were representative of the MND population in South Africa. A purposive sampling strategy is ideal where the selection of a desired population is rare or difficult to locate (Leedy & Ormrod, 2001; Trochim, 2006). Purposive sampling strategy does however introduce a source of bias (Trochim, 2006), since selected participants may not always approximate the characteristics of the total population of potential participants (Welman & Kruger, 1999). In a bid to enhance the representativeness of the selected sample, the researcher attempted to recruit participants from various sources at a level of private and government healthcare institutions.

Participants were pursued through neurologists in private practice, Chris Hani

Baragwanath Hospital and the MNDSA. The care worker of the MNDSA contacted possible participants and supplied names of the participants who consented to take part in the study. In addition, the researcher attended monthly MNDSA support group meetings on three occasions and posted an invitation to participate in the study on the MNDSA online forum. All the participants who met the selection criteria were contacted by the researcher.

Participant Selection Criteria.

Participants were selected based on the inclusion and exclusion criteria as outlined in Table 7 and Table 8.

Table 7
Inclusion Criteria

Criteria	Justification	Method
Neurologist confirmed diagnosis of MND	Participants selected for involvement in this study were isolated to the sample population with MND. This ensured that findings were representative of the MND population and were not impacted by the inclusion of neurodegenerative population groups such as multiple sclerosis or MND 'mimic' syndromes (Swash & Desai, 2006).	The MND care worker accessed neurology reports confirming the diagnosis of MND.
Stage of MND	Participants in Stage I-IV were included in this study. The classification of participant stage of disease was done according to the Classification of MND (Appendix A) (Riviere et al., 1998). Due to the limited acquirable sample and the scope of the current study, participants at any stage of disease were extended the invitation to participate in this study. The current study, being at a preliminary level of determining the presence of hearing loss in MND did not explore the relationship between MND stage and severity of hearing impairment. Sample size, designated time frames for data collection and research scope did not support the exploration of this relationship. As a result, willing individuals at any stage of disease progression were accepted into the current study.	This information was acquired through the MNDSA care worker and through direct contact with the patients prior to selection.
Age	Participants within the age range of 30 – 66 years were included as studies suggest MND typically presents during the fourth to sixth decades of life (Shaw, 2005)	Participant age was acquired from the MND care worker and through a case history questionnaire.
English proficiency	To ensure reliability of test results were not negatively affected by a limited understanding of the English language, only participants who were proficient in English were included in this study.	This was determined subjectively by the care worker. The researcher confirmed language proficiency at the initial visit through conversational interaction.
Gender	While an equal ratio of male and female participants was targeted for comparative purposes, the ratio documented for male and female MND is 1. 5:1 (Wijesekera & Leigh, 2007), thereby suggesting a likelihood of more male participants included in this study.	Participant gender was noted through a case history questionnaire (Appendix D).

Table 8
Exclusion Criteria

Criteria	Justification	Method
Participant age (<30 years)	MMND presents in the second decade of life (Swash & Desai, 2006). Hearing impairment is reported as the major initial symptom with sixty percent of patients presenting with known hearing impairment (Nalini, Yamini, Gavatri, Thennarasu, & Gope, 2006). Inclusion of patients younger than 30 years of age may indicate a juvenile onset of MND or MMNDA and may not have been representative of the typical presentation of adult onset MND.	This information was confirmed through the care worker and Case History Questionnaires.
(>66 years)	Approximately 25 - 40% of patients older than 66 years old present with a hearing impairment (Yueh et al., 2003). This figure continues to increase with increase in age. The exclusion of participants older than 66 years old, serves to lessen the involvement of age related hearing impairment from test findings. Consideration for presbycutic hearing impairment was accounted for as a co-variant during the statistical analysis of data. A presbycutic hearing loss was detected through objective measures: DPOAEs indicating involvement of the cochlear (Prieve & Fitzgerald, 2002) and neurodiagnostic ABRs assists in the isolation of site of pathology.	
Age at diagnosis (<30 years)	This serves to exclude any incidences of Juvenile / MMND. The South African population is rich and diverse in ethnicity, therefore although MMND is more typical in the Western pacific regions, it is important to acknowledge the suspected underlying genetic connection to this form of MND (Nalini et al., 2006) and the risk of South African participants with an early onset of MND having a genetic history to those areas. MMND reportedly initiates presentation from as early as 19. 1 years of age (SD - 7.94 years) (Nalini et al., 2006). This suggests symptoms may present from the age of 11.1 to 27 years of age - a decade earlier than the typical symptom presentation in MND. Furthermore MMND has been clearly associated with bilateral sensorineural patterns of hearing loss, while this current study aims to investigate the presence of hearing loss in patients presenting with typical form adult onset MND, excluding early onset/ juvenile forms (Nalini et al., 2006).	The care worker confirmed this information through review of the participants' neurological report.

Cognitive impairment	MND reports indicate subtle patterns of cognitive dysfunction with impairments in verbal fluency, word-finding difficulties and attention deficits; however this is rare (Phukan, Pender, & Hardiman, 2007). Hence, while many of these neuropsychological features are not often observed in MND patients (Abe, 2000), cognitive dysfunction in the participants would negatively threaten reliability and validity of the sample population resulting in exclusion from this study. The researcher therefore chose to exclude this category of participant due to the threats this would have on confounding the results of the study.	This information was acquired from the MND care worker who had access to neurology reports.
Co-morbid factors	Co-morbid factors linked closely to hearing loss, such as diabetes, ototoxicity and noise exposure will result in exclusion. Co-morbidity refers to the presence of more than one distinct condition in an individual (Valderas, Starfield, Sibbald, Salisburg, & Roland, 2009). Co-morbidity is of importance due its impact on the uniformity of participant selection. Without consideration of co-morbid factors erroneous assumptions about causality and findings may be reached, subsequently negatively impacting the validity of a study (Valderas et al., 2009).	Participants were required to complete a case history form (Appendix D). This form included a checklist of illness/ disease with known auditory involvement.
Pre-existing hearing impairment	The inclusion of participants with a known hearing impairment prior to the diagnosis of MND would negatively impact the validity of the findings, since the documented hearing loss cannot be viewed to be associated with adult-onset MND exclusively.	Participants were required to complete a case history form, probing into the presence of a known hearing loss and previous audiological evaluations.

Sample Size.

A total of eight individuals with MND (*N* = 8*)* participated in the study. The primary caregivers for all eight individuals were also involved in responding to the HEQ. While the decision making process linked to sample size is highly dependent on sampling strategies and economic concerns, practical viability of the target population remains an essential area of consideration (Schiavetti & Metz, 2002).

Although this study had a small sample size, it is important to place the sample size in a broader context by considering the plausible MND population in South Africa. MND incidence and prevalence in South Africa is suggested to be consistent with worldwide figures of 1.2 per 100,000 and 5.2 per 100,000 respectively (Wijesekera & Leigh, 2007; Logroscino et al., 2008). This suggests that 66 new cases of MND will be diagnosed in South Africa annually, with 158 people currently registered with the MNDSA. Table 9 represents the gender and age distribution of MNDSA members in South Africa and the site of this study, Gauteng, at the time of the current study. Due to the limited population size, further reduced by rate of attrition and geographical range, the results of the participants from the pilot study were included in the total sample size and analysis of findings.

Table 9

Gender and Age Distribution of MNDSA members

	Total members	Male	Female	Age range
South Africa	158	111	47	31 – 82 years
Gauteng	65	37	28	31 – 79 years

At the time of this study, 158 patients were registered with the MNDSA, 65 of who lived

in the Gauteng region, suggesting that purposive sampling was necessary to ensure the selected participants were appropriate to the established criteria and population group (Mark, 1996).

A total of 44 information packs (informed consent forms, case history questionnaire, franked envelope) were distributed to potential participants. These were distributed through the MNDSA care worker, at MNDSA support meetings, MNDSA online forum, private neurologists and an out-patient at the Chris Hani Baragwanath Hospital. Figure 2 provides a graphical depiction of the return and attrition rate of potential participants. The majority of the information packs (30) were distributed at the MNDSA support group meetings. This yielded a return of 13 informed consent forms of which only 11 met the inclusion criteria. Of the 11 individuals who met the inclusion criteria, four passed away before data collection commenced. One of the two participants who did not meet with the inclusion criteria was already fitted with hearing aids bilaterally. The hearing abilities of this individual were reportedly actively monitored by an audiologist thereby ensuring all hearing related aspects linked to communicative success were managed by a qualified professional in the field. The second individual who failed to comply with testing criteria presented with a known, acquired hearing loss as a result of childhood measles. This individual, although previously under audiological management and fitted bilaterally with in-the-ear hearing aids, requested testing despite previous management. She was issued with an updated audiogram and referral letter for hearing aid reprogramming. The test results of this individual were not included in the study as a result of the evident co-morbid factors linked to hearing impairment. Three information packs were issued to individuals via the MNDSA online forum of which one was returned and complied with the criteria outlined for inclusion in the study. Ten information packs were distributed to private neurologists in Gauteng.

Only one potential participant returned the informed consent form but did not meet the participant selection criteria. A single outpatient from Chris Hani Baragwanath Hospital volunteered participation in the current study, however criteria for participation was not met resulting in the exclusion of this individual from the total sample size.

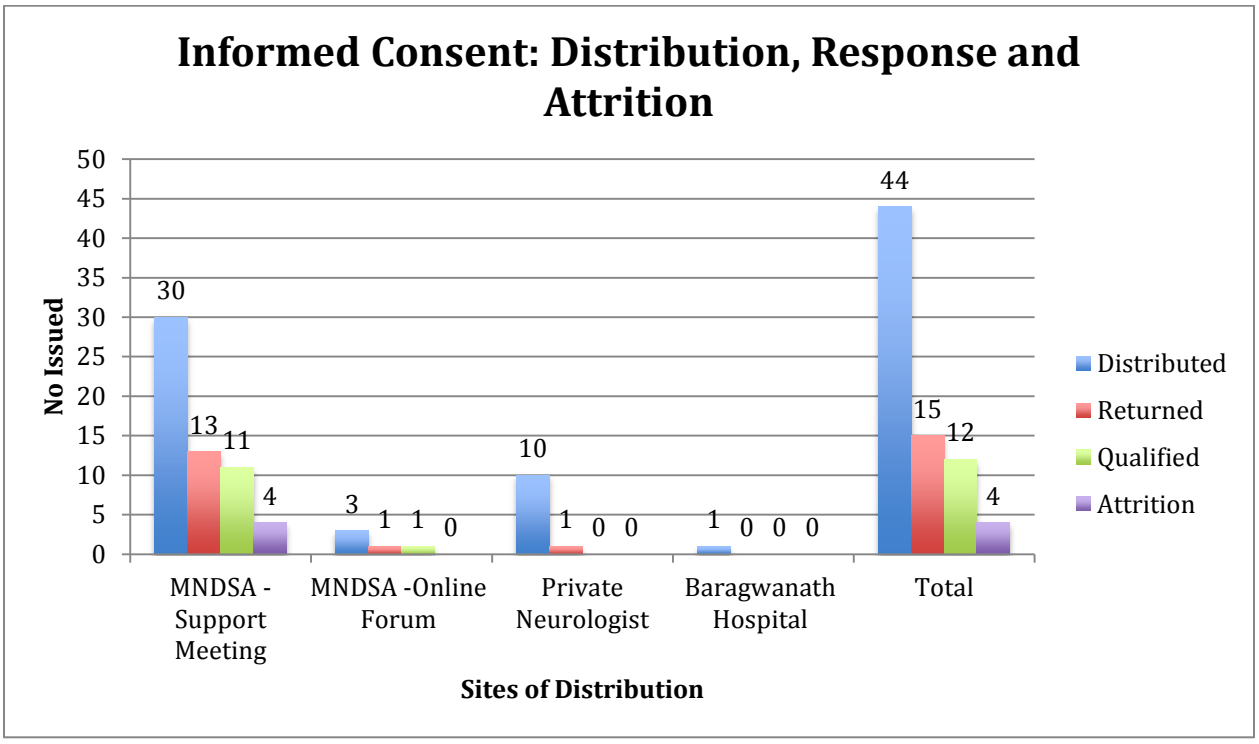

Figure 2. Informed Consent: Distribution, Response and Attrition

Participant Description.

A detailed demographic profile of each participant is set out in Table 10.

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1 Rife Therapy: the use of a Rife resonator generates resonance waves transmitted through handheld electrodes placed on the body, These electrodes create a negative polarity resulting in electromagnetic waves negating the reproductive ability and/ or the presence of a virus, parasite and/ or bacteria in the body.

Table 10

Participant and Caregiver Description

	P1	P2	P3	P4	P5	P6	P7	P8
Gender	Male	Female	Male	Female	Female	Male	Female	Male
Age (in years)	65,6	49,6	61,2	58,6	59,2	62,8	57,3	57,3
Since diagnosis	54 months	24 months	55 months	20 months	25 months	22 months	28 months	18 months
Since onset of symptoms	66 months	33 months	70 months	62 months	40 months	16 months	44 months	25 months
Type of MND	Mixed	Mixed	Bulbar	Spinal	Mixed	Spinal	Mixed	Mixed
Classification of MND	III	II	III	II	III	I	II	I
Primary caregiver	Wife	Husband	Wife	Husband	Life Partner	Wife	Husband	Wife
Caregiver Age	59,4	55,6	59,8	66,5	52,2	61,8	59,11	56,10
Reliance on others	Reliance for all activities except communication	Assistance for all activities except communication	Total reliance for all activities of daily living	Assistance with gross motor leg movement	Assistance for all activities except communication	Slight assistance required for fine motor skills	Requires assistance for fine motor skills	Mild weakness in leg movement, walking stick
Current medical & rehabilitative intervention	Neurologist and GP in the event of illness	Neurologist and GP in the event of illness	Neurologist and GP in the event of illness	Neurologist and GP in the event of illness	Neurologist and GP in the event of illness	Neurologist and GP in the event of illness	Neurologist and GP in the event of illness	Neurologist and GP in the event of illness
Previous rehabilitative interventions	Physiotherapy	None Reported	Rife Therapy 1 Physiotherapy Speech Therapy	Physiotherapy	Speech Therapy Physiotherapy	None Reported	Physiotherapy	Physiotherapy
Communication mode	Verbal – intelligible	Verbal – intelligible	Electronic Alphabet Board	Verbal – mildly dysarthric,	Verbal – intelligible	Verbal – intelligible	Verbal – intelligible	Verbal - intelligible
Auditory difficulty	No	No	No	No	Yes	No	Yes	No
Description of auditory Symptoms	Difficulty noisy environments Tinnitus Recruitment	Hyperacusis History of discharge	Hyperacusis Fluctuation (infrequent) History: right ear pain & discharge	Tinnitus Recruitment Difficulty noisy environments	Hyperacusis Tinnitus Recruitment Difficulty noisy environments	Hyperacusis Occasional Dizziness and Vertigo	Difficulty noisy environments + tinnitus	Dizziness Tinnitus Difficulty noisy environments

MND participants included an equal number of males and females. While this gender distribution offers balance for analytical purposes, it is not truly representative of the general population of persons living with MND, where a male to female ratio of 1,5:1 is typically reported (Wijesekera & Leigh, 2007). Of the caregiver participants five caregivers were female while three were male.

The average age for individuals with MND in the current study was 58.9 years (range: 49.6 – 65.6 years; standard deviation [*SD*] - 4.7 years). The average age of the sample was therefore consistent with the literature which identifies the forth to sixth decades of life being those typical of MND presentation (Shaw, 2005). The average age for caregivers was 58.8 years (range: 52.2 – 66.5 years; *SD* – 2.3).

Diagnostically, the average period since the initial presentation of symptoms to the month of data collection was 44.5 months (range: 16 – 70 months; *SD* - 19.87 months). The average period since the initial diagnosis to the month of data collection was 30.7 months (range: 18 – 55 months; *SD* - 14.97) Mitchell et al. (2010), identify an average time frame of 12 months separating initial presentation of symptoms from diagnosis. Donaghy, Dick, Hardiman and Patterson (2008) identified a median time from symptom onset to diagnosis at 15.6 months, although the literature generally identifies a median time frame from initial symptom to diagnosis at 10.6 – 17. 5 months (Rosatti et al., 1977; Donaghy et al., 2008). This supports the median time frame of 16. 5 months for participants in the current study.

Individuals varied in type of MND diagnosis, with 62.5% (*n = 5*) presenting with mixed

MND. Twenty five percent (*n = 2*) of participants were diagnosed with spinal MND and 12.5% (*n = 1*) were diagnosed with bulbar MND. Individuals also presented in varying stages of disease progression where 37.5% (*n = 3*) of the total sample presented in stage III and 37.5% (*n = 3*) in stage II. A further 25% (*n = 2*) of the participants presented in stage I of disease progression (Appendix A). All participants listed their primary caregivers as either husband, wife or life partner.

The acknowledgement of hearing loss and/or auditory symptoms demonstrated further variability amongst the participants in the current study. Twenty five percent (*n = 2*) of the participants acknowledged some experience of hearing difficulty while 75 % (*n = 6*) reported no auditory impairment. Despite this, when probed all participants were able to associate with varying forms and extents of auditory symptoms such as tinnitus, hypersensitivity to sound and recruitment.

Twenty five percent (*n =2*) of individuals were exposed to the discipline of speech-, language pathology, primarily for speech related management and compensatory strategies. No involvement from an audiologist was reported in terms of auditory evaluation or aural rehabilitation strategies paired with the set-up of communication systems.

Equipment and Measuring Instruments.

A discussion of the equipment and measuring instrumentation is provided in this section. These are outlined beginning with the subjective tools utilized in the form of a case history questionnaire (Appendix D), a self developed Hearing Experience Questionnaire [HEQ]

(Appendix E and Appendix F) and Hearing Handicap Invetory for Adults (HHIA) (Appendix G). This is then followed by the behavioural and electrophysiological audiological assessments. Special reference is made to the modifications required to response modes in the test battery due to physical and speech disabilities.

Case History Questionnaire.

Case history information forms an essential component to acquiring a thorough background into the participants' medical history (Bess & Humes, 2008). A self-developed case history questionnaire (see Appendix D) aided the selection of participants who complied with the stated inclusionary criteria. These were issued with informed consent packs. The content of this 27- item questionnaire is outlined in Table 11.

The questionnaire addressed six categories of information, all of which aided the researcher in determining candidacy and to make the necessary modifications for testing. For example, a participant with preserved motor function in the upper extremities would be able to complete testing using a push button response; while a participant whose upper extremities did not have functional use would have required modification of response modes. Advanced preparation for such modifications promoted efficiency of testing, which was essential in testing a population with a fatigable nature. This questionnaire additionally addressed the participants' desire for result feedback, which is highly relevant when considering the emotional threats an added diagnosis of hearing impairment may have on individual wellbeing. Where possible, close-ended information was presented using tick (√) responses to reduce the demand placed on participant fine motor skills. Categories of an open-ended question style were completed either

by participants or with the aid of caregivers if necessary.

Table 11

Case History Categories

Category	No of questions	Content
Personal details	3	Gender, home language and hand dominance
Auditory History	8	Perceived hearing loss, onset of loss, previous auditory testing, most recent auditory evaluation results (where applicable), use of hearing aids, laterality of fitting (where applicable), family history of hearing loss, surgery to head, neck or ear regions
Medical History	9	Onset of symptom presentation, age at symptom presentation, nature of medical contact at onset, year of diagnosis , diagnostic professional (general practitioner, neurologist), classification of type of MND, checklist of upper and lower extremity functional ability and speech production, medical checklist, review of medications
Communication	4	Preferred method of communication, extent of preserved speech production abilities, type of communication device use, alternative methods of communication (individual specific)
Results	2	Option to obtain test feedback, selection of feedback method
Transport	1	Individual transportation needs

Hearing Experience Questionnaire.

This self-developed questionnaire (Appendix E and Appendix F) comprised two primary categories: i) contact with the discipline of speech pathology and audiology and ii) communication and hearing. The first category served to establish whether individuals with MND were in communication with the relevant discipline as part of the management plan. This aimed to investigate the level of professional referral experienced. The second category to provide insight into individual subjective experiences associated with auditory abilities and their perceptions pertaining to hearing and communicative handicap levels as perceived by the individual with MND. A total of 13 open-ended participant directed questions were developed and reviewed against and alongside a selected three open-ended caregiver directed questions.

These questions directed greater focus towards participant and caregiver perceptions, functional priorities and perceived auditory impact. The three caregiver-directed questions mirrored three questions from the participant directed questionnaire to draw comparisons in the variations of perceptions between the MND participants' and their respective caregivers. The caregiver component of the questionnaire focused primarily on communication and hearing abilities. This served to gain insight into the perceptions an added diagnosis of auditory impairment would have on the social and functional ability of each MND individual. The categories included in this self-developed questionnaire are outlined in detail in Table 12. MND participants were required to return the completed HEQ to the researcher on the day of the test appointment, prior to the commencement of auditory testing. The return of completed forms prior to testing aimed to ensure that participant responses to HEQ items did not alter according to individual perceptions of test success and/ or result feedback in instances where this was requested. The caregiver component of the HEQ was completed by the end of the test session.

Table 12

Hearing Experience Questionnaire Categories

Category	Question Type	No. of Questions	Content
Contact with speech pathology and audiology disciplines	Open and close-ended questions	4	This information served to provide insight into patient contact with an allied health professional concerned primarily with the auditory system. This would provide additional information regarding hearing loss in MND and whether this is not readily detected as a result of limited inclusion of the auditory discipline as part of the multidisciplinary management team or the lack of necessity for this inclusion.
Communication and hearing (participant and primary caregiver)	Mixed open and close-ended questions	MND Participants: 9 questions *Caregivers: 3 questions*	MND Participant Only: Modes of communication, importance of communication versus hearing, identification of auditory symptoms. MND Participant & Caregiver Participants: rating of functional abilities linked to movement, speech and visual abilities, relevance of auditory diagnosis, implications of auditory impairment.

Hearing Handicap Inventory for Adults.

The Hearing Handicap Inventory for Adults (HHIA) (Appendix G) is a 25-item self-assessment checklist comprising two subscales evaluating social (12 items) and emotional (13 items) implications of hearing impairment (Newman, Weinstein, Jacobson, & Hug, 1990). This subjective questionnaire follows a closed-ended question style where participants are presented with the response options: yes, no or sometimes. This scale scores participant responses as follows: yes – 4; sometimes – 2 or no – 0. A total score of 100 can be obtained. Scores are calculated accordingly to reveal a 'significant handicap', 'mild-moderate handicap' or 'no handicap', while simultaneously providing insight into the nature of the handicap – be it on a social or an emotional level (Newman et al., 1990). The HHIA was developed to substantiate hearing difficulties identified in conventional auditory testing, guide decisions regarding hearing aid candidacy, facilitate the counseling process and shape the designing of client-centered rehabilitation programs (Newman et al., 1990).

The HHIA was developed as a modification of the Hearing Handicap Inventory for the Elderly (HHIE) and was designed for the evaluation of hearing impaired adults younger than 65 years of age, including two categories of employment. The HHIA was devised in the United States of America on a sample of 67 middle income, employed, non-hearing aid users. It is currently one of the most widely used instruments in English speaking countries (Monzani, Genovese, Palma, Rovatti, Borgonzoni & Martini, 2007). The HHIA has been further proven to be an appropriate measure for both clinical and experimental purposes in non-English speaking countries when directly translated e.g. Italy (Monzani et al., 2007). This suggests that despite

contextual differences and differences in individual perceptions of handicap across countries, the HHIA remains applicable and consistent as a self-report measure.

The HHIA scale is widely applicable as a means of gathering information pertaining to the various situational and emotional difficulties faced by individuals as a result of hearing loss. The application of the HHIA in the current study is therefore well supported for the purpose of reviewing individual perceptions of their personal levels of emotional and social handicaps for review against auditory test findings. Furthermore, the designated age group for use of the HHIA corresponds with the cut-off age for participant selection in the current study and as a result was deemed appropriate for use in this study.

Newman et al. (1990) evaluated the psychometric adequacy of this scale and its' audiometric correlates. Findings revealed internal consistency reliability of a high standard and a low standard error of measurement, thereby supporting the suitability of this measure for the current study. Furthermore, statistically significant relationships were recorded between the HHIA, pure tone sensitivity and suprathreshold word recognition abilities (Newman et al., 1990). The evidence collected by Newman et al. (1990) support the use of self-report handicap measures, specifically the HHIA, to support audiometric measures. In isolation, audiometric measures are identified as being inadequate in precisely recognizing a patients' reaction to hearing loss, supporting the importance of self-report questionnaires in audiological testing.

The HHIA was presented to the individuals in this study for completion prior to auditory testing and auditory diagnosis, thereby serving to ensure auditory test findings did not affect the

manner in which participants responded to the presented questions. Participants were requested to return the completed HHIA scale on the day of testing, prior to the initiation of testing. Participants were furthermore instructed verbally (via telephonic contact) and/or in written text (via email) that the HHIA served to explore emotional and situational difficulties that they personally *may* have experienced in daily living, despite the instructional and question format suggesting a definitive diagnosis of auditory impairment. An adaptation of the HHIA material would have been deemed more appropriate to limit any possible participant confusion pertaining to the completion of this form and is necessary to consider for the purpose of study replication.

Audiological test battery.

The test battery incorporated both behavioural and electrophysiological test measures to assess different levels of auditory function (Figure 3). The audiological procedures, equipment and modifications to the response modes are presented in Table 13.

Sequence of Testing

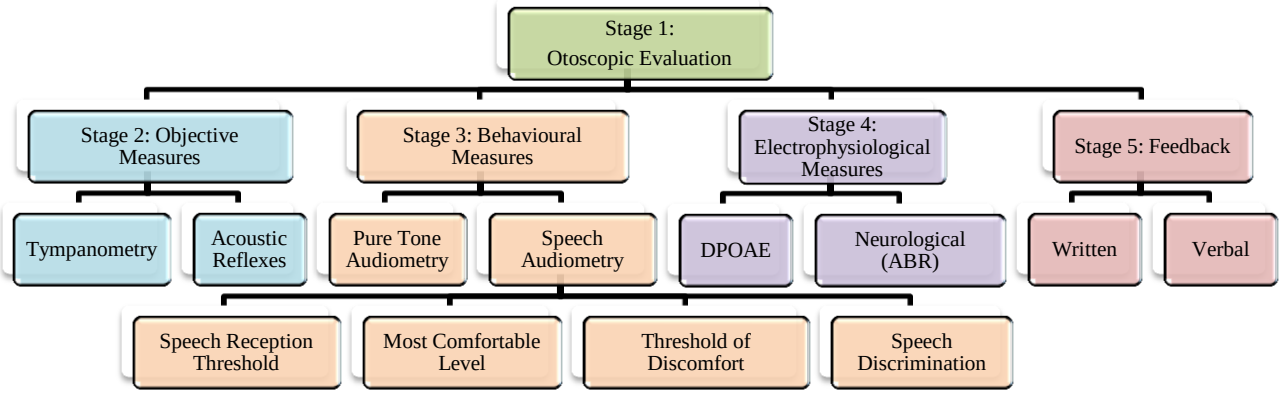

Figure 3. Sequence of Testing.

Pure tone and speech audiometry are reliant on motor strength for physical responses and speech production. Whereas pure tone responses are typically reliant on the physical action of a push-button response, speech audiometry testing relies on an individuals' ability to repeat verbally presented words. This repetition provides the audiologist with valuable information pertaining to speech reception and speech sound discrimination abilities. In turn, these results provide important diagnostic information validating the site of lesion responsible for auditory impairment and give an indication of the individuals' speech perception difficulties threatening communicative success.

Individuals with MND present with a gradual loss of motor function. The extent and site of this deterioration in muscular function varies based on the type and stage of the disease and therefore was not uniform across all participants in the current study. The speech system relies on muscular strength for movement of the respiratory muscles, laryngeal muscles and articulators. Weakness in one or all of these muscular systems threaten speech production abilities and consequently suggested the need for modifications in the various test procedures to be made to accommodate participant needs and abilities (Duffy, 2005). Response mode modifications are presented in Table 13. These modifications were applied only when necessary. The researcher was alerted to the need for possible response mode modifications upon receipt of case history questionnaires. The selection of an appropriate response mode was then confirmed and validated on the test day. Materials for modifications were easily accessible and available on the test day of each individual to ensure that efficiency of testing was not threatened by unaccounted for alterations in testing procedure.

Table 13

Equipment and Measures

Measure	Equipment	Rationale	Response Mode Modification
Otoscopic examination	• Otoscope • Speculae • Ultracide disinfectant	An otoscopic examination is performed to evaluate the state of the outer ear, and tympanic membrane for abnormalities and/or infection (Rappaport & Provencal, 2002).	None required
Immitance audiometry *(objective measure)* i) Tympanometry ii) Acoustic reflexes (Ipsi- and contralateral)	• GSI 33 Tympanometer • Tympanometry probe tips • Ultracide Disinfectant	Tympanometry evaluates middle ear status through the depiction of tympanic membrane motility as a function of variations in air pressure (Clark, Roeser & Mendrygal,2007). This test serves as an irreplaceable evaluative tool representing middle ear functioning (Clark et al., 2007). Acoustic reflex testing is a measure of stapedius muscle contraction as a response to intense sound stimulation. This is performed through ipsilateral and contralateral neural pathways, thereby facilitating in the differential diagnosis of middle ear, cochlear, retrocochlear pathologies as well as brainstem lesions (Block & Wiley, 1994).	None required
Pure tone audiometry *(behavioural measures)* i) Air conduction testing ii) Bone conduction testing	• Diagnostic Audiometer AC40 • Sound Proof Test Booth (double wall) • Headphones • Bone conductor • Response Button • Audiogram	Pure tone audiometry assesses hearing sensitivity as a function of frequency (Bess & Humes, 2008). This provides insight into the integrity of the auditory system as well as information relating to the symmetry, laterality, degree and configuration of a patients hearing thresholds (Harrell, 2002). Bone conduction audiometry allows for specified information pertaining to the type of hearing loss (Dirks, 1994). This information facilitates the making of a differential diagnosis through distinguishing between an outer, middle or inner ear pathology and assists in the classification of the type of hearing loss (Dirks, 1994).	• verbal response (yes/vocalization) • eye blinking response • visual gaze response • head nod

Measure	Equipment	Rationale	Response Mode Modification
Speech audiometry *(behavioural measures)* i) Speech reception thresholds (SRT) ii) Most comfortable listening level (MCL) iii) Threshold of discomfort (TD) iv) Speech discrimination (Sd)	• Diagnostic Audiometer AC40 • Sound Proof Test Booth • CID-W1 Wordlist (for SRT) • NAL-AB Wordlist (for Speech Discrimination)	The most important measurable component of human auditory function must be related back to the patients' ability to understand speech (Bess & Humes, 2008). This ability forms the foundation to participating in the majority of activities of daily living. i) SRT evaluates a patients' ability to detect and understand speech sounds at the lowest level possible (Bess & Humes, 2008). This test confirms the reliability of pure-tone audiometry results (Brandy, 2001). ii) MCL determines a level of speech listening, which affords the patient a comfortable listening experience, with an effortless ability to understand speech sounds (Brandy, 2001). iii) TD forms part of the calculation of the patients' dynamic range (Brandy, 2002). Dynamic range proves useful in the differential diagnosis of cochlear versus retro-cochlear pathology (Gelfand, 2009). iv) Evaluates a patients' ability to distinguish between sounds and recognize speech (Brandy, 2001). Speech discrimination scores guide the process of differential diagnosis associated with the possible site of pathology i.e. outer/middle ear; cochlear or retro-cochlear.	• visual gaze to picture cards • head laser to picture cards • alphabet board (Appendix I) (Appendix J) (Appendix K)
Distortion product otoacoustic emissions (OAEs) *(objective measure)*	• Biologic Diagnostic OAE • Disinfected nubs • Sound Proof Test Booth	This is a recording of sounds generated within the cochlea (Prieve & Fitzgerald, 2002). DPOAEs allow for information pertaining to the integrity of the cochlea to be obtained, therefore contributing to the differential diagnosis linked to auditory functioning at this level of the auditory system (Prieve & Fitzgerald, 2002). DPOAEs will be performed as these provide obtainable results for a wider range of hearing impairment (Prieve & Fitzgerald, 2002) and offer greater frequency specificity.	None required
Neuodiagnostic auditory brainstem responses (ABR) *(objective measure)*	• Electrodes • Eclipse ABR System • Plinth • Electrode gel	This is a highly specific, specialized measure capable of yielding unique diagnostic information in the differentiation of cochlear versus 8th cranial nerve pathology (Musiek, Borenstain, Hall, & Schwaber, 1994). Interpretation of ABR waveforms offer insight into the latency responses at a higher level along the auditory pathway, than that which behavioural test measures are capable of (Musiek et al., 1994). ABRs have detected waveform changes in progressive degenerative diseases such as multiple sclerosis (Musiek et al., 1994). The application of this test may therefore be useful in indicating the possible presence of auditory tract involvement in MND.	None required

Data Collection

Ethical and procedural considerations had to be taken into account with data collection to ensure reliability.

Ethical Considerations.

Data for this research study was obtained from human participants. The ethical and legal responsibility of the researcher to protect the rights of all participants was therefore critical (Schiavetti & Metz, 2002). A number of ethical considerations were taken into account. These were shaped around the theoretical foundations of the World Medical Association Declaration of Helsinki: Ethical Principles for Medical Research Involving Human Subjects (World Medical Association [WMA], 2000). These principles were applied to this study as a means of promoting its' execution in a manner that was honest and truthful to participants and valid in execution. The manner in which these principles were reviewed and applied to this study is delineated below.

Ethics Review Committee.

To assure protection of participants, a proposal, detailing the execution of the study aims, methodology, participant selection methods and instrumentation was submitted to the Medical Research Ethics Committee of the University of the Witwatersrand for ethical approval. This proposal additionally indicated the manner in which the Helsinki Declaration ethical principles were to be addressed (WMA, 2000). Ethical approval was granted in June 2010 with approval for title revisions made in October 2011. A clearance number M10569 was assigned to this study (Appendix L).

Participant information sheet for informed consent.

All participants were presented with a participant information sheet prior to the onset of any audiological testing (Sade, 2003). Informed consent encompassed more than an information sheet, but forms the fundamental basis of a respectful and trusted relationship between the participant and the researcher (O'Neill, 2002). Providing participants with information sheets afforded them the opportunity to accept or decline the invitation to voluntarily participate (Babbie & Mouton, 2001). Table 14 provides a detailed review of the ethical principles applied to the current study. Participants were requested to sign in acknowledgement of informed consent. In instances whereby upper extremity movement prohibited written consent, a thumb imprint representing participant consent was deemed permissible.

Table 14

Components of Informed Consent

Component	Rationale
Nature of the Study	Research participants were briefed on the nature, purpose and implications of this study (Sade, 2003). It was made clear prior to the signing of consent that this study was designed in an exploratory format which offered information about their auditory abilities, however did not offer personal rehabilitative benefits. Understanding the contribution this study may have on knowledge linked to MND and its implications on future MND auditory function studies afforded participants an opportunity to understand the relevance of their participation.
Autonomy and Confidentiality	Autonomy is highly reliant on individual perceptions and priorities (O'Neill, 2002). The autonomy of each participant in this study was unconditionally respected (Schiavetti & Metz, 2002). Confidentiality of all personal and medical details and information acquired through the data collection phase was guaranteed through ensuring that only the research team and supervisor had access to the data. Each participant received a participant identification number, which was used on all assessment related study material. Destruction of any data containing personal identifying information took place at the completion of the study. Data was only used for the specified study, and was not distributed for any other purposes. For the purpose of this study, anonymity and confidentiality of all participants was maintained by ensuring that a research coding system was utilized rather than participant name.
Withdrawal	Informing participants of their individual right to withdraw at any point without any negative consequences was clearly expressed. This knowledge was essential for maintaining participant autonomy and empowering participants with decision-making opportunity should they at any point have chosen to be voluntarily excluded from the study (Sade, 2003).
Compensation	A reimbursement model of participant compensation, where participants were compensated for actual expenses was applied (Grady, 2005). Research participants were provided with R100-00 financial compensation for travelling expenses to and from the USHC. Grady (2005) suggests that the reimbursement of participants for expenses incurred assists in making the process of participation a revenue-neutral activity. Reimbursement affords participants who otherwise may be unable to make the financial sacrifice, able to participate (Grady 2005), which may further assist in obtaining the desired sample size.
Non-Maleficence	Non-maleficence highlights the importance of bringing no harm to participants in human studies (Smith, 2005).The principles of beneficence and maleficence in medical ethics usually presents as a double effect whereby a single action is capable of bringing both good and harm to individuals (Smith, 2005). Due to the vulnerability of this population consideration of non-maleficence was of upmost importance. This study served to execute auditory testing in a manner that would yield reliable outcomes while simultaneously accounting for participant factors such as ability to engage in behavioural test procedures, emotional consequences of test outcomes and length of test concerns. Consequently, preparation for test modifications were made to accommodate individual motor and speech abilities and individuals were presented with the option to decline auditory test feedback, Lastly, testing procedures were carefully paced according to close caregiver and tester monitoring of individual fatigue and willingness to continue testing. The option of dividing test sessions over two consecutive days was presented to individuals, however this was not requested by participating individuals nor was this deemed necessary based on individual test performance.

Risks	Participants were informed that they would not be exposed to any harmful risks by agreeing to participate in this study. Testing procedures were non-invasive and test stimuli brought no known harm. Rest periods were offered to participants in the event that they experienced fatigue. In the event of impaired hearing ability being recorded, participants were placed at risk on an emotional level in terms of dealing with an additional impairment of ability. In line with this, participants were able to request that they did not receive feedback from their testing.
Inclusion	Research participants were informed that strict criteria were adhered to. Participants were informed that signed consent would not guarantee inclusion in the study. Criteria were not made known to participants as this posed the threat of participant dishonesty as a means of manipulating inclusion in the study.
Findings	Where results from a study were significantly meaningful to participants, investigators made participants aware that this information may be accessed and consequently, invited a request for those individual results (Shalowitz & Miller, 2005). In disclosing results investigators demonstrate respect for participants' autonomy and empower them with the knowledge to incorporate research results into their personal decision-making. This process acknowledges the participants presumptive entitlement to information about themselves and show gratitude for participants' voluntary participation in research (Shalowitz & Miller, 2005). The case history form provided participants with an option to select whether they would or would not like feedback of their results. Participants who chose to obtain test feedback were provided with verbal feedback at the completion of each assessment session. Participants were presented with recommendations and referring information where necessary, however it was clearly maintained that they were at no point obligated to follow-through with these recommendations, as autonomy continued to remain central to this study. Recommendations remained specific to individual participant findings. These included referrals for curative treatment in instances of active middle ear pathology, or longer- term intervention in the form communication training (Erber & Scherer, 1999). The latter may further assist in guiding communication specialists facilitating participants' use of AAC devices to consider exploring communication strategies paired with hearing impairment (Erber & Scherer, 1999).

Data Collection Procedures

The procedures for data collection during all three phases of the research are described below:

- Application to the Medical Committee for Research on Human Subjects. Prior to the commencement of any study, judgments need to be made by respected institutional ethics committees for approval (Babbi & Mouton, 2001). This is essential as opinions regarding ethicality differ (Babbi & Mouton, 2001). Ethical clearance was granted by the University of the Witwatersrand Committee for Research on Human Subjects (Medical) (Appendix L).
- Permission from MNDSA. An information sheet was issued to the authoritative figures of the MNDSA detailing the specifics and implications of the study (See Appendix B). This information sheet included a request for approval to locate potential participants through the MNDSA, which was approved shortly thereafter (Appendix C).
- A pilot study was conducted to finalize the measuring instruments, determine the equipment to be used in the study; determine the need for adaptation of test equipment; and determine the total testing time per participant.

During the main study phase the following procedures were followed:

- Participants were recruited in consultation with the MNDSA, private neurologists in Gauteng and the Chris Hani Baragwanath Hospital
- Participants were provided with informed consent and alerted to the objectives of this study as well as their ethical rights.

- Upon the acquisition of participatory consent, appointments were scheduled for testing at the University of the Witwatersrand Audiology Clinic
- Individuals were provided with the HHIA and HEQ for completion prior to the test appointment. Individuals were presented with these forms either via email or postal delivery. A verbal (telephonic) and/or written (email) outline of these forms was provided to individuals to support written instructions.
- Biological calibrations of the test audiometer took place prior to participant arrival on the day of testing to ensure consistency in test recordings were upheld for the duration of the data collection procedure.
- All audiometric measures were performed on the scheduled appointment day. Behavioural measures were conducted early in the test session followed by electrophysiological measures. This sequence of testing ensured that procedures reliant on active responses were executed early during the assessment, while electrophysiological measures, which were not reliant on active responses were conducted during the final portion of the appointments, ensuring participant fatigue did not influence the reliability of test findings (Figure 2).
- All participant questionnaires (HEQ and HHIA) were returned to the researcher on the scheduled appointment day.
- Caregivers were presented with and completed the caregiver portion of the HEQ during auditory testing of the MND participant. This occurred outside the testing booth.
- Verbal feedback was presented at the completion of the assessment session where requested by the individual. This was only conducted with participants who

indicated the desire for feedback in the initial case history questionnaire. Written audiological reports summarizing the findings from the auditory evaluation were compiled and issued with a two-week turnaround time from date of assessment. This was only performed for those participants who requested written feedback in the initial case history questionnaire.

- Participants were presented with R100-00 financial compensation in a sealed envelope at the completion of the assessment session.
- Participants were presented with thank-you letters at the completion of the assessment session.
- All the data was encoded and captured after which it was checked for any capturing errors.
- Accuracy of data capturing and thematic content analysis was then validated by the inter-rater.

Reliability and Validity

The underlying threats and methods to achieving reliability and validity in the current study are outlined below.

Reliability.

Reliability refers to the consistency and accuracy of a given measurement and determines whether or not a replica of the findings of the current study would be obtained if the same methodology were to be repeated (Schiavetti & Metz, 2002). Essentially, where reliability of findings are dependent on the recurrence of the original data and the manner in which this data

was interpreted, the various researcher-based restrictions placed on the data with regards to consistency and rigor of employment also required consideration (Lewis, 2003). The following components in the execution of this study were applied to address reliability threats i) once off assessments; ii) independent raters; iii) subjective assessments; iv) systematic errors.

Once-off Assessment.

Assessments for each participant took place in a single, once-off assessment session. The researcher considered the effects of a lengthy assessment paired with the high potential for participant fatigue during this lengthy test session. The researcher prepared for testing to be executed on two separate consecutive dates if necessary as a means of accommodating this obstacle. Despite this, all participants tested were willing and capable of completing the entire evaluation in a single assessment session. Six of the participants voluntarily remained at the test session following testing to discuss and share their experiences with MND. As a result of this positive response, multi-session assessments were not required for this study.

Independent Raters.

Inter-rater reliability refers to the extent to which two members of a research team evaluate the same results and provide identical judgments (Leedy & Ormrod, 2001). Two raters, the researcher and an independent rater were involved in the data analysis procedure to enhance reliability of the current study. Both raters were qualified with an Honours degree in Speech-Language Pathology and Audiology from the University of the Witwatersrand. Both raters had a total of 6 years clinical experience in the field of diagnostic audiology, with experience in the execution and interpretation of all behavioural and electrophysiological measures conducted in

this study. Audiometric results and HHIA scores were interpreted by the researcher and re-interpreted through the use of an independent rater. The completed audiograms, DPOAE recordings and ABR recordings were interpreted and findings were then verified. Agreement between the result interpretation of the researcher and the interrater was representative of a positive criterion. Of the two raters, only the researcher was present and involved in the data acquisition process. The researcher chose to provide the second rater with minimal information regarding the aims and content of the research study in an effort to limit the bias in data interpretation.

The normative data for the formulated test protocol was issued to this rater to ensure the normative data used for result interpretation remained uniform across all raters. A percentage of 25% of the interpreted data was re-analyzed by the other member of the research team (Silverman, 1993). The following formula was applied to establish a desirable level of inter-rater reliability (Alberto & Troutman, 2006):

$$\text{Percentage Agreement} = \frac{\text{Agreements}}{\text{Agreements} + \text{Disagreements}} \times 100$$

Inter-rater reliability percentages were calculated based on each audiometric test per individual. This was also applied to the HHIA scores to ensure reliability of scoring the categories of emotional, social and total impact of auditory abilities, as well as the coding of participant and caregiver statements extracted from the HEQ. Inter-rater percentage agreements are presented in Figure 4.

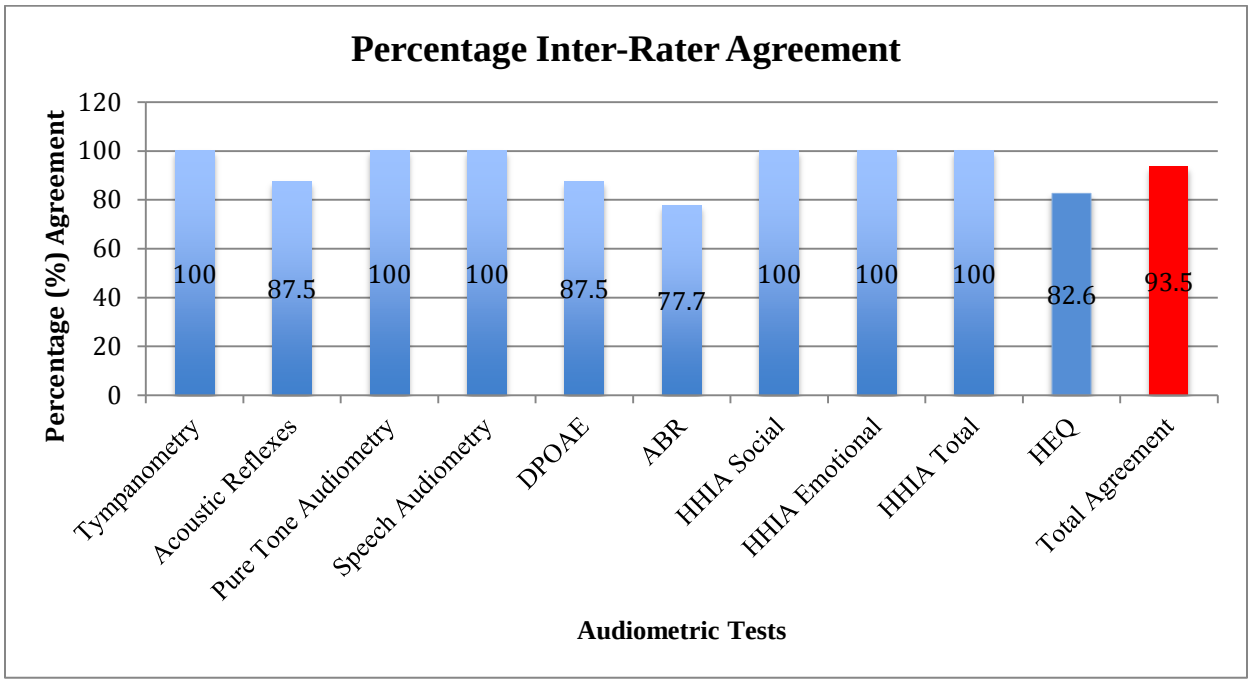

Figure 4. Percentage Inter-Rater Agreement.

Inter-rater reliability obtained for accuracy scores yielded a total percentage of 93.5%. Based on Silverman's (1993) classification system, this score was considered to be very good. Where raters disagreed, results were reviewed based on a theoretical and clinical debate between raters. This included a review of the documented interpretation guidelines and presentation of anonymous test results to a third party with audiological clinical experience for confirmation of result interpretation. The raters were able to reach mutual agreement following re-analysis of acoustic reflex and distortion production otoacoustic emission results, while a third rater was included to settle debate surrounding the identification of waveforms I, III and V for a single participant's results.

Subjective assessment.

Subjective assessment procedures further threatened the reliability of the current study.

Non-interactional observer bias suggests misrepresentation in the documentation and/or interpretation of test results (Silverman, 2000). A detailed test protocol with equipment, procedural and instructive specifications was formulated and closely abided to during participant testing.

Systematic Errors.

Evidence of measurement errors consistently reoccurring with each repeated measurement are known as systematic errors (Schiavetti & Metz, 2002). Since reliability essentially refers to the trustworthiness of research findings it was essential that instrumentation allowed for accuracy in results (Babbie & Mouton, 1998). Systematic errors threatening reliability of the current study were removed by ensuring that all test equipment was calibrated prior to initiating the data collection process.

Validity.

Validity typically refers to the 'precision' of a research finding and refers to the extent to which successful evaluation and measurement of the central theme is achieved (Lewis, 2003). The validity of the current study has been reviewed under the categories of i) internal; ii) external and iii) face validity.

Internal validity.

Internal validity refers to whether or not the researcher investigates that which he/she claims to investigate (Silverman, 2000). This is strengthened largely by reducing extraneous variables, narrowing participant characteristics and following strict measurement protocols

(Schiavetti & Metz, 2002). The internal validity of this study was therefore addressed through i) equipment calibration; ii) internationally applied test protocols and iii) elimination of known bias.

Equipment calibration comprised periodic electronic, biological and periodic calibration checks. Periodic electronic calibration involves an electronic calibration of all equipment ensuring that the minimum standards defined by SABS are met. Biological calibrations involve establishing a baseline measure on normal hearing individuals. Two normal hearing individuals were tested and baselines measures for both individuals were obtained. Prior to the assessment session, one of the two above-mentioned individuals was retested and comparisons in hearing thresholds were drawn (Roeser, Valente & Hosford-Dunn, 2000). A shift greater than 5dB at one or more frequencies from 500Hz – 6000Hz are typically indicative of the need for electronic calibration of equipment (Roeser et al., 2000). Test-retest sessions prior to the testing of all eight participants did not reveal shifts from the baseline measures. Daily listening check ensures the quality of the auditory signal is free from distortion and interference and ensures the sound signal is being delivered to the correct earphone. Elimination of malfunctioning equipment promotes the validity of test findings and removes the risk of inappropriate diagnosis and recommended management (Roeser et al., 2000). A daily listening check was conducted prior to the arrival of each participant on the day of testing.

Lewis (2003) indicates that internal validity is controlled through the elimination of known bias from the sample frame. The sample frame used for this study appeared to be largely free from known bias in that criterion sampling techniques led to the participants of this study being different ages, at different stages of MND progression and differing in the classification of

type on MND onset.

External validity.

External validity determines whether generated abstract postulates are applicable to the broader context (Lewis, 2003, Silverman, 2000). This refers to the generalizability of a study determining transferability (Schiavetti & Metz, 2002). Participants were largely selected from the same source i.e. MNDSA despite attempts to expand the range of sample recruitment. The narrow source of participant recruitment does pose a threat to external validity, however based on the limited population size of this target group of adults in South Africa and more so, in Gauteng, this could not be controlled. External validity is largely achieved through replication, thereby suggesting that if results from multiple cases are consistent findings may be considered more robust (Silverman, 2000). It is noted that MND-based studies on a worldwide scale have reported great consistency amongst patients with this disease in terms of incidence, prevalence and symptomatic presentation and progression (Swash & Desai, 2000; Wijesekera & Leigh, 2007). On account of the consistency of these reports on a global scale as well as the limited sample utilized in this study, the researcher made a cautious presupposition that result generalization may be validated on account of these factors. Furthermore, result interpretation was addressed in a descriptive manner, allowing for differences and similarities in test participants to be highlighted in a qualitative manner, where relevant. A multiple case study design does however pose potential biases. One such form of bias, holistic fallacy refers to a tendency to interpret data from individual cases as more similar than they really are (Kohn, 1997). This may be guarded against by engaging in team data analysis an approach that encourages multiple interpretations and the use of multiple ‘raters’ (Kohn, 1997). For the

purpose of the current study, acquired data was interpreted by the primary researcher as well as an independent research assistant in order to identify trends and patterns.

Face validity.

This is an estimation of whether a test is effective in measuring the desired criterion (Keller & Warrack, 2001). Lewis (2003) highlights the importance of examining the quality of questions that are presented in patient questionnaires to explore the perspective of research participants. To address this, the researcher paired with an audiologist with six years clinical experience in the audiological field divergently mapped out the information deemed as essential to fulfilling the target research aims. The pilot study then assisted in determining whether the content investigated in the Case History Questionnaire and HEQ successfully achieved that which it aimed to investigate. An audiologist external to the study thus reviewed these questionnaires and modifications were made accordingly to ensure face validity was achieved.

Trustworthiness.

The efforts implemented by the researcher to overcome the risks of subjective bias as a result of single researcher result analysis and interpretation relate to the trustworthiness of a study (Shenton, 2004). Credibility is highlighted as one the most important factors in establishing trustworthiness and serves to promote confidence in the accuracy of data and interpretation of the scrutinized area of research (Shenton, 2004). Triangulation, the method of combining a minimum of two theoretical perspectives, methodological approaches and/or data analysis methods in a single study, was selected to enhance the trustworthiness of this study (Thurmond, 2001). More specifically, analytical triangulation was applied serving to strengthen

confidence in the interpretation of results thereby increasing the validity of findings (Shenton, 2004). This was achieved throughout the process of thematic content analysis. The researcher translated participant and caregiver responses into a list of codes, which allowed for the identification of broader themes common across individual responses. A more detailed analysis then allowed for more specific and defined themes to emerge (King, 2004). The entire process of translating, coding and isolating broad and specific themes amongst individual responses was then re-executed by an independent researcher. This opportunity for reanalysis was based on the concept of circling reality, which relates to the necessity of acquiring more than one perspective (Shenton, 2004). This serves as a means of acquiring a more consistent and stable view of individual experience and perception of reality (Shenton, 2004). The participation of an independent researcher who re-translated and coded individual responses then allowed for a comparison of themes and patterns identified by both analysts, while an independent third party rater proceeded to further validate the emergent themes identified through the data coding process.

Data analysis and statistical procedures

This research study employed a paradigm of analytic triangulation to the data analysis. This involved interpreting certain aspects of the acquired data, particularly HEQ responses, utilizing a mixed method of both quantitative and qualitative analysis methods (Thurmond, 2001). Quantitative review of the data was conducted through statistical analysis using Statistica software (StatSoft, 2005). Mean and standard deviations and Pearson's product moment correlation (Table 15) comprised the descriptive and exploratory statistical tests utilized in the data analysis procedure respectively. The data was analyzed using **parametric statistics**

(Schiavetti & Metz, 2002) whereby inferences can be made regarding the parameters of data distribution i.e. correlation coefficient for the population correlation (Keller & Warrack, 2000). This form of analysis was further employed as it presents with greater statistical power (Keller & Warrack, 2000). Both mean and standard deviation analysis as well as Pearson's product moment correlation were deemed appropriate analytical techniques for the description and exploration of this single MND population.

Descriptive and inferential statistics were also applied to the data analysis procedure. Descriptive analysis allowed for the arrangement of data, numerically or otherwise and allows for meaningful extraction of the essential components for interpretation (Keller & Warrack, 2000). This was used to describe the audiological presentation based on the type, degree and configuration of the hearing loss for individual case studies. Inferential statistics, frequently used in hypothesis testing, was applied to yield conclusions extending beyond the immediate data and served to strengthen the internal consistency of this study (Trochim, 2000). Inferential statistics were applied through correlation analysis of the various tests executed in the study. A 0.05 significance level was used for all statistical tests, unless specified otherwise.

Table 15

Statistical Procedures.

Statistical procedures	Rationale
Mean scores, median and standard deviations were calculated where applicable to provide information on the spread of distribution	Information was obtained on the average of all scores as well as the average variability of scores (Maxwell & Satake, 2006).
Pearson's product moment correlation	This is a measure of the correlation between two variables and analyzes the linear dependence between two variables.

Qualitative methods were applied to the analysis and interpretation of subjective participant response in the third category of the HEQ. The analytical approach largely followed the conventions of thematic content analysis. Participant and caregiver responses were listed by the researcher and translated these into a list of codes representing themes identified in the textual data (King, 2004). Coding was executed in hierarchical order, allowing for data analysis at different levels of specificity (King, 2004). Broad themes based on the research objectives and questionnaire questions were identified from these codes to create an initial template. These themes were then subjected to a more detailed analysis leading to the formation of more specific, tightly defined categories within each theme (King, 2004). A comparison between caregiver and MND individual themes were explored (King, 2004). This procedure was replicated by an independent researcher. Following this, a discussion and modification of specific themes identified across both sets of analysis where necessary. An independent third party rater, corroborated the themes that emerged from the data coding process.

Conclusion

This chapter provided a detailed description of the methodology applied to this research study. Primary and secondary objectives were discussed followed by a description of the research design and phases. A description of the pilot study was provided with identification of methodological flaws and appropriate recommendations to rectify these obstacles. Participant selection criteria and participant description were additionally described in this chapter, followed by a description of the equipment and measuring instrumentation. This chapter reached its conclusion with a review of the data collection procedures and the data analysis applied in the current study.

Chapter Four

Results

Introduction

The results of the study will be presented in this chapter in accordance with the primary objective of the study, namely to determine the prevalence of hearing loss in individuals diagnosed with MND. Firstly, the audiometric test findings will be presented and then followed by the results of the HEQ and HHIA. The latter will serve to address the secondary objectives, namely the rating of functional abilities, perceived socio-emotional impact of hearing loss as value of auditory diagnosis as reported by MND individuals and caregivers. A 0.05 significance level was used for all statistical tests, unless specified otherwise.

Audiometric findings

The first secondary objective addressed the description of the audiometric findings of individuals with MND. This includes a description of participant-reported auditory symptoms, as well as findings from the otoscopic examination and acoustic immitance measures. The description of audiometric findings continues with a review of pure tone and speech audiometry results. This is then followed by a description of electrophysiological test measures in the form of DPOAEs and neurodiagnostic ABR findings.

Audiological symptoms.

Audiological symptoms described by participants included tinnitus, difficulty listening in noise and hyperacusis. Figure 5 represents the distribution of auditory symptoms as reported by participants in the initial case history interview and as part of the HEQ.

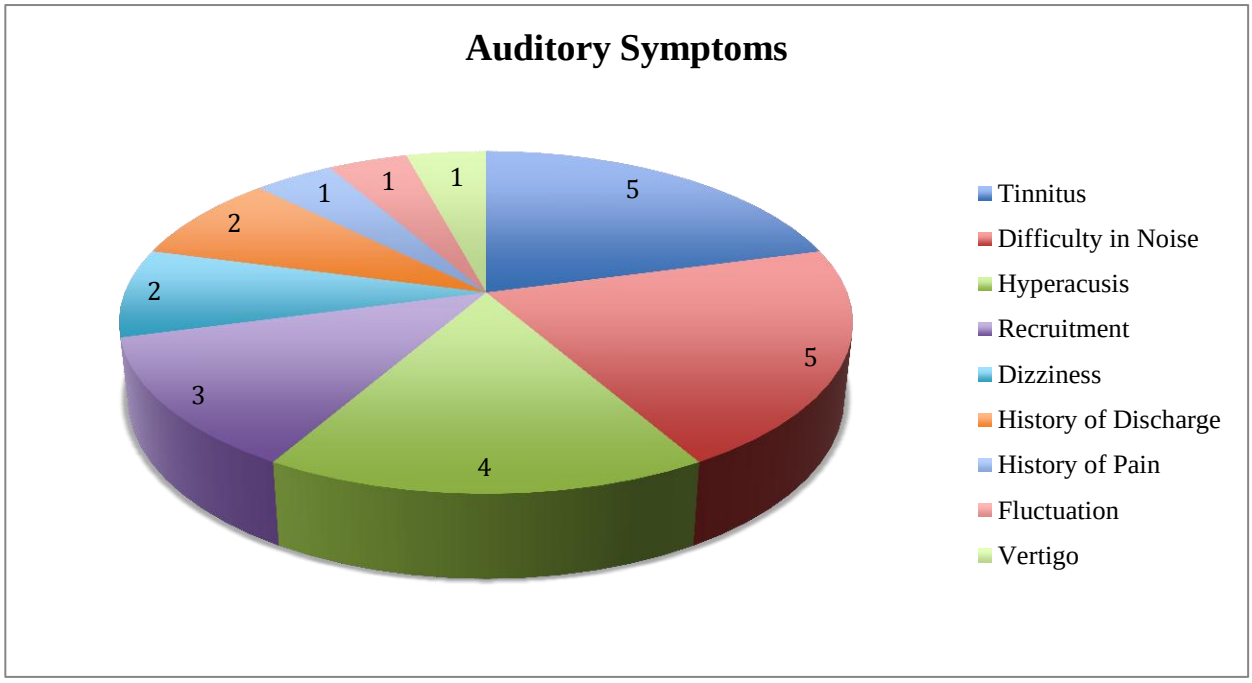

Figure 5. Participant Reported Auditory Symptoms.

A total of five participants reported tinnitus and difficulty listening in noisy environments, followed by four participants whose complaints included hyperacusis. A further three complained of recruitment. All the participants reported optimal listening abilities in quieter environments. Two participants dizziness and a history of ear discharge, while one indicated isolated experiences of hearing fluctuation, vertigo and a history of ear pain.

While the majority of the auditory symptoms reported by participants were typical and representative of the nature of their hearing impairment, a re-occurring report related to hyperacusis presented itself in four of the participants. These participants expressed particular concern with common sounds such as that of passing vehicles and barking of the neighbourhood dog to be a distressing and uncomfortable listening experience. Participants were able to isolate the onset of hyperacusis to have followed the diagnosis of MND, although exact time frames were not reliably acquired. While this percentage is not a reflection of the majority of participants, it is important that this is highlighted based on its unanticipated presentation. While evidence of hyperacusis remains anecdotal, this data invites itself to further exploration and research.

Otoscopic findings.

Otoscopic evaluation revealed clear ear canals, intact tympanic membranes and a visible cone of light in 15 of the examined ears. Only one participant (P3) presented with soft wax partially occluding the right ear canal.

Acoustic Immitance.

Tympanometry.

All participants (*N* =8) presented with a Type A tympanogram bilaterally suggestive of pressure (+50daPa to – 150daPa), static compliance (0. 28 – 1. 8 cm^3) and ear canal volume (0. 2 – 2. 0 cm^3) within the normal range (Hall & Mueller, 1997). Table 16 provides an outline of results obtained through tympanometric measures along with the average, range and *SD* for pressure, compliance and ear canal volume for both the left and the right ears.

Table 16

Summary of Raw Tympanometry Data with Average, Range and SD

Participants	Pressure (in daPa)		Compliance (in cm^3)		Volume (in cm^3)	
	Left ear	**Right ear**	**Left ear**	**Right ear**	**Left ear**	**Right ear**
P1	-30	-18	0. 28	0. 82	1. 2	1. 16
P2	-22	-40	0. 44	0. 31	0. 9	0. 92
P3	-32	-32	0. 89	0. 82	0. 98	0. 98
P4	-29	-64	0. 52	0. 45	0. 88	0. 75
P5	-27	-27	0. 9	0. 71	1. 14	1. 16
P6	-35	-40	0,86	0. 28	0. 88	1. 2
P7	-24	-42	0. 51	0. 37	1. 23	1. 04
P8	-32	-25	0. 48	0. 88	1. 04	1. 2
Average	-28. 9	-36. 0	0. 61	0. 58	1. 03	1. 05
Range	-28. 9 – 22	-64 -18	0. 28 – 0. 90	0. 28 – 0. 88	0. 88-1. 23	0. 75 – 1. 20
SD	4. 4	14. 1	0. 24	0. 25	0. 14	0. 16

Acoustic Reflexes.

Ipsilateral Reflexes.

Ipsilateral acoustic reflex results are outlined in Table 17. All ears (*N* = 16) presented with acoustic reflexes within the 70-90dBSPL range across the 500Hz-2000Hz range of ipsilateral reflexes. Absent reflexes were identified at 4000Hz ipsilaterally in six ears (37. 5%) specifically for P1, P6 and P7 bilaterally. P2 *(n = 1)* presented with normal ipsilateral reflexes for both right and left ears. This participant also presented with bilateral normal hearing thresholds.

Table 17

Summary of Raw Ipsilateral Reflex Data with Average, Range and SD (N=16 ears)

Participant	Ipsilateral reflexes (in dB)							
	Right ear				Left ear			
	500Hz	**1000Hz**	**2000Hz**	**4000Hz**	**500Hz**	**1000Hz**	**2000Hz**	**4000Hz**
P1	85	90	90	Absent	95	100	85	Absent
P2	95	90	95	100	90	95	90	100
P3	105	105	105	110	90	90	85	85
P4	105	95	105	105	95	95	100	100
P5	95	100	100	110	95	90	90	110
P6	105	100	100	Absent	100	105	100	Absent
P7	100	105	105	Absent	100	100	105	Absent
P8	100	95	100	105	100	105	100	105
Average	98. 7	97. 5	100. 0	106	95. 6	97. 5	94. 4	100
Range	85 – 105	90 – 105	90 – 105	100 - 110	90 – 100	90 – 105	85 – 105	85 – 110
SD	6. 9	6. 0	5. 3	4. 2	4. 2	6. 0	7. 8	9. 4

Contralateral Reflexes.

All ears (N = 16) presented with acoustic reflexes within the 70-90dbSPL range across the 500Hz-2000Hz range of contralateral reflexes. Table 18 provides contralateral reflex levels for eight participants and sixteen ears. Absent reflexes were recorded at 2000Hz in two ears specifically the right ear of P6 and the left ear of P7. Absent reflexes were noted at 4000Hz in ten ears bilaterally for P1, P5, P6, P7 and P8. P2 presented with normal contralateral reflexes for both right and left ears. This participant also presented with bilateral normal hearing thresholds.

Table 18

Summary of Raw Contralateral Reflex Data with Average, Range and SD (N=16 ears)

Participants	Contralateral reflexes (in dB)							
	Right ear				Left ear			
	500Hz	1000Hz	2000Hz	4000Hz	500Hz	1000Hz	2000Hz	4000Hz
P1	90	95	90	Absent	105	110	110	Absent
P2	95	90	90	95	90	95	95	105
P3	105	105	95	95	100	100	105	105
P4	105	105	105	100	95	100	100	100
P5	95	100	110	Absent	105	110	110	Absent
P6	105	100	Absent	Absent	105	105	110	Absent
P7	105	110	105	Absent	105	110	Absent	Absent
P8	95	95	110	Absent	100	105	110	Absent
Average	99	100	100. 7	96. 6	100. 6	104. 3	105. 7	103. 3
Range	90 – 105	90 – 110	90 – 110	95 - 105	100-105	105-110	105 – 110	100 – 105
SD	6. 2	6. 5	8. 9	2. 9	5. 6	5. 6	6. 0	2. 9

Pure Tone Audiometry.

The pure tone audiometry results for 16 ears will be presented in this section. The presented information will initially focus on findings of ear symmetry and laterality. A review of pure tone audiometry results for both air and bone conduction measures will follow. These findings will be reviewed in relation to the configuration, severity and type of hearing loss identified.

Laterality & Symmetry.

Bilateral presentation of hearing was recorded for all (N = 8) tested participants. Symmetrical hearing patterns were identified amongst six participants. Two participants (P3 and P6) presented with an asymmetrical hearing pattern, marked by a >10dB difference between ear PTAs.

Air and Bone Conduction.

The results revealed high frequency hearing loss for six individuals tested. The severity of the loss varied amongst participants. Two participants (P2 and P3) presented with normal hearing bilaterally, although it must be noted that P3 presented with a large air-bone gap in the right ear despite normal hearing thresholds. A total of 12 ears presented with hearing impairment isolated to the high frequencies. Table 19 provides an outline of pure tone air conduction findings per participant followed by Table 20 outlining the average, range and *SD* per frequency tested.

Table 19

Summary of Raw Air Conduction Threshold Data

Participants	Air conduction thresholds (in dB)															
	250Hz		500Hz		1000Hz		2000Hz		3000Hz		4000Hz		6000Hz		8000Hz	
	R	L	R	L	R	L	R	L	R	L	R	L	R	L	R	L
P1	20	10	25	15	20	10	25	25	45	45	50	55	60	50	65	45
P2	10	10	10	10	10	5	5	5	-	-	10	15	-	-	10	15
P3	25	15	20	10	25	5	25	5	25	-	15	0	25	-	30	-5
P4	0	0	10	5	5	5	5	15	-	-	20	20	30	25	40	55
P5	25	20	20	20	25	25	30	30	-	-	40	40	-	-	50	45
P6	10	25	10	20	10	20	15	35	5	30	5	30	25	45	55	70
P7	5	5	5	5	5	5	5	15	-	35	30	50	60	70	70	70
P8	0	0	10	5	10	10	5	15	-	-	25	30	40	40	50	60

Note: R represents the right ear and L represents the left ear

Table 20

Air Conduction Average, Range and SD

		Air conduction thresholds (in dB)							
		250Hz	**500Hz**	**1000Hz**	**2000Hz**	**3000Hz**	**4000Hz**	**6000Hz**	**8000Hz**
Average	R ear	11. 9	13.75	13.75	14.3	25. 0	24. 4	40. 0	46.35
	L ear	10. 6	11. 25	10. 6	18. 1	36. 7	30. 0	46. 0	44.4
Range	R ear	0 – 25	5-25	5 – 25	5 – 30	5 – 45	5 – 50	25 – 60	10 – 70
	L ear	0 – 25	5 – 20	5 – 25	5 – 30	30 – 45	0 – 55	25 – 70	-5 – 70
SD	R ear	10. 3	6.9	8.3	10.8	20. 0	15. 2	16. 4	19.4
	L ear	9. 0	6. 4	7. 8	11. 0	7,6	18. 3	16. 4	26.7

Note: R represents the right ear and L represents the left ear

PTAs are provided in Table 21. PTA values fall within the range of normal hearing (0 – 25dB) (Hall & Mueller, 1997). PTA is typically calculated based on the 500Hz, 1000Hz and 2000Hz frequency threshold average and therefore does not take into account high frequency hearing (Harrell, 2002). Applying a two frequency pure tone average calculation maintained indications of normal hearing due to the isolation of hearing impairment to the 4000 – 8000Hz range, which is not included in these calculations (Harrell, 2002). All individuals with exception to P2 and P3, who presented with normal hearing bilaterally, presented with a high frequency hearing impairment not detected by two and/ or three-frequency PTA calculations.

Table 21

Pure Tone Average (PTA)

Participants	PTA (in dB)	
	Right	Left
P1	23. 3	16. 6
P2	8. 3	6. 6
P3	23. 3	6. 6
P4	6. 6	8. 3
P5	25	25
P6	11. 6	25
P7	5	8. 3
P8	8. 3	10
Average	15	20
Range	5 -31. 6	6. 6 – 26. 6
SD	14.1	8. 7

Type of Hearing Loss.

Table 22 provides an outline of bone conduction thresholds, with range, average and *SD* in table 23. A total of six individuals and 12 ears presented with bilateral sensorineural hearing impairment. Two participants (P2 and P3) presented with hearing within the normal threshold range bilaterally. P3 did present with a large air-bone gap in the right ear, however thresholds for both air and bone remained within the range of normal hearing. P3 was the only participant to present with a diagnosis of bulbar onset MND, however the presence of the air-bone gap in the right ear may also be attributed to the presence of wax occlusion in the right ear canal.

Table 22

Summary of Raw Bone Conduction Threshold Data

Participants	Bone conduction thresholds (in dB)									
	250Hz		500Hz		1000Hz		2000Hz		4000Hz	
	R	L	R	L	R	L	R	L	R	L
P1	15	10	25	15	20	5	25	20	35	25
P2	5	5	0	5	5	5	0	5	5	10
P3	0	5	0	5	-5	5	10	5	-5	-5
P4	-5	-10	5	5	-5	0	5	15	20	15
P5	20	20	20	20	25	15	30	30	30	40
P6	5	10	10	20	0	0	15	25	5	10
P7	5	-10	-10	-5	-5	0	5	10	20	45
P8	0	-5	10	0	5	5	5	10	25	30

Note: R represents the right ear and L represents the left ear

Table 23

Bone Conduction Average, Range and SD

		Bone conduction thresholds (in dB)				
		250Hz	500Hz	1000Hz	2000Hz	4000Hz
Average	R ear	5. 6	7. 5	5. 0	11.9	16. 9
	L ear	3. 1	8. 1	4. 4	15. 0	20. 6
Range	R ear	-5 – 20	-10 – 25	-5 – 25	0 – 30	-5 – 35
	L ear	-10 – 20	-5 – 20	0 – 15	5 – 30	-5 – 45
SD	R ear	8. 2	11. 3	11. 6	10.7	13. 9
	L ear	10. 7	9. 2	5. 0	9.3	16. 6

Note: R represents the right ear and L represents the left ear

Configuration of Hearing Loss.

Twelve ears tested presented with a sloping configuration of hearing loss (P1, P4, P5, P6, P7, P8). A further four ears presented with a flat configuration of hearing thresholds (P2 and P3).

Degree of Hearing Loss.

Figure 6 outlines the severity of the hearing loss in 16 ears. All the ears presented with normal hearing (0 -25dBHL) in the low-mid frequency range, consequently presenting with normal PTAs. The isolation of hearing loss to the 4000-8000Hz range limited the value of PTA scores which tend to be isolated to the low-mid frequency range. Applying a two-frequency calculation of PTA furthermore failed to represent the hearing loss accurately. The severity classification below is thus based on high frequency hearing thresholds, the region where 12 of the total ears presented with hearing impairment. The remaining four ears presented with normal hearing thresholds across the frequency range.

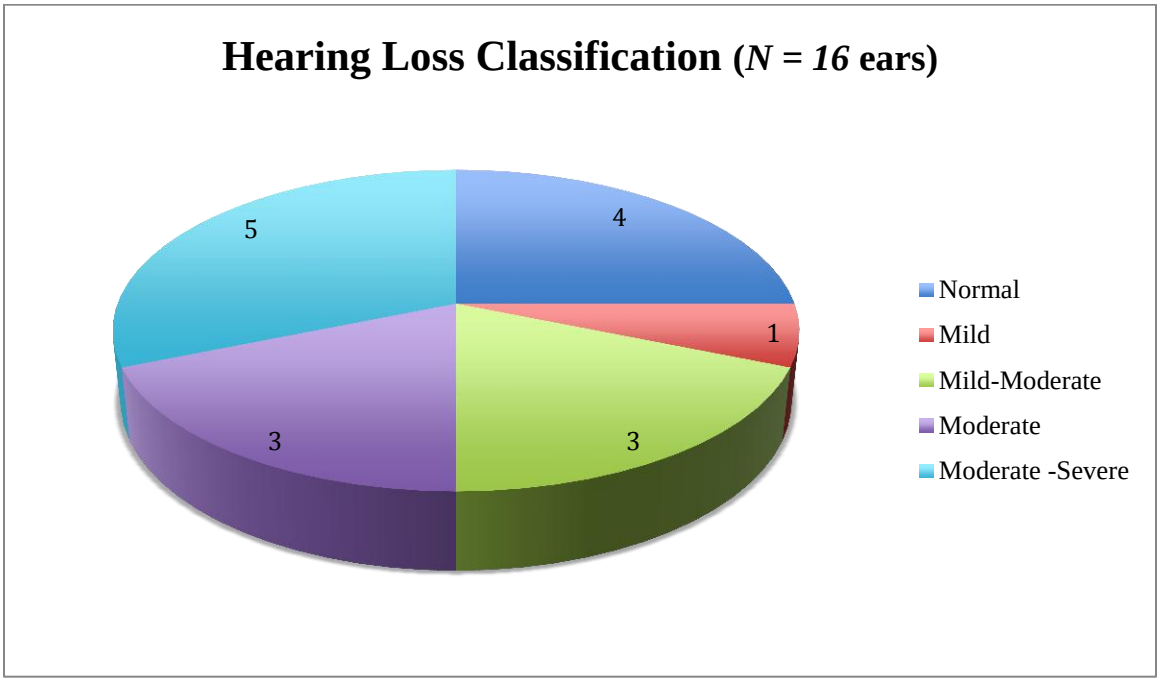

Figure 6. Hearing Loss Classification (*N* =16).

A total of four ears tested presented with normal hearing thresholds. One ear presented with a mild high frequency loss, followed by three ears presenting with a mild-moderate hearing

impairment in the high frequencies. Three ears presented with a moderate high frequency hearing loss, while the remaining five ears presented with a moderate – severe hearing loss in the high frequencies.

Speech Audiometry.

A summary of speech audiometry results is presented in Table 24 with average, range and SD in Table 25. These results include findings from speech reception thresholds (SRT), most comfortable levels (MCL), threshold of discomfort (TD), dynamic range (DR) and speech discrimination (Sd) testing.

Table 24

Summary of Raw Speech Audiometry Data (in dB)

	Speech audiometry (in dB)													
	SRT (in dBSL)		**MCL (in dBHL)**		**TD (in dBHL)**		**DR (in dBHL)**		**Sd (SRT+5dB) (%)**		**Sd (SRT+25dB) (%)**		**Sd / TD – 10dB) (%)**	
	R	L	R	L	R	L	R	L	R	L	R	L	R	L
P1	25	15	60	50	100	105	75	90	38	44	95	80	90	90
P2	5	10	40	40	85	80	80	70	82	79	97	97	100	100
P3	15	20	60	70	80	95	65	70	46	6	94	97	97	100
P4	20	20	60	65	95	95	75	75	79	64	100	97	100	100
P5	35	60	70	70	80	85	45	25	64	64	80	93	93	85
P6	10	10	60	55	70	80	60	70	40	40	80	100	100	90
P7	35	20	65	55	85	70	50	50	56	0	97	77	90	97
P8	25	20	55	50	90	85	65	65	72	60	88	84	100	97

Note: R represents the right ear and L represents the left ear

Table 25

Speech Audiometry Average, Range and SD

		Speech audiometry (in dB)						
		SRT	**MCL**	**TD**	**DR**	**Sd (%)/ SRT+5dB**	**Sd (%) / SRT+25dB**	**Sd (%)/ TD -10dB**
Average	R ear	21. 3	58. 8	85. 6	64. 4	59. 6	91. 3	96. 3
	L ear	21. 9	56. 9	86. 3	64. 4	44. 6	90. 6	94. 9
Range	R ear	5 – 35	40 – 70	70 – 100	45 - 80	38-82	80 – 100	90 - 100
	L ear	10 – 60	40 – 70	70 – 105	20 - 90	0 - 79	77 – 100	85 - 100
SD	R ear	10. 9	8. 8	9. 4	12. 4	17. 3	7. 8	4. 6
	L ear	16. 0	10. 7	11. 3	19. 4	28. 5	8. 9	5. 8

Note: R represents the right ear and L represents the left ear

Speech Reception Threshold.

Speech reception in normal hearing individuals typically presents at 25dB sensation level (SL) (Brandy, 2001). Thirteen ears (n = 13) presented with SRT scores representative of the normal range (Table 24). In total, three ears presented with SRT scores outside the normative range. P5 experienced difficulty with SRT testing particularly for the left ear, where a SRT of 60 dBHL was recorded. This is a marked discrepancy when reviewed against a PTA of 25 dB in the corresponding ear. P7 presented with a SRT of 35 dBHL in the right ear further showing discrepancy between the PTA of 5 dB obtained for the same ear.

Most Comfortable Level).

The MCL fell within the normative data range of 40 – 60 dBHL for all participants (N =8) (Table 24 and Table 25).

Threshold of Discomfort.

TD levels typically present within the 80 – 100 dBHL for individuals with normal hearing (Thibodeau, 2007). TD averages fell within the normative data range, with the right ear average of 85.6 dBHL and the left ear average of 83.6 dBHL (Table 25). P6 and P7 were however noted to present with TDs at 70 dBHL in the right and left ears respectively, which falls below the normative range of discomfort levels (Table 24). This suggests sensitivity to increased loudness intensity (Thibodeau, 2007). Discomfort levels below 85 dBHL are reported to be indicative of varying severity levels of hyperacusis (Vernon, 2002). These are evident for P2 left, P3 right, P5 right and P6 bilaterally all of whom reported complaints of hyperacusis (Table 24). P7 also presented with TD below 85 dBHL in the left ear, however this is not paired with individual reports of hyperacusis.

Dynamic Range.

These results are typically noted to present above 60dBHL in instances of normal hearing (Thibodeau, 2007). An average DR of 63.1 dBHL and 63.8 dBHL was recorded in the right and left ears respectively for all the participants (Table 25). These fall within the lower range of acceptable dynamic scores. Five ears presented with DR scores below the 60 dBHL minimum. This was observed in the right ear for P6 and bilaterally for P5 and P7 (Table 24). These scores are suggestive of recruitment and support the presence of cochlear pathology (Thibodeau, 2007).

Speech Discrimination (Sd).

Sd testing revealed findings supporting normal hearing abilities. In normal hearing subjects, speech discrimination scores are anticipated to reach between 88 – 100% at the level of

SRT +25dB. Scores below this are associated with a conductive or sensorineural (cochlear or retrocochlear) pathology. Rollover is not typically expected in normal hearing cases and is more highly associated with sensorineural hearing impairments. A rollover percentage of less than 20% supports the presence of cochlear site of lesion, while a rollover percentage greater than 20% provides evidence of retrocochlear pathology (Brandy, 2001).

Scores typically revealed improved speech discrimination abilities at increasing intensity levels. Average scores for right and left ears support the Sd abilities expected of individuals with normal hearing (Table 25). However, examination of individual ears revealed three ears achieved percentage scores below the 88% minimum at SRT + 25dB supporting some extent of hearing impairment. This was identified for the left ear of P1 and the right ears for P5 and P6 (Table 24). Participant averages at TD – 10dB were 96.3 dBHL and 94.9 dBHL in the right and left ears respectively (Table 25). These averages, when compared with SRT + 25dB averages do not suggest the presence of rollover, however evaluation of individual scores reveal less than 20% rollover in four of the ears tested. This was noted in the right ears of P1 and P7 and the left ears of P5 and P6 (Table 24). These findings support the presence of cochlear pathology.

The recorded SRT scores for P5 (left ear) yielded thresholds higher than anticipated (based on PTA values). This discrepancy in findings is further highlighted by pure tone findings classifying hearing impairment to be of mild-moderate severity. Furthermore, Sd performance for the left ear indicated scores within the normative 88-100% range for both SRT +25dB and TD -10dB and no rollover, suggesting normal speech discrimination abilities.

A small difference of 4.9% and 4.3% can be observed between SRT+25 dB and TD-10 dB averages for the right and left ears respectively. It is important to note that many participants presented with hearing impairment isolated only to the high frequency region of 6000-8000Hz, while a majority of speech sounds fall within the 2000-4000Hz range. Sd scores suggested discrimination abilities within the normal range at SRT+25dB even though hearing impairments were identified for all, but two participants in the sample. The presence of high frequency hearing impairment limited to the 6000-8000Hz range may account for these discrepancies.

Distortion Product OAEs.

The average results obtained for DPOAE measures are outlined in Table 26.

Table 26

DPOAE Frequency Specific Average, Range and SD

			DPOAE Averages (in dB)							
		Ear	**750Hz**	**1000Hz**	**1500Hz**	**2000Hz**	**3000Hz**	**4000Hz**	**6000Hz**	**8000Hz**
Amplitude	Ave	R	12. 1	11. 3	11. 7	9. 8	9. 6	9. 5	7. 0	5. 8
		L	11. 8	11. 1	10. 3	8. 2	9. 5	7. 9	5. 8	0
	Range	R	10. 0–15. 3	4. 4–16. 8	9. 2– 8. 0	5. 2– 3. 3	6. 4– 1. 2	3. 9 – 4. 3	4. 3 – 3. 3	3. 5 – 3. 4
		L	10. 1 –6. 0	8. 3 –15. 0	7. 7 –11. 6	0. 0 –11. 4	4. 6 –14. 2	-7. 8 –14. 5	-0. 9 -12. 4	-12. 2–13. 9
	SD	R	2. 0	3. 5	2. 7	2. 8	1. 8	3. 6	3. 2	3. 3
		L	1. 9	2. 0	1. 3	3. 7	4. 0	4. 6	6. 6	9. 8
SNR	Ave	R	8. 75	8. 09	8. 9	7. 2	7. 8	8. 3	5. 6	4. 4
		L	7. 3	8. 1	7. 9	9. 1	9. 6	8. 1	6. 8	3. 1
	Range	R	6. 5 –10. 8	6 – 15. 7	6. 3 - 18	6. 1 – 8. 9	6 – 12. 6	4. 4 –19. 6	-5. 5 –13. 3	-2 – 7. 6
		L	6. 3 –10. 2	6. 1 –13. 9	6. 1 –10. 2	6. 8 –15. 7	3. 5 –15. 8	1. 5 –16. 5	-1. 8 –16. 5	-5. 8 – 7. 2
	SD	R	2. 7	3. 3	3. 8	1. 0	2. 1	4. 7	5. 2	2. 9
		L	1. 2	2. 6	1. 6	3. 0	4. 6	4. 7	6. 2	5. 3

Note: Ave (average); *SD* (standard deviation); R (right); L (left); SNR (signal to noise ratio)

Based on the averages provided, it is evident that DPOAE amplitudes were present across the low-mid frequency ranges of 750Hz-1500Hz, while an increase in abnormal outer hair cell function was documented for high frequency ranges 2000-8000Hz. These averages represent a gradual decline in distortion product amplitude outside the >10 dB norm applied to this study in relation to increasing pure tone frequency. Signal-to-Noise Ratio (SNR) is considered as a means of differentiating impaired versus normal DPOAEs, although more specifically may be used as a means of determining the reliability of the DPOAE levels measured (Gorga, Neely, Dorn, & Hoover, 2003). A SNR of >6 dB was achieved consistently throughout the frequency range with exception to those high frequency ranges of 6000Hz – 8000Hz. Absent DPOAEs were recorded in the left ear at 8000Hz for P6 and P7 at 6000-8000Hz in the left ear. The remaining participants all obtained DPOAEs, although not all of these were normal emissions. P2 and P3 presented with normal hearing thresholds and acquired DPOAE recordings that corresponded with all emissions were recorded across the frequency range with a SNR above 6dB. The remaining participants achieved emissions across the frequencies ranging from 750Hz-4000Hz, while SNR showed deterioration in the 6000Hz-8000Hz range. Abnormal DPOAEs in this frequency region is suggestive of impaired functioning of the outer hair cells of the cochlea, isolating the cochlea as the potential site of lesion.

A high correlation between DPOAE and audiometric thresholds is typically noted in the mid-high frequency range (Gorga et al., 2003). This is supported in these findings whereby DPOAE SNRs correspond with the increasing loss of hearing in the high frequency range. Outer hair cell function is isolated to approximately 50 – 60 dB hearing loss resulting in less reliable DPOAE recordings as hearing loss approaches these threshold levels (Bartnik et al., 2009). The

increase in hearing loss noted in the high frequencies resulted in a SNR falling below the 6dB range suggesting outer hair cell damage in the range of 50-60 dB and beyond.

Pearson's product moment correlation was applied to the data to best determine correlations. DPOAE testing revealed moderately positive correlations between all consecutive frequencies tested, namely 1000Hz and 1500Hz, 2000Hz and 3000Hz, 4000Hz and 6000Hz and 6000Hz and 8000Hz. These correlation coefficients ranged from 0.51 – 0.69, while strong positive correlations were revealed for 6000Hz and 8000Hz (R = 0.93). Moderately negative correlations were identified between pure tone audiometry and DPOAE measures at 1000Hz, 2000Hz and 4000Hz. These correlation coefficients ranged between R = -0.59 to -0.73, while strong negative correlations at 6000Hz and 8000Hz ranged between R = -0.89 to -0.91. Ongoing consideration for the low number of data points applied to these analyses prevents generalization of these findings.

Neurodiagnostic ABR.

The summary of neurodiagnostic ABR results are outlined in Table 27. When applying a *SD* of 0.2 msec to inter peak latencies (IPL) normative data (95% confidence interval) suggests that only I-III and III-V IPLs exceeding 2. 2 msec and IPLs I-V exceeding 4. 2 msec are considered abnormal (Don & Kwong, 2002). As is evident in Table 27, IPL averages were within the normal range for all participant ears in the current study (N = 16) and did not exceed the time frame outlined in the protocol applied to this study. Absolute wave latencies (AWL) for wave I, III and V were present at 1.6 msec, 3.6 msec and 5.6 msec (*SD* of 0.2msec; 95% confidence interval) in individuals without retrocochlear pathology (Don & Kwong, 2002). The

averages calculated for participants in the current study reflect these norms accordingly with AWL for waves I, III and V not exceeding these parameters. Inspection of each case in isolation brings to light a number of factors that are not revealed when reviewing combined test averages.

IPL for wave I-V fall within the normal range (4. 0 msec with 0.2 msec *SD*). P2 and P5 (25%) present with slightly shorter wave I–V IPLs. Five participants (P1, P4, P5 bilaterally, P3 right, P6 left) and eight ears presented with longer wave I-III IPLs, while the remaining participants presented with I-III IPLs within the normative range. The five participants presenting with slightly delayed IPLs also presented with the more severe high frequency hearing impairment. All participants (N = 8) presented with wave III-V IPLs that were shorter than the outlined normative range (1.8 – 2.2 msec).

Table 27

Summary of raw neurodiagnostic ABR data with Average, Range and SD.

	Ear	Interpeak latency (IPL) (in msec)			Absolute Wave Latency (AWL) (in msec)			IAWLD of Wave V (in msec)
		Waves I-III	Waves III-V	Waves I-V	Wave I	Wave III	Wave V	
P1	L	2.3	1.6	3.87	1.33	3.6	5.2	0.13
	R	2.3	1.7	4.0	1.33	3.6	5.33	
P2	L	2.0	1.67	3.67	1.2	3.2	4.87	0.0
	R	1.9	1.65	3.54	1.33	3.22	4.87	
P3	L	2.1	1.67	3.87	1.3	3.4	5.1	0.17
	R	2.3	1.67	3.93	1.33	3.6	5.27	
P4	L	2.3	1.78	4.11	1.27	3.6	5.38	0.18
	R	2.3	1.6	3.87	1.33	3.6	5.2	
P5	L	2.4	1.27	3.67	1.2	3.6	4.87	
	R	2.3	1.37	3.64	1.33	3.6	4.97	0.10
P6	L	2.3	1.73	4.0	1.33	3.6	5.33	0.23
	R	2.0	1.77	3.8	1.3	3.33	5.1	
P7	L	2.0	1.77	3.77	1.33	3.33	5.1	0.1
	R	1.7	1.78	3.9	1.3	3.42	5.2	
P8	L	2.1	1.7	3.73	1.29	3.4	5.1	0.00
	R	2.2	1.56	3.73	1.37	3.54	5.1	
Average	L	2.17	1.64	3.83	1.28	3.46	5.12	0.11
	R	2.10	1.64	3.80	1.31	3.28	5.13	
Range	L	2 – 2.4	1.27-1.78	3.67– 4.0	1.2 – 1.33	3.2 – 3.6	4.87 – 5.38	0 – 0.23
	R	1. 72 -2.27	1. 37–1.78	3.54– 4.0	1.27 – 1.33	3.22 –3.6	4.87 – 5.33	
SD	L	0.15	0.16	0.16	0.05	0.16	0.19	0.08
	R	0.21	0.13	0.16	0.02	0.15	0.15	

The AWL for waves I, III and V for all the participants (N = 16 ears) were within normal limits (see Table 27), although slightly shorter latencies were consistently noted particularly for wave I and V AWL recordings. The shorter latencies identified throughout AWL measurements may be associated with the selection of a rarefaction polarity, which typically present with shorter latencies of 0.1 – 0.2 msec from the normal range (Don & Kwong, 2002).

An interaural wave latency differences (IAWLD) for wave V of >0.4 msec is associated with retrocochlear pathology (Hall & Mueller, 1997). It is evident from Table 27 that the IAWLD for all participants were within normal limits (average = 0.11 msec). It can thus be concluded that based on the results of the neurodiagnostic ABR test, that none of the participants presented with auditory neuropathy or pathology at the site of the brainstem.

These findings correlate with pure tone audiometry and Sd results, which through the absence of >20% rollover for the total sample further eliminated any audiometric indicators of a retrocochlear site of lesion. These findings, when interpreted with DPOAE results, suggest that where SNHL is indicated, a cochlear site of lesion is most probable for participants in the current study.

Perceived Psychosocial Implications of Hearing Loss on Daily Functioning

This section will present the findings from the HHIA inventory related to socially and emotionally perceived implications of hearing loss. It will in addition explore the relation between the HHIA and audiometric test measures obtained for participants in this study. This section will then conclude with a review of participant responses to the HEQ and participant and caregiver perceptions related to the value of auditory testing and diagnosis alongside the MND.

HHIA Review.

The HHIA reviews individual hearing experience on both a social and an emotional scale to ascertain the psychosocial level of functioning of the hearing impaired individual (Newman et al., 1990). Participant scores are added resulting in a handicap classification of ‘none’,

suggesting no handicap related to hearing impairment or ‘mild’, suggesting limited effects of hearing impairment on social and emotional wellbeing. A ‘moderate’ score suggests limited or average success in functional activities related to hearing ability, whilst a ‘severe’ score is indicative of critically harmful effects on individual social and emotional wellbeing as a result of hearing impairment (Newman et al., 1990). Participant responses to the HHIA are outlined in Table 28, followed by a statistical review of the correlation between participant auditory test findings and the associated emotional and/or social implications thereof.

Table 28

HHIA Participant Results (N=8)

Q	Nature	Content of Question	P1	P2	P3	P4	P5	P6	P7	P8
1	Social	Less phone use	Y	N	N	N	N	Y	S	N
2	Emotional	Embarrassment	N	N	N	N	N	N	N	N
3	Social	Group Avoidance	N	N	N	N	S	Y	N	N
4	Emotional	Irritability	S	N	N	N	S	S	Y	N
5	Emotional	Family Frustration	S	N	N	N	N	N	Y	N
6	Social	Difficulty at Party/ Social Function	Y	S	N	N	S	N	S	N
7	Social	Understanding clients/coworker	S	N	N	N	S	N	N	N
8	Emotional	Handicap	N	N	N	N	N	N	N	N
9	Social	Difficulty visiting doctors, family	S	N	N	N	N	N	N	N
10	Emotional	Co-Worker Frustration	N	N	N	N	N	N	N	N
11	Social	Difficulty in Movie/Theater Setting	N	N	N	N	Y	Y	S	N
12	Emotional	Nervousness	N	N	N	N	N	N	N	N
13	Social	Less frequent visiting	N	N	N	N	S	Y	N	N
14	Emotional	Family Arguments	S	N	N	N	Y	N	S	N
15	Social	Difficulty TV/radio	S	N	N	N	Y	S	Y	S
16	Social	Less frequent shopping	N	N	N	N	N	N	N	N
17	Emotional	Difficulty hearing – upsetting	S	N	N	N	Y	Y	S	N
18	Emotional	Isolation/ Desire to be left alone	N	N	N	N	N	N	N	N
19	Social	Less frequent communication with family	N	N	N	N	N	S	N	N
20	Emotional	Hampers social life	S	N	N	N	Y	Y	S	N
21	Social	Difficulty in restaurants	Y	N	N	N	Y	S	Y	Y
22	Emotional	Cause depression	N	N	N	N	N	N	N	N
23	Social	Less frequent TV/radio use	N	N	N	N	N	N	N	N
24	Emotional	Discomfort communicating with friends	N	N	N	N	S	N	N	N
25	Emotional	Isolation when in social groups	S	N	Y	N	Y	N	S	N
		SOCIAL TOTAL	18	0	4	0	20	22	14	6
		EMOTIONAL TOTAL	12	0	0	0	20	10	16	0
		GRAND TOTAL	30	0	4	0	40	32	30	6
		HANDICAP RATING	MILD	NONE	NONE	NONE	MOD	MOD	MILD	NONE

Note: Y represents ‘Yes’ responses, N represents ‘No’ responses and S represents ‘Sometimes’ responses

The results indicate that four participants reported no auditory handicap. Two participants reported a mild hearing handicap, while a further two reported a moderate hearing handicap impacting social and/or emotional functionality. Only two participants (P5 and P7) acknowledged hearing difficulties in the initial case history interview, with the remaining six reporting no auditory impairment.

The HHIA scale revealed wide variation in scores across participants, ranging from no reported handicap to moderate reported handicap. Four participants (P1, P3, P6 and P8) reported greater handicap at a level of social functioning over emotional wellbeing. One participant (P7) reported greater handicap at a level of emotional functioning. Three participants reported no differences between emotional and social functioning, of whom two (P2 and P4) presented with no handicap and one participant (P5) presented with a moderate handicap on the HHIA.

Correlation analysis for social versus emotional participant responses was performed using Pearson's product moment correlation coefficient. A 0.05 significance level was used for this statistical measure. The social versus the emotional responses showed good correlation ($r = 0.86$; $p = 0.006$) suggesting participants with higher social handicap typically were noted to have higher emotional handicap scores as well. Across the entire sample, MND participants responded '*yes'* to 12.5% (12 responses) of the twelve social questions, while a response of '*sometimes'* was associated with 15.6% (15 responses) of these questions. The remaining 71.9% of participant responses indicated that participants did not associate with the remaining social handicaps outlined in the HHIA. A total of 7.69% (8 responses) out of thirteen questions linked to the emotional effects of hearing impairment received '*yes'* responses from participants, while a response of '*sometimes'* was associated with an

additional 11.5% (12 responses) of the thirteen questions. The remaining 80.81% of participant responses indicated that participants did not experience the remaining emotional handicaps outlined by the HHIA.

Despite the good correlation between social and emotional scores, the percentage of positive scores from the HHIA suggests that the social implications of hearing impairment appear to be more readily acknowledged and reported than those at an emotional level. Participants reporting occasional difficulties in certain domains may represent the total of the sample experiencing initial markers suggestive of hearing impairment and/or denial related to the vast range and extent of deterioration in bodily function brought about by MND.

Difficulties experienced in different contexts were also probed. Four participants agreed that they experience social difficulty in a restaurant setting. Two participants reported difficulties in social activities including telephone use, listening success in movies, and television or radio use. A further one participant indicated that difficulty with telephone use and listening in movies was 'sometimes' problematic. Three participants indicated television and radio use to be an occasional area of difficulty, alongside difficulties in loud noise situations e.g. party/ social groups. The latter was met with firm agreement by two participants in the total sample.

At an emotional level the most common participant concerns were related to items 17, 20 and 25 where hearing difficulties were identified to be 'upsetting', a hindrance to social activity and result in isolation from groups of people respectively. In these instances, two of the participants positively identified with these difficulties, with a further two reporting occasional difficulties in each of the above listed instances.

HHIA and Auditory Testing.

A comparative review of the recorded HHIA responses and the auditory findings (PTA and speech audiometry) was executed as a means of determining the psychosocial impact of hearing impairment in the functioning of the participants in this study. Pearson's product moment correlation coefficient was used as the statistical measure for the review of HHIA findings and pure tone averages, with a confidence interval of 0.05.

HHIA and Pure Tone Average.

Statistical review of the correlation between PTA averages against HHIA social scores are indicative of a relatively strong, positive correlation ($r = 0.71; p = 0.050$) (Table 29). This suggests that for those participants presenting with higher pure tone averages, (i.e. poorer auditory abilities), the level of reported handicap at a social level of functioning increased. A poorer correlation was observed between pure tone averages and HHIA emotional handicap ratings ($r = 0.63; p = 0.095$). It is postulated that MND individuals in this study do not readily acknowledge or report the emotional implications of hearing loss (e.g. feelings of embarrassment, self-isolation, nervousness and/or perception of auditory handicap), despite audiometric evidence of hearing impairment. Review of the total HHIA scores revealed a positive correlation ($r = 0.69; p = 0.056$), indicating that the higher the total pure tone average, the higher the total HHIA level of handicap are.

Table 29

Statistical Correlation between HHIA and Pure Tone Average

Category	*r*	*Correlations*
HHIA Social	0. 71	*0.050
HHIA Emotional	0. 63	0.095
HHIA Total	0. 70	*0.056

*significant at the 90% confidence level

The PTA revealed moderately positive correlations with social and total HHIA scores and a poor correlation for emotional handicap ratings. Pure tone audiometry is therefore an adequate indicator of the level of auditory impact at a social level for individuals with MND, although the limited sample size precludes the identification of any strong, definitive trends.

HHIA and Speech Audiometry.

The correlation between the average speech discrimination scores and the HHIA totals revealed significant negative correlation of r = -0.74 (p = 0.034). This suggests that an increase in speech discrimination abilities corresponded with lower HHIA scores. The correlation between speech discrimination and social HHIA rating revealed a significant negative correlation of r = -0.74 (p = 0.038), while emotional ratings from the HHIA supported a negative correlation of marginal significance (r = -0.70; p = 0054).

Hearing Experience Questionnaire (HEQ)

The HEQ comprised a participant- as well as a caregiver component. The participant questionnaire explored (i) contact with the discipline of speech pathology and/or audiology, (ii) rating functional skills in order of importance, and (iii) the importance of receiving an auditory diagnosis.

Contact with Speech Pathology and/or Audiology.

Of all the participants, only two participants (P3 and P5) had attended consultations with a speech pathologist at the onset of MND to facilitate AAC. None of the participants in this study reported contact with an audiologist, or had been referred for an auditory evaluation or undergone previous auditory examination. None of the participants reported receiving information on hearing loss or the effects this may have on QoL from a speech-language pathologist, audiologist or another professional in the medical management team. As participants in the current study voluntarily remained an average of 45 minutes following test time to address concerns related to auditory symptoms and discuss stressors related to auditory difficulties, this suggests a need for such support.

Functional Rating Scale.

The categories of communication; arm movement; hearing; leg movement; vision; chewing and swallowing for nutrition were included in this section. Both MND participants and their caregivers were required to rate various functional abilities according to personal order of perceived importance when related to living with MND. Figure 7 outlines participant and caregiver responses to this rating scale.

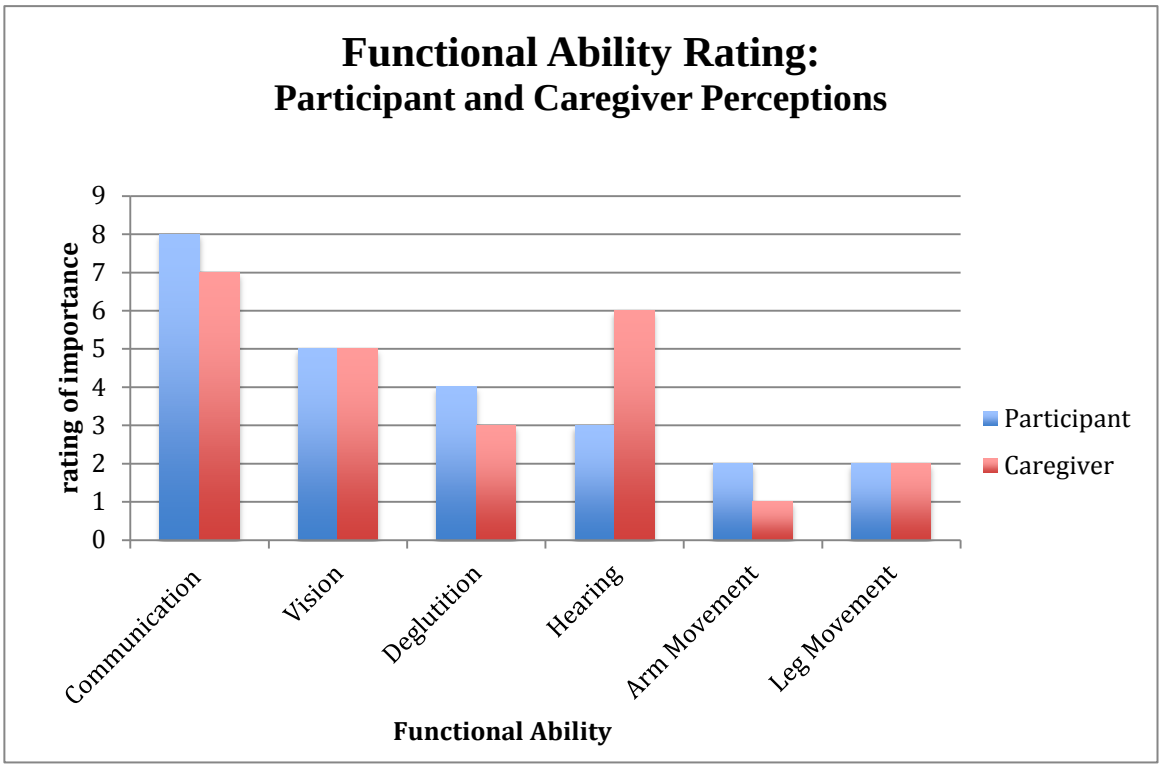

Figure 7. Functional Ability Rating: Participant and Caregiver Perceptions.

Results revealed variations in the perspective of participants and caregivers. Both caregivers and participants rated vision and leg movement similarly. Communication, deglutition, hearing and arm movement however presented with more differences in opinion. Amongst these differences, the rating of hearing ability presented with the greatest contrasting views between the two groups.

Communication rated highly amongst both participants and caregivers with all MND participants (N = 8) and seven caregivers identifying communicative skills as a priority functional ability. The general consensus amongst individuals with MND was that communication remained a more important functional ability than hearing. Participants suggested a loss of hearing could be accommodated for through the use of compensatory techniques such as lip reading, while the loss of communication was suggested to be more

devastating and less easily compensated for since loss of motor ability eliminates gesture as a compensatory technique for verbal communication. Participants did not readily acknowledge AAC devices as a form of facilitating communicative breakdown, which essentially could accommodate for expressive loss of verbal communication. Individuals with MND then described visual ability and deglutition to be amongst their most valued abilities, with five participants (n =5) highlighting the value of vision and four (n =4) emphasizing the importance of deglutition. Arm and leg movement were not rated highly amongst participants, six (n = 6) of whom indicated that adjustments to lifestyle can and in many instances have been made to accommodate the loss of these abilities. The general perception that communication and vision were central to maintaining contact with the outside world was shared amongst participants, while deglutition was identified as the third critical functional domain being connected to a number of varying explanations and participant perspectives.

Participants in this study highlighted the connection between eating and maintenance of social wellbeing. Participants reported that swallowing difficulties were harmful to social opportunity and since most adult socialization occurs alongside dining activities a great deal of anxiety and distress is brought on. Participants felt that swallowing difficulties in social settings were isolating and damaging to their self-esteem and dignity, more so than auditory impairment. When probed further regarding the option of non-oral feeding methods, these were also declined as a socially acceptable solution with suggestions that this gives rise to feelings of social exclusion. Participants were noted to associate deglutition very closely with social success.

Where participants rated visual ability to be the second most important requirement contributing to quality of daily living, caregivers interestingly rated hearing ability as a key

ability that would have influence on the QoL of their MND spouse/partner. Hearing ability was viewed as important by six caregivers, in contrast to three MND participants. The impairment of hearing ability was identified to be essential for maintaining QoL, reducing burden and maintaining a sense of involvement. Visual ability rated third highest by caregivers of MND participants with five caregivers identifying the importance of visual abilities. This was equal to those provided by participant responses. A firm awareness of the benefits of preserved visual acuity was expressed by caregivers particularly in connection with the use of AAC devices and the use of these devices as a means of maintaining expressive communicative function. Arm and leg movement contributed to the lowest rating by caregivers. Leg movement was rated as a marginally more important ability by two caregivers in contrast to arm movement which was only isolated by one caregiver. The remaining caregiver's responses to arm and leg movement supported the theme of adjustment and indicated that current technology allows for livable modifications to be successfully achieved overcoming difficulties with mobility therefore making this loss of function secondary.

Identifying Importance of Auditory Diagnosis.

Individuals with MND and their caregivers were questioned on the relevance of an added diagnosis of hearing loss superimposed on the diagnosis of MND. Five participants with MND reported that the added diagnosis of hearing loss would be of no relevance to them. Hence, acquiring information about auditory function would not be pursued individually. One MND participant reported that these findings would be '*interesting'* to know about, while the remaining two MND participants reported this information to be of extreme importance (Figure 8). Of the caregivers, five individuals reported this information to be of extreme importance, while a single caregiver agreed that this information would be

'*interesting'* and the remaining two MND caregivers reported that this information would be irrelevant (Figure 8).

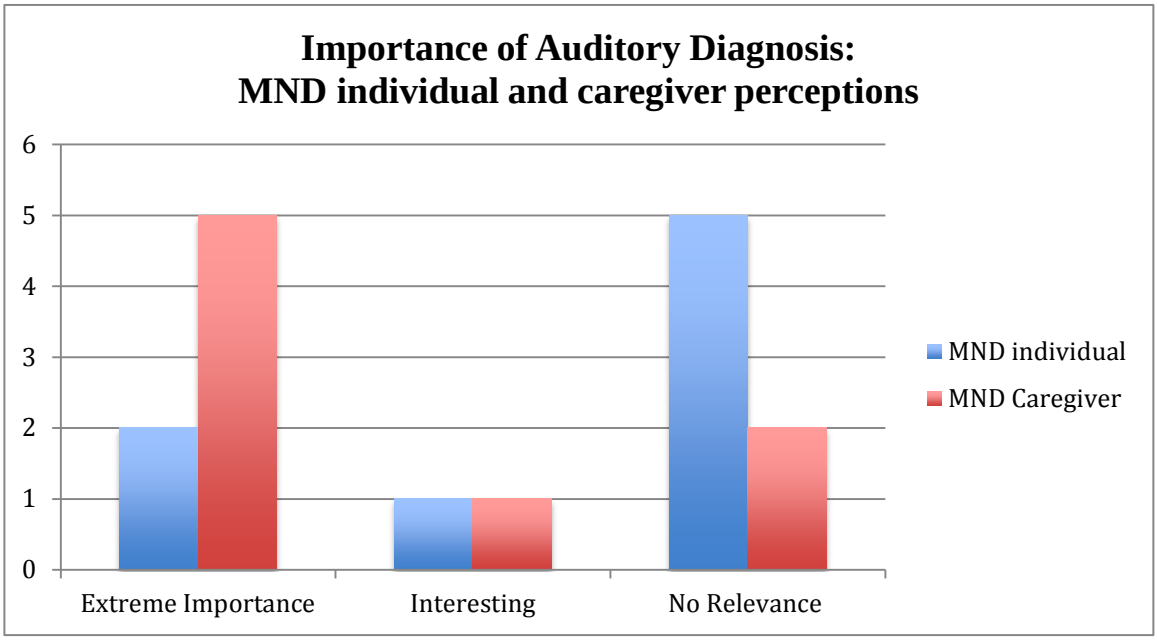

Figure 8. Importance of Auditory Diagnosis: MND individual and caregiver perceptions

Thematic mapping of participant and caregiver responses at this level of questioning revealed variations in perceptions. However, a number of key elements overlapped. Three primary themes emerged following the coding of responses. These included that of positive perceptions relating to control, self preservation and autonomy; negative perceptions relating to burdens, vulnerability and depersonalization and general awareness towards auditory impairment and management benefits.

Negative perceptions towards auditory diagnosis.

Five MND-individuals and two MND caregivers expressed the diagnosis of auditory impairment to be of no relevance to them indicating that the presence of a diagnosis of auditory impairment would serve more harm than value. Individuals with MND related

narratives of negative perceptions towards auditory diagnosis and placed emphasis on the sub-themes of burden, vulnerability and fear. Similar themes were identified in the responses provided by MND caregivers. Table 30 provides illustrative examples of caregiver and MND individual negative perceptions towards auditory diagnosis.

Table 30

Value of auditory diagnosis: Negative perceptions

	Individual with MND	MND Caregiver
Burden	* 'it's just another thing to deal with when there is already so much else to cope with' (P7) * 'with my lifespan being cut short, I don't want to spend my time at doctors unless it will improve things for me, not introduce new problems' (P4)	* '..will need more support from my side' * 'It will add to more stressful communicating' (CP1) * 'Every extra thing costs and these costs add up so if its something she can cope without then that's how it needs to be' (CP4)
Vulnerability (physical and psychological)	* 'finding out I have a hearing loss would just add to the loss of my legs, arm and talking. I'd be better off not having a proper diagnosis and be ignorant to it.' (P4) * '…have enough to deal with.' (P2)	* 'Everyday is a battle. Adding a new diagnosis …. will be a blow and emotionally devastating to everyone.' (CP2) * '…the relevance of hearing loss diminishes' (CP8)
Fear (isolation and depersonalization)	* 'finding out I have a hearing loss as well would rob me of yet another piece of who I am' (P2) * 'All my life I said hearing was my most important sense. To lose that as well would be devastating.' (P3) * 'I'd rather not know.' (P7)	* 'Music is his life!' It's how he copes with the isolation.' (P3) * '…it would be devastating to him and would take away one of the few joys left in his life.' (P3)

Individuals with MND and caregivers alike expressed concerns relating to the financial burdens. An added diagnosis of hearing loss was indicated to impose further expenses on the hefty medical expenses faced across the span of disease progression. Individuals with MND and caregivers further expressed that hearing impairment was something that could go without formal diagnosis and consequently medical expenses would

be spared for symptoms and/ or conditions that did not pose direct threats to life. In addition to the financial burden. MND caregivers further expressed concerns linked to the added responsibilities hearing loss would place in the hands of the caregiver such as adopting the role of social coordinator facilitating communication and managing communication breakdowns and attending audiology appointments. Both caregivers and individuals with MND acknowledged that the pursuit of a diagnosis that is not directly linked to life sustaining purposes would be an undesirable use of valuable time, especially in light of the progressive nature of MND.

Vulnerability of individuals with MND at physical and a psychological level were raised as an important sub-theme. This vulnerability contributed to the overall lack of desire to pursue auditory diagnosis. Concerns linked to individual vulnerability were further supported by a number of caregiver responses to the HEQ section of questioning. Individuals with MND typically emphasized the progressive nature of the disease and the ongoing loss of function parallel to motor neuron degeneration. Individuals with MND highlight the vast range of physical adjustments and readjustments to change required over time, alongside the emotional devastation that arises each time. It was expressed that the results of this concrete representation of disease progression leads to individuals repeatedly confronting mortality throughout the course of the disease. MND individuals therefore indicated that it is not only the process of adjusting to the physical and physiological changes occurring, but also to the emotional consequences of these. Individuals with MND described the experience of emotionally adjusting to physical changes to be '*exhausting'* and *'demoralizing'* and therefore indicated that this process of pursuing further diagnosis and confronting loss of function enhanced their vulnerability. Furthermore, individuals with MND suggested that this could negatively impact their mental and emotional state, both of which were highlighted as

necessary for deflecting depression and negative thoughts about mortality by participants in this study. MND caregivers acknowledged the emotional vulnerability in the face of progressive loss of function. They agreed that the emotional turmoil of adding further diagnosis to an already diagnostically untreatable degenerative disease serves no benefit for psychological and emotional wellbeing.

Fear as it relates to isolation and depersonalization emerged as a common theme amongst caregiver and MND-individual responses. It was raised by caregivers as a validation for the value of auditory diagnosis. Only two individuals with MND expressed understanding of the role of hearing in maintaining a sense of involvement and a sense of self in life. The rest of the individuals with MND reported varying degrees of fear for exclusion/ isolation and a loss of their sense of self. Caregivers expressed that hearing as it relates to recreational activities and hobbies e.g (music and/or television) aids individuals with distractions from negative circumstantial thoughts. This was further viewed to provide individuals with engaging experiences deviating from persistent feelings of loneliness and isolation even if only temporarily. Social activities as they relate to maintaining a sense of normalcy and promoting a sense of involvement, inclusion and value in in life were further highlighted as essential element to which hearing ability forms a foundation. While such fears were expressed by individuals with MND, only two individuals were able to relate these fears as factors that would be negatively impacted as a direct consequence of hearing loss. Five caregivers raised the awareness linked to fear of isolation brought about by hearing loss and the manner in which this poses the risk of depersonalization.

Positive Perceptions towards auditory diagnosis

Positive perceptions relating to the value of auditory diagnosis amongst the MND population exposed sub-themes linked to control and the need for self- preservation. Autonomy emerged as an additional sub-theme with the analysis of caregiver responses. Table 31 provides illustrations of extracts from caregiver and MND-individual raw data.

Table 31

Value of auditory diagnosis: Positive perceptions

	Individual with MND	MND Caregiver
Control	* 'hearing … my most important sense. At the first sign of hearing loss I would do everything I could to control this.' (P3). * 'There is so much I can't control in this disease. Hearing is something we can improve so if not, why not.' (P6)	* '…would strip him of his control over social situations' (P1)
Self-Preservation	* 'it's very frustrating not being able to hear properly…. You feel out of control in a group...You don't feel like your normal self.' (P6)	* 'If he couldn't hear I'd imagine this would make him self-conscious and withdraw. This would be uncharacteristic of his lively, joking nature.' (P1)
Autonomy	----------	* 'She would need me to step in and translate for her so she doesn't get embarrassed. She won't react to that well. She needs her independence' (P7) * 'He would need more help to socialize' (P3)

Individuals with MND who readily acknowledged the relevance of auditory diagnosis emphasized the role this would have on control and maintaining their sense of self, largely within the context of socialization. Similarly, caregivers supported this notion although they further emphasized the positive impact hearing would have on individual autonomy. Most caregivers acknowledged the increased social and communicative benefits afforded to affected individuals with managed hearing impairment. Two individuals with MND expressed the necessity of having control over loss of function. This highlighted the manner

in which maintaining a locus of control on oneself serves in terms of acquiring greater ownership of the disease. In support of this perspective, both individuals consequently supported the pursuit and value of auditory diagnosis and management where indicated. Caregivers demonstrated a stronger support for auditory diagnosis with corresponding views relating to individual empowerment and control in social contexts. They further emphasized the risks hearing impairment poses on individual character and personality, threatening a loss of self. Consequently the harm this poses on the independence of the individual with MND at a level of communicative success and reward was highlighted. The positive role of auditory diagnosis was hence seen to be of particular importance for MND caregivers forming a central element contributing to the positive psychosocial experience of the individual with MND. These responses demonstrate a strong caregiver focus and understanding of the social and emotional needs of the affected individual over and above the physical consequences of MND.

Awareness: Perceptions towards auditory diagnosis

The third theme that emerged from the HEQ responses related to the overall awareness of auditory diagnosis and rehabilitation. Table 32 illustrates selected extracts from caregiver and MND individual HEQ responses.

Table 32

Value of auditory diagnosis: Auditory awareness

	Individual with MND	MND Caregiver
Awareness of Benefits	* 'Hearing tests will tell me what I can do to make sure I am always able to hear people talking. I love being around people and if my hearing were to go it would be very hard for me to cope.' (P6)	* 'It will really affect his quality of life and without this knowledge we wont be able to get help ith making the adjustments to help him' (CP1) * 'Maybe he can get a hearing machine to stop his life from becoming even harder' (CP1)
Lack of Knowledge (education of auditory benefits)	* 'personally I can't see it making a difference in how I get by with MND' (CP4) * '…would have no positive impact' (P8)	* 'It's interesting for the people who study MND, but nothing more than that.' (CP8) * 'There is always the option of lip reading' (CP4)

Five caregivers supported the process of pursuing auditory diagnosis in contrast to only two individuals with MND. Caregivers demonstrated a more advanced level of understanding and awareness of the benefits auditory diagnosis would bring to affected individuals in terms of QoL enhancements, rehabilitative opportunities following auditory diagnosis and psychosocial benefits as a result of these opportunities for sensory regulation. In contrast individuals with MND presented with more limited insights into the awareness of auditory abilities. This supports the need for further patient education directed towards the role and involvement of the audiologist in the identification, diagnosis and management of auditory impairment. Further to this, a majority of individuals with MND (n = 5) reported that diagnosis of hearing loss could not yield positive outcomes. This highlights the importance of patient education regarding the role auditory ability and rehabilitation play in maintaining control, autonomy, social and emotional wellbeing. These expressed perceptions suggest the need for the redefinition of the medical management team at a secondary management level, to promote and refine a holistic multi-disciplinary care approach.

Summary

Six out of eight participants presented with hearing loss, comprising 75% of the total sample. whilst two participants in the sample presented with hearing within the normal limits. The degree of hearing loss in those hearing impaired individuals ranged from mild to moderate-severe high frequency loss. Prominent auditory symptoms reported by individuals in this study include difficulty listening in noise, tinnitus and hyperacusis.

Four individuals in this study reported no auditory handicap as revealed by the HHIA, followed by two participants reporting a mild handicap and an additional two reporting a moderate handicap. Social functioning appeared to be a more prominent area of handicap than emotional functioning as reported by the sample. No individuals in this study had previous contact with an audiologist nor had they been referred for audiological testing throughout the course of the disease. Individuals with MND rate communication, vision and deglutition as the most important of functional skills, while caregivers rate communication hearing and vision to comprise the most essential functional skills.

The majority of individuals with MND (n = 5) did not acknowledge the importance of auditory testing and diagnosis, whilst the most of caregivers (n =5) perceived it to be very important. There was an overlap in the key themes identified between caregivers and individuals with MND, namely negative perceptions, positive perceptions and auditory awareness. Negative perceptions relating to auditory diagnosis were isolated to the sub-themes of burden, vulnerability and fear. Burdens were described as they relate to financial, emotional and caregiver factors, while vulnerability was described in relation to physical and psychological vulnerability. The expression of fear was related to isolation and threats of depersonalization. Positive perceptions revealed subthemes of control, self-preservation and

autonomy. Caregivers presented with more realistic understanding of the benefits of auditory diagnosis and rehabilitation. Individuals with MND generally displayed limited awareness and insight into the role of audition as it relates to social and emotional wellbeing. Individuals with MND expressed desires for control, self-preservation and limiting communicative obstacles as valued components of their lives. The role of hearing ability as a factor implicating these elements was not however acknowledged by individuals in the study.

The rating of communication as the most important functional skills by both participants and caregivers aligns closely with the emergent themes of control, autonomy and self-preservation. Caregivers rate hearing amongst the most important of functional skills, which support the isolation of autonomy and self-preservation as central caregiver themes in the HEQ questionnaire. Caregiver responses to the HEQ further represent a greater understanding of the impact of hearing impairment with regards to maintaining autonomy and self-preservation suggesting consistency across these findings.

Conclusion

This chapter provided a description of the prevalence of hearing loss in the current sample. This included a detailed description of the pattern of hearing loss identified, as well as findings of both behavioural and electrophysiological auditory assessments. A review of HHIA findings related to the perceived social and emotional effects of hearing impairment in this MND population followed. The chapter concluded with a review of the qualitative data obtained in the HEQ. This was threefold and firstly included a bried review of MND individual contact with the audiological discipline. This was then followed by a review of MND individual and caregiver perspectives related to the rating of functional abilities. The

final component of this questionnaire explored the common themes that emerged in participant and caregiver responses to the perceived value of auditory diagnosis in MND.

Chapter Five

Discussion

Introduction

This chapter serves to provide an analytical review of the results provided in chapter four with support of the available literature. Results will be discussed in accordance with the primary and secondary objectives outlined in the methodology chapter of this report. The relevant literature will be applied to highlight trends and patterns in the acquired data.

Hearing Loss and MND

The section to follow provides a review of auditory test findings, drawing from specific cases in this 8- sample group. The following discussion is based on reported auditory symptoms, otoscopic observations, immitance and pure tone audiometry patterns, DPAOE findings and neurodiagnostic ABR results. This section will extract discussion points that are case-specific, highlighting and providing possible explanations for commonalities and variations in results across individuals in this 8- sample group.

Reports of tinnitus and difficulty listening in noise are typical complaints in individuals presenting with SNHL of a cochlear nature. These symptoms are further supported by audiological test results indicating cochlear pathology. Hearing-impaired individuals most frequently experience difficulty in the understanding of speech in adverse listening circumstances (Averill et al., 2007). A reduced ability to understand speech in these listening situations is documented as one of the primary and most limiting consequences of hearing impairment, that ultimately impact on the quality of communicative interactions (Nachtegaal et al., 2009).

A number of social and emotional effects may arise as a consequence of these symptoms, including depression, withdrawal and social isolation. In association with hearing loss, many individuals present with complaints of tinnitus, as indicated by individuals in the current study. Tinnitus gives rise to negative emotions in 25% of individuals including increased anxiety and distress and exasperates difficulties listening in noise (Jastreboff, 1999). Difficulty listening in noise creates further problems with communication in social settings (such as restaurants and family gatherings). These hindrances evolve into individual withdrawal from communicative situations, feelings of isolation, embarrassment in moments of misperceived sounds/ words and the need for frequent repetition (Jastreboff, 1999). A total of five individuals with MND (n = 5) complained of tinnitus and difficulty listening in noisy situations. It is ultimately the combined effects of these symptoms that lend themselves to reduced participation and limited engagement in activities of daily living (Jastreboff, 1999). Participants in the current study reported a range of auditory symptoms, however underreported the consequences of these. The limitations caused by underreporting auditory symptoms and effects inevitably reduce the QoL of the affected individual. While it is important to reinforce the wide range of physical (mobility, dysphagia, dysarthria) and emotional (loss of autonomy, dependency) factors that threaten the MND individuals QoL the effects of auditory symptoms such as tinnitus and difficulty listening in noise may further impose communicative and social difficulties inevitably exacerbating the extent of difficulty, often unbeknown to the affected individual.

Hyperacusis, a condition relating to a collapse of loudness tolerance, presents with increasing frequency as a symptom of hearing impairment. This was further reported by MND individuals in the current study. Hyperacusis may be reported by hearing-impaired individuals, however is also documented in normal hearing individuals (Hesse et al., 1999;

Vernon, 2002). This supports complaints of hyperacusis amongst participants in the current study, some of whom presented with hearing impairment and some of whom presented with normal hearing abilities (P2 and P3). The cause of hyperacusis remains unknown, although Vernon (2002) suggests a loss of functioning in the olivocochlear bundle that supplies the efferent innervation to the cochlea and exerts a suppressive effect to incoming sounds (Hesse et al., 1999, Khalfa et al., 1999). The impaired function of these nerves provides an explanation for normal sounds to be perceived as louder than usual. Hesse et al. (1999) isolate the cause of hyperacusis to be a disturbance in central auditory processing with an inhibitory deficit in the auditory pathway, hypothesizing this deficit to be in the region of the cochlea.

Hyperacusis is frequently reported alongside complaints of tinnitus, although tinnitus is recorded to be less severe of the two symptoms (Vernon, 2002). Hyperacusis patients also present with abnormal hair cell hypermobility across a wider frequency range, whereas hypermobility in tinnitus is reported to be isolated to the tinnitus frequency (Hesse et al., 1999). The severity of hyperacusis can be identified by TD and DR audiometric findings, where decreased TD is directly related to the presence of hyperacusis (Vernon, 2002). Four participants in the current study reported hyperacusis of a mild severity as measured on the TD-hyperacusis severity rating (Vernon, 2002). This suggests good correlation between participant reports and auditory testing. The auditory symptom of hyperacusis has marked negative effects on individual QoL and daily functioning reportedly resulting in avoidance of music, cinema, groups and festivities (Vernon, 2002). This is evident in participant complaints of distress and discomfort in typically normal listening environments and/ or previously tolerable listening situations. Avoidance behaviors may ultimately lead to complete social isolation and feelings of generalized fear (Vernon, 2002). In certain instances,

the symptoms of hyperacusis additionally give rise to depression and in some instances the intolerable nature of this gives rise to thoughts of life-ending pursuits (Vernon, 2002). MND individuals place emphasis on maintaining autonomy and preserving self-image thereby shifting focus from loss of physical function to maintaining interactive function. The presence of the above named auditory symptoms may consequently prove to be isolating and have devastating effects on individual self-worth and inclusion in previously pleasurable activities (Vernon, 2002).

Upon visual inspection, participant ears (in the current study) did not present with notable abnormalities. This indicates that structurally, MND has no visible effects on the structure of the outer ear through to the level of the tympanic membrane. Participant 3 presented with soft wax partially occluding the right ear canal as well as a history of recurrent ear discharge and pain in the right ear. The effects of chronic ear infections are reported to increase with age (Tambs, Hoffman, Engdahl, & Borchgrevink, 2004). It is reported that the earlier age of onset of persistent ear infections, the more substantial the effects are on hearing ability in later life. Findings for this participant revealed borderline normal hearing thresholds, although a history of middle ear infection is likely to be associated with the increased air-bone gap observed in pure tone audiometry.

Type A tympanograms supported the presence of normal middle ear functioning. This suggests that no conductive involvement was evident in participants of this study. The presence of a type A tympanogram isolates the site of lesion in hearing impaired ears to the cochlear and retrocochlear region (Hall & Mueller, 1997).

Acoustic reflexes cannot be applied as an isolated diagnostic measure due to the high variability in findings across individuals, hearing loss severities and sites of lesions (Bess & Humes, 2008). As a result, these findings should always be analyzed alongside individual case history, tympanometry, pure tone and speech testing to reach a differential diagnosis (Bess & Humes, 2008). Absent acoustic reflexes were evident for some participants, with these being noticeably absent in the high frequency range only. Absent reflexes at higher frequency levels correspond to the increasing severity of hearing loss in these frequencies. This suggests that higher intensity levels (exceeding the parameters of the equipment) are required to elicit an acoustic reflex, resulting in failure to obtain a reflex recording (Bess & Humes, 2008; Emmanuel, 2009). Where a cochlear hearing loss is indicated, reflexes may be elicited at normal levels of 70-80 dBSL typically up to 50 dBHL. As hearing thresholds increase above this level, the chances of absent and/or elevated reflexes increases, as evident in the findings from the current study (Emmanuel, 2009). An additional consideration is the well known variability in reflex recordings at 4000Hz. At 4000Hz absent or elevated reflexes are relatively common even amongst non-hearing impaired ears, further reinforcing that caution needs to be applied in interpreting acoustic reflexes as an isolated measure (Bess & Humes, 2008).

The first of the secondary objectives outlined in this study served to provide a description of the hearing loss trends of individuals diagnosed with MND. Two participants (P2 and P3) presented with normal hearing thresholds bilaterally, while the remaining six presented with varying extents of hearing loss isolated to the high frequencies. P3 presented with a large air-bone gap in the right ear despite borderline normal thresholds being recorded. Participant 3 was the only participant in the total sample to have a diagnosis of bulbar MND, also presenting with a soft wax occlusion in the right ear alongside a history of ear discharge.

Any of these factors in isolation and/or combination may be associated with the large air-bone gap.

In a study that investigated the differences in auditory thresholds ranging from 500Hz - 4000Hz before and after wax removal, it was found that hearing improved by an average of 5 dB across the frequency range analyzed (Sharp, Wilson, Ross, & Barr-Hamilton, 1990). This change in hearing ability, although small, may negate the need for amplification for an individual that is a borderline hearing aid candidate (Sharp et al., 1990). When a 5 dB improvement across the frequency range of P3's right hearing thresholds is applied the extent of the air-bone gap lessens, although remains relatively large with a 15-20dB gap between air and bone thresholds across the low – mid frequency range. This suggests that wax occlusion may not be the only factor contributing to this air-bone gap. Furthermore, P3 reported and demonstrated only mild consequences of recurrent infrequent infections during auditory testing thereby validating the borderline normal hearing thresholds recorded for the right ear. A history of ear infections has damaging effects on hearing levels later in life and typically has more harmful effects on QoL amongst adult subjects in contrast to younger subjects (Tambs, 2004). Management of this is therefore of upmost importance.

Interestingly, P3 was the only participant in the sample to present with a diagnosis of bulbar MND, which may serve as an additional factor contributing to the increased air-bone gap. Bulbar MND is typically characterized by motor neuron degeneration in the regions of the cerebral cortex, brainstem, spinal cord and the pyramidal tracts. These regions typically involve cranial nerves IX (glossopharyngeal), X (vagus) and XII (hypoglossal) (Snell, 2001). Further review into the innervations of the glossopharyngeal nerve reveals supply to the middle ear region. While the glossopharyngeal nerve carries largely sensory functions, a

visceral motor component exists, exiting the inferior ganglion emerging as the tympanic nerve, serving the tympanic cavity of the middle ear (Snell, 2001). The involvement of this nerves' innervations to the middle ear region as well as it's relation to bulbar neuron degeneration highlights the need for further investigation into it's involvement in the onset of conductive hearing impairment in individuals diagnosed with bulbar MND.

Conductive hearing loss in MND may further be attributed to the paralysis of the tensor and levator veli palatini muscles. These muscles play a crucial role in controlling the mechanical properties of the Eustachian tube. Paralysis of these muscles, on account of motor neuron degeneration linked to bulbar symptoms, prevents the active dilation of the Eustachian tube resulting in increased lumen in these regions (Ghadiali et al., 2003). The outcome of such paralysis leads to the development of negative middle ear pressure, ultimately increasing individual risk for developing otitis media with discharge (Ghadiali et al., 2003). This hypothesis supports the presence of a large air-bone gap in P3's hearing thresholds and is supported when viewed in conjunction with reports of 'infrequent' recurrent ear infection presenting since MND onset. Additionally, it is viable that thresholds remained within the normal limits, since the most recent occurance of discharge was reported to have occurred over 8 months prior to the test date. Therefore, since P3 did not display signs of active infection (discharge, pain, lack of tympanic membrane clarity) at the time of testing, test results reflected normal auditory abilities. Tympanometry measures were further indicative of normal functioning in the region of the middle ear, although this may be the result of active infection not being present at the time of testing, thereby having no negative effect on the middle ear pressure.

Six participants presented with bilateral, symmetrical sensorineural hearing losses

isolated to the high frequency range. These audiometric findings were paired alongside subjective concerns linked to listening experiences in noisy environments and suggestions of recruitment supporting the presence of sensorineual hearing impairment.

The speech discrimination scores of these participants typically indicated an improvement as presentation levels increased, although rollover was recorded in some instances (P1 and P7 L; P5 and P6 R). The presence of rollover however remained <20% supporting a hearing loss of a sensorineural nature linked to a cochlear site of lesion. For the rest of these participants, very slight improvements in Sd abilities were noted between the SRT + 25dB and TD – 10 dB. These participants (P2, P3, P4, P6, P8) presented with hearing impairment isolated only to the high frequency regions. The majority of speech sounds fall within the 2000-4000Hz range. For participants in this study, this was a range where hearing thresholds typically fell within a normal levels. As a result, the extent of the impairment at these speech frequencies was not significant enough to cause greater growth in Sd scores as most individuals scored within or near normal levels at SRT + 25 dB. One may expect that alongside progressive deterioration of hearing abilities encompassing the mid-high frequency range, Sd scores, particularly at SRT+25 dB level may deteriorate. Affected individuals would then rely on higher intensity levels to achieve success in sound discrimination abilities.

Discomfort levels of > 85.1 dB are suggestive of normal loudness tolerance. Discomfort levels between 65.1 to 85 dB is suggestive of mild hyperacusis, 45.1 to 65 dB is suggestive of moderate hyperacusis, whilst 25.1 to 45 dB and <25 dB is suggestive of moderate-severe and severe levels of hyperacusis respectively (Vernon, 2002). For all participants in this study with complaints of hyperacusis (n = 4), discomfort scores between 65.1 dB and 85 dB were obtained, suggestive of mild hyperacusis. Reduced TD scores

resulted in a smaller DR (<60dB) amongst certain participants (P5 and P7 bilaterally; P6 right) and is therefore suggestive of recruitment, a phenomenon closely linked to a lack of outer hair cell modulation in inner ear diseases of the cochlea (Hesse et al., 1999). These findings were consistent with the remaining test battery isolating the site of lesion to the cochlear region.

The relation between auditory thresholds and DPOAE thresholds indicate significant correlations, with the majority of cases who fail to meet a SNR of > 6dB presenting with a hearing loss (Gorga et al., 2003). Similarly, in the current study, participants failing to meet the 6 dB SNR criteria, presented with hearing impairment at the corresponding frequencies suggesting impaired function of the cochlear outer hair cells in the high frequencies. Decreased DP levels are reported to be proof of diminished sensitivity and tuning of the cochlear amplifier (Hesse et al., 1999). DPOAEs were successfully recorded with a slope-like decline in outer hair cell function mimicking the decline in auditory function identified through pure tone audiometry testing. This supports impaired outer hair cell function in the region of the high frequencies and provides further confirmation of a cochlear site of pathology (Bartnik et al., 2009). It was further found that DPOAE findings vary depending on the symptoms of tinnitus and hyperacusis (Hesse et al., 1999). Where tinnitus occurs as an isolated symptom, impaired function of the hair cells is indicated at the specific tinnitus frequency, whereas hyperacusis as a symptom affects outer hair cell mobility across a wider frequency range arising as a result of cortical disturbances in auditory processing (Hesse et al., 1999). This trend of a more vastly affected frequency range linked to hyperacusis was not noted in the current study. However, the mild level of hyperacusis recorded and the small sample size may have limited the identification of such trends. While the values in this study demonstrate a consistent trend in test findings supporting cochlear impairment, consideration

of the limited sample size should be applied to avoid over-interpretation and generalization of findings.

Participants in the current study presented with neurodiagnostic ABR findings consistent with individuals without impairment at the level of the brainstem. IAWLD for wave V fell below 0.4 msec for all participants, eliminating any indication of retrocochlear lesions. IAWLD V is reported to have clinical significance, suggestive of retrocochlear lesion, only when exceeding 0.4 msec (Don & Kwong, 2002; Musiek et al., 1994). Reliance on a smaller IAWLD V differences (e.g. 0.25 or 0.3msec) leads to an excessive proportion of false-positive ABR outcomes and hence lesser values cannot be considered clinically significant (Don & Kwong, 2001; Hall & Mueller, 1997).

IPLs were recorded between waves I-V, I-III and III-V. The presentation of ABR waves in the presence of a cochlear impairment typically reveal prolongations of wave I-III IPLs, normal or slightly shorter wave I-V IPLs and shorter wave III-V IPLs as a consequence of the prolongation of wave III-V. Findings in the current study were consistent with this, further validating the cochlea as the site of lesion. IPLs for wave I-V fell within the target range for six participants, while shorter wave I-V IPLs were recorded for two female participants (P2; P5). The reduction in IPLs for these two participants may be attributed to gender differences. It is reported that females present with shorter latencies than those outlined by the norms. Typically, the IPLs of females are reported as 0.1 – 0.2 msec shorter than the norms applied in this protocol and therefore remain within a normative region not suggestive of retrocochlear lesion (Don & Kwong, 2002). Prolonged wave I-III IPLs were recorded for two thirds of participants in the study, particularly where high frequency hearing loss was more severe. As a consequence of wave I-III prolongations, latencies were shorter

for all participants between wave III-V. Musiek et al. (1994) support this with the suggestion that even in normal hearing subjects the IPL's for wave I-III may be slightly longer than III-V. ABR recordings thereby provided further validation for a cochlear site of lesion.

Cochlear hearing loss and the effects this has on wave V amplitude are difficult to predict (Don & Kwong, 2002). While it is anticipated that the loss of activity as a result of cochlear impairment should affect ABR amplitude, it is difficult to predict the extent due to various factors, such as synchronization and phase cancellation (Don & Kwong, 2002). None of the AWLs recorded for participants exceeded the normative range. Wave I latencies for all participants appeared shorter than the guidelines previously established, which may be further explained by the choice of polarity applied to ABR recordings in this study. The use of rarefaction polarity typically reveals latencies that are marginally shorter than the norms provided (Don & Kwong, 2002). This latency reduction usually does not exceed 0.1-0.2 msec from the standard range described above. Don and Kwong (2002), further report that latencies for females are typically shorter than those of male individuals, providing possible validation for the slightly shorter wave III latencies observed in female participants (P2) in this study.

Summary of Audiometric Findings

Seventy five percent of individuals in this study presented with hearing impairment, the extent and severity of which, varied. Two participants presented with normal hearing thresholds (P2, P3), one of which presented with borderline normal thresholds and a large air-bone gap (P3). This participant, with bulbar onset MND, also presented with a history of *'infrequent'* infection and discharge. Six participants (P1, P4, P5, P6, P7, P8) presented with a sloping SNHL isolated to the high frequency range. This was confirmed by the DPOAE

results that supported the presence of outer hair cell cochlear dysfunction and the findings of the neurodiagnostic ABR tests, providing further support of cochlear site of lesion. Immitance audiometry suggested normal middle ear functioning.

Review of all test results in combination support the presence of a high frequency hearing loss isolated to the region of the cochlea. Caution is however required in isolating the etiology of this hearing loss as a definitive factor related to MND. The nature of the loss identified in this study corresponds to the pattern of presbycusis – a hearing loss arising as a result of the natural aging process (Dalton et al., 2003). This is of particular relevance since the onset of MND (fourth to sixth decades of life), coincides with the onset of age related hearing impairment (48-87 years) (Dalton et al., 2003).

Presbycusis is characterized by loss or death of the hair cells of the inner ear and/or atrophy of the auditory nerve in the basal region of the cochlea, presenting as a SNHL (Weinstein, 2002). The onset is typically around middle age with a gradually progressive nature. Individuals with an age related hearing loss express a number of primary complaints, the foremost of which relates to difficulties discriminating speech in noise (Weinstein, 2002). This corresponds with the reports of P1, P4. P5, P7 and P8. Presbycusis may or may not co-occur with tinnitus, however this symptom was reported by five participants in the current study (P1, P4, P5, P7 and P8). Tinnitus and hyperacusis frequently co-occur (Vesterager, 1997). Hyperacusis was reported to co-occur with tinnitus by four participants in the current study. The resultant increase in loudness perception reported amongst four participants, further supports the cochlea as the primary site of lesion for hearing loss in MND-participants in the current study. Presbycusis presents as a sloping loss where hearing thresholds are typically normal between 250Hz – 2000Hz, thus affecting the high frequencies first. These

findings correspond to the pattern of hearing loss noted in P1, P4, P5, P6, P7 and P8 in the current study.

It is evident from the above description of presbycusis and its' related audiometric features, that the six participants in the current study who presented with bilateral sensorineural impairment presented with remarkable similarity in presentation to that of individuals with presbycusis. It is also relevant to note that while P2 did not present with hearing impairment, she was the youngest (49.6 years old) of all the participants. A 7.7 year age difference existed between P2 and the next participant when reviewed in increasing chronological order. The onset of presbycutic audiometric signs may not yet be subjectively or objectively apparent at P2's age, as she is at the lower end of the age spectrum for presbycutic onset. It is furthermore documented that the hearing levels of males are typically poorer and difficulties present earlier than those of females, providing additional support for the absence of hearing loss in P2 at the time of the study, based on her gender and age (Weinstein, 2004). In conclusion, although hearing loss has been identified in the majority of the sample population, it is not possible to firmly conclude that the presence of hearing loss identified in this study exists primarily and definitively as a symptom of MND. This is particularly due to the limited sample in this study.

Although a single retrospective study (Maier et al., 2009) provides initial evidence of the presence of hearing loss in individuals with MND cannot be reliably generalized due to the limited sample size. The current study therefore served to add to the limited body of available research relating to hearing loss and MND. In addition, the current study highlights the importance of expanding the current patient management protocols to include

audiological assessment as the consequences of hearing loss may increase participation restrictions for individuals with MND.

Hearing Handicap Inventory for Adults (HHIA)

The HHIA served to provide insight into the subjective emotional and social auditory experiences of individuals in daily life situations - the negative impact of which is well documented (Dalton et al., 2002). The severity of hearing loss is positively associated with decreased function at a psychosocial level, which has greater implications in the MND population due to the added physical deterioration further compounding ADL and QoL. The HHIA scale revealed wide variation in scores across participants in the current study, ranging from no reported handicap to moderate reported handicap. It is important to bear in mind that all HHIA scores within this sample fall within the bottom 40% of the inventory scale. Caution must therefore be exercised in the use of these findings as a reflection of the general MND population due to the limited sample size and thus the limited power of generalizability.

It is interesting to note that while two participants initially reported some extent of hearing impairment via initial case history questionnaire, five presented hearing impairment as revealed through audiometric testing. Four participants also reported an auditory handicap on the HHIA. This demonstrates the discrepancy between initial reports of hearing impairment and audiometric findings; and between audiometric findings and perceived social and emotional handicap. These discrepancies are supported by the literature, which reveal that self- report measures typically underestimate the prevalence of hearing loss, resulting in the underreporting of hearing related difficulties (Dalton et al., 2003; Hallberg et al., 2008; Newman et al., 1990). Newman et al. (1990) consequently report that self-report measures such as the HHIA are insufficient in describing an individuals reaction to their hearing loss

alone. It therefore needs to be interpreted alongside pure tone and suprathreshold speech recognition test results for an integrated, holistic understanding of auditory handicap. It has been found that only 22% of individuals with a mild hearing loss, and 56% of individuals with a moderate-severe hearing loss, report a hearing handicap (Dalton et al., 2003). In contrast, 59% and 80% of individuals with mild and moderate-severe hearing losses respectively, isolated communication difficulties as a concern using the same self-report format. This suggests that individuals view communication difficulties to be of greater importance than auditory abilities, a notion further supported by individual opinions expressed in the HEQ for participants in the current study. Only two participants in the current study reported moderate difficulties related to hearing loss and socio-emotional factors. Nachtegaal et al. (2009) report that psychosocial health related to hearing impairment is reduced amongst older adults aged 65 years and older. The cut off age for this study was set at 66 years of age, thus it may be inferred that the low acknowledgement of hearing loss amongst the participants in this study is possibly a result of greater success at a psychosocial level than that of older adults.

Cross-sectional studies have revealed emotional disorders linked to anxiety and depression paired with MND diagnosis (Goldstein & Leigh, 1999). This suggests that depression and low self-esteem have a direct relationship to the effects of MND on everyday functioning and QoL (Goldstein & Leigh, 1999). It may be additionally important to consider that the participants in the current study perceive emotional handicap to be more closely related to the diagnosis, progression and severity of neuro-degeneration rather than auditory impairment. The latter suggests that individuals associate socio-emotional handicap as a direct result of MND, but fail to consider secondary or external impairments such as hearing ability. This results and validates the lower scores allocated to emotional hearing

handicap ratings in the HHIA.

It is further postulated that self-deception regarding hearing loss is often adopted as a means of protecting individual identity and image (Hallberg, 1999). This is particularly true in instances where there is a discrepancy between ones reality and desired self-image. Individuals with MND deal with a steady and gradual loss of function placing emphasis on the need to constantly redefine self-image to match reality (McLeod & Clarke, 2007). In instances where individuals deal with an ongoing need for readjustment, denial and failure to acknowledge these difficulties occurs. Individuals hence consciously or subconsciously underestimate the negative effects of hearing loss despite the external changes in communicative behaviors that arise as a direct consequence (Hallberg et al., 2008).

An additional explanation for HHIA responses revealing minimal socio-emotional concerns (with exception to P5 and P6) may relate to the extent of hearing impairment. Hearing loss across the entire sample was isolated to the high frequencies between 6000-8000Hz. It is possible that the frequency range affected by hearing loss is not yet broad enough to impose a more significant impact on the social and emotional functioning of the individual.

Hearing Experience Questionnaire (HEQ)

Contact with Speech Pathology and/or Audiology.

At the time of the study, none of the participants had been referred for auditory testing, educated on auditory impairment or counseled regarding the effects of auditory impairment on QoL. A study exploring the perspectives of services for MND by individuals with MND and their caregivers, revealed that while individuals praised the efforts made by members of

the management teams, deficiencies in professional knowledge linked to MND were highlighted (Brown, Lattimer & Tudball, 2006). In addition to this, the organization of secondary health services, counseling and emotional support to families and individuals with MND emerged as limitations. Brown et al. (2006) conclude that emphasizing the quality of inter-professional and multi-agency co-operation is an important means of promoting and enhancing the quality of care given to individuals with MND. Furthermore, knowledgeable care teams who consider primary and secondary consequences of MND additionally contribute to enhancing the support, management and overall experience of living with MND (Brown et al., 2006; Ward et al., 2003).

Findings of the current study cannot conclusively isolate hearing impairment to MND despite the presentation of hearing impairment in the majority of participants. As the age of MND onset, correspond with the onset of presbycusis, individuals with MND may also present with hearing loss. It is therefore imperative that these individuals' hearing should be monitored throughout the course of the disease. It is postulated that this will serve to reduce the damaging effects of hearing loss on individual QoL and ultimately the willingness to pursue life-prolonging measures (Ward et al., 2003).

Functional Rating Scale.

Academic and medical perspectives on MND focus mostly on the physical aspects of MND, while psychosocial aspects are accorded secondary importance (McLeod & Clarke, 2007). Individual and caregiver responses to the Functional Rating Scale applied in this study highlight this aspect. More attention needs to be paid to the wider spectrum of the needs of individuals with MND. This can be achieved by implementing multi-disciplinary approaches, with emphasis on enhancing overall QoL of the affected individual aside from

the physical aspects of the disease.

Over 80% of individuals with MND experience impairment in verbal communication (Leigh et al., 2003), an ability that was rated by all participants and caregivers in the current study as the most important ability to preserve. Individuals with MND frequently shift conversations from health-related symptoms to social and relationship issues. This highlights the importance of preserving communication and modifying it according to the individuals' level of communicative ability (Hardiman et al., 2004). AAC strategies are used to facilitate communication and address the loss of expressive verbal communication and are readily acknowledged within the MND population (Leigh et al., 2003). However, considerations for the effects of auditory impairment and the associated devices to enhance listening experience are not viewed as equally important. The progression of degeneration gives rise to an increased loss of function and a steady decline in individual day-to-day activity and participation. Hearing loss, be it a direct consequence of MND or not directly disease-related, poses significant threats to the overall QoL. When reviewing the effects of disease related symptoms alongside the additional loss of function, such as hearing loss, the increasingly detrimental effects of failing to manage any manageable symptoms on individual QoL become apparent. This in turn affects the individuals' experience of the disease thereby impacting their ability to manage it (Hardiman et al., 2004; McLeod & Clarke, 2007). The fact that individuals with MND fail to acknowledge the importance of audition as a key requirement for effective communicative exchange represents a gap in the awareness and management of secondary symptoms in MND (Hardiman et al., 2004). In contrast, the majority of caregivers in the current study, acknowledged the importance of hearing as a functional skill central to maintaining communicative success. Studies consistently report that significant others are typically more objective in acknowledging the negative effects of

hearing impairment and the importance of this on the communicative process (Dalton et al., 2003; Hallam et al., 2008; Starck & Hickson, 2004). Where caregivers are reported to be more realistic about the impact of hearing impairment on communicative functioning it highlights the necessity for caregivers to be actively involved in guiding the process of pursuing audiological consultation. The burdens hearing loss places on communicative and social success for both the affected individual as well as the caregiver are well documented (Boi et al., 2011; Paolo et al., 2008). In line with the latter, the reported benefits of early detection of auditory difficulties, management and rehabilitation are known to enhance the QoL for both caregiver and individual, enhancing the quality of socialization, the extent of social inclusion and reducing the social demands carried by the caregiver (Boi et al., 2011; Paolo et al., 2008). Where MND is a disease with devastating and life-threatening consequences, affected individuals understandably place focus on life sustaining needs such as respiratory function and swallowing. Caregivers are consequently better able to objectively identify secondary features such as hearing loss and the psychosocial effects these have on individual QoLsuggesting that awareness of auditory symptoms and management counseling may be more effective presented alongside caregivers of affected individuals, rather than MND individuals alone,(Hallam et al., 2008),

The importance of eating for nutrition to maintain nutritional wellbeing was highlighted by participants in the current study. As individuals' associate deglutition very closely with social success, there is a positive relationship between swallowing success and psychosocial functioning (Ekberg et al., 2002). Participants in the current study alluded to the fact that incompetency in eating is harmful to their individual QoL, and impact on their ability to maintain individual inclusion in social interactions, which are frequently set around a dining experience. This is supported by the findings of Ekberg et al. (2002) who reported

that over 50% of patients with MND reported eating less throughout the course of the day and found eating to be a displeasurable experience, while 44% of patients reported weight loss within the initial 12 months of symptom presentation.

Arm and leg movement were rated least important in the current study, amongst both individuals with MND and their caregivers. This suggests that individuals with neurological disease are more readily able to adapt to physical decline by lowering their expectations of physical ability and redirecting this to other areas of life (such as cultural, social and interactive domains). These reports are consistent with Foley et al. (2007) who suggest that the coping strategies implemented by the affected individual reinforces the need for health professionals to consider individuals with MND beyond their physical disability and in the context of their social and psychological systems.

Communication, hearing, swallowing and vision all contribute to the maintenance of human contact and closeness. Caregivers in the current study viewed hearing as very important. These findings indicate that both individuals with MND and their caregivers place greater focus on functional abilities (e.g. communication, hearing, swallowing and vision) that would have a direct impact on inclusion and involvement in daily life rather than those abilities related to mobility. Despite increasing disability, individuals with MND seek to maintain identity, self-worth and respect, of which are achieved through socio-emotional accomplishment (Foley et al., 2007). Although in this study audition is not equally important for individuals with MND versus caregivers, the shared notion remains that affected individuals seek to be purposeful, both existentially and socially to maintain psychological wellbeing. In order to achieve this, the primary focus is redirected towards the social and emotional aspects of disease rather than those linked to physical ability (Foley et al., 2007).

Identifying Importance of Auditory Diagnosis.

The initial case history interview revealed that the majority of participants in the current study did not report experiencing hearing difficulties, despite auditory testing revealing that they present with hearing loss. This finding is supported by Rawoon and Kiehl (2009) who found that 66.6% of participants in their study had a hearing loss, in contradiction to self-reports suggesting normal hearing. The effects of hearing impairment result in distorted communication, stigmatization and social isolation leading to emotional disturbances as readjustment to the impairment of function is realized (Tambs, 2004). These consequences appear to be highly underestimated by participants in the current study. It has been found that 20.7% of adults with hearing loss seek rehabilitation in the form of hearing aids (Popelka et al., 1998; Rawool & Kiehl, 2009). This indicates that only a small percentage of hearing impaired individuals pursue rehabilitative support, despite research showing that a reduction in depressive symptoms is reported in affected individuals once hearing aid fitting occurs (Tambs, 2004). The decline of these depressive symptoms following the fitting of a hearing aid further highlights the benefits of hearing loss diagnosis and management in individuals with MND.

In line with the reported psychosocial benefits of hearing aid use, it is relevant to consider the fine motor skills and manual dexterity required for hearing aid manipulation and use. When considered alongside the upper motor extremity difficulties experienced by the MND individual the handling obstacles become clearly apparent. Successful hearing aid usage is highly reliant on non-auditory factors including cognition and manual dexterity. The ability to manipulate a hearing aid is required in tasks such as inserting and removing the aids, operating various controls, adjusting volume controls and changing batteries to name but a few. Singh (2009) reports that evidence exists supporting the extent to which manual

dexterity predicts hearing aid manipulation and use. Several studies support an increase in hearing aid use when individuals are able to better handle the hearing device and a corresponding decline in hearing aid use in those individuals less able to manipulate the devices (Kumar, Hickey & Shaw, 2000; Meister, Lausberg, Kiessling, von Wedel & Walger, 2002; Singh, 2009; Wilson & Stephens, 2003). Difficulties with device manipulation poses direct threats to hearing aid use amongst the MND population and hence cannot go without consideration as a possible factor reducing recommendations for amplification amongst MND individuals. In consideration of the MND population it becomes evident that the combined effects of diminished hand function and the fine features of hearing aid devices pose a potential crisis for handling difficulties. This consequently translates into limited use and benefit from the hearing aid device. These difficulties in part, provide an explanation for the low hearing aid adoption rates even amongst the normal aging population (Singh, 2009). These low rates of hearing aid use may hence be further exacerbated amongst individuals experiencing limitations in upper extremity function as a result of progressive motor cell death, as in the instance of MND. The role of the caregiver in assisting individuals lacking the manual dexterity to manipulate and handle hearing aid devices thus requires consideration.

Paulo, Teixeira, Jotz, de Barba & Bergman (2008) report on the QoL of caregivers of individuals with auditory impairment. The role of the caregiver in dealing with progressive loss of function in MND leads to less involvement in social activity, ability to problem solve to overcome barriers in daily life and endure ongoing stressful moments during their adjustment to new life routine. When dealing with hearing impairment, the role of the caregiver is often extended to that of ‘interpreter’, aiding the hearing impaired individual in situations where speech recognition is necessary. Such situations may include hearing on the telephone, in doctors appointments, social engagements, making the caregiver liable for the

participation of the hearing impaired individual in social activities and daily routine (Paulo et al., 2008). Results indicated that the QoL of the caregiver was significantly hindered particularly in the social domain where social relations, social support and social activities were explored. In a comparison of caregiver QoL between caregivers who care for elderly patients with and without hearing aids, caregivers of patients without hearing aids scored significantly lower in social and psychological domains (Paulo et al., 2008). These findings may be attributed to the fact that hearing aid use introduces improved speech recognition making independent communication and ability to engage in social activities possible (Paulo et al., 2008). This reduces the level of isolation caused by hearing impairment and lessens the load placed on the caregiver, affording him/ her increased opportunities in social life and enhancing QoL for both the caregiver and the affected individuals (Paulo et al., 2008). The consequent benefits of caregivers aiding individuals without the manual dexterity for hearing aid handling are thereby reported to outweigh the social and psychological burdens (Paulo et al., 2008). In a disease such as MND where caregivers adopt the responsibilities for caring for the affected individual across multiple domains such as self-care (brushing teeth, bathing), mobility (directing wheelchairs, driving) and day-to-day activities, the added responsibility of handling hearing devices appear minimal when the outcome of this promotes alleviation of one aspect known to add to the social burdens placed on both the caregiver and the affected individual.

Despite the benefits of amplification, the underreporting of hearing loss is prominent (Tambs, 2004) and typically arises as a result of two factors. The first of these may be a result of the individual being unaware of the loss. The severity of communicative difficulties brought about by hearing loss vary according to the severity of the hearing loss, the associated symptoms and the communicative strategies unknowingly implemented to

compensate for this (Hallam et al., 2008). Amongst all hearing impaired participants in the current study, hearing loss was isolated to 6000Hz – 8000Hz. A loss in these frequencies tends to impact less on speech recognition abilities in quiet settings. The impact increases though in noisier listening environments and/or as the loss progresses. Hearing loss in the high frequency range may affect individual ability to detect and discriminate certain consonant sounds and individuals may rely on combined clues from patterns of speech sounds to understand what is said. The extent of individual reliance on combined cues such as visual and contextual clues is largely dependent on the severity of hearing loss in the frequency range and individual compensation. The majority of the participants in the current study reported that listening experiences in noise proved to be a greater challenge for them, a complaint that is consistent with the isolated high frequency impairment identified across six individuals in the study. It is therefore a likely possibility that hearing loss failed to be acknowledged by individuals in the current study as a result of the mild effects of the loss and limited frequency range of speech sounds affected, at the time of testing.

As the consequences of motor neuron degeneration extend from a physical level, emotional and social consequences continue to arise. Underreporting of hearing loss may also be linked to denial of auditory abilities and declining communicative function (Hallam et al., 2008; Hallberg, 2008; Rawool & Kiehl, 2009). Denial is a typical response that protects individuals from stress-provoking situations and serves as an adaptive means to prevent social stigma, and preserve mental integrity and capability (Rawool & Kiehl. 2009). Hallam et al. (2008) support denial as a defense mechanism against accepting hearing loss and suggests that hearing impaired individuals cope with loss through avoiding social situations. Avoidance or minimizing the effects of hearing loss are reported to be more common amongst males than females and occurs as a means of protecting self-image (Garstecki &

Erler, 1999; Hallam et al., 2008; Hallberg et al., 2008). This leads to individuals pretending to hear, guessing what was said and/or avoiding interactions. Only 25% of the participants in this study acknowledged experiencing hearing difficulties prior to testing, both of who were female. Interestingly, the male participants in the study who presented with a hearing loss denied the presence of hearing difficulties. This demonstrates the profound psychological burden of MND in light of the ongoing deterioration of function and the associated experiences of denial linked to new diagnosis. Since hearing loss is invisible it is simpler to deny than the loss of limb function. It is therefore postulated that individuals with MND may consciously or unconsciously fail to acknowledge hearing difficulties for fear of the negative consequences and the added emotional burden of loss of function beyond the known loss brought about by MND.

The majority of participants in the current study reported that the knowledge of auditory impairment would be of no importance with a range of negative perceptions linked to burden, vulnerability and fear emerging from responses in the current study. Participants approach a diagnosis of hearing loss as an additional negative, untreatable effect. This frame of thought may not be surprising in view of the diagnosis of MND and its limited rehabilitative opportunities beyond symptom management. It is however suggestive of the need for more active involvement of audiologists in the MDT. This role should include patient education, diagnosis, management and counseling as a means of enhancing patient QoL. It is postulated individuals who are psychologically isolated and do not believe their symptoms can be helped, will not complain profusely to health professionals in search of alleviation of concerns, but rather become fatalistic about their condition (Ekberg, 2002). Other professionals' lacking the knowledge and understanding of the profound social and psychological effects of hearing impairment, may further negatively impact the early

diagnosis and management of hearing loss. Raising health professionals' awareness of the detrimental effects of hearing loss on the QoL of the individual with MND, may result in a marked decline in the extent of psychological and social damage that may otherwise arise. Participants in the current study voluntarily remained at the test site for an average of 45 minutes with discussions based on individual auditory symptoms and experiences. This is further testament to the dire need for education, counseling and support related to communication and hearing abilities expressed by these individuals and their caregivers.

The acceptance of hearing loss is directly related to the severity of symptoms (Hogg, Goldtein, & Leigh, 1994). These symptoms are often more recognizable to caregivers and surrounding communicative partners, than the hearing impaired individual himself (Hogg et al., 1994). This was evident in the current study, where the majority of caregivers identified auditory diagnosis to be of much greater importance when compared to the individuals with MND themselves with emergent positive perceptions of auditory diagnosis being linked to autonomy, control and self-preservation. While the severity of functional impairment has been proven to be harmful to psychological wellbeing in neurodegenerative patients, failure to manage symptoms that are to varying extents manageable, further aggravate harmful consequences. The foremost of these consequences is the amplified the levels of distress and emotional anxiety faced by the affected individual. Alongside a gradual progression of hearing impairment, functional and social communicative abilities gradually prove more challenging, leading to experiences of distress and isolation. When failing to address these symptoms, in addition to the less manageable symptoms of MND, the psychosocial disadvantages are likely to increase. Thus, the ability to manage the presentation of any symptom, whether it is related or unrelated to the disease itself serves to enhance and promote better QoL for the individual.

It has been found that patients with terminal illness report improved coping resources, closer interpersonal relationships and newly defined life priorities as a result of counseling and understanding symptoms (Averill et al., 2007). In a study conducted by Rawool and Kiehl (2009) it was found that counseling plays an important role in the acceptance of hearing loss. Counseling allows for the exploration of the emotional impact of a diagnosis of hearing impairment and equips the individual with the necessary coping and compensatory strategies as well as amplification necessary to enhance auditory performance. Counseling for hearing loss when viewed in conjunction with the range of difficulties faced by the MND individual, is essential as a means of educating and empowering individuals with knowledge of the risks and rehabilitatory strategies available to facilitate the difficulties experienced (Hallam et al., 2008). Equipping the individual with this support, knowledge and resources promote improved communicative behaviours which may be implemented by both the caregiver and the hearing impaired individual (Hallberg, 1999). The latter serves to limit the demise of interpersonal relationships on account of hearing impairment guiding the process of mutual acknowledgment and understanding of the associated burdens faced by all individuals affected by the hearing impairment. This facilitates a recreation of respect for dignity, autonomy and capabilities of the affected individual, while alleviating some burden on both communicative partners (Hallam et al., 2008). This is particularly relevant where the deterioration of speech ability in MND brings along a different set of obstacles detrimental to the individuals autonomy, involvement and communicative inclusion. It has also been reported by Rawool and Kiehl (2009) that 20% of participants in their study who initially accepted their diagnosis, reverted to their original state of denial of the hearing loss one month after counseling. This further emphasizes the importance of including the audiologist as a permanent member of the multi-disciplinary management team, with their role including

the ongoing provision of counseling and support to facilitate individual success at a social level of engagement.

Activities of daily living relate to global functioning activities, including bed-chair mobility or short distance mobility to reach a toilet (UNESCAP, 2008). Although hearing loss cannot be isolated as a direct cause of individual failure to accomplish these goals, impaired hearing ability is firmly acknowledged as an additional element exasperating the levels of difficulty experienced (Dalton et al., 2003). This supports the notion that hearing loss substantially enhances the challenges faced by the MND individual when paired alongside general functional decline and the frailty that accompanies disease (Dalton et al., 2003). It is postulated that as the life expectancy of individuals with MND increases (through medical advances and symptom management), a greater prevalence of hearing impairment will be documented.

Individual differences such as personality and methods of coping are potentially critical for understanding the high variability in disease course (Averill et al., 2007; UNESCAP, 2008). Willingness to remain engaged in daily life, motivation to explore compensatory techniques, devices that accommodate loss of function, positive attitudes and an internal locus of control form essential traits that contribute to living successfully with MND (Averill et al., 2007). It has been found that individuals with hearing impairment wait for 5 – 15 years before seeking professional help to improve hearing difficulties (Rawool & Kiehl. 2009). This further highlights the lengthy process involved in acknowledging hearing loss and suggests that this process often exceeds the lifespan of a MND individual. It is therefore of upmost importance that MND management plans emphasize the importance of optimizing functional abilities (such as hearing) that are to a certain extent manageable and

that this is done with time urgency. This invites positive improvements in QoL limiting the extent of communicative difficulties faced by the individual with MND. It further facilitates lessening the social burdens faced by the caregiver who ultimately adopts the role of 'translator' to guide social success (Boi et al., 2011; Paulo et al., 2008).

Studies indicate that significant others tend to be more objective with regards to acknowledging and reporting the negative impact of hearing loss on communication, suggesting that the role of pursuing audiological consultation should rest more heavily caregivers (Dalton et al., 2003; Hallam et al., 2008; Starck & Hickson, 2004). Evidence of interpersonal stress is reported in 53% of marriages where one individual is affected by hearing loss (Hallam et al., 2008). The impact of hearing impairment on the significant other has been observed to be more severe than that of the individual with the hearing loss. It is reported that caregivers often participate less in social activities and are required to continually solve problems and adjust to changes in their life routine as a result of their partners hearing loss (Boi et al., 2011; Hallam et al., 2008; Paolo et al., 2008). Caregivers hence become liable for including the affected individual in social activities limiting their own communicative opportunities (Paulo et al., 2008). Pursuing auditory diagnosis benefits both members of the couple as it improves individual QoL and alleviates the detrimental social consequences of hearing loss (Paulo et al., 2008). The acknowledgement of the benefits of the diagnosis and management of hearing loss supports the findings of this study whereby caregivers were generally more supportive than the participants with MND to pursue auditory diagnosis.

The impact of hearing loss on close relationships emphasizes the need for professionals to provide family-based support to facilitate adjustment in contrast to isolated

patient-centered support (Hallberg, 1999). Changes in previous communicative patterns and domestic, social and leisure activities require modification in the presence of hearing loss for both the affected individual and caregiver. The role of the caregiver in family-based support is particularly relevant since it is caregivers who are more readily able to identify auditory difficulties in the affected individual and encourage the process of auditory diagnosis. The mutual willingness of the individual with hearing loss and family members to engage with the problem is paramount (Hallam et al., 2008). This is of particular importance since the stressors experienced by the different parties are often vastly different, creating the opportunity for misunderstanding, conflict and blame. Hogan (2001) report that where a mutual willingness to accept, acknowledge and manage the presence of hearing loss is not shared, 47.3% of relationships between caregivers and hearing-impaired individuals, suffer.

The management of auditory impairment through aural rehabilitation strategies has contributed to improved individual success in existential and social contexts, ultimately enriching individual QoL (Starck & Hickson, 2004). The psychological benefits of managing new symptoms therefore outweigh the perceived psychological harm of diagnosis, despite the overwhelming rejection of this claim from affected individuals (Rawool & Kiehl. 2009). These benefits cannot however be achieved without establishing a level of equilibrium between perspectives of hearing loss as viewed by both parties. Since the life-threatening effects of MND understandably lead to individual focus directed towards life-sustaining success, the role of the caregiver becomes of utmost importance in guiding a desirable QoL. Achieving these desirable levels of QoL are affected by secondary impairments such as the presentation of hearing difficulties, therefore placing the responsibility of pursuing auditory diagnosis on caregivers who are able to report more objectively on the impact of hearing impairment. Paulo et al. (2008) therefore encourage that auditory diagnosis is pursued in the

earliest of stages as a means of ensuring prompt management before the consequences of hearing impairment introduce further negative effects to the QoL of both the MND individual as well as the caregiver.

Conclusion

The involvement of certain pathways and lack of involvement of others plays an important role in the diagnosis of MND, however prominent sensory symptoms complicate the ease of diagnostic clarity (Pall, 1995). There is a growing body of evidence that suggests the involvement of sensory pathways in MND. As life-prolonging measures continue to expand lifespan of individuals with MND, the sensory, autonomic and oculomotor pathways may emerge as more prominent features of MND than currently accepted (Pall, 1995). The involvement of the auditory pathways when superimposed on the current features of MND may prove to be catastrophic to the maintenance of individual wellbeing and QoL.

Furthermore, as the life span of individuals with MND extends, the possibility of acquiring a presbycutic hearing loss increases. This could be a major factor influencing individual involvement in communicative contexts, whereby the vast adverse effects of hearing loss on social and emotional health lead to a decline in societal and social engagements, withdrawal and loss of autonomy (Rawool & Kiehl, 2009). Where QoL and the ability to adjust to loss of function are described as the foundation to maintaining individual desire to be socially engaged at a familial and a community level, the additional consequences of sensory effects such as auditory impairment become of critical importance. The importance of auditory assessment to promote early identification of hearing loss, even in the absence of specific patient complaints is supported by Paulo et al. (2008). Early diagnosis of a hearing loss will facilitate access to aural rehabilitation that could promote the

maintenance of QoL for both the affected individual and as well as the caregivers (Paulo et al., 2008)

In the face of incurable, terminal illness, control over manageable symptoms need to grasped with urgency and become of an obligatory nature within the multi-disciplinary management plan. Minimizing the devastating consequences of the disease as a whole and strengthening the involvement and inclusion of the affected individual in day-to-day interactive processes shapes desire to live, which should remain a priority in disease management. Hearing ability affords individuals enhanced quality of experiences in life and considerations for this category of disease management should never be undermined in the management process.

"I stand alone in a great crowd of MND 'sufferers', but to be acknowledged and have my opinions respected reminds me that I am still here. I don't know what tomorrow brings, but as long as I am seen I will wake up each morning with more anticipation (than the last) and look forward to the contributions (small or big) that I can make. My body will stop fighting only when my value in the world is abandoned"

Quote from P**3**

Chapter Six

Conclusion

Introduction

This chapter provides an outline of the rationale for the current study followed by a summary of the findings in accordance with the primary and secondary objectives outlined in Chapter 2. This chapter then concludes with a critical review of the study, while recommendations for future research will mark the close of this chapter.

Summary of Rationale and Findings

Progressive, neurodegenerative diseases such as MND result in severe negative effects on the affected individual. These effects expand across the entire ICF framework impacting the individual at both a level of physical structure and function as well as participation and activities of daily living. All of these categories have a marked negative impact individual QoL.

The inclusion of sensory neuropathy as a clinical feature of the ALS spectrum remains a cause of diagnostic uncertainty. However, with enhancements in disease duration and the consequent prolongation of lifespan, a wider spectrum of MND related signs and symptoms are anticipated to become clinically apparent (Isaacs et al., 2007; Shaw, 2005). Since curative treatments for MND have yet to be discovered, the challenge in terminally, progressive disease lies with optimizing individual QoL. This is largely achieved through maintaining a sense of autonomy and independence – two components that are negatively affected by hearing loss. A loss or decline in auditory function is a key marker associated

with declining individual independence and is typically associated with withdrawal and voluntary or involuntary exclusion from social situations, hence threatening individual QoL.

Management of diseases of a multi-systemic nature, such as MND, is best met by multi-disciplinary teamwork. Management should focus on symptomatic and rehabilitative measures, encompassing physical and environmental factors that span across the spectrum of MND (Ng & Khan, 2011). Investigation surrounding the atypical features of MND is therefore essential for the purpose of refining the standard clinical description of MND and redefining management plans for each individual. This aims to address and accommodate disease features at both a primary and secondary level as far as medical and rehabilitative management currently allow (Isaacs et al., 2007). This further validates the need for research into less explored domains of MND - in this instance, auditory function and the associated implications.

Hearing loss in individuals diagnosed with adult onset MND in this study was six from a total of eight tested individuals The descriptions of audiological findings of this study follow:

- Auditory symptoms described by participants included tinnitus *(n* =5*)*, difficulty hearing in noise (*n* = 5), hyperacusis *(n* = 4*)*, recruitment *(n* = 3), dizziness and history of discharge respectively *(n* = 2*)*, history of pain, fluctuating hearing ability and vertigo *(n* = 1*)* respectively.

- Six participants presented with a high-frequency sensorineural hearing loss, whilst the

remaining two participants had hearing within normal limits. The configuration of hearing loss was typically sloping in twelve of the ears with a flat configuration observed in four of the ears respectively. In the ears that presented with a hearing loss, one presented with a mild high frequency hearing loss, three presented with mild to moderate high frequency hearing loss, three presented with a moderate high frequency hearing loss and five presented with a moderate-severe high frequency loss.

- The pattern and description of hearing impairment appears to follow the pattern of impairment typically associated with age related hearing loss and hence cannot be isolated to MND based on the limited sample size of the study.

The perceived psychosocial implications and level of handicap related to hearing impairment amongst MND participants were explored with the HHIA:

- Four participants (*n* =4) reported no handicap, whilst two participants (*n*=2) presented with mild handicap and an additional two (*n* = 2_with a moderate handicap relating to hearing impairment and its impact on social and emotional wellbeing.

- Four participants (*n =4)* reported greater difficulty at a level of social functioning versus one (*n = 1*) who reported greater difficulties at an emotional level of functioning.

- The remaining three participants *(n* = 3*)* described equal levels of social and emotional functioning relating to hearing ability, two of whom presented with no handicap as reported by HHIA and one of whom presented with a moderate handicap.

- Positive correlations were found between pure tone audiometry and HHIA social scores, while less significant correlations were noted when comparing pure tone audiometry to HHIA emotional scores.

Contact and/or referral to an audiologist and functional abilities as rated by participants and their caregivers were further explored with the HEQ:

- None of the affected individuals in this study had contact with an audiologist or been referred for auditory testing by a speech-language pathologist and/or another professional in the multidisciplinary team.

- The HEQ revealed that all participants with MND rated communication as the most important functional skill, followed by vision *(n* = 5*)* and deglutition *(n* = 4*)*.

- Caregivers rated communication *(N* = 8), hearing (*n* = 6) and vision *(n* = 5*)* as the most critical functional skills.

- These findings indicate that caregivers and participants with MND share equal views regarding the importance of maintaining communicative ability in the face of degenerative disease that limits ones physical capabilities.

- Both groups view vision as another fundamental ability contributing to the maintenance of social inclusion and general wellbeing, more so than arm or leg movement.

- Views differ in relation to deglutition and hearing, where MND individuals identified the ability to chew, swallow and eat for pleasure to be a functional skill of more importance than auditory function.

- Participant caregivers in contrast indicated hearing ability to be the more important skill highlighting the importance of social inclusion and sense of belonging.

The findings of the final aim, to explore the perceptions of MND participants and their caregivers relating to the perceived value of auditory testing and possible diagnosis, are as follows:

- The majority of the participants (n = 5) with MND reported the diagnosis of auditory impairment to be of no relevance, followed by two who viewed this information to be extremely important and one participant who viewed this as something interesting to know.

- Conversely, the majority of caregivers *(*n = 5*)* reported the diagnosis of auditory diagnosis to be of extreme importance, followed by one who viewed this information to be interesting and two who reported this knowledge to be of no relevance to them, their MND spouse/ partner or their current lifestyle.

- Translation of responses followed by data coding revealed the emergence of three main themes namely, positive perceptions, negative perceptions and awareness.

- Positive perceptions linked to auditory diagnosis related to elements of autonomy, control and self-preservation.

- Negative perceptions linked to auditory diagnosis related to elements of fear, burden and vulnerability.

- Aspects of awareness related to lack of knowledge of diagnosis and management approaches to auditory impairment and conversely strong understanding of auditory importance and benefits of diagnosis.

- Results suggested that caregivers were more readily able to acknowledge, accept and understand the value of auditory testing and diagnosis when superimposed on living with a neurodegenerative disease such as MND. In contrast, participants tend to present with more resistance, denial and reluctance in accepting the impact that such diagnosis may have on QoL if left undetected.

Strengths of the Study

- The test protocol was isolated to a once-off test session, thereby serving to limit the effects of attrition on a data collection process as far as possible. This is in contrast to testing individuals with MND extending over multiple appointments per individual, which was likely to present with greater attrition rates than those experienced in the execution of the current study.

- Equipment calibration was confirmed prior to all testing appointments to promote

uniformity of all test measures across MND individuals in the study, ensuring consistency of results across all individuals.

- The sample was representative of the MND population in South Africa (varying ages, races, genders, and as far as possible, types and stages of MND). The latter suggests that these results are representative of the broader population of MND individuals in South Africa, although the limited sample size does warrant caution in overgeneralization of findings.

- The test protocol for auditory assessment included both behavioural and objective test measures thereby strengthening the consistency and cross-checking of findings from which conclusions were drawn.

- The test protocol was designed to ensure more objective test measures were performed towards the end of the data collections session. This ensured that individual fatigue did not influence responses to behavioural test measures. All individuals tested in the current study were capable of completing the full test battery and benefitted from the passive involvement in electrophysiological measures scheduled at the end of each session.

- Inter-rater reliability was established to strengthen the reliability and validityof the documented findings. The auditory test battery further supported this by providing cross-checking opportunity as a confirmation of the reliability of individual responses to various audiological assessments.

- Caregivers were presented with the HEQ questionnaire to complete during test time. This allowed for active caregiver involvement in the current study and provided valuable contribution from caregivers during the waiting period of auditory testing with the affected MND individual.

- Inclusion of caregivers in this study allowed for the comparison of views between the participants with MND and their caregivers, the latter of whom are commonly overlooked in the MND management process.

Limitations of the study

The current research was a preliminary effort to deal with the obvious lack of information on the prevalence of hearing loss in South African individuals diagnosed with adult-onset MND. A number of limitations were however identified as factors hindering the complete realization of the objectives outlined by this study. These include:

- The limited sample size is one of the primary limitations of the study. Notwithstanding the efforts by the researcher, the unique circumstances of each participant, the unpredictable nature of MND, and a high attrition rate, only a limited the number of participants could be included in the study. Hackshaw (2008) identifies a benefit of a small sample size to be associated with quicker participant enrollment; however this was not true for the current study. The process of recruiting individuals to participate in this study was of a lengthy nature and had a significant negative effect on the designated timeline for data collection procedures. Factors such as locating MND individuals representative of the general population and encouraging individual involvement proved challenging.

- The limited sample size eliminated the researchers' ability to obtain a sample representative of all stages of disease progression, which would have allowed for further analysis into the impact of MND on the auditory system at the various stages of disease progression. Exploring the effects of MND on auditory function at various stages of the disease therefore fell outside the scope of the current study.

- Similarly, the limited sample size further eliminated the ability to review results according to the different types of MND (spinal, bulbar and mixed).

- An additional limitation in terms of participant recruitment to the study was linked with determining an appropriate test site. This dealt with drawing a comparison between home testing, the more desirable and comfortable setting for participants versus clinic testing, the more reliable test environment for the purpose of result reliability. Ultimately, test reliability was deemed central to maintaining the integrity of this study and the clinical test setting was selected.

- The inclusion of speech-in-noise testing furthermore would have been beneficial for acquiring further information pertaining to the abilities of those participants who reported difficulty hearing in background noise.

- The use of a limited sample size affects statistical analysis procedures linked to achieving confidence intervals of 95% and interpretation of *p*-values, emphasizing the need for calculated balance in the interpretation of the results (Hackshaw, 2008).

- Replication of the current study would warrant adaptation of the HHIA questionnaire

to ensure greater specificity of test instructions. It is suggested that for the purposes of a similar study, the instructions must highlight that the HHIA is utilized as a means of exploring the various auditory difficulties one *may* experience as a result of potential hearing loss, rather than auditory difficulties experiences as a definitive consequence of hearing loss.

- Participants in the more terminal stages of the disease were not included due to the marked deterioration of physical function linked to mobility, speech and respiratory function as well as the fatigue and emotionally taxing demands experienced during this time. This eliminated the researchers' ability to obtain a sample representative of all stages of disease progression, which would have allowed for further analysis into the impact of MND on the auditory system at the various stages of disease progression.

While the above limitations are critical considerations to hold for future studies, small samples are possibly a more desirable route when examining new research hypothesis, such as in the current study (Hackshaw, 2008). One should consider that the use of a small sample size may not reveal a relationship which would only be exposed when applying the same methodology to a larger sample.

Recommendations for future research

Research on MND as it relates to the discipline of audiology is limited, and as a result there are many unanswered questions linked to atypical symptomatology in MND. A number of opportunities for further research surrounding MND and the auditory system include:

- First and foremost, replicating this study using a larger sample size, within a broader geographical context. This may be done using a collaborative approach amongst different academic/clinical institutions researching the auditory involvement in MND. Consistency in equipment and calibration would be critical, however would also allow for test sites to be more easily accessible to willing individuals from a wider geographical area.

- Alongside the recruitment of a larger sample size, future research may wish to explore the differences in audiological presentation in the various types of MND (spinal, bulbar and mixed).

- Future research may aim to conduct a longitudinal study tracking auditory function across the disease progression will provide further information on the auditory function as the disease progresses.

- It is further recommended to expand the audiological test battery used in this study to include the assessment of the higher levels of the brainstem and the cortex with measures such as the mid-latency response and the late-latency response. This would further aid auditory testing in this population, by eliminating the requirement for more demanding and physically taxing behavioural test measures and replacing these with less strenuous electrophysiological measures.

- Anecdotal evidence of hyperacusis was reported in this study, which may also serve as a worthwhile area of exploration in future MND based studies.

- A qualitative research study exploring the understanding of the benefits of audiologists as part of the MND management team from the (i) perspective of the persons with MND, (ii) their caregivers, (iii) audiologists, and (iv) the MDT involved in the management of the individuals with MND.

- The mean time participants voluntarily remained at the test site to discuss communicative and auditory-based experiences in daily life warrants futher investigation. This suggests that further research exploring and validating the role of speech-language pathologists & audiologists in counseling as well as their knowledge and skill when working with individuals with MND and their families, is indicated.

Clinical Implications

- Individuals with MND face a vast range of life threatening consequences (e.g. pneumonia, respiratory difficulties, risks of aspiration and physical concomitants). Within the ICF framework however, QoL is paramount to this holistic approach to individual management. Hearing loss and the threats this presents to social, emotional and communicative success has significant effects on QoL. Hence alongside the devastating physical consequences of MND, this study served to bring to light the profound impact hearing loss has on the QoL of a population facing repeated redefinition of self and ability in accordance with the holistic framework outlined by the ICF.

- The most important clinical implication of the current study is that MND individuals are likely to experience hearing loss to various extents and severity during the course of the disease. While it cannot be ascertained with such a limited sample size whether

hearing loss is directly related to MND or natural aging, indicators suggest hearing impairment is likely to pose a threat to communicative success at some stage over the course of the disease.

- A majority of current MND research relates to the physical consequences of motor neuron degeneration. While the significance of these physical symptoms should not be undermined, it is apparent that individuals with MND shift their focus away from physical abilities and towards maintenance of QoL, dignity and autonomy. This study exposes the devastating outcome of hearing loss on individual autonomy and interpersonal relationships and highlights the importance of acknowledging the effects this poses on maintaining QoL despite deterioration of physical ability. This knowledge serves to contribute to the body of information directed at promoting enhanced QoL for MND individuals.

- The benefits of amplification equipment such as hearing aids and communication-based strategies are undermined. This study draws attention to the positive effects of achieving regulation of sensory dysfunction and stabilization of the affected individuals' environment, particularly in the face of non-curable progressive decline of physical function. This and the impact it has on individual QoL often supersede individual desire for mobility. Further to this, it highlights the benefits of establishing enhanced functionality of the individual within communicative contexts.

- Understanding of the presentation of hearing loss amongst the MND population further assists in the process of selecting AAC devices accommodating for receptive and expressive communicative impairments.

- As a result the likelihood of hearing impairment and the limited reported involvement of the audiological discipline, this study highlights the need for expansion of the multi-disciplinary MND team to include those services of the audiological discipline, as well as empower individuals with knowledge and understanding of the detrimental effects of hearing impairment. Identifying this gap is of great value for building a body of support to promote an increase in audiological counseling and rehabilitative services available to populations in need of multi-disciplinary management.

- The large variations in caregiver versus MND-individual perspectives relating to auditory diagnosis emphasize the need for counseling units equipping families to succeed and maintain QoL in the presence of hearing impairment. These should be aimed at the immediate family unit as a whole rather than isolated symptomatic management of the affected individual and be of an ongoing nature as opposed to a once off opportunity. This would serve to take into account the burden experienced by the MND caregiver in combination with the affected individual.

- This study further exposed caregiver and MND-individual desire for ongoing counseling opportunities, allowing for caregiver and MND-individual experiences, queries and concerns to be accounted for. This suggests the need for a holistic, united approach to managing hearing loss, accounting for the frustrations and challenges faced by both parties rather than those faced by the individual with MND alone.

- This study also served to emphasize that while QoL is deemed of utmost importance to all individuals, in the face of such a multi-faceted degenerative disease where life

threatening physical and physiological management is of primary importance, secondary elements that threaten QoL fail to be as readily acknowledged. This aimed to bring to light the devastating effects hearing loss can have on a population with an already diminished QoL hence exacerbating the difficulties faced. Awareness of and addressing functional elements that can be rehabilitated therefore become imperative in the management plan of the individual. This is particularly true for auditory ability, since communication is reliant on verbal and auditory ability. MND individuals face a loss of physical function for gestural communication and speech ability for verbal communication, suggesting auditory ability to be the final element promoting some level of interactive success. Where MND individuals are afforded prolonged lifespan as a result of medical advancements, maintaining a desirable QoL increases in importance for this population.

Summary

This final chapter provided a summary of the rationale and the results for the research study described in this report. A critical review of the strengths and limitations identified in the execution of this study as well as practical recommendations for future research in this domain were provided. This chapter concluded with a review of the clinical implications arising as a consequence of the present research study.

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Appendix A: MND Severity Classification

STAGE CLASSIFICATION OF MOTOR NEURON DISEASE

Riviere, M., Meininger, V., Zeisser, P., & Munsat, T. (1998). An Analysis of Extended Survival in Patients with Amyotrophic Lateral Sclerosis Treated with Riluzole. *Archives of Neurology*, 55, 526 - 528.

Stage	Severity Classification	Clinical Features
I	Mild	- mild deficit presenting in one of three regions (speech, arm or leg) - Functionally independent in: - speech - upper extremities of daily living - ambulation
II	Moderate	- mild deficit in all three regions (arm, leg and speech) **OR** - moderate to severe deficit in one region - remaining two regions remain normal or mildly affected
III	Severe	- requiring assistance in two or three regions - dysarthric speech production and/ or - assistance required for walking and/ or - assistance required with upper extremities of daily living
IV	Terminal	- non-functional use of at least 2 regions - moderate or non-functional use of the third region

Appendix B: MNDSA Information Letter

SPEECH PATHOLOGY AND AUDIOLOGY
School of Human & Community Development
Faculty of Humanities
University of the Witwatersrand
Private Bag 3, WITS, 2050
Tel: (011) 717 4577; Fax: (011) 717 4572

15 April 2010

Dr. F. Henning

National Chairperson

MNDSA

Re: Permission to conduct research with adult-onset Motor Neuron Disease Patients Associated with the MNDSA

Dear Dr. Henning,

I am conducting research as part of the requirements for a Masters degree in Audiology at the University of the Witwatersrand. The primary aim of this study is to determine the prevalence and perceptions of hearing loss in individuals' diagnosed adult-onset Motor Neuron Disease (MND).

Studies have indicated that MND patients' regard autonomy and maintenance of identity as key factors influencing quality of life. A critical cornerstone for the maintenance of autonomy is communication. A reduced ability to perceive speech clearly as a result of auditory decline threatens success within communicative contexts. Auditory involvement in patients' with MND is yet to be documented, despite the implications this may have on a patients' quest to maintain a sense of self and belonging in the face of a gradual loss of independence in motor function.

I would like to obtain your permission to invite members of the MNDSA to participate in this study. Participants would be required to travel to the University of the Witwatersrand Audiology Clinic for a 90 minute audiological evaluation. The entails a complete evaluation of auditory function from the outer ear to the brainstem. This would include behavioural measures in the form of otoscopic evaluation, pure tone and speech audiometry. Test modifications to these procedures will be made to accommodate the physical and

communicative abilities of participants. Objective measures, requiring no physical responses, in the form of immitance testing and auditory brainstem responses (ABR) will also be performed. A questionnaire relating to auditory function will also be presented to MND-individuals and caregivers, for completion.

The participants for this study are required to comply with the following selection criteria: i) neurologist confirmed diagnosis of MND; ii) spinal or bulbar onset; iii) MND classification at stages I, II, III or IV; iv) older than 30 years; v) proficient in English; vi) living in Gauteng. Persons with MND who present with instruction following difficulties and/ or the presence of co-morbid factors with known auditory involvement (e.g diabetes) will be excluded from the study.

Participants will be offered financial compensation for travel expenses on the day of testing. The testing site is accessible to wheelchair users and has nearby parking. The sequential order of tests will be arranged in a manner that ensures behavioural measures are obtained first and the pace of testing will be designed to accommodate potential fatigue experienced by participants. Significant findings linked to auditory thresholds will be dealt with accordingly. This may include ENT specialist referral and/ or group session attendance addressing facilitative communication-based strategies aiding communicative success in the presence of hearing loss. These recommendations are dependent on the analysis of auditory findings and will be patient specific.

Ethical considerations will be of utmost importance. Participation is voluntary and informed consent will be issued to all prospective participants. Confidentiality will be guaranteed. The right to withdraw at any time without penalty will be clearly expressed. No harm will come to participants. Ethical clearance will be pursued through the submission of a study proposal to the Medical Research Ethics Committee at the University of the Witwatersrand, prior to data collection.

Your permission for granting the participation of the adult-onset MND patients associated with MNDSA would be greatly appreciated. In this event, I would request written and signed permission to be faxed to 011 740 2319 or e-mailed to the address provided below. Provisional permission pending ethical clearance would be admissible.

Should you require any further information regarding this proposed study, please feel free to contact me on 083 235 5629 or e-mail: ephilippou@webmail.co.za

With Appreciation,

E.Philippou
Masters Student
E-Mail: ephilippou@webmail.co.za
Tel: 083 235 5629

Dr. Karin Joubert
Research Supervisor
E-Mail: Karin.joubert@wits.ac.za
Tel: 011 717 4561

Appendix C: MNDSA Letter Granting Permission

Appendix C: Permission for Research Study from MNDSA

MNDA
OF SOUTH AFRICA
MOTOR NEURONE DISEASE ASSOCIATION

P.O. BOX 789,
HOWARD PLACE
7450

6 May 2010

Miss Elena Philippou
Audiology
University of the Witwatersrand

Dear Elena

Thanks for your email and attached letter. I am happy with the format of your study and am willing to grant you permission to continue. May I ask that it is made quite clear to the MNDA of SA members that the study has no relation to the MNDA of SA and that you merely obtained their contact details from us. We would like to ensure that members do not to feel obliged to participate in the study.

Regards and good luck with the study

Dr. Franclo Henning, FC Neurol (SA),
Neurologist, Division of Neurology, Tygerberg Hospital / Stellenbosch University
Chairperson: MND Association of SA

Appendix D: Participant Case History Questionnaire

CASE HISTORY QUESTIONNAIRE

Participant Number: ____ Date of Birth: _____ Interview Date: _____

1. PERSONAL DETAILS (please tick (√) the relevant detail)

1.1 Gender	Male	Female		
1.2 Home Language	English	Afrikaans	Zulu	Other (specify)
1.3 Hand Dominance	Right	Left		

2. AUDITORY HISTORY

2.1 Do you believe you have hearing difficulties? If yes, please describe the difficulties you have below. _____ _____ _____ _____	Yes	No

2.2 When did your hearing difficulties begin?	

2.3 Have you had a hearing test?	Yes	No

2.4 Please provide the results of your most recent hearing test below _____ _____ _____

2.5.1 Do you wear a hearing aid?	Yes	No	
2.5.2 If yes, on which ear?	Right	Left	Both

2.6 Do you have a family history of hearing loss?	Yes	No
If yes, please provide details below. _____ _____ _____ _____		

2.7 Have you had any surgery to the head, neck or ear?	Yes	No
If yes, please provide details below. _____ _____ _____ _____		

3. MEDICAL DETAILS

3.1 When did your symptoms begin?	

3.2 What was your age when your symptoms began?	

3.3 Where did you first seek medical advice?	General Practitioner (GP)	Neurologist	Other (specify)

3.4 When were you diagnosed with Motor Neuron Disease (MND)?	

3.5 Who diagnosed you with MND?	General Practitioner (GP)	Neurologist	Other (specify)

3.6 Which type of MND were you diagnosed with?	Mixed	Spinal	Bulbar

3.7 Please tick the detail that describes your ***current abilities*** in the following areas:

ARM MOVEMENTS	**√ / x**	**LEG MOVEMENTS**	**√ / x**	**SPEECH**	**√ / x**
No difficulties with arm movement		No difficulties with leg movement		No difficulties with speech production	
Able to move arm, forearm and wrist (upper extremities) independently		Able to walk independently (ambulation)		Able to produce speech independently	
Mild difficulty with arm movement		Mild difficulty with leg movement		Mild difficulty with speech production	
Moderate – Severe difficulty with arm movement		Moderate- Severe difficulty with leg movement		Moderate-Severe difficulty with speech production	
Assistance needed with upper extremities of daily living		Able to walk with assistance		Dysarthric Speech (slurred, unclear sounds when spoken)	
No functional arm movement		No functional leg movement		No functional use of speech	

3.8 Please mark which illnesses you have suffered over the past 5 years:

Asthma		Influenza		Otosclerosis		Blood Pressure	
Chronic Otitis Media		Tinnitus (ringing in the ear/s)		Hormone Replacement Therapy		Loud Noise Exposure	
Earache/Pain		Diabetes		Menieres Disease		Tubercolosis	
Ear Discharge		Mastoiditis		Malaria		Cancer	
Recruitment (sudden jumps in loudness)		Hyperacusis (sensitivity to regular sounds)		Meningitis			

3.9 What medications have you been on (in the past 5 years)?

Name of Medication:	Prescribed For:	Name of Medication:	Prescribed for:

4. COMMUNICATION

4.1 What is your preferred method of communication at present?	Speech	Writing	Eye tracking	Communication device

4.2.2 If speech, are you able to express yourself easily using:	Vocal Sounds	Single Words	Short Phrases	Sentences

4.3 If you use a communication device, is it:	Low technology			High technology electronic
	Picture Board			Computer-Based
	Alphabet Chart			

4.4 What alternate methods do you use to help your communication with others?

_____
_____
_____
_____

5. RESULTS

5.1 Would you like to find out the results of your hearing test?	Yes	No

5.2 If yes, would you prefer:		
a) to have results explained to you immediately after the test session?	Yes	No
b) to have your results posted to you?	Yes	No
c) both of the above	Yes	No

6. TRANSPORT

6.1 Please tick below

I am able to arrange my own transport to the test site.	
I am unable to arrange my own transport to the test site, although I would like to participate in the study	

Thank-you for taking the time to complete this questionnaire.

Appendix E: Hearing Experience Questionnaire (HEQ) (MND Individual Component)

HEARING EXPERIENCE QUESTIONNAIRE

1. CONTACT WITH THE SPEECH PATHOLOGY AND AUDIOLOGY DISCIPLINE

1.1 Have you consulted with a speech therapist since your diagnosis?	YES	NO
1.2 Have you consulted with an audiologist since your diagnosis?	YES	NO

1.3 Please provide a year for your first consultation with a speech therapist?	
1.4 Please provide a year for your first consultation with an audiologist?	

1.5 How often did/do you consult with the speech therapist?	ONCE OFF CONSULTATION	REPEATED CONSULTATIONS
1.6 How often did/do you consult with the speech therapist?	ONCE OFF CONSULTATION	REPEATED CONSULTATIONS

1.7 Describe the contribution made by the speech-language therapist and/ or audiologist in assisting your communicative abilities.

_____
_____
_____
_____

2. COMMUNICATION AND HEARING

2.1 What is your preferred method of communication at present?	Speech	Writing	Eye tracking	Communication device

2.2 If speech, are you able to express yourself easily using:	Vocal Sounds	Single Words	Short Phrases	Sentences

2.3 If you use a communication device, is it:	Low technology		High technology electronic	
	Picture Board		Computer-Based	
	Alphabet Chart			

2.4 Would communication be than hearing?	MORE IMPORTANT	LESS IMPORTANT	JUST AS IMPORTANT

Please provide a reason for your answer.

_____
_____
_____
_____

2.5 Rate the following abilities in order of importance to you (1-most important 6-least important)

COMMUNICATION		VISION	
HEARING		EATING AND SWALLOWING	
LEG MOVEMENT		ARM MOVEMENT	

2.6 Describe how the changes in your communicative abilities have affected your social and functional abilities in daily life.

_____
_____
_____
_____
_____
_____
_____
_____

2.7 How do you believe an added hearing loss would affect these abilities?

_____
_____
_____
_____
_____
_____
_____
_____

2.8 Would the added diagnosis of a hearing loss be:

EXTREMELY IMPORTANT FOR ME TO KNOW ABOUT		NOT GREATLY SIGNIFICANT TO ME	
INTERESTING FOR ME TO KNOW ABOUT		IRRELEVANT TO ME	

Please explain the reason for you choice:

_____
_____
_____
_____
_____
_____
_____
_____

2.9 Do you experience any of the following:

RINGING IN THE EARS		**SENSITIVITY TO LOUD NOISES/ SOUNDS**	
DIFFICULTY HEARING IN A QUIET ROOM		**SENSITIVITY TO REGULAR NOISES/SOUNDS**	
DIFFICULTY WITH HEARING IN GROUP SITUATIONS		**FLUCTUATIONS/ CHANGES IN HEARING ABILITY**	
EAR PAIN		**SUDDEN BURST OF LOUDNESS**	
EAR DISCHARGE			

2.9 Please complete the attached questionnaire: ***Hearing Handicap Inventory for Adults***

THANK YOU FOR YOUR ASSISTANCE!

Appendix F: Hearing Experience Questionnaire (MND Caregiver Component)

1. COMMUNICATION AND HEARING

1.1 Rate the following abilities in order of importance (1-most important 6-least important). List these items in the order you perceive to be most – least important to your spouse/ partner.

COMMUNICATION		VISION	
HEARING		EATING AND SWALLOWING	
LEG MOVEMENT		ARM MOVEMENT	

1.2 Describe how the changes in your partner/spouse's communicative abilities have affected your social and functional abilities in daily life.

_____
_____
_____
_____
_____
_____
_____

1.3 How do you believe an added hearing loss in your partner/spouse would affect these abilities?

_____
_____
_____
_____
_____
_____
_____
_____

THANK YOU FOR YOUR ASSISTANCE!

Appendix G: Hearing Handicap Inventory for Adults (HHIA)

HEARING HANDICAP INVENTORY FOR ADULTS

Instructions: The purpose of the scale is to identify your experiences in different listening situations
Check Yes, Sometimes, or No for each question. Do not skip a question if you avoid a situation because of a hearing problem.

	Yes	Sometimes	No
1. Does a hearing problem cause you to use the phone less often than you would like? (s)			
2. Does a hearing problem cause you to feel embarrassed when meeting new people? (e)			
3. Does a hearing problem cause you to avoid groups of people? (s)			
4. Does a hearing problem make you irritable? (e)			
5. Does a hearing problem cause you to feel frustrated when talking to members of your family? (e)			
6. Does a hearing problem cause you difficulty when attending a party? (s)			
7. Does a hearing problem cause you difficulty hearing/understanding co-workers, clients, or customers? (s)			
8. Do you feel handicapped by a hearing problem? (e)			
9. Does a hearing problem cause you difficulty when visiting friends, relatives, or doctors? (s)			
10. Does a hearing problem cause you to feel frustrated when talking to co-workers, clients, or doctors? (e)			
11. Does a hearing problem cause you difficulty in the movies or theater? (s)			

12. Does a hearing problem cause you to be nervous? (e)			
13. Does a hearing problem cause you to visit friends, relatives, or neighbors less often than you would like? (s)			
14. Does a hearing problem cause you to have arguments with family members? (e)			
15. Does a hearing problem cause you difficulty when listening to TV or radio? (s)			
16. Does a hearing problem cause you to go shopping less often than you would like? (s)			
17. Does any problem or difficulty with your hearing upset you at all? (e)			
18. Does a hearing problem cause you to want to be by yourself? (e)			
19. Does a hearing problem cause you to talk to family members less often than you would like? (s)			
20. Do you feel that any difficulty with your hearing limits or hampers your personal or social life? (e)			
21. Does a hearing problem cause you difficulty when in a restaurant with relatives or friends? (s)			
22. Does a hearing problem cause you to feel depressed? (e)			
23. Does a hearing problem cause you to listen to TV or radio less often than you would like? (s)			
24. Does a hearing problem cause you to feel uncomfortable when talking to friends? (e)			
25. Does a hearing problem cause you to feel left out when you are with a group of people? (e)			

For Clinician's use only: *Yes = 4 Sometimes = 2 No = 0*

Total score: ____ Total score for e-questions: ____ Total score for s-questions: ____

Appendix H: Participant Informed Consent

SPEECH PATHOLOGY AND AUDIOLOGY
School of Human & Community Development
Faculty of Humanities
University of the Witwatersrand
Private Bag 3, WITS, 2050
Tel: (011) 717 4577; Fax: (011) 717 4572

THE PREVALENCE AND PERCEPTIONS OF HEARING LOSS IN INDIVIDUALS DIAGNOSED WITH ADULT ONSET MOTOR NEURON DISEASE

Good day,

INVITATION TO PARTICIPATE IN POSTGRADUATE RESEARCH STUDY

Thank you for taking the time to read this letter. I am conducting research in the field of Motor Neuron Disease (MND) as part of the requirements for a Masters degree in Audiology at the University of the Witwatersrand. Medical research is done to enhance the understanding and knowledge in the field of medicine and disease. The aim of this study is to determine the prevalence and perceptions of hearing loss in individuals' diagnosed adult-onset Motor Neuron Disease (MND).

I would like to invite you to participate in this study. As part of this study you will receive a complete evaluation of your hearing abilities. Participation will involve a once-off test session, lasting a period of 90 minutes. Intervals and rest periods will be offered throughout the evaluation, if needed. Assessment procedures will include tests that rely on your response, while other measures will not require a direct response from you. Each test will be modified to best accommodate your physical and communicative abilities. The pace and order of tests will also be designed to best accommodate your needs. A brief questionnaire will also be issued to you and your primary caregiver (partner, spouse etc.) for completion.

You would be required to travel to the University of the Witwatersrand Audiology Clinic on the day of testing. If you are eager to participate, however lack the means of transport, kindly make note of this on the Case History Form and possible transport arrangements may be made to assist your participation in the study. Participants who are able to come to the clinic using their own means of transport will be offered R100-00 financial compensation for travel expenses on the day of testing. The testing site is accessible to wheelchair users and has nearby parking.

POTENTIAL BENEFITS AND RISKS
There is no direct benefit to participating in this research, with exception to getting a confirmation of your hearing ability. The information collected will however be valuable in understanding the vulnerability of the hearing system in patients with Motor Neuron Disease,

which is currently an area of research that has been neglected. This information will also serve to guide communication specialists in modifying the communicative environment to help the process of communicative exchange. This in turn may present further opportunities for enhancing quality of life and your interactions with communicative partners.

There are no known risks associated with your participation in this research.

HEARING TEST RESULTS

The findings from your hearing test may or may not indicate the presence of a hearing loss. It is entirely your decision whether you would like to know the results of your hearing test or not. Should you wish to find out the results of your hearing test, this may be done on the day of testing. You may alternatively request that results are posted directly to you. Where recommendations can be made, they will be given to you based on your individual hearing test results. You are at no point obligated to follow-through with these recommendations. You may also request that results from the hearing test are not shared with you and the research team will comply with this request.

PARTICIPANT RIGHTS

- Participation is entirely voluntary and you will not be forced to participate in this study without giving your full consent. You are not obligated to participate in this study.

- You have the right to withdraw from this study at any time without penalty, even if you initially agreed to take part.

- Anonymity will be guaranteed. There will be no identifying information on Case History forms and/ or assessment forms. No identifying information will be used in the research report.

- Every effort will be made to guarantee confidentiality. All personal information will be treated with the utmost confidentiality and will only be reviewed by the research team (researcher, research assistant and academic supervisor). Personal information will be safely stored in a locked cabinet and no other parties will have access to this.

PARTICIPANT CONSENT

Should you wish to participate in this study, please ***sign*** and complete the informed consent document below and the enclosed Case History Form. Kindly post this in the self-addressed envelope at your earliest convenience. Please ensure this is returned no later than 10 November 2010.

Should you require any further information regarding this proposed study, please feel free to contact me on 083 235 5629 or e-mail: ephilippou@webmail.co.za . You may alternatively contact the Chairperson of the Medical Ethics Committee at the University of the Witwatersrand, Anisa Keshav on 011 717 1234 or e-mail: Anisa.Keshav@wits.ac.za for additional enquiries.

With Appreciation,

E.Philippou
Masters Student
E-Mail: ephilippou@webmail.co.za
Tel: 083 235 5629

Dr. Karin Joubert
Research Supervisor
E-Mail: Karin.joubert@wits.ac.za
Tel: 011 717 4561

By signing below, I (name)_____, (contact number) _____voluntarily agree to participate in the study as outlined above.

Participant signature: _____ Date: _____
Please Note: an imprint of your finger (thumb) may be used should written consent not be possible.

Researcher signature: _____ Date: _____

Appendix I: Most Comfortable Level Response Modification

Appendix J: Threshold of Discomfort Response Modification

UNCOMFORTABLY LOUD

Appendix K: Alphabet Chart

A	**E**	**I**	**O**	**U**
B	**C**	**D**	**F**	**G**
H	**J**	**K**	**L**	**M**
N	**P**	**Q**	**R**	**S**
T	**V**	**W**	**X**	**Y**
Z	**YES**		**NO**	

Appendix L: Ethical Clearance Certificate

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

HUMAN RESEARCH ETHICS COMMITTEE (MEDICAL)
R14/49 Miss Elena Philippou

CLEARANCE CERTIFICATE	M10569
PROJECT	The Prevalence and Perceptions of Hearing Loss in Individuals Diagnosed wit Adult Onset of Motor Neuron Disease (MND) (Revised Title)
INVESTIGATORS	Miss Elena Philippou.
DEPARTMENT	Speech Pathology & Audiology
DATE CONSIDERED	28/05/2010
DECISION OF THE COMMITTEE*	Approved unconditionally

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

DATE 10/10/2011 **CHAIRPERSON** .....
(Professor PE Cleaton-Jones)

*Guidelines for written 'informed consent' attached where applicable
cc: Supervisor : Dr K Joubert

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...