

**Clinical and audiological features of Ménière's disease:
Insight into the diagnostic process.**

by

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ABSTRACT

Ménière's disease is the third most common inner ear disorder. The individual course of Ménière's disease in different patients makes it difficult to diagnose on the basis of symptomatology alone. The impact of Ménière's disease on quality of life has highlighted the importance of an additional tool to support the diagnosis of Ménière's disease. Apart from the patient's history, audiological data provide the most relevant information for confirming the diagnosis. The aim of this study was to analyse and describe the clinical and audiological features of a cohort of subjects diagnosed with Ménière's disease, in order to develop understanding of the pathophysiology of the disease and to facilitate the diagnostic process. The research is based on a retrospective study of the medical records of 135 subjects with Ménière's disease which were selected according to a non-probability sample. Descriptive statistics were used to organize, analyse and interpret the data. Sixty one percent of subjects presented with definite Ménière's disease, 14 % with probable Ménière's disease and 25 % with possible Ménière's disease. The results showed a higher incidence of Ménière's disease in females especially in the vestibular type. Three percent of subjects indicated a family history of Ménière's disease. Bilateral Ménière's disease presented in 39 % of subjects. The results confirmed that vertigo was the most debilitating symptom in Ménière's disease. Correlating the clinical features of subjects with audiometric and vestibular tests highlighted the clinical value of an audiological test battery including the following tests: Pure tone audiometry, Speech discrimination, Oto-acoustic emissions, Electronystagmography and Electrocochleography. This confirms the role of the audiologist in the diagnostic and rehabilitation process in patients with Ménière's disease.

Terminology: Clinician, diagnostic process, differential diagnosis, endolymphatic hydrops, etiology, idiopathic, Ménière's disease, pathogenesis, recurrent vestibulopathy.

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OPSOMMING

Ménière se siekte is die derde algemeenste binne-oor afwyking. Die individuele verloop van Ménière se siekte in verskillende pasiënte maak dit moeilik om 'n diagnose alleenlik op grond van die simptomatologie te maak. Die impak van Ménière se siekte op kwaliteit van lewe beklemtoon die belangrikheid van 'n addisionele hulpmiddel om die diagnose van Ménière se siekte te bevestig. Naas die gevalsgeskiedenis bied oudiologiese data die mees toepaslike inligting vir die diagnose van Ménière's se siekte. Die doel van die studie was om 'n reeks proefpersone met Ménière's se siekte se simptome te evalueer in verhouding tot oudiologiese en vestibulêre toeste, om kennis uit te brei ten opsigte van die patofisiologie van die afwyking, asook om die diagnostiese proses te vergemaklik. Die navorsing is gebaseer op 'n retrospektiewe studie van die mediese verslae van 135 proefpersone met Ménière se siekte, wat volgens 'n nie-waarskynlikheids steekproef geselekteer is. Beskrywende statistiek is gebruik om die data te organiseer, te analiseer en te interpreteer. Een en sestig persent van die proefpersone is gediagnoseer met definitiewe Ménière se siekte, 14 % met waarskynlike Ménière se siekte en 25 % met moontlike Ménière se siekte. Volgens die resultate is daar 'n hoër voorkoms van Ménière se siekte onder vrouens, veral ten opsigte van vestibulêre Ménière's se siekte. 'n Familie geskiedenis van Ménière se siekte is in 3 % van die proefpersone vasgestel. Bilaterale Ménière se siekte was verteenwoordig deur 39 % van proefpersone. Die resultate het bevestig dat vertigo die hoofsimptoom is wat lei tot ongeskiktheid. Die verhouding tussen die kliniese kenmerke van pasiënte en oudiometriese, asook vestibulêre toetsresultate beklemtoon die kliniese waarde van 'n oudiologiese toetsbattery insluitende die volgende toetse: Suiwertoonoudiometrie, Spraakdiskriminasie, Oto-akoestiese emissies, Elektronistagmografie en Elektrokogleografie. Dit bevestig die rol van die oudioloog in die diagnostiese- en rehabilitasie proses in pasiënte met Ménière se siekte.

Sleutelwoorde: Klinikus, diagnostiese proses, differensiaaldiagnose, endolimfatiese hidrops, etiologie, idiopatiese, Ménière's se siekte, patogene, herhalende vestibulopatie.



"When you are inspired by
some great purpose, some extraordinary project
all your thoughts break their bonds
your mind transcend limitations
your consciousness expands in every direction
and you find yourself in a new,
great and wonderful world."

- Patanjali

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CHAPTER 1

INTRODUCTION AND RATIONALE

This chapter aims to outline the purpose and relevance of the study by discussing various concepts associated with the diagnosis of Ménière's disease (MD), by presenting a rationale for the study, stating the research problem, clarifying terminology, and providing an outline for the organization of the dissertation.

1.1 INTRODUCTION

*“Don't shake your head, point and stare,
at the stumbling man over there.
You think he is drunk and that may be,
but then again it could be me.”*
(Haskins, 1996:1)

MD has been the focus of considerable attention since its initial description by the French otologist Prosper Ménière in 1861. It is the third most common inner ear disorder after presbycusis and noise-induced hearing loss, but it is often a wastebasket diagnosis for conditions of dizziness where the cause is unknown (Hall & Mueller, 1997:714). To guide clinicians (Ear-, Nose- and Throat specialists (ENT), neurologists and other medical personnel) in making an accurate diagnosis, it would be desirable for this disease to have a common base for management as many other specific diseases have. In MD, however, this is complicated by the presently unknown etiology of the disease (Schessel, Minor & Nedzelski, 1998:2677). Despite this there is agreement on the pathogenesis of the symptoms of MD (Paparella, 1985:445).

It is accepted that endolymphatic hydrops is the most significant change in the inner ear in MD, and at present it appears that the pathological mechanism of MD is related to a change in the volume and composition of endolymph. This results in oversecretion or underabsorption of endolymph (Jerger & Jerger, 1981:101). This state in the inner ear is known as

labyrinthine hydrops or endolymphatic hydrops, and affects the ability of sensory hair cells to respond to changes in position, movement and hearing (Li, 2002:2). Endolymphatic hydrops can occur secondary to infection, congenital hypoplasia, trauma, and inflammatory or idiopathic causes (Sperling, Paparella, Yoon & Zelterman, 1993:277). The cause of endolymphatic hydrops in MD is considered to be idiopathic (Jacobson, Newman & Kartush, 1997:339).

Sixty years after the demonstration of endolymphatic hydrops as the pathologic correlate of MD, there is still confusion about its precise role in the pathophysiology of the disease. Endolymphatic hydrops is the most consistent finding in the temporal bones of patients with MD, but it can exist without clinically manifesting the symptoms associated with MD. It seems that symptomatic endolymphatic hydrops is associated with more advanced stages of hydrops generally involving the cochlea and otolithic organs (sacculle and utricle) (Sperling et al., 1993:284).

Despite a wealth of scientific publications, the true nature of this unique cochlear and labyrinthine disorder remains a controversial issue for both diagnosis and treatment (Schessel et al., 1998:2673). As a result of several investigations, researchers have proposed a wide range of etiologies for MD (Dickins & Graham, 1990:55). For such an intractable disease, however, it is possible that an endless chain of unknown etiologies will be postulated even though the cause is occasionally confirmed in some cases (Schessel et al., 1998:2678). Although this reflects a continued lack of understanding, it also suggests that MD may be the common endpoint to a variety of factors influencing the normal function of inner ear structures.

Despite different clinical views regarding the diagnostic protocol used to identify MD, it is generally accepted that the most important diagnostic features of MD are found in the patient's clinical history. This includes a detailed description of the symptom presentation and the pattern of the disease (Schessel et al., 1998:2679). There are two clinical patterns of MD.

The first pattern is the so-called classical or typical MD, which involves both the cochlear (pars inferior) and vestibular (pars superior) labyrinth. By definition, all patients diagnosed with classical MD will experience the symptom triad of recurring episodic attacks of vertigo,

hearing loss and tinnitus (Ginsberg & White, 1994:20). The clinical course of MD, however, is characteristically variable. At the onset, some individuals with MD may experience only one or two of these symptoms. Later on in the course of the disease some of the symptoms such as vertigo and nausea may cease spontaneously (Silverstein, Smouha, & Jones, 1989:6).

The second pattern is referred to as atypical MD where the patient presents with either cochlear symptoms (hearing loss) or vestibular symptoms (vertigo) first, usually to be followed by the complete symptom complex of MD months or even years later (Hall & Mueller, 1997:714). Cochlear MD is thought to be as much as ten times more common than classical MD (Shea, 1975:263). Studies that have followed patients with cochlear and vestibular disease have found that 80 % of patients with cochlear disease develop classical MD, but only 20 % of patients with vestibular disease seem to develop the classical form (Kitahara, Takeda, Yazawa, Matsubara & Kitano, 1984:53; Paparella & Mancini, 1985:149). These atypical forms of MD may confuse the clinical diagnosis and a clear understanding of the different clinical patterns is necessary for an accurate differential diagnosis.

The symptoms, both in classical and atypical MD, may be sudden in onset, with little or no warning, or be preceded by a sensation of fullness in the ear, increasing tinnitus and a decrease in hearing sensitivity (Gibson, 2004:37). Patients may be entirely asymptomatic between the attacks, or some describe periods of dysequilibrium or light-headedness (Paparella, 1984:13). The severity of the symptoms varies. Patients may be minimally inconvenienced or completely incapacitated (Conlon & Gibson, 2000:480). In severe cases, the disease may lead to disability to the extent of curtailment of work, as well as certain social activities (Vesterhauge, 1996:11).

Symptoms such as nausea, vomiting, diarrhoea, and sweating often occur in association with the attacks. Presumably connections exist within the brainstem to link the vestibular, parasympathetic systems and vomiting centres (Barber, 1983:29). These symptoms may cause greater subjective distress than the vertigo itself (Barber, 1983:28). The severity of these symptoms varies both within a given patient in different attacks, and from one patient to the next. Usually, the more severe the vertigo, the greater the accompanying symptoms (Barber, 1983:29).

This variability in the initial presentation of symptoms in patients with MD may lead to uncertainty regarding the diagnosis. The diagnostic process is further complicated by the fact that many of the reported symptoms in MD are also consistent with other diseases (Table 1.1) (Schessel et al., 1998:2679). This is especially true if patients present with atypical forms of MD.

Table 1.1: Disorders to exclude during the diagnosis of MD (Weber & Adkins, 1997:978).

Central	Metabolic	Peripheral
❖ Acoustic neuroma	❖ Diabetes	❖ Benign paroxysmal positional vertigo
❖ Multiple sclerosis	❖ Hyperthyroidism	❖ Labyrinthitis
❖ Vascular loop compression syndrome	❖ Hypothyroidism	❖ Autoimmune inner ear disease
❖ Aneurysm	❖ Syphilis	❖ Perilymphatic fistula
❖ Vascular insufficiency	❖ Cogan's syndrome	❖ Otosclerosis
❖ Arnold-Chiari malformation	❖ Anaemia	❖ Migraine induced vertigo
❖ Cerebellar or brainstem tumours	❖ Autoimmune	
❖ Cervical vertigo		
❖ Transient ischemic attacks		
❖ Cerebrovascular accidents		

As indicated in Table 1.1, clinical investigations should exclude a multitude of central, peripheral and metabolic disorders that affect balance and/or hearing and may masquerade as MD (Li, 2002:9; Weber & Adkins, 1997:978). It is important to note that neurological abnormalities associated with central lesions are absent in MD (Barber, 1983:27). Clinicians must always maintain a high level of suspicion to make this diagnosis. Careful attention to patient history together with diagnostic testing will help to differentiate MD from other medical conditions (Table 1.1).

1.2 PROBLEM STATEMENT AND RATIONALE

Personal clinical experience as well as an extensive review of literature (Ferraro, Best & Arenberg, 1983:77; Ng, Srireddy, CCC-A, Horlbeck & Niparko, 2001:792; Schessel et al., 1998:2673) revealed that the diversified and individual course that MD takes in different patients makes it difficult for medical clinicians to identify the disease on the basis of symptomatology alone. It may take some time before the diagnosis is confirmed and too many patients live with chronic symptoms without knowing the diagnosis. Friberg, Stahle and Svedberg (1984:73) reported that the mean interval between the age at onset and the age at definite diagnosis is three years. The disease itself is unpleasant, but it may be even more stressful for the patient to have no explanation for the symptom. This elevates the level of stress that the patient experiences, affecting quality of life even more (Vesterhauge, 1996:11).

The impact of MD on quality of life has highlighted the importance of quantitative physiological tests as an additional tool to support the diagnosis of MD (Morrison, Moffat & O'Connor, 1980:710). Apart from the patient's history, audiological data provide the most relevant information for confirming the diagnosis of MD. Audiological data also provide insight into the pathogenesis and natural history of the disease (Lee, Paparella, Margolis & Le, 1995:527).

There is to date no single diagnostic test on which a definite diagnosis of MD can be based, and therefore it is important to establish the diagnostic value of different audiological tests that enable clinicians to compile effective test batteries (Schessel et al., 1998:2678). Insight into these tests will allow clinicians to arrive at the final diagnosis more rapidly, resulting in more effective treatment (Arts, Kileny & Telian, 1997:988). Diagnostic features such as laterality of the disease, which is indicated by some audiological tests such as electronystagmography and electrocochleography will influence choices regarding possible surgery (Moffat, Baguley, Harries, Atlas & Lynch, 1992:372). Audiological tests will also assist in the differential diagnosis, where the patient's clinical history mimics that of MD (Schessel et al., 1998:2679). Finally, the use of a reliable objective audiological test battery will enable the critical evaluation of treatment regimens (Arts et al., 1997:987).

The aim of this study is to evaluate the symptoms of MD in a series of subjects in relation to audiological and vestibular tests, in order to develop understanding of the pathophysiology of

the disease and to facilitate the initial diagnosis of MD. Since most research articles focus either on symptomatology or on the use of only one audiometric test this study will focus on the relationship between symptoms and different auditory diagnostic tests. The dynamic nature of MD makes an accurate diagnosis extremely difficult and the inclusion of reliable audiological tests as part of the diagnostic process can assist the clinician in determining appropriate medical and surgical management (Goin, Staller, Asher & Mischke, 1982:1389). To define an audiological approach to investigate MD and facilitate the clinical diagnosis the question remains: What are the clinical and audiological features of MD that provide insight into the nature of the disease and facilitate the diagnostic process?

1.3 TERMINOLOGY

The following terms are defined alphabetically according to their specific use in the study.

❖ Clinician

This refers to an expert clinical practitioner of medicine (*Oxford Paperback Dictionary, 1988:148*).

❖ Diagnostic process

This process is a means of making a statement of the nature of a disease or other condition made after observing its signs and symptoms (*Oxford Paperback Dictionary, 1988:222*).

❖ Differential diagnosis

This involves the diagnosis of a condition where the signs and/or symptoms are shared by various other conditions (*Oxford Dictionary of Nursing, 1996:125*).

❖ Endolymphatic hydrops

This is a pathological state in the inner ear caused by oversecretion or underabsorption of endolymph – the fluid contained within the membranous labyrinth of the inner ear in both the auditory and vestibular portions (Martin, 1994:310).

❖ Etiology

This is a scientific account of the causes of any disease (*Oxford Paperback Dictionary, 1988:12*).

❖ Idiopathic

This refers to a disease or condition where the cause is not known or where the condition arises spontaneously (*Oxford Dictionary of Nursing, 1996:228*).

❖ Ménière's disease

This is an acquired clinical disorder of the inner ear defined as the idiopathic syndrome of endolymphatic hydrops. The symptoms associated with the disease are recurrent episodic vertigo, hearing loss, aural fullness and tinnitus (Monsell et al., 1995:181). Ménière's disease presents in two clinical patterns namely:

- *Classical Ménière's disease*

Patients experience both the cochlear and vestibular symptoms, namely recurring episodic attacks of vertigo, hearing loss and tinnitus (Ginsberg & White, 1994:20).

- *Atypical Ménière's disease*

Patients present with either cochlear symptoms (hearing loss) or vestibular symptoms (vertigo) first, usually to be followed by the complete symptom complex of Ménière's disease months or even years later (Hall & Mueller, 1997:714).

❖ Pathogenesis

This refers to the origin and development of a disease (*Oxford Dictionary of Nursing, 1996:345*).

❖ Vestibulopathy (recurrent)

This refers to an unknown illness with recurrent episodic vertigo characteristic of MD but without auditory or clinical neurologic symptoms or signs (LeLiever & Barber, 1981:1).

1.4 ABBREVIATIONS

❖ ABG: Air-bone gap

❖ ABLB: Alternate binaural loudness balance

- ❖ ABR: Auditory brainstem response
- ❖ AP: Action potential
- ❖ BC: Bone conduction
- ❖ BPPV: Benign paroxysmal positional vertigo
- ❖ BSL: Baseline
- ❖ CM: Cochlear microphonic
- ❖ dB: Decibel
- ❖ DPOAE: Distortion product oto-acoustic emission
- ❖ EcoG: Electrocochleography
- ❖ EM: Effective masking
- ❖ ENG: Electronystagmography
- ❖ ENT Specialist: Ear-, Nose- and Throat Specialist
- ❖ Hz: Hertz
- ❖ IA: Interaural attenuation
- ❖ MD: Ménière's disease
- ❖ NTE: Non-test ear
- ❖ OAE: Otoacoustic emissions
- ❖ OHC: Outer hair cells
- ❖ PB: Phonetically balanced
- ❖ PBHL: Phonetically balanced hearing level
- ❖ PT: Pure tone
- ❖ PTA: Pure tone average
- ❖ PTT: Pure tone threshold
- ❖ RV: Recurrent vestibulopathy
- ❖ SASS: Statistical Analysis System Software
- ❖ SASLHA: South African Speech- Language Hearing Association
- ❖ SD: Speech discrimination
- ❖ SISI: Short increment sensitivity index
- ❖ SFOAE: Stimulus frequency oto-acoustic emission
- ❖ SOAE: Spontaneous oto-acoustic emission
- ❖ SP: Summation potential
- ❖ TE: Test ear
- ❖ TEOAE: Transient evoked oto-acoustic emission

1.5 ORGANISATION OF THE STUDY

The aim, results as well as the implication of the results of this study will be presented in the following chapters:

Chapter 1: Introduction

In this chapter the relevance of the research is outlined, and the reader is provided with background information leading to the specific research problem. The rationale of the study together with the problem statement is provided. Concepts related to the study are defined and an outline of the study is presented. This chapter is important in order to orientate the reader regarding the purpose of the study as well as the background upon which it is based.

Chapter 2: Perspective on the nature of Ménière's disease

This chapter presents a review of the literature and a discussion of theoretical underpinnings related to MD. Existing knowledge and research regarding the disease is presented critically. Current audiological assessment for the diagnosis of MD as well as the audiological profile of these patients is reviewed. This chapter aims to develop an understanding in terms of the nature of the disease as well as the importance of a reliable objective audiological test battery.

Chapter 3: Methodology

In this chapter the main aim and sub-aims of this research project are clarified. The research design is discussed and motivated from sources in the literature. The sample as well as the selection process is described. This chapter includes a description of apparatus and material used for participant selection, data collection as well as data analysis and processing. Data collection, recording and analysis procedures used in this study are also discussed. A description of the research methodology allows for understanding of the nature of the study and also allows for the study to be repeated by other researchers in exactly the same way.

Chapter 4: Results and discussion

All the collected and processed data are discussed, supported by figures and tables according to the nature of each question identified in the sub-aims. This makes trends and features of

observed data easy to understand and interpret (Johnson, 1994:15). Explanations are given if atypical results occur. The importance of this chapter is to provide answers to the research problems presented in this study.

Chapter 5: Conclusions and recommendations

The conclusions drawn from the results according to each sub-aim as well as the answers to the research problem are presented in this chapter. The implications of the research results are also addressed. A critical evaluation of the strengths and limitations in this research project forms part of this chapter, to provide valuable information for future research in this field. The statistical analysis of data allows certain dynamic and potential forces to identify areas that warrant future research according to which recommendations for future research are made (Neuman, 1997:295).

References

A complete list of all the references used in the thesis is presented in alphabetical order.

Appendices

Following the thesis, supplementary material valuable to understand the text more completely is included.

1.6 SUMMARY

MD was first identified over a century ago as an acquired disorder of the inner ear. There has been considerable research into this complex disease, but despite the large number of scientific contributions published, the pathophysiology and histopathology of the condition are still not completely understood. Disagreement and controversy regarding both diagnosis and therapy continue to surround the subject (Schessel et al., 1998:2681). Although clinical evaluation is improving, there is still no uniform agreement on the underlying causes of MD, and it is quite difficult to determine its origin (Schessel et al., 1998:2677). There is no reason for the diagnosis of MD to be a dumping ground for dizziness where the diagnosis is uncertain. Effective patient management requires a better understanding of the physiological

nature of the disease, as well the relationship that exists between the clinical and audiological features of the disease. The development of a generally acceptable plan for diagnosis and treatment poses a challenge for clinicians involved in the care of this condition. Research focusing on the relationship between the patient history and audiological features will facilitate the differential diagnosis of MD. Therefore, we must constantly update our knowledge through research for the benefit of our patients (McCartney, 2002:1). With increased understanding of the pathophysiology of MD patient management will become more effective.

CHAPTER 2

PERSPECTIVES ON THE NATURE OF MD

The aim of this chapter is to present a review of the literature as well as a discussion of concepts related to the nature of MD. This chapter will focus on both the symptom complex and the different audiometric tests, as well as the clinical value of each of these tests in identifying MD. It will highlight controversy that exists regarding diagnosis and treatment. Although treatment is not the focus of this study, different treatment approaches will be mentioned, since there is no universally accepted medical or surgical treatment for MD at this time (Vesterhauge, 1996:11). Efficient treatment depends on the correct diagnosis, again highlighting the importance of understanding the symptom complex of the disease (Schessel et al., 1998:2681).

2.1 INTRODUCTION

MD is an acquired clinical disorder of the inner ear, referred to as the idiopathic syndrome of endolymphatic hydrops (Monsell et al., 1995:181). The disease is associated with a symptom complex consisting of attacks of episodic vertigo, sensorineural hearing loss that may fluctuate, tinnitus and often a sensation of aural fullness (Ginsberg & White, 1994:20). Despite the fact that the symptom complex is well known, it remains a controversial and often a difficult disease to diagnose, which leads to difficulty in optimal management of the disease (Schessel et al., 1998:2673).

2.2 INCIDENCE OF MD

MD predominantly affects adults from the fourth decade of life, although a few cases in children have been reported (Paparella & Sajjadi, 1999:30). The difficulty in clearly diagnosing patients leads to a corresponding difficulty in determining the incidence of the disease. This is evident in the variation that exists in published reports regarding the incidence of MD (157 MD patients per 100 000 in England, 46 MD patients per 100 000 in Sweden and 7.5 per 100 000 in France) (Schessel et al., 1998:2673). Such figures are difficult

to analyse, because several variables may affect the results, such as differences in access to health care in the different countries, differences in diagnostic criteria, differences in data gathering techniques, as well as true epidemiologic differences among people around the world (Dickins & Graham, 1990:51). Genetic and environmental factors probably influence differences in incidence among countries and sections of countries (Schessel et al., 1998:2674). Despite a thorough investigation the author could find no reported figures for Africa or South Africa, emphasizing the need for this study.

A **sexual preponderance** of the disease has not been identified. Although several reports suggest opposing conclusions, it is mostly accepted that men and women are equally affected or there may be a slight bias toward female patients (Martin, 1994:311; Paparella, 1985:446; Watanabe, 1981:511). There is great variation regarding **familial occurrence**, but it is estimated that 10 % to 20 % of patients have a family history of MD (Paparella, 1985:447).

Issues related to the **laterality** of the disease are unclear. Paparella and Griebie (1984:233) found that nearly half the patients diagnosed as having unilateral MD developed bilateral symptoms after five years. Balkany, Sires and Arenberg (1980:605) found that 25 % of patients suffering from bilateral disease have the same severity of disease in both ears. In cases where the one ear was more affected than the other, they reported the right ear to be affected in 53 % of cases. The reported variance of bilateral involvement is between 6 % and 78 % or an average of 42 % (Balkany et al., 1980:603; Friberg et al., 1984:73; Wada et al., 1990:118). A low incidence of bilaterality may be accounted for by a number of factors. One explanation might be that patients who suffer from bilateral involvement are often unaware of a slight deafness or symptoms such as tinnitus in the contralateral ear. It is probable that studies showing a higher incidence of bilateral involvement are based on increased awareness of the possibility of bilateral disease, earlier detection of manifestations of the disease, and longer follow-up (Balkany et al., 1980:606). An extremely high incidence of bilaterality must also be viewed with caution, because hearing loss and tinnitus may be caused by other conditions apart from MD (Paparella & Griebie, 1984:233). This big difference in reported variance for the same disease process also strongly suggests the need for stricter diagnostic criteria. Without uniform criteria a valid assessment of laterality as part of the natural history of the disease is impossible (Balkany et al., 1980:606). Much research regarding the frequency of bilateral disease still needs to be conducted especially on the long-term course of the disease (Kitahara, 1991:74).

2.3 ETIOLOGY OF MD

The hallmark of MD at a histopathological level is the presence of endolymphatic hydrops within the inner ear. Although many possible etiologic factors can lead to endolymphatic hydrops, it is the hydrops that generate the symptoms of MD (Merchant, Adams & Nadol, 2005:1).

Obstruction of the endolymphatic duct is the basis for development of hydrops in experimental animals (Schessel, Minor & Nedzelski, 2005:3215). This is accomplished by any lesion that can produce failure of duct function including mechanical blockage, chemical fibrosis of the endolymphatic duct, viral inoculation of the endolymphatic sac, immunologically induced inflammation, and ischemia induced by mechanical obstruction of the sac vasculature (Merchant et al., 2005:1). Animal models only serve as histologic equivalents and cannot be interpreted to reflect the actual cause of the disease in humans, but the animal models do demonstrate that virtually any insult that results in failure of the endolymphatic absorption system can produce hydrops. Autoimmune processes have been suggested as etiologic and may also be related to the familial or genetic occurrence of the disease (Schessel et al., 2005:3216). Viral infection such as herpes simplex virus type, ischemia of the endolymphatic sac and the inner ear as well as a common vascular mechanism for migraine headaches and MD have also been proposed as possible etiologies. The recent discovery of a mutation in a brain calcium-channel gene in families with hemiplegic migraine and in families with episodic vertigo and ataxia suggests a possible mechanism for neurotologic symptoms in patients with more common varieties of migraine. A defective calcium channel, primarily expressed in the brain and inner ear, could lead to reversible hair cell depolarisation and auditory and vestibular symptoms similar to those found in MD (Baloh, 1997:615).

A number of processes seem to be associated with the development of hydrops. It is possible that MD may be multifactorial and precipitated by a variety of events such as autoimmune, viral, traumatic, vascular, endocrine and even congenital anatomic and molecular variations (Schessel et al., 2005:3216).

2.4 CHARACTERISTICS AND SYMPTOMS OF MD

The diagnosis of MD is based primarily on patient history. This includes a detailed description of the clinical presentation of the disease related to the triad symptom complex of vertigo, hearing loss and tinnitus (Schessel et al., 1998:2679). It is therefore important for the clinician to be familiar with the characteristics of MD (Barber, 1983:27).

2.4.1 Non-auditory characteristics and symptoms of MD

The most characteristic symptom and distressing complaint of patients with MD are the repeated attacks of incapacitating vertigo with intervals of complete freedom from dizziness between attacks (Paparella, 1984:13). Vertigo is the illusion of movement, either of oneself or of the environment, and is quite distinct from mere dizziness, unsteadiness or giddiness (Martin, 1994:321). True vertigo is an unpleasant sensation of imbalance, spinning and disorientation (Barber, 1983:26). Very typical of the vertigo experienced in MD is that it may occur at any time, unrelated to position or activity (Paparella, 1984:13). The attacks of vertigo may even waken the patient from sleep, unlike psychogenic vertigo (anxiety attacks) that may mimic the vertigo of MD (Barber, 1983:27). It is important for clinicians to document the duration of the individual attacks of vertigo, the frequency, effect of head movement, associated aural symptoms as well as associated ear disease in order to make a differential diagnosis (Schessel et al., 1998:2673).

MD is the most common cause of peripheral vertigo and as is typical with peripheral vestibular dysfunction, the symptoms get worse with any head movement (Schessel et al., 1998:2679). Typical attacks last for approximately two to three hours, but some patients have attacks of shorter or longer duration (Oosterveld, 1980:887). If the dizziness lasts for only a few seconds, or more than three days it is not believed to be MD (Vesterhauge, 1996:4). Symptoms of dysequilibrium may, however, persist for several days after the vertigo has resolved and for this reason it is relatively easy for patients to define when an attack starts, but not so easy to describe when it ends (Vesterhauge, 1996:4). Attacks vary in severity; the intensity of this symptom depends on the extent of disturbance of afferent vestibular discharge from the affected ear. A marked reduction of discharge activity causes severe vertigo, and less marked, milder vertigo (Barber, 1983:26).

During the attack there may be involuntary jerking eye movements (nystagmus), which are caused by a reflex (vestibulo-ocular reflex) from the balance organ in the inner ear (Shepard & Telian, 1994:426). Nystagmus always occurs with vertigo whether one can see it with the naked eye or not (Martin 1994:282). According to several researchers, rotatory nystagmus, as found in benign paroxysmal positional vertigo (BPPV), is uncommon in MD (Barber, 1983:26; Schessel et al., 1998:2688). Paparella and Mancini (1983:1011), however, found that positional vertigo was a prominent complaint especially in patients with vestibular MD, where positional vertigo presented as a severe problem between typical Ménière vertigo attacks (Paparella & Mancini, 1983:1011). Benign paroxysmal positional vertigo (BPPV) originates from the posterior semicircular canal and is a common vestibular disorder characterized by sudden onset of intense vertigo triggered in certain head positions (Katsarkas 1991:193). A latency of five to twenty seconds precedes the onset of transient vertigo that quickly increases in intensity and fatigues on repeated position changes (Gross, Ress, Viirre, Nelson & Harris, 2000:655). These episodes usually last for a few seconds and are accompanied by a sensation of rotation, postural instability, or occasional falling, nausea or even vomiting, and nystagmus. In a retrospective review of a large population of patients with BPPV, a high rate of association between MD and BPPV has recently been found (Karlberg, Hall, Quickert, Hindson & Halmagyi, 2000:380). BPPV developed in the Ménière's affected ear after the onset of endolymphatic hydrops in the majority of these patients. In view of these recent results, it is important to investigate this phenomenon (BPPV) in patients with MD.

Visual blurring may occur with active nystagmus in a severe attack, but it never occurs between spells (Barber, 1983:27). Some patients also experience the Tullio phenomenon. This term is applied to vertigo induced by intense sound stimulation (Ballantyne & Groves, 1979:572). There is a tendency for the attacks of dizziness to become less frequent and less severe over time, leaving hearing loss and tinnitus as the main symptoms of complaint for these patients (Vesterhauge, 1996:7).

2.4.2 Auditory characteristics and symptoms of MD

MD is characterized by progressive sensorineural hearing loss, usually with a history of fluctuation, tinnitus, loudness intolerance, diplacusis, and aural pressure (Lee et al., 1995:527).

2.4.2.1 Sensorineural hearing loss

The deterioration of hearing in patients with MD is known to be highly variable and dependent on the individual. In most cases, however, it leads to moderate hearing impairment (Kotimaki, Sorri & Muhli, 2001:14). In the early stages of the disease, most patients with MD suffer from hearing loss only during the attacks of dizziness. After a few attacks, however, patients also start to experience loss of hearing between attacks (Vesterhauge, 1996:6). Although sudden sensorineural hearing loss has been reported, acute attacks usually lead to a gradual deterioration in sensorineural hearing in the affected ear (Selmani, Timo, Marttila, Pyykko & Ishizaki, 2002:106). A sudden sensorineural hearing loss that responds to medication should not be considered to be MD (Monsell et al., 1995:182).

The sensorineural hearing loss associated with MD is usually fluctuating and progressive (Schessel et al., 1998:2680). According to Sakurai, Yamane and Nakai (1991:92) as well as Schessel et al. (1998:2680) fluctuation in hearing thresholds are especially found in patients whose audiograms are characterized by low-frequency sensorineural hearing loss. Although fluctuation of hearing is particularly noticeable early in the course of the disease, it can also be present even when the hearing loss is more severe (Barber, 1983:28). The hearing loss often occurs together with aural pressure or fullness in the affected ear (Schessel et al., 1998:2680). Patients who have had the disease for quite some time may develop rather significant hearing loss, which affects quality of life (Vesterhauge, 1996:6). The pace at which hearing deteriorates is more rapid in the first five years of the disease than later, and the hearing level then tends to stabilize leaving the patient with a moderate to severe sensorineural hearing loss (Barber, 1983:28). Greater losses may certainly occur, but patients with MD rarely become profoundly deaf (Schessel et al., 1998:2680). Only one to two percent of severely affected patients demonstrate a profound sensorineural hearing loss (Stahle, 1976:113).

Many patients do not understand the change in their hearing function. On the one hand, it is obvious to them that a hearing loss is present, since there is a reduced ability to discriminate and understand speech. The patient may, however, also experience recruitment, which makes them oversensitive to loud sounds and noise. They may also report the presence of diplacusis, which is a difference in the perception of pitch between the affected and non-affected ear

(Paparella, 1985:447). At the same time, or soon after hearing loss occurs, patients may start to complain of tinnitus in the affected ear (Barber, 1983:28).

2.4.2.2 Tinnitus

All sounds in the ear that do not originate from an outside sound source in the surrounding environment are referred to as tinnitus (Jastreboff, Gray & Mattox, 1998:3198). Patients will have different descriptions for this sound, but it is most often referred to as a buzzing, roaring, hissing, or ringing sound in either the ear or the head (Martin, 1994:310). In MD tinnitus tends to be a non-pulsating low-pitched whistling, roar or buzz (Barber, 1983:28). Oliveira, Bezerra and Araújo (1998:132) found that 79 % of patients with MD described the sound as a white noise. It may be continuous or intermittent. Tinnitus often begins, gets louder, or changes pitch as an attack approaches. After the attack there is frequently a period of improvement and relief from the tinnitus (Jerger & Jerger, 1981:101).

Some patients' complaints of tinnitus are much more severe than the complaints of others, even though the hearing loss is similar (Martin, 1994:454). Schechter, McDermott and Fausti (1992:34) explain this by comparing tinnitus with the experience of pain. They suggest that given a certain amount of neural activity stimulating the pain centres of the brain of two individuals, the experiences might be quite different. It is modified by higher brain centres and by such factors as mood, personality and culture. They also suggest that the effects of tinnitus on any individual's personal life may, at least in some cases, be influenced by the person's psychological state. According to Lebisch (2001:23) there is a chain reaction that exists between perception, emotion and attention. Stress, over-exertion as well as the amount of attention you give to the tinnitus will have a direct impact on the patient's perception of the tinnitus. Interviews with individuals who suffer from tinnitus show that in most cases stress, internal apprehension, subconscious conflicts, problems with spouse or partners or a mild depression were at the onset of tinnitus, not implying such were causative, but certainly contributing factors (Lebisch, 2001:24). Given the interactions between the often-unknown physiological base of tinnitus and the psychological reaction to it, the phenomenon of tinnitus continues to be very complex. In the long run, tinnitus is very often the symptom that causes the Ménière patient the most discomfort (Vesterhauge, 1996:7).

2.4.3 Course and development of MD

Once begun, MD pursues an unpredictable course marked by periods of exacerbation and remission. Periods of presenting symptoms (blocked ear, hearing loss, tinnitus, vertiginous attacks) are mysteriously followed by remission. The periods of active illness are likely to last for weeks or months, while periods of remission is often months or years, sometimes many years (Barber, 1983:31). Exacerbation of symptoms may occur in some individuals under conditions of stress (Jerger & Jerger, 1981:102). Early in the illness, hearing may return to normal and tinnitus disappears during remission, but later, significant and disabling hearing loss and tinnitus persist even when vertigo is absent. Vertiginous attacks may then still occur, but for the most part with lesser frequency than in the first five years. At this stage, nausea and vomiting are less prominent than earlier. According to Barber (1983:31) spontaneous cure, defined as absence of any of the three of its symptoms for at least ten years, is extraordinarily rare.

2.4.4 Emotional impact of MD

Apart from the incapacitating effects of the physical manifestations of MD, it is also reported that the disease is emotionally disabling (Barber, 1983:29). Patients experiencing the symptoms associated with MD are often quite worried and disturbed. They feel a sense of loss of control over their lives and this leads to a certain degree of uncertainty and instability in their psychological well-being (Paparella & Sajjadi, 1999:36). Interference in sleep pattern, change in social contact and performance at work can lead to significant anxiety and depression (Vesterhauge, 1996:11). During an attack of MD it may seem as if the person is drunk and it can be difficult to explain to employees, family and friends that it is a disease like so many others, brought about through no fault of one's own. Kinney, Sandridge and Newman (1997:67) reported that patients associated the physically disabling effects of MD (vertigo, hearing loss, tinnitus and aural fullness) as to be similar to that of a relatively minor medical problem. However, the emotional impact of the disease in the same patients was equivalent to that of a major medical problem.

2.5 DIAGNOSIS OF MD

The dramatic variability and protean manifestations of this disease can make it difficult to diagnose MD on the patient's history alone. Clinicians need to make the diagnosis of MD as early as possible to ensure the psychological and physical well-being of their patients. Both patient history and audiological tests are necessary to develop insight into the nature of the disease and establish effective patient management. This emphasizes the need for quantitative tests to support the clinical diagnosis.

As discussed earlier, endolymphatic hydrops has been a consistent finding in the temporal bones of patients with MD (Paparella, 1991:27). Unfortunately, there is no way to confirm the presence of hydrops other than by histologic evaluation of the labyrinth at autopsy. Most of the symptom complex is subjective in nature, rendering critical analysis problematic. The one objective finding, sensorineural hearing loss, is non-specific since it could be the result of other diseases related to the inner ear. Therefore, literature concerning the pathophysiology, diagnosis, clinical course, and treatment of endolymphatic hydrops abounds with controversy and speculation (Schessel et al., 1998:3210). For many reasons, it would be desirable to have an objective, premortem clinical test for the presence of hydrops. Numerous difficult treatment decisions would be simplified by such a test. Even if one could reliably depend on the presence of hydrops in patients with the classic symptom complex, it would be reassuring to confirm the diagnosis prior to destructive surgery such as labyrinthectomy (Paparella & Sajjadi, 1999:36). Additionally, it would be quite useful to be able to exclude the possibility of contralateral disease. Frequently patients present with only portions of the classic symptom complex, such as in vestibular or cochlear MD, and in these patients confirmation of the presence of hydrops would be beneficial. Particularly in patients with presumed vestibular MD, such a test would enable the clinician to lateralise the pathology to one ear that would in turn enable the use of potentially destructive therapy in that ear. Few other diseases have defied attempts at rigorous clinical testing as have MD. Even the best tests yield positive results in only about two thirds of patients with classic MD. MD is characteristically associated with frequent fluctuations in severity as well as spontaneous remissions. These facts seriously confound attempts to assess the efficacy of any clinical test or treatment modality and emphasize the importance of applying a test battery approach (Arts et al., 1997:987). Diagnostic testing for endolymphatic hydrops can be divided into non-specific tests and those thought to be specific.

In the non-specific category, the audiological tests most commonly used in establishing the diagnosis are pure tone (PT) audiometry (air- and bone conduction), speech discrimination (SD) scores, oto-acoustic emissions (OAEs) and electronystagmography (ENG). Specialized tests such as the alternate binaural loudness balance test (ABLB), short increment sensitivity index (SISI) and the immittance test may be used to describe diseased otologic hair cells. Auditory brainstem response (ABR) and immittance testing (Metz- and tone decay tests) may be performed to confirm the site of lesion (Ginsberg & White, 1994:20; Schessel et al., 2005:3218).

Specific tests for detection of endolymphatic hydrops include electrocochleography (EcoG) and dehydrating agents such as glycerol in combination with audiological tests. The purpose of dehydrating agents is to reduce the endolymphatic pressure in the inner ear and produce a measurable change, that is, improvement in pure tone thresholds (PTTs), reduction in summing potential negativity (as recorded with EcoG) and a change in the gain of the vestibule-ocular response to rotational stimulation. Critics of the glycerol test note that the test is unpleasant, possibly dangerous to patients, insensitive, impractical and subject to significant placebo effects. The reported sensitivity and specificity of the test varies widely and at this point, the clinical use of dehydration testing is unclear (Arts et al., 1997:992).

The clinical challenge is to know when a patient requires more than the basic hearing test battery, and which procedures will best define the patient's auditory status. In this era of health care cost containment, this entire process is a balancing act in which the consequences of not enough diagnostic assessment (specialists fail to identify serious auditory dysfunction) are weighed against the consequence of performing unnecessary procedures (additional costs to patient or third party payer) (Hall & Mueller, 1997:565).

The diagnostic audiometric test battery has evolved considerably since the traditional site-of-lesion test battery of the 1960s. Tests such as the SISI, ABLB, tone decay tests, and Békésy audiometry were performed, analysed, and interpreted in an attempt to differentiate cochlear versus retrocochlear auditory dysfunction accurately. Today, instead a variety of other procedures such as ABR and other evoked responses, OAEs and behavioural measures of central auditory function are performed. The goal remains essentially the same, namely the description of the site of auditory dysfunction (Hall & Mueller, 1997:565).

2.5.1 Audiometric tests

Recent areas of interest in diagnosis of MD have focused on auditory and vestibular test results and relating them to pathophysiology of the disease. Correct diagnosis of the disease will be assisted by a detailed description of the case history, physical examination and audiological diagnostic test results (Dickins & Graham, 1990:57). Audiometric data should include routine measurement and documentation of PTTs with its audiometric configuration and measurement of SD. A number of audiological tests, for example tests for recruitment and determination of site of lesions, that were extensively employed in the past, are less often used today. They have been replaced by more accurate alternative tests such as magnetic resonance imaging and EcoG (Lee et al., 1995:527).

2.5.1.1 PT audiometry results

Sensorineural hearing loss is a diagnostic feature of MD (Ginsberg & White, 1994:20). The hearing loss is usually of moderate severity (Kotimaki et al., 2001:14). There is, however, great debate regarding the audiogram configuration typically associated with MD. Elichar, Keels and Wolfson (1973:41) found that flat or rising curves with predominantly low-tone hearing loss are the two most common configurations. On the other hand, Goodman (1965:992) thought that configurations were characterized by peaks and dips, while rising, flat or falling curves were relatively infrequent. Barber (1983:27) found two audiometric configurations to be most common. The first is a low-frequency sensorineural hearing loss involving especially 250 and 500 Hertz (Hz), with frequencies above 1000 Hz relatively normal. The second is an inverted “V” curve also described by Paparella, McDermott and deSousa (1982:558). This peak configuration pattern involves a low-frequency fluctuating hearing loss, together with a coincident non-changing, high-frequency loss, with the peak classically at 2 kHz. Lee et al. (1995:527) also suggested this type of configuration to be a diagnostic feature of MD. Despite the different views regarding initial audiogram configuration it seems that peaked and low-frequency patterns are most prevalent. It is generally accepted that over time the hearing loss flattens and stabilizes, leaving the entire frequency range affected (Barber, 1983:28).

Despite different views regarding audiogram configuration associated with MD there are certain shapes more commonly found in the disease. It is also important to document reported

hearing loss, since patients may confuse a sensation of ear blocking (aural pressure) with hearing loss as found in vestibular MD or Eustachian tube dysfunction (Barber, 1983:27). PT audiometry is valuable in the differential diagnosis and helps to exclude other causes such as total or unilateral hearing loss which could arouse suspicion of another active process, such as acoustic neuroma (Barber, 1983:28). PT audiometry results will also help to distinguish between different forms of MD by revealing the presence or absence of cochlear involvement (Hall & Meuller, 1997:715).

2.5.1.2 SD results

SD is often normal early in MD, but as the hearing thresholds deteriorate, there is progressive decline in the patient's ability to discriminate words (Ginsberg & White, 1994:20). Mateijsen, Van Hengel, Van Huffelen, Wit and Albers (2001:384) found that although SD scores are in keeping with the degree of sensorineural hearing loss, the maximum SD score decreases as hearing loss increases. They found almost 100 % SD in patients with PTTs up to 40 dB. For larger losses, the maximum discrimination decreased. They concluded that although the degree of the hearing loss had an effect on SD results, the audiogram shape did not have a significant effect on SD scores.

2.5.1.3 OAE testing

OAEs are defined as low-level acoustic energy generated within the cochlea, either spontaneously or on response to acoustic stimulation, and recorded in the outer ear canal (Norton & Stover, 1994:448). OAEs are an objective, non-invasive and quantitative measure of sensory cell function in the cochlea. It provides information regarding outer hair cell activity, and is regarded as a reliable test for determining the structural basis of a hearing problem, particularly if it involves the outer hair cell system (Cianfrone, Ralli, Fabbricatore, Altissimi & Nola, 2000:111).

Four types of OAEs have been measured, namely spontaneous (SOAE), transient evoked (TEOAE), distortion product (DPOAE), and stimulus frequency (SFOAE) (DeVries & Decker, 1992:15). The clinical value of SOAEs and SFOAEs appears to remain in question, but TEOAEs and DPOAEs are proving to be useful clinical techniques to measure the response of the outer hair cells (OHC) and the status of the cochlear efferent system

(Cianfrone et al., 2000:111). DPOAEs are relatively narrow-band responses and considered to be the most valuable OAE for investigating frequency-related phenomena and for correlating audiometric findings (Cianfrone et al., 112). DPOAEs as well as other OAEs (transient OAEs and spontaneous OAEs) show sensitivity in detecting early cochlear dysfunction since they can be evoked by low and moderate level stimuli. Detection of these emissions offers a rapid, non-invasive means of screening for hearing disorders as well as assessing cochlear (dys)function (Sakashita et al., 1998:70).

In the clinical application of DPOAEs, two types of test protocols have commonly been used. The protocol more frequently used is referred to as the “DP-gram” in which DPOAE amplitudes for a single primary intensity are plotted as a function of frequency of the primary tones. The other is measurement of the growth function, which plots DPOAE amplitudes as a function of intensity of the primary tones (Sakashita et al., 1998:70). DPAOEs are sensitive to cochlear pathology such as MD and therefore research has indicated that in most ears with MD DPOAE amplitudes are reduced in the presence of elevated behavioural thresholds indicating affected OHC (Cianfrone et al., 2000:112). Changes in the activity of OHC following glycerol ingestion can also be verified with DPOAE measurements. Concerning monitoring of cochlear function in the glycerol test, it has been reported that DPOAE amplitudes increase with hearing improvement following glycerol ingestion for diagnostic purposes in patients with MD (Cianfrone et al., 2000:111). Changes in cochlear function may even be observed in the absence of detectable changes in behavioural audiometry (Sakashita et al., 1998:70). These findings have been reported in case studies, and results of large-scale investigations on the usefulness of DPOAE measurement in the glycerol test are clear (Cianfrone et al., 2000:112).

2.5.1.4 ENG

A test for vestibular function should always be carried out when MD is suspected, as it may give the only physical sign of a disordered vestibular labyrinth. ENG is a method to electrically monitor the amount of nystagmus occurring spontaneously or from caloric stimulation (Martin, 1994:319). Electro-oculographic recording of eye movements after caloric and rotational stimulation constitutes the most reliable method of assessing vestibular function (Shepard & Telian, 1994:431).

Spontaneous, eyes closed, unprovoked eye movements (spontaneous nystagmus) and eye movements provoked by changes in the orientation of the vestibular end organs relative to gravity (positional nystagmus) are recorded (Shepard & Telian, 1994:432). Rapid positioning into specific head positions (Hallpike maneuver) is performed to provide evidence for one specific condition – BPPV. A measure of the responsiveness of one horizontal semicircular canal relative to the other is typically performed through thermal caloric irrigations. Essentially, the caloric test measures the performance of the lateral semicircular canal and the cristo-ocular reflex. The caloric test remains the most useful test of the integrity of an individual labyrinth and is the most reliable for determining the affected ear (Rizvi, 1986:1262). Various studies show a significant caloric response reduction of 50 % to 74 % in patients with MD (Black & Kitch, 1980:636; Jacobson et al., 1997:197). Complete loss of vestibular function, as elicited by the caloric test, is a relatively rare occurrence and only reported in 6 % to 11 % of patients with MD (Jacobson et al., 1997:197; Schessel et al., 1998:2680). It occasionally happens that the normal ear, or the ear with better hearing, reveals abnormal results. It is important to be aware that the two ears may have dyssynchronous malfunction. One ear may have fluctuating hearing levels with relatively stable vestibular function, while the other ear has normal stable hearing with wildly fluctuating vestibular dysfunction. The balance dysfunction may have its origin in the ear with normal hearing (most of the time), yet the ear displaying hearing loss may be blamed for the entire set of clinical problems (Balkany et al., 1980:607). In such cases further careful enquiry may reveal former attacks that could suggest bilateral disease (Cawthorne & Hewlett, 1954:25).

ENG allows for objective measurement of vestibular function, differentiation between peripheral and central pathologies and elimination of central suppression. ENG is well tolerated by patients, monitors patient progress and allows accurate measurement of slow phase nystagmus (Shepard & Telian, 1994:431). Unfortunately only eye movement greater than one to two degrees can be measured, torsional movements cannot be measured and eyelid closure artifacts can interfere with the recording of reliable results (Marsh, Zane & Jenkins, 1991:451). ENG results cannot differentiate among involvement of the labyrinth, cranial nerve VIII, or the vestibular nucleus and does not provide for testing of the capacity for integration of all sensory input information used by the balance system. Experience and formally acquired knowledge is also necessary to obtain reliable results and diagnostic

interpretation thereof (Shepard & Telian, 1994:433). Understanding the limitations of ENG interpretation significantly improves the appropriate use of this tool.

2.5.1.5 EcoG

EcoG refers to the method of measuring stimulus-related electrophysiologic potentials of the cochlea and auditory nerve (Ruth, 1994:339). Tympanic EcoG is a non-invasive technique for recording cochlear potentials with an electrode placed on the tympanic membrane. The stimulus related electrical potentials associated with this part of the auditory pathway include the cochlear microphonic (CM), summing potential (SP) and the compound action potential (AP) of the auditory nerve. These potentials arise from the hair cells and the first order neurons in the inner ear (Levine, Margolis, Fournier & Winzenburg, 1992:614). This test is employed to evaluate cochlear function in patients with symptoms associated with MD.

A number of studies have demonstrated distinctive EcoG patterns in patients with MD as compared to normal ears. This difference is primarily in the form of an enlarged SP amplitude relative to the AP amplitude (Schessel et al., 1998:2680). The absolute amplitude of the SP and AP both show considerable variability across subjects and therefore a more consistent amplitude feature is the SP-AP amplitude ratio (Ruth, 1994:345). It has been postulated that an increase in endolymphatic volume alters the mechanical characteristics of the basilar membrane motion. Inasmuch as the vibratory asymmetry of the basilar membrane is thought to cause the SP, enhancement of this asymmetry by hydrops could explain the enlarged SP seen in MD (Ruth, 1994:340). An increase in the SP amplitude in relation to the AP amplitude has been reported in 62 % of patients diagnosed with MD (Schessel et al., 1998:2680). The reason why all patients with MD do not demonstrate abnormal EcoG results is unclear. It may reflect the fluctuant nature of the disorder or the deterioration of outer hair cells known to occur in more advanced stages of MD. The limitations associated with EcoG are firstly that full cooperation from the patient is mandatory for obtaining reliable tracings. It is important for the patient to remain still during testing, especially with respect to movement of the head and jaw. Slight movement or displacement of the recording electrode can have dramatic effects on the amplitude of the EcoG components (Ferraro et al., 1983:78). In addition, background noise level and impedance-matching difficulties are encountered much more frequently with EcoG than with other auditory evoked potential tests. In general, the EcoG clinician will require considerable technical and scientific expertise in order to

recognize and correct for these problems and to provide an adequate interpretation of the overall response. Finally, reliable click-evoked EcoG responses cannot be recorded in patients with severe to profound hearing losses (Ferraro et al., 1983:79).

Despite these limitations, EcoG remains a valuable test in the diagnosis of MD. At present it is the only proven investigation that can demonstrate objectively the presence of endolymphatic hydrops, which is the underlying pathologic finding in this disease (Moffat et al., 1992:370).

2.5.2 Clinical application of audiometric tests

The audiological tests discussed above can provide valuable information regarding the diagnosis, treatment and monitoring of patients with MD (Ferraro et al., 1983:77). It is valuable to use these tests as part of a test battery and not to rely on the results from one single test only (Ginsberg & White, 1994:23). The challenge is that not all audiologists have access to specialized equipment such as ENG and EcoG, and not all have received adequate training to perform these tests. These tests are also expensive and not accessible to all patients (Hall & Meuller, 1997:315).

2.6 MANAGEMENT OF MD

The ideal treatment of any disease is to remove the cause of the disease. Unfortunately it is quite rare to find the cause of MD in an individual patient (Vesterhauge, 1996:11). Due to the variability of etiology, diagnosis and course of MD, treatment is usually symptomatic and not always successful (Schessel et al., 1998:2681).

Although there is currently no known medicine or operation that can safely and permanently cure MD, there is still much that clinicians can do to help patients. Whenever a disease has an unknown cause, there are many different approaches to its treatment and each approach has its own proponents. Many treatment modalities have been proposed for patients with MD, but because of its extreme clinical variability, difficulty exists in quantifying treatment effectiveness. Most treatments, however, aim at reducing the pressure of the endolymph in the inner ear. Current therapy is aimed at the reduction or elimination of associated symptoms experienced by the patient with MD. Both cochlear components (hearing loss,

tinnitus and aural pressure) and vestibular symptoms (vertigo, imbalance and disability) must be treated. MD can be progressive (become severely intractable and disabling), or remain non-progressive (intractable vertigo or severe deafness may never arise) and medical management varies accordingly. In progressive MD where the hearing deteriorates, patients need more aggressive medical therapy and surgical therapy may be offered earlier. With non-progressive MD the treatment can be less aggressive (Li, 2002:3).

Since vertigo is typically the most debilitating symptom in the early stages of MD, the most important symptom to target is almost always the attacks of dizziness. Patients are understandably terrified and frightened, therefore recurrent vertigo can be devastating to the patient's well-being. Currently almost all proven therapy is directed at relieving vertigo (Schessel et al., 1998:2681). In a review of the available treatments for those with vertigo associated with MD, Torok (1977:1870) found that all treatments, medical and surgical, reported significant improvement of vertigo in 60 % to 80 % of patients. Treatment may include a combination of surgery, medication, change in diet and behavioural therapies.

2.6.1 Surgical treatment

Surgical treatment should be managed by an experienced ENT. Surgery is reserved for those patients who have failed prolonged conservative medical management and is estimated to be necessary in 10 % of patients (Schessel et al., 1998:2681). Several types of surgery are effective for treating the balance problems resulting from MD. Surgical techniques applied can either preserve existing hearing thresholds (vestibular neurectomy and endolymphatic sac decompression) or be destructive of hearing (labyrinthectomy). Taking into consideration the fact that it is not a life-threatening situation in the majority of patients, in whom it can be controlled with a combination of medical therapy and dietary control, aggressive and destructive surgical treatment should be viewed with caution (Paparella & Sajjadi, 1999:36). Medical management together with psychological and family support should always be utilized before turning to aggressive surgical or medical treatment (Vesterhauge, 1996:15).

2.6.2 Medication

Medication is used to relieve symptoms such as nausea, vomiting and vertigo, but also to prevent further attacks of MD (Merchant, Rauch & Nadol, 1995:69). These drugs are usually

prescribed by general practitioners, ENTs and neurologists. Diuretics, vasodilators, antivertiginous medication, antiemetics, sedatives, antidepressants or herbal remedies may be given to control allergies, reduce fluid retention, or improve the blood circulation in the inner ear relieving associated symptoms experienced by the patient (Schessel et al., 1998:2681).

The use of diuretics is believed by many to be the best medical therapy for MD. The goal is to reduce endolymph volume by fluid removal and/or reduced production. This form of treatment does not lead to permanent cessation of the disease progression, especially with respect to hearing loss, but it is an effective option for the control of symptoms in many individuals (Schessel et al., 2005:3219). Symptomatic treatment with antivertiginous medication, antiemetics, sedatives and antidepressants have also been reported to be beneficial in reducing the severity of the vertigo and in improving tolerance of symptoms associated with MD. The value of vasodilating agents as well as holistic treatments such as herbal remedies are poorly documented and long-term follow-up studies of the efficacy of this treatment are not presently available (Schessel et al., 2005:3220). In most patients, it is possible to alleviate symptoms associated with MD by using prescription drugs (Vesterhauge, 1996:15).

2.6.3 Dietary changes

Eliminating caffeine, alcohol, refined sugars and salt may reduce the frequency and intensity of symptoms (Martin, 1994:310). A low-carbohydrate diet has also been suggested to prevent sodium retention (Proctor & Proctor, 1992:635). Dietary modifications have proven successful in controlling vertigo and even stabilizing hearing in some patients. In most patients, however, a combination of dietary changes together with the use of diuretics are needed for the successful treatment of the symptoms associated with endolymphatic hydrops (Schessel et al., 2005:3219). Personal clinical experience has shown this type of treatment to be successful in the majority of patients.

2.6.4 Behavioral therapy

Therapy helping to reduce stress may lessen the severity of the disease symptoms. (Vesterhauge, 1996:11). Counseling forms an important part of treatment in the patient with MD. Medical professionals involved in the treatment of these patients (general practitioners,

neurologists, ENTs, psychologists and audiologists) should attempt to break the vicious circle of stress. Such a process begins by allocating the necessary time to listen to the patient's complaints, examining the patient's hearing and balance function thoroughly, and explaining to the patient the exact nature of MD. The natural history of the disease should be discussed with the patient and their family, stressing that there is not yet a medical or surgical cure, but for many, we can successfully alleviate symptoms and improve quality of life (Vesterhauge, 1996:11). Most are comforted by a reassuring explanation of the disease process, which should strongly emphasize that it is not life threatening, but chronic. More professional psychological support and treatment with antidepressants may be needed for some who have great difficulty in coping (Torok, 1977:1873).

Reversing hearing loss is an important part of optimal management of the disease. Unfortunately, long term hearing impairment does not seem susceptible to current methods of treatment (Kinney et al., 1997:67). Therefore, hearing rehabilitation with the use of hearing aids forms an important part of the treatment of a patient with MD, emphasising the role of the audiologist in the management of patients with MD (Vesterhauge, 1996:16).

2.6.5 Hearing rehabilitation

The emphasis of any rehabilitative effort must begin with an understanding of the person with whom we are dealing (Ross, 1994:587). The role of the audiologist becomes that of assessing the handicapping effects of hearing impairment in terms of communicative efficiency and evaluating the success of aural rehabilitative procedures in reducing these handicapping effects. The purpose of rehabilitative intervention is to maximize the reception of speech, reduce the impact of psychosocial factors and imparting information about hearing loss, use of hearing aids, assistive listening systems and communications strategies (Ross, 1994:592). This process involves a thorough evaluation of both the hearing impairment and the hearing handicap. This orientation to the rehabilitation process requires a commitment on the part of the audiologist that goes beyond determining the extent of the hearing loss and the site of lesion. It means that there must also be a strong commitment to the communication and other needs of the person with the hearing loss, that is, these needs cannot be met by only defining the parameters of the hearing impairment and by determining the amplification needs (Giolas, 1994:777).

Professionals involved in fitting hearing aids experience primarily two difficulties when fitting patients diagnosed as having MD with hearing aids. The first challenge is the presence of a fluctuating hearing loss (Barber, 1983:27). Follow-up audiograms are necessary and only once the hearing loss stabilizes can a hearing aid be fitted successfully. The second challenge is that typical of a cochlear sensorineural hearing loss, namely recruitment and distortion of speech. Patients with MD can have a decreased dynamic area for comfortable speech, and SD ability may be affected. This combination of reduced SD, diplacusis and recruitment, with the narrowed dynamic range of sound tolerance implied in the word recruitment, makes hearing aid use difficult (Barber, 1983:28). With proper testing and counselling these problems can, however, be overcome in most cases (Vesterhauge, 1996:17). The use of hearing aids also has the advantage of reducing the inconvenience of tinnitus (Vesterhauge, 1996:16).

2.6.6 Treatment of tinnitus

The main focus of treating tinnitus is to provide tinnitus control or relief, as there is no cure for tinnitus at this time (Goldstein & Shulman, 1999:8). Basic science and clinical efforts with patients who experience tinnitus, suggest tinnitus to be a complex symptom. Tinnitus control is most effective with combined therapy as part of a multidisciplinary approach. This has resulted in the development of a multi-factorial, multidisciplinary approach for tinnitus treatment including conventional (medical, audiological) and unconventional (alternative medical/non medical) approaches for treatment and control. Significant advances have been made with instrumentation including amplification, masking, habituation and electrical stimulation providing tinnitus relief (Shulman, 1996:95). Tinnitus retraining therapy as proposed by Professor P.J. Jastreboff (1990) is receiving a lot of attention and found to be successful in tinnitus control in 80 % of patients (Lebisch, 2001:25). The goal of retraining is to habituate tinnitus, in other words, lower the patients' awareness level to the point where tinnitus no longer dominates their perception. Retraining does not promise cure, but one benefit is that it works regardless of the cause of tinnitus. The focus of retraining is the improvement of the general quality of life and is harmless and free of side effects. No medication is given and hearing is not impaired. This approach does, however, require active patient participation demanding commitment, time, patience and determination (Lebisch, 2001:26). Counselling forms an important part of retraining therapy and aims at developing a thorough understanding of the mechanisms leading to the development and amplification of

tinnitus. Only a good understanding of the condition will allow the patient to develop a personal strategy of coping with it (Lebisch, 2001:25).

2.6.7 Conclusion

The uncertain and changeable behaviour characteristic of MD makes rational assessment of most forms of treatment of the condition remarkably difficult (Schessel et al., 1998:2681). MD is cyclical and has many spontaneous remissions and therefore whatever medication or diet was last used might be thought to be effective (Martin, 1994:311). Specific treatment will be determined by the clinician(s) based on the patient's age, overall health and medical history, extent of the disease, expectations for the course of the disease, tolerance for specific medications, procedures, or therapies as well as the patient's (or family's) opinion or preference (Merchant et al., 1995:72). Optimal management should stop vertigo, put an end to tinnitus and reverse hearing loss (Schessel et al., 1998:2681). A multidisciplinary approach should be encouraged to ensure effective patient management.

2.7 SUMMARY

MD is a disorder of the inner ear, which is associated with a symptom complex consisting of attacks of episodic vertigo, sensorineural hearing loss and tinnitus, often accompanied by a sensation of aural fullness (Monsell et al., 1995:181). Despite the fact that the symptom complex is well known, it remains a controversial and often difficult disease to diagnose, which leads to difficulty in defining optimal management (Schessel et al., 1998:2673). This might be, in part, a result of the dramatic variability and individualized nature that is characteristic of this disease, but also emphasizes the need for further research into the nature of the disease (Schessel et al., 1998:26673). The pathogenesis of MD is still not clear, and numerous factors are implicated as the cause. It is probably the result of a variety of etiologic processes all having in common the pathophysiologic process of endolymphatic hydrops (Dickins & Graham, 1990:55). There is no single test that can make the diagnosis of MD, and diagnosis is primarily based on patient history (Schessel et al., 1998:2678). This includes a detailed description of the clinical presentation of the disease related the triad symptom complex of vertigo, hearing loss and tinnitus. For an accurate diagnosis symptoms should be defined by an audiological test battery including PT audiometry, SD, OAE, ENG and EcoG testing. To date, there is no proven medical or surgical cure for those with MD, but there is

still much that clinicians can do to alleviate the symptoms experienced by patients with this disease (Vesterhauge, 1996:11).

CHAPTER 3

METHODOLOGY

The aim of this chapter is to describe the research design, material, methods and procedures used during the execution of this study, to provide sufficient information so the reader can understand the methods and procedures of the study to judge their appropriateness to investigate the research question asked. This will also permit direct and systematic replications of this study (Hedge, 2003:226).

3.1 INTRODUCTION

This chapter provides a detailed description of the methods and procedures used to perform this study. It outlines the aims and design of the study, and also describes the way in which data was collected and analyzed. The purpose of this study was to analyse and describe the clinical and audiological features associated with MD to assist in the diagnostic process. This was done by conducting a retrospective descriptive research study of patient files that comply with specific selection criteria. Relevant data was extracted from the files and plotted on a datasheet (Appendix A). The data was analyzed and interpreted quantitatively through descriptive statistics.

3.2 AIMS

The main aim of the study is to analyse and describe the clinical and audiological features of a cohort of subjects diagnosed with MD, in order to develop further understanding of the pathophysiology of MD and the diagnostic processes involved.

3.2.1 Sub-aims

The main aim is realized in the following sub-aims:

3.2.1.1 To describe the clinical profile of a group of subjects diagnosed with MD, with reference to the following factors:

- ❖ Gender;
- ❖ Age of onset;
- ❖ Family history;
- ❖ Type of MD;
- ❖ Laterality of the disease.

3.2.1.2 To investigate the incidence of symptoms experienced in each type (definite, probable and possible MD) of MD, with specific reference to the following manifestations:

- ❖ Vertigo;
- ❖ Nausea or vomiting;
- ❖ Tinnitus;
- ❖ Fullness in the ear;
- ❖ Tullio phenomenon;
- ❖ Fluctuating/fixed hearing loss.

3.2.1.3 To determine the significance of audiological and vestibular tests in the diagnosis of MD in a cohort of subjects, with reference to the following investigations:

- ❖ PT audiogram:
 - Degree of hearing loss (pure tone average);
 - Audiogram configuration;
- ❖ SD scores;
- ❖ DPOAE
 - Amplitude;
 - Frequency;
- ❖ EcoG
 - SP-AP amplitude ratio;
- ❖ ENG:
 - Saccades;
 - Optokinetic tests;
 - Tracking;
 - Positional test;
 - Gaze test;

- Bithermal caloric test result;
- Dix-Hallpike test.

3.2.1.4 To define a systematic audiological approach to the investigation of MD that will facilitate the clinical diagnosis.

3.3 RESEARCH DESIGN

The research is based on a retrospective study of the medical records of subjects diagnosed with MD by an ENT. This study largely relies on the statistical investigation of data and therefore it is based on a quantitative research approach. The use of this approach allowed for converting the clinical profile of the patients into numerical format (Steyn, Smit, du Toit & Strasheim, 1994:7), which allowed the researcher to generalize the findings from the subject profiles by employing statistical analysis techniques. This type of research also explores the possible relationships that exist between phenomena and allowed for careful description of the symptoms and clinical tests that assisted in the diagnostic process (Leedy, 1997:106). The main obstacle of this type of data collection is that it can be time-consuming, which stresses the importance of clearly defined aims and selection criteria which has been set out elsewhere in the study (Neuman, 1997:32).

Applied research was used because it investigates a phenomenon within the practical field and primarily aims to apply and tailor knowledge to address a specific practical issue. The main strength of applied research is its immediate practical use. The research design followed during this study is descriptive since applied research is frequently descriptive research (Neuman, 1997:22). This approach allows for the description of relationships, the documentation of information and provides new explanations for data that contradict prior beliefs. Through descriptive research one can provide an accurate profile of a group and also create a set of categories or classify types (Neuman, 1997:20). This allowed for data coding in grid format for data recording purposes (Appendix A). Descriptive research differs from explorative studies in that it is more structured and focuses on only a few dimensions of a clearly defined entity, in this case the clinical features of MD (Singleton, Straits, Straits & McAllister, 1988:90). These features are important for this study because of the variability and individualized nature of this disease as well as the protean features involved in the diagnosis.

Correlational study analysis can be successfully used in case history analysis, since it is a statistical investigation of the relationship between more than one factor and investigates surface relationships (Thomas & Nelson, 2001:288). It does not, however, necessarily search for the causal reasons underlying such factors (Leedy, 1997:111). This method has been used to answer the questions related to this study by describing the relationship between clinical and audiological features of subjects with MD.

3.4 ETHICAL CLEARANCE

Once the aims and design of the study was decided, a research proposal was submitted to the University of Pretoria Faculty of Humanities' Research and Ethics Committee for approval. The letter of ethical clearance and approval from the committee is included in Appendix B.

As professionals in the field of Speech Language Therapy and Audiology we have a responsibility towards the advancement of knowledge (Pannbacker, Middleton & Vekovius, 1996:9). One way of doing this is to conduct research that has been approved by the relevant ethical committee and to make the results known to colleagues. The ethical principles for Speech Therapists and Audiologists as identified by the South African Speech- Language Hearing Association (SASLHA, 2000) include beneficence and non-maleficence, autonomy, justice, veracity and fidelity. The principles applicable specifically to research include beneficence and non-maleficence, autonomy and fidelity. Implicit in the SASLHA Code of Ethics is that an investigator is responsible for protection of the rights and welfare of all participants (McCartney, 2002:1). The main ethical issues identified in this research addresses autonomy and fidelity including informed consent, disclosure of information and patient confidentiality.

This is a retrospective study of subject records. A letter was obtained from an ENT specialist in private practice wherein he confirmed knowledge of the research project and also gave written consent for access to the subject files (Appendix C). It was not possible to obtain individual consent from the specific subjects, because of reasons such as insufficient subject details, subjects not updating their details and emigration. The ethical guideline for research states that informed consent must be obtained from subjects when possible, but that signed informed consent statements are optional for most survey, field, and secondary data research

(Neuman, 1997:450). The proposed study falls into this category of secondary data analysis. The risk for potential harm to the subject will also determine the need for written consent (Neuman, 1997:450). The statistical analysis of administrative and biometric data is entirely impersonal and would therefore not be harmful to any of the subjects.

The following steps were taken to ensure disclosure of information and subject confidentiality:

- ❖ No personal information that could link the patients with the study was documented or used.
- ❖ The files of all subjects are secured in a confidential system within the ENT practice. Access to the files is limited to the ENT specialist, audiologist and specific assistants. The files of patients were not removed from the ENT practice.
- ❖ During the data collection phase the subject's names were only recorded to prevent double entries. This list remained in the care of the ENT for record purposes.
- ❖ The coding process ensured patient confidentiality.
- ❖ All information gained in the study was treated as confidential.
- ❖ Storing data on a computer package with a secret user password ensured the security of research records (Neuman, 1997:453).

3.5 SAMPLE

A sample is a set of data taken from the population of interest (Neuman, 1997:201), which in this case constituted 135 subjects diagnosed with MD. The sample size was chosen using a conventional method, where the sample size is based on past studies and experience with samples that have met the requirements of the statistical method (Neuman, 1997:222). Review of previous research papers on MD indicated an average sample size of 116 cases per study.

3.5.1 Population

The clinical data from this group of patients diagnosed with MD by an ENT specialist in private practice was used as subjects for this study. The group consisted of subjects with

typical and atypical idiopathic MD. Since the sample is representative of both forms of the disease, the full spectrum of this disease presentation was evaluated. The exact sample size was determined by the number of cases in the private practice diagnosed with MD.

3.5.2 Selection criteria

A convenience non-probability sampling approach was employed to select the sample, where samples of people of different ages, sexes and other classifications were selected (Leedy, 1997:204; Thomas & Nelson 2001:289). This approach was selected because the diversity regarding the signs and symptoms of MD makes it difficult to locate patients assessed and diagnosed by different medical professionals according to the same diagnostic criteria (Schessel et al., 1998:2681). It is also less time consuming and can be used to select members of a difficult-to-reach, specialized population (Neuman, 1997:204). The main disadvantage of this approach is that data may not be representative of the larger population (Leedy, 1997:204). The researcher addressed this problem by selecting a larger sample, including subjects diagnosed by the ENT specialist over a period of three years.

Candidates considered for possible selection had to meet the following criteria:

❖ **ENT examination**

Subjects had to be examined and diagnosed with MD by an ENT specialist based on the American Academy of Otolaryngology- Head and Neck Surgery's general guidelines (Monsell et al., 1995:182). This excluded other causes of vestibular dysfunction and hearing loss, which may mimic MD, from the study (Weber & Adkins, 1997:978). A question sheet was used by the ENT to facilitate the history taking process (Appendix F).

❖ **Acoustic trauma**

The case history of subjects had to exclude acoustic trauma in order to describe the effect of MD on hearing thresholds more specifically (Ginsberg & White, 1994:21).

❖ Presenting symptoms

A clinical description of the subject's presenting symptoms had to be available, since it is considered to be the most important tool for diagnosis of MD (Paparella & Sajjadi, 1999:31).

❖ Availability of audiological test results

The following audiological test results had to be available to define the symptoms associated with MD: PTTs, SD scores, EcoG, ENG (Saccades, Optokinetic, Tracking, Positional / Gaze / Spontaneous nystagmus, Bithermal calorics and Dix-Hallpike) and DPOAE test results (vestibular and classical MD group), as it is accepted that after patient history audiological data provide the most relevant information for confirming the diagnosis (Lee et al., 1995:527). These tests are thought to be the most reliable audiological measurements to determine cochlear and vestibular functioning in MD (Schessel et al., 1998:2680). Specialized audiological tests such as ABLB, SISI, ABR and the immittance test that were extensively employed in the past are less often used today (Lee et al., 1995:527) and were not included as part of this study.

Table 3.1: Summary of selection criteria.

- | |
|---|
| <ol style="list-style-type: none">1. Subjects were examined and diagnosed with MD by an ENT surgeon based on the American Academy of Otolaryngology- Head and Neck Surgery's general guidelines.2. All the subjects' case history excluded acoustic trauma.3. Subjects' files included a description of the presenting symptoms.4. Audiological tests results: PTTs, SD, EcoG, ENG and DPOAE were available. |
|---|

3.5.3 Sample size

One hundred and thirty five patient files between January 2001 and February 2004 complied with the selection criteria and were used for the study.

3.5.4 Description of sample

The sample case material is based on 135 subjects (57 males and 78 females). Their ages ranged from 20 to 68 years. Eighty two subjects (115 affected ears) were clinically categorized as definite MD, with documented sensorineural hearing loss, and with an

association of episodic attacks of vertigo, tinnitus, and pressure in most cases. Nineteen subjects (23 affected ears) were clinically categorized as probable MD, also with documented sensorineural hearing loss, but who has only experienced one attack of vertigo, tinnitus, and pressure in most cases. Thirty four subjects (50 affected ears) presented with a non-typical history and were diagnosed with possible MD. These cases presented with either a hearing loss, or vestibular symptoms in association with tinnitus and pressure in most cases.

Table 3.2: Summary of subjects.

Number of subjects	Age range	Number of participants in each type of MD
135 subjects: 57 (42 %) male 78 (58 %) female	20 – 68 years	Definite MD 82 (61%) Probable MD 19 (14 %) Possible MD 34 (25 %)

3.6 MATERIAL AND APPARATUS

This subsection describes the physical setting of this study as well as the names and model numbers of equipment used to obtain information relating to the study.

3.6.1 Material and apparatus during subject selection

The material comprised of the case history as well as the audiometric and medical test results of 135 patients with MD. These files were selected from an ENT specialist’s available client database by using a convenience non-probability sampling approach. All the files of patients complying with the specified selection criteria were used for analysis in this study.

The following material and apparatus were used to obtain and analyse information regarding the case history and audiological test results of all subjects:

3.6.2 Material and apparatus used during data collection

A datasheet (Appendix A) was compiled to organize the:

- ❖ case history as documented in the ENT specialist’s notes and
- ❖ audiological (PTT, SD, DPOAE, ENG, EcoG) test results

in tabular form for comparison and analysis.

3.6.2.1 Audiometry results

- ❖ The MAICO Audiometer GmbH MA52 with MAICO TDH-39 headphones was used to perform air conduction PT audiometry. The audiometer was calibrated in August of each year by NS Clinical Technologies. The calibration certificates are available on request.
- ❖ Bone conduction PT audiometry was performed using the above mentioned audiometer together with the Radioear B-71 bone conduction receiver. The bone conduction receiver was calibrated in August of each year by NS Clinical Technologies. The calibration certificates are available on request.
- ❖ The TDH-39 headphones were used to present narrowband noise to the non-test ear during masking.
- ❖ The monitored live voice function on the MAICO Audiometer GmbH MA52 was used to obtain SD scores with the use of a phonetically balanced (PB) word list. All the patients were English speaking and therefore the PB word list from the Central Institute for the Deaf Auditory Test W-22 list 3A was used during testing for all subjects (Appendix D).

3.6.2.2 OAE Analyzer

- ❖ The Madsen Celesta 503 Cochlear Emissions Analyzer (Version 3.xx) was used to perform DPOAE testing. The system consists of a software package installed on a compatible computer, a probe assembly and a signal-processing unit. The lightweight probe assembly was mounted on a shoulder harness and stabilized in the patient's ear with Madsen Electronics color-coded eartips.

3.6.2.3 ENG system

- ❖ The ICS Medical computerized CHARTR compact-2 ENG system program (Version 5.71) was used to evaluate vestibular function.
- ❖ A two-channel Electronystagmograph was used for simultaneous recording of both horizontal and vertical eye movements.

- ❖ The subject's skin was prepared for electrode application with Omni-Prep surgical scrub on gauze pads.
- ❖ An ICS Medical disposable EP-ENG pre-gel, silver/silver chloride electrode with attached safety leadwire was used to record eye movement.
- ❖ The ICS Medical water caloric stimulator (Model NCI-480) was used for caloric testing.

3.6.2.4 EcoG system

- ❖ The BIO-LOGIC Evoked Potential program system (Version 5.00 Model) was used to evoke electrocochleographic responses.
- ❖ Omni-Prep surgical scrub and gauze pads were used to prepare the subject's skin before the electrodes were applied.
- ❖ Ten20 conductive EEG paste was used on the two surface electrodes.
- ❖ A tympanic membrane electrode (TM-EcochGtrode) with FCG electrode gel was used to record EcoG results. A tympanic membrane electrode is a flexible tubelike structure with a gel substance at the tympanic membrane end and a wire connected to an electrode in at the other end (Hall & Mueller, 1997:308). It is the ideal choice for audiologists since it is non-invasive and the test can be performed without otologic support or anaesthesia. It also allows for reliable recordings of large amplitudes for SP and AP components in patients with mild to moderate hearing loss. The main disadvantages of tympanic membrane electrodes are limited commercial availability, high interelectrode impedance and patient discomfort. Accurate placement of the electrode also requires some technical skill (Hall & Mueller, 1997:315).

3.6.3 Material and apparatus during data analysis and processing

An electronic spreadsheet (Appendix A) was used to analyse and describe data by applying certain statistical formulas to the data (Leedy, 1997:271). A specialized statistical program on a mainframe computer was used by the statistician to complete this process (Leedy, 1997:273). The specific statistical software package used to organize the data into tables and figures and to perform statistical calculations through interactive processing was the SASS (Statistical Analysis System Software). The use of such a program increases the speed with which data can be processed (Leedy, 1997:274).

Datasheets allow for sorting of all the data based on separate categories, searching for information regarding specific measurements or variables, and compilation of graphs from parts of the data (Leedy, 1997:271). The speed and ease with which the electronic spreadsheet can calculate and recalculate formulas creates an important tool for the researcher to gain further insight into the data. It is easy to do additional comparisons between groups that might prove interesting. This allows for the researcher to ask ‘what if?’ questions for example: “What if the data were analyzed on the basis of gender and other variables”. “What if...” questions allow the researcher to look closely at alternatives that were always available but that may not have been examined because of the time and effort required to complete the calculations (Leedy, 1997:270).

3.7 PROCEDURES

This subsection describes in detail how the study was implemented, providing all the information necessary to replicate the study.

3.7.1 Procedures during subject selection

The researcher has worked in a practice in association with an ENT specialist, and was allowed access to the files of patients who consulted him with complaints of hearing and balance problems.

- ❖ Files between January 2001 and February 2004 were reviewed with the help of the ENT surgeon to select subjects who were diagnosed with MD.
- ❖ Data that complied with the selection criteria was separated and used for answering the research questions.

3.7.2 Data collection procedures

The quantitative data collection technique that was used is existing statistics research, where a source of previously collected information is located and used for statistical analysis (Neuman, 1997:32). Existing statistics is most frequently used for descriptive research (Neuman, 1997:32). This was done by gaining permission from an ENT specialist in private practice to conduct a retrospective review of patient charts. The data included 135 subjects

diagnosed with so-called “idiopathic” MD, including both auditory and vestibular symptoms and audiological test results.

The following data was extracted from the files and plotted on a datasheet (Appendix A):

- ❖ Complete case history documented by ENT specialist
- ❖ Subject’s age and gender
- ❖ Audiogram
 - PTT (air and bone conduction)
 - SD scores
- ❖ DPOAE test results
- ❖ EcoG results
- ❖ ENG results

The audiological tests were performed, by the researcher, on the same day as the ENT specialist’s examination to allow for comparison between test result and symptoms experienced by the patients. The following procedures were followed during audiological testing:

3.7.2.1 PT audiometry test procedure

PTTs (air- and bone conduction) were determined by following the test procedure proposed by Martin (1994:77,83), and interpreted according to Selmani, Pykko, Ishizaki and Ashammakhi (2002:174) and Lee et al. (1995:528).

❖ Positioning of the subjects during testing:

Patients were seated in a soundproof room, at a right angle to the audiometer. Earphones were positioned so that their diaphragms were aimed directly at the opening into the ear canal (Martin, 1994:74).

❖ Instructions to the subject:

“You will hear sounds in the right and left ear alternately. Some sounds will be low pitched and other high pitched. Please press the button every time you hear a sound in either ear even if it is very soft”.

❖ Procedure for air-conduction audiometry:

The “better” ear was tested first. In cases where subjects could not identify the “better” ear, the right ear was tested first. Pure tones were initially presented at 1000 Hz, proceeding to the lower frequencies in descending order, retesting 1000 Hz and then the high frequencies in ascending order (1000 Hz, 500 Hz, 250 Hz, 1000 Hz, 2000 Hz, 4000 Hz, 6000 Hz, 8000Hz). The PT was initially presented at 30 dB HL. If no response was observed, the level was raised to 50 dB, and then raised in 10-dB steps until a response was obtained or the limit of the audiometer was reached. After a response was obtained, the PT level was lowered in 10-dB steps. When the tone was lowered below the subject’s threshold, it was raised in 5-dB steps until the 50 % threshold response criterion was reached. Each frequency threshold was recorded on an audiogram (Martin, 1994:77).

❖ Masking:

Procedure: Narrowband noise was presented in the non-test ear when the intensity of the test tone minus 40 dB (interaural attenuation) was louder than the bone conduction (BC) threshold of the non-test ear at the same frequency. Since the BC threshold was not yet available the air conduction threshold was used. The masking levels were reviewed once BC thresholds were obtained. The noise was presented 10 dB above the threshold of the non-test ear. If the subject could not hear the sound in the presence of the noise, the noise was increased until the subject could identify the test tone with the noise in the non-test ear increased in two steps of 10 dB at each frequency (Martin, 1994:95).

Instructions to the subject: “You will hear a noise in the one ear. Please ignore the noise and only press the button when you hear the pure tone in the test ear”.

❖ Procedure for BC audiometry:

The test procedure was identical to that of air-conduction audiometry, apart from the frequency range that was more limited (1000 Hz, 500 Hz, 250 Hz, 1000 Hz, 2000 Hz, 4000 Hz) (Martin, 1994:85).

❖ Masking:

Procedure: Narrowband noise was presented in the non-test ear when there was an air-bone gap (ABG) of more than 5 dB in the same ear. The masking technique was the same as applied for air conduction (Martin, 1994:101).

Instructions to the subject: Same as with air conduction masking.

❖ Interpretation of audiogram:

The pure tone average (PTA) = PTT 500 Hz + PTT 1000 Hz + PTT 2000 Hz + PTT 4000 Hz ÷ 4 (Lee et al., 1995:528). The scale of hearing impairment was based on the PTA (Monsell et al., 1995:182; Selmani et al., 2002:174). The standard calculation of PTA at 500 Hz, 1000 Hz and 2000 Hz (Martin, 1994:78) was not used, because it does not truly reflect the degree of hearing loss in subjects with high frequency loss only (Lee et al., 1995:529).

Table 3.3: Interpretation of PTT (Martin, 1994:78).

Normal = 0–15 dB HL	Very mild = 16–25 dB HL	Mild = 26–40 dB HL
Moderate = 41–55 dB HL	Moderate-severe = 56-70 dB HL	Severe = 71-90 dB HL

3.7.2.2 SD test protocol

SD scores were obtained and interpreted according to the test protocol proposed by Martin (1994:136-139).

❖ Positioning of the subject during testing:

Positioning of the subject was the same as that for PTT measurements (Martin, 1994:74).

❖ Instructions to the patient:

“I am going to read a list of words to you, please repeat whatever you hear. You can guess if you are not sure. Please do not look at my mouth”.

❖ Procedure for obtaining SD scores:

Monitored live voice was used. Twenty-five words from a PB word list were presented 30 dB above the PTT. If the subject made more than two discrimination errors the

presenting level was increased with 10 dB until a level of 100 % SD, a plateau or maximum outset of audiometer was reached. The level where the subject achieves 50 % SD was also determined and at least three points were plotted on the speech audiogram (Martin, 1994:129).

❖ Masking:

Procedure: White noise was presented in the non-test ear (NTE) whenever the hearing level of the phonetically balanced words (PBHL) in the test ear (TE) minus the interaural attenuation (IA) equalled or exceeded the BC thresholds of the NTE at 500 Hz, 1000 Hz and 2000 Hz, expressed as a formula $PBHL (TE) - IA \geq \text{best BC (NTE)}$. Effective masking (EM) was presented at a level equal to the hearing level at which the discrimination test is performed, minus 40 dB for interaural attenuation, plus the largest ABG at 500 Hz, 1000 Hz and 2000 Hz in the masked ear, expressed in the formula $EM = PBHL (TE) - IA + ABG (NTE)$ (Martin, 1994:135).

Instructions to the subject: “You will hear a noise in the one ear. Please ignore the noise and repeat the words as before”.

❖ Interpretation:

Table 3.4: Interpretation of speech discrimination results (Martin, 1994:139).

<u>SD scores</u>	<u>General SD ability</u>	<u>SD curve</u>	<u>Site of lesion</u>
90% –100%	Normal limits	Normal S curve (100% at 20–30 dB HL)	Normal hearing
75% – 90%	Slight difficulty	Normal S curve (100 % above 30 dB HL)	Middle ear: Conductive hearing loss
60% – 75%	Moderate difficulty	Plateau	Cochlea: Sensorineural hearing loss
50% – 60%	Poor SD (difficulty to follow conversation)	Roll-over (20% deterioration in SD within a 10 dB increase in intensity)	Retrocochlear: Sensorineural hearing loss
<50%	Very poor SD unable to follow running speech)		

3.7.2.3 DPOAE test protocol

DPOAE recordings were obtained and analysed according to the *Madsen Operational Manual* (1995:25-43). The recommended system parameters were used during testing (Table 3.5).

❖ Positioning of subject during testing:

The subject was seated in a soundproof room, with either the front or the back of the subject facing the cochlear emissions analyser, depending on the side of the TE. An eartip was inserted into the subject’s ear to create an airtight seal in the external ear canal. The shoulder harness was placed on the shoulder on the side of the TE (*Madsen Operational Manual, 1995:26*).

❖ Instruction to subject:

“You will hear different sounds in your ear; you do not have to respond to these sounds. Please try to sit as still as possible”.

❖ Test procedure:

The software allows the detection of the DPOAE amplitude as a function of the frequency of the primaries (f1 and f2) at fixed intensity levels and the measurement of the growth function. The geometric mean of f1 and f2 was analysed at frequencies between 0.5 and 6 kHz in half-octave steps in order to correlate DPOAE threshold and PTT. Input-output functions were recorded at six geometric mean frequencies between 0.5 and 6 kHz. Emission responses are automatically plotted on two graphs, one depicting the input/output (growth) function, the other relating response amplitude to frequency (DP-gram). Colour coded responses are automatically recorded and presented in the cursor box (*Madsen Operational Manual, 1995:40*).

Table 3.5: Madsen technical system parameters used during DPOAE recordings.

SYSTEM PARAMETERS:	
F2/f1 1.22	2*f2-f1 654 Hz
F0 500 Hz	F1 Level: 65 dB SPL
F1 452 Hz	F2 Level: 55 dB SPL
F2 553 Hz	Gain (A) 27 Db
2*f1-f2 351Hz	Sweep Acc. Rej. 250 / 89 SD2

❖ Interpretation:

OAE data for amplitude and noise floor was analysed as a function of stimulus frequency and interpreted in relation to the normative data (Hall & Mueller, 1997:253). The DPOAE normative data for the Madsen Celesta is presented in Table 3.7. The normative data was compiled through the testing of thirty ears from young audiometrically normal adults (hearing threshold levels 15 dB or better from 250 to 8000 Hz and normal type A tympanograms (compliance values of between 0.3 and 1.75 cc, ear canal volume between 1.0 and 1.4 cc and middle ear pressure between -100daPa and +100daPa) in a quiet, non-sound treated room. The test protocol included: $f_2/f_1=1.2$; $L_1=65$; $L_2=55$ dB SPL. Frequency is the geometric mean of f_1 and f_2 (*Madsen Operational Manual, 1995:42*). Emissions were interpreted as normal if the DP amplitude minus the noise floor difference exceeded 5 dB, if the DP amplitude was greater than -10 dB, if the DPOAE data was replicable with amplitude values within 3 dB to 5 dB, and if the DP amplitude was within the normative region as presented in Table 3.6 (Hall & Mueller, 1997:255).

Table 3.6: DPOAE Normative data for Madsen Celesta (Hall & Mueller, 1997:261).

Frequency	Mean	SD	1%ile	5%ile	10%ile	99%ile	95%ile	90%ile
500 Hz DP*	4.9	4.92	2.42	3.06	3.37	7.38	6.74	6.43
NF*	-9.97	4.33	-12.14	-11.58	-11.31	-7.79	-8.35	-8.62
753 Hz DP	11.3	5.44	8.56	9.27	9.61	14.04	13.33	12.99
NF	-5.63	6.08	-8.69	-7.9	-7.52	-2.57	-3.36	-3.75
1006 Hz DP	11.9	6.98	8.39	9.29	9.73	15.41	14.51	14.07
NF	-8.23	4.48	-10.49	-9.91	-9.62	-5.98	-6.56	-6.84
1512 Hz DP	7.9	6.97	4.39	5.3	5.74	11.41	10.5	10.06
NF	-11.43	6.59	-14.75	-13.89	-13.48	-8.12	-8.97	-9.39
2011 Hz DP	4.8	5.39	2.09	2.79	3.13	7.51	6.81	6.47
NF	-17.03	3.32	-18.7	-18.27	-18.06	-15.36	-15.79	-16
3023 Hz DP	.6	4.5	-1.66	-1.08	-.8	2.86	2.28	2
NF	-22.43	3.19	-24.04	-23.63	-23.42	-20.83	-21.24	-21.44
4036 Hz DP	8.03	5.75	5.14	5.89	6.25	10.93	10.18	9.82
NF	-19.3	3.72	-21.17	-20.69	-20.46	-17.43	-17.91	-18.14
6060 Hz DP	14.17	8.03	10.12	11.17	11.67	18.21	17.17	16.66
NF	-16.6	3.42	-18.32	-17.88	-17.66	-14.88	-15.32	-15.54

*DP = amplitude of DP in dB SPL / *NF = amplitude of noise floor in dB SPL

Once the DPOAE results have been calculated, it was interpreted in relation to the PT audiogram to determine the site of lesion:

- ❖ Normal cochlear function was indicated by normal hearing thresholds together with normal emissions.
- ❖ Early microvascular damage (cochlear site of lesion) was indicated when the hearing thresholds were within normal limits, but some or all of the emissions were absent or display lower than normal DP amplitudes (Hall & Meuller, 1997:254).
- ❖ A retrocochlear lesion was suggested when normal emissions were elicited in the presence of a hearing loss exceeding 25 dB (Hall & Meuller, 1997:254).
- ❖ A cochlear site of lesion (possible retrocochlear in the case of severe sensorineural hearing loss) is indicated when a hearing loss is present with absent emissions (*Madsen Operational Manual, 1995:42*).

The presence of DPOAE was examined by interpreting the results independently for each subject, determining if emissions were normal or abnormal. Emissions were analysed in terms of amplitude and frequency. The DPOAE test results were recorded on the electronic spreadsheet.

3.7.2.4 ENG test protocol

The ICS Medical Electronystagmography instructional guide test protocol was followed to record and interpret ENG results (Stockwell, 1991:27-52).

❖ The examination room:

A patient examining table was placed in the centre of the room, with a visual display mounted on mounted on the wall beside the examining table. The subject was seated sideways on an examining table facing a visual display during the initial preparations for testing and during the saccade test, gaze test, tracking test, and optokinetic tests. The subject laid lengthwise on the table for the remainder of the examination (Hallpike maneuver, positional test, and bithermal caloric tests). During the caloric test the subject was positioned so that a line from the external ear canal to the outer canthus of the eye was vertical (30° angle) (Stockwell, 1991:2).

❖ Application of electrodes:

A small amount of Omni-Prep was placed onto a gauze pad and rubbed on the subject's skin at the sites of electrode application. A dry gauze pad was used to wipe off excess Omni-Prep. The first two electrodes were placed so that a line between them exactly bisects the subject's corneas as the patient gazes straight ahead. The third electrode was placed on the subject's forehead. The fourth and fifth electrodes were placed alternatively above and below the subject's left eye. These two electrodes also bisected the subject's corneas with the subject gazing straight ahead (Stockwell, 1991:33).

❖ Instructions to the subject for each test category:

Calibration and Saccades test: "Please look straight in front of you. The red dot will jump from the one side of the screen to the other. Please follow the dot, keeping your eyes on it without moving your head. After this, the dot will jump randomly on the screen for one minute and 42 seconds. Again, follow the dot without moving your head" (Stockwell, 1991:34).

Gaze test: "Look at the red dot. Once the test starts, the dot will jump to the right side of the screen. Without moving your head, follow the dot and keep your eyes fixed on it until the dot returns to the middle of the screen (approximately 20 seconds). We will repeat the test to the left side, upwards and downwards" (Stockwell, 1991:35).

Tracking test: "The dot is going to slide along the screen. Follow the dot without moving your head for two minutes" (Stockwell, 1991:38).

Optokinetic: "You are going to see a whole lot of dots moving across the screen. The test will be performed twice, first to the right and then the left. Choose one dot, ignoring the rest, and follow it until it disappears from the screen. Choose another dot and repeat the process until the test stops" (approximately 20 seconds) (Stockwell, 1991:39).

Hallpike Maneuver: "On the count of three lie down quickly. First, with your head turned to the right side and then the left side. Your head will hang off the bed, but relax your neck. Keep your eyes open". (Eye movement should be observed for at least 20 seconds) (Stockwell, 1991:43).

Positional test: “Look straight in front of you (eyes open/closed); straight up while lying down (eyes open/ closed); to your right while lying down (eyes open/closed); to your left while lying down (eyes open/closed). Each position will take approximately 20 seconds” (Stockwell, 1991:46).

Bithermal caloric test: “I am going to irrigate each ear twice with water, once with cool distilled water (30°C) and once with distilled warm water (44°C). We will wait five minutes before irrigating the other ear. You will probably feel dizzy, but it will only last a few minutes. After thirty seconds the water will stop and I will ask you to close your eyes. Close both eyes and start to count in threes. I will tell you when you can open your eyes again” (Stockwell, 1991:51).

❖ Interpretation of each test category

ENG is a physiologic test and does not measure the functional abilities of the patient. The results, however, often help to explain the symptoms that the patient has been experiencing. Discerning abnormal from normal or artifactual eye movement is predominantly a pattern recognition task requiring an experienced audiologist with formally acquired knowledge (Shepard & Telian, 1994:432).

Saccades: The response is automatically recorded and interpreted by the computer software as normal or abnormal. A 50 % overshoot from the normal square wave is considered to be abnormal and indicative of a central cause (Stockwell, 1991:35).

Tracking test results: Over or undershooting of the target sine wave is recorded by the computer software as an abnormal response (Stockwell, 1991:38).

Gaze/positional test results: A flat response with no nystagmus is normal. If nystagmus is present the slow phase velocity of the nystagmus beat is measured. The nystagmus intensity is considered significant if the slow phase velocity exceeds 6 deg/sec in at least one head position (Stockwell, 1991:37).

Optokinetic: Small nystagmus amplitudes are elicited. The slow phase velocity of the nystagmus beat is measured and the left and right side compared. It is normal if the

intensity of nystagmus is approximately the same in both directions and the patient is able to keep up with the stimulus (Stockwell, 1991:41).

Dix-Hallpike: The diagnosis of BPPV from the semicircular canals is based on the clinical finding of a transient, up-beating, torsional nystagmus (Stockwell, 1991:45).

Caloric test results: Each irrigation affects the vestibular receptors of that ear and provokes a horizontal nystagmus response. The responses provoked by the right ear irrigation are compared with those in the left ear, to determine whether the sensitivities of the left and right vestibular mechanisms are equal. The slow phase velocity of the nystagmus beat is measured for each response to calculate if a unilateral caloric weakness is present using the following formula: $(\text{Right ear warm response} + \text{right ear cold water response}) - (\text{left ear warm water response} + \text{left ear cold water response}) \div (\text{Right ear warm water response} + \text{right ear cold water response} + \text{left ear warm water response} + \text{left ear cold water response})$. The final figure is multiplied by one hundred to reveal a percentage. A 25 % interaural difference was considered significant or abnormal. A bilateral weakness is identified when the slow phase velocity of the nystagmus beat is smaller than 7 deg/sec in both ears (Stockwell, 1991:53).

Each subtest was analysed individually and the results were recorded on the electronic spreadsheet. The results of the different subtests were also combined to determine the site of pathology (central / peripheral).

3.7.2.5 EcoG protocol

EcoG recordings were performed and analysed according to the Vanderbilt Audiology Clinics' protocol (Hall & Mueller, 1997:300-305).

The following technical parameters as recommended by Hall and Mueller (1997:301) were used for the recording of the responses:

Table 3.7: Technical system parameters used for EcoG recordings.

STIMULUS:		FILTER SETTINGS:	
Type of stimuli	Clicks	Hi filter	1500.00
Stimulus phase	Alternating	Low filter	3.00
Intensity	Maximum (90 dB)	Channel	1
Masking	None	Maximum stimuli	1500
Stimulus rate	7.1/s	Electrode placement	Cz/A1/A2
Stimulator	Telephonics TDH-39 P Headphones	Type of electrode	Bio-logic Specialty TM-EcochGtrode

The objective is to record a clear and reliable SP and AP for right and left ear stimulation, to calculate the SP/AP ratio for each ear, and to categorize the SP/AP ratio as normal versus abnormal based on symmetrical and normal expectation. The decision regarding the type of electrode used should be based on hearing sensitivity. It is important to use an electrode option that produces a clear SP and AP at the 7.1/sec click rate. Tympanic membrane electrodes usually reveal a clear SP and AP component and can be used in the presence of a high frequency sensorineural hearing loss (Hall & Mueller, 1997:304).

Table 3.8: Technical parameters for the TM-EcochGtrode.

Gain	50 000
Channel	1
Window	9.984 ms
Pre/post	-2 ms
Stimulator	Headphones
Stimuli	Clicks
Rate	7.1/s
Polarity	Alternating
Intensity	90 dB
Montage	Non-inverting (input 1) = contralateral ear / surface electrode Inverting (input 2) = ipsilateral tympanic membrane electrode Ground (Cz/Fpz) = forehead, surface electrode
Classification	Baseline (BSL) Summation potential (SP) - Action potential (AP)

❖ Positioning of subject during testing:

All the subjects were asked to lie flat on a bed placed in a sound proof room.

❖ Instructions to the subject:

“I am going to prepare your skin with a special scrub, place one electrode on your forehead, another behind your ear and a third electrode will be inserted into your ear canal to touch the eardrum. The electrode is soft and cannot cause any damage. Earphones will be placed on your ears. During the test you will hear click sounds first in the one ear and then the other. Close your eyes and relax. Try not to move or speak while the test is in progress” (Hall & Mueller, 1997:304).

❖ Position of electrodes:

A small amount of Omni-Prep was placed onto a gauze pad and rubbed on the subject's skin at the sites of electrode application. A dry gauze pad was used to wipe off excess Omni-Prep. Ten20 conductive EEG paste was applied to electrode one and electrode two. These were alternatively placed on the mastoid of the NTE (A1/A2) and the forehead (Fpz). The TM-EcochGtrode was dipped into FCG electrode gel and inserted into the ear canal touching the tympanic membrane. All electrodes were secured with surgical tape (Hall & Mueller, 1997:309).

❖ Measurement:

Eighth nerve and cochlear potentials elicited by broad-bank click stimuli were recorded from the external auditory meatus. At least three replicated waveforms were obtained for each ear at a rate of 7.1/sec with 1500 stimuli. An alternating stimulus phase with broadband clicks was used to minimize the cochlear microphonic for optimal recording of AP/SP ratios. The normal or least involved ear was tested first (Hall & Mueller, 1997:301).

❖ Analysis and interpretation:

SP and AP amplitudes were identified and the ratio calculated from a common stable baseline. Interaural asymmetry was calculated and reported if the difference was more than 10 %. The SP/AP ratio is automatically calculated by the BIO-LOGIC Evoked Potential program software and was considered abnormal if the ratio exceeded 40 % (Hall & Mueller, 1997:303).

3.7.3 Data recording procedures

Data coding was necessary to systematically reorganize raw data into a format that was machine-readable for statistic analysis (Neuman, 1997:295). The main advantage of data coding is that data can be compared with the use of frequency and cross tabulations (Neuman, 1997:322). Unfortunately, this type of coding system did not allow for statistical calculations such as statistical significance (p-values) to be performed. This was, however, not the purpose of the study.

The coded data was presented in grid format where each row represents one case and each column a data field (Appendix A). A column was assigned to each variable (Neuman, 1997:296). Eighteen variables were identified namely type of MD, presenting symptoms, sex, age of onset, hearing level, audiogram configuration, laterality of the disease, family history, SD scores, OAE, EcoG ENG, Dix-Hallpike, number of attacks, frequency of attacks, duration of the disease, follow up audiogram level of hearing and the duration between audiograms. Most of these variables are self-explanatory, but the classification and coding system that was used for column one, five and six needs further clarification and is discussed below. A codebook (Appendix E) was compiled to describe this coding process (Neuman, 1997:295).

3.7.3.1 Type of MD (codebook column one)

Column 1 is concerned with the diagnosis of MD. The subjects with MD were divided into three groups (Table 3.9) as identified by the Committee on Hearing and Equilibrium of the American Academy of Otolaryngology-Head and Neck Surgery (Monsell et al., 1995:182). This allowed for differentiation between classical and atypical MD.

Table 3.9: Diagnostic classification of MD (Monsell et al., 1995:182).

Definite MD	Probable MD	Possible MD
1. Two or more definitive spontaneous episodes of vertigo 20 minutes or longer 2. Audiometrically documented sensorineural hearing loss (unilateral/bilateral) on at least one occasion 3. Tinnitus or aural fullness in the affected ear 4. Other causes excluded	1. One definitive episode of vertigo 2. Audiometrically documented sensorineural hearing loss (unilateral/bilateral) on at least one occasion 3. Tinnitus or aural fullness in the treated ear 4. Other causes excluded	1. Episodic vertigo of the Ménière type without documented hearing loss (vestibular MD), or 2. Unilateral/bilateral sensorineural hearing loss, with dysequilibrium but without definitive episodes (cochlear MD) 3. Tinnitus or aural fullness in the affected ear 4. Other causes excluded

According to the Committee on Hearing and Equilibrium of the American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) only subjects in the definite category should be reported as having MD (Monsell et al., 1995:183). There is especially controversy regarding the inclusion of a vestibular type of MD. The Committee on Hearing refers to episodic vertigo without hearing loss, tinnitus, or aural fullness as recurrent vestibulopathy (RV) (Monsell et al., 1995:182). Schessel et al. (2005:3217) also refer to RV and suggest that use of the term vestibular MD is an inappropriate application of the diagnosis. This argument is based on the fact that follow up studies on patients with vestibular MD have found that only 10 % to 20 % of the vestibular Ménière’s patients develop definite MD (Paparella & Mancini, 1985:149). In addition, Rauch, Merchant and Thedinger (1989:876) found no pathologic correlation (anatomical evidence of endolymphatic hydrops) for vestibular MD.

On the other hand Paparella and Mancini (1985:149) thought that some subjects with RV presented with features of vestibular MD. Rutka and Barber (1986:106) reported that 15 % of subjects with RV undergo diagnostic change to classic MD with follow-up. With regard to the temporal bone study by Rauch et al. (1989:876), only three cases presented with episodic vertigo of a nature indistinguishable from that of clinical MD. This constitutes only a small percentage of subjects with vestibular symptoms.

This study included these subjects with vestibular symptoms without hearing loss to investigate this entity and to compare data with other studies. It is hoped that inclusion of this group of subjects with typical MD symptoms, but without demonstrable hearing loss will shed light on whether vestibular MD should be included in the general classification of MD or as suggested a different entity such as RV. Rutka and Barber (1986:106) suggest that the key for diagnosing patients with vestibular MD or RV may be the presence or absence of tinnitus and aural fullness. Therefore the presence of tinnitus and/or aural fullness was included in the original classification of the Committee on Hearing and Equilibrium AAO-HNS to make the diagnosis of vestibular MD.

It has also been suggested that some patients in this category of vestibular MD / RV might have vestibular migraine (Schessel et al., 2005:3271). Both categories of patients present with episodes of vertigo. Episodic vertigo occurs in about 25 % of migraine patients (Baloh, 1997:615). A possible explanation for the association between migraine and MD is the development of endolymphatic hydrops in an ear previously damaged by migraine. Vasospasm could lead to ischemic damage to the endolymphatic duct and/or sac leading to the eventual development of hydrops (Baloh, 1997:617).

3.7.3.2 Pure tone average (codebook column five)

In Column 5 the hearing loss was classified according to the Committee on Hearing and Equilibrium of the American Academy of Otolaryngology-Head and Neck Surgery (Monsell et al., 1995:182). Two changes were made to allow for better description of the audiogram. The first stage of 0 – 25 dB had been sub- divided to allow differentiation of “no hearing loss” and “very mild hearing loss” (Selmani et al., 2002:174). The four-tone average used to determine the stage of hearing included 4 kHz instead of the recommended 3 kHz to reveal more high-frequency information (Lee et al., 1995:528).

3.7.3.3 Audiogram configuration (codebook column six)

The classification of audiogram shapes used in this paper is not unique. The choice was made to take the smallest number of categories that would still allow describing the characteristics of audiogram shapes seen most in this group of patients and those described in literature. The classification system resulted in five categories based on a research paper by Lee et al.

(1995:527). Rising, falling, and dip configurations are defined as at least a 10 dB difference in the low-frequency, high-frequency, and mid-frequency range, respectively. Flat configurations are defined as thresholds within a 10 dB range including 500 Hz to 4 000 Hz. Peak audiogram configurations, seen at 2000 Hz, are defined as at least 10 dB better than all other frequencies. There are three subcategories under peak configurations. Type I is within normal hearing limits, with the PTT for each frequency above 25 dB and the threshold for 2000 Hz 10 dB above the best threshold. Type II peak audiogram thresholds range between 25 to 60 dB. In type III peak configuration the PTT range within the 61 to 90 dB range (Lee et al., 1995:528). All hearing losses were of sensorineural origin. This was determined by comparing air- and bone conduction thresholds, which were found to be equal for the same subject (Martin, 1994:87).

Any coding errors threaten the validity of measures and cause misleading results. Therefore the process of cleaning data as explained in Table 3.10 was applied to ensure accuracy of the data (Neuman, 1997:297).

Table 3.10: Data cleaning process.

<p>After careful coding, the accuracy of coding was checked by applying:</p> <ul style="list-style-type: none">❖ wild code checking (involves checking the categories of all variables for impossible codes such as double digit entries in single digit slots) and❖ contingency cleaning techniques (involves cross-classifying two variables and looking for logically impossible combinations) to 15 % of the data for a second time (Neuman, 1997:297). If any coding errors appeared, the researcher rechecked all the coding.
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3.7.4 Data analysis procedures

Statistics is a science of collecting and interpreting data (Dietrich & Kearns, 1989:2). The data from the datasheet was quantitatively summarized and analysed with the help of a statistician. In correlational research information is reorganized and combined to address the research question (Neuman, 1997:294).

Descriptive statistics is a method involved in measuring variation and correlation, and was used to organize, analyse and interpret the data to provide answers to the sub-aims of the

study (Steyn et al., 1994:5). This method was used in order to predict or estimate a population parameter from the sample used in the study by looking for patterns and relationships between variables (Dietrich & Kearns, 1989:7). Identifying patterns and relationships between the clinical and physiological presentation of the disease leads to a better understanding of the clinical picture as well as the diagnostic process involved in this disease. This method also allowed for logical conclusions to be drawn from the data and permits inferences from a sample to a population (Johnson, 1994:2). The results of this study therefore represent the nature of MD in the general population.

The nonparametric statistic technique “chi-square” was used to compare data and present it in frequency and cross tabulations (Neuman, 1997:322). This was necessary to describe the clinical profile and categorize the symptoms experienced by patients with MD according to Ménière’s type and incidence as well as to determine the importance of audiological tests in the diagnostic process. This technique allowed for differentiation between groups, and is commonly used in comparative studies (Leedy, 1997:269).

Statistical analysis organized the data so that the results could be discussed, supported by figures and tables according to the nature of each sub-aim. Presenting results in this way provides quick, visual summaries of information about the phenomenon of interest. Graphic presentations make tendencies and features of observed data easy to understand and interpret (Steyn et al., 1994:77). It is intuitively appealing descriptive devices that may be used to describe samples (Dietrich & Kearns, 1989:1).

3.8 SUMMARY

The main aim of the study is to analyse the clinical and audiological features of a cohort of subjects diagnosed with MD, to provide insight into the nature of MD and the diagnostic processes involved. This study is based on a quantitative research approach. The sample consists of subjects with typical and atypical MD complying with specific criteria. Data was collected through the use of existing statistical material to conduct a retrospective review of 135 subject charts. Selected information was plotted on a datasheet and analysed quantitatively with the help of a statistician. Nonparametric statistic techniques were used to organize and

analyse the data to provide answers to the sub-aims of the study. Inferential statistics were used to interpret the data. The results are discussed in Chapter 4, supported by figures and tables.

CHAPTER 4

RESULTS AND DISCUSSION

The clinical and diagnostic characteristics of 135 subjects with MD are reported. The collected and processed data is presented in tables and figures. Each presentation is followed by an interpretive discussion of the data in relation to a voluminous literature on the classification of MD, which remains uncertain.

4.1 INTRODUCTION

According to the SASLHA Code of Ethics, professionals in the field of Speech Language Therapy and Audiology have a responsibility towards the advancement of knowledge (Pannbacker et al., 1996:9). One way of doing this is to conduct research as it involves discovering and learning new things (Neuman, 1997:1). Research also helps us to better plan and deliver services, to educate others as well as to build our basic knowledge to assist both professionals and patients in making informed decisions (Neuman, 1997:15).

It is important to conduct research specific to the South African context since several variables such as differences in access to health care in the different countries, differences in diagnostic criteria, differences in data gathering techniques, as well as true epidemiologic differences among people around the world may affect research results (Dickins & Graham, 1990:51). Genetic and environmental factors probably influence differences in incidence among countries and sections of countries (Schessel et al., 1998:2674). Seeking to discover the relationship among the clinical profile and audiometric test results and how all of this relate to the individual who has come to us for help, will encourage and motivate audiologists to do further and specialized testing in order to improve their accountability and reliability of the diagnosis as well as to function more effectively as part of the multidisciplinary team.

Data for this study was obtained by conducting a retrospective descriptive research study on the case history and test results of 135 subjects with MD complying with the specified

selection criteria. These files were selected from an ENT specialist's available client database. Sub-aims were identified to provide a systematic approach to answer the main aim of the study. The statistical method used to analyze data was descriptive. The results were obtained through the analysis of the spreadsheet (Column 1 – 32) with the use of the SASS program and presented in frequency and cross tabulations (Leedy, 1997:273). Differences among groups were tested for discrete characteristics such as age, degree of hearing loss and for presence or absence of a symptom or MD type by applying the chi-square test to the frequency and cross tabulations (Neuman, 1997:322).

4.2 PATIENT ANALYSIS

The results relating to the first and second sub-aims are discussed in this section. The first sub-aim was to describe the clinical profile of a group of subjects diagnosed with MD, with reference to the following factors: Gender; Age of onset; Family history; Type of MD and Laterality of the disease. The second sub-aim relates to the investigation of the incidence of symptoms experienced in each type of MD, with specific reference to the following manifestations: Vertigo; Nausea or vomiting; Tinnitus; Fullness in the ear; Tullio phenomenon and Fluctuating/fixed hearing loss.

4.2.1 Clinical profile of a group of subjects diagnosed with MD

The patient history was obtained through the analysis of Column 1: Type of Ménière's disease, Column 9: Sex, Column 10: Age of onset, and Column 16: Family history of the electronic spreadsheet (Appendix A). These columns were identified to assist in finding an answer to the first sub-aim in identifying the clinical profile of the patients diagnosed with MD. Other retrospective studies have used the same data collection procedure to compile and analyse the clinical profile of patients with MD (Meyerhoff, Paparella & Gudbrandsson, 1981:220; Wada et al., 1990:115). The results are summarized in Table 4.1.

Table 4.1: Summary of patient history in MD (n=135).

Definite MD	Probable MD	Possible MD	
		Vestibular MD/RV	Cochlear MD
1. 61 % (n=82/135) subjects 1.1 Males 49 % (n=40/82) 1.2 Females 51 % (n=42/82)	1. 14 % (n=19/135) subjects 1.1 Males 32 % (n=6/19) 1.2 Females 68 % (n=13/19)	1. 21 % (n=29/135) subjects 1.1 Males 34 % (n=10/29) 1.2 Females 66 % (n=19/29)	1. 4 % (n=5/135) subjects 1.1 Males 20 % (n=1/5) 1.2 Females 80 % (n=4/5)
2. Average age: 48 years Males 49 years Females 48 years	2. Average age: 55 years Males 54 years Females 56 years	2. Average age: 41 years Males 45 years Females 41 years	2. Average age: 51 years Males 33 years Females 56 years
3. Family history: 3 % (n=4/135) subjects	3. Family history: 0 %	3. Family history: 0 %	3. Family history: 0 %

The results obtained according to Table 4.1 are discussed individually in 4.2.1.1 to 4.2.1.4 according to the different aspects relating to the clinical profile of the subjects as identified in the sub-aim.

4.2.1.1 Type of MD

The majority (61 % - n=82/135) of subjects forming part of this study were diagnosed with definite MD, where subjects experience that classical symptoms triad of vertigo, hearing loss and tinnitus. Cochlear MD were found in only five subjects (4 % - n=5/135) in this study, with vestibular MD/RV being second in frequency (21 % - n=29/135) to that of the classical form.

Kitahara et al. (1984:52) found similar result with 62 % of subjects in their study diagnosed with definite MD. This contradicts the findings of Shea (1975:263) who found cochlear MD to be as much as ten times more common than definite MD. The findings of this study correlates with the findings of Kitahara et al. (1984:52) who found that 32 % of the subjects participating in their study were diagnosed with vestibular MD and only 6 % with cochlear

MD. The high incidence of cochlear MD in the study performed by Shea (1975:263) might be explained by the fact that some subjects only experience cochlear symptoms in the early phase of the disease (Kitahara et al., 1984:52; Meyerhoff 1981:222; Schessel et al., 2005:3217). Since it has been found that 80 % of subjects with cochlear MD go on to develop definite MD, the subjects in the study of Shea (1975:263) might have been diagnosed with definite MD if they were seen later in the course of the disease. The second possible explanation might be that it may be difficult or clinically impossible to differentiate certain cases of sudden deafness from cochlear hydrops revealing a higher incidence of what seems to be cochlear MD (Barber, 1983:29).

4.2.1.2 Sexual preponderance

This study found the incidence of MD in females to be higher than in males, with a female to male ratio of 1.4:1. This correlates positively with the 1,5:1 ratio of Wada et al. (1990:119) as well as with the findings of Meyerhoff et al. (1981:220). Oosterveld (1980:885) and Martin (1994:311), however, suggest a male predominance. Combining the results of the above studies, one is led to believe that there is probably no predilection for either sex in MD. Factors such as accessibility of medical services and geography will determine incidence outcomes of different research studies. It is however interesting to note that the female/male ratio is markedly higher in the cases of atypical MD. Watanabe (1981:512) also noted this preponderance especially in relation to vestibular MD/RV.

In the group with vestibular MD 66 % (n=19/29) of the subjects were female. Paparella and Mancini (1985:149) found 75 % (n=39/52) of subjects with vestibular MD/RV to be female, while Watanabe (1981:512) found a male to female ratio of 27:100. The reason and significance of these results are unknown.

The cause for the difference in incidence related to gender in MD is unknown. Genetic factors would appear to be irrelevant, since this incidence has been reported in several studies in different population groups (Schessel et al., 2005:3210; Watanabe, 1981:511). Epigenetic factors such as hormonal factors are likely to be responsible. In particular the earlier age incidence in females might indicate a hormonal background that is more favourable to the development of MD. It can be postulated that fluid retention in females on the basis of hormonal effects might aggravate fluid retention in the vestibular system and endolymphatic

hydrops might be more severe. Further studies, in particular related to levels of oestrogen and progesterone, might provide useful information in this regard (Watanabe, 1981:514).

4.2.1.3 Age of onset

It is well accepted that MD is an illness primarily affecting individuals in the fourth and fifth decade of life (Meyerhoff et al., 1981:222). The average age for subjects to develop MD in this study was 48 and ranged from 20 to 68 years. The distribution peaked between the ages of 40 and 60 years. This age distribution correlates well with those previously published in the literature (Meyerhoff et al., 1981:222; Oosterveld, 1980:886). Meyerhoff et al. (1981:222) reported an average age of onset of 45 years and Lee (1995:527) an average age of 42.6 years. Sakurai et al. (1991:93) reported a peak distribution of subjects with MD between the ages of 41 and 60 years. The average age of onset for males and females in this study is similar (45 years for males and 48 years for females) and in agreement with other reports indicating that there is no statistical significant difference between age onset in males and females (Balkany et al., 1980:605; Lee et al., 1995:527; Wada et al., 1990:116). There does, however, seem to be a tendency for subjects with vestibular MD/RV to present at an earlier age than subjects with definite MD. The age distribution peaked for the age group 35 to 45 years, while the peak for definite cases of MD was in the age group 45 to 55 years. Watanabe (1981:512) found similar results with the age group for vestibular MD/RV peaking at 30 to 39 years and for subjects with definite MD 40 to 49 years. This might be explained by the fact that in MD, vestibular symptoms can develop first, to be followed by cochlear symptoms leading to definite MD in later years (Paparella & Mancini, 1985:148). Subjects with vestibular MD might be experiencing early symptoms of definite MD which might only be identified years later.

4.2.1.4 Family history

This study only indicated a family history in four subjects (3 % - n=4/135). Paparella (1985:447) found approximately 15 % (n=75/500) of subjects in his study to have a family history of MD. Although these percentages are fairly low, it indicates that genetic MD remains a possibility for some patients with MD. The main reason for the discrepancy between these two studies can probably be related to genetic differences among the population groups from which the results were gathered. The population group represented in

the study done by Paparella (1985:447) is largely Scandinavian, whereas the current study reflects a Dutch, French and English population.

4.2.1.5 Summary of the clinical profile of subjects with MD with reference to type of MD, gender, age of onset and family history

These results clearly show the diversified and individual clinical course of MD in different subjects. Although the majority of subjects (61 % - n=82/135) experience the classical symptoms associated with the disease, some individuals only presented with part of the symptom complex. The incidence of males and females as well as age of onset correlates well with previous research. The results also indicate the possibility of some people to inherit a predisposition to MD. Although further research is necessary to confirm it, the possibility arises that females might be more susceptible to the atypical forms of MD.

4.2.1.6 Laterality

Information regarding laterality of the disease was analysed with the help of frequency and cross tabulations by applying the SASS program (Leedy, 1997:274) to Column 15: Laterality of the disease of the electronic spreadsheet. The possibility of bilateral involvement is important when considering different treatment approaches (Paparella & Griebie, 1984:223). The diagnosis of MD was established independently for each ear. Both ears had to meet with the criteria outlined in Table 3.10 in order for a case to be considered bilateral MD. The diagnosis of MD is questionable in cases with bilateral sensorineural hearing loss where the hearing loss progresses in a stepwise fashion over a short period and responds to immunosuppressive medication as well as in cases with a PTA worse than 70 dB or less than 50 % correct SD at the PB max (Monsell et al., 1995:182). Subjects presenting with this clinical picture of hearing loss were not included in this study. A comparison of unilateral and bilateral disease in relation to patient history is presented in Table 4.2.

Table 4.2: Comparison of unilateral and bilateral disease in relation to patient history (n=135).

Unilateral MD	Bilateral MD
61 % (82/135) 26 % (35/82) left ear / 35 % (47/82) right ear	39 % (53/135)
Males 60 % (34/57) Females 62 % (48/78)	Males 40 % (23/57) Females 38 % (30/78)
Average age of onset: 42 years	Average age of onset: 47 years
Average duration of symptoms: 2.5 years	Average duration of symptoms: 1 year

With regard to laterality, on the basis of the clinical symptomatology, 26 % (n=35/82) of subjects in this research study were diagnosed with MD in the left ear, 35 % (n=47/82) in the right ear and 39 % (n=53/135) patients with bilateral disease. This finding correlates with other studies (Balkany et al., 1980:604; Paparella & Griebie 1984:234). On the other hand, Wada et al. (1990:118) found 41 % of the subjects in their study to have right side MD, 53 % left side, and 6 % bilateral. Meyerhoff et al. (1981:221) found similar results with the left ear affected in 46 % subjects, 42 % right ear and the symptoms were bilateral in 12 % of subjects. According to the results of this study as well as a literature review it is clear that MD is not associated with a pattern in terms of laterality.

Watanabe (1981:512), found a significant preponderance of female cases (66 %) in terms of bilaterality. This particular series, as well as a study done by Paparella and Griebie (1984:236) did not find a significant sex difference between unilateral and bilateral cases. Both studies did reveal a slight increase in age of diagnosis for the bilateral subjects as a group.

The time lapse from the time of presentation of initial symptoms to the time of diagnostic evaluation was studied and some differences were noted. The average time lapse for subjects with unilateral MD was 2.5 years and one year for bilateral disease. This differs from the study by Paparella and Griebie (1984:234) who found an average of five years (one month to 25 years) for unilateral and 8.4 years for bilateral disease (four months to 23 years). It is, however, clear from the wide range indicated in brackets, that the average time lapse would depend on the selected sample. It is also possible that during future follow-up, some subjects might be diagnosed with bilateral disease changing the duration averages of this study.

The results presented in Table 4.2 does not reaffirm the increase incidence of bilaterality with increasing duration of disease. The reason for this is probably because 94 % (n=127/135) of subjects in this study have had MD for less than 5 years. The bilateral nature of MD is most likely to become manifest within five to 10 years after unilateral onset (Paparella & Griebie 1984:236).

The incidence of unilateral and bilateral disease in the different types of MD is presented in Figure 4.1.

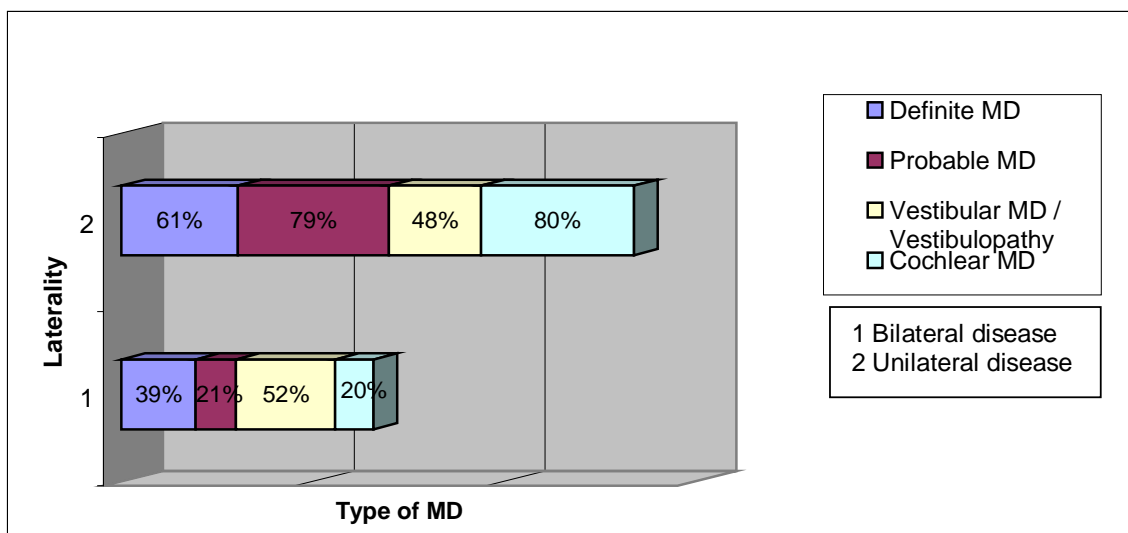


Figure 4.1: Laterality in MD (n=135).

Although most subjects with MD experienced symptoms for less than five years, an interesting feature is the high incidence of unilateral disease in cochlear MD in comparison with the other types of the disease (Figure 4.1). The incidence of bilateral disease in subjects with vestibular MD/RV was also high (52 % n=15/29) in comparison with the other types of MD. It is possible that cochlear MD is more likely to develop in one ear while vestibular MD/RV tends to affect both ears. The laterality of the disease will also be influenced by the duration of the disease in the subject. Another possible explanation for the high incidence of unilateral cochlear MD is that the diagnosis for cochlear MD depends on PT audiometry results. If abnormal results are only found in one ear at the time of testing it would be very difficult to diagnose bilateral MD. It is also important to note that only five subjects in this study were diagnosed with cochlear MD and that results might have been different for a larger sample. In vestibular MD there is little audiological information to reveal information

regarding laterality and the diagnosis is largely based on the subjects' perception of symptoms experienced. This might lead to inaccuracy when diagnosing the affected ear, leading to more subjects being diagnosed with bilateral disease. Paparella and Mancini (1985:149) found the incidence of bilaterality in the group with vestibular MD/RV to be much lower (14 %) than in typical MD. Since bilateral involvement increase over time (Balkany et al., 1980:606), the time period for which subjects have been experiencing symptoms related to MD will probably influence the figures related to bilateral involvement reported in research studies. The criteria used to define bilateral MD will also influence reported figures (Balkany et al., 1980:606).

Information regarding the degree of sensorineural hearing loss and audiogram configuration is used as one of the criteria to identify bilateral MD. The subjects' cochlear profile is used to determine auditory dysfunction and allows for early detection of bilateral MD (Balkany et al., 1980:606). Results regarding the auditory characteristics of subjects with bilateral disease are summarized in Table 4.3.

Table 4.3: Audiometric characteristics in bilateral disease (n=53).

	Symmetrical audiogram configuration
Bilateral symmetrical hearing loss (85 % - n=45/53)	96 % (n=43/45)
Bilateral asymmetrical hearing loss (15 % - n=8/53)	62 % (n=5/8)

The majority of the subjects with bilateral disease had symmetrical sensorineural hearing losses both in terms of degree of hearing loss and audiogram configuration. Only 15 % (n=8/53) of subjects with bilateral disease had significant asymmetric sensorineural hearing loss. It is of interest that there was greater variation (38 % - n=3/8) in the audiogram configurations of subjects with asymmetrical sensorineural hearing loss than in subjects with bilateral symmetrical sensorineural hearing loss (4 % - n=2/45). The difference in the audiogram configuration obtained for the two ears in the same patient probably indicates affected ears in different stages of the disease, as it is generally accepted that over time, as the hearing loss progresses, the audiogram configuration can change to become more flat and stabilized leaving the entire frequency range affected (Barber, 1983:28). On the other hand, Balkany et al. (1980:605) found that 25 % (n=26/104) of subjects with bilateral disease had roughly the same degree of hearing loss in both ears, with 75 % (n=78/104) presenting with

significant asymmetric hearing loss. The most likely reason for the higher incidence of asymmetrical hearing loss in their study is related to the criteria used during the diagnosis of the disease. Balkany et al. (1980:607) considered hearing loss as sufficient auditory evidence for considering bilateral involvement. The diagnosis of bilaterality in subjects in this study also considered early clinical manifestations such as tinnitus and aural pressure together with audiological and vestibular test results. Stricter diagnostic criteria would explain the lower incidence of bilateral disease with asymmetrical hearing loss in this study. Both studies indicated symptoms to be worse on the right side, although it is not of clinical significance. The presence of asymmetrical hearing loss and audiogram configuration differences in the same patient emphasizes the need for clinicians and audiologists to maintain a high level of suspicion for bilateral disease, for the early manifestations of inner ear dysfunction can easily be missed.

4.2.1.7 Summary of laterality in MD

The subjects in this study represent a group of patients with the clinical diagnosis of MD. In this research project 39 % (n=53/135) of subjects were found to have evidence of bilateral disease based on the clinical case history, including audiograms in conjunction with histories and vestibular tests. This is a conservative estimate, and is in good agreement with several other long-term studies (Balkany et al., 1980:603; Stahle, Friberg & Svedberg, 1991:78). The longer the duration of the disease, the greater the chances that the second ear will become involved (Balkany et al., 1980:605; Friberg et al., 1984:73). The prevalence of bilateral involvement has been the subject of several research papers and has been shown to increase with time from the initial diagnosis (Moffat et al., 1992:370). It is therefore expected that the proportion of subjects with bilateral disease would increase with continued follow-up, since most of the subjects have been experiencing symptoms related to MD for less than five years when they entered this study. The high frequency of bilaterality in MD has to be taken into consideration in the natural history and pathogenesis, and ultimately in treatment of this ailment. In patients with progressive MD where the symptoms are disabling and conservative medical management failed, surgical therapy may be offered. Surgical techniques which may be effective to preserve existing hearing thresholds such as vestibular neurectomy and endolymphatic sac decompression should be offered instead of destructive techniques such as labyrinthectomy, since bilateral involvement would complicate the patient's long-term treatment.

4.2.2 Investigation into the incidence of symptoms experienced in each type of MD

The reported symptoms of all subjects were recorded in Column 2 to 8 of the electronic spreadsheet. A column was assigned to each symptom generally experienced by patients diagnosed with MD. The symptoms include vertigo, hearing loss, tinnitus, fullness in the ear, nausea or vomiting, fluctuating hearing loss and the Tullio phenomenon. The occurrence of these symptoms as the subjects in this study experienced it, is summarized in Figure 4.2 and Table 4.4.

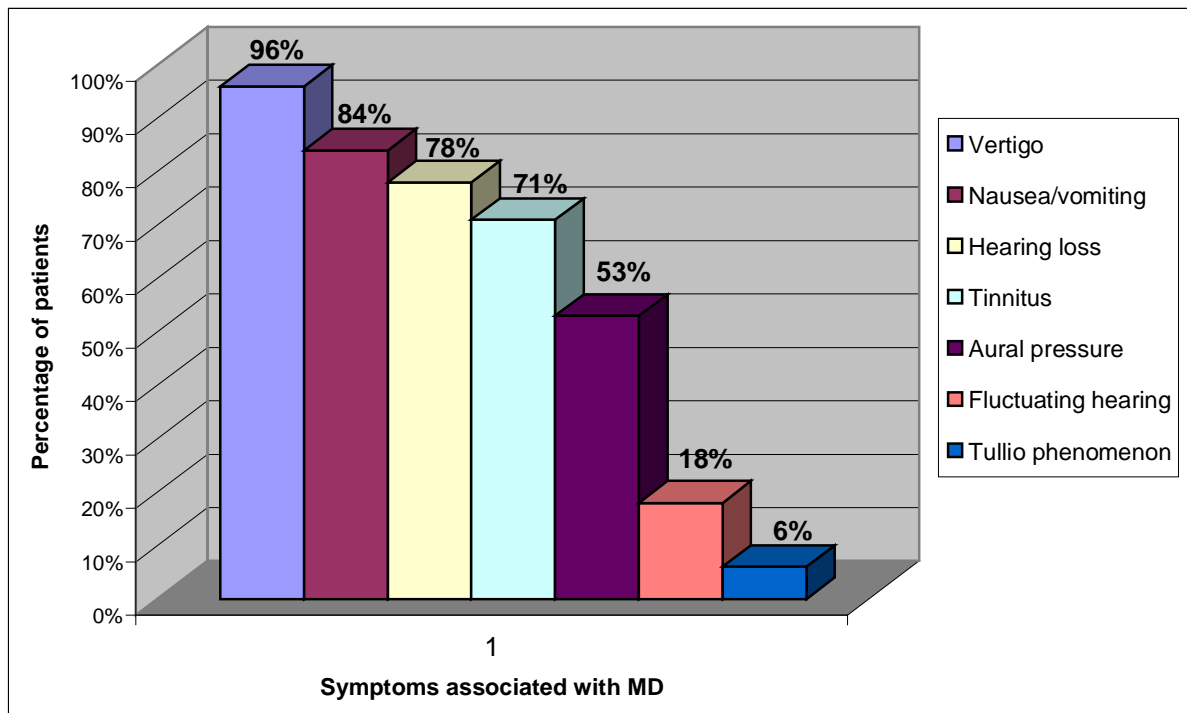


Figure 4.2: Symptoms experienced by subjects diagnosed with MD (n=135).

In this study 96 % (n=130/135) of all subjects experienced vertigo, 84 % (n=113/135) nausea and/or vomiting, 78 % (n=105/135) hearing loss, 71 % (n=96/135) tinnitus, 53 % (n=72/135) aural pressure, 18 % (n=24/135) fluctuating hearing loss and 6 % (n=8/135) the Tullio phenomenon as either primary or secondary symptoms. The majority (75 % - n=101/135) of patients described vertigo as the predominant or primary symptom, while only 22 % (n=30/135) complained of hearing loss and 3 % (n=4/135) of tinnitus as the main symptoms. The high incidence of nausea and/or vomiting correlates with the percentage of subjects experiencing vertigo. This is probably a result of connections that exist within the brainstem to link the vestibular, parasympathetic systems and vomiting centres (Barber, 1983:29).

Figure 4.2 indicates the importance of the symptom triad (recurring episodic attacks of vertigo, hearing loss and tinnitus) in the diagnosis of MD (Ginsberg & White, 1994:20). Seventy five percent (n=101/135) of the subjects experienced all three of the classical Ménière symptoms. Meyerhoff et al. (1981:221) also found that more than half (n=64/126) of the subjects in their study suffered from all three of the classical Ménière symptoms.

Not all subjects experienced all of the classical symptoms, and therefore the symptoms were divided into the different types of MD as presented in Table 4.4.

Table 4.4: Summary of symptoms experienced in each type of MD (n=135).

Symptom	Definite MD	Probable MD	Possible MD	
			Vestibular MD /RV	Cochlear MD
<u>Episodic Vertigo</u> Male (n=56/57) Female (n=74/78)	100 %	100 %	100 %	0 %
<u>Tinnitus</u> Male (n=42/57) Female (n=54/78)	83 %	79 %	38 %	40 %
<u>Hearing loss</u> Male (n=47/57) Female (n=59/78)	100 %	100 %	0 %	100 %
<u>Aural fullness</u> Male (n=25/57) Female (n=47/78)	40 %	37 %	97 %	80 %
<u>Nausea/vomiting</u> Male (n=52/57) Female (n=62/78)	91 %	100 %	62 %	40 %
<u>Tullio</u> Male (n=6/57) Female (n=2/78)	7 %	0 %	7 %	0 %
<u>Fluctuating hearing loss</u> Male (n=10/57) Female (n=14/78)	23 %	11 %	0 %	80 %

All the subjects in this study, except for those with cochlear MD, complained of episodic vertigo during their first visit to the ENT practice. Most of these subjects also experienced nausea or vomiting. Cochlear symptoms such as tinnitus and hearing loss were primary symptoms in the definite and probable MD groups. These results are in keeping with literature findings (Paparella & Mancini, 1985:148).

As shown in Table 4.4, it is of interest that 38 % (n=11/29) of subjects with vestibular MD experienced tinnitus. Paparella and Mancini (1985:149) found that 84 % (n=43/51) of subjects in their study with normal audiometric results complained of tinnitus. Tinnitus in the absence of hearing loss may be attributed to a moderate increase in potassium concentration of the perilymph of the scala tympani towards the apex of the cochlea, causing an increase of spontaneous activity of nerve activity (Horner, 1991:154). Another interesting finding is the high incidence of aural fullness in both vestibular MD/RV and cochlear MD. Paparella and Mancini (1985:149) found the symptom of aural pressure in almost every patient with vestibular MD. This might be explained by the anatomical site of endolymphatic hydrops, possibly as a result of malfunctioning of the utriculo-endolymphatic valve resulting in utricular and/or saccular hydrops to the relative or absolute exclusion of cochlear hydrops (Paparella & Mancini, 1985:150). Well-documented temporal bone studies are needed to confirm this suspicion. According to literature patients should probably be diagnosed with RV if no auditory signs or symptoms are present (Rutka & Barber, 1986:106). Patients with auditory symptoms such as tinnitus and aural fullness probably have a form of MD (Paparella & Mancini, 1985:150).

The Tullio phenomenon (vertigo provoked by intense sound stimulation) is not commonly mentioned in literature or studies regarding MD. Only patients with vestibular and definite MD experienced this phenomenon, which constituted 14 % (n=8/135) of all subjects used in this study. Kacker and Hinchcliffe (1970:155) mentioned the occurrence of the Tullio phenomenon in some subjects with MD. They explained that this phenomenon was sometimes observed in association with MD because endolymphatic hydrops distended the saccule to such an extent that it was in direct contact with the stapedial footplate without being cushioned by the perilymph. This creates direct contiguity of the ossicular chain with the membranous labyrinth. A literature study revealed no statistics to compare the incidence of this phenomenon with other studies. Personal experience, however, has shown that the Tullio phenomenon does affect patients with MD in their day to day activities and forms an

important part of patient management. The Tullio phenomenon can occur with or without sensorineural hearing loss.

The presence of fluctuating hearing loss in this study was very low (18 % - n=24/135), except in subjects with cochlear MD (80 % - n=4/5). Kitahara et al. (1984:52) noted cochlear MD to be characterized by fluctuation in sensorineural hearing loss. The cause for these results is not known yet. Possible explanations might firstly be that in definite MD, although both the cochlea and the vestibular system are involved, it seems that the vestibular system is more affected. In cochlear MD the primary site of lesion affected by endolymphatic hydrops is the cochlea and the effect of endolymphatic hydrops during the different stages of the disease are more noticeable. Secondly, subjects with cochlear MD might be more aware of hearing changes where patients with vestibular disturbances are often more concerned about the attacks of vertigo. Lastly it is possible that cochlear MD could be the early phase of definite MD where fluctuation in hearing is more common than later in the disease (Barber, 1983:28). More research is needed, but this might prove to be a diagnostic feature of patients with cochlear MD.

In summary, the morbidity caused by Ménière symptoms as determined in this study confirm the results of Meyerhoff et al. (1981:222) and Wada et al. (1990:118), who concluded that vertigo was the most dramatic and debilitating symptom in MD, with most subjects complaining of severe attacks of vertigo at the onset of the disease. Although several research papers indicate a fluctuating hearing loss as a diagnostic feature of MD, fluctuation is not universally present and is not essential to the diagnosis of MD (Monsell et al., 1995:182; Oosterveld, 1980:885; Schessel et al., 1995:2679). Further research might indicate aural pressure a diagnostic feature of vestibular MD/RV and fluctuating hearing loss diagnostic of cochlear MD.

Apart from the patient's history, audiological data provides the most relevant information for confirming the diagnosis of MD. Both patient history and audiological tests results are necessary to confirm the diagnosis of MD and establish effective patient management (Lee et al., 1995:527).

4.3 THE SIGNIFICANCE OF AUDIOLOGICAL AND VESTIBULAR TEST RESULTS IN THE DIAGNOSIS OF MD

The results relating to the third sub-aims namely to determine the significance of audiological and vestibular tests in the diagnosis of MD in a cohort of subjects with reference to PT audiometry, SD scores, DPOAE test results, EcoG results and ENG results are discussed in this section leading up to the end of the chapter. A better understanding of the audiological profile seen in MD is important, especially for diagnosing bilateral disease (Paparella & Griebie, 1984:233). The results are also discussed in relation to the clinical profile as identified in the previous section, since both patient history and audiological data need to be combined to confirm the diagnosis of MD (Lee et al., 1995:527).

The test procedure proposed by Martin (1994:77,83) as outlined on page 44 was used to obtain PT audiometry results. These results were interpreted according to Selmani et al. (2002:174) and Lee et al. (1995:528) and discussed in detail on page 47 and page 59. The degree of sensorineural hearing loss is based on the PTA of the hearing thresholds 500 Hz, 1000 Hz, 2000 Hz and 4 000 Hz. To characterize the shape of the audiogram in each ear, a classification system was introduced taking into account both the curvature and the slope of the audiogram. The classification of the different audiogram configuration types is explained on page 59 to 60.

4.3.1 The significance of PT audiometry and SD scores in the diagnosis of MD

PT audiometry and SD scores were obtained for all subjects and recorded in Column 11: Pure tone average left ear, Column 12: Pure tone average right ear, Column 13: Audiogram configuration left ear, Column 14: Audiogram configuration right ear, Column 17: Speech discrimination left ear and Column 18: Speech discrimination right ear of the electronic spreadsheet. The results are summarized in Figure 4.3 to 4.6 and Tables 4.5 to 4.15. These diagnostic test results are discussed in relation to the clinical features and symptoms presenting in the subjects diagnosed with MD.

4.3.1.1 PT audiometry results in relation to patient history

The influence of the type of MD (definite, probable or possible MD) as well as the age of the subjects on the PTA and audiogram configuration was investigated and the results are presented below.

4.3.1.1.1 The relationship between PT audiometry results and the type of MD

Most research studies, which investigated the influence of MD on PT audiometry results, did not distinguish between the different types of MD. This study differentiates between the different types of MD and its impact on hearing to clearly demonstrate any effects that change in site of pathology might have on audiometric results.

Table 4.5: PTA in different types of MD (n=135).

PTA	Definite MD (n=82)	Probable MD (n=19)	Possible MD	
			Vestibular MD/ RV (n=29)	Cochlear MD (n=5)
Normal (0-15dB)	0 %	0 %	100 % (n=29/29)	0 %
Very mild (16-25dB)	20 % (n=16/82)	16 % (n=3/19)	0 %	0 %
Mild (26-40dB)	56 % (n=46/82)	74 % (n=14/19)	0 %	67 % (n=3/5)
Moderate (41-55dB)	13 % (n=11/82)	0 %	0 %	33 % (n=2/5)
Moderate-severe (56-70dB)	8 % (n=7/82)	11 % (n=2/19)	0 %	0 %
Severe (71-90dB)	3 % (n=2/82)	0 %	0 %	0 %

Table 4.5 shows a wide distribution of degree of hearing loss, with a definite increase in mild hearing loss for subjects in this study. The average loss of hearing for each group in this

study, except for vestibular MD/RV where the subjects have no hearing loss, however, was between 40 to 50 dB. Severe hearing losses occurred only in subjects with definite MD, constituting 3 % (n=2/82) of all subjects. This correlates with the findings of other research where only 2 % of severely affected subjects experience severe to profound sensorineural hearing loss (Stahle, 1976:113). A severe hearing loss usually occurs in more advanced stages of MD (definite MD) (Schessel, 1998:2680). It is also evident according to the PTT that cochlear symptoms were more severe in subjects diagnosed with definite MD than the other types of MD. Kitahara et al. (1984:55) attribute this finding to the spread of biochemical alteration in perilymphatic fluid, which is limited in cochlear and vestibular MD/RV compared to definite MD. The anatomical site of the endolymphatic hydrops may also influence the audiogram configuration found in subjects with MD (Schuknecht & Gulya, 1983:1).

Table 4.6: Distribution of audiogram configuration in MD types (n=135).

Audiogram configuration	Definite MD	Probable MD	Possible MD	
			Vestibular MD/RV	Cochlear MD
Rising 1	8 % (n=7/82)	16 % (n=3/19)	0 %	20 % (n=1/5)
Flat 2	38 % (n=31/82)	32 % (n=6/19)	83 % (n=24/29)	60 % (n=3/5)
Falling 3	28 % (n=23/82)	26 % (n=5/19)	0 %	0 %
Peak 4	23 % (n=19/82)	21 % (n=4/19)	17 % (n=5/29)	20 % (n=1/5)
Dip/trough 5	2 % (n=2/82)	5 % (n=1/19)	0 %	0 %

Key:

- 1 Rising: PTT worst in low-frequencies with at least a 10 dB difference in the low-, high- and mid-frequencies.
- 2 Flat: PTT within a 10 dB range including 500 Hz to 4 000 Hz.
- 3 Falling: PTT worst in high-frequencies with at least a 10 dB difference in the low-, high- and mid-frequencies.
- 4 Peak: Hearing loss present in both low- and high-frequencies with PTT at 2000 Hz at least 10 dB better than all other frequencies.
- 5 Dip: PTT worst in mid-frequencies with at least a 10 dB difference in the low-, high- and mid-frequencies.

There is a wide distribution of audiogram configurations in the different types of MD, especially in subjects with definite MD. Of interest is that some subjects with vestibular MD/RV, with hearing thresholds below 25 dB, displayed a peak type audiogram configuration. Although hearing thresholds were regarded as normal (0 to 25 dB), it shows the importance of describing the audiogram configuration even when hearing is apparently normal, as it may be indicative of early cochlear involvement in an apparently normal hearing ear.

4.3.1.1.2 The influence of age on PT audiometry results

MD was first described by Prosper Ménière in 1861 as a combination of hearing loss, tinnitus and vertigo. Although the hearing loss may not be the most disabling symptom, most patients regardless of age describe their hearing loss as problematic (Mateijsen et al., 2001:379). A visual representation of the PTA in different age groups is presented in Table 4.7. The results were calculated for all 135 subjects diagnosed with MD, representing 188 diseased ears.

Table 4.7: PTA in the different age groups representing the subjects.

Age (in years)	Normal Hearing (0-15dB)	Very Mild Hearing Loss (16-25dB)	Mild Hearing Loss (26-40dB)	Moderate Hearing Loss (41-55dB)	Moderate-Severe Hearing Loss (56-70dB)	Severe Hearing Loss (71-90dB)
20-30	13 % (n=6/45)	12 % (n=3/26)	2 % (n=2/86)	6 % (n=1/17)	0 %	0 %
30-40	7 % (n=3/45)	12 % (n=3/26)	12 % (n=10/86)	12 % (n=2/17)	9 % (n=1/11)	0 %
40-50	56 % (n=25/45)	42 % (n=11/26)	26 % (n=22/86)	29 % (n=5/17)	46 % (n=5/11)	0 %
50-60	24 % (n=11/45)	23 % (n=6/26)	31 % (n=27/86)	41 % (n=7/17)	9 % (n=1/11)	67 % (n=2/3)
60-70	0 %	12 % (n=3/26)	29 % (n=25/86)	12 % (n=2/17)	36 % (n=4/11)	33 % (n=1/3)

There is a wide distribution of degree of hearing loss among the different age groups of subjects diagnosed with MD. All age groups, apart from the 20 to 30 years group displayed hearing losses ranging between very mild and moderately severe. The results did not indicate a correlation between the age of subjects and the degree of hearing loss. Mateijisen et al. (2001:381) found similar results. The effect of age on audiogram configurations was also investigated. The results for 135 subjects, representing 188 diseased ears are summarized in Table 4.8.

Table 4.8: Distribution of audiogram configurations in different age groups.

Age	Audiogram Configurations						
	Rising ¹	Flat ²	Falling ³	PeakI ⁴	PeakII ⁵	PeakIII ⁶	Dip ⁷
20-30 years	8 % (n=1/13)	13 % (n=8/63)	4 % (n=2/47)	0 %	23 % (n=7/31)	14 % (n=1/7)	0 %
30-40 years	8 % (n=1/13)	10 % (n=6/63)	11 % (n=5/47)	35 % (n=7/20)	0 %	14 % (n=1/7)	14 % (n=1/7)
40-50 years	38 % (n=5/13)	43 % (n=27/63)	16 % (n=7/47)	20 % (n=4/20)	45 % (n=14/31)	14 % (n=1/7)	29 % (n=2/7)
50-60 years	30 % (n=4/13)	23 % (n=15/63)	35 % (n=16/47)	20 % (n=4/20)	23 % (n=7/31)	29 % (n=2/7)	14 % (n=1/7)
60-70 years	16 % (n=2/13)	11 % (n=7/63)	35 % (n=16/47)	25 % (n=5/20)	9 % (n=3/31)	29 % (n=2/7)	43 % (n=3/7)

Key:

- 1 Rising: PTT worst in low-frequencies with at least a 10 dB difference in the low-, high- and mid-frequencies.
- 2 Flat: PTT within a 10 dB range including 500 Hz to 4 000 Hz.
- 3 Falling: PTT worst in high-frequencies with at least a 10 dB difference in the low-, high- and mid-frequencies.
- 4 Peak: Hearing loss present in both low- and high-frequencies with PTT at 2000 Hz at least 10 dB better than all other frequencies.
- 5 Dip: PTT worst in mid-frequencies with at least a 10 dB difference in the low-, high- and mid-frequencies.

Falling hearing losses were present in all age categories, but the occurrence did slightly increase with age. It is important to differentiate between high-frequency sensorineural hearing loss in MD and presbycusis. Ninety five percent (n=45/47) of subjects with falling losses were under the age of 65 years. Also, 64 % (n=30/47) of subjects with sloping audiograms had unilateral losses, which is not characteristic of presbycusis. According to Weinstein (1994:568) there is only a statistic increase for high-frequency sensorineural

hearing loss due to presbycusis from the age of 65 years. The high-frequency hearing loss experienced by the subjects can therefore be ascribed to MD and not presbycusis. The high incidence of unilateral hearing loss increases the risk of a retrocochlear suspicion and emphasizes the need for differential diagnosis (Weber & Adkins, 1997:978).

4.3.1.2 Relationship between PT audiometry results and the symptoms experience by the subjects

Any change in the auditory function of patient's with MD can result in the manifestation of other symptoms such as tinnitus, reduced SD ability and recruitment (Vesterhauge, 1996:7). Understanding the changes occurring in the cochlea of patients with MD can help to explain some of the symptoms experienced during the course of the disease (Vesterhauge, 1996:6). The calculations in Table 4.9 were based on 135 subjects with MD, representing 188 affected ears.

Table 4.9: Distribution of audiogram configurations in subjects with fluctuation hearing loss, tinnitus and Tullio phenomenon.

Audiogram configuration	Fluctuating hearing loss (n=30/188)	Tinnitus (n=127/188)	Tullio (n=12/188)
Rising	7 % (n=2/30)	3 % (n=4/127)	0 %
Flat	36 % (n=11/30)	44 % (n=55/127)	41 % (n=5/12)
Falling	10 % (n=3/30)	25 % (n=32/127)	8 % (n=1/12)
Peak I	7 % (n=2/30)	6 % (n=8/127)	17 % (n=2/12)
Peak II	30 % (n=9/30)	12 % (n=15/127)	17 % (n=2/12)
Peak III	7 % (n=2/30)	7 % (n=9/127)	17 % (n=2/12)
Dip	3 % (n=1/30)	3 % (n=4/127)	0 %

Table 4.9 shows that fluctuating hearing losses occurred in combination with all five audiogram configuration types, but was most evident in flat and peak II types (25 to 60 dB). This study as well as that of Mateijsen et al. (2001:385) could not support the finding, as suggested in the literature, that low-frequency hearing loss tends to fluctuate and a flat loss tends to remain stable (Sakurai et al., 1991:92; Schessel et al., 1998:2680).

Forty four percent (n=55/127) of the subjects with tinnitus had flat audiogram configurations. This high incidence is probably due to the fact that as mentioned earlier in the results that a large number of subjects with normal or very mild hearing losses experienced tinnitus. This group with normal or very mild hearing losses is also represented by a large number of subjects with flat audiogram configurations.

MD pursues an unpredictable course marked by periods of exacerbation and remission of symptoms. In most patients, however, it seems that during the course of the disease, cochlear symptoms (hearing loss and tinnitus) increase and vestibular symptoms (attacks of vertigo with nausea and vomiting) become less prominent (Barber, 1983:31). The distribution of symptoms experienced in each level of hearing was investigated to determine if a relationship between the level of hearing and the different symptoms experienced by patients with MD exist.

Table 4.10: Distribution of symptoms compared to degree of hearing loss (n=135).

Symptoms	Normal Hearing 0-15dB	Very Mild Hearing Loss 16-25Db	Mild Hearing Loss 26-40dB	Moderate Hearing Loss 41-55dB	Moderate-Severe Hearing Loss 56-70dB	Profound Hearing Loss 71-90dB
Vertigo (n=130/135)	29 % (n=38/130)	21 % (n=27/130)	40 % (n=53/130)	5 % (n=6/130)	5 % (n=6/130)	0 % (n=0/130)
Tinnitus (n=96/135)	15 % (n=14/96)	17 % (n=16/96)	41 % (n=39/96)	13 % (n=13/96)	8 % (n=8/96)	6 % (n=6/96)
Aural pressure (n=72/135)	45 % (n=32/72)	14 % (n=10/72)	30 % (n=21/72)	6 % (n=5/72)	5 % (n=4/72)	0 % (n=0/72)
Nausea / vomiting (n=114/135)	21 % (n=24/114)	20 % (n=23/114)	40 % (n=46/114)	10 % (n=11/114)	7 % (n=8/114)	2 % (n=2/114)
Fluctuating hearing loss (n=24/135)	0 % (n=0/24)	30 % (n=7/24)	46 % (n=11/24)	12 % (n=3/24)	12 % (n=3/24)	0 % (n=0/24)
Tullio (n=8/135)	25 % (n=2/8)	37 % (n=3/8)	13 % (n=1/8)	0 % (n=0/8)	25 % (n=2/8)	0 % (n=0/8)

The degree of hearing loss of subjects in this study was not directly related to the symptoms that they experienced. It is interesting to note that 73 % (n=69/96) of subjects with either normal hearing or mild hearing loss experienced tinnitus. This is not surprising taking into account the high percentage of subjects with vestibular MD/RV also experiencing tinnitus (Table 4.4). This finding correlates well with the research results of Paparella and Mancini (1985:150) who found 84 % (n=43/51) of subjects with vestibular MD to have tinnitus. These results confirm that tinnitus in MD is not related to hearing loss and is probably, as mentioned earlier in the study, caused by a moderate increase in potassium concentration of the perilymph of the scala tympani towards the apex of the cochlea (Horner, 1991:154).

There was also a high incidence (45 % - n=32/72) of aural pressure in subjects with normal PTT. This high number is expected since 97 % (n=28/29) of subjects in the vestibular MD/RV group also complained of aural pressure (Table 4.1). Paparella and Mancini (1985:149) also found a very high percentage (98 % - n=50/51) of their subjects with normal hearing complained of a pressure or fullness in the ear.

Although fluctuating losses did occur in subjects with different degrees of hearing loss, they were more common in the very mild (16 to 25 dB) and mild hearing loss (26 to 40 dB) categories. This finding supports previous reports indicating that fluctuating hearing loss is more common in the early stages of the disease or when only a mild hearing loss is present (Barber, 1983:28; Mateijssen et al., 2001:379). Sixty two percent (n=5/8) of subjects who experienced the Tullio phenomenon had hearing thresholds below 25 dB, with half (4/8) of the subjects audiograms presenting with a peak type audiogram. No figures for comparison could be obtained from previous studies and the significance of these results is not clear.

In summary, it is clear that PT audiometry results are not related to the symptoms of subjects with MD, but both PT audiometry results and the symptoms experienced by the subjects will be influenced by the spread of biochemical alteration in perilymphatic fluid in the cochlea. This shows the importance of combining the case history with PT audiometry results to accurately describe patient's cochlear status at the time of clinical examination.

4.3.1.3 Degree of hearing loss and audiogram configuration in MD

Although many earlier studies tended to describe audiometric configurations of subjects with MD as being a low-frequency loss during the early stage of the disease and flat in later stages, this study and others have identified a wide variety of configurations, including peak configurations with losses in both the low- and high-frequencies, audiograms with high-frequency losses, dip or trough patterned losses, and audiograms showing rising and flat losses (Barber, 1983:27; Elichar et al., 1973:41; Goodman, 1965:992; Lee et al., 1995:527; Paparella et al., 1982:558). A summary of the audiogram configurations as obtained in this study is presented in Figure 4.3. The calculations were based on 135 subjects with MD, representing 188 affected ears.

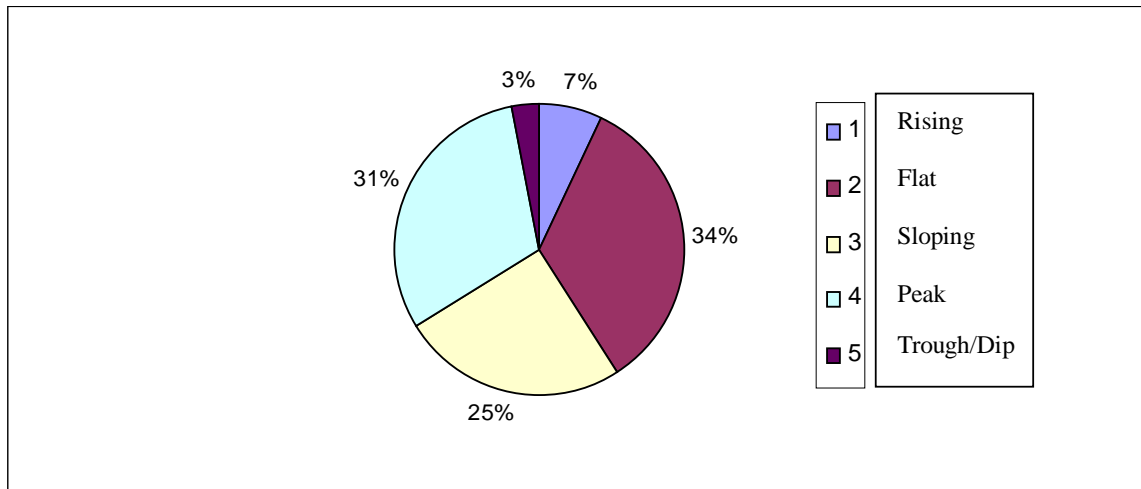


Figure 4.3: Distribution of audiogram configurations.

As depicted in Figure 4.3, the hearing impairment in the affected ear(s), as measured by PT audiometry, could be differentiated in five different types of configurations namely 7 % (n=13/188) rising, 34 % (n=63/188) flat, 25 % (n=47/188) falling, 31 % (n=59/188) peak and 3 % (n=6/188) dip.

The most common type audiogram configurations obtained in this study were flat and peak audiograms, which occurred in 65 % (n=122/188) of the affected ears. Of the 188 ears affected 59 (n=31 %) presented some evidence of a peak type audiogram, with 11 % showing a peak audiogram although hearing thresholds were above 25 dB. The classic low frequency sensorineural hearing loss appeared in only 7 % (n=13/188) of the cases.

These results correlate with studies performed by Meyerhoff et al. (1981:221) and Oosterveld (1980:888) who found that flat and peak types audiograms occur in about 70 % (n=320/457) of cases with MD. They also found high-frequency losses to be the third most common configuration in their study. According to Ginsberg and White (1994:20) these high frequency losses will flatten as the disease progresses and the low frequencies are also affected. Paparella and Griebie (1984:235) also found a high incidence of peak type audiogram configurations (42 % - n=151/360) in subjects with MD. Since the peak audiogram, especially type II peaks (25 to 60 dB), contains predominantly low-frequency losses, it is possible that earlier authors referred to these as low-frequency losses, not taking into account the high-frequency component. Similarly, flat losses could have been interpreted to include type III peak audiograms (61 to 90 dB) as described herein.

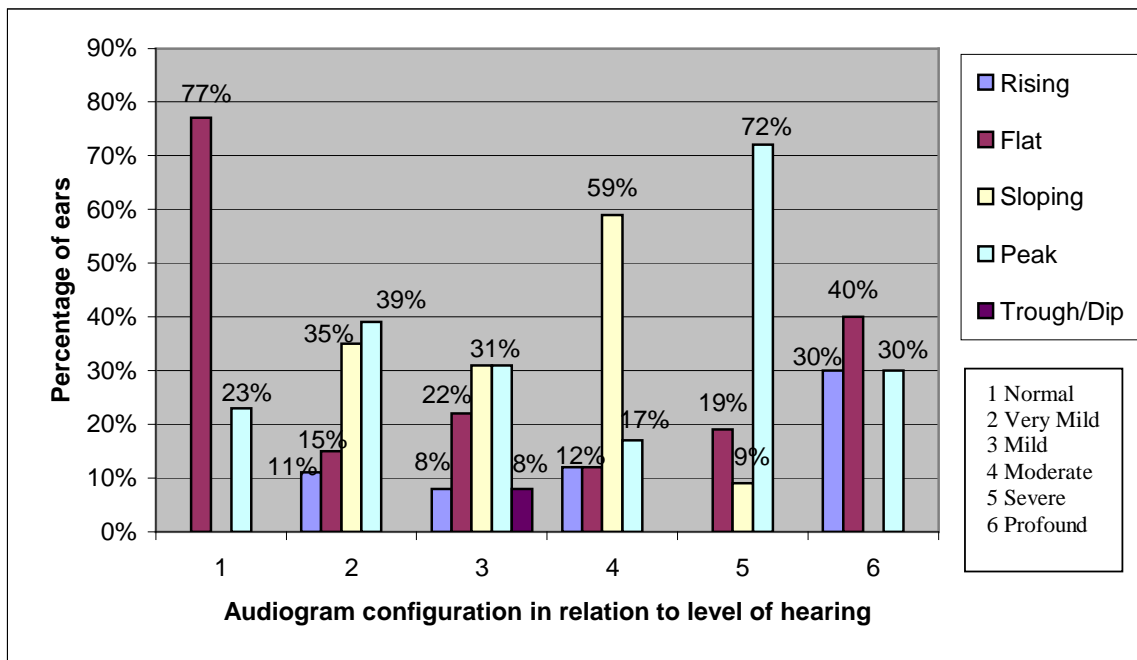


Figure 4.4: Audiogram configurations and the degree of hearing loss (n=188).

Figure 4.4 does not clearly show this correlation between audiogram configuration and average hearing loss. On the other hand, Mateijsen et al. (2001:385) found a significant correlation between audiogram shape and the average hearing loss in subjects with MD and therefore further investigations were performed.

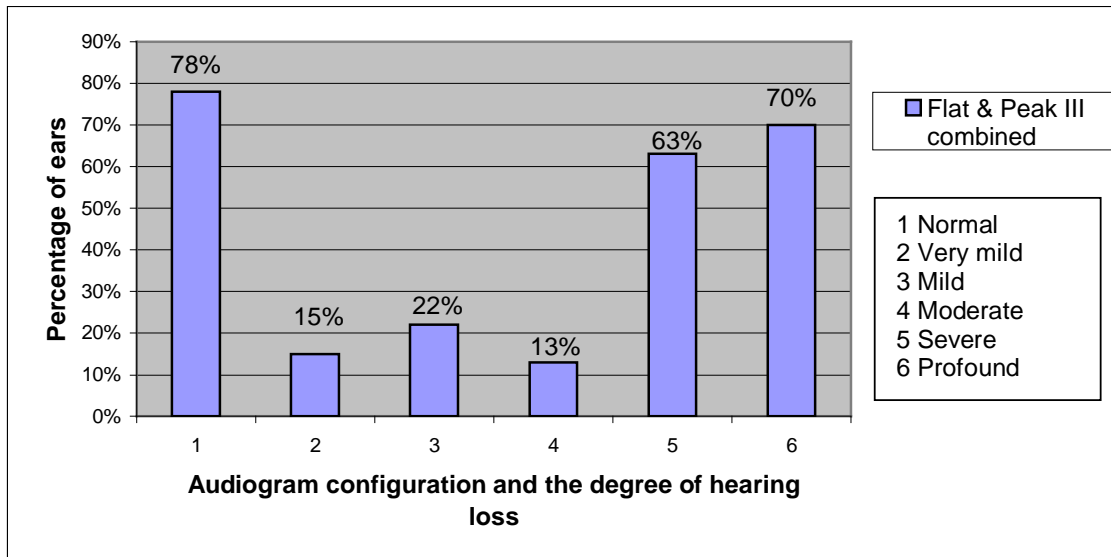


Figure 4.5: Combination of flat and peak III type configurations (n=70).

Lee et al. (1995:530) suggested that it is possible that in the past type III audiograms were considered as flat types and therefore the results were also interpreted with type III audiograms considered part of flat type audiograms (Figure 4.5). Reviewing the results in this way combining type III and flat type audiograms does show a significant increase of these types of audiograms as hearing loss progresses. The high incidence in subjects with normal hearing is not surprising, since normal PTTs are presented by flat type audiograms. This is in accordance with the description generally given of the course of MD (Barber, 1983:28). This study further confirms the findings of audiologic data reported in the literature on MD (Lee et al., 1995:530; Mateijssen et al., 2001:385). Of interest in this study was the finding that of all patients who underwent audiologic assessment 31 % (n=59/188) demonstrated a peak audiometric pattern. This study, together with Lee et al. (1995:527), suggests that this type of configuration may be a diagnostic feature of MD.

Although PT audiometry results should not be used in isolation it will, as part of a test battery, assist clinicians to determine which ear, if not both, is affected by MD (Balkany et al., 1980:603). Apart from type and degree of hearing loss, PT audiometry can also provide valuable information in determining the laterality of the disease.

4.3.1.4 PT audiometry results and laterality

PT audiometry is routinely used in the investigation of MD. Considering both PTT and audiogram configuration can, as part of the test battery, assist in determining which ear, if not both, is affected by the disease (Lee et al., 1995:527). Figure 4.6 provides a summary of the distribution of the audiogram configurations in unilateral and bilateral disease.

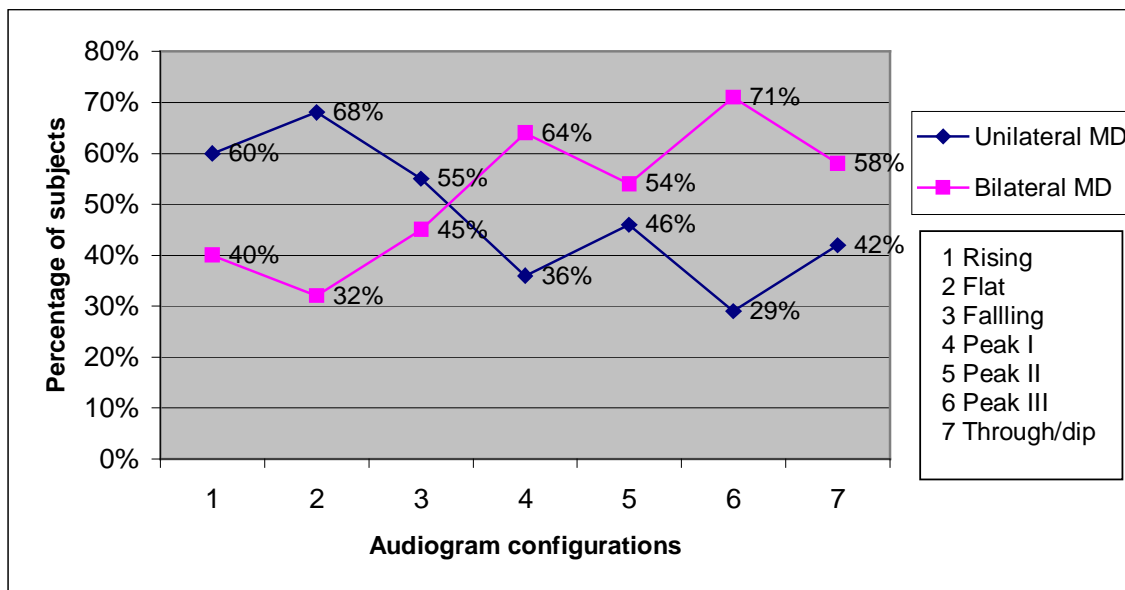


Figure 4.6: Distribution of audiogram configurations in unilateral and bilateral disease (n=135).

In this study, rising and flat audiogram configurations were more common in unilateral disease, with the peak type audiograms (especially type I and III) more common in bilateral disease. All types of audiogram configurations were found in both unilateral and bilateral disease and no distinction between unilateral and bilateral disease can be made on the basis of audiogram configuration alone. Mateijsen et al. (2001:385) found similar results, confirming that there is no significant difference between audiogram configurations in unilateral and bilateral disease.

It is interesting to note that 2 % (n=3/135) of subjects in this study exhibited a peak type audiogram configuration in the contralateral ear, but these were not included among the confirmed bilateral cases. Since the incidence of the peak audiogram has been found to be significantly greater in MD subjects than in non-MD groups (Paparella et al., 1982:555), it

was felt that many of these subjects might be showing early end-organ involvement of the opposite ear. If all of these subjects were to develop clinical disease in this opposite ear, the percentage of bilateral involvement would approach 41 %. This correlates with the findings of Paparella and Griebie (1984:235) who reported that 35 % (n=126/360) of patients with PTT below 25 dB revealed a peak type audiogram.

4.3.1.5 SD results in subjects with MD

SD scores were obtained and interpreted according to the test protocol proposed by Martin (1994:136-139). The protocol is described on page 47. The effect of the level of hearing loss on SD scores is summarized in Table 4.11.

Table 4.11: The relationship between SD scores and PTTs (n=188).

PTT	Normal SD	<50 % SD	50-70 % SD	>70 % SD
Normal Hearing (0-15dB)	100 % (n=45/45)	0 %	0 %	0 %
Very Mild Hearing Loss (16-25dB)	100 % (n=26/26)	0 %	0 %	0 %
Mild Hearing Loss (26-40dB)	97 % (n=83/86)	0 %	0 %	3 % (n=3/86)
Moderate Hearing Loss (41-55dB)	76 % (n=13/17)	0 %	18 % (n=3/17)	6 % (n=1/17)
Moderate-severe Hearing Loss (56-70dB)	11 % (n=1/11)	0 %	78 % (n=9/11)	11 % (n=1/11)
Severe Hearing Loss (71-90dB)	0 %	100 % (n=3/3)	0 %	0 %

Some researchers found poor SD scores as a common symptom of MD (Merchant et al., 1995:68; Shea, 1993:224). It is important to attempt to differentiate if a deterioration in SD ability is caused purely by a deterioration in PTTs or if the effect of MD on the cochlea results in lower SD scores than can be attributed to the degree of hearing loss only. The results in Table 4.11 show that SD scores were only affected in subjects demonstrating a hearing loss of above 40 dB. For more severe losses, the discrimination ability decreased with increasing hearing loss. The results of Mateijssen et al. (2001:386) also correlated with the findings of this study. Ginsberg and White (1994:20) also noted SD to be affected in subjects with MD, but found it to be in keeping with PTAs. According to Schuknecht (1963:658) SD is particularly affected during and immediately following an attack. SD scores will probably vary depending on when last the subject experienced an attack and when the next one will occur. Subjects who did not achieve a 100 % SD score demonstrated a plateau curve speech audiogram, which is in keeping with cochlear disease (Penrod, 1994:159). None of the patients presented with a roll-over (retrocochlear) type curve, further confirming that the sensorineural hearing loss found in MD is a result of cochlear rather than a retrocochlear pathology (Ginsberg & White, 1994:20). The degree of sensorineural hearing loss as well as the audiogram configurations could influence a subject's ability to discriminate speech. A visual representation of the effect of audiogram configuration of SD scores is presented in Table 4.12.

Table 4.12: Audiogram configuration in SD (n=188).

Audiogram configuration	Normal SD	<50%	50-70%	>70%
Rising	71 %	9 %	9 %	9 %
Flat	97 %	1 %	1 %	1 %
Falling	93 %	0 %	5 %	2 %
Peak I	100 %	0 %	0 %	0 %
Peak II	46 %	50 %	4 %	0 %
Peak III	12 %	17 %	42 %	29 %
Dip/trough	100 %	0 %	0 %	0 %

SD scores deteriorated in subjects with peak II and peak III type audiogram configurations. This can be explained if the PTAs for these groups are taken into account. Most patients in these two groups demonstrated a moderate to severe sensorineural hearing loss. Therefore it

seems that SD is more affected by the degree of hearing loss and not by audiogram configuration. This correlates with the results of Mateijssen et al. (2001:385) who concluded that the audiogram shape did not have any significant additional effect on SD.

A literature review of audiometric tests results in MD revealed that the duration and frequency of the attacks of vertigo can influence the PTT as well as the patient’s SD ability and therefore the effect of the duration of the disease on the subjects were included in this study (Sakurai et al., 1991:92).

4.3.1.6 The effect of the duration and frequency of attacks of vertigo in MD

The duration of the disease and the frequency of vertiginous attacks were recorded in Column 28: Frequency of attacks and Column 29: Duration of disease of the electronic spreadsheet. An understanding of the effect of the duration of the disease as well as the frequency of attacks on both the subject and auditory function is necessary to plan effective patient management. Table 4.13 provides a summary of the duration of the disease experienced by subjects in this study.

Table 4.13: Duration of the disease (n=135).

Period	Percentage of subjects
<1 month	21 % (n=28/135)
1-3 months	17 % (n=23/135)
3 months – 1 year	30 % (n=41/135)
1 year – 5 years	27 % (n=36/135)
> 5 years	5 % (n=7/135)

The period from the first attack of vertigo until subjects visited the ENT clinic varied greatly (Table 4.13). Thirty-eight percent (n=51/135) of the subjects in this study consulted the ENT within the first three months of experiencing attacks of vertigo, whereas 5 % (n=7/135) of the subjects came more than one year after the onset of vertigo. Wada et al. (1990:116) found similar results. Although this study as well as the study of Wada et al. (1990:116) indicated that a large number of subjects were diagnosed within the first year, there are still a percentage of subjects who experience symptoms for more than five years without a diagnosis. It has been reported that patients who experience a set of symptoms without any

explanation relating to the cause experience increased levels of stress affecting quality of life (Vesterhauge, 1996:11). This emphasizes the need for clinicians to improve the efficacy of diagnostic protocols to provide patients with an accurate diagnosis as soon as possible. The effect of the number and frequency of the attacks on the PTT were investigated and the results are presented below.

Table 4.14: The effect of duration, number and frequency of attacks on PTAs (n=135).

	Normal Hearing 0-15dB	Very Mild Hearing Loss 16-25dB	Mild Hearing Loss 26-40dB	Moderate Hearing Loss 41-55dB	Moderate-Severe Hearing Loss 56-70dB	Severe Hearing Loss 71-90dB
<u>Duration of disease</u>						
0 – 1 months	23 %	28 %	17 %	0 %	9 %	0 %
1 –3 months	18 %	17 %	20 %	19 %	9 %	33 %
3 months – 1 year	44 %	24 %	24 %	19 %	45 %	0 %
1 – 5 years	11 %	7 %	28 %	49 %	45 %	33 %
> 5 years	4 %	10 %	6 %	0 %	0 %	34 %
<u>Number of attacks</u>						
No attacks	0 %	0 %	63 %	37 %	0 %	0 %
1 attack	4 %	12 %	77 %	0 %	7 %	0 %
2 attacks	31 %	37 %	32 %	0 %	0 %	0 %
3 – 10 attacks	30 %	15 %	40 %	10 %	3 %	2 %
> 10 attacks	25 %	10 %	36 %	12 %	10 %	7 %
<u>Frequency of attacks</u>						
No attacks	0 %	0 %	62 %	38 %	0 %	0 %
1 attack	4 %	12 %	77 %	0 %	7 %	0 %
Weekly	55 %	19 %	31 %	4 %	2 %	0 %
Monthly	26 %	9 %	37 %	13 %	10 %	5 %
Yearly	10 %	20 %	52 %	10 %	0 %	8 %

The results presented in Table 4.14 do not indicate a relationship between number and frequency of the attacks and the degree of the hearing loss. In some subjects the sensorineural hearing loss appeared to exceed 56 dB after only a few attacks, while in other patients who had suffered more than ten attacks displayed PTA of not more than 20 dB. Oosterveld (1980:888) also did not find a relationship between the number, duration and frequency of the attacks and permanent damage to the hearing. The degree of permanent hearing loss is

probably related to the severity of the attacks and the specific anatomical site of endolymphatic hydrops (Paparella & Mancini, 1985:150).

Although the duration of the disease did seem to have an influence on the hearing thresholds in some subjects, there was no clear relationship. Some subjects with a severe hearing loss have experienced symptoms for less than three months and some subjects with mild loss for more than five years. The lack of correlation between audiometric data and the duration of unilateral disease is not unexpected. Previous studies have also shown little or no correlation between subjective or objective measures of the severity of the disease and the duration (Mateijssen et al., 2001:385). It remains crucial, however, to perform follow up audiograms since the hearing loss associated with MD may take one of the following four difference courses: Loss of hearing increases as the disease develops, hearing remains unchanged, hearing improves or progresses from unilateral to bilateral disease with involvement of hearing in both ears (Sakurai et al., 1991:95).

Although only 36 subjects returned for follow-up audiograms a comparison was still made of the hearing thresholds obtained at first and last examinations in all subjects during the remission period. According to the results, the subjects were classified into one of the following three groups: hearing deteriorated/unchanged/improved. These groups were also compared in terms of the frequency of vertigo attacks and the duration of the illness.

Table 4.15: Influence of frequency of attacks on hearing thresholds over time (n=36).

Follow up audiograms	Frequency of attacks	Average duration of disease
Deterioration in hearing threshold 38 % (n=14/36)	1 attack (22 %) weekly (34 %) monthly (44 %)	2 years
Unchanged hearing 54 % (n=19/36)	1 attack (23 %) weekly (45 %) monthly (32 %)	2.5 years
Hearing improved 8 % (n=3/36)	Weekly (100 %)	1 year

Hearing thresholds deteriorated in 38 % (n=14/36) ears, remained unchanged in 54 % (19/36) ears and improved in 8 % (3/36) ears. No significant correlation was noted to exist between

hearing loss type at first examination and subsequent progress of the hearing loss. This is in keeping with the result of Sakurai et al. (1991:95).

A comparison of the groups with deterioration in hearing thresholds and those with unchanged hearing, showed few differences in respect of the duration of the disease, and frequency of attack (Table 4.15). The main difference noted between the first two groups and the group with improved hearing is the shorter duration of the disease with regular attacks of vertigo. This correlates with other studies (Sakurai et al., 1991:97). The frequency as well as duration of attacks in the group with improved hearing might suggest that the improvement is caused by remissions that occurred earlier in the disease (Barber 1983:31).

Only 41 % of the subjects diagnosed with vestibular MD/RV returned for follow up audiometric testing and treatment. Follow-up audiograms were recorded between three and six years after the initial diagnosis. A hearing loss was recorded in 33 % (n=8/24) of these subjects, confirming that although no hearing loss was recorded, MD was the underlying cause for the initial symptoms experienced by these subjects. Paparella and Mancini (1985:149) also found that approximately 20 % (10/51) of their subjects with vestibular MD/RV develop definite MD with follow-up examination.

4.3.1.7 Summary of the significance of PT audiometry and SD scores in the diagnosis of MD

Concepts of pathogenesis and considerations of pathophysiology relative to symptoms can be inferred from the variety of audiometric configurations in this study. Although low-frequency losses occurring alone were uncommon, low-frequency losses together with high-frequency losses were common. Furthermore, in some cases, high-frequency losses were seen to occur alone. The main correlation between studies of temporal bones of patients with MD has been endolymphatic hydrops of the pars inferior (cochlea and saccule), with hydrops present in the basal, middle and apical turns of the cochlea (Rauch et al., 1989:882). In recent physiological studies of animals with induced hydrops Horner (1991:150) has demonstrated high-frequency hearing losses. He attributed this to the scala media that has expanded maximally into the scala vestibule due to increased volume of the perilymph (Horner, 1991:153). This occurrence, in association with endolymphatic hydrops of the cochlea (especially including the basal turn of the cochlea), could interfere with conduction of the sound wave and possibly explain the high-frequency losses observed in this study.

It seems that the average loss of hearing that can be expected by patients with MD is a moderate sensorineural hearing loss. This hearing loss will occur regardless of the patient's age. Symptoms seem to be more related to the different stages of the disease than the level of hearing loss. PT audiometry is sensitive to demonstrate early cochlear damage and assist in diagnosing laterality of the disease. Follow-up audiograms also proved valuable especially in subjects with vestibular MD/RV. These do not indicate a relationship between number and frequency of the attacks and permanent damage to the hearing.

4.3.2 The significance of DPOAE test results in the diagnosis of MD

DPOAE testing was performed on all subjects presenting with a sensorineural hearing loss, which constituted 130 ears diagnosed with MD. This is an objective, quick and non-invasive measure of determining the structural basis of a hearing problem and to detect early cochlear dysfunction (Cianfrone et al., 2000:111). Since it is also used to determine the primary site of pathological involvement, retrocochlear pathology can be ruled out at the initial consultation with the patient (Cianfrone et al., 2000:112). The DPOAE test results were recorded in Column 19: DPOAE left ear and Column 20: DPOAE right ear of the electronic spreadsheet.

The presence of DPOAE was examined by interpreting the results independently for each patient, determining if emissions were normal or abnormal. Abnormal emissions are indicative of cochlear damage specifically to the outer hair cells (Cianfrone et al., 2000:111). Emissions for each frequency were analysed individually. Emissions were interpreted as normal if the DP amplitude minus the noise floor difference exceeded 5 dB and if the DP amplitude was within the normative data for the Madsen Celesta 503 Cochlear Emissions Analyzer (Hall & Mueller, 1997:255). Emissions with decreased amplitudes were classified as emissions 5 dB above the noise floor, but below the normative data and an absent emission was classified as an emission with an amplitude less than 5 dB above the noise floor. Emissions were analysed in terms of amplitude and frequency. Seventy five percent (6/8) of the emissions had to be present for the DPOAE to be considered as normal for the test ear (Hall & Mueller, 1997:255). The results are summarized in Figure 4.7 and Figure 4.8.

4.3.2.1 DPOAE amplitude of MD subjects

DPOAEs are an objective, non-invasive and quantitative measure of sensory cell function in the cochlea. It provides information regarding outer hair cell activity, and is regarded as a reliable test for determining the structural basis of a hearing problem, particularly if it involves the outer hair cell system (Norton & Stover, 1994:448).

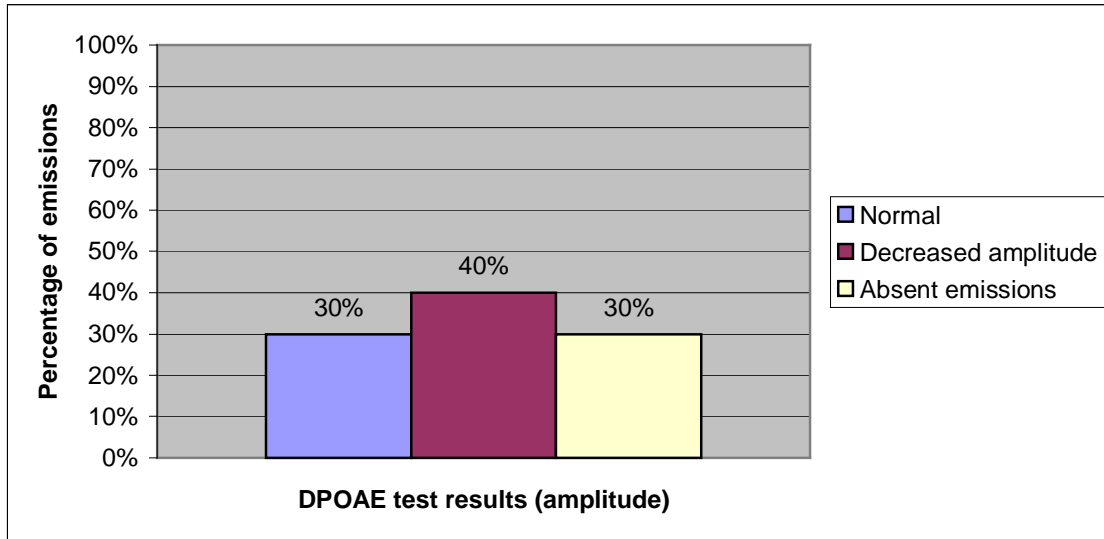


Figure 4.7: Presentation of DPOAE in terms of amplitude (n=130).

According to Figure 4.7 30 % (n=39/130) of the emissions were normal, 40 % (n=52/130) were present but with decreased amplitudes and 30 % (n=39/130) were absent. Cianfrone et al. (2000:111) also found more than 60 % of the ears with MD displayed abnormal DPOAEs despite the presence of an average PTT level below 40 dB HL. The high incidence of abnormal emissions indicates abnormal functioning of the outer hair cells. This is to be expected in subjects diagnosed with MD which leads to a cochlear site of lesion (Ginsberg & White, 1994:20). The presence of some emissions indicates that not all the hair cells are damaged at the same rate.

DPOAEs are considered to be valuable for correlating audiometric findings (Cianfrone et al., 2000:112). The presence or absence of DPOAEs was correlated with audiometric threshold values. The comparison between DPOAEs amplitudes and PTT displayed the presence of greater cochlear activity in the region where the hearing threshold was within normal limits. This was clearly demonstrated in peak type audiograms where low- and high-frequencies

were more affected than mid-frequencies. These results are compatible with those found by Cianfrone et al. (2000:114). According to the DPOAE results, 50 % (n=13/26) of subjects with very mild hearing loss showed evidence of cochlear pathology. Sakashita et al. (1998:76) also found that measurement of the DPOAE amplitude can detect small changes in cochlear function which do not lead to changes in hearing threshold, and has higher sensitivity than conventional PT audiometry in the monitoring of cochlear function. The results of this research project also suggest that cochlear function may change in subjects with MD, even within the frequency region in which hearing thresholds do not change, and that DPOAEs should be included as part of the test battery in the diagnosis of MD.

4.3.2.2 DPOAE test results in terms of frequency

DPOAEs are frequency related and can therefore be correlated with audiometric findings to identify the site of lesion of the hearing loss (Cianfrone et al., 2000:112). Emissions were therefore described in terms of the frequency at which they were elicited. The frequencies were divided according to the f2 value with low frequency from 500 to 1006 Hz, mid frequency 1512 to 2011 Hz and high frequency from 3023 to 6060 Hz. One hundred and thirty ears were tested at six different frequencies. Two hundred and sixty emissions were tested for each frequency category (low, mid and high).

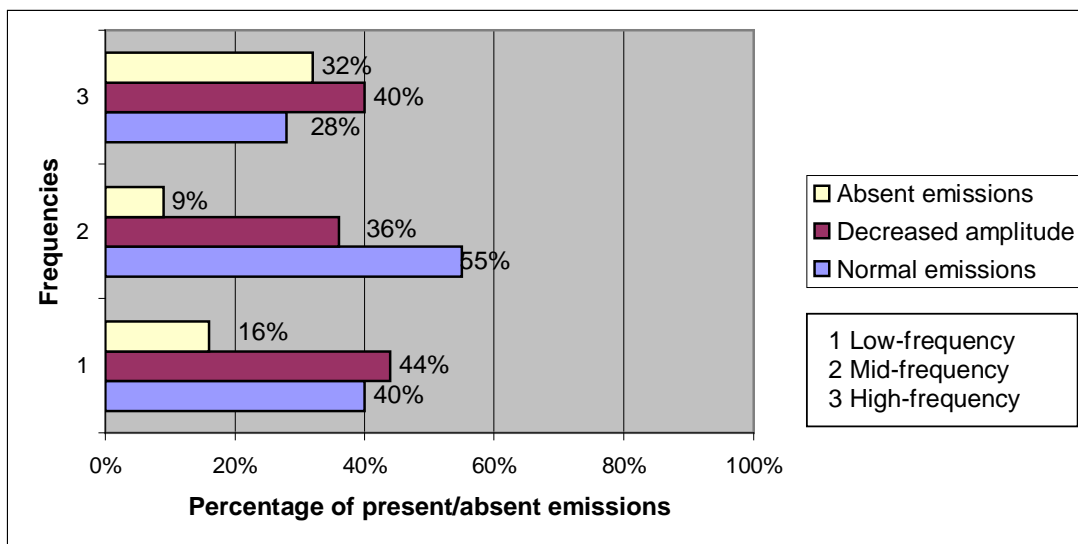


Figure 4.8: Presentation of DPOAE test results according to frequency (n=130).

Only 40 % (n=104/260) of the emissions at the low-frequencies were normal. Forty four percent (n=114/260) was present but with decreased amplitudes, and 16 % (n=42/260) was absent. In the mid-frequency range 55 % (n=143/260) were normal, 36 % (n=94/260) present with decreased amplitudes and only 9 % (n=23/260) absent. Twenty eight percent (n=73/260) of emissions in the high-frequency range were normal, 40 % (n=104/260) showed a decreased amplitude and 32 % (n=83/260) of emissions were absent. The increase in normal emissions in the mid-frequencies is in accordance with the high incidence (31 % - n=59/188, Figure 4.3) of peak type audiograms in this study.

4.3.2.3 Summary of the significance of DPOAE test results in the diagnosis of MD

Abnormal OAEs have been shown to be a highly sensitive, but non-specific indicator of cochlear dysfunction (Sakashita et al., 1998:70). Patients with MD cannot be distinguished from patients with other sensory hearing loss etiologies based on DPOAE findings alone (Hall & Mueller, 1997:273). DPOAE is useful to confirm cochlear pathology and exclude retrocochlear pathology. It is also useful to indicate early damage to the cochlea confirming the site of lesion and should be used as part of a test battery for the diagnosis of MD. DPOAEs test results should be viewed in relation to other tests for example EcoG, to evaluate the cochlear function in patients with MD is EcoG.

4.3.3 The significance of EcoG results in the diagnosis of MD

There has been an increased interest in EcoG for the diagnosis of patients with endolymphatic hydrops. Although there are different ways to interpret EcoG results, the SP-AP amplitude ratio obtained from alternating polarity click responses seems to be the most efficient diagnostic measure to differentiate patients with and without MD (Goin et al., 1982:1386; Levine et al., 1992:615). The EcoG results were recorded in Column 21: EcoG result in left ear and Column 22: EcoG result in right ear of the electronic spreadsheet (Appendix A). The results are summarized in Table 4.16 – 4.18 and Figure 4.9. The results are based on 135 subjects diagnosed with MD, representing 188 affected ears.

Table 4.16: EcoG results in subjects with MD.

MD TYPE		Positive EcoG result in affected ears (88%)	Positive EcoG result in at least one of subject's affected ears (96%)
Definite MD		85 %	98 %
Probable MD		83 %	100 %
Possible MD	Vestibular MD	96 %	100 %
	Cochlear MD	75 %	99 %

Eighty eight percent (n=166/188) of all subjects had a positive EcoG result in the ear diagnosed with MD. The majority (96 % - n=129/135) of subjects diagnosed with unilateral or bilateral MD had a positive EcoG result in at least one of the affected ears. The group with a negative EcoG result in an ear diagnosed with MD (3 % - n=4/135) fell into either the definite MD category or the possible (cochlear) MD category. In 1 % (n=2/135) of the subjects EcoG could not be performed for reasons such as low hearing thresholds or subject's discomfort.

According to Ferraro et al. (1983:69) a negative EcoG in the presence of MD symptoms is not uncommon. It is believed that it may be due, or at least in part, to the dynamic characteristics of the disease which would in turn vary the electrophysiologic status of the inner ear. Morrison et al. (1980:710) suggested that the more certain the diagnosis and the more established the disease, the greater is the likelihood of finding positive EcoG results. On the other hand, Margolis, Rieks, Fournier and Levine (1995:55) suggest that the EcoG test is also most sensitive in early stages of the disease before permanent damage to cochlear structures has occurred. This finding is also demonstrated in this study where subjects with vestibular MD showed a higher percentage of positive EcoGs. The influence of symptoms experienced by subjects with MD in relation to EcoG results is presented in Table 4.17.

Table 4.17: Comparison of EcoG results with regard to the symptoms complex (n=135).

	TRIAD OF SYMPTOMS (Vertigo + tinnitus + hearing loss)	NOT CLASSICAL TRIAD (Atypical MD)
Positive EcoG	85 % (n=115/135)	91 % (n=123/135)

Table 4.17 confirms the results of Margolis et al. (1995:55), indicating that a positive EcoG result does not depend on a diagnosis of definite MD (vertigo, tinnitus and hearing loss) as suggested by Morrison et al. (1980:710). Ninety one percent of patients who did not experience the classical triad symptom complex presented with positive EcoG test results, indicating the presence of endolymphatic hydrops at the time of testing. It is therefore possible to detect endolymphatic hydrops with EcoG testing in subjects with atypical forms of MD.

The presence of endolymphatic hydrops can cause a feeling of aural fullness in the ear (Schessel et al., 1998:2679). Since EcoG is the only proven investigation that can demonstrate objectively the presence of endolymphatic hydrops (Moffat et al., 1992:370) the symptom of aural fullness in conjunction with EcoG results were investigated.

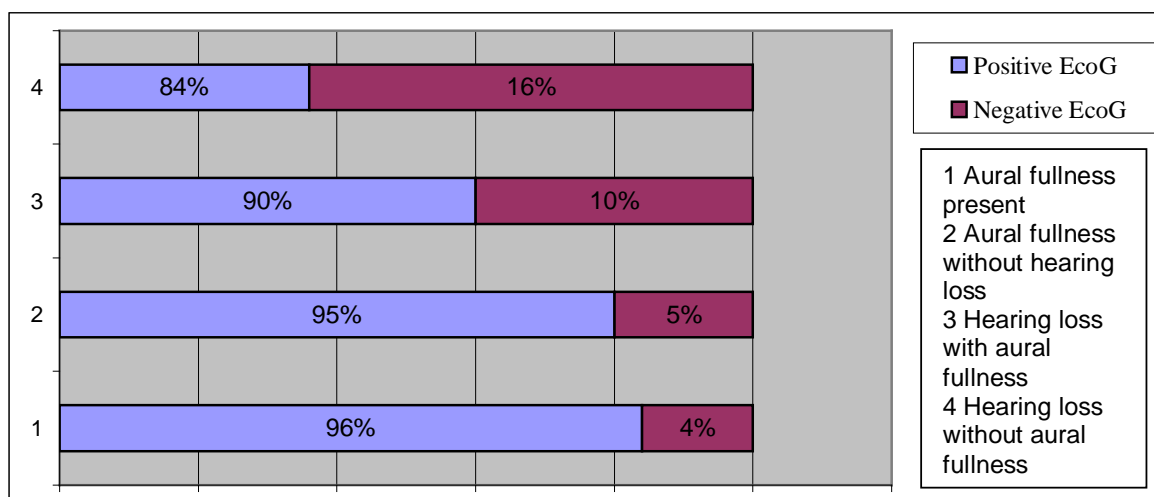


Figure 4.9: Comparison of EcoG results in subjects with aural pressure and hearing loss (n=135).

Ninety six percent (n=129/135) of subjects who complained of fullness in the ear did have a positive EcoG. The results also indicate that aural pressure seems to correlate more strongly with positive EcoG results than hearing loss in subjects with endolymphatic hydrops. Ferraro and Tibbils (1999:22) also found greater sensitivity for EcoG in subjects whose symptoms at the time of testing included hearing loss and aural fullness.

Another interesting occurrence in MD is positive EcoG results in asymptomatic ears. Moffat et al. (1992:370) recorded bilateral abnormalities with EcoG in 35 % of cases diagnosed with unilateral MD. Morrison et al. (1980:711) also noted symptomless ears to show positive

EcoG results and therefore evidence of hydrops. The EcoG results of subjects in this study were reviewed to investigate the manifestation of bilateral abnormalities in unilateral MD. The results are based on 135 subjects diagnosed with MD, constituting 188 affected ears and 82 normal ears. The results are presented in Table 4.18.

Table 4.18: EcoG results in affected and unaffected ears (n=135).

LEVEL OF HEARING	ECOG RESULT
Normal hearing (vestibular MD)	Positive EcoG results in 97% of affected ears
Hearing loss	<u>Unilateral</u> Positive EcoG result in 88 % of affected ears Negative EcoG result in 12 % of affected ears Positive EcoG result in 5 % of unaffected ears <u>Bilateral</u> Bilateral positive EcoG result in 75 % of ears Unilateral positive EcoG result in 25 % of ears

In this study, 97 % (n=28/29) of subjects with vestibular MD displayed positive EcoG results. If the possible diagnosis of vestibular MD is made on clinical grounds, usually on the basis of the history, it may be difficult to exclude vestibular neuronitis (Paparella, 1985:149). Assessing features from the clinical history in conjunction with audiological tests such as EcoG may allow differentiation of these patients. The confirmation of endolymphatic hydrops in patients who present with only portions of the classic symptom complex confirms the diagnosis of MD and also enables the clinician to lateralise the pathology to a specific ear.

EcoG was positive in 88 % (n=72/82) of subjects with unilateral MD. Bilateral abnormalities indicative of endolymphatic hydrops were recorded in 5 % (n=31/53) of subjects with unilateral MD. This might be an indication of a higher incidence of bilateral disease than originally recorded. These results emphasize the need for clinicians to monitor the normal ear in patients with apparent unilateral MD. All patients with bilateral disease had a positive EcoG result in at least one ear. Only 75 % (n=40/53) of subjects with bilateral MD had positive EcoG results in both ears. Although EcoG results are depended on the electrophysiological status of the ear at the time of testing, it still proved to be an effective tool to assist in the diagnosis of MD.

The results were not affected by either the audiogram configuration or the degree of hearing loss.

4.3.3.1 Summary of the significance of EcoG results in the diagnosis of MD

EcoG is a sensitive indicator of electrophysiologic events occurring in the cochlea and first-order cochlear neurons and provides responses of relatively large amplitude (Moffat et al., 1992:372). This study showed EcoG to be an effective diagnostic tool for the diagnosis of MD. Ninety six percent (n=129/135) of subjects clinically diagnosed with MD demonstrated positive EcoG finding. EcoG seems extremely valuable as part of the test battery especially in the diagnosis of patients with vestibular or cochlear MD.

The question arises as to whether these SP changes reflect endolymphatic hydrops per se or some other pathophysiologic phenomenon of MD. Although many studies report enlarged SP's it cannot be said that endolymphatic hydrops per se causes the SP changes seen in MD. Other factors such as biochemical changes, altered circulation or an interaction of factors may play a role (Goin et al., 1982:1386). However, regardless of the specific pathophysiological correlates of the observed changes, they are infrequently seen in other conditions and until further investigations shed light on this issue, EcoG offers the clinician a valuable tool in the diagnosis of MD.

Although PT audiometry and EcoG provide valuable information regarding the functioning of the inner ear, it is important to be aware that the two ears in patients with MD may have dyssynchronous malfunction. The one ear might present with a cochlear site of lesion, while the other ear is responsible for vestibular dysfunction (Balkany et al., 1980:607). A test for vestibular function should always be carried out when MD is suspected, as it may reveal the only physical sign of a disordered vestibular labyrinth.

4.3.4 The significance of vestibular function tests results in the diagnosis of MD

The most common cause of a unilateral weakness in terms of vestibular function is end organ disease. The most common ear disease that causes a unilateral weakness is MD (Jacobson et al., 1997:197). ENG was used to assess vestibular function in 135 subjects with MD. The ICS Medical Electronystagmography instructional guide test protocol as discussed on page 51

was followed to record and interpret the ENG results. The results were recorded in Column 23: Electronystagmography results of left ear, Column 24: Electronystagmography results of right ear, Column 25: Dix-Hallpike left ear and Column 26: Dix-Hallpike right ear of the electronic spreadsheet. The results are summarized in Table 4.9 to 4.28 and Figure 4.10.

Various studies show abnormal ENG results in patients with MD (Black & Kitch, 1980:636; Jacobson et al., 1997:197). Table 4.19 provides the results obtained of the ENG results for subjects in this study.

Table 4.19: Abnormal ENG results in subjects with MD (n=135).

VESTIBULAR TESTS	TYPES OF MD		
	Definite MD	Probable MD	Vestibular MD / RV
Saccades/Optikokinetic/ Tracking	0 %	0 %	0 %
Abnormal caloric response (39 % - n=53/135) - male 48 % (n=25/53) female 52 % (n=28/53)	24 %	6 %	9 %
Observed nystagmus (9 % - n=12/135) - male 42 % (n=5/12) female 58 % (n=7/12)	7 %	2 %	0 %
Positive Dix-Hallpike (16 % - n=22/135) - male 25 % (n=5/22) female 75 % (17/22)	8 %	0 %	8 %

Saccades, Tracking and Optokinetic tests were normal in all these subjects. This is not surprising since abnormalities in these tests are indicative of a central site of lesion (Shepard & Telian, 1994:425).

The presence of canal paresis in this study was determined during the period of nystagmus following irrigation with water. In 39 % (n=53/135) of cases the caloric test showed a

difference in excitability between the labyrinths of more than 25 %, to the detriment of the affected side, with only 1% (2/135) of subjects exhibiting a complete canal paresis. The reported incidence of canal paresis is anywhere between 50 % and 70 % (Black & Kitch, 1980:636; Jacobson et al., 1997:197). Wada et al. (1990:119) reported a lower incidence of 28 % and Oosterveld (1980:889) a slightly higher incidence of 48 %. Variations in the reported incidence of canal paresis is probably due to the use of different criteria for case selection and differences in the technique of ENG and caloric testing as well as criteria for abnormality. The low incidence of complete loss of vestibular function, as elicited by the caloric test, is supported in literature (Jacobson et al., 1997:197; Schessel et al., 1998:2680). There was no significant difference for abnormal caloric results or the presence of nystagmus between males and females. More detailed information regarding the nature of nystagmus is displayed in Table 4.20 and Table 4.21. Twelve subjects presented with *one or more* types of nystagmus.

Table 4.20: Summary of subjects presenting with different types of nystagmus.

Type of nystagmus	Subjects	Type of nystagmus	Subjects
1. <u>Spontaneous nystagmus</u> Present	1 % (n=1/135)	3. <u>Positional nystagmus</u> Present	8 % (n=11/135)
2. <u>Fixation nystagmus (gaze)</u> Present	2 % (n=3/135)	4. <u>Positioning nystagmus (BPPV)</u> Present	16 % (n=22/135)

Positioning nystagmus (BPPV) seems to be the most common type of nystagmus to be observed in patients with MD. This occurrence was also noted in other studies (Hughes & Proctor, 1997:610; Karlberg et al., 2000:382). Only 11 % (n=15/135) of all subjects experienced spontaneous, positional and gaze nystagmus. A detailed description of these subjects is presented in Table 4.21.

Table 4.21: Description of subjects with spontaneous, positional and gaze nystagmus (n=12).

Direction of nystagmus	Type of nystagmus	Type of MD	Affected ear	Gender
Mixed	Spontaneous	Definite	R	F
Right beating	Positional	Definite	L/R	M
Right beating	Positional	Definite	L/R	M
Right beating	Positional	Definite	L	M
Right beating	Positional	Probable	R	F
Right beating	Positional	Definite	R	F
Right beating	Positional	Definite	R	F
Right beating	Positional	Definite	L	F
Left beating	Positional	Definite	L/R	F
Right beating	Positional & gaze	Probable	L/R	F
Right beating	Positional & gaze	Definite	R	M
Left beating	Positional & gaze	Definite	R	M

Nine percent of subjects with vestibular complaints demonstrated nystagmus (Table 4.19). Of interest is that nystagmus only occurred in subjects with definite and probable MD where both cochlear and vestibular symptoms are present. In this study spontaneous, fixation, positional, and positioning nystagmus of the subjects were measured. Positional and positioning nystagmus types were observed more frequently in comparison with spontaneous and fixation nystagmus. This confirms the results of Wada et al. (1990:118). No correlation existed between the direction of the nystagmus and the side of the affected ear. This confirms the finding of Oosterveld (1980:889). The vestibular system of these subjects was studied during a symptom-free interval. The results are in accordance with Meyerhoff et al. (1981:223) who found positional nystagmus and spontaneous nystagmus to be unusual during symptom-free intervals.

In this analysis of 135 subjects with MD, 16 % (n=22/135) (Table 4.20) were found to have a codiagnosis of BPPV (positive Dix-Hallpike test result). A summary of these subjects is presented in Table 4.22.

Table 4.22: Summary of subjects with BPPV (n=22).

Positive Dix-Hallpike	Gender	Laterality of disease	Positive EcoG result	Caloric weakness
<u>Right ear</u> 64 % (n=14/22)	<u>Female</u> 68 % (n=15/22)	<u>Unilateral</u> 27 % (n=6/22) Right 18 % (n=4/22) Left	<u>Unilateral</u> 27 % (n=6/22) Right 18 % (n=4/22) Left	50 % (n=11/22)
<u>Left ear</u> 36 % (n=8/22)	<u>Male</u> 32 % (n=7/22)	<u>Bilateral</u> 55 % (n=12/22)	<u>Bilateral</u> 55 % (n=12/22)	

This study indicated a higher incidence of BPPV in female subjects. BPPV was limited to the same ear as MD in subjects with unilateral disease. All these subjects had positive EcoG results on the side diagnosed with BPPV. The subjects with bilateral disease had positive EcoG results on both sides, but experienced unilateral BPPV. In cases with asymmetrical hearing thresholds BPPV was found in the poorer ear. In 50 % (n=11/22) of these subjects an abnormal caloric response was recorded on the same side as BPPV.

This correlates well with the study of Karlberg et al. (2000:381) who found 19 % of patients with BPPV to have MD. Gross et al. (2000:657) and Hughes and Proctor (1997:610) also found higher incidence of females with BPPV, the significance of which is unknown. Karlberg et al. (2000:382) as well as Hughes and Proctor (1997:610) also found that BPPV was limited to the same ear as MD in patients with unilateral disease. These studies also indicated positive EcoG results on the side diagnosed with BPPV. This indicates a relationship between the two disorders and argues against the assumption that the combination of the two disorders is merely coincidental. The relationship between level of sensorineural hearing loss and BPPV was investigated and the results are presented in Table 4.23.

Table 4.23: PTTs in subjects with MD and BPPV (n=22).

Hearing threshold	Patients experiencing MD and BPPV
Normal Hearing (0-15dB)	40 % (n=9/22)
Very Mild Hearing Loss (16-25dB)	21 % (n=5/22)
Mild Hearing Loss (26-40dB)	19 % (n=4/22)
Moderate Hearing Loss (41-55dB)	10 % (n=2/22)
Moderate-severe Hearing Loss (56-70dB)	5 % (n=1/22)
Severe Hearing Loss (71-90dB)	5 % (n=1/22)

Most subjects (80 % - n=18/22) with MD and BPPV presented with either normal hearing or a mild sensorineural hearing loss. Paparella and Mancini (1985:1011) also found an increased incidence of BPPV in vestibular MD. Since most subjects with BPPV had normal hearing no correlation between audiogram configuration and BPPV was found.

It has been suggested that the free floating otoconial debris causing BPPV could obstruct the flow of endolymph and affect reabsorption, thus producing hydrops (Paparella, 1991:31). On the other hand, endolymphatic hydrops might damage the utricle, resulting in loose otoconia and BPPV secondary to the MD (Hughes & Proctor 1997:612). Another possible explanation of this association is that a common underlying disorder both produces hydropic distension and damages the otolithic apparatus separately (Hughes & Proctor, 1997:612).

The above results indicate that ENG is a method to electrically monitor the amount of nystagmus occurring spontaneously as well as from caloric stimulation (Martin, 1994:319). The caloric test, however, remains the most useful test of the integrity of an individual labyrinth and is the most reliable for determining the affected ear (Rizvi, 1986:1262). A summary of the caloric tests results is presented in Table 4.24.

Table 4.24: Caloric test results in the different types of MD (n=135).

MD Type	Normal caloric response	Mild weakness	Moderate weakness	Severe weakness	Profound weakness
Definite	80 %	13 %	4 %	2 %	1 %
Probable	79 %	11 %	10 %	0 %	0 %
Vestibular	80 %	13 %	5 %	2 %	0 %
Cochlear	100 %	0 %	0 %	0 %	0 %

This study revealed an almost equal distribution of caloric responses across different types of MD except for cochlear MD where the cochlea is the only site of lesion. The only significant feature is that only subjects with definite MD experienced profound caloric weakness. Subjects with definite MD tend to present with lower PTTs, which might also have an influence on these results (Schessel et al., 1998:2680). The effect of the degree of sensorineural hearing loss on the caloric test results is summarized in Table 4.25 and Table 4.26.

Table 4.25: Abnormal caloric responses in relation to the degree of hearing loss (n=52).

Level of hearing	Abnormal caloric response
Normal hearing (0-15dB)	27 % (n=14/52)
Hearing loss (16-90dB)	70 % same side (n=36/52) 3 % opposite side (n=2/52)

Seventy three percent (n=38/52) of subjects with abnormal ENG presented with a hearing loss. It seems that patients with both cochlear and vestibular problems are more likely to demonstrate a reduction in the caloric response (greater than 25 %) as elicited by bithermal caloric testing. The classification of caloric responses is presented in Appendix E.

Except in 3 % (n=2/52) of subjects, cochlear and vestibular symptoms were located in the same ear. It occasionally happens that the normal or better hearing ear reveals abnormal results (Balkany et al., 1980:607). The left and right ear may have dyssynchronous malfunction. The balance dysfunction may be emanating from the ear displaying normal hearing (most of the time), yet because of the severe fluctuations in hearing the other ear is blamed for the entire set of clinical problems (Balkany et al., 1980:607). In such cases further careful enquiry may reveal former attacks that could suggest bilateral disease (Cawthorne &

Hewlett, 1954:25). This indicates the clinical value of ENG results in diagnosis laterality relating to the disease, especially as part of the test battery.

Table 4.26: Caloric test results in relation to PTT (n=135).

PTT	Normal caloric response	Mild weakness	Moderate weakness	Severe weakness	Profound weakness
Normal hearing (0 – 15 dB)	54 %	29 %	14 %	3 %	0 %
Very Mild Hearing Loss (16 – 25 dB)	46 %	42 %	6 %	6 %	0 %
Mild Hearing Loss (26 – 40 dB)	73 %	15 %	8 %	2 %	2 %
Moderate Hearing Loss (41 – 55 dB)	54 %	20 %	20 %	6 %	0 %
Moderate- severe Hearing Loss (56 – 70 dB)	56 %	44 %	0 %	0 %	0 %
Severe Hearing Loss (71 – 90 dB)	100 %	0 %	0 %	0 %	0 %

The degree of hearing loss in subjects did not influence the caloric response. All of the subjects with moderate to severe hearing losses either had normal caloric functioning or a mild caloric weakness with some subjects with normal hearing experiencing severe caloric weakness. The effect of MD on the cochlea and the vestibular system will probably depend on the anatomical site of endolymphatic hydrops and the sensitivity of the individual's cochlear and vestibular system to the effect of hydrops. The effect of audiogram configurations on caloric responses were also investigated and the results are summarized in Table 4.27.

Table 4.27: Abnormal caloric responses in relation to different audiogram configurations (n=52).

Audiogram configuration of affected ears	Abnormal caloric response
Rising	14 % (n=7/52)
Flat	14 % (n=7/52)
Falling	21 % (n=11/52)
Peak	51 % (n=27/52)
Dip/trough	0 %

There was a fairly even distribution for rising, flat and falling audiogram configurations for subjects with abnormal ENG results in this study. It is interesting to note that 51 % (n=27/52) of subjects with an abnormal caloric response presented with peak type audiograms (Table 4.27). No figures for comparison could be found in other studies. Lee et al. (1995:530) suggested that audiogram configuration is influenced by varieties and degrees of hydrops, which would probably have an effect on vestibular functioning of patients with MD. It is, however, to determine the pathological site and degree of the hydrops without post-mortem studies.

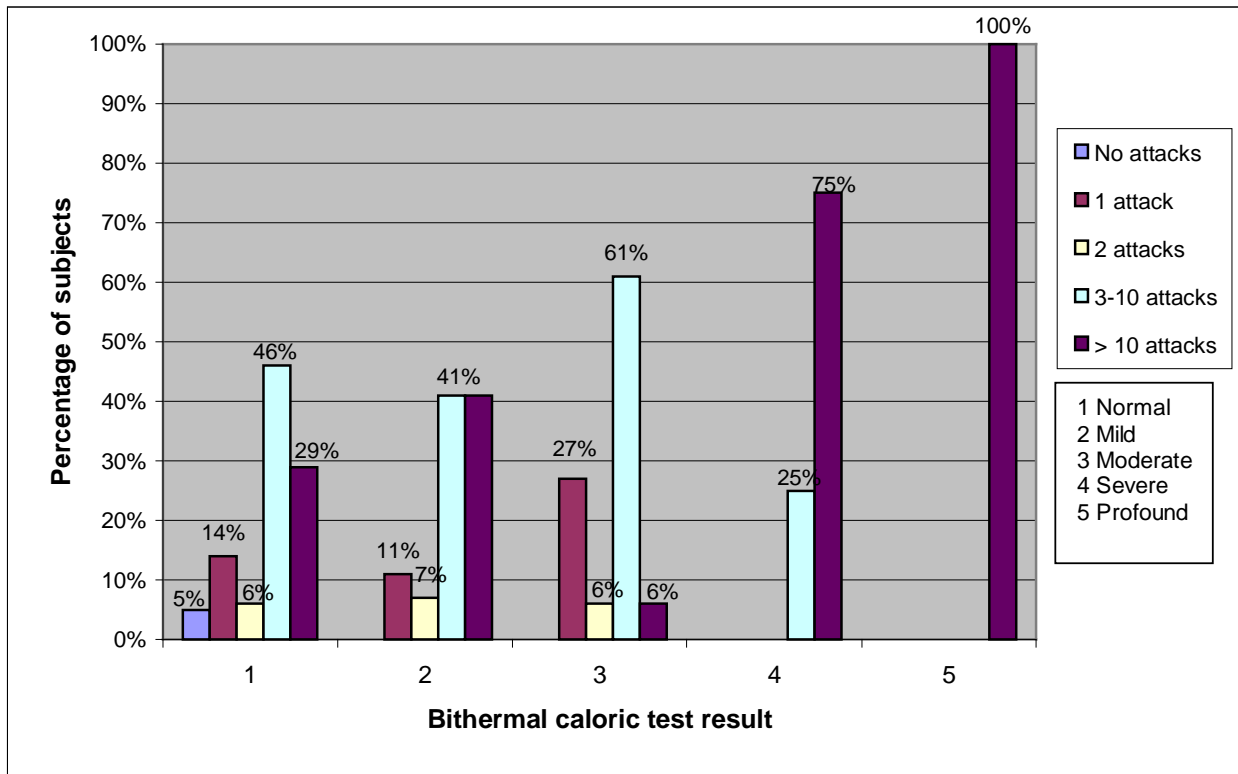


Figure 4.10: Caloric test results in relation to number of vertiginous attacks (n=135).

This study did not indicate a relationship between the degree of caloric weakness and the duration or frequency of vertiginous attacks. Some subjects demonstrated moderate caloric weakness after only three months of attacks while others demonstrated mild or no caloric weakness even after 5 years. Although there is no direct correlation between number of attacks and severity of caloric weakness, there is an indication that subjects experiencing more attacks have a greater chance of experiencing greater caloric weakness. Apart from the attacks of vertigo, it is suggested that other symptoms might also influence caloric test results (Balkany et al., 1980:607). The influence of other symptoms experienced by subjects in this study on caloric results is summarized in Table 4.28.

Table 4.28: Abnormal caloric responses in relation to symptoms experienced by subjects with MD (n=135).

Symptom	Abnormal ENG
Aural fullness (n=72/135)	20 % (n=14/72)
Tinnitus (n=96/135)	18 % (n=17/96)
Nausea / Vomiting (n=114/135)	19 % (n=22/114)
Hearing loss (n=106/135)	19 % (n=20/106)
Fluctuating hearing loss (n=24/135)	10 % (n=2/24)
Tullio phenomena (n=8/135)	12 % (n=1/8)
Vertigo (n=130/135)	40 % (n=52/130)

There is a fairly equal distribution of symptoms in subjects with caloric weakness. It is, however, interesting to note that despite vestibular complaints only 40 % (n=52/130) of subjects complaining of vertigo had abnormal caloric test results. The permanent effect of MD on the vestibular system is probably related to the anatomical site and degree of endolymphatic hydrops. According to Rizvi (1986:1268) an abnormal caloric response is the result of endolymphatic hydrops causing distension of the utricle that sufficiently distorts the ampullary walls. The results indicate that it would not be possible to assess the effect of MD on the vestibular system of a patient without a vestibular function test, emphasising the clinical value of ENG results in the diagnosis of MD.

4.3.4.1 Summary of the significance of ENG results in the diagnosis of MD

This study confirms that ENG findings are helpful in establishing the diagnosis of MD and confirming the affected ear. Caloric testing is effective in testing the integrity of an individual labyrinth and determining the affected ear. ENG is also an important test to confirm either central or peripheral causes for vestibular dysfunction assisting in the differential diagnosis of MD.

4.4 SUMMARY

In this chapter the clinical profile of 135 subjects with MD were described. The significance of audiological and vestibular tests in the diagnosis of MD was investigated and the results of this study confirmed that a systematic audiological approach will assist clinicians in the diagnosis of MD.

CHAPTER 5

CONCLUSION AND RECOMMENDATIONS

The aim of this chapter is to highlight the most important findings of the study, to explain the implications of these findings to the field of audiology, to evaluate the research project and identify the limitations of the study as well as possible directions for future research.

5.1 INTRODUCTION

The dynamic nature of MD may lead to uncertainty regarding the diagnosis in patients, increasing stress and affecting quality of life (Vesterhauge, 1996:11). A better understanding of MD will facilitate the initial diagnosis of MD and assist the clinician in determining appropriate medical management (Goin et al., 1982:1389). The main aim of the study was to analyse and describe the clinical and audiological features of a cohort of subjects diagnosed with MD, in order to develop understanding of the pathophysiology of MD and the diagnostic processes involved.

5.2 SUMMARY OF THE RESEARCH FINDINGS

The most important findings regarding the case history as well as audiological and vestibular tests during the diagnosis of MD are highlighted. A summary is presented according to the sub-aims of this study.

5.2.1 Clinical profile of a group of subjects diagnosed with MD

It is generally accepted that the most important diagnostic features of MD are found in the patient's clinical history (Schessel et al., 1998:2679). Careful attention to patient history supplemented with diagnostic test results will aid in the diagnosis of MD.

5.2.1.1 Gender

This research study showed a slightly higher incidence of MD in females (ratio 1,4:1). The increase in incidence for females in relation to males is more noticeable in atypical MD, especially of the vestibular type. Although, the reason for this is not known, it might suggest that females might be more likely to develop atypical MD, especially of the vestibular type. Further research is needed to confirm this and to investigate possible reasons for this observation. The results are in keeping with other studies regarding gender and MD, which also noted a slight preponderance to females especially in relation to vestibular MD (Meyerhoff et al., 1981:220; Wada et al., 1990:119; Watanabe, 1981:512).

5.2.1.2 Age of onset

There is a definite increase in the distribution of subjects between the ages of 40 years and 60 years. There seems to be a tendency for subjects with vestibular MD/RV to present at an earlier age than subjects with definite MD. This might be because these subjects present with vestibular symptoms early in the course of the disease before classical MD develops. The results are similar to those found by Meyerhoff et al. (1981:222), Oosterveld (1980:886) and Watanabe (1981:512).

5.2.1.3 Family history

It was found in this study that only 3 % (n=4/135) of subjects indicated a family history related to MD. This figure is low in relation to other research (Paparella, 1985:447), which states that 10 to 20 % of patients present with a family history of MD. This study does, however, still indicate that genetic inheritance of MD remains a possibility in some cases.

5.2.1.4 Laterality of the disease

Patients can present with either unilateral or bilateral MD. Combining evidence of audiometric tests and patient history, it was found that 39 % (n=53/135) of subjects in this study seem to have bilateral MD. This study did not find a significant sex difference between subjects with unilateral and bilateral disease. The increase of bilaterality with increasing

duration of the disease was not reaffirmed in this study, probably because only 6 % (n=8/135) of subjects have had MD for more than five years.

5.2.1.5 Diagnosing different types of MD according to the symptom complex

All the subjects diagnosed with definite MD experienced episodic vertigo with sensorineural hearing loss. Most subjects complained of tinnitus (71 % - n=96/135) and nausea (84 % - n=113/135). The results show a definite clinical pattern with these subjects. This confirms that it is usually not difficult to diagnose MD in patients with definite MD (Schessel et al., 2005:3217). There are, however, subjects who presented with only cochlear or vestibular features of MD. Eighty percent (n=4/5) of the subjects diagnosed with cochlear MD experience fluctuating hearing loss and aural fullness. Although more research is needed to confirm this, fluctuating hearing loss and aural fullness might be diagnostic features of cochlear MD. The diagnosis of vestibular MD / RV is more complex, because of the controversy that exists regarding the diagnosis of a vestibular type of MD (Monsell et al., 1995:183). In this study 21 % (n=29/135) of subjects presented with MD type vertigo without apparent hearing loss.

5.2.2 Investigation into the incidence of symptoms experienced in each type of MD

The results and discussion in the previous section (Chapter 4) demonstrated that 65 % (n=88/135) of subjects complained of episodic vertigo, sensorineural hearing loss and tinnitus. It also highlighted that vertigo was the most dramatic and debilitating symptom in MD. An interesting finding was that 38 % (n=11/29) of subjects with normal hearing experienced tinnitus. Further investigations might indicate aural pressure as a diagnostic feature of vestibular MD and fluctuating hearing loss diagnostic of cochlear MD. The Tullio phenomenon was present in 14 % (n=8/135) of subjects.

5.2.3 The significance of audiological and vestibular tests in the diagnosis of MD

MD is variable in its clinical presentation, but despite this, MD has a precise identity. A better understanding of the audiological profile seen in MD is valuable for confirming the diagnosis of MD (Paparella & Sajjadi, 1999:30).

5.2.3.1 The significance of PT audiometry in the diagnosis of MD

Sixty one percent (n=82/135) of subjects presented with a unilateral hearing loss. The average hearing loss found in these subjects with MD, except vestibular MD where hearing is normal, was moderate sensorineural hearing loss of between 40 to 50 dB. A fluctuating hearing loss was demonstrated in 18 % (n=24/135) of the subjects. Only 3 % of subjects presented with severe sensorineural hearing loss. It is interesting that 10 % (n=14/135) of subjects with hearing loss reported their hearing to be normal, indicating the importance of obtaining PT audiometry results for all patients suspected of having MD.

Flat and peak type audiogram configurations were found in 65 % (n=122/188) of affected ears. Two percent of subjects with unilateral MD also exhibited a peak type audiogram configuration in the contralateral ear.

5.2.3.2 The significance of SD scores in the diagnosis of MD

This study found SD scores to be only affected in subjects with thresholds above 40 dB, as is found in non-Ménière ears. No indications were found of reduced SD relative to the expectation based on PT loss. The audiogram shape does not appear to play any additional role in SD. The usefulness of SD scores as part of the test battery lies in confirming PTT as well as differentiating cochlear from retrocochlear site of lesions (Penrod, 1994:159).

5.2.3.3 The significance of DPOAE test results in the diagnosis of MD

Abnormal DPOAE results were found in 70 % (n=91/130) of subjects in this study. There was an increase in normal emissions in the mid-frequency range which was in accordance with the high incidence (31 % - n=59/188) of peak type audiogram configurations in this study.

5.2.3.4 The significance of EcoG results in the diagnosis of MD

EcoG testing proved to be a reliable diagnostic tool to assist in the diagnosis of MD. Eighty eight percent (n=166/188) of all subjects had a positive EcoG result in the affected ear, indicating that EcoG is of great value especially early in the disease when the clinical picture

is not clear as well as in patients without hearing loss. The results of this study indicated that a positive EcoG result does not depend on a diagnosis of definite MD where patients experience vertigo, tinnitus and hearing loss. Ninety one percent (n=123/135) of subjects with atypical MD displayed positive EcoG results. A significant finding in this study is the presence of endolymphatic hydrops indicated by EcoG in subjects with vestibular MD / RV. Careful attention to the clinical history suggested that subjects without hearing loss with a positive EcoG result display some cochlear symptoms such as tinnitus and aural fullness. The presence of aural fullness seemed to have a positive influence on EcoG results. Ninety six percent (n=129/135) of subjects who complained of aural fullness had positive EcoG results. Another interesting finding in this study was the presence of positive EcoG results in asymptomatic ears.

5.2.3.5 The significance of ENG results in the diagnosis of MD

Saccades, Optokinetic tests, Tracking, Positional and Gaze tests did not reveal any diagnostic information to assist in the detection of endolymphatic hydrops, but because these tests are expected to be normal it is valuable to perform these tests in order to differentiate between central and peripheral disorders. A caloric weakness was found in 39 % (n=53/135) of subjects. Except in 3 % (n=2/52) of subjects, cochlear and vestibular symptoms were located in the same ear. The results did not indicate a significant difference for abnormal caloric results or the presence of nystagmus between males and females. Horizontal nystagmus was observed in 9 % of subjects. A rotatory nystagmus (BPPV) was found in 16 % (n=22/135) of cases with a higher incidence in female subjects.

5.2.4 **Systematic audiological approach to the investigation of MD that will facilitate the clinical diagnosis**

The last sub-aim formulated in relation to the main aim of the study was to define a systematic audiological approach to the investigation of MD that will facilitate the clinical diagnosis. The results of this study revealed the importance and relevance of certain tests in the diagnosis of MD. It is clear that audiologists play an important role as part of the multidisciplinary team involved in the management of MD. The challenge in choosing a battery of tests is to exclude certain diseases in the process of diagnosing another. Based on

the results and conclusions of this study, a systematic approach for the investigation of patients with vestibular complaints is proposed in Figure 5.1.

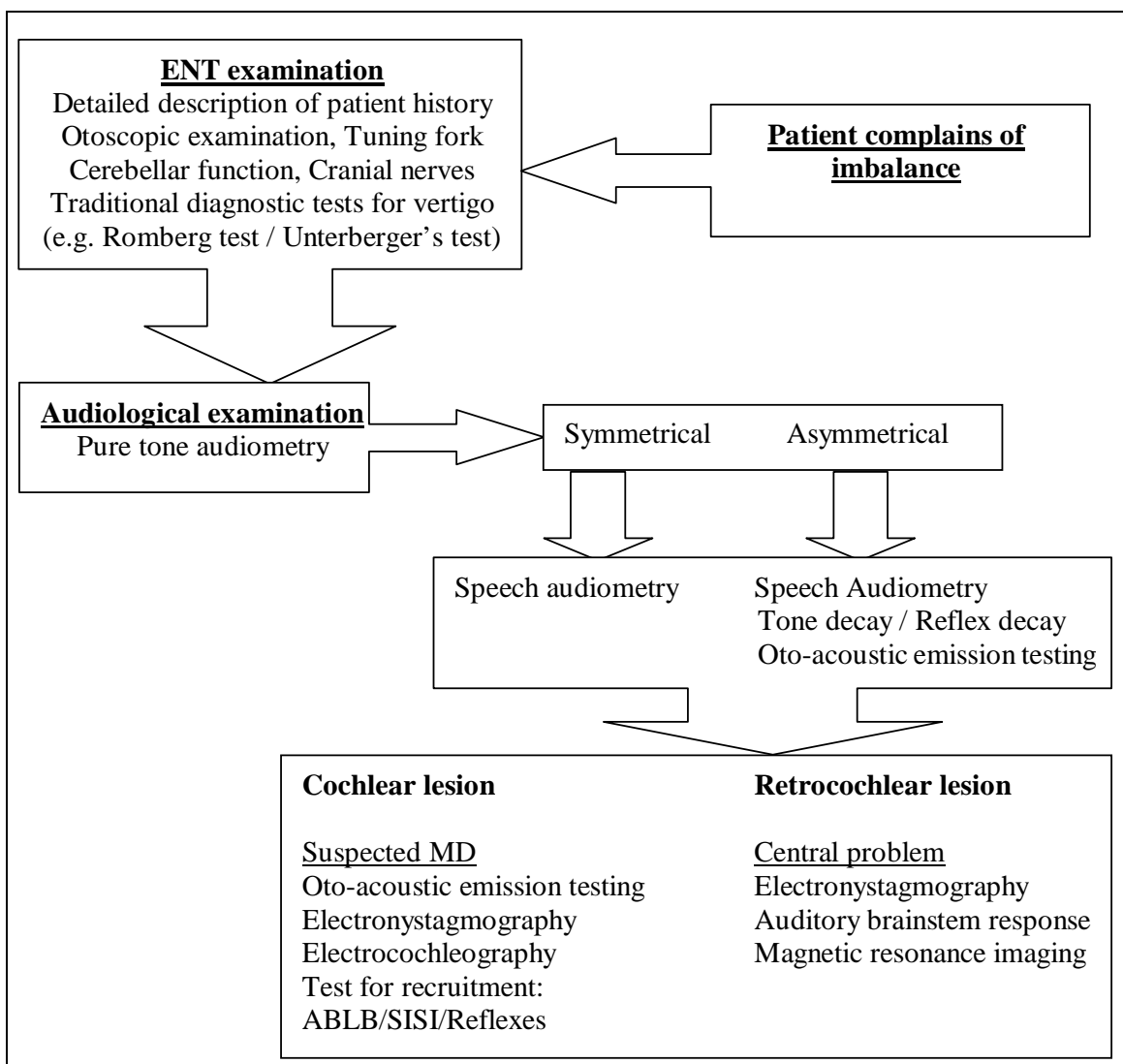


Figure 5.1: Examination of patients complaining of episodic vertigo.

The above presentation indicates that in following a systematic approach in the diagnosis of any vestibular disease, will reveal the site of lesion and assist in formulating a diagnosis. There is no single test that can be used to make the diagnosis of MD, but a combination of both ENT and audiological information will provide valuable information to identify the clinical features that will lead to the diagnosis of MD.

5.3 IMPLICATIONS OF THE FINDINGS

The diagnosis of MD is complex. The process from initial consultation to final diagnosis is often very long, and leaves many patients worried and despondent (Vesterhauge, 1996:11). By correlating the clinical features of subjects with both typical and atypical MD, with audiometric and vestibular tests, this study highlighted the clinical value of an audiological test battery in assessing patients with MD. The addition of a reliable physiological measure to the diagnostic battery will assist medical professionals in determining appropriate medical or surgical management (Goin et al., 1982:1389). This confirms the role of the audiologist in the diagnostic and rehabilitation process as well as encouraging a multidisciplinary outlook in effective patient management.

5.3.1 The clinical profile of a group of subjects diagnosed with MD

Although MD predominantly affects adults from the third and fourth decade of life it can present at any age and affects both males and females. It is clear that atypical forms of MD exist and therefore central nervous system problems such as acoustic neuroma and vestibular migraine should be ruled out by thorough neurological investigation, since many of the reported symptoms in MD are also consistent with other diseases (Baloh, 1997:616; Weber & Adkins, 1997:978). The possibility of genetic inheritance in MD should be identified in the patients' case history. This will alert the clinician to early symptoms in the patient as well as other family members.

The prevalence of bilateral involvement has been shown to increase with time (Moffat et al., 1992:370), and it is therefore expected that the proportion of subjects with bilateral disease would increase with continued follow-up, since many of the subjects were still young when they entered this study. This suggests that follow up of patients presenting with unilateral MD is essential to monitor the uninvolved ear. Early recognition of incipient MD in the asymptomatic contralateral ear of a patient with known unilateral disease has obvious and profound implications for patient management. A conservative method of treatment should be considered, especially when considering surgical intervention for intractable disease since there is no evidence that surgical treatment of one ear delays the potential involvement of the second ear. Treatment should aim at retaining or restoring labyrinthine function. When the

different treatment options are discussed with patients, possible involvement of the contralateral ear should not be disregarded.

5.3.2 Investigation into the incidence of symptoms experienced in each type of MD

From the results it is clear that the clinical history is still the most important feature when diagnosing MD (Schessel et al., 1998:2679). There was, however, subjects (35 % in this study) who did not present with the classical picture and could not be diagnosed on symptomatology alone. The key to the diagnosis should, however, be found in the patient's case history. It is important to describe the clinical signs and symptoms of the inner ear disease, which will result in greater understanding of the clinical manifestations and natural history of all inner ear disorders and specifically MD. This will also guide the clinician in terms of compiling a test battery that will lead to an accurate clinical diagnosis.

There should be an awareness that the Tullio phenomenon can occur in MD. Its presence can be determined by simply asking the patient if they experience vertigo in the presence of excessive noise. This is important especially when a hearing aid is considered. Patients will have to be counselled, explaining the benefit (improved hearing) and potential disadvantage (vertigo) of amplification. Most hearing aid companies in South Africa offer a 30-day trial period for patients to test the success of hearing aids for their specific hearing needs, and audiologists can offer patients the opportunity to experience the effect of amplification without the obligation of purchasing something that might not benefit their specific situation.

5.3.3 The significance of audiological and vestibular tests in the diagnosis of MD

The clinical value of **PT audiometry** is recognised to determine the type, degree and audiometric configuration of the hearing loss. PT audiometry demonstrated sensorineural hearing loss in 10 % (n=14/135) of subjects who did not, on initial examination, complain of hearing loss. Also, some subjects with aural fullness without demonstrable hearing loss complained of a hearing loss. The audiogram configuration also reveals valuable clues to assist in making a diagnosis. These results indicate the importance of determining PTT for every patient regardless of their subjective assessment of their hearing function.

The high incidence of unilateral loss increases the risk of retrocochlear suspicion and emphasizes the need for differential diagnosis (Weber & Adkins, 1997:978). This emphasizes the importance of a full diagnostic test battery to exclude retrocochlear pathology. The occurrence of a fluctuating hearing loss also necessitates retesting and follow-up audiograms. The audiogram configuration should be noted even when hearing is apparently normal. Moffat et al. (1992:372) found early evidence of endolymphatic hydrops in the asymptomatic contralateral ear of patients diagnosed with unilateral MD. Slight asymmetry may be indicative of early cochlear involvement in patients with apparently normal hearing. Certain audiogram configurations (such as peak type audiograms) are more commonly seen in MD and may also indicate early involvement of the opposite asymptomatic ear. The early recognition of incipient MD in the asymptomatic contralateral ear is important to identify the best treatment option for these patients.

The absence of **DPOAEs** have been shown to be a highly sensitive, but non-specific indicator of cochlear dysfunction (Sakashita et al., 1998:70). Patients with MD cannot, however, be distinguished from patients with other sensorineural hearing loss etiologies based on DPOAE findings alone (Hall & Mueller, 1997:273). DPOAE test results are useful to confirm cochlear pathology, exclude retrocochlear pathology and to indicate early damage to the cochlea even when hearing thresholds are normal. DPOAEs should therefore be performed even if patients' audiograms demonstrate normal hearing thresholds, as 50 % of subjects in this study showed evidence of cochlear pathology despite normal audiograms. Sakashita et al. (1998:74) also noted a significant change in DPOAE amplitudes even though hearing thresholds were normal.

EcoG is a valuable objective otoneurological technique used to assess endolymphatic hydrops and should form part of the assessment protocol in patients with MD. The results of this study were in agreement with Schuknecht and Kitamura (1981:18) who found that patients with vestibular MD display positive EcoG results and experience early cochlear problems such as tinnitus and aural fullness. Patients without these clinical signs should be referred to as RV. This will help to differentiate between vestibular MD and RV.

It seems that the ideal time to perform EcoG testing would be when patients experience aural fullness. Since it is believed that enhancement of the SP will vary according to or with the changes in intra-labyrinthine fluid pressure and volume that characterize the disease, EcoG

should be done during or as close to an attack as possible (Ferraro et al., 1983:78). EcoG should be performed bilaterally, even if only unilateral MD is suspected. Some evidence of endolymphatic hydrops was found in asymptomatic contralateral ears, which might be an early indication of bilateral involvement. EcoG measures may offer some insight to the diagnostic process where certainty of the diagnosis is less clear. Even when the diagnosis is clear it will be reassuring to confirm the presence of endolymphatic hydrops with a clinical test. The value of EcoG in this study has been the demonstration of abnormalities compatible with hydrops in the asymptomatic contralateral ear, as well as that together with a detailed clinical history it would be useful to differentiate between vestibular MD and RV. Evoked response audiometry, both brainstem and electrocochleographic responses, forms an important part of the modern clinician's tools for investigating sensorineural hearing loss (Hall & Mueller, 1997:565). It should be noted, however, that the 'diagnostic' electrical response features are common to all diseases causing endolymphatic hydrops regardless of whether it is idiopathic or secondary to or associated with such disease as late syphilis, otosclerosis, old labyrinthitis, or on occasion acoustic neuroma. A wide diagnostic horizon must be envisaged.

This study confirmed that **ENG** findings are helpful in establishing the diagnosis of MD and confirming the affected ear. Furthermore, it is an effective tool to determine if the episodic attacks have caused permanent damage to the balance system. Although caloric weakness might not be present in all patients, when it occurs it is accurate to identify the involved ear especially if the cochlear and vestibular symptoms are caused by separate ears. A significant finding in this study is a positive Dix-Hallpike in some subjects with MD. The clinical implication is that that patients with recurring BPPV should be monitored for MD and all patients with MD should be tested for BPPV. The Dix-Hallpike test usually forms part of the ENG test protocol, but audiologists without this equipment can perform this test for benign paroxysmal positional vertigo as a stand-alone test.

5.3.4 The role of the audiologist as part of a multidisciplinary team

The American Speech-Language-Hearing Association (ASLHA) states that "the practice of audiology includes ... screening, identifying, assessing, and interpreting, diagnosing, preventing, and rehabilitating peripheral and central auditory system dysfunctions...providing and interpreting behavioural and (electro)physiological measurements of auditory and vestibular functions...selecting, fitting, and dispensing of amplification, assistive listening

and alerting devices...” (Hall et al., 1993:254). The South African Professional Board for Speech-Language and Hearing Professions further stipulates that determining appropriate assessment protocols, applying appropriate assessment procedures in order to determine nature, degree and site of a hearing impairment, assessing the need for and then planning and providing individual counselling as well as consultation with professionals to provide information on issues pertaining to aspects of hearing and vestibular development, normal functions and disorders forms part of the minimum competency requirements for the practice of Audiology (“Competency profiles, 2005:12”). Audiologists should follow a heuristic point of view, where they go beyond the mastery of test techniques and careful recording of data, although these skills are vital.

Audiologists should be inspired, imaginative, and compassionate, seeking to discover the relationships among the procedures and how all of this relates to the individual who has come to us for help (Robinette, 1994:191). The clinical implication of these results is to encourage and motivate audiologists, especially in the South African context, to do further and specialized testing in order to improve their accountability and reliability in the diagnosis of MD. MD is a diagnosis of exclusion and many disease entities can mimic MD. Thus, it is essential to eliminate other possible etiologic agents for complaints that could be related to MD before making this diagnosis. Therefore the use of a test battery and cross-correlation of test results are essential.

All clinical audiologists should be able to perform the basic test battery. Evoked potential and vestibular tests, however, require some special training and skill. The software needed to perform these tests is expensive and not many governmental institutions or private practices have access to all the tests. Even if audiologists have the equipment and skill to perform the tests, many patients cannot afford to pay for extensive test batteries. Audiologists have a responsibility to know what tests are available and where they can refer patients should they not be able to perform the tests themselves. Continued Professional Development (CPD) offers a unique opportunity for audiologists to develop their current skills and acquire new ones. In cases where patients cannot afford to pay for expensive diagnostic tests two possibilities might be considered for the future. Firstly, universities and audiologists in private practice who perform these tests could work in partnership to perform tests at a reduced rate as part of student training. Secondly, audiologists and ENTs in government hospitals should motivate the use of specialized equipment and attempt to include these tests

in their yearly budget. Witbank Hospital is currently in the process of applying for such a grant.

5.4 CRITICAL EVALUATION OF THE STUDY

The value of this study lies in the information that was gained from a specific group of subjects with MD regarding the clinical presentation of the disease. By determining the clinical value of an audiological test battery in assessing subjects with MD, and describing the clinical features of these subjects, an attempt was made to contribute to the development of MD research in SA. The results of this study made it possible to identify important features that leads to the diagnosis of MD. This research has contributed to the development of knowledge of this group of subjects with MD that can be implemented to improve patient management. The results of this study also confirmed the role of the audiologist in the diagnostic and rehabilitation process as well as encouraging a multidisciplinary outlook in effective patient management. Results of this study also confirmed that a systematic audiological approach would assist clinicians in the diagnosis of MD.

The group of subjects selected for this study reflects a good representation of patients with MD because these subjects were tested and diagnosed before this study commenced, therefore ruling out biased results. The disadvantage of a retrospective study of patient records is that patients do not keep their information updated, making it difficult to contact people at the time of the study, making it difficult to obtain informed consent to use the data as well as to arrange follow up testing. There was no control group for this study to compare results in patients with and without MD. Although the sample size is statistically significant (Neuman, 1997:222), it represents a homogenic population of European origin, representing the white population in South Africa. The data obtained from this study cannot be applied to other population groups until they have been included in similar studies.

During the literature study it was realised that although loudness recruitment is common to most forms of cochlear losses, over-recruitment is frequent in cases with milder losses and this seems to be fairly specific for hydrops (Arts et al., 1997:103). According to Ginsberg and White (1994:20) complete or over recruitment is the rule in this disease and it would have been valuable for the study to include recruitment tests. Recruitment can be determined by the ABLB test or a high SISI score. The presence of recruitment is also indicated by a positive

stapedial reflex produced by stimulus levels in the affected ear at or nearly equal to those levels required to illicit the reflex in the unaffected ear. This occurs despite the reduced hearing levels (Ginsberg & White, 1994:20). Since the test protocol was established before the study started, these tests were not included as part of the test battery.

DPOAEs were only performed in patients with PTTs exceeding 10 dB HL. According to the results of this study, it is clear that DPOAE can detect cochlear dysfunction even before detected with behavioural audiometry. It would be valuable to test all patients with a cochlear disease such as MD to determine early cochlear changes. It would also, as in the case of EcoG be valuable to test the asymptomatic ear in unilateral MD to indicate early cochlear involvement which might be indicative of possible bilateral MD.

Margolis et al. (1995:54) found EcoG to be most sensitive in the diagnosis of MD in the early stages of the disease before permanent cochlear damage occurs. They recommended that three indicators be used for evaluation of patients with endolymphatic hydrops, namely SP-AP ratio, AP latency difference to condensation and rarefaction clicks, and tone-burst evoked SP. Levine et al. (1992:614) also recommend separate examination of the rarefaction and condensation click recordings. In this study, only the SP-AP ratio was investigated, and further examinations might have provided further insight into abnormal response patterns in patients with MD.

5.5 RECOMMENDATIONS FOR FURTHER RESEARCH

There is still information that should be obtained regarding the diagnosis, treatment and quality of life of patients with MD in South Africa. During statistical data analysis certain potential areas for further investigations were identified to formulate future research proposals.

5.5.1 Investigate the diagnostic criteria and clinical picture of Tumarkins and Lermoyez MD. These types are not commonly described in literature and more information on these two types would be valuable for diagnostic purposes.

- 5.5.2 Distribute questionnaires among different ENT and Audiology practices to determine the test protocol and insight into the value of different audiological tests regarding the diagnosis of MD.
- 5.5.3 Perform EcoG studies on patients diagnosed with RV and vestibular MD to gain more information regarding this category of patients.
- 5.5.4 Research is needed to investigate the impact of MD on quality of life to gain understanding into the psychological aspects of this disease.
- 5.5.5 Repeat the methodology of this study on other population groups, to investigate the incidence as well as the impact on quality of life of these patients.

5.6 CONCLUSION

MD is a chronic illness that often affects individuals in the prime of life. Many authors believe that the disorder has a serious impact on the psychosocial status of individuals and their families leading to a significant decline in quality of life for patients with this disorder (Harris & Anderson, 2000). The psychological aspects of the disease such as anxiety, depression and inability to concentrate becomes the responsibility of the treating physician to develop and offer treatment options to their MD patients (Torok, 1977:1873). Effective patient management to improve quality of life can only be planned once the diagnosis is made. It is believed that the information gained from this study will assist clinicians to compile a test battery that leads to identification and diagnosis of the disease as soon as possible.

MD should be a clinical diagnosis based on the history and general examination, supplemented by audiologic and vestibular findings. Complete quantification of each inner ear dysfunction and elimination of the possibility of a central pathologic process must be accomplished prior to establishing a diagnosis (Balkany, Pillsbury & Arenberg, 1980:589). The difficulty in attempting to determine what is essentially a histological diagnosis (endolymphatic hydrops) in patients exhibiting only partial symptomatology has led to the development of EcoG as an objective tool, to aid the diagnosis of endolymphatic hydrops. Evoked response audiometry forms an important part of the modern clinician's tools for investigating sensorineural hearing loss and MD (Selmani et al., 2002:98).

5.7 SUMMARY

The true nature of this unique cochlear and labyrinthine disorder remains a controversial issue for both diagnosis and treatment (Schessel et al., 1998:2673). Despite the fact that MD is the third most common inner ear disorder there is still confusion about its precise role in the pathophysiology of the disease (Hall & Mueller, 1997:714). Although medical science still has not conclusively elucidated the underlying cause of MD and there is no single test that provides a sufficient level of sensitivity and specificity to be routinely useful in the diagnosis of MD, the combination of different audiological tests provide valuable information to assist in the diagnosis and patient management. Clinicians should adopt a problem-oriented approach, where the patient history should provide enough clues as to which tests are needed and how to combine the relevant diagnostic information to make an accurate diagnosis as soon as possible.

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APPENDICES



**APPENDIX A:
DATASHEET**



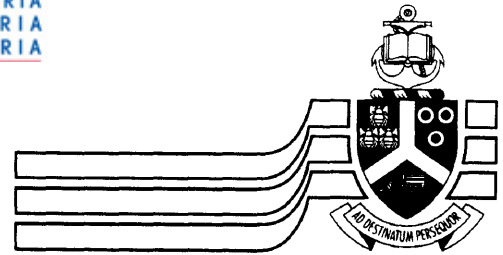
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**APPENDIX B:
ETHICAL CLEARANCE LETTER**

Studenteno: 9701698
Ons verw: Me P Woest
Tel: 012 420 2736
Faks: 012 420 2698
E-pos: petru.woest@up.ac.za



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20 Oktober 2004

Me AM Naudé
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Fakulteit Geesteswetenskappe

Geagte me Naudé

TITELREGISTRASIE: STUDIERIGTING – MKOMMUNIKASIEPATOLOGIE (OPSIE 1)

Met genoegte deel ek u mee dat die volgende goedgekeur is:

ONDERWERP: Clinical and audiological features of Meniere's disease: Insight into the diagnostic process
LEIER: Me C Avenant
MEDE-LEIER: Dr ME Soer

U aandag word in besonder op die volgende gevestig:

1) TERMYN VAN REGISTRASIE

- (i) U moet vir minstens een akademiese jaar as student vir die magistergraad geregistreer wees voordat die graad toegeken kan word.
- (ii) U registrasie moet jaarliks voor April van elke akademiese jaar hernu word totdat u aan al die vereistes vir die magistergraad voldoen het. Geen herregistrasie sal na 31 Maart aanvaar word nie. U sal slegs geregtig wees op die leiding van u leier indien u jaarliks bewys van registrasie aan hom voorlê.

2) GOEDKEURING VIR INDIENING

Vir eksamendoeleindes moet u voldoende eksemplare vir elke eksaminator indien, tesame met 'n skriftelike verklaring van u leier dat hy/sy die indiening van die verhandeling goedkeur sowel as 'n verklaring deur u, wat voor 'n Kommissaris van Ede geteken word, wat by die Fakulteitsadministrasie ingehandig word.

3) VOORSKRIFTE IN VERBAND MET DIE VOORBEREIDING VAN DIE VERHANDELING/SKRIPSIE ASOOK DIE SAMEVATTING IS OP DIE KEERSY VAN HIERDIE BRIEF UITEENGESIT.

Die uwe

nms **DEKAAN: FAKULTEIT GEESTESWETENSKAPPE**

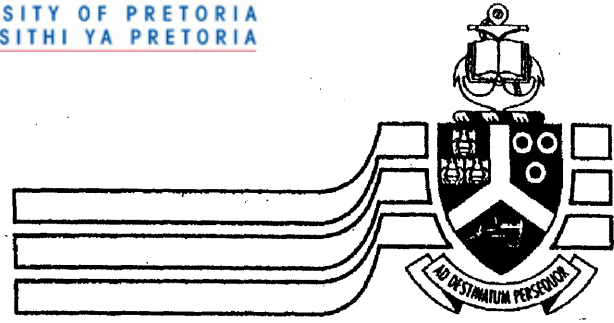


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**APPENDIX C:
PERMISSION FROM ENT TO STUDY
AND REPORT ON PATIENT
RECORDS**



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University of Pretoria

Department of Communication Pathology
Speech, Voice and Hearing Clinic

Dr. B.L Wolfowitz
PO Box 2310
Houghton
2041

PERMISSION TO STUDY AND REPORT ON RECORDS

I am currently a Masters student in Audiology at the University of Pretoria. In order to comply with the degree demands, I have to complete an extensive research project resulting in a dissertation.

The main aim of the study is to describe and analyze the clinical and audiological features of a cohort of patients diagnosed with Meniere's disease, in order to develop further understanding of the pathophysiology of Meniere's disease and the diagnostic processes involved. The research will be based on a retrospective study of the medical records of these patients.

In order to perform this study I will need access to files of at least 100 patients diagnosed with typical or atypical Meniere's disease. I request your permission to use the files of patients in your private practice. The following data will be extracted from the files and plotted on a datasheet:

- ◆ Complete case history of patient;
- ◆ Patient's age and gender;
- ◆ Pure tone audiogram results;
- ◆ Otoacoustic emission test results;
- ◆ Electrocochleography results as well as
- ◆ Electronystagmography results.

The statistical analysis of administrative data is entirely impersonal and would therefore not be harmful to any of the patients, but the following steps will also be taken to ensure patient confidentiality and anonymity:

- ◆ No personal information that could link the patients with the study will be documented or used.
- ◆ The files of patients will not be removed from the ENT practice.
- ◆ During the data collection phase the patient's names will only be recorded to prevent double entries. This list will not be kept for the study and will remain in the ENT practice as a reference list.



- ◆ The coding process ensures patient confidentiality.
- ◆ All information gained in the study will be treated as confidential.

Regards

Alida Naude

Alida Naude
Student

Carina Avenant

Carina Avenant
Supervisor

B Louw

Prof. Brenda Louw
HEAD: DEPARTMENT OF COMMUNICATION PATHOLOGY



Dr. B.L Wolfowitz
PO Box 2310
Houghton
2041

Mrs. A.M Naude
PO Box 2048
Wingate Park
0153

PERMISSION TO ACCESS PATIENT FILES FOR MASTERS DEGREE RESEARCH PURPOSES

Herewith I grant Alida Naude permission to access patient files from my private practice in order for her to complete her study on:

“Clinical and audiological features of Meniere’s disease: Insight into the diagnostic process”.

I hereby acknowledge that the purpose and procedures of this study has been fully explained to me. The expectations, benefits and implications of the results were also discussed.

I further understand that:

- ◆ I can withdraw permission for access to my patient files at any stage;
- ◆ There will be no reimbursement for participating in the research study;
- ◆ All information for research purpose will be kept confidential and anonymous;
- ◆ The abridged version of the results of the study will be made available to me on request.

I, B. L. Wolfowitz, hereby give permission for access to my patient records for the purposes of the abovementioned study.



Dr. B.L Wolfowitz



Mrs. A.M Naude

DR. BRIAN WOLF

M.B., B. Ch. (Wits)
Ph.D. (Wits)
F.R.C.S. (Edin.)



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FACIAL PLASTIC
HEAD & NECK SURGEON

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2004-06-07

Ethical consideration related to the proposed thesis of Alida Naude.

Alida Naude works in my office as an audiologist and has done so for some years.

The audiological tests that she and previous audiologists have performed have been filed in an entirely confidential system.

The research purposes for which this clinical material may be used is entirely ethical, does not breach patient confidentiality in any way, the names and identities of the patient's remain undisclosed to any other party, and the research programs are purely analytical, are not in conflict with any peer reviewed ethics parameters, and are in the best interest of patient care and the development of improved therapeutic options.

None of the patients have objected in any way to the use of such material.

I will personally ensure that this research project conforms to accepted ethical standards and the nature of the study may be submitted to any ethics board or committee for approval.

Yours faithfully

BRIAN WOLFOWITZ



**APPENDIX D:
PB WORD LIST**



PB WORD LIST

CID AUDITORY TEST W-22

LIST 3A

- | | |
|------------|------------|
| 1. Bill | 26. Aim |
| 2. Add | 27. When |
| 3. West | 28. Book |
| 4. Cute | 29. Tie |
| 5. Start | 30. Do |
| 6. Ears | 31. Hand |
| 7. Tan | 32. End |
| 8. Nest | 33. Shove |
| 9. Say | 34. Have |
| 10. Is | 35. Owes |
| 11. Out | 36. Jar |
| 12. Lie | 37. No |
| 13. Three | 38. May |
| 14. Oil | 39. Knit |
| 15. King | 40. On |
| 16. Pie | 41. If |
| 17. He | 42. Raw |
| 18. Smooth | 43. Glove |
| 19. Farm | 44. Ten |
| 20. This | 45. Dull |
| 21. Done | 46. Though |
| 22. Use | 47. Chair |
| 23. Camp | 48. We |
| 24. Wool | 49. Ate |
| 25. Are | 50. Year |



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**APPENDIX E:
CODEBOOK**



CODEBOOK

Column	Variable name	Description
1	Meniere's type	A = Definite (>1 episode of vertigo lasting >20 minutes; audiometrically documented hearing loss; tinnitus/ fullness in the ear; nausea/vomiting; other causes excluded) B = Probable (A but with only one attack) C = Possible (A without hearing loss - vestibular) D = Possible (A without vertigo - cochlear)
2	Presenting symptoms	1 = Vertigo 2 = Hearing loss 3 = Tinnitus 4 = Fullness in ear 5 = Nausea/Vomiting 6 = Fluctuating hearing loss 7 = Tullio phenomenon
3	Sex	1 = Male 2 = Female
4	Age of onset (years)	1 = 20-30 2 = 30-40 3 = 40-50 4 = 50-60 5 = 60-70
5	Hearing level	1 = 0-10 dB 2 = 11-25 dB 3 = 26-40 dB 4 = 41-55 dB 5 = 56-70 dB 6 = 71-90 dB
6	Audiogram configuration	1 = Rising 2 = Flat 3 = Falling 4 = Peak (1-3) 5 = Dip/trough
7	Laterality of disease	1 = Unilateral 2 = Bilateral
8	Family history	0 = No 1 = Yes
9	Speech discrimination	1 = S curve correlates with pure tone 2 = Plateau - 1 <50% discrimination 2 50-70% discrimination 3 >70% discrimination
10	OAE	0 = Test not performed 1 = Normal 2 = Cochlear site of lesion
11	EcoG	0 = Test not performed 1 = Positive 2 = Negative
12	ENG (caloric weakness)	0 = Normal (0-25%) 1 = Mild (26-45%) 2 = Moderate (46-65%) 3 = Severe (66-85%) 4 = Profound (>85%)
13	Dix-Hallpike	0 = Negative 1 = Positive
14	Number of attacks	0 = no attacks 1 = 1 attack 2 = 2 attacks 3 = 3 - 10 attacks 4 => 10 attacks
15	Frequency of attacks	0 = No attacks (cochlear) 1 = 1 attack (probable) 2 = Very frequent (weekly) 3 = Not so frequent (monthly) 4 = infrequent (yearly)
16	Duration of the disease	1 = 0 - 1 months 2 = 1 - 3 months 3 = 3 months - 1 year 4 = 1 - 5 years 5 = >5 years
17	Follow up audiogram level of hearing	1 = 0-10 dB 2 = 11-25 dB 3 = 26-40 dB 4 = 41-55 dB 5 = 56-70 dB 6 = 71-90 dB
18	Duration between audiograms	1 = 0 - 6 months 2 = 6 months - 1 year 3 = 1 - 2 years 4 = 2 - 3 years



APPENDIX F:
ENT PATIENT HISTORY CHECKLIST



SPECIFIC FORMULATED QUESTIONS USED DURING THE HISTORY

TAKING PROCESS.

Vestibular History

1. Ask the patient to describe the vertigo (light headedness/ swimming sensation in head/ blacking out/ tendency to fall/ spinning);
2. When did the patient experience the first episode of vertigo?
3. How many attacks have occurred subsequently?
4. Is the dizziness constant or paroxysmal, coming and going (attacks)?
5. How often do they experience these attacks of vertigo?
6. How long does the vertigo last?
7. Have the attacks been accompanied by nausea and/or vomiting?
8. Does anything precipitate an attack such as loud noise (Tullio phenomenon), dietary intake or positional change of the head?

Cochlear History

1. Has the patient noticed a hearing loss? Does it worsen during or before the attack?
2. Have they noticed a fluctuation in their hearing?
3. Have they experience tinnitus (describe the sound and severity)? When was it first noticed? Does it change during or just before an attack of vertigo?
4. Has the patient noticed an inability to tolerate loud sounds?

History of aural pressure

1. Has there been a sensation of fullness or pressure in the ears or head?
2. If yes, is it there all the time? Does it change before or during an attack?

Additional questions covered in the history

1. Identify medications taken presently and in the past.
2. Is there a family history of hearing loss or vertigo?
3. Have they sustained any injuries to the ear or head?
4. Do they or any family member have a metabolic or relevant systemic disease?