Chapter 1

General introduction
Introduction

In 1861 Prosper Menière was the first one who described a series of patients with the classical triadic symptomatology of hearing loss, vertigo and tinnitus. The essence of his hypothesis was that the symptoms were caused by a disorder of the labyrinth instead of a vascular cerebral dysfunction, as had previously been thought. In his paper published in the ‘Gazette Médicinale de Paris’ [1] Prosper Menière concluded that the inner ear may be suddenly affected causing tinnitus, diminution of hearing and attacks of vertigo. The vertigo may be accompanied by nausea, vomiting and syncope. He described that the attacks were intermittently followed by hearing loss of increasing severity. In the 20th century the interest to establish an exclusive diagnosis and an adequate therapy was growing.

During the last decades a lot of different definitions of Menière’s disease for diagnostic, therapeutic and evaluation purposes have been used. In 1972, the Committee on Hearing and Equilibrium of the American Academy of Ophthalmology and Otolaryngology (AAOO-HNS) defined Menière’s disease as a disease of the membranous inner ear with a characteristic set of symptoms and signs and with a pathological correlate of endolymphatic hydrops [2]. This hydrops of the endolymphatic system in the temporal bones of patients with Menière’s disease was discovered by Hallpike and Cairns, and also Yamakawa, in 1938 [3,4]. Since then endolymphatic hydrops has been generally accepted as the basic histopathological substrate of Menière’s disease. The definition of the disease has been revised in 1985 and in 1995 by the Committee on Hearing and Equilibrium of the AAOO-HNS [5,6]. Despite defining and grading the different symptoms of Menière’s disease by the American Academy of Otorhinolaryngology in 1995 [6], there are still unresolved issues on the definition of the disease, especially on defining duration, uni- and bilateral Menière’s disease, and the necessity of the presence of aural fullness if tinnitus is absent. The above mentioned considerations have to be taken into account when defining Menière’s disease. Basically the definition of Menière’s disease must include the clinical symptomatology as the direct consequence of the idiopathic endolymphatic hydrops as the basic histopathological substrate. Furthermore a uniform applicability of the definition of Menière’s disease is obligatory for clinical and scientific purposes. The diffuse and complex symptomatology of Menière’s disease requests an accurate diagnostic program to confirm the diagnosis and to exclude other pathology. With such a program it might be possible to obtain more information about the cause of the disease and to establish staging, allowing more specific treatment modalities for each particular stage.
Objectives of this study

The Department of Otorhinolaryngology of the University Hospital Groningen reconsidered the definition of Menière’s disease taking the above mentioned considerations into account. The Definition Menière Groningen was developed. With this definition Menière’s disease was defined as a sensorineural (cochlear) hearing loss combined with tinnitus (present now or in the past), vertigo attacks (at least two, present now or in the past) and exclusion of other pathology through the Groningen Diagnostic Protocol. As part of the Definition Menière Groningen the Groningen Diagnostic Protocol was developed to confirm the diagnosis Menière’s disease and to exclude other pathology. The protocol allowed a prospective and systemic evaluation of the diagnostic value of each different test for the diagnosis of Menière’s disease. In addition, the contribution of each test separately and their correlations with regard to a possible classification was further subject of investigation, possibly allowing more specific treatment modalities for each particular stage.

Chapter 2 describes the history and the evolution of defining Menière’s disease. In Chapter 3 the shortcomings in defining Menière’s disease are discussed and the need for a more sophisticated definition of the disease is emphasized. The Definition Menière Groningen is introduced together with a description and definition of symptoms, affected and unaffected ear, uni- and bilateral disease and the duration of the disease. A detailed description of the Groningen Diagnostic Protocol is presented. Furthermore, data management is discussed, including the design of a Menière database and data analysis. The patient characteristics of the studied population are presented in Chapter 4, including the analysis of the start and duration of the disease and the severity of the perceived subjective symptoms. The effects of the four days hospital admission, required for the protocol, are also described.

Chapter 5 describes the results of pure-tone and speech audiometry of this cohort of patients. The shape of the pure-tone audiogram is analyzed. Results are also related to average hearing loss and to duration and severity of disease and symptoms. In Chapter 6 the prospective outcomes of electrocochleography of this cohort are given. The value of electrocochleography for analysis of Menière’s disease is discussed. Results are related to average hearing loss and to duration and severity of disease and symptoms. Chapter 7 presents the results of the prospective study of evoked otoacoustic emissions in this group of Menière patients. Click-evoked as well as distortion product otoacoustic emissions are measured. Results are related to average hearing loss and to duration and severity of disease and symptoms. Chapter 8 describes the results of the vestibular tests of the Groningen Diagnostic Protocol. Vertigo and electronystagmography results in uni-and bilaterally affected
patients are discussed. Results are related to average hearing loss and to duration and severity of disease and symptoms.

The outcomes of the prospectively performed three-dimensional DFT-CISS Magnetic Resonance Imaging study on the patient cohort are demonstrated in Chapter 9. The distance between the vertical part of the posterior semicircular canal and the posterior fossa is measured and compared with this distance of control ears with normal hearing. Also differences between affected and unaffected ears as well as differences between uni- and bilateral disease are studied. Results are related to average hearing loss and to duration and severity of disease and symptoms.

The results of perilymphatic pressure measurement using the tympanic displacement analyzer on the patient cohort are presented in Chapter 10. A group of normal hearing adults are used as a control. Results are also related to average hearing loss and to duration and severity of disease and symptoms.

Chapter 11 describes the results of the assessment of aldosterone in the patient cohort. Results are also related to average hearing loss and to duration and severity of disease and symptoms.

Chapter 12 finally is an evaluation of the Definition Menière Groningen and the Groningen Diagnostic Protocol as has been used in this thesis. Based on the results of this thesis and the literature a renewed version of the Definition Menière Groningen is introduced, called the Definition Menière Groningen 2001. The Definition Menière Groningen 2001 including a diagnostic protocol is introduced for clinical purposes. The Definition Menière Groningen 2001 including an extended diagnostic protocol is proposed for scientific analysis.

References


