

Chapter 2

History and definition

History

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. This was published in the 'Gazette Médicinale de Paris' [1]. In this paper Prosper Menière concluded that the sudden dysfunction of the inner ear is not only responsible for the symptoms of tinnitus and diminution of hearing, but also lead to attacks of vertigo, dizziness, uncertain gait, staggering and falling. The attacks may be accompanied by nausea, vomiting and syncope. He described that the attacks were intermittently followed by hearing loss of increasing severity.

Centuries before Prosper Menière described his classical triadic symptomatology Menière-like symptoms were already reported. Plutarch, a Roman historian, already suggested that Julius Caesar suffered from a 'falling sickness' which was characterised by 'giddiness' which was referred to by Shakespeare, who in addition made Caesar hard of hearing in the left ear [2]. Centuries later Martin Luther once wrote to someone who complained of the itch 'I would give ten florins to change with you; you do not know how distressing vertigo is. At this very moment I am unable to read a letter through at once, and indeed I cannot read more than two or three lines of my Psalter; for when I make the attempt such a buzzing comes on my ears that I am often on the point of falling off my seat. On the other hand the itch is a useful thing' [3]. Luther was convinced about the fact that the devil was violently attacking his ear [4].

In the 18th century paroxysmal vertigo was well recognised. It was not very well defined at that time. Doctors and scientists thought that the origin of it was central. In 1821 Itard described two groups of patients both with 'cerebral apoplexy'. One group recovered from their attacks without subsequent neurological deficit and the other group did not. He suggested that the attacks of dizziness in some cases could be caused by a disease of the ear, instead of a central nervous disorder. In detail he described a patient having characteristic symptoms of Menière's disease [5].

In 1881 Jean Marie Charcot, a leading neurologist of his time, mentioned Menière's communication to the Academy of Medicine in June 8, 1861. He did so in his 17th and 18th lecture, titled 'Menière's vertigo ('Vertigo Ab Aura Laesa')'. Charcot listed the symptoms of Menière's disease, charging his colleges to memorize the characteristics of the disease so that they should not continue to misdiagnose Menière with epilepsy [6]. The confusion regarding the diagnosis can be illustrated by the so called epilepsy of the famous Dutch impressionist Vincent van Gogh. Based on van Gogh's written statements in letters, he suffered from frightening attacks of disabling, recurrent vertigo, with nausea

and auditory and visual disturbances that were described as hallucinations. He used the French word 'vertige' to describe his attacks of vertigo. Between the severe attacks, persistent imbalance, motion intolerance, and positional dizziness accompanied by intolerance for loud sounds might cluster for several months. Van Gogh's handwritten statements describing his attacks and his illness are a compelling evidence for the diagnosis Menière's disease and not epilepsy [7,8].

In the 19th century a lot of important research on otologic and vestibular disorders was performed in France around Paris. By experiments on pigeons Flourens proposed that the semicircular canals were involved in the maintenance of posture and balance. He hypothesized that a lesion in the semicircular canals was responsible for the earlier described vestibular symptomatology [9,10]. In 1872, Simon Duplay coined the term 'maladie de Menière' for this dysfunction [11]. Mc Bride felt the need for a thorough neurological and otological examination to exclude non-labyrinthine disease [12].

Politzer of Vienna first described and quantified results of treatment for patients with Menière's disease. In 1902 he measured an improvement of hearing and a reduction of the number and the severity of the attacks. Menière's findings were critically discussed in his book 'Diseases of the ear' [13]. Dandy first noticed that the attacks lasted from days to weeks, with the patients feeling well between the attacks [14]. A fluctuating hearing loss was first described by Crowe [15] and Mygind et. al. [16]. Crowe also stated that the attacks occurred at irregular intervals at any time and patients were well between attacks. He personally felt that the diagnosis should be based mainly on the history. Aural fullness and a sensation of pressure in the ear were reported less than 10 years later by Lindsay [17] and Cawthorne [3].

In 1934 Furstenberg pointed out that many otologists had made their own classification at that time. In his opinion this made it impossible to have comparable groups for evaluating treatments. He defined Menière's disease as violent attacks of vertigo with nystagmus in a patient with deafness, associated with nausea, vomiting and tinnitus [18].

In 1927 Portmann treated patients having vertigo with endolymphatic sac surgery. He described his operating technique and poor results in 1927 [19]. For this he used the work of Knapp who described the complaints of vertigo, deafness and tinnitus as the Triad of Menière [20]. In 1927 Portmann was invited for the first 'Congress of the 'Collégium Oto-rhino-laryngologium'' by Professor Benjamins and Dr de Kleyn in Groningen, The Netherlands. In his oral presentation called 'Recherches sur le sac endolymphatique. Résultats et applications chirurgicales' he again discussed his endolymphatic sac surgery procedures and their poor outcomes [21].

In the past therapeutic options in Menière's disease were legion [22]. Until now a real causative treatment for Menière's disease has not been found. By necessity, treatment

was, and still is, mainly symptomatic. It is well known that the placebo-effect of any treatment modality is large.

Definition

During the last decades many different definitions of Menière's disease have been used. In 1902, for example, the Dutch handbook of Otolaryngology defined Menière's disease as follows: 'The typical spells of 'ear dizziness', known as Morbus Ménièrei can give symptoms of an acute balance disorder, causing sudden dropping attacks with nausea and vomiting. During the attack the patient is conscious. Even when the dizzy spell is mild, it will still appear in attacks; only occasionally the balance disorder is permanently present. Typical is the subjective sensation of rotation and the appearance during sudden head movements' [23].

Although the definition of Menière's disease as a specific entity was worldwide recognised, confusion in the terminology remained. There was a great deal of variability in the vast literature on Menière's disease concerning its epidemiology, clinical features and natural history. Some of this variability may be real but a significant part must be regarded as the result of differing diagnostic criteria.

It was therefore essential that strict diagnostic criteria were developed. In 1972 the Committee on Hearing and Equilibrium of the American Academy of Ophthalmology and Otolaryngology (AAOO) defined Menière's disease as a disease of the membranous inner ear with a characteristic set of symptoms and signs (table 1) and with a pathological correlate of endolymphatic hydrops [24]. 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. Since then this endolymphatic hydrops has been generally accepted as the basic histopathological substrate of Menière's disease [25,26].

Since 1972 there have been two updated versions by the AAOO of these criteria. The first one in 1985 is listed in table 2 [27]. The later version of 1995 is listed in table 3 [28]. The 1995 criteria aimed to simplify the definition. They allowed more flexibility, making it more usable in a wide range of settings. The patient's history became more important. Menière's disease was redefined as the idiopathic syndrome of endolymphatic hydrops and the importance of exclusion of other possible causes were discussed.

Table 1. 1972 Criteria for the diagnosis of Menière's disease. Committee on Hearing and Equilibrium; report of subcommittee on Equilibrium of the American Academy of Ophthalmology and Otolaryngology, AAOO.

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- Fluctuating, progressive, sensorineural hearing loss.
 - Episodic, characteristic definitive spells of vertigo lasting 20 minutes to 24 hours.

The patient remains fully conscious with no neurological accompaniments or sequelae; during the spells vestibular nystagmus is always present.

- Usually tinnitus.

The attacks are characterised by periods of remission and exacerbation.

Table 2. 1985 AAOO-Criteria for the diagnosis of Menière's disease.

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- A fluctuating, sensorineural hearing loss associated with tinnitus - the deficit is characteristically of low frequency or flat type.
 - Vertigo - spontaneously occurring sensation of movement that is accompanied by unsteadiness and lasts from minutes to hours. More than one attack needed to establish diagnosis

Definitive spell - often prostrating, often accompanied by nausea and vomiting. Patient oriented and conscious, no neurological sequelae. Horizontal or horizontal rotatory nystagmus is always present during the definitive spell.

Table 3. 1995 AAOO-Criteria for the diagnosis of Menière's disease.

- Recurrent, spontaneous episodic vertigo. definitive spell - spontaneous rotational vertigo lasting at least 20 minutes (commonly several hours), often prostrating, accompanied by disequilibrium that may last several days, usually nausea, commonly vomiting or retching, no loss of consciousness. Horizontal or horizontal rotatory nystagmus is always present.	
- Sensorineural hearing loss (not necessarily fluctuating).	
- Either aural fullness or tinnitus (or both).	
- Certain Menière's disease:	'Definite' disease with histopathological confirmation.
- Definite Menière's disease:	Requires two or more definite episodes of vertigo of at least 20 minutes with hearing loss plus tinnitus and/or aural fullness. Audiometrically documented hearing loss on at least one occasion
- Probable Menière's disease:	Needs only one definitive episode of vertigo and the other symptoms and signs. Audiometrically documented hearing loss on at least one occasion.
- Possible Menière's disease:	Definitive episodic vertigo without documented hearing loss or documented sensorineural hearing loss, fluctuating or fixed, with disequilibrium, but without definite episodes (= non-definitive vertigo).

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