

Meniere's disease: is it a bilateral disease?

F. SALVINELLI, M. TRIVELLI, F. GRECO, M. SILVESTRINI*, E. FERNANDEZ**, R. PALLINI**

Institute of Otolaryngology, "Campus Bio-Medico" University - Rome (Italy)

*Institute of Neurology, "Tor Vergata" University - Rome (Italy)

**Institute of Neurosurgery, Catholic University "Sacro Cuore" - Rome (Italy)

Abstract. – **Background.** Bilateral Meniere's disease (MD) is still controversial due to different criteria used to assess the involvement of the primarily affected ear and the contralateral one. We evaluated the percentage of bilateral forms in 49 patients with MD.

Methods. 49 patients with (MD) were studied. All were selected according to the following requirements: history, tonal audiometry, glycerol test, Auditory Brainstem Response (ABR), vestibular examination. Magnetic Resonance (MR) was performed in 14 patients.

Results. A raised hearing threshold in the contralateral ear was found in 23 patients, but only 7 (14.3%) fulfilled the requirements to be considered affected by bilateral MD. The delay of occurrence in the contralateral ear was 7 years (from 5 to 12 years). Submillimeter Magnetic Resonance is determinant for differential diagnosis with Meniere-like syndromes.

Conclusions. A conservative approach in surgical treatment of unilateral Meniere's disease is recommended because of the possibility of evolution in a bilateral form, that can occur even after 10 years from the onset of the disease.

Key Words:

Vertigo, Hearing loss, Hydrops, Magnetic resonance, Endolymphatic duct, Surgery.

Introduction

The pathoetiology of Meniere's disease (MD), identified more than 100 years ago, remains controversial^{1,2,3}.

Researches don't pay much importance to the problem of its bilaterality even if it is relevant for a correct therapy^{4,7} and data in literature are still controversial (Castellano 2%; Jonkees 78%)^{6,8}. Different data could be a consequence of the criteria used to confirm

the secondary involvement of the ear contralateral to the primarily affected one.

Greven⁹ on 292 patients observed a high percentage (73%) of bilateral MD regarding the hearing loss; while this percentage was significantly reduced (29%) when also tinnitus and recruitment were considered. Paparella⁷ on 360 patients observed a high percentage (78.8%) of bilateral ear but only a small percentage (32%) presented a Meniere's clinical picture. Camarda⁵ on 28 patients affected by MD for at least 5 years observed only 2 cases of bilateral disease.

Different data in literature depends probably from different criteria used for diagnosis of MD. In fact patients with symptoms of MD beginning from a primarily affected ear, underestimate the involvement of the contralateral ear¹⁰.

We consider the percentage of bilateral forms in 49 patients with MD. This study took place in our Audiology Department.

Material and Methods

We evaluated 49 middle-aged patients whose average age was 51.57 years (from 28 to 77 years). The group was comprised of 19 males and 30 females who had been affected by unilateral MD for at least 5 years. All cases were selected according to the following items:

- History to investigate typical triad: tinnitus, fluctuating hearing loss at pure tone audiometry with recruitment¹¹ and vertigo. Aural fullness was included in the evaluated symptoms;
- Glycerol test;
- Auditory Brainstem Response (ABR) to confirm the cochlear nature of sensorineural hearing loss;

- Vestibular examination by bithermal caloric test (Fitzgerald-Hallpike 30/44°C) and nystagmographic recording;
- Magnetic Resonance (MR) in order to exclude diseases of the cochlear nerve and to investigate the vestibular aqueduct and endolymphatic sac, which authors report to be clearly narrowed in MD patients^{12,13}.

Great attention was paid to find signs of involvement of the contralateral ear.

The criterion used by us to make diagnosis of bilaterality was the presence of tinnitus and cochlear sensorineural hearing loss with objective (pure tone audiograms) or subjective (anamnesic response) signs of fluctuating hearing loss in the opposite ear. The fluctuation in hearing level, as a consequence of MD crisis or glycerol test, was regarded as positive when its level, at least for low-tones, was 15dB lower and returned to its original values¹⁴.

Results

A raised hearing threshold in the contralateral ear was found in 20 patients but only 7 were found affected by bilateral Meniere's disease.

The patients with unilateral MD (42/49; 85.7%) consisted of 18 males and 24 females with the average age being 51 years. The average duration of the disease was 12 years (from 11 to 33 years). The mean age of the patients at the onset of the disease was 42 years (Table I).

Hearing loss of the hydroptic ear affected was considered slight in 11 patients, moderate in 26 patients and severe in 5 patients;

Table I. Epidemiologic data.

	Unilateral	Bilateral
No. of cases	42 (85.7%)	7 (14.3%)
Males	18	1
Females	24	6
Age at presentation (yrs)	51 (28/77)	55 (42/73)
Age of onset (yrs)	42	38
Duration of the disease (yrs)	12 (10/31)	19.1 (10/39)

Table II. Severity of hearing loss.

	Unilateral		Bilateral	
	PA-Ear	CL-Ear	PA-Ear	CL-Ear
Slight	11	10	0	5
Moderate	26	7	3	2
Severe	5	3	4	0
No. of cases	42	20	7	7

PA-Ear = primarily affected ear; CL-Ear = contralateral ear

hearing loss in the contralateral ear was considered slight in 10 patients, moderate in 7 patients and severe in 3 cases (Table II).

Vestibular examination by bithermal caloric test was carried out of Meniere's fit and it revealed an unilateral labyrinthine depression in 31 patients, bilateral depression in 2 patient and a normal caloric response in 11 patients (Table III).

In the patients affected by bilateral MD (7/49; 14.3%) 6 were females and only 1 was male. They were middle-aged, 55 years (from 42 to 73 years) (Table I).

Hearing loss of the ear primarily affected was, moderate in 3 patients, severe in 4 patients. In the contralateral ear it was slight in 5 patients, moderate in 2 cases (Table II).

Vestibular examination in bilateral disease showed a normal labyrinthine response in 4 cases, a bilateral depression in 2 cases and a bilateral hyperactive response in only 1 case (Table III).

Glycerol test was found positive in 4 patients.

The average duration of the disease was 19.1 years (from 10 to 39 years) in middle-aged patients of 36.9 years (Table I).

In the remaining cases the average duration between the involvement of the first and the second ear was 7 years (from 5 to 12 years).

Table III. Bithermal caloric test (FH 30/44°C).

	Unilateral	Bilateral
No. of cases	42	7
Normal caloric responses	11	4
Hyperactive caloric responses	0	1
Depressed caloric responses	31	2

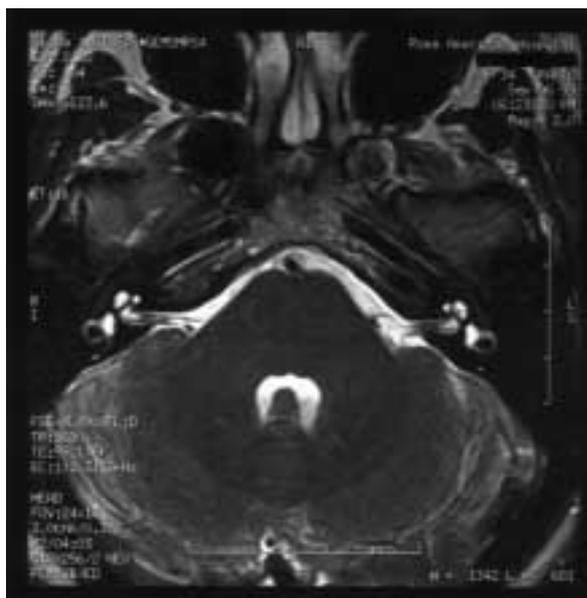


Figure 1. MRI with contrast enhancement. The membranous labyrinth has normal CSF signal. The endolymphatic duct is visualized on the right side, while on the left the endolymphatic duct is not visible and the sac is reduced in volume.

On MR the degree of visualization of the membranous endolymphatic duct and sac was found significantly hypoplastic in the MD group (Figure 1). A normal vestibular aqueduct was found bilaterally in two patients with unilateral MD.

Discussion

Data regarding incidence and evolution of bilateral Meniere's disease are still controversial, due to different criteria used to assess the involvement of the primarily affected ear and the contralateral one^{7,8,10,14-19}. We observed 7 cases of bilateral MD, 14.3% of the MD patients.

Deafness, tinnitus and recruitment are not signs to be considered in the involvement of the contralateral ear, as they are common in most cochlear hearing loss. On the contrary, the presence of fluctuating sensorineural hearing loss, after a fit of vertigo or glycerol test, should be a rigorous criterion in order to make a diagnosis of involvement of the contralateral ear.

Meniere's syndrome, incorrectly thought as Meniere's disease, is to be studied too. The number of patients affected by Meniere's syn-

drome is growing thanks to the latest techniques of investigation. In our study 4 cases were not considered by statistics. At the beginning they were classified as bilateral Meniere's disease but then they were classified as:

- Chiari's syndrome type I (2 cases);
- abruption of blood-brain barrier (1 case);
- multiple sclerosis (1 case).

MR is determinant for differential diagnosis with some of Meniere's syndrome¹⁵ and it appears important the evaluation with a submillimeter MR of endolymphatic duct and sac²⁰. In our case load, it was never completely detectable the entire membranous labyrinth on the affected size of patients with MD, and the same marked failure to visualize it was found bilaterally in bilateral Meniere patients. It was always possible to visualize the membranous endolymphatic duct and sac on the healthy side of patients with monolateral disease.

Therefore it would be right to include this study in the diagnostic protocol of Meniere's disease and to revise the statistics in order to obtained results.

As well as the incidence, the lenght of time occurring between the involvement of the ear primarily affected and contralateral is subject to debate. Our data show a longer duration of bilateral MD (19.1 years) in relationship to unilateral form (12 years) and a changeable involvement of the contralateral ear to that primarily affected. The average duration of transformation of unilateral Meniere's disease in bilateral form was 7 years (from 5 to 12 years)(Table IV). This outcome, in spite of a small sample group, would seem to show that most conversions in bilateral forms verify in the first 10 years from the onset of the disease. On the contrary, some authors^{7,14,21} think that the in-

Table IV. Delay of the occurrence in the contralateral ear (7 cases).

	Delay (yrs)	No. Cases
	5	3
	7	2
	9	1
	12	1
Average	7.1	

volvement of healthy ear occurs after 5 years. Others think that the longer time passes from the onset of the disease, the higher is the number of conversions^{10,18,19,22-24}.

A conservative approach in surgical treatment of unilateral MD is recommended because of the possibility of transformation in a bilateral form.

The advancement of imaging modality, with the use of submillimeter MR imaging of the temporal bone in Meniere's disease, allows to detect the smaller dimension of endolymphatic ducts and sacs in MD patients²⁵⁻²⁷. The majority of subjects with Meniere's disease have non visualized endolymphatic ducts bilaterally in the symptomatic ear. This finding suggests that the membranous endolymphatic duct and sac collapse and hypoplasia of the vestibular aqueduct occurs in Meniere's disease patients. This finding represents a significant help in this. The membranous endolymphatic duct and sac is not well detectable with MR on the affected side in the majority of patients, while it is possible to visualize it, at least partially, on the healthy side. This may explain the unilateral MD disease while the degree of visualization on the healthy side may allow to forecast a predisposition to the development of bilateral disease in Meniere's disease group²⁸.

On the other side, the primary cause of the increased endolymphatic volume appears to be not yet completely clear. In our study two patients presenting with a classic unilateral disease, showed a normal MR evidence of membranous labyrinth bilaterally. Further investigation need to be performed in order to understand the precise causes that lead to an imbalance between secretion and resorption of the endolymph in MD.

References

- 1) MENIERE P. Sur une forme de surdit  grave dependant d'une lesion de l'oreille interne. *Gazette Medicale* 1861; 16-29.
- 2) ARSLAN M, SALA O, MOLINARI GA. Experimental reproduction of Meniere's disease. *International Symposium on Meniere's Disease*. Italy: Padua, 1962.
- 3) PAPARELLA MM. The cause (multifactorial inheritance) and pathogenesis (endolymphatic malabsorption) of Meniere's disease and its symptoms (mechanical and chemical). *Acta Otolaryngol (Stockh.)* 1985; 99: 445-451.
- 4) BOHMER A, FISH U. Bilateral vestibular neurectomy for treatment of bilateral Meniere's disease. *Third international Symposium on Meniere's disease*. Italy, Rome, 1993.
- 5) CAMARDA M, DI BLASI A, GIORDANO M, SABATO M, ANGELINI F, POLLA A. Is Meniere's disease bilateral? *Third international Symposium on Meniere's disease*. Italy: Rome, 1993.
- 6) CASTELLANO R. Meniere's disease and its surgical treatment. *Neurosurgery* 1951; 8: 173.
- 7) PAPARELLA MM, GRIEBIE MS. Bilateral Meniere's disease. *Acta Otolaryngol* 1984; 97: 233-237.
- 8) JONKEES LB. Some remarks on the patients suffering from Meniere's disease. *Trans Am Acad Ophthalmol Otolaryngol* 1971; 75: 374.
- 9) GREVEN AJ, OOSTERVELD WJ. The contralateral ear in the Meniere's disease. A survey of 292 patients. *Arch Otolaryngol* 1975; 101: 608-612.
- 10) KITAHARA M, MATSUBARA H, TAKEDA T, YAZAVA Y. Bilateral Meniere's disease. *Adv Otorhinolaryngol* 1979; 25:117.
- 11) DEL BO M, GIACCAI F, CALOGERO R, ARAN JM, PORTMANN CL. Problemes actuels en audiologie clinique. *Rev Laringol* 1986; 107: 171.
- 12) HEBBAR GK, RASK-ANDERSEN H, LINTHICUM FH Jr. Three-dimensional analysis of 61 human endolymphatic ducts and sacs in ears with and without Meniere's disease. *Ann Otol Rhinol Laryngol* 1991; 100: 219-225.
- 13) TANIOKA H, ZUSHO H, MACHIDA T, SASAKI Y, SHIRAKAWA T. High-resolution MR imaging of the inner ear: findings in Meniere's disease. *Eur J Radiol* 1992; 15: 83-88.
- 14) THOMAS K, HARRISON MS. Long-term follow-up of 610 cases of Meniere's disease. *Proc Roy Soc Med* 1971; 64: 853-856.
- 15) PULEC JL. Meniere's disease: Results of a five years study. Read before the Fourth Extraordinary Meeting of the Barany Society. Los Angeles 1974.
- 16) WRIGHT AJ. The deafness of Meniere's disease. *J Laryngol Otol* 1943; 58: 379-380.
- 17) BALKANY TJ, SIREB B, AREMBERG I. Bilateral aspects of Meniere's disease: an underestimated clinical entity. *Otolaryngol Clin North Am* 1980; 13: 603.
- 18) ANDREOLI C. La bilateralit  della malattia di Meniere. *Audiol Ital* 1986; 3: 286-291.
- 19) GREEN JD JR, BLUM DJ, HARNER SJ. Longitudinal followup of the patients with Meniere's disease. *Otolaryngol Head Neck Surg* 1991; 104: 783-788.
- 20) PALUDETTI G, COLOSIMO C JR, DI NARDO W, GALLI J. Neurovascular anomalies in Meniere's disease. *Meniere's disease Pathogenesis, Pathophysiology, Diagnosis and Treatment*. Third international symposium on Meniere's disease. Italy: Rome, 1993.

- 21) ENANDER A, STAHL J. Hearing in Meniere's disease. A study of pure tone audiogram in 334 patients. *Acta Otolaryngol* 1967; 64:543-556.
- 22) STAHL J, BERGMAN B. The caloric reaction in Meniere's disease. *Laryngoscope* 1967; 9: 1629-1643.
- 23) MORRISON AW. Management of sensorineural deafness. London: Butterworth & CO Ltd, 1975: 145-173.
- 24) FRIBERG U, STAHL J, SVEDBERG A. The natural course of Meniere's disease. *Acta Otolaryngol Suppl* 1984; 406: 72-77.
- 25) WELLING DB, CLARKSON MW, MILES BA et al. Submillimeter magnetic resonance imaging of the temporal bone in Meniere's disease. *Laryngoscope* 1996; 106: 1359-1364.
- 26) TANIOKA H, TERAHARA A, FURUTA A et al. MR imaging of the inner ear; findings of Meniere's syndrome. *Rinsho Hoshasen* 1989; 34: 1365-1370.
- 27) SANDO I, IKEDA M. The vestibular aqueduct in patients with Meniere's disease. A temporal bone histopathological investigation. *Acta Otolaryngol (Stockh)* 1984; 97: 558-570.
- 28) ALBERS FWJ, VAN WEISSENBRUCH R, CASSELMAN JW. 3DFT-magnetic resonance imaging of the inner ear in Meniere's disease. *Acta Otolaryngol (Stockh)* 1994; 114: 595-600.