

Migrainous vertigo and Ménière`s disease: Is there a common genetic background?

Neuhauser et al.¹ prospectively evaluated 200 patients from a dizziness clinic and 200 ones from a migraine clinic. Prevalence of migraine according to International Headache Society (IHS) II criteria was 38% in the dizziness clinic and 24% in sex and age matched controls ($p < 0.01$). Migrainous vertigo was present in 7% of patients of the dizziness clinic and in 9% of the ones in the migraine clinic. In 15 of 32 patients vertigo was always associated with migraine during the acute attacks. In 16 patients this association was irregular and 2 patients never had both migraine and vertigo together.

Radtke et al.² studied 78 patients (40 male and 38 female) aged 29 to 81 years old, all with idiopathic uni or bilateral Ménière`s disease according to the AAO-HNS criteria. Lifetime prevalence of migraine with and without aura was 50% among Ménière`s patients and 25% among normal controls ($p < 0.001$). Furthermore 45% of the patients with Ménière`s disease always experienced at least one of the migraine symptoms (headache, photophobia, aura) symptoms during the acute attacks. They postulated a pathophysiologic link between migraine and Ménière`s disease.

Urkur et al.³ studied VEMP parameters in Migrainous Vertigo, Ménière`s disease and migraine and found very similar results for all the patients. They also postulated an etiologic link for these syndromes.

Gazques et al.⁴ published a paper on recent Advances in the genetics of recurrent vertigo including familial episodic ataxias, Migrainous Vertigo and Ménière`s disease. They found that 10 to 20% of Ménière`s disease patients have positive family history for this disease.

Cha et al.⁵ described 6 families with index patients affected by Ménière`s syndrome and Migraine. There were 56 siblings affected. Of these, 26(41%) met the IHS II criteria for Migraine. Fifty per cent had Migraine with aura. Three patients had typical aura without headache. Sixty three family members had recurrent spells of spontaneous vertigo. There were 3 twin pairs, 2 mono and 1 dizygotic. One homozygotic pair had Migraine and Ménière`s syndrome while the other had migraine and episodic vertigo without auditory symptoms. They proposed a common heritage for Migrainous Vertigo and Ménière`s disease with variable penetrance.

Bertora & Bergman⁶ studied using quantitative EEG (qEEG) 120 patients who had Ménière`s disease and Migraine and 85 Ménière`s disease patients without migraine. Eighty five per cent of the Ménière`s disease patients had haemodynamic brain variations like the

ones found in Migraine. Brain electric depolarizations and cortical irritative focuses are common to Migraine and Ménière`s disease patients. However Ménière`s disease patients had important hyperactivity in the limbic lobe.

Brown in 1941⁷ and 1949⁸, Bernstein in 1965⁹ published several families with hereditary Ménière`s syndrome transmitted in an autosomal dominant mode. All siblings of these families had strong headaches associated with the Ménière`s symptoms. They did not attempt to characterize the headache according to the IHS criteria.

In 1992 Oliveira & Braga¹⁰ published one family with hereditary Ménière`s syndrome in which the index patient had full blown Ménière`s syndrome associated with very strong headache and drop attacks. One daughter and two sons of this patient had exactly the same clinical symptoms as their father. All of them had low tone sensorineural hearing loss and an endolymphatic sac procedure done in the index patients cured the drop attacks and made the vertigo attacks very mild. Twelve years later these results were unchanged. Again the headache was not studied and classified according to IHS criteria.

In 1997 Oliveira et al.¹¹ published another family with Ménière`s syndrome in the index patient and in some siblings, headache alone in others, atypical vertiginous symptoms in others associated with headache in some of them. This time a careful study of the headache was done and it was classified as migraine according to the IHS II criteria.

In 2002 Oliveira et al.¹² published 6 families from Brasília with siblings displaying a spectrum of symptoms from Migraine alone to full blown Ménière`s syndrome and migraine. Migraine with atypical vertiginous crisis, vertiginous crisis alone and Ménière`s syndrome without migraine were seen in siblings of these six families.

The 1997 family was followed during 12 years and the natural history of the migraine-vertigo syndrome was established: most of the times migraine would come before the vertiginous symptoms and the intensity of the migraine would diminish as the vertigo became more intense.

Later this family was studied from the molecular genetics stand point and a locus in chromosome 5-(5q35) showed a high lod score for the gene¹³. Efforts to identify this gene are under way at this point.

All the families described above had an autosomic dominant mode of genetic transmission with variable penetrance.

Vestibular symptoms associated with Migraine is called Vertiginous Migraine and vestibular symptoms associated with auditory symptoms (tinnitus and low tone fluctuating sensorineural hearing loss) is called Ménière`s disease. In the families described above we found siblings with migraine and both auditory and vestibular symptoms as well as with vestibular symptoms alone. Other combinations of symptoms were found in other siblings. A spectrum of symptoms seems to be caused by the variable penetrance of the gene in these families.

In the 1997 family who was followed for 12 years we could see that as time went by these combinations of symptoms could change. For instance the index patient -who had absolutely classic Ménières syndrome including fluctuating low tone sensorineural hearing loss and was indeed disabled when first seen today has very little annoyance from vertigo and migraine. Three sons of the index patient had intense migraine alone and sometimes had to be hospitalized to treat the migraine crisis.

All these observations led me to think that Migraine, Vertiginous Migraine and Ménière`s disease may be part of a spectrum linked by a common genetic origin.

Of course the multifactorial etiology for these symptoms is real and life style, diet and other factors modify the expression of the genetic trait¹⁴.

Identifying the gene will probably prove or discard this theory.

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