

Chronic Cerebrospinal Venous Insufficiency (CCSVI) IN Meniere Disease. Case or Cause?

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Abstract

CCSVI is the acronym for Chronic Cerebrospinal Venous Insufficiency, initially described by P.Zamboni, as being strongly associated with multiple sclerosis (MS). It is a syndrome characterized by stenosis of the internal jugular veins (IJVs) and/or azygous vein (AZ) with opening of collaterals and insufficient drainage.

Bavera PM carried out 823 Duplex exams on a control group of 60 patients without MS. As expected CCSVI was found only in few subjects of the control group, three, two females and one male, but all affected with Sudden Sensorineural Hearing Loss (SSHL).

Successively, we reported a case of bilateral SSHL with vertigo, showing evidence of the CCSVI pattern at Duplex examination (not associated with MS). To the best of the authors' knowledge, this kind of association has never been reported.

We studied 52 patients affected with cochleo-vestibular disturbances subdivided into two groups of out-patients: Definite unilateral Meniere (Men): 12 subjects (8 males and 4 females, mean age 41,6.yy) according to international AAO-HNS 1995 diagnostic criteria - No-Meniere (No-Men): 14 subjects (6 males and 8 females, mean age 44,7.yy) affected with unilateral cocleo-vestibular impairment

A third group of subjects have been considered, as a "normal" group, 13 patients (8 females and 5 males, mean age 45,5 yy) affected with Benign Paroxysmal Positioning Vertigo (BPPV) with cochlear involvement

Asymmetrical arterious flow in VA or CA was revealed in 2 Men 9 no-Men and 1 BPPV, respectively 12,5 - 60,7 - and 8,6 %. Differences between Men and No-Men and between each of this group with respect to BPPV were highly significant ($p < 0.001$). Asymmetrical venous flow in IJV or VV was detected in 9 patients in MEN group and in 4 in no-MEN and 2 BPPV, respectively 79 - 28,5 and 13 %. Differences between Men and No-Men and between each of this group with respect to BBV were highly significant ($p < 0.001$)

Key words: *CCSVI, Meniere Disease, Vertigo, Hearing loss*

Introduction

As a matter of fact the venous outflow of the neck has aroused interest, and importance, mostly in these last three years after the so-called Zamboni method aimed to detect and study “Chronic Cerebrospinal Venous Insufficiency” (CCSVI) [1]. This type of exam and problem, based on detection of abnormal venous outflow from the head and neck, is principally linked with multiple sclerosis disease. It is characterised by multiple stenoses of the extracranial venous draining pathways, i.e. the internal jugular veins and the Azygous veins, which lead to collateral formation, alteration of the blood-brain barrier. It could be diagnosed by means of MRI digital Venography and, especially, Duplex examination of cerebral and cervical vein system. [2,3]

Abnormalities of cerebro-spinal venous flow have been described also in no-MS patients. For example Koerte et al. [4] performed a 2D time-of-flight MR-venography of the upper neck region to visualize the venous vasculature in patients with migraine. They showed a higher prevalence of dense secondary extracranial venous networks and a significantly larger percentage of venous outflow through secondary channels, through epidural, vertebral, and deep cervical veins.

Bavera PM [5] carried out 823 Duplex exams on a control group of 60 patients without MS. As expected CCSVI was found only in few subjects of the control group, three, two females and one male, but all affected with Sudden Sensorineural Hearing Loss (SSHL).

Successively, we reported [6] a case of bilateral SSHL with vertigo, showing evidence of the CCSVI pattern at Duplex examination (not associated with MS). On the basis of these experiences we decide to investigate Cerebrospinal Venous system in Otoneurological patients even if not associated with Multiple Sclerosis.

In order to clarify the role of venous drainage in Meniere Disease we compared cervico-cephalic emodynamic (arterial and venous flows) in three groups of patients affected with vertigo, Meniere Disease (MD), no-Meniere and Benign Paroxysmal Positioning Vertigo (BPPV).

Material and Methods

We studied 52 patients affected with cochleo-vestibular disturbances subdivided into two groups of out-patients:

- Definite unilateral Meniere (Men): 12 subjects (8 males and 4.females, mean age 41,6.yy) according to international AOO-HNS 1995 diagnostic criteria [7]
- No-Meniere (No-Men): 14 subjects (6 males and 8 females, mean age 44,7.yy) affected with unilateral cocleo-vestibular impairment

A third group of subjects have been considered, as a “normal” group, 13 patients (8 females and 5 males, mean age 45,5 yy) affected with Benign Paroxysmal Positioning Vertigo (BPPV) with cochlear involvement

All of them underwent to a complete audio-vestibular investigation by the same audiologist (MV) according to international accepted protocol [8] . Diagnosis was posed by the same otoneurologist (A.DC).

ECD was performed by the same specialist (BPM) and two test were performed:

- 1) Arteries test. Flow of vertebral artery (VA) of both sides have been investigated both

in static and dynamic positions in order to evaluate the variations of arterial flow due to head and neck movements. The flow, static and dynamic, of the cochlear artery (CA) of both sides have been investigated, too, according to the method described by Belcaro and Nicoalides [9] with the patient is completely lying flat on his back in the most comfortable way, with a low pillow, searching absolute neck relaxation to avoid possible muscular contraction. The Duplex access to “find” the cochlear arterial system may differ from one patient to another and should be carried out from a posterior and anterior projection of the ear and “working” a lot on the machine’s capacities. It isn’t only a matter of what is seen but also of what is heard and transformed into a spectral flow analysis. The blood flow exam is mostly based on the CW scale more than on the imaging itself because of the small vessel dimensions.

ECD was considered abnormal when the vertebral and/or the cochlear artery presented a reduced flow in static and/or dynamic conditions at the same side of the affected ear at least of 50% with respect of the controlateral side

2) Veins test, according to the protocol described by Zamboni et al. [1]. The CCSVI protocol exam at 00° and 90° was carried out on informed and willing patients according to CCSVI diagnosis procedure.

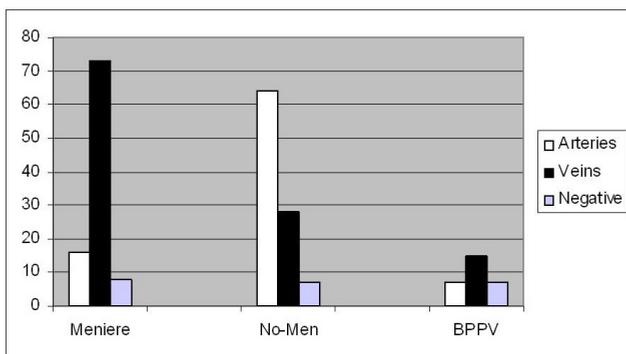
ECD was considered abnormal when at least two out five of Venous Haemodynamic Insufficiency Severity Score (VHISS) criteria were satisfied.

Results were discussed with an independent well-trained otoneurologist (HA). MEN vs no-MEN vs BPPV findings were compared by means of Student t-test and considered as significant at 0.005 p level.

Results

Asymmetrical arterious flow in VA or CA was revealed in 2 Men 9 no-Men and 1 BPPV, respectively 12,5 - 60,7 - and 8,6 %. Differences between Men and No-Men and between each of this group with respect to BPPV were highly significant ($p < 0.001$).

Asymmetrical venous flow in IJV or VV was detected in 9 patients in MEN group and in 4 in no-MEN and 2 BPPV, respectively 79 - 28,5 and 13 %. Differences between Men and No-Men and between each of this group with respect to BBV were highly significant ($p < 0.001$)



Tab.1: Figure shows the distribution of ECD abnormal tests in the three groups of patients. Arterious abnormalities are significant more represented in no-Men, venous abnormalities in Men, normal results in BPPV

Conclusions

Meniere's disease is a chronic illness that affects a substantial number of patients every year worldwide. The disease is characterised by intermittent episodes of vertigo lasting from minutes to hours, with fluctuating sensorineural hearing loss, tinnitus, and aural pressure. The primary histopathological correlate is endolymphatic hydrops. Paparella [10] used the notion of "lake-river-pond" to explain the occurrence of malabsorption of endolymph leading to hydrops. This notion describes the endolymphatic sac as a pond, with the vestibular aqueduct (the river) connecting it to the endolymphatic fluid space that is like a lake. When there is an obstruction near the endolymphatic sac or duct, a backlog of endolymphatic fluid is created, leading to hydrops.

The vertebral artery is a major artery in the neck, It branches from the subclavian artery, where it arises from the postero-superior portion of the subclavian artery. It ascends through the foramina of the transverse processes of the sixth cervical vertebrae. Then, it winds behind the superior articular process of the atlas. It enters the cranium through the foramen magnum where it unites with the opposite vertebral artery to form the basilar artery (at the lower border of the pons). At the junction between the medulla oblongata and the pons two vertebral arteries join into the basilar artery, forming the so-called the vertebrobasilar system, which supplies blood to the posterior part of circle of Willis and anastomoses with blood supplied to the anterior part of the circle of Willis from the internal carotid arteries.

The basilar artery ascends in the central gutter (sulcus basilaris) inferior to the pons and divides into the posterior cerebral arteries and the superior cerebellar artery. From the basilar artery arises the anterior inferior cerebellar artery (supplying the superior and inferior aspects of the cerebellum), as well as smaller branches for the supply of the pons (the pontine branches). In under 15% of people the basilar artery gives rise to the labyrinthine artery while, generally, the labyrinthine artery (then subdivide into cochlear and vestibular arteries), is a long slender branch of the anterior inferior cerebellar artery (more or less 85% cases) or basilar artery (<15% cases), arises from near the middle of the artery; it accompanies the vestibulocochlear nerve through the internal acoustic meatus, and is distributed to the internal ear.[11]

The veins of the [vestibule](#) and semicircular canals accompany the arteries, and, receiving those of the [cochlea](#) at the base of the [modiolus](#), unite to form the internal auditory veins (or veins of labyrinth) which end in the posterior part of the [superior petrosal sinus](#) or in the transverse sinus. [12] The common modiolar vein enters the bony channel immediately adjacent to the aqueduct to become the vein of the cochlear aqueduct which in turn drains via the inferior petrous sinus into the Internal Jugular Veins (IJVs). Injury or occlusion of this vessel would be particularly significant since it is widely believed to provide virtually the entire venous drainage of the cochlea. [13] The cochlear aqueduct and the internal auditory canal communicate with the subarachnoidal space; in the guinea pig model, an occlusion of the veins of the cochlear aqueduct results in an increase of perilympatic endolymphatic pressure, a decrease of cochlear blood flow and endolymphatic potential. Furthermore, since many of the venous vessels in the scala tympani have little or no bony covering and are essentially exposed to the perilympatic space, the venous system is a route of entry for the cells participating in the inner ear inflammatory process. [14]

Another interesting point is that the blood leaves the brain by using the back propulsion of the residual arterial pressure (vis a tergo), complemented by antero-

grade respiratory mechanisms (*vis a fronte*). The latter consist of the thoracic pump increased venous outflow during inspiration: the increase of negative thoracic pressure improves the aspiration of blood toward the right atrium. In addition to *vis a tergo* and *vis a fronte*, postural mechanisms play a fundamental role in ensuring a correct cerebral venous return.

The pattern of cerebral venous drainage changes, even under physiological conditions, is thus depending on the body position. In the prone or supine position, the outflow through the IJVs is favoured, whereas passing to the upright position transfers most of the encephalic drainage to the vertebral veins. [15]

In recent years, together with gradual improvement of Duplex machines, interest has been gained in searching anatomical diagnostic areas that appear both critical and challenging, for the operator and the apparatus that is being employed. The vascular anatomy of the cochlea is the most unusual and probably the most difficult to evaluate. The blood flow is segmentally, centrifugally, and centripetally arranged, with many spirally perpendicular vessels shunted in, suggesting good possibilities for variations of circulation. There is a pronounced apical simplification of the vascular pattern probably more than corresponding to the decreasing volume of the turns. The blood supply of the scala vestibuli is mainly arterial; that of the scala media is capillary or by-pass, and that of the scala tympani is venous. For this last reason, and more recently, the Duplex examination is carried out also in an upright sitting position, always kept comfortable for the neck, so to register the difference of the venous outflow according to postural pressure and different breathing activity.

The cochlear artery, often correlated with severe sudden deafness [16,17], appears one of the most difficult vascular exams to carry out. Moreover, this exam isn't easy to transform into a standard path, like for example the most common Duplex exam of the carotid arteries, because the anatomical access is extremely difficult and varies from one patient to another, or even gender since women generally have a smaller structure and window access to the area. The two sides appear very much like a stereophonic Hi-Fi apparatus and so both should register similar bloodstream haemodynamic flow. An important predominance of one side upon the other clearly is abnormal.

The most frequent abnormalities detected with the internal jugular veins along their extra-cranial way. In case of a pathologically slow jugular outflow and secondary venous hypertension [18], other veins have to increase their normal activity of venous drainage and Duplex is the only possible exam, up till now, that allows imaging in lying and sitting positions and also requires patient interaction. The vertebral veins appear to be among those that increase, as far as it is anatomically possible, the venous drainage. Not rarely, also the vertebral outflow appears insufficient and, consequently, venous hypertension can be present also in this segment.

It is now being demonstrated that venous outflow problems are present in several progressive neurologic diseases. Symptoms may vary in percentages but, those regarding postural and/or listening problems seem to be always more frequent in all the diseases. [4]

The echo-colour Doppler (ECD) is the ideal tool for dynamic assessment of both cerebral arterial inflow and venous outflow. In our patients ECD indicated dynamic abnormalities of blood venous cerebro-spinal drainage quite exclusively in Meniere Disease and arterial alterations in no-Men group and normal cerebro-cephalic pattern in BPPV.

On the basis of our results we can suggest that blood outflow could play a pathogenetic role in Meniere Disease [19] while in no-Meniere Disease cochlea-vestibular damage may be mainly sustained by a reduced blood inflow. Substantial normalities of ECD in BPPV confirm that BPPV is a vestibular dysfunction and not a lesion of the inner ear, at least in the majority of the cases.

To the best of the authors' knowledge, the association between Meniere Disease and CCSVI has never been published.

CCSVI might explain the anatomical background which provides a predisposing factor for cochlea-vestibular diseases, in general, and Meniere Disease, particularly. ECD if aimed to detect specific blood flow abnormalities is a specific tool for the pathogenetic differential diagnosis in cochlea-vestibular disorders

Further studies in larger groups of patients are needed to investigate the exact mechanisms and correlation between inner ear diseases and Cerebrospinal Arterio-Venous flow abnormalities.

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