**Neurovascular “compression” of cranial nerves**

Functional microneurosurgery of posterior cranial fossa cranial nerves

**Introduction**

The cranial nerve dysfunction syndromes are related, in most cases, to a "vascular compression" along the nerve, usually at the level of the dorsal root entry zone (DREZ) or exit zone of the nerve from the brainstem.

The compression by a vessel can also be linked to the presence of a vein or to an association of artery and vein. For the past few decades have been developed methods of "micro-neurovascular decompression" (MVD), which allowed the surgeon to detach from the surface of the vessel the compressed and irritated nerve, thus eliminating the pathological contact.

There is therefore a range of symptoms, mostly exuding in adulthood, which can rightly be regarded as exaggerated expression of the normal function of certain cranial nerves.

These are distributed in the cerebellopontine angle, anatomical recess between the pons and cerebellum, located bilaterally in the posterior cranial fossa, close to the brainstem.

The cause of these manifestations symptom, along with other additional etiologic factors, it found its proper interpretation in the last three decades, especially after the introduction of the operating microscope and microsurgical techniques. These have confirmed the occasional observation of some neurosurgeons in the first half of the century, which had reported the existence of "abnormal relations" between vessels and nerves during surgical procedures conducted in the cerebellopontine angle for different diseases.

This has allowed the refinement of methodologies to alleviate the "conflict" between a vessel and a nerve, taking care clinical manifestations sometimes disabling.

The most important among these, subject to surgical treatment, are:

- The typical Trigeminal neuralgia ("tic douloureux");
- The abnormal muscular contraction of the hemiface (Hemifacial spasm);
- Tinnitus (noise, ringing in the ears) with or without unilateral progressive hearing loss and vertigo;
- The pharyngeal pain (usually triggered by swallowing), called Glossopharyngeal Neuralgia;
- The disabling positional vertigo (DPV);
- Some cases of neurogenic hypertension;
- The spasmodic torticollis;
- Cyclic oculomotor spasm with paresis.
The concept of neurovascular compression

The unifying element, in terms of the causative factor of all of these clinical manifestations is constituted by the presence of a contact, that will define abnormal for morphological and anatomical position, between the emergence of the nerves from the brainstem and a specific vascular structure, arterial or venous. Typically the "abnormal contact" is configured as a cross compression, that is, as a compression variable angle between the two structures as a compression or "sandwich", namely a nerve entrapment between two different vascular structures.

These are located predominantly at the level of the REZ, the root entry zone, this small portion of the nerve located in the proximity of the brain stem, still held by the central myelin, found within the brain and particularly delicate (oligodendroglia). Myelin is the central nerve fiber coating material, which allows them to properly transmit impulses.

We have to recall that the brainstem consists of the midbrain, the pons, the bulb and part of the medulla oblongata and that from both sides of this structure out (or enter, depending on the nature of sensory or motor) the various cranial nerves of which we will talk.

The authors who have contributed most to the clarification of this entity called "neuro-vascular compression" as a primary cause of specific clinical symptoms depending on the nerve subjected to compression, they were, in chronological order: Walter Dandy in the early '30s, James Gardner in the early 60's, and finally, Peter J. Jannetta in Pittsburgh, which, after the first experiences with Robert Rand conducted in Los Angeles, confirmed, systematizing and reporting, the existence of a neurovascular cause of irritative cranial nerves syndromes.

Was routinely Jannetta to apply the surgical microscope to the treatment of these diseases and is also its merit having a widespread technique approach to the cerebellar-pontine angle perfectly adequate to perform the 'micro-neurovascular decompression' (MVD).

This term refers to the surgical procedure intended to alleviate the abnormal contact between vessel and nerve, with resolution of symptoms of hyperfunction / neural hypofunction.

Many surgeons around the world, including in Italy, confirmed with their case studies and their results as provided by the 'pioneers' of this surgery.

The types of anomalies produced by vessels nerves are typically formed by deflection and angle of their course (bending); footprints left on their surfaces with discoloration of myelin (grooving) splitting of the nerves by arterial loops penetrating inside their thickness, with distortion effect - stretching of the fibers, and more.

The compression of the vessel on the nerve must be a cross pulsatile compression, prevailing at the root entry (or exit zones) of the nerves from the brainstem.
Symptoms of neurovascular conflict

**Typical trigeminal neuralgia** or 'tic douloureux'.
The incidence per year is 3.4 cases per 100,000 men and 5.9 per 100,000 women.
The age of onset is usually after the fourth decade of life. The pain is most frequent on the right side of the face, especially in women, and affects the middle or lower face, the less often the upper part around the eye.
The patients affected by this event experience pain perhaps one of the most devastating that nature can produce: Attacks usually last very little, can be repeated several times in the day, even at night, are characterized by a throbbing pain, such as stab or electric shock. The intensity of the pain is extreme.
The subject, in time, it becomes absolutely conditioned by his pain, do not feed more, do not care of the cleaning of the face, until to become a human larva. This is because there are skin staples, said trigger zones, whose contact or even slight stimulation triggers the pain. The trigger zones are located around the nose, between the nose and upper lip, at the level of the gingiva. Even a breath of air, the contact of the face with water of different temperature, chewing, talking, swallowing, sneezing can trigger a bout of excruciating pain.

After the crisis, or the pain disappears completely or it may be a milder algia, deep and disturbing. The patients live in constant fear of hitting a new ruinous crisis of pain. Medical treatment is usually effective in the early onset of the crisis, it may not control them on an ongoing basis.

The surgery arises at this point as the only possible alternative: Various methods have been proposed to alleviate or cure trigeminal neuralgia, their list would go beyond our discussion. With the microsurgical operation of "micro-neuro-vascular decompression" (MVD) it is usually possible to eliminate the causative agent of the problem and the effect of "compression button" on the nerve. The postoperative result is excellent in over than 95% of cases of the "typical" form, the so-called "tic douloureux".

Is relevant to remember that there are atypical forms of trigeminal neuralgia caused by acoustic neuroma, meningioma or other processes expansive own or seat close to the trigeminal nerve and that a low percentage of neuralgia is given by multiple sclerosis or other inflammatory reasons. In these cases, evidently, it is not possible to consider the neuro-vascular conflict as before described.
In large case studies provided by Peter Jannetta relapses are possible: early after surgery, because the vessel causing the cross compression have not unpacked and rightly moved from the nerve, or later, at a distance of more than 10-15 years. These are likely to be induced by a reorganization of the neuro-vascular relations or dislocation of Teflon. Treatment of Trigeminal Neuralgia, by MVD, offers the best guarantee of resolution of symptoms, compared to other forms of surgical treatment that can produce the so-called painful dysesthesia, linked to the destruction of the trigeminal fibers. Surgical methods alternative to MVD in the posterior fossa exist and have been described by numerous sources. Anyway,
MVD, that is the "micro-vascular decompression" in the posterior fossa, if properly conducted, decompress and respects the sensory functions of the trigeminal nerve, including that of the cornea.

**Hemifacial Spasm (HFS)**

This disorder has an incidence of 0.74 per 100,000 males and 0.81 per 100,000 females per year (28). In its typical variant, is characterized by involuntary spasmodic contractions onset from the muscles surrounding the eye, which then, in time, gain the lower muscle territories of the half of the face, until the platysma which extends in the neck.

In the advanced stages of the disease, the spasms alternating with tonic contraction of the muscles, with obvious deformation of the hemiface, of variable duration.

In the atypical variant, muscle spasm starts from lower hemiface and extends later to the eye, but this condition is much more rare. With meticulous intraoperative microsurgical observation, also in this case is possible always identify a vessel compressing the nerve.

Given that the hemifacial spasm is a debilitating problem for the patient, with a serious psychological effect on his social life, MVD is the only one suited to achieve the resolution, sometime instant, of the disorder.

For the MVD surgery of Hemifacial Spasm, the retrosigmoid craniotomy must be performed lower than that used for the trigeminal nerve, but for the rest of the neurosurgical technique is the same.

Even hemifacial spasm, which prefers a younger age of onset than the trigeminal neuralgia, is more often observed in females and on the left side of the face.

During the microsurgical decompression, the risk of producing hearing damage is higher, given the proximity of the emergency of the cochlear nerve. For this reason, more and more frequently is used the intraoperative auditory monitoring (BAER). This provides immediate information about surgical maneuvers at risk for hearing loss. Even in the case of Hemifacial Spasm a complete resolution of symptoms occurs in 95% of cases. Nerve palsy as a complication of the intervention is less than 0.25%.

**Tinnitus** (noises, ear ringing) with or without the vertigo and progressive unilateral hearing loss.

When a posterior cranial fossa tumor has been excluded as a possible cause, also these symptoms have to be considered "hyperfunction" of the cochlear and vestibular nerves. The central myelin is represented by a long stretch in the course of the vestibular-acoustic bundle and for this reason vessels in conflict can occur not only at the level of the REZ. When it is not generated by a disease of the inner ear or of the hair cells inside, tinnitus can be determined by vascular compression along the entire course of the nerve and not just the portion closer to the brainstem. The vertigo is most often caused by situations of iuxtacentine conflict, on the vestibular nerve. Frequently, together with vertigo, imbalance disorders are present. If there is only imbalance without true vertigo, the vessel will be found on the vestibular division of the eighth cranial nerve just at the level of the REZ. The cases that are successfully treated by a MVD are associated
with abnormalities in the brainstem auditory evoked potentials on the same side of the symptoms. At this level, compressions by veins are found more easily than arteries, such as the structures responsible for the conflict. In surgical procedures performed for relieving the tinnitus alone, this can not be resolved completely after surgery, but patients may also benefit from a reduction in symptoms, more or less marked compared with the preoperative status. Remission of imbalance and dizziness, even after a long time after surgery (weeks to months) may be expected in 90% of patients, the tinnitus (even after several months after surgery) in 60% of cases.

**Neuralgia of the glossopharyngeal nerve**

It is a violent pain in poussees, which occurs in the tonsillar region, triggered by swallowing and often accompanied by throbbing pain in the throat and pain deep in the ear. This clinical entity is a symptomatic indication for neurovascular decompression of the ninth cranial nerve (glossopharyngeal). The exposure of the nerve occurs in the same way as described for the facial nerve in the course of hemifacial spasm. Here also is used to unpack or move the arteries in contact with the nerve and put some Teflon in between. Working on the left side, there may be a cardiovascular response in arterial hypertension, which should be monitored and treated.

**Disabling positional vertigo (DPV)**

This syndrome has been identified recently by the group of Moller and Jannetta in Pittsburgh. It consists, from the point of view of symptoms, of the following characteristics: vertigo or severe disequilibrium, almost continuous (therefore, absence of periods of paroxysmal symptoms and absence of hearing fluctuation as occurs in Meniere’s disease); vertigo and/or disequilibrium triggered by any movement of the head and not subject to “fatigue”; ineffectiveness of vestibular suppressants, drugs used in the treatment of other forms, vertigo can be objective, worsened by certain head movements or body positions; feeling sway, to be "in boat" (the whole can improve during movement), difficult gait with wide basis, uncertainty and unsteadiness; patients have an almost constant feeling of nausea and vomiting during movement; they present tinnitus, with pain, occasionally short and small muscle twitches around the eye. This Syndrome, so special, but definite, has been and is treated by MVD of the cochlear nerve. In some cases it was noticed intraoperatively as the vessel causing an indentation, that is a groove, on the surface of the nerve, especially on the vestibular one. The healing and disappearance of symptoms occurs gradually over time, faster for vestibular symptoms than tinnitus.

**Neurogenic hypertension**

There are cases of so-called essential neurogenic hypertension, caused by a vascular loop of the vertebral artery or basilar artery creating a real impression on the underlying nerve at the level of the left dimple side of the bulb. The vascular compression causes a dysregulation of the neural mechanisms that maintain the balance of tone sistemi arterial pressure, causing a facilitation of
pressure and inotropic centers (ie that provide contractile force) of the heart. After MVD and liberation of the nerve by mechanical pulsatile and compressive effect, the blood pressure values are reported in the standard gradually. The first experiments conducted by Peter Jannetta, have been proven by experimental models (only the dimple left side of the bulb causes this effect) and have been confirmed by other authors.

**Spasmodic torticollis**

Among the most rare events resulting from vascular compression of the root entry zone of a cranial nerve is the "spasmodic torticollis". This event, when it does not fall within the broader context of dystonia from a disease of the basal ganglia, can be favorably treated by neurovascular decompression of the eleventh cranial nerve (accessory nerve) at the level of its emergency from the medulla oblongata, with a marked reduction or resolution of this debilitating psychologically and physically problem, within three years after surgery.

**Oculomotor paresis with cyclic spasm**

The pathophysiology of cyclic oculomotor spasm with paresis is extremely rare and it is similar to hemifacial spasm. It is probably due to compression of a portion of the oculomotor nerve, while turns out to be, at the same time, a functional disorder of the central nucleus.

**The microsurgical procedure of decompression (MVD)**

The microsurgical vascular decompression is performed under general anesthesia, syarting with an incision behind the ear, on the side of the pain (in trigeminal neuralgia cases) and the removal of a small piece of cranial bone (minimal craniotomy, about 3x3 cm), exposure and opening of the dura mater, exposure of the cerebellopontine angle in the direction of the trigeminal nerve (supracerebellar type approach). Recognized and exposed the nerve, which can also be hidden by a major venous structure called Dandy petrosal vein, we proceed to explore the course and to identify the vessel (usually the superior cerebellar artery) and with one or more of its looping branches, throbbing, distorts its surface and the regular course of the nerve. The loop is gently detached from the nerve with high-magnification microsurgical instruments, without exerting traction on the branches which are detached from it and maybe supplying the brainstem. At this point we introduce one or more layers of Teflon, of adequate size, with the intent to permanently detach the contact and isolating the surface of the nerve from the artery. Sometimes you can retract the vessel with Teflon or silk strings, which are then anchored to the dura mater (the tentorium in general) with biological sealants. Before closing the operative field it is important to check once more, in order not to miss out on other vessels, artery or vein, which contribute significantly, with the main vessel, in compressing the nerve and in altering its structure and function. The operative technique described is used not only in the treatment of trigeminal neuralgia, but also for the treatment of all other syndromes in which one or more vessels compress a nerve in the posterior fossa.
The most impressive results are obtained after the decompression of the trigeminal nerve in case of typical neuralgia (tic doloureux), of the facial nerve in case of hemifacial spasm, of vestibular-cochlear nerve in cases of disabling positional vertigo with or without tinnitus, and in cases of neuralgia of the glossopharyngeal nerve.

Concluding Remarks

The concept of neurovascular decompression is nowadays well established and is presented as a method applicable to the treatment of many syndromes of hyperfunction of the cranial nerves in the posterior fossa. Recent researches point out that, in conjunction with the event of mechanical and anatomical vascular compression on the nerve, there must be, at least in certain cases of hemifacial spasm and of trigeminal neuralgia, also a facilitation of the central nuclei of the nerves involved. The facilitation of these nuclei, through a mechanism of "kindling" (facilitation) would occur as a result of the presence of one or more vessels compressing a nerve, as a functional reorganization of the the nucleus, from which the nerve takes origin or in which it is contact. The third factor that still escapes to an identification of the nature is the so called suitable substrate, a factor congenital or acquired over time, that makes it possible in a given individual the realization of those mechanisms which, given the presence of a conflict neurovascular, influence by a retrograde way the corresponding nucleus. In the nucleus would occur an anatomical and functional reorganization able to trigger (like a "epileptic crisis" and that is to trigger abnormal and uncontrolled impulses) hyperactive symptoms attributed to the nerve who has contact or origin with/by this nucleus.