The Mystery of the Vestibular Labyrinth, a Family Passion

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In memory of André Montandon (1905–1983), pioneer in vestibular investigation

The field of craniofacial surgery, as it is usually defined, deals with the skull, facial bones, and soft tissue disorders of the surrounding structures. There is 1 part of the cranium left apart in this type of surgery, the petrous bone, which is almost dealt with exclusively by ear specialist. Every physician has learned, during his studies, the anatomy and physiology of the inner ear: the complex mechanism of the transmission of sound to the brain, the intricacies of the vestibular labyrinth, which is the key to our equilibrium. It is, however, very unusual for 2 brothers to have been introduced during their adolescent years to notions like nystagmus, rotating stools, Menière disease, and Barany test. And what an exceptional experience for the 2 boys to become witnesses of the venue of a Russian delegation in their father's office in 1961, coming to evaluate the possible problem of equilibrium of future cosmonauts before the first spatial flight of Yuri Gagarin. This early exposure to the field of equilibrium and vertigo has certainly played a role in the final orientation of André Montandon's sons: Pierre became an ENT, specialized in ear microsurgery, Denys, preferring a more macroscopic field, became a plastic surgeon specialized in craniofacial surgery.

The role of the ear canal as a vector for the transmission of the sounds was recognized since Antiquity. The physician, poet, and philosopher Empedocles (490–430 BC) was a precursor in the elaboration of a physiology of the senses: vision, audition, olfaction, taste, and touch. For the hearing, according to his animist philosophy where everything had a soul, the pulsating air (the sounds) introduced inside the ear rings small bell (ὠ) stimulating a fleshy branch (ἀ ὀ), [the cochlea?] corresponding to an equivalent function of our soul, to ensure the perception of the sounds. For Galen, 500 hundred years later, The Great Designer who made our body, placed a very hard and solid bone in front of the nerves of hearing, preceded by an oblique foramen and a conduct in the form of a labyrinth full of detours to lessen the impact of the cold air, and prevent anything to enter into the brain.

Since the Renaissance, several anatomists have studied more carefully these small organs located inside the petrous bone. Interestingly, the first to describe briefly the malleus and the incus and to note their theoretical association with the transmission of sound was a surgeon, Jacopo Berengario da Capri (1460–1530), well known for his publications and lectures on anatomy and surgery at the university of Bologna. His *Commentaria supra Anatomiam Mundini*, published in 1521, constitutes the first example of an illustrated anatomic textbook ever printed. Although the management of cranial fractures had been described before by several authors, the *De Fractura Calvae sive Cranei* (On Fracture of the Calvaria or Cranium), published in 1518, is the first printed treatise devoted to head injuries with several illustrations, including an entire surgical kit for performing cranial operations, and a step-by-step procedure of craniotomy for the management of skull fractures along with the sequential use of the previously presented instruments[1] (Fig. 1). Another Italian anatomist, Gabriele Faloppio (1523–1562) added much to what was known before about the internal ear and described in detail the tympanum and its relations to the osseous ring in which it is situated. He also described minutely the circular and oval windows (*fenestræ*) and their communication with the vestibule and the cochlea. He was the first to point out the connection between the mastoid cells and the middle ear, and naturally the canal which bears his name *(aquaeductus Fallopii),* through which the facial nerve passes after leaving the auditory nerve. Another known anatomist of this period is Bartolomeo Eustachio. The most important treatise of Eustachio in relation to otology is the "Opuscula anatomica," published in 1563. It contains the section *Epistula de auditus organis*, in which Eustachio wrote that he discovered the presence of the stapes before his other colleagues. He provided an unequivocal description of the tensor tympani and established that the chorda tympani is a nerve branch arising from the facial nerve. Eustachio's most known contribution is the description of the structure that holds his name (*The tube of Eustachio*), acting as a valve between the ear and the pharynx. Eustachio also contributed to the study of the semicircular canals and the cochlea.

In spite of these important contributions to the anatomy and physiology of the ear, the role of the semicircular canals remained a

FIGURE 1. Skull fractures by Berengario da Carpi, 1518.

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mystery and no one would establish the relation with the equilibrium. The Roman philosopher Seneca (4 BC–65 AC) gives a riveting description of seasickness in one of his letters."I was suffering the torments of that sluggish brand of seasickness that will not bring one relief, the kind that upset the stomach without clearing it." He added as a joke: "You can take it from me that the reason Ulysses got himself wrecked everywhere was not so much because Neptune was against him from the day he was born, but because he was given to seasickness like me—it'll take me twenty years to reach my destination, too, if I ever have to journey anywhere by sea!" Of course, Seneca never made the link between seasickness and the inner ear, preferring to attribute it to the stomach.

In 1683, the French Joseph-Guichard Duverney wrote a whole treaty on hearing and the anatomy of the ear, *Traité de l'organe de l'ouïe*, where he attributes to the semicircular canals a function similar to the cochlea, for a better hearing. The Napolitan Domenico Felice Antonio Cotugno was only 24 years old when he described the labyrinth, the spiral lamina, the modiolus and the endolymphatic sac in his famous dissertation *De Aquaeductibus auris humanae internae* (On the Aqueducts of the Human Internal Ear) published in 1760. This treatise is a masterpiece of 80 pages that describes the inner ear and its various fluid-filled spaces. Significantly, Cotugno is also recognized as the discoverer of the permanent presence of labyrinthine fluid in the various cavities of the inner ear and the existence of 2 aqueducts: the cochlear and the vestibular: "These things make me think that I am not misusing the term aqueduct when I apply it to the canals, which themselves reveal the shape and function of the term. There is, indeed a path that begins in the vestibule and passes through the petrous bone, which at a certain moment fills with humor from the cavity and carries it to the lateral sinus of the dura; a path that I call the vestibular aqueduct (aqueductus vestibuli)." As his predecessors, Cotugno could not explain the function of the vestibule and the semicircular canals.

Jean Flourens who was a professor of comparative anatomy in Paris published in 1824 his experimental results on pigeon semicircular canals: "If the membranous ducts are injured, a painful sensitivity to tones is observed, accompanied by abrupt and violent movements of the head... If the horizontal canals are severed, the animal turns on its vertical axis; if the posterior vertical canal is severed the animal rolls over backward, and if the anterior vertical canal is severed the animal falls forward. . .." Flourens concluded that the semicircular canals influenced the direction of motion, rather than playing a role in balance. His work had been largely ignored but was known to the French otologist Prosper Menière and acknowledged in his final paper in 1861, which made definitely the link between the equilibrium and the semi-circular canals.[2] Commenting his observations on cases of severe deafness associated with vertigo, he wrote: "These functional problems located in the inner ear may be linked to accidents attributed to the brain, such as vertigo, dizziness, uneven walking, spin, and falling sensation; in addition they are accompanied by nausea, emesis and syncopal condition. . . There are good grounds to believe that the material lesion of these functional problems is located in the semi-circular canals."

Present Knowledge of the Vestibular Labyrinth, Our Sixth Sense

The vestibular system is the organ of equilibrium and spatial orientation. This sense organ, also called the Vestibular Labyrinth, is located in the temporal bone in continuity with the cochlea, which is the sense organ of hearing and they share fluids called perilymph and endolymph. The vestibular system consists of 3 semicircular canals and 2 otolithic organs, the utricle and the saccule (Fig. 2).

All movements of the head cause slight displacements of the endolymph and the result of these movements is a slight flexion of

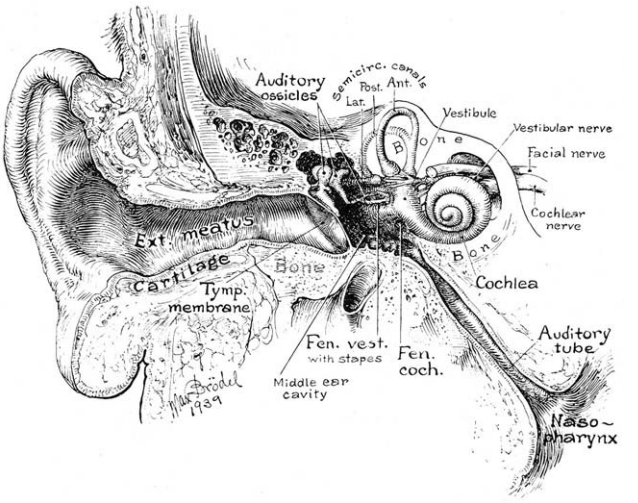

FIGURE 2. The hearing system by Max Brödel, 1939.

the little hairs located at the top of the sensory cells. This changes the rhythm of the electrical discharges transmitted to the brain by the vestibular nerve. This mechanism is very similar to what happens to the cochlea in response to sounds. Any movement of the head causes the vestibular system to correct the position of the ocular globes to avoid that the eyes follow the head movements instead of keeping a stable axis of vision. In other words, the gaze of the eyes remains independent of the head movement with a normal functioning vestibular system. Contrarily, in a malfunctioning vestibular system, the ocular globes tend to move in synchrony with the head causing oscillopsia.

Symptoms elicited by alteration of the vestibular function tend to be perceived as a horrifying brain disturbance. Vestibular disorders can cause a multitude of symptoms including vertigo, spatial disorientation, and motion sickness, including also nausea and vomiting as in seasickness.

Vertigo is a frequent motive for consultation with general practitioners and referral to neuro-otologists. Lesions can be diverse: inflammatory, mechanical, traumatic, tumoral, or chemical. Neuro-otologists use sophisticated tools to establish a precise diagnosis and choose a proper treatment. Treatments include a broad range of procedures: physiotherapy, medications such antihistamines, corticosteroids, and antiviral. Surgical procedures can be conservative or ablative such as decompression and drainage, total labyrinthectomies or vestibular nerve section.

The Geneva Otology School

In 1945, John Erath, one of the first ENT specialists practicing in Geneva, reported the results of his 30 years' experience with an original method that he used on a number of patients suffering of tympanic perforations. His nephew, André Montandon, undoubtedly oriented to this field of medicine by his uncle, completed his training in otorhinolaryngology in Geneva, Paris, and the USA, and was appointed professor and head of the ENT Department at the University Hospital of Geneva from 1947 to 1974. Among his main interests, he developed the surgery for tumors of the larynx and described a special cannula that bears his name and is still used routinely in Europe for anesthesia during total laryngectomy. Microsurgery of the middle ear was his surgical field of predilection, and his main research interest was concentrated on the vestibular system. Among his multiple publications he wrote already in 1956 a manual on the functional tests of the inner ear,

which entered the bookshelves of most French speaking ENT specialists.[3]

Trained in Basel, Zurich, and Boston, his son Pierre Montandon was promoted Staff Surgeon and Assistant Professor at Harvard Medical School, before being solicited to come to Geneva in replacement of his father to take over the Geneva ENT Department in 1975, a position that he hold until 2000. Pierre's main interests and achievements concentrated on the research and development for better understanding and treating hearing and vestibular disorders. His most outstanding contributions concern experimental and clinical recordings of cochlear potentials,[4] functional surgery of the middle ear,[5] the elaboration and introduction of cochlear implants, [6] and treatment of Menière disease with minimally invasive and conservative procedures.[7,8] These topics have been the subject of a number of publications in international journals and book chapters. Reviewing these articles, one is stricken by the fact that most of them emanate not only by the senior author, a physician, but also by a team of other scientists, engineers, physicists, and specialized technicians. This collaboration denotes the complexity of the research in the field of hearing and equilibrium, which can only be apprehended by a local or international team mastering different domains.

Professor Jean-Philippe Guyot succeeded later to Pierre Montandon in 2004 and pursued the research in the field of what he likes to call *the sixth sense* that is the vestibular system and its relationship to the brain functions. As he explains it for the general practitioners:[9] "If one can maintain a position or the stability of the vision on an object and orient oneself in space, it's because this sense is intact. It is when this vestibular system is impaired that one realizes that it merits the appellation of the sixth sense." "We don't stay in equilibrium, we just fight against falling." To manage a critical situation, our brain takes into account several parameters: proprioception, central and peripheral vision and the vestibular system of the inner ear. When all these information are concordant, we feel all right; if not we might feel very bad. Fortunately, our brain can adapt and manage a few discordances; otherwise, we could never hold on a boat without being very sick. Vertigo is very variable, which makes it difficult to diagnose, and difficult for the patients to describe their troubles. Consequently, patients may consult several GP or even ENT specialists before having an accurate diagnose. Medical or surgical treatments are not always very successful. It is however possible to correct a few troubles thanks to vestibular physical therapy and play on the capacities of adaptation of the central nervous system.

New Discoveries: Link With Dyslexia and Neuro-Prosthesis

Several studies have suggested a link between dyslexia and vertigo, leading to possible therapeutic consequences. A Finnish research identified benign paroxysmal positional vertigo causing dyslexia through increasing the amount of involuntary rapid eye movement, making it difficult for patients to focus on a word prior to treatment. Treating the vertigo reduced the involuntary eye movement, easing reading. A few Finnish schools are in the process of testing vertigo treatment to help dyslexic children.

Jean-Philippe Guyot and his team in Geneva, in collaboration with Boston at MIT and Maastricht in Holland, have recently developed a vestibular implant in the treatment of vestibular function impairment. Based on devices similar to the captors, which change the screen's direction of the smartphones, this prosthesis may be implanted like a cochlear implant. It provides electrical stimulation directly to the vestibular nerve with parameters that reproduce what the labyrinth normally transmits to the brain (Fig. 3). This vestibular implant has been already successfully implanted since 2007 on a number of patients who were incapacitated by total and bilateral loss of vestibular function without compensation by the central nervous system.

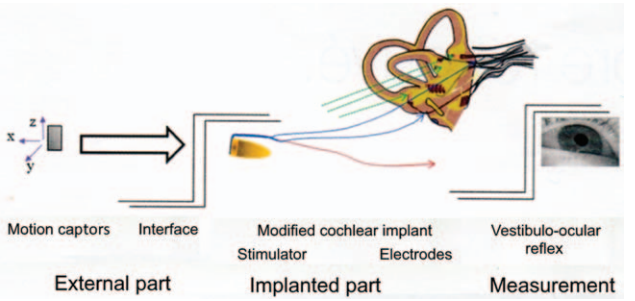

FIGURE 3. Vestibular implant. Patent Geneva University Hospitals. (European patent application EP13153300.2. 2013-01-30).

CONCLUSION

The collector of fluid-filled tubes and chambers, which contain the receptors for the senses of equilibrium and hearing, is often called the labyrinth, because of its maze-like structural anatomy. In Greek mythology, the Labyrinth was an elaborate, confusing structure designed and built by the legendary artificer Daedalus for King Minos of Crete at Knossos. Its function was to hold the Minotaur, the monster eventually killed by the hero Theseus. If the anatomical structure of the vestibular labyrinth is complex, its function and physiopathology are even more complex and confusing. No wonder, it is only since the last decades that neuro-otologists have started to apprehend the mechanism of this sixth sense, which concerns every one of us and is particularly debilitating in case of dysfunction. The research in this field is now in full expansion. With the development of spatial flights, the vestibular equilibrium of cosmonauts in microgravity and many other neuro-otologic reactions are still to be discovered.

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