Selective Canal Impairment on Video-HIT in Peripheral Vestibular Diseases

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- Welcome everybody, and thank you very much for being with us even today. My name is Anna and I'm the Education Manager here in Inventis. Welcome to this first appointment of the year with our webinar campaign on balance campaign, which is part of the Inventis project called Academia. In collaboration with the Vestibology Italian Society, directed by Professor Augusto Pietro Casani, professor at the Pisa University who is with us today, we announce the webinar "Selective Canal Impairment on video-HIT in Peripheral Vestibular Diseases." The webinar is led by an excellent doctor, Dr. Andrea Castellucci, ENT consultant at the Santa Maria Nuova Hospital in Reggio Emilia, Italy. Thank you very much, Professor Casani and Andrea, for accepting our invitation.

It is always much appreciated. So your contribution is fundamental to expand the borders of academia. For this reason, after the event, you will receive a survey I kindly ask you to fill out. The survey will help us to understand your needs, to get your ideas in order to provide the best educational support ever. And now I leave the word to Professor Casani first and to Dr. Castellucci immediately after. Enjoy the webinar. Bye for now.

- Oh, thank you, Anna, and thank you, Andrea. Only few words to introduce Andrea Castellucci. Dear colleagues, for me, is an honor, is a great pleasure to introduce Andrea Castellucci. Andrea is a young, but already established expert in otoneurological field, but presenting Andrea, I cannot forget and express a pleasant memory of his teacher, Professor Giovanni Modugno, professor of otolaryngology at the University of Bologna. Giovanni Modugno was a great otoneurologist and I think that Castellucci is the continuous doctor of his research. And this webinar is organized and sponsored by Inventis, but the organization involved also the Italian Vestibology Society. And thanks to the manager of Inventis Academia, Anna Scala, I would like to express a thanks for her competence and availability.

Only few words about the Italian Vestibology Society. This society was born in 2013 and this is the first scientific association of Italy that brings together expert and scholars on function and disorder of the balance. The Italian Vestibology Society aims to involve all those interested in basic science and in clinical and application sciences concerning the vestibular system and its representation in the central nervous system. And so what our aim is to increase the knowledge about the vertigo and the imbalance as well related system and the symptoms. But another important purpose of the Italian Vestibology Society is the contribute to train new professionals and to evolve our specialty involving the vestibular system as a priority. But we would like to disseminate and validate current and up-to-date scientific information, both professional and both the patients suffering from problems related to disorder of the vestibular system.

And finally, this webinar deal with a very important topic, the selective canal impairment on video-HIT in peripheral vestibular disease, and Dr. Castellucci is surely one of the most expert about this field. So this webinar finally intend to promote an interface with other national and international physician to make known to an international audience the point of view of one of the most eminent Italian colleague, Andrea Castellucci, regarding this topic that surely is a very, very interest for our knowledge about the vestibular system. Please, Andrea, you can start with your speech. Thank you very much to all the colleagues that are involved in this webinar.

- Thank you, thank you very much, Professor Casani. It's a honor for me to be here. Good evening to everyone or good morning to everyone, depending on where you are. And firstly, I would like to thank Inventis and Anna Scala for Inventis for the kind invitation. I'm happy to be here to present this talk. And of course I would like to thank Professor Casani, the president of the Italian Society of Vestibular Disorder, VIS. I'm proud to be part of the directive board of this society and I'm sure that with this new deal, we're going to spread the knowledge, at least, the Italian knowledge on the vestibular science. And so I really would like to thank Professor Casani for his efforts in

encouraging his colleagues, Italian colleagues to spread the knowledge in the vestibular disorders in the vestibular field.

So as Professor Casani has already mentioned, I always need to thank my mentor, Professor Giovanni Carlo Modugno, he was my mentor in Bologna when I used to be a student and resident and he was a great scientist and he told me everything about vestibular disorder. He was the first one who introduced me in this kind of field. So every talk that I give at the beginning, I need to thank him because it's because of him that I'm here, it's thanks to him that I'm here talking with you. And secondly, I really like to thank my close colleagues Pasquale Malara from Switzerland, actually he's Italian, but he works in Switzerland, and Salvatore Martelluci from Latina, Italy. They are close colleague of mine who we are working together very close, and we're doing some research together.

And this talk should be co-authored by them because actually it's a series of patients. It's our experience in selective canal loss in video head impulse test in vestibular disorder. So this kind of talk should be co-authored by them. And I also would like to thank Enrico Armato from Italy and I would like to thank him for his efforts in spreading knowledge about video head impulse test in last days, the French edition of his book on video head impulse test has been released, so I really like to congratulate with him because of his great efforts on this. So let's go with the introduction of this topic. So we'll speak about selective lesion at the video head impulse test and we'll try to see, we'll try to see which are the disease that lie behind a selective lesion of video head impulse test.

We'll try to match each selective lesion of the video head impulse test with the rest of the vestibular assessments, to try to help clinicians that face selective lesion on the video head impulse test to try to help them to understand what's going on in this kind of patients. And of course, this is my experience, this is our experience, my experience

and Pasquale and Salvatore's experience. So it would be nice if at the end of this talk, we have some time for discussion. Probably not all the disease that I'm introducing you reflect the same experience of other people from the audience, so I really would like to make some discussion at the end and I would like to encourage you to making questions and comments on our experience.

Of course, I left something away from this talk. So I beg your pardon if I forgot something in this kind of talk. Before understanding which are the disease behind a selective lesion of the video head impulse test, we have to remind anatomy of inner ear. We have to remember this is a very famous picture taken from the literature. We have to remember that the inner ear is made by three ampullary receptors and two otolith receptors, okay, the utricle and the saccule. And we have to remember that the innervation is given by two branches of the vestibular nerve, so the superior branch and inferior branch. So since the beginning, since from this picture, we can understand there are two difference.

There are two different part of the labyrinth, a superior part of the labyrinth that involves anterior and superior canal efferents and utriclar efferents because they run through the superior vestibular nerve, and the inferior part of the labyrinth that involves the posterior canal and the saccule. So from this picture, we can see from the start, we can see that the posterior canal, it's a little bit separated from the other canals, from the innervation, okay? And the same kind of differentiation between superior labyrinth and inferior labyrinth is also given by the arterial supply of the inner ear. We know that there are two main arterial supply, two main arterial branches that branch from the internal auditory artery. The superior part of the labyrinth is supplied by the anterior vestibular artery.

So the anterior vestibular artery supplies mainly the horizontal, the superior canal, and most of the utricle, probably something also goes to the saccule, but more or less, it

supplies the same receptors that are innervated by the superior vestibular nerve. While the common cochlear artery supplies mostly the saccule, a small part of the utricle probably, and the posterior canal, so the same sensors that are innervated by the inferior part, the inferior branch of the vestibular nerve. But with a big difference, the common cochlear artery goes all the way also to all the cochlear parts. So this is one of the main difference between innervation and vascular supply. And this will be very important where we'll face lesions that involve a selective canal, selectively a canal, and also the cochlea.

And also from this differentiation between superior and inferior part of the labyrinthine, depending on the vascularization, we can see that again the posterior canal is separated from the other canals. So from this preface, we can already understand how the posterior canal will be selectively lesioned much more often compared to the other canals because innervation is separated from horizontal and superior canal, and also the vascularization is separated from horizontal and superior canal. So this is a very important preface. Additionally, we have to remember that the posterior canal lies in the undermost part of the labyrinthine with the patient both in upright position and also in supine position that posterior canal lies always in the undermost part of the labyrinth.

So diseases related with auditory dislodgements that might lead to selective impairment of a canal, and also hydrops, endolymphatic hydrops due to distension of the endolymphatic space probably will lead more often to involvement of the undermost part of the labyrinthine. Again, posterior canal lies in the undermost part of the inner ear. So posterior canal loss, a selective lesion of the posterior canal in the video head impulse test would be much more frequently detected by clinicians compared to the other canals. So thanks to the introduction of the modern and fast tools for vestibular testing, we know a lot of information on the functional status of peripheral vestibular receptors and afferents, and it's quite easy to have information about these sensors.

We know that cervical VEMPs and ocular VEMPs test the otolith organs and their afferents, cervical VEMPs the saccule, and ocular VEMPs mainly the utricle. And the video head impulse test can give us important groups on the high frequency response for the three semicircular canals, so we can know everything about the five vestibular receptors. And this is a famous picture taken from papers of Professor Curthoys. So we know that we can interpret what's going on in an inner ear if we match cervical and ocular VEMPs with the results of the video head impulse test for the three canals. So combining the results of this test, we can have a more accurate assessment of vestibular function, enabling the identification of specific lesion patterns affecting the inner ear sensors or the vestibular nerve lesions.

So we know that if we have a selective lesion of the horizontal and superior canal and the utricle, we know in an acute phase, in acute patients with probably also horizontal nystagmus, we'll have a superior vestibular neuritis. And if we have posterior canal with the saccule affected, we'll have an inferior vestibular neuritis and so on. And let's say we can play with this kind of results to understand what's going on in the inner ear. So it's very, very, I would say not so very easy, but we are getting very close, we are getting closer and closer to understand what's going on in the patients when we examine the patient, both in acute phase and also in the chronic stage.

So let's go with the lesions, with the disease that can lead to a selective lesion of the horizontal canal at the video head impulse test. So this is bedside head impulse test, this is a horizontal bedside impulse test, We can see corrective saccades after horizontal head impulse, normal on the left side. It was abnormal on the right side, but it would be nice if we try to perform also bedside head impulse test for the vertical canals in the RALP and the RALP plane. You can see here on the slow motion that there are no corrective saccades after head impulse for the vertical canals. I will go back again in this bedside head impulse test also for the vertical canals, it would be

nice that after this talk, if someone is not familiar with this kind of bedside test, if after this talk, clinicians start to perform also bedside head impulse test to detect selective lesion also bedside.

Of course, they will have to confirm their findings also with the video head impulse test, but it would be nice if already from a bedside examination, people could think that probably a selective canal lesion, probably they will get close to the final diagnosis. Looking through the literature, we know that there are also central disorders that can lead to selective lesion of the horizontal canal, Wernicke encephalopathy due to the deficiency of thiamine B1 vitamin is known to lead to a selective lesion of the horizontal canal, but we'll not speak about central disorders even because they're not so common that lead to a selective lesion of the semicircular canal. We'll just speak about peripheral disorders that can lead to selective lesion of the semicircular canal of the video head impulse test.

Looking through the literature, we can see that there are some papers saying that in the acute stage of vestibular neuritis, we can test patients with a selective lesion of the horizontal canal. So they explained this with a partial vestibular neuritis. But actually, if we read these papers, if we go to the results, all of these papers say that patients with a selective lesion of the horizontal canal due to a vestibular neuritis, so an acute unilateral vestibular loss, all of these patients recovers very fast, that horizontal cannot recovers very fast. And this paper from Bela Büki says that when a vestibular neuritis on the same side had really occurred years before, all semicircular canals recovered completely. So probably some of these patients with selective lesion of the horizontal canal, they recovered fast.

Some of these patients already had something similar in the past and it's not that typical for vestibular neuritis. We know that vestibular neuritis in most patients just results in unique episodes that last days, weeks already, also the patient is disabled for

weeks and also for months. And some of these patients recovers, most of these patients do not recover, they just compensate. So this is another interesting paper from Professor Casani and his colleague from Pisa, Elena Navari. And they say that slightly more than this is a nice paper about lesion patterns and possible implication in recovery in acute vestibular neuritis. And they say that more than half of patients seem to be associated with, have developed lesion patterns that are associated with the nerve lesion or lesions of the inner ear, of the sensors innervated by the superior branch of the vestibular nerve or the total vestibular nerve.

And this patient has a worse clinical outcome, while the remaining patients who exhibit selective involvement of vestibular receptor, they have a pattern of spontaneous recovery and the hypothesize that probably, they're due to intralabyrinthine lesion patterns. Probably there are some lesion patterns due to intralabyrinthine mechanism. And I think this is probably one of the most important thing we have to keep in mind when we have selective lesion of receptors in acute vestibular loss. Probably it's not the typical vestibular neuritis, probably it's something different. Let's try to see which is our experience with this patients with the selective lesion of their horizontal canal. Because if we know, if we have in mind this kind of scheme, acute vestibular loss could be mainly due to a sudden functional loss of the vestibular nerve division, usually the superior vestibular branch, generally attributed to a viral reactivation.

Or we can also have a selective lesion of the anterior vestibular artery that supplies the horizontal superior canal and also the utricle, a selective ischemia of a terminal branch of the internal auditory artery. But we should have at least horizontal and superior canal both involved at the same time. And also the usual when in case of unusual combination of instrumental findings in acute settings, we also need to investigate alternative diagnosis compared to acute vestibular loss because probably there are something different from a typical vestibular neuritis or a typical ischemia of the

anterior vestibular artery. So let's go with the first case. This is a lady with an abrupt onset of acute vertigo, typical for an acute vestibular loss.

She doesn't have auditory symptoms, her brain CT scan was normal, neurological examination was unremarkable. We know that brain CT scan doesn't give us a lot of clues, but actually she was normal. Look, she has a spontaneous horizontal nystagmus beating to the left, so we hypothesized right vestibular loss and she had a horizontal bedside head impulse test that was positive on the right side with re-fixation saccades, so it was a typical diagnosis of right acute vestibular loss. But if we look at her instrumental work up in the acute stage, we'll see that, okay, auditory function is okay. Caloric test is difficult for an acute vestibular neuritis. There is a severe right canal paresis on the caloric test, so horizontal canal in low frequencies is damaged, okay.

But if you look at the ocular VEMPs and the cervical VEMPs, we can see that while the cervical VEMPs are symmetrical and it would be okay for a superior vestibular neuritis because okay, the saccule is spared, but look also the ocular VEMPs are symmetrical, so there is no utricle loss in this kind of patient. And look at the video head impulse test, we just have a selective lesion of the horizontal canal, a slight selective lesion of the horizontal canal. So which kind of disease are we facing in this case? Could it be a selective partial neuritis involving just the right horizontal canal ampullary nerve? It might be, but it's not that typical for a vestibular neuritis, or could it be a selective lesion, ischemic lesion of the arterial terminal branch that supplies the horizontal canal?

It could also be, but we need to think something different because they're not very typical. Probably in this case, the patient had a right horizontal canal pseudo-hyporeflexia due to a spontaneous jam of the horizontal canal, inducing a selective plug of the canal lumen. In this case we hypothesized that a clot, an otolith clot, was entrapped in the membranous duct of the right horizontal canal leading to a continuous negative pressure, leading to a continuous bending of the cupula away

from the utricle, so leading to inhibitory stimulus for the horizontal canal, that's why we have a spontaneous horizontal nystagmus. And the cupula is totally bent forward, is continuously bent because there are continuous negative pressure between the cupula and the otolith clots, so the cupula does not respond to low and high frequency stimuli.

So we have a selective lesion of the horizontal canal on the video head impulse test and also a lesion, a reduced response on caloric test. And it was actually a good hypothesis because after repeated head-shaking, look, we transformed this kind of spontaneous horizontal nystagmus in a direction-changing geotropic horizontal nystagmus with paroxysms, of course. So it means that we were facing a jam of the horizontal canal and we transformed this jam in a typical BPPV of the horizontal canal because the jam crumbled and otoliths fell in the non-ampullary arm of the horizontal canal. So we have a geotropic direction-changing horizontal nystagmus with crescendo decrescendo course, so paroxysmal course. And after reposition maneuver, we used the Gufoni maneuver for the horizontal canal, the patient subsided.

And the most interesting thing is that after this maneuver, look, we performed again the caloric test and the caloric test was normal. There was a complete recovery of the right canal paresis and there was also recovery in high frequency for the lateral canal, also the video head impulse test. So we need always to keep in mind that when we have a selective lesion canal, so for the horizontal canal with a spontaneous nystagmus, probably there is a canalith jam. Canalith jam was introduced by John Epley at the end of the 90s. He faced this kind of condition when he was performing kind of a repositioning maneuver and there was a sudden conversion from positional nystagmus to a direction-fixed nystagmus, and he hypothesized that otoliths could aggregate in a clot and remain entrapped somewhere probably in another portion of the canal.

And usually, he described this as a complication of canal reposition maneuver, but we can also face patients with a spontaneous jam. That was BPPV or paroxysmal

nystagmus before coming to our observation. And when we examined the patient in this particular situation, otoliths are entrapped and unjammed somewhere. This is one of the first report of spontaneous jam of the horizontal canal. And in this particular situation, colleagues from Germany noticed this deficit nystagmus, paretic nystagmus. So they hypothesized a continuous negative pressure between the ampulla, between the cupula of the lateral canal and the otolith clot, and it was exactly as our case, but in their case they had a canalith jam as a complication or canal reposition maneuver, so it was more easy for them to hypothesize a canalith jam.

And this is another report hypothesizing the opposite scenario. In this scenario the otolith clot is compressing, is pressing toward the ampulla, so the ampulla is continuously bent toward the utricle. So in this case, there is an irritating nystagmus because utriculopetal flows are excitatory for the horizontal canal. And so both of this scenario could be possible, which we have to keep in mind this. We also have to thank Professor Leonel Luis first, who first reported canalith jam with the video head impulse test with a selective lesion of the video head impulse test. And later on, a lot of colleagues, also Dr Marcelli from Italy with Schubert and other colleagues, Dr. Comacchio from Italy, and also we reported other patients with probably a jam of the horizontal canal.

Look at this interesting case of my colleague, Oswana Malara, a similar patient with an acute vertigo, and nausea and vomiting, so very similar to an acute vestibular loss. He had a slight spontaneous horizontal nystagmus beating to the left, he didn't have auditory symptoms, brain scan was negative, neurologic examination was unremarkable. But in this kind of patient, actually Pasquale detected a positive bedside horizontal head impulse test toward the left, toward the same side where the nystagmus was beating. So it's not very typical for an acute vestibular loss, it's opposite actually, so which is the diagnosis? Could it be something peripheral, could it be something central? Probably. Let's have a look, let's have a look. Let's try to use the

instrumental test we have in our hands to understand what's going on in this kind of patient.

So cervical and ocular VEMPs were totally symmetrical. And again there was a selective lesion just for the left horizontal canal. This is the over canal round and this was just a selective lesion on the left side. So probably in this case, there was a left pseudo-hyporeflexia due to a spontaneous jam that was pressing in this case in the right horizontal canal sorry, in the left horizontal canal that was pressing toward the utricle, it was pressing the cupula of the left horizontal canal toward the utricle. So we had an irritating nystagmus, let's say, at the same time paresis, apparent paresis for that canal because the cupula is continuously bent and it does not respond to high frequency inputs that are the head impulses.

So that's why we had a paresis, a selective paresis of the left horizontal canal. And he also tried to think about this scenario and he's now doing some maneuvers to try to make that clot crumble after this maneuver. Look, nystagmus changes, nystagmus changes depending of the head position of the patient. So look now there is a huge right beating nystagmus. So probably the otoliths crumbled and went in the ampullary arm of the left horizontal canal. And the interesting thing is that the instrumental workup, actually the video head impulse test after this maneuver resulted in a total functional recover of the left horizontal canal of the video head impulse test. Another case of a patient, typical case of acute vestibular loss, 64-year-old woman with abrupt onset of vertigo, she didn't have any vestibular symptoms previously and she just had her arterial hypertension, CT scan was normal, neurologic examination was normal.

I can say that subsequently, she performed brain MRI and it was totally normal, and bedside head impulse test horizontal was positive on the right. So it's a typical clinical feature of right acute unilateral vestibular loss, of course, but even in this case, we just had a selective lesion just of the right horizontal canal, no lesion of the superior canal.

Could be partial neuritis, it could be, but actually when we have selective lesion, we always need to think something else, something else, something else than typical neuritis because usually, superior neuritis involves horizontal and superior canal. And in fact, the day after, we checked her the day after and nystagmus was reversed. Now nystagmus is going toward the right side, and the interesting thing is that the video head impulse test normalized, the reduced VOR gain for the horizontal canal normalized.

So could it be this a neuritis that just last one day, with an irritative nystagmus the day after? Hmm. Or a recover nystagmus the day after? Okay, it could be. But actually the patient developed a typical Meniere's disease on the right side with auditory symptoms and recurrent spells of acute vertigo. And she developed the typical dissociation between low and high frequency for the horizontal canal on the right side, so reduced, extremely reduced response on the right side at the caloric test, and a normal head impulse test on the right side. So it was typical scenario of Meniere's disease. So we always need to think about Meniere's disease when we have a selective lesion of the horizontal canal.

Even if the patient is the first time he has or she has vestibular symptoms, probably he or she will develop a typical symptoms of Meniere's disease and this kind of fluctuation of the VOR gain, it's a new concept, it's a new concept because we all knew that Meniere's disease is a fluctuating disorder. We know that fluctuation of the low frequency in the audiometry is a typical pattern for Meniere's disease. But since the introduction of these tests, this fast new test for the vestibular assessment, we could record fluctuation of the vestibular sensors during attack and in the interictal stage. Also before the use of the video head impulse test, other colleagues using the rotatory chair noticed that during the paretic nystagmus, the VOR gain in the affected side became lower than the intact side.

So they put a side alteration in the dynamic property of the peripheral vestibular system that might correlate with the direction change of spontaneous nystagmus, but probably there is a increased mechanical resistance or a transient hair cell dysfunction probably due to a ionic mixture due to the rupture of membranes that goes to mixed ions inside the endolymphatic space. One of the first report of fluctuation during the acute stage and the interictal stage on Meniere's disease was reported by Professor Manzari with Professor Burgess back in 2011. They noticed a rapid vestibular fluctuation documented with the video head impulse test in Meniere's disease and they hypothesized the intoxication of ionic and chemical elements and a collapse on the endolymphatic space due to membrane rapture during the attack.

And also Professor Yacovino recently published papers of this rapid fluctuation of the horizontal canal during the acute stage of Meniere's disease. So we have in our hands a very important tool to record day by day, I would say hour by hour, what's going on in the patients during Meniere's disease. This is another report with several patients with Meniere's disease started during the acute phase or in the interictal stage. It's a very important significant cohort of patients started by Professor Kim from South Korea, and they found that the impaired gains in the paretic stage usually involves more in the horizontal canal compared to vertical canals. And during the irritative or recovery stage, VOR gains are normal, so there is a restoration of the VOR gain during the interictal stage or the post-acute stage, let's say.

This is another patient studied by my colleague Pasquale. He's a a 35-year-old man with a rapid progressive onset of disequilibrium and nausea. He had the chance to examine him immediately. Symptoms started just two hours before and symptoms were unaffected by change of head position, so it was not a positional vertigo, it was an acute vertigo. No auditory symptoms, and he could record this slight left beating spontaneous nystagmus, and at the bedside head impulse test on the horizontal plane was positive on the left side. And it's quite interesting. It's not very typical for acute

vestibular neuritis because the head impulse test was positive on the same side toward which it's beating, the spontaneous nystagmus. So what is going on?

Is it a central pathology or it could be a jam as we saw before or could be something different? Audiometry was normal, so it doesn't give us important clues. The left horizontal canal was selective lesion as we noticed, but he performed also the cervical and ocular VEMPs. And look, there was an increased amplitude and enhanced amplitudes of the ocular VEMPs on the side where the horizontal canal was impaired, so it's definitely different from an acute vestibular loss. So probably there is something different. And after three days, audiometry was still normal and the video head impulse test normalized for the horizontal canal and also the ocular VEMPs normalized again. So there was something fluctuating, spontaneously fluctuating. And after one week he had a similar attack, and Pasquale had a chance to examine him immediately.

He said, "Come to my office in case you have another attack." Immediately, the patient immediately went to Pasquale and he found the same finding, so probably there is some Meniere's disease going on. And he also had left aural fullness and tinnitus this time. And the audiometry was like this, so typical for the early stage of Meniere's disease and the video head impulse test detected an impairment of the left and the posterior of the horizontal and the posterior canal on the left side, so it's not very typical for an acute vestibular loss in this case, so it's something different. And in fact, it was an acute stage of Meniere's disease, and again, the ocular VEMPs were enhanced on the left side compared to the cervical VEMPs was more or less the same.

And in fact, later on, he performed an MRI with hydrops protocol with late acquisition after four hours from the administration of the gadolinium, And he could detect an endolymphatic hydrops also in brain MRI. So how can we explain this enhancement of utricular response in acute stage? It's something that was already published by Professor Manzari and Professor Curthoys. So an hydropic state of the membranous

labyrinth might lead to enhanced utricular dynamic response probably due to pressure changes of the endolymph. So this fluctuation of macular function during Meniere's disease could be recorded if we had the chance to record, to test the patient in the acute stage. So probably dynamic utricular function in the affected ear is enhanced, and the saccule is not, usually is not similarly affected probably because there are two different sensors, utricle is differently attached to the inner ear respective to the saccule, so probably there is a different activity of the jerk sensitive of type 1 receptors that is enhanced in the utricle and it's not that enhanced in the saccule.

And we can also face this situation, this is a lady, a 61-year-old woman with an acute vertigo typical for an acute vestibular loss on the right side. I had a chance to examine her the following day from the onset of symptoms. Look, there is a spontaneous left beating nystagmus. Neurologic examination was unremarkable. The bedside head impulse test on the horizontal plane was positive on the right side, so it was a typical clinical scenario of a right acute vestibular loss and also mass vibration increased this pattern. So it was a typical diagnosis of right acute vestibular loss and in fact she had both superior and lateral canal loss on the video head impulse test. So we do not have doubts in this case on the diagnosis.

We might hypothesize probably a selective ischemia of the anterior vestibular artery. We cannot know if there was a vestibular neuritis or a selective ischemia. We should perform a hyperventilation test because probably if there was selective neuritis of the superior branch, next time we could reverse as Professor Califano teach us, but we didn't perform hyperventilation test, so we know this is a typical acute vestibular loss due to a superior vestibular neuritis or selective lesion of the anterior vestibular artery. We also confirm the utricular loss with the ocular VEMPs, cervical VEMP was symmetrical, selective lesion of the ocular VEMPs on the right side. And after one week she recovered, she recovered. She was much better, she felt much better.

You see the nystagmus is much more reduced and you can see here that the superior canal perfectly recovered while the horizontal canal did not recover very well. So in post-acute stage after a few days, we can face this kind of scenario with a selective lesion of the horizontal canal, probably due to an asymmetrical recovery, a different recovery between the vertical canal and the horizontal canal. But if we face this kind of pattern in the acute stage, we could not be sure about the vestibular neuritis because vestibular neuritis in the acute stage should always lead to both impairment of the superior canal and the horizontal canal. So in the post-acute stage, we can face a selective lesion of the horizontal canal due to an asymmetrical recovery.

This is a nice paper from colleagues from Switzerland, John Allum from Switzerland, that test patient in the acute stage of acute vestibular loss. And after five weeks, they noticed that the peripheral improvement and central compensatory mechanisms are similar across the three planes, but actually they found difference at the video head impulse test. They found that the posterior canal is the lowest, is the slowest to compensate. So probably there are slight difference of recovery process among canal. So we can detect the patient with just the anterior canal record, but not the horizontal canal. So in the post-acute phase of acute vestibular neuritis, we can face this kind of different impairment of the video head impulse test.

Okay, so we saw the selective lesion of the horizontal canal. Let's go to selective lesion of posterior canal, and it's probably the biggest part of this talk because as we said in the preface of this talk, posterior canal is much more frequently involved in this lesion respective to the other canals. Again, I would like to stress again on the utility of doing the head impulse test also clinically, also bedside, without the use of the instrument to see if there are corrective saccades. Look, this patient had corrective saccade just for the left posterior canal, look this huge downward corrective saccades of their head impulse in the RALP of the left posterior canal. We know that selective lesion of posterior canal may be due to aging.

Usually it's bilateral and symmetric. I will not speak about aging. It's probably due to chronic hypofunction, chronic hypoxia of the lowest part of the labyrinth. It's probably an idiopathic etiology that presents together with progressive sensorineural hearing loss for the high frequency and also with positional downbeat nystagmus, so probably there are both peripheral and central issues that goes together and it's called presbyastasis. We'll not speak about this and we'll not speak also and we'll need to speak about the selective lesion that we can face in case of internuclear ophthalmoplegia due to a lesion of the medial longitudinal fasciculus that serves as the main passage for the high-acceleration VOR for the contralateral posterior canal because it's a central issue.

We'll speak about peripheral disorders that might lead to a selective lesion of the posterior canal. This is one of the most important paper in my opinion from colleague from Switzerland, Alexander Tarnutzer and Professor Weber. This is a cohort of 52 patients with selective lesion of the posterior canal and they perform all this kind of tests in these patient and they found that the most frequent diagnosis behind a selective lesion of the posterior canal was a history of an acute vestibular loss with or without sensorineural hearing loss. And the second more frequent diagnosis was Meniere's disease and also vestibular schwannoma. So of course we can also face a vestibular schwannoma that just involves the inferior vestibular branch, the inferior branch of the vestibular nerve.

And they found a greater involvement of additional receptors, so saccule, utricle, cochlear, in vestibular schwannoma, and in patient with a history of an acute vestibular loss. We published a similar paper a few years later and we found more or less the same, we found that the most frequent diagnosis in our 51 patients with selective lesion of the posterior canal were patient with a history of an acute vestibular loss mostly with sensorineural hearing loss. And we'll see which are the causes of this. And

then Meniere's disease and again vestibular schwannoma. And let's start with some case. This is a patient with acute vestibular symptoms, started two days before with right sensorineural hearing loss, profound sensorineural hearing loss. Actually not profound, but severe sensorineural hearing loss.

He had arterial hypertension, arterial fibrillation, he was in treatment with oral anticoagulants. His neurological examination was unremarkable, but actually he has a slight downbeating nystagmus, I'm not sure if you can appreciate this slight downbeating nystagmus. He had a normal bedside head impulse test, normal horizontal bedside head impulse test in the horizontal plane, but he had this kind of severe right sided flat sensorineural hearing loss, flat or down sloping sensorineural hearing loss. We also performed vibration and with vibration, the slight downbeating nystagmus was increased. So for most of us, this patient should be checked for central pathology, of course, he might have something in the brain stem or in the cerebellum. It's important to always think about central pathology.

Spontaneous downbeating nystagmus should always check for central pathologies. We also had this reinforcement after horizontal head-shaking, so we had that kind of perverted head-shaking nystagmus. But if we check with the video head impulse test of this patient, we noticed that he had the selective lesion of the right posterior canal on the same side of the right sensorineural hearing loss. And with the cervical VEMPs and ocular VEMPs, we noticed that the ocular VEMPs were near symmetrical, probably they were greater on the left side, but the cervical VEMPs were absent. So the saccule is also affected in these patients compared to the left saccule, so probably we are in front of a common cochlear artery ischemia, why?

Because we know that common cochlear artery is one of the maintainal branch of the internal auditory artery that goes to supply the cochlear turns, most of the saccule, the posterior canal, some part of the utricle probably. So in this case, we had this kind of

situation. The patient also had the vascular risk factors, so this will probably the most likely pathomechanism behind this kind of scenario. In fact, if you look at the literature, we can see a lot of series of patient, a lot of investigation of patient with sudden sensorineural hearing loss and vestibular symptoms. And most of the paper go to the same conclusion. Almost half the patient have cochlear and posterior canal selective loss.

This was a paper from 2005 as far as I remember in that period, there was not a video head impulse test, there was a search try, but this is another interesting paper from colleagues from Australia, from Professor Halmagyi, and Professor Miriam Welgampola and colleagues. And they found that patients with vertigo and sudden sensorineural hearing loss, most of these patients have a lesion pattern that reflects the vascular supplies of terminal branches of the interior auditory artery. And in 30% of this patient, they had a lesion pattern that was consistent with a common cochlear artery ischemia, as the patient that I just showed you. We also published this paper of a patient with a clinical picture consistent with a common cochlear artery ischemia.

And we noticed that this patient had a selective filling defect on the left posterior canal on the same side of the hypothetical ischemia of the common cochlear artery. So this was his MRI years before that he performed for other symptoms and this was the MRI that he performed a few months after the onset of this sudden sensorineural hearing loss on the left side and vertigo with a selective lesion of the left posterior canal and a selective lesion on the left saccule. So there was a typical filling defect that is very consistent with an ischemic lesion. So we had, let's say, a confirmation of this kind of vascular mechanism. And this is another lady with similar symptoms, acute vestibular symptoms and right sensorineural hearing loss, but the sensorineural hearing loss in this case was just for the high frequency domain on the right side, and she had a slight horizontal spontaneous nystagmus that was going away from that part, so it was a paretic spontaneous nystagmus, a slight paretic nystagmus on the horizontal plane.

And what's going on here, she didn't have a positive bedside head impulse test on the horizontal canal. So we had a slight spontaneous nystagmus with hearing loss and with a normal head impulse test, and we should think about an ischemia in the territory supplied the ICA artery. So we had to check her for central pathologies. But again, with the vestibular assessment, a complete vestibular assessment, we had the chance to understand that there was a selective lesion in a particular part of the inner ear, a vascular lesion in a particular part of the inner ear because the posterior canal was impaired and the saccule was impaired because cervical VEMPs were impaired, nothing on utricle. So we hypothesized a selective lesion of the vestibulo-cochlear artery because the vestibulo-cochlear artery branches from the common cochlear artery and gives vascular supply to the basal turn of the cochlear, so the high frequency domain of the auditory spectrum in the saccule and the posterior canal again.

So it perfectly matches our findings. This is another patient that presented very similarly to the patients that I showed you before with that downbeating nystagmus, a spontaneous downbeat nystagmus. The torsional component is not very easy to detect, no auditory symptoms, he was a smoker but he didn't have cardiovascular risk factors. And is it central? Of course, the horizontal bedside head impulse test was normal, spontaneous downbeat nystagmus should always be checked for central pathology. We checked him, we tested him with bithermal caloric test, it was normal, so horizontal canal was normal. Cervical VEMPs were impaired on the left side and ocular VEMPs were totally symmetrical, and the posterior canal on the left side was isolately impaired, so probably in this case, we had an inferior vestibular neuritis because we did not have auditory symptoms, we just have vestibular symptoms with a selective lesion of the posterior canal that might lead to a spontaneous downbeat nystagmus because there is a tonic prevalence of the contralesional anterior canal, so there is a downbeat nystagmus.

And we also had the impairment of the saccule on the same side. So in this case, we had an inferior vestibular neuritis. Inferior vestibular neuritis might occur, it's not that common actually. Usually when we have a typical pattern consist with inferior vestibular neuritis, usually we have also auditory symptoms, and in this case, it's more probably an ischemic lesion of those tender branches that I showed before. But it might happen. This is one of the first report on inferior vestibular neuritis by Professor Halmagyi and Professor Karlberg and Curthoys. And they found that this patient with an acute onset of vertigo, typical of an acute vestibular neuritis, they didn't have normal calorics, normal horizontal head impulse, but a selective lesion of the posterior canal and absent cervical VEMPs, so saccule is impaired in these patients.

So those sensors that are innervated by the inferior branch of the vestibular nerve. And this is probably one of the biggest set of patients with inferior vestibular neuritis. This is a paper by Professor Kim from South Korea among more than 700 patients with an acute vestibular loss, nine patients presented with downbeat nystagmus in an acute stage with a selective lesion of the posterior canal and absent air conductor cervical VEMPs on the same side. Among these nine patients, three had a sensorineural hearing loss on the same side, so probably there are lesion of the vascular patterns. They had patterns consisted with vascular lesion, but the other six patient were typical vestibular neuritis. We had another patient that actually behaved very similar to the patient that I just showed you.

Look, this is a patient with an abrupt onset of symptoms of vestibular symptoms with a spontaneous downbeat nystagmus, horizontal bedside head impulse test were obviously normal, no auditory symptoms. Neurological examination was unremarkable, audiometry was normal. And look, there was a slight downbeat nystagmus that was actually increased by skull vibration as you can see here, you can see the slight downbeat nystagmus. It's not very clear because the patient was always blinking. He

was very disappointed by these kind of symptoms. Of course, he needs to be checked for central pathologies because spontaneous downbeat nystagmus should always be checked for central pathology. But we had a chance to test him with our vestibular assessments. And we noticed this severely reduced VOR gain for the left posterior canal.

You can see here the right anterior canal, so the contralateral pair canal is also a little bit reduced, probably there is a loss of the push pull mechanism in this case or probably it was already downregulated by central compensating mechanisms. The cervical VEMPs, sorry, the ocular VEMPs, were totally symmetrical while the cervical VEMPs were impaired in this patient. So it's actually a clinical scenario consistent with acute inferior vestibular neuritis. But we followed up this patient and we performed a brain MRI in one week. He was quite anxious to know if he had a central pathology, but his neurological examination was normal and also brain MRI was actually normal. But he performed months ago because he still had a downbeat nystagmus that was increased by skull vibration.

He performed a CT scan of the temporal bones and we noticed what? We noticed an ossification of the left posterior canal that we found impaired at the beginning in the acute stage. So we reviewed the CT scan that he performed, he performed a brain scan in the acute stage to know if he had something going on in the brain stem or in the cerebellum. And we saw that that posterior canal was actually patent, it was normal in the acute stage. So he developed an ossification of the posterior canal on that side. So probably we had the chance to record another vascular lesion of the inner ear because we know that vascular lesion of the inner ear might lead to an ossification of that canal.

Neuritis usually do not lead to ossification. And probably we had the chance to record a selective vascular lesion of the posterior vestibular artery. The posterior vestibular

artery branches from the vestibulo-cochlear artery and this tiny branch of the vascular tree of the inner ear does not go to the cochlear. So it selectively supplies the sensors that are innervated by the inferior vestibular nerve. So it actually perfectly overlaps the inferior vestibular neuritis. So we always have to keep in mind that when we have a typical scenario of vestibular neuritis that could be a superior vestibular neuritis with horizontal and superior canal impaired with the utricle, or also if we have selective lesion of the saccule and the posterior canal, we always have to keep in mind that it might be vestibular neuritis but also a selective lesion of those branches that supply the same structures that are innervated.

So the anterior vestibular artery for the superior part of the labyrinth and the posterior vestibular artery for the inferior part of the labyrinth. Okay, it was quite a strange case, I would say, but quite huge case. Let's go to vestibular schwannoma. Vestibular schwannoma of course might involve just the inferior part, the inferior branch of the vestibular nerve. This is a typical case of a lady with a right-sided tinnitus and progressive right hearing impairment with slight unsteadiness. Usually patients with vestibular schwannoma do not have acute vestibular symptoms usually, they just have progressive symptoms or very few symptoms of the vestibular symptoms. This is her audiogram and the most important thing is that after hypermethylation, she developed an important, rapidly an important recovering nystagmus, nystagmus beating toward the lesion side.

This is typical for neuro issues, typical for neuro. And we performed an accurate vestibular assessment for her and bithermal caloric test was normal, so horizontal canal was normal in low frequency and the video head impulse test was selectively lesioned on the right side for the posterior canal. And of course, cervical VEMPs were impaired on the right side, and ocular VEMPs were normal on the right side. So of course, she had a vestibular schwannoma on the right side, you can see here. And in this case, we had a typical clinical picture consistent with vestibular schwannoma that

selectively involves the inferior branch of the vestibular nerve. Of course, it probably compresses the cochlear artery, leading to a co-existent hearing impairment, progressive hearing impairment on the same side.

There are plenty of papers on the vestibular assessment of patient with vestibular schwannoma. This is just one of those of Professor Taylor from Australia, with Professor Welgampola. And they say that in particular, if we have little vestibular schwannoma, in the case of small vestibular schwannoma, it is possible to detect a selective lesion of the superior vestibular nerve in 12% of patients and in 10% of patient an inferior vestibular nerve involvement, while in most of the other patients, probably there is a mixture of involvement between superior and inferior vestibular nerves. This is another patient with not in acute stage, but in a chronic stage, and I would say in intrerictal stage because it's a patient with diagnosis of right definite Meniere's disease.

He was about to evaluate in the interictal stage, this is his auditory function. He had typical right-sided recurrent cochleo-vestibular symptoms. He had a progressed hearing impairment. But look, this kind of patient had a normal bedside head impulse test on the horizontal part, but look, after head-shaking, he had the downward components of the post head-shaking nystagmus. And this is quite typical for patients with Meniere's disease, I guess, I'm sure that most of patients that now are in the audience agree with me that a big portion of patients, a consistent portion of patient with Meniere's disease after head-shaking nystagmus, they really do develop a downbeat nystagmus. Why? Why do they develop downbeat nystagmus after horizontal head-shaking?

Because probably, beside having the typical low frequency impairment for the horizontal canal typical for Meniere's disease, most of these patient do have selective reduction of the posterior canal on the same side. And also of course, this patient also

had lesions of the auditory receptors. But this kind of downward component, vertical component in that post head-shaking nystagmus are quite typical for Meniere's disease. This is one of the paper from colleagues from South Korea that says the same thing, the downbeat components are present in 97% of patient with Meniere's disease and also patient with vestibular migraine. And these vertical components are possibly related to the involvement of vestibular apparatus and compensation, probably both mechanisms, but this probably reflects an asymmetrical impairment of vertical canals or misorientation of the velocity-storage mechanism.

Probably in our opinion, a subset of patients with this kind of downward components are probably due to this asymmetrical impairment of vertical canal. Usually, the posterior canal of that side is impaired and we know that if we perform horizontal head-shaking, we just do not, I would say we just not excite and inhibits the horizontal canals because a little bit exhibition and inhibition activation of the vertical canals occur during horizontal head-shakes. So if we charge the velocity-storage with the horizontal head-shaking and we stop, probably there is an asymmetrical release from the velocity-storage and we might face this kind of downbeat nystagmus. The posterior canal loss in endolymphatic hydrops Usually in the interictal stage in chronic stage is quite common.

I'm sure that most of you have detected this kind of finding, and this is probably due, these are just some records that I would like to mention is most frequently the most frequent detected abnormality is selective posterior canal loss followed by horizontal canal loss. Usually, posterior canal disorder might cause subjective sensation of dizziness rather than acute vertigo, but yes, it could be also my experience. I also notice patient with acute vertigo with a selective lesion of the posterior canal also in the acute stage with acute vestibular symptoms. And this is probably due because, excuse me, probably because the posterior canal lies in the undermost part of the labyrinth. So probably the hydropic mechanisms, hydropic abnormalities, alteration, on the

endolymphatic space, probably they aggregate, they are more prominent in the undermost part due to gravity probably this might be one of the possible explanation or it might be probably due to a continuous enlargement, herniation of the saccule within the posterior canal.

We know that the saccule is very close to the ampulla of the posterior canal, so probably an herniation of the saccule, progressive hernation of the saccule might lead to a selective lesion of the posterior canal, a little bit similarly to what has been published recently about the reduced VOR gain for the horizontal canal in Meniere's disease probably due to an hernation of the endolymphatic space of the utricule inside the horizontal canal. This is another case of a patient with a diagnosis of probable left side Meniere's disease because she just had an acute sensorineural hearing loss years before with this kind of pattern in the high frequency domain. And since then, she developed recurrent vertigo attacks without typical cochlear symptoms.

So it's not a typical Meniere's disease, so it might be diagnosed as a probable left-sided Meniere's disease. This is her video head impulse test in the interictal or chronic stage, but she developed a recurrent vertigo attack and I asked her to record, I asked actually her caregiver who was her daughter to record the eye during attacks. And look, there is a slight downbeat nystagmus. I'm not so sure if you can appreciate this slight downbeat nystagmus in this first video and also in this other video, she develops always acute downward, with downbeat nystagmus. Is it central? What is it? Is it vestibular migraine? It's something strange. It's probably another, I would say another manifestation of Meniere's disease that acutely leads to a selective lesion, a selective impairment of the posterior canal.

I asked her to come one time during this attack, she came with a wheelchair. Look, there is a slight downbeat nystagmus that could be recorded by video oculoscopy. And look, this kind of downbeat was much more appreciated in supine position, and also

with the vibration, it was much more increased. What is it, is it something central, a recurrent central pathology that are associated with that left previous sensorineural hearing loss on the left side? Hmm. But the most interesting thing is that in acute stage, she developed a selective lesion for the left posterior canal. So a selective VOR gain impairment for the left posterior canal at the video head impulse test. And looking in the literature again, the group of Professor Kim from South Korea published a series of patients with downbeat nystagmus in the ictal stage, in the acute stage of Meniere's disease.

It is quite interesting finding because it's probably a subset of patient that behaves differently from the other Meniere's disease patients. Usually, these patient have a simultaneous impairment also of the cervical VEMPs, and besides the decreased gains for the posterior canal. So probably also this patient do have endolymphatic hydrops, and probably there is an acute herniation, I'm sorry, an acute herniation of saccules within the posterior canal or something else. It's not easy to understand, it's not easy to know, but it's interesting that this kind of findings are replicated also in literature. This is another patient that came to my office actually with a history of recent positional vertigo that started like positional vertigo, but became worse and worse.

It became unsteadiness, it became something worse for him since the morning that I examined him. He had a long standing left sided hearing loss, he had a brain MRI that was negative, he had a horizontal bedside head impulse test that was pretty normal. So also, this patient needs to be checked for central pathology. Of course, his neurologic examination was normal. But look, this downbeat nystagmus was enhanced by this positioning. It was probably also a slight right torsional component. I'm not sure if you can appreciate it. What is going on here, is this central, is this peripheral? What else? I performed in acute an instrumental workup and ocular VEMPs and cervical VEMPs were symmetrical, but he developed a selective lesion of the left posterior canal.

And since he had positional vertigo before, I performed some maneuvers, I performed some maneuvers hypothesizing something that was related to an otolith clot in the posterior canal. And in fact, after one week from this current reposition maneuver, look, his spontaneous nystagmus subsided totally, and I will rapidly go to the interesting part, but if I put it on the left side, we can appreciate paroxysmal horizontal geotropic nystagmus. Now it's beating toward the left side on the left side, so it's geotropic and it's transient, and after a while, we'll go to the right side and it's transient again toward the right side. So probably he had something that was definitely related with this positional nystagmus. This is typical for BPPV.

After this maneuver, the video head impulse test normalized for that posterior canal. So we hypothesized what? We hypothesized a selective jam of the posterior canal that in this case was continuously pushing the cupula, was continuously leading to a positive pressure toward the cupula, or the cupula was bend toward the utricle so inhibiting the posterior canal. So we had downbeat nystagmus, spontaneous downbeat nystagmus with a right torsional component, it was on the left side. And we also had a left hyporeflexia that was transient, transient because as soon as we made maneuvers to detach that clot from the wall of that membranous canal, we had a conversion from posterior canal jam to BPPV involving the non-ampullary arm of the horizontal canal of the same side.

So we had a canalith switch, let's say, with the normalization of the VOR gain for that posterior canal. But it was quite rare case. I will not say that posterior canal jam are quite rare. What is much more frequent is this scenario. Look, this is a patient, this is Pasquale, this is a patient with a typical positional nystagmus I would say with position vertigo, but not typical positional nystagmus. We did not have a typical upbeat nystagmus with ipsilesional torsional component, this is a right Dix-Hallpike maneuver. I will run again the video. Look, we not have the typical upbeat nystagmus with the

paroxysmal course but we have a downbeat nystagmus with right torsional component with persistent course.

And this is not rare at all. This is quite often to have this kind of patient. What's going on in this patient? Could it be central? Hmm. Could it be peripheral? Yes, probably it's peripheral, but is it a BPPV? Is it right anterior canal BPPV or left BPPV involving the non-ampullary arm of this canal because both of these BPPV, both of these scenario lead to the same downbeat nystagmus with the right torsional component because if it is a right anterior canal, during the Dix-Hallpike maneuver, we have an excitation of that canal, so downbeat with the right torsional component. But if you have a left apogeotropic posterior canal BPPV due to an involvement of the non-ampullary arm of that canal, we have an inhibition of the left posterior canal that results in the same downbeat nystagmus with the right torsional component.

So my friend performed an accurate assessment of this patient and he found no alteration, no abnormalities of the cervical and ocular VEMPs, but just a slight reduction of the posterior canal on the left side, so probably it was a left apogeotropic posterior canal. And in fact after maneuver, he obtained a conversion from left apogeotropic posterior canal in a typical geotropic posterior canal. Look, now the nystagmus is upbeat with left torsional component typical for BPPV of the left side. So he made maneuvers and after those maneuvers, we had the normalization of the VOR gain for that left posterior canal. So also BPPV not in the typical jam scenario, but also in this kind of scenario when we have a positional persistent downbeat nystagmus, also in this case, we might have typical impairment of the VOR gain in high frequency for that canal.

Of course, my friend performed an epi maneuver to restore her symptoms. So we published these papers because in case of persistent position of downbeat nystagmus due to either anterior canal BPPV or apogeotropic posterior canal BPPV, in particular

when there is downbeat nystagmus with persistent course, not with paroxysmal course but with persistent course, probably debris, otoliths are thought to alter endolymphatic dynamics and cupula response mechanism, resulting in high frequency VOR deficit for the involved canal because probably they are organized in a kind of positional jam. They are not jammed when the patient is upright position, they just lead to a positional jam, when we bring the patient in the diagnostic tests, in the diagnostic position, they lead to a positional jam on that canal.

So they allow the cupula to be activated by low frequency stimuli that are otoconial shifts, but they impede the ampullary receptor to respond to high frequency inputs. So during the head impulse, there is a reduced response, that's why we call it a low-pass filter. So this condition may probably occur when otoliths are close the canal here, close to the common crus for the posterior canal leading to a persistent downbeat nystagmus. Or when the otoliths are in the arterial canal, probably in another portion the anterior canal, where they are partially entrapped there, leading to a persistent downbeat nystagmus. In this kind of condition, in this condition, it's quite easy to find a slight reduction selectively of the canal, posterior canal in case of non-ampullary arm, posterior canal BPPV, or anterior canal in case of anterior canal BPPV.

And this is what I've just said. So in this cases we might have this kind of slight reducement of VOR gain. Okay, so the last minutes for selective lesion of the anterior canal, I will be very fast because it's not very common to find anterior canal loss. And if we find anterior canal loss, we always have to keep in mind that we might have in front of us a superior canal dehiscence because superior canal dehiscence is probably the most important disease that might lead to a selective lesion of the superior canal. I'm not sure if you had the chance to see this bedside head impulse test for the horizontal canal is normal, now in the RALP, normal, right anterior left posterior, now in the LARP, right posterior left anterior, look this huge upbeat corrective saccades after head impulse for the left anterior canal.

So superior canal dehiscence is one of the most important for my window lesion that lead to vestibular symptoms and cochlear symptoms, typical symptoms due to a reduced impetus on the inner ear. And of course there was the first report of Professor Miner from Baltimore, and you can see here a typical patient with this kind of conductive hearing loss for the low frequencies, but with normal impedance on the otometer with preserved acoustic reflex, so there is nothing going on on the middle ear, it's inner ear conductive hearing loss. And look, this is a positive Valsalva maneuver with closed nostrils with downward nystagmus due to an increased pressure of the middle ear. These patients usually have typical torsional components at the skull vibration tests, most of them have horizontal components beating toward lesion side.

And this is thanks to Professor Duma that we have this kind of notions. And most of these patients, of course these patients do have an increased amplitude of ocular VEMPs and increased amplitude of cervical VEMPs with the reduced thresholds due to a lower impedance of the inner ear. This is the typical scenario of superior canal dehiscence. This is the images, the CT scan of the patients, and most of these patients who have a reducement of the anterior canal VOR gain. Why? Why do this patient develop this reducement? Because probably, okay, this is just review of the possible sites of superior canal dehiscence. But why do patients with superior canal dehiscence developed a reducement of the VOR gain for that canal?

Because probably the wider is the dehiscence, no problem. This is what we found, we found that the wider is the dehiscence, the lower is the VOR gain for that canal. So there is an inverse correlation, a strong inverse correlation between SCD size and VOR gain for that superior canal. Why? This is our results of a paper that we published some years ago. There was no modification of the VOR gain of the other canal, but just for that canal probably or there is auto-plugging process of the middle cranial fossa content of the superior canal, and this was something that was already hypothesized

from colleagues from the group of Professor Miner and John Kerry, probably there is a dissipation of mechanical energy through the dehiscence.

So when we perform the head impulse test during this kind of downward head impulse, some of the mechanical energy is wasted through the dehiscence because probably, there are some pumping effect on that dehiscence that is not covered in that part of the canal. When the canal is not covered, probably the membranous duct is behaving differently, allowing a dissipation of that energy that does not go to move the ampulla correctly. So probably there is a dissipation of energy during the head impulse. And of course, this is much more likely to occur when we have bigger dehiscence. But we also have to keep in mind that this kind of scenario might be reproduced also in posterior canal dehiscence.

Look, we have a typical pseudo-conductive hearing loss on the right side in a patient with right pulsatile tinnitus with a selective lesion of the right posterior canal and this patient's had a posterior canal dehiscence due to a right high-riding jugular bulb. So also dehiscence, depending on the canal effect, it might lead to a selective lesion, a selective apparent lesion of the VOR gain for that canal. Of course, a selective lesion of the anterior canal might be due to an anterior canal lithiasis, an anterior canal BPPV where probably there are otoliths partially entrapped in that anterior canal. Look at this lady with positional downbeat nystagmus. Look, there is a positional persistent downbeat nystagmus with slight left torsional components.

What is going on here, it's a central pathology or it's a peripheral pathology due to a apogeotropic right posterior canal BPPV or left anterior canal BPPV? We cannot know, but if we have in our hands the video head impulse test, we might understand what's going on. Cervical and ocular VEMPs were normal, but the video head impulse test was slightly impaired for the left anterior canal. So probably she had left anterior canal BPPV, that's why after the Yacovino maneuver, we had a conversion from anterior

canal BPPV to a posterior canal BPPV on the left side and we had a normalization of the VOR gain for that anterior canal. We also might have the chance to record a complete jam of the interior canal.

This is quite unique. This was a case that we published last year with a patient with a recent history of posterior canal BPPV that was submitted to repositioning maneuver because it was quite refractory and she become non steady. Normal position of vertigo, but just unsteady. Of course, this kind of spontaneous upbeat nystagmus is not very easy to interpret. You see this kind spontaneous upbeat nystagmus. of course, this patient needed to be checked for central pathologies. But we performed an accurate vestibular assessment and we found a selective lesion of the anterior canal on the same side of the recent BPPV that she had on the posterior canal. So we hypothesized this kind of jam that was continuously pushing the cupula toward the anterior canal on the utricle, so inhibiting the anterior canal, so that's why we had a spontaneous upbeat nystagmus.

And after a repositioing maneuver, actually after Brandt-Daroff maneuver, we suggested her to perform Brandt-Daroff maneuver to recover, we had a normalization of the anterior canal VOR gain. Of course, she was scheduled for central pathology, nothing was going on. I think this is one of the last case. We can also have the chance to record a selective spontaneous upbeat nystagmus in some patient with recurrent vestibular symptoms. This could be due to vestibular migraine or also to Meniere's disease. These patients had the typical symptoms of Meniere's disease on the left side, of course also, she had to be scheduled to be checked for central pathologies, and she had a selective lesion of that left superior canal.

So probably in that particular moment, she had a selective impairment of that canal, leading to spontaneous upbeat nystagmus. And based on MRI according to this research, endolymphatic hydrops in the semicircular canals is distributed more

dominantly in the superior canal and the posterior canal. So in some cases, this might reflect this kind of spontaneous upbeat nystagmus. And this is also a nice paper from Professor Nicola Perez-Fernandez. He recorded an acute spell of Meniere's disease leading to an irritating upbeat nystagmus, an irritating nystagmus with upbeat components and a selective reduced superior canal VOR gain on the video head impulse test. So we also have to keep in mind that quite rarely also endolymphatic hydrops might lead to this kind of scenario.

And of course, and this is really the last case we might face, a patient with a recent history of acute vestibular loss due to superior vestibular neuritis with an acute stage, a horizontal and a superior canal VOR gain loss, like in this case, on the right side. But we might have the chance to see these patients after a few months and we might get the chance to see that on the horizontal canal, she's recovered, but on the anterior canal, she's not recovered. So in the post-acute phase, in the chronic stage, we might have the chance to record an asymmetrical recovery from an acute vestibular loss, but not in the acute stage. And if you can see here, she has a typical upbeat components after vibration tests.

And this is why, this is because there is a predominant posterior canal components, posterior canal function compared to the anterior canal function. That's why we have upbeat nystagmus. Okay, so in conclusion, and I'm very sorry to be so late. So after this talk, I would like to focus on a few things. So thanks to modern tools for vestibular testing, it's nowadays possible to have detailed and comprehensive measurements of reflexes generated by each vestibular end-organs. So it is possible to detect selective lesions and dysfunction, transient or permanent, for a single receptor and is not rare at all. These findings have allowed to obtain new insights on the changings in labyrinthine micromechanics and vestibular afferents function in patients with acute/chronic interictal states of inner ear disorders.

So according to our findings we can conclude that the posterior canal represent the pattern most frequently detected. The selective lesion of the posterior canal represents the pattern most frequently detected with the video head impulse test, likely due to a separate innervation and vascularization of the posterior canal compared to the horizontal and anterior canal. Endolymphatic hydrops in the acute stage, in particular in the acute stage, and BPPV with a complete or incomplete jam can result in selective lesion for each of the three canals with different pathomechanism, but on a transient base, and each case, excluding long-lasting hydrops where posterior canal loss usually persists. So a selective horizontal canal loss may be due to Meniere's disease. When we have a selective lesion in acute stage of a patient, always think to Meniere's disease in the acute stage, or a BPPV with a horizontal jam.

In this case, we have to ask the patient if he had previous BPPV or he suffered from BPPV or we try to transform this kind of spontaneous nystagmus in a typical BPPV. Or we can face in the post-acute/chronic stage an incomplete or asymmetrical recovery following an acute vestibular loss due to a superior vestibular neuritis or an anterior vestibular artery ischemia. In case of selective anterior canal loss, we always have to think about superior canal dehiscence or sometimes more rarely, BPPV leading to position of downbeat nystagmus, so an anterior canal BPPV, or Meniere's disease, but it's quite rare I would say, or sometimes in a chronic stage following an acute vestibular loss, selective lesion of the superior canal because there was an asymmetric or an incomplete recovery with just the horizontal canal recovered and not the anterior canal.

And in case of posterior canal loss, we have to think a lot of possible diagnosis. So labyrinthine ischemia is probably the most important diagnosis to think about in particularly if we have sensorineural hearing loss on the same side, sudden sensorineural hearing loss on the same side. Inferior vestibular neuritis is not that common, but it might happen. Meniere's disease, a lot of Meniere's disease both in the chronic stage, I would say much more common in chronic stage and more rarely in

acute stage. But it might happen that recurrent attacks of Meniere's disease may lead to downbeat nystagmus and selective lesion of the posterior canal. Of course vestibular schwannoma, if we have typical symptoms, chronic symptoms of vestibular schwannoma, and also BPPV, in particular apogeotropic form of the posterior canal where otoliths are partially entrapped in another portion of that canal.

Or also posterior canal dehiscence if we have other symptoms and other signs typical of thermal reader. So thanks to an extensive vestibular assessment, it's possible to differentiate also central from peripheral acute vestibular dysfunction. If you saw, a lot of patients have the question mark, is it central? Is it peripheral? But thanks to the assessment, we had the chance to be sure that all this patient was peripheral, and of course an extensive evaluation of all vestibular centers and afferents should be always given to avoid misdiagnosis. So I would like to suggest you to stress, I would like to stress the importance of a comprehensive assessment also in the acute stage for this patient with atypical scenarios. So I would like to thank all of you, and I would like to thank again my friends, Salvatore and Pasquale, and I'm here for questions and I will be happy to answer you.

Okay, so. I will go through through the questions. Pleasure to meet you both masters, thank you very much for your interest. I can see the paper, okay. Regarding your position, so Mayada Shelif, thank you very much for being here, Mayada. We met in Dubai, she's a very nice, very talented physician from Egypt, so I really say hi to her. So Antonis Vasiyalis, yes, regarding your position that hydrops may affect more the posterior canal, do we have any data that selective loss of posterior canal can be found more often in hydrops compared with the horizontal canal hyperfunction that we already know that is not very often dissociation of caloric head impulse test. Well, I would say that in the high frequency domain, it's much more common to have posterior canal loss of the video head impulse test.

So in the high frequency domain with a video head impulse test, I would say one third of patients in chronic stage of Meniere's disease, they do have a posterior canal loss. On the contrary, in the low frequency domain, of course we do not have a similar test like a caloric test for the vertical canals, caloric test just test the horizontal canal. Most of patients, I would say, more than the health of patients, I would say almost all patients with Meniere's disease do present with the impaired caloric results. I don't have an answer for this. I don't have an answer for this. I think that the MRI will give us important clues, important answers for this, probably there are different compartmental dysfunction.

Probably not even MRI will tell us the answers, but yes, it's a good question, but I'm not sure I can answer you, but it's a really good question. So how long in your opinion could last since canalith jam goes back to classic BPPV triggered by symptomatology? In this case, usually canalith jam does not last very much. Usually it's something about days, I will not say more than one or two weeks because in one way or in the other way, in some ways, that otolith clot crumbles and goes to switch to a typical form of BPPV. Usually patients do not have the typical symptoms of an acute vestibular loss because it's just a selective lesion on that canal without utricular otolith, I would say macular deficit.

But when I have a selective lesion of the horizontal canal or some other canals with a history consistent with positional vertigo, I always try to do maneuvers to detach that otolith clot. This is a nice case, but actually I will like to see the instrumental assessment of this case. Not sure I can answer you, I'm sorry for this. Again, Antonis, ischemic lesion of the right is better results actually follow up head impulse test series? Yes. Which one has best results? In my opinion, the real neuritis do not recover, the real neuritis, acute neuritis, in my opinion, they do not recover. They are still impaired even after months. Ischemic lesions, probably because there are some gray areas

probably because there are some collateral flows probably, they tend to recover instrumentally a little bit better than the real acute neural lesion.

But we have to think also another thing that's mainly elderly patients develop lesions due to ischemic issues. And we know that elderly patients do not recover very well compared to young patients. So we have to consider both of this condition, and you know, elderly do not recover because probably they do not move very well, so they do not stimulate the compensation and also they do not stimulate the recovery of that function. So we have all to think of this condition both. But I would say that in a middle-aged patient, in age 50, if he had vestibular neuritis, I think he would recover worse than an acute ischemic lesion. My first question has been answered by the slides.

Okay. Ah, okay, do you see the K of nystagmus when finding canalith jam, and do you see the K of nystagmus when finding canalith jamming? The K of nystagmus, you mean probably, he is Kim Fox, the K probably in nystagmus is changes. No, usually when there is a canalith jam, nystagmus is spontaneous and it does not depend, it is not affected by change of head position. So usually it does not change. Jerry Kaplan, it is possible to get a transcript of this class? A lot of material is covered. Sure, probably we'll organize something. Andrea, what is your opinion about, Maxine Suduko, oh hi, Maxine, very nice to have you here. A very nice friend from Ukraine.

What is your opinion about the three different otoliths induced endolymphatic hydrops? Maxine, this is very interesting. It is a very interesting topic because we know that there are some research that thing that some hydrops might be due to detachment of otoliths from the saccular macula. I think that if we do not find some kind of maneuver that can make the patient recover from hydrops, it'll be very hard to prove this kind of hypothesis, but it's actually a very interesting thing. It's interesting because we know that a significant portion of patient with hydrops do develop typical BPPV because of

utricle detachment. So it is easy to think that probably some saccular otoliths detached from the saccular macula endo probably toward the cochlea.

And it's my experience that some patient with hydrops, with spontaneous nystagmus due to an acute stage of hydrops, when you perform some kind of maneuver, just diagnostic maneuver, some of them say that they feel better, and some of them also have a reduced nystagmus, but it would be interesting to find abnormalization also of the hearing function. So in my opinion, we are still a bit far from the answer of this nice question. Paolo Cruz, this was a true masterclass. Thanks. Thank you very much, Paolo. I think we met in Facebook somewhere, so I'm very glad you appreciate it. Thank you very much. Second question from Maxine, how to explain spontaneous nystagmus of the healthy side in Meniere's disease, has improved to normal.

How to explain spontaneous nystagmus towards the healthy hair, so paretic spontaneous nystagmus in case, has improved or no? Ah, so not if it's reduced, but just if it's normal or improved. Actually it's quite common to find an irritated, excuse me, a spontaneous paretic nystagmus in patient with hydrops with no positive head impulse test on both sides. And it goes obviously in differential diagnosis with central pathologies. Of course, if a patient has typical recurrent attacks, we do not schedule him acute neurological examination or MRI in a few days. So in my experience, most of patients with spontaneous nystagmus in Meniere's disease, also paretic nystagmus, do not have impaired VOR gain at the video head impulse test. Some patients I showed you had.

In this case, there is just probably endolymphatic, I would say, issues about rupture on membranes with ionic mixture that probably do not lead to a lesion on the high frequency response, but just lead to a transient hypofunction of that canal leading to paretic nystagmus. And probably it's not so detected by the high frequency in the high frequency domain. Real answer is probably not so easy to give a real answer, probably

there is a transient distention of that canal that do not lead to a real VOR gain reducement. I think in literature, there are a lot of theories and would be nice probably in another webinar to see and to try to discuss about this because thanks to these modern tools, it's very easy to record in the acute stage people with Meniere's disease.

I have a bit serious patient with acute attack of Meniere's disease without impairment of the VOR gain, which you see also in literature with other different theories. And to better answer to your nice question, Maxine. Can we watch the record? For sure, Ram De Silva, for sure. Thank you very much. Thank you very much really. I really appreciate your comments. Thank you very much, Marie Cecilia, thank you very much, Yu. Informative webinar, thank you very much. Oh, Professor Azuma, it was honor for me to have you here. So if you're still here, I give you a big hug and I thank you for your great lectures. I always learn from you. So thank you much, it's honor for me to have you here.

Is it typical to see spontaneous downbeat nystagmus in post horizontal downbeat nystagmus in Meniere's patient both during and between episodes? Yes, Dr. Kim Fox. Yes, in my opinion, from my experience, well, spontaneous downbeat nystagmus is not that common. I would say it's quite rare. I had the chance to record at least four or five patients with acute downbeat nystagmus in the acute stage with selective transient posterior canal lesion. This is not very common. Also, Professor Kim from South Korea, with a huge series of patient they have, they just published I think a series of 10 or 12 patients or probably 7 patients. But the thing that is very, very common in my opinion is the post head-shaking nystagmus with downbeat components.

This is quite common in my opinion. And this is, in my opinion, it's due to an asymmetrical activation of the vertical canal doing horizontal head-shakings. And this somehow goes together with something that we are writing with Professor Azuma. So probably it would be nice to come again to this focus, to this topic when we'll be ready

the paper with Professor Azuma. Hey Catarina, thank you very much. Thank you very much for your comment. If you're still here, I say hi to you. How does the partial focal damage of labyrinth caused by ischemia in the treatment protocol? Well, this is a nice question. Of course if we know with video head impulse test and cervical and ocular VEMPs, the lesion pattern, we have to schedule a treatment for protocol, especially for this kind of lesion.

Of course, if we have just a lesion of the posterior canal, we will have to insist on vertical movements, vertical type one and type two exercises, especially with the movements from upright to supine, much more than horizontal movements, and then horizontal treatment protocol. This is at least what I think. Actually, I'm not involved in first person in the treatment protocol of patients. I usually give other colleagues my patients to be treated, but I would suggest my colleagues to insist on vertical movements. Thanks for a great presentation. Thank you very much, Mina Duna. Genie, she's a very close friend of mine. I'm very happy to have you here, so hi. What do you mean by selective BPPV?

Yes. Yes, when BPPV or in a complete jam or a partial jam involve just one canal, we have a selective lesion of that canal on the video head impulse test. Genie, it would be nice to speak about this with you to have the exams of these patients. It would be interesting to know if this patient have a spontaneous nystagmus. Probably they have some jamming process, or I don't know. It would be interesting to discuss together about this patient and to know which other findings you had the chance to detect in this kind of patient. Thank you, Jerry Kaplan, for your nice comments. Sorry. Ah, okay. Maxine, to the bedside. Yes, probably there are always issues about transient, okay, so the irritating nystagmus, you mean probably, Maxine?

Yes. Irritating nystagmus, yes. In the first phase, in the early acute stage of Meniere's disease, sometimes we have transient irritating nystagmus. And irritating nystagmus

sometimes can occur with a selective lesion of that canal or sometimes a normal function of that canal on video head impulse test sometimes an increased function of that canal on video head impulse test. I think that with Meniere's disease, we can detect whatever we want because Meniere's disease we know is a fluctuating process depending on the moment that we test the patient, we have different results depending on the patient. You know, Meniere's disease is a multifactorial disease with so many disease behind, so many pathomechanism that lead to so many etiologies that lead to Meniere's disease.

So probably it can depend on the particular type of patients in this case. I'm not sure if I correctly answer your question, Maxine, but I hope we have a chance to speak about this in other situations. Thank you very much, thank you very much for your presence. Yes, we met through Facebook and we would like to collaborate, so it's a pleasure to have you here. Thank you, Rosa. Thank you, Antonis. This was the last comment. I really thank you. Yes. Okay. Yes, Anna, probably I'm too late.

- Oh no, you are not. But I can see loads of congrats from the audience. In fact, I think that our audience enjoy this fantastic learning opportunity. A lot of questions, a lot of questions. So thank you very much, Andrea. Thank you very much. It was great. And I keep getting messages even from Academia email, asking for the recording, for the PDF for your presentation and so forth. And so the audience really enjoyed this opportunity.

- Thank you very much.

- So we will see for sure Dr. Castellucci soon, very soon because he is a key member of the Inventis Academia and author of several content, which will appear on AudiologyOnline very soon. So thank you very much, Andrea, once again.

- Thank you very much, Anna. May I say that probably in the future, some of the topics that I just mentioned will be displayed better. The theory of jam, canalith jam, will be the topic of another webinar.

- Fantastic. Fantastic. In fact, keep in mind for the audience that you will receive a survey after the webinar that I kindly ask you to fill out because this survey will help us to understand your needs and your ideas in order to provide the best educational support ever. And so thanks to your opinions, we can create a new webinar with Andrea, okay? In order to meet your needs. So thank you, Andrea, once again. A couple of notes for the audience. I just want to remind you that next February the 9th, Professor Samit Desgupta will manage the seminar, "Etiology, Diagnosis and the Management of Vestibular Disorders in Children." It is the first lecture that Professor Desgupta will manage for Inventis Academia. Andrea, thank you very much once again, and I hope to have you as the main speaker for future webinars as soon as possible, let me say. Okay.

- Thank you very much.

- Thank you. Thank you very much. Thank you everybody, and bye for now.