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Meniere's Disease in Children

Presenter: Soumit Dasgupta, MD

Greetings everyone and a warm welcome to Inventis Academia and thank you for joining us live today. It's my great pleasure to introduce Professor Sami Dasgupta, consultant audio vestibular physician and clinical lead in Pediatric audiology at Alder Hay Children's NHS Foundation Trust in Liverpool. We are truly grateful for his continued collaboration with Inventis Academia. So professor, thank you very much. We are also delighted to have with us Dr. Andrea Castellucci from Regemelia Hospital, a long time friend and colleague of Professor Desgupta and a member of the Vestibular Italian Society.

Thank you very much Andrea for coming for introducing Professor Dasgupta during this session. Today's lectures, Meniere's disease in children is the first of two sessions Professor Dasgupta will deliver for us in this 2026. Well, while Menard disease is well recognized in adults, in children it is rare and often secondary. Accurate diagnosis is essential to prevent mismanagement and ensure appropriate care. Professor Dasgupta will guide us through phenotype diagnosis and management supported by cases from his extensive pediatric clinical experience experience.

Please feel free to type your questions in the Q and A section. We will address them at the end of the webinar and you will also receive a short survey afterwards. Your feedback is always greatly appreciated. I now invite Dr. Castellucci to take the floor. Thank you very much, Anna.

Thank you very much for your kind introduction and hello to everyone. And I'm really pleased to introduce today a friend first of all, but also a great physician, renowned worldwide for his interest in pediatric vestibular disorders and for his studies in, in the diagnosis in the treatment of vestibular disorders in children. We know how they could be so difficult to be diagnosed and we know how pediatric population are not friendly for doctors as they don't tell you what they have. So we need to discover, we need to.

You need to open some dark shadows, we need to put lights in dark shadows of their disorders. We have to use all we have in our hands. So bedside vestibular tests and of course a pediatric background

that only a few of us have. So very, very curious to know what Professor Tasgupta will tell us tonight about Meniere's disease and lymphatic hydrox in children. Very, very tough topic.

So thank you very much Sumit for your wonderful lecture that you will have and I will be very pleased to listen. Thank you very much. Thank you very much Anna and Inventis for giving me this platform to preach a very difficult subject. As my very good friend Andrea has been Saying about pediatric vestibular medicine, this is the best way I have found to reach a global audience and it is paying off. I was told that nearly there are more than 700 registrations and already 214 have joined by far the biggest webinar I have ever done with the maximum audience.

So thank you, guys. Without you, this webinar has got really no meaning. And of course, my grateful thanks to my friend as well as a very esteemed colleague, Andrea Castellucci, who I know for some time now. And his work, of course, is mainly in the adult section. And he is a pioneer.

In fact, we call him the new God. Him and another colleague, Salvatore Martelucci, they are kind of advancing the science like no one else. So today, as Andrea and Anna have said, that I'm going to talk about a very difficult topic because Menieres as an entity is very controversial in adults. And to make a diagnosis in children and to treat them is even. It was even more complex and probably a little bit controversial as well.

First of all, I'd like to take your attention to this title. What I said here was Meniere's syndrome, endolymphatic hydrops. I haven't used the word Meniere's disease. So what is the difference between a Meniere's disease and a Meniere's syndrome? That will become apparent as we go through.

So I'm trying to cover as much as possible. It's a very complex topic and my group is actively looking into it. And I'll present some preliminary data later on as to where my group is at. We hope to publish sometime this year. So we are going to look into the brief history of Meniere's disease, not syndrome.

I'll tell say I'll give you the difference later, but Meniere's historical perspective is absolutely fascinating. And as you guys by this time would know that I'm very fond of history of vestibular medicine. And we have got a historical group who regularly publish. We will look into pathophysiology of Menieres, which is of course the controversial, because still nothing is finalized, nothing is certain. We'll talk about diagnostic criteria and then we'll delve straight into the pediatric domain, pediatric world.

We'll talk about epidemiology and etiology of pediatric Meniere syndrome, how to diagnose, how to treat. A very brief idea about our Alder his series, which is still ongoing. And finally we'll conclude. So let's start with the history of Menier's disease. You'll be surprised to know that as early as the first century A.D. there was a Greek physician who was from Syria and who actually worked with.

At the. At the court of the great Roman emperor Trajan.

So he was accurately described describing the very first symptoms of many years. So as you can see that this is pretty much ringing of the years, pain and heaviness and pretty much the whole works that he has described. And this was 1st century A.D. right. And there were sensory symptoms. So probably he was the first one in history to actually tell you about not only.

And not only a phenotype of Meniers, but also probably sensory issues during such spells, which vaguely hints at a vestibular migraine. And then it was lost. Not quite. Because Galen, who followed him, who was the court emperor of Marcus Aurelius in about. Round about 150ad he quoted Archegidis and described similar symptoms in subjects.

Then of course, the Middle Ages hit, the Roman Empire fell and it was the Italian monks from Salerno who went to Arabia to pick up texts and came back at great peril and the science flourished in Arabia. And two Arabian physicians, one was a Persian and one was from. From the Arab world. Raises or

Rashis and Avicenna or Ibn Sina, who was from Persia, they picked up and they also quoted phenotype and artigenesis description. So yes, there was knowledge that something in the background happens, but no one researched into it properly.

And then we had to wait a long, long time until the 19th century.

And in 19th century, remember, vestibular physiology was still in its infancy. Right. Because 1841, Jean Marie flew off from Paris was first said or first showed that the semicircular canals are very important for balance in his classical and seminal PGN experimentation. And then in 1860, Friedrich Golz from Berlin, he ex. He kind of expanded on Floss's theory and he did some experimentation and he confirmed it that there was some kind of hydrostatic mechanism within the lymphatic fluid which drove vestibular sensation.

So that was what was known about vestibular physiology. That's it. And then erupts into the scene a Parisian gentleman who worked in the deaf and dumb school. So obviously plenty of hearing losses. So, you know, admittedly there would be balance problems.

He was a gentleman called the Prosper menier, who in 1861 published this in a French scientific journal. And I've translated this and you can see for yourself that he has picked up where Artigenus has left. But he was the first one in history to actually ascribe such a Symptom galore. Such a symptom spectrum which involves dizziness, balance and hearing to the inner ear. Everybody knew by that time that the inner ears were hearing.

But notwithstanding flir and galls, people hardly knew that the balance is also coming from the inner ear. And he was the first person in history who ascribed an inner ear pathology to deal with balance. So his discovery was, you know, it was way ahead of his times. And his discovery still today rings a huge bell because he was a pioneer. Unfortunately, look when he died 1862 and in 1861 he published it and

he's theory was completely rejected by the French Academy of Sciences, which was, you know, as famous as the Leopoldina in Germany or the Royal School of Medicine or RSM in, in England or the Royal Society.

So people rejected and you know, the tragedy was that he died a very, very broken man. So he was. He got depressed, he lost the will to live and he died just within a year because his theory was completely rejected. And I haven't shown it here, but there was his friend, Armand Trousseau. You remember Trousseau's sign?

Hypocalcemia, Armo Trusso or hypercalcemia. Don't remember when it. When your face ticks. He was the one who took up many earth's cause and propagated what many of us seeing to be finally picked up by the great physiologists like Joseph Breuer, Ernest Mack and Alexander Crumb Brown. They also believe that many a does really have a case.

So whenever you talk about Meniere's nowadays, it's one of the commonest disorders in adults. Whenever you speak Meniere, Meniere's disease, Meniere do give a thought for this absolute pioneer and also think about Armo Trusso. And because it's so complex, 165 years on, we still do not know much as to what is going on out there, although there is some knowledge. So what really happens in the Meniere's? I'm sure that most of you would know about this, but what is important is I'm just going to give you a resume as to what are the accepted theories of the pathophysiology of Meniere's disease.

So it is a clinical syndrome. Why do I call it a syndrome? Because it is a disease of the inner ear. But it has got a global manifestation. In other words, it causes more than one symptom.

It consists of episodes of spontaneous vertigo, classical spinning associated with unilateral fluctuate hearing loss, which waxes and veins Comes and goes tinnitus. And if there is no hearing loss, or if there is a hearing loss, there is an accompanying oral fullness. The accepted theory is, and this has

been proved by postmortem examinations, artificially induced is that the endolymph chamber inside the inner ear labyrinth, the membranous labyrinth, is in the cochlear duct. And in the vestibular membranous labyrinth, it hugely expands and increases its volume, which is the. Which is responsible for pretty much most of the symptoms.

In order to understand what really happens, you need to have a good idea as to how the endolymph moves. You know the metabolism of end, as you know that it's actively secreted by the stria vascularis. That's the origin and that's in the scala media of the organ of Corti in the inner ear. And it is a large compartment that is in the anterior part of the membranous labyrinth. Far bigger the compartment is far bigger than the vestibular compartment, which is in the middle and the posterior parts.

So this fluid freely flows, which comes from the stravascularis. And there must be a region which is over here, which is over here, the red one, where the two endolymphatic systems meet or converge, and that is the ductus, the sinus canalis, reunions. So the ductus reunions is formed by the saccule, which is this, and by the cochlear duct, whereas this is the utricle, the utricular duct. Then the all three of them meet together, where the endolymphatic flow becomes common, forms the endolymphatic duct, and finally it terminates in the endolymphatic sacrificial, where the fluid is reabsorbed and the endolymphatic dynamics is maintained. A key place or a key site in this whole area is the utricular valve of bust.

The utricular valve of bust is actually the junction of the utricular convergence to the ductus reunions. And it's the only place in the whole system which acts as a valve. It's called the valve. Valve means what it permits flow in one direction and it does not permit flow in the opposite direction, so it permits flow only towards the endolymphatic sac. And this is a very crucial concept for understanding Meniere's disease, the valve of bust.

And indeed it has been studied widely by researcher who research on Meniere's. So in other words, the valve of bust is critical to regulate endolymphatic fluid Pressure. However, of course, valve of bust is very important for the kind of physical and structural endolymphatic flow movement. But there is a finer aspect to the endolymphatic flow. You see, the endolymphatic chamber and the perilymphatic chamber in the inner ear bony labyrinth is very much compartmentalized, which means they are separate from each other for the simple reason they are completely different fluids.

And if they have, somehow they mix, they kind of lead to an annulment of each other's electrolyte ionic movement, and therefore there is no function because they neutralize each other. However, the inaya has got a very small area right at the apex where there is a connection between the scala tympani and the scala vestibule. Scala tympani in the scala media. And that's what is called the helicotrema. And the helicotrema is where the perilymphatic and the endolymphatic fluid meets.

So the scala Tympanian scala vestibuli, they communicate to each other through this very tiny gap where the endolymphatic fluid from the scala media in a way contaminates. But in life, in actual physical situations, this is too small an area to make any significant, significant either deficiency or an alteration in function. But it's important because it does explain one particular phenotype of Menieres.

So the three of us, Marco, Mandala, any Salpin, Goneri, you know, Marco from Italy, from Siena, any Salpin is from Izmir. And myself, we wrote the latest Scott Brown chapter on Meniere's disease. And we. We had a very big brainstorming session, and this was what we came up with. That Meniere is either due to increased striavascular activity when the secretion is far more than the reabsorption.

So, so obviously there is a collection or there is something at the endolymphatic sac end where reabsorption is impeded, or there is an obstruction somewhere in the entire duct system. And these small blue marks, they, they. They kind of denote the different obstructions. And that's the valve of bust. So that downstream flow of endolymph from the ductus reunions is somehow impeded.

So the fluid collects on the other side, the squid, the fluid collects away from the endolymphatic sac. Now, this is an important concept because something is causing this, and we still do not know what is causing this. So this figure tells you the different ductal system, and somewhere there's an obstruction. Increased secretion decreased absorption and the causes postulated and numerous. It can be genetic, it can be inflammatory, for example autoimmune, it can follow infection.

Infection, for example, a simple viral, viral labyrinthitis can cause it of course, tumor, trauma, migraine, vestibular migraine, because of its vascular manifestations of vestibular migraine, similarly might affect the endolymphatic flow movement. It could be metabolic, it could be aging presbycosis, age 65 plus. But we still do not know what really happens. We have got an inkling that this might happen and our colleague Jose Antonio Esquemes from Sydney, his group has actively been researching on genetics of Meniere's for a long time and indeed has managed to identify quite a few genes which runs in families, which I've also seen in my families in the pediatric population. So there is convincing evidence that there is an endolymphatic expansion, what now we will term as endolymphatic high drops, which the reason is not clear, but there is a fluid expansion.

What happens to this fluid? That's the important question, isn't it? What happens to the fluid? Is it contained? Now look, membranous labyrinth is a very delicate duct system.

These duct systems are very delicate. If there's too much pressure from the endolymphatic fluid, then there's high chance that the system will rupture. Right? It's a logical, it's, it's a logical inference and that's exactly what happens. So the Ruser membrane bulges up hugely, right?

As you can see, huge collection of the Rus fluid which pushes the R R membrane up and it fills, in fact in some instances it fills the scala vestibule. And then the important thing is the helicotrema mixture of perilymph and endolymph increases. It reaches a stage where it's no longer just a trivial mixture, it's a

significant mixture because of the volume of the fluid. And that leads to a very important phenomena. In early parts of many years when this happens, the kind of hearing loss you get is a low frequency hearing loss.

And there is a postulation that this low frequency hearing loss reflects this mixture of two fluids which are toxic to each other. So then the thing ruptures. And what happens when it ruptures? It ruptures into the perilymph. So once again there is mixing of fluids.

And this mixing of fluids leads to a profound problem in sensory function because of the toxic environment that is created, which is responsible for all the ver symptoms and, etc, etc. But nature has got a wonderful way of self correcting. So after this admixture, the whole thing gets reabsorbed once again by the whole system, especially the internal effect sack symptoms subside and you go back to square one when the rupture heals. Now once again, this is a very important concept. Healing of the, the rupture itself and healing of the rupture.

There are of course other theories as well. For example, a hydrostatic theory says that there is no rupture, it's just the pressure of the whole fluid which causes sensory problems. And the reason for in that case, if I'm saying here that the rupture is responsible for vestibular symptoms, the hydrostatic theory says that in the hydrostatic, because there is no rupture, what happens is the valve of bust is severely compromised. There is a lot of buildup of fluid in the utricle. And what is next to the utricle?

The lateral semicircular canal. So the lateral semicircular canal, sensory epithelia is, is affected and you get a typical vertiginous spell. Which brings me to a very interesting point. When you do proper vestibular quantification in long standing venues, the lateral semicircular canal is the commonest to be affected. And of course utricular and saccule, it could be vascular and ischemic sharing a common pathology.

With vestibular migraine, it could could be an electrolyte imbalance. And the last one, which I am not very clear about, but there is evidence postulated is BPPV can cause Menieres. You know, because BPPV is usually utricular otolith stasis. Right. But in many years what happens is when the saccule sheds the particles.

And because the saccule is very important for the ductus reunions, these debris, they clog the ductus reunions. As a result, a similar problem happens, utricular bulge and everything. So I am not very clear about this, but it has been postulated, certainly theoretically it's very much possible that there could be a saccular autolith debris which might present as BPPV as the initial period. But as I said, evidence is not clear. So this is a pictorial, graphical representation of what happens with the rupture theory, which is the commonest.

So as you can see, the endolymph, the perilymph in the two, two compartments, this bit, this bit will now expand hugely. As you can see, it's expanding.

Mixture of perilymph and endolymph and then voila the RUS membrane ruptures. The whole thing is this is kind of leads to this toxic environment. The whole place is replaced by endolymph and that is a detriment to proper function either hearing on balance. And then after some time the rupture heals and it goes back to square one. I think this graphical representation is extremely clear and it tells you the most accepted theory of pathogenesis.

Now, did you see that rupture? What happens when a membrane ruptures? There's a hole. And what happens if there is a third hole? That's a third window.

So many years in its early stages, as long as there is a rupture behaves like a third window. In fact there is good evidence that the initial phases many years presents with a conductive hearing loss and third window features. In fact I have seen, I, I couldn't show you here. I've seen hen positive Henber

sign in the first stage of men's disease, which is absolutely great. In fact J also told me an also told me that the first stage of any third window is actually a menier, which is, which is a very, very fascinating point.

So how do we diagnose MENAs? Now I'm not going to go through this in detail because this is well established and this is Bani Society 2015 plus the European association of Neurotologists, you know where I a member. So two or more spontaneous episodes, 20 minutes to 12 hours. Look at the duration. The duration is obviously far more investibular migraine.

And for fully blown Menieres you need an audiometric documented low to medium frequency since renal hearing loss during the attack or one of the episodes of vertigo. I do not know about Andrea, but my experience with adult Menieres is I seldom catch them when they're having an acute spell. They don't want to come to the hospital because they are so ill. So fluctuating oral symptoms, you know, it initiates oral fullness and not better accounted for anyways. Now that's, that's the, that's a more kind of less frequent variety than a probable Menieres in the pediatric population because you know they possibly can't describe to you that this 12 hour rule they wouldn't be able to tell you or even the parents would be able to tell you exactly of criteria.

And that makes the diagnosis so difficult. And for probable Menieres it's fluctuating oral symptoms. You do not need to demonstrate a hearing loss. But chronicity is when the ruptures, when the basement membrane ruptures become so thick and fibrotic following repeated ruptures, it doesn't rupture anymore and the fluid goes on collecting. That's the chronic phase and that is where permanent sets in.

And you get irreversible, irreversible hearing loss. And actually the vertigo symptoms get better. Not only that, there is some, there is after some time when the vestibular system completely loses function, that's a burnout, and the other side takes over. So there is sudden relief of symptoms. But I'm not going into that because you already know that.

And unfortunately, 165 years on, we still don't have any biomarker for MD just like vestibular migraine. So it's very much historical based. And that's where the challenge in the pediatric population lies.

So you might not be able to glean a good history, you know, this classical episodic vertigo like vestibular migraine, lasting from minutes to 12 hours. A child might not be able to describe a hearing loss. And there is a very, very skilled art to understand what's going on by just talking to the parents, by asking the right questions. You see hearing loss, the child will behave very differently from a child who's hearing. If a child develops a hearing loss and the parents, there has been studies, about 85 to 90% of cases, the parents, they can make out that the child is not hearing that well.

In fact, I'm a parent myself, I'm sure all you guys who are parents here, you would feel the same, that, you know, something is not right. Something, some, you know, he used to answer to questions, he used to answer to a definite tone of voice he's not answering. So this is a very, very arduous process and you need to be very skillfully trained in it. You need to, you know, kind of see many children because you see children with their parents and you need to develop this insight.

The other very interesting thing which I found in my series is in a developing year, remember the membranous labyrinth in a child is developing. It's still not completely developed, by which I mean, you know, it hasn't attained its adult size. The metabolism is still not the same as in adults. Hormonal regulation of endolymphatic hydrop secretion is different because that's maturing as well. And as a result, it doesn't behave the same way as an adult because I have seen in my series spontaneous remission of Meniere's.

Now let me just now once again reiterate, what is a Meniere syndrome? Meniere's syndrome is Meniere's, but where you find an obvious secondary cause, the other name is secondary endolymphatic hydrops. And in children, primary Menieres, what you find typically in adults is virtually

non existent. It can happen in older children, but it's virtually non existent. So in children it is secondary endolymphatic hydrops or Meniere's syndrome rather than Meniere's disease.

We have mentioned this third window. In the early phases, there's a third window created. Not one third window, Numerous third windows, numerous. And when they heal the fibros, they don't become third windows anymore.

And classical features, they become apparent in the mid to late teens. You know, 16, 15 year old, 16 year old might actually physically tell you the exact Bharani's criteria history, which will lead you to believe that there could be a Menieres and it may exist as a part of a syndrome. There are many syndromes which can present with the secondary hyundrolapetic hydrops, which I'm going to mention a few. So how common is it? It's unilateral in 66% and my series, only about a, I would say a fifth proceed to a bilateral phase because it takes 15 to 20 years for a unilateral to convert to a bilateral one.

Even in adults, 5 to 20 years, 20 years is the maximum. So far as I see it, 1/3 of them spontaneously remit. But this is anecdotal. Hopefully we'll publish this. And this is because of the resolution of the primary cause.

Whatever has caused this corrects itself. It could be maturity, it could be some metabolic problem and things like that. And of course the child's brain is highly plastic. And remember the travascularis also grows and molds itself as the child grows older, older. And the, and the biovestibular function tests are quite variable because you will get vestibular hypo function during an active phase.

But as I said, I hardly see them in active phase. I see them in the quant phase where you might, you might not get them. And here you see a rough distribution of prevalence. So as you can see, it is mighty, mighty, mighty rare. In our series it's 0.7 and by far we are the largest series.

It constitutes only a very small percentage of pediatric vestibular, whereas vestibular migraine constitutes about 38% of pediatric dizziness. In case of Menieres, it conceals only up to about roughly average 1%. So it's very, very rare. So once When I found this, that I'm not seeing as many Menieres, I don't see many BPPVs, I asked myself as to what can be the cause of this lack of presentation of this disease in the pediatric population. So I looked in the publications and there has been a systematic review which was done by my fellow Jayasakti Sanyasia, who's doing brilliant work in, in Malaysia, if she's here.

Jay Sakthi, my salutation to you. They have done a beautiful literature review which was solely needed and you can see the very, very small number of studies, right? It's a very small number of study. 57 children from 1958 to 2024. That's incredible.

That's very small. And none of these studies I had, I went through all the studies, none of these studies actually went through finding an etiology. So they keep on saying Meniere's disease. But in my opinion, the way my series is looking, it's very likely it's ms, not md. You need to do the etiology.

They all had normal CT and MRI. That's a screening one. So MRI is CTE standard 0.6 millimeter, 0 point, I think 0.8 to 1.2 millimeter without reformatting. And the MRI was a standard T2 weight. Familial patterns were observed, which we have also observed.

And One study in 87 that was a long time back has actually mentioned that a third of them are secondary. And he also identified a few, few, few causes like infection, like head trauma, like structural problems in the inner ear and so, and so forth. But many studies that did observe overlap with vestibular migraine.

So what else, what, what else does, what else? Do the publication say that, yes, proposed etiologies would be genetic? Quite a few. And some of the genes, as you can see, they also present with

sensorial hearing loss. MYO 7A, for example, they can present with sensorial hearing loss.

They are at the moment classified as dfnb, autosomal recessive, but I would call them a syndromic one because they also present with the Meneus like picture. And all this, for example, usher one A tecta tecta is autosomal dominant cock, autosomal dominant. So all these genes they present with a Meniere's like picture along with the hearing loss. Autoimmune is very strongly incriminated, especially in the pediatric population, which we shall see Allergy which acts through an autoimmunity bias. Infection, structural defects, trauma.

A vascular theory has been proposed where the blood flow itself leads to an abnormal regulation of endolymphatic flow. It's very much under the influence of endocrine and autonomic. There are some endocrine syndromes which can lead to increased trivascular flow, for example some adrenal conditions. Dietary. Yeah, well, anecdotal but you know, same kind of triggers as migraine chocolates and all those things.

And the commonest variety in the adult population is there is endolymphatic hydrops. But it's primary because we do not know a reason. Sometimes we do, but most of the times we do not. So that's why it's important to distinguish pediatric Meniere syndrome with adults, Meniere's disease.

Now these two which we have come across is very important syndromic conditions. One is a von Hippel Lindau disease that's an autosomal dominant condition with a tumor suppressor gene mutation and they generate roundabout mid teens to early twenties and endolymphatic sac tumor which presents like a Meniere's syndrome. And the VFT is variable. If you are interested, we have published on this, please type my name and Fon Hippilindao. This was actually published by a fellow of mine from Turkey, Samsung, Nasrettin Fatih.

So Nasrettin, if you are in this audience, my salutation to you as well. And the second one which is a very interesting conditions is X linked type of phosphotomic rickets xlhr. Now this is as you see, as you can see, X linked, right? And the entity was notified, was notified to me by my colleague pediatric endocrinologist saying that this child is diagnosed with this and she is extremely dizzy, she can't go to school and so on so forth. Can you please check her out?

So I checked her out and clearly she fulfilled the criteria for probable Menieres and she was put on an immune modulator called burosumab. This is a phosphate deficiency. So burosumab has got a role too in phosphate metabolism and with burosumab the whole condition completely reversed and she did not have any Meniere's like symptoms from then on. And we have also published this, but I haven't, I was part of the author team. But this was published in the Journal of Endocrinology.

So but now we have collected a series which we aim to include in our series. So it presents with a many a syndrome phenotype and these two conditions are very, very interesting conditions.

So why would many years, let's call it endolymphatic hydrops differ in pediatrics and adults. Now look, this is something which has come out of years of reading of various publications talking to the top brains in many years. Most of them are adult men years. But this is what I think I can safely say and conclude.

So the integrity of the stravascularis is very, very, very robust. Far more so if there is an abnormality, it tries to repair itself far better than the adult. If menus is due to autoimmunity, then the immune system is being primed during the pediatric developmental age. And that means that the stravascularis or the whole membranous endolymph might not react to an immune reaction to the same way as that of an adult. It's far more metabolic, obviously, because pediatric systems generally are far more metabolically active, which acts as a protector.

So overall, it's far more resistant to endolymphatic fluid changes than adults. Unless. Unless it's important, something messes it up. So that's this. Unless actually tells you that very likely a secondary cause is messing this up.

In the natural situation, it won't get messed up, but in that abnormal situation, it does get messed up. Whereas it's exactly the opposite in adults, because there, the immune system is primed, the hormonal regulation is there. You know, its metabolism has steadied, it has been stable, less trivascular integrity. That thing will again backfire once one reaches the age of 70 because of wear and tear. I don't know.

I don't know whether anyone has studied. The second important point is it's very difficult to fit the, the criteria because as I said, your pediatric patient will not talk back to you and accurately describe that I had a tumor against crisis. I fell suddenly. Parents will tell you, mother will tell you that I have seen this strange, odd expression coming to my child's face, who then doesn't respond, although he's not lost consciousness, just drops to the floor as if he has been hit by a sledgehammer. This can be BPVC as well, but BPPC is up to the age of four.

So yes, the parents will describe the hearing loss may be normal, because as you know, that in the first early phases it can be variable. It's waxing and waning, which means the system can still repair itself in adults. Similarly, it can present like this, but usually when you're actually catching them in the right period, you will get a low frequency. I tell you the reason. And then it of course evolves.

This hearing loss in the chronic phase, vestibular function is variable in adults. Vestibular function is much more involved, is much more affected than, than the, than the, than the pediatric variety. And well, it's almost always secondary, but the majority is idiopathic. In case of adults you do not find a real cause. Sometimes you are lucky, you can find a trauma.

And the chances of post traumatic Menieres, if I'm not mistaken, is about 4 to 10% after because it, it kind of messes up the vascularity of the trivascularis. And for children it is a very slow progression over several years, is far more quick in adults. And yes, there is evidence of spontaneous recovery.

These are the differential diagnosis in children. Vestibular migraine coexists in 50%. The way you know it's both vestibular migraine as well as meniers is vestibular migraine comes with its set of own autonomic symptoms. So you get a patient with episodic dizziness, tinnitus, hearing loss as well as photophobia and phonophobia, then you have got your case. Third window syndromes are absolutely crucial and important, as I said, that they're intimately connected to each other.

The first stage of superior semicircular canal dehiscence, I didn't know this. My friend Gerard Giannotti told me is actually a Meniers Lica syndrome which was then confirmed by Eugene in school. And this has really opened my eyes because this will also help you immensely from the clinical point of view. Peripheral cochlear vestibulopathy, as we said, trauma, tumor, infection, etc. But they can all lead to mania syndrome.

Please don't confuse Ms. With multiple sclerosis. It inadvertently happens, but as long as you are categorical about it, shouldn't be a problem. And in central vestibular symptoms with dizziness, usually audiological symptoms are missing and you get the classical central eye movements. Now comes the crunch. How do you diagnose a population where you do not get a proper history?

Not talking about older children, please. I'm talking about children up to the age of maybe 13 or 14. 14. 14 to 16. They are good with history.

Except sometimes boys don't talk and the girls, they come up in monosyllabic answers. So the boys will say huh? And the girls will say yes, maybe a bit. So even they are difficult, you know, even they are difficult.

So they might not be described. So once again you are depending on the parents to understand whether there is any hearing loss or a dizziness or a tinnitus. Tinnitus as a history from the pediatric population is surprisingly well obtained. You know, why? Because if they hear an ectopic noise in their ears, they're the first to tell you that I'm hearing something in my ear.

They are very good to tell you they're hearing something. What they are hearing, they can't. But they will tell the parents. And that's, that's a good piece of history. Muffled hearing, oral fullness.

Something is blocking my ear. Third window symptoms. The commonest I've found is autophony. That is they don't like their voice if they can describe it to you. And sometimes you also get a vestibule.

You also get an audio vestibular behavior. When previously a child was very vocal, a child was kind of very expressive. Suddenly he starts to talking in very low tones. That's, that's. Now these are fine traits of examining children which can only come if you have seen many, many children, they're interested in the behavior.

And yes, cognitive and psychological reaction can be quite severe. The way they react to their condition. Any dizziness, pediatric dizziness generates a lot of psychological morbidity which we have seen. So that's why you need to diagnose them. So it is.

And also the other thing which is very important, that this condition generates a lot of fatigue. That's the other thing. And sometimes I get patients with dizziness, not just menus from the chronic fatigue service because of this.

Yes, I have seen Hennibert sign in our series and it's well established. The early phases of Menieres normal or abnormal PTA depends on the stage of the disease. Starts with low frequency and then it just

kind of proceeds to a flat sensory, neural, later variable vestibular function tests. Vestibular behavior is deranged, plenty of balance problems and stuff and so forth. And of course cognitive and psychological reactions.

So this is my famous vestibular gram. I have done this numerous times in the past. But as you can see, we do a full vestibular assessment focusing on frequency specific information. And of course we do all audiometric evaluation. So autoac estimation, ECOG is very invasive in children, so we don't use that anymore.

Just a little bit on the different bioma or different vestibular function tests. So calorics in md. Now this has been traditionally historically the benchmark of diagnosing vestibular weakness. And it was used universally in many years to demonstrate that there is a vestibular weakness so it can show a canal paresis. And the reason that calorics is indicative of ultra low frequency response, close to the static response.

So because ultra low frequency Same as the helicotriuma mixture. It is a lower frequency pathology rather than a higher frequency pathology, because high frequency pathology is mainly the rapid semicircular canal. But it is promising that there is a dissociation between calorics in and vhit. The latter is normal, while scalorics are not, because you might catch a Meniere at a stage when the low frequencies are affected, only sparing the higher frequency. So we still haven't got into that utricular valve of bust ballooning of the endolymphatic chamber to the lateral semicircular canal.

This is promising, but in no way is it universal and it's found in everybody. It depends on the stage and therefore calorics is not a good biomarker. For many years ecog, SP and ap, of course it was postulated it's a biomarker, but there is hardly any sensitivity and specificity. But recent shows that with ECoG, rather than using a click with a tone burst stimuli, it might have a high sensitivity. But this is very invasive for children and I do not recommend this in children.

Adults you can do it if you want, but not in children. Video head impulse test obviously picks up beautifully semicircular canal weakness, mainly the laterals. VOR gain is not a good indicator, but it may show fluctuations. So it might improve, it might improve and restore itself, or it might show an obvious pathology and it depends on the fluctuation of the disease. So Yasovino, our colleague from Buenos Aires, has conclusive, has shown very beautifully that if you catch the Menieres in active phase, then there would be a VOR gain which is reduced and also in quiescent stages as well.

But nowadays it seems that. Look, initially when there is a weakness, right, the vestibular lateral semicircular canal is weak, let's say. So the view organ is gone down and there are lots of saccades. But because after the system heals itself, the vor gain recovers completely. But saccades may persist, which is a sign of old vestibular weakness, which has been conclusively shown by Nicola Pere Fernandez Ruth Korsager and quite a few colleagues.

And at the end of the day I don't know what Andrea will say, but because he's a big boss in vhit, I don't know what he'll say. But you know, to me I don't consider VHIT as a biomarker for many years.

This is also not a biomarker. They are very conflicting results. You can have all sorts and however, and this is what I found in my series, WIMS will show third window features in the early stages. In fact, I'm going to show you a figure where the first stage of menus were diagnosed in a child where the WIMS had low thresholds and high amplitudes. So it might indicate different types of disease, but not well studied.

But this test which we have started doing looks very, very promising, that is frequency tuning of VMP. So you do a WIMP CVEMP and OEMP at 500 hertz and at 1 kilohertz and then take a ratio of 1 by 1 kilohertz by 500 hertz and you know, usually at higher frequencies the WIMP starts to lose its amplitude. So the 500 Hz amplitude will always be more than the 1 KHz, which is therefore a kind of a

decimal ratio. But in many years, in pretty much all stages, what happens is the whole thing reverses so 1kHz amplitude becomes more than 500hHz amplitude. But overall of course it's not a biomarker for md.

And this has now taken the worldwide storm. Of course this is a good test and it is, it follows a high drops protocol which is quite complex which I'm not going to discuss here because of lack of time and it's still in its infancy but many places are using it and it cannot might not pick up an MD and in it might be positive in non MD as well. It's very time consuming and resource intense and in the pediatric population toxicity of contrast because it's a whopping dose of gadolinium, you are injecting and you're asking the child to wait for four hours after the first run. I tried it on one child of mine and that child and the mum parents were extremely not angry but they were quite uncomfortable and intolerant so we had to call it off. I don't think that we will get an ethical approval to perform pediatric hydrops MRI scans.

So I haven't. I have done it in adults, but I have never done it in children and I don't think that you should do it because nothing is established.

Now this is a test which I have taken from the literature, but I've never done it. And I'll be very, very kind of glad and open to the audience today whether you have got any experience with this in many years. That's a multi frequency wideband tympanometry and it's because of the alteration of Stepi's footprint pressure. It's still in its infancy, but people are working on it and I think that the machine itself to do this multi frequency is pretty expensive and it's too early to infer. But to be very honest and frank, I have never tried it in my population as yet.

So this is a useful protocol in the pediatric population. Detailed history. Then you make a diagnosis or you do you kind of think that there is a probable or or a fully blown Ms. or MD full clinical examination. You need to do audio vestibule examination, Cardiological, neurological, ophthalmological, musculoskeletal, general. Why these the first?

Apart from the first one, all the others have to fulfill Bharani's fourth criteria which is no other condition is explaining the signs and symptoms. Then you do a detailed audiometry, then you do vestibular. Okay, what is going to happen to the OEs? Of course in the non ictal period, in the interval period always might be completely normal. SRTs are usually always present unless of course there is any other pathology which explains a loss of this rt.

I have in fact seen ANST with Menieres as well and oas of course in the later stages they will lose O and this is a useful vestibular protocol. So remember there is no obvious biomarker. But try this at least in older children. And of course this is equally applicable to adults. Caloric dissociation, I.e.

v heat is normal, calorics are not. Do a very proper vng always, always perform the Heber sign always and record it. We hit might show saccades, CWMP tuning and O tuning and tone bust ecog. I don't do in adults at all I in pediatrics at all. And then you need to have an extensive diagnostic workup in the pediatric population.

This is where it's different from adults because you need to look into the autoimmune profile, immunoglobulins, pretty much everything. Mri, ct, different kind of different blood tests, then genetic testing and so on and so forth.

So this is our Alderhead protocol. High index of suspicion. Any two of episodic dizziness with all this. And remember the history might not come from the child or the young adult, but the history will come from in many cases from the parents. If there is added photophobia and phonophobia, you'll think about concurrent or simultaneous vestibular migraine.

Variable hearing loss, it could be normal in the inter rectal period or it could be a CHL in the stage of the third window when the ruptures are happening. Then it might become a mixed One it might become a

frank NSHL early phases it will mimic a third window so you might get negative bc. This is something which I think it might be quite new to you but there is good evidence to it. Varial OE as I mentioned balance function tests are abnormal and CBM tuning and of course you might pick up stigmata of medical condition. For example in the Excel HR you might pick this up.

How do we treat?

First thing is very important, not as much as in adults is lifestyle change. You know it works magic. So this is what first actually is. Not even lifestyle. It's a cognitive approach.

You'll have to take the child and the mother through the entire pathogenesis as to what is happening and that acts as a closure. And remember a child is a very resilient human being. So if the child can reconcile himself to the condition that leads to a sense of well being followed by lifestyle changes. First is diagnosis of allergy. Allergy has got a definite role to play in many years.

So you like to pick up the allergy. If sometimes you do not pick up an allergy, you might have to do a skin prick test and so on and so forth and try to actually manage it. Auditory rehabilitation of course, nothing more to say. DPOE growth and sometimes dead regions. If it's a severe hearing loss, tinnitus, retraining, of course you go for that sound enrichment and all those things.

Vestibular rehabilitation only when the disease in the chronic phase with clear vestibular weakness in your examination blanket. Vestibular rehabilitation for many years is a no go because it doesn't work. Vestibular rehabilitation is where there is a definite documented vestibular weakness and the disease is either quiescent or inactive or chronic. Stress management, as I said, they do react to it psychologically. You might have to address this in many.

In fact I have got kids from the CAM services. So many, many stress management routines. You have to follow plenty of hydration. Salt intake. Many people say that salt should be restricted, but now the philosophy has changed.

You can still eat or take salt, but you'll have to take adequate water with it because it's a redistribution of the salt which is important, not the absolute quantity of the salt. Low grade exercise are very good as well. However, the most important aspect of Meniere's is a pharmacological management and topping the list is betahistine. This is not validated by Cochrane. It it's empirical.

But if you talk to colleagues all around the world you will find out that betahistine is a time Honored highly effective way of treating Meniers. And our colleague Michelle Laur from Marseille has clearly shown that betahistine is a labyrinthine stimulant and it promotes vestibular compensation at the brain stem level. Now previously I've said that as well and I have gone into the debates about this, but I stand by what Michelle has said for the simple reason. I've got immensely good results with betahistine. You might use calcium channel blocker, which is once again empirical, but they do take away the sting of the dizziness when they're taken SOS diuretics.

They do work in adults quite well, but in kids I would be in two minds because they are strong medicines and they need proper supervision. But in older children I would be inclined to use a low grade thiazide diuretic. I will not use a carbonic anhydrous inhibitor because of the side effects. You can get various types of anti dizziness pills, for example ploperazine, phenothiazine, but there is of course there's only some, some evidence. None of them look have any effect on the hearing because in many years at the end of the day both, you know, endolipatic hydrops, secondary or primary, what happens is the distressing bit is the dizziness not as much as the hearing.

A child can live with the hearing loss and we can fit hearing aids, but the dizziness eats into his quality of life, prevents him from going to school, makes him withdrawing, makes him avoid stuff, lose out on

his friendships, lose out on playground activities. So the prime aim of this pharmacological intervention is to get rid of or give a symptomatic control of the dizziness. Steroids, oral or intratympanic? My experience with oral steroids is very minimal. But intratum panic?

Yes, A Cochrane validated study. In severe cases where all these things come into being, when it's a severe severe drain in quality of life, we would go for an intra tympanic and in some instances in about 40% the hearing can be restored provided you are treating the child in the active phase and not in the chronic or in the towards the burnout phase. But it leads to a very significant improvement of vestibular function. And if nothing works, once again, Cochrane validated, you do a medical labyrinthectomy with gentamycin, you take away the dizziness completely. But we very sure that the other vestibular system is working well.

And in bilateral menus, bilateral labyrinthectomy is a no go, is unilateral labyrinthectomy is A no go. You can treat the concomitant migraine as well because they do coexist in 50% of cases. There is one study which has shown that acyclovir as an anti herpes antiviral agent works because their theory is at the end of the day meniers is because of a post herpes infection. Well they might have a case but in no way is it universal. I have tried acyclobide only in one child with good results but of course you can't generalize this and occasionally you can improve the vascularity with calcium channel blockers e.g.

candice, Satan and all those things. Surgical management has a role to play as well. Ventilation tube application. Well, grommets are still done. A minute device is still used but the evidence is very very sketchy.

So I wouldn't be recommending this in the pediatric population. The next one. Tenatomy of the middle ear muscles, hardly done. So it's all with the postulation or all with the theory that if you alter the middle ear pressure then the stapes foot plate pressure will be relieved as a result that will lead to better endolymphatic flow. Well, it's very far fetched but in some instances work but I don't use them.

Our colleague, Professor Hisam Saliba from Morale, he has devised an operation where he actually physically clips the valve of bust just beyond because he thinks that if you can prevent this huge amount of endolymphatic fluid, the lack of endolymphatic fluid reabsorption at the endolymphatic sac level, then if you arrest it in the middle then that leads to an improvement. But I don't think it will work in everybody because as we have seen it has got multiple causes and it's very difficult to make out what is what. Endolymphatic sac surgery. Many colleagues do it and they swear by this. But my limit, my, my experience is quite limited with endolopharitic surgery because usually 95% times of the cases I, I've achieved very good symptom control with pharmacological management including intra tympanic steroids.

I don't need them to go to surgery in the pediatric population. I'm not talking about adults. You know, in all our series of many years in pediatrics there has been intra tympanic steroid injections but no, we haven't had any proper medical labyrinthectomy or an endolytic surgery or a vestibular neurectomy. Now here I'm going to tell you a very interesting story. I was once referred a patient who was about in early 30s, a top flying exec, made own diagnosis.

I'm not mentioning the sex because of GDPR made its own diagnosis and had a colleague neurosurgeon perform a vestibular neurectomy. The symptoms didn't go away, in fact the symptoms got worse. Vestibular neurectomy has been shown to be a sure shot, absolute sure shot sent to me and I was completely in a fix. So I asked a senior colleague in America and he told me a fantastic thing then that in as much as about 1/7 or 1/8 of the human population the cochlear nerve carries inferior vestibular nerve fibers, which is fantastic. So what are you achieving when you're cutting the inferior vestibular nerve?

Nothing. This is a normal anomal variation. It's almost like one side does not have a vertebral artery. The other side has got beautiful vertebral artery and you only pick it up when you routinely scan the

back of the neck. These freaks of human anatomy are present throughout the entire human population.

Remember this is absolutely important to know so things can go wrong. These are the outliers and you must believe that. You must, you must keep that in mind. And finally we'll come to the Alder Hay experience. That's my lab.

So our incidence is 0.7, average is about 14. We have done the whole bit. We have extensively investigated and we have found in 90% cases there is a secondary ELH. Only in one case. So far we haven't found anything and we have found loads of intracra, different pathologies, intracranial pathologies, for example Langerhans history, Langhan's histiocytosis into nastoid causing Menieres, a vestibular migraine, autoimmune disorder, systemic like for example rheumatoid or juvenile rheumatoid arthritis, an allergic spectrum trauma, two cases with trauma, some genetic.

We haven't typed the gene because we don't have the resources. But the familial history is very very interesting. And metabolic of course the xlhr, as I said. So these are symptoms. We have seen tulios, we have seen autophone, different kinds of balance symptoms, psychological, cognitive and a severe drain on the quality of life.

In our series unilateral is about 75%. There have been a few with bilateral different types of hearing loss, different types of vestibular loss, occasional V, heat and shim abnormalities, CWM tuning Abnormalities missing school. And we have done pretty much all the investigations. For example especially we concentrate on the autoimmune and we have picked up celiac disease from my clinic presenting with a manias phenotype. A few audiometry as you can see, normal audiometry in these two instances, a very mild conductive loss.

In the early phases you get a sensorine or hearing loss and you get a sloping mixed loss. Almost like a mixed loss. Different VHIs. Normal, normal. Just a few catch up suckers on the left posterior on the

right posterior.

As you can see that I haven't shown you any lateral semicircular canal which is the commonest canal to be involved. Shimp asymmetry in shimp is a con. Not a consistent feature but I don't find it infrequently. There is a significant asymmetry between the right and the left side. The right gain is far less for example this one too.

But the way it was normal by the way. And out here they had. We also have a similar kind of an asymmetry which is pathological. Now we have clearly set our shimp norms in a very very detailed publication which has been published in the Frontiers. Please have a look at our norms, especially asymmetry and peak psychotic velocity.

So different kinds of vems. As you can see that there is no vamp here. Out here this is pretty normal vamp, normal symmetry. And out here as I told you, I promised you, this is a huge amplitude with reduced thresholds. We always use bone conduction huge amplitude with reduced thresholds on one side which is the early third window.

These are all unilateral. These are all unilateral and this side is completely normal. So this is very interesting. Now this is CM tuning, right? So as you can see this one was actually told us that there is a third window going on.

And when we did the CM tuning the CM tuning was preserved. So this is less than 1 preserved. But on the same patient we did an O And look, the O clearly shows that there could be and endolipatic hydrops. The ratio between 1 and 500 is quite significant. 1.9.

Now I had a very good look of the OVIM literature in WIMP tuning and there are only a few studies. So this is something which would also highlight in our studies that a CWM tuning is not enough but you

have to also have to do an ovim. So that child, young person, I think 13 or 14 not only had the ruptures phase which is the best memory, the rhizome membrane rupturing and creating a third window effect. But also there is active endolipitic hydrops going on because of O tuning abnormality.

So these are the different managements we have seen. I've told you about the management and intra tympan panic Steroids are only in about two cases an excellent symptom control with this entire full force of multidisciplinary management. What? What are they? Cognitive lifestyle changes.

Beta histine abortive attempt. Abortive to to abort the acute spells with other medication. Intratympanic steroids. That's how we achieved symptom control. We did not require a unilateral labyrinthectomy medical or surgical.

Why? Because I told you that many years syndrome in children. Because the developing membranous labyrinth and the developing brain they do tend to get better over a period of time. Very much un adults. And I've also told you the reason why and how to identify it.

So this was a teenager with eventually find out had a celiac disease. Simple, many a phenotype, comprehensive history. You ask about dietary habits and I was told that this child whenever take something gluten gets a huge tummy upset. One thing led to the other. My rheumatology colleagues who beautifully diagnosed the celiac and this was what I found in the vestibular function test.

A very nice positive head shake, middle frequency involvement.

You see the post hetric nystagmus we hit was normal shimp. As you can see, this is asymmetrical. So there is a problem on the right side and normal wimp on the left. Absent wimp on the right. So this kid was didn't have a life.

Completely messed up. So we had to put intracympanic steroids. You might ask me the regimen and the dose it depends on different places, different kind of regimens. In the pediatric population it's every four weeks for the first three cycles. Then carefully monitor the situation.

So it's intranipanic dexamethasone. I don't inject them. But my colleague ENT surgeon I work very closely with does this for us. This is unique because this was a young person with xlhr, the one which I told you about. And dad and dad and the young person were very astute to send me a video of an acute spell.

Now how lucky am I? I don't get it. In ADA population I think you get it. But in the pediatric population, you see, this was what was sent to me just shot with a mobile phone.

She was hugely symptomatic and by the way, when you get an acute ictal phase like this, I'd like to ask the question and please write it in your chat. Which direction does the nystagmus localize to? Does it localize to the normal side or does it localize to the diseased side? Please let me know so you can see the hearing loss, you can see the shimp, which is pretty normal. Semicircular canals are not affected.

And as far as the WIMPs are concerned, this is also not affected. So largely intact vestibular system, which means you know that herniation is happening, it's releasing the endolymph and then the whole thing is healing, happening one again so you don't get over. But unfortunately the hearing has been affected, so it can happen either way. So finally, although it's rare, it can be made with a very rigorous clinical algorithm. It's a different phenotype and vast majority.

So you need to find out why this has happened and then treat the cause. For example, Langhan cystiocytosis, there was a case who was treated by my oncology department with good results. So you need to treat the cause. So we are lucky that we have a cause. In adults you don't have a cause, so they are not that lucky.

And your first and overwhelming priority is to give symptom control so that the kid gets back a life and of course finding out a cause and treated it. And they respond to pharmacological management extremely well and it's important to keep them under review. So finally, you are more than welcome to my pediatric vestibular course where we do cover many years in great detail. It's in Liverpool, of course. It's a.

It's a beautiful city with a very, very. It's a cultural city. But I'm Manchester United, so I don't support Liverpool United, although they're a great club. And recently we have floated this. It's about a year 10 of us from all over the world.

So you are more than welcome to join our cause and join our group. And that's why I'm so, so grateful to. I'm so, so, so grateful to invent is that Inventis always gives me the podium to propagate this entire pediatric vestibular movement, which is solely needed all around the world. Right, I'll end there. Thank you very much.

And of course I'm open to questions. So Anna, you can take over now.

Okay. Professor, can you hear me? First of all, yes, I can. Fantastic. Professor, thank you very much for such a clear and clinically valuable presentation.

And once again, we truly appreciate your expertise and your ongoing contribution to inventis academia. I. I can see three questions from the audience. May you be so kind, professor, to open the Q and A box and read the first one. Qn box is the chat. Is it?

Yeah. Yeah. Great. Yeah. Got it.

Great.

Yeah. Dr. Lasa. Yeah. Right. Okay.

So the first question is. I understand that there is a hater reproduced many years in such an earlier onset in children can be explained. However, there is not burden. Burden. It is still worth looking for genetics.

Well, you know, this is my opinion. Of course you might disagree with me. It's not necessarily explains an earlier onset. There are many genetic disorders which present as late onset. You know, I can often tell you at least about 100 across the board in audio vestibular medicine where the onset is late.

I mean, a typical example is some of the autosomal dominant hearing losses. They are all late. So a genetic predisposition doesn't necessarily indicate an early onset. Therefore, what I would say is if there is a positive family history, only then you go for genetics. Otherwise you don't.

It's as simple as that. And that's what I do. In my. My series, AVE is the common cause of dizziness. We see this on CT scans.

So what is ave?

Not sure. Anna, what is ave? To be honest, I have no idea. But we can ask to Dr. Nova. Okay.

Because I can see her live. She's attending. So can you please tell me what AVE is?

Because abbreviations are all different in different countries. I'm sorry. Sure. Okay. In the meantime, we can go home.

Yes, Professor. Yes. Yeah, I read about congenial cmv. Yes, As I said, you have got a good point. Any infection can cause mess up in all the secondary things.

So in our series, I think there was one cmv. CMV means non syndromic cmv, which is only hearing loss who presented with a phenotype. But there is hardly any evidence of, you know, kind of who has studied meners in. In. In cmv.

So yes, it can most certainly cause it. Because I did say any infection. In fact, starting from herpes to any. Any viral infection can cause it. Let me google AV and see what happens.

Wait, let me just see what AV means.

AV. Okay. Dr. La Nova has just reply. Professor. Yeah.

Wide vestibular aqueduct. My God. Professor, may you be so kind to open the Q and A box. Okay, you can see hitting the zoom lower bar on the screen. Q and A. Yeah, I don't have any Q and A.

Wait, no more. There is a more. Yeah, I've got a Q and A. Fantastic. Okay, if you scroll down at the end of the bar, you should see the answer from Dr. Anna Latinova.

Well, Eva, I know, but what is Ave? It's the other way around. Maybe it's the language. Enlarge vestibular. Okay.

Enlarged vestibular aqueduct is the most common cause of dizziness and hearing disorders. Once again, with due respect, I do not agree with. Vestibular migraine is the commonest cause of dizziness. Vestibular migraine. If you let aside vestibular migraines, then the commonest cause of dizziness and hearing disorder is very likely non genetic syndromic conditions.

And EVA is almost always. Well, it can happen in an isolated way, but it's strongly written 30% of cases with pendrin but functional enlarged vestibular aqueduct where there is actually no evidence of an enlargement of the bony vestibular aqueduct. No, because the whole Menier syndrome spectrum is limited to an expansion of the membraneous labyrinth, not of the bony labyrinth. So I don't think that you can say the expansion of an endolymphatic fluid chamber in case of a menus is actually a vestibular aqueduct syndrome. It's not.

That's. That's according to me. And of course you can differentiate it from many years disease. Because EVA classically will present with a third window in the early phases. Many as presents with third window, but the EV A third window is far more common.

And EVA is mainly diagnosed by the type of hearing loss and of course the vestibular function tests.

Thank you, Professor. We have received a lot of questions. I kindly ask you to pick up some. Right. Okay, Mehmet, 20 minutes to 12 hours.

How is it possible for rust membrane rupture to resolve within such a brief period?

Right. This is a very complex answer. The RUS membrane, how quickly it repairs itself is unknown. We do not know. So it can repair itself over 12 hours or it can take longer, we do not know.

But the reason why it recovers in the initial period is something completely different. And this is because of the enzymatic cascade which leads to generation of reactive oxygen species in the cochlea and the vestibular system following any assault or damage to the system. That system has got a potential to recover within 72 hours. Which we have said, which we call a normal human asymptote. So consider this.

There's a rupture, there's a toxic environment created. Reactive oxygen species pour along the whole thing. When Reactive oxygen species pour along the whole thing. There must be oxygen salvage radicals as well. There's a perfect balance.

As long as the salvage radicals can counteract this extra ros, the system recovers. If they don't, the system doesn't. In many years, in the early phases, the system does recover. So that must mean that the ROAS is actually being counteracted by the salvage radicals. And as I said, the rhizomes membrane, even if it doesn't repair by 12 hours, I'm sure that it would repair in days.

There is no at this, I don't know of any evidence. So yes, initially the system tries to counteract the system and then the system plugs the hole. The whole thing is, the whole thing recovers. It's important both with vestibular migraine and with many years, both the 72 hour period is critical. If the system can repair itself, it has to do within 72 hours.

That's why in many years in children, if there is a sudden drop of hearing, then in some instances you do need to see them urgently and consider steroids. That's the important thing. But unfortunately you don't get them like that.

Well, the next one. Paolo Jorge, as I said, I do not believe in ECOG because this is very, very resource intense and actually invasive. Not good for pediatric population. For adults it's a different, different kind of possibility. And once again, calorics.

I wouldn't use it in children, but adults you can do it, but I wouldn't do it in children at all. Glycerol test in children. Absolutely no role. Anything which is unfriendly to the child. No role, no go.

So huda falil, you use vamps now why do you use vamps? That's my question. Why do I use WIMPs? The thing is, the other thing is do you use, if you are using WIMPs, do you use positive frequency

tuning curves?

Okay, so I see the next part of your question. So okay, I now get an idea as to why you use WIMPs. Yes, WIMPs are very important for vestib for at all stages. Now what I would say is my experience so far and what the literature suggests is the CVEMP tuning and the OVM tuning, they are deranged when the disease is active active or quiescent, which means an attack can soon happen. In other words, it's not the chronic phase that you don't have any attack.

So if you get a CVEMP or a novemb tuning positive and if you see that the VEMP itself is also Deranged, let's say low, abnormal and low amplitudes. Or for example, say, let's say that they are asymmetrical, one side is weak than the other. Then the disease is active or quiescent. It's not the chronic. However, in the very early phase, your CFM tuning might be intact, but you will classically find the third window picture which is high amplitudes and low thresholds.

I'm sure you have found them. I'm sure you found them as well. So yes, CMP is very important. Gadolinium.

Well, I am not very clear about what happens to gadolinium in a child, but as far as I know, my radiology colleagues are quite skeptical in using anything more than the recommended dosage because they have led to different kind of kidney problems, including acute renal failure. I do not know exactly how long it stays, but yeah, it is a problem in children and no studies have been done. So you know, I'm not going, I'm not going to venture this at all. Okay, what dose betahistine. Betahistine in this country is licensed from the age of five or six.

You start with a very low dose, 8 milligrams, three times a day and then you gradually build it up, never more than 48 milligrams. Not from the German school who says in adults you can use 796 milligrams.

Ah, Hadas. Hello my friend. How are you?

The thing is, would you differentiate between vestibular migraine and msa? It's pretty much because, look, autonomic symptoms, they do not usually happen in Meniere. So if they happen with a Meniere phenotype, then sometimes vestibular migraine and vestibule and many years, many years disease, they merge together. So always keep a look for, look out for autonomic symptoms. For example visual symptoms, for example photophobia, phonophobia.

You know, in photophobia and phonophobia they have been reported with meniers, but it's not very common. So yeah, autonomic symptoms with a menus phenotype, very likely there could be an added vestibular migraine.

How best can we differentiate ANST with the fluctuating hearing loss? That's an absolutely magnificent question. So ANST is associated with. Look, menier is mostly, mostly vestibular because that's how the first thing was diagnosed. So ANST hardly presents with vestibular classical dizziness or vertigo.

ANST might present with balance problems. Yes, but yes, fluctuating hearing loss is a differential diagnosis. Absolutely. But in fact, but in case of ANST or stapidial reflexes will be absent in 82 to 86%. So always use stapedial reflex when you are investigating someone with Meniere's.

So in our, in our threshold is 40 for 40 decibels NHL. So 40, 45. I would say 45. We start at 60, then go down to 55, then 50, then 45. If your thresholds are maintained at 45, that's our lab norm, then that is a reduced threshold.

Positive beta Isin. I told you the dose. Oh, this is my. This is my colleague.

This is such an important point, Sudhira. That's exactly why I'm so happy so many people are joining this workshop or in this, this webinar that we need to tell our pediatric colleagues or other medical specialties that Meniers does have a cause to explain the Menieres, which is a systemic cause. It often gets missed and they end up in chronic fatigue syndrome. They end up in camps. So that's the reason why we are doing this.

So REM always do a full work workup for many years. No stones unturned, no shortcuts, no role of glycerol test. Invasive gluten ataxia. Good question. That kid, the one with the celiac, he also had a little bit of gluten ataxia because he had some gaze book nystagmus with rebound.

So yes, gluten ataxia, but the name suggests itself ataxia in many years in children you usually don't get a tumor can crisis which you get in adults. But in children you also don't get this typical ataxia. And gluten is cerebellitis. So this cerebral ataxia is absent in many years, but it can coexist at that kit. For example, how do you titrate the betahistine?

Betahistine, remember, is one of the safest medications around. So you can actually, you'll have to take regular updates. Our protocol, Sudhir and mine is every two to three months we telephone the children, telephone the parents and take an update and then accurately try to manipulate the pharmacological intervention. We might have to add new drugs and well, when the spells become less and less and less, then you start weaning the betahistine and finally stop it. And it's important to understand that the children usually do not reach the burnout phase.

Burnout phase means no hearing, no vestibular function on one side, all symptoms gone except the hearing loss. But the balance symptoms are gone. The children in our series, at least about 30, 35, we haven't seen a single burnout as yet. And some of them we are, we have been treating for quite a long time.

Okay, well the last one, 8mg TDS. Well, yeah, we start with the 8mg TDS and then take regular updates. It's variable. We do not know when the but majority, you know, up to the age of 13 or 14, they would respond to 8mg tedious. It's a fantastic drug.

Yeah. Okay. Professor, I sincere thank you for addressing such a rare and complex topic as many heart diseases in children. Let me say professor, this is a subject no one else has explored in five years of inventis academia. So let me say that your contribution has truly enriched our scientific journey.

Thank you once again, Professor. Thank you so so much for once again. No, I always thank you because otherwise I can't reach so many people. This movement has started. South Africa is even thinking North Africa event.

So yeah, we need to tell people that this is real, it exists. So please don't ignore the kids who are dizzy and who have got great difficult balance. That's the most important thing. Thank you Anna. Thank you everybody for this occasion.

I'm really very honored and I'm very grateful for all of. For all those who have attended because it only shows that the interest we are kindling all over the world is paying off. Thank you. Yes, last thing. I would also like to remind you all that the second session with Professor Dasgupta will take place later this year.

But our next academia webinar is scheduled for March 18th featuring Dr. Silvette Wiener Vache presenting why and how to test the vestibular function in hearing impaired children. So thank you all for attending and for being part of our academia community. We look forward to seeing you you at the next session. Have a thank you for evening. Bye.