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Third Windows in the Pediatric Population

Presenter: Soumit Dasgupta, MD

Greetings everyone. We extend a warm welcome to each one of you and express our gratitude for joining us. Today, inventis is privileged to introduce the third and final lectures of the series, featuring an internationally respected expert, Professor Soumit Dasgupta. Professor Dasgupta serves as a consultant, audio vestibular physician, neurotologist and clinical lead in pediatric audiology at Alder Hay Children's NHS Foundation Trust, Liverpool. We sincerely thank Professor Desgupta for accepting our invitation, but we are also delighted to have with us doctor Andrea Castellucci from Reggie Millia Hospital, a longtime friend and colleague of Professor Dasgupta and a member of the Vestibular Italian Society.

Andrea, thank you very much for accepting the academia invitation. So the title of today's lecture is third windows. In the pediatric population, vestibular disorders can significantly impact children's lives. Often going undiagnosed. With special expertise and a deep understanding of the inner ear's anatomy and its connections, Professor Desgupta will shed light on the often overlooked phenomenon of the third windows, a topic that has seen rapid research advancement since its discovery.

We encourage you to jot down any questions in the designated space on your screen and following the event you will receive a survey and we kindly request your participation because you know that your feedback will enable us to tailor our educational support to meet your needs effectively. And now I invite Doctor Castellucci and the professor Dasguchta to take the floor. Enjoy the webinar. Thank you, Juana, thank you very much. Very, very nice to be here with you.

And especially I'm very happy to introduce my friend Soumit Dasgupta. Actually, he doesn't need to be introduced because everyone knows him and his skills. I just would like to tell a few things about him. We met a few years ago in Barani society meeting in Madrid and since then I've been learning so many things from him. Especially I learned a lot of things about vestibular disease in children.

Because, you know, children do not come to your office and tell you what they have, so you need to have a holistic approach with them. And this is one of the main focus of Professor Dasgupta, holistic approach. If you want to know and if you want to understand what children have when they come to your, to your office and in particular, throwing those pathologies are tricky, are not very easy to be diagnosed, at least in adults. In adults are like this, but in adults, you know, we have a lot of symptoms and signs that we know that third windows can, can give, can read. So we have our keys to get inside the third window, pathology.

I'm sure that he will tell you that in children, third windows do not show up like they show up in adults. So you need to learn to know a lot about vestibular physiology in children. And again, you need to have an holistic approach to children if you want to diagnose this kind of rare disease that actually, they're not so rare. Probably. Probably they are a lot.

They are very, very less diagnosed, better than they are. So I'm very happy because through windows, it's one of the, let's say that it's a model, that it's a wonderful model to understand, to understand the physiology of the inner ear. So I'm sure that soumit will shed light into this kind of scenario. Big scenario, big group of syndromes. So thank you very much, Sumit, for being here.

So I will lead the podium to him. Bye.

Thank you very much, Andrea, for those kind words. And thank you very much, Anna, to facilitate this. Once again, you guys are doing an absolutely stellar job. And one thing we have learned from COVID is that, well, you can do this virtually without being physically present, which has actually brought the world closer together.

So essentially, today I'm going to talk about third windows in the pediatric population. And as Andrea rightly said, that they do not follow the same trajectory as you would expect for a third window in an adult. And the main reason is because a child's vestibular system is developing. And when I say it's developing, endolymphatic fluid metabolism and endolymphatic dynamics, that is, the flow of the fluid, is pretty different from that in adults. And a typical example of this is manifested by the way a child responds to a head injury.

So, yeah, it is quite fascinating and quite different. You know, when I first started in pediatrics, I knew about adult third windows, of course, because that's a very interesting condition. But I had no idea what I was stumbling into. I remember my first pediatric third window was so interesting that it got me hooked. And at the moment, I think, if I'm not mistaken, we have got one of the largest series of third windows in children.

I frequently get asked this question, how come you have diagnosed so many kids with third windows? As Andrea said, quite rightly, it's a tricky diagnosis. I said, well, you need to have a very, very low threshold. So that's what I'm going to tell you today. So hopefully, at the end of the lecture, you'll find out that this condition in children poses a diagnostic challenge, and it is a condition which has got a significant morbidity, not from as much from the physical point of view, which is very common in adults, but from an emotional point of view, because, you know, all those very, very exotic symptoms like gaze evokes, editors, conductive diacousis and all these things, they often get dismissed by the parents, by the school, by everybody.

So it takes a very strong, because the child suddenly finds that the child is not believed, which is a very, very dreadful thing to happen to a child. So at the end of the day, we need to understand the condition and we need to diagnose it.

So a very brief look into the pathophysiology. You know, you all know about this, I'm sure. What is a third window? Now, let me tell you guys that when I first started training, I took a long time to understand the pathophysiology, because if you, the moment you understand what happens, it's very, very easy for you to follow the disease trajectory. So you'll have to know what really happens with a third window.

So obviously, as you guys know, that there are two natural windows, the oval window and the round window, which is how our ears are created. The oval window is very important for preferential sound distribution, which is when the sound comes and hits the drum, transmit it through a circular vibration all the way to the stapes foot plate. When the middle ear does, all its beautiful transforming mechanisms, adds in a lot of energy so that the energy doesn't get lost in the fluid air interface at the oval window. And it transmits, the oval window transmits the sound to the cochlea, right? So that's what we call preferential distribution.

So the majority of sound is conducted like this. A part of the sound, however, is not conducted through the oval window, but it's carried by the ossicular inertia. The ossicles are bones, remember, they're attached to the skull, so when they vibrates, their vibrations are transmitted to the skull. The skull vibratess, which directly can generate a cochlear traveling wave, but the majority of the distribution is through the oval window. So once the oval window starts vibrating, the energy then sets up a cochlear traveling wave, which traverses through the whole length of a cochlea from the base to the apex.

And as the traveling wave spreads, it creates these undulations in the basement membrane, which is the very basis of outer cell activation, starting at the tectorial membrane. I think you will know all this. But remember, as the cochlear traveling wave trans moves from the oval window to the round window, it starts to lose its energy. So when it reaches the round window, the SPL, the sound pressure level, has dropped. So

in other words, the cochlear travelling wave travels from a zone of high SPL, the base, to a low split towards the apex, but it doesn't lose all its energy.

Now if this energy is contained within a highly delicate membranous cochlear duct system, then there is a high chance or a risk of the membrane rupturing. So there has to be a mechanism by which this excess energy is dissipated, and that is done through the secondary tympanic membrane in the round window. A physiological effect we call the tympanic baffle. That's how the cochlear traveling wave. If you'll have to know anything about the physiology of hearing, I think cochlear, the concept of cochlear traveling wave is the most crucial attribute that you need to know.

So that's the two windows. If you add another window, and that's our classical third window. Now that is a pathological third window. However, in the natural system there are a few areas which might act as third windows as well. It's spread all over the inner labyrinth.

However, these third windows, they are not pathological because they do not cause a functional effect. A pathological third window is when there is a functional effect which is felt, which is perceived, and it alters the endolymphatic fluid movement and pretty much everything else. So these are therefore conditions which are essentially a communication where the communication should not happen. You see, the oval window is a communication of sort between the inner ear and the middle ear. Now if you get a third window, which is on top of the oval window, a pathological connection that becomes a third window, or on the other end, more towards the brain, if there is a connection between the cranial cavity and the inner ear, that becomes a third window.

Now remember, there is a chance that in a third window there is a direct connection with the perily lymph, because perilymph is a typically brain fluid. So either inner ear cranial cavity or inner ear middle ear abnormal connection, that we call a third window.

What happens here is quite exciting because a pathological third window has got even a lower spl than that of the round window. So the cochlear traveling wave faces the path of least resistance. And that has got some fantastic and very, very interesting effects, which was first identified by Lloyd Minor in 1998 with his first series of the prototype third window, superior semicircular canal dehiscence.

And the third window accurately was described in 2008 by Samil Merchant. Both these papers I shared, both these papers are available online and they are open access. So because these are such seminal publications, they have changed the face. I mean, identifying a new disease, that takes a lot of vision. So Lloyd Minor can be justly said as one of the giants, one of the vestibular giants, just like Michael Halmaji and Ian Kurthoys.

So the first time that is identified was the superior semicircular canal dehiscence. So here is an animation of what I've just told you about tympanic baffle. Look, that's the oval window and that's the round window. So you see the dissipation of energy, and then this energy is transmitted through the cochlea as a cochlear travelling wave. So what happens when you subject this third window situation with a sound?

Let's say you are doing a pure tone audiometry when the sound travels through the middle ear, because the third window is actually a bodhi abnormality. Remember the preferential distribution, which I've talked to you about? I told you just now that preferential distribution is lost to a certain extent, because the third window absorbs that same sound which goes through the middle ear, through the vibration of the skull, because simply the third window vibratess as well. So the preferential distribution is lost. And because you lose the preferential distribution, that is manifested as a conductive hearing loss in your puritan audiometry, this is actually not a real loss.

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There is no disease in the middle ear, so it's not a real loss. It's rather what we would call a gain rather than a loss. So this is how the energy is dissipated through the bony apparatus, middle and inner ear. And then once the cochlear traveling wave forms, due to any reason, either due to direct ossicular inertia or bony skull vibration, or the classical oval window interface, that cochlear traveling wave then travels from the oval window to the third window and not to the normal and natural round window. So what happens?

It therefore flows from a zone of high spl to a zone of even lower spl, lower than the physiological SpL. So it's easy to travel. You know, it is less resistance pathway than the cochlear traveling wave faces. As a result, when you do a hearing test, a pure tone audiometry, the bone conduction improves. Now, is this because the cochlea suddenly gets some?

Is this because there is something which is happening in the cochlea which is improving your hearing. No, it's a spurious hearing. And this gain is also called a sensory gain or a cochlear gain. The net result is you get an airborne gap. Now, airborne gap is not specific for third window because you get airborne gaps in so many different middle ear disorders.

Right. But inner ear, third window is the only inner ear condition which generates an airborne gap, like a conductive hearing loss. And if you haven't already read, then Andrea and I, we edited a wonderful research topic in audiology research. We have got how many papers now? Maybe five or six papers and still coming where we discuss this, where researchers from all over the world, they have discussed this phenomena, inner ear conductive hearing loss.

And the moment I learned about this immediately, all my children with middle ear disorder, typical conductive hearing loss, which I cannot account for by a middle ear

disorder, I subject them to the full third window algorithm, which we will come. Now, this thing goes on. Yes, of course. But remember, at least in a child's brain, maybe not as much in an adult brain, which is far more robust from the anatomical and from the physiological point of view, in a child's inner ear. If this thing is continuous, then there is an effect on endolymphatic flow.

And when that happens, there is a permanent and irreversible damage which can affect any part of the inner ear labyrinth or cochlear. You can get sensorial hearing loss, you can get semicircular canal deficiencies. You can get utricular circular deficiencies, because that's when the function is compromised because of a chronic derangement of endolymphatic flow metabolism and of dynamics. And there is evidence to this effect. And you'll be surprised to know that early endolymphatic hydrops in both kids as well.

In adults, early Meniere's disease, they might act as a temporary third window as well. It's just the sheer volume of the endolymphatic fluid which causes this. So how common is this third window in the pediatric population? And you can imagine that we do not have many studies because this is not diagnosed to the same extent as that in adults, for the simple reason people are not on the lookout. You know, if you want to diagnose something, you need to actively seek for it.

And I've got a wonderful video just telling you the thought process which will make you seek for a third window. And here, I might add, that there are a group of people in the world, in fact, I would say about ten to 15, who we will call ourselves tri fenestrologists third window study group. So there's Ashley Wakim, there's Marco Mandala. Andrea, I think, will be joining the club soon of being a fenestrologist because we are passionate about third windows. And we pretty much see third windows everywhere.

You know, if anyone comes and there is a reason for it, guys, you know, because the original Johns Hopkins team with Lloyd Minor, who identified the superior semicircular canal decence, there's a huge expertise out there. And one of the other experts is, who still works in Johns Hopkins, I think Simon Carey. Now, Simon Carey says that third window is a condition which mimics any autological condition. So you get a situation where, you know, you think it's something which is not quite right. That might be actually a third window.

So it's very cryptogenic. So if I might digress a little bit, what is cryptogenesis? Cryptogenic? You have heard about a thing called cryptozoology. Right now, cryptozoology is the study of mythical monsters.

For example, the loch ness in Inverness, for example, the Bigfoot in America, the yeti in the himalayas, the Mokele mobembe in Congo. And I happen to be part of one of those groups. I remember the first time I went to Loch Ness in Inverness in Scotland, in a place called drumnad rocket. At that time, I went with my wife. We didn't have kids.

And I spent the whole night by the banks of the loch ness trying to figure out where the monster is. And my wife wasn't very happy. But this is something which we follow with passion. And there is only one mythical monster which has been identified so far. That's a tasman tiger.

There are skeletons found. Third window is like that. It's there somewhere lurking. We just can't prove it. And that's why it doesn't get proved.

That means that you pick up less and less unless you've got a completely open mind and follow an algorithm. So, in the adults, excluding endolymphatic hydrops, which act as third windows in the beginning, is 0.1% of the population in adults. So it's not terribly, terribly common. But if you calculate the entire adult population in the world,

it's a sizable number in children. Non syndromic, which means de novo, semicircular canal dehiscences.

That accounts, you know, about 1.2% out of 649. So semicircular canal dehiscences. But that was a study in 2014 where the other third windows were not identified. And the commonest pediatric third window is an enlarged vestibular aqueduct, which we call in this country, dilated vestibular aqueduct. So I'll call it EvA.

There is one third window which we have recently published in the same research topic. It about x linked gusha. About 90 kids have been reported, including mine, I think about 92 to 94. And others are extremely, extremely, extremely rare. So it's rare.

But I might say if you have got. If you've got the kind of the attitude to pick it up, it becomes not frequent, but not infrequent either. And I'll give you an example later. So these are examples, and these are examples which have been discovered by absolutely superlative experts. So obviously the prototype is the Lloyd minor one, which is superior semicircular canal dehiscence.

But you can get deheses from all canals. And in fact, the lateral semicircular canal dehiscence can occur after chronic separate buttitis media or longstanding middle ear disorder. That's a typical fistula, really. So where there are many, many bony canals in the inner ear, you get a carotid canal, you get a facial canal, and dehiscence might occur in any of the canals which will generate a third window effect. XIN Gasher syndrome is very interesting because this is an incomplete partition of the cochlea, which opens up the inner ear to the cranial cavity.

There. The modulus is missing, part of the modulus is missing, so that creates a third window effect. In fact, this was previously called perilymph gusha. You know why? They were diagnosed as otosclerosis.

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And the moment you try to put a piston in there is a huge perilympic gusher and the patient loses the hearing. So this started as a perilymph gusher, but now it's an x linked. It's a genetic mutation, x linked recessive genetic mutation called Pof 34. Perilum fistula is a very classical example of a third window. So all this now, one of our colleagues from Paris, from Lyon, actually, Eugene Anesco, a very good friend, we started talking in one of the meetings and he said that I have found out a third window, which I think a third window, because it.

It kind of manifests as a third window, is a dilated internal geometry. So I told him, that's very funny, because I've got about two to three kids where I have excluded everything else, but they present with third window features and they have got dilated internal mediators. So he and I, we are now thinking of collaborating to do a series. He has already done a series, I think he's already published, and that is very interesting. So dilated or internal geometers, how does that create a third window?

It can either be because the membranous labyrinth is so much kind of voluminous out there, because, remember it, the membranous labyrinth size is proportional to the bony labyrinth here. The bony labyrinth is huge. So the increased fluid volume, just like an enlarged vestibule aqueduct, increased full volume or a nola fatty hydrox can clear third window effect. Or there are subtle fistulae because of a dilate internal mutism between the internal mutism and the. And the brain, essentially the cranial cavity, which creates a third window effect.

Hypermobile stapes footplate is a new third window discovered by our very good colleague, Professor Arun Ghadre from Danville. And he gave us a talk last year which was absolutely phenomenal. And he narrated how he came across this hypermobile stapes footplate following a head injury. And he discovered this new. So you guys who

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are watching this, maybe one day you are going to discover a third window which would be phenomenal.

Well, otosclerosis, when you get a cochlear atherosclerosis, there might be microfistula between the cochlea and the cranial cavity or the middle ear even. And that might lead, for example, if there is a otosclerotic focus in the fissile ad fenestrum, which extends to the posterior fissilar, anterior to posterior, then that might create or open up a third window effect, which has been reported. So there are loads of third windows. The ones which are in red. Those are the ones which we have found in the pediatric population in our series.

So how does it behave? What is the natural history? So it may be congenital or it might follow other anatomical abnormalities. For example, high riding jugular bulb can erode the floor of the posterior semicircular canal. Various venous sinus abnormalities.

For example, the sigmoid sinus can generate a problem with the superior semicircular canal. It can be syndromic. It doesn't have to be de novo as you find in adults. It can be a part of pen red charge. Branchia otorenal.

We have reported a superior semicircular canal dehiscence on a Down syndrome kit. And we are about to report a posterior semicircular canal dehiscence on a DFNB 22, which is a non syndromic autosomal recessive hearing loss with a posterior semicircular canal. Dehesins. We have also reported a superior semicircular canal dehiscence in a very interesting cytokine mediated mutation, which is called Med 13 L. That's also available.

I think. I think, if I'm not mistaken, it's open access or sometimes we don't find a reason and they can be late onset. It's not essential that the child is born with it

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because, you know, the skull is continuously molding itself. You know, it's developing osteogenesis, osteoclasis. This process carries on until the age of at least 16.

And because it molds itself, there is a chance that the third window, because it's a bony abnormality, also changes over a period of time. You know, it can get better, it can widen up, it can get better, or it can stay dormant when it manifests again at the age of late twenties to early forties or even fifties, when you get a classical adult. Classical adult syndrome. So, very importantly, remember, the skull molds itself. And that is key to understand why a pediatric third window is different from adults, because, remember, all third windows are bony.

And very interestingly, in bony deficiencies, especially superior semicircular canal, if the dural plate is breached, then that is a bit of a problem because that not only exacerbates the symptoms, but it might also affect vestibular function, and that is very important. This was also from the Johns Hopkins team by Brian Ward. And we have in our series found out a traumatic third window in a child. So a child with a traumatic third window, a third window occurring after a trauma, so important points to consider in children. And this is where it's absolutely crunch, because we need to understand, why is it so different in children?

We have mentioned this. We have mentioned this, so the clinical features will differ. As Andrea said before, in an adult, it's not difficult because the adult will tell you classically that look, you know, I can hear my eyes move, I can hear my footsteps. You know, I don't like sounds because sounds make me dizzy. But remember, seven, eight year old kids, they wouldn't be able to tell you all this.

And the parents, you know, what exactly the child feels, they are very good in picking up this. There's something wrong, but it cannot pinpoint what is wrong. So it is your

entire clinical acumen to bring out this history from the parents. And that's not easy. You need to have a very solid pediatric experience.

I mean, I spent three years in pediatrics when I was training. And this pediatric experience is important because we need the insight to figure out a parent's communication. You talk to millions of parents and you know what they are talking about. For example, change of facial expression when a child loses balance is a mark of a dizziness in a child. Child won't tell you I'm dizzy.

Mom will tell you when he comes down a slide, when he goes to a swing, his facial expression changes, and that is a sign of dizziness. This comes with years of experience. And as I said, you know, it's not that if you have an practice where you see both adults and kids without any pediatric training, then you might miss things. And that's so important because I don't think, Andrea, anywhere in the world, ENT training. I have been an ENT trainee myself before I came to neurotology.

I don't think anywhere in the world, unless I'm mistaken. I might be mistaken. Forgive me if I'm mistaken, but ENT training does not include dedicated pediatric training with acute pediatrics. I have seen pediatric patients with meningitis, running influence. I was awake all night treating a septic child.

This is important, believe me, this is very, very important to understand not only the child, but the parents. Otherwise you wouldn't understand anything of child doesn't talk back to you. Very difficult to get a history and classical features when the child can actually tell you what's going on. That comes much later, round about the mid to late teens, because there they can actually describe to you what is going on. They become somewhat adult like yet because they tell this to their parents, to their schools and to everybody else.

They are not believed because they are so interesting symptoms. They are not just, I am, I can't walk or I'm having a hearing loss. They are far more than that. So these are very, very important points. And you can actually physically demonstrate that the vestibular system is normal or weak with all the beautiful tests that you have.

But remember, if you're seeing a child, you must factor in the point that their vestibular function may be deranged. That's because of altered endolymphatic fluid dynamics. There's one fluid which is circulating through the whole inner ear labyrinth, cochlea or vestibular system. That's the beautiful ducts and duct tubes that might get affected because it's a developing system. It's not a developed system as you get in adults.

And that's a crucial point to understand. So whatever symptoms you can glean from the parents or from the kids themselves, hearing loss, of course, they might tell you that my hearing loss is on and off, fluctuating not all sounds will create a third window effect. When it does create a third window effect, the child doesn't hear and then the hearing comes back unless it goes to the permanent stage. Vestibular symptoms like dysequilibrium, conductive disaccusis. So conductive dehiscence means magnification or amplification of sounds generated by the body.

For example, when you are walking, you usually don't hear your footsteps, but in third windows you can because the bony skeleton starts to vibrates. When you walk from your feet, the vibrations come, they stimulate your third window because remember, the bony skeleton is one continuous thing. So, and in a child, often this happens as a misophonia, distorted sound. So not only body generated sounds, but also simple, innocuous sounds, which can create a third window effect, quite distressing for the kids. And they perceive this not as a hearing loss, but as a misophonia.

For example, I've had kids who tell me I can't sleep with my brother in the same room because his breathing drives me nuts. I can't sleep because I can't sit on the dining

table eating with my mom and dad because the noise of eating, it drives me mad. And when the child can't say this, your parents will tell you he doesn't sit and eat dinner with us because the moment we start eating, he lives, he runs from the room, he avoids the sound. This leads to avoidance behavior. Gaze evoked induced is very interesting, doctor.

I can hear my eyes move in my head. Extremely distressing. And I've seen adults, you know, driven to the point of being really psychologically upset with this autophony. Now that's very interesting. When you speak, your larynx vibratess, vocal cords vibratess, and then the vibrations are transmitted to your mouth and the mouth is a bony skeleton.

That vibration goes all the way up to your inner ear, stimulates the third window, and you get an amplification or resonance of the sound. So you don't like your own voice. You just don't. You hate it, actually. And I've seen executives can't go to board meetings because they don't want to speak.

Their voice just doesn't seem so bad to them, so they don't want to speak to avoid it. You similarly positive intra abdominal pressure. You know, that, that kind of, if you are, if you are breathing against something or if you are coughing or you're sneezing, that might stimulate the third window and you get dizziness or dysequilibrium, you might get loud sound, which is classical tullio third window, where loud sound, because of its spL, it stimulates the third window and generates a dizziness. You might get pulsatile tinnitus or even sometimes oscillopsia. Pulsatile tinnitus can be central.

If it's peripheral, it's always a third window. And remember that. And Andrea, I think that's a very useful project for us. And the pulsatile tinnitus, inner ear causes of pulsatile tinnitus, that would be very, very useful to study. So that's the other thing, which is very important.

Pulsatile tinnitus can be found. The other thing, which is very interesting, and I'll show you a video, is a pulsatile nystagmus. So you see downbeats or upbeats, which pulsate with the heartbeat, which is synchronous with the heart. Because remember, the heart beats the excursions of the heart, systole and diastole, they are actually pressing against your chest. And what is a chest?

It's a bony skeleton. Right. So through your ribs, that might stimulate the third window. So all these are very, very exotic symptoms. You know, these are quite out of the mill, extraordinary symptoms.

And that's why in kids, at least not in adults. In kids, no one believes them. And you can imagine the kind of effect it has on a child when no one believes the child.

So how do we pick up a third window? There are objective signs and you can jolly well do them in kids. Henneput sign. Hennebert sign is increase the middle ear pressure by any means, sigil valsalva, trigal pressure to stimulate the third window. And you see a classical nystagmus appearing.

The direction of the nystagmus tells you which canal it's localized, but you also get in endolymphatic hydrox or menieres as well. So that's a Henny bird sign. Let me just tell you briefly the difference between a Hennebert sign and a fistula sign. A fistula sign was described by Pulitzer in the 19th century in a perforated ear drum. So a fistula sign indicated perilymp fistula in chronic middle ear disease.

However, Hennebert in 1912 discovered that without perforation in the drum, with a normal tympanic membrane, you can elicit any birth sign. And he attributed this to syphilitic otitic labyrinthitis. I'm sure there was some third window going on here. You do a mastoid vibration test. It is vin vibration induced test.

And you might get any stigmas. Now, video head impulse test in third windows, the evidence is emerging. We have done lots of work in kids, and our colleague Andrea has done lots of work in adults, and this might be deranged if there is a fundamental vestibular weakness, which does happen in kids, not as much in adults. And once again, it's attributed to endolymphatic fluid mechanics. Vemps, of course, are the King cornerstone.

That's how third windows were originally recommended to be diagnosed. Webs is the classical hyper response, but now more important, it is a hyper response, but now more importance is placed for low thresholds rather than high amplitudes. And therein lies a problem, because threshold studies in children are quite difficult. Remember, this active movement is a lot of strain, cWMP, I mean, for the kids are looking up for an ovum response, the children might not like it, and they get fatigued. So once again, it's difficult sometimes to get thresholds in kids.

But if a child has a third window, then vmps are deranged. Either they can be with huge amplitudes or they can be asymmetrically weak. Weak is indicated that this is going on for some time. So if the autolysis are fundamentally weak, they will not return a hypersignal, so expect anything. But what we have found in our series is WIMS are always abnormal of some sorts in a third window, and PTA puritan audiometries with variable loss.

In children, we have some quite as few sensorineural hearing losses. Even in non syndromic third windows, we have found sensorineural hearing losses, especially the dilatatory internal meters. So any kind of hearing loss you can find quite different from the adults, where you always find a low frequency conductive airborne gap in children, of course you can find it. But remember, sensory hearing loss does not exclude a third window. That's my point.

And we have identified syndromes like this.

So children present with conductive diseases, pulsatile, tinnitus, if they can describe, and all these kind of things which you see in the first bullet point history is often from the adult, from the parents. Final diagnosis is HRCT. And here I'd like to introduce a very interesting thing, which is called a CT negative third window. I'm sure, Andre, you have come across this. This is now a recognized entity.

You get all the features of third window symptoms, and you get a vamp derangement. You might get a VHIT derangement. However, when you do a CT scan, there is no third window, there is no frank dehiscence. So in that case, they are calling it a CT negative third window. And we have identified in our pediatric group this thinning has to be less than 0.4 millimeter to call it a thinning.

So, thinning. Thinning. Dehiscence. A thinning, not dehiscence. Dehiscence is not thinning.

So a thin roof of the superior semicircular canal or a thin floor of the posterior semicircular canal can generate exactly the same phenotype, but they are negative on the HRCT. Would you be brave enough to attempt surgery? Some people do and some people do not operate if CT is negative. So this is a thing which is being debated hotly, but the emotional reaction in children just cannot be overlooked. It's very, very, very important.

So how do we diagnose in children here? As I said before, I'm going to give an example that it's cryptogenic. You know, you need to be very careful as to what you hear. Every important. Every important piece of communication from the parent or the child is important.

So this is from a story by Arthur Conan Doyle featuring Sherlock Holmes. It's called Silver Blaze. Silver blaze was a prized race horse in Dartmoor. He was betted, or he was supposed to win a race with lots of money. One evening he was kidnapped and his carer, Stryker, was found dead, battered to death.

Head was smashed. So Sherlock Holmes was called in to investigate. And this is what was the communication between Sherlock Holmes and the local inspectors? Just listen to this. Is there any other point to which you wish to draw my attention, Mister Holmes?

To the curious incident of the dog in the nighttime? But the dog did nothing in the nighttime. That is the curious incident. No one noticed it, right? There was a dog in the stable when this thing was happening.

No one noticed anything. But Sherlock Holmes noticed that such a big thing happened at nighttime inside a closed stable. The dog did not bark. That's very odd. Now transpose this to this.

This is the story that I made up yesterday. And I loved making it up for you. Is there any other point to which you would like to draw my attention to, professor? Yes. To the curious incident of the child refusing to sing in the choir.

He loves. Many kids do not sing in the choir, professor. He does, and he wants to avoid it. That is the curious incident. And this was my 1st, 3rd window in all the hay.

Now that was a young lady who came from the mental health services because she suddenly stopped singing in the choir. She couldn't. And she told me, crying, that I just couldn't because my voice sounded horrible to me. No one believed her. Went to mental health services, an emotional wreck.

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Then came to me and I'll show you the slide. You'll be astounded. I'll show you the slide later. That's a suspense. That was a third window, by the way.

Of course, there were other symptoms, so high index of suspicion. When would you suspect in a child? Let's say you haven't got any history, but you have. Could manage some tests, a conductive hearing loss. Regardless of tympanometry, which cannot be explained by a middle ear disorder, the maximum hearing loss you get in a middle ear disorder is about 65 because that's about roughly the transforming mechanism which makes up for the refractor refractory error between oval window and cochlear.

But if it exceeds that, 50 decibels or more. 65 decibels or more. I have seen 80 decibels conductive airborne gap that is not coming from the middle ear. Mixed loss, remember, it mimics everything. Negative busy threshold.

So the third window is so efficient and such a low pressure zone that you actually get a bone threshold below zero minus five. And that's one of the criteria, actually. You need to demonstrate a normal middle ear. However, you know, we might get an abnormal middle ear, too. You might get a coexisting otitis media with effusion, longstanding otitis media with effusion.

With such a picture, right. Even if the tympanometry is flat or negative, you'll still suspect a third window. Long standing, that's the key word. And of course, varieties of balance systems.

Sensory hearing loss. Remember, third window can be part of a syndrome fluctuating. We have mentioned that rarely they will come to you with history. You might get a paradoxical cochlear involved vestibular involvement. So on one side you might get a cochlear sensory hearing loss, but the third window is in the other side.

Now, that is very classically syndromic. Disproportionate functional hearing. That's what I said. You get 80 decibels hearing loss, but the parents and the child tell you, no, I'm fine. Sometimes my hearing drops, but otherwise I can function quite well.

Disproportionate loss, because it's not a real loss, remember, absolutely crucial. And then finally you nail it with the high resolution CT scan. You might say six year old, seven year old. Do we really need to do an HR CT because of radiation effects? Let me tell you, nowadays HRCT is extremely advanced, especially with a technique called cone beam CT.

Plus the radiation which is afforded to the temporal bones, that is equivalent to a three year natural dose from environment. Does that mean that we stop walking? But even then, you do not take a chance. It's a very carefully clinically weighted decision. The only time when you'll have to do it is an enlarged vestibular aqueduct.

Why? Because that needs a quick diagnosis. Because every time there is a blow to the head, the hearing drops. You need to know whether there is an EVA, and EVA is usually a syndromic EVA, penrigs and everything else. But it can, of course, be de novo as well, as I said, and vamps are indicated in all cases.

And this is what I was talking to Andrea just a little while back, that every individual pediatric vestibular lab must have its own norms for Vhit and vampire crucial. You just can't transpose adult norms to the pediatric population because, remember, one is a developed system. One is a developing system. For example, in our lab, the threshold for cwms, we have. We are doing ovms now.

So threshold for cwms in our children is 85 decibels. Under 85 decibels at 80 decibels. If we find that's a low threshold by our standards, not in adults, we hit vertical canal

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gain is 0.6 to 0.7 in our lab. In adults, it's 0.8 to 0.9. So you see the difference, and that's a very crucial thing.

So these are our criteria, which we have formulated over the years. Just have a read. AB gap exceeding 60 decibels, normal stapedial reflexes, and auto acoustic emissions, indicating normal middle ear. But it's not exclusive. But as I said, middle ear disorder can exist.

Symptoms of dysequilibrium and, of course, when the complaint of typical third window features abnormal clamp, abnormal v hit, abnormal vng. If there is a vestibular weakness, you can pick up head shake, dynamic visual equity, and all the other different abnormalities. And you can also get a sensorineural hearing loss. But these sensorial hearing loss get. And have a normal MRI.

If sensorial hearing loss, have a normal MRI. Our association postulates that you should do a CT scan where you might unite many things, because CT is bony. A quick look at our alder Hay series. This is where I work. That's my lab and that's my hospital.

920 children we saw in three years time. We did everything possible from the audiological and the vestibular side. High resolution CT scan was done with on a 1.2 millimeter window, which was then reformatted at 0.5 and less in Stenver and postural views. Remember one thing, you are suspecting a third window, but your radiologist says there is no third window. Please discuss it with him, because unless you discuss with him and unless you tell him, please, can you please specifically look into the different canals and not just the semicircular canals, the facial canal, the carotid canal.

You'll see that your radiologist colleague will tell you, yes, there is something out there, because you'll have to marry the symptoms with the signs. And many times this happened. I frequently discuss with my radiology colleagues about this. The video

head impulse test can be localized to the same canal or involve a different canal. So posterior canal dehesin C with a lateral canal deficiency.

And that's because, once again, when there is a weakness in the system because of aberrant endolymphatic flow, so our total yield was 6.17 of 920. So roughly about 74. The commonest is enlarged vestibular aqueduct. Next, superior semicircular canal dehesins and the others, including posterior semicircular canal dehiscence, are quite rare. In the others, these are the different bony canal dehiscences.

For example, we had a facial canal. So these are the two Hennebert signs. Andre recognized the right one. I sent it to you. This is from the series.

This is Hennebert sign. This is all tragal pressure because Valsalva kids can't do very properly. So you press the Tragus and then watch the eyes as you would watch, as the hawk would watch the eyes. And you'll see there is a rotatory nystagmus, which appears, which, if beats up, would be a superior semicircular, and if it beats down, would be a posterior semicircular canal. Wait, I'll just advance this.

It's a long video.

There we go. That's a beautiful nystagmus. This was a superior semicircular canal. Adhesions. You see the up, up, tauting.

And this is a posterior one. Same maneuver, but here, you see it's moving down. And these were very trivial symptoms. For example, dysequilibrium. My, my son, my daughter can't ride a bike.

My daughter doesn't like me breathing. My daughter doesn't like when we are eating. She doesn't want me one. Mom told me she doesn't want me to brush her hair

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because every time I try to do this, he said, no, no, no, I don't. So all these very, very interesting symptoms, and that's why they're not believed but, and this is, guys, a pulsatile nystagmus.

Look, sorry for the poor quality because I was doing it with an analog video camera.

Nothing there. But look what happens when child looks down.

You see a beautiful downbeat.

And see, he said, I can hear my heartbeat when this happens.

Very interesting. Pulsatile nystagmus, some pure tone audiometry. I won't waste much time because all I wanted to show is the typical airborne gap here. And this is a mixed loss. So these are all different types of conditions.

Posterior semicircular canal, superior semicircular canal, enlarged vestibular aqueduct. This is a thinning, as I said, Ct negative. But you clearly see the airborne gap. And almost like a mixed loss here. This is an x length is a dilated iam, and that's an x length gusher.

So this was, I think, a superior semicircular canal. Dehesins, where you get. Oh, sorry. This one is first, so I'll just. Give me 1 second.

Yeah, this one was a dilated vestibular. Enlarged vestibular aqueduct. You see the coverts and some ovoids on the right posterior canal here. This was superior semicircular canal dehiscence, but with problem on the lateral semicircular canal. This was a posterior semicircular canal.

Adjacent. You see this beautiful coverts. Five out of five, this guy. Very interesting. Is the one we have discovered with the DFNB 22, the auto, a autosomal recessive mutation located in chromosome 16, I think, x linker, anterior canal, facial canal dehiscence, lateral and left posterior.

All this was obviously on the left. This is an enlarged vestibular duct. You can see the huge webs. And the threshold has been narrowed down to 50. The other side is normal.

Superior semicircular canal dehesins bilateral. Now, this is the girl that I spoke to you about. She had bilateral superior semicircular canal dehiscence, severe autophony, facial canal dehiscence. Here you see normal. And this side is lowered amplitude.

This is an asymmetry. So this is weakness, not a classical third window irritation. Excellent. Gusher again, same asymmetrical. Now, here the threshold is down to 75.

This is a classical enlarged vestibular aqueduct on one side. A useful way of noting this is the diameter of the posterior semicircular canal. Should be the same as the Vestibulo aqueduct, but here, clearly, it is larger. You need to know how to read a CT scan for third windows. And please, you know, go to one of your radiology colleagues and ask him to teach you.

I get. I got taught very early part, but I'm continuously learning for them. You need to know your scan because you are the best person to make a judgment, because you have got the insight. You have seen the patient. A radiologist hasn't seen the patient.

This is the girl I mentioned. My first one, beautiful bilateral, and she had surgery. My only case who required surgery. In pediatrics, you do not need surgery because the

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skull molds and becomes. The whole thing becomes dormant until it manifests again at a later state.

So you need to counsel them what to expect. But this kid needed surgery by a colleague down in London and complete cure. She got back to her choir singing and she still contacts me because she's so grateful without this diagnosis. Remember, she was an emotional wreck. It's one of the best that I've done in my life.

This is a high jugular bulb which has eroded the floor of the posterior canal. You can see the clearly the dehiscence. Oblique, sagittal, excellent gusher, no modulus. You get a Christmas tree appearance and a hugely dilated and bulbous internal tremetous dilate. Internemiatus, both sides dilated.

All third window features in an eight year old boy. Sea VEMP was deranged, Vhit was deranged, and this was the only thing I found. So obviously this has to cause it. And then out came Yujin, who said, you are right, shobhit. I found a few as well, because by that time, I wasn't really communicating this to many people.

I just passed it. But now I think we have got a case which we need. We are collaborating to write up facial canal dehiscence. You see that? Facial canal dehiscence.

This is the facial canal. That's the facial canal. This is breached over there. And also you get a hugely dilated internal humid thinning and clearly see, the bone is intact. There is no dehiscence.

But this is less than 0.4. And this was classical third window phenotype. This was a post radiation traumatic superior semicircular canal dehiscence. After proton beam therapy, normal one side, right side, no roof. This was after proton beam therapy.

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There was none. We have scanned her before. There was no dehiscence. So the key cornerstone of managing kids is an accurate diagnosis, because you inherit these kids from all over the place, general practitioners, pediatricians, neurologists, mental health services, everywhere, because their symptoms continue without any diagnosis. They get very upset.

The parents get upset the moment you tell them what's going on. That is the cognitive approach, which I insist you must do on all children, days are gone where intervention is only surgery or medicine. That is not the only intervention. The intervention is cognitive empowerment with information. This might sound cliche to you, but believe me, it's not.

Just ask yourself, if you get, let's say, a chest pain, you rack your brains, what can it be? Is it a heart attack? Is it a, you know, rib problem? Is it anxiety? Or is it a lung cancer metastasis to my ribs?

You ask yourself, this uncertainty drives you mad. Kids are very resilient. Even then they want to know. And when you are treating a child, you are treating the parents. So the moment you tell them that, if someone comes and tells me, like, look, you have got a pericardial effusion that explains your chest pain, my mind will be at peace.

I know what's going on now. I'm going to deal with it by going to relevant experts. But if you do not know what you are dealing with, that amplifies your disease cognition, your cognition, your thinking to disease much, much more. And that is something which we achieve when we tell the parents and the children, this is what is happening. You show them the scan, you show them the tests.

Hearing rehabilitation. When it's an irreversible hearing loss, of course it works. But remember, many of them are fluctuating. So we have devised a way of hearing giving these kids hearing aids, which is a bone conducted hearing aid. But we do it by trial and error, which means we set it conservatively.

We don't fit it just according to the hearing loss, because we know this will fluctuate and it might actually become less. Vestibular rehabilitation often works if there is a significant balance problem, which you have picked up with vestibular tests. So, vestibular rehabilitation does work because it reinforces your balance, but for distressing symptoms, which is luckily not very common in kids, and, of course, that first one is different. In our series, maybe one or 2% have got distressing symptoms where you need surgery, and surgery is either plug the third window or plug the canal, or convert the three window situation to a two window. This operation is discovered by our good friend and colleague Ashley Wakim from Rutgers, who just plugs the round window, right.

So the third window and the oval window, they act as the normal tympanic baffle phenomenon, and he has got extremely good results. But, of course, I'm not a surgeon. I'm not. I have. I'm not a surgeon for many, many years now.

So I don't have any experience. But there are varieties of ways. But surgery for kids is very, very rare because you achieve pretty much what I've shown you get a very good outcome. And there is evidence about the cognitive aspect of third windows, just because the symptoms are so, so very, very interesting. And situation counseling is very important.

All my kids, when they transition to adulthood, past 18, I give them a full list of likely third window features all, and tell them if you get any, you know what's happening, and

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if it's getting distressing, then I'm afraid that surgery will be the only option for you. So they all get this situational counseling. You'll have to do that.

So, yes, they are rare, but you do find them if you have got a strong index of suspicion. And currently our count in Alderay is about 115 over the last ten years, which means about 11.5 per year, which is common under many conditions, actually. So it is there very much there cryptogenic under the tip of an iceberg. You need to be very vigilant. And the most important thing, take home message.

They might not present with same features as that of an adult. That is absolutely crucial. So these are the three papers which we have done regarding our own series. This is the one which Andrea and I co edited. This is the topic.

So we have. Yeah, this is open access. And these are also open access because they appeared in frontiers. So this, we are doing a research now currently on Owens in children with third windows. So we have just started it.

So that will be a very interesting one. So it never forget the elusive third window. So let me show you a brief clipping which I suddenly came across. And this makes the point so beautifully. This is a famous movie called Rear Window by Alfred Hitchcock.

Just watch carefully.

So is the third window slowly sliding up the two windows? People will look because they are shut, but a third window is slowly opening up. Exactly. Happens in third window in adults and in child, and we must never, ever forget it. Thank you very much.

That's the end, I think, finished right on time.

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Thank you, Andrea. That's finished. Thank you very much. Sumit, can you hear me? Yes.

Yeah, it was great. It was fabulous. And as always, as always, we are. You go to the point. So it was actually a wonderful insight.

In the third windows in children, they are very, very not very easy to detect. But as you said, we need to be like Sherlock Holmes. Absolutely need to be like him. So. Absolutely.

That's great. That's great. So there are some questions, if you want. I can. I can tell you the questions.

So you can. Yeah, please, please do. Andrea, please. Great. So I'm very happy to have a question from Saulsan Abu Zayd.

She's a wonderful colleague from, from Jordan, from Oman, and we wrote some, we wrote a paper on posterior canal dehiscence together. So she's very skilled and she knows a lot of things about third window disease. So she's asking, I have a few cases of children with third window syndrome. In addition to the third window symptoms, they have vestibular migraine symptoms. Do you find that, do you find that in your patients?

So let me tell you, Saswan, that vestibular migraine accounts for nearly 33% of pediatric dizziness. In dysequilibrium, third windows would account for 7%. So vestibular migraine, if you find the third window child, it's not. No, t not linked to the third window. They are completely different conditions.

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However, because there is a third window, the vestibular migraine symptoms might be perceived even more. But pathologically or physiological? Not physiologically, pathophysiologically, they are not linked. They are two different conditions in the same child. Okay, wonderful.

So very clear answer. So we have a lot of congratulations. Thank you. So another question from Reem Abdullah. Does the treatment and recommendation applies also to the adults?

That's a very interesting point. No, I think, Andrea, you would probably be better to answer this, because I deal adults in only my private practice and I send them to my surgical colleagues, really, I go through the whole treatment, what I do with my kids. I tell them what's going on, show them what's going on. And many of them acclimatize or adapt to it. But there was one gentleman I remember whose only problem was, from the functional point of view, was he was unable to play golf because every time he hits the ball with the club, golf ball with the club, he was very, very dizzy and he just hated it.

That was getting in his way of life. And he had a huge web. So we eventually managed to find out a superior semicircular canal dehiscence. So I told him, look, this is your only problem. Why are you even bothering?

You are perfectly fine, otherwise, you are fine. And he said, no, sorry, professor, I can't live like this. I'll have to play golf. Had surgery. Wonderful result.

Back to golf. So it all depends on individual cases, really. But I think the treatment in adults would be different from that in pediatrics. But the general kind of empowering information remains the same, I think. So that's what I would do.

Yeah, I totally agree with you. Recommendations are the same. But you know, probably also, the expectation from the surgery is different between adults and kids. So it depends. So we have another question from Morasha Adsosa.

What results would we expect to see on the auto acoustic mission? I think, yes. Yeah. Okay, so interesting. So, basically, and there are, there are a few papers here, so.

But they're all in adults, remember? So what our experience is, if the child is a de novo semicircular canal dehiscence that is not linked to any syndrome, then the OAS should be intact. So in other words, the OAS would be normal because you do not get a cochlear pathology, and the middle ear is normal as well. However, lately, I mean, I would say about 2020, 5% of our kids are syndromic, either a chromosome abnormality or even the downs, as I said, in those cases, the sensorineural hearing loss, there is a sensory hearing loss which affects the cochlea. So obviously, in those cases, the OAS will be absent.

Great. Thank you. Thank you. Thank you for. And just, can I tell them a story?

Andrea, with your permission, when we were writing our first paper in 2018, I was severely researching into third windows, adults and children. And there I tried to find out what happens to tympanometry and reflexes in the middle ear in third windows. And I came across only one paper, which I've quoted in my paper, and that's the first thing which I told my colleague in Barani when I met him. I was introduced that you are the guy I know very well, because I have quoted your paper, and that guy is Andrea Castellouches sitting here. Thank you.

Thank you very much for this. It was actually, it was actually a great paper. The paper that you wrote about the winners in, that was our first. But that's how I knew about you, Andrea. You are famous.

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Yeah, it was very great, even, because I think it was one of the first who tested kits with third windows with the video heading pulse test. I think it was the first. Yeah, it was the first. It was great, actually. So another question, how bone conduction improves in third window syndromes.

I did have a slide explaining this in detail. It's essentially the cochlear traveling wave travels from a zone of high spl in the oval window to a very low spl in the third window and not to the round window. That's why it retains some of its energy. And as it retains some of its energy, there is more energy transmitted to the basement membrane for outer hair cell activation. And that improves the hearing threshold in the bone, not the air in the bone.

There was a slide. I think it was my 7th or 8th slide, where I've explained. Yes, that's the third window effect. That's the third window effect, yeah. Great.

So another question from Abdullah. Does hearing aids helps in case where the patient has mixed hearing loss in order to diagnose with the windows? Now, that is a very, very difficult proposition. Thank you for asking, because we find this problem. See, a mixed hearing loss in a third window can mean two things, that the conductive element is due to the third window and the sensory, not a neural, the sensory hearing loss.

The cochlear hearing loss is due to a problem in the cochlea or because it's a syndrome. Now, because the conductive element fluctuates. You see, fitting hearing Aids is mighty difficult because there might be a rejection, the child comes back or mom comes back, says that it's too loud. So, as I said, yes, they can be fitted based on individual cases, but your audiologist must be experts in this trial and error method, which we have kind of stuck. We do in all the hay after a lot of deliberations between ourselves, that you always.

So let's say that there is a mixed loss where the air is about 70 decibels. You would set the hearing aids at about 50 decibels. Let's say the bone is about 40, 30 decibels. Hair bone gap. You will be conservative in your approach so that you are allowing for this change in the conductive threshold.

And it does work, you know, it does work in the sense that some children get great benefits and some children reject it. So fitting hearing aids in third windows is difficult, let me tell you. You need a lot of experience. I mean, I don't fit hearing aids. My audiologists do, but they are brilliant in it.

Yes. Even. Because, you know, truly phenomenon by Microsoft. Exactly, exactly. Even, Andrea, I don't know.

Even sometimes hearing aids produced nystagmus. I've seen that. So it's not. Yeah, it's not easy. So it's a purely clinical decision, but it can be done.

Yeah. Yes, sure. So other. Congratulations. Sure.

And another question from Rosanna Rodriguez Villalba. Beautiful speech. Doctor Dasgupta, what incidence of perinephotic fistula do you have in children who attend your clinic? So something. So in that series.

Yeah, in that series of 920 kits that I mentioned, I only had two plfs, two post traumatic or spontaneous, that is, one is post infective, so chronic long standing Milia, and the other one is probably a de novo one, but I haven't seen any post traumatic one. Of course. It's a possibility. I've seen it in adults. In fact, in adults, Andre, I've seen people when they.

One guy came to me who was a saxophone player, so he blew on the saxophone player one day, and that's it. Oral fullness, autophony. And that was a PLF. But in kids, it's unlikely. I mean, I have seen a couple in our.

In my 115, I've seen only two, so maybe one less than 1%. Now, if you ask me why that PLF is so rare in kids, I'll have to think about it. I don't have an answer right away, but it's something to do with the bony skeletal anatomy or something. I don't know. Probably.

Interesting. Thank you. So another question from Faria labor and cochlear implants. Outcomes in superior canal dehiscence. In my experience, I've never had any child or adult ending up with a cochlear implant in superior semicircular connotations.

However, in third windows, cochlear implants work extremely well because I have got at least five or six. The majority of them. No, I have at least seven. Five of them with enlarged vestibular aqueducts, with cochlear implant, and two of them with. No, actually six.

And one of the. One of the x linked gushers, which I saw, which we published, has also got a cochlear implant. So cochlear implant can be used in syndromic third windows. Most certainly they can be done. Very good outcome, actually, because they deal with the hearing loss and even the cochlea, you directly stimulating the cochlea, due to whatever reason, there is some beneficial effect.

In fact, this is well known that the cochlea stimulation actually helps the vestibular system as well. So, Andre, I'll tell you something, that when we check vestibular function in children with about to gochlear implant, we see no function, bilateral areflexia, motor development and delay. They get the cochlear implant, their motor

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development shoots like that, no problem. You repeat the vestibular function tests. No change.

So peripheral, no change. Same. We hit huge saccades, absent webs, but function is huge. So I asked Sharon Cushing and I asked J. Rubinstein in Seattle.

I think you know them. They told me that it's at the central level, that the cochlear, the sound at the central level, especially in the temporal gyrus, you know, where this vestibular and the cochlear cortex are very close to each other, they feed into each other and that's what preserves the functional balance. That's what I've seen. So the cochlear implant in enlarged vestibular aqueduct and in excelling, they practically, they, both of them improve the balances in the kids. But so, you know, we have been talking about this, Andrea, frequently, that how can you objectively measure compensation?

You know, the dispersion, the pr score, dispersion, clustering of saccades. Then for example in the shimp, the peak saccadic velocity and everything, that's how you can objectively measure compensation. But with cochlear implant, the functions are exactly the same. Huge saccades are flexia, but the child who was unable to stand start standing, walking and running. I saw a kid even last week, he was running.

No vestibular function. So some, I believe Jay and Sharon, and they do lots more cochlear implants in kids. So they have said that it's something to do at the central level which kind of compensates for this peripheral deficit which is not reflected in the periphery. That is. That's the only way.

That's the only way. Very interesting. Yeah, yeah, yeah, definitely. Another question from Riehm Abdullah. If a patient refuses surgery but wants the symptoms to improve, what would you suggest?

Well, that's where we call situational counseling. Identify what makes you distressed and then try to mitigate it. It's some kind of rehabilitation. For example, if you hear your footsteps very loudly, wear some insoles so that the sounds get minimized. But we never advise avoidance behavior.

Avoidance behavior is a no go. So if you think that you'll get a tullio, do something to protect your hearing, put some earplugs or get the sound down. But we do not ask them to come out of an activity which they love.

Sure. So last question. It's yes, it's a nice question. From Szonette Dreyer. I'm an audiologist at a school for the deaf in Worcester in South Africa.

One of my learners is a profound hearing imperative. Eight year old. Both mom and dad are congenitally deaf. Home language is sign language. I try to feed him with hearing aids, but the moment the hearing aids are fitted he becomes dizzy and vomits.

No problem without hearing aids. Ct scan done, no abnormality found. I took away the hearing aids since the family is sign language users. Did you find similar symptoms in your vast experience and advice? That is absolutely brilliant Sinit and thank you so much for joining from South Africa.

For a moment I thought it was Boston, UK, which is our next door neighbor. So there are a couple of things which I must tell you. The first thing is, has any investigation been done to figure out why there is a hearing loss? In other words, it's clearly genetic, right? So it's probably autosomal dominant.

So have you actively sought out the cause, number one? Number two, have you done vestibular function tests, especially the VHIT and wemp? Because you might not get an nystagmus if it's bilateral. What about the motor development of the kid? Was he slow?

Did he walk, stand, run at the right time? At this point of time, how is he in his playground activities? You know, trampoline, bouncy, castle, swings, slides? How does he react to bright lights? Is there any visual over reliance?

All this is very important information now that will give you a full clinical picture or a phenotype. The only thing which is very suggestive here is when you put the hearing aids, he becomes dizzy and vomits. Now, just now, I told Andrea that I have seen this happen in a third window. CT scan is done. But look, Chanel, CT scan probably would have been done with a 1.2 millimeter slice in an axial or at the most, a coronal plane.

You get back to your radiologist, please, and you discuss with him. Ask him to specifically look into all the different canals, reformatting the whole city. Scan at 0.5 millimeter and then down to 0.3 millimeter, especially in the Stenber and Poshkal plains. I can tell you there is a 90% to 95% chance you will pick up a third window. This looks like a third window, an autosomal dominant third window.

This is very likely a chromosomal abnormality. Or this could be a pen red. You need to do the thyroid. There might be a dilated, enlarged vestibular aqueduct. So I think this is suggestive of a third window.

It can be other things, too, but this is you will. You are bound to find out. A mutation here.

I totally agree. There's. There's a nice paper from the group of Leon that you cited before. From.

Yes. And also Professor Giannoli from the US. And they say that multiple auticapsule dehiscence should be sought, should be looked for. Because even when you have a

patient with symptoms typical from superior canned dehiscence, and this patient has superior can dehiscence, but actually, you perform surgery, symptoms persists. So probably there are other adhiscience, as you said, in cochlear fascia dehiscence, horizontal and facial nerve dehiscence.

There are a lot many. And this is syndromic as well. So, yes, I'll be very, very interested to find out what syndrome, because I'll ask you strongly to do a chromosome karyotype. So usually, my experience, third windows, when they're syndromic in kids, are located around about 13 to 16 q, which is, I think the cue is the long arm, in my experience, that is. But as I said, some well established syndromes might also be present.

Brachio autorenal, for example, autosomal dominant with third windows. So maybe the parents have got it as well. And Bor doesn't have to be branchial and oto and renal. It can be a Bohr variant, which means you can only have a kidney abnormality or a branch abnormality or aortic abnormality and a kidney abnormality. I would also strongly suggest to you that you scan the kidneys of this kid.

See, I've given you so many advice. Sorry. Things. Yes, brilliant. Brilliant.

So me. Thank you very much. So I think. Thank you, guys. I think that Anna is coming to say hi to you and for the next appointment, of course.

Yes. So let me say that I'm very grateful to Professor Dosgupta and to doctor Castellucci for this wonderful presentation. This is the third webinar that Professor Dos Gupta has conducted, the DCR for inventives, academia. And with today we conclude the series. Professor, the passion you put into this presentation is always palpable and inspiration to everyone, in fact.

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And I hope to come and visit you soon. Thank you. Please. It's a beautiful city. I invite you especially.

I'm saying this in public, it's a beautiful city, but I don't like the football team, because I'm Manchester United. I'm die hard Manchester United. But it's a beautiful city. My folks are lovely, my staff are lovely, extremely competent guys. Beautiful hospital.

Please do visit Liverpool. Sure. And I also hope to have you once again in the near future as a speaker at an academia webinar. Absolutely. Thank you.

And let me say that I love Sherlock Holmes. And one of these quotes that I prefer is, you see, Watson, but you do not observe, and 16 and 13 are different concepts. I love. Fantastic. And did you like the story I made up?

I made up, sure. Yeah. I love making up. Making it up. Sure.

Oh, by the way, just let me tell everybody that the significance of the silent dog, this was because the dog knew who was kidnapping the horse and killing the carer. So immediately, your suspect list goes down hugely. Thank you, Professor Dasgufta. So, for everybody, please keep an eye on inventors social media pages for the latest updates and information on how to register for upcoming events by inventis academia. And please, please remember to watch out for the upcoming survey.

And we greatly appreciate your input and suggestions. So thank you once again. Professor Gupta. Thank you once again, Andrea. And thank you everybody for your fantastic participation.

Bye for now. Bye bye. Thank you. Sumit. Thank you, Andrea.

We'll talk to you again. Bye. Thank you. Bye.

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