

## Hearing Loss in Patients with Osteogenesis Imperfecta

**Tracy Hart:** Hello and welcome to the Osteogenesis Imperfecta Foundation's monthly podcast. My name is Tracy Hart and I am the CEO of the Osteogenesis Imperfecta Foundation. Each month, the OI Foundation brings you information about the diagnosis and treatment of osteogenesis imperfecta presented by an expert in the field of OI and rare bone disease. The podcasts are a part of the ongoing educational effort of the newly formed Brittle Bone Disorders Consortium, part of the National Institutes of Health's Rare Diseases Clinical Research Network. The professional education activities of the Brittle Bone Disorders Consortium are led by the OIF. Our podcast today will focus on Hearing Loss in Patients with Osteogenesis Imperfecta.

We're very excited to have with us today Dr. David Vernick. Dr. Vernick is associated with Harvard Medical School in Boston; he is a Surgeon in Otolaryngology at Massachusetts Eye and Ear Infirmary, Beth Israel Deaconess Medical Center, Brigham and Women's Hospital, and Children's Hospital in Boston, MA. Dr. Vernick is also a member of our Medical Advisory Council. Dr. Vernick, thank you so much for being with us today.

**Dr. Vernick:** It's a pleasure, thank you for asking me

Great. We're going to go right into our first question; can you tell us how common it is for people with OI to have hearing loss and why is this occurring in a disorder that is commonly thought of as brittle bone disease?

Well the incidence of hearing loss in OI increases with age. By 50 years of age, pretty much 50% or so of everyone who has OI has a significant hearing loss that is handicapping to them. The incidence increases with age, but it can start as early as birth so that children, teenagers, and young adults need to be conscious of the fact that they can have hearing loss too and may need to seek intervention much sooner than any of their peers would need to seek intervention for help for hearing loss.

Now, in your experience, do you see hearing loss is more prevalent in different types of OI?

I think most of the work is done certainly on the most common type of OI, which is type 1 OI. The numbers that I'm giving you are for Type 1 OI. The other types, the numbers are much smaller, but the incidence is at least that great. The progression as well is at least that specific. I missed part of your first question so let me go back a minute before we move on. And that was why does this disorder, commonly thought to be brittle bone disease, cause hearing loss. The answer to that really is a basic question about how our inner ear works. Our inner ear is an environment which is surrounded and shielded by bone and inside are the hearing and balance organs and they are held in a specific environment that nourishes them and keeps them healthy. The problem with OI is that the environment doesn't always stay as healthy as it should. The bone turnover, the changes in the bone, the changes in the bone texture all affect the environment that the inner ear is living in. with a less healthy

environment, the organ doesn't do as well, and that's thought to be the cause of most of the hearing loss that progresses with OI.

*That's interesting. I know you touched on this just a minute ago, but in your experience when do people really start noticing that they have hearing loss related to their OI? And if they start to experience that, what should they do? Who should they go to see? Should they go to their primary care doctor and say this is what I'm experiencing, and then what should they do?*

Ok, let me start at the very beginning, which is at birth.

*Lots of questions there*

In most states now, I'm not sure if it's all 50 states, but in almost all states now we do newborn screening and so that's very critical in picking up whether you have OI or whether you don't have OI or whether a child has significant hearing loss or not. And so I think that that process of screening for hearing loss begins at that point for everyone, and children and adults who have OI can have all the other causes of hearing loss as well, in addition to those which are caused by OI. So, typically if they have hearing loss at newborn stage, that should be picked up before they've have significant delays in speech and language development. The next step that usually happens is over the first couple of years with speech and language development, if it is delayed then those children should be seen by an otolaryngologist and an audiologist to have their ears checked to make sure they look fine and they don't have any infections or fluid going on in their ears and they should be screened with a hearing test to make sure they still have hearing in the normal range. It's not as easy to do a hearing test on a 1 year old or a 2 year old as it is on somebody who is a teenager or an adult but there are still methods of testing hearing even on newborns and 1 and 2 year olds before their responses can be accurate. There are ways using what are called auditory evoked potentials and otoacoustic emissions which are things that the ear can do with stimulation that we can record electrically and don't require the child to give a feedback. So we can get a pretty good assessment of what a child's hearing is, even before they can tell us that they're having troubles. As the children get older, the recommendation has been to have the hearing tested every three years to make sure they're not developing any slowly progressive hearing loss that might be interfering with the speech and language development or with their schoolwork or learning in their day-to-day settings. Many schools have a program where they do screen kids for hearing, it's important to check with the school where your child goes to make sure that's being done and that your child is passing those tests. There are also some simple things you can do at home to see if the child is turning up the TV too loud, not responding appropriately to questions where the response should be positive and it isn't. Those kinds of things should alert a parent that their child needs to be screened. Then, as we get older, just increasing troubles with communication, I think, warrant continued hearing screening periodically. And again, every three to four years is probably reasonable if you're not having an troubles, until such time that you either are doing great and you never need it, or you're starting develop some hearing loss, then the screening for hearing loss is present, probably should be once a year.

*Ok, thanks. For those listening, Dr. Vernick is a regular participant at our conferences and meetings and I know, Dr. Vernick, you get a lot of questions from adults that are experiencing hearing loss and are ready for some sort of intervention, what do you tell them? What are the choices? What can people look to to help them with their hearing loss?*

The types of intervention that are available really depend upon the type of hearing loss that the person has. We generally take hearing loss and divide it into two main categories. The first category is a problem with the ear itself, the eardrum and the little bones of hearing. The second category is the nerve of hearing; the inner ear function and the nerve of hearing back to the brain. The first part with the little bones of hearing where sound is conducted through the ear canal, the ear drum, the little bones of hearing into the middle ear, is called a conductive hearing loss. And the second kind, which involves the inner ear and the nerve of hearing, is called a sensory neural hearing loss. There are things we can do to help both of those kinds of hearing loss depending upon how severe they are. The easiest thing to do and the simplest thing to do, which is not interventional, if the hearing loss is mild from either kind, is to simply be aware that that's what's going on and to try to improve the listening environment. So, if someone is sitting up front in the classroom (that has a mild hearing loss) they can usually do much better than if they're sitting in the back. Carrying on conversations in quieter areas helps a lot, paying attention to people's lips to pick up additional clues helps. So those things will help with mild hearing loss of either type. If the loss gets to the point where that's not adequate, then the next intervention to consider, if it is a conductive hearing loss, which is a problem with the eardrum and the little bones of hearing, there are really two things you can do. One is considered an operation, where you can go in and try and fix whatever the problem is, and the other is hearing aids. It used to be said in the old days that hearing aids didn't work for sensory neural hearing loss, but that's just not the case, in fact, most people that wear hearing aids now are being treated for sensory neural hearing loss not the conductive hearing loss. So, that is certainly an option. When kids are under age 10 or 12, they can have troubles with fluid in their ears or recurring infections – tubes can help sometimes to drain off the fluid or help stop the ear infections and that may be all that's needed for intervention. If they have troubles with the little bones of hearing, then surgery on those is really not a good idea until they get to be a teenager or a young adult because the results are just not as good when they are little kids. The ear has not fully grown at that point and the success rate is significantly lower. So most children who have hearing loss, whether it's conductive or sensory neural, if it's not something simple like fluid in their ear, the best option is to get them hearing aids to boost up their hearing. Make sure their teachers know that they're having hearing problems and make sure they're sitting up front in the classroom so they don't have all the competing background noise when they're trying to get their lessons. As people get older, they have a conductive hearing loss, the bones that you can repair in somebody that has OI or not as good or not as sturdy as building block as somebody who doesn't have OI, but there are still operations to replace those little bones, where you can put in artificial prosthesis to try and replace the bones. The most common one is the littlest bone- the stapes bone- and in OI that bone can be even malformed or bony changes can be around the base of it and prevent it from vibrating normally. There's also problems at times with head injuries or even loud noises where the little bones can break in the middle ear and surgery can, many times, fix those. That's not always possible and those operations are not as successful as they are in people that don't have OI, but they are certainly something to look into and consider. If

that is not a consideration or if the loss is sensory neural, the answer is to get properly fitted hearing aids. As we get older, it's usually sensory neural hearing loss that becomes a major issue. For those people, hearing aids help a great deal and for some people as they get older and hearing loss becomes much more severe so hearing aids are not an effective means of treating their hearing loss, there is a device called a cochlear implant. That's a surgical procedure where you implant a little fine electrode array or a little fine tubing with wire in it into the inner ear directly. It is placed underneath the skin, behind the ear, then there is an external processor like a hearing aid that is attached to it with a magnet that stimulates the nerve directly and bypasses the whole ear, ear canal, eardrum and inner ear issues so it directly stimulates the nerve of hearing to give you hearing back. That has been successful in kids and adults. It has been done in patients who have OI and is very successful in them as well. The only consideration that is a little bit different with OI is that the bone is not as dense, so that these implants work by triggering little electrical impulses, and normally the bone around the ear will shield those impulses and act as a barrier so they don't spread too far, and since the OI bone isn't as dense or as favorable to shielding those sounds, you sometimes have to turn the current down a little but lower than you might for someone else. But you can still do very good stimulation of the inner ear to give back hearing in those patients.

*Is the cochlear implant probably the most aggressive type of correction for hearing loss, in your opinion, or what's on the horizon for new interventions?*

Cochlear implants, I think, are probably the fastest improving technology we have for hearing right now. I don't know that I would call them aggressive, because as long as you follow the criteria, which are that if you can't get anything effective with a hearing aid, then you're either deaf, or you have a cochlear implant to try and get some hearing back. Although they are as good as normal hearing, and not as good as you can get with a hearing aid, they're still far better than being deaf. So, the criteria for fitting them, I think, is justified by the results that we get from them- so that somebody that can benefit from a hearing aid isn't offered a cochlear implant, they're not a candidate for that. They're only a candidate for a cochlear implant if they failed the hearing aid use or in kids if they are born deaf

*And how long is that surgery, what is that surgery like, for a cochlear implant? Is somebody put under anesthesia, how long does that take?*

The surgery is actually pretty straightforward even though it sounds like a fairly complex procedure. It's a general anesthesia where you are put to sleep, there is an incision made behind the ear, there is some bone taken away from the bone behind the ear called the mastoid, there is an opening made into the inner ear and the electrode is inserted into the inner ear and secured in place. So, the surgery takes somewhere around 2-3 hours. It can be done either as a day surgery, so that people come in and have the surgery and go home the same day, or overnight stay. Some people come in and have it done and stay overnight and go home the next morning. The area is then allowed to heal up so the person has some stitches behind their ear that come out in a week, a dressing over their ear which comes off in a week, then they can pretty much do their normal activities for another three to four weeks, and usually at about a month they will meet with an audiologist and have the external device fitted and adjusted and start the process of learning how to use the cochlear implant. There are some complications with

the surgery that can occur. People can have some dizziness afterwards, which clears up. People can have rarely, but can have an injury to their nerve that supplies motion to their face called the facial nerve, and people very often will lose any residual hearing that they have in their ear at this point. So that's why you don't do it in someone that has good residual hearing or can use a hearing aid. They're working on some cochlear implants that are called hybrid implants where they are done in such a way to try to preserve any leftover hearing that's in the ear, but preserving that hearing is not a guaranteed thing, so they're usually not done. They're done with the hopes of preserving the hearing but they're not done in patients who that's critical for them to be able to do that, at least not yet.

*So, one of the goals of our medical education initiative here for professional education is to provide consultations with experts in the field. What would your recommendation be, Dr. Vernick, if a person with OI went into their primary care provider and said "I'm having hearing loss, this isn't right, I'm 43 years old, I have OI." What should that primary care provider do at that point, in your opinion, how should they advise them?*

Well I think the first thing is to take a look and make sure that their ears look ok. Make sure it's not wax that's block up their ears, it's not fluid blocking up their ear and giving them the hearing loss. People with OI can have all the regular hearing loss problems that everyone else does. If the ear looks perfectly healthy and normal... both ears look perfectly healthy and normal, since we usually have two... then they need to get a hearing test. I think referring them to an audiologist makes a lot of sense. Some of the primary care offices use these little screening devices where they play some tones in the ear to see if the person can hear them or not to try and get a generalized level of hearing and that's ok, but I don't think it's appropriate for people with OI because they can have fairly significant hearing loss in some frequencies and not all frequencies, so that may not be a really good screen for them. I think anybody with a history of OI ought to go directly from a normal exam to getting a hearing test.

*Ok, great. You mentioned hearing aids, you mentioned a little bit about hearing aids. Who fits hearing aids for people, would it be the audiologist? If somebody needs hearing aids, are there different types, how do they find the right one for them?*

Well I think the key to getting the right hearing aids has to do with first getting a good hearing test so you know exactly what kind of hearing loss you have and how severe it is. There are lots of different companies which make hearing aids and there are lots of people that dispense hearing aids. There are audiologists who are trained to do that, who have skill in dispensing and fitting hearing aids, and that's who I would recommend that people go to. There are stores that sell hearing aids, there are internet sites that sell hearing aids, there are salespeople who go around selling hearing aids to people and I would caution against that because you're getting somebody who has been trained, or not trained very well, to sell hearing aids, not to fit them properly and I think it's really important for a person that has OI, everyone in general, that their fitting be proper and be adjusted. You can take two people that have the exact same hearing level and they may need totally different settings for their hearing aids depending upon what their listening environment is. It's not like glasses, where you can go in and do your eye exam no matter where it is, buy your pair of glasses, stick it on and walk out the door and you're fine. To fit somebody properly with hearing aids and get them to get the most out of them,

usually it requires two or three, sometimes more, visits back in to the audiologist, to get the hearing aids adjusted once they've been set at a reasonable level given the hearing loss. Those kinds of things depend a lot on, again, what the listening environment is and what the requirements are of that person. There are lots of bells and whistles that can go on with hearing aids, including directional microphones, the ability to filter out background noise, Bluetooth connection to TV and phones... so there are a lot of different possibilities that you can use to improve your ability to use a hearing aid that the general salesman is not going to be able to help you with. So I recommend that everyone go in to see a trained audiologist for a proper fitting.

*Excellent, thank you. Well we have just a few minutes left and would love for you to share anything we haven't talked about that you think is really important for people to know or some last comments that you have for us?*

A few things – I think it's important, I think it's important that people be more assertive – I shouldn't say aggressive – but more assertive about checking hearing in people that have OI than the general population, especially in the children growing up. We tend to think a lot of times our kids don't pay attention to us, and they may not, but it's important to give them the benefit of the doubt and make sure it's not because they don't hear us, or they're not doing as well in school as they should because they can't hear the teacher. I think that's really a good take-home message. I think it's important as well to look at some very recent data that has come out that shows that not hearing well as we age give you a greater chance of developing earlier dementia. It's important for us to continue to interact socially with other people, and that requires hearing. It's important to do everything we can to maintain that connection, whether that includes hearing aids or surgery, it's just important. People that don't hear tend to isolate themselves from the general population and that leads to cognitive decline. I think one of the other take home messages is, if it's a problem that you can fix, and many times hearing loss is a problem that you can fix or at least help, that you need to do everything you can to do that and keep people interactive.

*Excellent, very good advice. Well Dr. Vernick, thank you very much for joining us. You can find out more information about the medical education initiative we have going on at the OIF by visiting our website at [www.oif.org](http://www.oif.org). Dr. Vernick is a member of our Medical Advisory Council, so his information is on our website as well. Dr. Vernick, thank you so much for this really valuable information.*

It was a pleasure, thank you for having me.

*Alrighty, thanks again*

Goodnight.

*Goodnight.*