

Hello. These are USH Talks. And I'm Marly Kenna. And I will be presenting information on Usher syndrome and cochlear implants. I am a pediatric ear, nose, and throat doctor at Boston Children's Hospital, and have the privilege of having taken care of many of these children.

So what is a cochlear implant? It is not a hearing aid. It does, however, provide auditory sensation, so a sensation of sound by stimulating the surviving elements of the nerve endings in the inner ear. As opposed to a hearing aid that just makes things louder, a cochlear implant is much closer to the way we hear than a hearing aid.

It, in general, supports development and maintenance of spoken language and provides environmental awareness, as well as access to spoken language. So this is what a cochlear implant looks like. On the left of your screen, you see an ear. This is a right ear. There's a piece on the outside. That is the transmitter. And so the sound comes in, it goes to a piece that looks quite a lot like a hearing aid.

There's a microphone there. This sound then goes across the skin to the internal piece, the receiver stimulator, where an electrical signal is generated. That then goes down the electrodes that are implanted in the cochlea, where a response is generated, which goes down the hearing nerve to the brain. And then you hear.

So many things have to line up for the cochlear implant to work correctly. It's very similar to the things we have to line up for us to hear without a cochlear implant.

So what are the potential communication options for a deaf child? Well, certainly spoken language, which is what we're really talking about today is a good option. But, certainly, manual communication-- American Sign Language, or Signed Exact English-- is also a terrific language, and allows the child to communicate with their parents and their friends, or with each other.

Many people do a combination of spoken language and manual communication, sort of a total communication approach. And one form of communication, for example, learning spoken language doesn't mean that you can't also learn and use manual communication or signed language.

So who are the candidates for a cochlear implant? Well, in general, there are children age 12 months or older, although in some cases, children may be a little bit younger. Children who are born with their hearing loss generally have a bilateral-- so both sides-- profound degree of hearing loss. These are pre-lingually deaf children, which means that they've either lost their hearing or they were born with a profound hearing loss before they acquired their spoken language.

Children who lose their hearing after they have acquired the spoken language are called post-lingually deafened children. And those children, in many cases, may also be candidates for a cochlear implant. In addition, children and adults who are hard of hearing-- so they have a little bit of hearing, but they're really not doing very well with their hearing aids-- may also, in some cases, be candidates for a cochlear implant.

What about children with Usher syndrome? Are they good candidates for cochlear implants? While, in general, people with Usher syndrome are good candidates for a cochlear implant, they have to meet the same hearing loss requirements as anybody else. And, of course, they or their parents want them to have a cochlear implant to support the development of spoken language.

If they, as a patient, or the parents, want them to use spoken language as a communication modality, that can be supported by cochlear implant. It doesn't have to be their only communication modality. But it has to be something that the patient and the family are willing to work on so that spoken language can develop.

So which patients who have Usher syndrome are candidates for a cochlear implant? So patients with Usher syndrome type 1, who are often born with a bilateral profound hearing loss, who are really deaf at birth, are most likely to be candidates during early childhood. These children are considered pre-lingually deafened children. That means that they already had their hearing loss before they were able to develop spoken language.

And, in general, most children with Usher syndrome type 1 are born with a very significant degree of hearing loss. Patients with Usher syndrome, who have Usher syndrome type 2 or type 3, or, occasionally, somewhat less typical, Usher syndrome type 1 patients, may start out with a hearing loss, but they still have a lot of hearing. They wear hearing aids.

And then over time, their hearing may start to progressively get worse. And in those cases, they will develop spoken language, initially, with their hearing aids. And then over time, they're really not using their hearing aids very well. And they will go on to become cochlear implant candidates, as well.

Have people with Usher syndrome received cochlear implants? Well, yes. Many have both-- children who are born deaf-- as well as older children and adults who had a progressive change in their hearing. In the right patients, under the right circumstances, these patients have developed a lot of good spoken language. And they really benefit greatly from these implants.

However, even if the patient receives a cochlear implant, no matter what their diagnosis-- and if they are-- especially if they're a young child, they will need speech and language therapy, generally speaking, for many years to come. In addition, although most patients do very well, there is a wide range of possible outcomes after a cochlear implant. And that's true, whether the patient has Usher syndrome, or has some other reason to have a

hearing loss.

So are there reasons why people with Usher syndrome may not be good candidates for cochlear implant? So there are many reasons why any patient with or without Usher syndrome may not be a great candidate for a cochlear implant. Every patient situation is different. And, therefore, this should be discussed with their cochlear implant team as part of an overall assessment.

Reasons why people, including patients with Usher syndrome, may not be good candidates are that they may be older children or adults who have mainly used sign language their entire life. They haven't worn or used hearing aids effectively. And they have not really developed usable spoken language. So those children or adults getting the cochlear implant when they're much older often have very limited benefit. And, generally, we would not expect that they can develop good spoken language.

There certainly can be medical reasons for anybody, whether they have Usher syndrome or not, that may make it riskier for them to have anesthesia, or depending on the anatomy of the inner ear, it may be riskier for them to actually have a cochlear implant. There are some special considerations, potentially, for cochlear implants in patients who have Usher syndrome.

For example, there may be effects on the patients' balance. We do know that the electrodes are implanted into the cochlea, which is part of the inner ear. Parts of the inner ear also control balance. It turns out that, although we do worry that people's balance could get worse after cochlear implant, there is actually some emerging information that Usher syndrome patients, especially type 1 patients, who often start out with balance that's not very good before the implant, actually may have better balance after their cochlear implant, especially when it's turned on.

And if that turns out to be the case, both hearing loss and, maybe, balance problems would become reasons to consider a cochlear implant. So should I wait or should my child wait for a cochlear implant? Well, people ask us this all the time. We know that on the horizon, there's gene therapy or potentially other types of therapy that may improve the hearing or prevent the hearing from getting worse. And so the question is, should the patient get one cochlear implant or two cochlear implants, or no cochlear implants, and wait for these therapies to become available?

I think that the current state of the art is that these therapies are not available, currently, for use in humans. Most of this therapy is still in the laboratory, in laboratory mice or other types of laboratory animals. So if a patient is otherwise a good candidate for an implant and it seems reasonable to go forward, it is, therefore, reasonable to consider at least an implant in one ear. And depending on circumstances, possibly, an implant in both ears. Down the road, this may change. But, currently, that's the state of the art.

What's the timing of a cochlear implant? So for a child who's born deaf or has very early onset profound hearing loss, generally speaking, the first two to three years of life are felt to be critical periods of time when the child will learn their language, whether that's spoken language or sign language. So if the parents really want the child to learn spoken language to get a cochlear implant in the first couple of years of life is ideal. Although, obviously, sometimes there are reasons that, that can't happen on that timeline. For patients who are concerned about saving an ear for therapies coming down the road, at least one side cochlear implant will provide access to sound and, therefore, the possible development of spoken language.

So in summary, patients with Usher syndrome may be very good candidates for cochlear implants. Making a definite diagnosis of Usher syndrome, actually may help in making the decision about a cochlear implant. For example, if the patient knows, or the parents know, the child's vision may start to get worse and make access to sign language more difficult, sometimes, knowing that a cochlear implant's a possibility, with access to sound, may be, actually, something very good to know.

And in addition, for all patients and their families who are considering a cochlear implant, whether or not they have Usher syndrome, they should talk to other people who have made this exact same decision, and about why they decided to go forward with an implant, why they perhaps didn't go forward with an implant, and what type of outcomes they were actually seeing in their own child or in themselves if they had an implant.

Like everything else with Usher syndrome, there's always excellent information available at the Usher Syndrome Coalition website, including some information about cochlear implants. This USH Talk, of course, will be on this website and available for viewing at any time.

Thank you very much. And I really appreciate you taking the time to watch this presentation. If you want to learn a little bit more about cochlear implants, these are two excellent sources of basic information about cochlear implants. The first one is the National Institute of Deafness and other Communication Disorders-- NIDCD-- at the National Institute of Health. And the second one is through the Food and Drug Administration. Both give very excellent basic information about cochlear implants and the devices themselves.

Thank you very much.