Engaging Families in Continuing to Improve EHDI Programs, in partnership with American Cochlear Implant Alliance

Presenter: Karl R. White, PhD

Thank you so much for joining us today. This is Donna Sorkin from the American Plant alliance, and we're thrilled to be partnering with continuing ed audiology online for this two part series on engaging families and continuing to improve EHDI programs in partnership with American Quigley Implant alliance. You could go to the next slide. And we're looking at the power of parents in fostering language in deaf and hard of hearing children. And this is the second of a two part series.

We did the initial one already with Doctor Deana Susskind. Go to the next slide. So let me just tell you a little bit about the American Cochlear implant Alliance, and you may wonder why. There's another organization in the hearing health field, and we came into being about twelve years ago. Our membership is very focused on cochlear implants and access to hearing care, and our members are from across the hearing health continuum.

We have a very extensive website for those who are in and out of cochlear implants, and we try to be highly collaborative with other organizations, such as our speaker today from Utah State, and of course, our partners in this endeavor, continuing education audiology online. So I've given you some ways to contact us and we go to the next slide. And that's our mission, to advance access to the gift of hearing provided by cochlear implantation through research, advocacy, and awareness. And we really focus very much on those factors that contribute to the underutilization of cochlear implants, both in children, as we'll be talking about today, and in adults. And we want to improve awareness regarding both the candidacy and outcomes for the general population, as well as for those who are in hearing health who are outside of CI.

Go to the next slide, please. So this topic is really critical right now because hearing loss in children has changed so dramatically. We now are able to find children early on in their lives. When I first started in this field some 30 years ago, we didn't find kids

with hearing loss until they were two and a half or three years of age. And that has changed dramatically due to the work of Doctor Carl White and others in this field.

And we're now able to fit technology routinely at two weeks of age and are able to recognize that parents are, in fact, a child's best and first teachers, just as they are for children who have typical hearing. And we're really emphasizing the fact that the language of the home allows integration into the fabric of the family. And we really adopted this term, language nutrition, that was originally envisioned for parents of all children by pediatric nurses who wanted to emphasize this whole concept of talking to your children and engaging with your children. And we found that 30 million words, a book that came out some 15 years ago and introduced by our speaker last week, Doctor Janice Eskine, was so important for lifetime achievement in children because parents already have the tools to move forward with this. So next slide, please.

And we have hence initiated our listening language literacy activities, which are a high priority for us at AC alliance. And we want families of children with all levels of hearing loss to be able to foster literacy in the language of the home in terms of talking and interacting, and also really emphasizing that when that happens and children are integrated into the fabric of the home, there are important social emotional benefits for the children and for the family, and the fact that hearing the sounds of language facilitates reading and writing and contributes to literacy outcomes. So with that, let's move on to our introduction of our speaker today. We are thrilled to have with us Doctor Carl White, who has been the director of the National center for Hearing Assessment and Management at Utah State University for, I think, 24 years. So really an important pillar for all of our work in early intervention.

And prior to getting involved at the national center, he was actually at National Technical Institute for the Deaf, working there on research and evaluation on many of these same issues. And he also served in Washington in a variety of ways with federal

agencies and on the Hill with congressional members. And he serves still on many international organizations and advisory groups, such as the World Health Organization, the US Department of Health and Human Services, March of Dimes, the American College of Medical Genetics, the National Academy of Sciences, Engineering and Medicine, and the American Academy of Pediatric. He is truly a treasure to our field. So with that, I'd like to turn the floor over to doctor Carl White.

Well, thank you so much, Donna, for that kind introduction. And it is absolutely a delight for me to be here today talking about something that I love to talk about. I'm going to talk a little bit about where we've come from, where we are now, and what some of the things that I hope will happen in the next decade. But woven throughout that, as you will see, is the role that families and parents have played in this amazing progress that has been accomplished during the last 25 to 30 years. As Donna said, we can now start very early with children because we're identifying them early.

And this little girl you see is wearing hearing aids, which surprises a lot of people. But this is what we would like to see happen, that parents have the option when their children are just a few days or weeks old, to provide them with the assistance they need to develop language and to thrive as all other children's do. So these are my disclosures and these are the learning outcomes for today's session. And I'd like to introduce that by talking about three historical people or events to set a context for what we'll be discussing today. So the first kind of historical theme I'd like to introduce comes from William Shakespeare in the Tempest.

And he said, the part that everyone's heard is what's past is prologue. In other words, where we're going to move in the future depends on what we've done in the past. But the part of that statement that most people are not familiar with is what Antonio said next. He said, what to come in yours and my discharge. And so, as important as it is to

build on what we've already accomplished, I think it's very important for all of us to recognize that what happens next is not going to just happen automatically.

It will happen because of what people like you and I do. I was reminded of another part of this that is important when I read a story about Supreme Court Justice William O. Douglas, who served decades ago on the Supreme Court. But he was riding in a cabinet from the airport in Washington, DC to the Supreme Court building. And as they passed the National Archives building, these words, what's passed is prologue, are chiseled on the front steps of that building.

And knowing that cab drivers in Washington, DC have a reputation for having an opinion about everything, Justice Douglas asked the cab driver what he thought those words meant, and the cab driver hesitated for a second, and then he replied, they mean we ain't seen nothing yet. And I think that's very apropos for where we are with early identification and treatment of hearing loss. We have made incredible progress in the last several decades, but we ain't seen nothing yet. The second perspective I'd like you to think about for just a moment comes from an old television series that Lily Tomlin was in. And in that series, she was a telephone operator that generally treated her customers with little sympathy and always told it like it was.

And once she Lily Tomlin, later, during an interview, said, talking about a lot of things that need to change in this world we live in, I always wondered why somebody didn't do something about that. And then she said, and then I realized I was somebody so similar to what Shakespeare said, we all need to realize the role we have to play in moving the field forward, and there is still a lot that needs to happen the third idea I'd like you to think about comes from this man. This is a picture of my father. He was the cowboy's cowboy. He was also a veterinarian and taught his children to love horses.

When I was about two years old, I spent as much time on horses as doing anything else. And as I got older, I learned to ride. And you can see here that the stirrups didn't quite fit me, but that didn't slow me down. And then, as I continued to grow older, I went on many pack trips with my brothers and my father in the Uintah mountains in Utah. Then I was lucky enough to meet a young lady who also loved horses, and she became my wife.

And we continued to ride. And then we had children, and they went on pack trips with me. And then grandchildren came along, and they too, have learned to love horses. Now, why am I telling you all of that? So let's go back to this picture.

When I was about eight years old, this was a horse that I had been working to train. And she was pretty good at this point, but not great. And one day my father said, we need to go out and check on the horses that are out on the back of the property. We had a place where it was about a mile ride back, and we had to cross a stream to get to these horses. And it was wintertime in northern California.

And as we rode out to check on the welfare of those horses in the back pasture, we had to cross a creek that usually didn't have much water in it. But because it had been raining a lot, it was much higher than I had ever seen it. And instead of being about 15ft across, it was about 75ft across. And I was on a half trained horse, but my father was leading me. And as we rode into the water, I said, how am I supposed to do this?

And he said, you'll be fine. Just lift your feet out of the stirrups, perch on top of the saddle, and you'll stay mostly dry. And so it happened just like he said. We crossed that creek and went out and back and checked on the horses. And then as we were returning, we came, of course, to the same creek, and I did the same thing.

As we rode down into the water, I lifted my feet out of the stirrups, but the horse I was on was a pretty smart horse. And as I did that, she realized that she had an opportunity of eliminating a nuisance in her life. And so she waited until we got down into the water pretty good. And then she shook and bucked a little and I fell off into the creek. And I was wearing my heavy winter jacket, and I was sure this was the end of me, that I was going to drown right there.

And so I started flailing in the water. And after a few minutes, it seemed like longer than that, but I reached the shore and I looked up and there was my father on his big black horse, just laughing and laughing. And I said, I was upset. I said, I could have drowned, and you think it's funny? And he said, well, I wasn't too worried about you because the creek is only about 3ft deep, and as soon as you got tired of flailing in the water, I was pretty sure you'd hit bottom and stand up and walk out.

So the reason I was having so much problem is that I had lost my perspective. And I think sometimes it's important for us, as we move forward in early identification of hearing loss, that we remember the perspective from where we are, where we've been, and where we need to go. So as I go through some more substantive matters now about early identification of hearing loss, please keep those three things in mind. What's past is prologue, but more importantly, what to come in yours and my discharge. Secondly, that somebody does need to do something about many of these issues we'll be talking about, but each of us is a somebody.

And then finally, that we need to keep our perspective as we move forward. So this graph represents that permanent hearing loss occurs more frequently than any other condition for which we can screen babies at birth. If we take a random sample of 10,000 babies and look at what kinds of congenital defects and diseases affect those babies, then permanent hearing loss would affect about 30 of those babies, compared to eleven with down syndrome or only one with Phenylketonuria. And so, even though

hearing loss occurs much more frequently and has serious side effects, as we'll see in a moment, it's not nearly as well known as many of these other conditions. It's also important to recognize that hearing status of children is a very heterogeneous variable.

The data in this graph come from the Centers for Disease Control reports from early hearing detection and intervention programs, or EHDI programs. And as you see, only about 14% of the children that are reported to CDC have bilateral profound hearing loss. What many people think of as deaf children. Over 50% of those children have either unilateral loss or bilateral slight or mild hearing loss. And yet these children with the milder forms of hearing loss also have serious negative consequences if we don't identify them early.

And so that's part of the perspective we need to keep in mind. Also, it's important to note that more than 90% of all children who are born with permanent hearing loss have two parents who are hearing parents. And in the vast majority of cases, those parents have never met a person who has permanent hearing loss, and they're not quite sure what to do. And so we need to provide them with support that is culturally sensitive and appropriate and provides them with all of the options that they will need to decide what's best for their child. Now, many people associate hearing loss with the use of sign language.

And although that's an appropriate approach, it's important to recognize that the world is changing quite rapidly with respect to how families choose to communicate with their children. This came from a study we did several years ago that was a national sample with parents from ten states. About two thirds of those children, all of these parents had children who were under six years of age. And about two thirds of those families used listening and spoken language only, or mostly listening and spoken language in conjunction with a little bit of sign language or cued speech. Only 6% of those families were using American Sign language primarily.

And so it's important that we keep in mind that parents do have choices, and those choices have important consequences. Now, this video focuses on why it's so important to learn to communicate. This is a video of Helen Keller, who, in spite of being both deaf and blind, learned to communicate very effectively with those around her, and later in life, became a very influential social activist and an advocate for people who are deaf or hard of hearing. Later in her life, Helen was asked whether hearing loss or blindness was the more serious disability in her life. And she surprised a lot of people when she said that hearing loss is much more serious.

And the reason she gave was that blindness separates people from things, but deafness separates people from people. So this ability to communicate is where we really need to focus. And it's what has changed so dramatically because of the success of newborn hearing screening identification programs. Now, people who are deaf and hard of hearing communicate in a variety of ways. Some use sign language, some use listening and spoken language, and some use other methods.

It's not that one of these is right or wrong, but it's what is most comfortable and successful for families that we should be focusing on as we look for ways to provide support to these children who are deaf and hard of hearing and their families. Now, I'm going to show you a couple of videos here demonstrating how young children communicate young children who are deaf are hard of hearing. The first one comes from a family where the father is profoundly deaf. The mother is hearing. But american sign language is the language in the home.

And they're just leaving a car repair shop. And there are two little boys, both of whom are adopted and are about two years old at this time. They were adopted from China. When they came to the US, they had no language at all. And because sign language is the language of the home in this case, look at how well they do.

So they're just leaving the garage. The one little boy says, the car's up high. It's going higher. The other brother says, hey, dad, look over there. I see a car moving up.

It's going higher and higher. Hey, mom, is someone driving that car? There's so many cars, and they're all shaking. Then he has a side conversation with his mom. I love you.

She says, I really love you. And he reports back, I really love you. Then his brother says, we'll be there soon. Hey, look over there. Yeah, it's an airplane.

I see it. And it's flying. Yeah, an airplane flying. Hey, dad, pay attention to me. Brother says, trains go fast.

I'm looking for a propeller. Where is it? Yeah, where is one? Yeah. I don't know.

Is it up in the sky? Oh, you're talking about a helicopter. A helicopter. A helicopter. Where's a helicopter?

Is it up in the sky? Where? Yeah. I don't know. Hmm.

I don't know either. Yeah, there was one here before. So I hope you see in this video that these little boys have an incredible language base, and they communicate very effectively. They're able to ask questions based on past experience. They describe what they see.

They jump around from one topic to another. In short, this is sophisticated language that they're using. It's not just labeling signs. It is a language that has its own grammar and morphology, and it can be used very successfully. But for that to happen, children

have to be identified early, introduced to that language, and have fluent language models.

Now, keep that in mind as we look at another situation here. This is a little girl from this classroom of children from the mid 1970s. And this classroom focuses on listening and spoken language. And I want to show you first where we were 35 or 40 years ago, and then we'll focus in on where we are today. So this provides you with a historical perspective.

Listen carefully to this little girl who has severe bilateral loss. So she has some residual hearing and is wearing the best hearing aids that were available at that time. And see if you can understand her.

I suspect for most of you, you couldn't understand a word of what she said. Let me play it again and give you a visual clue.

So with the visual clue, most of you could get most of what she said. So what you've heard here is what is often referred to as deaf speech. It has a certain tonal quality to it, and for people who were accustomed to it and knew the context where it was happening, they could understand a fair amount of what this little girl was saying. So her family members could understand quite a bit. But when she went to the neighborhood park to play or to the grocery store or to school, then most people only understood what you understood the first time you listened to it, which was almost nothing.

So I want you to compare where we were in the mid seventies, because this represents about as good as we were able to do. Compare that with what's happening today. So I'm going to show you a video of two six year old boys who have profound hearing loss

in both ears. So worse hearing loss than this little girl has. And the difference is that they were identified within a few days of being born.

They immediately began receiving intervention. They received cochlear implants when they were about eight months old. And this video was made by one of their mothers to introduce the boys to their classmates on the first day that they attended their neighborhood school. Hey, I'm AJ. Yeah, hi.

My name is Gibson. People are special in different ways. One of the things that makes me feel special is that I'm deaf. I'm deaf, too. And deaf means that your ears can't hear.

AJ and I have special things to show you are called cochlear implants. They help us here. Cochlear implant is a big word, so I call them c I s. C is for cochlear and the eye is for implant. CI's are amazing because they're like bionic ears.

Sometimes my friends have questions about my ci. How does that thing stick to your head? There's a magnet inside. The magnet here connects to the magnet in my head. The inside and the outside have to work together.

When I disconnect this, I can't hear. When it's on, we'll lock. I can hear. So according to AJ, it's all very simple. VoilÃ .

And everything's taken care of. Now, we know it's not really quite that simple, but it is amazing the progress that has been made during the last 35 or 40 years. And these two little boys are very representative of that progress. They aren't superstars. There are hundreds and thousands of children who are doing every bit as well as AJ and Gibson are, are doing.

And as early identification of hearing loss gets better and better, and we're reaching these children earlier and earlier, then we're making even greater progress. So it's important, in the words of Shakespeare, to reflect on what's happened in the past that is setting the prologue for the future. How did we get from where we were back in the mid seventies to where we are today? And it applies to children who are using listening and spoken language, as well as children who are using sign language or cued speech. So there are, in my mind, at least three factors that have contributed to that.

First is earlier identification of hearing loss, the result of newborn hearing screening programs that we'll talk about a little bit more. But at the same time, we were fortunate that technology was expanding so that we got better and better assistive hearing devices, better hearing aids that were focused specifically on children rather than adults, better cochlear implants, and we continue to see improvement in those areas. And then thirdly, we were learning a lot about how to teach language. And whether we're teaching Spanish as a second language, or American Sign language, or the first language of English, we know now how to do that a lot better than we did before. Now, all of this really started with earlier identification of hearing loss.

During the late eighties and early Nineties, Utah State University conducted the first large scale, systematic evaluation of universal newborn hearing screening in Rhode island. So the Rhode island hearing assessment project was of what was the first large scale effort to see if we really could identify children during those first few days and weeks of life. And it was incredibly successful. We identified about six children per thousand. Now, that number is higher than what you're used to hearing, because it was over represented with a population of children who were in the neonatal intensive care unit, but it included both, well babies and NICU babies.

And it became the foundation that many other programs built on and replicated over the next few years.

The results of the Rhode island hearing Assessment project led to the National Institutes of Health, who convened a consensus development panel in March of 1993 that looked at all of the evidence that was available at that time, not only from the Rhode island project, but from other projects as well. And they concluded that all babies should be screened for hearing loss before leaving the birth hospital, and that that would help to bring the age of diagnosis down dramatically so that we could be more effective in our intervention programs. Now, many of us who were involved in this were optimistic that we would see rapid change. And unfortunately, it didn't happen as rapidly as we had hoped. So when the NIH consensus conference concluded that all children should be screened for hearing loss, about 6% of all babies were being screened using some method.

And it took another ten years before we got to the point that we are, and remain today where we're screening 98% to 90%, 98% to 99% of all babies. Some of you probably haven't seen newborn hearing screening happen. This next video comes from a parent education video that we did many years ago, but the process remains essentially the same then as it is today. Sophia how can your baby's hearing be checked? Thanks to advances in technology, we can now check a baby's hearing shortly after birth.

Your healthcare staff is concerned about your newborn's hearing because one out of every 300 babies is born with a hearing loss. Children who cannot hear clearly will often have trouble learning to communicate. That's why it is important to check or screen each newborn's hearing using one of two safe, simple, and painless procedures. Let's watch. An example of each.

One method is called OAE, or otoacoustic emissions screening. In this process, a trained screener places a small earphone in the baby's ear that emits a series of soft sounds. The inner ear typically responds to these sounds by producing echoes called

otoacoustic emissions, which are analyzed by the screening equipment. Another method is called AABR, or automated auditory brain stem response screening. In this process, a training, the brain screener places band aid like sensors on the baby's head as soft sounds are played into the baby's ear.

These sensors analyze the brain's response to the sound, no matter which method is used. In a few minutes, the screening is finished and the result is displayed.

Now, as this equipment has gotten more widely available and less expensive, then universal newborn hearing screening has expanded not only throughout this country, but throughout much of the world, and that has led to dramatic reductions in the age at which babies are being identified. This compares the average age of identification from a number of studies done in the eighties and early Nineties at two and a half to three years of age, with studies in the late Nineties and early two thousands that we've reduced the average age of identification to two to four months of age. Now, these averages mean that there are still children who are not being identified until well after three or four months of age, and so we still have work to do. And that work is summarized by this little cartoon that when we first started doing newborn hearing screening. We put a lot of effort into figuring out who should do the screening, when it should happen, what equipment worked best, and we were able to resolve a lot of those issues.

And there was kind of the feeling that once we got all of these issues resolved, then a miracle would occur and we would have success. Babies would go on to achieve in similar ways as their hearing peers. But as is pointed out by this little fellow, he says, good work, but I think we might need a little more detail in this box where the miracle has to occur. And that miracle has required us to really re examine how we do diagnosis of babies, what happens with early intervention programs, how to connect those babies to their primary healthcare provider, how to create data management

systems that can track babies through the system and provide the basis for evaluating and improving the system. But most importantly, and this is woven through all of the parts of improvement in early hearing detection and intervention, has been how we support and involve families.

It really has been the extensive involvement of families in the early hearing detection and intervention program that has made all the difference in the world. So, as I now move on to talking about what's happening now and what needs to happen next, remember, what's past is prologue, but what's to come in yours and my discharge? That each of us is the somebody that can help to make things happen and how we keep all of these things in perspective, not only where we've come from, but where we're going. So I'm, I'm going to give you a few quick examples. This chart, which I know you can't read, comes from some material that we developed for the American Academy of Pediatrics quite some time ago that outlines the flow chart that should be followed when babies are identified with hearing loss.

The part I want you to focus on is right here, where this part of the chart talks about. To where should babies who are diagnosed with hearing loss be referred? For additional information. And the American Academy of Pediatrics recommends that there are three primary areas that an otolaryngologist, an ophthalmologist, and a geneticist should see the child to provide additional information. Now, several years ago, we wanted to look at how well that was happening, and we did one study in 2006.

We replicated that study in 2012, and the results were very stable. We asked the question, if you have a newborn in your practice that has been diagnosed with a moderate to profound bilateral hearing loss, to whom would you refer the baby now? It wasn't a multiple choice question because we wanted to know how much they would come up with on their own. And the results were alarming. Only less than 1% of physicians said that they would refer to an ophthalmologist.

Only about 9% said they would refer for a genetic evaluation. 75% said they would refer for an oligological evaluation, which sounds a lot better when you compare it to these two. But really, that's quite alarming. Also that fully a quarter of primary health care providers would not refer the child to an expert in hearing as a part of the workup and evaluation. We also asked that same sample, at what age can babies be fit with hearing aids?

And again, it was an open ended question. We know now, and from what you saw at the very beginning of this, we can fit babies with hearing aids when they're a couple of days old. It's really just a scheduling problem. But alarmingly, about 40% of these physicians didn't think you could put hearing aids on a baby until they were six months of age or older. So we've got a lot of work to do, and I think families are one of the groups that does that most effectively to help educate our primary healthcare providers about what the consequences of hearing loss are, but also what can be done to help these children, because, as you've seen, if they get appropriate treatment, they do, as well as any other child.

Okay, the second area I want to focus in is, in response to this question, what percentage of children who are deaf or hard of hearing at five years of age were born deaf or hard of hearing? The answer is about 50% of them, which means that there's twice as many children that become deaf or hard of hearing after being born with normal hearing as are born deaf or hard of hearing. This information is reinforced by findings from the National Health and Nutrition Examination Survey, which is a national survey that's done periodically by the Department of Health and Human Services. As you can see here, there are seven to ten babies per thousand who are deaf or hard of hearing, or children who are deaf and hard of hearing in the six to 19 year old range. So this is at least double, in some cases, triple what we're finding for babies who are born in the hospital.

So this raises the question of, what's our safety net? If we've screened all the babies in the hospital and identified all those who are deaf or hard of hearing at that point, how do we pick up the other 50% of babies? And the answer is that we can implement screening programs in head Start part C, daycare. One of the earlier studies that is cited here, they found about the same number of children in early head start programs as were being identified in newborn hearing screening programs. There are similar studies have been done.

Here's two others that found about the same number of children. So we can't afford to relax after we get hospital based newborn hearing screening programs implemented because there are at least that many babies, again, who acquire hearing loss during those early years. And we need to be more systematic and whether we're screening in head start programs or well, baby visits for physicians or daycare programs, and at this point, there are very few children who are being screened after the newborn period, there are a lot of resources available to people who can, many of which are on this website. Those resources, most of them are free, and there's a lot of information about how these programs can be effectively implemented. But similar to what happened in newborn hearing screening programs, the success of screening in the birth to three area is going to happen because of the advocacy and the support of families.

A third area I'd like you to think about for just a few minutes is the use of video, of Internet based video transmission to identify and to provide services to children who are deaf or hard of hearing. This picture is a family with a little boy who is wearing an implant, has bilateral profound hearing loss, and he's getting early intervention services through what we call tele intervention. I'd like to show you a video of him when he was just a little baby, about nine months old, of how he was getting intervention. Hi. Hi, Alex.

Hi. He's looking up. So why don't you just present the sound, Nancy, and then we'll see if he has any reaction just to your voice. No. No toy.

You just want to present the sound first and then we'll see if he looks and starts turning to you or anything like that.

Perfect. Awesome. Looked right at you. Awesome.

Yeah. So he looked up so he can the reward. That's right. Whoa, whoa. He's never done that before.

He shoots. He scores. Yes. That was amazing. That is really the first time I've seen that.

That's great. So I show you this video to emphasize two points. One is that we need to be more broad minded in thinking of how we can deliver services to children, whether they are early intervention services or diagnostic services. And that video conferencing technology has come a long ways and represents a very viable means of providing those services. But secondly, it's the role that families need to and can play in providing services to their children in normal and natural ways that fit right into their daily routines.

One of the questions we had when we started providing early intervention services over video conferencing equipment was whether they would be just as effective as face to face services. And we did a randomized study of 27 children, some of whom received face to face services, some of whom received tele intervention services. And we found that the tele intervention services, as shown in this graph, were actually more effective than the face to face services because they involved families. And those families were then able to integrate those strategies into their daily routine. And so the child was constantly being helped to develop and use language.

Again, there are a lot of resources available. This comes from our website. Most of these resources are for free, and this is an area where I think we need to significantly expand what's happening. The fourth area has to do with something called congenital Cytomegalovirus, and it is even though it's one of the most frequent viruses that affects human beings, many people have never heard of it. For most people who are affected with Cytomegalovirus, it has about the same effect as a mild cold.

Many people are affected and never know it. But if a mother develops or acquires Cytomegalovirus while she's pregnant, then there are serious consequences. About one in 200 children are born with congenital Cytomegalovirus. So that's about 20,000 children per year. And of that 20,000 children, about one in five of those children will develop permanent problems.

Unfortunately, this virus is not well understood, or is not in most people's awareness for taking care of it or doing something about it. This slide comes from a study done by CDC in 2016, where they ask a nationally representative sample of people if they had heard of these various conditions, and then they compared that to what the incidence of those conditions were. And as you can see here, even though Cytomegalovirus is one of the most frequent conditions, it is not. Most people have not heard of it and are not aware of it.

Of those 20,000 babies who are born each year with Cytomegalovirus, hearing loss is the most frequent consequence, although there are other very serious consequences as well. Interestingly, about four times as many babies who fail newborn hearing screening will have congenital Cytomegalovirus as in the general population. This has led more and more programs now to begin screening for Cytomegalovirus among those babies who fail their newborn hearing screening test. Although this is a very positive step forward, it still misses about half the babies who are born with congenital

Cytomegalovirus. So the real solution needs to be universal Cytomegalovirus screening.

Minnesota has started doing that. There's another pilot project in New York which is doing it. So screening for Cytomegalovirus is in a very similar situation right now as where we were with newborn hearing screening in the late 1980s. We know how to do it. We've demonstrated that it can be effective, but it's nowhere near as frequent yet as it needs to be.

Now. I want to take a slight detour now and talk about as just one example of how parents, individual parents, have affected where we've come to in our efforts to detect Cytomegalovirus and take care of it early. This little girl is named Daisy. She was born in March of 2011. She failed her newborn hearing screen, went back for an ABR a couple of weeks later, but they were unable to get it.

She had a number of issues during those early months in life, was eventually diagnosed with unilateral mild hearing loss when she was 15 months old, and then about three months later, was diagnosed using the stored dried blood spot that she had, Cytomegalovirus. Then that unilateral mild hearing loss progressed until she had a profound bilateral hearing loss, diagnosed at 20 months of age, and received cochlear implants at 21 months of age. What I want to talk about for a minute, though, is how Daisy's family was instrumental in moving forward with how children are identified and diagnosed with Cytomegalovirus virus and how they receive services. So this is Daisy with her brother and her grandmother, who just happened to be the majority leader in the Utah state Senate and a special educator by training. And Daisy's mother, who was also a special educator by training.

Daisy's mother and her grandmother went to work when Daisy was diagnosed with Cytomegalovirus and became profoundly deaf as a consequences and said, why

haven't more people heard about this? And they worked for several years to develop legislation in the state of Utah that was passed into law in March of 2013. And it was a law that required hospital or required the Department of Health to educate families about Cytomegalovirus and to create a public education program so that more people would know what it is and why it's important. It also, that law required the Department of Health to provide information to all of the daycare programs or other childcare programs, head start programs, about what Cytomegalovirus virus is and how it can be detected and what can be done about it. And so it focused on making people aware of what this condition is.

And then it also directed all of the hospitals in the state to test those infants who failed their newborn hearing screening test to receive or to have available testing for Cytomegalovirus at no cost to the family, and then to also educate the Department of Health that the Department of Health would educate medical practitioners about cmv testing requirements. Now, remember the slide I showed you a moment ago that said four times as many babies who fail the newborn hearing screening test have congenital Cytomegalovirus as in the general population.

It's been interesting to see what has happened with the support not only of the Department of Health, but with a lot of families in Utah advocating for and helping people to understand this. This shows how successful they were in testing babies who failed their newborn hearing screening test for Cytomegalovirus. So it started out in 2013. They could only do that successfully with 36%. But because of people at the Department of Health and because of family groups who became involved in this and continued to push for it, the number of babies who were successfully being screened rose over time to where, by 2018, 96% of the babies who failed their newborn hearing screening test were also being screened for Cytomegalovirus within the first 21 days of life.

They also implemented an extensive public awareness campaign. So there were materials printed on the sides of buses and light rail systems, billboards as you drove across the country, brochures that were distributed to families, seminars done for daycare providers, head start programs, and others who provide services to young children.

There's been a real effort to help people understand the consequences of Cytomegalovirus, and that has had a dramatic effect. It's interesting that it's not only affected what happens with children who are diagnosed with Cytomegalovirus, this shows when the law was implemented in 2013 and how many babies are successfully receiving diagnostic audiological evaluations after they fail their newborn hearing screening program. And so the emphasis on being able to test for Cytomegalovirus on those who failed has led to a dramatic increase in the percentage of babies who successfully complete their audiological diagnostic evaluation after failing a newborn hearing screening test. Similarly, the number of babies, the average amount of time it takes to complete that newborn audiological evaluation has gone down from an average of 40 days now to an average of 29 days. And this means that babies are able to receive early intervention and assistive hearing devices that much sooner.

It's also spread throughout the country. Now, this shows the number of states that have passed similar legislation, and most of this has happened because of a parent group called the National CMV foundation, which has taken what happened in Utah and been able to spread that to other states across the country. So coming back, the power of parent advocacy is something that we don't take enough advantage of. This shows the government of Utah signing the legislation that really started with this one little girl and was able to be implemented because of what her grandmother and her mother did. Now, obviously, they didn't do it all alone.

There were many other groups, university research centers, the Utah legislature, the Department of Health, the University of Utah, the primary children's hospital, which is the main level four NICU hospital in the state. But all of those groups, working together, have been able to move forward in a way that has dramatically changed the lives and the outcomes for children who are born with congenital Cytomegalovirus. But we still have a lot of work to do. We need to use this information in the past as prologue, but we need to move forward with what we're able to implement in similar situations. Now, the last area I want to talk about briefly is what's happened with advances in genetic screening and gene therapy.

Many people understand that about 60% of all congenital hearing loss has a genetic cause. Some of those are syndromic, some are non syndromic. We've made dramatic progress in the last 20 years in being able to test for those genetic mutations. There are over 200 different genetic mutations that are directly linked to congenital hearing loss. And if we can implement genetic testing, and I think we're on the edge of being able to do it in terms of both the technology and the cost, then there are dramatic benefits for children.

We can avoid unnecessary and often costly tests for those children who have congenital hearing loss. We can dispel information and offer emotional support for parents who are worried that their child may have hearing loss because of something they did. And perhaps in my mind, what's most important is we can anticipate other potential health problems. So we can identify children who are at risk for antibiotic induced deafness. We can monitor for other conditions, such as visual impairments or kidney impairments or things like Usher syndrome, which cause children to be both deaf and blind.

And then finally, we can provide information to families about the chances of hearing loss in subsequent children. I think it's important to note, though, that these benefits up

here really change the outcome of children who are born as deaf or hard of hearing. A number of people are now advocating that we take our physiological screens that are done with otoacoustic emissions and ABR. Those will not go away because there are still about 40% of children who don't have a genetic basis. But if we can combine the screening, we're doing with OAE and ABR, with screening for Cytomegalovirus and for genetic screens using the dried blood spot, then we can even further improve the outcomes for these children in dramatic ways.

Now, this is not something that's just an idea. In China, there are now a number of peer reviewed studies that are based on very large number of children. This study was based on about 240,000 children who received both physiological screening and genetic screening, and dramatically increased the number of children who were identified with hearing loss. As a result, this study had over a million newborns that were screened with both OAE and genetic screening, and again found a very significant increase in the number of babies who were being identified. So genetic screening is really on the cusp of becoming universally available.

I think with the right kind of advocacy and support from families and from professional groups, that we'll see these programs implemented just as widely as newborn hearing screening is now being implemented. Another very interesting aspect of the use of our understanding of genetics has just come out in the last few months, where a number of groups have been reporting the success of gene therapy with children who have one specific form of congenital hearing loss. It's a mutation of the Otoferlin gene, which causes auditory neuropathy, and only about probably 1% of children who have hearing loss would be eligible for this particular type of gene therapy. But it was first reported at the European Society of Gene and Cell Therapy last October that two groups in China had successfully treated children for the Otoferlin mutation, where they take a neutral virus that carries the correct DNA pattern for the Otoferlin gene. It's injected into the

inner ear, and then that correct genetic sequence multiplies and replaces the mutated sequence.

And children who had profound hearing loss before the injection are able to hear again or not again, they hadn't heard from the time they were born, so they were born with congenital hearing loss. But they go from profound hearing loss in some cases to almost normal hearing loss in other cases to a moderate hearing loss. But these are dramatic outcomes. The data were further reported in the New York Times in January, and then in a Lancet article, which reported several other centers that had found the same sorts of things in a journal editorial from molecular therapy journal in just this earlier this month, it was stated that the Oto Furlong gene therapy restores hearing in children. The data are compelling and astonishing and indelibly celebrate the ways gene therapy can change lives.

I think we have a long ways to go, but you're going to hear more and more about gene therapy. There are lots of issues that it raises, both from an ethical perspective and from a medical perspective and from a scientific perspective. But this is one of the areas where the past certainly is prologue. But what happens next is up to us, and we need to make sure that we have the right perspective. We need to make sure that we're doing what we can to move forward in a systematic and rational way with all of these things, whether it's how we educate physicians, how we use two way video conferencing to deliver services, how we better identify and treat Cytomegalovirus, or how we use genetic screening and gene therapy to successfully help these children and families.

I think we are on the verge of dramatic breakthroughs that will completely change how we identify and provide services to children who are deaf or hard of hearing. And the people who will continue to drive that, in my mind, are the families of children who are receiving services as they advocate for and work with agencies to implement these

things in a more widespread manner. Last slide is our website that contains many of the resources I referred to throughout this talk that are available for download. I look forward to working with all of you as we continue to explore ways to provide even better services and achieve better outcomes for children who are deaf or hard of hearing. So thank you for the opportunity to talk with you.

And Carl, thank you so much for an extraordinary history of where we've. Where we've been and where we've going, where we're going. And we all are so grateful for all that you've contributed to the progress that we have seen. I don't see any questions in the chat box. If you have one, you need to put it up there really fast because we are at the end of our hour.

And in the meantime, I think we should advance to the next slide, Carl, which has the questions and details on how to receive credit for having been part of this exam for your friends that haven't been able to attend today, this will be, it's been recorded and it'll be available and we'll have information about it on our website. And there'll be also information on the audiology online site as well, because this is a super important topic that all of us want to continue to be involved in. Let's see. There's one. Let me see if it's a relevant question that we want to talk about.

I don't think so. I think there's no other questions. So I think we'll just end here and thank Doctor White for being with us. And thank you to all of you for being part of this important course that we had today. It.