

Welcome everybody to this webinar on Childhood Hearing in the 21st Century, new frontiers in auditory development and recent discoveries in infant hearing loss. I'm **Jan Gatty** and I work for Clarke Schools for Hearing and Speech, and here I'm the Director of Services for Children and Families. I'll be your moderator for today and my first job is to introduce our other speakers.

Barbara Hecht is Director of Clarke Schools in Boston. She's currently President of the OPTION Schools International which is a consortium of schools for the deaf where children learn listening and spoken language. And she's also the co-director of project that we're doing with CREC-Soundbridge on telepractice and virtual intervention with infants and toddlers.

BARBARA: Welcome, everybody.

Our next speaker will be **Elizabeth Cole**. Elizabeth is the director of CREC Soundbridge in Connecticut, this is a public school program that serves children who have hearing loss and their families and are also choosing to use listening and spoken language as a mode of communication. Elizabeth has written a book, called *Children with Hearing Loss, Developing, Listening, and Talking* which is a mainstay in most graduate school curriculums for training teachers of the deaf.

ELIZABETH: Hi everybody, welcome.

You'll see on your screen two slides, one that talks a little bit about the Clarke Schools and one that talks about CREC. I'm not going to read the slides to you, but I want you to notice that our missions are very, very similar.

Both programs are designed to work with children from the time their hearing loss is diagnosed in infancy and to provide educational support for them throughout their academic career.

This is the first of a series of four webinars. It really was designed for early intervention professionals who are working with families of children hearing loss but do not have specialized training or expertise in that area. So it's an update for early intervention professionals. It's designed to be introductory in nature and to give us a global overview of current technology and practice and research in our field.

We know that children who have hearing impairment most of them have parents who can hear and have no experience with deafness. Part of their Early Intervention work is helping figuring out what kind of system of communication they want to use with their children. There are lots of choices, there are sign languages, there are spoken languages, and some families use a combination of the two. But our talk today is really going to look at listening and spoken language in children with hearing impairment when that is the choice of the family, because that's really the area of expertise of our speakers here today.

We've just been joined by another speaker, and I'm so glad Juliet could come by today to welcome you formally to our talk. I'm going to ask Juliet to welcome us and then I'm going to talk to her a little bit extemporaneously.

JULIET: I'd like to welcome you to the first of four webinars presented by Clarke Schools for Hearing and Speech and CREC Soundbridge. Childhood Hearing in the 21st Century. I hope you all learn a lot.

JAN: Thank you, Juliet. Juliet can you tell the folks a little bit about yourself, how old you are, what grade you are in school?

JULIET: I'm in the 3rd grade and I'm 9 years old.

JAN: Can you tell me some things you like to do in school?

JULIET: I like language arts and I also like...well math is okay.

JAN: That's a very politically good answer for this group! How about what you like doing outside of school?

JULIET: I like painting, gymnastics, I like to ice skate.

JAN: Thank you very much for coming. Juliet can walk to where we're presenting this webinar from school and she needs to get back for the afternoon. Thank you very much Juliet for coming.

Juliet was born with a profound hearing loss and she spoke clearly and fluently today. But if she took off her cochlear implants she would not be able to hear she would be completely deaf. I think the outcomes of children like Juliet are no longer the exception. Before the 21st century children with more hearing could learn to speak clearly, but today even children with severe and profound hearing loss can learn to listen and talk fluently proficiently and clearly. It isn't easy. It's not easy for the parents or the professionals, but in collaboration with each other, outcomes like Juliet can be typical for children born with hearing loss.

We're all teachers in this room today, and like good teachers we've come up with some learning objectives for you all. By the end of the hour, you should be able to describe 20th century advances in early hearing detection and intervention. You should also be able to understand the kind and type of tools that we use, instruments that we use, to diagnose hearing loss in children and what they tell us about that hearing loss and the child's capacity to hear. You should be able to identify hearing devices, sensory aids, that can be used to increase auditory capacity and what their purpose is and you should be able to identify all the necessary components of early intervention for deaf and hard of hearing children so they can learn to acquire spoken language clearly and easily through the sense of hearing.

The whole talk this morning is going to be looking at 21st century advances. I'm going to ask Barbara to take over now and sit in front of the microphone and talk a little bit about the first one.

BARBARA: Thanks, Janice. Again, welcome to everyone, whether you're following our slides or whether you're watching this webinar through our archive.

Since the turn of the millennium, we have made enormous progress in our ability to

detect, diagnosis, and support infants and toddlers with hearing loss and their families. The key to all of these advances is newborn hearing screening.

In the year 2000, just as the millennium turned, the very first federal grants were given to states to develop their Early Hearing Detection and Intervention programs. In the year 2000, there were only 21 states who received those grants, for the program that we now refer to finally as EDHI. In the past decade since the turn of the century we have made tremendous progress. In 2010, universal newborn hearing screening has become a reality in the United States.

Prior to the turn of the century, in 1999 the data we have showed that less than half of all newborns were screened for hearing loss. The infants who were screened either happened to be lucky enough to live in one of those pioneering states that had actually started to do hearing screening, or typically they were high risk infants, and infants who has spent some time in the neonatal intensive care unit.

By the year 2012, we really truly are at universal newborn hearing screening. Screening is absolutely critical as a first step, but screening is only a screening. Many infants who do not pass the screening actually have normal hearing, and so what's really critical is after screening, we need to complete a full diagnostic evaluation, and that brings us to our first opportunity for you, the audience, to participate.

We have a question for you that we'd like you to answer.

And if you'll look on your screen, you'll see that question: At what age can hearing loss be accurately diagnosed after screening?

If you just go ahead and click with your mouse or tap with your finger on your laptop, we would like you to let us know when you think, by what age is it possible to diagnosis hearing loss after newborn screening.

Okay. I think we're going to close our polls, so you can put your clickers down.

Let's look at the answers – most of you, about 54% of you said it's possible to diagnosis in the first days and weeks of life, and you are correct.

Some of you also said that it's possible to diagnosis at three months. And as you'll see in a moment, you're also correct. Our goal, really, is to diagnose as early as possible.

Let's see if we can go back a bit to the 20th century, in the old days, at that time only high-risk babies were screened and that meant that the average age of diagnosis was somewhere between 2-2½ years of age. And as many of know, that led to significant delays in their auditory speech and language development, and those delays in language development in particular had a very serious impact on all other aspects of children's development as well as their later academic and career opportunities.

Currently the EDHI standards, and remember EDHI is the Early Hearing Detection and Intervention standards. Those standards say that we should complete screening within the first month of life. By three months, we should have completed the diagnostic process. And by six months, a child should already be involved in early intervention.

So, for those of you that said we can do diagnosis by three months – three months is really our outside limit. Our goal is to complete the diagnosis before that time, especially because, as we'll discuss in a minute, some of the diagnostic tests that we use require the

baby to be asleep and after the age of three months, a baby might require sedation to be able to participate in those tests.

So, that leaves us then to the number two advance in the 21st century, having to do with infant diagnostics. Today, because the technology is so sophisticated and relatively inexpensive, we are routinely able, not only to confirm that an infant or newborn has a hearing loss, but we can actually report the degree of the hearing loss, the locus of the hearing loss, and we also have readily available technology that allows us to assess the structure of the ear, the middle ear, and cochlea. We're even at this point able to assess the integrity of the auditory nerve.

Just as a quick overview and a reminder for those of you that need a little refresher, this is a cross-section of the human ear. And as we look at this cross-section, we can see that sound basically, sound waves are collected by the outer ear and they're conducted right down through the ear canal, all the way to the eardrum. Vibrations are made when sound hits that eardrum, and those vibrations then are conducted along the little tiny bones/ossicles that are in the middle ear. The ossicles basically consist of the malleous, the incus and the stapes – that tiny little bone here – that sends vibrations along to the cochlea – to this snail shaped organ.

The cochlea is basically a spiral tube that is filled with liquid and lined with hair cells that are really microscopic nerve endings. When the vibrations hit the cochlea, the liquid and the hair cells begin to move. And it's the movement of those hair cells, those nerve endings that generate electrical impulses that are sent along the auditory nerve to our brain, and that is basically how we understand sound.

Traditionally we talk about two major types of hearing loss. The first type is what we call conductive and this is the type of hearing loss that has to do with conduction of sound on its way to the cochlea. So this would be hearing loss affecting the outer or middle ear. Often conductive hearing loss is curable or treatable through medications or surgeries or PE tubes, but there also are conductive losses that are due to structural differences in the ear that might be permanent, for example atresia. Sensorineural hearing loss is quite different and sensorineural hearing losses can range from mild to profound. They usually involve some damage to those hair cells in the cochlea or it's possible that the hair cells never developed. At this point sensorineural hearing loss and damage to those hair cells is not medically curable. Although there is on the horizon some research on hair cell regeneration.

But the new advance in the 21st century is that although we're still using this term "Sensorineural," we now actually can differentiate between a sensory loss, a loss that merely has to do with damage in some sense inside the cochlea and a neural loss, and the neural loss has to do with transmission of the sound along the auditory nerve and the auditory pathways in the brain. So, as we move forward, I think, in the next few years, you will see more and more discussion of sensory loss vs. neural loss. What's important to know is that most of the hearing losses that we're now calling sensorineural are really due to difficulties in the sense organ, in the cochlea.

So, what are the tests that we use? We have a wonderful array of electrophysiological tests for infants. Infants don't have to raise their hands and say "I heard that," or press a button - we have tests that require the baby simply to be still and the first of these is what

we call otoacoustic emissions. We also have auditory brainstem response, something called ASSR and some other tests we'll discuss in a moment.

Otoacoustic emissions indicate whether the cochlea is functioning. And I want to show you actually, in this slide we have a little animation that basically shows you how that test works. We put a tiny probe into the baby's ear, sound is played into that ear, and the sound reaches the cochlea.

What's fascinating about OAEs is that this technology is based on the discovery that a healthy cochlea echoes back sound that comes to it. So as you see that little blue ball moving along, it's really indicating that the cochlea has received a sound, is echoing back the sound, and the otoacoustic emission equipment picks up that little echo.

This OAE, Otoacoustic Emission test is not a hearing test, but it tells us whether all the parts of the ear are really functioning.

The gold standard and in many ways the test that you're likely to hear a lot about is the auditory brainstem response test. There's also new related technology called Auditory Steady State Response - and both of these tests indicate whether sound that enters the ear can be transmitted not only through the ear, but along the auditory nerve.

The way that these tests are done is by sending a sound into the baby's ear and then measuring whether the brainstem responds to that sound. It's important to note that the ABR really could be and should be done not only by putting a probe into the baby's ear, but also by using what's called an otoconduction, a conduction ABR – and in that case with the conduction ABR we're able to circumvent a possible problem in the outer or middle ear and look at whether sound that reaches the cochlea by vibrating the skull – whether that sound is able to reach the brainstem.

ASSR is really a new technology that many centers are beginning to use. It allows us to get very quick, ear-specific bit of information about how the baby is actually hearing by looking simultaneously at sound that's transmitted to both ears and looking at a range of thresholds.

So, here again, is a little animation to show you how ABR and ASSR work. Sound is played into the ear and we're looking at that ABR to see whether that sound is transmitted to the brain.

Key to all assessment ultimately is behavioral testing. We want to know the child actually is receiving sound. Behavioral testing is certainly not new in the 21st century. It was new actually in the 20th century, but we really do want to incorporate behavioral testing into our battery of high-tech tests. There are a number of ways actually that we conduct behavioral testing and it really depends a lot on the age of the baby, and the baby's ability to respond.

Other assessments that are part of the whole battery of tests that need to be conducted include tympanometry and bone conduction ABR. Those really tell us whether there might be a middle ear or conductive component to the hearing loss.

Also, acoustic reflexes are conducted; they are often part of the tympanometry evaluation. And those acoustic reflexes tell us whether the pathway to the brainstem in

back is functioning well. If a hearing loss is diagnosed, it is very often the case that CT scans are done in order to look at the integrity of the bone and the structure of the ear and MRI imaging can be used to look at the auditory nerve itself.

So, as a result, all of these advances, we can detect an infant's capacity to hear at birth, we can determine the nature of the hearing loss, and we can monitor the stability of that hearing loss over time.

It's important to remember that we should be monitoring that hearing loss not only at the time of diagnosis, but over many weeks and months. When we're testing tiny little babies, a lot can change, and so the process of diagnosis really is a process. It begins right after birth, but it goes on. And once the hearing devices are fitted, it's important to keep monitoring, to see how the baby or young child is learning to respond to sound.

So that brings us then to the third advance in the 21st century. We now have really increased the capacity to provide auditory access to spoken language to young children. There are a variety of devices that we can use to ensure that infants and toddlers, even infants and toddlers with profound hearing loss can have access to sound.

The particular type of device that we use depends a great deal on the structure of the hearing mechanism itself. And it's important to remember that the fitting of those devices really must be done by somebody who has expertise in pediatric audiology and the new art of fitting very young babies and children with hearing devices.

That brings us then to our next polling question. Jan, do you want to read this?

JAN: To read the question? Okay. At what age can a child be fitted successfully with a conventional hearing aid? Your choices are on the side, and waiting. Pick up your mouse or your pad and help us know what you know about how old the child can be for hearing aids. Okay. The votes are still coming in. We've heard from about 70% of you. Hit your button if you haven't. Okay. Pencils down; mice down.

BARBARA: Okay. So about a little more than 40% of you said it's possible to fit a conventional hearing aid within the first days or weeks of life and another 45% of you said it's possible to fit a device by three months, and then some of you said six months, and a couple of you thought it might be at a year or two.

Really, we've had a lot of discussion among the three of us. It is possible now to fit a hearing aid -- let's see if we can get to our next slide -- to fit a hearing aid within the first month of life; at 4-5 weeks. And it's certainly more common to see that hearing devices are fitted later.

The disadvantage of that, of course, is that if we fit those devices later, we're really giving the infant less access to sound and the brain less opportunity to begin to make use of that sound. So, hearing aids can be fitted in the first weeks of life. They basically are amplifiers. Hearing aids aren't new in the 21st century, but what is new is how miniaturized they've become and they can be fine-tuned through all kinds of digital technology to match the child's level of hearing. And we also have wonderful ways now of looking at what the child is actually hearing with the device, not only through behavioral testing, but also by doing some probes into the child's ear to see what sound is being

transmitted.

JAN: And because the process takes a while, the sooner you get started, the better. There are lots of appointments, so it's good to get started early. So, by three months and listening consistently.

ELIZABETH: And one of the other things, of course, sometimes it's taking a while to get the ABR testing done in order to have results that give you information that you can use in setting the hearing aids the first time. There are challenges with really tiny babies, but it's absolutely worth it, because the whole point is to get sound to the brain, just as Barbara said, and you lose less listening time the earlier that the child receives amplification.

BARBARA: And this, of course, assumes that parents have chosen to provide hearing aids and hearing devices for their children.

Absolutely.

BARBARA: And that really is a parent choice, and parents may choose to wait a little bit. They also may choose not to use a hearing device. But if spoken language is one of the goals, then getting that hearing device on early is critical.

There is also something called a bone conduction hearing device. This is used for some conductive or unilateral losses. It is delivering sound through a bone oscillator that the baby wears on the head and that oscillator transmits sound to the skull to the cochlea and you might hear Softband or Baha, and Baha is a particular brand, but there are some others, but this device is used more and more frequently and you will see -- you will begin to see babies, if you haven't already, you will begin to see babies in your early intervention programs wearing a Softband or even possibly wearing a bilateral Softband. Cochlear implants, I'm sure you've all heard about, and you know that those are for children with severe and profound hearing loss. They are FDA approved for children beginning at 12 months of age. We are now finding that in some cases, cochlear implants might be -- the survey might be performed on children or babies as young as nine months, if there is a medical indication that that is important.

In some countries, in some European countries, we're seeing babies receiving implants at six months of age if the diagnostic process is pretty accurate and confirmed. It's important to remember with all of these devices, they don't make a deaf child into a hearing child. These are still deaf children who are using devices that don't provide sound the way hearing people perceive sound.

And also, these devices must be continually reassessed, remapped, reprogrammed, and monitored. And that brings us to the fourth 21st century advance and I'm going to turn the mic over to Elizabeth Cole.

ELIZABETH: Thank you, Barbara.

So, I'm going to -- I have the privilege of talking a bit about this number four and also

number five for the 21st century advances. What's just wonderful is that as a result of all of the technological advances that Barbara was just talking about, fluent, clear spoken language is possible for most children born with any degree of hearing loss, as long as the hearing loss has been identified early and there's also appropriate technology, specialized early intervention services, and, of course, the very huge item that's needed is parent education and support and engagement.

So the opportunity is there technologically and the other pieces need to also be there, too, of course. This, I think, is a wonderful graphic showing that spoken language, early spoken language, is possible with the three things that we just mentioned. It's like three legs of a three-legged stool, and without one, the stool will fall over, basically. I'm going to be now talking about the professional expertise that's involved in order to help the parents be as engaged as they need to be and provide them with the support that they need.

So, what we're suggesting here is that what's really needed is that early intervention specialists need to be the ones who are kind of driving the bus for children with hearing loss. Luckily there are some states, Connecticut being one of them, that have specialty programs for children with hearing loss, which is absolutely incredibly important and we're really lucky that that's the case.

The fact is that new expertise is needed in the professionals who work with children with hearing loss. And because of two of the factors. One of them is that now we have more younger children, more babies and toddlers than we did ten years ago, even, that we're working with, and that's very different from working with a child who's two and/or older, which used to be the age of identification of hearing loss.

The other thing is that parents really need to be the central focus of the intervention. Helping the parents help the child and that, again, is very different from most people's training where you worked either with the child -- well, you worked in the child, or you worked in the classroom setting, so this is a different landscape

The focus -- so, we're describing this as a new body of knowledge, but I think really it's not -- some parts of it are new. Certainly the technological parts are new, but many of the other parts have been there for some time and there are plenty of professionals out there who have much of this information, but focusing it specifically on working with children who have hearing loss is perhaps what's new for some professionals.

What's really wonderful is that the Alexander Graham Bell Association through the academy has developed an association, it's new right now, but it's a way that parents and school districts are going to know whether or not the person that they're hiring is someone who really has the knowledge and credentials to help the children learn to listen and to talk.

The certification is called listening and spoken language specialist, shortened to LSLS and there are two types of certifications, either auditory verbal therapist or auditory verbal educator and that means they are working auditorily to help the child learn spoken language. The auditory verbal therapist works with children and parents primarily in that sort of setting. But auditory verbal educators are more likely to be working with children in specialized or general education settings. So, that's just a quick definition, I suppose,

for all of those parts that have to do with this new certification. That really does embody the information and knowledge that is needed in order to do this work appropriately and well.

So, again, the focus is on the same things we've already talked about, early diagnosis and assessment. The focus of the certification, I mean, is on those areas. Plus parent education and guidance and coaching and support. And a piece there we haven't talked quite a lot about is the parents need to learn what to do to maintain an acoustically-appropriate environment in all of the child's listening environments and also help them learn to use hearing as a primary sense modality, which is not intuitive because you're thinking the child cannot hear and it's important for the child to watch me all the time. Nowadays we don't think in those terms, we think in terms of giving the child lots of opportunities, not listening alone, and given the fact that they can listen and understand without having to watch. Obviously much of life includes watching, too, but as a technique and strategy, that certainly is an important thing for the professionals and the parents to keep in mind when working with the children.

And, of course, using auditory verbal techniques and advocating for the child's unique needs as an individual with hearing loss. Those are all part of what the focus is of this new certification. I'm going to go into a little bit of detail here because I think it's important for everyone to understand how vigorous it is in order for professionals to get the information and get the certification that we'll give them what they need in order to be able to do this work appropriately.

There is a written exam, which is a four-hour exam, I believe, and in order to be able to even sit for the examination, one of the requirements is that you have to have a master's degree in either deaf education, speech language pathology, or audiology. So this is a post-graduate kind of training that's required in order to prepare yourself for the LSL. Obviously you need to have the license or credentials to practice, but the post-graduate training is 80 hours in post-graduate study in auditory studies as well as professional experience, have 900 hours of working with children with hearing loss in the past three years and working with children with hearing loss and you have to have practicum with a person who is supervised and the recommendation from the parents.

The content areas on the exam include the ones that are on this list, and these, of course, are areas that any, really, excellent professional working with babies and children and toddlers who have hearing loss need to have, but these are the ones for the actual test. So, hearing science and technology plus auditory functioning, this is the science background, the scientific knowledge that's needed to help the professional understand and also exploit that incredible technology that we have right now, the flexibility of it and the capability of the technology we have for individualizing that technology to meet the child's hearing needs.

And obviously this is done in partnership with the audiologist, but it's often the professional who's working with the parents of the child who has the information from observation and from their own kind of informal experimentation with the child to notice things like the child not responding in ways that they should be by that point in their listening experience, and to put forward, you know, to have in their mind what the possible explanations for that might be based on their knowledge of what children ought to be able to do by that length of time of listening and using that technology.

It could be the child isn't, you know, attending, it could be that they simply need more gain; it could be a number of things.

Also, for example, if the child cannot say very many words together in a sequence, it could be that a change in the child's cochlear implant, programming them would give them greater access to a larger chunk of information and that something like that is the kind of knowledge that the professional needs to have to be able to tell their audiologist, "Hey, do you think we can do something about this? Because it really needs to be -- the child needs to have appropriate access."

Another area is spoken language communication, and on the next slide you'll see auditory verbal strategies for listening and spoken language development and there's scientific knowledge that has to do with language development and also normal language development and normal speech development, but then how do you help the child acquire it and help them become informed and be intelligible and how do you help them move into language abilities and complex language and being able to deal with abstract language and thought and higher-level thinking.

And, of course, child development, obviously that's an incredibly important area and I've already talked a little bit about the parents' guidance that's needed. How do you work with the parents? Because most people don't have that kind of education as part of their training, and so it's learning about family systems and also learning about different adult learning styles and being able to meet those demands or needs of specific families. And then other areas on the exam include the history, philosophy and professional issues. And the last two on there, education and emergent literacy are really aimed at providing that knowledge that you need and the skills and helping the child develop auditory and language skills that will support them and the general curriculum as a foundation for reading, as well as for all academic abilities.

So, I really like this, what do you call it, a graphic, because it shows that what we're doing is drawing on information from audiology, from speech language pathology, and from education, in creating this new professional, and all of that information drawn from the three areas is focused on helping children with hearing loss learn to listen and talk. So, that professional certainly is not working in a vacuum by any means. The LSLS specialist, listening spoken language specialist is absolutely working in partnership with other early intervention professionals.

And, you know, which includes someone who is, let's say, a general early intervention specialist or some other profession that includes OTs and PTs and speech language pathologists, sometimes co-treating using similar strategies and making sure that everyone is working with the child auditorily.

But the person that the LSLS professional works with closely is absolutely as a team is the pediatric audiologist, since fundamental to everything else, making sure the child's auditory access is as good as it possibly can be, so the wonderful therapy and talking that everyone is doing with the child is reaching the child and reaching the child's brain. One of the wonderful things about the new technology is that it allows us, new technology and new techniques and strategies, the wonderful thing and results of all of those things we're working developmental and not working remedially. Remedial work is what we used to have to do when there were already established patterns, so one has to sort of

stop using those patterns and then substitute new ones, but now working developmentally, it's possible to establish the right, appropriate patterns from the very beginning with children who have the advantages of early notification and appropriate interventional technology.

So, that's the end of my little piece here, on number four and five. I'm going to hand the microphone back to Barbara who's going to talk about some new exciting research findings that are informing the field today and into the future

BARBARA: So, as a result of all of the advances in the 21st century, we now have access to some new data, some new things that we've learned, things that we've learned as a result of being able to detect, diagnosis, and begin early intervention quite early. And I'm going to talk about a few of these today

Our next webinar that will be occurring in January will give us some opportunities to continue on this track and especially to look at what we're learning about the auditory brain.

Up until the time that we were able to routinely diagnosis hearing loss at birth and to screen babies at birth, we rarely were able to detect unilateral or one-sided hearing loss, and so children who had a hearing loss, even a very significant hearing loss in one ear, were often not diagnosed until they were well into the school years.

They notoriously had difficulties in school, they had behavioral problems or children with attention disorders or learning disabilities and the difficulty was they had a lot of trouble hearing in classrooms and noisy environments and may have even had significant difficulty in acquiring language to begin with.

And Fred Bess about 20 years ago kind of drew our attention to this group of children and I paraphrased what he used to say, but basically what we know from a lot of research is minimal or mild hearing loss as well as unilateral hearing loss can be extremely consequential and very, very important in the child's development, but because we're now able to test children at birth, we are routinely picking up those children who have a unilateral hearing loss. Less often we're picking up children with mild losses, because in some cases the screening measures we use are not sensitive enough.

So, what have we found about this group of children? It turns out in those -- there are a few areas in the country where people have been studying this group and depending on which center you look at, anywhere from 1/3 to over 40% of the children who are in early intervention programs have unilateral hearing loss

In some states and in some regions, children with unilateral hearing loss are not eligible for early intervention and this is something that we really need to all work on changing because we know that even after we have identified unilateral hearing loss, just identifying it is not enough, and the children who did not receive early intervention in the past few years are showing significant developmental problems.

Interestingly, some of the research is suggesting that a right ear unilateral hearing loss may be a little bit more problematic than the left ear unilateral hearing loss and that really has to do with the fact that the right ear is transmitting directly to the left hemisphere, to

the language centers of the brain.

We know from parents that any degree of hearing loss is significant for parents and one parent said to me, "The grief that I feel is not related to the degree of hearing loss, it's not -- you know, we don't experience the shock and the concern as parents when more mildly, it is a mild loss or it's a unilateral loss."

So we do need to provide parents support and there are a number of centers around the country that have actually provided some very innovative programs, specifically for parents who have had children with unilateral or mild hearing loss.

We also know that unilateral hearing loss might develop into bilateral hearing loss, and one center in the Boston area actually found that up to about 40% of children who were initially diagnosed with unilateral hearing loss had progressed to bilateral hearing loss. One of the difficulties we have right now is that there's a lot of controversy still about the best way to provide audiological support and early intervention support for children with hearing loss. Do you provide hearing aids or not? Under what circumstances? Are FMs enough? So we have a lot to learn still about the best treatment options and the best ways to support families of children with unilateral loss.

Another thing that we've learned, and that we really would not have known, although I think we suspected it, is that hearing loss can develop after the newborn period even in the early weeks and months of life.

This is an important thing to remember because there's a very recent study, Nancy Young who is a cochlear implant surgeon in the Chicago area has just recently published a study where she looked at all of the children who had received cochlear implants and then looked back retrospectively to see what had happened on their newborn screenings, and 1/3 of the children that we see with cochlear implants had passed newborn screening. So, what does that mean? It means that we need to now, as we move forward with our EDHI programs, we need to really be thinking about not just newborn screening and diagnosis, but early and periodic monitoring and screening.

A study in the Los Angeles area and one that has just recently been published, we'll post the link for you, one study found that about three in 1,000 toddlers who had sensorineural hearing loss after screening, when they were screened in well-baby clinics and one direction that we seem to be going in is look at ways to provide early intervention screening as part of well-baby care, in daycare centers and in other places that the parents take their children.

As early intervention professionals, especially early intervention professionals who are not necessarily working in the field of hearing loss, it's important to always investigate hearing if there are concerns about language development for behavior and attention.

ELIZABETH: That could be parent concern or school concerns, people who work in infant care are the people who notice that one child doesn't seem to be responding as well as another, but it can also be parents and, you know, you can get a hearing test without, you know, anybody else, you just call up and get a hearing test from a hospital or another center that does testing like that.

BARBARA: We are also now making great advances in our understanding of genetics in general, and the genetics of hearing loss in particular. I don't think we mentioned early on, but the statistics vary a little bit, but we know that more than 90% of parents who have children with hearing loss are themselves hearing and don't necessarily have any known family history of hearing loss.

But 50-60% of children with severe and profound hearing loss have a genetic cause of hearing loss and that's because it's usually a recessive genetic trait. We have now identified, partly through the human genome project and other research, NIH, we have identified genes associated with hearing loss that is not part of a syndrome and we know about other causes of hearing loss that are associated with certain syndromes. And interestingly, this is fairly new research, we have now actually discovered one of the genetic mutations that causes Usher's Syndrome and that causes hearing loss and gradual loss of vision in childhood and that genetic mutation that has been discovered has actually been turned into a test. It's not a test that detects all types of Usher's but it is now available if that is a concern.

About 1/2 of hearing loss has a non-genetic cause and the most important and the most common cause of non-hereditary hearing loss is cytomegalovirus, CMV, and the vast majority of babies show no symptoms of CMV at birth and it is often experienced by the mother as a mild cold or a little virus and so the parent really may not -- the parents may not be aware that the infant was exposed to CMV in utero.

CMV is often associated with hearing loss that develops after the newborn period and so there is now some very, very interesting research ongoing looking at the efficacy of combining CMV screening along with hearing screening in the hospital so we will be better able to follow and monitor those newborns who may have been CMV-exposed. As I mentioned earlier, we can now differentiate between sensory hearing loss and neural hearing loss. And we're learning a lot about a condition that has a number of names, but it's often called auditory neuropathy spectrum disorder and it may be responding well but there is transmission of sound and impulses along the auditory nerve.

Auditory neuropathy can range from a profound hearing loss to a mild hearing loss and even normal hearing and what is interesting about auditory neuropathy is the child's ability to perceive speech sounds is often worse than you would think based on their audiogram, based on the thresholds appearing that they are able to detect. We also know that auditory neuropathy is quite variable in how it progresses and there are actually cases where auditory neuropathy may be present at birth and disappear in the early months of life. It also can get worse.

And there's a lot of concern and discussion about the best ways to intervene. We do know that many, many children with auditory neuropathy do quite well with cochlear implants and that's not an intuitive finding, but it turns out, we think that cochlear implants will often help because they help to synchronize the transmission of sound through the auditory nerve.

This is an area you will be hearing more and more about. We don't know yet what percentage of the hearing loss population has auditory neuropathy. Some studies suggest it may be as high as 10%, others say at more, like, 4-5% and I think to some degree that is on how auditory neuropathy is defined and what criteria are used to

diagnose.

ELIZABETH: And you may know that it comes earlier.

BARBARA: And very quickly I just want to give you a little teaser for our next webinar. We do know that there are sensitive periods, sometimes we used to call them critical periods, for development, and especially there are sensitive periods for auditory development and the impact of auditory stimulation

We've seen in children like Juliet, who you heard earlier, that if children with profound hearing losses get early access to found and expert intervention, they can really learn to listen and talk quite well.

We now have, at the biological level, some of the evidence that this really is part of neurological development. So if you look to your left on this slide, this is actually a cross-section -- or a section of an auditory nerve fiber in a normally-hearing cat. This research was done on cats.

And if you follow along here, you'll see this is an auditory nerve fiber and where those little stars are, those are synaptic connections across the nerve fiber, they're transmitting sound information, and this is what an auditory nerve fiber looks like in a hearing cat, or in a person.

This is what the auditory nerve fiber looks like in a deaf cat and you can see that the fiber sort of peters out, and what's important is the areas that show none of the synaptic characters. The deaf cat was given cochlear implants quite young and this is now the auditory nerve fiber in a deaf cat who received an early cochlear implant and you can see the development of that auditory nerve fiber looks almost identical to the auditory nerve fiber in a normally-hearing cat. You see the vestibules.

What we're learning about auditory stimulation, it really does change the way the brain develops and grows and changes the auditory pathways in the brain, and this will be the topic of our next seminar.

JAN: Okay. I understand that my voice falls off at the end, so I'm going to make an effort here to summarize.

First of all, I want to thank both of my colleagues, we had a really good time working on this project together and it was nice to hear it today. I'm just going to summarize quickly the landscape for deaf and hard-of-hearing children has really changed in the 21st century and this is really because of universal newborn screening, at birth, the ability to diagnosis hearing loss in young children at a very young age, the ability to provide them with auditory access for spoken language so they can hear the sounds of speech, and differentiate them well enough to actually learn to produce them clearly.

We had new professional expertise in this area, we had a new certification. We're really prepared to help people working with children who don't have this specialization to work with them as a team, and there are also new research findings simply because we know what the population is from a very early age.

We began with Juliet, she's pictured here with her sister Sophia who also has a hearing

loss, and she really is a product of all of this new work, and she's the kind of child we're looking at and continue to find out how she can learn to talk and hear.

We have one bit of housekeeping before you leave. You will be receiving a questionnaire in your e-mail, if you participated in this seminar today, you will receive this tomorrow, and we really -- it's really important to us that you respond. It's the only way we have to know how we're doing and what you need, and so your responses will be collected by a third party, they will be anonymous, we encourage you to be kind, careful, and critical, but it's important to us to know.

I think -- I will just leave you with this last message, we'll have to deal with your questions by e-mail, I think, but we hope to see you again in January when we will again be together talking about from the ears to the brain, auditory perception in infants and toddlers and the development of the listening brain.

And for a repeat of this webinar, if you had to leave or were eating your sandwich and you missed anything, you can go to this website, which is part of the Clarke School website and the webinar will be online for your reference.

Anybody else have anything to say?

Thank you everybody. Thank you.

JAN: Thank you.

Bye-bye.

(End of presentation).