\* \* \* \* \* \*

Congenital Cytomegalovirus (cCMV) - The Most Common Cause of Non-Genetic Hearing Loss

&

The Missing Link: Early Intervention Services for Babies Born with Congenital CMV

A black background with a black square

Description automatically generated with medium confidence March 19, 2024

11:00 AM - 11:25 AM | Congenital Cytomegalovirus (cCMV) - The Most Common Cause of Non-Genetic Hearing Loss | Mineral Hall B/C

&

11:30 AM - 11:55 AM | The Missing Link: Early Intervention Services for Babies Born with Congenital CMV | Mineral Hall B/C

\* \* \* \* \*

This text, document, or file is based on live transcription. Communication Access

Realtime Translation (CART), captioning and/or live transcription are provided in

order to facilitate communication accessibility and may not be a totally verbatim

record of the proceedings. This text, document, or file is not to be distributed or

used in any way that may violate copyright law.

\* \* \* \* \*

(Captions provided by a live Captioner.)

»  Hello. I think we're going to go ahead and get started. I want to give Ruth all of the time for her presentation, I'm Sonia Richardson, moderator for the session and this is Ruth Marin, she's going to tell us about Congenital Cytomegalovirus, the most common non genetic reason for hearing loss, and we may or may not have time for questions, but Ruth?

»  Thank you all, as Sonia said I'm Ruth Marin, Pediatric Audiologist in the Washington D.C. area and have been since 1988. Some of you may not have even been born yet.

[Laughter]

About 13 years ago I was hired to develop and manage a newborn hearing screening at a hospital outside of DC, so that's my focus, not doing diagnostic or hearing aids anymore. In 1990, I had a patient in DC multiply involved, and somehow, we knew she had Congenital Cytomegalovirus in 1990, I don't know now, we didn't hear the test and really, I didn't hear much about CMV until a few years ago.

When I read about how prevalent it was and how much it impacts us, I reached a colleague in Delaware and formed the Mid Atlantic CMG Consortium, and we realize the legislation paths broke off, so into two groups. We have audiologist, doctors, parents, we have over twenty members and now 21 to 22.

Because I met Amy back there who also is in Maryland and I, like, grabbed her and said she needs to be part of our group.

[Laughter]

So anyone in Maryland avoid me or you'll be involved.

[Laughter]

It's a hot topic, there are four sessions at this for CMV, and I always learn something new. A lot of my slides the in presentation are backgrounds, and I won't have time going in—depth, but if you have seen other presentations or attended workshops CMV, you will see the slides.

And finally, the best appropriate inclusive terminology to describe hearing status, and I'm going to do my best to honor that. But I'm old. It's hard for me to get used to using old terminology, so I will do my best and I apologize in advance if I slip up.

Okay. I'm sure all us know CMV is common and part of the herpes family, most people, down to 80%, most clear up, but, —— sorry; the virus was first isolated in 1956. And named in 1960. So it's taken 60 years to really get to the point where we have so much information, and it's the most common non genetic cause of deafness.

To diagnose CMV, it has to be made in the first three—months of life to separate congenital for acquired. I'm old—school, I have notes on paper. And CMV is transferred through bodily fluids and anyone with kids, we know there is no lack of fluidly, we're digesting what we know that and so the women are at risk because of how it transmits, you can get Congenital Cytomegalovirus from mom's primary infection or reactivation of virus in her system. Just like if you have had chicken pox years later it reactivated and becomes shingles, that's the best I can explain it probably completely accurate.  But when a women contracts CMV, there's a 1 in 3 chance to transfer it to the baby. Research suggesting that we can give medications in the first trimester, but we don't test mother's for CMV so how would we know? I don't know. That's more common an all those combine 1 in 200 babies will have CMV, and some will have asymptomatic, the specialist on your committee said that no clinical manifestations means there may be things there you just don't see. So I'm going to go back and forth with the two terms but they're not completely the interchangeable.

More children —— whoops sorry —— so this is a list of some of the complications and not all, but sensory neuro—changes from birth or progress are the most common. It's a lot. Having been a pediatric audiologist for such a long time, and having seen children with multiple involvement, how many might have had, how children might have had CMV and we didn't know to do anything about it.

More children have other disorders because CMV, but the CDC reports 90% of women have never heard of CMV, and we can't prevent it but we can reduce the risk with of basic precautions. Don't put your children's pacifier in your mouth. I never understood when parents thought that was cleaning it. Don't use your child's spoon, wash it all the time. Just very simple things we do to avoid other illnesses. I think I missed —— sorry, there's a slide? No, sorry.  My papers are flipped.

We have different ways of screening for CMV. And the first is the hearing—targeting screening.

There are some states and locations that do that. And that is just screening for CMV on the basis of a failed newborn hearing screen.

The problem is it's not very effective.

Because most babies who don't have other symptoms, or other manifestations may pass the hearing screening and it occurs over time, this is about 7% of babies that have a change in hearing status. I started the first targeted program in the State of Maryland listed in the CMV website and over a year and a half we have not identified any babies just on the failed screening. The next step is expanded screening, and we are doing this at the hospital also. Based on other manifestations, babies with seizures, intrauterine growth and things like that. For example, we have a baby born in NICU, and small for the age and a CMV screening was ordered and when it was positive and then that baby failed the, the newborn hearing screening.

So at that point we knew to refer the child to the infectious doctor, for results and getting into the remain for all the different evaluations and treatments that would need to be, one scheduled.

And then finally, universal, based on your screening experience, we hope that will be the gold standard. For everybody to do the universal screening.

So this is the map of the United States and where the states are in legislation for the CMV screening. The teal, darker color those states have some or the of legislation for CMV whether it's education, screening or both.

And just we're so excited that about, I don't know, two—weeks ago, Senator Blumenthal from Connecticut, anyone here from Connecticut? They introduced a bill for screening, my understanding is it's similar for the EHDI funding for states to set up their own programs.

I have not read the bill yet, so I can't tell you the differences there.

The first state to pass the legislation was Utah in 2013, I don't have time to go into the story but sufficed to say don't mess with a grandmother in the legislature. The gray with the states with stakeholder interest. They may not have gotten as far as actual laws, but are working toward it and now thanks to our group, I'm proud to say our state is gray. And Minnesota was the first to have universal, and that's the gold standard. Just a conversation with me and someone in the program in, they identified 40 newborn babies, that's positive but only 2 failed the screening, so that's 38 babies missed, to see if they have Congenital Cytomegalovirus and would develop symptoms, because not every baby does.

So I mentioned we do in my hospital a hearing targeting screening, but we know it's not enough. And it's not enough, because any child that doesn't fail the, the newborn hearing screening, and also does not have any other complications will get missed.

So about half of the children who ultimately develop a change in hearing status, will have passed their -- the newborn hearing screening.

So for any of you who are the newborn hearing screening audiologists, we're missing so many babies by not doing the universal screening, so versus universal screening, we again, all we all know what that means. But it helps us identify children with asymptomatic children with CMV status as well, as other issues and co—morbidities, and allows for all monitoring and facilitate servicing and treatment. I don't necessarily mean medication when I say treatment. Although, that can be part of it. Anti viruses are being used to me and probably to all of you, early intervention, access to language, hearing aides, cochlear implant, to me that's treatment and if we don't know about it we can't work with the families.

  In August, a few of us in my consortium met to discuss, and we met the panel, and they were receptive. The chair was brilliant but had no knowledge of hearing in screening and she said we should not recommend screening for children because it places undue burden on partners. Pediatrics always screen babies' hearing.

And parents will know when their children's hearing status is changing.

I was dumbfounded. And I did not say what I wanted to say.

[Laughter]

I won't say it here either.

[Laughter]

But we were just blown away by that. We're going to go back, gather more information and go back to the counsel to rebut their issues and questions and hopefully be able to move forward. I was really surprised because Maryland is typically known as a more progressive state. We all know the benefits of early intervention so we're not talking about that. Screening methods: Saliva is standard, $15 covered by most insurances urine testing is... What's the word? Not effective, we're not going to catheter rise maybes or have nurses squeeze diapers, so that's not effective. However, newborns have the blood spot tests, and the Minnesota data, it shows that, that the sensitivity is really improving. So hearing status. 25% of all sensory hearing differences are thought to be due to CMV. That's huge, with or without symptoms and this got me when I first starting, learning, it can be congenital biological rate, and fluctuating, and for any audiology, pediatric audiologists in the audience, I I'm sure being you think about cases where kids hearing were fluctuating and didn't know if they were playing around with you, suspecting middle—ear changes. We just didn't know. We know the hearing can progress through five, and have years but it can continue and can continue and we see the hearing has changed and we don't understand why.

Sorry.  So there seems to be a higher proportion of hearing losses in the systematic babies, however there are so many maybes that are asymptomatic, so the numbers are higher. I hate quoting studies because it's boring, but there's a few I wanted to show you and are links if you want to look more. This is the Minnesota data, tests 444 babies and about 40% of the babies passed the newborn hearing screening and later determined to have a change in status. 52% with a unilateral status developed bilateral status over time and those that received antiviral medications did have some positive hearing but it's not standard of care to give antivirals with hearing, there's a lot of risk with antivirals and we're not yet. From the CDC, over 366,000 could be born with CMV, that's not all having changes in hearing status; but that's a lot of babies for us to monitor.

The next two studies are, um, from the same group. And you can see here that um, out of 92 babies 65% had a progressive change in hearing, 25% passed the newborn hearing and then developed a change in status, most progressed to bilateral and I'm seeing the same things over and over again in the research. This is estimated; 900 babies annually have hearing loss greater than 70 DB in one year by 12 months so again, it's progress so if we don't know we can't monitor. The same group studies symptomatic versus non, and 75% did have central hearing differences, so again looking back to all the numbers and the incidents, it's really a lot of babies.

All the babies identifies based on the newborn hearing screening had a decrease in hearing status. All of them in this group and again the greatest was found in 5 to 6 years, but also other brain abnormalities were also associated with hearing differences, so I already mentioned this, this is just a summary to say multiple studies with unilateral, most develop to bilateral.

I don't know if there's any physicians in the group to explain that to me, but I'm not able to explain that. And in most cases the poor—hearing ear worsens more quickly.  So the recommended timeline for screening for hearing, there's no standard but the American Academy of Audiology just releases a few months ago have guideline. Minnesota is age ten and AAA is age 6, we're using the American Audiology guidelines. When I understand don't, if we know hearing can change until age 18, why not just screen? How hard is it to screen once a year, you know, even after age 6? I don't know who to ask that question to, but that is something that's bothering me.

Antiviral therapies are being investigated and used but only with kids to moderate to receive, and like I said, there's a lot of risk and you just don't want to do it without considering them. There are vaccines in tribal and showing promise, I attended an NIH conference a few months ago where I understood almost nothing, it was all molecules and way above my head but they talked about if we have this, do you vaccinate kids, women? The informal was to vaccinate children to not pass it along, about the you don't vaccinate every child, so if you are a women who's working or have children, you're still not going to be completely protected.

So there's a couple of organizations that have been supporting CMV screening. American Academy of Audiologist recommends early identification, but doesn't go as far as saying they recommend universal screening. My personal opinion, just mine, they didn't go far enough with this. They had the ability to have this platform and didn't do. At the Academy of Otolaryngology, over, February something, a month ago, did recommend universal screening and we're awaiting the pediatrics’ statement, we don't know what it's going to say. The doctors on my committee don't know, but we're all very, very anxiously awaiting. CMV, it's something like, every other workshop here as been about CMV and with good reason. We know the cost of early identification and intervention is cheaper than later and very briefly, I kept this in my presentation, there are flyers.

This is about audiologist monitoring and all available on the website for free, you can print them out. Some are at the exhibit hall, but all of the funding comes from donations, so if you can printout your own, that would be great.

They have supports and next steps in English, my hospital paid for the translation into Spanish, about 95% of my patients are Spanish, and we're donating this to the national CMV Foundation but haven't put it on the website yet.

Again, all the evaluations you need and again in Spanish and hopefully be on the website, there's links and resources with and a QR code and about 40 articles I have read and been working on, hopefully you go back and work on CMV screening in your state, if it's not already in process. And I know we don't have a lot of time, so I'll stand outside and if anybody has a question, great. If not, e—mail me.

» Okay.  I think we're going to go ahead and get started. I'm Sonia Richardson, I'm the moderator for this talk and I'll like to introduce The Missing Link, Early Intervention Services for Babies with CMV.

»  Welcome, everybody, thank you for coming. With me today is Meredith from Boston Children's Audiology, give a wave. Next to her is Brenda Allair, Assistant Director for the Consortium of New England Deaf and Blindness. Before we jump into today, we're really here for parents.  Lisa Saunders is a leading advocate for CMV screening and taught us about hope, perseverance, collaboration and supporting each other. Daughter Elizabeth was born with CMV and passed away at 16 in 2006. When I first met Lisa, she asked what I did and I told her deaf blind program, and she said I never realized my child was blind.

Vanessa's son Logan was born with congenital CMV, and her son passed away at 4 months and she is advocate for CMV awareness, and these parents teach us every day about collaboration. Our objective today is to show you the collaboration with parents, healthcare and educators and will highlight across disciplines for early referral and timely services for children and families.

As I began this journey with the coalition, I stood back and thought okay, I'm a federal grant. Where do I fit in here? This is for my brothers and sisters out here corrected to deaf—blind projects and thinking why is CMV so important? As I started working with the CMV coalition, we ban to focus on what's really important. How can we move the process for children and families? And we identified educating providers around CMV and congenital protocols and practices and finally, identify champion for screening and legislation.

As a deaf blind director, I realized our charge, this population we have very shared common goals around intervention, at—risk youth, family support and feel oh, poll change around CMV screening.

And I started to look at CMV as a wicked problem.

I know in Boston that is overused, but it is a wicked problem.  We know it's preventable and 1 out of 3 women who become infected with CMV will pass that onto their child. We know 1 child is infected with CMV as we sit here, we know 1 in a 100 children are born with CMV each year and 1 out of 5 will become disabled. Yet so many states don't have screening.

So CMV, as a wicked problem, is difficult to solve. It has interconnecting facts and issues, it is a interdisciplinary approach. It's time consuming and interive; meaning we have to relook at protocols and finally, across many disciplines.

So this slide, again, underscores why CMV is a wicked problem. If you look at things such as aids, and spinal issues, this is so much.

We did a workshop recently and asked how many people heard of CMV, only one person raised her hand, and she was from outside of the United States. It was shocking to us, because CMV is such a prevalent issue. So, we started thinking how to educate folks around CMV, and highlight areas and they're really complex kids impacted by CMV, often hard to monitor and have frequent evaluations and hospitalizations and we also found an incredible amount of these families have multiple contacts not communicating with each other, and who better than intervention providers really on the front lines working with these families on the day—to—day basis?

Another important piece I think is really important for today's take away is around late identification.

Symptoms of CMV may be overlooked leading to diagnose disordered without appropriate treatment.

An example of that I want to share, a little guy I have been working with about 6 months and I worked with his team last week, he has seizures, and cortical issues and the team reported he's not interested in music as much as he used to be. So I looked at the clusters of issues being presented, and said when was his hearing last checked? He's 7 now and was last checked at 3, so again, having all these issues and later Brenda will talk to issues related to the visual impairment. So for us, the link is the early intervention, earlier training and that includes staff, DLI, SOPS, therapists, you name it, we wanted it to be concise and often for families and really, ideally, what different providers should be focused on. So our module has assessments, PowerPoint, key domains such as auditory, vision, communication, tactile, motor, social—emotional and self—help. And with that, Becky and Meredith are going to be going into more, on some different aspects of the module.

That's right, Tracey, as introduced I'm part of the Massachusetts Coalition but also a medical student so they asked me to talk about CCMV, so what we really wanted to provide a snapshot of the information in the module, if you are able to use it and again, identifying that missing link Tracey and team identifies for one aspect of the CMV kiddo but don't see the other symptoms or are unable to link them. So impacts, as listed, microcephaly, seizures, periventricular calcifications, sorry I'm going a little quick.

[Laughter]

[Laughter]

But a number of um, MRI abnormalities and one of the important things that you can look out for, when MRI abnormalities appear, that can be worse outcomes in the long term so that's just the information we haven't to provide to give early intervention providers background of what to except, and what to look out for. And we think this is an important component, as about 50% of CMV children that are asymptomatic have these, so just so you are just aware.

And as Tracey mentioned, there are long—term CMV that may not present initially but may come out later on, like with the hearing.

So here's another snapshot of the module, and things you may see come out later on with CMV.

Interestingly, CMV is the most common use of delayed psycho—motor, which I did I want know, so this is something to be aware of and have eyes out for.

»  Hello, everyone, I am Meredith, I am an audiologist and I'm here to very briefly share some hearing differences absorbed with C—CMV and now being at the EHDI conference and those hanging around for the session before us, you are experts in this area perhaps, but to get everybody on the same page and shared in the module, C—CMV is the leading non genetic hearing, accounting for 25% of hearing differences diagnosed in children. We also know that C—CMV related hearing differences can develop with time and that really prompts the recommendation to hearing testing in the kids. C—CMV do are varying presentation across kids and even across time. So these hearing give retentions an be for transit or can be progressive developing over time and can fluctuate over time. It can affect one ear. It can affect both years. And we can see it at birth, or we can see it develop later on. So again this really prompts the need for us as audiologist to regularly monitor these kids.

And some signs and symptoms of these hearing differences, we want to share with the early hearing interventionists, we want to list them here. For instance, infants surprised by loud noises, not responding to their name, it's not all of them, but no things for generalists to look out for and we also know also that C—CMV they may be hard of hearing—plus, so not turning the head would be not necessarily only hearing but maybe motor skills, so it's something an interventionalist can say hey, let's look more deeply.

I get to be the final hitter here, I guess. One key we wanted to focus on the module was helping those children with vision concerns who have been diagnosed with C—CMV there's a lot of conversation, and rightfully so, for the hearing issues with C—CMV but for the children with the neurological impact, some kids are vision issues is less researched, as we were developing the model with A, finding the information about C—CMV and hearing there's a lot of robust information and that's wonderful, there are mentions through the that hearing research around vision but the really intense research just hasn't been done quite as well. So some of the ocular vision conditions that can be present, these are things obvious in the eye on an eye exam. So sort of a cross eye, that shaking eye. Issues with the optic nerve. And very specifically with C—CMV is damage to the retina. These are things that can be diagnosed with an eye exam. But again, these are children with those neurological impacts it might not be as obvious so they're getting so many other interventions this is far down on the list and we really want interventionalists to be aware of that, additionally we know brain—based impairment is significant for these children. Tracey mentioned children she's work, I, I have several I'm working with as well, and this is often attributed to other needs, maybe the hearing difference that's been diagnosed and we really want early interventionalists to be aware and to monitor these children. So what do we know about C—CMV and the CVI, which is cortical brain-based impairment? We know CMV is the most common viral infection and CVI is the most common visual impairment, that's the two most common and we want to be aware of that and due to the nervous central damage, which obviously weary of we've talked about.

You may see a different MRI for there's children and often have severe loss in the vision field and there are really spare reports in the literature, but we do suspect this is happening for children, part of the module obviously we'll talking to early interventionalists about these impairments so they can be aware and monitor, these would be things symptomatic of ocular impairment and—or a brain—based impairment, because we want them to be aware. So things like visual attention, preference for highly colored toys and falling with something that's atypical for a child with their level of motor involvement.

As Tracey said, this is really about families and we want families to know there's support for them, so those positions by relationships with the families, early intervention with provide clear concise information for families. We know here at EHDI the critical difference early intervention can make and we want them to have support along the way, I didn't hear about CMV, I never heard it before? Is it my fault? Right? All the feeling families have and there's not places to go for information and resources, we want to make sure families are connected. One of the key parts of this module is going to be family stories. And so as part of the module, participants will have access to videos and information directly from families themselves. Talking about how CMVs impacted them and their child.

So overall, we had some targeted outcomes. Um, for developing this module. We want to increase the awareness of C—CMV. Alongside medical providers. Along the conversation here at EHDI has been wonderful, there's so much C—CMV information getting to families and we want that information getting to children and families and we want to make sure these children diagnosed and especially for hearing and deaf blind, and we want to make sure they're referred early to appropriate programs, programs to deaf and hard of hearing for language acquisition even for these really complex children, we want to talk about follow.  You and intervention and we really want to make sure we're following best practice. We know children impacted by C—CMV benefit from sensory, and neuro follow up, but if nobody's look for those things, they're not going to get into the programs. So being aware of possible hearing differences and being aware that could progress. As Tracey said before, it progresses and making sure ocular differences are being assessed and being assessed by people that understand the impact of C—CMV so this is a QR code that you can scan. And you can get registered to know when the module is finalized. We're still tweaking a few little things. We also have contact information, on the back, so if you want e—mail or additional information. And Tracey, I think I actually left the memory stick over there?

[Laughter]

[Laughter]

I put, together, just a quick view. Of the module itself. So show you the module itself. So if I'm lucky... I might be able to open it? We'll see. It's not opening.

Yeah, what's what I'm wondering. Let's try. Anybody want to try?

[Laughter]

[Laughter]

[Laughter]

Is that here? On the screen it doesn't show. We'll try. Give us a minute sorry.

[Laughter]

[Laughter]

It worked when I was in the presenter room, so of the course it won't work here. Okay.

»  Try that?

»  Technologies are great. There we go. Part of this is pre, and post survey. We may not be quite as successful as I had hoped here. This will be shared when you scan the QR code.

[Laughter]

We were very excited to show you guys today, but there is a pre, and post survey and check for the providers, so as the completion of the module, there's preexisting checklist for kids with dual needs, and our hope is this will be embedded in existing training, so state—mandated training, right? So we're not asking for additional, we just want to embedded in systems that exist. I'm sorry we couldn't show you today but thank you very much for your attention today.

[Applause]

[Applause]