How to Start a cCMV Screening Program, in partnership with the American Academy of Audiology

Presenters: Angela Shoup, PhD, Maggie Kettler, AuD, Megan Pesch, MD

At this time, it's my pleasure to kick off today's webinar, How to Start a CCMV Screening Program. Today's webinar is in partnership with the American Academy of Audiology. We have three presenters. I'm so thrilled to have them with us today. Dr. Maggie Ketler, Dr. Angela Shoup, and Dr. Megan Pesch.

And I invite you to read more about our presenters on our website. In the interest of time, I'm going to turn the webinar over to our presenters now.

Great. Thank you so much for this opportunity. Let's jump right in. Before we start, just a quick word from the Academy.

Welcome. I'm Tish Gaffney, the president of the American Academy of Audiology, and I'm here to share important information about the organization. The Academy is dedicated to achieving recognition of audiologists as the experts in hearing and balance care and the field of audiology as an independent and clinical specialty that is integral to the healthcare team. Through our partnership with Audiology Online, we're excited to offer courses like this one to help you broaden your knowledge and skills. We also offer the latest resources in practice management and evidence based clinical practice so you can grow professionally and improve patient care.

We advocate for our members in Washington and across the country to ensure that the voices of audiologists are heard and we help connect audiologists to one another through our valuable networking opportunities like our annual convention. To learn more about the Academy, make sure to visit audiology.org thank you.

Here are our disclosures, limitations and risks and then our learning outcomes.

So today we're going to talk about why congenital cmv? Why should we care and what is it? How do we screen for congenital cmv? Making a plan, gathering your crew, who should be screened, screening

tests. And we'll talk also about follow up.

We're also going to be real about bumps in the road and some tips and tricks that we've learned from our experiences.

So I'm a developmental pediatrician by training. CMV was not in my repertoire at all until about seven years ago now when my third daughter was born. Throughout my pediatric training we only had two slides about congenital cmv. I actually went back and looked at my notes, but it took me a while. There was nothing in there about the transmission or really screening.

But things have come a long way since then. So seven years ago I was pregnant with my third daughter. Healthy pregnancies, or so it seemed. I gained weight, the baby gained weight, and she was born looking perfect. I mean she was about just under eight pounds.

A good sized Baby, normal head circumference. She did have some little dots on her face, but it was a bit of a rough delivery, like a little ugly. So that can happen sometimes when babies get stuck, but she didn't have them on the rest of her body. And when she referred on her newborn hearing screen, we were like, oh, no big deal. I mean, I'm a pediatrician, so I had spent my career talking down anxious moms.

And so I was like, this is my third kid, I'm not going to be worried. This is like obviously fluid. And we took home our baby girl and just loved on her. Around 10 weeks was when we went back for our repeat screen, which turned into a full diagnostic. And of course, at that time we learned that she had profound bilateral sensory neural hearing loss.

It took an additional, gosh, eight weeks or so for us to figure out that she had congenital cmv. And that's because she wasn't screened at birth. And when you are testing babies that are a little bit older for cmv,

there are a lot more steps you have to take and it takes some time, which we'll get into in a few minutes. But she was outside the window for antiviral treatment. She had findings on her head ultrasound and this was really like a missed opportunity.

And I had spent 10 weeks looking at this baby 24 7, and not once did it cross my mind. And it was just a real wake up call for me and not to be angry at the other providers that took care of her because, yes, this was a miss. But you just cannot pick up that these babies have congenital CMV for the most part, unless you're screening them. So that's why I kind of pivoted my career and I'm now seeing patients with CMV and doing research in this area. So what is congenital cmv?

It's a common DNA virus. It can cause mild, like cold symptoms. Not a runny nose, more like maybe a sore throat and some lymph nodes. But really most of the time it causes zero symptoms. And usually I say it's generally a no big deal virus.

It's something that's passed around. Very commonly, most US adults have been exposed and it's only a big deal if you are a growing fetus or if you are an immunocompromised adult. Otherwise, which, you know, the most of us, it's just a no big deal virus and you would never know.

Now, there are different types of transmission of cmv. Horizontal transmission is from person to person, so born person to born person. So often Happens is a baby or a toddler at daycare. You know, toddlers are putting everything in their mouth and sharing all the germs around. And they'll.

And I'm sure what happened in my case, because I had two kids at daycare at the time that I was pregnant with my third daughter, was I would eat off their plate. So some they bit a waffle. I would be like, I'm finishing that waffle. I was hungry and busy and tired. So of course I was eating that waffle.

But I was also eating probably their CMV germs. And then vertical transmission is when it's transmitted from a pregnant mom, pregnant person, to the growing fetus. And so that is transmission from me to my daughter. And in that case, when it's transmitted in utero, that is congenital cmv.

So usually we put CMV into these two buckets, and we recently started to change up the names of these two buckets, but they're still generally two to three buckets. We really do a disservice to the condition and thinking about it this simplistically, because babies can look like there's just this whole spectrum of kids who are running around typical, typical hearing is fine, living their lives. I mean, maybe there's someone on this call who has congenital CMD and doesn't even know it's possible. And then there's the other end of the spectrum where these babies die in utero or they're so sick they pass away in the NICU or in early childhood. So this huge spectrum.

But still, when we think about it more simplistically, it's symptomatic and asymptomatic. Symptomatic really is 10% of the kids, and it means that they are born with visual symptoms. Like, if you were really discerning, you could tell that they had, you know, oh, like my daughter had little spots on her cheeks. Okay, that was a symptom. They can have sensorineural hearing loss, but that's actually not a criteria.

And most of those symptomatic kids go on to have atypical development. And then the other end of the spectrum are most of the kids we say born asymptomatic. 90%. These are clinically in apparent. You look at this baby, sometimes people will call this a congenital cmv.

Infection versus disease is symptomatic. These babies can have sensorial hearing loss as well. Although what irks me is it doesn't count as a symptom. You know, it's like, it's hard for parents Right. And they're like, what do you mean my baby's deaf?

And they're asymptomatic. But, you know, nobody asked me when we meet these criteria. But most of those kids go on to have difficult development, aside from those who do have hearing loss.

So this is a little girl. Her Name's Robin. She's 12 now. I emailed her dad to ask if I could use her picture. And he said, he wrote back and said, Robin said, of course.

So this is Robin when she was born. She has what we would typically call a blueberry muffin appearance. You know, in pediatrics we make everything sound cute. So she has symptomatic disease. So she was small for gestational age.

She had a small head circumference. You can see here, she has those little spots called petechia or purpura. This baby is obviously very jaundice. She had hepatosplatomangaly, means enlarged spleen, enlarged liver. I don't know actually if she had seizures, but that's something we can see differences in the brain anatomy.

And I'll say too that microcephaly, that small head size and is a result of the brain anatomy being different. So sometimes we'll talk about microcephaly being kind of a poor prognostic sign, but it's really, it's a marker of the brain anatomy and then laboratory abnormalities from that high abnormalities, liver or some blood counts. But again, this does not include isolated sensorineural hearing loss.

And then asymptomatic. These babies, normal physical exam, normal labs, no brain effects, but they can have hearing loss.

All right, now I'm going to turn it over to Dr. Hsu. Thank you so much. Now we're going to talk a little bit about why we screen for cmv. And I think that part of the answer you already know because of the fact that sensorineural hearing loss can co occur as a symptom of cmv, even though it's not considered enough to make them a symptomatic child. So when we think about audiologists and our focus on CMV and why we became so involved in the congenital CMV care process is that congenital CMV is actually the most common non hereditary cause of congenital sensorineural hearing loss.

About 4 to 6% of congenital sensorineural hearing loss is, has been associated with congenital CMV. And it's also been estimated that about 20% of all pediatric sensorineural hearing loss, including those who had delayed onset of hearing loss after birth, is associated with congenital cmv. So One of my favorite resources as I try to wrap my head around and talk to families about expectations with children with congenital cmv, especially since we've got that kind of complicated picture of calling a child asymptomatic or symptomatic based upon their visual signs and symptoms. It comes from an article by Kabbani and Ross in 2020 and I find this table to be very helpful. What we do know about congenital CMV is that hearing loss is the most common long term sequela.

It affects both our symptomatic and our asymptomatic children. And in infants with symptomatic congenital CMV, sensorineural hearing loss develops in approximately 30 to 40% of these children during the newborn period or early childhood years. The hearing loss in this group, as you can see from this table, is often very severe, with about 75% of the affected children experiencing bilateral, severe or profound sensorineural hearing loss. And as you can predict, we're not going to miss those children. Typically they have a pretty significant developmental concern.

Delayed onset of hearing loss also occurs in this population in about 18 to 27% of the symptomatic infants and and we also typically will see a progression of hearing loss over time of about 20 to 54% of these babies. So once again, we don't have the comfort level of telling a parent really solid expectations early on in the process, even for a symptomatic child with congenital cmv. So even though we associate asymptomatic congenital CMV with a lower overall risk, these infants do remain vulnerable to hearing loss, particularly the delayed onset and progressive sensorineural hearing loss. So approximately 10% of the asymptomatic infants are going to be diagnosed with sensorineural hearing loss at birth or they're going to develop hearing loss within the first several years of life. So once again, this is going to be developmentally significant for these children.

And more than half of these cases will involve only one ear. So what we often see is those who are symptomatic will have bilateral involvement. Those who are asymptomatic, maybe some of our unilateral hearing loss kids. But remember that delayed onset hearing loss in asepsymptomatic children also occurs, but it typically occurs a little bit later than for children that have symptomatic CMV, with about 38% of those developing sensorineural hearing loss, as you can see on the table, at about age of 44 months. In addition, hearing thresholds frequently fluctuate and this is a key issue to Consider as we talk further about screening as well.

Hearing thresholds are going to fluctuate in this population and about half of the affected children may demonstrate some partial improvement in hearing over time. Whereas we also recognize, though, that others will have a fluctuation in the other direction. So the potential for delayed onset and progressive hearing loss in both our symptomatic and our asymptomatic babies with congenital CMV really underscores the importance of us designing long term audiologic surveillance programs to ensure that we have timely diagnosis of these children as well as timely intervention so that we can help to improve their developmental outcomes. All right, so why should audiologists be concerned about congenital cmv? I hope that these statistics have helped convince you that we're concerned because both infants with overt and without overt clinical signs at birth may have sensorineural hearing loss.

Many will have delayed onset of sensorineural hearing loss. So it's not like we screen them and we're ready, we're finished. And we know that we're not screening and finished with any child, but in. But this is a case where we need to have a heightened degree of concern about this potential delayed onset of hearing loss. They may have unilateral or bilateral impact, and it may.

Many are also going to have progressive hearing loss, which can progress to profound sensory neural hearing loss, especially in those that are symptomatic at birth. So why would we engage in newborn congenital CMV screening? Well, I think that as audiologists, we recognize that most of the time what

we are doing is with any patient that we're seeing for screening or diagnostic testing. We're essentially providing two functions. One is to help identify, and in this case, we want to identify infants with congenital CMV and determine which of these infants with congenital CMB have possible congenital hearing loss because of it.

But we also then want to provide education and support to the families. We want to make sure we've educated them fully about the disease process itself and the potential impacts on hearing, that we're making very clear treatment recommendations and that we're connecting them. Interprofessional engagement is extremely important with all of our patients, but even more so with those with congenital cmv. And parents are begging for us to engage interprofessionally with our colleagues to make sure that we're making appropriate referrals, that we're clear with everybody about what needs to occur to support this family more comprehensively and effectively. We need to design and implement an appropriate monitoring plan and make sure the family understands that clearly and provide what prognostic information we can.

So as we start planning on offering congenital cmv, one of the first pieces that we have to look at is who's important to be on your team. And this will change potentially at different stages of your process of supporting a family who has a child with congenital cmv. But let's talk about what team you really need to pull together just for the screening process. So in the screening process, we recognize that the audiologist needs to be involved, but we also need to always involve our newborn nursery staff. Our attending physicians are a huge part of supporting a CNV screening program.

And we need to recognize that as audiologists, we're working within an environment, but it's not our home environment, it's their home environment. And we need to make sure that they support the process and they help us understand how to insert ourselves as fluidly as possible into the nursery ecosystem for this process. Microbiology lab, pediatric infectious diseases, otolaryngology, and the

primary care provider. And then I would circle back again to that primary care provider. If you're fortunate, you will have a primary care provider who has experience with congenital CMV or more education in congenital CMV.

And but as Dr. Pesh mentioned before, even in people studying in developmental pediatrics who are specializing in this area, it's a very limited part of the course load in their educational process. So you, as an audiologist who becomes passionate about congenital cmv, may also take on the role of gently helping to support education of others that are working with the family. One of our important questions who should be screened? There are three essential approaches to congenital CMV screening. I'm going to talk right now about hearing targeted CNV screening, and then we will cover later expanded targeted CNV screening and universal screening.

Hearing targeted CNV screening is becoming more and more common. It is legislatively mandated in several states and has been also taken up by many health and hospital systems nationwide that are not mandated or required to to implement hearing targeted congenital CMV screening. I was fortunate in that in the 90s, when I was working with my colleague Chris Owen, and all of our nursery group that we had pulled together, just like on the previous slide, to design our newborn hearing screening program, we had an infectious disease neonatologist, Pablo Sanchez, who was passionate about congenital cmv. And as we were designing our newborn hearing screening program, he said, why don't we do screening for congenital CMV on babies that don't pass the newborn hearing screening? And after discussing it with him, we went back to our group we had convened of an interprofessional support group for planning the newborn hearing screening program.

And we were able to get agreement amongst the whole group that we would implement hearing targeted congenital cmv. To our knowledge, we are the first hospital to actually do this. I've been told that by other people. I don't know for sure if that's correct, but it was all due to having an infectious

disease physician involved in our interprofessional planning for newborn hearing screening that we implemented there this. And so our protocol was from early in the 90s to first do it, or late in the 90s to do first an inpatient hearing screening by a hearing screening technician.

If the baby passes the screening, we go into evaluating the medical and birth history to look at if they're at risk for delayed onset of hearing loss. And then we'll provide parents with either information that their baby passed the hearing screening. We don't know of any indicators for delayed onset of hearing loss, but our educational materials always suggest to parents that if their child's not meeting the developmental milestones that we provide for them that they have their child reassessed. And then if they do actually have a delayed onset of hearing loss risk factor, we put them into our monitoring program and provide the family with information about their risk factor and recommendations for follow up. If the baby doesn't pass that initial screen, which is in the first few hours of birth, we then do and our screening program is 100% screening ABR.

If they don't pass that first screening, we then have them screened, hopefully close to discharge by an audiologist. And this is to one give them a little bit more time. If we still have a little bit of fluid in the ears after the birth process, Vernix in the ear canals or something of that sort that may be causing conductive pathology to allow that to clear if possible. And then the audiologist will be checking to see if there are any issues with placing the couplers appropriately, making sure that we're getting a good screen. If the baby passes, they go through the same process as if they would have been passed in the initial screen.

But this is where we diverge. If they refer, they then get a hearing targeted congenital CMV screening. Dr. Pesch will be talking later about our type of screenings, but at the Parkland program, our preferred screening is a urine cmv. And then in some cases, if we're not able to get a urine cmv, we will do a saliva swab, we collect the urine sample before discharge and then we schedule them for their

outpatient hearing screening and to get their congenital CMV screening test results at the outpatient hospital within 10 to 12 days after discharge. If they pass the outpatient hearing screening and they're at risk for delayed onset of hearing loss, they're going to go into the parent education that we have for.

They're going to receive their information about the delayed onset of hearing loss program and recommendations. If they do not, they'll then be discharged from the program with information about developmental milestones. If they refer or do not pass, they then get a diagnostic eval. But what's important here is that when we have a patient refer and they get a hearing targeted CMV screen, we now have another delayed onset of hearing loss indicator to use to determine if they should be followed more closely. But they're also going to get a diagnostic evaluation if they have a positive CMV screen.

So when they come back to the birth hospital in 10 to 12 days after discharge, as I mentioned, we hopefully at that point we will have verified that we have our congenital CMV screening test results. And if we do not have a valid sample when we review the test results before their outpatient visit, we'll collect a new sample for congenital CMV at that outpatient visit. And that's extremely important because we want to make sure that we're able to get a saliva sample or a urine sample within that early process. As you'll find out more about when Dr. Pesch talks about the screening tools we use. If the congenital CMV screen results were positive when we reviewed them prior to the outpatient hearing screening result visit, we then will also arrange with a medical provider to meet with the family and this medical provider will be able to discuss the congenital CMV test results, the recommended follow up plan for medical management and evaluation of congenital CMV, and as well as make recommendations and set up referrals for the parents.

At that point, the audiologist is continuing with the recommendations that we would be making for audiologic evaluation, but we're collaborating, working interprofessionally with infectious disease and with the medical providers that are now going to be following the child for congenital cmv. The goal

here is also to connect the family with pediatric infectious disease before they leave so that they have all of their follow up in place and they can be considered as possible candidates for antiviral therapy.

So after engaging with our program for about seven years, we went ahead and did a study about seven years in of the first five years of our congenital CMV screening program. And so in this case we looked at all the babies that received a congenital CMV screen after referring on newborn hearing screen screening. And we found that about 5% of those babies that received a congenital CMV screen were positive for congenital CMV. Of the babies that were positive for congenital CMV, 67% had a confirmed auditory impairment and 25% had normal hearing. We lost 8% to follow up.

Notice that of the 67% with confirmed auditory impairment, when we looked closely in a chart review, 25% of these had clinical signs that prompted the congenital CMV screen. Because our hospital kind of does an expanded targeted. We were already screening babies that had clinical signs and symptoms as well as those that were born to HIV positive moms. And so in this case, 25% of our babies would have also gotten a screen because they were ordered based on their clinical signs and symptoms. But 75% of those with confirmed auditory impairment had just failed the newborn hearing screening.

That's the only reason they received a congenital CMV screen. Now, important to look at here is that 33% of those that received a screening because they failed the newborn hearing screening. If you really did a deep dive into their medical chart, you would find that 33% of that should have been picked up for clinical signs and symptoms. So that's a consideration when you look at expanded targeted screenings is that sometimes you miss those clinical signs and symptoms. As Dr. Pesch mentioned, she didn't automatically see that early on in her baby as well.

And then 67% had no clinical signs or symptoms of congenital CMV. If those with normal hearing, 83% received the congenital CMV screening because of their hearing screening test results, at first we were like, okay, these must have been conductive hearing loss babies because they passed their outpatient

rescreen and or ended up with a normal diagnostic. But now what we know, and we didn't, I didn't know this at the time, was that actually these were some of those babies probably that had fluctuating hearing loss due to the congenital CMV. And then 17% had clinical signs that prompted a congenital CMV screening. So notice that 67% of our babies that were had confirmed auditory impairment due to congenital CMV would have been missed if we were just looking at signs and symptoms that were visually evident.

So then if we went back, I went back later and just did a casual review of our data to see if it held up with the data that we had seen. After a five year review of the data, and after 17 years of universal newborn hearing screening and hearing targeted congenital CMV, we still had about 89% of our babies that were referred on hearing screening received a congenital CMV screen. Of those, 0.3% of those screened had confirmed auditory impairment. But what's important to recognize is that 6% of our infants with confirmed auditory impairment had congenital CMV. When you think about the statistics early on about what percentage of babies with sensorineural hearing loss actually have congenital cmv, this is pretty much in line with that.

It's also pretty much in agreement with data from Barbie et al out of Italy. In 2000. They reported on their test results of kids that were identified with sensorineural hearing loss at less than 2 months of age. 10% of those infants had congenital CMV as confirmed by dried blood spot samples. So we've had some consistency in our statistics.

When we look at a program where we're doing urine screening at birth and comparing that to the data with Barbie et al. And they're very similar. Remember, ours are just looking at birth, not within two months of age, the first two months of age. Then the question becomes, well, that's great. We're already doing newborn hearing screening.

Pretty easy to grab a urine or a saliva sample if a baby doesn't pass newborn hearing screening. A pretty small population to have to look at. This looks like the answer, but is this really the answer? I think that you probably already know from some of what we talked about before that maybe it's not the perfect answer, but we were privileged to be able to participate in the CHIME study that was led out of the University of Alabama at Birmingham. And this involved a number of different sites that were able to do almost universal CNB screening for a period of time.

So for about, for a few years, we were actually able to look at babies that screen. Babies in the hospital that had passed their newborn hearing screening also received a congenital CMV screen. Not 100% of them, but those that agreed to participate in the study. So we looked at our data a few years later and looked at, okay, how many of our babies with CMV do not pass newborn hearing screening? But how many babies with congenital CMV were picked up because we were doing the CHIME study and actually they had passed their newborn hearing screening.

So how effective is it? So what we can see is that of those that had congenital CMV and were screened for CMV because they did not pass hearing screening? 7% of all infants with congenital CMV were picked up with this, 5.5% in the well Baby Nursery and 20% in the NICU. And then we can look at those who passed and would not have been screened for CMV. So this would be 93% of all infants with congenital CMV, 94% in the well baby nursery and 80% in the NICU.

So when we go back and look at the data, what about infants with CMV who passed the newborn hearing screening? What we can see here is we do have that breakdown of unilateral versus bilateral loss. But essentially our babies were about 57% of infants who had CMV related sensorineural hearing loss in the newborn period were identified by newborn hearing screening. But that meant that if we looked at babies within the newborn hearing period or the newborn period, that about 43% of infants with congenital CMV who developed sensorineural hearing loss within that two month period that

Barbie's group was looking at or 60 days of life past their newborn hearing screening. So these would be babies that we would have missed for monitoring or providing information about developmental potential of delayed onset of hearing loss.

So I'm going to turn it on over now to my colleague Dr. Ketler to talk about the other two approaches to congenital CMV screening. Thank you. So it is exciting. We are seeing many more babies that are screened through the hearing targeted initiatives. But just as Dr. Shoup said, we recognize that there are still many babies that are going to be missed by this type of screening.

We've seen some steps that have been moving forward to really start an expanded targeted hearing screening, Hearing plus screening. And this has been adopted in many institutions and, and there is continuing efforts across the country to have this expanded targeted implement implemented into organizations. And when we talk about expanded targeted screening, we're really referring to kids who do have symptoms at birth. Usually the symptoms for the expanded targeted include the hearing screening results as well as the child's if they are SGA or small for gestational age, have microcephaly, any of the petechiae, the jaundice, the enlargement of the liver and spleen, seizures, the intracranial abnormalities or laboratory abnormalities, these are the most common signs and symptoms that babies who have CMV may have at birth. When we look at expanding to this group, we will catch more children who have congenital cmv.

But unfortunately, this isn't still going to allow us to catch all of the children with cmv. So while this identifies more of our symptomatic babies, we still expect to only catch about 10% of all cases of congenital CMV using these signs and symptoms. In addition to the hearing loss. We also know that these babies who have expanded targeted who are missed on the expanded targeted still are at risk for a later onset sensorineural hearing loss. So it gives us still a gap in the patients that we can treat and the care that we can provide to those who need it.

We are seeing more and more discussions on all fronts about universal CMV screening. And what universal CMV screening really means differs from place to place.

So this is essentially a test all of the babies. Just like the evolution that we've had with the newborn hearing screening programs and the newborn hearing screening legislation. Across the country, we are working towards efforts of universal congenital CMV screening. We are seeing states like Minnesota and Connecticut passing legislation that will require the CMV screening. And this is done routinely in several Canadian provinces.

Now, what is important to note is how we screen the babies for congenital CMV when we're talking about universal testing is still something that is widely debated and often not supported by all of the experts in the field. So we are talking about most of the time dry blood spot testing, and Dr. Pesh is going to talk more about the sensitivity and specificity of those specific tests. But there is some argument about really the need of PCR testing universally to be done in order to catch all of the babies who are in need of follow up care for congenital cmv.

When we consider the congenital CMV universal programs, one of the questions that really comes up is the cost. So who pays for the universal screening of babies who do not have risk factors? Now, insurance is really good about covering CMV screening in children who fail their hearing screenings and children who are at risk or children who have risk factors to start. But when we talk about children who are not at risk, there is a question about who will pay for the actual test that is completed. Will insurance companies be willing to pick this up?

Are hospitals able to support the screening at their local level? Is this something that needs to be done at a department of Health? There are many questions that are unknown when you talk about universal CMV screening as well as really how are we going to implement and follow these babies long term? Again, we talk about the kids who are asymptomatic at birth and potentially asymptomatic for the rest of

their life. Is detecting the congenital CMV infection in those babies something that's beneficial to them and to their families?

It's really still something that's widely debated. Many will argue that it's still the parent's right to have that information. And then others will mention parental stress and concern of knowing a diagnosis that may not impact their child.

Now when we talk about targeted and compare it to universal screening. It's important to recognize that there is a lot of support around the country and the world for targeted CMV screening. It's supported by acia, aao, aaa. We are hoping to get some guidance from AAP in the next few months to year. Those organizations provide us some good input about what who should be screened and why they should be screened.

But when we talk about universal, it really is just so much unknown that the level of support for this screening isn't something we see as widely applied. And when we talk about the babies who are identified through universal screening who are asymptomatic, we are forced to answer questions about how do we treat these asymptomatic children. Children. Children who are asymptomatic will likely have parents who have questions about what this means for the future. And those questions really don't have answers.

So the question is, do these children need additional testing? Do they need expensive lab work? Do they need MRIs or head ultrasounds? Is there an argument to have these children tested? There is a cost to the family and to the health care system for testing children that may not need testing done as well as there is the burden that it puts on families just to have this additional worry and stress.

Parents with their newborn children already have a lot of anxiety about the development of their child and is it important to give them this information again, considering that many of the children who have

congenital CMV won't need additional medical or developmental follow ups.

When we talk about newborn hearing screening, one of the things that I get asked regularly is how does it get onto everybody's panel? And what I've learned through my work and is different for every state and every province has a different way to really evaluate what is on their screening panel or their dry blood spot panel. There is a national CMV or national RUSP recommended Uniform Screening Panel that does not currently include cmv. But when a disease or disorder is added to the rusp, most of the states will add it to their individual panel based on that RUSP recommendation. Another thing I will add is that sometimes these recommendations need to be done through legislation and in other states this is not required through legislation.

In Ohio, we do not need legislation to add disorders or screen for conditions on our newborn screening dry blood spot. We have a panel that sets the recommendation and that is what we follow. So the pathway is different from state to state for how this gets added. We are working with the National CMV foundation on having CMV added to the rusp. It takes a Lot of work to get an additional disease or disorder added, including a lot of research, a lot of funding, dollars and advocacy.

So while it hasn't been added yet, there are still a lot of efforts being made in order to have this added into the future.

And here is a great representation. It's always nice to show the map of, of what is happening around the states, what is happening in the country really for the kids with cmv. And you can see here, the states that are colored are states who currently have active legislative activity for cmv. Now again, I always like to highlight that this isn't really all of the work that's being done. As Dr. Shoup mentioned before, many organizations have adopted CMV screening without legislation.

And there can be an argument for this should be done as a grassroots effort. This should be done because it's the right thing for children. We don't always need to legislate all of the health care that we provide for patients and children. So there are ways to get CMV screening programs started in your area. Legislation is one avenue, but definitely not the only avenue where you can go.

I'm going to pass this over to Dr. Pesch.

Okay, great. Let's talk about choosing your test. The thing about congenital CMV is you have to test on a sample that was collected before 21 days of age. And that's because when kids go home and they're excited, exposed to breast milk or here's my daughter being exposed to slobbery sister, they can catch CMV in other ways. Postnatal CMV generally not a big deal.

So if you were to test a baby after 21 days and they came back positive for CMV, it's really tricky to know if that is a slobbery sister giving them kisses, which no big deal, or if it's congenital cmv, which is potentially a really big deal. So the sample has to be collected before 21 days of age. Oops, there we go. So there are three different sample types that are kind of the main ones that we see. Urine, saliva and then dried blood spot.

And that's from the heel poke test that is done for general newborn screening. But there are pluses and minuses of each of these. For urine testing, it's a basic CMV PCR test.

There isn't a high complexity of the test. It's often a send out test. At least it was at our hospital until maybe your internal lab, if you have access to that at your hospital or health system, can develop it in house. But for us, when we implemented this at the University of Michigan, our turnaround time was like two to four days. So those babies that are in the, well, newborn nursery are long gone and home, and sometimes they can be lost to follow up the testing.

Really the cost is super variable and it depends on the lab. So I've seen it as low as $10 and as high as $200 a test. It is not a viable option for newborn for universal screening. And that's because these babies do not pee in a cup, right? They pee.

They have to stick a little bag under there and say a prayer because they only pee usually one to two times in the first 24 hours of life. So if you don't catch that first urine, you're out of luck in terms of screening in a hospital. So it can really delay discharge. Even though this is a fantastic test, this test, we would say, is the gold standard diagnostic test because it is so sensitive and specific, meaning that it catches almost. I think it's like 99% of cases of congenital CMV if done before 21 days of age.

And when you get a positive test, it means it's cmv. It doesn't mean, oh, it could be CMB or it could be another virus. It's cmv. So it's a really good test. Oh, there we go.

That's a little baggie.

So what we did in our hospital, we didn't have saliva available and we didn't have dried blood spot available. So what we did to start off is, you know, sometimes you just have to improvise improvised. We made, we called them pee pee kits. And then we would send families home with this little takeout bag. And we're like, oh, we think your baby should be screened for cmv.

Here are some instructions. Here's gloves, here's a little urine collection bag or two. Collect it at home, pour it in a cup, and, like, return it back to the lab when you can. CMV is a very hardy virus, so it'll stick around for a long time, even if the urine isn't refrigerated or if it's in, like, the car for a few hours. No big deal.

Of course, this is a lot to put on a family, but we did it when we didn't have any other options and we just felt like screening was important.

Saliva testing is another great option. Really. It's like a Q tip. In our hospital, we use the same swab as we do a Covid nasal swab. And you just put it in the gums and get that baby drool on it.

So easy. And then send that off the equipment to get this done. You can either do it on your floor or if you're in a hospital setting or in a lab. It's about $4,000, which, as I've learned in terms of microbiology testing equipment is not very much. Some companies will actually donate it because then you have to buy their tests and their products.

The swaps are about 6 to $25 each. And if done in the hospital, it's billed to. Oh, sorry. It's billed to the insurer. It's done in a hospital.

It's part of this bundle payment it's made to the insurer. That's part of all the hospital childbirth care. But if it's done in an outpatient, like they do in Utah, they are able to bill it separately to the insurer.

There's just an example of a type of saliva swab. This baby is like, what are you doing?

Before I go into that, I should say the biggest drawbacks of saliva swabs. So it is very sensitive. They will pick up cmv, but it's not very specific, meaning they will pick up CMV that sometimes maybe was in mom's breast milk, which is very common. And again, generally no big deal. Or that's the big.

So we'll see false positives. So what we do is if you have a saliva swab that's positive, we always, always follow that up with a urine or a dried blood spot that's positive. We always follow it up with the urine before 21 days of age, if we can get that.

Oops, got the clickies here. And then there's dried blood spot testing. So this can take about two to eight weeks to turn around. In Michigan, it requires a release of information so that the parents say,

yes, we agreed to release my child's biological materials. And then we.

In Michigan, I think, actually around the country, there are only about five labs that do this. So you have to send it out, usually to another state. The turnaround time can be upwards of eight weeks. There's a low sensitivity. So some labs have really great technology, and they're able to pick up CMV about 80% of the time in true positive cases.

So if you tested 100 babies with congenital CMV, it would pick up 80 of them. So that's good, but not fantastic. Other labs, their test would only pick up 40 of those babies. And that's because CMV is just not present in high concentrations in the blood.

The nice part about dried blood spot testing, though, is if your state Saves the dried blood spot. You can get it later on. So this is a sample that was collected before 21 days of age. So in my daughter's case, she was, by the time we got around to the CMV testing, she was four months old. And we were able to get her dried blood spot and tests for that.

But this is just an outline of the whole process and the players that are involved. It's not straightforward.

Some states, like Minnesota and I believe Connecticut are, and provinces in Canada are universally screening using dried blood spot. And that is really nice because it's already a specimen that they have. So it's just adding on a test at a level like a central lab where most of the babies have specimens going anyways. So from a screening standpoint, public health wise, I think this is the way to go, just for ease, but knowing that we are going to miss minimum of 20% of the babies, which is not great. So there are some technological advances that we're still waiting on.

Okay. Things to avoid. Please don't test antibodies, IgM antibodies, those are the short term antibodies. They don't cross the placenta. So they're not necessarily moms.

But not all babies are able to make them right away. And IgG is Mom's antibodies. So mom might have this like gorgeous prior immune response, the CMV and like, you know, some immunity. And so knowing the baby has it's not helpful serum from a lab draw so you can test CMV in the blood. It's not terrible, but it's never been, it's never been piloted for universal or CMV screening.

And that's because of the low concentration of the blood or of the, the virus in the blood. And then specimens drawn after three weeks of age. I mean, honestly, sometimes you have to make a guess if you don't have the dried blood spot, if the dried blood spot comes back negative and you're like, this looks and smells like cmv, you can test to see if the baby has antibodies or if they still have a virus in their system and make an educated guess. But for screening, it is not the way to go.

And here is just an overview of the algorithm that we use for congenital CMV screening. This. I think we'll have the references later available as a second document. This is from a paper I wrote that was in bmj and I believe it's like freely accessible now because it's old enough, but it's just a flowchart that really helps us decide, you know, what is what, because it's not a simple process to screen for cmv. The screening is just the first start and then, then what do you do?

Perfect. All right, so now we're just going to touch very briefly on establishing your protocol. And I think the important thing to remember here is that you do need to think through all of these different steps, regardless if you're doing, I mean, specifically if you're going to be doing hearing targeted screening or an expanded targeted. Because in a lot of those cases, the audiologist may be driving a lot of the follow up that's occurring. So you need to make sure you know who's going to be keeping track of the test results.

From our standpoint, when we started the program at Parkland years ago, it was the audiologist. So the audiologist was scheduling the outpatient rescreen 10 to 12 days after the baby was discharged, partly

so we could capture that second specimen, if needed, below 21 days of age. But the audiologist was would then go in and check the CMV results before the outpatient visit. Who's going to order referrals? Who in your state has the ability to order referrals?

And so partnering with somebody who's in the clinic that has the ability to order the referrals. If you do identify a child with congenital CMV is extremely important. Determining who's going to order what the audiologist is driving information about what additional audio evaluations need to occur. But you need to have agreed with your interprofessional team on what type of workup you're going to have for children that are identified with congenital cmv. And then that you also need to make sure you're communicating very clearly with the PCP, recognizing, as Dr. Pesch alluded to earlier, that the training for PCPs on congenital CMV is widely variable.

So you may be sending the child's information to a PCP who's never seen a child or doesn't know that they've seen a child with cmv. We can probably bet they've seen a child with cmv, but they may not realize it. I cannot stress enough how useful the article by Dr. Pesh and colleagues is. This is a great way to start looking at how you're going to set up your protocols. I'm not going to go in depth in this because I know we're reaching the end of our time with you, but please go through and take a look at it.

It is on your reference sheet. It does have algorithms for every step of the process in working with families with a child with congenital cmv, including recommendations for the types of additional evaluations that need to occur for a baby that's identified with cmv, and then also information about how we need to monitor not just hearing, but vision and other areas developmentally for the child with congenital cmv, recognizing that, once again, you may be the person that's the expert in congenital CMV in your facility, and so being aware of at least who else needs to be part of that conversation to support this family more comprehensively and effectively is important because you may be responsible for trying to pull together this interprofessional team. So just quickly, a couple of really important

lessons learned, especially if you're going to be working with hearing targeted cmv, is you can sometimes run into resistance in your nursery if you're coming in, as the old audiologist saying, okay, I've screened this baby's hearing, they didn't pass the hearing screening. I need you to put in an order for congenital CMV testing, and we need you to bag the baby. Some providers may be resistant to ordering the congenital CMV screening, especially if they think they know why the child has hearing loss.

So if they have microtia, they may say, well, that's why they have it. If they have other diseases and symptoms that are associated with hearing loss, well, that's why they have hearing losses. I think it's important to remember one of the pediatricians that was also a faculty medical teacher at the med school at UT Southwestern would always refer to ticks and fleas. He would try to train his students that don't assume that just because you come up with one reason for something occurring, they can't have something else co occurring. So many of the signs and symptoms people may look at and say, well, that's why they have.

Hearing loss could be part of symptomatic congenital cmv. So we need to make sure that we receive support for that. And you may be the person kind of educating them about that. I would recommend, if you're running into resistance, that once again, you go back to that planning process in the beginning of who needs to be on your team. And you need to have an infectious disease physician, a developmental pediatrician, or a neonatologist who is championing the congenital CMV screening program so that maybe they can have a conversation with their peers.

If you're having a struggle with getting people to order the test for you, another piece of information about how to have a successful program is make sure you're educating everybody. You're developing good, clear guidance and education about the program, why you have the program, educate them at the onset of the program, and then have periodic re education to your nursery providers about their

responsibility for ordering the congenital CNV test. Make sure you're providing the protocol, a written protocol, and the rationale for the congenital CMV screen. And having that established protocol that people can refer to really does lead to less resistance in implementing the program. Some of the things that we will want to include in our protocols is start with what you are going to do, what you can do.

If you're just starting from scratch. You can make a proposal to screen all infants with confirmed sensorineural hearing loss is just a starting point. And then including a statement in the discharge summary for the PCP is important to make sure they're aware that the baby was screened and what the screening results were. Have a solution for if there are delays in testing the dried blood spot. You can file an incident report or a patient safety report if you're not getting cooperation with the screening program and it's documented that this is a protocol that our hospital follows and request a head ultrasound.

In the meantime, work with somebody because inside some cases you can find intracranial indicators of congenital CMB that will help guide your recommendations for this family. Some hospitals make a saliva swab a standing order, but once again, remember that within your protocol, then you need to have a statement that if you have a positive swab, saliva swab, you need to follow with the urine. Rethink the handouts you're giving parents and the type of education you're providing about monitoring for delayed onset of hearing loss and things of that sort if they don't pass a newborn hearing screening. And then reach out to your state aap, EHDI Champion, to get support in setting up your program as well. All right, I'm going to turn it back over to Dr. Ketler and for the last few minutes I'd like to talk about what happens if you're not affiliated with a birth hospital.

So in a lot of situations, audiologists are leading this work around the country and may or may not be directly in a birthing hospital where they can have some more input. I would tell you that even if you are not directly affiliated, this is still a challenge that you can take on and you can overcome. It's going to

involve developing even more partners and making sure that you understand the process and the administration of each individual organization. I have seen in my work that people are very receptive to communication, especially when it is focused on what's best for the babies. So by having your partners and finding your partners and Making sure you can answer and find answers to the questions that they have is going to be really helpful in making these communications and really making making headway in this.

There's going to be some challenges that will happen in this situation. The biggest one is going to be communication because if you think about it, most of our organizations have communication systems that are very unique and locked into that organization. So obviously there's HIPAA laws and there are concerns with sharing of information. You'll need to work with your partners to overcome those. Again, very doable and very possible.

If you have the right team that's really willing to help. You're going to want to make sure you're able to communicate with babies again and families through these different pathways. They're going to be a patient at this hospital and then at a children's hospital. Likely. Making sure that that communication can be seamless for families will go a very long way.

Another big concern when you're doing this is setting up a way to identify and track families. I will tell you, when we started here in Cincinnati, this was one of our biggest concerns was how do we not lose families along this pathway so that constant communication can be really helpful and doing that on the front end saves a lot of headaches and really makes sure that all the kids get the best care possible. If you cannot make the headway into the birthing hospital, it is really important that you as audiologists at least know that you have the part for the babies who come to you as an outpatient. If those babies have not been screened, doing a saliva while they are in the office for their follow up hearing test is a really great way for us to take control of making sure these children get tested. You're going to need an order

to be placed by a physician.

So keep in mind that it is really important to have that relationship. And also really managing your access to appointments becomes critical. So as Dr. Pesch said, we need to get these appointments in by three weeks to of age. Many audiology clinics have wait times of four months before they can actually be seen. That doesn't really help our families who we need to get that as quickly as possible.

So from the birth hospital education of getting appointments as quick as possible and then your access improvement really can make it so that the outpatient option is a viable option for patients and families that can't get it done on the inpatient setting.

And then I will mention that for the kids who are older than that six month age where CNV testing may not be as important, it is or may not be possible. It is still really important for us to get this information for kids to be able to have the appropriate follow up and recommendations and expectations, expectations in the future. So we will really work with our Department of Health to access those dry blood spots whenever we can so that hopefully we can get that information. You need to know per state. That varies.

So in Ohio, we keep them till their second birthday. If it's two weeks in a month past, they won't have it. But if it is in that timeline, that's a great option. And then for children who are older than six months, they could have the IGG screening as well.

The last few slides really want to focus on the need of support for our kids who have cmv. So our patients who have congenital CMV are going to need a village that is there for them that can really provide everything they need. You're going to want to be working with multidisciplinary team of therapists, keeping the parent and family at the middle but making sure that they have access to all the different professionals that may help. And we are all volunteers with the National CMV foundation and if

you haven't visited the CMV foundation website, I encourage you to do so. There is plethora of information that you can print and give to families or hard copies can be sent to you to give to families.

The resources that they have and the supports that they have for families is really invaluable for people going through this process. There are also support groups. There is a Care to Talk which is run by the National CMV Foundation. Hands and Voices offers CMV resources and Parent Guide and there is a CMV Mommies Facebook group. There's a warning on that that they post a lot and so it may be overwhelming on your feed, but.

But at least for families who are really interested in getting that information initially, it's something we would absolutely encourage to do. I'm going to skip our reflections because we are over on time, but really is such important work that we are seeing spread throughout the country. It's an exciting time for congenital CMV awareness. I think we are making some really good efforts moving forward and hope to continue, continue that in the future. All right.

And I'm going to open the questions. Okay. We have no questions. The only question we have is that Connecticut started universal CMV screening in July of last year. Thank you very much for that comment and that clarification.

Thank you all. We'll just give it another minute. People may be typing in. I just wanted to say thank you so much for such a kind, comprehensive talk. You got so much in the hour, so much actionable information here, great resources and references.

So I can't thank you enough for bringing this course to Audiology Online. And I am not seeing any other questions come in, so we will wrap for today. Thank you again to our presenters. Thanks to everyone who participated. Also, a reminder that courses on audiology online are open access.

So if you want to share these with your team, with your state association with other audiologists to help move this forward, please send the link. The recording will be posted in a few days and that's a wrap today. Thanks everybody.