Critical Congenital Heart Disease Webinar
April 2016

Presenters: Lisa (Hom) Wandler, Ruthanne Salsbury, Jamey Kendall, and Dr. Merlin Ariefdjohan

Please direct all comments/questions pertaining to this webinar to Thalia Wood at Thalia.wood@aphl.org or 240-485-2701.

Lisa : CCHD Critical Congenital Heart Disease screening webinar. We have two great presenters on this afternoon, Jamey Kendall and Merlin, from the University of Colorado.

Can you go to the first slide, Ruthanne, with the FPS update?

Ruthanne S: Sure.

Lisa : Great. Thanks so much.

Last February, at our February webinar, just two months ago, Dr. Hokanson presented, and one of the things he shared was an update on the Society Thoracic Surgeons public reporting and their database.

The public reporting aspect is pretty new. I think they just started last year, however, the surgical database for congenital heart surgeries has been in existence for many, many years. Over a decade.

The link that he shared and that Thalia shared after the webinar lists the public reporting of FPS outcomes for surgical centers. We had a couple questions, including if they didn't see the center or hospital that they were interested in, folks should do. We followed up a little bit and this website where FPS publicly reports outcomes on surgical centers is updated only once a year. I believe the next time they're going to update it is this summer. Because a lot of the big centers weren't on it last year. If you check again, I'm not sure the exact date of when they'll update it, but perhaps late summer, you may be able to find more information on other centers.

Another great resource would be to go to the website of the hospital itself. A lot of information on outcomes is typically displayed on hospital websites, if you go to their cardiovascular surgical page, you can find information there, as well.

We just wanted to give that brief update because we got some questions after our February webinar.

With that, we will go ahead and get started with our April webinar, and I think Ruthanne's just going to give a quick overview of the APHL CCHD presentations that occurred in February.

Ruthanne S: Great, thank you so much, Lisa.

This year's new born screening and genetic testing symposium took place in St. Louis on February 29t through March 3rd. This meeting was sponsored by the Association of Public Health Laboratories and co-sponsored by the Missouri State Public Health Laboratories, as well as Health Resources and Services Administration and the International Society for Neo-Natal screening.

This year, the symposium's theme was New Born Screening: Gateway to Healthy Babies. It was wonderful having international experts present on new and emerging technologies, candidate conditions, quality improvement and testing and clinical outcomes and their therapies.

Now I'd like to talk a little bit about the CCHD presentations in particular. This year, the point of care testing received more attention than in the past, which was really exciting. CCHD was linked with early hearing detection and intervention. There's a total of seven oral presentations and four posters.

These topics are often tied together in people's minds, as well as in databases, so it was really interesting hearing the connection between those. Many of the topics were reflective of our CCHD bi-monthly webinars, and included data collection, use of birth defect registries, home birth, and alternative algorithm.

The administration of CCHD programs continue to remain a challenge nationwide, but these are making headway in various areas, and it was really valuable just having the face to face meeting in order to facilitate discussions and discuss a lot of great, new ideas.

We're really excited to share several of these presentations in future webinars, and we're delighted to welcome Merlin and Jamey to this month's webinar to share a brief summary of their presentations.

So with that, we can move on to our first presentation. Jamey?

Lisa : Our first presentation is going to from Jamey Kendall, and she is a nurse from the Kansas Department of Health and Environment, and has over 27 years of experience in Kansas. She started with new born screening in 2008, when Kansas expanded the screen panel to 28 conditions.

Previously, she worked on the Special Healthcare Needs program as a case manager, and then as their director. She graduated from Wichita State University, and has always loved working in public health.

Thanks so much for joining us this afternoon, Jamey.

Jamey: You're welcome. Can you hear me?

Lisa : You're coming through perfectly.

Jamey: I knew I had un-muted my phone, and I wasn't sure if you re-muted me or no.

Today I'm going to talk about Kansas and our experience with the implementation of CCHD screening. We use a quality improvement method versus legislation, and I'll just go through this a little bit.

Our administration at the time did not want to legislate the CCHD screening because they felt like it was a practice ... It was supposed to be a ... I can't think of the term. The practice to do that.

Lisa : The standard of care, is that ...

Jamey: Thank you, yes.

If I don't write everything down, I have trouble.

You can go to the next screen, and everyone's aware in 2011, CCHD was added to the core panel. In 2013, our new born screening advisory council recommended addition of CCHD to our panel. In Kansas, we're unique because we do not have a fee for our new born screening, and we don't require each legislation ... We don't require change each time we add a new condition. But CCHD was unique because it is not on the blood spot card, obviously, so that put a few more barriers up for us.

It's like I said, the secretary recommended a quality initiative, versus legislation. We had a lot of lobbying from the Heart Association, which I know every state had their lobby from them. We developed a sub-committee of our advisory council that started meeting, and that includes pediatric cardiologists, pediatric surgeons, somebody from the hospital association, and parents, March of Dimes and others.

They met on a regular basis trying to determine what our plan was going to be.

You can go to the next slide.

So we hired a staff member, and [Janay Kellen-Shultz 00:07:48] was that staff member. She was very involved in CCHD screening on a national level, also, but she was hired to make education visits to each birthing facility in the state.

We started with a survey of all our birthing facilities to see what the status was of screenings at that time. So we knew the ones that were screening, and the ones that were not screening when we started.

She developed two different trainings. The full training was performed at those facilities that were not screening at the time of the visit. Then she also made visits to each facility that reported screening, to make sure that they were screening appropriately, and to share additional information that had been developed by Kansas.

We had some tools and draft protocols that we provided them. Some of our Kansas City hospitals had protocols, and the other hospitals that weren't screening could use those as a template to develop their own.

We started a reporting mechanism through red cap, and asked all of our hospitals to report the CCH screening through there. We did not ask our large birthing facilities, because we knew in the future, we were going to go to the birth certificate, and use that as our screening mechanism, or we were hoping we were, and we knew we would just get a lot of push back right away with large birthing facilities in reporting, also.

You can go to the next screen.

When Janay made the visits to the facilities, she did use that time to address some other issues related to new born screening. She would take out information about their unsatisfactory rates regarding the blood spot cards, and talk to them about those issues in addition to just CCHD.

From May 2014 through last July, we made great progress in the implementation of CCHD without a mandate. We started with 30 percent of our birthing facilities screening, and that went to 100 percent. We estimate, in our birthing facilities, 100 percent of our infants are screened at this point.

We did a few trainings with midwives. We had a large training in the Kansas City area that those Missouri and Kansas new born screening programs worked with a midwife there. And we had the request from midwives in Wichita, also, to do a training down there.

That's just where we had a lot of trouble with the midwives in our screening rate with the blood spot cards, also, so we of course have challenges like everyone else for that.

We have the interim online reporting mechanism, the red cap, which we could monitor. Blood results were coming in, and to make sure things were coming in and looked right, and would address that if it didn't.

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This is one of our most helpful tools that we developed, and this was out of all the interactions that Janay had with the national committee. This still ... I have it right in front of my desk as I'm talking to birth clerks and other providers at this time, and sharing that with the birth clerks right now, who are entering our data into the birth certificates.

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This just is a graph of our implementation and how we hit 100 percent over that time period.

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This is just a quote from Dr. Cane, who is at Children's Mercy Hospital, and they found that over the time period where we began with the CCHD screening that they saw a dramatic decrease in the number of newborns presenting critically ill due to their CCHD. The impact of [inaudible 00:12:29] symmetry on early diagnosis of CCHD is clearly saving lives.

We've had some really wonderful providers to help us through our implementations.

So our next step ...

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Like I mentioned, we do have module now in the same area where they, in Kansas, they report the newborn hearing screening on the birth certificate. So they have a separate screen for CCHD screening that started in October. We did not get a live feed into our data system until last month, or back in February. We are just now able to see the kids that are not passing the CCHD screen, or the ones that are not showing up as screened.

It's very obvious that it's still a lot of our larger facilities are not reporting the screening when they are done. That's one of our next steps to work on. We do link what's vital, of course, with our newborn screening system. Beginning a year ago, the first part of 2005 and used the same system as our newborn hearing screening, so we're able to track all the babies that are born in Kansas, and finally see which ones aren't getting the screens at all.

I have a birth clerk conference and a training that I'm going to go to at the end of this month. We have also developed a draft reporting form. Now that we have all the information in our data system, the next challenge is, really, how to follow up on that. Who to follow up with. We have an advisory council meeting coming up in two weeks, and we'll address some of those.

What I have done so far, limited ... I've had probably three of the reports that actually have the baby as failing the newborn screen. But the percentages that they had listed in the report would have made it a pass. So I had contacted the birth clerks and verified that, and many of them thought any time a baby had a 94, that they were going to fail, so they were automatically putting a fail in for that.

Another couple of facilities were putting in ... Switching around the foot results and putting in one percent difference, and so they had 99 percent for the right effort [inaudible 00:15:23] and then one percent for the foot. So they had a variance of 98 percent. So we worked with them on that, but we've only had two or three of those that have turned up like that.

Just our process at this point now ... Just trying to figure out our next steps, and we have gotten a few that did fail that did not have CCHD. I haven't actually found one since we've been doing the reporting that has actually been confirmed with a diagnosis. That doesn't mean there isn't one out there, because like I said, the large facilities are not doing their reporting at that time.

We have a new staff person that's starting on the 18th that will take this and run with it. We will get it sorted out, and I'm sure she will make great strides with it.

I think that's about all that I have. Are you guys doing questions now, or ...

Lisa : Thanks so much, Jamey. That was a fantastic presentation. We really appreciate the update on what's going on in all the QI work you've done in Kansas.

I think what we're going to do is hold questions to the end, so if you're able to stay on until the end of the webinar, that would be fantastic.

Jamey: Sure.

Lisa : Does that work for you? Great. Thanks.

So next we will be hearing from Merlin, who is a research specialist affiliated with the Department of Psychiatry, the child and adolescent division at the University of Colorado, as well as the Pediatric Mental Health Institute and Innovation Center at Children's Hospital in Colorado.

She received her PhD in clinical nutrition from Purdue, and now an MPH epidemiology candidate at the Colorado School of Public Health, at the University of Colorado, and will be graduating this May.

Merlin has extensive research experience ranging from bench science to clinical studies, and her study presented at this webinar was completed as part of her Capstone project at CSPH.

So welcome, Merlin. Thanks so much for agreeing to share with us some of the things you've been doing in Colorado.

Merlin: Hello, can you all hear me fine?

Lisa : Yes, you're coming through perfectly, thank you.

Merlin: Excellent. Good day, everyone. And thank you to [inaudible 00:17:51] for giving me this platform to share results, this research.

Before I go on further, I need to apologize in advance if I sneeze of cough during my presentations, apparently I'm allergic to spring this year. There's only so much medication can do. Hopefully we won't have too much of that.

Thank you for the introduction, and I'm excited to share this data with you.

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As you all know, one of every four newborns with heart defects have CCHD, and this translates to approximately 7,000 to a hundred newborns with CCHD delivered in the US every year. Newborns with this condition will require cardiac surgery within the first year of life, and stresses the importance of diagnosing as early in life as possible.

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Newborns with CCHD can appear healthy at birth, however, due to the [inaudible 00:18:54] their heart structure, they will soon have low blood oxygen saturation levels attributing to cyanosis. Due to this, this condition used to be labelled as blue baby syndrome.

I'm certainly not trying to make light of this condition, but a cyanotic newborn easy to spot if they turn as blue as this.

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In reality, though, newborns with CCHD look more like this.

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Which makes CCHD more difficult to diagnose, especially in newborns that have darker skin tones. And both of [inaudible 00:19:31] become a valuable screening tool, and I think you would share my sentiments here, highlights it by three exclamation marks, just to rate the importance of this screening effort, considering late diagnosis will lead to severe health consequences to the newborns, and [inaudible 00:19:49] the family in health care.

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Although CCHD newborns screening is in place, there are many gaps in information, especially at altitude. We all know that partial pressure of oxygen decreases with increasing elevation. This is physiologically reflected in babies delivered at altitude. They have lower oxygen saturations than those born at sea level, even when healthy.

Yet current newborn algorithms are set and spaced on studies done at sea level, which raises potential efficacy issue when applied at altitude. Due to this, [inaudible 00:20:28] request that additional studies be done at altitude.

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This brings about to our research questions. First, we would like to know what are the pulse ox values of this newborn with CCHD delivered at altitude? Can we alienate these values reflecting their condition? Secondly, how did the newborn was born with a screening algorithm? In other words, we would like to know how well the current AP algorithm performs at altitude.

This is important, because if the algorithm's not catching this newborn at an acceptable level, then we may need to revise it.

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In some methodology, this is a unique retrospective study because the [inaudible 00:21:17] comprised of newborns already diagnosed with CCHD, so essentially, we look at the data backwards. In which case, we extract the data from medical charts to identify true positive CCHD cases, and then recreate the screening protocol using the AP algorithm on these cases.

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Just to be clear, in reality when these newborns were admitted to the hospital for treatment, they were not screened using the screening protocol. But due to their condition, their oxygen saturations were monitored using pulse oximetry. We took those values, and then applied the screening algorithms to answer our research questions, in which case from this point onward, I will refer to our observations on the simulated screens as screening results.

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We defined our cases as term and neo-term newborns who were diagnosed with any of the 70 CCHD diagnoses listed in the screening guidelines, and were treated at Children's Hospital in Colorado from 2003 to 2013.

Children's biomedical informatics, [inaudible 00:22:30], identified 343 cases. However, we have to exclude some of them due to these issues listed on the slide. We ended up with 158 cases that were then analyzed for demography, risk factors, oxygen saturations at 24, 28, 36 and 48 hours after delivery, and screening results.

This [inaudible 00:22:54] were selected because we wanted to mimic the screening process. I'd like to point out that altitude in this study is defined as approximately 5,000 feet above sea level, which reflects the altitude of Aurora, Colorado, where Children's Hospital is located. And here are some of our key findings.

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So who were these cases admitted to Children's for the past decade? They were mostly male, white, Caucasian, and not Hispanic or Latino, which I expected, since 78 percent of cases were from Colorado.

Remaining cases were from the surrounding states listed here, and were transferred to Children's for higher level of care within 24 of their birth.

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Health variables, such as having family history, having genetic abnormalities, and having diabetic mother, have been observed as respecters at national level. However, this does not seem to affect predisposition to CCHD in our cases.

The top three diagnoses are HLHS, TTA and TOF, which is similar to trends observed at national level. This may imply that altitude does not influence the type of heart defect that can manifest during fetal development. Based on our data, Children's Hospital treated approximately 30 cases per year for the past decade.

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In [inaudible 00:24:26] oxygen saturation, here are the results. This is a [inaudible 00:24:30] table, but let's break it down and take a closer look.

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First, this is the value for healthy newborn at sea level. Based on the work by Samuel Edall, it averages approximately 98 percent with about five to six percent difference between minimum and maximum level.

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This is the value for healthy newborns at altitude, based on Right at All's group, which has 97 percent. It's lower than healthy counterpart at sea level. They also have a wider range.

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However, based on our study, newborns with CCHD at altitude show a much lower average with even wider range than their healthy counterparts, regardless of elevation.

You may be wondering, then, what about the values on newborns with CCHD but at sea level? Surprisingly, we don't know because to the best of our knowledge, this is the first study evaluating the pulse oximetry values of a large group of newborns with known CCHD diagnosis.

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I did mention earlier that we simulated screening protocol on these newborns, even though they were already diagnosed with CCHD. This was done in an effort to determine the utility of the algorithm. Let me walk you through the results.

Here, the red bar denotes those who failed screening. In this case, red is a good thing, because that means the algorithm caught these true positive cases at their first screen. On the contrary, green bar denotes those who passed screening without any concern for CCHD. This represents false negatives in our model.

Keep in mind for now, red is good and green is bad. Yellow bar represents those who produce borderline screening results. According to the screening protocol, this needs to be re-screened for confirmation for up to three times.

As you can see here, our first screen resulted in 23 borderline cases that went on to second screen.

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Out of that 23, it remained borderline and had their third screen.

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At the last screen, these three cases split off as failed and passed accordingly.

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If you look at the graph as a whole, we are most concerned about the greens. This [inaudible 00:27:16] line represents our false negative cases, reflecting 80.5 percent screen capture rate, which turns out to be comparable to national data.

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We also looked into the effects of prostaglandin and oxygen supplementation among these cases. Even after receiving these treatments, these newborns still failed at screening. This may imply that newborns with CCHD born at altitude may present most of the defects, but it also implies that even though our current restraining algorithm was based on studies at sea level, it is sufficiently effective at altitude.

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To summarize, this study is the first comprehensive review of newborns with CCHD at altitude. Our unique study design and reasonably large sample size allowed us to further delineate the average and range of oxygen saturations of this population, and to directly evaluate the problems of current screening algorithm.

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Overall, knowing that applying current algorithm at altitude produced approximately 80.5 percent capture rate, gives the [inaudible 00:28:31] assurance in moving forward with CCHD newborn screening in Colorado. It is definitely not a perfect screening algorithm, but it seems to perform as comparable at sea level.

Where do we go from here? In a separate study being conducted by our collaborator, we have data on the true negative cases from a hospital, so we may combine this data to get a more complete picture on the performance of the screening algorithm when applied at all [inaudible 00:29:01].

And before I end, I'd like to thank my collaborators. Chris, John and Marcy, who have been instrumental in this project.

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Last but not least, I'd like to thank all of you for listening in and for your efforts in ensuring the progress of newborn screening. Thank you very much.

Lisa : Merlin, that was fantastic. Thank you so much. We definitely learned a lot from your interesting, novel research project. I love your picture of the cute baby seat to end on.

Now we definitely have some time for questions. If folks can just feel free to either type into the presenter box, questions or you can feel free to ask them out loud.

I did have a brief update. I know Jamey, you showed what we affectionately call the pantyhose diagram, or the screening tool, which was the chart with the red, yellow, green, listing out the algorithm, depicting it in a diagram. I just wanted to mention that that is something that Cynthia Mueller, a Virginia nurse, shared with the National Collaborative, so I wanted to make sure that folks knew where that came from. Thanks for highlighting that as a helpful tool that's kind of spread nationally, after being developed first in Virginia by a nurse.

I had a couple follow up questions for our presenter. Jamey, I know you mentioned that some of the initial work you did in Kansas was making sure that centers that didn't have procedures could sort of share with what other centers who had been screening for a while were using, which kind of made me wonder in Kansas, do you have an idea of whether most hospitals are using the same protocol? Specifically, do you guys use the American Academy of Pediatrics endorsed screening protocol, or do you see significant variation between centers on which algorithm they use?

Jamey: I think that across the board they are using the same one. I know when Janay did the teaching, that that's one that she would have been presenting to use. We haven't seen a lot of change from that.

Lisa : Interesting. That's good news. I know it's easier for results reporting if the centers are using the same algorithm.

I also was wondering with the roll-out of the results on birth certificates, what kind of information will you be asking centers to report? Will it strictly be pass/fail, or are you looking for other information as well, such as the pulse oximeter screening readings for pre and post-ductal stats?

Jamey: We are asking for the number of times that they screened, we are asking for the date and time of the screen. I have one right here I'm going to pull up.

Lisa : Sorry to put you on the spot.

Jamey: That's okay.

Lisa : It's definitely great work you're doing in Kansas.

Jamey: How many screens were completed, the date of the ... And then we have them report on the final screen. The date of the screening, the time, and then whether it was military, or regular time. Then the screening results, the upper extremity than the foot and the difference, pass or fail, and that's a drop down box. And then if it's fail, they get a further drop down box whether they were referred to a cardiologist or facility, where they were referred, and the name of the cardiologist.

Then there's a box for a reason for not screening, so we have a list, a drop down box there, and then other if they have something else.

Lisa : Sounds great. Thank you.

I wanted to at this time see if other folks had questions for Jamey and for the Kansas team.

Ruthanne, do you have any questions that have been typed in through the webinar ready talk?

Ruthanne S: No, I do not see any.

Lisa : Okay.

Merlin, thank you so much. I know, certainly, this is very novel work and congratulations to you and the team in Colorado for examining CCHD screening at moderate altitude.

I know you mentioned that there hadn't been too many large population level studies at sea level, and I was wondering if you were familiar at all with some of the international publications? I think that's very true in the United States, we haven't seen any big population based studies with outcomes that are sort of multi-staged or have large volumes such as we've seen out of Europe, for example. The Granelli study, and the recent studies, from not Europe, but New Zealand, and Australia and China.

Do you have any familiarity with their work, and have you been able to compare that at all with what you're finding?

Merlin: I'm sorry, but would you clarify are those studies based on already diagnosed babies with CCHD or how do babies go through screening?

Lisa : I think a lot of the studies are looking, examining specifically, the CCHD screening results and then looking at the specific diagnosis of those who have cardiac diagnoses as a result of screening, and then a lot of time they will say ... A lot of them break down by defect what the pulse oximeter saturation readings were at the time of screening.

Was your study sort of a little bit of a different focus? Certainly at moderate altitude, definitely, it's novel.

Merlin: That's right. I'm afraid I'm not so familiar with the research but thanks for pointing them out to me. I can go look for them. But most is that I have, that I'm aware of, is based on how the babies are being screened for CCHD and then looking at this, creating results, and breakdown of the diagnosis after that.

I'm not aware of any other studies that look at it from our angle, in which case we are already looking at babies already diagnosed with CCHD, and then reapplying the screening protocol on them.

Lisa : It's fantastic work. Are you guys planning on publishing this soon?

Merlin: Yeah, yeah, definitely. We are currently drafting the manuscript, and hopefully it will be published soon.

Lisa : That's very exciting. Congratulations to you team. We'll all keep our eyes out for that manuscript in the near future.

Any questions for Merlin before we can consider wrapping our webinar a little bit early?

No questions? Ruthanne, do you have anything? Hello.

Ruthanne S: Hi there. I just received a comment, give me one moment.

This question is unrelated to today's presentation, but someone was curious if any of the states are collecting perfusion index in addition to pulse ox results in their screening. It's saying there some young infants who have screened fine, but then a few days later present with critical corrosion of the aorta, so this person is wondering if a perfusion index would help detect these babies? We have also seen false negatives with HLHI.

Lisa : So do either of our presenters care to weigh in on this topic? I know it's a little bit more general to CCHD screening rather than the specific work going on in Colorado or Kansas, but certainly ...

Merlin: I'm afraid I'm not in that area to be able to address the question.

Lisa : Sure, no problem.

Jamey, did you care to comment on the question about peripheral perfusion index and ...

Jamey: I don't have anything to add.

Lisa : I think it's a fantastic question, and it really is a topic that we hear about at the national level. I actually have spoken with some of my colleagues who have been working in the field for quite a while on this topic. I know in some of the initial studies coming out of Sweden, the Granelli work, she really felt that peripheral perfusion index might be helpful in identifying co-arc patients, specifically because I think healthy babies generally have a perfusion index of one or higher, and co-arcs may, you might see closer to the point seven value.

I spoke with Dr. Oster at the CDC in Atlanta recently, and we had a conversation around this topic, and right now, I think most of the experts agree that there really isn't enough data to answer that question at this time, which I know probably isn't what we're all hoping for.

Certainly I think as more research comes out and as other investigators examine these issues, we'll have more to be able to comment on as in terms of whether or not we can get closer to detecting those left sided out flow tract defects like coarctation.

Did anyone else ... I know we have a lot of screening experts on the call. Did anyone else care to weigh in on this question? I'd be happy to open it up to others, especially the pediatric cardiologists that might be on the call.

Ruthanne S: This is Ruthanne. I did receive another comment in the comment box.

Someone suggested that they hadn't seen any reports of implementation on perfusion index, which shouldn't be implemented without study. The problem is the timing. Co-arcs usually doesn't become detectable until after duct closes.

Lisa : I would absolutely agree with that. Thank you to the person that added that comment.

Do you know who it was, Ruthanne?

Ruthanne S: Yes, it was John.

Lisa : Thanks, Dr. Hokanson, appreciate it.

Any other questions for either our presenters or just other things you'd like to discuss in relation to CCHD screening?

Well, we will be ending a little bit early this afternoon. We actually had originally lined up three speakers, but due to scheduling conflicts, we were able to only have two this webinar, but we will look forward to sharing with you more information on CCHD screening and examining further topics during our June webinar. So please look out for the emails announcing our June webinar, and thank you so much for joining us.

Thanks, everyone, thanks so much, Merlin and Jamey, we really appreciate it.

Merlin: Thank you so much.

Jamey: You're welcome.