

MCH EPI Conference

Plenary II: MCH in Indian Country - Partnerships to Identify and Address Health Disparities

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THALIA WOOD: Thank you, Judy. Actually, I want to thank Judy and Laurie both for inviting me to be here today to talk to you. I am actually now at the MCH epidemiologist but that unit is within the same section I work for in The State of Alaska and we work very closely with them, and I'm thrilled to be here. Now, those of you who've heard me speak in other states, because this is my fifth conference this year that I've spoken nationally and I always show these slides, if you've seen this before I apologize. But I really get of having Alaska being shown as, like, really tiny and south of Arizona because we're really not that small. We take up four time zones, and there were four time zones when I moved there in the '70s. Now, we just have one because it's light all summer, dark all winter so it doesn't matter what time it is. And you probably all heard by now that we actually are up north between Canada and the Russia. And that's, between those two countries, so.

I really like maps, so I'm actually going to show a few maps because I decided I missed my calling and I should have been a geography teacher. But this is just a map of, another map of Alaska that shows broken down by the major tribal groups. And part of what I'm going to talk about today from the newborn

screening perspective are two conditions that are found mostly in the Northern and Western parts of Alaska. And then, here's another map and this is broken down by Alaskan native regional corporations.

And interestingly enough, what happened then back in 1997, the Alaska Native Tribal Health Consortium was formed. And what it was, it's a nonprofit health organization, owned and managed by the Alaska Native tribal governments and the Regional Health Organizations. It was created to provide statewide native health services and what they had established as part of that group then is the MCH epi unit as part of their consortium. And of course, we have our own MCH epi unit at the state. And what's happened is that they started to kind of work with each other. They are on some advisory groups for each of those two entities, but what I had learned today from somebody from my unit is that they tend to divide the state up by service regions when they're talking about MCH epi data, whereas the state tends to go by geographic region. So, there's a little bit of a disparity. And I think there's a lot more room for collaboration. And I think Stacy is right, I think we all need to work together and work on some of these issues. But I also hear that we, on the State of Alaska's MCH epi unit, has put up several data books, and they do have a future data book that's just like looking at the Alaskan Native data point.

Now, I have just a few population estimates for you. This is based on data from the Bureau of Vital Statistics. Even though we're a huge state, we take up four

time zones, there's less than 700,000 in our entire state. And so, and in a total by Alaskan Native is about 17 percent, or almost 18 percent are Alaskan natives this is from 2005 data. And then we have, I usually, when I talk about newborn screening, I talk about the fact that we have about 10,000 births a year. It's always between about 10,000 or 11,000 every year. So that year again, it was 10,453 and about almost 26 percent were identified as Alaskan Natives. So, even though they're only 18 percent of the population, they do have about 25 percent of the births. And that tends to remain pretty steady from year to year from what I've seen.

So, what I'm going to talk a little bit about today is, in Alaska we do call it newborn metabolic screening. And I just spoke at the National Newborn Screening and Genetic Symposium in San Antonio last month and they're trying to get away from calling metabolic because we screen for hemoglobinopathies, and endocrine disorders. And they want us call it the blood spot screening and, but my advise -- was established awhile ago when we're just going to stick with metabolic because that's what we've always called it. So, we know we screen for those other conditions.

And I was asked to speak at the MCH EPI Immunization conference that was put on by Alaska Native Tribal Health Consortium and that kind of also helped opened the door, I think, to also talking to them more about some of these issues as well. I happen to know one of the pediatricians at Alaska Native Medical

Center in Anchorage who asked me to be part of that plenary speech to talk about just this topic. And I'm just going to gloss over just a few things because I, since the time is little bit shorter today but for those of you that don't know about the blood spot or the metabolic screening, it was begun in Massachusetts, in Oregon in '63. We actually started screening for PKU only in Alaska in '68. And all states currently test PKU and hypothyroidism although most of the states are starting to expand their panels way beyond that, and we did as well. Up through the '80s and '90s, we were screening for six disorders. And then in October 2003, we started screening for more than 30 disorders with it expanded technology and that includes CPT1 and you will find out in a minute why I just picked that one disorder, and then we added cystic fibrosis last year.

On that first six things that we were screening before we expanded in 2003, one of them was congenital adrenal hyperplasia. It's an endocrine disorder that's inherited, defect of cortisol synthesis. And then part of the reason we added it early on, and a lot of states didn't have it added is we actually added this on to our panel in the early '80s because there was, they noticed an increased incidents in the Yupik population out in the Western Alaska. Some studies were actually done in the late '60s by Dr. A.J. Hirschfeld. And just as a side note, his son Matt Hirschfeld is a pediatrician currently at Alaska Native Medical Center in Anchorage. And Dr. Hirschfeld noticed that there was this increased incidence of this particular disorder in the Yupik population but at the time, there wasn't a screening test that would pick it up. And it wasn't until the late '70s that they

started to develop a screening test that could be used right off the blood spots that would pick up this disorder.

So then there was a paper written by Sonya Ping in 1982, where she discussed the fact that they had run a pilot in Alaska just to determine that it was feasible to pick this up on the blood spot test. So, we immediately added it to the panel. And of course, there's an increased sensitivity whenever something's picked up that's just predominant in one population, as you can well imagine. I mean, anything that kind of sets a population apart is going to be a little bit more sensitive especially to the people in that population. So, then what was next, like I said, when we expanded the panel in 2003, we didn't know what we were going to find. Everybody thought that since you expanded the panel to all these other disorders, the most the common disorder that people would find would be MCAD.

But what we found, is we started finding that CPT1 or Carnitine palmitoyltransferase I deficiency and that's why we call it CPT1 because we don't want to say that obviously all the time. It's an enzyme that aids in fatty acid metabolism. And it feeds, feeding inhibits the activity while fasting activates it. So, if you ever reduce enzyme activity, it can result in severe hypoglycemia in the fasting state. And the consequences could be serious illness and seizures. So, a little bit about the history of this occurrence. It was first described in 1981 and between '81 and '99, there was only 16 patients, that's worldwide. Before expanded screening, the total had increased to about 30 patients throughout the

world. However at this, about that time Canada had noticed an increased in their native population all in their Inuits. The first case in Alaska then was diagnosed in the beginning of 2004 right after we expanded the panel. We currently have more than 100 infants detected through newborn screening. But we also know we're not finding all of them. We did a kind of validation study of the screening panel using tandem aspect just this year. And we now think that we're probably missing about 90 percent of the kids. So, I mean, this year we've only have had 60 identified so far. And due to the estimates we find from that -- look at the validation study we think there's probably about 700 kids a year born with this. Now that's just, don't quote me on that because we're still looking at that. It's just to opened up lots more question that it has answers and so we just really don't know yet what it all means.

And there is also – I have cases identified in the Greenland Inuit population. So, we had another map and the only reason I'm showing this map is because all the phase, people, let me see this is, I hope that works, where is it? Anyway I, all of the people, children identified with this are all around the top of the world. So, if you look around the top of the world, they're all in Northern Alaska, in Canada, in Greenland. I don't think they're screening in Siberia and in Northern Russia yet, but if they were screening for this disorder, I think they'd find it in that population as well. So basically, they're finding it all around the top the world around the Artic Circle.

So, what does this mean? Like I said, it's definitely opened up more questions than answers. We really don't know what it means. Sometimes getting IRB approval can be a challenge because there's lots of people that are proposing studies and wanting to look at this. And it's becoming quite the buzz word in the newborn screening world about but what this means at Alaska. Fortunately, the pediatric clinicians at Alaska Native Medical Center are engaged in looking for answers and so they can sometimes help push some of this stuff through the Alaska Native IRB to have us look to see what this means.

Other physicians around the state are being educated on this condition. So, when they see children out in the villages, especially if they come in to the emergency rooms and they're hypoglycemic and non-ketotic, but they were considered screening for this and using -- we have an order form that they can use. They just need one blood spot off a finger stick. They can send it into Oregon and, well, let me back up on that. Oregon actually does our screening for us because we don't have enough babies born in Alaska to have our own public health lab do the screening. So, we have an intergovernmental agreement with Oregon. So, Oregon has developed the DNA test for this. And so, they can send a blood spot down to them and get the screening done on these children if they show up in emergency room. And the doctors are really -- do see a lot of kids showing up that are hypoglycemic and non-ketotic.

Right now it's too soon to tell if education will reduce the high incidents of infant mortality in rural Alaska. This is another slide that I borrowed from our MCH epi folks, it's from '92 to 2004. But if you looked at the numbers on here, you can see that the numbers for the infant mortality rate are much higher in northern and southwestern Alaska than they are from the rest of the state. And that's, right now, where all these children lived. They have been identified through newborn screening. And they don't live in the rural, I mean, excuse in the hub communities where there's a hospital normally like Nome or Bethel. They live Kwethluk, in Shishmaref, in little tiny villages and most of the villages out there have a community health aid.

So, I've done a lot of education to the community health aids as well to try to make sure they understand what this condition means. The management of CPT1 is pretty simple actually. There's no medications to take for this, it's just frequent feedings. So, it was determined that newborns to six months age should be fed every three to five hours. So basically, if the child starts sleeping all night, we encourage moms to go in and wake up the child and give them, it doesn't have to be a lot. We don't want to see obese babies, but at least to get their blood sugar back elevated. So, if an infant hasn't eaten for more than eight hours, then the advice is to say is to be seen right away by a physician or a health aid. And they really should not go more than 12 hours without any food. And if, they will need an IV. And it's also very important if the child's going to have surgery that they don't go n.p.o. that they actually have to have at least a

glucose IV pre-surgery to keep their blood sugars elevated or they can start having seizures before they go into surgery. A diet low in fat and high in carbohydrates may be recommended.

So, then we decided how are we going to reach the target audience? How are we going to reach the families out there with these infants who've been identified and the health aids who take care of them in most of these communities. So, we decided to make brochure to educate the families on CPT1. The collaboration included the metabolic nutritionist from University of Washington who comes up to Alaska to do metabolic clinics and the metabolic physicians from Seattle Children's. They did some focus groups with the Alaska native population mostly over Alaska Native Medical Center on the information and the layout in the brochure to make sure that they would feel that they were comfortable receiving something of this detail. Because this has got -- you can't do low literacy brochure with something called CPT1, I mean, you're spelling it out. You have to, but you have to get the information to them. So, they did some focus groups to make sure that they would find it usable.

And then, after it was developed, we distribute it widely to rural health clinics. And again, like I said, I've talked to the health forums that come to Anchorage, when their health aids all come into Anchorage for further training and try to make that they were aware of the brochure. I mail out -- frequently out to the

villages, especially if there's been a newly identified family in the village. And this is just to show what the brochure itself looks like.

The one thing that's incorrect on this right now is over in lower left hand corner where it says metabolic clinic. We actually are no longer seeing these children in Metabolic Clinic mainly because there's just too many of them, and it would overwhelm our clinics. So, instead we're really targeting education. We just feel that education is the way to go for the families, for the practitioners, for the physicians in those areas. This is the inside of the brochure. So, it's got the guidelines for feeding and what to do if your child is ill, and what -- make sure they get something to drink right away, some fruit juice or something, and to try to get their blood sugars up. Now under a treatment over there on the left hand side, it does say 10 to 25 percent of enzyme activity should be enough to maintain good health. And that was one thing we found when we first started discovering this condition.

Initially, there wasn't a confirmatory test for this. So, the first seven children all had to have tissue biopsies. And as you can imagine, that's pretty traumatic and invasive procedure to obtain tissue biopsies on these little babies. But what they found when they were doing the confirmation studies, that they all had some residual enzyme activities so they weren't totally deficient. So, we didn't know if that was somehow protective or not but what we have found over time is that some of these children have been symptomatic. If they go too long without

eating, or one mother was diluting the formula way back, way, way back because she started running out of money, and the child started having seizures because they weren't getting enough to keep their blood sugars elevated. So, it does, it can be symptomatic.

Now, I'm going to shoot over really quickly to newborn hearing screening because I do oversee the manager of that program as well. With newborn hearing screening, all (inaudible) hospitals were on board with screening by 2003, screening equivalent was placed in eight public health nursing centers in areas high home or midwifery center births or low hospital births, so that we can make sure that the children in those areas were getting screened as well. And then it was a pass by our legislature that -- mandating newborn hearing screening in 2006 with it actually taking effect just this year in January.

Now, we do have a high number of rural Alaskan infants being born. Approximately 40 percent of the babies born at Alaska Native Medical Center in Anchorage are the families from outside of the Anchorage service area. Often, like I said, from those rural areas so they come into Anchorage to deliver but then they go home pretty quickly out to their villages. So, if these infants have a misscreen before discharge, or an abnormal hearing screen, the goal is to have them re-screened by one month of age. But this isn't often possible due to the remoteness of some of these villages.

Okay, here's one way you can get the patients to the clinic, and this is actually mode of transportation in rural Alaska. In a lot of places they just traveled, there's no road system as you can imagine. Our state is really large but we don't have a lot of road systems. So you can get from village to village with the dogsled. I've been out to villages that only have dogsleds in the winter and four wheelers in the summer to get around. So, next steps for following up on hearing screening, we had to make a decision on how we would reach the most families after they went home to the villages especially if they should have had a re-screen and didn't get one done. So we decided to produce both in VCR and DVD form because this is back before, everybody just went to DVDs. We have lots of VCR format too.

We made a video specifically for the community health aids and practitioners in rural Alaska. We chose a vendor to produce the DVD and it outlines the milestones that should be reached at various ages and children with normal hearing. So, now it tells you at one month of how they should be responding at three months, at six months. So that the health aids who are examining these children can kind of look for these milestones. We've also distributed the DVD to public health nurses, to families, to anybody that's interested on having it. And it's been widely distributed throughout the state.

And again, we often talked about this at the health aid forums, and they've been all very receptive to getting this DVD and so they could use these milestones as

they look at these children. This is a picture of the front of the DVD. It's actually about 14 minutes long, and it starts off with just the sound of the birds that you see there, and talking about how, this is what we would normally hear and yet the child who can't hear wouldn't be able to hear the sounds of the birds out there. And it's just a very beautifully done DVD. We get lots of request for information. We get lots of request coming in for the DVD on the hearing milestones, for the CPT1 brochure.

And right now, we're also developing a DVD on CPT1. What we decided to do there is we've decided that the brochure, yes, the brochure is widely distributed but we also thought maybe it'd be more important, since I've had focus groups with Alaska natives and what they talked about, they were more receptive to seeing something, watching something than reading something. So, we thought, well, that may be a better way to reach this population. So, right now I'm helping co-produce a DVD on CPT1. It's going to be about 14, 15 minutes long. It's designed for families and providers out in rural Alaska where we talk about the feeding and the frequency of feeding, and symptoms to watch for, and when you should get that child right into a provider, and how important it is. And I think that we've actually used families who have infants with CPT1. And I think that's another important thing to do, is we engage those families in producing the video. So, they are actually starring, if you will, in the video. And so, that's really good to have them right there and they can talk about what they felt when they found out

that their child had this deficiency and what it meant to them. And that's just my contact information. So, thank you.