Bayou to Beltway
Pearson Cross' (PC) radio program on KRVS
Interview with Phyllis Griffard (PG) and Conni Castille (CC) about DeafBlind Cajuns
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Transcript

PC Hi friends, welcome to Bayou to Beltway. I'm your host, Pearson Cross, and today we are joined by two experts to talk about Acadian Usher Syndrome. The first person we'll talk to is Conni Castille, born in Breaux Bridge, got a degree in Philosophy at UL Lafayette, got a Masters in English with a concentration in Folklore, was a graduate of the Moving Image Arts program, created a film and is now Director of the Moving Image Arts Program. We are also joined by Dr. Phyllis Griffard. She was born in Morgan City, raised all over Louisiana and Mississippi, came to USL, got a degree in Zoology, a BS, then she got a Masters at Purdue in Medicinal Chemistry, taught at Xavier and then also at LSU, where she got her doctorate, and then she has been at UL Lafayette since 2015. Thanks for coming to the show.

CC Thank you.

PG Thank you.

PC So for people who, and that includes me, for people who are not necessarily that familiar with Acadian Usher Syndrome, what is Acadian Usher Syndrome?

PG Usher Syndrome is a set of conditions of deafness that is eventually followed by blindness. Many forms of blindness in this category are genetic, and it's a deterioration of the retina from the outside in. And this particular kind of Usher Syndrome, it's deafness that's followed by blindness starting in teen years, but profound deafness from birth. So all of these people are full members of the Deaf community. And it just so happens here in Lafayette or in the Acadiana area, because of the migration of the Cajuns, the gene, the allele that causes this condition, is just in the highest concentration in America. This created the largest population of DeafBlind individuals. So Usher Syndrome is any genetic condition of deafness that's followed by progressive blindness. And there's different levels, but the one we have here, Ush1C, is one of the most profound.

PC Now, does geographical isolation have something to do with this? So I'm thinking about the population of Acadians that moved here that carried that allele, that propensity, right? But then they got here and they concentrated and married each other. Did that create conditions for more people?

PG It could have but that's not really well documented. We don't have enough data about that. But it makes a lot of sense from a population genetics point of view, that that would be the case and a lot of that was because of, at least as much about language and religion and family connections.

PC Okay, I guess what I'm trying to get at is blindness and deafness, there are other conditions besides Usher's that do the same kind of thing?

PG They're all called Usher's.

PC Oh they're all called Usher's.

PG	Ours has a particular gene that does this, and there's others in other places in the world that do similar things.

PC	Yeah. Let's talk about that. Where else in the world do you find Usher Syndrome?

PG	Well, they tend to be concentrated in places, well, from what I understand, one of the biggest populations is in England. There's a Pakistani family that had settled there that has a lot of consanguinity is what we prefer to say.

PC	The commonality of blood.

PG	Yes. Exactly. And that seems to be the biggest population in the world, but ours is second largest in the world here in Acadiana and the largest in America. We don't really know the numbers…

PC	What are those numbers?

PG	If you ask the DeafBlind community, they will tell you it could be up to about a thousand and if we play with the numbers of what we think the allele frequencies are, that's about what it comes out. In the high hundreds to a thousand people currently living.

PC	So I was getting, reading some of the material associated with the show. I was getting back into my Mendelian genetics, you know, and thinking about how the syndrome works. So two parents both carry the recessive gene. The allele is recessive, that means you have one of their four offspring would then have, in all likelihood, be carriers, is that correct? So it's a 25 percent chance of.

PG	of having the condition.

PC	Yes, having the condition. And 50% chance of having a carrier, right? Yeah interesting. So it's one of those rare cases where the Mendelian stuff works out, right?

PG	It really does.

PC	It makes eighth grade worthwhile again. So we said between, you think, about a thousand people. There are differences between the ages of the people in terms of culture, and I want to get culture into this because people aren't Blind and Deaf in a vacuum. So it's here in Acadiana. It's a culture living in a community. How has that affected the development of the syndrome here?

PG	Well interestingly, you know, the Deaf community has many people who use sign language as their primary language here in America, and the causes of deafness are varied, but the majority of them elsewhere don't also go blind. But here in Louisiana because of the prevalence of this allele, about, we think, about 10% of the Deaf community here in Acadiana either will become blind or, and the blindness is progressive, so some people don't experience it profoundly until their 50s, some experience it earlier. But what has been interesting to me is learning that some of the Deaf people in Louisiana who have travelled elsewhere have seen that the way we sign here in Louisiana, the way the Deaf community uses sign language, is in a smaller frame, a smaller space in front of them, and that is because there's always been enough people in the Deaf community with a narrow field of vision as they get older that the sign language is…

PC	Needs to happen in a fairly concentrated... I noticed that you said earlier the retina fails from the outside in. So as it fails on the outside, the range of vision gets smaller and smaller, like a tunnel effect?

PG	Exactly.

PC Okay, so and then that affects the way that people sign here, so people make very small signs in a way

PG Maybe not very small, and they wouldn't know it unless they've traveled to other parts of the country where the signing is much bigger. The first person I heard this from was a DeafBlind man who had gone to school out-of-state and participated in sporting events at other schools. And he just noticed that, wow, these people in other places just sign bigger. And so he attributed that dialect to this history.

PC Well, let's talk about this and I want to turn over to Conni just for a second. But the question I have for you is how did each of you become aware of and interested in the Syndrome. Conni, you're a filmmaker. And how did you find out about this and think about this?

CC Well, I had the pleasure of meeting Phyllis. Phyllis had returned back to Louisiana, and she had contacted me regarding some of my films, one in particular I believe was King Crawfish. She's got a really cool program that I hope she talks about called OurBio where you use, how does it go? It's so poetic. You learn the biology of a culture through the geography of the people, something like that. So bringing real life experiences into the classroom, and I didn't know anything about Acadian Usher Syndrome or Cajun Usher, and I was just kind of surprised that, I mean I was born in Breaux Bridge, I grew up here. I'm Cajun and I didn't know anything about this subculture. So it definitely piqued my interest, and then we started working together. I started to incorporate it into some classes that I was teaching here at UL to get students involved. So we did pre-production basically in a classroom situation through a documentary class I was teaching and kind of using the students as a crew for pre-production and then here we are a few years later, still working on the project and making some progress.

PC And it should be mentioned that this film, the working title right now is The Quiet Cajuns.

CC Right. That's correct.

PC And when do you think it's going to be finished?

CC I would hope, I'm really hoping for this year, 2020, hopefully by summer, spring, something like that.

PC And hit the festival circuit at that point?

CC Perhaps. Yeah. And hopefully Louisiana Public Broadcasting. We see it as a UL Moving Image Arts production, that's, you know, has a lot of students involved in it on purpose. It's kind of like a service learning for them. But also having the experience of working on a documentary that has meaningful content, I believe. So that's the idea.

PC So, putting a plug-in for the Moving Image Arts Department. You know that many people may not know that that resides kind of within the bounds of the English Department here, but MIA, it's kind of its own department. But it's also a focus on storytelling. They say, well, why is there a movie, films hanging out in English? It's because you all are about telling stories, writing scripts, working with language.

CC Right. Professor Charles Richard was brought on to UL years ago to start a film program. And he wanted to put it in the College of Liberal Arts for that exact reason, for the storytelling aspects. And actually Allison Bohl Dehart and I were the first, kind of, graduates of it, well, I graduated in the English Department with the folklore concentration, but she was in the visual arts, and it was a perfect example of what Charles, what Professor Richard had in mind where you had these interdisciplinary students coming together to make media. And so our first film was *I always do my collars first*. And that was the first film to come out of the Moving Image Arts program.

PC Wow, it's a great resource. Phyllis, how did you get interested in this issue?

PG Well, I actually have Usher Syndrome in my family. I have two first cousins with the condition, and as a biologist and as an educator, it's a great way for humanity to shine a light on science and science to shine a light on humanity. There are too many stories and it's a culture. And it's easy for students, especially freshmen, to be able to see someone in their daily lives. After I teach about this condition or about the molecular architecture of the hair cells of the cochlea or whatever, I'll have students come back and say, “Oh, I was waiting tables and I noticed these people were using sign language in each other's hands and touching. I had never noticed that before” and so suddenly they become aware of a culture that was under their noses that helps them understand science and vice versa.

PC Very interesting. So a quick follow up there, we’re about to go to break. You have two first cousins, so are you positive for the allele as well? Or do you have any idea?

PG I don't know, but we always do the math in my classroom and figure out what my chances are.

PC Really, wow.

PG Yeah I make them figure that out

PC That really brings it up close and personal. Well, we're talking today to Conni Castille from the MIA Department here at UL and Phyllis Griffard from the Biology Department about Acadian Usher Syndrome, Deaf and Blindness. We'll talk some more when we come back.

Break

PC Hello, and welcome back to Bayou to Beltway. I’m your host, Pearson Cross. We’re talking today to Conni Castille from the Moving Image Arts Program at UL Lafayette and Dr. Phyllis Griffard from the Biology Department about Acadian Usher Syndrome. Question. How does the DeafBlind community in Acadiana address this syndrome? What's life like for someone in this community?

PG The DeafBlind community here in Acadiana, and really it's not just here, it reaches all across Louisiana and there are spotty cases around the country, obviously. But here in Acadiana, they are first and foremost part of the Deaf community. The older people especially, if they've gone Louisiana State School for the Deaf or they've been in the immersive Deaf program, for example at Lafayette High, they're part of the Deaf community. And they might use both sign language and speech, depending on their age. But the younger members of the Usher Syndrome community may not be deaf because the parents have chosen to use cochlear implants. So there's lots more, the answer to this question about what life is like really depends on whether they are part of the Deaf sign language community or the oral community or both. But for someone who is deaf, most who don't have vision loss, or vision loss yet, they have jobs, they have job training, they have support for education opportunities. But for those who begin to lose their vision, they're encouraged to participate in training that’s sponsored by the state for preparation for blindness, in household challenges, getting around, navigation challenges. So even here on the campus of UL Lafayette, we often see people walking with white canes across St. Mary and down the boulevard, and that's because these are all people who are training at the Affiliated Blind of Louisiana facility right on St. Mary. Now, that's for anyone who's losing vision, but because we have such a concentration of DeafBlind, there's a lot who go for residential training in Braille and using the cane and household services.

PC A number of things come to mind here, one is that if you told me today that I was going to lose my vision in a progressive manner, I would become incredibly, I think, depressed about it. I mean, is that an aspect? How do people deal with this knowledge that, gee, you're not only deaf but, you know, at some point yet to be determined, as you get older, you'll lose your vision too. I mean it seems those are the two most important senses that we have.

PG Most people who are deaf and are part of this community, most of them know if they're going to have Usher Syndrome. Nowadays children, are if they're found to be profoundly deaf at birth, are given a genetic test soon after birth. But in older times, people who were about 50 now probably didn't find out till they were in high school that they would be losing their vision and they started to have night blindness. But what I've found by talking to lots of people in this community is, in some families, there's just lots of deaf people, and deaf blind people. And so they have always had lots of sign language in family events. And they're very independent, fiercely independent, and in fact…

PC Well let's talk about that. What does it mean to be Cajun? And what does it mean to be, you know, how do you live in this community?

CC Right. Well, that's one of the, hopefully, what the film will kind of reveal, this subculture of Acadian culture, of Cajun culture. And Phyllis has relatives that have Usher. Acadian Usher. I did not grow up around it. And I was, I was just blown away of the joy and happiness and the liveliness, and how just the energy in her family. We have two people in the documentary that we follow, representing both generations. The older generation that as Phyllis said, didn't even perhaps know about Acadian Usher and that they would lose their vision later and certainly didn't have the technology of the cochlear implants. And then we have someone representing the younger generation that can take advantage of the cochlear if they choose to, if their parents choose to. But as far as the culture, I noticed they enjoy their boiled crawfish more than you and I do (laughing). The other senses are I guess more enhanced, as you would expect.

PC Do you think that's the case? Is there any data that suggest that that's true?

PG I don't know of any official data, but anecdotally, this is the other part that happens, is if you talk with your hands, you can't eat and talk at the same time. So you have to eat, and really enjoy it, take your time, and even if you can't see those crawfish, I've seen them go through some crawfish without seeing them.

PC That was funny. I had this vision of someone [eating crawfish]. Now the way it works is if you're talking with your hands, to someone who is blind that person has to actually touch your hands in order to tell what you're saying, right? Is there a way that you sign that you make what you're doing more clear to someone who's seeing or hearing with their hands as opposed to with their eyes?. Because when you make a sign of a letter with your hands, I mean that's a different thing than having someone do that on your hand or something, right?

PG It is, but I'm when I watch people using tactile ASL, tactile American Sign Language, it's a really beautiful thing to see because they're really feeling the motions of the arms. In ASL you're using two hands, but when they're holding your hand, they're only using one, so they have to fill in lots of gaps. And so it usually works best when the interpreter for the DeafBlind person is someone they're used to interpreting for them, because they're very used to how it feels when they're being interpreted for.

PC Is that interpreter someone who's provided by the state? or is that usually a family member? How does that work?

PG It's almost always someone provided by the state. Actually there's been a program that was advocated by the DeafBlind community itself. Some of these leaders that pushed, I guess it's now 20 years ago, maybe less, that pushed to get funding for what are called Support Service Providers. And these are people who can use sign language. Some are deaf, and not blind, but can drive, can help interpret and have a lot more visual support for these people. So they have a certain number of hours, depending on funding per month. And so that one of the things the DeafBlind community is constantly doing is having to advocate to the legislature or to the Deaf Commission, the Commission for the Deaf, for more hours so that they have more independence, more mobility, more participation in jobs and schools, doctors' appointments, etc.

CC And is it, once a month, they have their social at a coffee shop here in Lafayette, and it's really nice that they all come together. They communicate, it's a nice thing to witness.

PC That's amazing. Let's talk about cochlear implants. Has that been those are you typically do it fairly young. And you put it in your ears and then people can hear but there's some good things about it and some drawbacks, could you talk about that a little bit?

PG Well at a technical level, what it does is it brings only about... the best ones now bring about 32 frequencies of sound into the cochlea. And so what people hear is not a complete spectrum as someone who has normal hearing has. So there's always intensive speech and hearing therapy for anyone who's been born profoundly deaf and never heard anything, to learn how to interpret those 32 frequencies. And so there's a big concern about...

PC Can I stop you just for a second, how many frequencies does a person hear?

PG Ooh, A thousand frequencies?

PC So is it, when you say 32 frequencies, is it in a particular range or is it all over the place, or is it just kind of like a fence with a whole bunch of missing rungs or something?

PG That's what I picture, exactly like that, and so a lot of the speech and hearing therapy is about using other cues to interpret and add to, so watching your mouth and these sorts of things. The Deaf community has always been reticent to support cochlear implants, because they are very concerned that people who are confined to using only hearing with cochlear implants don't get the full richness of language that they have with sign language. And they really want them to be a part of their community.

PC I would think, you need both really and lip reading or something, you know the whole spectrum but I can understand. So are you going to be in the film, are you addressing people who are at various stages of all this? You said you're going to follow a young and old person?

CC Right

PC So are those people, do either of them have cochlear implants or not?

CC The young boy does, and he plays piano, he belongs to a dance troupe. In fact when he, I guess, they get upgraded a lot, these cochlear implants. His mom told us the story of the Bluetooth now with the cochlear. And they're driving along and she's got her, you know, GPS mapping going on, and it wasn't coming through the car radio. But he started saying, "Mom take a left here, Mom take a right there", because it was going through his cochlear Bluetooth. And then the older, the other person that in the film represents the older generation that did not have access to cochlear implants. He's actually a big advocate for the DeafBlind community. And so we kind of talked about all his work with that as well.

PC So if this condition is a recessive trait and its conditions are so severe. I mean, I would think Deafblindness is pretty severe, how come it didn't die out through natural selection?

PG Ooh, that's a great question. That's not how the genetics works, unfortunately, if you play with the math. And also the allele is silent. So it's often there, and also people who are deaf and go blind still have families, and their children can be carriers, or even if they both have Usher Syndrome, their children will all be deafblind, and it's not lethal. So there are many conditions like that

CC The mother of the young boy in the film says, you know, there's so many other things that we could be dealing with, and this is really not that bad when, you know, in retrospect.

PC You know, I'm kind of thinking like some of the same things that are raised by other genetically inherited diseases, like where parents have to, I mean, I would imagine now that people can get tested for this perhaps? And so if you meet someone in the Cajun community or someone who has it in their family, you can find out what your odds are and if you find out, I mean, if you both have it, you would have to maybe talk about, well do we want to have children or not, or how do we feel about this? Is that part of what it means to be part of this community?

CC I think so, and I think if you've grown up with family members that have Acadian Usher, you don't really see that it changes your life in such a meaningful way that you would not, it would not make a difference in who you decide to marry and have children with because it's such a rich community and a rich family with love, happiness, joy. You see that. But if somebody who never had that experience, or was ever around Acadian Usher, I could see what would be more daunting and like whoa, wait a minute. But that's kind of one of the things too that the film explores as well.

PG We did ask that question directly, maybe not. I don't know if we got it on film but the people in DeafBlind Community just kind of shrug their shoulders and go, “we wouldn't change a thing. This is just, yeah... Well, we’d kind of like our vision, to hold onto our vision longer because we can drive…”. So they wouldn't mind having a cure or a fix for slowing down the blindness but as far as regretting what their hand was dealt…

CC I think the parents of children with Acadian Usher have a harder time, maybe because they don't have that experience and understand that, hey, it's okay. Just like all of us that can hear and see, we don't really know what it's like.

PC We're coming right to the end, we’ve got about a minute left. Tell me, is there hope of a cure? Will gene therapy do something? Can CrispR splice in some new genes and get us out of this?

PG Actually there’s a very promising, several promising therapies. We have a symposium coming up every couple of years. LSU Health Science Center brings in a researcher, Dr. Jennifer Lentz, to update the community on what's happening. And she has a very promising gene therapy. The problem is, and it works great in mice, they have the exact same gene. It cures them of blindness and deafness and their vestibular disorders. But yet it's a bit of an orphan disease, and so making it to the next step of FDA approval or getting a pharmaceutical company to do this, especially when just a few doses will actually prevent the progression of the disease. So it's a complicated answer, but yes, there are solutions on the horizon.

PC Wow. Well, let's certainly hope so. In the meantime, it sounds like a fascinating film and certainly discussion. We've been talking this morning with Ms. Conni Castille and Dr. Phyllis Griffard about Acadian Usher Syndrome, a film called The Quiet Cajuns. It's coming out in the Moving Image Arts Department and the Biology Department. So thank you for joining us this morning.

CC Thank you.

PG Thanks for having us.