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Brian Martin: From UPMC Children's Hospital of Pittsburgh, welcome back to That's Pediatrics. I'm Brian Martin, Vice President of Medical Affairs here at Children's.

Carolyn Coyne: I'm Carolyn Coyne. I'm a scientist in the division of pediatric infectious diseases.

Brian Martin: We're happy to welcome Dr. Ken Nischal today to the program. Dr. Nischal's the Chief at the Division of Ophthalmology, Strabismus and Adult Motility. He is the director of the pediatric program development at the UPMC Eye Center. His research interest include craniosynostosis, ultrasound, and pediatric anterior segment therapy. Welcome Dr. Nischal.

Ken Nischal: Thank you.

Carolyn Coyne: Welcome. Thank you for joining us. So, I'm a basic scientist and that sounded very complex and complicated to me, what Brian just introduced you as. Why don't you tell us a little bit about what you do.

Ken Nischal: So, I'm an ophthalmologist who did a fellowship after my resident training specializing in pediatric ophthalmology. And I did that at Sick Kids in Toronto, and I developed a specialist interest in anterior segment developmental anomalies as well as craniosynostosis problems. And part of that I developed an interest in ocular genetics. So, my role at the moment is I'm the director and the division chief of pediatric ophthalmology, strabismus, which is when the eyes are not aligned in children, and adult motility. So the same thing can happen in adults as well. I've been here since 2011, the end of 2011. I was the chief at Great Ormond Street Hospital Children in London. And then I got recruited by UPMC almost eight years ago. And my remit when I was recruited was to build the division, because it had lost its previous chief who had moved away, and I was asked to come in and build up the division, which I think I've done over the last eight years.

Ken Nischal: One of the interesting things is that traditionally most ophthalmologists have super specialized into areas of the eye. So you have cornea specialists, cataract specialists, glaucoma specialists, retina specialists, neuro-ophthalmologists, uveitis specialists, and in the United States, and most parts of Europe, pediatric ophthalmologists have really done a little bit of everything, which has been counter to what the rest of ophthalmology was doing. And part of the reason is that pediatric ophthalmologists are not common. And so if you're working in a rural setting, for example, at the middle of Oklahoma, and you are a pediatric ophthalmologist ... I don't know why I chose Oklahoma by the way. [crosstalk

00:03:20], because my geographic understanding of the United States is very limited. Maybe I should've said Nebraska.

Carolyn Coyne: You can apologize to [crosstalk 00:00:03:29].

Ken Nischal: Yeah.

Brian Martin: I'll take rural for 200.

Ken Nischal: What is Oklahoma? So, if you have one pediatric ophthalmologist, often adult ophthalmologists don't want to deal with children, because it brings with it its own stresses and difficulties, because firstly, children won't sit still for you to examine them. Then you have to deal with the parents, because the understanding and the conversation is not only with the child, but with the parents. And so a lot of adult ophthalmologist will have very busy, fast turnover practices. And if you have a child come in, it could take 45 minutes out of their whole day, which ... And they don't want to do that. So, if you have one pediatric ophthalmologist, then often they're forced to do a little bit of everything. But if you work in a center of excellence, like UPMC Children's, then we attract patients from quite far away as well as locally. And you then have the option of getting individuals who are in our department to super specialize in one area.

Ken Nischal: So for example, Ellen Mitchell is a pediatric neuro-ophthalmologist. Very difficult to come by, but we have one here. And then we have a new addition called Dr. Patel who is not only a new ophthalmologists, but a specialist in nystagmus. So, that's when the eyes don't hold still and wobble. And there are various treatment regimens now that can be applied to help those children. And in fact, the sooner we see them, the better. Firstly, you make the diagnosis as to why it's going on. And secondly, you then can offer various treatments. We have Matt Pihlblad, who has a special interest in retinopathy of prematurity and adults strabismus. So he does a lot of the adult strabismus. We have two pediatric optometrists, our newest edition, Erin Keim has a fabulous reputation in concussion management. So, she manages a lot of the children who have been concussed in sports injuries and she really is making a big difference to our population that before we didn't really know quite what to do with and she's been trained and done a fellowship in that.

Ken Nischal: Then we have Craig Luchansky who's a contact lens specialist. And so it may seem a bit bizarre, but the epidemic of myopia in the whole world, but also the United States means that we've set up a myopia treatment center. And so children who are progressively becoming myopic, nearsighted, can actually have interventions and one of those interventions is to use corneal reshaping therapy. So, it's really interesting, somebody said to me once that your mother tells you something when you're 12 and you actually don't hear it or understand it until you're over 40 and that actually is very true. So my mother used to say, "Don't wear your glasses all the time. They'll make your eyes weak." Well, scientifically that's true.

Carolyn Coyne: [crosstalk 00:06:40], take my glasses off.

Ken Nischal: She used to say, "Don't read in dim light, your eyes will get weak." And scientifically we've proven that that's true. So, there are various environmental factors and other interventions that Craig is leading in the myopia treatment center. So, I do a lot of the pediatric cataracts, believe it or not, children can get cataracts. And I do corneal transplants in children. So, there are only a handful of doctors in the United States that do that. And as a result, we have a lot of patients coming from all over the United States and all over the world for corneal transplantation and also glaucoma. So those are my special interests here. Dr. Patel, who is starting, not only will she be doing nystagmus, but she'll be taking on the children who have craniosynostosis, mainly because I have so much to do in terms of my other roles here at Children's that I need someone to be taking on some of those craniosynostotic projects.

Carolyn Coyne: I'm thinking about diagnosing a lot of these things. So I'm the mother of a 10 year old who constantly actually says that he needs glasses despite the fact that there's actually no data to support that he needs glasses. I think he just sees that mommy and daddy wear glasses. So I'm thinking about the difficulties you mentioned before, why people don't go into pediatric sort of specialty looking at these things and if part of this is just diagnosing it in children. So when does this become obvious when children need to see you and is that part of the problem even getting children in to see you?

Ken Nischal: Well, one of the things that's quite interesting is that we have a school vision screening program in Pittsburgh and in the surrounding areas and most actually States have a mandated program. The problem is that the quality of that screening is not always the best. So you often get lots of false negatives, sorry, false positives, and that's not, that is a problem and it isn't a problem. I'd rather have a false positive and then say everything is okay then have somebody missed. So, the first thing is that the school screening program is pretty good. What's really interesting is my more recent hat is medical director for telemedicine at Children's. And Dr. [inaudible 00:09:11] and myself have been talking to the Pittsburgh schools about how we can deliver telemedicine to the school population. And within that remit would be how to provide better screening.

Ken Nischal: There are automated photo screeners, there are also apps that you can put on a regular iPhone or an Android or a Samsung or whatever you like, which will allow the child to take a picture of themselves or a health professional take a picture of the child and work out if they have an eye problem. So, I think that the first thing is that we need to improve how we screen children. And that's one issue. The other issue is parents are normally very good at picking up that something's not right. So if a child's squinting their eyes and sort of piercing their eyes closed or trying to see the television or sitting up close or bumping into things, all of these have been common reasons why children will get sent to us for an evaluation. We often ... We have a very close relationship with our genetics department and the genetics department send us lots of patients

because the eyes sometimes can clinch the diagnosis in a child with multiple eye, sort of nonocular problems and we often will make the diagnosis of a condition where they have several possibilities and we look at the eyes and go, actually I think it's going to be, this is going to be the most likely to help navigate the molecular testing.

Ken Nischal: One of the things I'm not sure people realize is that, and I didn't realize until I came here, I've worked a lot of what I considered prestigious hospitals. Hospital Sick Children in Toronto is very well known. Great Ormond Street Hospital Children is the oldest children's hospital in the English-speaking world. In fact, a brick was taken from Great Ormond Street and laid in the foundation of the children's hospital of Philadelphia when that hospital was built, but the pathology that I've seen here at Children's Hospital of Pittsburgh, and more importantly, the expertise outstrips all of those hospitals. It's really quite extraordinary. The breadth of patients with unusual problems has led to us developing the Center for Rare Disease Therapy, of which ophthalmology's is an active participant, because we'll often get children with a rare disease and it needs metabolic medicine, genetics, neurology, ophthalmology to work together to create not only a treatment plan, but sometimes even a diagnosis.

Ken Nischal: One of the things that I think is really important is that when patients come here that they, and their parents, they understand that we, all the practitioners in my division work to common, consensus led, evidence based protocols. So if I'm away and they've been seeing me and I'm on vacation, because occasionally I like to go away for seven days and relax by the beach, then if one of my colleagues, he's one of my patients, they get the same treatment, the same treatment protocol. And that's really important. Because we have treatment protocols, we're able then to assess outcomes and we do that. Our outcomes are on the website so that people can have a look and see what our strabismus surgery outcomes are, what our pediatric cataract outcomes are. And it's ... I think that's really important. It's something that was a bit of a culture shock to me when I came to the United States, that everybody says they're really good, but there's no evidence.

Ken Nischal: And I think that that ... I think parents should ask for evidence and should ask ... Shouldn't be afraid of saying to their practitioner, well, okay, my child, you're saying my child has a pediatric cataract? How many have you done and what are your outcomes?

Carolyn Coyne: Especially when it's your eye. You want to make sure the person operating on your eye knows if he or she [crosstalk 00:13:27].

Ken Nischal: Right. Well, it's your child's eye, it's even worse because you have no control over what's going on. So I think that that's really important. You talked about diagnosis. One of the things that's really important, and I've ... It's taken me eight years, but I managed to bring from Great Ormond Street, a gentleman called Dr. [inaudible 00:13:48]. He's probably one of the world's leading visual electrophysiologists. So what that means is this, he can tell if a child has an

intact visual pathway and has the potential to see, even if the child can't verbalize anything. And he can also tell if the retina, which is the photosensitive layer in the back of the eye, is functioning. So we now at Children's have an enormous data bank and treatment opportunities for children with retinal dystrophies. You may have heard recently that the FDA approved Luxturna, which is a treatment for labors congenital amaurosis. That is an early onset retinal dystrophy that is blinding and Luxturna is an injection of a type of virus that has been ... All the nasty bits of the virus have knocked out so that it acts as a carrier to take the healthy gene into the retina. We have several companies working with our division and our adult division working with children who have other types of retinal dystrophies, and because we have doctorly assets and we can do their retinal function, we know which ones are going to benefit.

Ken Nischal: But we also have dedicated genetic counselors, very unusual for a pediatric ophthalmology department to have dedicated genetic counselors. And that team is led by Hannah [inaudible 00:00:15:29]. So we're able to not only make a diagnosis clinically, but we can from a molecular point of view, make a diagnosis, and then see if there's a gene therapy that's available for these children, either in Pittsburgh or across the country. We've sent two children to Portland for a treatment of a particular type of retinal dystrophy. So, I think that we really are providing what I would consider comprehensive pediatric ophthalmology care at the division.

Brian Martin: That's exceptional story and Dr. Nischal, as somebody who has had my own child in your clinic and has benefited from your care, one of the things I was really struck by was the, what I would describe as the marriage of technology and a great sort of human touch in the care that you provide. Reaching back a little bit to the myopia center, would you mind, sir, sharing with the listeners some of that story. I was struck by the adage is like you said, that your mother tells you when you're young, that you realize late, particularly with something as common as myopia. Could you speak a little bit about the history of that, of the center?

Ken Nischal: Yes.

Brian Martin: It's reasons and really what we're doing here.

Ken Nischal: Yeah. So we ... So, in 2000, in the year 2000 I was at Great Ormond Street. I was attending there and I had a fellow who came to me from Singapore whose name was Audrey Chia. And Audrey told me about the tremendous epidemic of myopia. She was saying that 85 to 90 percent of 12 year olds were myopic in Singapore. And the problem with Singapore, or not with Singapore, with myopia, is that it has a tremendous economic burden. If you think about the number of glasses people have to wear, the lenses have to be changed, contact lenses, contact lens solutions, and then you go into LASIK or refractive surgery, it's a very expensive problem. In fact, it's been estimated in 2014 that the economic burden of myopia is 265 billion U.S. dollars worldwide. And now if you add to that the fact that the longer the eyeballs get, so when you have a long

eyeball, you're nearsighted. The longer the eyeball gets, the greater the risk of common problems like glaucoma.

Ken Nischal: So, if somebody comes in and they're at 40 and they're being checked for glaucoma, the first question we ask them is, are you myopic? Because that increases your chances by two to four times. The next problem is cataract. It increases the chances of cataracts. Sort of, you might get a cataract when you're 60 or 70. If you're myopic, you might get it when you're 50 or 60. Retinal detachment. Depending on the studies you read, it can increase the risk of retinal detachment from anything from seven to 15 times the normal population. And of course if you get a retinal detachment that doesn't get fixed, you lose your vision. The most devastating risk is if the eyeball goes longer than 26, 27 millimeters. It should be 23 and a half. If it goes longer than that, and you're myopic greater than minus six to minus seven, you can get something called myopic maculopathy, which is an accelerated form of age related degeneration.

Ken Nischal: So you lose your central vision. So, when you take all of that into account, it makes sense that if there's a way of stopping the eyeball from growing that you do that. So the Singaporeans and particularly Audrey Chia, and I'm very proud of this. This is a fellow that I never taught anything about myopia too, but she's really become one of the leading forces about myopia treatment. And one of the studies that she did was called ATOM2, where they basically found that a very low dose of atropine, 0.01%, that doesn't cause any accommodation problems can actually arrest the progression of myopia in up to 90% of children who have it placed in their eyes daily. I mean that really was revolutionary. And so now there are lots of studies going on not to replicate what they found, because that's already been done.

Ken Nischal: But to see if the effect of atropine is the same in a heterogeneous population like the United States rather than a homogenous one like Taiwan or Singapore. But what's really interesting is that the environmental factors that we submit our children to are much more important. And this is only relevant during the growing period of the eye. So the eye has three growth phases. From birth to 18 months, 18 months to eight years, and then the one we're interested in is eight years to 16 or 17 years, if you're a male, and some people say 21 years if you're a female. And during that time, if you don't control the environmental triggers, your eyes can really grow rapidly. And so the sorts of things we know that protect you from developing myopia is two hours daylight exposure daily. If you get two hours daylight exposure, you don't have to be outside, you can be by a window.

Ken Nischal: In fact, in Singapore now, they passed a law that no schools can be built with opaque walls. All their walls have to be translucent to let light in, to reduce the myopic effect. So, there's two hours daylight, don't read in dim light. When you read in dim light at night, children, we all did it right? We would lie down and we put the book between the light and us and we'd read in a shadow. Well, you put your peripheral retina into darkness and it drives myopia. So, when children

come to us, we instigate these environmental factors and then we think about whether we can use atropine, whether they're going to use a contact lens, and there is something else that's available but not available in the USA at the moment. These glasses, and I'm taking my glasses off at the moment, the typical concave glasses that myopic people wear when you look through them, everything looks smaller.

Ken Nischal: They make everything focused in the center of your vision at the back, but in your peripheral retina, they drop the focus to just behind the retina, and in the growing eye there's something that makes the retina the eyeball think, well, I better grow longer so I can catch those rays of light in my peripheral retina, because it turns out your peripheral retina plays a huge role in refractive error. And just to sort of close this topic, I'm minus five, my wife is minus six. None of my children wear glasses, because from a very young age, all of this experimental data was available and I was able to ban them from taking their phones into their bedroom.

Carolyn Coyne: So I was just about to ask screen time, because my ... When you were talking about reading and light, I'm thinking of my son who like the iPad, it's a constant fight to get the iPad away.

Ken Nischal: I know and they do it ...

Carolyn Coyne: So ...

Ken Nischal: And they do it in pitch black. They lie in their beds and what's happening is that they're stimulating their central retina, but not their peripheral because it's in darkness. So that's not good. It's clearly not good. I mean, there's a whole other issue about screen time, right? Whether you're in bright light or not, but there is definitely environmental triggers that you have to control. And I think it's really important. And the other thing was my oldest son was minus three quarters, which is not very much when he was about 10, and I used to make him take the glasses off when he got home, because he didn't need them. He liked them, but he didn't need them. He needed them at school. So I think it's important to do these sorts of things. I don't want people listening to this thinking that, oh, they shouldn't wear glasses if they're nearsighted. You have to be able to cross the road safely, right?

Ken Nischal: So we have to have ... You have to be sensible about this. They have to wear the glasses to cross the road, see the board at school, watch a movie. But we have to be careful about what they do. There's plenty of evidence that near work triggers myopia as well. If you .. In Taiwan, they looked at all the sons and daughters of Taiwanese university lecturers, and then looked at the children of farmers, all the farmers children were about plus one, all the university lecturers children were about minus five. So it's .. There is no doubt that near work plays a role. And I'm not saying they shouldn't do it, but the lighting has a tremendous effect and two hours daylight playing outside as well has a protective effect.

Carolyn Coyne: This gives me even more of a basis to go home tonight and say, "Put the iPad away."

Brian Martin: Exactly.

Carolyn Coyne: Well, thank you so much for joining us. This was really educational. Very interesting.

Brian Martin: Thank you very much.

Ken Nischal: My pleasure. Thank you.