ANNOUNCER: Welcome to Akron Children's Hospital. Over the next hour, you'll see treatment to correct a rare eye condition known as congenital nystagmus. The condition causes rapid, uncontrollable eye movements that often blur vision. Since it effects the nerve behind the eye rather than the eye itself, glasses and contact lenses cannot correct the condition. The procedure is intended to trick the brain by detaching and reattaching horizontal eye muscle to the same location, slowing down the uncontrollable darting eye movement. OR-Live makes it easy for you to learn more. Just click on the "request information" button on your webcast screen and open the door to informed medical care. Now, let's go to the moderator of our program.

AMARPREET SINGH, MD: Welcome to Children's Hospital Medical Center of Akron in Akron, Ohio, where today we'll be witnessing a new and exciting procedure that's aimed at treating a condition that causes one's eyes to wiggle uncontrollably. Good afternoon. I'm Dr. Amarpreet Singh, and I'm a pediatric ophthalmologist here at Akron Children's Hospital. My partner, Dr. Robert Burnstine, who is the chief of pediatric ophthalmology here at Akron Children's Hospital is currently performing the procedure in the O.R. right behind us. Before we get to the procedure, I urge you to use -- take full advantage of our presence here and use your -- use the MD--MDirectAccess button to please e-mail us your questions, and we will be sure to answer them in the order they are received. Without any further ado, we're going to go into the O.R. and see how we're doing. Boomer, how are you doing this afternoon?

ROBERT BURNSTINE, MD: Hello, Amar, we're doing fine. We already started the case. We're on the second eye muscle out of the four. What I just did was detach and reattach the outside muscle in the right eye, the right lateral rectus muscle.

AMARPREET SINGH, MD: Can you tell me a little bit about this patient?

ROBERT BURNSTINE, MD: Yes, he's an 11-year-old -- 11-month-old, sorry. Eleven-month-old little boy with -- that came to me five months ago with nystagmus. And with no family history of nystagmus. We did get an MRI on him to prove that he didn't have any serious problems, and he has what's called infantile nystagmus syndrome, or congenital nystagmus.

AMARPREET SINGH, MD: Okay, and right now, you said you are detaching -- you're just -- detached the muscle?
ROBERT BURNSTINE, MD: Well, I'm just -- I'm just now opening the muscle, or the conjunctiva to the muscle, on the left medial rectus muscle, the left inside muscle.

AMARPREET SINGH, MD: Okay. Okay. Let's talk a little bit about what congenital nystagmus is and what the definitions are of this. Congenital nystagmus is a -- is a condition which causes the eyes to move back and forth. This results in an inability of the eye to focus on any particular object and decreases one's vision. This -- the estimated incidence is about 1 in 3,000 people.

ROBERT BURNSTINE, MD: Other hook.

AMARPREET SINGH, MD: Oftentimes, due to the shaking, one can -- one will adopt an abnormal head posturing, turning to the left or to the right in order to dampen the shaking. Let's take a look at a video that shows exactly what it looks like in a patient of ours. Here is a child in our practice who, as you can see, as they're looking forward, their eyes are shaking uncontrollably back and forth. Right now they're looking straight ahead, and the visual acuity is actually fairly poor in this particular patient. You see that even when they look off to the side, it may increase, but the shaking is always there, no matter which way they're looking. So there have been a lot of different treatments for a nystagmus that have been done in the past, and none of them have actually had very much success until recently. The Dell'Osso procedure has come up. Did you want to tell us a little bit about the procedure?

ROBERT BURNSTINE, MD: Sure, I'd be happy to. Dr. Dell'Osso is a PhD engineer, he is not a medical doctor, and he has dedicated the last 35 years of his life towards the study of nystagmus. And he is a very brilliant and keen observer, and he observed that if we take the muscle off the eye and reattach it, for some reason, we don't know exactly why, it slows the amplitude of the nystagmus, which improves the vision.

AMARPREET SINGH, MD: When we talk about -- when Dr. Burnstine is talking about the amplitude of the nystagmus, what he means is this: when we have somebody's eye -- we have a diagram of an eye right here -- if you take a look at this eye, what we -- the way that vision works is light is passed through the cornea and through the crystalline lens, which then focuses the light onto the posterior part of the eye next to the optic nerve. This is called the fovea, or the macular area. So light comes through and is focused clearly onto the fovea. With a patient that has nystagmus, they have oscillations of their eye, so the eye wiggles back and forth as we just saw in that video, back and forth. Well, an image is never able to stabilize onto the fovea and create a good image there, so their visual acuity is decreased. What the purpose of this procedure is is to, instead of having the image go back and forth this fast, what it does is it slows the oscillations or slows -- decreases the amplitude and allows that image to stay on the fovea a little bit longer, improving the visual acuity. During the actual surgery, what's done is -- we have a diagram here showing a view of the eyeball with some attached muscles, if we could zoom in on that. What we see is the muscle origin is back in the posterior part of the orbit -- these are bones here - - and the origin of the muscle, the optic nerve coming through, and the muscle originates in the posterior part of the orbit and courses until it attaches onto the eyeball itself. In this surgery, what we do is we detach the muscle here and then reattach it to its exact same location. By doing this, we think that the proprioreceptors are reset and the amplitude of the jerk is decreased. Let's see how we're doing here on the -- in the procedure.
ROBERT BURNSTINE, MD: What I've done, Amar, is I've taken the muscle off the eye. I have it on some sutures. And now I'm going to reattach it and pull it back right up to where it was before and tie it down. I've done this on about 65 patients now, and all of them have either been better by at least 20% or the same. Nobody's been worse, but almost everybody's been made better by the procedure. So I'm tying it down back to the original insertion site. These are absorbable sutures; they will dissolve by themselves in about three to four weeks.

00:09:04

AMARPREET SINGH, MD: It looks like we have our first question in right now. The question is: is there any medical treatment for nystagmus? Well, the answer, as we sort of alluded to, is there have been certain things that were tried in the past, including some drugs that decreased neural activity, such as Baclofen, however they all have limited success. Even previous surgical techniques that have been tried, such as Kestenbaum Procedure, met with limited improvement as well until -- until this new procedure, the four-muscle horizontal tenotomy, or the Dell'Osso Procedure, was -- was thought up. And we have another question. The question is: my child has albinism, and will this procedure benefit him? Would you like to take that?

00:10:13

ROBERT BURNSTINE, MD: I have done about five patients with albinism, ranging in age from 3 years to about 10 years. They did not -- because their eye is not a normal eye, they did not benefit as much as somebody who has a normal eye who has infantile nystagmus, but all of them have had a definite improvement of their vision, especially in the area of peripheral recognition.

00:10:47

AMARPREET SINGH, MD: So this not only improves the central visual acuity, but also expands their peripheral vision and their peripheral visual acuity, is that correct?

00:10:54

ROBERT BURNSTINE, MD: It does. It actually basically sweetens the sweet spot if you want to -- and it broadens the sweet spot. So instead of looking directly straight ahead, their peripheral vision has been improved by broadening the sweet spot, if you understand what I'm talking about.

00:11:13

AMARPREET SINGH, MD: It's like in tennis, right?

00:11:15

ROBERT BURNSTINE, MD: In -- in tennis, yeah. You have a bigger racquet.

00:11:18

AMARPREET SINGH, MD: A bigger racquet. So right now, it looks like what we've done is we've finished with one of the muscles, it's been reattached onto the globe, and the conjunctiva, or the covering of the -- of the eyeball has been reattached as well with absorbable sutures that Dr. Burnstine mentioned will dissolve on their own. We're now starting on the opposite eye. Are we starting on the opposite eye, or the same?

00:11:50

ROBERT BURNSTINE, MD: We're going to be back on the left eye, on the outside muscle on the left eye.

00:12:00

AMARPREET SINGH, MD: Let's go -- I want you to meet a patient of Dr. Burnstine's named Jordan, who was affected with -- who was affected with congenital nystagmus. This patient had very poor visual acuity of about 20/80 to 20/100, and I want you to see how it affected his life.

00:12:29
JORDAN: It pretty much affected me, like, is -- when I was at, like, basketball games or if I was sitting high up, I really couldn't make out people's faces or, like, the numbers of the players. And -- or when someone was far away from me and my friend said, "hey, look, there's whoever," I really couldn't tell who it was because it was all blurry to me, so I never really could see who it was until we got closer to him. Because all my friends, we just joke around about it. I always kid around with them about it, and they just joke around with me about it. They just, kind of like, always, like, if my eyes just start doing that, they're just like, "whoa, what'd you just do?" or kind of just kidding around with me. It never really bugged me that much until about the time I was ready to get my license, and then I went to driver's training, and they pretty much told me that with how bad my eyesight was, I wasn't going to be able to get it. I knew there was a decent chance going into it that I wouldn't be able to get my permit, but I always kind of just had high hopes that I would be able to get through and be able to get my license and then when they told me that I couldn't get it, I was pretty upset about it because driving's a pretty big thing and I wouldn't be able to do it. I just kind of felt like everything I had tried to do or everything I did kind of didn't matter anymore, like everything just kind of caved in on me. It didn't feel very good at all. It just kind of made me realize that I had something that made me different than everyone else.

AMARPREET SINGH, MD: So this is what Jordan had -- had to say about this, and obviously you see that after he's had this surgery, he plays pool better than I do. But seriously, the -- it's amazing, the impact that it's had on him when he thought that he couldn't do the things that his friends were able to do, and you can see the kind of disappointment that he -- he experienced. Imagine having your -- your freedom or a significant portion of your freedom taken away, which is exactly what we sort of take for granted when we realize that, you know, we drive, and it's such an integral part of our own -- of our own freedom. We have another question that I'm going to answer here, and it says: can this procedure be done on a patient with nystagmus? And the answer to that is yes, they can be -- can be performed on adults. And I'll go to Dr. Burnstine to elaborate a little bit on that.

ROBERT BURNSTINE, MD: I'd like to just back up for just one second and go back to Jordan. Jordan came to me when he was 15. He had applied for his temporary license or -- his driving a car, and unfortunately, he didn't pass that, and that was pretty devastating to him. His vision at that time when I -- when I first saw him about a year ago was best corrected -- that means with glasses and contacts or whatever -- he was about 20/80. He -- I performed the surgery on him, I think, back in April or May of last year, and his vision -- I just spoke to his dad today, and he's driving, he's got an unrestricted license, and his visual acuity is 20/40, so he was -- he's a great story. The answer to your question about adults -- yes -- the oldest adult I've done is age 60. Can you pull this back a little bit? And I -- I usually start doing this as early as age 1 year, so we generally think the earlier we can reset the computer, so to speak, or reboot the computer, the better the results, but I have had good results with adults. Quite a few, actually.

AMARPREET SINGH, MD: Okay. So when we talk about -- we can sort of address who the ideal candidates are for surgery, and what we see is ideal candidates are patients that are between the ages of 1 and 50 who have a diagnosis of congenital nystagmus. The patients should also not have had any previous eye-muscle surgery and have very little abnormal head posturing. Again, if you remember, we said,
sometimes, in order to decrease the amplitude of the shaking, they adapt a -- a posture, maybe, to the left or to the right to decrease the -- the jerking movements. This can still be performed. This actually doesn’t even preclude future surgery in order to address the head posturing at a later date. So here we’ve just started by exposing the opposite muscle, the muscle has been removed from its insertion, and now sutures -- temporary -- or the dissolvable sutures are being placed into the sclera and the muscle will be brought back up to where it was originally taken off from. So, again, an overview of this -- of this particular surgery is that we detach the horizontal muscles from the eyes and reattach them to the original insertion site. Somehow, the proprioceptors are reset and the nystagmus is dampened. Some of the risks of the surgery are -- with any surgery, there's always risks. General anesthesia patients usually have to have general anesthesia, so there's always the risk of general anesthesia. Very rarely, you can get an infection at the surgical site, and there's always a risk that you could rupture the globe, however, that's extremely unlikely in the -- in the hands of a skilled surgeon. The other risk is that the procedure doesn't work, however, again, as we alluded to, almost all the patients have experienced some sort of improvement in their visual acuity. On average, you get a 20% increase in the central visual acuity and a 50% increase in the peripheral visual acuity.

00:19:49
ROBERT BURNSTINE, MD: Could you cut this a little bit shorter? Oh, never mind. I need you to pull this back a little bit.

00:19:59
AMARPREET SINGH, MD: We'll take another question here. The question is: are there any other treatments that someone with albinism can benefit from? Our son's almost at a driving age and really disappointed that he may not be able to drive. He's also worried about his independence and being able to go off to college, seeing as he's a straight-A student. So the question, Boomer, is are there any other treatments -- are there any other treatments that can benefit a patient with -- with albinism?

00:20:33
ROBERT BURNSTINE, MD: Well, certainly the mainstay of somebody with albinism would be sunglasses or sun protection because it -- they get an enormous amount of light. They're almost like dazzled like a flashbulb going off inside -- or going off around them all the time, so sunglasses. I like to use contact lenses, colored contact lenses. I do perform this surgery. I -- we make sure they're in their absolute best prescription as far as for seeing. I have had one child years ago that I did this surgery on that got, like, a telescope to drive, and she's actually driving. She got straight A's in college. So there is definite hope for somebody with albinism, no question about it.

00:21:30
AMARPREET SINGH, MD: Okay. Well, we heard from Jordan just a few minutes ago on how it has -- how nystagmus has affected his life. Now I'd like you to hear from his mother, Lynn, about what she did when she learned that her child was not going to be able to get a license. Can we roll that video, please?

00:21:53
LYNN: I was just going page by page. I had actually Googled then "nystagmus" and it pulls up like, you know, 7,000 different pages, and I thought, "I'll go through every page if I have to," and I started at page one, and after about two -- I think I was on about two and a half hours when I came across one for the Akron Children's Hospital, so I was -- I almost couldn't believe what I was seeing, really, after being told for years, you know, "there's nothing out there." Then when I saw Dr. Burnstine's website, it was just like, "oh, this is so wonderful." I was so excited I could hardly sleep that night. I just think it was the best thing we could've ever
I don't care what it would've cost, I'd have paid 10 times more to have it done, you know. It was so worth it for him, I mean, it basically changed -- changed his life. I mean, I know you can get along in life without a driver's license, but it's so much -- gives you so much more freedom to have that driver's license, so it -- it's made a big impact on his life.

AMARPREET SINGH, MD: It most definitely has made a big impact on not only his life but obviously on his whole family's life. He's able to now do what his friends can do, go on with a very productive life, and live a normal -- a normal social life as well without having to rely on somebody else for -- for transportation. So again, in this particular case for those of you who are just joining us, we are performing a surgery called the Dell'Osso Procedure, which is a four-muscle horizontal tenotomy, which basically means detaching the horizontal muscles from the eye -- two on each eyeball -- and then reattaching them to where they were removed. Somehow this resets the proprioreceptors in the eye and slows the oscillations and the amplitude of the jerking. And we are now -- I believe we're on the fourth -- fourth muscle?

ROBERT BURNSTINE, MD: That's correct.

AMARPREET SINGH, MD: Okay. Right now what we're doing is Dr. Burnstine is taking down the conjunctiva, which is the covering of the eyeball, in order to get exposure to the horizontal eye muscles. While he's isolating the eye muscle, we'll go ahead and take another question here that just came in. It says: if new treatment is discovered years from now, would this procedure preclude or prevent any additional or new surgery? And the wonderful thing is since we are just detaching the muscle and reattaching it to the -- its exact same location and not removing any of the muscle -- this is a muscle-sparing procedure -- it will not preclude any future surgeries. Do you have anything to add to that, Boomer?

ROBERT BURNSTINE, MD: No, that's exactly right. I think that there's no reason in the world why a new procedure wouldn't be able to be applied to these eyes if something develops later on.

AMARPREET SINGH, MD: Let's take a look at this actual procedure right now. If you look over here, this is -- the muscle is being shown underneath the -- the hook, and there are sutures that are being passed through the tendon of the muscle in order to isolate it and keep control once it's removed from its insertion. This is done on both aspects, the superior and inferior aspect of the muscle. There's the -- the dissolvable suture that's being passed through the muscle, and it'll be tied off to prevent any bleeding, to have good control of the muscle itself. About how long would you say this surgery takes, Boomer?

ROBERT BURNSTINE, MD: I would say roughly about an hour.

AMARPREET SINGH, MD: Okay. It takes about an hour under general anesthesia. It's done as an -- on an outpatient basis, which means your child would be going home that same day. And postoperatively, what's your postoperative regimen, Dr. Burnstine?

ROBERT BURNSTINE, MD: They just use some eye drops for about five days. These kids do very, very well.
AMARPREET SINGH, MD: And there isn't a whole lot of pain associated with this, is there?
00:26:56
ROBERT BURNSTINE, MD: There is not. I will -- I do have a moment here to show you -- here's the outer covering right here, it's called the conjunctiva, and there's two layers to the white part of the eye. The outer layer is what you look at, the inner layer -- this layer right here -- is what I look at. This is the muscle, and we've been - - on the other three muscles, we've detached this muscle and I have sutures in here so I can reattach the muscle. So I'm going to be cutting the muscle off the eye right now. I am using an operating microscope, as you can see, so I can see very precisely where I'm cutting and where I'm suturing. There is always some blood when you cut an eye muscle. But we control that readily with cautery. It's an almost painless operation because the only part of the eye that's really sensitive is the cornea, which is the front part up in here, the window, if you will. The crystal on the watch. So as you can see, the muscle has been completely taken away from the eye there, and now we're going to reattach it.
00:28:34
AMARPREET SINGH, MD: So as you can see, when you cut the muscle off, it sort of slips back and that's why the -- the muscle is originally tied with sutures in order to control it. So this is a partial thickness bite through the white part of the eye called the sclera, and it's being passed at the original insertion of the horizontal rectus muscle. Again, this is reattached both inferiorly and superiorly.
00:29:08
ROBERT BURNSTINE, MD: There's a little piece of fuzz on that.
00:29:12
AMARPREET SINGH, MD: And we'll take another question here. This patient is saying: my nystagmus is not just back and forth like the patient you showed, instead it rocks. Can the surgery be done on me? I also had crossed eyes and surgery was done when I was 18 months old. So this is a patient who's had previous eye surgery and has some type of oscillations. I'm not exactly sure what the rocking -- what the rocking means, whether it's a back-and-forth or a up-and-down or a see-saw type, but that's basically the question: can it be done on someone who's had previous eye surgery? I think the answer to that is -- I'd be happy if they'd forward me their records, and I can look at them and get back to them. I would need to know what actually has been done. If -- if the horizontals have already been done, than this procedure probably would not offer them much help here.
00:30:16
AMARPREET SINGH, MD: So anyone that is interested in actually making an appointment or getting or -- or having a referral, you can use the appropriate buttons on your web browser as well to send us an e-mail. If we -- if we don't get to all of your e-mails right now, please be assured that we will respond back with -- with an e-mail promptly, as well. So as we were just saying, Dr. Burnstine has just reattached the eye muscles and now is going ahead and closing up the conjunctiva.
00:30:57
ROBERT BURNSTINE, MD: Well, I've got one more end of the muscle to reattach, and then I'll be doing that.
00:31:01
AMARPREET SINGH, MD: Okay, can we go back to that -- the close-up here. So as you see, when he pulls on the suture, you'll see the muscle coming back up to its original insertion where it had started out. Now, is there any difference if you have it done earlier or later? Meaning like, let's say you're 50 -- the 60-year-old patients that you've done or the 50-year-olds, is there a difference in their outcomes compared to the 5-year-olds or the 2-year-olds?
ROBERT BURNSTINE, MD: I think the answer is probably yes. In medicine, it's always better to get treated early than late, but obviously we can't turn back the hands of time, so, you know, the best time to have had this surgery is when you're 1, the second best time is today. And now we're closing the muscle -- or the conjunctiva back over the muscle.

AMARPREET SINGH, MD: Okay. We have another question here that says: does the time interval between muscle disconnection and reconnection have any influence on the final result? Meaning if it's detached for, let's say, five minutes as opposed to, you know, 30 minutes or --

ROBERT BURNSTINE, MD: I'm not aware of that.

AMARPREET SINGH, MD: Okay. So here, this is the last step of the procedure where Dr. Burnstine is just reapproximating the conjunctiva to where it originally came from with dissolvable sutures. We have another question here. Somebody is asking: what is the cost and is this covered by insurance, or is it considered cosmetic?

ROBERT BURNSTINE, MD: It is covered by insurance. The cost of the procedure, including the hospital and anesthesia, probably, depending on where you are, $4,000 to $5,000.

AMARPREET SINGH, MD: But again, that's going to be covered by the insurance, and the families usually see a very, very small amount of that.

ROBERT BURNSTINE, MD: Hopefully.

AMARPREET SINGH, MD: So this procedure is just about at its conclusion, and it's really taken much less than an hour to perform this.

ROBERT BURNSTINE, MD: Okay. So I just want to show you what the eye -- completed eyes -- let's take these -- these off, and we can move the microscope out of the way. And I don't know if you can get a close-up of these eyes, but I think you can see, they don't look too bad considering he's had four eye muscle operations -- four muscles done.

AMARPREET SINGH, MD: Yeah, there sure isn't -- there's not a lot of blood there, it doesn't look irritated too much, it's --

ROBERT BURNSTINE, MD: And we're going to put some drops in now, and I'll give these to the family to use for the next five days. You can keep your e-mail questions coming in and we'll be happy to answer them, and thank you all for joining us today.

AMARPREET SINGH, MD: We have another question here as well. It says: what are -- what causes nystagmus? What are some of the causes of nystagmus? What are things that we look for when a patient comes in to see us? Well, some of the things that we look at, we want to make sure we exclude things like cataracts or glaucoma, things that are -- that are related to the optic nerve, like atrophy, any macular diseases or retinal causes as well. We also have to make sure the patient doesn't have albinism. And in certain types of oscillations, we have to preclude any tumors. So our patients usually get an MRI as well just to make sure that we're not missing -- missing something in the 0.1% chance that there's actually a mass. And we have
another question here that says: does it run in families? The answer is yes, there are actually both recessive and dominant forms of congenital nystagmus that have been found with mappings to the different -- to various chromosomes, both the sex chromosomes and other -- other chromosomes, like chromosome 6.

ROBERT BURNSTINE, MD: Might I -- I have -- I have one family that's got five brothers and sisters with nystagmus. The parents did not. I just tell people, somebody has to start a problem. The mother could be a carrier, the father could be a carrier, they produce children with it, and they have nystagmus.

AMARPREET SINGH, MD: Right. Well, thank you for letting us join you today in this surgery. It was very --

ROBERT BURNSTINE, MD: My pleasure.

AMARPREET SINGH, MD: And thank you all for joining us as well. If you have any further questions, please keep the questions coming. We'll be happy to answer them if not live here, then back via e-mail. Thank you very much and have a great afternoon.

ANNOUNCER: This has been treatment for congenital nystagmus performed live from Akron Children's Hospital. To obtain more information, to make an appointment, or make a referral, please click the appropriate buttons on your screen.