2 February 2024 / A new treatment for deafness

[HALF SECOND OF SILENCE]

[BILLBOARD]

SEAN: [00:00:00] Something truly remarkable happened in Philadelphia recently.

*<<CLIP>> NFL COMMENTATOR: [00:00:04] Here's a sack by this defense!*

SEAN: [00:00:07] Not the collapse of the Philadelphia Eagles, though. Ouch.

*<<AMBIENT SOUNDS FROM CHILDREN'S HOSPITAL OF PHILADELPHIA>>*

SEAN: [00:00:12] An 11 year old Moroccan boy traveled to Philly via Spain for a procedure.

*<<TAPE>> CHILDREN'S HOSPITAL OF PHILADELPHIA:*

*DOCTOR: [00:00:19] You know, I feel weird, but.*

SEAN: [00:00:22] Aissam Dam was born deaf. Totally deaf. But he went to the Children's Hospital of Philadelphia for a clinical trial.

*<<TAPE>> CHILDREN'S HOSPITAL OF PHILADELPHIA:*

*DOCTOR: [00:00:31] This will be where the gene therapy drug will go into that tubing there.*

SEAN: [00:00:35] A new gene therapy offered something miraculous. It allowed Aissam to hear for the very first time.

*<<TAPE>> CHILDREN'S HOSPITAL OF PHILADELPHIA:*

*DOCTOR: [00:00:44] I asked if ‘you heard that,’ and he told me yes. Pretty cool!*

SEAN: [00:00:49] Today, Explained, we're going to hear all about it, but we're also going to ask whether deafness is something that needs to be treated in the first place.

[THEME]

SEAN: [00:00:03] Antonio Regalado is a biotech reporter at MIT Technology Review. Not so long ago, he was contacted by some scientists in China who said they could make deaf kids hear for the first time, and he had to know more.

ANTONIO REGALADO (MIT Technology Review reporter): [00:00:20] Well, they had pursued a gene therapy to reverse deafness, congenital or inborn deafness in children. And so they had been working on a gene therapy that would treat basically one type of inherited deafness, just one type. But it's really cool, and, what was cool about the story is we got to interview the mother of a child who had the treatment and heard about the amazing results. A girl who couldn't hear, and then within a month of getting this gene therapy into her ear that was injected, she could hear out of the treated ear.

SCORING IN Cloud Seven (Flute Version) BMC

ANTONIO: [00:00:56] I can tell you the story about a little girl named YiYi, who lives in China, and who is one of the kids that got this treatment. She's really cute. Her mother, Qin, makes lots of videos about her and puts them on the internet. And they're really remarkable videos that demonstrate that this girl who was born deaf can now hear her mother.

*<<CLIP>> QIN LIXUE: [00:01:18] <<speaking Chinese>> Is my voice too loud?*

*YIYI: <<speaking Chinese>> Yes, loud.*

*LIXUE: Loud?*

*YIYI: Medium loud*

*LIXUE: Medium.*

*YIYI: Yeah.*

*LIXUE: What day is it today?*

*YIYI: Hmmmm… Saturday!*

ANTONIO: [00:01:23] A couple of years ago, YiYi did get a cochlear implant in one ear. So that's a device that helps her hear out of that ear. But that kind of hearing is not that natural. It has a kind of a metallic sound. You don't hear details like the wind through the trees.

*<<CLIP>> COCHLEAR IMPLANT RESEARCH LABORATORY:*

*Woman’s voice as heard through implant: It was a full moon, three nights ago*

ANTONIO: So the family was glad to try gene therapy in YiYi’s other ear, and that worked.

*<<CLIP>> QUIN LIXUE: [00:01:49] <<speaking Chinese>> Was it fun when you were in Shanghai?*

*YIYI: <<speaking Chinese>> It was fun*

*LIXUE: What did you do there?*

*YIYI: I did… hmmm… the squid!*

*LIXUE: It’s called “eat the squid”*

*YIYI: I ate the squid!!*

SCORING OUT

SEAN: [00:01:58] How many people have received this kind of gene therapy where they may have been deaf or hard of hearing and can now hear?

ANTONIO: [00:02:09] Well, this is extremely new. The first cases we heard about were from China this fall, and at that time, that group had treated about ten children, and they were ready to report on the results in five. And then just recently, just a couple of weeks ago, we heard about another case in the U.S. with similar results.

*<<CLIP>> CBS NEWS: [00:02:27] An 11 year old boy received the experimental treatment back in October. The boy who was born deaf in both ears now only has mild to moderate hearing loss in the ear that was treated.*

ANTONIO: [00:02:39] That we know of, six children so far treated in this way for deafness in the world. Right. All these studies, they've just started to launch recently, within the last year. And it happens that there's several groups doing it. So they're all in a bit of a race, to be first and to demonstrate these incredible results. There's at least three teams doing this particular kind of gene therapy.

*<CLIP> REGENMEDGLOBAL: [00:02:39]*

*Dr. Zheng-Yi Chen, Harvard Medical School: We’ve been working in the field long enough to know, we have come a long way, now is really the tipping point, so are going to have some spectacular success as a field… and also is incentive for us to work even harder.*

SEAN: [00:03:20] And what is this experience like for, for a child to go from not hearing to all of a sudden having this fifth sense?

ANTONIO: [00:03:30] So I really wanted to know what her experience was. Now, this girl’s only 5 or 6, so it's hard to get, you know, even her mother, has trouble getting her to express to her clearly exactly what it's like. But, you know, there are a few details that were compelling. For instance, they live in this apartment block, in a noisy area of town, and the little girl has started to complain that she hears sound at night. right? It's too noisy at night, she says, which she wouldn't have said before because she wouldn't have heard the noise.

SEAN: [00:04:01] Never move to Brooklyn, Yiyi.

ANTONIO: [00:04:03] Another story was, you know, when she went to school, in her little grammar school, they would have a nap time. And when the nap time was over, a bell would ring, you know, a little bell and say, nap time's over, kids. And before, Yiyi would, you know, her friends had to tell her that the bell had rung, but now she can hear it.

SEAN: [00:04:21] Is her hearing at 100%, 50%? 25%? What?

ANTONIO: [00:04:25] Well, of course, the doctors and scientists involved are measuring her hearing. And the way they expressed it to me was that her hearing is, is it about 60 to 65%. So it's close to normal speech. It's like having moderate hearing loss. Now, it can differ at, you know, the frequency, you know, high frequency or low frequency noises. But the amazing thing is that, you know, she would not have heard anything sitting in the movie theater with the loudest sound possible before.

*<<CLIP>> Lucasfilms THX intro*

ANTONIO: And now she can hear, you know, essentially a normal voice, but you might not hear a whisper.

*<<CLIP>> Nokia phone plays careless whisper ringtone*

SEAN: [00:05:11] Will her hearing continue to improve? Or is this it? How does it how does it work?

ANTONIO: [00:05:17] I think that's one of the unknowns. Will it continue to improve?

SEAN: Hm.

ANTONIO: Maybe not. But it's an incredible success that she can hear, you know, natural sounds already. That's a big success. I don't think it's going to improve, uh, that much further, but that's my guess.

SEAN: [00:05:35] Can you tell us what this gene therapy is doing, Antonio?

ANTONIO: [00:05:39] One of the cool things about writing this story is you learn about the ear. It's not, we don't, I don't really think about my, what's going on in my ear that much, but it's really amazingly cool little organ.

*SCORING IN – Little Sparklers*

ANTONIO: Inside of it are these hair cells. They're not actually hairs. They just kind of look like hairs because they have these little extensions, and there's about 16,000 of these cells,

*<<scratching sounds>>*

and each one is sort of tuned to a different frequency. So the sounds that come into your ear, they vibrate these little hair-like structures…

*<<vibration sounds>>*

ANTONIO: …they vibrate them, and that information is turned into a chemical signal and then an electrical signal and goes to your brain…

*<<phone dialing, electronic sounds>>*

ANTONIO: …you know, where you where you experience the sensation of hearing.

*<<man’s voice says ‘Hi, Hello!>>*

ANTONIO: But it’s actually, because of these little wiggly antennas, that you can hear at all. People have a mutation in their gene that makes a crucial part of that system. It's a it's a molecule that is basically the the chemical transmitter of the signal.

*<<modem booting up>>*

ANTONIO: And so in this form of hearing loss, the problem is that people just don't have this chemical.

*<<photocopier making copies>>*

ANTONIO: And the gene therapy basically puts new DNA for this gene into people's ear cells. And so, with that copy of the gene, the cell can then make this molecule.

SCORING OUT

SEAN: [00:06:58] And when people hear gene therapy, I think they think Crispr. But this isn't Crispr.

ANTONIO: [00:07:04] This is not Crispr. This is yeah, you can think of gene therapy as adding a new gene, a whole copy of a gene. And in fact, this gene is pretty big. Whereas when people talk about Crispr, that's gene editing. That's sort of that's putting in some molecules that could change a gene, often break a gene, which could also be useful. In this case, you have to add the gene back. And you really couldn't even do that with Crispr at this point. So this is more classic gene therapy.

SEAN: [00:07:31] And would this classic gene therapy be able to help any deaf person, or is it better suited to kids?

ANTONIO: [00:07:39] This gene therapy would be suitable for only a small fraction of the people who, of people who are born deaf, which because only only 1 to 3% of those people, nobody's quite sure because it's rare enough have this particular genetic mutation. Like there's lots of different genetic mutations that cause deafness, and some of them are much more difficult to deal with. This one happens to be the sort of most, most straightforward, the easiest one to deal with. And this would account for about 1 to 3 cases of inborn deafness or congenital deafness, not counting people who go deaf for other reasons.

SEAN: [00:08:15] But you did mention that there's more trials going on. There's there's at least one in the United States as well. Does that bode well for the sort of spread of this treatment, the more wide application of it?

ANTONIO: [00:08:29] I think what, what this does is it's going to inspire and excite scientists who are working on kind of harder gene therapies for other forms of deafness. Right? There's going to be more money, more excitement, more interest, now that they've done this. I should say that a lot of gene therapies, as miraculous as they are, they fail in the marketplace. And the reason they fail is that there's too few patients. These are for ultra rare diseases. So it's just, you can imagine, you have a company, a biotech company of 50 people, and there's only 50 patients in the world. So that's … it doesn't make economic sense. So a lot of these gene therapies, as amazing as they are, they undergo a form of market failure. They, they, they actually can't be sold. Some of them are retired and, and patients can't get them. So with this one for hearing, we'll have to see. There's still there's just not that many people can benefit.

SEAN: [00:09:28] It's interesting. So there's a bunch of competitors entering this market which you say doesn't have, you know, super broad potential. Why are there so many competitors trying to get in on a small market?

ANTONIO: [00:09:43] A lot of people think gene therapy is the future. And it may be. I mean, it is dramatic in its essence, right? I mean, you are changing or adding DNA to human cells. I mean, that is kind of profound because DNA is the molecule of all life. And now scientists can change it and add to it and engineer it. And they can treat all kinds of diseases that they couldn't treat before. So it's just exciting. And the technology makes it possible. And so that animates biotech companies. But for the scientists involved, it's just super exciting, not only from a technological point of view, but also, you know, from a medical point of view. Many of them are doctors, you know. To, to be able to cure someone, to be able to help someone, in a disease that has never before…like there is no treatment for deafness using pharmaceuticals or pills. There is a cochlear implant that you can get, but there's no pill that you can swallow that will help restore hearing. None.

SCORING IN River Keys (BMC)

ANTONIO: If you're a doctor, that is something profound to help someone that was beyond help. So that is another reason to do it.

SCORING BUMP

SEAN: [00:11:03] You can read Antonio Regalado at technology review dot com.

Just because you can do it, should ya? We’re going to ask a philosopher, who happens to know a thing or two about being deaf, when we’re back on Today, Explained.

[BREAK]

[BUMPER] [00:00:02]

PROFESSOR TERESA BLANKMEYER BURKE (Gallaudet University): [00:00:06] Hello my name is Teresa Blankmeyer Burke and I am full professor of philosophy at Gallaudet University. Gallaudet University is a bilingual institution with American Sign Language and English language in use for instruction, and we also have deaf and hard of hearing and hearing students.

SEAN: [00:00:27] Professor Blankmeyer Burke is Deaf herself. She signed this interview and her responses were voiced by an American Sign Language Interpreter.

PROFESSOR BLANKMEYER BURKE: [00:00:36] My work in gene therapy has the potential to impact ethics and certainly the Deaf community themselves… and my work, my research for the last 20 years has involved consultancy in any number of different groups, such as the United Nations, the World Federation of the Deaf, the National Association of the Deaf of this country, the NAD, and others. So this is a topic that is my jam.

SEAN: [00:01:04] <chortles> Well, I'm so glad because I think when people read the news that science has advanced to the point that gene therapies can restore hearing for children who have never fully or even partially been able to hear anything, they think it's nothing short of a miracle. But I want to ask how you, someone for whom this is their jam, someone who thinks about these questions on a philosophical level, react to such news.

PROFESSOR BLANKMEYER BURKE: [00:01:39] There are two stories that were reported in the New York Times last week about the 11 year old boy from Morocco. There were also six children in China. One thing which we have seen from the reports, as well as a review of the study, is that these seven children, taken together, who had gene therapy, are all still hard of hearing or deaf. And these children need support and access to a language. As an ethicist, I think that it is hugely important for those children. Yes. Gene therapy is taking place. From the perspective of the public, it’s seen as a panacea, but that is not anywhere near the case. So I think it's the nature and expectation of people who don't have the lived experience within the Deaf community to think,<<gasps>> it is miraculous. And I would also add that I am not against gene therapy in and of itself. I think it's important for us to consider all of the potential impacts of this technology.

SCORING IN Cloud Seven (Flute Version) BMC

PROFESSOR BLANKMEYER BURKE: One thing that's part of the history in the signing Deaf community, is the experience of miracle cures that are imposed upon us, and a history of people who see a development – a recent example might be cochlear implants – and how that can impact the lives of children. These developments are of a concern in the Deaf community, because the priority generally of deaf people and for a deaf child, would be to say you're okay and that you do have a language, you have access to your language. Sometimes what happens with medical interventions and technologies is that you would have an issue of, say, treating a child with whatever that intervention might be, but there's very little attention given to what might happen to that child if that intervention or therapy doesn't work. We see that within the Deaf community, certainly, it is a source of concern because children need to have full access to language from the get go, of course, for the purposes of learning, and in these scenarios, they don't. And they experience what is known as language deprivation. And language deprivation is a term of art that we see in the literature surrounding the Deaf community. And it is the notion that people who don't have access to language during the critical window of development or language acquisition, will never really gain full access to a language. They might have the ability to communicate gesturally in some way, but as far as their critical thinking ability, their interpersonal ability, there are permanent deficits.

SCORING OUT

SEAN: [00:04:35] So if I'm understanding you correctly, for you, it's less of a miracle. It's less of a end of a story and more of a midpoint where, okay, yes, these children have had some hearing restored, but how are they communicating? Are they still learning language? And your, your chief concern here is sign language.

PROFESSOR BLANKMEYER BURKE: [00:05:07] For me, I think it's the most important thing that the deaf children get a language, full stop. It's certainly nice for deaf children to have both, to be sure, access to a signed language. I can use myself as an example. Having grown up in a mainstreamed environment educationally, learning to speech read use what hearing I had, and figuring out how to navigate the hearing world. When I came to the sign language community, for the first time in my life, I noted that I had full access to a language and I was no longer playing guessing games. Of course, now when you're learning a new language, you are playing guessing games. But that all evened out. So the experience was actually understanding and understanding the feeling of not having to struggle to understand. It was just there. So part of my question remains for those children, maybe they will acquire spoken language, which is all well and good, but for other children, that might not happen. And we have to consider what would happen for every and all deaf children in this environment of gene therapy. Whatever happens, I'm more concerned about shining a light of ethics on people who are deaf, and my overriding concern about the decisions made about whether to administer gene therapy or not. I think that one thing that really warrants close attention is the word “serious conditions.” This includes things such as sickle cell anemia, cystic fibrosis, Tay-Sachs, and other things. These conditions cause a tremendous amount of pain and suffering or could result in premature death. And this is the cohort that is always within the discussion of prioritization to receive gene therapies.

SEAN: [00:07:12] Right. I think you're leaning towards something I want to ask you about, which is the simplest way to think about this treatment is … is as one that is curing deafness. And I imagine you don't think deafness is a thing that needs to be cured.

PROFESSOR BLANKMEYER BURKE: I think for people whose experience is within the hearing world and they don't have an experience in assigning deaf context in that community, I think that it would be a matter of course, that they would think they'd like to see this particular, sensory disability cured. At the same time, I think that it is important to note that we've had conversations about difference. I see the signing Deaf community as a thriving cultural linguistic community that carries their own ethical norms, their own cultural norms, their own cultural contributions to art, to history, to any number of domains, the sciences. This community is part of our human diversity.

SCORING IN Gon(e)athan, The Reprise

PROFESSOR BLANKMEYER BURKE: [00:07:32] I am always concerned when I see a challenge, or an attempt, to dilute human diversity. I would think, yes, that people think it's easier, people who are hearing, would think that it is easier to not be deaf. But within the community, I know many people who are deaf, who have intergenerational deaf families, and they have purchase in a part of community with deep roots and a deep reverence for that experience and that community. I recognize that, of course, there are tensions between parental right to make a decision for their children and a community who wants to preserve itself. And the parents' decisions ultimately impact the ability of that community to maintain itself and indeed, thrive, going forward. So where we are now is, well, I've been thinking about this problem for 20 years and I still don't have an easy answer.

SCORING BUMP

PROFESSOR BLANKMEYER BURKE: Yes, we need to support parents in making decisions for their children, however, at the same time, I think it's important that parents are fully informed before they make such a decision. And it's important for people to understand the value of a signing Deaf community and what that brings to the world in terms of human diversity.

SCORING BUMP

SEAN: [00:10:04] Professor Teresa Blankmeyer Burke. Gallaudet University. Washington, District of Columbia.

Our program today was produced by D.C.’s Victoria Chamberlin. We were edited by Matthew Collette, mixed by Patrick Boyd, fact checked by Haleema Shah and Avishay Artsy. Welcome back, Avishay!

The rest of our team includes Amina Al-Sadi, Hady Mawajdeh, Amanda Lewellyn, Miles Bryan, Laura Bullard, Jesse Alejandro Cottrell.

We use music by Breakmaster Cylinder.

Our executive producer is Miranda Kennedy.

My co-host is Noel King.

I’m Sean Rameswaram.

<< For podcast:>> Today, Explained is distributed by WNYC. This show is a part of Vox, which is totally free thanks in part to contributions from our listeners. Join us at vox.com/give. >>

<< must cut this out for radio >>

[10 SECONDS OF SILENCE]