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Patient-Centered Research in NF2: Improving Quality of Life**
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Good afternoon, ladies and gentlemen. We're going to begin with our next presenter, Amanda Bergner; was kind enough to come all the way from Johns Hopkins. We're delighted to have her. She wears many hats, apart from being the senior counselor at the NF clinic there, currently the director of operations for the NTAP program. Amongst other things, she will be reporting on the longitudinal study on the quality of life. This is a topic we've never addressed here at our conference. So this should be very exciting, please welcome Amanda Bergner. (Applause)

AMANDA BERGNER: Thank you. So especially with the smaller group I'd rather be more informal. If people have questions as we go, go ahead and ask them along the way. I think we have about an hour to go through talk and questions. So just shoot up a hand if anyone has questions as we go.

I'm talking this afternoon about a couple of different things that all fall under the umbrella of patient-centered research in NF2. I like this schematic. It helps us think a little bit about a reorientation in research and medicine that's ongoing. If we think about back 50 years, I'm not quite sure this is how people would think about research, where the patient is actually in the center with the provider, the clinicians, researchers are around the outside, all focusing inward on the patient and the patient's needs. In many ways that's the way research has always been. In other ways we've taken great strides recently to really bringing this to the forefront and making it a priority.

So I want to start off talking a little bit about what quality of life is. I imagine all of us, if I say what's your quality of life, something's going to come to mind. But as we begin really talking about it, I want to think a little bit about being sure that we're thinking often as expansively as we need to about quality of life. So sometimes we think, oh, yeah I feel pretty good today. But is that really encompassing everything that could fall under quality of life?

This slide gives us a concept of many of the aspects that go into quality of life. If you think of self in the center, often as we think of this type of research we think of mental health, physical health and spiritual/emotional health. Around the outside, recreation, family, vocation, social, community and finances. We all have needs in these realms, how we meet those needs can look different from person to person.

But we all have these needs.

And so when we think about quality of life measurement, how we measure quality of life, often we're looking at what have become standard indicators. And some of them are listed here if we think what's the quality of life of people with NF2, we could look at how much income people with NF2 have over their adult life. We could look at how much education people with NF2 obtain. We could study how much people with NF2 feel like they belong to various social groups.

There are a lot of different ways to look at quality of life. And recently researchers have begun

distinguishing between two aspects of well being. Emotional being and emotional experiences can change from day to day. And then overall life evaluation. How is your life in general as compared to a particular scale?

So thinking just about the emotional aspect and thinking about the entire aspect of life. So if we take another step closer to what we're working on and talking about here, quality of life in healthcare, it becomes even more specific and there's another new acronym, health related quality of life. Not necessarily income and education, but health aspects of quality of life. Here's a sample quality of life question... how have things been going for you during the past four weeks?

“Very bad, could hardly be worse. Pretty bad, good and bad parts equal. Pretty good, very well, could hardly be better.”

It's not asking how have things been go over the past four weeks with your NF2 or how has your health been over the last four weeks, it's in general how has life been going. So as we get more specific, the questions that are related to quality of life and healthcare actually will get more and more specific to health.

So health related quality of life can be measured in many ways. I broke it down for us today into two ways to think about this. The first is quantitative research; things that can be measured or calculated, scores that can be added together, anything that's going to produce a number of sorts on a scale. And so this often comes in the form of questionnaires. These are generic questionnaires, some of you may have heard of some of these. There are disease-specific questionnaires. And this is an example of a NF2-specific questionnaire that's related to health and quality of life. These quantitative methods of study, they are often what's called multi dimensional, looking at a lot of different aspects of life that are specific to health. So maybe therapy induced side effects or maybe the financial impact of a medical conditions.

The other big category of quality of life measurement is qualitative. Versus quantitative, we're trying to understand why and how someone's making a decision. Not just when or what or where.

This tends to be more one on one interviewing. Maybe focus group interviewing. And we're trying to look for general themes that when we get a group of people with NF2 together and say, "What are the things that are the most difficult for you because of your NF2?", people talk more openly and start to see that many people say the same types of things over and over. That would be a result from a qualitative research study. Those are the two big types that we'll talk about today.

Have people been involved in this type of research for NF2?

No?

I know some people -- okay.

So I always like this question why. What difference does this make?

AUDIENCE: Know who to contact to get into these studies.

AMANDA BERGNER: Yeah. We're going to actually talk about that. So that's a good question.

So why is this even important?

And a lot of these reasons are starting to come more to the forefront. We understand this relationship between cost and values. We're going to spend a lot of money, we want to spend it in the way that makes the most sense. And how are we going to decide what makes the most sense?

Is that what makes the most sense to us as clinicians, or to colleagues that are researchers, or to families and patients?

Who's deciding what makes sense to spend money on? If we can only spend money on certain things, how do we prioritize? Often quality of life and assessment of quality of life from families is really critical as we think about where to spend money and time and effort.

It's also becoming more and more important for regulatory agencies like the FDA in the United States and the EMA in Europe as they begin making decisions about what types of drugs to approve. So again, even if a drug doesn't necessarily impact the size of tumor, but pain gets reduced and the patients are reporting they have less pain in a questionnaire and we can show that over time, it may be that that's enough of a reason for the FDA to say, okay, while the tumor is the same size, but patients are feeling better, we approve this drug to be used for pain control.

So we're starting to understand the impact it can have on the clinical trial process as well. This was a 2010 survey that was done in a specific pediatric community asking how patient-centered is today's funded research. And what we saw is that 57 percent of the studies at this point included patient outcomes, which is great. A little over half are asking how patients are actually doing, rather than just measuring something on an MRI.

About 18 percent were meeting actual patient needs. So that's a big drop-off. So about a fifth of studies were -- if you asked the patients what they needed improved in their lives, the studies were addressing those specific needs. And actually what this study showed it was that none of the research studies that were surveyed at this point indicated that patients were involved in the research proposal.

The research -- the generation of research questions were otherwise involved in the conduct of the study except the research subjects.

So when we're designing research, what this is saying is we're not asking patients and families what should be studied, how do you think we should study it, what difference will it make to you?

And as we go doing the research, not involving people, at least at this point -- haven't been involving people in how the research is conducted. What we've actually seen is that -- this was a huge problem, it's beginning to change.

So there was an aspect of the Affordable Care Act that was passed in 2010 that awarded primary stakeholders, which is patients and families, a direct and meaningful role in setting research priorities,

overseeing the research program and communicating study results. And this Affordable Care Act funded the Patient-Centered Outcomes Research Institute (PCORI).

We've been in a little bit of a different situation in NF2 only because for the last 10 or 15 years, patient have been involved to some extent with the DOD clinical trials process. And so there have been some patients and families that have served on the review committees in the last few years to take a look at some of the research proposals that have come in which is providing a voice to patients and families at the level of funding which is great.

There are organizations such as yours, NF Midwest, Children's Tumor Foundation where patients and families are providing funding that's been given to researchers and you can have a little bit of a say about how that money is spent. Overall we needed to make a really big change.

So I want to shift a little bit to talking about specifics around NF2. At this point there's one scale that has been developed to be specific to NF2. Called the "nifty" quality of life, or the NFTI-QOL. Designed by the NF group at Guy's Hospital in London. It's available in U.K. English and U.S. English right now. And it actually consists of eight questions. It's meant to be a very brief screening to help establish whether or not people have concerns related to NF2 that's impacting their health. Their mental health and physical health. And can be used in a clinical setting to try to identify specific concerns people with NF2 might have. Not terribly comprehensive. A more comprehensive NF2 instrument is being developed. Dr. Maura Cossetti, developed a quality of life tool in NF2. That's not available yet.

AUDIENCE: What is Dr. Cosetti's background?

AMANDA BERGNER: That's a good question, she's an otolaryngologist. An ENT doctor.

So often when you're developing quality of life instruments specific to a condition, you'll start off with the qualitative research. Pull together small groups of patients and families and ask them what's of most interest to them, what's concerning, troubling. And develop questions, and then go back to the group with questions and ask does this question accurately reflect what you were expressing?

And then once the questionnaire is put together, it goes out to the field. And you know, some 200-300 people need to fill it out and give it back. And then the researchers will take a look at the answers and figure out if the questionnaire can really function as a questionnaire.

So it takes several years to develop a really good quality of life instrument.

Again, they can be used in clinical care. So often they're used as screening tools to offer helpful intervention. If you answer a handful of the same questions every time you come in, your doctor can start to see, well, usually this person answers a 5. Everything's really good on all of these questions. Now this time they answered a 3. So we need to spend a little more time on this aspect during the visit today because this looks like it's changed for this person.

You can also track progression of symptoms over a stretch of time using the same questionnaire.

It also can be used in clinical trials. So there's an initiative that's been put together by Dr. Scott Plotkin and Brigitte Widemann... the REiNS Collaboration--Response Endpoints in Neurofibromatosis and Schwannomatosis, and there's a committee on patient reported outcomes. This group is looking at all the available patient reported outcomes for quality of life and figuring out which ones are best to use in clinical trials for NF2. We actually just evaluated the NFTI-QOL tool last month. I think people thought it was helpful in many ways, but left a lot to be desired because it really didn't address a lot of the symptoms that people with NF2 ultimately experience.

So really the goal is to identify a pool of measures that already exist that are appropriate to use in clinical trials. We are also finding gaps and things that need to be developed that we don't have right now.

I wanted to shift from here to talking some about some of the specific work that I've been involved in. I'm sure you all have heard of Avastin. This has been thought to be effective for some people against their vestibular schwannomas. This was first out in the world as an observational paper by Dr. Scott Plotkin at Mass General. He had given this drug to a handful of patients and seen improvement in hearing in some of them, not all of them. In order to show whether or not that was just a coincidence or it actually was something that could be replicated, we needed to run a clinical trial. So between Hopkins, Mass General and NIH we ran a trial where we gave 14 patients Avastin for one year and asked these questions primarily... Does their hearing get better, and do their tumors shrink? Just the vestibular tumors. The only ones we were looking at.

And so as a part of this study we also looked at quality of life. We measured quality of life at baseline, which means at the beginning when people came onto the study for all 14 of the people enrolled. And then we measured it three more times. Halfway through getting the drug, all the way through the time that you get the drug, and then we took people off the drug for six months, and then we studied again quality of life when they weren't on the drug.

The questions here are just -- does taking this medication improve quality of life? Do people with improved hearing also experience improved quality of life? Does improved quality of life come along with tumor reduction? And what happens when people stop taking this drug? These were the main questions we were looking at.

We actually -- used several actual tools. The first one is the SF-36, probably the most widely used in the world to study quality of life of any other tool. Its weakness is that it's not specific to NF2, and its strength is that this allows us to compare NF2 with a lot of other medical conditions. And so it assesses a lot of different domains. And here are two example questions. In general, would you say your health is: Excellent, very good, good, fair, poor. And compare today one year ago, how would you rate your health in general now?

So you can see that these are not specific to NF2, they're overall general health-related questions.

So people took this survey four times while they were on the study. We also used the speech and spatial qualities of hearing measurement, which is designed to measure auditory disability really by asking people about very specific everyday circumstances they might find themselves in.

Here are some of the questions.

So if we look at No. 1, you're talking with one other person and there is a TV on in the same room. Without turning the TV down, can you follow what the person you're talking to says?

And people can score anywhere along this scale from not at all to perfectly. If it doesn't apply, they can check not applicable. Everyone in our study answered somewhere along this line.

And it goes on -- I think there are about 35 or 40 questions that are asked that provide very specific scenarios. And so people answer this survey four times, at four different time points throughout the study.

And the third quantitative measure we used was the tinnitus reaction questionnaire. This is designed to find out what sort of effect tinnitus has on lifestyle and well-being. We know it's a real problem for people with NF2.

So here are some example questions. My tinnitus has made me irritable. So you can say, not at all, all the way up to all of the time.

And there are 20 questions on this scale. My tinnitus has made me unhappy or angry.

And so people filled this out four times throughout the study. We're incredibly appreciative for people being patient and filling it out and giving us some feedback.

The other aspect of the quality of life arm that we did on this study was qualitative. So I did interviews with everyone when they first started on the study and asked three questions.

Why did you choose to participate?

Do you think you'll have a positive outcome?

And how would you describe the impact NF2 has had on your life so far?

Now, the interviewing is a little different. So it can be more conversational. I asked more questions than this. But ultimately everything stemmed out of one of these three questions.

And a lot of what we were wondering was really, for instance, if people think they're going to have a positive outcome, and then after three or four months taking the drug, they don't have any improvement in their hearing, is their quality of life score going to go down after that?

Because they wanted a positive outcome and they're not seeing it. So that's one of the things I was interested in.

I also did a study with everyone as they left -- an interview as they left the study. Asking them if they felt they had an improvement in their symptoms, did they have any side effects, did they ever consider stopping the drug because of the side effects?

Did they ever consider stopping the drug for other reason?. And one of the questions I found really fascinating in the response is now that you've been through this and you've done all of this and you've come every three weeks and had an infusion and all these MRIs, would you do this again? Would you put yourself through this again?

Because at the time this study launched, it was the first clinical trial in NF2 with a drug that was being conducted. And so it is of interest to see really what's motivating people to join these studies. How much patients' intolerance are people going to have with various studies as they're designed?

We need to think about making it reasonable for people who are going to participate. And what impact did participating in the trial have on you in terms of your resources, time, energy, money?

So what I really was hoping actually when Diana and I were first talking four or five months ago is that we would have analysis done enough that I could give you some general ideas of what we're seeing. But we're still working through that. One of the things I did want to talk about that has come up that I found really interesting was, whether or not people had hearing response, almost uniformly people were mentioning in this end of study interview the relationship that they had formed with the study team and how important that was to them. And how it was going to be not to see these people that they had been seeing every three weeks anymore? And coming off of a clinical trial you're not going to see this group that you had really come into relationship with and had been taking care of you.

And it spoke to me -- I'm still really working on this aspect of how to present this -- but it speaks to me about relationship building and how in the setting of NF2 the hearing loss we can often see an impact in communication and ability to form and maintain social relationships. And so by joining a clinical trial, it's sort of a situated relationship that you're a part of. And does this speak to the fact that people, you know, with NF2, as they go on in life with more and more hearing loss and more and more difficulty with relationship, are they really thirsting for that? Is that a need in the community?

That's one of the interesting things that came out that I wasn't expecting and just wanted to share that with you. We're hoping that, again, as -- I think it was Andrea this morning who was saying that publishing studies is really critical as a way to get information out. We're hoping these papers will be out by the near future.

This leads me to one of the other studies that is a little more recent. And with one of these ideas that I sort of put together I would be really interested to have feedback in the community about this. But I've witnessed this in clinic and in some of the other work we've done. That having NF2 can eventually lead to hearing loss, which can lead to impaired communication...which can lead to decreased social connections, which can lead to isolation, which can lead to depression.

And that this is one of the quality of life paths that we feel concerned about. It's not the only thing operating. But it's one possible path that people can follow.

And so you know, as I thought about the research that's ongoing in the clinical setting, not the -- clinical research and treatment, we are at this point intervening somewhere here. We're working on it.

If we give Avastin or lapatinib or one of these other drugs, I didn't list them all, can we stop the impact of NF2?

Through hearing aids and cochlear implants and ABIs, can we work at this level? Can we work against the hearing loss? So what can we do here?

So say you have NF2...there's nothing to prevent that...And you develop hearing loss. These things may or may not help you. What if you get to here? Then what happens?

So this really became an area that I have been interested in for a stretch of time but this last year applied for a small grant from the Children's Tumor Foundation to look at whether we can impact things here.

So I just want to talk to you guys a little bit about that work today too.

But -- I love starting off with definitions, because then we're all on the same page, at least a little bit.

So if we think about communication and what it is, it's really a meaningful exchange of information between two or more living creatures. It's really inherently a social interaction. It requires the ability to engage in an interaction with another being.

And often communication is thought of as -- I'm sure there are many ways to define it. If we think about really four primary skills. So the process of speaking, listening, then considering what's being listened to and what you're going to say. And as well taking in all this nonverbal information.

And so I was thinking about NF2. It doesn't impact this. It may impact this depending if you're face to face. We're in the era of texting and all these things that aren't happening face to face and you're missing a lot of nonverbal communication. It can impact speaking. Right?

So we understand about vocal fold tumors and the tumors that can impact speech. Certainly the facial palsy that can come from removal of some of the tumors or the tumors themselves. And I think we all can agree it can impact listening and hearing. And that's why we're here.

There can be a number of different realms that are impacted. For people who are fairly isolated, if they've gone down that stair step pathway, they may be left primarily with this realm of communication. Which has a lot to do with in an internal feeling of isolation.

So I also -- I don't know, I couldn't resist this. I had to put this in. The communication between girls. When they say hi to each other. They're checking out their hair, and their skin and status -- is that a tan line, and comparing their figures. And all this is going on. And here's guys....hi.

I do put this in because I think of the nonverbal communication aspect, when you're not doing this as well and not doing this as well, -- you get really good at this, actually. I always think -- I always think of it as like an undercover secret agent kind of training that you go through. You get really good at nonverbal communication. Some people are already doing it. And some people not as much. Of course this doesn't apply to everyone.

So we talked a little bit about this already. But what can impair communication for people NF2? Certainly speech problems, hearing problems. Some people eventually or currently have motor or dexterity issues as well. Sometimes people will say just text. And then you know, there are these tiny little buttons that you're trying to get your fingers to -- right. Right. And balance issues that can all be related to difficulty with motor and -- motor skills.

So I did an online survey this summer. Many of you hopefully participated. It wasn't seemingly around the world. Because I asked people where they were from and probably 25 percent of people were not from the U.S. And I think we hit four continents at least. About 225 people completed the survey. And I asked a lot of questions. But I just wanted to present some basics today. So I asked people how are you communicating? If you have NF2 and you have hearing loss, how are you communicating?

I think what's really important for us to look at is that these are all the people that say they're communicating using auditory and verbal communication...that is, speaking. These people are using cued speech, which is -- if you haven't heard of it, it's like a short hand sign language that goes along with lip reading to help make some things more understandable. So this whole part of the graph right here, all these people here are relying on some type of auditory-verbal communication.

Here is our sign language group. 12 percent. And this 17 percent here I put an arrow, because this is all put together people who said I communicate using pen and paper, computer, or texting. This is primary communication. So I put an arrow there because I'm not at all clear that we would consider a pen and paper communication as a reasonable way to communicate with everyone that you come across with in your life.

This is a group that I have a lot of interest in understanding more. I think this is also interesting because when you -- when I talk to clinicians, I say I just tell everyone to learn sign language. And part of the educational piece is that only a smaller percentage of people are using it in this community.

Now granted, I can do the whole bias thing, this was an online survey and certainly may not reflect the entire community. Keep that in mind. But this is actually our experience in our clinic as well and in talking to some other people. I just don't think there's a huge uptake, certainly not enough to keep us from needing other methods of communication. And what we have left is that the vast majority of people are really relying on auditory/verbal communication. And I think we need to keep that in mind.

Interestingly, no matter what the mode of communication, 93 percent of people in this survey, which ends up being about 200 and change, 200 people, said that they have difficulty communication in multiple settings, restaurant, home, school, phone.

It's a huge problem. No matter what option people are using. It's a problem.

So as part of the money I had gotten to do the survey, actually it was intended to take a look at apps that are available that could potentially support communication for people. And so I'm working with an audiologist at Towson University at Johns Hopkins. Apps that can translate speech into text so you can read it and text that you type into speech if you're having difficulty speaking.

We started with the Apple platform and products, because what I would say coming into our clinic is that almost everyone has an iPhone. And then when I did the online survey, we actually saw everyone had a smart phone, I asked them what type. And half have an Apple and half have a droid and a small percentage with other products. We had already planned the whole study when we learned that actually there's a good split in the market not only in the general population, not only in the NF2 but the general population.

We looked at 10 different apps that fell into one of these categories and assessed them. With me thinking about utility from an NF2 perspective and Dr. Smart thinking about the utility from hearing and hearing loss perspective.

Really this is the question, can technology fill some of this gap? Can things we already have be brought into service?

So again from our survey we found that, you know, the majority of people who completed the survey - again, an online survey. These are our tech people. But 72 percent own a smart phone and 92 percent reported they have an iPad or some other sort of tablet. The technology is already out in the community. Can we harness this?

So I just wanted to go over a little, because this aspect of the study is a little further along... that we reviewed four -- we could locate four speech-to-text apps for the Apple platform. You're probably familiar with Dragon dictation, we looked at voice dictation, V lingo, and Paper Port Notes. Happily they're all free except this one that is a dollar. Available for sure. And available for most of them for multiple platforms.

They don't require a lot of memory. Dragon has done a nice job of translating into a lot of different languages. These guys are catching up a bit. They all need Wi-Fi to operate. That's one of the drawbacks, if you don't have Wi-Fi and you're just with your phone, these won't work.

And I did want to make a note, I discovered here this summer that Dragon is owned by a company called Nuance, and that company just bought Vlingo and Paper Port Notes. It may be the voice recognition system inside these two apps will get a lot better very quickly. Because they were just purchased by the same company.

So here's how we did this. I created this paragraph as a trial -- you can read it. The text was created as a trial of the voice recognition accuracy of this app. So I would read this to the app. and it would spit back the dictation. This one made 11 errors. If you can see -- I tried to put them in bold. The places that made the errors. Here's vestibular. I tried to use a medical term, a schwannoma, from endymoma. From an astrocytoma. And this is the NF2 gene.

It did pretty well. This was in a medical office. I then went on to do it in a sort of busy way and it degraded considerably. As you can imagine with a lot of background noise.

And so the next step that we're going to do is to actually use a blue tooth, when I'm talking. And put the iPad further away and see if putting something right by our mouth will make it more like a quiet

medical office. All in all if you were sitting with your doctor in a quiet medical office and they were speaking and you were holding the iPad and doing your best to lip read and looking down to double-check things, it might work as a way to supplement your understanding of what's being said.

The other thing that we looked at, all of our reviews are available online. I'll tell you how to get them. How to use the computer system is if you program the app. ahead of time with questions you have, then you can put the cursor right underneath your question and as the doctor answers, it gets recorded right under your question. And then you can ask the next question and it records the answer. And then you have a perfect transcript with a few errors of what was said at the appointment.

And that way you can remember the questions you want to ask and have them answered. We have some suggestions like that that we've made as well in our review.

STEVE: This was done with dragon dictation on like an iPad or iPhone?

AMANDA BERGNER: Uh-huh.

STEVE: I have both, Dragon dictation on my iPad, try to use, and I have Dragon naturally speaking on my Windows-based laptop. And with that I use a blue tooth microphone, works really well because it will give you continuous dictation versus this, dragon dictation, I don't know how you did it, but on mine he can only -- you can only do about a paragraph, two or three sentences at a time, and then you have to stop and let it process. Were you able to do this all on one?

AMANDA BERGNER: And I did a rather lengthy paragraph.

STEVE: That's interesting. Maybe that's a newer app. then.

AMANDA BERGNER: That's possible.

STEVE: That's a lengthy paragraph. Yeah. Maybe it's a newer app. I'll check into that.

AMANDA BERGNER: It's possible there was an update.

STEVE: That would operate the same way as my Dragon naturally speaking. It is continuous, no lapses or no breaks.

AMANDA BERGNER: One of the things we recommend in terms of communication with people in the clinic who have hearing loss is that we -- as clinicians, we don't just talk for a long time. That we talk for a shorter time and check in and be sure that there's good understanding. And then say something -- you know, so we go in smaller pieces anyway. When there's communication, when there's a higher risk for miscommunication.

But I think that's really important. Some of the other apps did the same thing you're talking about. We could only get a little bit. And then a little bit more. But this one we did all at once. But thank you for sharing that.

So just to compare with the Vlingo, there were 50 errors in the same paragraph. With Vlingo. I like this one. This is by lateral vestibular schwannoma -- really to the point where this is not understandable here. Hearing loss and balance dysfunction. We'll see if this program can tell -- I can't even tell what I said here.

Oh, tell a schwannoma from an ependymoma. We got "panda mama." And it had trouble with NF2 gene. This becomes not as helpful in terms of supporting people's understanding because there are so many errors. So that the accuracy of each of the programs was assessed in this way.

Again, this is just one aspect of the review. We talked a lot about the things that you brought up, about how long you can talk, and some other tips about using the app. Is in our complete review. Which is downloadable from the Web.

So then we looked at six text-to-speech apps. There were many more text to speech apps, many of these have been created for people who have stroke or kids with autism, that have difficulty with verbal language. These are quite a bit more expensive. This one is free. But the upgrade, which is totally worth it in our opinion, is \$100.

Available for -- mostly for all of the systems. Verbally is only on the iPad. Only in English. But you don't need Wi-Fi to operate them, which is nice. Because then you can have them and use them anywhere.

Several of them use pictures. Because they're created more for children. And several of them don't have any pictures.

So just to give you an idea of how we did the review, each of our reviews contains -- this is just a piece of it. Several paragraphs that talk about the positive aspects of each app. If we took about proloquo2go. Specific categories and -- this is set up for preprogrammed phrases and you can click on it and it will ask out loud whatever the phrase is or say it out loud. You can add easily your own preprogrammed phrases.

You can -- with this app. for instance, you can go back and see what you said in the last 15 minutes, hour, day, and earlier. It saves phrases that you've used before.

We in our review, we tried to hit on a lot of issues that we found. When using the keyboard to type sentences, the app. will speak everything that's written on the page each time. If you've already asked a question and you type another one, it will ask both questions and that is not really what you want to do. Then we would try to put in when we found solutions.

We would put in what we found as a work around. And then provide some other tips.

This is just a quick idea of what's included in the review.

And here are some pictures of proloquo2go. This one has pictures. Maybe certainly better for kids that are having difficulty with communication. You can preprogram phrases and different like whole

sentences or small phrases. The beginning of a lot of phrases, I want, and a list of things that you can click on really quickly. You can see up top where it's creating a sentence. I feel happy, I want to go outside. And then when you press play, it speaks the entire sentence for you.

This is a picture of verbally, which is a non-pictorial app. This is all language and vocabulary. And creates the sentence in here which then you can choose to speak. Then you can save that sentence if it's something you want to use regularly.

One of the things that we were wondering about is whether this would be helpful for people in the hospital right after surgery. Maybe not even for a long stretch of time. But just for that stretch where they're recuperating from surgery and having difficulty speaking. That an iPad at a hospital center could be preprogrammed with phrases that are used pretty regularly right after surgery and provided to patients to support communication with the people rounding in the hospital as they come to see them.

AUDIENCE: You mentioned the preprogramming of questions. What comes to mind for me is in addition to that idea about post surgery, I'm sure everyone in this room has been to a number of medical consults, which are daunting and laborious and difficult not only for the patient to hear but then difficult for the medical professional to communicate. You know, I mean, it's both ways sometimes. And we get various patient-doctors who have found ways to -- and it seems to me another suggestion for preprogrammed questions would be the basic bottom line consult questions.

You know, what is the outcome, if I do this procedure, whatever. There's a litany of questions every patient walks into.

AMANDA BERGNER: Right. Right. And I have lots of ideas. But I would think, you know, one thought I have is whether we're going toward making our own app. for instance. Or something that would attach to one of these apps with a preprogrammed bank of phrases and questions. I'm getting an ahead of myself. I think there are lots of potential applications.

Where are we headed?

What we're doing now as we completed this review of all of the apps and it's available on the Web, and what we're going to do is take the ones that we thought were the best -- again, this hasn't been tried out with patients or families, and go to patients and families. And teach them how to use the apps and then try speaking.

We'll go to different parts of the hospital, medical setting, in a cafe, in a hallway. When you're walking, when you're sitting mainly and try it out. And see how it works and get feedback from people that have NF2. That will still be at the size of a pilot study. Put in an application to the Department of Defense medical research program to increase the size of the pilot study to add the droid products because we're seeing half of the NF2 community that has -- is on the droid program. And specifically study apps in a clinical setting. Not just the feedback one on one from a handful of patients, but to actually pilot using an iPad in the setting, in a clinical setting for six months and to use it on inpatient rounds and outpatient consults. That's the ask at this point for the funding from the DoD.

There have been many people that asked about this. I'm working with a young man who's in high

school in New York and is entering a science competition who wants to design glasses that -- like the Google glasses where the language comes up on the actual lens of the glass. And so he's putting together a group of advisors for this project. And I'm on the board. It's way over my head, but it sounds very awesome.

And then actually we also -- Jennifer and I also put in a funds request to Advocure. They have a small grant in and I don't know if it was awarded or not. The application was due in August. Our idea was to assess the accuracy of speech reading. A lot of people use lip reading and we're curious how accurate it is. When we think about these medical settings, people are making huge decisions about their family, about themselves, about surgeries and interventions and drugs.

And a lot of them are relying on a lot of clinicians -- clinicians are presuming that people are understanding because they're lip reading. And we're wondering if that's true or not. We'll see if that gets funded. That's one of the other directions in which we're thinking about.

So when I come to talks like this, I always want to know like, great, I get all excited but I don't know what I can do. And I leave and think what can I do to help? So I thought I would give a couple suggestions. I'm sure there are many more things. But I would love to hear from anybody about your ideas.

I think I said it this morning in the NF1 talk. Great ideas can come from anywhere. Not every idea is great, but great ideas can come from anywhere. When you have your own experience, like you were sharing, I really -- I would love to hear from people about that.

If you wanted to participate in our pilot study of apps and you can get yourself to Baltimore, Maryland, I would be delighted to have you. Unfortunately right now we don't have travel funds. But if you're coming through D.C. in the next five months...

Everybody can take a role in supporting the various funding sources that exist now. And helping generate more funding.

So supporting the programs that exist now, letting, you know -- especially for the federally funded programs, letting people that are responsible for making decisions about that, now how important they are.

Certainly foundation funding. And then I had a lovely conversation with the gentleman who's in the room here, last night about wanting to start his own funding stream and revenue. That's of course always an option that can be dedicated specifically to NF2.

I really would encourage you to talk, talk, talk to people about what a problem this is. I think it's not -- I guess I've just been really surprised within the NF2 world how little we're talking about the actual communication problems that people are having.

And so the more we talk to people, clinicians and ask, you know, what can I do, what are my options, are there other ways we can communicate, the more awareness will be spread about the need for that.

So back to patient-centered research, I'm not sure you can appreciate this, but all of these are people that come together. And how to find me. I think there's a stack of my cards downstairs. I think -- yeah, we have maybe five minutes or so for questions. Thank you very much. I really appreciate your being here. (Applause)

AUDIENCE: Right now I have trouble with loss of hearing. I do have problems like comprehension and communication skills. Well -- where will that fall? And do I have to worry about future hearing loss as I get older? And should that be a major concern?

AMANDA BERGNER: I think that's one of the things that people with NF2 face is, you know, understanding that hearing potentially and probably will get worse. And making plans for how to communicate down the road even if communication now is okay. I think that's one of the things that families ask a lot about, people ask a lot about. I would encourage you to talk with your audiologist. Do you have an audiologist?

AUDIENCE: Not yet. First time I ever heard of one.

AMANDA BERGNER: Audiologists are trained specifically about hearing, and they often can be very well plugged in to lots of different ideas about how to assess whether your hearing is the problem or whether the connection between the hearing and the way your brain processes is working correctly.

And there are some questions like that. If it's specific to hearing, which we know can happen a lot for people with NF2, then yeah, I think it's reasonable that you would have questions about how -- you know, how can I plan ahead for this down the road as it might get worse. Does that answer your question?

AUDIENCE: Sort of kind of, yes.

AMANDA BERGNER: I'm happy to talk to you separately too.

AUDIENCE: You talked a lot about hearing loss and ways that people communicate... about cochlear implants - are those fairly effective? Are we finding good research with those?

AMANDA BERGNER: That's a great question. And I'm not an audiologist and I don't specialize in cochlear implant or auditory brainstem implants. Certainly a lot of people use those. And you know, it's a really critical intervention in this pathway. Like how can we impact the hearing loss?

And so I didn't really talk about the drugs that are available, I didn't talk about these. I was more focused here on this talk. But that's a huge part of NF2 and the auditory aspect of that. NF2 is people are looking at the utility of cochlear implants. But that's just not my area of expertise. But it's a good question.

AUDIENCE: We can talk about that.

AMANDA BERGNER: Great. Thank you guys very much. I'd be happy to hear from any of you.
(Applause)

AUDIENCE: Can I -- I have a question. Did you say that the money coming out of CTF is to develop classes that can decode speech reading? Is that happening?

AMANDA BERGNER: Not yet. It's an idea. But it hasn't happened yet.

Additional Resources

[Review of Apple Apps](#) - This is a summary of Apple based applications that help support spoken communication for people with NF2 and hearing loss or vocalization problems.

[Apps Overview Chart](#)