

**(Kimberley Smith) Thank you!
Okay, can people hear me okay? I
got the worst spot, nobody wants
to have the spot before lunch be-
cause all anybody can think about
is the food, they are about to eat.**

So, I try and get through this as quickly as I can for you. So, I am here to talk to you today about 'Quality of life and psychosocial wellbeing in Usher syndrome'. To give you a little bit of background about myself: I am a lecturer in health psychology at the University of Surrey. But prior to working there, I worked as a lecturer at Brunel University London. And I taught on the master's in health behavior and psychology.

And it was while that I was approached by a student called Gavin Dean who has Usher syndrome himself, who really felt very strongly about doing a project around psychological wellbeing in people who have Usher syndrome. So, I think, before anything else, what I would like to get across is, this is very much Gavin's project and I am here representing it on his behalf.

Gavin himself wrote a blog for the limping chicken website. And that blog gives you a really nice insight as to why he feels that this was an important project to do. He says, having Usher syndrome himself since he was a teenager,

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he often found himself in many situations where his deafness and blindness left him feeling depressed, anxious, isolated, hopeless and frustrated because of the many challenging and uncertain experiences that he encountered in his everyday life.

He felt that these feelings came from barriers in communicating with other people, barriers in mobility, barriers in accessibility and also a general lack of understanding and appreciation and even acknowledgement about what Usher syndrome is and how it effects the people throughout their lifetime, both physically and psychologically. So, by undertaking this study, it allowed us to get some - he calls it - ,hard scientific evidence in the public eye on the impact of Usher syndrome on psychological wellbeing'.

And we published a paper on his work in 2017 in a journal called BMJ Open. And, anybody can access it. So, if you feel that you want to, you can go and download the study that myself and Gavin did. But before we get into the study, I would like you all to think about what health is.

If I was to ask each one of you: „How would rate your one health on a scale that runs from excellent, very good, good, fair or poor?“ „How would you rate it?“ And then think about, what is it that is influencing how you rate your own health. There are lots of different things that are influencing how people think about their own health. There might be physical issues that are happening, but with other people it might be linked to functioning, not to

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being able to physically do, what they want to do or need to do. For some people it is social, they can't go out and engage with people, as they want to. For some people it is much very much linked to how they feel psychologically. And this is reflected in the World Health organization definition of what health is. It is not just the absence of disease or infirmity.

It is a state of complete physical, mental and social well-being. So, when we bare that in mind, and we know that health is this really multi-dimensional complex thing, why is it that - and no offense to any other Usher researchers here, who do this - so much Usher syndrome research is focused on the biology of Usher syndrome and finding a treatment. And there is no doubt that this is incredibly important. But Usher syndrome is not just biology. Usher syndrome is something that is affecting people. And it will affect them psychologically and socially and it will do more than affect them just biologically.

It might have an impact on something, that we in health psychology call 'quality of life'. And this broadly is the satisfaction a person has with their own life. And it is influenced by lots and lots of different things. So, it is influenced by our psychological wellbeing, how happy we are, whether we feel depressed or sad. It is influenced by our environment. This work is showing that, if you are in an area where is lots of green space, that is better for your wellbeing, than being in an environment that is very built up and polluted. Spirituality will have an impact on people's quality of life, as well. Spirituality can really help

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enhance people's quality of life. Another thing that can influence is the level of independence, whether you are able to do things for yourself or not. When you actually ask people, „What is the main thing that influences your quality of life?“, social relationships will often come out as the top thing.

We are social creatures, we need to be around other people and so social relationships do explain a lot of quality of life. And finally, physical health is a part of quality of life. But it is not all of quality of life. So, when we consider - wrong way - quality of life and what it is, we can see that, well, there is lots of things that are on that diagram that might impact people who are living with Usher syndrome. So, let's consider a few of those. Firstly, level of independence, will be something that might affect people who live with Usher. And Watters-Miles did a very nice thesis a few years ago, where they talked about something called ‚independent dependence‘. Where to maintain quality of life and to maintain independence, sometimes people would have to ask others for help with certain activities.

Something else that might affect level of independence in Usher syndrome is change and uncertainty. So, Usher syndrome is very different depending on the type that you have. But it is also something that changes over time, the longer that you live with it. There is a lot of uncertainty around, how that change might affect you. And there is a possibility, that as the condition progresses, that it might have an effect on level of independence. But ability to

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personally manage goals is associated with a better quality of life in people with Usher syndrome. And adaptation in particular, is something that can really help. So, there is research out there that has been done, interview studies, where people have almost prepared themselves for the fact that they might lose their sight.

So, they teach themselves Braille and they adapt to what is going to happen, so that they can maintain independence. And these ways to maintain independence without - I don't know - I totally lost my train of thought here, I got up 4.30 this morning to catch to the plane. And my brain is completely fried, I am really sorry. I just smoothly go on to the next slide. So, environment is something else that can impact quality of life. So, navigating environment is gonna be a key thing. When you look at interviews that have been done in people with Usher syndrome, uneven pavements can be something that can be hard to navigate, branches that hang down, cyclists, these are all things that can impact on quality of life.

Lighting is something that has been shown, can improve quality of life in people with Usher. So, something as simple as turning the light on in an indoor environment can really help. Communication is gonna be key, as well. So, not everybody will be able to communicate with sign language. And that can be quite difficult, when out and navigating in a new environment. Assistive equipment is something that can be really helpful, so there are various things that can help you use computers, people can find that canes can help, when they go out. Change and

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learning skills will also help, as the condition progresses. Adapting to and learning new skills will be important to help you to navigate your environment.

In terms of physical health, a lot of people with Usher syndrome have balance difficulties, which will invariably impact on their ability to go out and about. There is also work showing that fatigue and headaches can be issues, as well. And that is proposed to be linked to the fact that you have to concentrate so hard, to think on things that it can be really, really tiring, when you have Usher.

And there is also associated co-morbidities. So, some people might also have intellectual disability. Psychologically, there is work showing that Usher syndrome can be linked with higher rates of depression. In particular, depression is linked to the diagnosis, when you are told that you have this condition. That can be really hard for people. And also as the condition progresses and you start losing independence. So, I think, not being able to drive anymore for a lot of people is a key event that can lead them feeling quite depressed. There is also anxiety, stress and fear that can arise, as well. This is linked a lot into the uncertainty of what might happen in the future and the progressive nature of the condition. But it is not all about news.

If you look at interview studies, a lot of people with Usher syndrome have an incredibly positive outlook, they see it as a challenge. Something they can - they try - like this diagnosis empower them, rather than beat them down.

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So, there is work that shows us, that is not all doom and gloom. I think, that is one thing, I really want to get across here. A lot of researches focus only on the negative for some reason, but there is positive stuff out there, too.

Usher syndrome also has a big impact on social relationships. First thing that will come to most of your minds, will be communication. Whether you can communicate with other people and how you do so. People with Usher syndrome can also report feelings of loneliness and isolation which are linked to deafness.

But again, it is not all bad news. When you look at what is out there, people with Usher syndrome, peer connections, having friends, who also have Usher syndrome can be a really beneficial thing. To have someone else there, who understands what living with this condition is like, is good. And then your friends and family and social support networks are something that will really help improve your quality of life. It is something that will also have an impact on your family, as well. So, when somebody is diagnosed with Usher syndrome, they often are worried, about how it is gonna impact on their family, on their ability to look after their parents, when their parents get older or their ability to look after their children.

So, in terms of relationships there is a lot of work that showing that Usher syndrome and social relationships are linked with wellbeing. So, when we go back to this quality of life figure, we can see there is a lot of things that influence quality of life in people with Usher syndrome. So,

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myself and Gavin - (someone sneezing repeatedly) - bless you, I hope you are okay.

Really, we want to look at everything, but you can't look at everything. It is really ridiculous to expect people with Usher syndrome to sit down for two hours and answer question after question for you. So, we decided to focus our study on two main areas that we are interested in. Social relationships and psychological wellbeing. We wanted to see, how these were linked with quality of life.

The aim of this study that we did was to determine whether psychosocial wellbeing is associated with physical and mental quality of life in a UK-resident population of adults with Usher syndrome. We asked for people to take part, who had a diagnosis of Usher syndrome, were aged 18 or older and also lived in the UK. And, Gavin had links with SENSE, the charity in the UK. He had links with meet-up groups, as well. So, we were able to advertise through a lot of deafblind charities, through magazines advertisements, meet-up groups. We managed a 120 people, that said that they were interested and we had 90 people who ended up completing the survey for us.

The first thing that we measured was called 'health-related quality of life'. We looked at mental quality of life and physical quality of life. And we wanted to see what predicted these in people with Usher syndrome. So, we looked at their characteristics, their age, their gender. We looked at health-related characteristics, sight-registration status, deafness level and associated co-morbidities. We

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also measured depressive symptoms, we measured loneliness and we also measured social support. So, apologies for the next table, but I will just tell you about the main things to take away from this. In terms of age, most of the people that we had in our study were around 36 to 45.

60 % of our sample were female, more women than men took part in our survey. And most people, 43 %, were employed or self-employed. Most of our sample had Usher syndrome Type II, just under half of the sample reported having that. But 11 % reported that they didn't know or they hadn't been given a type of Usher syndrome. Most people had severe hearing loss, 63 % of our sample. 70 % were blind or had a severe sight impairment. And most reported, that they did not have other disabilities or illness, 63 % had no other illnesses or disability. So, when we then looked at physical quality of life, we found that this was linked with how depressed people felt. Whether they had another chronic illness or disability and being older. So, this meant that the higher people's depressive symptoms were, the poorer their quality of life was.

Those people who reported having another chronic illness or disability were more likely to have a poorer quality of life. And those people who were older were more likely to have a poorer quality of life than people who were younger. When we then focused specifically on mental quality of life, we found that this was predicted by depressive symptoms, social support and loneliness. The higher somebody's depressive symptoms, the lower the mental quality of life. The more social support they

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had, the better their mental quality of life. And the more lonely they were, the poorer their mental quality of life. So, depression and loneliness, we found, were linked with a poorer quality of life and social support with a better quality of life. And I don't think, these results will surprise anybody in the room. But, what we really wanted to get across was that psychosocial wellbeing seems to be important for quality of life in people with Usher syndrome.

So maybe, it is time to start thinking to about health more broadly in people with Usher syndrome and to start thinking about psychological wellbeing and their social relationships. It has been good points to the study, we did. This is estimated, that there are around 10,000 people in the UK who have Usher syndrome.

So, the fact we managed to recruit 90 people, I think, is a really good sample size. And we had a wide range of ages who took part, too. But we can't necessarily take these results and apply them to every single person who has Usher syndrome.

A lot of the people in this study were employed, a lot of them were female. So, we probably had an overall better functioning Usher population, than you might normally find. These were also self-selected people, there are issues with that, too. People who have more issues tend to put themselves forward for studies, where they can talk about those things. The questionnaires that we used, were short and self-report.

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To get a clinical diagnosis of depression, you are given an interview by a psychiatrist, and obviously that wasn't feasible for what we did here. So instead, we used a symptoms questionnaire of depression, where people would rate different symptoms. So, we might have overestimated depression slightly. And this is also a cross-sectional study, which means, it is a study that was just done at one point in time. So, we don't know, what it is leading towards. But I think, the main question is: Can we improve quality of life for people with Usher syndrome? And something that comes out of the literature quite a lot is that we need to promote the positive.

As all of us move through our lives, we are gonna encounter transitions that will affect us psychologically and socially. And for every single person, resilience is one of the main things that we can build up, that will help us to cope with these changes, as we age. Coping is gonna be really important, too. So, having a problem-focused approach to coping, has been shown to be much better for psychological wellbeing, than having what we call an 'emotion-focused coping strategy'.

And empowerment is really important, too. If you feel like your independence has been taken away from you by this condition, it is really important that you try to empower yourself somehow to improve your quality of life. So, take up a new hobby, do something that makes you feel good. Redirecting goals and adjusting is going to be key, too. So, it's not just Usher syndrome. All of us as we age are going to find that we can't do certain things.

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So, we might have to readjust our goals and adjust because of that. The key thing is: Don't just focus on the negative. Now, I am a researcher who has made their living looking at depression. And I am as guilty of this, as everyone else. But as researchers, we just tend to focus on what is bad. And I think, if we are going to improve mental wellbeing, quality of life, we also need to look and really focus on what is good. And peer and social support is one of those things that is good. It comes out time and time again, as being really important for the quality of life in Usher syndrome.

There is also a lot of work, showing that cochlear implants can benefit people in terms of quality of life, too. So, what we need to do as researchers, we need to take a more positive psychological approach. I think, this claim „seek and you shall find“, applies, when it comes to psychological research. If you look for the bad, you will find the bad. But if you look for the good, you will find that, too. So, we need to take a more balanced approach to how we look at this. We need long-term work. Most of what we have in psychological wellbeing in Usher syndrome is cross-sectional. But this is a condition that worsens over time.

So, how does that impact people psychologically? We need more work to tell us about that. And I am aware, that I have just talked about Usher syndrome, as though it is one condition. I am very aware that there are very different types that affect people very, very differently. So, doing more work across different types in severities of Usher syndrome and looking at, how that is linked with

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quality of life, will also be really important. I think, the key message, I want you to go away with, as you go to lunch, is that health is more than physical. It is not just biology, health is how you feel psychologically and socially, as well. So, finishing off, I just liked to firstly thank the UsherVibe Group. They very kindly gave us root money, so that we could do this study. SENSE played a really big role in helping us to recruit people. Gavin, thank you so much for allowing me to do this study with you on Usher syndrome. And finally and most importantly: Every single person who took part in this study! And thank you all as well for your attention! (applause)