Introduction

Life and Change with Usher is a research study about the lives of people with Usher syndrome. Over two years we studied the lives of people with Usher, first in books, articles and blogs, and then by asking them about their lives.

Thank you very much to all the people who answered our questions and gave their time, and all those people who helped us to find people with Usher – without you we would not have been able to complete our project.

We interviewed 44 people with Usher 1, 2 and 3 who lived in the UK. They were men and women, from all over the country, and from different ethnic backgrounds, aged between 14 and 59. Some of them were sign language users, including hands-on signing, and others used spoken English. We interviewed some young people twice, and some adults were part of focus groups. They talked about what they thought about Usher, what changes they had been made in their lives, and how they worked, learnt and enjoyed time with family and friends. We used these stories to identify some key pointers about how people with Usher in the UK today live their lives, what they think and what they feel. We grouped these into 7 topics, and we describe each one in a little more detail below.

Diagnosis

The adults in the project said being diagnosed with Usher was a shock. They often felt emotional and sometimes became depressed. Some of the older adults said they were told in a very harsh way that they were going blind. They wanted clear information on how their visual impairment would progress – although it depends very much from person to person. Young people with Usher couldn’t remember being told that they had Usher. It seemed easier for them to accept than for their parents. People also wanted information about other eye conditions such as cataracts and cystoid macular oedema which are more common in people who have Usher.

There are also questions about the information about the different types of Usher. For example we saw teenagers with Usher 2 with a lot of vision loss, and people in their 50s with Usher 1 with not that much vision loss.

When people are first told they have Usher, things often do not change very much –vision (and hearing) does not change overnight. How people feel about themselves can change very quickly. People spoke about living with Usher, learning to accept it, and making adjustments in order to achieve what they want to in life.

Mobility

Mobility was a very important issue for participants and impacted on many areas of people's lives. One simple thing that people did was to make sure that they switched on lights and had bright lights. People with type 1 Usher and some of the people with type 3 spoke about balance difficulties and worried about looking clumsy.

Some young people had received specific mobility training on public transport which was useful, and most people felt confident in using public transport. There were difficulties when support for travel, such as passenger assistance failed. Also people found it more difficult sometimes living in areas with unreliable and irregular public transport. People living in bigger towns and cities were more positive about public transport. They said that one of the good things about having Usher was free travel on public transport.

The decision to use a cane or a guide dog was a very significant moment for people with Usher. People found it hard because it showed others that they were blind. This made them feel vulnerable, and like a fraud because often they could still 'see'. People sometimes felt they had to change and use a cane after something had happened, e.g. bumping into people in shops. Some people only used a cane in certain situations e.g. in crowded places. A lot of people thought guide dogs were more acceptable than using a cane, although they recognised that they involved a lot of work and not everyone liked dogs. People who were guide dog owners were worried about attacks on guide dogs and discrimination. Some people were happy to carry on without using any aids – for other people it was a big relief when they did.

Communication

People in the project communicated face-to-face using spoken English and sign language, including hands-on signing. People who used spoken English did not change to using sign language. Some people did learn some sign or manual language (hands on or visual frame, or the deafblind alphabet) to help with communication when they could not wear their hearing aids or it was dark. Because more young people with Usher 1 are having cochlear implants at a young age, only one of the 5 young people with type 1 Usher were sign language users. All 10 of the adults with Usher 1 were sign language users.

People used standard print, large print, paper or ink of different colours, to read written documents. Some used audio materials, and only two used braille all the time for written materials. One more person used braille for labels. A number of the adults wanted to learn braille, to prepare for the future, but found it difficult to get information about courses and funding.

Quite a lot of the people used the same technology as people without hearing impairment or visual impairment, such as smartphones, E-readers, and iPads. People with Usher used equipment with enlarged fonts, used built in torches and magnifiers, changed the text background and colours. A smaller number of people used specialist technology such as braille note, Dolphin, radio aids, audio description on the TV. Some found it difficult to know how this equipment worked and some found it difficult to afford.

Family, friends and support networks

Overall family and friends supported people well. People with Usher worried about telling others about having Usher, but after they had told them, there was an adjustment and then people's relationships did not change. Sometimes friends and families would 'forget about' a person's Usher which could be annoying! People found asking others to drive them especially frustrating.

Deciding to have a child and being a parent were important issues. A few participants were very worried about passing on Usher to their children. Other people said that there were worse things that could happen, and if their child did have Usher they would be in the best position to help them. Some people were worried about coping as a parent with Usher and decided to have a child quickly whilst they had more vision, or not to have a child at all. The people who were parents were very happy and for the most part coped successfully. A small number of people were parents before they knew that they had Usher.

People received formal and informal support. Formal support included schemes such as Access to Work, communicator guides and specialist advice from organisations like Sense. Informal support came from local blind societies, Deaf clubs and Facebook groups. Most people, but not all, looked for support at particularly difficult times. It was important for people to meet other people with Usher who were like them in terms of age, communication preferences, and vision loss. People wanted to talk about the positive achievements of being a person with Usher, as well as the difficulties.

Education

People attended different types of schools. All of the adults with Usher 1 went to schools for the deaf and were sign language users (even though the schools were oral schools). They all knew that they had Usher when they were at school (except one whose parents knew but they did not). Most of the adults with Usher 2 did not know that they had Usher when they were at school, and went to mainstream schools and had support from Teachers of the Deaf. Some had a lot of support, some had very little. A couple of the adults attended special schools for children with learning difficulties even though they felt that they did not have learning difficulties. Two people with Usher 3 swapped between mainstream schools, a school for the deaf and a school for the blind as their hearing and visual impairments changed. Most of the young people with Usher 1 had cochlear implants, they were more likely to be speech users and attend mainstream schools.

Some teachers understood the needs of people with Usher syndrome. Those people had good experiences at school. Others struggled to make their needs known and to be understood. Specific support from teachers of deafblind children was unusual. Visits from teachers for children with visual or hearing impairments was more common. People had difficulty getting appropriate careers advice.

Some people were bullied and worried about looking different – some people had loads of friends and had a very happy time. Some young people who were English language users found it difficult to fit in at schools for the deaf where the majority of pupils were signers.

Employment

There were 32 adults in the project. 17 (53%) were employed, 9 (28%) were unemployed, 4 (13%) were full time mothers. One person (3%) was at university and one person (3%) was unable to work due to illness. 8 people (25%) worked full time and 9 (28%) worked part time.

Many participants worked in organisations for people with disabilities. Half (9) of the people who were working, worked for organisations that provided services and information for people with disabilities, particularly those with sensory impairments. A further 4 were employed by the NHS. Of the 9 unemployed people 3 were looking for work with, or wanted to work for an organisation specialising in hearing and/or visual impairments. 7 sign language users were working full time or part time. Six of these were employed in organisations for people with disabilities. People wanted to work in these organisations because they were more understanding especially with communication. The young people looked forward to all sorts of careers in the future, although a couple also mentioned that they wanted to work for deafblind organisations.

People who used Access to Work found it useful but reassessments were stressful. Sometimes there were difficulties in getting payments in time. People said employers usually understood their difficulties but sometimes they thought employers were just following the rules, rather than understanding completely. Some people did not want to tell others about the full extent of their vision and hearing impairments so this made work stressful. Other people said that as soon as you let other people know that you have Usher life becomes a lot easier.

Leisure Activities

People took part in lots of different activities such as martial arts, going to the pub, baking, composing music, and rock climbing. Some of the parents with young children said that they did not have much free time!

After being diagnosed with Usher some people wanted to do as much as possible whilst they could still see well enough to do so (even though most people with Usher do not lose their sight completely). Many people were determined that Usher would not stop them from taking part in activities. In order to continue participating they had to make adaptations and adjustments to the ways that they did things and how they felt about it, for example – taking a friend’s shoulder at the cinema, starting tandem cycling, using taxis more often. Sometimes Usher did stop people taking part in activities (especially driving) but generally people tried to be positive and focus on new activities and ways of doing things. Some older adults who had more vision loss were more likely to take part in activities at home or to rely on friends and family to take them out and about.

Some people with Usher took part in activities specifically for people with visual impairments - these activities focused on sports, e.g. cricket, archery, goalball. They found that they were supported through equipment, coaching, and understanding. A smaller number of people took part in activities with other d/Deaf people. People did meet up with other people with Usher but in general this was for support rather than leisure activities.

SIP

41 participants filled in a survey called SIP (self image profile) about their self image and self esteem. We used different surveys for the adults and for the young people. People were asked to show on a scale of 0-6, whether different characteristics were ‘not at all like me’ (0) through to ‘very much like me’ (6). The different characteristics included things like happy, hard working, and confident. Participants did the survey twice – first they had to say ‘how you think you are’ (self image), then they had to say ‘how you would like to be’ (self esteem).

People with Usher gave answers similar to the rest of the population. Overall the adults and young people with Usher had ‘average’ levels of self image and self esteem. There is a small suggestion that adults with Usher 2 might have a more positive ‘outlook’ than people with other types of Usher. Also it was interesting to see that about a third of the adults with Usher wanted to be less sensitive, whereas people in the general population usually want to be more sensitive. It could be that adults with Usher wanted to feel less emotional about Usher or maybe they had a different opinion on what ‘sensitive’ means. The young people saw themselves as friendly, helpful, and fun however they also worried a lot. Some of the young people felt different from others, and others wanted to feel more different. This is interesting and might be an area to look at in the future.

Having Usher in the UK today

The stories people told began a picture of what it is like to have Usher in the UK today. We saw that people with Usher enjoy life. Most had already accepted being deaf – they did not want to become visually impaired, but that was the situation. Some were, or had been, depressed. They may have been depressed when they were diagnosed, but also they became depressed at moments of change, for example, giving up driving.

People with Usher also managed, and learned to manage. They used specialist aids and equipment such as guide dogs and iPhones, and accepted and asked for support. Sometimes of course, they did not manage. There were understandable and often avoidable frustrations. Some of these were with people and others were with systems. Sometimes the environment was against them. However usually people were able to deal with these obstacles.

Also things develop – things changed for individuals and things changed within society. For example nowadays young people with Usher are more likely to go to mainstream schools, young people with Usher 1 are more likely to have cochlear implants, and the understanding of the different types of Usher is changing. One thing that was difficult for some people is that at the moment there is no treatment for Usher and no matter what they do their Usher will not get better.

In the 7 topics mentioned above, we found that the themes of change, predictability/uncertainty, and diversity helped us understand what is unique about Usher – the Usherness of Usher. Change is at the centre of Usher. For some, things changed when they were diagnosed, for most people things changed when something happened which they could not avoid e.g. when they started using a cane. They needed support, which was often available only through family and friends. Some people had to change jobs when workplaces and managers were no longer prepared to meet their needs. People also all knew they would change in the future. Some might decide to get on with it, and cope with each change, through new aids, strategies, and support. Others might hang on as long as possible in the old way, before an accident or similar forced them into something new.

The second theme of Usher is uncertainty and predictability. At diagnosis, predictions were often made, but in many cases, they had turned out to be inaccurate. Participants worried sometimes because of the uncertainties of Usher – what would happen, how would their friends and families react? Others decided to beat the predictions, for example by deciding to have a family quickly, or to go skydiving. They challenged the predictions and the fact that they might not be able to do things. Some people wanted to take control of the uncertainties; others decided to let things happen.

Finally, the participants and their experiences are very different – there was lots of diversity. The progress of their visual impairment (and hearing impairment) is also very individual and diverse. What type of Usher they had or what had happened to other family members with Usher did not predict how fast and how much visual impairment an individual would have. Individual diversity is important - what was an obstacle for one person might not be for another. Some people had negative experiences, and some people were more depressed, or felt more isolated, or have given up more activities than others. They had different amounts of support. Some find adapting hard. But many had positive experiences as well. People with Usher can do many of the things they want to – they sometimes have to find a way round it – but they can still do it.

Once again thank you - the project depended on the willing participation of people with Usher syndrome. We would like to thank all those who took part in interviews and shared their time with us. Everyone's views were interesting and we have learnt from them all.

If you would like to know more about the project or to ask questions or make comments, please contact Liz Ellis on usher.project@contacts.bham.ac.uk , by telephone on 0121 414 4392, or by text on 07591 126370.
We will be happy to send you a complete version of the final report, either electronically, or by post. It is in font size 14. We can send it in bigger font, audio, coloured paper, or braille – let us know what you would like.

You can also read the final report online:
http://www.birmingham.ac.uk/research/activity/education/projects/usher-syndrome.aspx

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