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Usher 2 – how is it for *you*?

Usher 2 sometimes seems like an invisible condition. It has no outward signs, and many people don't realise they have it. And plenty of doctors have never heard of it. What is it like to live with this hidden syndrome?

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All forms of Usher cause impaired hearing and sight, so it might not seem significant which type of Usher people have. Whether they have Usher type 1, 2 or 3, people often talk most about the challenges presented by their visual impairment, which develops similarly in both types. But there is an important difference when it comes to hearing, because whereas people with Usher 1 are born profoundly deaf, those with Usher 2 are born partially hearing, and people with Usher 3 are born hearing and sighted.

This difference can have far-reaching effects on people's early lives and identities. Crucially, people with Usher 2 usually wear hearing aids or other devices and live in the hearing world. Most attend mainstream schools, perhaps within a specialist unit, and very few use British Sign Language. Unlike most



A meeting of the HSI group

people with Usher 1, therefore, they are not part of what is often described as the 'Deaf community'.

When I meet Des Lucy, who has Usher 2, there is no sign that he has an impairment, apart from his discreet hearing aids – and Des's lipreading is so fluent that I am unaware of his hearing loss as we talk. Des wears glasses, but then so do I. But while I can see Des, the table between us, and the wall behind him, Des can see only my face. I only know →

What is Usher 2?

People who have Usher type 2 are born with a partial hearing loss. In their teens they develop Retinitis Pigmentosa (RP), which causes visual impairment. The severity of symptoms varies: some people have a profound hearing loss, while others have more hearing, and the degree of visual impairment is not the same in every individual.

However, most people with Usher 2 wear hearing aids, use speech, and lipread. In most cases, their hearing loss generally doesn't change over time, although in some cases it does.

Initially their visual impairment may not cause too many problems – many people adapt to it. However, the effects will become more severe. The first symptom most people notice is nightblindness. Their side vision may also begin to give them problems, so that they knock into

things or fall over them. In time, moving around – especially in unfamiliar places – may become more and more of a problem.

Other effects of RP can include difficulty in adjusting to changes in the brightness of light, visual disturbances such as flashing lights and floaters, and cataracts.

Most people with Usher 2 continue to use their central vision well into middle age – they will probably still be able to read, for example. Eventually their field of vision may narrow so far that it becomes hard for them to read or lipread. They may have to learn other methods of receiving information, such as deafblind manual, or Braille. Very few people become totally blind. There is no treatment for RP, but it is possible to learn techniques for maximising the use of the remaining vision.

this because he tells me. If you meet someone with Usher 2, you probably won't realise it.

They may not realise either – many people with type 2 don't find out they have Usher until they are well into adulthood, although they may have had symptoms since childhood.

“ Fear of the unknown is one of the hardest things to cope with ”

Colin Bennett for example, had always found it impossible to see at night – he remembers as a young child in the war being sat on the doorstep to eat carrots because his mother had heard on the radio that pilots ate them to help them see in the dark. But Colin was not diagnosed with retinitis pigmentosa until 1986, aged 45. And it wasn't until 1989, when he went to get better hearing aids, that the consultant diagnosed his Usher.

‘I didn't know there was a thing called Usher. He was the first person who linked my sight problems to my hearing problems. That changed my life because I got in touch with Sense.’

The frequent failure to diagnose Usher 2 is largely down to it being so rare. Maybe one in 100,000

Colin Bennett



people have Usher, almost half of whom will have Type 2. As few children with Usher 2 go to schools for Deaf children, where teachers are more likely to be Usher aware, their Usher is rarely picked up at school age. Once out of school, it is even less likely to be spotted. Most GPs have never heard of Usher. Most optometrists have never seen it.

Mandy Priestman is unusual as she went to a school for Deaf children from the age of 11, and that was where her Usher was picked up, when her PE teacher saw her begin to miss the ball in rounders. Diagnosis followed a year and a half later, at the age of 15. When asked how she felt at such a diagnosis, Mandy replies ‘It worried my parents. I just thought it was something else to take on alongside my hearing.’

Gloria Ward runs Sense's Usher peer mentoring scheme, and meets many people with Usher 1 and 2. She comments that ‘it seems to be less traumatic if someone is told before they become a teenager. The implications don't really hit you when you're young.’ This is tough on people with Usher 2, who are often in their 40s before being diagnosed, and have to deal not only with the diagnosis but the with the effect of it on their already well-established work and family lives.

“ Many people with type 2 don't find out they have Usher until they are well into adulthood ”

Of course, everyone is different, and some people cope better than others with their failing vision. Most people cope well most of the time. The trouble with Usher though is that it is not a static condition. You can't just get used to it and move on: each time the impairment worsens it can restart emotions such as grief, anger or fear.

Interestingly, people interviewed for this piece all talked of their Usher with acceptance, and were determined that they would live good lives no matter what happened. They had all had dark moments, but had consciously moved on, and adapted. Nadeem Alvi, for example, went into depression when he was diagnosed. But he says ‘I came out of it and decided I couldn't sit back.’ He decided to find out everything he could about Usher, and to restart his education. Now he says that his studying is what keeps him going.

Asked whether he knows anyone who hasn't been able to deal with finding out about their Usher, Nadeem says that he doesn't, but points out that he'd be unlikely to meet them. The very nature of depression means that people are likely to withdraw and stay at home, and if they're not in touch with an organisation like Sense or Deafblind UK, they may find it hard to recover alone.

Sense's National Usher Co-ordinator, Marylin Kilsby says that she has reassured people over the years that it's a normal reaction to be depressed at a diagnosis of Usher. 'Because obviously some people

do get depressed. Naturally. They've had the shock of the diagnosis, and they don't know what the future holds. That's a biggie with Usher: you don't know a prognosis. You don't know how quickly your sight is going to go down.'

Fear of the unknown is one of the hardest things for someone to cope with. Listening to people with Usher 2 talking about their lives, it seems that they deal with this fear by focussing instead on the current reality of their condition.

Mandy Priestman is now 50, and has just had to retire from her work at the British Library in Boston →

"The most important thing is to feel accepted"

Des Lucy is 46 and has Usher 2, like his older brother Kevin. He is married to Diane and has three sons. Des is a railways rolling stock Standards and Document engineer, commuting daily to London from his home in Kent.

Des started wearing hearing aids aged five, and learnt to lipread at primary school, where he split his time between mainstream classes and a specialist unit. Des doesn't know if his secondary school had a hearing aid unit – he was so proficient at lipreading by then that he didn't need one. The only exception the school made for Des's hearing was letting him off French.

Des's sight didn't begin to deteriorate until his teens, after he'd left school, but it didn't affect him too much, and he continued to play football – one of his passions. He started work for the railways as an apprentice.

"He continued to play football – one of his passions"

He lost his night vision first, noticing it when he went out with the lads in the evening: 'I didn't see the steps, or the bush. I didn't question why – just assumed they could see better. And I didn't notice my visual field shrinking.'

For many years, Des's sight didn't cause him too many problems. He played squash right up until his oldest son was born in 1984. 'I realised that I couldn't see well, though – as did my opponents, who learnt that I had a blind spot!' He gave up squash and football, but occasionally tries his luck. 'Now I only play with my boys, but it's safer for all parties if I stay in goal. The last time I



Des Lucy

played I was in goal but I decided my team needed help so I ran down the wing, and straight into a bush.'

Des wasn't diagnosed with retinitis pigmentosa until about nine years ago, when he began to find it hard to see the computer screen at work.

Des's vision is changing, and nights are getting harder. His visual field is down to 10-15 degrees, and now he has macular degeneration in one eye. He says 'You start to accept there are things you can do and things you can't. There are times when you feel down. But you just move on. I've had lots of distractions – wife, children, change in the railways – and I've had to adapt, make new friends. You've got to accept it. And the most important thing is to feel accepted, and to feel that you have some value.'



Carol Pollington

Spa, so her life has taken a new turn again. She says 'I'm very philosophical about it. I think I always have been. I'm more aware of my Ushers now, as it's getting harder year after year. But when I was younger, at university, I just concentrated on being as normal as possible. I think I got away with it because I could still hear to speak and have conversations.'

When Colin Bennett was diagnosed with RP in his 40s, he says 'I felt pretty awful. It was a bad time – I'd lost my job and my marriage and I was homeless. He says 'The rest of my life has not been easy either. But,'

he adds, 'it taught me to appreciate things – just how good it is to be able to walk along the promenade.'

Unusually for someone with Usher 2, Carol Pollington always knew she had Ushers, as her much older brother had already been diagnosed. She says that it has sometimes been hard to cope – especially as she was growing up in the 1940s, 50s and 60s, when there was 'an appalling lack of information. Given the choice I would rather not have known I had Usher when I was growing up. I grew up with a nightmare

hovering over me. Even if I would not let myself think about it, I knew it was there. The reality has been entirely different. There have been bad times but only for very short periods and I have always been so lucky to have a family, including my siblings, who have allowed me to be normal.'

Mandy Priestman agrees, saying that her family were enormously supportive. They were determined that she should get the best possible education, and sent her away to a selective school for deaf children when she was 11.

Diagnosis

Diagnosis of Usher of all types is by observation, as there is no clinical or genetic test available so far. It is not unusual for someone with Usher 2 to be diagnosed with retinitis pigmentosa as an adult, but for the connection with Usher not to be noticed for some years.

There are probably about 3 people with Usher for every 100,000 in the general population. Although we believe that about 40% of people with Ushers have type 2, this is a rough figure as there is a high chance that Usher 2 is under-diagnosed. There are several reasons for this:

- many people with Usher 2 are such good lipreaders that the doctor diagnosing their RP may not suspect that they have a hearing loss
- most people with Usher 2 attend mainstream schools, where awareness of Usher is much lower than in the schools for Deaf children attended by most Usher 1 people
- there is still low awareness of Usher among doctors.

There is a blurring in the diagnosis of Usher 2 and Usher 3, so that someone who has Usher 3 may initially have been diagnosed as having Usher 2. They are usually born with normal sight and hearing, and develop hearing and visual impairments in adolescence or later. Their hearing may continue to deteriorate. Usher 3 is much rarer than Usher 2.

Colin Bennett's family were supportive in a different way. He says 'Disability was not something that was tolerated in the family. We were six boys – weakness was not encouraged.' Though there have been times when he's wished for a more protective family, he feels that it would have been a mixed blessing – because no compromises were ever made, 'it made me get on with my life, so I had a normal education and career, married and had children.'

“ Did I hide it? Oh yes – I never spoke about it to anyone”

This powerful desire for a normal life is echoed by everyone. But there is a flipside, as it may also lead people to hide their impairments for as long as possible. One of the realities of RP is that it increasingly makes it harder to get about safely. Eventually many people with Usher use a cane or a guide dog, or both. But accepting that the time has come to use a cane or a dog can be a major hurdle. It means telling the world at large that you have a

Nadeem Alvi



visual impairment, which you may have more or less successfully concealed for years, both from yourself and from others.

Many people with Usher 2 treasure its invisibility, regarding their sight and hearing problems as strictly personal and private. Carol Pollington admits this freely: 'Did I hide it? Oh yes. From an early age I never spoke about it to anyone. I knew I had it but it wasn't affecting my life – it was nothing to do with anyone else. I did not then, nor do I now, feel that self pity gets you anywhere. Also, I felt that if I talked about it, then it was real.' →

Why do people get Usher?

Usher Syndrome is passed on genetically. For someone to get Usher, both their parents must pass on the same Usher gene. Their parents will probably not have had Usher themselves, but will have been carriers.

A carrier is someone who has only one copy of the mutated Usher gene. They are unlikely to be aware that they carry this Usher gene.

The Usher gene is called a recessive gene because it only causes the disease to develop when two matching genes are present. Everyone carries at least two or three recessive genes, but this does not affect their health. These recessive genes only become important if someone with a particular recessive gene has children with someone with the same recessive gene. This is extremely unlikely, because we have over 100,000 genes.

There are several types of Usher, each with a different mutation to the gene. If parents have different types of Usher, their children will not develop Usher.

Can we test genetically for Usher?

Genetic testing can show whether a sibling who does not have Usher carries the same recessive gene as their sibling with Usher. It cannot be used on someone unrelated to the Usher person, such as a potential marriage partner. Genetic testing is not used in the diagnosis of Usher.

Inter-family marriages

Some parts of the population may be more vulnerable to Usher because of their preference for marrying within a family. After he was diagnosed with Usher 2, Nadeem Alvi, a British Asian who has Usher, found that there has been a lot of inter-marrying in his family. As a result, he and his family have got involved in genetic research into Usher.



Like Carol, Des Lucy has an older sibling who was diagnosed before him. He too avoided acknowledging his Usher for years. 'I heard about his diagnosis, but being ignorant – or complacent – I thought I was all right.' Eventually though Des too had to seek a diagnosis as his sight worsened.



In common with many people, Des's decision to seek a diagnosis was sparked off by a change in his vision – in his case, he found that he could no longer see the VDU screen at work in bright summer light. One of the effects of the diagnosis was purely practical – he had to give up driving, which hit him

“I took a good look at myself and knew I had to do some

“ I have early memories of my mother waving her finger to my left and right and asking if I could see it”

Carol Pollington was born in 1939. She was originally from Manchester, but moved to Carmarthenshire when she retired age 63. She worked as a secretary until her second daughter was born, and went to university almost 20 years later to take a degree in psychology. She then specialised in deafblind work for social services and Sense. She is married, with two grown-up daughters.

'I've always known I have Usher, despite having very mild symptoms, as my brother who is fifteen years older than me had much more severe Usher 2. I have early memories of my mother waving her finger to my left and right and asking if I could see it, and asking me to listen to the ticking of a watch – which I could hear.

I'm pretty sure people didn't guess that I couldn't hear and see well. From my early teens I realised I didn't hear as well as everyone else because if I was at the back of the school

assembly hall I couldn't hear all the headmaster was saying, but otherwise I had no problems. I noticed in my early twenties that I often didn't see that someone was offering me a cigarette, and was aware of nightblindness. I had problems with sun glare in my mid-thirties, and real problems in my mid-forties – mainly due to cataracts that I wasn't aware of. They were diagnosed late and I was virtually blind for three years. By this time my hearing was deteriorating.

I had been told by my doctor that I would go blind and deaf ... when I was older. But when is "older"? I also believed, because no one had told me otherwise, that I would be completely deaf and blind. My brother was far worse than me and I did not want to be like that. So I pushed it all to the back of my mind and would not think about it. I was frightened. I tied it up in a box and now and then I would carefully undo the bow and look inside and then quickly tie it up again.

hard: 'At the time I felt that was the biggest thing.'

Des still misses driving. He looks back at the days when he could just say that he was popping out to see his parents, or down the tip – everyday things – and says 'I have lost the freedom to do things my own way.'

"I'd advise anyone to have a cane. It's quite good for chatting up women too – it's an ice-breaker!"

Mandy Priestman has never been able to drive. In fact she met her husband because he was asked to give her a daily lift to work when she first started at the British Library. She was spurred on to get a guide dog by their then nine-year-old daughter Holly. 'She started to complain about having always to go clothes shopping with her dad tagging along.' Holly also wanted her mum to be home after school, but Mandy had to wait at work for Paul to drive her home at the end of the day. The arrival of her dog

Unis meant that Mandy could go home by bus at lunchtime, and was mobile on her own again. 'The regained independence was a lovely feeling.'

Not everyone wants a dog. Colin Bennett doesn't like them much, but loves his symbol cane, having resisted it for years. He no longer has to explain why he's falling over other people's children, and it stops others bumping into him. 'I'd advise anyone to have a cane. It's quite good for chatting up women too – it's an ice-breaker!' He doesn't mention what the women of Finland, Holland, Denmark and Sweden made of him and his cane when they travelled through their countries this summer by bus and train. But mobility clearly isn't a problem in daylight.

On the other hand Colin terribly misses the ability to read: 'My eyesight is deteriorating remorselessly. The biggest problem is that I can no longer read, and I'm a voracious reader. I can only read 18 point bold now. So I have to pay someone to read for me, which takes most of my money.' →

"We don't want to be a community. I'm a citizen of the UK"

thing"

As Usher progressed I hid it from my husband and family. As it grew worse, I still would not talk about it. I was not denying it, I just was not prepared to talk about it. It was my problem and I would deal with it.

In my forties, my brother died of lung cancer. I said to myself that I had to learn from his life – he had little social life, had not married, spent most of his time in his room listening to the radio on headphones – he would not use a hearing aid so conversation was very hard. I finally took a good look at myself and knew I had to do something. I contacted Social Services. I was offered cane training – which stopped after a couple of lessons when the trainer realised I could not hear a stream beneath a bridge! – and Braille lessons, which were brilliant. I had learnt to read again. All I needed now was mobility so I applied for and got my first guide dog in 1990. As I did not want to fail the assessment, I knew I had to do something about my hearing – so I finally got a



Carol Pollington

hearing aid, which I have used ever since.

I then attended college, got three A levels and a teaching certificate, started teaching Braille at college, went on to university, got jobs right away and my life had turned around.'

"I pushed it all to the back of my mind – I was frightened"

“ You meet people with similar conditions, and I enjoy the social meetings ”

Colin’s readers have to plough through mountains of documents, as he is a vocal campaigner for disability rights and other issues, regularly taking on his local council. He’s an active member too of the National Federation of the Blind, as well as belonging to Sense’s Hearing and Sight Impaired group (HSI UK), which brings together people with Usher 2 or 3. ‘It’s for people who live in the hearing and seeing world and who use English.’ He adds ‘Without Sense I wouldn’t have known any other Usher people – it’s a tremendous way of ameliorating isolation.’ Des Lucy belongs to the HSI too. He says ‘I get the support I need from the HSI. You meet people with similar

conditions, and I enjoy the social meetings.’

But both Colin and Des – and most other people with Usher 2 – are absolutely clear that though they need to know other people with Usher, and their Usher is an integral part of their personalities, their Usher and their impairments are not what define them. In this, they are unlike the majority of people with Usher 1, who often identify themselves as Deaf people with Usher. As Colin – always a plain-speaker – says, ‘We don’t want to be a community. I’m a citizen of the UK.’

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Sense services for people with Usher

Information, advice and support

The Usher team based in London has a wealth of experience of Usher syndrome and has known many individuals with Usher, and their families, for many years. They offer advice and information about a wide range of subjects – everything from the effects of Usher to moving into higher education and employment.

In addition, Sense Outreach Workers based around the UK, support individuals and their families. This might include carrying out assessments, helping someone to access education or leisure opportunities, linking them into other sources of support or advocating with local authorities on their behalf.

Courses and training

Training is tailored to meet the needs of different groups – from basic Usher awareness to providing training for teachers, hearing therapists and other professionals. Specific training is also provided to centres and groups – for example on communication through touch or on environmental modification.

Conferences and social events

Weekend conferences are organised from time to time. These have included a weekend for hearing and sight impaired people, a weekend for parents and a creative arts weekend, as well

as the recent European Acquired Deafblindness Conference – which you can read about on page 18.

Research

Sense initiates and been closely involved in research activities – for example, recent research into the development of hands-on signing. Currently it is co-ordinating the National Collaborative Usher study – the first of its kind. See page 15 for details.

Sense supports two branches which are largely organised and run by people with Usher themselves:

Usher UK

Brings people with Usher together to offer information and advice, provide mutual support, campaign for change and to have fun through social activities and events. See Usher UK’s website – www.usheruk.org.uk – for further information.

The Hearing and Sight Impaired Group

This is a self-help and support group for people with impaired sight and hearing – most of whom are hearing aid users. It has regular meetings and social events.

For more details about Sense’s Usher services tel: 020 7272 7774, text: 020 7272 9648, fax: 020 7272 3862 or email: info@sense.org.uk