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Usher syndrome, an unseen/hidden disability: A phenomenological study of adults across the lifespan living in England.

Usher syndrome is a rare, unseen, inherited genetic disability and is a main cause of acquired deafblindness in the United Kingdom. The aim of this descriptive phenomenological study was to develop an understanding of the experiences of diagnosis and living with Usher syndrome, from the perspectives of adults living in England. The social model of disability and Erik Erikson's identity development theory provided theoretical perspectives pertinent to deafblindness/Usher syndrome. Individual interviews were conducted with 20 adults, using participant-led multi-sensory communication and interview methods. Communication methods included British sign language, deafblind manual, hands-on signing and clear speech. Interviews were conducted, face-to-face, telephone, email and Skype. Following thematic analysis, the findings revealed that this unseen sensory impairment affects everyday life and future plans, significantly impacting on individuals and their families, friendships, new and existing relationships. These findings are discussed from the perspective of the social model of disability and identity development theory.

Keywords

Unseen disability, Usher syndrome, D/deafblindness, descriptive phenomenology, Social model of disability, Identity development theory

Points of interest

- The literature review demonstrates the limited previous research about living with Usher syndrome.
- The study findings provide an understanding of what life is like for people who experience diagnosis of, and are living with, an unseen condition, such as Usher syndrome, from the perspective of adults living in England.
- The study findings revealed that although Usher syndrome is not life threatening, the condition affects everyday life and future plans.
- A diagnosis of Usher affects both new and existing relationships for people across their lifespan.

Introduction

Usher syndrome, which is one of the main causes of acquired deafblindness in the United Kingdom (UK), is a rare inherited genetic condition (Sense 2014). This article reports on a qualitative study, which aimed to increase understanding about experiences of people living with Usher syndrome. The social model of disability and Erik Erikson's identity development theory provided theoretical perspectives pertinent to deafblindness/Usher syndrome.

The UK Equality Act 2010 extended the range of discriminations, referred to as protected characteristics, which are protected by law; one of these is disability (Johns 2020). The Act states that a person is disabled if they have a physical or mental impairment that has a substantial and long-term negative effect on their ability to do normal daily activities. Negative attitudes still exist despite legislation to

challenge public attitudes and discrimination against disabled people (Aiden and McCarthy, 2014). Some impairments are not obvious and as such are referred to as ‘non-apparent impairments’ (Evans 2019, p726), unseen or hidden disabilities (Couzens et al. 2015). Hidden disabilities can be developmental or acquired but rarely have a ‘physical presentation’ (p24), which can result in lack of awareness and consequently lack of support (Couzens et al. 2015). People with Usher have previously referred to their condition as a ‘hidden disability’ as their impairment was generally not obvious from appearances (Ellis and Hodges 2013, p177). With Usher syndrome there is no physical presentation, unless a person uses sensory equipment, such as a red and white symbol cane, which denotes a sight and hearing impairment. Nonetheless, there are tensions between whether or not to display an unseen impairment. Displaying an unseen condition by, for example, using a guide dog or red and white cane, can lead to discrimination and prejudice. However, choosing not to display an unseen condition can lead to confusion or misunderstanding of behaviours, such as bumping into people or objects, due to a visual impairment.

This article refers to both small ‘d’ and capital ‘D’ D/deafblindness, which are relevant to perspectives on disability. Capital ‘D’ Deafness applies to a person who would not consider themselves to be disabled, but part of a cultural, social, and linguistic group (British Deaf Association 2015). Deaf people consider being Deaf as part of their heritage and are likely to be proud to be Deaf (Evans and Whittaker 2010). Deafblind with a capital ‘D’ refers to people who consider themselves to be culturally Deaf but also experience sight impairment. Small ‘d’ deafness refers to a person who was previously a hearing person or was born deaf but grew up in a hearing family with

oral communication being the primary communication method; they would most likely consider deafness a disability. Likewise, people who are deaf and then experience sight impairment, termed deafblind, usually consider their sensory impairment a disability. People are regarded as D/deafblind if they have combined visual and auditory impairment resulting in difficulty with communication, information and mobility (Deafblind Services Liaison Group 1993). When D/deafness or blindness is combined, the complexity increases especially for communication, mobility and access to information (Simcock 2017, Kiekopf 2007). Consequently, people who experience deafblindness are noted to be 'some of the most vulnerable members of society' (Simcock and Manthorpe 2014, p2325).

Usher syndrome

People who have Usher syndrome experience sensorineural hearing impairment and progressive sight impairment (Wallber 2009), which is caused by a degenerative retinal disease: retinitis pigmentosa (RP). The primary symptoms are night blindness and peripheral vision impairment, which affects people who are initially sighted but whose vision gradually, deteriorates because of RP (Wallber 2009). Organisations such as Retina UK, a UK charity, work for people affected by inherited sight impairment such as RP and invest in medical research for treatments (Retina UK 2018). However, there are currently no proven treatments to decelerate or stop deterioration.

There are three clinical types of Usher syndrome with definitions for each type. In type 1, people are born profoundly d/Deaf and start to experience reduced vision as a child, sometimes experiencing difficulties with balance, walking and the vestibular

system (Pennings et al. 2003). In type 2, people experience reduced hearing but are not born d/Deaf; there does not seem to be obvious balance difficulties. Later in life people with Usher syndrome type 2 start to experience sight impairment (Sadeghi et al. 2004). In type 3, people may not realise they have Usher until later in life because there are no obvious hearing, sight, or balance impairments, rather the impairments occur gradually; some people may think sensory impairment is due to advancing years and do not realise they have Usher (Moller et al. 2009). Regardless of the type, each person's experience is unique, and the prognosis is unpredictable; a person could become blind very quickly or they might still see to read at 50 years old.

Usher syndrome is degenerative and is detected when a person, or the parents/caregivers of children or young people, notice deterioration in vision and seek advice. Whilst Usher syndrome is manifested by various hearing impairment levels or D/deafness from birth, this condition progresses to additionally affect sight. The combined hearing and sight impairment is caused by gene mutation and whilst research has engendered an advanced understanding of genetic causes of Usher (Yoshimura et al. 2015), this has not provided everyday practical assistance for people who experience Usher or their families (Cohen et al. 2007). Usher syndrome is a recessive disorder which means that a person must inherit a change in the same gene from each parent for them to have Usher (National Institute on Deafness and other Communication Disorders 2017).

Most literature that discusses Usher syndrome is clinical (Cohen et al. 2007, Moller et al. 2009, Sadeghi et al. 2004) and does not consider psychological or social

aspects. There were only five studies identified that focused on living with Usher syndrome (Ellis and Hodges 2013, Damen et al*.* 2005, Côté et al*.,* 2013, Wahlqvist et al*.* 2013, Högner 2013) and one further study focused primarily on Deafblindness, with some reference to Usher (Kyle and Barnett 2012). The literature review revealed that there had been limited methods used to study the experience of living with Usher syndrome. In the UK, there have been only three relevant research studies since 2005 (Damen et al*.* 2005, Kyle and Barnett 2012, Ellis and Hodges 2013). Damen et al*.*'s (2005) European study, which included the UK, identified that further research was needed concerning support for people with Usher syndrome to enable safe independence.

Due to the limited literature that focused on people's experiences of Usher syndrome, literature was also reviewed about deafblindness and sensory needs more generally. There were four main themes identified: the impact of diagnosis for people with Usher syndrome, the support and intervention for people with sensory impairment, the psychological impact for people with sensory impairment, and safety and risk for people with visual impairment. Of the studies that focused on support and intervention for people with sensory impairment (Kyle and Barnett 2012, Damen et al*.* 2005, Martens et al*.* 2014, Prain et al. 2010, Prain et al*.* 2012, Hadidi and Khateeb 2014, Argyropoulos and Thymakis 2015), only two were UK based (Damen et al*.* 2005, Kyle and Barnett 2012). The five studies that explored the psychological impact highlighted the ongoing physical and psychological effects (Ellis and Hodges 2013, Côté et al*.* 2013, Wahlqvist et al*.* 2013, Sheppard and Badger 2010, Högner 2013). As regards safety and risk, Kim et al.'*s* (2014) findings raised concerns about advancing technology, for example, electric low sound cars.

Theoretical perspectives

This study was based within the social model of disability, which arose because of the resistance of disabled activists, such as Paul Hunt, in 1972, and was continued by the Union of the Physically Impaired Against Segregation (UPIAS), which was 'founded by Hunt' in the mid-1970s (Beckett and Campbell 2015, p271). The document 'Principles of Disability' proffered that:

> *Disability is something imposed on top of our impairments by the way we are unnecessarily isolated and excluded from full participation in society. Disabled people are therefore an oppressed group in society.* (UPIAS 1976, p4)

The initial concept behind the social model of disability was that people were not disabled by their impairments but by the disabling barriers they faced, so disability is a socio-political issue rather than an individual one (Bricher 2000, Oliver 2013).

Shakespeare (1992) highlighted that the disability movement broke the connection between the physical body and the social situation; the social model thus distinguishes between impairment and disability. Impairment is bodily or biological, and disability is societal, or a social condition in which society disables people with impairments (Hughes and Paterson 1997, Evans 2019, Jenks 2019). Jenks asserted that 'the lives of people with disabilities are affected by their impairments' (2019, p466), for example, not being able to drive a car. Jenks further highlighted that 'impairment allows people with disabilities to demonstrate to those who are temporarily able-bodied the ways in which the world must be made more accessible',

for example, ‘buses need to be accessible and this is an impairment-based claim’ (Jenks 2019, p466). However, it could be argued that not all experiences fit with the social model, for example, where hearing impairment has been seen as medical and requires curing or treating, medical advancement and technology have made it possible for some people who may never have had the opportunity to hear, to attain a level of hearing through cochlea implants [An electronic medical device that replaces the function of the damaged inner ear]. The resulting impact of this medical treatment on their hearing impairment may then reduce the disability experienced.

The study was also informed by identity development theory, which was developed by Erik Erikson, a developmental psychologist and psychoanalyst who trained under psychoanalyst Sigmund Freud (Kroger 2007, Forber-Pratt 2017). Identity development could be defined as individuals coming to have an understanding of themselves, within the context of their experiences (Kruger 2007). Erikson described ‘both the developmental function and the individual differences in development’ (Dunkel and Harbke 2017, p58) and produced one of the ‘few human development theories that propose a lifespan perspective’ (Goodcase and Love 2017, p355).

Erikson’s life span perspective comprised an eight-stage cycle of development (Knight 2016) starting from infancy and ending in older adulthood and death and necessitating resolution at different stages of the life span (Erikson 1982, 1998). Identity development theory is therefore relevant to Usher syndrome, which is a progressive condition across the lifespan, with potential to affect identity development at different stages. Research suggests that stable identity makes people more resilient, reflective, and independent in making key life decisions (Flum

and Caplan 2006, Kroger et al. 2010, Verhoeven et al. 2019) with the process of reflection being an integral element of the ‘construction of personal identity’ (Flum and Caplan 2006, p105).

A critique of identity development theory is that there are socially constructed expectations and values of our society and if a person does not meet these expectations, they may feel they have failed (Crawford and Walker 2010, Arnett 2000). However, there are milestones to reach from the day that a child is born (Crawford and Walker 2010), with their growth and development being mapped to gauge development and prevent any medical issues being overlooked.

The research

The research questions were:

1) What is the experience of being diagnosed with Usher syndrome?

2) What is life like for people who are diagnosed and live with Usher syndrome, on a daily basis?

The study was conducted within a constructivist paradigm using descriptive phenomenology, as the aim was to develop an understanding of the experiences of diagnosis and living with Usher syndrome. Rogers and Pilgrim (2005) argued that constructivism holds a fundamental belief that reality is not already established and awaiting discovery, but rather is transitional as a result of our actions as humans and thus reality is constructed. Use of phenomenology enabled in-depth exploration of complex individual experiences of a phenomenon (in this study, living with Usher

syndrome) and then the analysis of the composite experiences of individuals to appreciate the essence of the phenomenon for all individuals. Descriptive phenomenology focuses on participants' experiences from 'their viewpoints as fully as possible' and aims to 'capture exact and deep layers of life', creating a thorough description of the phenomenon (Sorsa et al*.* 2015 p9). In terms of Erikson's (1982) identity development theory, descriptive phenomenology enabled participants to relate their developmental experiences associated with their impairment. The approach also gave participants opportunity to describe their experiences of impairment within society, as they talked about their everyday life, which could be later reviewed in the context of the social model of disability (Oliver 2013).

The UK sensory charity 'Sense', who work for and support deafblind people (Sense, 2019), was gatekeeper for the study and supported recruitment from the Sense database and the two Usher groups, one for Hearing and Sight Impaired (HIS) and another group who were mainly British Sign Language (BSL) users. Whilst most participants were introduced through Sense, four participants were recruited via snowballing, where current study participants recommend others. Purposive sampling, where the researcher chooses participants with a particular phenomenon e.g., living with Usher syndrome, was used to include people who would help the researcher better understand the condition and answer the research questions (Cresswell, 2013). Individual semi-structured interviews were conducted with 20 participants (n=10 male, n=10 female) aged 18-82 years, with different Usher types (n=2 type I, n=17 type 2, n=1 type 3), and of the following nationalities: British (n=15); Asia (n=2), British Caribbean (n=1), Irish (n=1) and Portuguese (n=1).

Semi structured interviews are compatible with phenomenology because they 'explore the richness of everyday life as it manifests to those who experience it' (Campbell and Scott, 2011 p6). Interviews were conducted using various communication methods and through face to face, email, Skype (with camera on or off) and telephone. The communication and interview method were chosen by the participant, not the researcher, leading to a change in power dynamics and unexpected equalising (Evans, 2017). For example, although the researcher had sensory communication skills, it was not her first language, therefore the balance of power was altered (Evans, 2017). This innovative way of combining participant-led communication and interview method, referred to as 'Multiple Sensory Communication and Interview Methods' (MSCIM), has been described in detail previously (Evans, 2017).

The interview topic guide linked to the study aim and concentrated on gaining participants' experiences of living with Usher syndrome. The interview questions were adapted according to communication used, for example where a person used BSL; the word order of questions was altered to reflect BSL grammatical structure. The interviews ranged from 40 minutes to one hour 40 minutes and were transcribed by the researcher; the length varied as interviews were participant led, so if a person wanted to talk longer the interview was not cut short. There were no incentives; participants contributed freely and willingly. Participants were given pseudonyms to ensure anonymity and confidentiality, while giving voice to individuals. The different communication methods used were: clear speech; written communication; visual frame BSL, which is signing within the individual visual field of the person with Usher syndrome, to enable use of residual vision; and hands on BSL in which the person

places their hands over those of the signer to feel the signs produced, and/or deafblind manual alphabet, which is a method of spelling out words directly onto the person's hand, as each letter is denoted by a particular sign or place on the hand. The data were captured using various methods: email (written), video camera (visual) and digital voice (audio) recordings. Data from email were already in written format and the researcher followed the descriptive phenomenological approach of taking participant experiences as a given. Data collected during face to face, Skype and telephone interviews were audio recorded and transcribed (type-written copy) verbatim. Data from participants who communicated using visual frame BSL, hands on BSL and/or Deafblind manual were both visually and audio recorded and included the use of voice over, which is where a hearing person used clear speech to translate visual communication such as BSL.

Data analysis

The data were analysed using Braun and Clarke's six phases for thematic analysis, which include identifying, analysing, and reporting patterns (themes) within data (Braun and Clarke, 2006). Thematic analysis aligned with descriptive phenomenology as participant experiences were central and the process enabled an understanding of how participants constructed and made sense of their experiences. Additionally, thematic analysis was compatible with the quality and diversity of the data and promoted a transparent and rigorous approach (Braun and Clarke, 2013, Clarke and Braun, 2017). The findings were then reviewed from a perspective of the social model of disability and Erickson's identity development theory.

Ethical considerations

The study was approved by a University Research Ethics Committee; confidentiality and anonymity were central to the research process. Each participant was allocated an anonymous identifier, any identifying details were removed during transcription and no personal details were included. Pseudonyms were asigned for reporting the findings. Participant information sheets and consent forms were provided and individual communication requirements were considered, for example the consent form and participant information sheet were read to people whose sight impairment prevented them from reading it, provided in large print if required, and translated for people who used sign language. Each person was given details for them to consider within their individual time frame and then provided written consent to participate. All data collected were stored securely on an encrypted device**.**

Findings

The findings are next presented in four themes: Diagnosis is the start of the experience; Familial relationships across the lifespan; A sense of belonging; Experiences of professional support. These findings are discussed in relation to theoretical perspectives and previous research in the section 'Discussion and conclusions'.

Diagnosis is the start of the experience

The age of diagnosis varied considerably: a few participants were diagnosed as teenagers; most others in early adulthood but some were diagnosed in their 30s or later. For those who were younger at diagnosis, their parents were involved in how, and whether, they were told. One participant related the importance of telling a child about their diagnosis or they may jump to the wrong conclusion:

I approached my parents and said I had been thinking I was going to die. They were quite upset obviously and horrified and I said to them look you have to tell me everything you cannot hide things from me….I thought it's only my eyes I am not going to die after all. (Kate)

Prior to the diagnosis of Usher, all participants experienced hearing impairment ranging from mild to profound, however, additional visual impairment led to the diagnosis of Usher syndrome. Most participants had experienced impaired night vision or nightblindness, where people cannot see well at night or in dark conditions such as the cinema or at a club (Sense 2014). One participant commented:

I know and understand that nightblindness is probably the earliest symptom to manifest itself and didn't realise that I didn't see as well as my peers at night or in different lighting situations, that my eyes took longer to adjust, hence more accidents. (Pam)

While nightblindness was an indication of visual impairment, most participants were not aware of the severity of their condition. For example, Sara, who had experienced sight impairment prior to diagnosis, thought '*there is nothing wrong you know what I mean, it's just one of those things I was just a bit clumsy*'. Prior to diagnosis, many participants described clumsiness, and several were involved in accidents, which they now realised was due to their increasingly limited vision:

I did have a couple of silly little car accidents and that was only purely because I did not see the car in the field of my vision and went into it. Silly but that's erm at the time I didn't really twig I wasn't seeing very well. (Jeff)

Despite pre-diagnosis deafness and experiences of visual impairment, the reality of being diagnosed with a degenerative condition such as Usher syndrome, with the complexities of dual sensory impairment, began at the point of diagnosis.

Participants responded differently to their diagnosis; some described relief at knowing but others expressed anger followed by sadness. Profound shock was also expressed, with the realisation that one sense being impaired, for example, hearing, can lead to personal vulnerability, to lose both affects everyday life and future plans:

> '*I felt like someone had punched me in the stomach […] you have grown up being deaf and you always think there is a worse thing that could have ever ever happened and that's your sight! Because you rely on your sight so much for communication if you're deaf or partially deaf for lip-reading, sign language all that sort of thing. You think oh God if I lose the sight life won't be worth living.* (Sara)

Some participants experienced feeling low or depressed and some had suicidal thoughts, requiring psychiatric admission in some instances, and long-term use of anti-depressants. These experiences highlight the psychological impact when a person is diagnosed with Usher thus counselling services should be available following diagnosis.

Factors affecting a person's response to their diagnosis included their age, gender, individual personality, family support and familial attitudes. Receiving a diagnosis of Usher had practical, emotional, and physical implications. While some participants became more cautious following diagnosis, others had a different outlook:

Make most of life be happy because there are worse things happening than losing sight and hearing. It's not the end of the world it really isn't erm I don't know I've learnt to live with it. I've done abseiling I've done everything. (Ruth)

Whilst participants shared varying experiences post diagnosis, for everyone, there was the realisation of the implications:

Growing up in the past year or so has made me realise some of the restrictions that go with Ushers, although I have lots of coping strategies I will never drive, walk anywhere on my own, read a book again and that makes things different. I am also over reliant on my parents, teachers and support workers and that can be hard. (Tia)

Familial relationships across the lifespan

The theme of 'familial relationships across the lifespan' included the impact on families when a child is diagnosed with Usher, experiences of being a parent who has Usher, and relationships and family support. Some participants described how their parents found the diagnosis of Usher difficult, particularly as the condition is genetic/hereditary and the parents may be carriers. The fact that Usher syndrome is a genetic/hereditary condition arose frequently during the interviews, one participant relayed:

The Usher gene was passed on to me because both my mum and dad were carriers of the gene but did not have the disease itself. (Harry)

The word 'carrier' used by several participants means 'a person or animal that may transmit a disease or a hereditary characteristic without suffering from or displaying it' (Thompson 1996, p144). Where participants mentioned that their parents were

carriers of Usher, they did not appear to attribute any blame. While the parents are often completely unaware that together they have a gene that can lead to their child having Usher syndrome, the realisation can lead to guilt. These emotions could impact on communication and relationships:

> '*If I'm honest I think my parents have found it really hard to cope with what has happened to me. Obviously it is genetic and I think guilt has played a large part in it all […] I've found it hard to express my difficulties as I have not wanted to upset them. Whilst they definitely want what is best for me, it has been hard for us to support each other when we have each been struggling with it individually.* (Tia)

Overall, participants had differing experiences of family support, ranging from overprotectiveness to a lack of support. They identified how attention and support in the family was given to the child with the highest needs, as parents had limited time and resources. Of the 20 participants interviewed, five had siblings with Usher, thirteen had siblings without Usher and two had no siblings. Some participants discussed how supportive their siblings without Usher were to them.

Two individuals described how, when they had children, professionals considered they would be unable to cope as parents due to their sensory requirements. However, with family support, these mothers were able to keep and bring up their children. These two mothers were now 82 years and 69 years of age respectively and so their experiences must be contextualised within society's attitudes and legislation of that time. Today, with current UK legislation such as the Local Authority and Social Services Act 1970 s7 (to identify and provide an assessment for people who experience D/deafblindness), Equality Act 2010 and Care Act 2014, parents

experiencing impairment have legal rights and are entitled to an assessment of the needs and protection against discrimination on the grounds of their impairment. In this study there was no mention of whether the diagnosis impacted any participants' decisions to have children, thus this may be an area for further research.

For some participants knowing they had a degenerative but unseen condition affected how they felt about building new relationships; for others socialisation in certain environments was a barrier and for another maintaining relationships was impacted. One participant shared how he felt about knowing that the condition was degenerative but unseen when forming new relationships:

> '*I had a horrible dark secret, I knew I was diagnosed with RP [*retinitis pigmentosa] *and I knew what RP meant, it was like a horrible burden on my back that I couldn't quite get off. You know it made me nervous, very nervous*'. (Gareth)

Two other participants, Kate and Pam also discussed the impact of Usher on forming new relationships. Although Kate was successful from a professional perspective, she relayed the following experience:

> *'I have not even had a proper boyfriend I might you know gone out once or twice with people but I never had a boyfriend'.*

Pam had a similar experience:

> *'Everybody else had a boyfriend I didn't have boyfriend like in the same way and the thing is people know you are different'.*

Some participants with Usher expressed that they were not 'normal' because of their sight and vision impairment but they did not want to be labelled as being different,

especially as the stigma attached to the sensory impairment could impact on relationships. Some participants explained that they would rather appear drunk than reveal they had a sensory impairment as '*it's quite alright to be drunk, but it's not OK not being able to see'* (Quentin). Usher also affected maintaining existing relationships, one participant conveyed:

> '*It's ongoing all the time, constant changes. The other family and spouses find it difficult to cope with changes'.* (Fred)

A sense of belonging

Two aspects to this theme were interrelationships between communication, culture and community, and friendships and shared experiences. Overall, there were varied experiences of belonging to the Deaf community, but these were closely linked with communication methods used by individuals. Essentially there are two groups of people who experience D/deafblindness: those who are culturally Deaf and whose first language is BSL, and those who previously had speech and were part of the hearing community. However, being part of a culturally Deaf community was not merely a shared communication method, but entailed a shared understanding of culture, linguistics, history, storytelling, and humour, all contributing to a sense of belonging, for example one participant said: *'All Deaf, same culture same, happy together*' (Debra). Even after a diagnosis of Usher, capital 'D' Deaf people saw themselves as culturally Deaf and the shared communication with other people whose first language is BSL created a feeling of belonging.

As BSL is a visual form of communication, acquired deterioration of sight affects communication. Thus, some participants described how being part of the Deaf

community changed with the progression of Usher, as their sight impairment affected communication through BSL. For example, one participant who had felt part of the Deaf community said:

> *'Yes I did but now I don't. Deaf people see things in a different way. Now I can't see that well. I haven't seen any of my friends from [name of school] for a very long time'.* (Ben)

Another participant whose first language, used from birth, was BSL, was educated in a school for Deaf children and had experienced a life-long sense of belonging as part of the Deaf community. However, when her eyesight started to deteriorate, it became harder for her to communicate with her friends as sign language is a visual language. Her friends stopped communicating with her and she became isolated and not wanting to attend the Deaf club, even though she had attended it her whole life. With support from her daughter and a sensory worker, her friends' awareness of Usher and how they could communicate with her eventually resolved the communication difference, so she could continue to feel part of the Deaf community.

The findings also indicated how people living with Usher are united by shared experiences within support networks. Whilst a diagnosis of Usher can lead to isolation, sensory charities helped to overcome societal barriers, enabling meetings with other people with Usher, and empowering people to feel less alone while also learning coping mechanisms from shared experiences. There were specific groups aimed at young people and their parents as a role model peer group. Meeting up with other deafblind people could have a positive and inspirational impact:

'It wasn't until I met the deafblind people and they were all doing things and I thought I want to have a go at doing it. If they can do it I'm doing it and I did. (Ruth)

Some participants travelled long distances to meet other people with Usher with whom they could share their experiences.

For many participants, the use of technology, such as social media, videoconferencing, and email, enabled interaction with people with Usher across the world, providing support and friendship. As social media platforms are available 24/7, they were accessible whenever the person with Usher is ready to seek information or support or meet others with Usher. Technology also enabled wider connections:

'Technology important for someone with Usher it keeps us in touch with the sighted/ hearing world. For me it allows me to write (touch type) read (screen reader) and braille (braille note) easily in a mainstream environment'. (Tia)

The type of technology chosen by individuals depended on their level of sight and hearing impairment.

Whilst technology had great potential to reduce barriers, there were some experiences of where societal failure to use technology acted as a barrier to a sense of belonging. For example, one participant (Quentin) expressed the disabling effects of being unable to hear the eulogy at a funeral because there was no induction loop at the crematorium.

Experiences of professional support

Experiences of professional support affected education, employment, mobility, and socialisation. Thirteen participants had required reasonable educational adjustment to meet their sensory requirements and some shared positive experiences of support received in the partially hearing unit (PHU) in primary school. However, others had less positive educational experiences:

> *'I did have problem hearing and understanding what teachers was saying – Fell behind with my education'.* (Nora)

Others were bullied when they wore sensory equipment such as hearing aids so would not wear them at school, with adverse effects on education.

From a mobility perspective, there were examples of disabling, rather than supportive, attitudes encountered by participants:

> *I have experience that the bus drivers are not always aware how important for deafblind people to get off the stop they need to get off I had a driver refuse to stop at my stop….. This had now put me off travelling on buses on my own which is now two years ago. (Iris)*

The impact of such experiences regarding the opportunities for people with Usher to remain independent cannot be underestimated.

As people are diagnosed with Usher at varying ages and their condition continues to progress, sometimes people who had been in employment became medically retired, earlier than they would otherwise have been. Others had not been able to continue with their career plans at an early stage, for example, Pam had hoped to be a professional dancer but:

I had to give up dancing any way so...so um you know it did affect me. (Pam)

Some people who could no longer work experienced considerable distress, compounded by negative experiences of benefits applications. A new system that used a Work Capability Assessment (WCA) was implemented by the UK's coalition government as part of an austerity programme to reduce benefit costs (Cross, 2013). One participant described the impact on himself:

> *I was forced to go to the job centre; my brother had to take half a day off. I went there with a red and white stick and a personal listener wires I used headphones, she still said to me at the end there "could you please look at the monitor and check what I have written". I started waving my stick around and said I can't see the monitor let alone the text. She said we still think you should apply for work. I don't need all this sorry I put letters in front of her from [specialist eye and Ear, Nose and Throat] hospitals and then she just ignored it all*. (Gareth)

Thus, even though this participant's Usher impairment was evidenced by medical reports, the societal systems and attitudes of professionals posed a disabling barrier to financial support.

Participants described how sensory equipment provision had a significant impact on their lives. For some people, if their sight was more impaired than their hearing, a red and white or white cane was paramount for safe orientation and alerting others to their sight impairment. However, sensory equipment could increase their sense of vulnerability as it made their impairment more visible: '*I sometimes feel vulnerable in*

the street with a red and white stick' (Gareth). Some participants who had experienced profound hearing impairment from birth found flashing doorbells/telephone/fire alarm or pager systems were useful. Several participants had guide dogs which enhanced everyday functioning as well as being companions:

> *I can ...go a bit further than I do and go to unfamiliar places. I can go to familiar places my guide dog knows where to take me.* (Pam).

However, whilst a guide dog was an enabler for mobility and being able to work, it also made visual impairment more obvious to the public and colleagues. Sara shared how she had experienced rude comments at work when people saw the guide dog and realised she was severely sight impaired, for example, one person said: '*I think it's lovely they have given you this job*'. However, Sara felt under pressure to suppress her feelings:

> *I do a lot of tongue biting you know what I mean particularly with a guide dog, I am aware I am one of those public faces of guide dogs for the blind.*

Discussion and Conclusions

The complexity of losing both sight and hearing is described as not impairment of sight *and* hearing but sight *times* hearing (Sense 2014), which highlights how the impairment of a second sense has a profound impact for people affected. Consequently, although Usher/D/deafblindness is not life threatening, it affects everyday life and future plans, as highlighted in the current study, and in previous research (Ellis and Hodges 2013, Kyle and Barnett 2012, Högner 2015, Wahlqvist et al. 2013). Whilst the social model of disability provided the wider context for the research (Oliver 2013, Bricher 2000), Erik Erikson's identity development theory (Kroger 2007) was particularly pertinent to deafblindness/Usher syndrome because

Usher syndrome progresses across the lifespan. The findings are next discussed from a perspective of these theories, with reference to previous research where relevant.

The findings support Erikson's identity development work as, when an individual experiences a life-long, degenerative condition (such as Usher syndrome) there is an inevitable impact on transitioning from early to later life (Eriksson et al. 2020). Most participants were diagnosed as teenagers or in early adulthood, which is an important time for relationships and identity development. The findings revealed how participants' expectations about their lives were altered following their diagnosis, with effects on relationships, careers and independent adulthood, which are usual developments across the lifespan. Thus, the life that people plan prior to the diagnosis of Usher syndrome is often very different to the life that will be their future. For example, learning to drive is a key transition point for many people but participants described how a diagnosis of Usher either prevented learning to drive or led to them being unable to continue driving, which affected their mobility and independence.

Erikson considered that ideal identity development included finding social roles that enable that person to fit into the wider community (Kroger 2007). However, for a person who experiences Usher syndrome, the community they may have been part of is affected by changing sensory requirements, mainly due to the impact on communication. The findings highlighted that if a person is part of the Deaf community and uses BSL as their first language, as BSL is a visual form of communication, sight impairment due to Usher affects communication and thus their

sense of belonging to that community. Similarly, Kyle and Barnett (2012) reported that participants who were deafblind felt less a part of the Deaf community. Nevertheless, in the current study, some culturally Deaf participants still referred to themselves as Deaf after a diagnosis of Usher, not capital 'D' Deafblind or small 'd' deafblind. This notion of self-perception linked to identity was also discussed in Kyle and Barnett's (2012) study where participants who had Usher, but were previously Deaf, referred to themselves as Deafblind, as opposed to having Usher.

Erickson's theories around relationship developments and impact on identity development were illustrated well by the current study's findings, which revealed that a diagnosis of Usher affected relationships across the lifespan including friendships, partners, marital relationships, children, and parents. These findings support those from earlier studies that explored disability over the life course (Priestly 2003), effects on the family when a child experiences impairment (Reichman et al. 2008) and the impact of Usher on existing relationships (Ellis and Hodges 2013). For some participants in the current study, the guilt expressed by their parents, as carriers of the Usher gene, impacted on communication and relationships.

Eriksonian concepts consider the goal is the ability to form intimate relationships while still retaining one's own identity (Kroger 2007). If these negotiations are successful then our thoughts, emotions and personality will develop but a person who encounters difficulties in forming other relationships may experience transitional issues in forming their own identity, thus affecting developmental stages in adulthood (Kruger 2007). Whilst Dunkel and Harbke (2017) acknowledge that there are individual differences in development, the findings particularly revealed difficulties in

forming new relationships, as well as maintaining existing relationships. The participants' sight and hearing impairments impacted on socialisation, while their knowledge of their degenerative condition, not visible to others, posed a barrier to developing new relationships. Two of the female participants revealed that they had never had intimate relationships, but they described success in other life stages, such as in their professional lives. As there does not appear to be previous literature directly relating to how Usher impacts new relationships, this may be an area for further research.

Some participants discussed how charities such as Sense enabled them to meet other people with Usher. The use of technology also enabled them to feel part of a wider community and contributed to their identity development by reducing isolation and increasing inclusion. These findings support previous research with people who have Usher syndrome, for example, Ellis and Hodges (2013) reported that social media was a useful tool for people living with Usher to meet other people with the same condition and not feel alone. Maiorana-Basas and Pagliaro (2014) highlighted that although technology has the potential to reduce isolation and increase independence for people with hearing impairments, accessibility needs to be available in a format that meets sensory requirements and does not pose a barrier.

As previously highlighted when considering the social model of disability, people are not disabled by their impairments but by the disabling barriers faced in society (Oliver 2013, Bricher 2000). Several participants discussed the barriers to independent mobility, including being unable to drive and difficulties with public transport. One participant's experience illuminated how the attitude of bus drivers

had profoundly impacted on her confidence to use public transport; this example highlighted that whilst accessible buses are important (Jenks 2019), without changing public attitudes, people with impairments will still be deterred from using them independently. In an example illustrating disabling attitudes of professionals, two older participants discussed that when they became mothers, there were assumptions that they would not be able to care for their children. Fortunately, legislation now offers protection against such discrimination (Care Act 2014, Equality Act 2010).

Where participants were unable to work, they described poor experiences of benefits applications, in particular, through the Work Capability Assessment (WCA). The UK's WCA has received much criticism, with media reports of individuals who have been declared fit for work and had benefits stopped, against medical advice, resulting in harm, and even death, due to serious financial problems (Warren et al. 2014). Shakespeare et al. (2017) noted that reports from consultants and general practitioners who know the person's health status are superseded by the company administering the WCA. In the current study, a participant gave an example of just such an experience, where letters from experts about his condition were ignored. This experience illustrated how the disabling attitudes of the assessors compounded the impairment experienced from Usher syndrome. Cross (2013) asserted that the WCA has been discredited by disability activists and has had a devastating impact on the lives of disabled people. Furthermore, the process of undergoing a WCA has been found to be a demeaning experience (Burgess et al. 2014); the current study revealed that people living with Usher have had similarly poor experiences.

In the current study, participants described experiences illustrating the stigma associated with sensory impairment. Goffman (1963) defined stigma as 'an attribute that is deeply discrediting' (p3). It could be argued that 'many of the difficulties that people with disabilities face are, in fact, a product of stigma, not of their disability' (Pérez-Garín et al. 2018, p1). Usher is largely an unseen disability, and as such some participants referred to it as being a 'secret', and described their dilemmas about disclosing their condition, particularly when meeting new people, which affected new relationships, as discussed earlier. However, they also identified that the visual display of their impairment, such as through having a guide dog, could lead to discriminatory attitudes from the public or colleagues. Other studies have also revealed stigma associated with impairment (Tsatsou 2020) and with sensory impairments specifically (Pérez-Garín et al. 2018, Hess 2010, Soffer 2019). Pérez-Garín et al. (2018) found that people with hearing impairments reported barriers in leisure activities but people with visual impairments further revealed lack of equal opportunities, mockery and/or bullying, and overprotection. Some participants in the current study experienced over protection from families, but this was probably well-intended rather than discriminatory.

Goffman (1963) theorised that people made adjustments to their social identities to fit in with the imagined community of non-stigmatised people, referred to as 'normals'. In the current study, participants' descriptions echoed the idea that people without Usher were 'normal' and that they wanted to appear 'normal'. Goffman (1963) used the term passing to describe how people would try to pass as 'normal' to avoid stigma. His theory was illustrated in the current study, where participants described how it was preferable to pass as drunk, due to perceived normality, in

contrast with visual impairment, which they considered stigmatising within their age group. Passing can be complicated and both physically and mentally exhausting (Kermit 2019) yet some participants found it preferable as a technique to appear 'normal' rather than divulge their visual impairment to peers. The phenomenon of passing, to avoid stigma, has been observed in different age groups and in people with varied impairments, for example young children with hearing impairments (Kermit 2019) and young people with cerebral palsy (McLaughlin 2017).

One of the reasons that Usher is not always recognised as a serious debilitating condition may be because it is mainly unseen (Sense 2014, Petersen 2006), unless the person uses sensory equipment or has a guide dog. Prain et al. (2012) identified that even where workers have some sensory expertise, services for people with sensory impairments can be lacking, with disabling effects. The UK's guidance and legislation (The Special Educational Needs and Disability Regulations 2014, Education Act 2011) and special education needs under part 3 of the Children and Families Act 2014 outlines that local authorities and educational provision must take into account a child's sensory or special educational needs (National Deaf Children's Society 2020). Argyropoulos and Thymakis (2015) proffered students with a disability who are well equipped, are better able to achieve their educational and vocational goals; although their research was conducted with students with multiple disabilities and visual impairment, these principles would apply to students with Usher. Participants in the current study had mixed educational experiences but some had experienced effective adjustments which helped them to achieve educationally.

To conclude, the aim of the study was to develop an understanding of the experiences of diagnosis of and living with Usher syndrome, from the perspective of adults in England. Their experiences illustrated how, from a social model of disability perspective, the impact of their combined visual and hearing impairments was often compounded by their experiences in society, particularly through perceived stigma and discriminatory attitudes, which affected their independence. Participants also gave examples of their strategies to pass as 'normal' and thus avoid the stigma associated with Usher. The findings supported Erickson's social identity development theories, as a diagnosis of Usher syndrome affected identity development across the lifespan, impacting on their sense of belonging to a community and relationships, both existing and new. Throughout this study it was clear that people diagnosed and living with an unseen condition such as Usher syndrome want awareness raised and support to empower them to live with the everyday practicalities of living with Usher and to achieve their goals across their lifespan.

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