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What is the lived experience for people with Leber Hereditary Optic Neuropathy?

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## Abstract

This study aimed to hear the voices of seven people living with Leber Hereditary Optic Neuropathy (LHON), to examine their lived experience. The participants were recruited through a website and social networking sites designed for this community; they discussed their experiences in semi-structured interviews. The analytical process was informed by Interpretive Phenomenological Analysis (IPA) principles. This process illustrated a lived experience that was initially consumed by psychosocial losses and conflicts around identity. However through pragmatic and resilient attitudes the participants developed practical methods and emotional coping mechanisms that permitted their adjustment to sight loss and progression in life. Consistent with other studies, the fundamental finding was that the associated feeling of loss and frustration are chronic if, episodic, throughout life.

## Key words

*Visual impairment, LHON, lived experience, loss, identity*

## Introduction

### LHON (Leber Hereditary Optic Neuropathy)

Millions of people worldwide are affected by visual impairments and many of these are incurable (Merabet, Rizzo, Amedi, Sommers & Pascual-Leone, 2005). One of these is Leber Hereditary Optic Neuropathy (LHON), which affects about 1 in 25,000 people in the UK (Man, Turnbull & Chinnery, 2002). LHON is a mitochondrial genetic disease that preferentially causes blindness in young adult males. It is characterised by bilateral sub-acute loss of central vision owing to focal degeneration of the retinal ganglion cell layer and optic nerve. It is an incurable condition that leaves people within the criteria for blind registration (Man et al., 2002).

Only 50% of male and 10% of female carriers actually develop the optic neuropathy (Man et al., 2002). It mainly affects young adults in their second and third decade of life (Man et al, 2002 & Kirkman et al. 2009). The current research on LHON focuses on its biology, but provides no insight

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3 into the psychological impact of the disease. Van Senus (1963) briefly addressed the social  
4 consequences for sufferers, in his thesis on LHON in the Netherlands. He gives details on  
5 occupations and the impact on the social position of 208 participants. The results show a spectrum of  
6 social consequences, with a small majority (23%) stating that their status had slightly deteriorated. To  
7 address the gap of literature in this area research into the psychological impact of other forms of  
8 acquired visual impairments have been reviewed to look for possible commonalities. Permeating this  
9 body of literature are the associated psycho-social losses and the impact on identity caused by the  
10 acquired visual impairment.  
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### 20 *Loss*

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22 A key finding in this literature is that the psycho-social losses are chronic, if episodic, throughout life  
23 (Hull, 1990; Hewson, 1997; Hainsworth, Eakes & Burke, 1994; Murray, McKay & Nieuwoudt, 2010;  
24 Moore, Constantino & Allen, 2010). The onset of a visual impairment disrupts how a person feels  
25 about themselves and their ability to act within the world. There is clear linkage between loss of ability  
26 and confidence or self-esteem as shown in the Network 1000 project, which consulted that number of  
27 visually impaired people on a range of issues and found a connection with the loss of independent  
28 living skills and confidence or self-esteem (Douglas, Corcoran & Pavey, 2006). Additionally to  
29 independence the literature shows that people regret the loss of status, hobbies, abilities and social  
30 skills (Devenney & O'Neil, 2010; Moore et al., 2010; Stanford, Waterman, Russell & Harper 2010;  
31 Thurston, Thurston & McLeod 2010). Through practical adjustments, developing emotional coping  
32 mechanisms and supportive networks participants learn how to live as blind people (Baus, 1999;  
33 Fourie, 2007; Murray et al. 2010; Moore et al., 2000).  
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46 Attempts to generalise this adjustment have been made, with authors drawing on Kluber-Ross's  
47 (1969) grief model, to describe this psychological process (Atkinson, 1990; Baus, 1999; Bergeron &  
48 Wanet-Defalque, 2013; Schainholz, 2000). However this model proposes a staged time bound  
49 sequence that does not consider the dynamic, changing circumstances of a person's life experiences.  
50 These have been acknowledged by authors who appreciate that a new experience and how an  
51 individual appraises their ability to manage it can cause an apparent regression in their adjustment  
52 process (Hayeems, Geller, Finkelstein & Faden, 2005; Hewson, 1997; Tuttell, 1994). It therefore  
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3 seems most appropriate to use Hewson's (1997) Episodic Stress Model to explain how people  
4 experience the associated losses and how and why they reoccur. She proposes that the loss of an  
5 ability results in two levels of reaction; primary and secondary. The first refers to the period after  
6 diagnosis when huge emotional and practical demands are placed on the individual, requiring  
7 recognition (an awareness of the condition and its implications) and lifestyle adjustments. The  
8 secondary stresses refer to daily hassles and frustrations that can be dealt with but their culmination  
9 can result in a reaction reminiscent to a primary stress, externally suggesting a complete regression.  
10 However it is proposed that this is overcome by reducing the external stresses to allow coping  
11 mechanisms to deal solely with the manifestation of the secondary level stress.  
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### 21 *Identity*

22 It is salient to acknowledge the role identity plays in this adjustment process. Hayeems et al. (2005)  
23 found that behavioural, lifestyle and psychological adjustment is contingent upon shifting to a blind  
24 identity. However unlike Baus (1999) and Dale (2010), Hayeems et al. fail to recognise the changed  
25 relationship with the world. In Dale's narrative exploration of the experience of living with a visual  
26 impairment and its impact on identity, a participant described how it was not a lack of sight but  
27 relationships with sighted people that affected their identity. This reflects the reality that although a  
28 visually impaired person may be able to master the practical limitations of their disability, through the  
29 continual use of assistive technologies and acceptance of support, the consequence is being instantly  
30 categorised as a disabled person with limited capabilities.  
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42 The conflict between positions, caused by the sporadic use of assistive devices was well illustrated by  
43 Dale (2010). One participant described feeling like a fraud because he would keep his white cane in  
44 his bag unless he felt a situation, like a busy street, required it. Conceptually this could be because he  
45 knew that the public perception of a blind person was someone who was in complete darkness. When  
46 he was using the white cane he was fitting that image but once he returned it to his bag he knew he  
47 was then in opposition to this image.  
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55 The above highlights the dilemma between concealing blindness to avoid the stigma and revealing  
56 blindness to receive full support. For the visually impaired who have residual vision the question is  
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3 more prominent. Senus (1963) reported how, when the need to read or see things in detail was  
4 removed, people with LHON weren't disabled. He based this statement on the mobility and  
5 independence that people can achieve through mastery of their peripheral vision. Many of the  
6 participants in his study were proud that they could hide their blind identity. Despite the improvement  
7 in society's image of disabled people, the literature would suggest that the public's perception of  
8 blindness still has a negative stigma.  
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16 Studies of the lived experience for people with acquired visual impairments paint a picture of great  
17 loss initially, but through mastery of the limitations, the negative emotions can be suppressed.  
18 However the self's reflection within society will always allow this to resurface. Hewson (1997)  
19 suggests that the process of dealing with the loss cannot be generalised, or placed within a grief  
20 model, because the appraisal of the impact is idiosyncratic and dependent on various factors.  
21 There is a dynamic process of adjustment to living with a visual impairment and a key theme is the  
22 complex issue of identity. Authors see this as pivotal to learning how to function as a blind person  
23 (Hayeems et al., 2005; Zaborowski, 1997). The visual impairment forms a new position of the self. Its  
24 initial prominence is overcome through mastery of the limitations of the disability, but it is always  
25 present. Decisions about concealment of this identity are influenced by the reactions of sighted  
26 people, which can create internal conflict for the visually impaired.  
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38 The current study has explored the experiences of people living with LHON, examining their  
39 perceptions of the associated losses, how it has impacted upon their identity and the journey of  
40 adjustment to living with a visual impairment.  
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## 45 **Method**

### 46 *Methodological stance*

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48 Qualitative data were collected from seven participants using narrative semi-structured interviews,  
49 informed by interpretive Phenomenological Analysis (IPA) principles (Smith & Eatough, 2007; Smith &  
50 Osborne, 2003; Willig, 2008). This approach gave the participants the opportunity to speak openly  
51 and frankly about their experiences of life with LHON. This was deemed to be the most appropriate  
52 style of analysis because the research was aimed at explicating the essential meaning the  
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3 participants are making of their conscious worlds. Narrative, semi-structured interviews were used as  
4 they are viewed as the exemplary method to capture these experiences (Smith & Eatough, 2007;  
5 Smith & Osborne, 2003). The interview schedule was developed based on suggestions from Smith  
6 and Osborne (2003). It began with an open question allowing the participant to describe their  
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8 experience in their own words, before returning to specific topics to explore their meaning in greater  
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10 detail.  
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### 13 14 15 16 *Analytical procedure*

17 The reflexivity of the researcher plays a critical role in both the collecting and analysing of the data.  
18 For the reader to fully appreciate the interpretations of the data, it is salient to introduce the  
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20 researcher's values and assumptions (Elliot et al., 1999).  
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25 The field researcher and first author of this paper has LHON, which has aided his ability to explore  
26 participants' narratives and to appreciate the basic assumptions about medical, social and personal  
27 experiences within the participants' discourse. However because he has his own lived experience of  
28 life with LHON, a potential pitfall was that he found his preconceived ideas within the text and wasn't  
29 open to uncertainty and adaptability within the analytical process. To address this, the second author  
30 moderated this process and a thorough and systematic analytical procedure was undertaken.  
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38 Conforming to IPA principles the data were approached ideographically, with each transcript being  
39 analysed in a stepped process as suggested by Willig (2008) and Smith and Eatough (2007). The  
40 transcript was read numerous times to familiarise the researcher with the data. All interesting or  
41 significant comments and units of meaning were recorded; these were then interpreted and organised  
42 into emerging themes. Lastly all the emerging themes were clustered together to find super-ordinate  
43 themes. During this process it was necessary to return to the transcript as themes were enriched and  
44 developed to ensure they represented the participant's subjective experiences. A table was created  
45 containing verbatim to display the super-ordinate and subordinate themes to ensure the themes were  
46 grounded in the participant's language and accurately represented their experiential interpretations.  
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48 This procedure was repeated across all participants, before common themes were identified within  
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50 the corpus.  
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### *Sample and recruitment*

Eight people were recruited for the study, with the first being used as a pilot interview to test the effectiveness of the schedule, and thus was not included in the analysis. No alterations were made to the questions as a result of the pilot. All the seven interviews included in the analysis were conducted with male subjects, reflecting the predominately male LHON population (Mann et al., 2002).

Participants were recruited by publicising the study on social networking groups and directly emailing people registered with an LHON website. At time of recruitment there were no UK specific online forums and all sources were open internationally. The researcher identified two social networking sites that each contained an LHON group; a brief message, outlining the purpose of the study, was posted on each site. The same message was emailed to people, based in England, registered with the website. Interested individuals were requested to email the researcher. Eleven people responded, the majority (5) to the website email, the researcher chose to proceed with the respondents (8) who lived in the UK for consistency of social experience. They were then sent further details of the research, for their consideration, before agreeing to participate. Elliot et al. (1999) recommends that details are provided on participants and their life circumstances permitting the reader to assess the relevance of the findings. To protect the anonymity of the interviewees, their relevant details are provided without any reference to the individual. The sample covered a range of ages, 21 to 62 and length of time with LHON, 1 to 43 years. Prior to diagnosis all were occupied in either education (2) or employment (5); subsequently three are now unemployed, two are in education or training and two, who have lived with LHON for the longest period, are employed. All interviewees were given a leaflet for the Royal National Institute for the Blind's (RNIB) emotional support service, in preparation for any distress that may have been caused by the discourse. These measures helped the study to gain ethical approval from Oxford Brookes University.

All the interviewees were based in England and the interviews took place in locations that were convenient for the participant including; café's, their homes, local libraries and local pubs. One participant was unable to meet the researcher in person, so the interview was conducted through the

1  
2  
3 internet on Skype. The interviews ranged in length from 41 minutes to 104minutes (M= 69 minutes).  
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5 The interviews were digitally recorded and fully transcribed. All participants gave informed consent.  
6  
7 To protect the anonymity of the participants pseudonyms are used in this research paper.  
8  
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## 10 **Findings**

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12 The analytical processes produce six key themes that illustrate the participants' lived experience of  
13  
14 LHON; psycho-social losses, attitudes and coping strategies, development of practical skills, identity,  
15  
16 regaining independence and recurrent loss.  
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### 20 **Theme 1 – Psycho-social losses**

21  
22 A key aspect of the lived experience was the psycho-social losses associated with the visual  
23  
24 impairment. The loss of vision as a sensory tool affects the person's capacity to live and act within the  
25  
26 world, evoking frustration at their reduced ability.  
27

28 This is well demonstrated by the loss of social/communication skills that five people discussed in their  
29  
30 discourse, which made them ignorant of non-verbal communication, preventing them from acting in  
31  
32 the appropriate and desired manner. Paul's experience with his neighbour illustrates this:  
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36 My neighbour recently fed back to me that "when you first moved in I thought you were really  
37  
38 rude" and I said "why is that?" and she said "well we smiled at you a few times and you never  
39  
40 acknowledged us"  
41  
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43 Chris recalled how he explains the functional impact of his loss of vision:  
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46  
47 One of the ways that I used to describe is that it is kind of like being in a bubble of naivety sort  
48  
49 of thing. You have got this little bubble where you can see so far, for such and such a  
50  
51 distance of about half a metre or so pretty well-ish. And then other than that, you are  
52  
53 completely naive to everything else going on  
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56 The loss of independence and freedom was a strong theme permeating the interviews, with six of the  
57  
58 participants raising this issue. For Alex in the first two years of his lived experience the difficulty of  
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3 independent travel tested his resolve. The frustration of continually getting the wrong bus or train  
4  
5 culminated in suicidal thoughts. Determined to continue travelling independently he had resorted to  
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7 walking four or five hours to get home:  
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9

10 (... ) taking four and five hours to get home because I got fed up getting on the wrong train.

11  
12 Many a time I'd walk along the river and thinking "shall I just jump in and forget it?"  
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16 This exemplifies how individualised the lived experience is. How a person appraises the various  
17  
18 challenges associated with living with LHON will be reflected in their emotional reaction. Two  
19  
20 participants recalled only being depressed in a single moment, whilst four described an initial  
21  
22 depressing period without discussing any particular low points. Positive attitudes and coping  
23  
24 mechanisms were used to manage these challenges, but across the discourse was the reality that  
25  
26 daily hassles and associated losses could always culminate in frustrations:  
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28

29 You live with it and learn to cope better and you still get good days and bad days. (Paul)

30  
31 I still get periods of frustration, you probably do. (Martin)  
32  
33

34 These will manifest through experiential losses and the inability to complete small tasks. The  
35  
36 participant's history as sighted people compounds this frustration because they are aware of the ease  
37  
38 with which they could live and act within the world.  
39  
40

41  
42 I would say the biggest frustration is knowing that I have put something down somewhere and  
43  
44 when I am reaching for it, putting my hands on the work surface for example and saying to my  
45  
46 girlfriend "it's not there, I can't find it" (Martin)  
47  
48

49 Understanding the pleasure they are missing exasperates the loss of visual aesthetics that three of  
50  
51 the participants bemoaned:  
52  
53

54  
55 You enjoy it because you've seen the scenery. If somebody says "that mountain looks great"

56  
57 then "fine, thank you, it does look..." you know but I can't really engage (Martin)  
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4 All the participants expressed stress and frustration at their loss of skill, ability and independence.  
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6 These were most pronounced at the start of their lived experience because with time they developed  
7  
8 the tools to manage daily challenges and their memories of living and acting as sighted people faded.  
9  
10 However the culmination of losses will always resurface in frustrating moments, dictated by an array  
11  
12 of factors influencing the individual's mentality in that period.  
13  
14

## 15 16 **Theme 2 – Attitudes and coping strategies**

17 Threaded throughout the interviews was a pragmatic approach to living with LHON, as the  
18  
19 participants discussed the tactics they used to manage their environments and the attitudes they  
20  
21 embodied which allowed them to deal with the recurrent frustrations. Chris's practical strategy for  
22  
23 meeting people was echoed by other participants:  
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25

26  
27 I tend to call people up before meeting now and sort of "where are you? Are you in the  
28  
29 garden?"  
30  
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32 Six of the participants demonstrated a determination and resolve that enabled them to continue with  
33  
34 their lives and overcome the frequent challenges:  
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37  
38 (...) you know I am a great believer in there not being such things as mistakes just learning  
39  
40 opportunities. There was a lot of opportunities shall I say. I had to dig deep and do it for  
41  
42 myself. (Paul)  
43  
44

45 Humour was used by three of the participants to reframe embarrassing or frustrating moments,  
46  
47 alleviating potential stress and anxiety.  
48  
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50  
51 (...) and then you start finding that there is a humour side to it. You get loads of tins in a  
52  
53 cupboard and if my wife was away for a day or two days or something, the fridge would be full  
54  
55 of bowls of things that I didn't want like, buttered beans and I thought they were baked beans,  
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3 pineapple chunks instead of peeled tomatoes. She'd come home and she'd laugh her head  
4 off, and we'd laugh our heads off (Alex)  
5  
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7

8 Below, Dominic and Martin reveal their attitude that has enabled them to progress and move forward  
9 despite the adversity of a visual impairment.  
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13  
14 You get on with it or you sit in the corner and weave a basket and I was never prepared to do  
15 that. (Dominic)  
16  
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18  
19 There are so many things that one can do, you just need to adapt the tools... (Martin)  
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### 22 23 **Theme 3 - Development of practical skills** 24

25 The pragmatism described above was demonstrated through the practical methods the participants  
26 used to adjust to life without their main source of sensory information. Adjusting to life with LHON  
27 requires the individual to overcome the barriers surrounding their competencies. They develop new  
28 skills and use other senses as tools to obtain the information. For example, developing the skills to  
29 use assistive technologies or to 'touch type' relies on using sound or touch as a tool. Martin clearly  
30 understood the principle of replacing lost visual information and found he was still able to fly planes, if  
31 a partner could provide him with the relevant details:  
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39 I have been ballooning since, I have been in the Alps, I have landed the balloon and all I need  
40 is the information in that the field is clear, we are now 10ft up, 6ft up, 4ft up and then I am able  
41 to touch down.  
42  
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46  
47 Three of the participants gave examples of using other senses as tools to obtain information, as Paul  
48 details:  
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51  
52 ...but now I can put my hand in my pocket and tell you how much money I have got just by  
53 feeling it, you know?  
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3 The skills to live as a visually impaired person can also be developed by the person's experience of  
4 new activities which increase awareness of their capabilities. Meeting other visually impaired people  
5 can inspire the individual to overcome barriers around their abilities and try new things, as  
6 demonstrated by Alex:  
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12 Through them I took up sport and went to cricket, football, and athletics with them. I think it  
13 improved my mobility, my spatial awareness, my much sort of VI skills  
14  
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#### 16 17 18 **Theme 4 - Identity**

19 The level of residual vision that remained post LHON onset appeared to influence the decision to  
20 display their blind identity. All participants were on the disability register, suggesting an  
21 acknowledgement of their blind identity. The discussion deliberating the portrayal of this identity was  
22 intertwined with a theme of the reaction of others, rather than an internal resistance.  
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28 Salient across the discourse on identity was the public's image of a 'blind person' and subsequent  
29 reactions to symbols of this perception. The white cane is possibly the most powerful representation  
30 of the blind position and all participants were aware of this symbolism and used the cane as a non-  
31 verbal form of communication. Alex reported that it helped him to cross the road safely:  
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38 I'll have a white cane and purely get it out to get across the roads.  
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41 The symbolism of the white cane also had negative consequences which limited its use by some  
42 participants. Five participants' sparing use of the cane was based on these factors; dominance of their  
43 personality, vulnerability and losing control of situations, as the following extracts describe:  
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48  
49 Well I use it when I want people to know. I'll get it out folded up when I want people to know,  
50 like if I go into a supermarket or something or when I'm getting the train like that, and when I  
51 need help. But the rest of the time, I want my friends to know sure, but for everyone else it is  
52 not that I don't want people to know but it's not important sort of thing and it is not who I am  
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3 and it is not what makes me who I am, it is just something that occasionally helps me out.

4  
5 (Chris)

6  
7  
8 There is also the vulnerability side of it as well if you are out and the long and short of it is that  
9  
10 it is not a very nice world out there and there are places that are not very nice and I would  
11  
12 probably prefer to struggle than walk down the road with a long cane saying "Hey I am blind,  
13  
14 come and mug me" (Paul)

15  
16  
17 The participants' residual vision placed them in conflict with the public perception of a blind person,  
18  
19 which is of someone in complete darkness. Their experience of having lived as a sighted person  
20  
21 helped them to empathise with the ignorance of the public about the range of visual impairments. Paul  
22  
23 explained the consideration behind his use of language:

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26  
27 I try not to use the word blind because that is what my understanding of a lot peoples thinking  
28  
29 is that I don't have any sight at all. They can't comprehend that like "how did he walk out of  
30  
31 the door? How did he know where the door handle was?" so I just generally try to say to  
32  
33 people in a dismissive way "my eyesight isn't very good" and just leave it at that really.

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35  
36 Three participants spoke about providing explanations to people who questioned their blind status,  
37  
38 but showed tolerance towards their doubters because of the empathy they possessed:

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41 Sometimes I guess they are quite surprised, like "really, are you blind?" and sometimes they  
42  
43 try to test things out with me like "can you see this?" (Oliver)

44  
45  
46  
47 The results within this section suggest that overtly displaying a blind identity places the individual into  
48  
49 a broad category of blind people that doesn't accurately represent the participant's skills, abilities and  
50  
51 attitudes. This decision reflects an acknowledged changed status within and relationship with the  
52  
53 social world.

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56 **Theme 5 - Regaining independence**  
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3 As part of this process of adjustment all but one participant talked about regaining their  
4 independence. This context provides an appropriate time to raise the topic of the participants' varying  
5 visual fields. While no scientific measurements were used, each participant described the extent of  
6 their field of vision during the interview. The scope of the visual field that is affected ranges from one  
7 participant having just the centre blurred with clear peripheral vision to another whose entire visual  
8 field is blurred. The clarity of a participant's visual field influences how they view regaining their  
9 independence. Those (2) whose visual field is most affected became reliant on other tools for  
10 information, in this example extending their dexterity through the use of a cane:  
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20 It was really good learning to use the white cane and learning routes and things like that  
21 because it just gave you that independence and was able to sort of help you get out there and  
22 do things again. (Oliver)  
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25  
26

27 Two other participants explained practical techniques they used to allow them to live and work  
28 independently:  
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32 So I had it in large print on the outside of the spine so that I could find the folder if I wanted  
33 and still keep a degree of independence. (Alex)  
34  
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38 As I knew that it was long term I could go out and buy adaptive technologies and look into that  
39 sort of stuff and look into getting my independence back really in various different ways.  
40  
41 (Chris)  
42  
43  
44

45 The last two participants' notion of regaining independence was focused on placing themselves in  
46 unfamiliar situations to gain confidence in their abilities as a blind person:  
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48

49 I like to be outgoing even if I have to struggle a little bit. I like to get out there and do these  
50 sorts of things for myself. (Paul)  
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55 However, despite all participants travelling on a similar journey of adjustment, the question of whether  
56 it ever ended was disputed:  
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5 I would say that if somebody says that they have totally adjusted to their sight loss then I  
6 would say that they are not being truthful and honest with themselves really. I just think that it  
7 is an on-going experience really (Paul)  
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11  
12 However Dominic believed that through his extensive experience of being visually impaired he had  
13 developed all the necessary practical skills and made all the possible lifestyle adjustments.  
14

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16  
17 Oh yeah. I can't think of any area that needs some adjustment. Come on I've been a blind  
18 person for 43 years, if I wasn't adjusted by now then I have got a real problem. (Dominic)  
19  
20

### 21 **Theme 6 Recurrent loss**

22  
23 Regardless of the participants' opinion on the process of adjustment it was acknowledged that they  
24 will always experience moments that evoke a feeling of loss for their sight. The practical and  
25 emotional coping mechanisms outlined above permit the individual to overcome barriers and lead a  
26 fulfilling life. However their limited sight will mean that they have fewer options in life and reduced  
27 visual experience of a moment. At intermittent times this can lead to a feeling of loss and frustration  
28 reminiscent of their early period of adjustment.  
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35  
36 Interviewer - do you think that you ever do fully get over losing your sight?  
37

38 Dominic – No, no you don't.  
39  
40

41 Alex provides examples of experiences where the feeling of loss will reoccur:  
42  
43

44  
45 Especially with my son like "I wish I could see him more" or I'll go to a wedding and you want  
46 to see something that is there or happening, or you want to enjoy what everybody else is  
47 looking at, and there are certain sporting events where I'll get invited to things and you'll just  
48 want to see.  
49  
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54  
55 It is appropriate to conclude the findings by revisiting the theme of loss because this highlights that  
56 the lived experience will always be vulnerable to a situation that encapsulates what they have lost.  
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## Discussion

This research paper has recorded the lived experiences for seven people with the visual impairment Leber Hereditary Optic Neuropathy. It has aimed to make the unfamiliar experiences of people with LHON familiar for its readers by providing insight into what meaning they have attached to instances in their lives since diagnosis and how they interpret the world they now inhabit. It has revealed that the course from being fully sighted to living with LHON requires resilience, pragmatism and determination to develop the practical and emotional coping skills to manage their environments and progress with their lives. A fundamental finding is that the process of adjustment has no conclusion, as the challenges and restrictions of their visual impairment will occasionally overwhelm the coping mechanisms they have developed, reflecting the findings of Murray et al. (2010). This substantiates Hewson's (1997) Episodic Stress Model, as the evidence from this research shows that how people appraise both primary (being diagnosed) and secondary (experiential frustrations) stresses is dependent on their context and personality making it a very individual experience. An essential reason for using this over a grief model (Atkinson, 1990; Baus, 1999; Schainholz, 2000) is that it helps to explain why the feeling of loss and frustration are chronic and will be episodic throughout life. For some participants the frustration arose from the inability to complete routine tasks, whilst for others it was the loss of visual information hindering how they experienced a specific moment. As Hewson proposes, living with the loss of ability makes the individual susceptible to occasional frustrations. Periodically a culmination of these stresses can lead to a feeling of loss reminiscent to that experienced during the primary stress.

Concurring with the discourse in Dale's (2010) narrative exploration of the effect a visual impairment has on identity, it was not the sight loss but relationships with sighted people that affected the self-image. Several participants' acknowledged the practical benefits of using a white cane but used them sparingly due to the reaction of others, citing factors such as dominance of their personality, vulnerability and losing control of situations. There is a clearly defined stereotype of a blind person of someone who is in complete darkness and many of the participants acknowledged this when considering how to explain their visual impairment. The question of how does LHON affect identity is not purely an internal question about the self but is compounded by how others interact with and help



1  
2  
3 to construct a person's identity. Participants described how communicating their blind identity, either  
4 verbally or through symbols, alters relationships with sighted people. It drew out intrigue or  
5 assumptions about their capabilities but it often dominated perceptions of them. Different tactics were  
6 described to manage this interaction depending on the context, the other and the individual. One  
7  
8 tactic was to address the misunderstanding by discussing the condition and educating the other, the  
9  
10 second was to be passive in the moment, reluctant to extend a brief encounter, and the third was to  
11  
12 conceal their visual impairment to avoid the situation. Hayeems et al. (2005) and Zaborowski (1997)  
13  
14 suggest that adjustment to a visual impairment is contingent upon accepting the blind identity, which  
15  
16 enables the person to utilise all possible help. The interviews detailed how the individuals' accepted  
17  
18 their blind identity and recognised how this could shift their position in society. When they chose to  
19  
20 conceal this it was temporary and did not mean an internal resistance.  
21  
22

#### 23 24 25 *Limitations and recommendations for future research*

26  
27 It would be erroneous to generalise the results of this study across the LHON population, because it  
28  
29 should be considered that those who volunteered to participate are actively involved in the LHON  
30  
31 online community. The implication is that those interviewed represent a sample of the population who  
32  
33 have highly developed coping mechanisms allowing them to live a fulfilling life. It is also difficult to  
34  
35 compare and contrast themes within the stages of the adjustment process because the range of time  
36  
37 living with LHON (1 to 43 years) is large for such a small sample. To better understand the lived  
38  
39 experience for people with LHON it would be appropriate to recruit a more representative sample.  
40  
41 Working in partnership with medical professionals to contact patients from their database would allow  
42  
43 for a recruitment programme that aimed to capture a cross section of the population at different times  
44  
45 in their lived experience by performing a longitudinal study. This would allow for a more systematic  
46  
47 investigation into the development of attitudes and coping resources. It would be of interest to  
48  
49 discover if these attitudes were uniquely related to the participants' prior life trajectories or whether  
50  
51 the challenge of an acquired visual impairment forces the birth and/or growth of such an attitude.  
52  
53 Additionally research with a larger, more representative sample could provide greater insight into the  
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55 chronic but episodic losses and if and when they might resurface.  
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The empathy of the field researcher for their situation encouraged the participants to discuss their stories in a free and open manner. However, despite the rigorous analytical process it is impossible to eradicate the influence of his lived experience on the data collection and potentially in the analysis. It would therefore be appropriate in future research to add a sighted field researcher to the process.

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