

RESEARCH PAPER

Life on hold: the experience of living with neuromyelitis optica

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Abstract

Purpose: Neuromyelitis optica (NMO) is a rare autoimmune condition characterised by acute relapses of optic neuritis and extensive transverse myelitis. The aim of this qualitative study was to develop an improved understanding of the experiences of people living with NMO. **Method:** Fifteen participants completed a semi-structured interview and data were interpreted using a constant comparative method. Participants were recruited from the Northern UK NMO Service. **Results:** Five major themes were identified: diagnosis and treatment, symptoms, adjustment, identity and support. **Conclusion:** Results suggest NMO is a difficult condition to live with due to the unpredictability of relapses and accrued disability of visual or spinal symptoms occurring with each relapse. Poor vision, reduced mobility, bladder dysfunction and pain affected participants' independence and experience of living with NMO. Participants reported that during relapse and recovery they would "put their life on hold". They identified the importance of periods of stability to enable them to adjust to their condition and therefore aim for "normality" of life that they believed was comparable to their peers.

Keywords

Daily activities, lived experience, neuromyelitis optica, qualitative research, relapses

History

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► Implications for Rehabilitation

- Disability due to NMO has a major impact on an individual's life, abilities and identity.
- Timely treatment of relapse is essential to reduce or prevent disability.
- Health care professionals need to coordinate care locally and nationally.
- Multi-disciplinary work is vital to promote adjustment, coping strategies and support for people living with NMO.

Introduction

Neuromyelitis Optica (NMO) (also known as Devic's disease) is a rare autoimmune condition characterised by relapses of the optic nerves (optic neuritis) and spinal cord (transverse myelitis) [1]. Historically, NMO was viewed as a subtype of Multiple Sclerosis (MS) [2] and was therefore treated in similar ways. The identification of a specific antibody (anti-Aquaporin 4) for the antigen aquaporin-4 [3] (present in approximately 70% of people with NMO) provided an increased understanding that NMO is a separate disease to MS. It has now been shown that many treatments such as beta-interferon used in MS may increase relapse rates in NMO [4,5]; therefore, it is essential to confirm the NMO diagnosis to prevent disability caused by incorrect treatment.

Worldwide there are few epidemiological studies on NMO; the prevalence in the UK is estimated at 1 per 100 000 [6]. People of an Asian, African or South American ethnicity have a higher

incidence of NMO [7,8], and predominantly females are affected, with a gender ratio of 5:1 [6]. The typical onset age of NMO is in the fourth decade but the first attack may occur at any age from early childhood through to the elderly [1].

The UK Department of Health Specialist Services have classified NMO as a rare neurological condition and consequently funded two national centres. Their aim is to provide expertise in diagnosis, relapse management and treatment in collaboration with patients' local neurological services throughout the UK. The service offers a multidisciplinary approach including a range of professionals. For further information on NMO visit www.nmouk.nhs.

The onset of NMO is usually through a devastating relapse of optic neuritis (ON) or transverse myelitis (TM) causing significant disability. Relapses can happen within hours and may have a similar impact to a spinal cord injury. However, unlike a spinal cord injury NMO is a relapsing condition, where every relapse causes further disability, requiring a period of rehabilitation. ON (unilateral or bilateral) is associated with blindness, loss of colour vision, central scotoma and pain on eye movement [1]. Longitudinal extensive TM (more than three vertebrae) is associated with bilateral motor weakness, sensory loss including numbness, banding, intense paraesthesia, paroxysmal tonic

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Table 1. Characteristics of NMO participants.

Participant No	Sex	Age (yrs)	Yrs from 1st Symptoms	Aquaporin-4 Status	Clinical presentation
1	F	48	4	Positive	Motor/sensory/visual loss
2	F	18	2	Negative	Motor loss
3	M	74	34	Positive	Complete visual loss
4	F	45	1.5	Positive	Motor/sensory/visual
5	M	64	21	Positive	Motor loss
6	F	28	13	Negative	Complete motor/sensory loss
7	F	54	8	Positive	Motor/visual loss
8	F	33	5	Positive	Motor/visual loss
9	F	43	4	Positive	Visual loss
10	M	65	10	Positive	Motor loss
11	F	41	6	Positive	Motor/sensory/visual
12	F	54	2	Negative	Motor/visual
13	F	25	10	Negative	Visual/motor loss
14	M	52	5	Negative	Visual/motor loss
15	F	54	5	Negative	Visual/motor loss

spasms, neuropathic pain, itching and bladder and bowel dysfunction. Brain stem involvement may occur, and may cause prolonged hiccoughs, nausea, vomiting, vertigo or respiratory failure [2,9].

Residual disability in NMO is acquired through relapses resulting with severe visual loss and/or paralysis of limbs, highlighting the importance of timely and effective treatment. There are four aims of treatment for NMO: acute treatment of relapses, prevention of relapses, symptom management and rehabilitation [10–12]. First-line therapy for acute relapses is high dose methylprednisolone [2,11], and plasma exchange [12] may be considered if there is a poor response to steroid treatment. Steroids and immunosuppressants are used for the prevention of relapses [2,11,12].

Each individual experiences a range of symptoms such as visual loss, mobility impairment, pain, fatigue and continence issues. Symptom management is provided by the relevant health care professional including ophthalmologists, physiotherapists, occupational therapists, nurses and dietitians. A person-centred approach is necessary, with an action plan tailored to the needs, aims and goals of each individual. This requires shared decision making to ensure that goals are relevant to the individual, in the context of their current situation and life experience. Due to the rarity of NMO specialist assessments are conducted at the national centres; however, to provide a holistic care package with good continuity of care, it is essential for strong liaison and coordination with local care services such as neurologists and therapists. Contemporary research on NMO has focused on the clinical, radiological and immunological aspects of the condition, as well as symptoms experienced by patients such as pain [13], itching [14] and tonic spasms [15]. Previous research has highlighted some of the difficulties of living with a relapsing condition [16]. However, factors unique to NMO such as the distinctive pattern of optical and spinal symptoms, the high levels of residual physical disability, reduce the extent to which NMO is comparable to other neurological relapsing conditions. The aim of this qualitative study, the first of its kind, was to gain an insight into the experience of living with NMO.

Method

Sampling and participants

Participants were recruited from the northern NMO centre. Twenty-eight patients were invited to the study through an information sheet. They were selected to ensure a varied sample of age, gender and disability. Nineteen patients agreed to participate, although four withdrew before participating in the

interview. Reasons given for withdrawal were family difficulties ($n = 2$), work schedule ($n = 1$) and ill health ($n = 1$).

All 15 participants who agreed to be interviewed (Table 1) had residual disability from previous relapses of optic neuritis and myelitis. Nine participants tested positive for aquaporin-4 antibodies. Visual loss ranged from partially impaired vision in one eye to complete loss of vision in both eyes, and four participants were registered blind. Motor loss ranged from poor balance and weakness in one leg requiring sticks/crutches to tetraplegia (one participant), and four participants were wheelchair users. Sensory loss included complete numbness, neuropathic pain, tight banding around the trunk and paroxysmal spasms. Seven participants had stopped working due to their NMO, six were in part-time work, two participants had never worked and three were studying part-time. Nine participants were married and had longstanding relationships (minimum 20 years). Two participants lived alone and four lived with their parents.

Approval for this study was granted by a local research ethics committee. Informed consent was obtained from all participants. Interviews were held at a convenient location for the participant, either at the hospital or at home.

Data collection

Interviews were conducted by the NMO nurse specialist and assistant psychologist. Each interview lasted between 30 and 60 minutes and was digitally audio-recorded. Participants were interviewed using a semi-structured interview schedule that aimed to elicit participants' feelings, their experience of diagnosis, experiences of health care, changing nature of, and their attitude to, NMO over time, daily impact of symptoms (both physically and psychologically), and adjustment and coping methods. Anonymity of the participants was essential, so all identifying features including names, locations and treatment centres were removed during transcription.

Data analysis

Audio files were transcribed by the authors to ensure familiarisation with the data. Data analysis using a constant comparative method [17,18] was an on-going, iterative process. The interviews produced a rich data set of approximately 30 pages per interview. Each transcript was numbered for identification purposes to ensure anonymity and confidentiality. Transcripts were analysed and coded by the two interviewers independently, to ensure reliability of analysis. The coding process involved attributing a code to a

Table 2. Themes and subthemes of results.

Themes	Subthemes
Diagnosis and treatment	<ul style="list-style-type: none"> • Initial symptoms • Getting a diagnosis • Relapse recovery • Treatment and medication side effects • Fear of future relapse
Impact of symptoms on everyday life	<ul style="list-style-type: none"> • Visual loss • Sensory loss • Continence • Fatigue
Adjustment	<ul style="list-style-type: none"> • Coping • Frustrations • Life on hold • Mood • Finances
Identity	<ul style="list-style-type: none"> • Independence • Interests • Image
Support	<ul style="list-style-type: none"> • Family • Pets • Close friends • Informal societies

sentence or section to represent an idea or concept associated with the research question. Codes were generated from the data not imposed on it and identified by listening to the audio recordings whilst reading the transcripts. Researchers met to discuss potential codes and interpretations of the data. Once codes were finalised through consensus, they were grouped into sections discussing similar issues or meanings and then themes emerged from the data with the assistance of thematic maps. These themes comprised diagnosis and treatment, impact of symptoms on everyday life, adjustment, identity and support. Within these themes subthemes emerged and are presented in Table 2.

Results

From the data, five major interconnected themes (Table 2) were identified: diagnosis and treatment, symptoms, adjustment, identity and support. These themes contributed to an individual's experience of living with NMO.

Diagnosis and treatment of NMO

This theme included initial symptoms, getting a diagnosis, treatment of relapses and recovery and the fear participants lived with of future relapses, due to the potential disability that could accrue. The initial symptoms of myelitis or optic neuritis occurred quickly, yet resulted in the participant requiring rehabilitation for many months. The process of getting a diagnosis for such a rare condition as NMO was arduous and learning to live with a disability had a major impact on participants.

Initial symptoms included double vision, pain upon eye movement, loss of vision in one eye, sudden vision loss, intense neuropathic pain, tight banding sensations, paraesthesia, and loss of sensation in arms and/or legs, lack of coordination, paralysis of limbs and retention of urine. The majority of participants ($n = 11$) highlighted how rapidly initial symptoms presented and reached a peak within 48 hours with no previous illness or warning signs.

So I was driving through town. . . I noticed right there in front of the Mosque where they were praying, I had to pull over and 'phone someone to come and get me, I'd gone blind, that's how that happened. (Participant 14a)

. . . started off I had a bath one Tuesday night and noticed my right leg from my foot to the ankle when I put it in the hot

water the water felt cold . . . by Thursday it had gone right up to the top of my leg. I couldn't stand up and I had lost use in my left leg altogether. (Participant 10a)

In comparison to initial symptoms of visual or mobility impairment, five participants started with intense neuropathic pain as an initial sign, prior to myelitis occurring.

Experiences of health care were varied, though the majority of participants voiced negative encounters. Thirteen participants complained that their doctors were not knowledgeable about NMO. This lack of knowledge had a potentially serious impact on outcomes, and retrospectively, four participants wondered if earlier steroid provision might have prevented their current visual or mobility impairments. The length of time and amount of recovery following relapse varied between participants. Some regained nearly full function whilst others were left with severe disability.

The vision started to come back within a month, little bits of the darkness started to subside. The ophthalmologist said by 6 months he would get a good idea of how much was going to return. I'd say it's about 50%; it's difficult to measure because it's like looking through a colander. (Participant 4a)

Thirteen participants reported difficulty attaining a diagnosis of NMO, and four sought a second opinion either privately or abroad. These participants reported misdiagnosis, most commonly MS ($n = 12$). Other alternative diagnoses included transverse myelitis, cerebral vasculitis and meningitis. Two participants who were diagnosed with NMO during the last two years considered themselves to "be lucky", as they were seen by neurologists who were knowledgeable of NMO which improved the speed and accuracy of the diagnosis.

Following initial symptoms, 13 participants required rehabilitation in hospital. Several participants when given the diagnosis whilst in hospital had little concept of the impact of NMO. However, upon arriving home, participants started to appreciate the severity of their condition and far-reaching changes to their life. They then felt scared and many felt depressed as they feared another relapse.

Participants reported fear of the future, fear of the unknown, fear of death and the realisation that further relapses could happen, meaning they would have to change their life plans. Despite this, the majority of participants reported relief at finally having a name for their condition.

After a series of tests they came up with a diagnosis, phoned me and said you've got Devic's syndrome, you can look it up online. So I looked it up and I found that I was likely to go blind, stop breathing and drop dead and I thought this is not the best way to tell somebody. But then it was a great relief to have a name for whatever condition I had. (Participant 5a)

As time progressed, participants realised their initial fears were not justified. The majority ($n = 9$) of participants reported that although symptoms had not improved, they had adjusted and now managed their symptoms on a daily basis.

All participants reported wanting more support than they had received, especially during the early stage of diagnosis. Due to the rarity of NMO, many Health Care Professionals (HCPs) did not understand the consequences of relapses that is, visual and spinal symptoms accrue during relapses if not treated quickly. Participants also reported lack of knowledge on NMO and two participants incorrectly believed that as one NMO feature was present (i.e. optic nerve or spinal cord damage) then they would not experience NMO-related damage to another part of their body.

Participants reported they appreciated HCPs who made a point of researching NMO, providing the correct treatment, working in partnership with them and referring to other specialists. Some ($n=4$) participants reported good access to services such as immediate admission upon relapse and being able to contact their consultant directly for quick referral. Participants differed in what they wanted from their HCP, though all appreciated honesty alongside listening to their needs. A major frustration for all participants was not feeling listened to by their HCP.

Once diagnosed with NMO, patients were likely to have further relapses. Immunosuppressants and long-term steroid treatments used to prevent relapses in NMO had unwanted side effects such as weight gain, indigestion, osteoporosis, acne and stretch marks. Medications such as antacids, calcium and vitamin D supplements were given to reduce the risks of side effects. All participants were affected by side effects; younger females in particular were concerned about their image, weight gain and the views of their peers.

The weight thing really upsets me, I see all these slim girls and they're able to wear what they want, look really nice and I have to wear a jacket. So the weight thing is still a big issue. I looked like a hamster I was really ballooned out. I've got the steroids so they make you hungry and they make you just want to eat loads but I have to have them because otherwise I'd probably be poorly again. (Participant 2a)

In addition, medications given to manage symptoms such as pain, spasms and bladder dysfunction were also associated with additional side effects such as drowsiness, dry mouth and indigestion. Consequently, previously fit and healthy participants took 6–10 different medications, which created additional responsibilities and constraints on normal daily life.

Participants worried about the future, especially regarding further relapses, symptom progression, recovery and hospitalisation. A major concern for them was what the next relapse could do, especially the thought of losing sight or mobility. This appeared to be linked to the fear that they could not adapt to a different set of symptoms although they had adapted to the symptoms they already had.

... Sometimes I do think "oh what's going to happen to me if I can't see or blind or I cannot walk" but in other ways try to stop myself thinking about it, ... that's, one day I don't know! (Participant 9a)

Impact of symptoms on daily life

The most prevalent symptoms reported were visual disturbances due to optic neuritis, including blurred vision, lack of colour vision and complete blindness. Spinal lesions caused bilateral motor weakness, sensory loss including numbness, banding sensations, intense paraesthesia, neuropathic pain and bladder or bowel dysfunction.

The majority of participants ($n=13$) reported visual impairments including two with no perception of light. Some ($n=3$) did not see their visual impairment as a handicap and coped well, but stated they would never get used to their visual impairments even after a long period of blindness (30 years in one case). They reported frustration at having to concentrate and plan everything in advance, and at times this was so mentally exhausting that they reduced their social activities. A major frustration for these participants was being dependent upon others. Participants reported detrimental physical consequences including banging into doors, bruises, cuts and falls. Participants learnt to adapt and utilise strategies to manage their visual impairment; however, they

reported not feeling safe going out alone and this fear then had an impact on their self-confidence, causing social isolation. Participants developed other senses as compensation such as increased hearing and relying more on memory, touch and vibrations.

... it's not like I expected to be blind but actually being unsighted isn't as scary as sighted people think it is, that's my own personal view. ... as long as I can maintain independence, hearing kicks in and other things come into play. Because you can still be independent sightless and sightlessness isn't black. I hope that I shall be gardening for years. (Participant 15a)

Twelve participants had neuropathic pain that was constant and/or paroxysmal, using descriptions such as burning, intense paraesthesia, tight banding, cramps, spasms, electric shock, icy or scalding sensations or numbness. Areas of worst pain were in the legs (predominantly bilateral), central back and arms. Due to numbness in their legs, seven participants reported problems with balance and a fear of falling. Pain significantly affected participants' daily life by impacting upon their daily activities, mood, walking ability, enjoyment of life and relationships.

The pain would come on for no reason at all. Start from your toe to ankle to mid-thigh. It would only last 40 seconds until it goes off but it's terrible pain that pulls my foot up it was that bad. It may occur several times an hour or day; it's so unpredictable. (Participant 10a)

Bladder issues such as urgency, frequency, hesitancy of micturition, nocturia and incontinence were a major concern for nine participants. They reported difficulties accessing toilet facilities, and this was a major concern in all journeys and event planning. Five participants used intermittent self-catheterisation and reported mixed responses ranging from the benefit of increased independence, to increase of urinary tract infections that exacerbated NMO symptoms resulting in hospitalisation. Three participants used continence pads, which were reported to increase independence. Bladder problems caused embarrassment, humiliation, decreased self-confidence and reduction in social activities, thus affecting the participant's independence.

... I have to plan everything, if I was to go out shopping, we have to make sure that I've done my catheter before we go out, make sure there are toilets about, I have to take spare clothes in the car in case I have an accident. (Participant 2a)

Fatigue affected seven participants, causing restrictions on activities of daily living including employment or education. Participants discussed their need for pacing and planning activities to prioritise their energy usage for the most important tasks, thus causing frustration due to lack of spontaneity and a dependency on others.

Even walking my dogs, I have to know where I'm going, I have to know there are walls that I can sit on, my legs go really stiff and I just have to sit down to let life get back into them. And then it affects my back and oh it's just so frustrating. I know my limits and I know how far I can go. (Participant 1a)

Adjustment

Participants had to feel they were in a "stable phase" without relapses before they could adapt their lifestyle, make adjustments and start coping with their situation. Participants felt their "life was on hold" as they could not plan for the future, due to waiting

for their next relapse, this phase caused many frustrations. The adjustment period varied for each individual but was helped by increased lengths of time without relapses.

Participants reported a dichotomy between adjustment to, and acceptance of, living with NMO. Several participants reported they had adjusted to their condition, although they could never accept it. Seven participants reported learning to live with NMO, as there was nothing else they could do; others felt they would be able to adjust provided that their symptoms were stable and without relapses; however, they could not predict when this would be. The aim for all participants was to live a life as normal as possible compared to their peer groups.

... I had to get my degree between relapses, I was only just starting at looking to get back into teaching and now I've relapsed again and it's so unstable, to get some stability with it would be something. (Participant 13a)

The need to "just get on with life" was highlighted by 14 participants. The uncertainty of the condition meant they could not predict what was going to happen tomorrow or in the future, and so they preferred not to think about it and live for today.

Now, I sort of deal with it. There's no point thinking about it all the time because it's not going to go away so I think I'm getting better at dealing with it than I was at first, like a couple of years ago. (Participant 7a)

Eleven participants retained hope that their situation would improve and get better. Participants reported they felt their "life was on hold" and they wanted to match the "normality" and life goals and concerns of their peers and family members such as independence, attending university, employment and relationships.

I just feel I've missed out on all my 20's like you're almost left behind. What I mean is in the scheme of life I'm 33 soon so you don't want to be living with parents. I'd have liked to have had a family, I'd have liked to have had my own place, be capable, I've lost a lot of confidence through having NMO and the treatment with the weight gain and things. (Participant 8a)

The main frustration for all participants was a reliance on others and a lack of independence due to their physical limitations. Frustrations were often due to a change in role including limitation of physical activities, inability to complete activities perceived as their duty, driving restrictions and resulting financial pressures. Others' ignorance of NMO, difficulties socialising and negative reactions of others towards them added to their frustrations.

My world gets smaller because everything that I would like to be doing seems to be closing off. I think my independence attached to my physical ability has been the most frustrating, far more than my sight if I'm honest. (Participant 15a)

Despite the highly prevalent attitude of "just get on with it; try not to think about it", many participants reported still having days where they felt very down. Ten participants reported experiencing low mood, depression and suicidal thoughts in the first couple of years and then more recently experiencing low mood intermittently, often linked to a relapse and worsening of symptoms. The main reason for low mood was the realisation of their current situation and their problems for the foreseeable future. Low mood was found to have an impact on participants' relationships and day-to-day living.

... I think probably it has affected me more mentally than physically.

Well I'm a wreck emotionally, I worry about everything and I get so wound up, I can't seem to do things quick enough. . . (Participant 12a)

Participants reported having days where they felt they could not cope but reported an active effort to change their mood and focus on the positives. Good family support was essential in helping them to cope and improve their mood.

The lack of financial independence was worrying for participants especially those whom had worked. Financial concerns included paying the mortgage, bills, supporting children and living with no income. Participants reported going without luxuries such as holidays, to manage financially. However, three participants reported they were diagnosed with NMO, at a time when they were financially secure.

I'd like part time work if I'm ever going to feel better. But never full time, every time I've worked full time I've had a relapse. I think with NMO you can feel useless because you're relying on people and if you're not working and things. Then I'd feel more useful, a sense of purpose. (Participant 8)

All participants reported lengthy difficulties navigating the benefit system and understanding what they were entitled to, participants with visual impairments found the system especially difficult. Participants were unanimous in reporting that a confirmed diagnosis of NMO made it much easier to receive benefits. NMO-related disability reduced income and increased expenses such as taking taxis instead of driving, fitting adaptations and buying equipment.

... I've always been in control, working and then suddenly you're not earning money and you just wonder what the hell is going to happen. Family and kids were small. We had a bit of savings but it got used up. The worst thing is not knowing what to do because nobody tells you what to do. If you're not familiar with the benefit system it's frightening, especially if you can't see anything, and you can't read anything. And people don't want to read things to you; they just shove forms at you. (Participant 3a)

Identity

"NMO" became participants' identity as they became known as a "blind person" or a "paralysed person". Participants' identities were found to be composed of their beliefs or values, perception of self as independent, positive perception of themselves, ability to complete meaningful activities and judgement by society.

Participants' self-worth appeared to be related to their ability to do the tasks they saw as being their role and part of a normal life. NMO was found to affect self-confidence, self-worth and self-belief and ultimately their independence. This then influenced participants' personality and attitude to life.

I can see that my confidence is diminishing, not driving, I've driven for 36 years and I am very aware that if I am anywhere on my own, I actually felt quite vulnerable. (Participant 1a)

Participants, especially females, were concerned about their image compared to peer groups and press publicity about ideal figures, and were self-conscious about weight gain and changing shape as a consequence of steroid treatment. They compared themselves to a time before NMO when they were slim and could wear fashionable clothes. These changes in their figures and

clothing caused embarrassment, which in turn had an impact on their social skills, resulting in a fear of socialising and eventually self-imposed isolation. Participants were appreciative of those who saw beyond their symptoms, such as visual loss or wheelchair use and valued friendships from people who knew them before their NMO diagnosis.

All participants had interests and hobbies that were not limited by NMO, including gardening, music, decorating, dancing, exercise, cake-baking, audio-books, TV and poetry. All participants had to juggle their symptoms, particularly fatigue, alongside their interests and routine chores and duties on a daily basis.

Support

There were many contributing sources of informal support that participants used to meet their needs including family, pets, close friends and charities.

Families were generally supportive, although participants reported sadness regarding their dependence upon family members. Participants worried about how they would cope with aging, how they would cope if they lost their partner, and the well-being of their spouse or parents due to the duties of caregiving.

... he works full time, it's his own business and he's not getting enough sleep in the night, sometimes I'm up twice in the night. I'd be totally lost without him.

... I think, what if he died before me, I do worry that, if anything happened to him
I don't know how I'd manage. (Participant 7a)

Older children provided help including physical assistance, housework, caring for younger siblings and emotional support. Family dynamics were found to change over time and difficulties were experienced within the family when roles were reversed or changed.

For nine participants, pets provided benefits including a meaning for life, a routine, companionship, safety, security and increased independence. Dogs and cats were classed as a "best friend" who provided unconditional support and affection regardless of participants' disability.

Participants reported a few close old friends that understood their symptoms, knew them before their disability and provided social support, although many friendships had declined over time. Participants generally reported difficulties making new friendships that did not judge them on their NMO symptoms; however some participants had achieved this.

I don't want to be treated differently and at work no one treats me differently, I've got really good work friends, they are so used to me, they go out with me and don't think of me as having a wheelchair they just think of me as me and that's really nice. (Participant 6a)

All participants reported they did not want contact with informal societies or support groups. Some participants did not want sympathy and others had negative perceptions of societies such as the Royal National Institute for the Blind and Multiple Sclerosis Society and felt they did not belong to these groups. Participants suggested that the desire to meet other people with NMO was high shortly after diagnosis but grew less over time. Several participants expressed an interest in meeting others with NMO, although the majority of participants did not want to dwell on the condition or be reminded of the potential severity of symptoms in others. Participants frequently reported social comparisons, which were usually divided into one of two

themes: either "people are better off than me" or "there is always someone worse off than me".

For me to meet other people with NMO and much more serious versions of the illness was an eye opener for me and helped a great deal to put things into perspective and to find out. I think I don't feel lucky to have NMO but I feel lucky to have got away with a light version of it. (Participant 5a)

Discussion

Theoretical saturation was reached in this sample of 15 participants, suggesting that despite their very different physical symptoms and life circumstances there are many issues of common importance to this clinical group. Similar themes were found for both people who were aquaporin-4 antibody positive or negative, suggesting that despite differences in diagnosis, individuals' experience were similar. NMO is characterised by recurrent relapses of optic neuritis and transverse myelitis causing significant disability. This study highlighted the fear and worry that future relapses may cause loss of sight or paralysis. This concern is different to people living with other relapsing conditions such as multiple sclerosis or rheumatoid arthritis [16,19,20], as although there are similar concerns regarding unpredictability of relapses they do not have to contend with the prospect of such sudden onset of disability.

First symptoms in NMO had a devastating impact, causing severe disability within a short period of time, requiring extended periods of rehabilitation. As participants initial symptoms usually presented in just one area, for example, vision or mobility they were usually investigated by a specialist in that area (e.g. ophthalmologist) who did not have the specialist neurological knowledge to suspect a diagnosis of NMO. Therefore, participants usually had at least two or three relapses before a neurological connection was made, thus causing accumulation of disability due to the difficulty and increased length of time obtaining a diagnosis of NMO.

Few other conditions have such diverse presenting symptoms, each of which initiates a different care pathway. The closest similarity is multiple sclerosis where diagnostic criteria are well established, leading to a quicker diagnosis [9,21]. The majority of participants were initially diagnosed with MS; however, it has now been shown that NMO is a different disease to MS in both management and pathway of the conditions [2,4,5,11] and this misdiagnosis caused difficulties in achieving successful treatment plan for many participants. Although NMO is a difficult condition to diagnose, patients emphasized the need for neurologists and other HCP to listen to them, empathise and show understanding of their individual circumstances. As NMO is such a rare condition, many HCP have poor knowledge, and little understanding or appreciation of the speed with which relapses need to be diagnosed and treated. Patients with NMO tend to be under the care of MS neurologists where the emphasis of relapse treatment is to speed up recovery [22] rather than preventing disability [11]. In MS, progression of the condition is usually the cause of disability, compared to NMO where relapses are the cause of disability, therefore the emphasis of treatment is very different [1,2,10]. There is therefore a need to provide information, education and increased awareness of the differences between NMO and MS for HCP [23–25]. Participants adhered to prolonged steroid regime due to the fear of future relapses and potential disability of other symptoms. The observable side effects of steroids particularly weight gain and significant stretch marks had a large impact on a person's image and confidence, causing reduced socialisation and subsequent lack of peer support that ultimately negatively affected their daily life. They required

dietary and exercise input from a multi-disciplinary team to minimise the impact. This is in comparison to MS where disease modifying treatments have minimal effect on the outward appearance.

Participants experienced feelings of low mood and anxiety, particularly after diagnosis, or on return home from hospital, similar to studies investigating MS [16,20] and spinal cord injuries [26–29]. The difference between a spinal cord injury (single event) and NMO (relapsing condition) is that in NMO this low mood appears to reoccur with relapses and therefore clinicians need to be aware of these symptoms, even in patients who have lived with the condition a long time. The impact of visual loss on daily activities and psychosocial well-being made adjustment difficult due to increased dependence on others requiring additional support [30,31]. Participants used support from HCP, combined with self-management strategies, to help them match the achievements and goals of their peers. Jannings and Prior [26] found that people with spinal cord injury (SCI) adjusted to their new lifestyle as time increased from the initial event. Our study supported this and found that if relapses occurred frequently, participants had difficulty accepting their situation and were unable to plan for the future resulting in them “putting their life on hold”. In an extended period of time without relapses, participants developed coping strategies to effectively manage both psychological and physical difficulties of NMO in daily life. Naturally, therefore, the process of adjustment followed a similar pattern in all participants but the time taken to adjust differed individually and appeared to be dependent upon the frequency of relapses. It may therefore be necessary for HCPs to provide additional support to all participants in times of relapse (and particularly with frequently relapsing patients), to provide an increased focus on proactive not just reactive support, thus enabling patients to develop personalised coping strategies and limit the amount of time spent putting their life on hold.

This study identified the difficulty of accessing services, therapists and financial benefits, and participants unanimously reported that access was made more difficult without a conclusive diagnosis. Although many HCPs may not be familiar with NMO, the problems caused by symptoms such as urinary, neuropathic pain, mobility or balance problems should be treated in similar ways to the same symptoms caused by a different condition such as MS or SCI. The findings of this study suggest that meaningful goals are different for every individual with NMO and require explicit discussion and objectives in the care planning of people with NMO. This requires multi-disciplinary care and coordination of specialist care and local services such as housing, benefits and financial aid to enable patients to reach their goals and adjust to living with NMO. As participants also reported difficulties accessing credible information on this rare condition, similarly to studies reporting that a lack of information has also been identified as a major issue in MS [24,32], there is therefore a need for HCPs to act as information providers for these patients.

The opportunity to participate was offered to participants with a range of demographic and clinical variables including recent relapse (within 3 months), disability and length of time since illness onset, age, gender and employment. Being currently unwell or feeling low due to NMO symptoms were reasons for declining participation in this study. Also, similarly to physical symptoms, it may be that people experiencing symptoms of depression or low mood, or who felt they were not coping effectively, did not volunteer for this study.

The minimum length of time since onset of symptoms was 18 months. Therefore, it was possible that many participants chose to participate as they felt they had adapted to living with NMO. Including people with a recent illness onset may uncover different themes, as a large section of our findings discussed

strategies for adaptation and managing life with NMO, which may come with time. The authors acknowledge they may have their own bias from prior knowledge of NMO gained through working in a specialist NMO team. As the researchers came from different clinical backgrounds, it is hoped that any biases were discussed and the impact on results was therefore limited.

Conclusion

This study provides valuable and unique findings on the experience of individuals and living with NMO. Although NMO shares similarities with other relapsing conditions, it is differentiated by the speed and severity of relapses, causing significant disability at the onset of the condition. These results suggest that NMO has a major impact on people’s identity through the physical and emotional changes it causes, that adjustment is a lengthy and individual process and that people with NMO utilise support from various sources of which health care services are essential.

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Declaration of interest

The authors report no conflict of interest.

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