

RESEARCH ARTICLE

Service delivery for people with hereditary spastic paraparesis living in the South West of England

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Abstract

Purpose: Hereditary Spastic Paraplegia (HSP) is an inherited nervous system disorder characterized by development of leg weakness, spasms and stiffness. While generally acknowledged that health and social care services can minimise symptoms and improve quality of life, there is a lack of research exploring this from the perspective of people affected by HSP. This qualitative study explored the users and providers experience of using rural services. **Method:** Focus groups and interviews were undertaken of people with HSP ($n = 14$), carers ($n = 6$) and professionals ($n = 12$), to describe their experience of service provision and to suggest improvements for care. These were taped, transcribed and analysed. **Results:** Four themes emerged: (1) Diagnosis, symptoms and finding support; (2) Therapy, treatment and the delivery of care; (3) Managing the disease together; and (4) The way forward. **Conclusions:** Rehabilitation and support for self-management is valued by those affected with HSP throughout the disease trajectory from diagnosis onwards. Key to this is the development of a partnership approach which includes carers. Single point, well-informed, gatekeepers may enhance the coordination and delivery of care in rural areas. These findings underline current guidance promoting a holistic approach for people with neurological conditions.

► Implications for Rehabilitation

- People with HSP see as a high priority the need for additional neuro-physiotherapy services and emotional support services.
- Single point, well-informed, gatekeepers are essential to coordinate care, especially with rare diseases.
- Health professionals need more education and training about the disease process and its consequences for people affected by HSP, including the carer burden.
- People with HSP require better self-management advice, information and support.

Background

Hereditary Spastic Paraplegia (HSP) (also known as Familial Spastic Paraparesis) is an inherited central nervous system disorder typically characterized by gradual development of muscle weakness, spasms and stiffness of the legs, which may be first noticed in early childhood, or at any age through adulthood [1]. It affects approximately 3–9 in 100 000 [2] of the population. In some forms of the disorder, bladder symptoms also occur, or the weakness and stiffness may affect other parts of the body. These symptoms can impact significantly on the person's life, for example, by causing difficulties with their balance and mobility. People with HSP do not die of the disease itself but live with increasing deterioration in physical and sometimes neurological ability [3]. Much of the research in

relation to HSP discusses the genetic and physical manifestations of the disease [1]. There is some evidence that health care services and other supportive agencies can provide helpful input to minimise the impact of these symptoms, and to improve the quality of life of persons affected by HSP [4]. However, our literature search highlighted a lack of studies which have explored the perceived usefulness and availability of these services for HSP, particularly in rural areas.

Access to appropriate health care in rural areas is a major issue for health care providers across the Western world [5,6]. In a review of rural service provision in the United Kingdom, accessibility was shown to affect health status [7], and an American study found that access to transport improved health status in rural areas [8]. Recently, in the United Kingdom, attempts have been made to describe urban/rural differences using a range of classification based on population size and such descriptors as ‘hamlet’ and ‘village’ [9], but this definition did not consider access to health care. However, in a review of the literature conducted in Norway [5], the authors found that, despite the problems of definition of the term *rurality*, several approaches seemed to have relevance to the provision of health

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care for rural communities and these included telemedicine, managed care pathways and integrated medicine. These were not cheaper alternatives but enabled continuity of care for people living in rural areas. With chronic conditions such as HSP, these approaches may have value. Understanding the experience of living with HSP in a rural area and the specific issues relating to access will help to identify planning priorities for health care providers.

Study aim

This study aimed to explore the experience of health care provision, in a rural area of the United Kingdom (UK), from the perspective of both the service users and providers. A qualitative approach is widely recognised as being an effective methodology for achieving this aim [10]. To achieve this, we intended to ask participants about their degree of satisfaction with current service delivery; the mechanisms of support available for people with HSP and their carers; and current management strategies for people with HSP. Following these discussions, we were interested in gathering suggestions for service improvements from professionals, carers and people with HSP.

Research approach and methods

The project was overseen by a research group which included a person with HSP, a specialist in HSP a specialist in rehabilitation and two independent qualitative researchers.

A qualitative research approach, using focus groups and interviews was used. This methodology has been demonstrated to facilitate problem identification and the exploration of different types of solutions [11]. Importantly, focus groups have shown to be useful in examining not only what people think but how they think and why they think that way [12]. It was our initial intention to use focus group sessions to actively encourage participants to talk to one another, to ask questions, exchange anecdotes and to comment on each other's experiences. However, following several unsuccessful attempts to organise a suitable time for neurologists and medical practitioners to meet as a group, it was decided instead to conduct individual interviews with these professionals to ensure their perspective was included. The same topic guide was used for the focus groups and for the individual interviews. The benefits of individual interviews are they offer the researcher a greater opportunity to explore, in depth, specific issues raised by the interviewee [10]. The limitation of using this method was that there was no possibility of creating debate about the issues raised.

Sampling

Purposive sampling [13] was used to gather the views of participants from different perspectives. Our aim was to achieve

maximum variation by including people with HSP with a wide range of ages and disability, carers of different genders and relationships (e.g. husbands, wives, daughters) and professionals with a range of experience and from different disciplines.

Recruitment

People with HSP were recruited via three avenues: (1) a letter sent to all members of the HSP society within the rural area of the UK ($n = 30$), (2) following discussion about the aims of the study at a local HSP Support Group meeting and (3) via an advert on the HSP Society website and in the National HSP newsletter. In each situation, interested individuals were provided with information sheet and contact name should they wish to participate in the study. Any person with a confirmed diagnosis of HSP or their carer, a minimum age of 18 years, who were able to travel to a focus group session and provide informed consent, was eligible for inclusion.

Any health care professional who was currently involved in providing services to people with long-term neurological conditions, and who at some stage had provided care for someone with HSP, was eligible to participate in this study. They were recruited via adverts placed on the University website, and the websites of a range of professional bodies and special interest groups (Table 1).

Ethical approval

This study was ethically approved by the South West NHS Ethics Committee (Rec ref 08/H0202/108) and Plymouth University, Faculty of Health Research Ethics Committee.

Data collection

The focus groups. Fourteen people with HSP, 6 carers and 12 health professionals took part in this study. Six focus groups were conducted: two of each sub-group, in an attempt to capture a broad spectrum of views. This was a pragmatic decision based on the relatively small number of people affected by this condition. In line with the literature [14], we endeavoured to ensure a minimum of 4 and maximum of 12 participants per focus group; however, one of the carer groups only involved 3 people. We felt it was essential to run two separate carers groups to compare and contrast the responses from the two groups and to reduce the possibility of one-person dominance which sometimes occurs in focus groups [15].

The focus group sessions, which lasted approximately 90 minutes, were conducted by experienced qualitative researchers, one acting as observer. The role of the observer was to study the interaction amongst the focus group members, especially focussing on body language. Her notes were used during analysis to consider the context in which comments were made, whether

Table 1. Characteristics of health care professionals.

| Profession | No. participants | Gender | Client load | No. years in practice | No. pwHSP seen each year |
|----------------------------------|------------------|--------|-----------------------------------|-----------------------|--------------------------|
| Specialist Neuro-Physiotherapist | 1 | Female | Mixed neurological | 10–15 | 0–5 |
| Specialist Neuro-Physiotherapist | 1 | Female | Mixed neurological | 15+ | 5–10 |
| Physiotherapist | 1 | Female | Mixed neurological | 5–10 | 0–5 |
| Physiotherapist | 1 | Male | Mixed neurological | 5–10 | 0–5 |
| Physiotherapist | 2 | Female | Mixed neurological | 10–15 | 0–5 |
| Physiotherapist | 1 | Female | Mixed neurological | 15+ | 0–5 |
| Specialist Neuro-Nurse | 1 | Female | Mixed neurological rehabilitation | 20–25 | 0–5 |
| Specialist Genetic Nurse | 1 | Female | Genetic diseases | 10–15 | 5–10 |
| Neurologist | 2 | Female | Mixed neurology | 15 | 5–10 |
| General Practitioner | 1 | Female | Generic | 5–10 | 0–5 |

Table 2. Topic guide for people with HSP.

| Tell me about: |
|---|
| – your experience with health care services that you have used in relation to management of Familial Spastic Paraparesis. |
| – how satisfied you are with the health care services you currently receive. |
| – whether there are any additional/alternative health care services that you might find useful to help you manage your condition. |
| – the mechanisms of support that are available to you and/or your partner/family members/close friends. |
| – whether there are other mechanisms of support that you would find useful in helping to manage your condition. |
| – whether you have any suggestions for improvements to these support services. |
| – the current strategies you use to manage your condition. |

or not participants showed an emotional response. The observer's comments added breadth and depth to the data. To maximise recruitment and minimise burden of extra travel for the participants affected by HSP, the focus groups were undertaken on the same weekend as the HSP residential Annual General Meeting. This enabled the people with HSP to be interviewed on the first day and their carers on the second. Although there was a risk that issues from the focus groups would be discussed overnight, which might affect the data, our primary responsibility was to the participants' well-being. The focus groups of health care professionals were undertaken at a site close to their place of work for their convenience.

The interviews

Three interviews were undertaken; two neurologists and one medical practitioner. These interviews, which lasted up to an hour, were conducted by the qualitative researcher who facilitated the focus groups (JG). They were held at a time and a place convenient to the interviewees, typically their work place.

Demographic data was gathered in all participants via a brief self-report questionnaire, enabling us to describe the sample and monitor the effectiveness of our maximum variation sampling strategy. The topic guides, which comprised a series of open-ended questions, were based on (1) a review of the literature and (2) discussions with people with HSP, carers and health professionals. Having prepared a draft topic guide the questions were discussed with the lead for the HSP local society, and a nominated health professional. By ensuring close user involvement from its outset, we were confident that the research topic and focus group questions were meaningful and relevant. Refer to Table 2 for the topic guide used with people with HSP.

Analysis

Focus group audio digital recordings were transcribed verbatim and checked for accuracy. Names of persons and places anonymised. We used thematic analysis to code the data [9]. Two transcripts were coded by two experienced members of the research team, one of whom had not been present at the focus group, and compared for accuracy. This involved the researchers identifying parts of the text which identified the primary content, or meaning, relevant to our research aim. The coding was similar, and the meaning derived the same by each of the researchers. Following coding of all transcripts, the codes with similar meanings were grouped into first themes, which were then discussed with the research group, who used their clinical and personal experience to decide whether the themes reflected experience. The themes were then re-categorised and named by again re-ordering the codes.

Table 3. Characteristics of people with HSP.

| Age | No. participants | Marital status | Time since diagnosis |
|-------|------------------|----------------|----------------------|
| 26–45 | 2 | Married | 0–5 |
| 26–45 | 1 | Married | 6–10 |
| 26–45 | 1 | Single | 11–25 |
| 46–60 | 2 | Married | 6–10 |
| 46–60 | 2 | Married | 11–25 |
| 46–60 | 1 | Divorced | 11–25 |
| 46–60 | 1 | Married | 26–40 |
| 61–90 | 1 | Widowed | 6–10 |
| 61–90 | 3 | Married | 11–25 |

Table 4. Characteristics of carers.

| Relationship | Age (years) | Marital status | Time since diagnosis of partner (years) | No. participants |
|--------------|-------------|----------------|---|------------------|
| Spouse | 61–90 | Married | 11–25 | 3 |
| Spouse | 61–91 | Married | 26–40 | 1 |
| Spouse | 26–45 | Married | 0–5 | 1 |
| Spouse | 61–90 | Married | 6–10 | 1 |

Results

In total 14 people with HSP, 6 carers and 11 professionals participated in this study; their characteristics are summarised in Tables 1, 3 and 4.

Four overall themes emerged: (1) Diagnosis, symptoms and finding support; (2) Therapy, treatment and the delivery of care; (3) Managing the disease together; (4) The way forward.

Theme 1 – “... the first word that I heard was ‘spastic’”: diagnosis, symptoms and finding support

The genetic nature

This theme explored the genetic nature of HSP, the methods used by people with HSP and their carers to manage their condition and their support mechanisms. Considerable discussion centred around the genetic consequences of the condition. Some had first noticed symptoms in their extended family and had decided to investigate the causes of their own symptoms. Others were referred to a geneticist post diagnosis to discuss the possibility of passing on their affected genes to their children. Understandably, this was a stressful time, which was aggravated by the time taken for the test results. The two main areas of concern were the uncertainty of not knowing if and how the disease would manifest itself and the degree of likelihood the disease markers would be passed on should they have children.

The neurologists interviewed recognised these anxieties and viewed the genetic service both as an important initial stage in the patient pathway post diagnosis, and as a resource for patients and their relatives to ask questions. The specialist nurses felt their responsibility was to work with families and maintain their own knowledge about the disease so they could respond effectively to the information being sought by those affected by the disease. This required maintaining a long-term relationship with the families by “keeping the families on their books” and being aware that several members of one family may need their advice and support over many years.

Two different approaches to discussing their diagnosis and its implications with their children were described by people with HSP and their carers. While some chose not to tell their children about the faulty gene, others felt it is important to do so in order that they could “be prepared” and “live life to the full before and if symptoms appeared”.

The accurate diagnosis of a long-term degenerating neurological condition is often complicated, requiring exclusion of a range of differential diagnoses. Nevertheless, the delay in determining the cause of the presenting symptoms was viewed by the participants as unnecessarily slow, and was sometimes interpreted as showing a lack of understanding from the professionals. However, the interviews with the neurologists conveyed a clear understanding of the recognised need for an accurate diagnosis to be provided in a supportive and timely manner. Their key concerns were that, once diagnosed, their patients would not have the specialist support services they needed locally.

The difficulty in sensitively conveying the diagnosis of a disabling long-term condition is widely acknowledged [16]. In the case of HSP, the shock of diagnosis was further emphasised by the name of the disease, which strongly reminded people of childhood taunts:

... the first word that [I] heard was “spastic” because in our age group a spastic was, you know you laugh at kids down the street, you know ha ha you’re a spastic, and you know it was a rude name that you called somebody and so ... the only word he heard was spastic. (Carer 1)

Potential stigma associated with the disease was not confined to the initial diagnostic phase:

Because spasticity is incredibly visible as a disability and because of the spasms it draws attention. So you can’t hide... But with HSP you can be sat, maybe sat here now, and you would not know. And then they stand up and you know the gait... I think they probably had to build up a lot of defences over their lives about people looking at them with a disability out there in society. (Health care professional)

Symptoms

The people with HSP also described the many everyday difficulties they faced in managing their progressively deteriorating physical symptoms, from the first signs ‘I started to walk a bit funny’ to the later symptoms such as falls and loss of balance. To avoid tripping, adaptations had been made within their own homes, such as removing mats and excess furniture. At work, managing symptoms could be more problematic. For example, incontinence meant the need for planning ahead and dealing with the symptoms in an environment where people might not be so supportive. It was clear that the issue caused embarrassment:

Increasing incontinence is an issue with this thing and it’s one that I don’t think any of us like to talk about. I don’t know how many times we talk about it in our group but I would be very reluctant to and yet in my mind it’s a hot issue. (Person with HSP 1)

Preparation and constant planning, by both the person with HSP and the carer, were essential in effectively managing the physical aspects of the condition; various ways of coping were described. Planning ahead on a daily basis involved factoring in preparation time, gathering information about the accessibility of the destination and the availability of toilets and parking. In the long term, the planning involved access to a range of mobility aids and home adaptations. Even with careful planning participants talked about the difficulties facing unforeseen obstacles. The response to such unplanned events

was often described as “frustrating” or “depressing”. For example:

... We were struggling, it was freezing cold, we had to walk around quite a way. And then we were faced with this huge array of steps. There was no entrance for disabled, there’s no wheelchair access and I can’t, that’s a public building that annoys me that there’s nothing. (Carer 2)

The rarity of the condition meant that there was a need to respond to the perceptions and enquiries of others and this was dealt with in various ways. Participants used various strategies to avoid long explanations. For example, some chose to say they had a more common condition such as arthritis that might be more readily understood, others blamed their symptoms on accidents. They recognised this was unsatisfactory but described it as easier than repeated efforts to explain and deal with further questions.

Finding support. In response to these pressures, people with HSP and their carers expressed the need for psychological and emotional support, ideally by someone who understood the condition, but at least by someone with experience in neurological conditions. The professionals were also keen that psychological support should be provided by neurologically trained professionals.

... we have to think about what type of psychologist. Because we don’t want a psychologist who comes in and just does testing and then doesn’t feed back to the team anything relevant. (Health care professional)

The need for professionals who understood neurological conditions was constantly revisited. Lack of local specialists was blamed on the rarity of the condition and the problems of living in a rural location. In response to the lack of availability of local expertise, many people had turned to the internet for information. Others had travelled hundreds of miles to access specialist care, despite this being both onerous and inconvenient. With these issues in mind, it was clear that all the participants, both those affected by HSP and the professionals, recognised the value of the local HSP support group which was seen as central to sharing information and experience.

I find it (the HSP Support Group) invaluable as a carer because we’ve spent a lot of time talking and you know other carers we all talk. You know we all talk and we’re all going through exactly the same frustrations and everything else and the anger and the depression we get as carers. (Carer 2)

Theme 2 – “I think our coordination could be much better”: therapy, treatment and the delivery of care

Gaps in service

While positive stories of care delivery were recalled by those with HSP and their carers, nevertheless it was not uncommon that services failed to deliver the care they hoped for. The professionals identified a lack of specialist services for this client group and highlighted a general lack of coordination of health care for people with long-term conditions. One neurologist commented:

I think our coordination could be much better. I don’t meet regularly with the therapists involved in seeing my patients so we communicate at an arm’s length, really by letter. ... the communication is rather distant and not particularly swift. (Neurologist)

Lack of time, a “tradition” of poor coordination across disciplines and a lack of electronic patient notes were some of the reasons cited as contributing to this confusion in coordinating care.

Participants commented that communication and coordination of services became even more problematic when they had to access care from both the public and private health care sectors. Dealing with the differing philosophies of these sectors raised an added complication for people with HSP. For example, some people had used various complementary therapies even when they might conflict with treatment and advice from the medical health care system.

Access to services

Another issue of concern was the level of care provision for people with HSP, and the variation in access to care services. The majority of the professionals interviewed felt that people with HSP should be responsible for managing their condition on a day-to-day basis and only access specialist services, for example, when they needed prescribed treatments or exercise programmes. This “patient-led” service had mixed responses from the participants with HSP and their carers. They felt this pressure to always find care and search for therapy themselves created a tension between being independent and yet wanting to be cared for in a more hands on way.

I think once you’ve come out of their office and you’ve been diagnosed they don’t want to know, you hit a brick wall, they’ve done their job, that’s your condition and that’s it, you’re back to your GP and your GP’s got 2000 odd patients and that’s it. (Person with HSP 2)

Helpful therapy

All the participants stressed the importance of physiotherapy in relation to accessing up-to-date information and support to relieve symptoms, especially in the absence of drug treatment. However, finding the “right” physiotherapy service was a major problem. While some advantages were cited in favour of hospital-based physiotherapy (such as ease of communication with neurologists in monitoring medication), there was a general agreement by the professionals that community-based physiotherapy may better meet peoples’ needs by bringing care closer to home. The physiotherapists themselves emphasised the need for specialist over generic approaches to care, in part due to the general lack of understanding about the condition given its relative rarity. There were many descriptions of people’s condition deteriorating when in hospital, because of a lack of appropriate rehabilitation and/or inappropriate drug treatment.

A wide range of privately provided complementary therapies (e.g. acupuncture, hydrotherapy and hypnosis) and leisure services were accessed to reduce the effect of symptoms such as pain or fatigue they felt that the relaxing effect of some therapies such as massage helped their mobility in the short term. Participants also described the problems they had finding regular fitness classes appropriate for their symptoms and the need for independent physiotherapists who understood their condition.

Theme 3 – “I’m not his carer I’m his wife”: managing the disease together

Carer definition

During the course of the carer’s focus groups, it became clear that the people we had described as “carers” rejected this description, believing that this disease needed to be managed together as a couple. They described how both of their individual lives had

been limited by the condition in different ways. They cared for each other and worked together to best manage the disease. There were many descriptions of how HSP had affected the experience of being a couple:

I know that he will never dance with me again and that’s heart-breaking. (Carer 3)

Together they had found new interests and adapted their home to meet the increasing needs of the disease and to maintain a normal family life in which the person with HSP could be completely engaged. Nevertheless, alongside this the carers noticed a change in the balance of power in the relationship. They observed that this was not only because they were the able-bodied partner, but also because the person with HSP appeared to be less interested or able to make decisions. There were general discussions amongst the carers about whether this was related to the condition itself or whether their partners were “losing interest in the world”. Carers used words such as “patience” and “frustration” to describe their experience of how their roles had changed, and the restrictions that had been placed upon both of their lives:

You can’t let it eat you up. You’ve got to; you’ve to let the anger out haven’t you? Because otherwise you can’t function with the job that you’ve got to do you know? (Carer 4)

Carer needs

Despite the constant anxiety caused by being vigilant about the needs of the person with HSP, the carers were rarely asked by health care professionals about their own needs. They recognised that in terms of priority:

We come bottom of the pile. (Carer 4)

When carers were asked what services they would like, there was general interest in keep fit classes because they felt they had to be the “muscle” in the relationship, for example being strong enough to push wheelchairs or lift their partner. The classes would need to be at a time that would fit with the primary focus of their attention, their partner.

Theme 4 – “. . .there’s no point in managing a void, lets fill it”: the way forward

Care coordination

All the participants were in agreement about the need for a “named gatekeeper” to coordinate care and be a central reference for people with HSP, their carer’s and the professionals providing care. This person could be either from health or social care services, but they had to understand the condition and be well-informed about local and national services available to meet the needs of their client group.

In terms of clinical specialists, participants recognised that it was unlikely that there would be a local specialist nurse or doctor for this specific condition alone because of its rarity, but that there were possibilities for neuro-specialists to take a special interest in the disease. The location of these neuro-specialists was particularly relevant to people with the condition living in more rural areas. Many described how hard it was to get to the specialist centres which were always located in large cities, and how difficult it was to get around once there. Whilst they recognised that specialist centres would provide them with the latest treatment they wanted on-going treatment in the form of neuro-physiotherapy and neuro-rehabilitation closer to home.

Poor coordination

In discussing how best to develop local secondary care services able to respond to the needs of this particular group, the professionals raised a number of concerns, often related to poor communication and a lack of coordination of care. For example community physiotherapists were not always informed when patients were admitted to hospital, which prevented them from providing advice to the hospital-based clinical teams about the individual's needs.

The instigation, in some areas, of a neuro-care pathway had been successful in enhancing the cross flow of information between community and acute care, although it was acknowledged that better systems were required throughout the patient pathway from the point of diagnosis to ensure people did not “fall through the net”.

When asked about what might be the causes of poor information and coordination of care, one professional said:

Well, money number one, two, three, four and five... If you've got a big patient group like MS patients you can set up an MS service in [the South West]. Something as rare as HSP over an area like the peninsular [a large geographical area], well that's tough, that's difficult. (Health care professional)

Discussion

The aim of this research was to understand the experience of people affected by HSP living in a rural area, and investigate how current service delivery was meeting their needs. The physical restrictions and symptom manifestations created daily challenges wherein the carers and people with HSP work together to manage their everyday lives. This is a familiar situation to others with neurodegenerative diseases [17–19].

There was a wide range of physical manifestations reported which reflected the neurological effects of the disease; fatigue, trips, falls and incontinence were amongst the most often highlighted. The participants discussed how these symptoms restricted their lives, for example, making outings something to be planned well in advance, and provoking anxiety for the carers when leaving their spouse alone. This “constant awareness” and living with disease affected both the person with HSP and the carer, and there were requests for, amongst others, physical support services and emotional support. Finding support services appropriate to the needs of specific disease groups has been shown to be an extra stress on people already having to manage a range of problems [20]. The professionals agreed that appropriate emotional support should be available, suggesting it should be more akin to a listening service than one that measured levels of depression in individuals. There was listening support available through the HSP group and there was general agreement that the support group offered a chance to share problems and experiences and this was beneficial.

The patient journey for many long-term neurological conditions can be difficult and has been well described [21,22]. People with HSP are no exception. Many described the anxiety created firstly in finding the right diagnosis and then in attempting to find treatment. In line with other studies exploring rural services for people with long-term conditions [23], the tendency towards more centralised rehabilitation services meant long distances had to be travelled to access services, with fewer opportunities to receive rehabilitation services closer to home. As mentioned at the beginning of this article, tele-health is a potential method of closing these gaps in service delivery allowing access to city-based services in more distant locations [24,25], although the participants in this study, in the absence of alternatives, were keen

to access face-to-face consultations sometimes travelling from the South West to London to do so. It also has the additional potential value of allowing rurally based health professionals to become better informed about the disease and its management options by attending the tele-based consultations. A systematic review of cost-effectiveness studies of telemedicine interventions found that this area needs to be further investigated [18].

In line with the experience of others living with a rare condition [26], those affected by HSP felt that local medical practitioners and therapists often did not have sufficient knowledge about the disease to provide appropriate guidance. Therefore, once their condition had been assessed and initial treatment offered people with HSP and their carers felt they were “on their own”. The lack of specialist knowledge about the condition may result in poor self-management advice being provided.

To maintain control in dealing with health service providers, people with HSP need a clear and positive sense of themselves [27]. In achieving this they are in a better position to negotiate appropriate care. As their condition deteriorates, this sense of self is constantly changing and the health care provider's role is to help enhance the possible by offering rehabilitation and maintaining existing abilities. In addition, access to programmes such as the UK-based expert patient programme that aims to improve peoples' self-efficacy by enabling them to manage their health condition in collaboration with health care professionals may help to reduce and avoid these feelings of isolation in managing their condition [28] and their health.

With regard to rehabilitation interventions, people with HSP often tried a multiplicity of local complementary therapies. Some found these of short-term benefit but they were expensive and decisions had to be made about what had to be removed from the family budget in order to pay for them. The balance between professionals feeling the “onus was on the patients” to access care when they needed it and the person with HSP's need to feel they were “doing something to help themselves” caused distress, which was further increased because of the travel involved in accessing care.

Several people with HSP commented that clinicians' lack of knowledge of HSP had negatively affected their hospital experience and continuity of care following discharge. Of concern was the need to track HSP patients when admitted to hospital. Early physiotherapy intervention was considered important to maintain their physical strength, flexibility and mobility and to liaise with the medical team to ensure appropriate drug treatment.

In combination with pharmacological management of symptoms such as spasticity and incontinence, physiotherapy is a key component of care [29]. Both people with HSP and the health care professionals considered neuro-physiotherapy to be crucial to maintaining their mobility. Specific treatments such as FES were not always available and while acknowledged as not being suitable for everyone, people with HSP felt they wanted better access and availability. This finding is in line with the views of people with other neurological conditions [30].

In the third theme “managing the disease together” carers discussed the negative emotions they felt when caring was described as a burden. They felt it de-personalised the act of caring and believed that their partner would have cared for them had they been in a similar position. Nolan [31] talks about this shared care as “reciprocity” and points out that each family situation needs to be assessed independently to understand the caring dynamics. Some studies have shown that the long-term caring role can be associated with significant depression [32], suggesting that even though the caring may be conducted out of love and have mutual benefits, the carers' quality of life can be significantly diminished in long term. Finding the right balance

between attentiveness and enabling their partners to still feel in control was a concern for carers and the need for a balance is confirmed in other research [33].

In conclusion, accessing appropriate treatment whilst living in a rural area appears to be a major concern for people affected by this rare progressive neurological condition. Some suggestions to improve the quality of care include: the allocation of a named coordinator of care to optimise coordination of services and enhance communication; improved education of health professionals regarding the assessment and management of both the physical and emotional aspects for people with the condition and carers alike, in part through access to tele-health services; and increased access to neuro-physiotherapists locally.

Limitations of the research

As with all qualitative research, this study reports on the views and opinions of a self-selected group of people with HSP, their carer's and health care professionals. As such the results are not generalisable to all people with HSP living in rural localities. What has become clear however is that many of the points raised have been identified as important by government guidelines [34], and research into people with neurodegenerative diseases [35]. The value of this research therefore is to further emphasise the need for appropriate timely information, coordinated care and services provided by staff who are willing and able to listen to the individual needs of patients.

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Declaration of interest

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