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ORIGINAL RESEARCH

The Lived Experiences of People with Progressive Supranuclear Palsy and Their Caregivers

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ABSTRACT

Introduction: Progressive supranuclear palsy (PSP) is a neurodegenerative disorder initially characterised by disturbances in gait, balance and posture, with death occurring after several years of progressive physical and cognitive decline. This, along with a low index of suspicion, a high degree of diagnostic uncertainty and no approved treatment options, greatly impacts the lives of patients and caregivers. This research was conducted to (i) gain insight into the clinical and emotional journey of patients with PSP, (ii) assess experiences and perspectives, (iii) understand disease impact and (iv) identify key challenges and unmet needs.

Methods: A literature search and qualitative interviews with six PSP experts were conducted to map the clinical pathway and identify breakpoints. The pathway was validated by key opinion leaders in seven countries. Qualitative research was conducted over 6 months in seven countries with PSP stakeholders ($N = 112$) to explore the emotional journey. The approach included self-ethnography, 60-min telephone interviews and the completion of 7-day smartphone diaries.

Results: The current PSP clinical journey can take many different pathways, with patients

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cycling through the healthcare system before a correct referral is made and a possible/probable diagnosis received. Breakpoints contribute to delays in accessing appropriate clinical care, a high degree of diagnostic divergence and suboptimal management of the disease. The emotional journey is dominated by negative feelings, although some moments of positivity were noted. The research highlighted a lack of disease understanding amongst all stakeholders and a lack of support for patients/caregivers. The authors make a number of recommendations for care improvements, including longer consultation times, closer collaboration among healthcare professionals and patient organisations, and more varied support and information for patients/caregivers.

Conclusion: This work represents a major collaborative effort to understand the lived experience of PSP. The research illustrates that a coordinated effort from all stakeholders is required to address ongoing needs and challenges within PSP.

PLAIN LANGUAGE SUMMARY

Progressive supranuclear palsy (PSP) is a rare brain disorder caused by damaged nerve cells. PSP is often misdiagnosed as Parkinson's disease. Sufferers typically have issues with walking, eye movement, mood and memory, all of which worsen over time, and they often become entirely dependent on caregivers. Sadly, there is no cure, but day-to-day living can be supported. In this study, the researchers wanted to understand the lived experience of patients and families. First, researchers collected information from published sources about what it is like to live with PSP. Then they spoke with PSP experts, key opinion leaders, patients, caregivers, patient organisations, neurologists and nurses in the UK, France, Italy, Germany, Spain, the USA and Japan. This revealed important learnings about the clinical and emotional journey in PSP. There is a need for patients, caregivers and healthcare professionals to have open dialogue and build trust. Moreover, a closer collaboration between patient organisations and healthcare professionals could lead to improved care. Caregivers emerged as invisible heroes, and PSP care must prioritise support for them, in addition to patients. This study provides invaluable insights into the lived experience of patients and caregivers, as well as recommendations for supporting their clinical and emotional journey.

Keywords: Progressive supranuclear palsy; Neurodegenerative disorder; Unmet need; Quality of life; Clinical journey; Emotional journey; Lived experience

Key Summary Points

Progressive supranuclear palsy (PSP) is a rare neurodegenerative disorder that is under-recognised and under-researched and, accordingly, the unmet needs in PSP are vast and multifactorial. There is little published literature about the experiences and perspectives of people living with PSP, caregivers and healthcare professionals (HCPs) throughout the patient journey.

The current clinical journey of a patient with PSP is characterised by a pattern of cycling through the healthcare system, a delayed and terminal diagnosis, absence of treatment and rapid disease progression. The emotional journey is dominated by negative feelings, especially around diagnosis, although there may be some moments of positivity. It is essential that patients, caregivers and HCPs take the diagnostic journey together and that HCPs communicate clearly what is known and unknown at specific timepoints, and future prognostic expectations, even if not optimistic. Tailored education is needed for all parties and a coordinated effort required from all stakeholders in the lobby for better, more supportive care of people living with PSP and their caregivers.

INTRODUCTION

Progressive supranuclear palsy (PSP) is a rare, relentlessly progressive neurodegenerative movement disorder associated with a range of motor and cognitive symptoms, as well as shortened survival [1–6]. It is an under-recognised and under-researched disease, and accordingly, the unmet needs in PSP are vast and multifactorial. Obstacles to improved management of PSP include a low index of suspicion (due to its rarity), a high degree of diagnostic uncertainty and delay, lack of approved therapeutic options, validated biomarkers or specific imaging, as well as inconsistent access to appropriate care; all of these greatly impact the lives of people living with PSP and their caregivers [5, 7–9].

The sensitivity and specificity of clinical PSP diagnoses can be poor, particularly for the rarer or more atypical subtypes of PSP, and the resulting diagnostic delay may impact prognosis and management [5]. The diagnostic accuracy for PSP-Richardson's syndrome is higher than for other clinical syndromes of PSP [10], where definitive clinical features may not occur for many years, or ever. This period of uncertainty is challenging for patients and caregivers and, while diagnostic care for PSP has reportedly improved over the last 3 decades, some may still wait more than 2 years for a diagnosis [11]. Studies looking at the accuracy of PSP diagnosis found that PSP was correctly diagnosed at the first clinical visit in 19% [12] and 25% [13] of patients, respectively; 63% of patients were correctly diagnosed at the final visit and Parkinson's disease (PD) was the most common clinical misdiagnosis [13]. It is worth noting that while clinical diagnosis may have improved over the years, the most accurate and authoritative diagnosis derives from examination of brain tissue, and resultant autopsy report.

A survey conducted in 2016 by PSPA (PSP Association), a UK-based charity, identified a lack of understanding from healthcare professionals (HCPs) that at times led to an incomplete or insensitive explanation of PSP at diagnosis by the neurologist. This impacted how patients and caregivers coped with diagnosis and disease management moving forward. Survey results are available directly from PSPA (info@pspassociation.org.uk). The unrelenting disease progression affects not only the patient but also their caregivers, as the patient's loss of independence means increased reliance on assistance with the normal activities of daily living [14–16]. In addition, symptoms such as behavioural and mood changes, difficulty interpreting emotions, cognitive decline and sleep disturbances adversely affect interpersonal relationships and can cause psychological strain in all parties involved, further increasing the burden on family and caregivers [7, 14, 17–20]. A broadening awareness of these burdens has led to caregivers being described by the clinical community as the 'unrecognised sufferers', 'silent heroes' or 'invisible patients'.

PSPA and CurePSP provide online resources and support services for people with PSP and their caregivers. PSP-focused education for HCPs, including for allied HCPs and the primary care team, is also available through both charities. Specifically, PSPA has an interactive, evidence-based online resource for HCPs that provides information on PSP, sets out the standards of care, outlines symptoms and provides links to additional resources [21]. CurePSP have developed their 'signature publication' guidebook, a 150-page reference document for patients, their families/caregivers and HCPs that provides the latest information on PSP, including chapters on the management of symptoms, managing care and getting professional support, being a caregiver and making plans [22]. There is also research underway at Cardiff University (Cardiff, UK) looking at the impact of caring responsibilities for people living with a progressive condition. Caregivers of those living with PSP have been invited to take part [23].

A deeper level of understanding into how a person living with PSP moves through the healthcare system, touchpoints with HCPs and, importantly, key breakpoints within this pathway would constitute a valuable contribution to existing available resources. Additionally, a clearer view of the emotional journey, both positive and negative, that patients and their caregivers may experience could aid dialogue

between the patient/caregiver and the physician and provide much-needed support to those impacted by PSP.

Current Study Objectives

To obtain deeper insight into the clinical (healthcare) and emotional journey of patients with PSP from early symptoms to end-of-life. To assess experiences and perspectives of patients, caregivers and HCPs throughout the patient journey, to understand how the disease impacts on each and to identify their key challenges and unmet needs. To identify how needs can be met, how disconnects can be overcome and how patient and caregiver experiences may be improved.

METHODS

Mapping the Clinical Journey

Firstly, a literature review was conducted in 2020 to help understand the clinical journey and identify breakpoints (described as 'a current situation that is believed to represent a barrier to optimal care for people living with PSP'). PubMed, Google Scholar, the Education Resources Information Centre (ERIC) and the Cochrane Database of Systematic Reviews were searched for "Supranuclear palsy, progressive" AND any one of the following: "guideline" OR "guide" OR "education" OR "training" OR "learning" OR "need" OR "educational needs" OR "needs assessment" OR "healthcare needs" OR "health services needs" OR "challenge" OR "challenges" OR "professional practice gap" OR "clinical practice gap" OR "practice" OR "programme" OR "continuing education" OR "professional development" OR "continuing professional education" OR "disease management" OR "gaps" OR "barriers" OR "stakeholder" OR "knowledge" OR "attitude". Only English-language results were included. Randomised controlled trials, systematic reviews, meta-analyses, literature reviews, HCP surveys and patient surveys were all eligible for inclusion, provided that they contained information reporting on practice gaps. Following this, a series of 1-h qualitative phone interviews was conducted with six experts in PSP (four clinical experts [based in Belgium, Germany, the UK and the USA], a clinical social worker [based in the USA] and a representative from the PSPA [based in the UK]) to discuss the common clinical pathway in PSP, key breakpoints, insights into how best to address the breakpoints and whether these aligned with the six experts' experiences. The clinical journey was updated following these discussions and was presented to key opinion leaders (KOLs) in seven countries (UK, France, Italy, Germany, Spain, the USA and Japan) for subsequent validation.

Exploring the Emotional Journey

Secondly, qualitative research was conducted in seven countries (the UK, France, Italy, Germany, Spain, the USA and Japan) in the latter half of 2020. A holistic approach to the research was taken with the inclusion of different PSP stakeholders, resulting in 112 respondents overall; these included 21 pairs of patients/caregivers, seven patient organisation (PO) representatives, 42 KOLs, and 21 nurses. KOLs were recruited if they were neurologists or movement disorder specialists (MDSs), treatment decision makers, not affiliated with the pharmaceutical industry, in clinical practice for 3–40 years and spent at least 50% of their time in direct patient care. Similarly, nurses were recruited if they were practising between 3 and 40 years and caring for people living with PSP on a regular basis. Self-ethnography was used to encourage patients/caregivers to use self-reflection to capture their personal experiences of the disease. This research was conducted over 6 months in close collaboration with POs who identified patients living with PSP and their caregivers. The approach included 60-min in-depth telephone interviews in each country and 7-day smartphone diaries from patients/caregivers. The smartphone diaries sent daily prompts to participants reminding them to 'Capture a moment' and 'Complete my daily survey'. 'Capture a moment' required

participants to upload a video or photograph from the previous 24 h that described the patient's life and to supplement this by typing or recording a description of the emotional impact (either positive or negative) this moment had on the patient or caregiver. The daily survey comprised nine questions, taking no more than 5-min to complete, evaluating daily condition, mood (feelings and emotions), motor and non-motor symptoms, daily activities and time spent providing care. A simplified version of Robert Plutchik's psychoevolutionary theory of emotion was used to classify general emotional responses [24].

An overview of the methodological approaches used to investigate the clinical and emotional journey in PSP is shown in Fig. 1.

Compliance with Ethics Guidelines

According to criteria within the EPHMRA code of practice (Sects. 1.1 and 1.2), this study is defined as market research. As such, consistent with the guidance issued by the British Healthcare Business Intelligence Association (BHBIA) and the EPHMRA code of practice (Sect. 1.3), neither ethics committee approval nor clinical trial registration was required. Informed consent, including consent to publish the study findings, was obtained from all study participants prior to the study. Additionally, consent was sought from all participants to publish verbatim responses.

RESULTS

The Clinical Journey with PSP

The initial literature review yielded 195 unique 'hits' that, upon review of the abstracts, provided 25 relevant articles (Supplementary Material). These contained little information on the clinical pathway for patients living with PSP, highlighting the lack of published evidence in this area. Our qualitative interviews were informative, providing insight into how an individual living with PSP progresses through the healthcare system, the clinical perspective of the disease and the lived experiences of those with PSP (Fig. 2). A number of key breakpoints were also identified throughout the journey (Table 1).

The clinical pathway was validated by KOLs in the seven countries and considered an accurate representation of the patient journey through the healthcare system with some country-specific differences noted. In some countries, patients were more likely to present to their primary care physician (Germany, Spain, UK and the USA) or the neurologist (USA) than attend the emergency room, except in the case of falls, once symptoms become apparent. In the UK and Japan, there are very few PSP experts and patients are more likely to be treated by general neurologists who may have experience with PD. In Japan, healthcare is access-free and patient routes into care can vary, coming via the primary care physician (PCP), neurologist, orthopaedist or PO. In Germany, specialised neurologists tend to be responsible for patient management.

The Emotional Journey

Five stages of the patient journey were identified (Fig. 3): first symptoms and route to diagnosis; diagnosis; treatment initiation; living and struggling with PSP; end-of-life. Three of the stages (first symptoms and route to diagnosis, diagnosis and treatment initiation) were topics for discussion included within the structured research and standard within a patient journey; two of the stages (living and struggling with PSP and end-of-life) emerged from the analysis. Our research highlighted six major 'pain points' from the patient/caregiver perspective, defined as particularly difficult milestones in the patient journey that require dedicated support and that are common to all patients and across the geographies:

1. Fighting for an answer
2. No longer you
3. Rapid decline
4. Confronting your mortality
5. Loss of independence/total dependence
6. Irreversible decline

Fig. 1 Methodologies used for investigating the clinical and emotional journey in PSP. *KOL* key opinion leader, *PO* patient organisation, *PSP* progressive supranuclear palsy

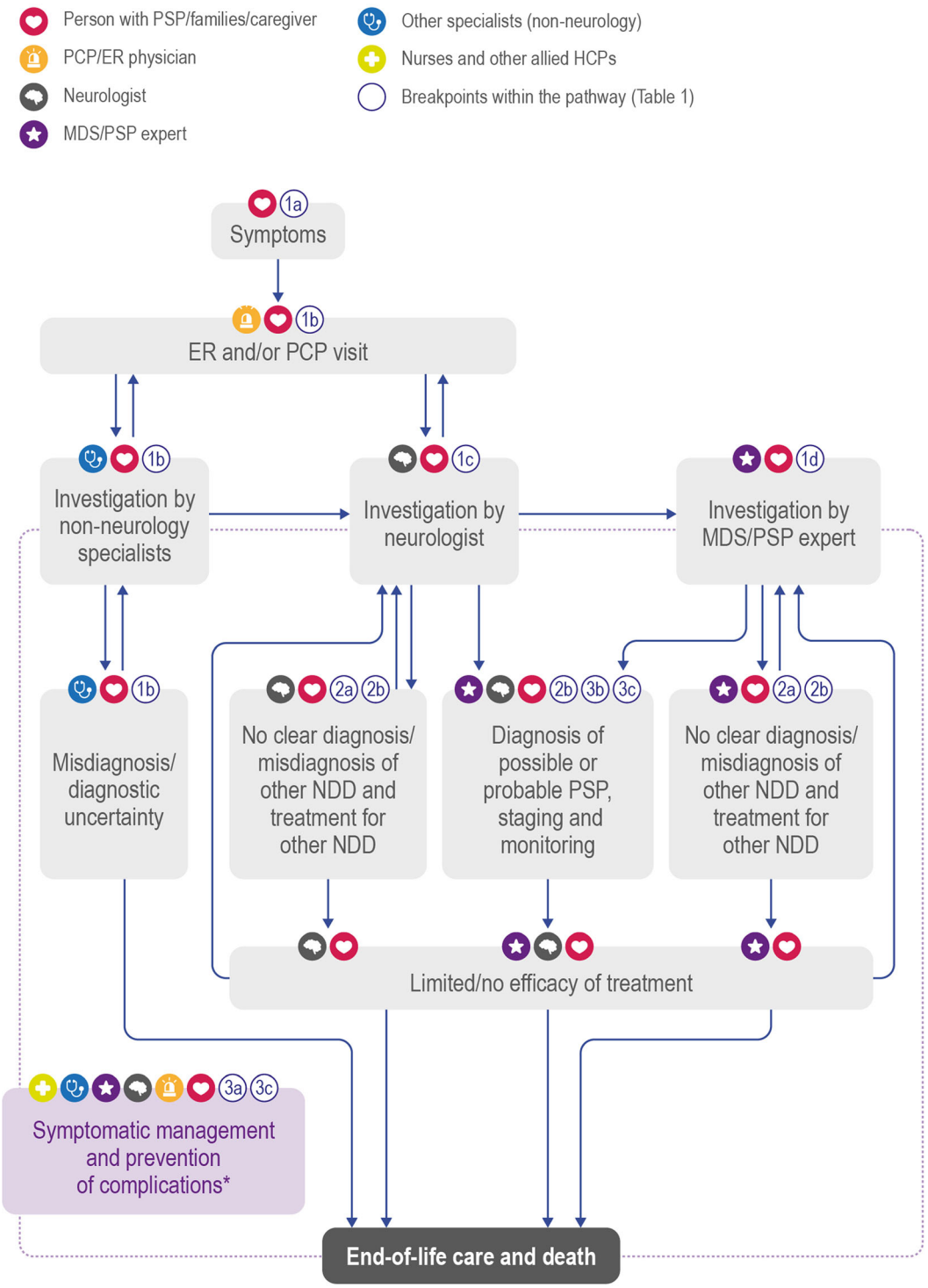

Fig. 2 The clinical journey with PSP [12, 32, 33]. *Extent of involvement of other HCPs in the management of people with PSP will be dependent on individual circumstances and geographies. *ER* emergency room, *HCP*, healthcare professional, *MDS* movement disorder specialist, *NDD* neurodegenerative disorder, *PCP* primary care physician, *PSP* progressive supranuclear palsy

The patients' and caregivers' experience of these 'pain points' at the five stages of the patient journey are shown in Table 2. A sample selection of quotes obtained during the research and grouped according to the five stages of the emotional journey is provided in Table 3. A key distinction between the clinical journey (Fig. 2) and the emotional journey (Fig. 3) is that the emotional journey is time-based from first symptoms to end-of-life, whereas the clinical journey highlights movement through the healthcare system and touchpoints with HCPs. They are not intended to be used synchronously but instead to complement each other and to provide a holistic view of the lived patient experience.

Time to Diagnosis

Based on feedback from HCPs, it was found that an average duration of 1.5–3 years was spent in the healthcare system before people with PSP receive the most accurate possible diagnosis,

Table 1 Breakpoints identified throughout the clinical journey

Patients with PSP reaching and accessing appropriate HCPs for correct referral	1a	Delay in patients seeking medical advice due to multiple potential reasons, including misattribution of symptoms, lack of awareness as the patient adapts to the symptoms, or apathy associated with the condition
	1b	Delay in referral to correct specialist due to presentation with unspecific symptoms and lack of awareness of the symptoms indicating a neurological condition amongst PCP, ER doctors, non-neurology physicians, nurses and other allied HCPs
	1c	As a result of the rarity of the disease as well as disease heterogeneity, neurologists do not suspect PSP until hallmark symptoms arise, contributing to further delays in diagnosis
	1d	Point at which patients are referred to MDS/PSP experts differs widely. Delay in referral of patients to PSP experts can lead to suboptimal management and patient/caregiver frustration
Diagnostic divergence	2a	As a result of a low index of suspicion and an overlap in symptoms, PSP is associated with a high degree of diagnostic uncertainty
	2b	Lack of diagnostic biomarkers, treatment options and/or disease understanding/experience, as well as disease heterogeneity, means diagnosing physicians may feel unable to provide patients with a possible or probable PSP diagnosis
Management of patients with PSP	3a	As a result of lack of treatment options, people with PSP (pre- and post-diagnosis) are treated for symptoms versus targeting disease pathology
	3b	Lack of consistency in the grading and monitoring of PSP
	3c	Lack of guidelines and quality standards for the management of PSP may lead to inconsistency of care, suboptimal management and increased burden on both the patients and caregivers

ER emergency room, *HCP* healthcare professional, *MDS* movement disorder specialist, *PCP* primary care physician, *PSP* progressive supranuclear palsy

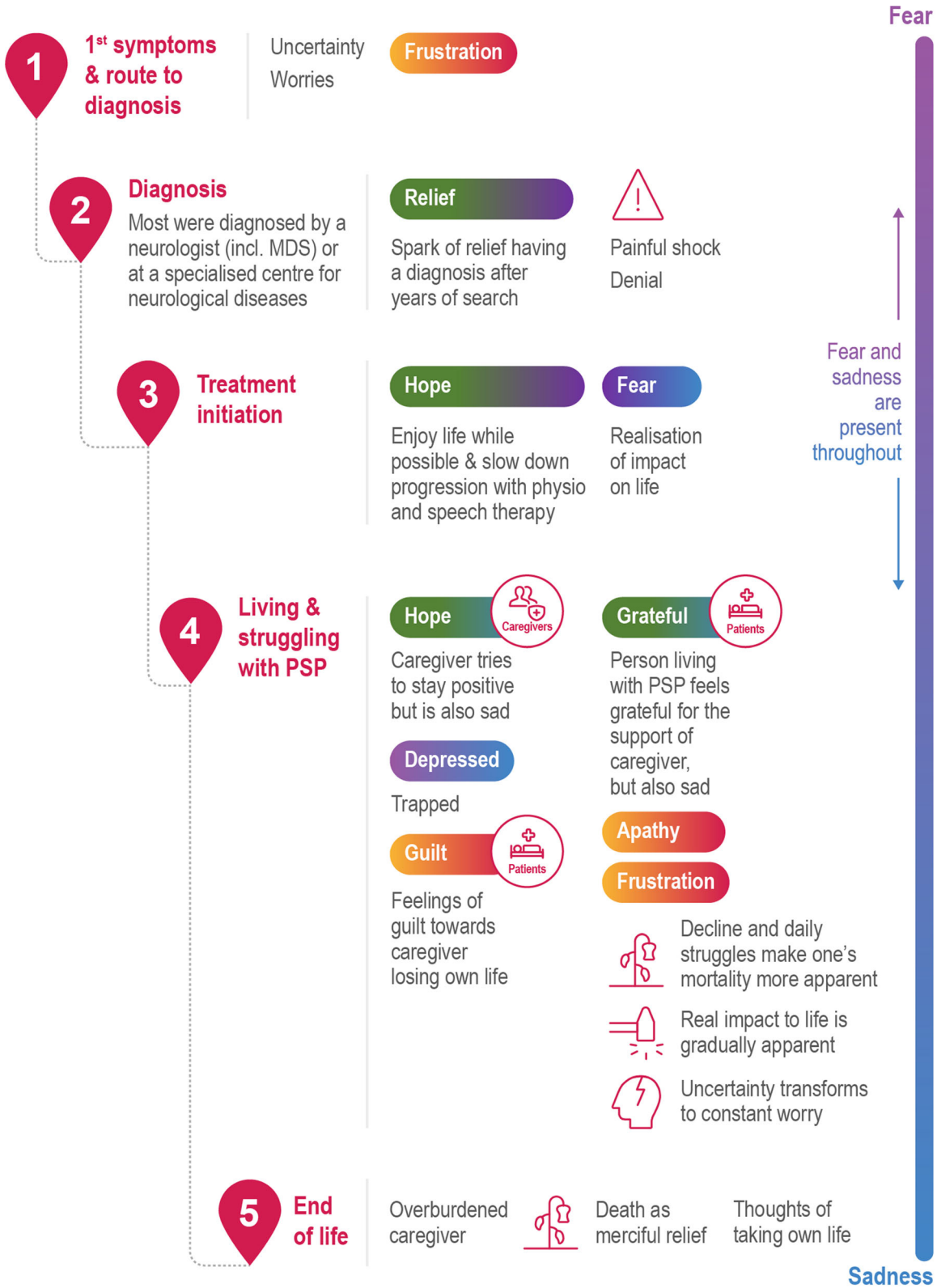

Fig. 3 The emotional journey with PSP. *MDS* movement disorder specialist; *PSP* progressive supranuclear palsy

Table 2 Six major 'pain points' over five stages of the clinical journey

'Pain point'	Experience of pain point for patients and caregivers	Stage of patient journey				
		1. 1st symptoms & route to diagnosis	2. Diagnosis	3. Treatment initiation	4. Living & struggling with PSP	5. End-of-life
1. Fighting for an answer	People exhibiting symptoms of PSP and their caregivers are fighting for an answer to understand what is wrong with them. This includes seeing multiple HCPs and sometimes waiting for years for the correct diagnosis	×	×		×	
2. No longer you	Some of the first symptoms reported include incontinence and changes in behaviour. Throughout the progression of PSP, new symptoms occur and/or existing ones become more prevalent	×			×	×
3. Rapid decline	At certain stages of the disease the decline in functional ability can be rapid	×	×		×	
4. Confronting your mortality	Once the diagnosis is made, people living with PSP and their caregivers begin to understand the impact of PSP and that there is no happy ending		×		×	×
5. Loss of independence/total dependence	Patients lose their independence in the course of the disease. This represents one of the biggest concerns for people living with PSP				×	×
6. Irreversible decline	The point at which symptoms progress in severity, little if any 'quality of life' remains and the end-of-life is near					×

HCP healthcare professional, *PSP* progressive supranuclear palsy

which ultimately can only be confirmed upon autopsy. For all countries, a duration of 3–-9 months to diagnosis was considered the best-case scenario. This was more likely if symptoms are typical, if patients are self-aware and seek medical help early, and if they see a movement disorder specialist. France, Spain and the USA did not report beyond 4 years as the worst-case scenario; however, all other countries reported a higher range. The rarity of PSP, disease heterogeneity and overlapping symptoms with other conditions, such as PD or other atypical parkinsonism, contribute to the diagnostic delay. It was noted that the uncertainty around diagnosis is often one of the worst times for families.

Perceptions of HCP Support

The majority of patients/caregivers felt that their PCPs had limited disease understanding and that their neurologist did not give them sufficient information at diagnosis (e.g. details on how individuals with PSP should be cared for and information on disease progression). However, people living with PSP and their caregivers also reported feeling overwhelmed by the diagnosis and unable to process the information until later. Only a few reported that their neurologist was empathetic or made them feel comfortable during the interaction, although the authors propose that patient state-of-mind following the negative diagnosis may also contribute to this feeling. In Japan, the diagnosis dialogue appears to be better handled than elsewhere, with HCPs providing patients/caregivers with more information and proactively supporting networking opportunities. Furthermore, the system in Japan seems to offer better quality support options, including access to local care managers and home visits from clinicians.

HCP Perspectives

Sources of education and information were the main unmet needs identified by HCPs. Suggestions to improve this deficit included conferences, webinars, brochures, websites, events to exchange ideas, digital tools to improve multidisciplinary team coordination and recordings for people living with PSP/caregivers who cannot come to the healthcare centre. Multidisciplinary teams are not always 'institutionalised' but, depending on location, may take the form of a collaboration between HCPs on an as-needed basis. Nurses were considered underutilised in creating awareness and education about PSP, although in many countries a lack of government and public funding limits dedicated nurse support. Physiotherapy is also becoming an important resource given the lack of medication available. Patients and caregivers described beneficial outcomes from patient targeted physio- and speech-therapy sessions. However, quicker, on-demand support to help reorganise patient homes, to anticipate a placement or to access palliative care is also required.

Caregiver Perspectives

Over the course of the patient journey, the need for support increases for both the patient and caregiver. Many caregivers will give up work to care for their loved one which, along with out-of-pocket costs not covered by insurance, can result in a financial burden. Caregivers often feel frustrated, sad, alone or unsupported and feelings of guilt arise from self-perceptions of giving inadequate care. Caregivers try to stay positive and remain in control, denoting satisfaction from helping their loved one to maintain some quality of life. However, the progressive cognitive limitations of the patient result in increasing patient dependency and apathy, thereby contributing to the struggles of the caregiver. While negative emotions dominate in patients, some draw joy from the support of their caregivers. Nonetheless, caregivers are also in need of support as their own lives may be diminished as the disease progresses. Despite caregivers having a high risk of developing depression themselves, there is little support available to them. Towards the end-of-life, the burden on the caregiver increases both physically and psychologically, where the patient may be bedridden with limited ability to communicate. Caregivers will provide constant

Table 3 Illustrative quotes to support the emotional journey

Stage of the emotional journey	Source	Quotes
1. First symptoms and route to diagnosis	UK caregiver	Patient "had unexplained severe falls and personality change. We were running a business together and things were going badly wrong, I couldn't understand why he was making such bad choices"
	USA clinician	"It's a very emotional type of disease. The family and caregivers have gone through, not just the disease itself per se but just the experience of getting there, to the diagnosis"
2. Diagnosis	Japan caregiver	"My feeling was falling from a cliff […] At that time, I thought my husband won't last for 12 months. Everything went blank. I did not know what to do"
	German clinician	"They are shocked. Sometimes they cry. Especially if the diagnosis is early, if patients with 63 to 65 have it. […] a wife with a husband who still works, 63 or 64 years old, who are looking forward to retirement and then they get news like this, then it is a shock"
	USA caregiver	"I felt alone, I felt that there's nothing I can do, or anybody can do to change this"
	USA caregiver	"I was very relieved when [the] doctor diagnosed it, that he'd figured out what it was. Up till then it was like, is it cancer? Is it a brain tumour, just what is going on? So just knowing is better than not knowing"
	France clinician	"They're relieved because we've put an end to a long phase where they didn't know what the patient had. Now they know what they have. We've given it a name. Psychologically that's very important for the family"
	USA nurse	"Sadness […] It's hard to have to finalize this diagnosis and tell them because it is their life, it is their independence being stripped away from them at that point. It's very unsettling as a provider. You almost feel helpless"
	Germany caregiver	"The neurologist in the clinic did the final PSP diagnosis. It was just the neurologist, there weren't other parties involved. No, they didn't tell me anything about the progression of the disease, I knew more than the doctors. I, myself, knew a lot already and I brought it up at the clinic. So, they could assume that I was quite informed already"
	Spain nurse	"Well, the thing is, not always, but sometimes they come from the neurologist office and they are so confused (sometimes there is not enough time to explain everything or the terms used are very technical). So frequently, they come to us asking for things"
	Spain clinician	"We must be honest with the patient, but we must not tell them all the truth, because it is not nice. We all know that the prognosis is bad and life expectancy is short, but we mustn't say that to patients, let alone in their first visit. […] One of the key things we must do is to keep patients in good spirits, because that will have an impact on their health condition"
	France clinician	"I never talk about life expectancy. I never tell the patient 'You have 5 or 6 years left', which is the average life expectancy for PSP patients. That's indeed a piece of information I would tend to omit"
	Japan caregiver	"Yes, it was very easy to understand. The doctor didn't use very difficult words and explained in an easy-to-understand way. Because there was a lecture meeting on PSP, we learned there are so many patients with PSP. I think that was very good as well. We felt very anxious as if we were alone. There was nobody with PSP around us. Nobody knew about the disease. And we were worried about what will happen to us"
	UK clinician	"I mean, the worst thing you can do is tell somebody you have an incurable disease and there's nothing I can do"
	Germany clinician	"Experience has shown that even in the early stages of the disease, the cognitive impairments will make it impossible for the patients to absorb everything that is said, that's why you need their caregivers present as well"

Table 3 continued

Stage of the emotional journey	Source	Quotes
3. Treatment initiation	Germany caregiver	"At the beginning, we had the feeling, it is bad, but we did not have an urgent need for treatment. After half a year […] it hits you what people tell you […] It was a trigger for us that we look for patient care, power of attorney. We even went further; we went to the lawyer and made a will testament"
	Japan clinician	"It is important for patients to have hope. For intelligent patients, latest news on treatment under development can give motivation"
	USA caregiver	"To live our life full of joy, enjoy things we enjoy together while my husband can't walk like we used to, we got a wheelchair, a transfer chair. I take him for walks around the island or wherever we go, it's something, we like being by the beach so we can still enjoy that together"
	Italy patient and caregiver	[Caregiver] "I would say, stay positive and enjoy what you can until you are in the conditions to do it and take life one day at a time. Do everything you can do on that day. This is what I think. So that there are no regrets later on'
		[Patient] "I would say the same thing. It is what I try to do: I try to live in the moment"
	Italy nurse	"To slow down the progression of the disease and improve the quality of life in the best possible way"
	Japan nurse	"The treatment goal is to slow the progression and maintain the good function as much as possible with rehabilitation. When patients have a fall, some break the hip and others hit the head. If that happens, they experience a sudden decline in function and could possibly become unable to move or bedridden. Therefore, fall prevention is one of the treatment goals'
4. Living and struggling with PSP	German caregiver	"I had the feeling my own life was taken"
	USA patient	"She's very caring. She understands me in the right ways, putting me to bed, getting me up in the morning and to the bathroom. She feeds me breakfast. How does that make me feel? It's good, it makes me feel good'
	UK patient/caregiver	"What future? There isn't any for me here. He says his mind is healthy, he sees things people are doing and thinks he can do it, but he can't. He'll often say 'Get me up I'll do that,' but yeah, he can't so, yeah"
	UK clinician	"If I can make them stay at home and as independent as possible for as long as possible. So, that, that is, you know, the best we can achieve and it's the most rewarding"
	Japan nurse	"When patients come to the hospital for rehabilitation, some of them can improve their function even a little and go home looking happy and saying thanks. When I see such patients, I feel it was good to be involved in everyday care for them"
	France clinician	"The big frustration is that there is no effective symptomatic or disease-modifying treatment"
	UK clinician	"This is not a single disease affecting a single person; it's affecting the whole family group"
	France nurse	"What they struggle with most is the guilt for not looking after the patient, well not being able to look after the patient. They are scared they won't be able to do it properly'
	France caregiver	"I asked myself should I take my granddaughter on holiday for a week or is that cruel, going to beach to build a sandcastle? It meant leaving my wife behind for a week. It was complicated"

Table 3 continued

Stage of the emotional journey	Source	Quotes
5. End-of-life	Spain caregiver	'I feel my mother is going to die any minute. I honestly tell you I wish for that to happen. I don't want to see her suffering… and she has suffered. We had to hospitalise her with septicaemia the first week in July'

PSP progressive supranuclear palsy

care for patients at this stage of the disease, and it is not uncommon for them to wish for the situation to come to an end.

The Role of POs

POs play a crucial role in providing patients and caregivers with support from diagnosis through to end-of-life. They act as a trusted source of information and provide guidance on how to navigate the healthcare and financial systems. Caregivers can access support groups and network with other carers through POs or use hotlines for queries. Many caregivers will do voluntary work for POs once their loved one dies, often becoming ambassadors for the disease and sharing their experiences to help others. Unfortunately, not every country has a PSP PO, and, in some cases, PSP may be integrated into the PD PO. Proximity may also limit caregiver access to POs with some caregivers unable or unwilling to travel to access the support.

Table 4 provides a summary of areas of concern raised by the survey respondents above.

DISCUSSION

This work represents a major collaborative effort to try to understand the lives of people living with PSP and their caregivers, providing valuable information and validation of the lived experiences of those involved throughout the PSP journey. It provides a holistic view of the disease through both a clinical and an emotional lens, incorporating insights from HCPs, patients and caregivers, as well as the published literature. The current clinical journey with PSP can take many different pathways and patients tend to cycle through the healthcare system before the correct referral is made and a possible/probable diagnosis is received. Breakpoints contribute to delays in accessing appropriate clinical care, a high degree of diagnostic divergence and suboptimal management of the disease. A lack of consistency in grading and monitoring of PSP, made complicated by the differential clinical presentation and progression of PSP, was identified as one breakpoint within the clinical journey. The PSP Rating

Scale (PSPRS) is a prospectively validated, disease-specific scale for PSP that yields reproducible annual progression rates [25]. However, it can be time-consuming to use and its utility in clinical practice is the subject of ongoing debate [25]. Simplified versions of the PSPRS, including the PSP Clinical Deficits Scale [25] and versions modified for use within virtual settings [26], may offer reliable alternatives.

This research into the emotional journey with PSP highlights some moments of positivity, despite being fraught with negative feelings. Consistent with published literature [11], it may take many years to receive a diagnosis and the support that is currently offered by HCPs to patients and caregivers is not reaching its potential. Based on the perspectives described the authors' suggestions to improve HCP utility would include longer consultation times to allow HCPs to adequately explain what the disease is, how it progresses and how it can be managed. This would also give HCPs enough time to use language that is appropriate for people with PSP and allow patients/caregivers the space to process information and have their questions answered. However, this may be limited by lack of government funding, long waiting lists, and specialists' time. Given the limited time that specialists have available, both in terms of frequency of visits and duration of consultation (which may be cut short as a result of fiscal pressure), the authors would also suggest extending the dialogue with caregivers following the initial diagnosis to continue to provide explanation, answer questions and share resources and information. If an appointment with the primary HCP is not available, then this role could be fulfilled by a nurse and social worker or allied health professional, in-person or remotely. A government-funded specialised nurse practitioner or nurse specialised in PD could also be a point of contact for patients with PSP. The authors also suggest that closer collaboration between POs and HCPs may be beneficial, with POs providing intensified outreach to time-constrained HCPs. This could be pivotal in sharing vital support and information with patients and caregivers, across the patient journey.

The study findings suggest that compared with other countries, patients/caregivers in Japan receive more information upon PSP diagnosis and better support options. In 2015, the Japanese government released its 'New Orange Plan' to prepare for a wave of dementia expected with an ageing population, followed by the Framework for Promoting Dementia Care in 2019, requiring government ministries to raise awareness of cognitive impairment across various societal sectors [27]. Moreover, the Japanese authorities encourage research into intractable diseases and the provision of financial support for affected patients [28]. In 2000, a nursing care insurance system was implemented, and in 2003 PSP was listed in the government's Specified Disease Treatment Research Program, enabling patients to access publicly funded medical support [29]. We speculate that these initiatives could be driving the supportive nature of Japan's healthcare system towards the patients with PSP and their caregivers observed in this study.

Throughout this research, the caregivers emerged as the invisible sufferers and unmentioned heroes of PSP.

As the disease progresses, the patient becomes almost entirely dependent on their caregiver. As such, information and support needs are varied and change over time. More information on available financial support, life expectancy, nutrition and tube feeding and how to prepare for end-of-life is needed. Improved access to emotional and psychological support (such as adult day programmes and support groups) for both the patient and the caregiver, home healthcare, social-care support and respite care will alleviate some of the struggles highlighted within the research. The authors cannot emphasise enough the enormous impact PSP has on the caregiver and would suggest more helplines are set up for on-demand support.

The strengths of the study include its number of respondents (N = 112) and collection of responses acquired from across seven countries, together representing a solid base for sampling within a rare disease area. This study adds a rich patient-centric context to the recently published consensus outlining best practices in the

Table 4 Identified areas of need

Diagnosis	Diagnostic delays are common
	Limited disease understanding among HCPs, including PCPs and general neurologists
	Patients/caregivers not provided with sufficient information at diagnosis
	Poor handling of the diagnostic dialogue by the specialist
Patient management and care	Lack of HCP education and information on PSP
	Nurses are underutilised in creating awareness and education about PSP
	MDTs are not always institutionalised
Support for patients/caregivers	Quick, on-demand support for patients/caregivers is lacking
	Caregivers remain largely unsupported throughout the PSP journey
	Limited access to PSP POs

HCP healthcare professional, *MDT* multidisciplinary team, *PCP* primary care physician, *PO* patient organisation, *PSP* progressive supranuclear palsy

clinical management of PSP [30]. It supports the calls from Rowe et al. for early active management of people living with PSP, starting with an early diagnosis, proactive management of symptoms, and preservation of quality of life, by drawing on the basic principles of good, multidisciplinary medical care and acting as a champion for those affected [31].

The study was limited in that while we have tried to account, as far as possible, for geographical differences regarding routes into care, patient evaluation, HCP involvement and treatment approaches, we cannot account for differences in countries that were not assessed. Instead, the research offers what the authors consider a holistic view of how a person living with PSP moves through the healthcare system, touchpoints with HCPs and key breakpoints within this pathway. Furthermore, the emotional journey does not distinguish between emotional changes as a symptom of PSP and the emotional response to diagnosis and rapid disease progression. However, the analysis accounts for the views of not only people with PSP but also caregivers, HCPs and POs providing multiple perspectives on the patient's lived experience and the emotional journey.

CONCLUSIONS

The person living with PSP, their caregiver and specialist neurologist and care team must go on the diagnostic journey together.

PSP is associated with a high degree of diagnostic uncertainty. As such, specialists should be honest and open with patients and caregivers from the beginning and throughout the diagnostic journey, including communicating clearly what is known and unknown at specific points in time and future prognostic expectations, even if not optimistic. This can also help build the relationship between practitioners and patients/caregivers.

Education and information are needed for all stakeholders, providing the right level of support at the right time, and tailored to the needs of the person living with PSP and their caregiver.

There is a need for all stakeholders, including payers and policy makers, to be educated around the atypical parkinsonisms, including PSP. Specialists have an enhanced understanding of the PSP disease area and there is an opportunity to share knowledge with non-specialists to improve differentiation of PSP from PD, or other less common neurodegenerative movement disorders. Nurses, who may have less

constrained schedules, tend to offer more time and empathy to patients and caregivers, and report that they are additionally eager to be better trained in the needs of patients living with PSP. Diagnosis can be overwhelming for the patient and their caregiver. It takes time to build an understanding of what PSP is and the impact the disease will have on daily life, the full extent of which is often only realised later in the disease course. Support needs to be given at the right time and information tailored into 'bite-sized chunks' (however, this does not currently happen outside of POs) and made available for HCPs to share with the patient/caregiver. Patients and their caregivers need links back to neurology teams that can provide access to more information in the weeks and months following diagnosis. This could be conducted by a clinic coordinator, nurse, social worker or allied health professional. HCPs would welcome specific funding for such resources from governmental and other sources.

Caregiver health, as well as the patient's health, is a major public health concern that physicians and other HCPs need to address.

People living with PSP and their caregivers could be better supported through a variety of activities including psychological support, provision of information (brochures, videos, etc.), PSP training for caregivers, including administering medication, at home and respite care, practical services for the management of basic needs (e.g. nutrition, mobilisation and in-home safety), better access to rehabilitation, occupational therapy ('tele rehab'), financial support and family and community support networks.

The authors are aware that many of these suggestions may require more funding and time, and better reimbursement models. Thus, a coordinated effort from all stakeholders is needed in the lobby for better, more supportive care of people living with PSP and their caregivers.

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Compliance with Ethics Guidelines. According to criteria within the EPHMRA code of practice (Sects. 1.1 and 1.2), this study is defined as market research. As such, consistent with the guidance issued by the British Healthcare Business Intelligence Association (BHBIA) and the EPHMRA code of practice (Sect. 1.3), neither ethics committee approval nor clinical trial registration was required. Informed consent, including consent to publish the study findings, was obtained from all study participants prior to the study. Additionally, consent was sought from all participants to publish verbatim responses.

Data Availability. The datasets generated during and/or analysed during the current study are available from the corresponding author on reasonable request.

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