Movement Disorders
CLINICAL PRACTICE

RESEARCH ARTICLE

Challenges Faced by Patients With Progressive Supranuclear Palsy and their Families

Theresa Moore, RN, MScN,[1,] Mark Guttman, MD, FRCPC[1]*

Abstract: The literature is inadequate for understanding the challenges experienced by people with PSP and their families. Therefore, the aim of this study was to understand the challenges of people with PSP and their caregivers and identify their priority need. In this qualitative study, five focus groups were conducted with people with PSP and/or their family caregivers, one group with long-term care staff, and one with community caregivers. Data were analyzed using fundamental qualitative description. Four themes were identified: knowledge, services, research, and symptoms. Knowledge challenges were identified as the priority need, with the most common challenges in this category being lack of knowledge of PSP among community workers, physicians, patients, and family members. Service challenges involved service access and interactions with physicians, community workers, private caregivers, and long-term care staff. Research challenges related to the lack of research and the failure of health care providers or PSP organizations to communicate research findings. Symptoms most often identified as challenging were falls, mobility, vision, mood or thinking, speech, and swallowing. Participants identified their priority need as dissemination of information about PSP. This has not been captured in previous research. This information needs to reach doctors, long-term care staff, community workers, patients, families, and the general public. Subsequent activities to meet this need are summarized. These activities resulted in three new resources: a brochure for patients and families; an information packet for physicians; and a webinar for staff in long-term care and community.

PSP is the most common type of atypical parkinsonism. The estimated prevalence is 6.5 individuals per 100,000[1,2] and the average survival time is just 5 to 7 years.[3,4] The diagnosis of PSP is challenging, especially in the early stages. The PSP Association (United Kingdom) have identified that three quarters of people with PSP are initially misdiagnosed.[5] It is possible that misdiagnosis was most often by family physicians, who would have little awareness of PSP. This impression is supported by the North American organization, CurePSP, where many patients report that their family doctors knew nothing about PSP.[6]

Three studies have explored the needs of people with PSP or their families. The first was a survey of 180 family caregivers who were members of the North American Society for PSP (now known as CurePSP).[7] The survey including ratings of the frequency of burden related to 22 caregiving domains. The researchers found significant burden, which was related to disease severity, disease duration (peaking at 18 months postdiagnosis and then plateauing), and female caregiver gender. A Swedish study[8] examined symptoms, services, quality of life, and information needs of patients with parkinsonism; however, just 4 of the 23 subjects had PSP. Finally, a British study[9] of 27 PSP patients included interviews, physical exam, and quality-of-life measures and found problems in all five quality-of-life domains, with more than half of subjects reporting problems with walking/balance/falls, depression or apathy, meeting people, pursuing hobbies, going out, and household chores.

The literature is inadequate for understanding the challenges and needs of people with PSP and their families. This understanding is needed to determine how best to improve support to those affected by this rapidly progressive disease. Therefore, the aim of this study was to understand the challenges of people with PSP and their caregivers and identify their priority need.

[1]Center for Movement Disorders, Markham, Ontario, Canada

*Correspondence to: Theresa Moore, Center for Movement Disorders, 2780 Bur Oak Avenue, Markham, Ontario, Canada L6B 1C9;
E-mail: tmoore@movementdisorders.ca
Keywords: supranuclear palsy, progressive, needs assessment.
Relevant disclosures and conflicts of interest are listed at the end of this article.
Received 5 February 2014; revised 14 April 2014; accepted 15 April 2014.

doi:10.1002/mdc3.12037

Methods

Focus groups were chosen because they are cost- and time efficient, allow participant interaction, and promote participant comfort and openness.[10]

Participant Recruitment

The local research ethics board provided ethics approval before participant recruitment. The opportunity to participate was offered to all the families of the 36 PSP patients of the Center for Movement Disorders in Markham, Ontario, Canada, as well as any of these patients who could communicate clearly. Requests for participants were also made to nine other local movement disorder specialists and, to reflect diversity of care received, 15 neurologists in a southern Ontario city without movement disorder specialists. In addition, the opportunity was publicized by CurePSP and Parkinson Society Canada. To obtain diverse perspectives, professional caregivers of PSP patients in long-term care and community settings were also recruited. Potential participants were approached in clinic or by phone. All interested patients, family members, and caregivers completed a written consent and were accepted into the study, resulting in a convenience sample of 45 participants. Primary reasons for declining to participate were poor health, difficulty speaking or traveling, language barrier, and inability to find a respite caregiver.

Data Collection

Between December 2009 and June 2010, five focus groups were conducted with people with PSP and/or their family caregivers, one group with long-term care staff, and one with community caregivers. The participants included 9 people with PSP, 16 spouses, 10 adult children, 6 long-term care staff, and 4 community workers. Of the 22 patients represented, 13 were from the Center for Movement Disorders in Markham (and therefore known to the primary and/or second investigator), 5 were clients of other movement disorder centers, 3 received their PSP care from general neurologists, and 1 from their family physician. The 6 men and 3 women with PSP ranged in age from 63 to 79 years and had been diagnosed between 6 months and 5 years earlier. All but 1 still resided at home. The long-term care group included two personal support workers (PSWs), two recreation therapists, one registered practical nurse, and an assistant director of nursing. The community caregiver group included two PSWs, an occupational therapist, and a respite program manager.

The focus groups were moderated and tape recorded, without field notes, by the primary investigator, a master's degree trained nurse, with 3 years of experience in movement disorders, a family member with PSP, and two previously published qualitative studies. Patient/family groups lasted 2 hours, whereas professional caregiver groups were 1 hour. The long-term care group was held at the long-term care facility. Four of the patient/family groups were held in person at the movement disorder center, whereas one patient/family group was conducted by video conference for individuals from five northern Ontario communities, and the community caregiver group was held by conference call. At the outset, participants were informed that the researchers' overall goal was to improve support for those affected by PSP. Therefore, participants' responses would be used to identify the priority need so that programs or resources could be developed to meet this need. Open-ended questions were used to explore challenges faced before, during, and after diagnosis as well as responses to these challenges. Probes to seek additional depth and clarity were used sparingly in light of time constraints. On closing, each participant was asked to identify the most important need. During professional caregiver groups, participants were asked what patient/families told them about their challenges and what they noticed about their challenges and responses. Although theme saturation was achieved before the final focus group, data collection continued to include all of the participants who had been recruited.

Data Analysis

Data were analyzed using fundamental qualitative description, employing low inference interpretation to present a comprehensive summary using participants' words. This method seeks descriptive validity—an account that most people would agree is accurate—and interpretive validity, an accurate accounting of the meanings participants attributed to those events. Codes are generated from the data themselves and responses are counted so that themes are ordered according to prevalence.[11,12] In this study, participants' reports of relative importance of needs were also taken into account when identifying the priority need. Analysis involved the primary investigator: reading each transcript; rereading the transcript and highlighting statements about challenges; identifying emerging themes; developing a handwritten coding template to organize data; and counting and ordering theme statements and priority needs. Analysis also included calculation of interrater reliability and consultation with the second investigator.

Trustworthiness and credibility were ensured by use of verbatim transcripts, rigorous checking of the transcripts while listening to the tapes, and attending to the complexity of human experience evidenced by contradictory observations. In addition, interrater reliability was calculated for two of the transcripts with an independent rater who was a masters' prepared physiotherapist with movement disorder expertise. The number and content of the phrases extracted by the raters were compared and discrepancies discussed. Prediscussion agreement was 83% for the first transcript and 89% for the second. Postdiscussion agreement was 100%. Neither transcripts nor findings were reviewed by participants.

Findings

Four themes emerged related to patient and family challenges: lack of knowledge; service access and interactions; inadequate research and research dissemination; and symptoms.

doi:10.1002/mdc3.12037

Lack of Knowledge

The most prevalent theme identified as the priority need was lack of knowledge, with 36 of the 45 participants identifying this as a challenge and 32 as the priority need. Common concerns were lack of knowledge of PSP among community workers (20), general practitioners (17), neurologists (6), emergency room physicians and other specialists (4), movement disorder team members (2), and unanswered questions among patients and family members (10). For example, one participant said: "What is critical is information…the necessity of information going out to the community support services, information going to the doctors so that they recognize it." Whereas another shared: "He didn't have much energy. The family doctor said 'he's getting older.' He said 'no, this feels different.' And then he started to fall…Two years later, she finally sent him to a neurologist who thought he had a stroke. When we came to see the doctor at the movement disorder center, he thought it was PSP."

After diagnosis, families found themselves responsible for educating community workers, family physicians, emergency room physicians, specialists, long-term care staff, and others. For example, one wife noted, "I have to explain what it is every time we go to the emergency room." Whereas another shared: "You have to train the support worker how to handle them and what are the symptoms…It doesn't give me relief that I can leave my husband because she doesn't have that knowledge." And another said: "Even after diagnosis, my family doctor admitted he knew nothing. I said 'I'm going to make an appointment I'll give you time to research.' When I went back in 4 months, I don't think he could have even named the illness." And another shared: "The neurologist was young and new…He felt it was PSP …but he doesn't know a lot about it either. He said 'go on the internet.'"

Questions and comments from the participants revealed their own continuing needs for knowledge. Some were unaware of available resources, such as CurePSP, respite care, PSP support groups, speech resources, and exercise programs. Many asked the moderator questions about PSP etiology, prevalence, and progression. A few participants expressed contradictory views and found information "overwhelming" and "debilitating." They preferred to "react to the situation as it is happening today." One spouse shared: "Physicians and care people need to recognize not everyone wants to know the full prognosis. She wants a bit more hope."

Service Access and Interactions

Twenty-nine participants identified challenges related to service access and/or interactions with physicians, private caregivers, long-term care staff, and Community Care Access Center (CCAC) workers (CCAC is funded by the Ontario Ministry of Health and Long-Term Care and provides in-home support services). Eleven identified services as a priority need. Common concerns included failure or delays in referring for assessment, the number of physicians seen over several years to obtain the PSP diagnosis, insufficient service hours, inadequate staffing, and the long wait times for referrals, community services, and long-term care. Also of concern was physician advice that "nothing can be done" for PSP. One daughter shared that in "trying to get a diagnosis we went to the family physician, a geriatric doctor, a neurologist, back to the family doctor and then finally…the movement disorder doctor." For many, the challenges with providers persisted after diagnosis, such as this widower who shared: "The second time we visited the neurologist she said 'there's no need for me to see you again.' We felt as though we were cut adrift…like outcasts. CCAC came and said money is very tight, they can't help us. We desperately needed help." And this daughter who said: "The message at the movement disorder center was 'there's nothing we can do.' There didn't seem any point to going back. All they could offer was Botox."

Inadequate Research and Research Dissemination

Although none of the health care provider participants spoke about research, 9 of the 35 patient/family participants felt challenged by the lack of PSP research, lack of funding for such research, and/or failure of health care providers or PSP organizations to communicate findings. For example, one said, "It's very frustrating that there isn't a lot of research being done…there should be more lobbying to the government to have them fund research." Whereas another countered, "They are doing the research, they just don't communicate it…if we send more money, we would like to hear what success there is." And another added: "The information could have been printed on the back of an envelope. Hope would have been very helpful."

Symptoms

All but 2 of the participants identified symptoms as a challenge, though none identified symptom management as the priority need. The symptoms most often identified as challenging were falls ($n = 35$), mobility (33), vision (27), mood or thinking (26), speech (24), and swallowing (18). Participants often spoke about how these symptoms evolved. For example, one reported, "First, I fell gardening. Then I fell backward all the time. I used to fall down the stairs, too. I couldn't see the last step. Now I can't turn my eyes down and don't see any of them." Caregivers were also affected, such as this wife who said:

"The most difficult part, it's not the physical disability, it's the mental. Now in the mind he cannot pass the messages to say, wait for a second, I am coming. He is like 'I can do it,' and he will do it and then we have the fall…How do I cope with the frustration and stress?" And another who shared: "What's troubling me now is his choking if he eats. He chokes a lot and he coughs a lot and he really can't bring it up."

Discussion

This study of PSP patients, family members, caregivers, and health care professionals found that the most prevalent priority

doi:10.1002/mdc3.12037

need identified by participants was lack of knowledge about PSP among health care providers, patients, and families. This has not been captured in previous research, likely because the current study used a qualitative method and was designed to explore challenges of any nature, not just those related to symptom management, caregiver burden, or typical quality-of-life domains.

Lack of knowledge is an important finding because it offers clinicians a way to improve support to patients and families through education even when symptom management approaches are limited or suboptimal. In particular, we can target education to the three groups most frequently identified by participants as in need of knowledge—community workers, family physicians, and patient/families. A quality-of-life study published after the completion of our focus groups suggests that educating patients about PSP could improve health-related quality of life because a low level of education was an independent determinant of decreased quality of life.[13] Our findings support the call for patient education, but also point to the need to expand efforts to include education of health care providers.

An in-depth review of the CurePSP, Parkinson Society Canada, and PSP Association websites followed by telephone interviews with their education representatives revealed that there is, in fact, a considerable amount of information available for patients, families, and health care providers. Few, if any, of these resources have been formally evaluated by their target audiences. The primary investigator then reviewed all of the PSP resources from these organizations. Several gaps were identified that could be contributing to the lack of knowledge among health care providers, patients, and families. These gaps included (1) a succinct overview of symptoms and care options, (2) a concise information packet for Canadian family physicians (CurePSP has a lengthy packet that includes U.S. resources), and (3) a webinar that would make information more accessible to health care teams in long-term care and community settings. To fulfill our ultimate aim of improving support to those affected by PSP, the researchers then addressed these gaps through developing, testing, and distributing a new brochure, physician packet, and webinar. The specific activities associated with these new resources are summarized in Tables 1, 2, and 3.

Limitations

Despite the researchers' recruitment efforts, several groups were underrepresented in the focus groups, specifically patients and families from the other large local movement disorder center and individuals who have not been referred to a movement disorder specialist. It is possible that patients and families who had not been seen at an interdisciplinary movement disorder center may have different experiences and even greater needs. Future research could make further efforts to recruit these underrepresented groups. In addition, research should explore the barriers that exist to the awareness and use of PSP resources among family physicians and other health care providers.

TABLE 1 Development, testing and availability of PSP brochure

Title	Getting Help for PSP: A Guide for Patients and Families
Target audience	Patients and families
Content	Concise overview of symptoms and care options for each
Pretest	Yes
Items included in pretest and testing	Helpful at and after diagnosis Helpful in talks with team Provides hope Length and scope Clarity Suggestions
Testing method	Ontario: mailed survey CurePSP: Survey Monkey
Response rate	Ontario: 44/74 (59%) CurePSP: 105/1189 (9%)*
Respondents	16 patients 133 family members
Major testing results	Helpful at and after diagnosis Would help in talks with team Provides hope Length and scope "just right" Content clear except for three words Need to add resource section
Major revisions made	Added resource section Improved clarity of introduction
Availability	Parkinson Society Canada by mail or download (English and French) at www.parkinsoncno.ca *or* www.parkinson.ca To our patients and their families at clinic visits Mailed to family physicians of our patients, upon diagnosis, as part of a physician packet (see Table 2)

*Emailed to CurePSP constituents. Date of CurePSP involvement not tracked. No reminders sent.

Conclusion

The many challenges faced by people with PSP and their families are diverse and evolving. These challenges include lack of knowledge, service access and interactions, inadequate research and research dissemination, and symptoms. Often, sources of help were unknown, inaccessible, underutilized, or inadequate. Although there are multiple opportunities for improving support, participants identified their priority need as dissemination of information about the disease and about diagnosis and care of the person with PSP. This information needs to reach doctors, long-term care staff, community workers, patients, and families. Phase II of this study endeavoured to address this need, resulting in the availability of three new resources: a brochure for patients and families; a packet for family physicians; and a webinar for staff in long-term care and community. It is expected that improved knowledge will translate into more skillful, timely, and supportive care for those affected by PSP.

Author Roles

(1) Research Project: A. Conception, B. Organization, C. Execution; (2) Statistical Analysis: A. Design, B. Execution, C. Review and Critique; (3) Manuscript Preparation: A. Writing of the First Draft, B. Review and Critique.

TABLE 2 Development, testing, and availability of physician packet

Target Audience	General Practitioners (GPs) Nurse Practitioners (NPs)
Content	Diagnostic DVD (from CurePSP) Parkinson's vs. PSP Diagnostic Criteria PSP Overview (from CurePSP) Brochure (see Table 1) PSP: Some Answers (from CurePSP) Service Organizations
Pretest	Yes
Items included in pretest and testing	Helpful for recognizing PSP Helpful for care of newly diagnosed patient Helpful for ongoing care Missing content Suggestions
Testing method	Mailed survey
Response rate	GPs 15/42 (36%) Geriatric NPs 12/48 (25%)
Respondents	15 GPs of PSP patients 12 Geriatric NPs
Major testing results	All packet components very or somewhat helpful for recognizing PSP, caring for newly diagnosed patient, and/or ongoing care Some revisions suggested for CurePSP materials (forwarded to CurePSP) Questioned whether some medications are contraindicated and if diagnostic criteria are most current version available
Major revisions made	Added contraindicated medication list Substituted diagnostic criteria, which have greater sensitivity and reliability
Availability	Packet mailed to general practitioners of our PSP patients upon diagnosis Individual components by download from Parkinson Society Central and Northern Ontario at www.parkinsoncno.ca CurePSP authored components also available from www.curepsp.org

TABLE 3 Development, testing, and availability of PSP webinar

Title	A Guide to PSP for Staff in Long-Term Care and Community
Target audience	Long-term care staff and community workers
Content	Parkinson's vs. PSP Symptoms Care options Patient-centerd care Family care Resources
Pretest	Yes
Items included in pretest and testing	How helpful Most helpful aspect Changes needed
Testing method	Online or emailed survey
Response rate	31/49 (63%)
Respondents	31 staff from community and long-term care
Major testing results	Helpful for learning about PSP care: Very (71%) Somewhat (29%) Most helpful was symptom information and care strategies Least helpful was poor sound quality
Major revisions made	Added resources to provide to patient and family at diagnosis Sound corrected in archived version
Availability	CurePSP at www.curepsp.org (archived webinars June 19, 2012) Parkinson Society Canada Central and Northern Ontario may also present this PowerPoint presentation live upon request.

T.M.: 1A, 1B, 1C, 2A, 2B, 3A
M.G.: 1A, 1B, 2C, 3B

Disclosures

Funding Sources and Conflicts of Interest: Financial support was gratefully received from Parkinson Society Canada, Central and Northern Region. The authors declare that there are no conflicts of interest relevant to this work.

Financial Disclosures for previous 12 months: Theresa Moore is employed by the Center for Movement Disorders, Markham, Ontario, Canada, and received a grant from the Parkinson Society Canada. She has no financial disclosures in the following categories: stock ownership in medically related fields; intellectual property rights; consultancies; expert testimony; advisory boards; partnerships; contracts; honoraria; royalties; or other. Mark Guttman serves on the advisory boards of Teva Canada, UCB Canada, Roche, and Bristol Meyers Squibb. He has contracts with Merck (clinical trials in Parkinson's Disease), Chelsea (clinical trials in Parkinson's Disease), and Teva (clinical trials in Huntington's Disease). He received a grant from the Parkinson Society Canada and funds from Novartis Canada. He has no financial disclosures in the following categories: stock ownership in medically related fields; intellectual property rights; consultancies; expert testimony; employment; partnerships; honoraria; or royalties.

References

1. Nath U, Ben-Shlomo Y, Thomson RG, Morris HR, Woods NW, Lees AJ, Burn DJ, The prevalence of progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome) in the UK. *Brain* 2001;124:1438–1449.
2. Schrag A, Ben-Shlomo Y, Quinn BNP. Prevalence of progressive supranuclear palsy and multiple system atrophy: a cross-sectional study. *Lancet* 1999;354:1771–1775.
3. Diroma C, Dell'Aquila C, Fraddosio A, et al. Natural history and clinical features of progressive supranuclear palsy: a clinical study. *Neurol Sci* 2003; 24: 176–177.
4. Litvan I, Mangone CA, McKee A, et al. Natural history of progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome) and clinical predictors of survival: a clinicopathological study. *J Neurol Neurosurg Psychiatry* 1996;60:615–620.
5. A guide for people living with PSP, CBD, and other atypical parkinsonian disorders. Available at: www.psp.org/file_download/bdbad4-d5-6215-4624-9aec-6f909f459fb5 accessed (17 April 2011).
6. What is PSP? Available at: http://www.pspassociation.org.uk/what-is-psp/ accessed (22 May 2011).
7. Uttl B, Pilar MA, Litvan MD, Grafman J. Caregiving in progressive supranuclear palsy. *Neurology* 1998;51:1303–1309.
8. Sjöström AC, Homberg B, Strang P. Parkinson-Plus patients—an unknown group with severe symptoms. *J Neurosci Nurs* 2002;34:314–319.
9. Schrag A, Selai C, Davis J, Lees AJ, Jahanshahi M, Quinn N. Health-related quality of life in patients with progressive supranuclear palsy. *Mov Disord* 2003;18:1464–1469.

doi:10.1002/mdc3.12037

10. Vogt DS, King DW, King LA. Focus groups in psychological assessment: enhancing content validity by consulting members of the target population. *Psychol Assess* 2004;16:231–243.

11. Maxwell JA. Understanding and validity in qualitative research. *Harvard Educ Rev* 1992;62:279–299.

12. Sandelowski M. Whatever happened to qualitative description? *Res Nurs Health* 2000;23:334–340.

13. Winter Y, Spottke AE, Stamelou M, et al. Health-related quality of life in multiple system atrophy and progressive supranuclear palsy. *Neurodegen Dis* 2011;8:438–446.

doi:10.1002/mdc3.12037