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Usage of Support Services in Primary Lateral Sclerosis

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Abstract

Patients with Amyotrophic lateral sclerosis (ALS) rely on a variety of support services during the course of their illness. Patients with Primary lateral sclerosis (PLS) have a slower progression of disease and different clinical spectrum. Their needs for allied health services and social support have not been well characterized. To investigate these needs, 25 patients with PLS and caregivers were surveyed about the use of assistive devices and support services. Needs for assistance changed as the disease progressed. Their greatest need was for gait assistive devices and home help for activities requiring mobility. As in other chronic diseases, there was a striking use of the internet to gather information and for patient support groups.

Keywords

Amyotrophic lateral sclerosis; internet support groups; Primary lateral sclerosis; wheelchair; survival

Introduction

Patients with Primary Lateral Sclerosis (PLS) have spasticity and weakness but slower decline than patients with Amyotrophic Lateral Sclerosis (ALS). In ALS, allied health and social support services that are typically required include physical therapy, nutritional and respiratory support, speech pathologists, patient education, psychologists, physicians, in-home assistance, nurses, and hospice (1). Most of these services are coordinated through multidisciplinary clinics where ALS patients receive care. There are no studies of the services that patients with PLS require throughout the course of their illness, and it is unknown whether they need or use similar services. Needs for wheelchair and gait assistive devices may occur later in their illness or may differ as the illness progresses. To investigate this issue, we conducted a brief survey of patients with PLS followed in our clinic.

Material and Methods

The questionnaire was developed from a pilot study with several open-ended questions that were sent to 45 patients. The pilot data and questions from the ALS literature on social services and wheelchair usage to derive a 17-item closed ended questionnaire (2–4) (see supplementary material). The questionnaire was approved by the Institutional Review Board. Forty PLS patients and their caregivers were mailed the questionnaire as part of an annual mailing. All patients conformed to the criteria proposed by Pringle (1992) for PLS (5). Twenty-five PLS patients returned the questionnaire. Twenty-two were contacted by telephone for additional follow-up. Medical charts were reviewed to obtain further information about age at the onset

of symptoms, age at the time of diagnosis, and history of onset of the use of canes, walkers, and wheelchairs.

Results

Subjects

Of the 25 patients, 13 were men and 12 were women. The mean age was 59 years old, with a mean age of symptom onset at 45 years, without differences between men and women.

Physical/occupational therapy needs

None of the patients initially reported receiving care through a multidisciplinary clinic. However, on the telephone follow-up, 10 said that they had attended ALS or Muscular Dystrophy Association (MDA) clinics at least once and 5 received continuing care through such clinics. Three people did not find such clinics to be helpful. 24% of PLS patients reported receiving regular physical therapy, 12% of patients received speech therapy, and 8% of patients received occupational therapy. More than half of the patients received some form of alternative medicine.

The most prominent need overall was for assistance with activities of daily living that involved mobility (76%). Forty percent of patients reported needing household help with chores such as cleaning, 36% needed help with cooking, 32% of patients needed help with dressing/personal hygiene, and only 16% reported being independent. Most of the PLS patients (88%) had serviceable speech and nearly all (96%) had good respiratory function, and required no ventilatory assistance.

All of the patients indicated that they used some form of a gait assistive device. The type of assistive device was not predictable solely by the duration of disease symptoms (Figure 1A), with similar mean disease duration in those who used a cane alone (14 years), a walker (12 years) and wheelchair or motorized scooter (16 years). From the telephone follow-up, we found that 14 out of 22 patients used gait assistive devices prescribed by physicians or physical therapists, eight patients personally chose their current gait device with or without the agreement of their doctor. The review of charts of individuals who were currently using a wheelchair, however, generally showed a clear progression from cane to walker to wheelchair over the course of several years (Figure 1B).

Psychological support

The long duration of symptoms in PLS raises the question about where patients and caregivers turn for support for the emotional and psychological issues. None of the patients or caregivers sought support from social workers, psychologists, or psychiatrists. Just over half (60%) of patients reported that they used no external resources for support. On the telephone follow-up, most patients indicated that this was by choice and not because of lack of access. Twelve percent of the PLS patients belonged to a patient support group, and 16% utilized internet discussion and support groups. Twelve percent reported receiving emotional support from other miscellaneous sources, such as family and friends. Similar trends were seen among the caregivers: 44% had no resources for support, 12% belonged to a patient support group, 16% used internet support groups, and 16% turned to other sources. Patients with PLS typically sought information about their disease from multiple sources: their physicians (52%), newsletters (36%) and the internet (68%). The internet was used as a source of information as well as for participating in online discussion groups.

Burden of Illness

PLS has an impact on the lives of patients and caregivers. Forty-eight percent of patients retired because of their illness, and 32% of patients or caregivers reported changing their jobs. Sixty-eight percent made changes to their households, such as modifying showers or tubs for safety, adding ramps and lifts. Most of the patients in this survey had medical coverage for their healthcare, through combinations of insurance (44%), Medicare (44%), social security (24%) or from charitable organizations (28%). Most patients had made advance directives, with a durable power of attorney (60%) and with living wills (60%). None of our patients reported preparations for hospice care.

Discussion

Several findings emerged from this study. We found that almost all patients with PLS needed gait assistive devices over an extended period of time - a cane, walker or wheelchair. In contrast to patients with ALS, who typically need a wheelchair by two years after symptom onset (4), only a third of the patients with PLS reported currently using a wheelchair, and many had symptoms for a decade or more. In PLS the type of gait assistive device needed changed over a time span of years as mobility became more impaired, highlighting the need for durability and proper fit of walkers and wheelchairs. Only a few patients with PLS needed speech assistive devices or respiratory assistive devices. Discussion of issues such respiratory needs and speech devices are advised early in the course of ALS (3); in patients with PLS such discussions are less urgent.

Families and caregivers of patients with PLS made modifications to their homes, retired and/or changed their jobs. Surprisingly, despite the emotional toll, few patients felt the need for psychological or emotional support. The use of internet support groups and online discussions allows patients with PLS to interact with other PLS patients, a phenomenon that has been reported in other chronic illnesses (6,7).

The needs of PLS patients are distinct from ALS patients. They report needing fewer types of support services, but those that are needed are needed over an extended period of time. Our clinic population may be unusual, as they are actively involved in clinical research. If their pattern of primary care is similar to other PLS patients, however, the mechanism for obtaining support services is less systematic than for patients with ALS. The PLS patients in this survey rely to a lesser degree on organized clinics, generally by choice, seeking out help on their own, by referrals from their primary physicians, and/or using internet sources for advice.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgements

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Figure 1.

A. Assistive device used for gait by PLS patients, graphed by duration of symptoms. Each point represents the current use by an individual patient. If more than one device was used, the most dependent one is indicated.

B. Progression of the use of a cane, walker and wheelchair over time in individual patients in the subset that currently use a wheelchair.