ORIGINAL ARTICLE

Profile of Patients with Amyotrophic Lateral Sclerosis Across Continuum of Care

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ABSTRACT: ***Objective:*** This study describes the socio-demographic and clinical profile of persons with amyotrophic lateral sclerosis (ALS) in home care, nursing homes and complex continuing care settings in several Canadian jurisdictions. ***Methods:*** A cross-sectional study was conducted using available Resident Assessment Instrument (RAI 2.0 and RAI Home Care) national databases from 1996-2011. The profile of ALS patients was compared with patients without pre-specified neurological conditions. ***Results:*** There were 2,092 ALS patients identified in these settings. Persons with ALS were more likely than those in the comparison group to suffer from health instability (25.4%) and minor to major depressive symptoms (27.2%) , to experience falls (44.0%) and weight loss (22.9%), to require extensive assistance in activities of daily living (54.9%), and to receive rehabilitation services: physical (23.9%), speech language pathology (8.9%), and occupational therapy 43.3%). ***Conclusions:*** The ALS population in this study are greatly affected by a number of health issues. They are more likely than the comparison group to require therapies, medical interventions, and psychotropic drug use. While persons with ALS have a poor prognosis, a great deal could be done to enhance their quality of life and the quality of care they receive.

RÉSUMÉ: Profil des patients atteints de sclérose latérale amyotrophique dans l'ensemble des différentes étapes de soins. ***Objectif :*** Cette étude décrit le profile sociodémographique et clinique d'individus atteints de sclérose latérale amyotrophique (SLA) qui reçoivent des soins à domicile, en établissement de soins prolongés ou en milieu de soins continus plus complexes dans plusieurs juridictions canadiennes. ***Méthode :*** Nous avons effectué une étude transversale au moyen de l'Instrument d'évaluation des résidents (IÉR 2,0 et IÉR soins à domicile) dans des bases de données nationales de 1996 à 2011. Le profil des patients atteints de SLA a été comparé à celui de patients sans problème neurologique prédéterminé. ***Résultats :*** Nous avons identifié 2 092 patients atteints de SLA. Les individus atteints de SLA étaient plus susceptibles que ceux du groupe témoin d'avoir un état de santé instable (25,4%) et des symptômes dépressifs allant de mineurs à majeurs (27,2%), de présenter des chutes (44,0%), de perdre du poids (22,9%), d'avoir besoin de beaucoup d'aide pour les activités de la vie quotidienne (54,9%) et de recevoir des services de réadaptation : réadaptation physique (23,9%), orthophonie (8,9%) et ergothérapie (43,3%). ***Conclusions :*** La population de patients atteints de SLA est très touchée par plusieurs problèmes de santé. Ces patients sont plus susceptibles que ceux du groupe témoin d'avoir besoin de différents traitements, interventions médicales et médicaments psychotropes. Bien que le pronostic soit réservé chez ces patients, il y a beaucoup à faire pour rehausser leur qualité de vie et la qualité des soins qu'ils reçoivent.

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Amyotrophic lateral sclerosis (ALS) is a fatal degenerative disease involving motor neurons in the brain and spinal cord[1-3]. The annual age-adjusted incidence rate for ALS is about 2 per 100,000 in Canada[4]. Similar estimates are reported for Europe[5]. The age of onset of ALS symptoms is between 40-70 years[3] with reported peaks at 47-52 years and 58-63 for familial and sporadic disease, respectively"[1], and it is more common in men[1,3].

Amyotrophic lateral sclerosis primarily results in physical disability, but can also cause behavioural and cognitive impairment[6,7]. Depression, anxiety and sleep disorders are common[1,8]. It is a fatal disease (typically due to respiratory failure) with average survival of three to five years from the onset of symptoms based on clinic experience[1,3].

Large scale studies of persons with ALS across the continuum of care are scarce. To provide appropriate care and treatment for persons with ALS, it is necessary to have a comprehensive understanding of the characteristics of persons with ALS across care settings. This knowledge is particularly important considering how disabling ALS is. This study aims to provide a comprehensive profile of the socio-demographic, clinical characteristics, and health resource utilization of three distinct groups of persons with ALS, that is those in: (1) home care (HC); (2) nursing homes (NH); and (3) and complex continuing

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care (CCC) hospitals/units compared to persons without ALS or selected neurological conditions.

Methods

Design, Sample and Source of Data

This cross-sectional study forms an integral part of interRAI Canada's initiative, "Innovations in Data, Evidence and Applications for Persons with Neurological Conditions" (ideas PNC). This larger initiative aims at identifying the prevalence, profile and needs of persons with neurological conditions, such as ALS, dementia, epilepsy, multiple sclerosis, and stroke across the care continuum (http://interraicanada.uwaterloo.ca/).

The sample for this study was drawn from two large clinical databases: The Complex Continuing Care Reporting System (CCRS) and the Ontario Association of Community Care Access Centers (OACCAC) database.

The CCRS is managed by the Canadian Institute for Health Information and contains data on demographic, clinical, functional characteristics and resource use on persons receiving continuing care in either CCC hospitals/units or NH settings[9]. In both settings, interRAI's Resident Assessment Instrument 2.0 (RAI 2.0) is used as the standard assessment in normal clinical practice (www.interRAI.org). Only Ontario and Manitoba have CCC hospitals/units. In Ontario (173 CCC hospitals/unit), the CCRCS database includes quarterly and full RAI 2.0 assessments from April 10, 1996 to March 31, 2011. In Manitoba (one CCC hospital), where RAI assessments began in July 1, 2008, the data covers the period from July 1, 2008 to March 31, 2011.

The NH data include quarterly and full annual RAI 2.0 assessments over different time periods: Ontario (635 facilities) and Nova Scotia (8 facilities) from July 1, 2003 to March 31, 2011; British Columbia (126 facilities) from July 1, 2006 to March 31, 2011; Manitoba (38 facilities), Newfoundland (seven facilities), Saskatchewan (168 facilities), and Yukon Territory (two facilities) from July 1 to March 31, 2011. RAI 2.0 has been fully implemented in Ontario and Yukon, whereas its implementation is still underway in the other provinces.

The OACCAC database contains demographic, clinical, functional characteristics and service use data for persons in Ontario only who are expected to be receiving home care services for 60 days or more using interRAI's Resident Assessment Instrument–Home Care (RAI-HC)[10]. The home care RAI assessments are conducted twice yearly and include full assessments of persons residing in the community from January 1, 2002 to December 31, 2010.

For the purposes of this study, the most recent RAI 2.0 and RAI-HC assessments were used.

Ethics Approval

The ideasPNC research project received ethics approval from the Office of Research Ethics, University of Waterloo (Project #17045).

Case Ascertainment

The diagnostic coding for ALS was based on written text in the OACCAC database. To obtain accurate diagnostic information from free text entry fields, a detailed iterative process was undertaken. Initially, neurological experts provided lists of free texts used in clinical practice to identify all the neurological conditions of interest. Then, analysts searched the database for similar key words and identified additional similar terms for verification by the neurologists for possible inclusion. Finally, based on an approved list of terms, a macro was applied to pick out each client with a given free text diagnosis (if applicable) and assign the appropriate diagnosis in the final dataset.

In the CCRS database, "Amyotrophic Lateral Sclerosis" was selected from a pick list of diagnoses provided in the RAI 2.0 form. The interRAI assessment instruments used in the three databases do not provide specific diagnostic information on frontotemporal dementia (FTD) and, thus, our ALS data do not specify FTD diagnosis.

Gambassi and colleagues (1998) reported high positive predictive values for neurological diagnoses in the RAI 2.0 when linked with other clinical datasets[11]. Similar findings were reported in a Canadian study on diagnostic coding in patients admitted to CCC hospitals/units from acute care[12]. However, neither of these studies included ALS as a diagnosis.

The profile of persons with ALS was examined relative to a comparison group that included patients without ALS or any of the following pre-specified neurological conditions identified using a combination of written texts, pick lists and ICD-10-CA codes: Alzheimer's disease/other dementias, epilepsy, cerebral palsy, Huntington's disease, multiple sclerosis, muscular dystrophy, Parkinson's disease, stroke, spinal cord injury, and traumatic brain injury.

Clinical Measures

The interRAI family of assessment instruments offers a system of integrated health information on individuals' performance in terms of their physical, psychosocial and cognitive function, health status, and treatments or interventions needed[13]. The suite of assessment instruments includes a variety of clinical scales, such as the Cognitive Performance Scale (CPS)[14], the Depression Rating Scale (DRS)[15], the Changes in Health, End Stage Disease and Signs and Symptoms (CHESS) Scale[16], the Activities of Daily Living Hierarchy Scale (ADLH)[17], the Aggressive Behaviour Scale (ABS)[18], the Pain Scale[19] and the Instrumental Activities of Daily Living Scale (IADL)[20]. The reliability and validity of the interRAI instruments have been widely demonstrated[21-24].

Analysis

All analyses of the results were conducted using SAS version 9.2, SAS Institute Inc., Cary, North Carolina (http://www.sas.com/). Descriptive analyses were conducted to describe the profiles of persons with ALS and those of persons in the comparison group. Chi-square tests of significance were calculated to compare diagnostic groups (ALS vs. comparison group) within each of the three care sectors. Significance was assumed at the $p < 0.05$ level.

Table 1: Demographic Profile of Patients with Amyotrophic Lateral Sclerosis (ALS) and those in the Comparison Group^

Demographic Characteristics	HC		NH		CCC	
	CG	ALS	CG	ALS	CG	ALS
N	320,581	1,454	51,000	247	95,338	391
Female	64.7%	45.7%	69.5%	55.1%	61.0%	47.6%
Age Group						
0-44	4.9%	5.6%	0.7%	2.8%	2.0%	4.6%
45-54	6.6%	14.4%	1.6%	9.3%	3.9%	11.0%
55-64	11.1%	26.2%	4.4%	10.5%	8.7%	23.0%
65-74	16.6%	29.3%	9.3%	23.5%	19.4%	25.6%
75-84	31.6%	20.9%	26.5%	28.3%	36.7%	27.9%
85+	29.3%	3.6%	57.5%	25.5%	29.4%	7.9%
Married						
Male	56.3%	76.8%	23.3%^{ns}	20.7%	45.4%	42.0%
Female	29.4%	56.9%	8.0%*	16.9%	22.5%	30.1%
Overall	38.9%	67.7%	12.7%*	18.6%	31.5%	36.3%^{ns}
Diagnosis						
Heart Failure	13.5%	1.7%	19.7%	6.2%	15.8%	4.1%
Emphysema/COPD	18.6%	7.8%	19.5%	13.2%	18.5%	6.2%
Diabetes	24.9%	9.7%	24.7%	15.8%	22.9%	15.9%
Cancer	24.2%	3.7%	13.0%	5.8%	36.0%	7.5%
Any psychiatric	22.9%	31.9%	48.5%	56.3%	36.6%	60.9%
Other CVD	63.5%	36.9%	60.6%	46.9%	48.5%	34.7%

COPD=Chronic Obstructive Pulmonary Disease; CVD=Cardiovascular Disease. HC=Home Care. NH=Nursing Home. CCC=Complex Continuing Care. CG=Comparison Group. ^The Comparison Group comprises persons without any of the following neurological conditions: Alzheimer's disease/other dementias, Cerebral Palsy, Epilepsy, Huntington's Disease, Multiple Sclerosis, Muscular Dystrophy, Parkinson's Disease, Stroke, Spinal Cord injury, and Traumatic Brain Injury. Unless otherwise noted, the p values for chi-square tests of significance in this table are less than or equal to .0001 for tables comparing diagnostic groups within sectors. The symbol "ns" refers to values that are not significant at the .05 level and "*" denotes those that have significant p values below .05 but above .0001.

Table 2: Profile of Functional Performance of Patients with Amyotrophic Lateral Sclerosis (ALS) and those in the Comparison Group ^

Functional Performance	HC		NH		CCC	
	CG	ALS	CG	ALS	CG	ALS
N	320,581	1,454	51,000	247	95,338	391
Cognitive Performance Scale						
0 (Intact)	63.5%	55.6%	32.6%	31.6%	34.3%	24.3%
1-2 (Borderline intact-mild impairment	33.1%	36.3%	35.6%	28.7%	35.2%	36.6%
3-4 (Moderate-Moderate severe impairment)	2.4%	6.1%	22.2%	29.6%	20.3%	24.0%
5-6 (Severe-Very severe impairment)	1.0%	1.9%	9.6%	10.1%	10.2%	15.1%
ADL Hierarchy Scale						
0 (Independent)	73.1%	22.9%	11.9%	2.4%	8.5%	2.8%
1-2 (Supervision required-limited impairment)	18.1%	22.2%	23.7%	8.9%	24.9%	4.3%
3+ (Extensive assistance required – total dependence)	8.8%	54.9%	64.4%	88.7%	66.6%	92.8%
Pain Scale						
0 (No pain)	29.7%	40.9%	41.7%	42.9%	20.6%	27.9%
1-2 (Less than daily pain-Daily pain not severe)	54.7%	47.9%	52.3%	51.0%	66.7%	64.7%
3+ (Daily severe pain)	15.6%	11.1%	6.0%	6.1%	12.7%	7.4%
CHESS Scale						
0 (Not at all unstable)	31.2%	10.2%	39.0%	36.4%	15.8%	14.6%
1-2 (Little - Some instability)	54.4%	64.4%	47.1%	47.4%	42.1%	47.1%
3+ (Moderately -Highly unstable)	14.4%	25.4%	14.0%	16.2%	42.1%	38.4%
IADL Items						
Any Impaired IADL	86.5%	97.0%	n/a	n/a	n/a	n/a
Meal Preparation	77.9%	95.3%	n/a	n/a	n/a	n/a
Managing Finances	53.3%	74.9%	n/a	n/a	n/a	n/a
Managing Medications	41.3%	70.5%	n/a	n/a	n/a	n/a
Transportation	66.6%	87.4%	n/a	n/a	n/a	n/a

IADL=Instrumental Activities of Daily Living. HC=Home Care. NH=Nursing Home. CCC=Complex Continuing Care. CG=Comparison Group. NA=Not Available. ^The Comparison Group (CG) comprises persons without any of the following neurological conditions: Alzheimer's disease/other dementias, Cerebral Palsy, Epilepsy, Huntington's Disease, Multiple Sclerosis, Muscular Dystrophy, Parkinson's Disease, Stroke, Spinal Cord injury, and Traumatic Brain Injury. Unless otherwise noted, the p values for chi-square tests of significance in this table are less than or equal to .0001 for tables comparing diagnostic groups within sectors. The symbol "ns" refers to values that are not significant at the .05 level.

RESULTS

Sample Size

In the three care settings, the sample of persons with ALS in this study was 2,092 with 69.5% (n=1,454) of cases in HC, 11.8% (n=247) in NH, and 18.7% (n=391) in CCC hospitals/units. The ALS sample of 2,092 comprised 0.23% of the total number of individuals with one of the pre-specified neurological conditions in all three settings (N=892,312). The comparison group (n=466,919) comprised all persons without any of the pre-specified neurological conditions (see above). All comparisons described in this paper are significant at the $p < 0.05$ level (unless otherwise specified).

Demographic Characteristics

Table 1 provides the basic demographic and diagnostic information for ALS patients and the comparison group. In all three settings, those with ALS were less likely to be female consistent with other reports that the incidence of ALS is more frequent in men with a ratio of 3:2[3]. Those with ALS were generally younger than the comparison group. Except for those in CCC settings, persons with ALS were more likely to be married than those in the comparison group.

In all three settings, persons with ALS were less likely to have one of the somatic comorbidities examined but more were more likely to have "any psychiatric diseases" (including anxiety

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and depressive symptoms, delusions and hallucinations) than the comparison group.

Clinical and Functional Profile

Table 2 shows the distributions of clinical scales available in each of the interRAI assessments. In all three settings, persons with ALS were more likely than those in the comparison group to have moderate to severe cognitive impairment (CPS scores of 3 - 6) with increasing prevalence across the more resource intensive care settings (HC → NH → CCC). Functionally, persons with ALS were more likely than the comparison group to require extensive assistance or to be totally dependent (ADLH scores of 3+).

In all settings, persons with ALS were less likely to have pain but more likely to have high health instability (CHESS scores of 3+) than the comparison group.

Health and Social-Economic Profile

Table 3 shows the health, social and economic profiles of the study sample. Those with ALS in HC settings were more likely to experience higher rates of falls than the comparison group despite increased wheelchair use. Persons with ALS were less likely than the comparison group to walk and bathe independently in all settings. Also, those with ALS in NH settings were substantially more likely to have pressure ulcers, and they were much more likely to have swallowing problems than the comparison group in all settings. In HC, persons with ALS were more likely than the comparison group to report fair/poor health. Persons with ALS in all settings also had higher rates of bowel incontinence, while those in NH and CCC settings were more likely to have bladder incontinence than the comparison group. In all settings, the ALS group was more likely to have unintended weight loss of 5% or more in last 30 days or 10% or more in the last 180 days than the comparison group. Except for those in CCC settings, the ALS group was more likely than the comparison group to show a noticeable decrease in the amount of food or fluid intake. With respect to IADL, persons with ALS in HC settings were more likely to have any IADL impairments than the comparison group. In terms of communication issues, the ALS group was much more likely than the comparison group in all three settings to experience communication impairments, including expression and comprehension.

Those with ALS were also more likely than the comparison group to exhibit moderate or worse depressive symptoms (DRS scores of 3+) or anxiety symptoms than the comparison group. Only those with ALS in NH settings were more likely to manifest aggressive behaviours than the comparison group; however, the rate of aggressive behaviour for persons with ALS was much higher in facility-based settings than in HC settings. In all three settings, persons with ALS were more likely than the comparison group to have been physically restrained and received antidepressants and anxiolytics. The ALS group in CCC settings also was more likely to receive sedatives than the comparison group.

With respect to social and economic issues, those with ALS in HC and CCC settings were more likely to experience conflict with others than the comparison group. In all settings, those with ALS were less likely to experience social isolation than the

Table 3: Health, Social and Economic Profile of Patients with Amyotrophic Lateral Sclerosis (ALS) and those in the Comparison Group ^

Health, Social and Economic Profile	HC		NH		CCC	
	CG	ALS	CG	ALS	CG	ALS
N	320,581	1,454	51,000	247	95,338	391
Health Issues						
Fell in last 90 days	28.8%	44.0%	14.8%	13.0%	24.5%	11.0%
Unsteady gait	54.6%	70.9%	35.0%	29.1%	42.4%	23.3%
Shortness of breath	28.0%	29.8%	18.3%	17.8%	32.9%	39.9%
Loss of appetite	12.8%	11.8%	29.2%	32.0%	39.1%	30.2%
Pressure ulcers	9.0%	7.2%	8.3%	9.3%	14.5%	21.2%
Trouble swallowing	6.5%	60.8%	13.0%	51.8%	18.6%	75.4%
Weight loss	11.4%	22.9%	11.4%	16.2%	17.3%	20.7%
Nutritional problems	4.2%	6.9%	29.2%	32.0%	39.1%	30.2%
Fair/Poor self-rated health	22.7%	35.7%	n/a	n/a	n/a	n/a
Occasional/worse Incontinence						
Bladder	20.2%	17.5%	53.9%	58.7%	32.9%	40.9%
Bowel	6.1%	9.5%	26.9%	36.8%	27.4%	48.1%
Communication Impairments						
Expression	3.0%	21.7%	9.8%	26.7%	11.6%	38.6%
Comprehension	3.7%	4.1%	10.8%	12.6%	14.1%	18.2%
Mobility						
Uses wheelchair	6.8%	36.7%	56.7%	73.1%	44.7%	67.5%
Walks independently	35.1%	13.8%	25.0%	10.1%	13.6%	5.9%
Bathes independently	33.8%	13.7%	1.3%	1.2%	5.0%	1.8%
Mental Health						
Anxiety Symptoms	12.8%	15.0%	34.3%	39.7%	23.4%	34.8%
Delirium	2.9%	1.6%	9.9%	9.7%	16.7%	16.1%
Any Aggressive Behaviour	3.3%	3.0%	27.1%	33.2%	20.3%	21.0%
Hallucinations/Delusions	1.5%	0.6%	3.8%	2.4%	5.3%	2.6%
Depression						
0 (Not depressed)	63.6%	46.6%	42.6%	33.5%	50.7%	32.0%
1-2 (Some depressive symptoms)	22.0%	26.3%	29.5%	28.2%	28.6%	29.2%
3+ (Potential minor – Major depressive episode)	14.5%	27.2%	27.9%	38.4%	20.8%	38.8%
Physical Restraint Use	0.2%	1.0%	8.4%	16.6%	9.2%	14.1%
Psychotropic Drug Use						
Antipsychotics	5.6%	4.1%	20.2%	15.4%	14.6%	13.8%
Antidepressants	19.9%	30.6%	41.0%	51.4%	25.3%	52.7%
Anxiolytics	17.4%	19.1%	19.6%	32.4%	34.6%	48.3%
Sedatives	14.3%	12.9%	12.5%	10.1%	15.7%	23.3%
Social & Economic Issues						
Made economic trade-offs†	2.3%	2.5%	n/a	n/a	n/a	n/a
Conflict with others	11.1%	12.2%	13.9%	14.8%[ns]	11.8%	18.3%
Social Isolation	18.8%	6.3%	5.0%	2.3%	6.2%	4.1%
Caregivers distressed/overwhelmed	12.8%	29.4%	n/a	n/a	n/a	n/a

HC=Home Care. NH=Nursing Home. CCC=Complex Continuing Care. CG=Comparison Group. NA=Not Available. ^The Comparison Group comprises persons without any of the following neurological conditions: Alzheimer's disease/other dementias, Cerebral Palsy, Epilepsy, Huntington's Disease, Multiple Sclerosis, Muscular Dystrophy, Parkinson's Disease, Stroke, Spinal Cord injury, and Traumatic Brain Injury. Unless otherwise noted, the p values for chi-square tests of significance in this table are less than or equal to .0001 for tables comparing diagnostic groups within sectors. The symbol "ns" refers to values that are not significant at the .05 level. †Because of limited funds, during the last 30 days, the patient made trade-offs among purchasing adequate food, shelter, clothing, prescribed medications, sufficient home heat or cooling, OR necessary health care.

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Table 4: Health Resource Utilization of Patients with Amyotrophic Lateral Sclerosis (ALS) and those in the Comparison Group ^

Health Resource	HC		NH		CCC	
	CG	ALS	CG	ALS	CG	ALS
N	320,581	1,454	51,000	247	95,338	391
Any Rehabilitation						
Physical Therapy	12.2%	23.9%	55.1%	64.0%	63.2%	61.6%
Occupational Therapy	10.7%	43.3%	4.8%	4.9%	47.4%	55.8%
Speech Language Pathology	0.3%	8.9%	0.2%	1.6%	6.5%	27.9%
Social Work/Psychologist	1.6%	7.0%	1.4%	1.2%[ns]	11.7%	15.6%
Psychiatrist	n/a	n/a	n/a	n/a	n/a	n/a
Other physician visits	36.7%	39.1%	66.8%	68.4%	93.4%	93.9%[*]
Personal Support/Homemaking	55.7%	0.0%	n/a	n/a	n/a	n/a
Recreation therapy	n/a	n/a	12.2%	15.0%	31.3%	30.9%
Medical Interventions						
Respirator/other device	0.4%	6.1%	0.3%	4.5%	0.5%	19.2%
Oxygen/Respiratory therapy	10.3%	16.5%	11.6%	17.0%	27.4%	39.9%
Intravenous	4.9%	1.7%	2.0%	4.0%	12.7%	10.0%
Nurse monitoring	38.3%	35.5%	n/a	n/a	n/a	n/a
Enteral/Tube feeding	1.0%	21.5%	0.9%	19.4%	2.9%	45.5%
Tracheostomy care	0.3%	2.1%	0.2%	2.8%	1.1%	17.4%
Wound care	27.0%	34.1%	20.7%	27.1%	39.5%	37.1%
Emergency Department Visits						
None	78.6%	85.7%	79.9%	85.4%	54.4%	67.5%
1	15.8%	11.6%	17.1%	13.8%	36.5%	26.7%
2+	5.6%	2.7%	3.0%	0.8%	9.1%	5.8%
Hospitalizations						
None	64.7%	81.4%	82.4%	88.1%	38.8%	68.1%
1	29.2%	17.0%	15.6%	11.9%	43.0%	24.9%
2+	6.1%	1.7%	2.0%	0.0%	18.2%	6.9%

HC=Home Care. NH=Nursing Home. CCC=Complex Continuing Care. CG=Comparison Group. NA=Not Available. ^The Comparison Group comprises persons without any of the following neurological conditions: Alzheimer's disease/other dementias, Cerebral Palsy, Epilepsy, Huntington's Disease, Multiple Sclerosis, Muscular Dystrophy, Parkinson's Disease, Stroke, Spinal Cord injury, and Traumatic Brain Injury. Unless otherwise noted, the p values for chi-square tests of significance in this table are less than or equal to .0001 for tables comparing diagnostic groups within sectors. The symbol "ns" refers to values that are not significant at the .05 level and "*" denotes those that have significant p values below .05 but above .0001.

comparison group. In HC settings, caregivers of persons with ALS were more likely than caregivers of persons in the comparison group to be distressed or overwhelmed.

Health Resource Utilization

Table 4 shows the health resource utilization of the study sample. Those in HC settings were more likely than the comparison group to receive all of the rehabilitation services or therapies listed. Those in CCC settings were more likely than the comparison group to receive occupational, speech language pathology, and social work/psychologist services/therapies. In all settings, the ALS group was more likely than the comparison group to receive oxygen/respiratory therapy, make use of a respirator or other devices, receive tube feeding and tracheostomy care than the comparison group.

Despite their general higher scores on clinical and functional indicators in all three settings, the ALS group was less likely to have made emergency department visits or required hospitalizations than the comparison group.

Discussion

This very large and unique study, describing the socio-demographic and clinical characteristics and health resource utilization of persons with ALS across the continuum of care, may be the largest of its kind. Only a very small proportion of individuals with neurological conditions in the three study settings had a diagnosis of ALS consistent with the low prevalence of ALS in the general population. However, ALS is associated with substantial morbidity and mortality[4]. The widespread adoption of interRAI assessment instruments in Canadian provinces/territories has made it possible to obtain standardized clinical information for persons with ALS with sample sizes not typically seen in the literature. The important clinical differences among ALS populations in the three health care settings underscore the importance of having common, standardized function and clinical measures across the continuum of care. Although the ALS diagnosis points to a range of important clinical consequences for persons with the condition, the present findings demonstrate substantial heterogeneity within the ALS population between service settings.

Amyotrophic lateral sclerosis is an adult disease with an onset peak age between ages 55 and 74. Within this age group, the majority of ALS patients remain in the community receiving home care. It is interesting to note that most of those in HC are married (67.7%), which may suggest the important role that spousal support has in preventing institutional care. In contrast, only 18.6% of persons with ALS in NH are married.

A review of the clinical profile of the ALS group shows a severity gradient across the three settings. For instance, the highest proportion of ALS patients having any psychiatric comorbidities (Table 1) is situated in CCC (60.9%) followed by NH (56.3%) and HC (31.9%). A similar gradient is also noted with depressive symptoms and functional impairments. The majority of ALS patients requiring extensive assistance or totally dependent are in CCC, followed by NH and HC, as would be expected with decreased levels of disability through this continuum of care. Persons with ALS in HC were more likely to have moderate to high health instability and to have self-rated their health as fair/poor compared. Since pain is not a primary symptom in ALS, it is not surprising that ALS patients were less likely to have pain. However, 59% in HC, 57% in NH, and 72% in CCC reported having pain (less than daily to severe daily pain) highlighting the need to maintain a high index of suspicion for presence of pain symptoms in ALS patients. This finding is consistent with a prior study where 60% of ALS patients reported experiencing pain[25].

The high rates of falls and unsteady gait in the study sample is expected, given that weakness is associated with ALS. Those in HC experienced higher rates of falls and unsteady gait compared to CCC and NH settings. It is not surprising that ALS patients in CCC and NH were more likely to be in wheelchairs than those in HC, and those in HC were more likely to be able to walk independently. These findings suggest that as ALS patients become weaker and unable to walk independently their likelihood of requiring either NH or CCC for increased level of care and supervision increases.

As expected, ALS patients in this study were more likely to experience breathing difficulties[1]. Patients with ALS in CCC

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settings were most likely to have shortness of breath and, not surprisingly, were more likely to receive oxygen therapy and or require a ventilator than those in either HC or NH settings. It is not surprising that the rate of dysphagia was also higher in ALS patients in all three settings given that this is a common symptom seen in clinical practice[1]. An interesting finding was the higher rates of pressure ulcers in NH and CCC settings in ALS patients compared to the comparison group. While the development of pressure ulcers may be a rarity in ALS patients, the data show that the risk is present and clinicians should be mindful of it.

In this study, 21.7% of ALS patients experienced weight loss. This finding may be associated with loss of muscle bulk, or as a complication of breathing and swallowing difficulties. Other researchers have reported decrease in body fat, lean body mass and muscle power in those with ALS[26]. The results of the current study point to the need for appropriate management of ALS patients' nutrition, including an assessment by a nutritionist. Persons with ALS in this study also showed higher rates of bowel and bladder incontinence. This may relate to their being less likely to be independent in locomotion and greater overall ADL dependence.

Consistent with other research[27], caregivers of ALS patients in the community were more likely to be distressed. Aside from the physical aspects of providing care to their family member who is affected by ALS, factors such as cognitive impairment, falls, and communication impairments may all contribute to caregivers' psychological distress.

It was interesting that the ALS group in all three settings was least likely to have made emergency department visits or required hospitalizations. This may be related to more access to a multidisciplinary team, including physical and speech therapists, and physicians. However, other research has shown inconsistent results of the benefits of multidisciplinary services in ALS patients[28,29].

Strengths and Limitations

The use of validated national multi-jurisdiction clinical databases involving very large sample sizes is one of the strengths of this study. This study demonstrated the feasibility of using the interRAI suite of assessment instruments to assess the socio-demographic characteristics, health profiles and health resource utilization of people with ALS across the continuum of care.

This study also has a number of limitations. Its cross-sectional design limited the ability to estimate the incidence of clinical change in ALS patients; however, future research may readily address this limitation by making use of the longitudinal assessment records available in the databases described here. Data were only collected in the participating Canadian provinces, but not in the rest of Canada where the interRAI assessment systems have not yet been implemented. As well, the study sample is limited to ALS patients who are accessing the health care settings studied, but there are no comparable data on those who may have undiagnosed ALS or those who are not yet receiving home care or more complex continuing care. An important opportunity to remedy this information gap would be to ensure that any registries developed for neurological conditions such as ALS include measures that would be compatible with the interRAI instruments already in widespread use to assess persons in HC, LTC and CCC settings. One other limitation is that the data available could not specify if patients with ALS also had FTD. Future studies should endeavour to secure such data.

Conclusion

In summary, the ALS population described in this study has exceptional needs that require substantial resources from service providers spanning the continuum of care. Clearly, persons with ALS are more likely to require therapies, medical interventions, and pharmacotherapies than the comparison group. The functional and clinical consequences of ALS also place a substantial psychological distress on caregivers. While persons with ALS have a poor general prognosis, a great deal could be done to potentially enhance specific aspects of their quality of care.

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