Research Paper

Clinical Autonomic Research 1999, **9**:1–4

Occurrence of depressive symptoms in Shy-Drager syndrome

Corinne E. Gill, PH.D.[1], Ramesh K. Khurana, M.D.[2], and Russell J. Hibler, PH.D.[2]

[1]Datahr Rehabilitation Institute, Brookfield, Connecticut, USA

[2]The Union Memorial Hospital, Baltimore, Maryland, USA

Address correspondence and reprint requests to Dr. Ramesh K. Khurana, Chief, Division of Neurology, The Union Memorial Hospital, 201 E. University Parkway, Baltimore, MD 21218, USA
Tel: (410) 554-2286; Fax: (410) 554-2179

Received April 8, 1998; accepted as revised October 25, 1998

We investigated the frequency and severity of depressive symptoms among patients with Shy-Drager Syndrome (SDS) and correlated depression with the extent of the patients' disability. Data were collected from 15 patients and their spouse caregivers through a mailed questionnaire. The patients were asked to complete the Beck Depression Inventory (BDI) questionnaire, while caregivers were asked to complete the self-assessment Parkinson's Disease Disability Scale and The Northwestern University Disability Scale for Parkinson's Disease. Data were statistically analyzed using descriptive statistics and Pearson-Product moment correlations. The prevalence of depressive symptoms was 85.7%; 28.6% of SDS patients scored in the moderately to severely depressed range. There was no significant correlation between the severity of depressive symptoms and disability ($r=0.02$, $p=0.94$) and the ability to perform activities of daily living ($r=0.0$, $p=1.0$).

The prevalence of depressive symptoms in patients with SDS is common. The patient's level of depression does not correlate with physical disability. Pharmacologic management and interventions aimed at increasing active coping methods should improve quality of life.

Key words: multiple system atrophy, Shy-Drager syndrome, depression, disability.

Multiple system atrophy (MSA) encompasses the term Shy-Drager syndrome (SDS) and presents with a spectrum of neurological features due to the involvement of extrapyramidal, autonomic, cerebellar, and pyramidal systems. It is a progressive neurodegenerative disease that leads to severe physical disability and reduced life expectancy [1]. Despite the clinical predisposition to depression, only a few reports describe the occurrence of depression in SDS. These reports provide limited information. In 1984, Kwentus and co-workers reported the first instance of an autopsy-proven SDS presenting with an insidious depression 6 years before the onset of the first neurological symptom [2]. The patient suffered a depressed mood that was characterized by sadness, reduction of libido, weight loss, insomnia, apathy, and eventual suicidal thoughts (toward the end of his illness). Ruxin and Ruedrich in 1994 described a 78-year-old male who was diagnosed with multiple system atrophy (MSA) based on his progressive neurological dysfunction, Parkinsonian symptoms, orthostatic hypotension, and an inadequate response to L-dopa. The patient had a concomitant diagnosis of major depression with dysphoric mood, poor appetite, sleep disturbance, loss of pleasure, and guilt [3]. In 1996, Pilo and co-workers examined depression in patients with multiple system atrophy and Parkinson's disease (PD) [4]. The researchers reported that there was a higher percentage of disability and disease progression in MSA patients, but there was no greater incidence of depression and no clinically significant depression in any of the patients.

These initial case reports raise the question of whether there is a co-occurrence of depression in SDS. Our clinical experience suggests that the complaint of depression is common among SDS patients. The objectives of the present study were as follows:

1) Determine the frequency and severity of depressive symptoms among SDS patients.

2) Consider the characteristics of depression.

3) Examine the relationship between depressive symptoms and the extent of the patient's disability.

4) Investigate the association between the onset of illness and depressive symptoms.

Methods

Fifteen patients with SDS and their spouse caregivers participated in the study. They were referred from an SDS support group and were diagnosed as having SDS by their neurologist. No attempt was made to control for medications or for the presence of other diseases. Patients were predominantly white (93.3%), married (93.3%), and male (73.3%). Their average age was 69 years ± 13.41 (SD). The mean duration of illness was approximately 5.27 ± 4.46 years (SD). The majority of patients had graduate school education or professional training. Twelve of the 15 patients responded to a follow-up questionnaire. Eight of these 12 patients reported depressive symptoms. The onset of the depressive symptoms preceded the diagnosis of SDS in four patients, began after the diagnosis of SDS in two patients, was observed concurrently in one patient, and one patient was uncertain. None of them were on medications that can exacerbate depression. Four patients were taking tricyclic antidepressants or serotonin reuptake inhibitors.

Independent variables included caregivers answering questions about demographics, including the patient and caregiv-

ers's age, sex, race, marital status, and duration of the patient's illness.

The *Beck Depression Inventory* [5,6] was completed by the patients. This is a self-report screening measure of depression. This instrument is comprised of 21 four-choice responses that index the number and severity of depressive symptoms. A total score is obtained by adding the highest score for each of the 21 items and ranges from 0 to 63. According to Spreen and Strauss, the cut-off points for the normal range are 0–9, for minimal depression 10–15, for mild to moderate depression 16–19, for moderate to severe depression 20–29, and for severe depression 30–63 [7]. The Spearman-Brown reliability has been reported as 0.93 and the internal consistency for the items as 0.86 [7].

The *Self-Assessment Parkinson's Disease Disability Scale* [8] was completed by the caregivers who rated how the patients performed numerous activities of daily living. The items focus on gross mobility and fine motor coordination tasks. The sum score varies from 24 to 120.

The *Northwestern University Disability Scale* (NUDS) [9] was also completed by the caregivers. This instrument indexes the patient's degree of disability in the domains of walking, personal hygiene, dressing, eating, feeding, and speaking. A sum score from 0 to 100 is obtained, with a lower score indicative of greater disability. Canter and colleagues noted that the inter-rater reliability for this scale was 0.95.

Data were collected through a mailed questionnaire. The protocol included the demographic questions, the Beck Depression Inventory, the Self-assessment Parkinson's Disease Disability Scale, and the Northwestern University Disability Scale. The Beck Depression Inventory was completed by the patient, and all other instruments were completed by the caregivers. Informed consent procedures were followed and consent was obtained from each patient and caregiver.

Data analysis

Descriptive statistics (frequencies) were obtained for the depression variable and for individual test items to summarize the data. Pearson product-moment correlations (2-tailed) were completed for each variable to investigate the relationships between depression and disability and activities of daily living. All variable were examined for normal distribution and linearity. One of the patients did not complete the BDI because he was too ill to respond to the questions. His missing score was replaced by the mean.

Results

The mean BDI score was 14.43 ± 6.67 (SD). The prevalence of depressive symptoms was 85.7%; 28.6% of SDS patients scored in the moderately depressed range. Depressive symptoms were characterized by fatigability, dissatisfaction, loss of libido, work inhibition, sadness, and feelings of hopelessness about the future (Table 1). On the self-assessment Parkinson's Disease Disability Scale, patients' mean score was 81.13 ± 29.11 (SD). On this scale, their greatest difficulties were: traveling by public transportation and writing a letter. The mean score for our SDS patients was higher than that reported previously for a group of Parkinsonian patients who had a longer duration of illness [8].

Table 1. Depressive symptoms identified by Shy-Drager patients on the Beck Depression Inventory (n = 15)

Fatigability	100%
Sense of dissatisfaction	92.9%
Loss of libido	85.8%
Work inhibition	85.8%
Sadness	78.6%
Hopelessness about the future	71.4%
Somatic preoccupation	64.3%
Indecisiveness	57.2%
Irritability	57.1%
Social withdrawal	57.1%
Self-accusation	50.0%
Loss of appetite	42.9%
Insomnia	42.8%
Crying	42.9%
Suicidal ideation	35.7%
Weight loss	28.6%
Self-dislike	28.5%
Unattractiveness	28.5%
Sense of failure	7.1%
Punishment	7.1%
Sense of guilt	0%

On the NUDS, the average disability score was 52.66 ± 12.28 (SD). Major areas of difficulty were eating and walking.

The results were further examined to evaluate association between depressive symptoms and disability ($r=0.02$, $p=0.94$) and the ability to perform activities of daily living ($r=-.00$, $p=1.0$). None of the correlations were significant. Of note, there was a negative but statistically non-significant correlation between depressive symptoms and duration of illness ($r=0.48$, $p=0.07$). See Table 2.

Discussion

In the healthy elderly population, 3% of men and 4% of women may suffer from depression [10]. Dooneief and colleagues reported depression in 47% of patients with Parkinson's disease [11]. Although Pilo et al. did not observe

Table 2. Correlations between depressive symptoms and patient disability, level of independent functioning, ability to perform activities of daily living, and duration of illness (n = 15)

Patient disability	−0.02
Patient's level of independent functioning	+0.08
Patient's ability to perform activities of daily living	0.00
Patient's memory and behavioral problems	+0.31
Duration of illness	−0.48†

†p = 0.07.

clinical evidence of depression among their MSA patients, their Beck Depression Inventory score was 18 or more in three out of 12 patients, indicating moderate to severe depressive symptoms [4]. In our study of 15 MSA patients, 28.6% had moderate to severe depressive symptoms characterized by dissatisfaction, loss of libido, work inhibition, and hopelessness about the future. The percentage of patients who acknowledged depression was noteworthy and higher than anticipated. This proportion was similar to an estimate of depressive symptoms reported in primary care settings throughout the United States [12]. This finding indicates that depression may be unrecognized and untreated in SDS patients. A high prevalence of depressive symptoms may suggest a more widespread involvement of systems critical not only to motor and autonomic activity but also to emotional functioning.

The depressive symptoms most frequently identified by patients were fatigue (100%), sense of dissatisfaction (92.9%), loss of libido (85.8%), work inhibition (85.8%), sadness (78.6%), and hopelessness (71.4%). Of these, it may be argued that fatigue, decreased libido, and work inhibition may be secondary to the effects of the vegetative disease process rather than from depression. In contrast, the feelings of dissatisfaction, sadness, and hopelessness are not necessarily concomitants of having a physical condition. These depressive symptoms may negatively impact on the person's well being, quality of life, interpersonal relationships, and response to medical treatment. This may exacerbate an already stressful situation for both the patient and the caregiver. Consequently, the identification of potentially treatable depressive symptoms in SDS patients is vital. If depression is suspected, simple screening methods and clinical evaluation may be used to diagnose the depression and subsequent pharmacological intervention should follow.

Our study reveals that individuals with a shorter duration of illness had a tendency toward more depressive symptoms. This was similar to findings obtained in another study that found a bimodal distribution of depression in Parkinsonian patients, with the highest frequency in the early and late stages [13]. This finding is not surprising, given that early in the disease process these patients often experience impotency, incontinence, and autonomic dysfunction. These symptoms respectively affect the patient's ability to have an intimate relationship with a spouse and to manage work responsibilities. For most adults, these capacities provide meaning and enjoyment to life. SDS, with its severe debilitating disease progress, robs the patients of basic abilities that are relied upon in daily interchange.

Is the co-occurrence of depressive symptoms and MSA an emotional response to the experience of physical disability, or is depression a primary symptom? Various studies in Parkinsonian patients have produced inconsistent results. For example, Starkstein et al. demonstrated a significant correlation between impairment and depression scores in Parkinson's patients [13]. On the other hand, Celesia and Wanamaker reported no significant relationship between these variables [14]. The results of our study were similar to those of Celesia and Wanamaker. Contrary to our expectations, the level of depressive symptoms in SDS patients did not correlate with physical disability. This suggests that patients did not experience depression due to the devastating motor disability, physical dependence on others, or the inability to perform basic activities of daily living. Depression and disability were either not linked or the increase in disability was slow enough to allow the patients to adapt.

This study has several limitations. The BDI, which is used to quantitate depression, is neither specific nor intended as a diagnostic instrument. A clinical psychiatric assessment would be required to establish the diagnosis of a depressive disorder [15]. In addition, we did not control for prior history of depression, antidepressants or other medications, diurnal mood variations, and presence of other diseases, that might affect the results. In a recent study, Berrios et al. compared psychiatric and autonomic tests in 32 Parkinsonian subjects and 32 controls. They reported a significant association of depression with the following autonomic symptoms: postural dizziness, frequency of micturition, urinary hesitancy, constipation, dry mouth, impotence, and loss of libido [16]. This suggests that the presence of autonomic failure may lead to an over diagnosis of depression. Future work in this area might explore the relationship between depression and impotency and hypotension in SDS patients. In order to generalize these findings, large scale studies of SDS patients are necessary to highlight profiles of mood changes in relation to progression in motor disability and identify the medications that are most effective for depression in these patients.

Acknowledgments

We thank Ms. Dorothy Trainor-Kingsbury and the Shy-Drager support group for referral of patients.

References

1. Wenning GK, Tison F, Schlomo B, Daniel SE, Quinn NP. Multiple system atrophy: a review of 203 pathologically proven cases. *Movement Disorders* 1997; 12:133–147.
2. Kwentus JA, Auth TL, Foy JL. Shy-Drager syndrome presenting as depression: case report. *J Clin Psychiatry* 1984; 45:137–139.
3. Ruxin RJ, Ruedrich S. ECT in combined multiple system atrophy and major depression. *Convulsive Therapy* 1994; 10:298–300.
4. Pilo L, Ring H, Quinn N, and Trimble M. Depression in multiple system atrophy and in idiopathic Parkinson's disease: a pilot comparative study. *Biol Psychiat* 1996; 39:803–807.
5. Beck AT, Ward CH, Mendelson M, Mock J, and Erbaugh J. "An Inventory for Measuring Depression." *Arch Gen Psychiatry* 1961; 4:53–63.
6. Beck AT. *Beck Inventory Depression: Manual.* San Antonio, TX: Psychological Corporation; 1987.
7. Spreen O, Strauss E. *A Compendium of Neuropsychological Tests: Administration, Norms, and Commentary.* New York: Oxford University Press; 1991:385–390.
8. Brown RG, MacCarthy B, Jahanshahi M, Marsden CD. Accuracy of self-reported disability in patients with Parkinsonism. *Arch Neurol* 1989; 46:955–959.
9. Canter GJ, deLaTorre R & Mier M. A method for evaluating disability in patients with Parkinson's disease. *J Nerv Ment Dis* 1961; 133:143–147.

10. Boyd JH, Weissman MM. Epidemiology of affective disorders. *Arch Gen Psychiatry* 1981; 38:1039–1046.
11. Dooneief G, Mirabello E, Bell K, Marder K, Stern Y, Mayeux R. An estimate of the incidence of depression in idiopathic Parkinson's disease. *Arch Neurol* 1992; 49:305–307.
12. Zung WW, Broadhead WE, Roth ME. Prevalence of depressive symptoms in primary care. *J Fam Practice* 1993; 37:337–344.
13. Starkstein SE, Preziosi TJ, Bolduc PL, and Robinson RG. Depression in Parkinson's disease. *J Nerv Ment Dis* 1990; 178:27–31.
14. Celesia GG, Wanamaker WM. Psychiatric disturbance in Parkinson's disease. *Dis Nervous Sys* 1972; 33:577–583.
15. Clinical Practice Guidelines, number 5. Depression in Primary Care: volume 1. Detection and diagnosis. U.S. Rockville, MD: Department of Health and Human Services (AHCPR Publication #93-0550); 1993.
16. Berrios GE, Campbell C, Polityńska BE. Autonomic failure, depression, and anxiety in Parkinson's disease. *Br J Psychiatry* 1995; 166:789–792.