Forum Reumatol.
2023, vol. 9, No. 3, 96–104

ISSN: 2720-3921, e-ISSN: 2720-3913
DOI: 10.5603/RF.a2023.0018

VIA MEDICA
www.journals.viamedica.pl

ORIGINAL ARTICLE

Klaudia Palka, Barbara Buc-Piorun, Karolina Nowak, Monika Bultrowicz, Martyna Kuczyńska, Przemysław Kotyla, Ewelina Machura

Department of Internal Medicine, Rheumatology and Clinical Immunology, Faculty of Medical Sciences, Medical University of Silesia, Katowice, Poland

Everything you always wanted to know about systemic sclerosis but were afraid to ask: Part 3. Vital activity of patients with systemic sclerosis

ABSTRACT

Systemic sclerosis is an inflammatory connective tissue disease of autoimmune origin, characterized by progressive fibrosis of the skin, internal organs and damage to blood vessels referred to as vasculopathy. Although the most visible symptom of the disease is hardening of the skin, the involvement of internal organs leading to their extreme insufficiency determines the severity of the disease, resulting in a severe course for the patient.

The method used to carry out this test is the diagnostic survey method. The paper used a questionnaire that contained 22 questions, including 7 open-ended questions and 2 multiple choice questions.

The aim of the work is to show how systemic sclerosis affects the patient's vital activity, with particular emphasis on those activities of everyday life that cause the greatest difficulty. In addition, an attempt was made to determine how the progression of the disease affects the physical and mental sphere of patients with systemic sclerosis.

Studies show that systemic sclerosis is a disease that affects the life activity of patients to varying degrees. It penetrates both the physical sphere of the patient, gradually limiting his independence, but also into his mental sphere.

Rheumatol. Forum 2023, vol. 9, No. 3: 96–104

KEY WORDS: systemic sclerosis; vital activity; inflammatory connective tissue disease

INTRODUCTION

Systemic sclerosis (SSc) is an inflammatory connective tissue disease of autoimmune origin marked by progressive fibrosis of the skin, internal organs and damage to blood vessels termed as vasculopathy. Although the most visible symptom of the disease is sclerosis of the skin, the involvement of internal organs leading to their extreme failure determines the severity of the disease, resulting in a severe course for the patient.

The first detailed description of SSc dates back to the 18th century. Dr Carlo Curzio of Naples published a monograph in 1753, where he gave a description of a seventeen-year-old patient, Patrizia Galiera. He described the patient's skin as dry, hard in the likeness of wood. In his monograph, he wrote "A patient with diffuse skin tightening and hardening of the skin all over the body and thickened eyelids…, cold skin, difficulty opening the mouth…".

The disease most commonly affects people aged between 30 and 50. This means that people become ill in the most active period of their lives. As it progresses, SSc affects the simplest activities of daily living. Patients often have to reduce their responsibilities or give up their jobs. Reduced independence is often associated with reduced contact with friends and family. Patients usually stay in a home or hospital environ-

Address for correspondence:
Klaudia Palka, MD
Department of Internal Diseases, Rheumatology and Clinical Immunology
Faculty of Medical Sciences, Medical University of Silesia
Ziołowa 45/47
40–635 Katowice, Poland
e-mail: klaudia.muszalik@tlen.pl

ment. All this has an impact not only on the physical well-being but also on the patient's depressed mood [1, 2].

The disease process can develop slowly and gradually or, in the case of an aggressive form of disease, manifest suddenly. The problems that patients will face are not only those from the physical sphere but also from the mental sphere. The period of the illness coincides with an age when the patients are economically active but also take an active part in family and social life, all of which means that the illness often limits their activities, bringing with it a depressed mood. This results in SSc patients becoming dependent on third parties at an age when their healthy peers are independent. Therefore, nursing care should focus on the patient's physical and psychological needs [1, 2].

The main problem is a limitation and, in advanced stages of the disease, a deficit in self-care. Activities of daily living (ADLs), such as toileting, putting on clothes and preparing meals, can be very difficult for patients whose limb mobility is restricted. Therefore, patients often have to use third-party assistance.

AIM OF THE STUDY

This study aims to show how systemic sclerosis affects the patient's ADLs, with a focus on those ADLs that cause the greatest difficulty. Moreover, an attempt was made to determine how the progression of the disease affects the physical and psychological spheres of SSc patients.

MATERIAL AND METHODS

METHOD, TECHNIQUE AND ORGANISATION OF THE STUDY

The method used for conducting this study is the diagnostic survey method. The study used a questionnaire that contained 22 questions, including seven open-ended questions and two multiple-choice questions. The questions in the questionnaire focused on the patient's assessment of their ADLs and the changes that had taken place in their lives after being diagnosed with SSc.

The surveys were collected *via* an online questionnaire. Participation in the survey was voluntary and anonymous. The questionnaire was completed by 77 people. The respondents are those who struggle with diffuse SSc (dSSc) or limited SSc (lSSc).

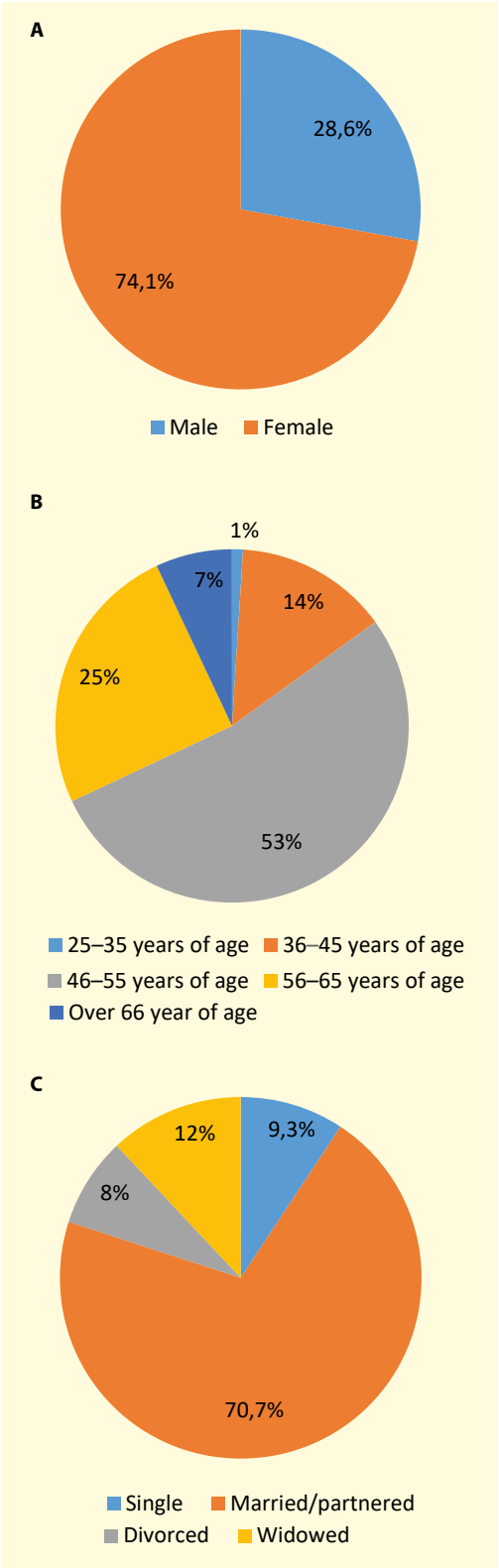

Figure 1. Demographics of patients with systemic sclerosis: **A.** Sex of respondents; **B.** Age of respondents; **C.** Marital status of respondents

The study group consisted of patients diagnosed with SSc based on European Alliance of Associations for Rheumatology (EULAR)/ /American College of Rheumatology (ACR)

classification criteria, including 56 women and 21 men aged between 25 and 68 years (Fig. 1A). In this group, 42 patients with dSSc were identified based on the Le Roy and Medsger criteria, while the remaining 35 were diagnosed with lSSc (Fig. 3A). Among this study group, as many as 46.8% of people were diagnosed with the disease between the ages of 36 and 45 years, while only 1.3%, or one person, was diagnosed between the ages of 18 and 25 years (Fig. 1).

CHARACTERISTICS OF THE STUDY GROUP

The largest group was those between 46 and 55 years of age — 53%. The majority of respondents were women — 71.4%. A significant proportion of respondents were married or in a civil partnership — 70.7%.

Respondents also provided answers regarding the fact of having children. The majority of respondents, 84.2%, confirm that they have children; the largest number of them have two children — 35.1% of respondents.

Respondents also identified their education, with the largest number of respondents having a secondary education, i.e., 45.5% (Fig. 2A). More than half of the patients surveyed are economically active with 26% working full-time and 24.7% undertaking part-time activity.

The impact of the illness on the difficulty of becoming gainfully employed resulted in 35.1% of respondents receiving disability benefits (Fig. 2B).

RESULTS

Systemic sclerosis was diagnosed most frequently between the ages of 36 and 45, with 46.8% of respondents giving this answer, followed by people diagnosed with SSc between the ages of 46 and 55, i.e,. 35,1%. Only one person surveyed was diagnosed earlier, between the ages of 18 and 25.

The results show that SSc is a chronic disease with 32.9% of respondents having been diagnosed with the disease for more than 6 years. The next largest group was found to be patients whose disease diagnosis had been known for three to 4 years — 27.6%.

Diagnostic difficulties in SSc patients are the reason for the significant delay between the onset of the first symptoms and when a formal diagnosis is made. In the study group, 19.7% were people who had known the diagnosis for 5 to 6 years. An identically sized group of patients, 19.7% , were diagnosed between 1 and 2 years after the onset of symptoms. It should be emphasised that the first symptoms and health problems that have been associated with SSc probably occurred much earlier in patients, while the time from actual diagnosis is relatively short. None of respondents had a confirmed diagnosis for less than a year (Tab. 1).

Systemic sclerosis, due to its significant musculoskeletal involvement, directly affects the patient's independence.

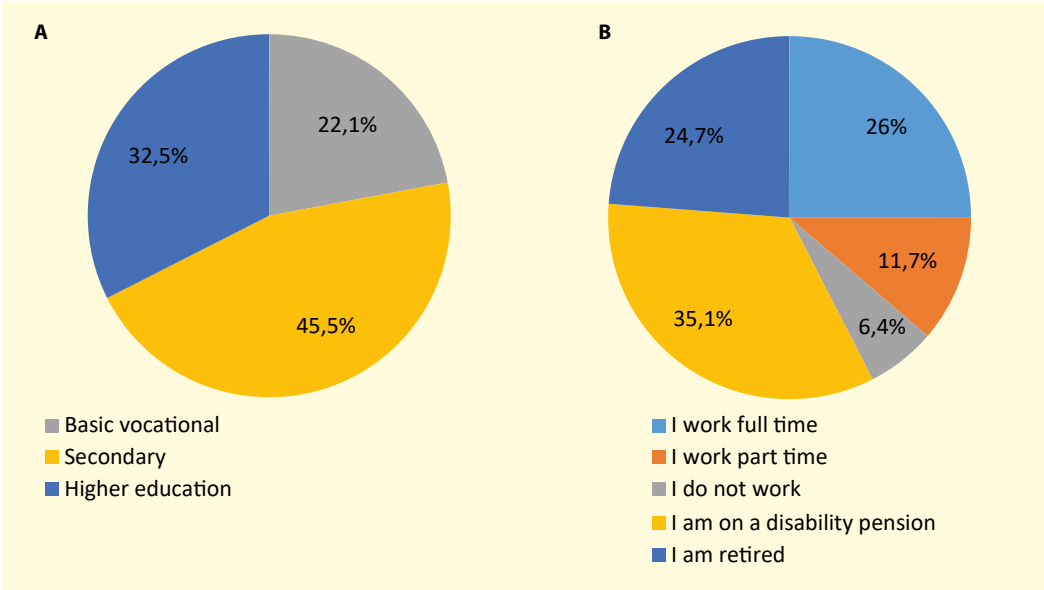

Figure 2. Education level and occupational status of respondents: **A.** Education level of respondents; **B.** Occupational status of respondents

In this study 55.8% of respondents declared completely independent mobility, while 42.9% of patients specified mobility with the help of orthopaedic equipment to facilitate movement, including 12 respondents using an orthopaedic cane or elbow crutches, and 6.5% of respondents using an orthopaedic walker. A wheelchair is a mobility aid for 18 respondents. One person among the respondents is a bedridden person (Fig. 3B).

Among the main groups, complaints occurring in the course of SSc were identified as 6 main groups. These included musculoskeletal symptoms occurring in 67.3% of respondents, pain affecting 62.3% of respondents. Cardiovascular symptoms in 55.1% of respondents, respiratory symptoms in 21% of respondents. Skin lesions in 42.4% of respondents, gastric symptoms in 14.5% of respondents.

In the surveyed population, the survey revealed the frequency of symptoms reported by the respondents. Among them, 58.4% of the patients reported experiencing the disease symptoms as quite frequent, while 32.5% of the participants described them as very frequent symptoms. This means that 70 participants out of 77 are significantly affected by their illness (Fig. 4A). It is also significant that none of the participants reported never experiencing the symptoms. They always accompany the patients to varying degrees. Physical well-being has a direct impact on mental well-being, which in turn affects family relationships and social interactions. Among the respondents, a positive result is that as many as 84.4% of respondents reported that the disease did not affect their family relationships. Three participants even noticed a positive impact

Table 1. Age at which systemic sclerosis was diagnosed among respondents and time since systemic sclerosis (SSc) diagnosis among respondents

Age at which the respondent was diagnosed with SSc					
Age	18–25 years of age	26–35 years of age	36–45 years of age	46–54 years of age	Over 55 year of age
% of respondents	1.3	3.9	46.8	35.1	13
Time since diagnosis of SSc among respondents					
Time since diagnosis	1–2 years	3–4 years	5–6 years	More than 6 years	
% of respondents	19.7	27.6	19.7	32.9	

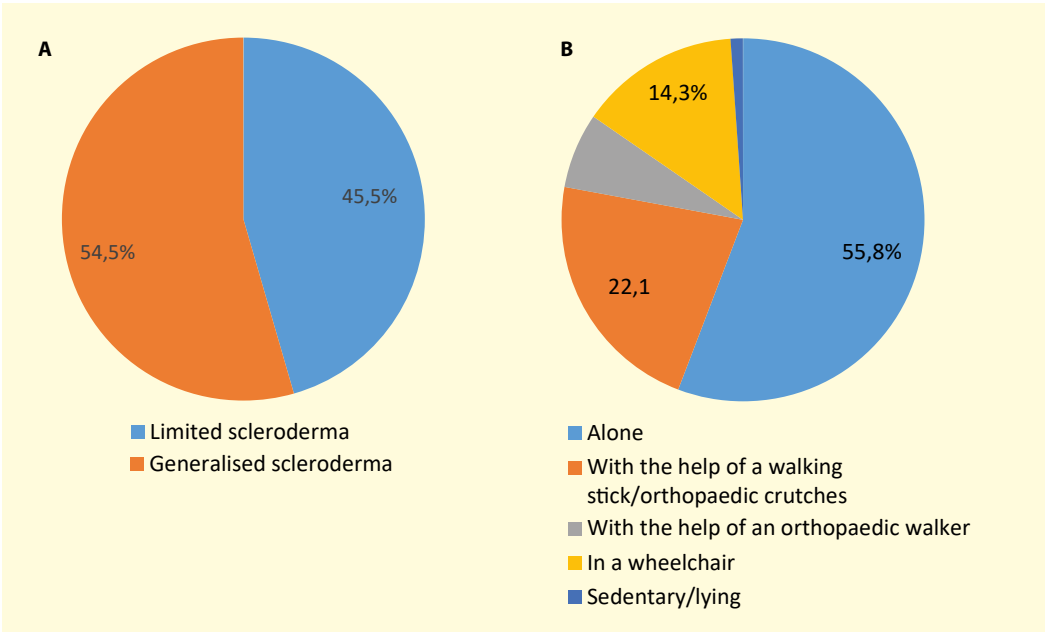

Figure 3. Characteristics of the type of sclerosis and the need for orthopaedic supplies of the respondents: **A.** Forms of systemic sclerosis among respondents; **B.** The way respondents move

of the disease on strengthening their family bonds (Fig. 4C). Given social interactions, 35 respondents perceived a negative impact of the disease, which they described as frequent. Moreover, 6.5% of the participants stated that the disease had a continuous impact on their social interactions. Rare impact was identified by 28 respondents, while 9 respondents stated that the disease had no impact on their social interactions.

The respondents specified in which ADLs they need assistance. The need for assistance in meal preparation was reported by 76.7% of the participants. A significant number of respondents, 46.8%, declared their ability to independently prepare and take medication. However, among them, 2.6% stated that they constantly require assistance, while 7.8% reported needing assistance in this activity all the time or frequently. Household chores are another ADL. Among the surveyed individuals, 13 respondents reported needing permanent assistance in household chores, while a significant percentage — 48.1% of the patients — stated that they require frequent help in this regard. Buying groceries is an activity during which assistance was required all the time or frequently by 54.6% of the respondents; 22 individuals rarely used this form of assistance. In terms of doing shopping, 15.6% of the respondents reported not needing any assistance. Moving around the home is another activity that can be difficult for SSc patients. Permanent assistance is required by 26% of respondents, frequent use of assistance is declared by 5.3%. Assistance is used rarely or not at all by 92% of respondents.

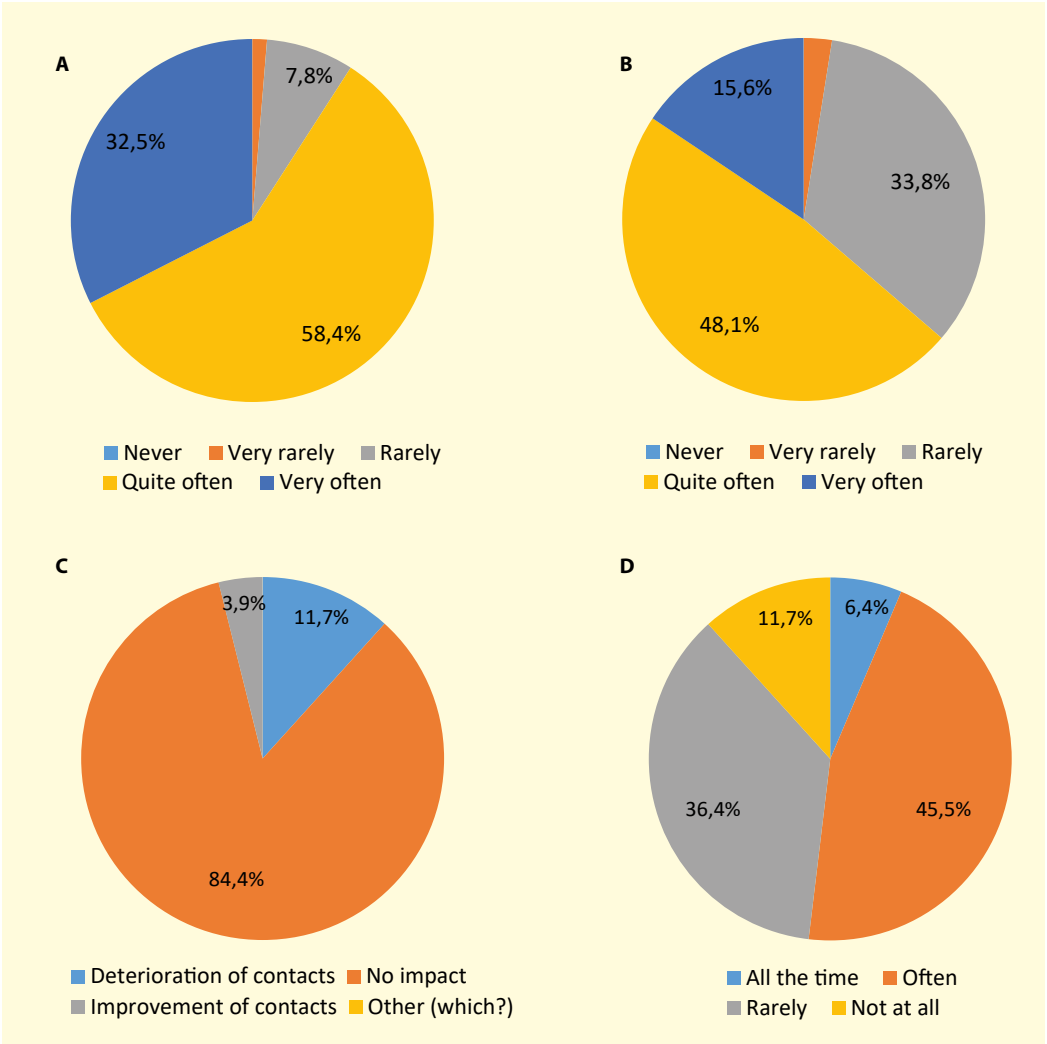

Figure 4. Emotional wellbeing of patients with systemic sclerosis: **A.** Frequency of perceived symptoms among respondents; **B.** How often do the experienced symptoms disrupt work among respondents?; **C.** Impact of the disease on family interactions of respondents; **D.** The impact of physical wellbeing and emotional wellbeing on social interactions of the respondents

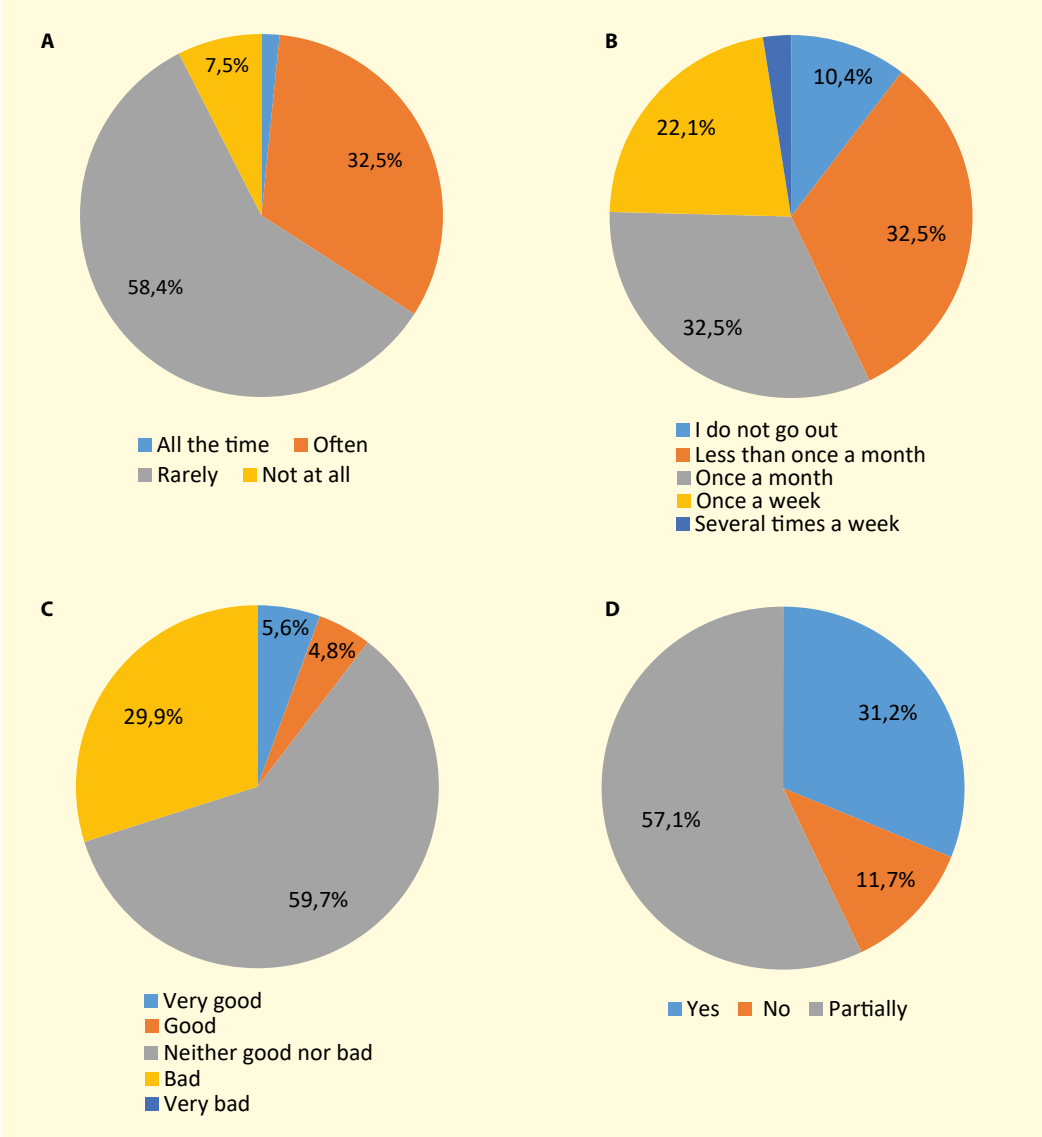

Figure 5. Social activity of respondents: **A.** Perception of loneliness among respondents; **B.** Frequency of leaving home among respondents; **C.** Assessment of own health status among respondents; **D.** Perception of life fulfilment among respondents

Moving outside the home can pose greater difficulties, as indicated by the results where 8 respondents require permanent assistance in this regard. Moreover, 20.8% of the participants described their need for such assistance as frequent. Maintaining personal hygiene is an important activity that affects the wellbeing of the body and the wellbeing of the patient.

Permanent assistance with hygiene is required by 3.9% of respondents, while 11 respondents stated that they need frequent assistance. A higher percentage of respondents, 45.5%, indicate that they do not currently require assistance in maintaining personal hygiene, while 37.7% of respondents reported rarely relying on assistance. Putting on and taking off clothes is a seemingly simple activity for healthy people. Among SSc patients, 2.6% of the respondents require permanent assistance, while 9 respondents frequently rely on assistance. Patients strive to remain independent in basic ADL for as long as possible, as reflected in the survey results. According to the survey, 41.6% of the respondents rarely rely on assistance when dressing up, while 44.2% do not require any assistance in this regard.

Using the toilet is one of the very intimate activities identified in this questionnaire, the results show that patients want to remain independent in this activity for as long as possible. Therefore, 71.4% of respondents do not require assistance, 19.5% rarely use assistance. Frequent assistance, or permanent assistance, is declared by 6 respondents. All the results are given in Table 2.

Table 2. Frequency of use of assistance in activities of daily living

	Meal preparation	Preparation and administration of medication	Doing the housework	Doing shopping	Moving around the flat	Moving outside the residence	Personal hygiene	Dressing up	Use of the toilet
All the time	5.2%	2.6%	16.9%	18.2%	2.6%	10.4%	3.9%	2.6%	2.6%
Often	27.3%	7.8%	48.1%	36.4%	5.2%	20.8%	14.3%	11.7%	5.2%
Rarely	49.4%	42.9%	28.6%	28.6%	40.1%	28.6%	37.7%	41.6%	19.5%
Not at all	18.2%	46.8%	6.5%	15.6%	51.9%	40.1%	45.5%	44.2%	71.4%

Questions about feelings of loneliness, frequency of leaving home, assessment of one's own health status and feelings of life fulfilment among respondents were also important points in the survey. Systemic sclerosis can affect all these areas of life. Loneliness and depression are rarely or not at all felt by 66.2% of respondents. Unfortunately, a sense of loneliness and depression often or constantly occurs in 33.8% of the respondents (Fig. 5A). Following that, the frequency of leisure outings is also varied among the respondents. There were 10.4% of respondents who did not leave their homes for recreational purposes. Moreover, 32.3% of the respondents reported going out for recreational purposes less than once a month. More than once a week, 2 individuals among the respondents engage in recreational activities (Fig. 5B). The self-assessment of health status revealed that unfortunately no one rated their health as very good. Their health status was rated as very bad or bad by 36.4% of respondents. A significant majority, 59.7%, rated their health as neither good nor bad, while only three respondents rated their health as good (Fig. 5C). The respondents were also asked about their feelings of fulfilment in life. Partial fulfilment was felt by 57.1% of respondents. Furthermore, 31.2% of the respondents reported feeling fulfilled in life. Lack of life fulfilment among respondents is felt by 9 respondents (Fig. 5D).

DISCUSSION

Research shows that SSc is a disease that affects patients' ADL to varying degrees. It penetrates both the patient's physical sphere, gradually limiting his or her independence, but also his or her mental sphere.

The majority of respondents were women. The questionnaire was completed by 55 women and 22 men. Systemic sclerosis is diagnosed more frequently among women than men. These results are consistent with epidemiological data indicating a 3–8 times higher susceptibility to illness among women compared to men [3–5] (Fig 1A). The age at which SSc is most commonly diagnosed is between 30 and 50 years of age [2, 6]. This fact was confirmed by the respondents' answers to the question about the age at which the disease was diagnosed in them. Among 46.8% of the respondents, 36 individuals, systemic sclerosis was diagnosed between the ages of 36 and 45. In 35.1% of respondents, 27 people, the diagnosis was made between the ages of 46 and 55. Systemic sclerosis is a chronic disease, and this is confirmed by the results of this survey as well. In the survey as many as 32.9% of the respondents (25 individuals) reported having the disease for more than 6 years. The length of patient survival depends on the form of the disease and its dynamics. The prognosis for systemic sclerosis is generally serious. It is estimated that 55% to 80% of patients survive ten years [7].

A larger portion of the respondents in this survey, 54.5% (42 individuals), have been diagnosed with dSSc . Almost half, 45.5%, of respondents struggle with lSSc. These data are in contrast to literature data indicating a higher prevalence of lSSc [8].

When analysing these discrepancies, it is worth noting that the surveyed group were patients of a unit specialising in the diagnosis and treatment of systemic sclerosis, which may have resulted in a "negative selection of patients" by accepting more severe and advanced forms of the disease. This fact significantly influenced the prevalence of different forms of the disease in the group described.

Systemic sclerosis is a chronic and progressive disease. Patients' awareness of the sequelae of systemic sclerosis and their current health status have an impact on their depressed mood. These data are in line with the results of this study, where as many as one third of pa-

tients feel lonely and depressed. In this study, physical well-being and emotional well-being were shown to have a significant effect on social interactions. Patients with systemic sclerosis are significantly more susceptible to depressive disorders, according to research. The percentage of patients with depression among those with rheumatological conditions is higher than the percentage of individuals with depression in the general population [9]. These observations align with the existing literature, which indicates a significant reduction in the quality of life among individuals with SSc.

Thirty-five individuals, 45.5% of respondents, state that their current health status often affects their interactions with friends. The disease has caused them to limit their contact with the external environment, and most often they spend their time in a familial environment. For the respondents, the disease has an impact on social relationships and social life. These observations are in perfect agreement with the results of a large Italian study showing a significant reduction in the social life of SSc patients [10]. The largest number of respondents also rely on family support. A total of 65 individuals responded that they use the help of their wife/husband, parents or children.

Assistance is necessary given the complications associated with systemic sclerosis, which gradually limit the patient's independence in performing ADLs. The complications experienced by SSc patients are related to the form they have been diagnosed with. Each of these complications also affects the quality of life of SSc patients [11]. Despite the discomfort experienced, 55.8% of respondents (43 individuals) move independently. However, as shown in the literature, individuals with systemic sclerosis significantly less frequently engage in physical recreational activities, which further contributes to a sense of reduced quality of life [12].

In this study, it was found that 37.7% of the respondents are employed, including 26% (20 individuals) working full-time. These data are consistent with the findings of a study from Sweden, which indicated a similar frequency of full professional activity among SSc patients. These data are in line with the results of a study from Sweden indicating a similar frequency of full professional activity of SSc patients [13]. Restrictions on the ability to perform gainful employment and social roles, which are also related to the mismatch between jobs and the needs of the employee. Despite their disability, patients strive for independence and self-sufficiency by taking up employment (Fig. 4B). The impact and significance of occupational work have been raised in an article discussing nursing care for patients with functional impairments. This study highlighted the positive impact of occupational work on reducing feelings of isolation and improving the quality of life for individuals across different age groups [14].

However, the factors limiting the ability to perform gainful employment still remain the axial symptoms of SSc, such as damage to the respiratory system, musculoskeletal system and weakness.

There are also symptoms that cause discomfort and affect the emotional and physical well-being of the patient; however, these symptoms do not limit physical activity. The majority of the respondents, despite being diagnosed with systemic sclerosis, try to maintain a positive outlook on life and appreciate their current health status, which may deteriorate at varying rates as the disease progresses. When asked about their health, the majority of respondents described their health as "neither good nor bad". The majority of respondents also feel fulfilled in life and such results can be seen as a positive. Despite the difficulties and negative assessments related to their own health and life, the research revealed that these respondents are a minority of individuals.

CONCLUSIONS

The survey results allow the following conclusions to be drawn:

1. Patients diagnosed with systemic sclerosis do not feel the impact of the disease on family interactions.
2. A chronic disease such as systemic sclerosis negatively affects the wellbeing of patients. Patients gradually limit social interactions and leisure outings.
3. Patients most often benefit from assistance during work-related activities that require the use of muscular force and activation of the musculoskeletal system. Most often they need help with various household chores and grocery shopping.
4. Patients wish to remain independent for as long as possible despite their deteriorating health and physical limitations. This is particularly true in the area of self-care and self-grooming.
5. The occupational status of the respondents also shows that patients try their best not to

give up their ADLs for as long as possible. Therefore, a large proportion of respondents continue to take up professional work. Certainly, the type of work performed and the progression of the disease do not always allow patients to continue working in their profession, so a significant portion is currently on disability pension.

6. Systemic sclerosis is a chronic disease. Its course and the symptoms that patients experience have a huge impact on their ADLs. The more advanced the stage of the disease, the more the patient's ADLs decrease.

ARTICLE INFORMATIONS AND DECLARATIONS

DATA AVAILABILITY STATEMENT

Not applicable.

ETHICS STATEMENT

Not applicable.

AUTHOR CONTRIBUTIONS

Concept: KP, BBP, KN, MB, MK, PK, EM; Data collection: KP, BBP, KN;

Data analysis: MB, MK, EM; Text creation: KP, PK.

FUNDING

Not applicable.

ACKNOWLEDGMENTS

Not applicable.

CONFLICT OF INTEREST

Not applicable.

SUPPLEMENTARY MATERIAL

Not applicable.

References

1. Pączek L, Mucha K, Froncewicz B. Choroby wewnętrzne. Podręcznik dla studentów pielęgniarstwa i położnictwa. PZWL, Warszawa 2014: 531–535.
2. Olesińska M. Reumatologia w gabinecie lekarza Podstawowej Opieki Zdrowotnej. PZWL, Warszawa 2019: 369–382.
3. Bromirska M, Smoleńska Ż, Zdrojewski Z. Etiologia i czynniki ryzyka rozwoju twardziny układowej-przegląd literatury. Forum Reumatol. 2018; 4(1): 52–56.
4. Zimmermann-Górska I. Reumatologia kliniczna. PZWL, Warszawa 2008: 603–623.
5. Snaith M. ABC reumatologii. PZWL, Warszawa 2007: 122–127, 141–150.
6. Sierakowski S, Sierakowska M, Zimmermann-Górska I. Postępy reumatologii klinicznej. PZWL, Warszawa 2014: 192–207. Lis-Święty A, Brzezińska-Wcisło L. Twardzina układowa – czynniki prognostyczne, aktywność i ciężkość choroby. Przegląd Dermatol. 2010; 97: 398–405.
7. Lis-Święty A, Brzezińska-Wcisło L. Twardzina układowa – czynniki prognostyczne, aktywność i ciężkość choroby. Przegląd Dermatol. 2010; 97: 398–405.
8. Peoples C, Medsger Jr TA, Lucas M, et al. Gender differences in systemic sclerosis: relationship to clinical features, serologic status and outcomes. J Scleroderma Relat Disord. 2016; 1(2): 177–240, doi: 10.5301/jsrd.5000209, indexed in Pubmed: 29242839.
9. Buras A, Waszkiewicz N, Szulc A. Depresja i proces zapalny w chorobach reumatologicznych. Postepy Hig Med Dosw. 2015; 69: 162–168.
10. Galetti I, di Nunzio S, Brogelli L, et al. How do systemic sclerosis manifestations influence patients' lives? Results from a survey on patients and caregivers. Curr Med Res Opin. 2021; 37(suppl 2): 5–15, doi: 10.1080/03007995.2021.1992371, indexed in Pubmed: 34726112.
11. Kowal-Bielecka O. Powikłania naczyniowe twardziny układowej. Reumatologia. 2012; 5(1): 1–8.
12. Liem SIE, Meessen JM, Wolterbeek R, et al. Physical activity in patients with systemic sclerosis. Rheumatol Int. 2018; 38(3): 443–453, doi: 10.1007/s00296-017-3879-y, indexed in Pubmed: 29151128.
13. Sandqvist G, Scheja A, Hesselstrand R. Pain, fatigue and hand function closely correlated to work ability and employment status in systemic sclerosis. Rheumatology (Oxford). 2010; 49(9): 1739–1746, doi: 10.1093/rheumatology/keq145, indexed in Pubmed: 20511345.
14. Haor B, Szambelan J, Pietrzak M, et al. Opieka pielęgniarska nad pacjentem z deficytami sprawności w czynnościach dnia codziennego w przebiegu twardziny układowej. Zeszyty Naukowe. Zbliżenia Cywilizacyjne. 2019; 15(2): 38–51, doi: 10.21784/zc.2019.010.