Nursing problems of patients with systemic sclerosis

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Abstract

Systemic sclerosis (SSc) is a chronic autoimmune disease connective tissue and one of the most common collagen diseases. There are several clinical types of scleroderma which differ in their course, possible complications and prognosis. The most characteristic form SSc is limited and diffuse systemic sclerosis. The SSc is characterized by the progressive fibrosis of the skin and internal organs, leading to their failure, morphology and blood vessels disorders.

Purpose: The aim of our work is to identify the main health problems of patients suffering from systemic sclerosis depending on its clinical form: limited systemic sclerosis (lSSc) and diffuse systemic sclerosis (dSSc); to determine the influence of disease duration on symptom intensification in patients with lSSc and dSSc.

Material and methods: The study group consisted of 63 patients with systemic sclerosis diagnosed according to the criteria of the American Rheumatism Association (ARA), 47 of whom had limited systemic sclerosis (lSSc) (74.6%) and 16 – diffuse systemic sclerosis (dSSc) (25.4%).

Conclusions: The key thing in the complex therapy is to recognize the individual care problems of the patient, to assess his ability to cope with the disease in daily life and to plan care, support, education and help of other professionals. The main aim of individual nursing care is to alleviate ailments, prevent infections, observe life-threatening conditions and to educate the patient as regards self-care and self-observation.

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Introduction

Systemic sclerosis (SSc) is a systemic connective tissue disease, characterised by a progressive fibrosis of the skin and internal organs leading to their failure, changes of organs morphology and their blood vessels as well as the immune system dysfunction [1].

The SSc most often affects people aged from 30 to 50. Women develop scleroderma 3-4 times more often than men.

Limited systemic sclerosis – lSSc, previously known as CREST syndrome, which is an acronym for its clinical symptoms: calcinosis – calcification in soft tissues, Raynaud's Phenomenon, which usually significantly (a few years or longer) precedes the disease, esophageal dysfunction, sclerodactylia – hardening of finger skin and teleangiectasia – the occurrence of blood-vessel lesions on the skin. Skin lesions affect the face and upper limbs – distal to elbows, and lower limbs – distal to knees. It is a milder form of SSc and particularly chronic. At first, it was believed that in this form of SSc the systemic lesions are limited only to the oesophagus, but it has now been proved that there are also lesions in other organs [2].

Diffuse systemic sclerosis dSSc was previously known as progressive systemic sclerosis. Symmetrical, widespread and quickly progressing skin lesion affect not only the face and limbs, sometimes except for fingers, but also the corpse. The hardening of limb skin involves the areas proximal to elbows and knees. Other characteristic features dSSc include fatigue, arthralgia, polyarthritis with swelling and itching fingers and toes and frequent joint contractures. The Reynaud's Phenomenon usually appears simultaneously with skin thickening. Diffuse systemic sclerosis is characterised by a greater than lSSc dynamics of the involvement of skin and internal organs: most often concern lungs, less frequently – kidneys and heart, very

Table 1. **Data of examined groups**

Age	**to 40 of years**		**41-60 of years**		**≥61 of years**	
	N	%	N	%	N	%
	12	19%	35	55.6%	16	25.4%
Education	**no more than the elementary education**		**at least the average education (college)**			
	29	46%	34		54%	
Civil status	**married**		**not married**			
	46	73%	17		27%	
Place of residence	**town**		**village**			
	38	60.3%	25		39,7%	
Duration of disease	**to 4 of years**		**5-14 of years**		**≥ 15 of years**	
	17	27%	29	46%	17	27%
Profesional activity	**working**		**not working (pensioner)**			
	23	36.5%	40		63.5%	

often – the alimentary tract. The onset of disease may be quite rapid, especially in the first stage (the first 3 years). The most dramatic complications dSSc include renal involvement in the form of scleroderma renal crisis – 80% within the first 4 years. As a result, dSSc is much more severe than lSSc [2,3].

A patient with systemic sclerosis requires permanent medical care, specialised diagnostics, monitoring of basic life parameters for life-threatening systemic complications, a systematic treatment and nursing. The patient and his family should be involved in the educational processes aiming at their preparation to non-professional care [4].

The objective of this study is to identify the main health problems of patients suffering from systemic sclerosis, with a differentiation between the limited and diffuse form, as well as to determine the influence of disease duration on symptom intensification in patients with lSSc and dSSc.

Material and methods

The study group consisted of 63 patients with systemic sclerosis diagnosed according to the criteria of the American Rheumatism Association (ARA), 47 of whom had limited systemic sclerosis (lSSc) (74.6%) and 16 – diffuse systemic sclerosis (dSSc) (25.4%). There were 60 women and 3 men. The basic research tool was a survey questionnaire drafted for the needs of this study. The analized data were presented in the statistical package 6.0, being parameters like percent, average, standard deviation for the studied parameters.The results were verified with the test t-student and χ^2.

Results

The majority of our group were patients aged 41-60 (55.6% of all subjects), with at least secondary education (54%) and married (73%). Over 60% of subjects came from towns. The patients who had been ill for 5-14 years constituted 46% of all our subjects. There were 36.5 % professionally active and 63.5 % inactive persons. Among the professionally inactive, 28 were pensioners (44.4%) and 12 were retired (19%) (*Tab. 1*).

It was observed that in the group with a longer disease duration, there were more professionally inactive persons. In the population with disease duration ≥15 years all the subjects were pensioners ($p \leq 0.05$).

Health problems of patients with systemic sclerosis

The Reynaud's Phenomenon was present in almost all subjects (93.7%), both in the case of limited systemic sclerosis (lSSc) and the diffuse form (dSSc), irrespective of disease duration.

The vast majority (61.3% of total) of patients with both forms of SSc reported *dysaesthesia in fingers,* which depends on disease duration. *Ulceration of fingers and toes* affected 53.2% of subjects; 48.9% of subjects of the lSSc group , and 66.7% of the dSSc group.

In over 65% subjects the *hardening of skin* was observed. Morning stiffness occurred in 43.5% of all patients, with no significant differences between the two types. It was noticed that morning stiffness occurs much more often in patients with shorter disease duration of 0-4 years (in the dSSc group – 100% of subjects, in the lSSc group – 72.7%), than in patients with longer duration of the disease.

Over 55% of patients reported difficulties with performing everyday tasks and claimed to need help. The longer the patient suffered from systemic sclerosis, the bigger self-care problems they had, especially in the case of limited type of SSc ($p \leq 0.05$).

The inability to cope with the disease had tendency to increase ($p=0.08$) in group of patients with lSSc (52.5%), especially in the first stage of the disease (72.7% in the group with 0-4 years of disease duration), in relation to the subjects with dSSc, where only 26.7% of patients reported the above mentioned inability.

The subjects, irrespective of disease type, declare that they do not smoke (82.3%), and a vast majority of dSSc patients do not drink alcohol (86.7%), the remaining 13.3% drink occasionally. More patients 53.2 % with lSSc declare that they do not drink alcohol at all and 46.6% that they do it occasionally.

Symptoms involving the alimentary tract

Alimentary tract problems affected much more the patients with limited systemic sclerosis than dSSc. The main problem

***Table 2*. Alimentary tract problems**

Alimentary tract problems	limited systemic sclerosis (lSSc)		diffuse systemic sclerosis (dSSc)		in all	
Appetite	**N**	**%**	**N**	**%**	**N**	**%**
lack of the appetite	18	38.3%	4	25.0%	22	34.9%
no appetite problem	29	61.7%	12	75.0%	41	65.1%
Swallow						
difficulties in the swallow	32	68.1%	6	(37.5%)	38	60.3%
no swallow problem	15	31.9%	10	62.5%	25	39.7%
Significance	$p \leq 0.05$					
Heartburn						
no problem	11	23.4%	6	37.5%	17	27.0%
feels the heartburn	36	76.6%	10	62.5%	46	73.0%
Diarrhoeas/constipations						
problems with emptying	26	55.3%	5	31.3%	31	49.2%
no problem	21	44.7%	11	68.7%	32	50.8%

***Table 3*. Health problems involving the respiratory system**

Problems on the respiratory system	limited systemic sclerosis (lSSc)		diffuse systemic sclerosis (dSSc)		in all	
Effort dyspnoea	**N**	**%**	**N**	**%**	**N**	**%**
does not appear	19	40.4%	6	37.5%	25	39.7%
feels the difficult breathing	28	59.6%	10	62.5%	38	60.3%
Cough						
heaps of times coughs	8	17.0%	2	12.5%	10	15.9%
seldom coughs	39	83.0%	14	87.5%	53	84.1%

***Table 4*. Blood pressure in patients with systemic sclerosis**

Blood pressure	limited systemic sclerosis (lSSc)				diffuse systemic sclerosis (dSSc)				in all
	Duration of disease				Duration of disease				
	in all	to 4 of years	5-14 of years	≥ 15 of years	in all	to 4 of years	5-14 of years	≥ 15 of years	
increased blood pressure	14 29.8%	3 27.3%	6 27.3%	5 35.7%	6 37.5%	1 20.0%	3 42.9%	2 50.0%	20 31.8%
regular blood pressure	33 70.2%	8 72.7%	16 72.7%	9 64.3%	10 62.5%	4 80.0%	4 57.1%	2 50.0%	43 68.2%

of all subjects were difficulties with swallowing (dysphagia), with statistically significant differences between the groups with lSSc and dSSc ($p \leq 0.05$). In the lSSc group with disease duration of 5-14 years, these ailments were particularly intensive (81.8%). Patients with lSSc also more often in comparison to dSSc reported the feeling of heartburn and problems with defecation (*Tab. 2*).

Symptoms involving the respiratory system

No statistically significant differences have been observed between lSSc and dSSc as regards the intensity of symptoms involving the respiratory system. More than half (60.3 %) of the persons with SSc studied suffer from effort dyspnoea (*Tab. 3*), which is more frequent in patients with 5-14 years of disease duration.

Over 80% of subjects, especially in the diffuse SSc (93.3%), reported quick fatigability and the feeling of tiredness, irrespective of disease duration.

Increased *blood pressure* was reported by 32.3% of subjects, more by dSSc patients (40% of subjects, with an increase related to disease duration) than those with the limited type (29.8% of subjects) (*Tab. 4*).

Pain is one of the dominating problems of patients with systemic sclerosis. Over a half of subjects with both types of SSc (67.7%) reported a very frequent feeling of pain, especially in the first stage of the disease (0-4 years). The sensation of pain affected most of all the joints of arms and legs (96.8%) (*Fig. 1*).

Psychological problems. 42.9% of patients (48.9% of subjects with the limited type and 25.0% of patients with the diffuse type reported bouts of low spirits, irrespective of disease duration. The vast majority of subjects in both groups (71.4%) reported a negative influence of their physical and mental health on social activity (in the lSSc group – 76.6%; dSSc – 56.2%). Disease duration has a significant influence on lowering social activity of patients with diffuse systemic sclerosis ($p \leq 0.05$).

Figure 1. **The location of pain in patients with systemic sclerosis**

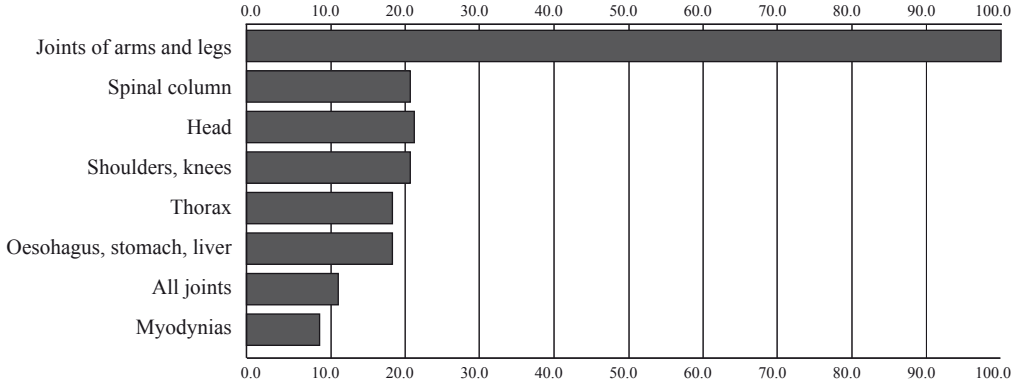

Table 5. **Psychological problems in patients with systemic sclerosis**

Psychological problems	limited systemic sclerosis (lSSc)		diffuse systemic sclerosis (dSSc)		in all	
Spirits	**N**	**%**	**N**	**%**	**N**	**%**
decreased spirits	23	48.9%	4	25.0%	27	42.9%
no problem	24	51.1%	12	75.0%	33	57.1%
Avoid people						
yes	27	57.4%	6	37.5%	33	53.2%
no	20	42.6%	10	62.5%	30	47.6%

Social activity	limited systemic sclerosis (lSSc)				diffuse systemic sclerosis (dSSc)				in all
	Duration of disease				Duration of disease				
	in all	to 4 of years	5-14 of years	≥ 15 of years	in all	to 4 of years	5-14 of years	≥ 15 of years	
problems with social activity	36 76.6%	7 63.6%	18 81.8%	11 78.6%	9 56.2%	1 16.7%	5 71.4%	3 100.0%	45 71.4%
no problem	11 23.4%	4 36.4%	4 18.2%	3 21.4%	7 43.8%	5 83.3%	2 28.6%	0 0%	18 28.6%
Significance	$p \leq 0.05$								

Over a half of subjects, especially those with lSSc (57.4%), declare that they avoid people (*Tab. 5*).

Patients believe that the biggest problems connected with their disease are chronic and progressive character (35% of all subjects), pain (31%), general weakness (27%) and difficulties in moving (26%). Patients also reported problems of psychological nature, bouts of low spirits, anxiety, fear (14%).

Discussion

Rheumatic diseases are one of the major health problems of the contemporary society due to the consequences arising from the dysfunction of numerous organs and systems. Systemic sclerosis is one of the diseases which causes numerous biological as well as psychological and social problems [5].

Out of 5 clinical types of SSc, the most frequent ones are the limited type (lSSc) and the diffuse type (dSSc) [2].

Both in the limited and diffuse form of SSc skin lesions undergo 3 stages: swelling, hardening, disappearance. At first, patients have difficulty to bend their fingers due to swelling, which poses the risk of dermatorrhexis. Later on appear partial contractures of fingers due to skin hardening, which makes bending and straightening of fingers difficult. Patients (especially those with limited SSc) often complain on pain connected with easily inflicted skin injuries and ulcers which are difficult to heal [6].

Reported 53.2% of patients finger and toe ulceration. A vast majority of patients (61.3% in total) with both types of SSc reported dysaesthesia in fingers, which is related to disease duration.

The Reynaud's Phenomenon is defined as episodic, usually bilateral, paroxysmal contractions of vessels in distal body parts and, in some cases, also the vessels of internal organs, as a reaction to inductive factors, which most often include cold, stress or medication. Patients complain on the sensation of cold in finger area, which is sometimes accompanied by pain, especially in winter [2]. The Reynaud's Phenomenon occurred in almost of all our subjects (93.7%), with both types of systemic sclerosis, irrespective of disease duration.

Joint symptoms of SSc typically accompany lesions in the skin and periarticular subcutaneous tissue. Our patients

complain on jointache of changing location, which is usually symmetrical; morning stiffness of fingers, wrists, elbows and knees, as well as temporary swelling [1,2]. Morning stiffness was observed in 43.5% of all subjects, without significant differences between the two types. It has been noticed that morning stiffness occurs much more often in patients with disease duration of 0-4 years.

Over a half of subjects with both types of SSc (67.7%) reported a very frequent feeling of pain, especially in the first stage of the disease (0-4 years). The sensation of pain affected most of all the joints of arms and legs (96.8%), and, to a smaller extent, joints of the spine, knees and head (22.2% of subjects).

The longer the patient suffered from systemic sclerosis, the bigger self-care problems he had, especially in the case of the limited type of SSc ($p \leq 0.05$). The inability to cope with the disease was more often reported by patients with limited SSc (52.5%), especially in the first 4 years.

The feeling of tiredness is a general symptom of SSc, where in 1/3 of patients with the diffuse form (dSSc) in its early stage (<3 years) and the advanced stage of the limited form (lSSc) (>10 years) the occurrence of tiredness is characteristic [2].

Quick fatigability and the feeling of tiredness were particularly frequent in the group with diffuse SSc (93.3%), irrespective of disease duration.

Numerous studies have shown that SSc patients present dysfunctions of internal organs, especially lungs, heart, alimentary tract and kidneys [6,7].

Problems involving the alimentary tract include difficulties with opening the mouth, pain during swallowing of food and a swallowing disorder (dysphagia), ill-being connected with the feeling of heartburn and belching, patient's discomfort due to problems with defecation (diarrhoea/constipation) [6].

Problems involving the alimentary tract, diagnosed in self-examination, affected more the patients with limited systemic sclerosis. The main problem of subjects were the difficulties with swallowing. In the lSSc group with disease duration of 5-14 years, these ailments were particularly intense (81.8%). Patients with limited SSc also suffered more from heartburn and defecation problems, which intensified together with disease duration.

Respiratory problems include the difficulties with breathing (dyspnoea, first during effort and in advanced stages also at rest, weakness, quick fatigability), patient's discomfort caused by a chronic, dry, non-productive cough, a risk of pulmonary arterial hypertension (a later stage of the limited SSc) [2].

We did not observed statistically significant differences between both clinical forms of SSc as regards the intensity of symptoms involving the respiratory system. Subjects, especially in the diffuse SSc (93.3%), reported quick fatigability, irrespective of disease duration and effort dyspnoea, which occurred in both studied clinical types of SSc (59.7%) and is more frequent in patients with 5-14 years of disease duration.

Scleroderma renal crisis develops in about 80% of dSSc patients within the first 4 years of SSc. It is manifested by quickly increasing arterial hypertension, sometimes accompanied by strong headache, vision disorders, convulsions, acute left ventricular failure and the symptoms of renal failure. Some patients with TPN have normal blood pressure and the crisis is manifested by the symptoms of quickly progressing acute renal failure [1].

Increased *blood pressure* was reported by 31.8% of subjects, more by dSSc patients (37.5% of subjects, with an increase related to disease duration) than those with the limited type (29.8% of subjects). It was reported that the awareness of the consequences of systemic sclerosis causes a worse psychic functioning of the patient, who goes through periods of resignation, bouts of low spirits, fear and depression. Especially women may suffer because of the changes in their facial appearance, which is so characteristic for systemic sclerosis and make them similar to other persons suffering from this disease [5,6,8,9].

Irrespective of disease duration 42.9% of patients, especially those with the limited form of SSc, reported bouts of low spirits. A vast majority of subjects in both groups (71.4%) reported a negative influence of their physical and mental health on social activity. Disease duration has a significant influence on lowering social activity of patients with diffuse systemic sclerosis ($p \leq 0.05$). Over a half of subjects, especially those with lSSc (57.4%), declare that they avoid people.

The biggest problem identified by patients is the chronic and progressive character of systemic sclerosis, the feeling of pain, general weakness and difficulties in moving. Patients also reported problems of psychological nature such as bouts of low spirits, anxiety and fear.

Conclusions

1. Skin hardening, ulceration, pain and morning stiffness of joints cause difficulties in self-care, which intensify with disease duration.
2. Patients in the first stage of limited systemic sclerosis presented an inability to cope with the disease.
3. An important problem of patients with diffuse systemic sclerosis are quick fatigability and the feeling of tiredness.
4. Problems involving the alimentary tract affect most of all patients with the limited type, in particular those with disease duration of 5-14 years.
5. Effort dyspnoea may indicate on lung lesions connected with systemic sclerosis.
6. The chronic and progressive character of systemic sclerosis has a great influence on the psychic conditions of patients as well as their social and professional activity, which decreases together with disease duration.

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