The effect of a self-management program on the quality of life of patients with scleroderma

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Abstract:

BACKGROUND: Scleroderma is a rare disease with complex disorders. It affects the quality of life with severe impacts on the skin and extensive complications in the internal organs, and does not have a definitive treatment. This study aimed to investigate the effect of a self-management program on the quality of life of patients with scleroderma.

MATERIALS AND METHODS: This was a clinical trial in which 54 patients with scleroderma were randomly divided into two groups of 27 each (experimental and control groups). The data were collected using the Systemic Sclerosis Questionnaire. A self-management program was sent to the experimental group via a mobile phone application (WhatsApp) every day for three months. Statistical analysis was performed in Statistical Package for the Social Sciences V21.

RESULTS: The Wilcoxon signed-rank test showed that the average overall quality of life score of the experimental group showed a significant increase after the implementation of the program (*P* value: 0.00). The average overall quality of life score of the control group also significantly declined after the intervention (*P* value: 0.00). The Mann-Whitney U test revealed that there was no significant difference in the overall quality of life score of the two groups before the intervention (*P* value: 0.31); however, after the implementation of the self-management program, a significant difference was observed between the two groups (*P* value: 0.00).

CONCLUSION: According to the results, the self-management program can help improve the quality of life of patients with scleroderma.

Keywords:

Quality of life, self-management, systemic scleroderma

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Introduction

Scleroderma is a rare disease[1] with complex disorders.[2] This disease, with an autoimmune etiology, is a rheumatic disease with no definitive treatment and is associated with complications and a high mortality rate.[3] By affecting the connective tissue and microcirculation, it causes fibrosis of the skin and internal organs.[4]

The local type is manifested with severe alterations in the skin of the hands and face, and the common symptoms of the systemic type are Raynaud's disease, ulcers, dry joints, reflux, pain, fatigue, loss of appetite, and weight loss.[1,5] This disease is usually associated with heart, lung, kidney, and sexual problems. Treatments can have side effects such as premature menopause and infertility.[6]

There are involvements specific to scleroderma in other areas besides the physical domain, which are often neglected.[7] These include stress that causes anxiety disorders, distress, reduced self-esteem, a sense of dependence on others, and embarrassment.[4,8,9]

In addition, doubt and uncertainty in the form of ambiguity and delays in diagnosis, unknown prognosis,[10] lack of a definite treatment, and self-treatment measures[11,12] may impact the quality of life of these patients.[13]

Although such problems in these patients are still not fully known, they generally challenge their mental health and reduce their quality of life.[14]

Recent studies indicate that compared to other rheumatic diseases, scleroderma has a greater impact on the average quality of life, and patients with scleroderma experience a poorer quality of life.[13]

Meanwhile, there is no known effective treatment for modulating the scleroderma process. Care and treatment measures are carried out with the main goal of reducing symptoms and disability and improving the quality of life.[6]

Therefore, patient-centered care, which includes strategies to improve self-esteem, resilience, and self-efficacy, may help improve satisfaction with treatment and the health and quality of life of patients with scleroderma.[8]

However, due to the lack of specific guidance and training, these patients face an information challenge and, consequently, various problems adapting to the disease. They try to improve their management to achieve a better quality of life. A proper self-management program can make it easier for these patients to cope with the disease and boosts their quality of life.[4]

Self-management, as a structured and developing program, empowers patients to carry out activities that help maintain and improve their health and manage the symptoms of the disease; such programs emphasize the management of the effects of the disease on patients' functioning, emotions, and communication.[15,16] They enable people to consciously choose a new perspective and a general skill when problems arise to improve their management and leadership with multiple strategies and appropriate functioning.[17]

Moreover, self-management components include education, physical activity, exercise, pain control, fatigue management, adjustment of risk factors, dyspnea management, ergonomics, relaxation methods, energy conservation, joint protection, and the use of assistive equipment.[17] Nutrition, medication, emotional or stress management, communication techniques, and cognitive-behavioral techniques have been mentioned as other components of self-management.[15,18]

In general, self-management strategies include interventions for behavior changes and coping principles, such as determining and setting goals, identifying obstacles, solving problems, attracting support, operational planning, self-regulation, and leading operations in different groups of diseases, which are applied as useful strategies.[17]

Therefore, it is expected that the training in self-management programs should help maintain independence, increase efficiency, promote responsibility and active participation in self-care, control the symptoms and complications of the disease, reduce dependence on others, and decrease the frequency of hospitalization and treatment costs.[19]

A literature review revealed that various interventions have different effects on the dimensions of quality of life in patients with scleroderma.

Jönsson *et al*.[20] (2018) showed that training and specialized exercises could help reduce pain, improve the quality of life, and promote self-efficacy.

Maltez *et al*.[21] (2020) stated that stem cell transplantation had a positive effect on the physical indicators affecting the quality of life of patients with scleroderma, but it did not affect their mental indicators.

In a study to manage and improve the indicators related to the quality of life in patients with scleroderma, Sierakowska (2019) considered it important to assess anxiety and disability and stated that identifying these factors helps design and develop non-pharmacological interventions with the help of teams with different specialties.[22]

Few studies have been conducted on scleroderma patients worldwide because its origin and treatment have not been determined yet. Only a few parts of the problems and symptoms of this disease have been examined, and despite the extent and depth of the problems faced by these patients in all aspects of life, they need to improve their quality of life and reduce challenges. Furthermore, most studies on self-management programs for patients with rheumatoid have focused on the general disease condition rather than focusing on systemic scleroderma. Therefore, to improve the quality of life of these patients through the management of their unique conditions, the present study aimed to determine the effect of a self-management program on the quality of life of patients with scleroderma.

Materials and Methods

Study design and setting
This was a clinical trial (IRCT id: IRCT20191021045178N4) with two groups (experimental and control) that were evaluated before and after the intervention.

The intervention relied on a self-management program designed based on the information obtained from qualitative research.

Study participants and sampling

The research population comprised all patients with scleroderma. The research setting was Hafez Hospital, affiliated with Shiraz University of Medical Sciences (Iran). After the project was approved, the permits were obtained from the Shiraz University of Medical Sciences, including the code of ethics, and were presented to the manager of Hafez Hospital. Using the census method for sampling, a list of all patients with scleroderma who had records in the rheumatology clinic and visited the rheumatology department and the clinic of Hafez Hospital in six months was prepared.

Patients with a definitive diagnosis of scleroderma who had been diagnosed a year ago, had sufficient experience with the details and problems of the disease, had the physical, mental, and cognitive ability to participate in the research, were able to speak Persian, and were willing to participate in the study were included. Patients would be excluded if they were unwilling to cooperate at any stage of the research or were unwell and unable to continue participating.

Blocked randomization list (Block size: 4) was used to allocate people into groups. The participants were divided into two groups, an experimental group, and a control group. Blocking was performed based on the random block generation software. The study was double-blind. The patients, the researcher who collected the data, and the statistician who analyzed the data were blinded to the groups.

The sample size was estimated using Gpower software with a type 1 error of 0.05 or 5%, power of 95%, and based on similar articles. The sample size of at least 22 people in each group was estimated. Considering a 20% risk of attrition, the final sample size was estimated to be about 27 people per group.

Measurements

The measurement instrument was the Systemic Sclerosis Questionnaire (SYSQ), designed by Ruof *et al.*,[1] which reports the functional impact of scleroderma and the general and systemic symptoms of patients with scleroderma. This questionnaire has 32 questions which are divided into four categories (general health, gastrointestinal, musculoskeletal, and cardiopulmonary symptoms), scored on a Likert scale (from 1 to 4), denoting the ability to perform an activity (1: "no problem," 4: "disabled"), the severity of symptoms (0: "no problem," 4: "very severe"), and the frequency of symptoms (1: "never," and 4: "always").

The validity of this questionnaire was examined and calculated with Cronbach's alpha. The questionnaire is reliable and repeatable and can be used to evaluate patients with scleroderma. Its internal consistency was reported between 0.73 and 0.93. The interobserver repeatability for all domains (0.786–0.983) and the intraobserver repeatability were very good. The agreement for the domain of general symptoms was very good (ICC = 0.916), and for the domain of musculoskeletal symptoms (ICC = 0.897) and cardiopulmonary domain (ICC = 0.842), and the range of gastrointestinal symptoms, it was reasonable (ICC = 0.686).[23]

Intervention

A self-management program was designed for patients with scleroderma using the information obtained about the patients' needs in the qualitative part of the dissertation. This self-management program was implemented for the experimental group for three months via mobile phones in an online group and was presented regularly and daily with the three main contents of spirituality, physical activities, and psychological skills. The self-management program was also implemented for the control group after the intervention group. During the intervention for the experimental group, the control group received routine treatment.

Procedure

For all the selected participants, the objectives of the study were explained, and informed consent was obtained from them; then, the questionnaire was administered to collect information before the intervention. Immediately after the intervention, the questionnaire was once again completed by the patients in both groups.

Ethical considerations

Ethical considerations were observed in this study:

All the participants were provided with the necessary information about the project. The participants signed the informed consent form before starting their collaboration.

The participants were assured that all their information would remain confidential, and if they wished, they would receive the results of the study. The principle of confidentiality was observed in the research, and data collection was performed confidentially and anonymously.

All the steps were carried out according to the Declaration of Helsinki.

This project was reviewed and approved by the Ethics Committee of the Shiraz University of Medical Sciences and received the code: (IR.SUMS.REC 1399.1211).

Data analysis
Data analysis was performed in SPSS.

The non-normality of the data was determined with the Kolmogorov-Smirnov test and the histogram. The difference in the quality of life of the two groups was evaluated with the Mann-Whitney U test before and after the intervention. Moreover, the difference in the quality of life before and after the intervention in each group was evaluated using the Wilcoxon test.

Results

Participants' characteristics and frequency
A total of 54 people were included in the study, 27 people in the control group and 27 in the experimental group. Most participants were women (*n* = 49, 90.7%), and five (9.3%) were men.

Their mean age was 47.68 ± 10.48 years. The patients were homemakers (64.8%), self-employed (13.00%), employees (11.1%), retired (7.4%), students (1.9%), or unemployed (1.9%). Their level of education ranged from elementary school (24.1%) to bachelor's degree (14.8%) and master's degree (1.9%), and the average duration of the disease was 11.99 ± 7.56 years.

Effects of the program
The average quality of life score in the control group was 8.61 ± 4.02 before the intervention, which increased to 9.89 ± 4.98 after the intervention; the Wilcoxon test showed a significant difference between before and after the intervention (*P* value: 0.00).

The average quality of life score in the experimental group was 7.36 ± 4.32 before the intervention, which was reduced to 5.23 ± 4.46 after the intervention; the Wilcoxon test showed a significant difference between these two values (*P* value: 0.00).

The average quality of life scores in the experimental and control groups did not significantly differ before the intervention; the Mann-Whitney U test showed no significant difference between the two groups before the intervention (*P* value: 0.31).

However, the mean score of quality of life in the experimental and control groups showed a significant difference after the intervention (*P* value: 0.00).

Table 1 shows the average overall quality of life scores before and after the intervention in both groups.

Effects of program on domains of SYSQ
The questionnaire included four domains: musculoskeletal, digestive, cardiorespiratory, and

Table 1: A comparison of the average overall quality of life scores before and after the intervention in both groups

Quality of life score/ groups	Before the intervention		After the intervention		Wilcoxon test
	SD	Mean	SD	Mean	*P*
Experimental	4.32	7.36	4.46	5.23	*P*: 0.00
Control	4.02	8.61	4.98	9.89	*P*: 0.00
P Mann-Whitney *U* test	*P*: 0.31		*P*: 0.00		

general health, each of which was evaluated before and after the intervention in both groups.

The difference between the mean musculoskeletal domain in the control group before the intervention (6.30 ± 5.09) and the mean musculoskeletal domain in the control group after the intervention (7.85 ± 6.53) was significant (*P* value: 0.03).

The difference between the mean musculoskeletal domain in the experimental group before the intervention (5.48 ± 5.84) and the mean musculoskeletal domain in the intervention group after the intervention (4.59 ± 5.68) was not significant (*P* value: 0.14).

The difference between the mean digestive domain in the control group before the intervention (7.50 ± 4.55) and the average digestive domain in the control group after the intervention (9.20 ± 5.71) was significant (*P* value: 0.00).

The difference between the mean digestive domain in the experimental group before the experimental (6.84 ± 4.37) and the mean digestive domain in the intervention group after the intervention (4.49 ± 4.74) was not significant (*P* value: 0.14).

The difference between the mean cardiorespiratory domain in the control group before the intervention (7.81 ± 5.47) and the mean cardiorespiratory area in the control group after the intervention (8.23 ± 5.31) was not significant (*P* value: 0.13).

The difference between the mean cardiorespiratory domain in the experimental group before the intervention (5.67 ± 4.48) and the mean cardiorespiratory domain in the experimental group after the intervention (3.67 ± 4.21) was significant (*P* value: 0.00).

The difference between the mean general health domain in the control group before the intervention (12.69 ± 4.75) and the mean general health domain in the control group after the intervention (14.58 ± 5.34) was not significant (*P* value: 0.13).

The difference between the mean general health domain in the experimental group before the intervention (11.81 ± 6.37) and the mean general

health domain in the experimental group after the intervention (7.50 ± 6.27) was significant (*P* value: 0.00).

Table 2 shows the difference in the mean scores of quality of life domains before and after the intervention in the two groups.

Discussion

The purpose of this research was to determine the effect of a self-management program on the quality of life of patients with scleroderma. In this study, we tried to use an instrument specific to these patients, which is more sensitive and accurate to clinical changes.[23]

The age of onset of the disease is 30–50 years; in this study, the average age of the patients was 47.68 ± 10.48 years. The highest prevalence of the disease is in women, and in this research, 90.7% of the patients were women. The maximum life span of affected patients is about 11 years, and in this research, the average duration of the disease was 11.99 ± 7.56 years.[1] The longer survival of patients can indicate the progress of medical care sciences and the improvement of patients' awareness to deal with their disease.

The quality of life score of the control group after the intervention was 9.89 ± 4.98, and the quality of life score of the experimental group after the intervention was 5.23 ± 4.46. The average quality of life score in the experimental group was 7.36 ± 4.32 before the intervention, and the average quality of life score in the control group was 8.61 ± 4.02 before the intervention.

The mean overall quality of life scores in the experimental and control groups did not significantly differ before the intervention; the Mann-Whitney U test did not show any significant difference between the two groups before the intervention (*P* value: 0.31).

Table 2: A comparison of the average domains of quality of life before and after the intervention in the two groups

Group/domains	Before the intervention	After the intervention	Significance level
Musculoskeletal			
Control	6.30±5.09	7.85±6.53	0.03
Experimental	5.48±5.84	4.59±5.68	0.14
Digestive			
Control	7.50±4.55	9.20±5.71	0.00
Experimental	6.84±4.37	4.49±4.74	0.14
Cardiorespiratory			
Control	7.81±5.47	8.23±5.31	0.13
Experimental	5.67±4.48	3.67±4.21	0.00
General Health			
Control	12.69±4.75	14.58±5.34	0.13
Experimental	11.81±6.37	7.50±6.27	0.00

Nevertheless, the average quality of life score of the experimental and control groups showed a significant difference after the intervention (*P* value: 0.00).

In fact, before the intervention, the patients in the two groups did not have a marked difference in terms of the overall quality of life score, but after the intervention, they showed a marked difference; after the intervention, the same test showed a significant difference between the two groups (*P* value: 0.00), which reveals the improvement of the quality of life and the effect of the self-management program on the experimental group compared to the control group after three months.

The findings of the study showed that the self-management program had a significant impact on the quality of life of the patients with scleroderma.

Furthermore, the self-management program has been effective in reducing cardiorespiratory symptoms and improving general health, but it did not have a significant effect on digestive and musculoskeletal symptoms; in other words, the patients in the experimental group improved compared to before the treatment in terms of cardiorespiratory (*P* value: 0.00)) and general health (*P* value: 0.00). The lack of effect of the self-management program on musculoskeletal (*P* value: 0.14) and digestive domain (*P* value: 0.14) demonstrates the importance of planning in the early days of the disease before the symptoms emerge and the problems set in.

On this, Doumen *et al*.[24] (2021) emphasized that self-management programs can create long-term self-efficacy in patients whose rheumatism is diagnosed early.

By showing the effect of fatigue on the quality of life of patients with scleroderma, Yakut *et al*.[25] (2020) suggested that managing fatigue by reducing respiratory problems and improving ventilation using various sports that can strengthen the respiratory muscles effectively improves the quality of life. Pettersson *et al*.[2] (2021) and Bongi *et al*.[3] (2009) showed that the management of scleroderma by rehabilitation treatments might be better therapeutic effects in the form of the prescribed action of a physiotherapist along with personalizing exercises.[26]

After three months, the average score of the quality of life in the control group decreased in all areas, and this decline was significant in the musculoskeletal (*P* value: 0.03) and digestive (*P* value: 0.00) domains. This reduction can be attributed to the progressive nature of scleroderma, which was greater for the musculoskeletal and digestive areas in these patients. This shows the necessity and importance of a self-management program to prevent and control the progress of complications.

Preis *et al.*[27] (2018) stated that malnutrition reduces the quality of life in patients with scleroderma, and to prevent this problem, standardized nutritional therapy should be followed.

Milette *et al.*[5] (2019) suggested that providing self-management programs as specific solutions to deal with the challenges of patients with scleroderma can empower them to better cope with and manage their disease and improve their quality of life.

Wojeck *et al.*[28] (2021) emphasized in their reviews that at least one self-management outcome, such as patient activation, self-efficacy, self-regulation, and global health comes after self-management intervention in a patient with systemic sclerosis.

To achieve a desirable and permanent quality of life in patients with scleroderma, self-management ability must be permanently and persistently established, requiring planned training and continuous evaluations. Shao *et al.*[29] (2021) note that the self-management program based on the theory of self-efficacy enhances self-management, self-efficacy, and physical performance, after six months of evaluation and support. They also emphasized the need for training and evaluation to change and improve lifestyles with longer duration in chronic diseases. As well as according to Bayati *et al.*[26] (2018) Enhancing health literacy and improving health behaviors significantly were affected by Educational interventions.

Limitations and Recommendations

Since scleroderma is a rare disease, it was difficult to find participants matching the inclusion criteria in most cities; thus, the researcher selected the sample by regularly visiting the rheumatology department and clinic.

The immunological nature of the disease, especially during the COVID-19 pandemic, limited the presence of patients in the treatment setting; thus, communication with the patients was made in compliance with all health protocols.

Conclusion

The results of this research showed that implementing a self-management program for patients with scleroderma led to positive results in terms of their quality of life. The program can be used in different medical and nursing departments. The results indicate to doctors and specialists that since medical treatment and pharmacotherapy alone cannot improve all aspects of patients' quality of life, by implementing a self-management program, in addition to the results of treatment, better outcomes can be achieved in terms of improving the quality of life of patients; with the improvement of quality of life, patients become more hopeful about treatment measures, and their treatment cessation rate will decrease.

For nurses in the clinical setting, especially in the rheumatology department, this program helps provide care and education to the patient. The program can also help with nursing education as a curriculum for students and community health during home visits. All this helps improve the independence and self-management ability of patients, prevent complications, and improve their quality of life.

The results of this study can be a basis for other studies in this field.
1. Providing a self-management program for the long term or forever
2. Teaching the use of smartphones to all patients
3. Evaluating patients over a longer period post-intervention.

Conflicts of interest
There are no conflicts of interest.

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