

RESEARCH ARTICLE

Fatigue and activity management education for individuals with systemic sclerosis: Adaptation and feasibility study of an intervention for a rare disease

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Funding information

Open access funding provided by IReL.

Abstract

Purpose: Fatigue is an overlooked symptom for musculoskeletal diseases, including rare conditions like systemic sclerosis (SSc). The purpose of this study were the following: (1) to adapt the content and delivery method of an existing fatigue intervention, and (2) to determine the feasibility of Fatigue and Activity Management Education in Systemic Sclerosis (FAME-iSS).

Methods: In Phase 1 adaptations were recorded using the Framework for Modification and Adaptations. In Phase 2, participants completed the 6-week FAME-iSS intervention and baseline, post-intervention, and 3-month follow-up assessments measuring fatigue levels, fatigue impact, mental health, self-efficacy, and use of energy conservation strategies. Qualitative data included a post-intervention focus group and 3-month follow-up interviews.

Results: Three main changes were made to the original intervention: (1) the content was adapted from management of Lupus-related fatigue to management of SSc-related fatigue, (2) context for delivery from Ireland to United States, and (3) in-person to online format. Participants ($n = 4$) were 51.8 ± 12.1 years old, had been diagnosed with SSc for 12.0 ± 8.0 years, were receiving disability support, and had college degrees. All participants had 100% attendance and completed all study activities. Participants had positive comments about the programme format, content, and implementation. Post-intervention, improvements were observed in most measures. Moderate effect sizes were noted in fatigue and self-efficacy scores.

Conclusion: Participants' positive responses supported the need for FAME-iSS for people with SSc. Even with the small sample, FAME-iSS led to improvements in fatigue and use of energy conservation strategies. The online format allowed for participation and sharing of ideas, especially during the global pandemic.

KEYWORDS

activity participation, fatigue, non-pharmacological intervention, occupational therapy, rheumatology, scleroderma, self-management, systemic sclerosis

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1 | INTRODUCTION

Fatigue is one of the most prevalent and disabling symptoms of systemic sclerosis (SSc or scleroderma), impacting all areas of life including activities of daily living, work, parenting, and social participation (e.g., Basta et al., 2018; Murphy et al., 2021). Recent literature has shown that various forms of non-pharmacological interventions are effective in reducing fatigue for persons with other rheumatic diseases (e.g., Hewlett et al., 2011; Katz et al., 2018). However, there are no fatigue-related programmes tailored to the rarity of SSc and its unique symptomology (i.e. skin thickening, vascular involvement, fibrosis of internal organs including the heart, lungs, kidneys and gastrointestinal tract; Denton & Khanna, 2017). This paper describes the process of adapting an existing fatigue management programme for individuals with Systemic Lupus Erythematosus (SLE) to the specific needs of people with SSc.

Although fatigue is a common symptom associated with various chronic conditions across many specialty areas, individuals continue to feel inadequately equipped to manage their fatigue and misunderstood by both their social supports and healthcare providers (Jaime-Lara et al., 2020). Fatigue is multi-faceted in both its causes and impact. For SSc, fatigue has been correlated with physical factors (e.g. pain, comorbidities, gastrointestinal symptoms, breathing problems, joint involvement), lifestyle factors (e.g. smoking, exercise), and psychosocial factors (e.g. illness behaviours and coping, depression, anxiety; Assassi et al., 2011; Azar et al., 2018; Del Rosso et al., 2013; Kwakkenbos et al., 2012; Strickland et al., 2012; Thombs et al., 2009). Studies in rheumatology show that fatigue impacts physical, cognitive, emotional, and social functioning (Connolly et al., 2014, 2019; Geenen & Dures, 2019; Murphy et al., 2021; Nicklin et al., 2010; Nikolaus et al., 2013). As such, rheumatic disease self-management programmes must take an integrated approach to fatigue management, allowing individuals to trial strategies from all areas and problem solve what works for them. However, many existing programmes only focus on one type of fatigue or one strategy that affects fatigue (e.g. increasing exercise to reduce physical fatigue). In addition, most programmes are delivered via a face-to-face format which may be impossible for individuals who have fatigue and mobility issues or a rare condition like SSc.

The Fatigue and Activity Management Education (FAME) programme was developed for individuals with SLE in Ireland by one of the authors (DC). In the original delivery of FAME, participants completed six 2.5-h in-person group sessions, led by occupational therapists, to learn about and discuss factors that increase fatigue for individuals with SLE (Table 1). In line with self-efficacy theory (Bandura, 1977), occupational therapists encourage participants to share their past experiences with fatigue management strategies, problem solve how to implement new strategies, and motivate one another to create and accomplish 'SMART' (specific, measurable, attainable, relevant, and time-based) goals each week. A pre-post study demonstrated statistically significant improvements in mood and quality of life, as well as positive trends in fatigue, activity participation, self-efficacy, and understanding of fatigue (O'Riordan et al., 2017).

Given the similar impact of fatigue on activity participation for individuals across rheumatic conditions, the three authors, all with research and clinical rheumatology experience, adapted the content of FAME to address nuances of living with SSc. The adapted programme, renamed FAME in Systemic Sclerosis (FAME-iSS), retained the weekly educational topics as per the original FAME but was adapted to reflect current research on factors that impact fatigue in SSc, and considerations for persons with rare diseases. Within the adaptation process, revisions were systematically documented and a feasibility study was conducted with a small group of individuals with SSc to answer the following questions:

1. Is the content of FAME adaptable and relevant for people with SSc?
2. Are the adaptations and online delivery of FAME-iSS acceptable to persons with SSc?
3. Is FAME-iSS potentially effective in managing and reducing the impact of fatigue?

2 | METHODS

Prior to this study, KC and JP conducted a literature review of fatigue and fatigue management in rheumatic conditions, as well as a needs assessment of young adults with rheumatic conditions who experienced high levels of fatigue (Carandang, Mehdiyeva & Poole, in preparation). FAME was identified as an intervention with promising effects that may be adaptable to people with SSc because of its focus on the multiple dimensions of fatigue and on strategies that promote activity participation. KC and JP contacted the FAME developer (DC) for more information, training on how to conduct the intervention, and a request to adapt for individuals with SSc. The international team collaborated on a two-phase process to develop FAME-iSS as described below.

2.1 | Phase One

The research team held meetings one to two times/month (schedule permitting) over a period of 7 months to discuss the core elements of FAME and iteratively edit the content of the slide deck and handbook. Each author was assigned two of the six weekly topics to complete the first draft of revisions using tracked changes and send to the other research team members for review. At each meeting, each line of the revised chapters and all slides of the six sessions were reviewed to collaboratively make final determinations of changes. Changes were documented using the Framework for Modification and Adaptations (FRAME; Stirman et al., 2019) and are reported in the results.

2.2 | Phase Two

A feasibility study was conducted with a small group of individuals with SSc to obtain their feedback about the content and delivery of

TABLE 1 Topics and content discussed at Each Fatigue and Activity Management Education session

	Topic	Content
Weeks 2–6 (interchangeable)	Week 1: Introduction	<ul style="list-style-type: none"> • Overview of programme • Definition of self-management • Relationship between myriad of factors that affect condition-related fatigue • Fundamentals of goal setting
	Energy management	<ul style="list-style-type: none"> • Energy conservation principles • Translating energy conservation principles into action within daily activities • Sleep hygiene strategies • Using a fatigue diary • Set weekly goal
	Physical activity	<ul style="list-style-type: none"> • Recommended exercises for individuals with disease • Benefits of exercise • Factors to support adherence to exercise and physical activity • Review last week's goal and set new goal
	Pain management	<ul style="list-style-type: none"> • Pain mechanisms in disease • Relationship between pain and fatigue in disease • Joint protection techniques, including body positioning • Set and review weekly goals
	Stress management	<ul style="list-style-type: none"> • Physiological responses to stress • Lifestyle factors that impact stress • Stress management strategies • Trial relaxation strategies • Set and review weekly goals
	Nutrition	<ul style="list-style-type: none"> • Relationship between diet, health, and energy/fatigue • Recommendations for healthy diet and/or dietary modifications • Strategies for managing diet and eating patterns • Set and review weekly goals (<i>If last session, discuss how to maintain progress and continue setting goals</i>)

FAME-iSS, explore the acceptability of the adapted programme, and determine the appropriateness of the programme evaluation process (e.g. outcome measures) for use in a larger clinical trial.

2.2.1 | Participants

Participants were recruited from local divisions and support groups of the Scleroderma Foundation in US Mountain and Pacific time zones (selected for ease in finding a common meeting time for delivery of FAME-iSS). Participant inclusion criteria were the following: (1) United States residents; (2) Physician diagnosis of SSc based on self-report; (3) Age 18+ years; (4) Access to an electronic device with videoconferencing capabilities; (5) Ability to communicate in English; (6) Be willing to attend all virtual sessions over the course of 6 weeks and (7) In order to provide informed feedback regarding the

relevance and potential benefit of the content of FAME-iSS for those with SSc, individuals with SSc who self-reported the presence of fatigue were required to participate in the study. The presence of fatigue was indicated by a score of four or above (total of ≥ 12) on the first three questions on the Multi-Dimensional Assessment of Fatigue (MAF; Belza, 1995); degree of fatigue (1- not at all to 10- a great deal), severity of fatigue (1-mild to 10- severe), and level of distress from fatigue (1—no distress to 10—a great deal of distress).

2.2.2 | Procedures

All participants completed a demographic questionnaire and a set of patient-reported outcome measures (PROM; described below) and selected their availability in terms of times and days for the 90-min weekly meetings. Study data were collected and managed using

REDCap electronic data capture tools hosted at the University of New Mexico (Harris et al., 2009). Once the programme day and time were agreed, all participants were sent an electronic and paper copy of the FAME-iSS handbook. At the end of the sixth session, study participants were invited to participate in a focus group discussing their opinion of the content and relevance of FAME-iSS, ideas for dissemination and implementation, and areas for improvement. Participants also completed another set of PROM. Those who completed all study activities to this point received a USD \$25 gift card. Three months after programme completion, participants completed a final set of PROM and an individual follow-up phone interview and received another USD \$35 gift card.

2.2.3 | Data collection

Participants completed the following PROM at baseline:

The Multidimensional Assessment of Fatigue (MAF; Belza, 1995) is a 16-item scale that has been used across many chronic diseases populations to measure four subjective domains of fatigue: degree and severity, amount of distress it causes, timing, and degree to which fatigue interferes with the activities of daily living. A global fatigue score is calculated with a total score ranging from 0 (no fatigue) to 50 (severe fatigue). A clinically meaningful score of 11.5 for the MAF was identified by Goligher et al., (2008).

The Modified Fatigue Impact Scale (MFIS; Fisk et al., 1994; Strober et al., 2020) is a 21-item self-report scale that was originally developed for persons with multiple sclerosis to evaluate the impact of fatigue on three dimensions: physical, cognitive, and psychosocial functioning. Total scores for the MFIS range from 0 to 84 with higher numbers indicating greater fatigue (Fisk et al., 1994).

The Hospital Anxiety and Depression Scale (HADS; Zigmond & Snaith, 1983) is a 14-item self-report measure with two subscales of depression and anxiety. Each item is rated on a four-point scale (0 = not at all, 3 = yes definitely). Scores from each subscale are summed to produce a total anxiety score and total depression score. Total scores for each subscale are categorized as 0–7 (normal levels of anxiety/depression), 8–10 (borderline anxiety/depression) and 11–21 (clinical anxiety/depression; Zigmond & Snaith, 1983). Although the HADS has been used with a wide range of patient groups, a clinically meaningful score is reported only for people with Chronic Obstructive Pulmonary Disease in the HADS-Anxiety scale. A change of 1.5 is considered clinically meaningful for this population (Puhan et al., 2008).

The PROMIS Self-Efficacy for Managing Symptoms (Gruber-Baldini et al., 2017) is an eight-item measure which examines participants' confidence in managing symptoms so as not to interfere with activities they perform in daily life. Scores range from 8 to 40 with higher scores indicating greater self-efficacy.

The Self-Efficacy for Performing Energy Conservation Strategies Assessment (SEPESCA; Liepold & Mathiowetz, 2005) is a 14-item assessment, originally developed for persons with multiple

sclerosis, that measures participants' confidence in using energy conservation strategies. Scores range from 14 to 140 with higher scores indicating greater self-efficacy.

Participants also completed two questionnaires at post-intervention and at 3-month follow-up:

An 8-item short course evaluation was included to provide feedback on acceptability of FAME-iSS. Likert rating scales from 1 (strongly disagree) to 5 (strongly agree) were used to measure participants' satisfaction with content and delivery format of FAME-iSS.

The Energy Conservation Strategies Survey (ECSS; Matuska et al., 2007) asks participants if they implemented 14 different energy saving strategies, how often they used the strategies (once/week; 2–3/week; 4–6/week; daily or more), and their perceived effectiveness (1 = not effective to 10 = very effective) of each strategy to reduce fatigue. For strategies not used, participants are asked to provide a reason for why they had not implemented the strategy.

2.2.4 | Qualitative data collection

Participants were asked for their opinions and experiences of FAME-iSS through semi-structured interviews at two timepoints:

Post-intervention focus group: Participants discussed their experiences of FAME-iSS, including programme strengths and areas for improvement. The discussion was led by DC using a structured interview guide (Figure 1). This interview guide was the same as that used in the post-programme focus group and follow-up interviews in the original FAME study (O'Riordan et al., 2017). It was designed by the FAME study research team. The discussion was audio recorded, transcribed (KC), and de-identified for analysis. The focus group duration was 25 min.

Individual follow-up interviews: Three months post-intervention participants completed a telephone interview to discuss the perceived impact of FAME-iSS on fatigue management and subsequent impressions of programme content and delivery. Interviews were conducted by JP using the FAME follow-up interview guide (Figure 1), audio recorded, transcribed (KC), and de-identified for analysis. Interviews ranged from 15 to 25 min with a mean interview time of 19.5 min.

2.2.5 | Data analysis

Descriptive analysis explored participant demographics, clinical characteristics, baseline fatigue measures and recruitment and retention rates. To explore potential effectiveness of FAME-iSS, Wilcoxon Signed rank tests were performed and Cohen effect sizes were calculated in IBM SPSS Statistics v25. In accordance with the short and structured nature of both types of qualitative data collection, KC developed a bulleted list of recommendations and strengths identified by at least two participants. In addition, KC

Focus group schedule:

What was your overall impression of the content of FAME-iSS?

What was your overall impression of the online delivery format of FAME-iSS?

Which aspects of FAME-iSS did you find *most* useful and why?

Which aspects of FAME-iSS did you find *least* useful and why?

Has FAME-iSS helped you to understand your fatigue any better?

If so, in what way?

If not, why do you think that it has not helped?

Has FAME-iSS helped you to manage your fatigue better?

If so, in what way?

If not, why do you think that it did not help?

Have you any recommendations for improving FAME-iSS for people with Scleroderma?

Follow-up interview schedule:

How are you managing your fatigue now?

Are you using any specific fatigue management strategies from FAME-iSS to help you manage your fatigue?

If so, which strategies are you using and how useful are they for you?

If not, why do you think that you are not using any of the strategies?

Did attending FAME-iSS help to improve your energy to carry out your daily activities?

If so, in what way?

If not, why do you think that it did not help?

Since attending FAME-iSS are you participating in any additional and/or 'new' activities?

If so, what are these activities?

Are you using any resources from FAME-iSS: goal setting, the FAME-iSS handbook?

If so, when and how often do you use them?

Thinking back on FAME-iSS, have you any recommendations for improving the programme for future delivery?

FIGURE 1 Focus group and follow-up interview schedules. FAME-iSS, Fatigue and Activity Management Education in Systemic Sclerosis

identified data that pertained to participants' use of fatigue management and energy conservation strategies to contextualize quantitative data.

3 | RESULTS

Because the planned programme commencement dates coincided with the beginning of the COVID-19 pandemic, recruitment was delayed for approximately 6 months. Nine individuals contacted the research team with interest in participating in the study. Eight individuals (88.9%) met the eligibility criteria and ultimately four

individuals (44.4%) signed consent and completed the pre-intervention questionnaires. The four individuals who withdrew cited unexpected commitments: one moved to a different state, one had a conflicting surgery, and two reported being over-committed. On average, participants were 51.8 ± 12.1 years and diagnosed with SSc 12.0 ± 8.0 years. All participants had a college degree (two Associates; one Bachelors; one Postgraduate). All participants reported receiving a disability allowance, and three participants were married.

1. Is the programme content adaptable and relevant for people with SSc?

Utilizing FRAME, Table 2 reports the adaptations made to the original FAME programme. All adaptations were pro-actively decided upon during the pre-implementation/planning phase by the three authors. Due to similarities between experiences of fatigue and factors that increase and exacerbate fatigue, it was not necessary to make substantial changes to much of the session on fatigue

management (week 2). As all members of the research team are occupational therapists, for content discussed in week 3 (exercise) and week 6 (nutrition), we requested a review of the PowerPoint slides and relevant handbook chapters by a physical therapist and registered dietician respectively, to ensure that adaptations made to these sessions were not contra-indicated for individuals with SSc.

TABLE 2 Adaptations to FAME to develop FAME-iSS

What is the adaptation?	What is the goal of the adaptation?	What is the rationale of the adaptation?	What concerns about fidelity to the original model need to be monitored?
Context (setting) was modified from in-person delivery to online delivery via Zoom conferencing platform.	<ul style="list-style-type: none"> • Increase participants' access to programme. • Increase participants' retention and completion rate. 	<ul style="list-style-type: none"> • Due to the rarity of SSc, it is difficult to find enough patients to hold regular in-person meetings. Virtual meetings allow for connection without the time and expense of travel. • Fatigue and mobility issues may hinder patients' ability to attend in-person meetings regularly. • Virtual meetings may be an accessible platform for patients to engage and try strategies in their own homes. 	Because videoconferencing does not allow for unstructured conversations and breaks, time for rapport and relationship building between participants was less. Will this affect participants openness in sharing with one another?
Two other changes were made to accommodate the switch to online. (1) The context (format) was shortened from 2.5-h weekly sessions to 1.5-h weekly sessions, and (2) The content (packaging of materials) was changed: slide deck was condensed, and some content moved to the handbook to cover the same amount of material.	<ul style="list-style-type: none"> • Maintain participants' capacity (i.e. motivation, physical/cognitive capacity) to participate in the programme. • Increase participants' programme satisfaction. 	To accommodate for the shortened attention span on e-learning platforms.	<p>Time for all components (i.e. didactic learning, group discussion, and goal setting) were all shortened.</p> <p>Content that was previously delivered during the module was moved to the handbook reducing opportunities for peer discussion.</p>
Context (population) was changed from patients with systemic lupus erythematosus to patients with SSc. Content was refined to consider symptoms that are especially prevalent within SSc (e.g. respiratory involvement, gastrointestinal issues, Raynaud's phenomena, social and emotional effects of having a rare disease, exercise precautions for heart and lung involvement). National Health promotion recommendations (nutrition) were changed from Irish recommendations to United States recommendations	Increase reach of programme	Content changes addressed population-specific factors.	None identified. Because rheumatic conditions often have overlap, weekly topics were kept the same and only minor editing was required. In addition, the same format was used to talk about relationships between fatigue-related factors with time allotted for conversations between participants, which by nature vary based on individual disease experiences.
Context (population) was changed from Ireland to United States. To make this change, content was slightly tweaked to reflect vocabulary and spellings used in the United States.	Increase reach of programme	Content changes addressed cultural factors.	None identified. The research team discussed potential cultural differences to monitor during feasibility study.

Abbreviations: FAME, Fatigue and Activity Management Education; FAME-iSS, FAME in Systemic Sclerosis; SSc, systemic sclerosis.

2. Are the programme adaptations and online delivery acceptable to people with SSc?

Participants rated high levels of satisfaction (median score of 5/5) on all questions related to FAME-iSS content and delivery. These questions included presentation of information; personal comfort with the learning environment; usefulness/helpfulness of programme slides, handbook, and discussions; accessibility of the handbook; and helpfulness of the goal setting process. Attendance was 100% by all four consented participants for all six sessions indicating high acceptance and relevance for individuals with SSc.

In the post programme focus group and individual interviews, participants had positive feedback about the programme format, content, and implementation. A list of programme strengths and areas of improvement are provided in Table 3. Participants emphasized that their perspectives on the programme were greatly influenced by their status as “SSc veterans,” or individuals who had been diagnosed with SSc for several years. Overall, participants stated the programme was useful, particularly to revise information that they may have already heard before but were not actively applying for

example “Some of the information that you gave is important because we know things are going on, but we can't explain it. And then when you see, ‘Well, if you do these two things, that's why this happens.’ Instead of just thinking ‘Why did this happen?’”. However, some participants also stated they likely adopted fewer strategies because “people who have had [SSc] for a long time have already adjusted their behaviours; it's hard to change them.”

Participants discussed how expanding the group, especially to those with diverse experiences (e.g. individuals with newly diagnosed disease; children/caregiver dyads), may be beneficial to the overall dynamic of sharing: “if you had a few more people, maybe there would be more goals that you can get ideas about and try.” Participants varied in their recommendation of an ideal group size ranging between 6 and 15 people; important considerations included the number of people simultaneously visible on the videoconferencing platform as well as participants' personalities, which would ultimately affect comfort levels in honestly sharing experiences during discussions. Participants agreed that increasing the number of attendees may require a longer session duration because of lengthier discussions and/or more questions from individuals with a recent diagnosis of SSc. Finally,

TABLE 3 Programme strengths and areas of improvement identified by participants

Programme strengths	Quotes
Online format with a small group minimized need to travel while retaining ability to build relationships.	<i>I felt very comfortable, and it was easier for me rather than going out and trying to meet... I like that it's a smaller group because you don't feel so outcast. It's almost the perfect group size, 10 and under... I could see people's expressions and not feel like I'm ignoring somebody.</i>
Accompanying handbook allowed for preparation prior to group and tangible materials to take notes during group.	<i>As we've gone along, I've taken notes about what people have said and different ideas that I might not have written down. I like something in my hands to work with.</i> <i>I read the section that we were going to do before we met. So usually an hour before the class, I made sure I read it and had my thoughts down.</i>
Interactive discussions fostered sharing of experiences and new ideas.	<i>[Other groups] tend to speak at you or they show you videos and discuss topics but there's not a lot of personal interaction as far as other people's experiences, where I felt with this, there was and that was helpful.</i> <i>There's things I would have never considered i.e. boxing or air frying and I just think [that] in numbers and in groups, there's power and knowledge and knowledge and power.</i>
Use of visual models facilitated holistic understanding of how many areas interplay to impact fatigue.	<i>At the beginning when we were talking about COVID and how that all affected everything and it all kind of intermingled, I like how you were open to those different changes of adding that to the flowchart.</i>
Weekly recording and reporting content-related goals acted as motivation and accountability to take action during the week.	<i>If I told you I would do this, then I have to do it without lying about it or something. Just like with any workout I would do or any challenges you may do with your friends, you're like okay this is what we're going to do this week and next week we'll build up.</i>
Areas for programme improvement	Quotes
Goal setting worksheet should include all areas covered in programme (e.g. nutrition, sleep) to track simultaneous to main goals. Consider integrating a fatigue diary format.	<i>I like the idea of incorporating all of the goal setting to include what's your fatigue level, how would you rate your eating scale, and your sleep, and your stress level because it all does encompass what this study is about. So having that as part of the goal rather than just what are you going to do this week.</i>
Incorporate sections on communication, relationships, and finding support. This may be a topic by itself or spread throughout the current weeks.	<i>I might suggest adding a relationship or support system as part of the programme. Because that does factor into how you manage your fatigue. I don't know how to go about doing that, but I think that's adding the support in there or your relationships with your physicians.</i>

participants stated the section on nutrition was the least helpful but attributed this to experiencing unique gastrointestinal symptoms and questioned if others would feel the same.

3. Is the programme potentially effective in managing and reducing impact of fatigue?

All participants fully completed the questionnaires at baseline, post-intervention, and at the 3-month follow-up period. Table 4 shows there were non-significant improvements in all measures from baseline to post-intervention with moderate effect sizes noted in the MAF global score and the SEPSCA total scores. Three of the six measures indicated a decline in fatigue impact and depression, and improved self-efficacy for managing symptoms between post-intervention to the 3-month follow-up.

The ECSS reflects a range of energy conservation strategies recommended to participants during FAME-iSS. Directly following the programme, all 14 strategies were used by at least two participants with seven strategies being used by three participants (Table 5). Of those who reported not using strategies, the most frequent reason provided by participants was that they were already using the strategy prior to attending FAME-iSS. Utilization of strategies declined between post-intervention and the 3-month follow-up as evidenced by three of the strategies only being used by one participant. Effectiveness scores for energy conservation strategies post-intervention ranged from five to the maximum score of 10 and from two to nine at follow-up.

At the 3-month follow-up interviews, all participants noted that the energy conservation strategy 'pacing' was a significant addition to their regular routines whether used within 1 day: "I'm planning my day better. I know the first half of the day I'm better and by afternoon I go downhill. So, I'm trying to prioritize my day" or throughout the week "If I over-exercise, if I walk too far, if I do too much in one day, [the fatigue] is

definitely worse the next day. So, there's a balance that you have to find". Although not asked directly, participants also identified reasons that may contextualize why there was a decline of strategy use from post-intervention to the 3-month follow-up, including restrictions during the COVID-19 pandemic (concerns about being exposed outside as well as the ability to do activities safely inside the home) and significant disruptions in routines, for example, moving house, unanticipated surgeries, holidays.

4 | DISCUSSION

The purpose of this study was to adapt an existing fatigue management intervention for the needs of persons with SSc and examine the feasibility of the new programme FAME-iSS. Through an international collaboration, the research team made three significant contextual changes to the original FAME programme: (1) modified content from that designed for individuals with SLE to those with SSc; (2) changed culturally specific terminology used in Ireland to that of the United States; and (3) re-packaged the programme from an in-person format to a videoconferencing platform. In doing so, the core elements of the programme that were based on self-efficacy theory were retained, including a combination of didactic education, participant discussion, and experiential learning through goal setting and accountability. Following these adaptations, a small group of individuals with SSc participated in a feasibility study to share their perspectives and ensure the programme's acceptability in order to continue improving upon the programme content and design. Including the perspectives of various stakeholders within early stages of programme design and considering barriers to participation are two potential strategies that may increase the likelihood that an evidence-based programme is implemented successfully in the future (Khanna et al., 2019; Poole et al., 2019).

TABLE 4 Baseline, post-intervention, and 3-month follow-up median scores for patient-reported outcome measures

Measure	T1 ^a Median	T2 ^a Median	p-value between T1/T2	Cohen's <i>d</i> ^b	T3 ^a Median	p-value between T2/T3	Cohen's <i>d</i> ^b
Multi-dimensional Assessment of Fatigue Global Score ^c	40.9 (20–46)	20.7 (17.8–28.1)	0.14	0.51	16.5 (14.8–25.4)	0.68	0.65
Modified Fatigue Impact Scale Total ^c	62 (10–67)	38.5 (27–42)	0.27	0.38	47.5 (26–61)	0.72	0.13
Hospital Anxiety and Depression Scale -Anxiety ^c	5.5 (1–5.25)	4 (0–4.25)	0.41	0.28	4 (1–9)	0.56	0.20
Hospital Anxiety and Depression Scale -Depression ^c	7 (3–6.25)	4.5 (3–4.75)	0.28	0.38	6 (5–13)	0.18	0.47
Self-Efficacy for Performing Energy Conservation Strategies Assessment	8.3 (5.4–8.0)	9.2 (8.3–9.2)	0.12	0.56	9.6 (4.4–10)	1.0	0.10
PROMIS Self-Efficacy for Managing Symptoms	29.5 (24–30.8)	31.5 (29–33)	0.18	0.47	31 (17–37)	0.11	0.57

^aT1: baseline; T2: post-intervention; T3: 3-month follow-up.

^bCohen's *d* effect sizes: 0.2–0.50 = small to moderate; 0.51–0.80 = moderate to large; and >0.80 = large.

^cLower scores indicate improvement.

TABLE 5 Use and perceived effectiveness of energy conservation strategies

Energy conservation strategies	Number of participants at T2	Number of participants at T3	Median effectiveness score T2	Median effectiveness score T3
Changed work heights	2	3	7.5 (6-9)	8 (6-9)
Changed location of equipment, furniture, etc	2	2	8 (8-8)	7 (4-8)
Used energy-saving equipment	2	2	10 (10-10)	9 (8-10)
Changed body position during activities	3	3	7 (6-8)	8 (5-9)
Eliminated activities	2	3	8.5 (8-9)	7 (6-9)
Delegated activities	2	3	7 (6-8)	8 (6-10)
Requested assistance	3	3	8 (7-9)	9 (2-9)
Adjusted standards for completing activities	3	1	8 (8-10)	2
Adjusted priorities	3	2	10 (8-10)	5 (1-10)
Simplified activities	3	2	7 (6-9)	5 (1-10)
Planned day to balance rest and activity	3	1	7.5 (6-9)	6
Changed time of day for carrying out activities	2	2	9 (8-10)	8 (7-9)
Took rests during day	2	1	5 (3-7)	2
Took rests during activities	3	2	7 (5-8)	7 (6-8)

Overall, participants had positive comments about the format and content of FAME-iSS. In line with our goal to increase access to the programme, participants agreed that the virtual format of FAME-iSS allowed them to participate in a fatigue management intervention regardless of geographical proximity to hospital-based programmes which typically deliver self-management interventions in the United States. Geographical distance was a barrier in the original FAME study, where over half of participants who expressed interest did not attend the in-person programme (O'Riordan et al., 2017). In addition, because recruitment for FAME-iSS took place in the early stages of the COVID-19 pandemic, the virtual format allowed supported participants to think about ways to manage their disease despite this unexpected disruption to their lives. Individuals with rare diseases, and groups/organizations supporting these individuals, have leveraged online resources to disseminate information even before the pandemic (Khanna et al., 2019; McMullan et al., 2020; Vicari & Cappai, 2016). While there is a lot of discussion about how to effectively deliver telehealth care for individuals with rare diseases (Castro et al., 2021; Chowdhury et al., 2021), the disadvantages of virtual services (i.e. reduced access to diagnostic tests) do not appear relevant to the design or goals of the FAME-iSS programme. The virtual setting did not interfere with goal setting processes or group discussions as participants stated these were highlights of the programme. Goal setting proved to be crucial in the success of the programme as participants reported that being accountable to goals set during the programme helped to sustain achievements. Group discussions allowed for participants to share their ideas about fatigue management strategies, which were often then attempted by others

in the group, as well as to share successes towards reaching their goals. These comments suggest high acceptability of the educational component of each module, wherein trained occupational therapists provided professional expertise about mechanisms of fatigue and self-management strategies, and individuals with experiential expertise living everyday with SSc contributed their invaluable experience and knowledge of living with fatigue. Finally, participants reported that having access to a tangible printed format of the handbook in addition to the online programme was effective to aid in notetaking and preparing for, and reviewing content after, each session.

Challenges were experienced recruiting for the feasibility study during the COVID-19 pandemic; however, this timing makes it difficult to determine if our recruitment strategy was effective or needs to be adjusted in the future. Even with restrictions due to COVID-19, recruitment efforts were limited to the Western part of the United States to make it easier to schedule a compatible time to meet. These efforts yielded one virtual group comprised of four participants out of eight who were eligible. All participants had been living with SSc for many years and reported already knowing many of the topics and strategies discussed during FAME-iSS. Therefore, more changes may have been observed in another sample of participants living with SSc for a shorter duration. Interestingly, participants' primary recommendations were about ways to expand the reach of the programme to individuals who may find the programme most useful (e.g. newer diagnosed; and families). Future efforts will include recruiting nationally to yield a larger sample size with more diversity in disease duration and other characteristics, which will

allow for further analysis of the underlying mechanisms of the intervention.

While limited conclusions about effectiveness can be drawn from this small sample size, it is worth noting that PROM showed improvements across all outcomes (i.e. fatigue, fatigue impact, depression, anxiety and self-efficacy) immediately post-intervention, but declines were observed at 3-months post-intervention for fatigue impact, depression, and self-efficacy outcomes. Similarly, participants reported a high use of fatigue management strategies directly after participating in the programme but these behaviours decreased after 3 months. These results show that the selected assessments are likely to be responsive to changes in a large sample size as part of a clinical trial. Paired with qualitative data, they also suggest that these participants perceived immediate benefits from engagement in the programme but that we may need to refine the programme to encourage sustainment of the use of fatigue management strategies and these positive outcomes. Within follow-up interviews participants described how life events (e.g. surgeries, holidays, transitions) interrupted their routines and habits developed during the 6-week programme, and subsequently that their ability to integrate fatigue management strategies required more effort as they were no longer at the forefront of their minds.

There are several study limitations that should be addressed in future studies. First, our current study contained only the perspectives of this specific team of occupational therapy researchers (with consults from a physical therapist and registered dietician) and four individuals with SSc to adapt FAME. Our next steps are to obtain input from a wider group of healthcare professionals and follow a process to systematically document the necessary intervention components and mechanisms needed for other occupational therapy practitioners outside of our research team to facilitate FAME-ISS groups. Second, due to the small sample size of participants we are only able to draw minimal conclusions about the effectiveness of the FAME-ISS programme. Because this feasibility study was <https://doi.org/10.5014/ajot.2013.006270> intended to inform a larger pilot study (Tickle-Degnen, 2013), the information gathered is an important foundation for this line of research especially because there is very little precedent for developing fatigue management interventions for rare diseases.

5 | CONCLUSION

Our feasibility study showed that the content and virtual format of FAME-ISS was acceptable and received positively by people with SSc. Even with the small sample, FAME-ISS led to improvements in fatigue and increased use of strategies to manage fatigue in people with SSc. The videoconferencing format allowed for social participation and sharing of ideas between individuals with rare diseases who may not otherwise have been able to connect, especially during the global pandemic. Future studies will further examine the feasibility and effectiveness of FAME-ISS.

ACKNOWLEDGEMENTS

We thank the Scleroderma Foundation and chapters for assistance in recruiting and our first group of patient participants for being willing to join us and learn together.

Open access funding provided by IReL.

CONFLICT OF INTEREST

The authors do not have any conflicts of interest to disclose.

ETHICAL APPROVAL

This study was approved by the University of New Mexico Institutional Review Board (Study 19-038). All participants verbally reviewed the informed consent with a research team member and signed an electronic informed consent form. This study is registered with [ClinicalTrials.gov](https://clinicaltrials.gov) (NCT04401943).

AUTHOR CONTRIBUTIONS

Kristine Carandang: Conceptualisation; Methodology; Data collection and analysis; Writing - original draft; Writing - review & editing. **Janet Poole:** Conceptualisation; Methodology; Data Collection; Writing - review & editing. **Deirdre Connolly:** Conceptualisation; Methodology; Data collection and analysis; Writing - review & editing.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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How to cite this article: Carandang, K., Poole, J., & Connolly, D. (2022). Fatigue and activity management education for individuals with systemic sclerosis: Adaptation and feasibility study of an intervention for a rare disease. *Musculoskeletal Care*, 20(3), 593–604. <https://doi.org/10.1002/msc.1617>