SCLERODERMA PATIENTS' COMMITMENT TO ILLNESS MANAGEMENT: STRATEGIES AND LEARNING

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ABSTRACT

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The management of chronic diseases is described as the "health challenge of the 21st century" by the World Health Organization. Patients' active role in managing their illness is considered, by many, as central in addressing this challenge. This study explored and described, through scleroderma patients' own perceptions and understanding, their commitment to illness management, including how they were involved in dealing with their illness and how they learned to do so. The role of social interactions, in particular, support groups, in this process was also investigated.

Using a mixed-methods approach, 201 patients were surveyed, and 25 in-depth interviews were conducted. The quantitative results of this study indicated that 64% of patients were committed in managing their illness by being highly active in dealing with their illness. An increase in activation was associated with longer disease duration in the first decade of illness. Additionally, the patients with high social support were more active. The qualitative findings showed patients engaged with various types of work to mitigate the physical, emotional, psychological, relational, and financial impact of the illness. In doing so, patients employed four problem-solving strategies that they had learned by confronting problems in daily lives. These strategies were at the heart of their incidental and tacit learning of how to manage their illness.

Only 32% of patients participated in support groups. Support group participants showed higher activation and considered these groups as providing support, learning opportunities, and venues to help other patients.

This study indicates that patients' commitment to management of their illness, far from being a static characteristic of patients, is a spectrum where patients are engaged in a process of complex negotiation with multiple needs of their illness, in tandem with their illness trajectory. Illness uncertainty, learning, and strategies to solve problems in managing the illness frame patients' commitment and engagement. A preliminary model delineating these elements is provided.

DEDICATION

To scleroderma patients

Your resiliency was the inspiration for this work.

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INTRODUCTION

The World Health Organization (WHO, 2002) has called the management of chronic diseases the "health challenge of the 21st century" and predicted that chronic conditions will "be the leading cause of disability throughout the world by year 2020" (p. 6). Including patients in their care, WHO suggested, is one of the core elements of tackling this challenge:

> Because the management of chronic conditions requires lifestyle and daily behaviour change, emphasis must be upon the patient's central role and responsibility in health care. Focusing on the patient in this way constitutes an important shift in current clinical practice. At present, systems relegate the patient to the role of passive recipient of care, missing the opportunity to leverage what he or she can do to promote personal health. Health care for chronic conditions must be re-oriented around the patient and family. (p. 5)

Since this statement was put forth, a body of research on ways to encourage patients to have an active role in their own care has come to exist, notably for conditions like diabetes, asthma, and cardiovascular diseases. Despite these efforts, a fundamental understanding of this phenomenon of an active role, variably referred to as activation, engagement, participation, involvement, empowerment, self-care, self-management, among many other terms used, is lacking. This study investigated this phenomenon in patients with scleroderma, a rare autoimmune disease, with the purpose of inquiring into how it looked among these patients, what strategies patients used, and how they learned them, in order to add to the body of knowledge in this area and possible implications in health care practice and policies.

Context and Background

According to the National Health Council (2012), chronic diseases, including chronic non-communicable diseases (CNCDs), affect approximately 45% of adult Americans and some 8% of children under the age of 18. In the U.S. annually, seven out of ten deaths and 75% of health care costs are attributed to chronic conditions. Chronic diseases are identified as those that have long durations (over 3 months), are not cured by medications, and are not preventable by vaccines. However, technical definitions vary considerably based on considered characteristics such as "duration or latency, need for medical attention, effect on function, pathology, departure from well-being, noncontagious nature, multiplicity of risk factors, and non-amenability to cure" (Goodman, Posner, Huang, Parekh, & Koh, 2013, p. 2). Some of the most prevalent chronic conditions are disorders such as high blood pressure, Alzheimer's disease, cardiovascular disease (CVD), stroke, cancer, chronic respiratory diseases, diabetes, and arthritis (CDC, 2010). By the year 2020, the chronic disease contribution to mortality rate is "expected to rise to 73% of all deaths and 60% of global burden of disease" (WHO, 2013), making it the "dominant global public health issue soon" (Greenberg, Raymond, & Leeder, 2011, p. 1386).

The tremendous economic and human costs and the sheer worldwide magnitude make the problem of prevention, management, and cure of chronic diseases an urgent one. This problem is compounded by the realization that the prevalent health care systems are designed to cater to the needs of patients with acute illnesses, and conversely, are ill-equipped to address the needs of chronic disease patients the world over (Pan American Health Organization [PAHO], 2013). In acute and urgent care, "the emphasis is on diagnosis, ruling out serious disease, and curative or symptom-relieving treatments" (Wagner, Austin, & Von Korff, 1996, p. 513). These interventions constitute the totality of care needed for acute illnesses, which are characterized by being episodic and limited

in duration. For chronic diseases, the acute phase only marks the initial stage of a multi-phase disease trajectory (Corbin & Strauss, 1988). Furthermore, the disease presentation at its earliest stages may be complex and baffling: some patients contend with MUS (medically unexplained symptoms) or *contested illnesses* (Barker, 2011; Daker-White, Sanders, Greenfield, Ealing, & Payne, 2011) for years or a decade in a state of "diagnostic limbo" (Corbin & Strauss, 1988, p. 27). More importantly, once diagnosed, many patients encounter medical practices that are not designed to provide for their ongoing needs.

To date, the inability to meet these needs is deemed as the "single greatest challenge facing organized medical practice" (Wagner, n.d.), where "coverage gaps and poorly organized care" (Schoen, Osborn, How, Doty, & Peugh, 2009, p. w13) are viewed to be at the heart of the problem. Many experts concede that ecological and political changes at the policy and organizational levels are sorely needed to tackle the problem, but also acknowledge that the required multidisciplinary and boundary-crossing teamwork required "will necessarily take a lot of time, require cooperation among a variety of folks generally unaccustomed to working with one another, and require disparate stakeholders to develop new ways of doing business" (Vladeck, 2001, p. 175). The proposed organizational care models for the management of chronic diseases, such as the Chronic Care Model (Wagner et al., 2001), Innovative Care for Chronic Care framework (WHO, 2002), and the Expanded Chronic Care Model (Barr et al., 2003), depict the multiple stakeholders that ideally should be involved, and their roles and relationships within this complex enterprise. Engaging the patients in their care and having involved and activated patients (and caregivers) constitute important elements of these models.

At the higher policy level, the National Research Council's (2001) report recommends a restructuring of the healthcare system that is patient-centered by "providing care that is respectful of and responsive to individual patient preferences,

needs, and values and ensuring that patient values guide all clinical decisions" (p. 39). The Patient Protection and Affordable Care Act's (2010) allocation of $3.5 billion in funding for a new Patient-Centered Outcomes Research Institute expands the patient and caregiver involvements to biomedical research. Globally, the Expert Patient program in the U.K. (Department of Health, 2001), the German der mündige (mature) patient (Dieterich, 2007), and the Canadian and Australian policy directives (Forbat, Hubbard, & Kearney, 2009) represent similar initiatives and debates, experimenting with and exploring patient involvement within a number of divergent and varied healthcare systems.

These policy and organizational initiatives have accelerated a trend for a bottom-up, patient-centric approach for service delivery (Wiig, Storm, Aase, & Gjestsen, 2013). In essence, they legitimize patients as *active participants* in their own care, in medical and cultural contexts that have traditionally treated them based on a paternalistic model of the patient-physician relationship (Emanuel & Emanuel, 1992; Robinson &Thomson, 2001). A paternalistic model privileges the physician's knowledge and views and decontextualizes the disease from the illness experience by considering the medical treatment as an objective and scientific undertaking to alleviate the symptoms.

Such a view has taken hold, overtime, as the practice of medicine transitioned from *bedside-medicine* to *hospital-medicine* and then *laboratory-medicine* (Jewson, 1976). These transitions have shifted the sources of information for doctors and narrowed the roles that patients play in the diagnostic process and knowledge creation (Wilde, 2007). Under bedside-medicine, the experiential perception of sickness by the patient and observations of the extended family and neighbors might have been the basis of conceptualization of the illness; under hospital medicine, "the sick-man became a collection of synchronized organs, each with a specialized function" (Jewson, 1976, p. 229). With the advent of laboratory medicine and the utilization of pathology as the determinant of a disease state, the "patient was removed from the medical investigator's

field of saliency altogether … by a relocation of the fundamental realities of pathology in microscopical events beyond the tangible detection of patients and practitioners alike" (p. 237).

Nettleton (2004) argued that the anatomical science that was privileged in hospital medicine, along with the physiological science that was the basis of laboratory medicine, has started to give way to *informational medicine.* Propelled by information and systems theory sciences and advances in technology, the mechanistic view of body is replaced with a "somatic system," where disease is "a form of information malfunction or 'communication pathology'" (p. 668).

Patients are affected by these technological changes as well. Not only do they have access to unprecedented amounts of medical information that was only available to medical professionals just a few years earlier; they are also increasingly the producers of information in online communities and have become creators of what Fox and Ward (2006) call *health identities* ranging "from a relatively medicalized 'expert patient' to an independent consumer of health information" (p. 463).

These technological changes, including self-tracking devices, information access, and connectivity to peer patients, have converged with a number of other factors to initiate a shift in how the current medical care models are viewed. Among these factors are: the worldwide scale of the problem of chronic conditions, the proposed health policy initiatives, the paradigmatic shift in how diseases are conceptualized, a move away from paternalistic models, the financially and economically unsustainable health delivery models, redefinition of patients as consumers, and an aging population often not with one but multiple chronic conditions. These factors have gathered force and contributed to a movement toward more inclusive and collaborative models of care where *not* having the patients involved is *not* an option (Greenhalgh, 2009; Reiser, 1993; Say & Thomson, 2003; Swan, 2009).

Consequently, the real-world problem of understanding how patients should be involved or choose to have an active role increasingly is viewed as critical from research, practice, and policy perspectives. This begs the question: What does it mean for patients to play an active role?

What Do We Know about the Active Roles Patients Play?

A scoping search of literature was conducted using CINAHL, PsycINFO, Medline, and ERIC databases and the key words 'patient,' 'involvement,' 'engagement,' 'activation,' and 'participation' for journal articles between 2000 and 2016. The following six categories were identified as areas where the notion of patient active role and involvement is explored in the literature:

1. Biomedical and Health Research: patient involvement in research agenda setting, defining and assigning importance to treatment outcomes, review of effectiveness of research, lay membership in research ethics committees and scientific advisory process; participation in clinical, non-therapeutic, genomic, and translational research and trials; raising funds and doing medical charities.
2. Social, Policy and Practice: patient involvement in enhancement of evidence-based- practices, input into development of guidelines for clinical practice, setting priorities for healthcare improvement, assessing office consultation quality, and taking part in participatory action research.
3. Patient Safety: including patient involvement in medical error disclosure, diagnostic error mitigation, and safety interventions and initiatives.
4. Products and Service Development: patient involvement in health services design and quality improvement; health technology design and medical device development; development and evaluation of patient support tools, patient

reported outcome measures, information leaflets, and rehabilitation questionnaires.

5. Education: patient involvement as educators in healthcare education curricula and health professional education programs for medical students and nurses, as lay-peer educators, and as participants in educational intervention / prevention / rehabilitation programs.
6. Involvement in their own health through: health care consultations, treatment decision-making and goal-setting for care, engagement with health information, as participants in self-help groups, online social network and self-management programs, involvement in diagnostic process and health screening, self-monitoring and use of health technologies.

The focus of this research is limited to item number 6 on this list: involvement of patients in their own care. The literature concerned with this type of involvement makes a number of assertions about patients who play an active role in their own care:

- These patients have better disease outcomes (Stewart, 1995), show higher self-efficacy (Ritter, Lee, & Lorig, 2011), and express higher levels of satisfaction with their care (Ashraf, Colakoglu, & Nguyen, 2013; Glass et al., 2012).
- Patients' education, age, race, gender, ethnicity and cultural background, and the severity of the illness have an impact on the levels of involvement. These levels may change over time and with increased knowledge. The younger, more educated, White, and female patients tend to be more involved. While those with lower education and those with severe life-threatening conditions prefer to defer to doctors (Arora & McHorney, 2000; Carey et al., 2012; DeWalt, Boone, & Pignone, 2007; Say, Murtagh, & Thomson, 2006; Smith, Dixon, Trevena, Nutbeam, & McCaffery, 2009).
- Across multiple chronic conditions that have been studied, all patients showed a high desire for getting information about their conditions and treatment

options, while levels of preference for being involved in decision-making varied (Chewning et al., 2012; Tariman, Berry, Cochrane, Doorenbos, & Schepp, 2010).

- The scope of active involvement goes beyond what occurs in clinical encounters and includes patients' daily lives (Audulv, Asplund, & Norbergh, 2012; Kralik, Koch, Price, & Howard, 2004).

  Contextual factors such as empowerment interventions, clinical setting and the physician's communication style, patients' social networks, patients' psychological states and levels of depression, anxiety and distress exert an influence on the active role patients take on (Arora, Ayanian, & Guadagnoli, 2005; Eldh, Ekman, & Ehnfors, 2008; Entwistle, Prior, Skea, & Francis, 2008; Sepucha & Mulley, 2009; Street, Gordon, Ward, Krupat, & Kravitz, 2005; Wensing & Grol, 1998).

In spite of these understandings, the conceptualization and operationalization of the notion of active role and involvement in care have been fraught with difficulties. Forbat, Hubbard, and Kearney (2009), based on research done on cancer patients, concluded that "one of the greatest barriers to truly integrating patient involvement into the health service, policy and research is the conceptual muddle with which involvement is articulated, understood and actioned" (p. 2553). In addition to a lack of definitional clarity and consensus, many of these conceptualizations are not from the perspectives of the patients themselves (Barello, Graffigna, Vegni, & Bosio, 2014), but represent the professional and medical view of how patients should be active and involved.

The Rationale for This Study

First, the conceptual muddle noted by Forbat et al. (2009) and the perceived need for clarifying and describing patients' involvement with their own care warrant further

research in this area at a conceptual level. Second, there is a need to understand these concepts from the patients' point of view and through studies that solicit patients' experience and commitment to the management of illness. Third, implications for practice and policy, including insights to facilitate participation, to remove barriers to involvement, and to deliver more responsive and tailored care, may be obtained through such explorations. Referring to patient engagement, Mostashari, the National Coordinator for Health Information Technology, is reported as stating "if it were a drug, it would be a blockbuster" (Schneider, 2012). This highlights the extent to which the significance of patients' active role in their own care is underestimated at the practice and policy levels. Last, insights from such studies might be beneficial to patient groups in empowering their memberships to take on more active roles in their own care. As face-to-face and virtual social networks of patients are becoming more influential for obtaining information, getting provider referrals, and experimenting with alternative therapies, the practice implications expand beyond the traditional venues. Understanding patient involvement may expand our understanding of these non-traditional patient habitats in empowering patients.

Problem Statement

Despite a general agreement that patient involvement and activation are critical components of the types of care models needed for chronic conditions, our understanding and articulation of the concept of patients' active role and involvement, and the operationalization of it as a construct are limited, in particular, from the perspective of patients themselves. In that light, this inquiry seeks to understand, from patients' perspectives, in what ways their commitment to their illness management is perceived and is indicative of taking an active role and being involved, and how it may be related to learning.

Although in many studies, the role of learning in patient involvement is implied, it is rarely explicitly explored. In some work, learning is referred to in the context of formal educational programs. For example, Lorig (1993) defines patient self-management "as learning and practicing the skills necessary to carry on an active and emotionally satisfying life in the face of a chronic condition" (p. 11). Charmaz (2003) refers to an illness experience as a conduit for learning "new definitions of self" and relinquishing the "'old ones'" (p. 170). Corbin and Strauss (1988) note, "No one chooses an illness trajectory, but having been caught up in it, one must try to learn to live with it" (p. 75). Thorne, Paterson, and Russell (2003), regarding the expertise that many patients develop in illness decision-making, state,

> In short, we need to know what expertise in such decision-making entails beyond adherence to medical recommendations, understand how it is learned and sustained, and develop strategies for how we can best support it as it evolves over time and circumstance in the life trajectory. (p. 1338)

Learning in this research is the explicit focus; the lens of adult learning theories is utilized as a framework for this exploration.

Purpose of the Study

The purpose of this study is to explore and describe, through scleroderma patients' own perceptions and understanding, their commitment to illness management, including how they are involved in dealing with their illness and how they learned to do so. In this exploration, the significant areas of commitment in engaging with disease and living with the illness, and the patterns in involvement and possible meanings ascribed to them by patients are examined and described, and beliefs, feelings, attitude, strategies, and contextual factors that may have shaped these are scrutinized through a number of theoretical lenses. Such an examination in scleroderma patients is suspected to contribute to outlines of a general model of involvement in one's care, applicable not only to this

group of patients, but also other chronic disease patients. The rationale for this argument is presented next.

Significance of Studying Scleroderma Patients

The rationale and significance of studying scleroderma patients are predicated on four characteristics of chronicity, autoimmunity, complexity, and the diversity of presentation in this disease. These are presented in this section, after a general introduction to scleroderma.

Scleroderma

Scleroderma, meaning hard skin, also known as systemic sclerosis (SSc), is an immune-mediated, connective tissue, inflammatory disease that involves fibrotic changes (scarring) in skin and/or the connective tissues of internal organs. An unknown trigger mechanism causes an inflammatory reaction by the immune system that is followed by fibrotic changes. Three processes in the pathology of the disease are observed: the fibroblast overproduction of extracellular matrix, vascular endothelial cell damage, and autoantibody productions due to immune response (Chung & Utz, 2004), although it has not been determined, to date, whether the autoimmune response is primary or secondary to the disease process.

There are great variations in manifestation of the disease in terms of organ involvement, and the severity and progression that ensue. The subtypes include localized, limited cutaneous scleroderma (lcSSc), diffuse cutaneous scleroderma (dcSSc), and scleroderma sine scleroderma (ssSSc). In the localized form, skin below elbows and knees is the only affected area. In its limited form, the build-up of collagen manifests outwardly as shiny, thick, and tight skin on face, trunk, hands, and feet. In this form, the disease is not fatal, but the skin deformity, joint and muscle pain, and resulting

disabilities greatly impact the quality of patients' lives. The systemic diffuse form of the disease progresses to one or more internal organs, such as lung, heart, kidney, and esophagus. Scleroderma sine lacks the skin manifestation, but similar to the diffuse form, impacts the internal organs. The end-stage organ failure is the cause of mortality in the progression of the systemic form of the disease.

The etiology and pathogenesis of scleroderma are poorly understood, and hence no treatment to counteract the primary cause(s) of the disease exists. Clinically, scleroderma is managed by therapies that address the secondary symptoms and organs' complications. For example, use of ACE inhibitors for kidney involvement and early treatment of pulmonary hypertension are attributed to considerable reduction of mortality in these subgroups of patients (Eckes, Hunzelmann, Moinzadeh, & Krieg, 2007). Largely experimental and with varying degrees of success, immunosuppressants such as chemotherapy and organ transplant anti-rejection drugs are used, off-label, to control the immune system's response. The goal of these treatments is to slow down the collagen build-up and the progression of the disease.

It is estimated that some 200,000-300,000 in the U.S. suffer from this illness, although estimates of prevalence from hospital-based data vary from 5 to 34 per 100,000 of the population (Cooper, Bynum, & Somers, 2009). Given the challenges in diagnosis and the overlap with other conditions, it is difficult to provide an accurate count. Similar to other autoimmune diseases, it is four times more prevalent in women in general, and some 7-12 times higher in women between the ages of 30 and 55 (Gottesman, 2003). The rarity and the lack of experience of many health professionals with the disease result in a prolonged diagnostic phase, anecdotally 5 or 10 years, where patients have to see between "two to five doctors before receiving the diagnosis" (p. 2). Primarily treated by a rheumatologist, depending on the number of organs involved, most patients have to be under the care of multiple specialists (dermatologist, cardiologist, gastroenterologist, nephrologist, and pulmonologist) and undergo various tests (blood tests, lung function

tests, echo-cardiogram, CTs, etc.) on a regular basis. Some might need mental health professionals' help to cope with the depression, stress, and the uncertainty of the disease. Tightening of the skin on hands leads to curling up of fingers, precipitating surgery for some patients. Skin tightening around the mouth necessitates seeing dentists with the knowledge and experience of working with such cases; pharmacists might have to be enlisted on how to manage prescription pills when the esophagus is affected.

The extent of the medical care needed and its coordination, how very little is known about the disease itself, the experimental nature of medical interventions employed, the considerable variations in its manifestation, the disability and disfiguration that afflict some, and the unpredictable disease course place scleroderma in the category of complex chronic autoimmune diseases. The confluence of characteristics of chronicity, autoimmunity, complexity, and diversity in presentation of the disease makes scleroderma patients a novel population to study, as this population encompasses many characteristics of chronic illnesses in one setting.

Chronicity

A Centers for Disease Control and Prevention (CDC) report (Ward, Schiller, & Goodman, 2014) places 117 million people, that is about half of the U.S. adult population, as having one or more chronic conditions. Sperry (2006) defines a chronic disease as:

> a disease entity that usually does not have a single cause, a specific onset, or a stable set of symptoms. Such a disease state is largely an objective entity. Although a cure may be possible for mild levels, it is unlikely for moderate and advanced levels of disease process. The disease course tends to be marked by periods of exacerbation and remission as well as progressive degeneration. (p. 6)

Scleroderma shares these defining characteristics with most chronic conditions. Lack of clarity on a cause(s), non-specificity of onset, a protean characteristic (absence of a set of stable symptoms), fluctuations in symptoms, and progressive nature of illness give rise to

subjective illness experiences such as a sense of uncertainty and confusion about the state of the disease and one's prognosis; interruptions of routines and unpredictability of one's health state due to fluctuation of symptoms; and stress and depression caused by the progression and downward decline of health. Although the intensity and timing of these experiences may vary among chronic disease sufferers, they do occur with all patients in some form or another, at one point or another, and may be viewed as shared commonalities among chronic disease patients.

Autoimmunity

Among the 117 million patients with chronic conditions, there are 23.5 million patients (National Institute of Health [NIH], 2012)—and by some accounts up to 50 million (American Autoimmune Related Diseases Association [AARDA], 2017)—who suffer from some 80 autoimmune diseases, including the better-known ones like rheumatoid arthritis, diabetes mellitus (type 1), multiple sclerosis, lupus, Crohn's disease, Hashimoto, celiac disease, Graves' disease, inflammatory bowel disease, psoriasis, rheumatic fever, and rarer ones like scleroderma, Kawasaki, Sjogren's and Still's diseases.

Autoimmunity, defined as the failure of body to distinguish between self and non-self (Anderson & Mackay, 2014), was inconceivable and resisted as a concept in medical community until fairly recently: "1955-1965 [was] the decade marked by the question, does autoimmunity exist?" (Rose & Mackay, 1985, as cited by Silverstein, 2014, p. xxxiv). Once accepted, it has been instrumental in shaping a re-emergent perspective on the concept of disease and its treatment. Anderson and Mackay (2014) argue that:

> As the concept of autoimmunity emerged in the middle of twentieth century, it therefore represented to a degree the return to older physiological or biographical concept of disease causation … before the bacteriological fall, when evil, in the form of germs, entered the world of etiological theory. (p. 15)

This earlier "constitutional model" of being ill, according to Anderson and Mackay, relied on restoring body's homeostats by individualizing, modulating, and adjusting medications, similar to how the modern approaches to autoimmune disease treatments have emerged and have been put into practice. These emergent holistic approaches have a better fit in chronic disease models and, it may be argued, provide a more natural context for expressions of involvement and active roles by patients. Studying patient involvement in scleroderma patients, thus, may prove to be a more meaningful undertaking due to this contextual factor, as opposed to inquiring into disorders where physicians come "to look for the disease, not the person" (p. 15).

Complexity

Scleroderma is a complex disease for a number of reasons, including multi-organ involvements. For patients with complex diseases, as Weiss (2007) notes, "clinical decision making and required care processes are not routine or standard. For complex patients, many recommendations from evidence-based medicine are unlikely to apply in a straightforward manner because of 'exceptions'" (p. 375). Scleroderma patients are not alone in this regard. This complexity extends to patients with comorbidity, such as the 25% of the U.S. adults who have two or more chronic conditions (Ward, Schiller, & Goodman, 2014). For these patients, providing information and involving them in self-management practices are not straightforward due to ambiguity in behavioral changes to recommend, and a lack of evidence-based knowledge to impart, making patient involvement or lack thereof more challenging to pinpoint. Scleroderma, where complexity plays a major role not only in treatment but daily struggles of illness, provides opportunities to look at involvement from a more nuanced vantage point by forcing us to look to other ways patients in complex situations engage with illness, specifically, when formal self-management practices are not readily applicable.

Diversity

Variations among diseases may present themselves in areas such as onset and length of time one is afflicted (from birth, childhood, adulthood, or at old age); ease of identification and diagnoses; etiology; genetic contribution; availability and efficacy of treatment; stigma and public manifestations of disease; disruption to everyday life and morbidity; and the prognosis, progression, and mortality rates. Using some of these factors, chronic diseases may be roughly grouped into four categories of (a) life-threatening diseases like cancer, (b) manageable condition such as diabetes, (c) progressively disabling diseases like Parkinson's and lupus, and (d) the non-life-threatening conditions with waxing and waning courses, such as fibromyalgia (Sperry, 2006). In scleroderma, within a single disease, the characteristics of the above four categories are represented. Consequently, in a single patient population, the breadth of issues affecting patients' involvement may be studied.

Altogether, the above four characteristics of scleroderma form the main rationale and significance for this research and its chosen population and may be summarized as:

- Studying a complex autoimmune disease with applicability to 23.5 million patients dealing with autoimmune diseases, and possible relevance to 117 million suffering from chronic conditions, a quarter of whom deal with the complexity of comorbidity.
- Studying involvement in an autoimmune disease where the conception of disease makes involvement a more natural occurrence, and where the diversity in presentation of the disease provides variability that generally requires studies across multiple disease populations.

In addition, this study consists of:

- an under-studied patient population,
- a multi-disciplinary approach to understanding and framing of the concept of involvement by considering both the illness experience and learning.

Three specific research questions, as outlined in the next section, motivate this study. How learning may underlie the engagement of patients impels a second dimension of the research questions posed.

Research Questions

The following research questions informed this study:

RQ1. How do scleroderma patients describe/understand their commitment to illness management? Did commitment to be involved change over time? If so, when and how?

RQ2. What strategies do scleroderma patients use to be involved, and how did they learn them? What do they perceive as impeding or facilitating their learning? How do they overcome the barriers?

RQ3. How are social interactions, particularly support groups' facilitations/ participations, perceived in terms of involvement and learning?

Researcher's Assumptions

Detecting early malignancies on mammograms is extremely challenging. As part a research in digital mammography to understand how a radiologist determined malignancies on X-rays, the researcher had worked with an extraordinary radiologist at a teaching hospital. The researcher has attributed the radiologist's high level of expertise and competence to how she was able to confirm or rule out her hunches by combining various sources of data: those derived from mammograms directly, the clinical and patient history data, her formal medical knowledge, her experiential and tacit knowledge of working with her patient population, as well as her openness to the observations and thoughts of patients themselves. Working with a population with a below average formal

education, talking to patients to get information, and not relying on what was written on forms were part of her routine. Observing how she engaged patients, in a specialty in which patient contact is not a normal part of the practice, led the researcher to believe that the medicine practiced well incorporates many sources of information and insights, including those from patients. Conversely, patients have to play their part in this exchange for better outcomes.

Fast-forwarding ten years, one of researcher's children was diagnosed with a serious condition. Contrary to advice given by a doctor, she researched the illness. Her research led to the recognition of misdiagnosis by the treating doctor, rejection of treatments offered, and seeking treatments at a specialty center, which diagnosed the illness as transitory and not needing medication. Since that encounter, the researcher has come across other parents with similar experiences who had learned on their own and had become experts in the conditions of their children. They had to learn how to navigate the healthcare system, what to demand, and how to actively be involved.

Recently, a relative was diagnosed with scleroderma. Many years have passed since the earlier isolated experience of searching for information for the researcher's child. This time, looking for information on scleroderma, the researcher was witnessing how other patients and caregivers grappled with similar issues through online communities. These experiences have led the researcher to assume:

- Patients want to be involved in their own care.
- Patients want to know about their diseases and treatment options.
- Whereas in acute illnesses a passive role may be a welcome option, with more serious conditions, most patients are compelled to become engaged with their illness.
- Commitment to illness management and involvement with one's care is not easy. People learn through the process of doing it.

Definition of Terms

Acute/Critical Condition—Illnesses that have curative or symptom-relieving treatments, or are self-limiting, and usually have short-term durations.

Adherence—"The extent to which a person's behaviour - taking medication, following a diet, and/or executing lifestyle changes, corresponds with agreed recommendations from a health care provider" (WHO, 2003, p. 3).

Chronic Conditions—Disorders characterized by long durations, being not curable, or preventable by vaccines.

Complex Disease—Diseases encompassing multiple chronic conditions, or a single chronic condition presenting unique needs in terms of functional limitations and disabilities (Bayliss et al., 2007)

Contested illnesses—Syndromes characterized by a cluster of non-specific symptoms like pain, fatigue, and mood swings that cannot be attributed to any organic abnormality. Examples include fibromyalgia, chronic fatigue syndrome, and irritable bowel syndrome (Barker & Galardi, 2011). Illnesses where sufferers claim to have a specific disease that many physicians do not recognize or acknowledge as distinctly medical (Conrad & Barker, 2010).

Disease—A physiological condition with possible corresponding therapeutic interventions to eliminate or ease the symptoms.

Evidence-Based Guideline—"Systematically developed statement to assist practitioner and patient decisions about appropriate health care for specific clinical circumstances" (Institute of Medicine, 1990, p. 38). These guidelines are developed by a panel of experts within a medical field.

Illness—The symptomatic human experience and suffering of having a disease.

Illness experience—The working definition used in this study is: the combination of physical impacts of the disease on the body, the emotional/psychological toll it

engenders, and the social repercussions such as those relating to managing others and dealing with life work, employment, and finances.

Patient Support Group (or **Peer Support Group**)—

> the provision of emotional, appraisal [affirmational], and informational assistance by a created social network member who possesses experiential knowledge of a specific behavior or stressor and similar characteristics as the target population, to address a health-related issue of a potentially or actually stressed focal person. (Dennis, 2003, p. 329)

Rare or Orphan Diseases—The Orphan Drug Act of 1983 defines these as the illnesses that affect fewer than 200,000 people in the U.S. It is estimated that 25 million Americans and 250 million people worldwide are afflicted with one of 7,000 orphan diseases (Lo, 2014).

Scleroderma—Also known as Systemic Sclerosis (SSc),

> is a chronic connective tissue disease generally classified as one of the autoimmune rheumatic diseases. The word 'scleroderma' comes from two Greek words: 'sclero' meaning hard, and 'derma' meaning skin. Hardening of the skin is one of the most visible manifestations of the disease. (www.Scleroderma.org)

Chapter II

LITERATURE REVIEW

The purpose of this study was to explore and describe, through scleroderma patients' own perceptions and understanding, their commitment to illness management, including how they were involved in dealing with their illness and how they learned to do so. To carry out this research, a literature review was conducted.

Patients' active role is conceptualized under terminologies such as patient empowerment, involvement, participation, activation, and engagement, among others. In common parlance, these words are considered synonymous. For example, involvement is understood to mean being a participant embroiled or engaged in an issue or activity. In contrast to their common meanings, there is a lack of definitional clarity in a large body of literature that has explored these concepts. Gallivan, Burns, Bellows, and Eignseher (2012), in a literature search of studies between 1995 and 2010, found 15 different terms utilized. Fumagalli, Radaelli, Lettieri, Bertele, and Masella (2015) noted, "These concepts are sometimes treated as synonyms, other times in contraposition, yet others as unrelated concepts" (p. 385). Gallivan et al. (2012) observed that

> there is broad support for the idea that patients should be "involved" and should "take part" in health care, but no clear consensus exists about what precisely this means and how much or to what extent patients are included. Although these terms [patient involvement, partnership, collaboration, participation, engagement] are used in the literature, they are not readily defined in most papers and thus the reader is not necessarily clear as to what they mean. (para. 14)

The lack of clear definitions and consensus among researchers on what these terms mean and how to meaningfully operationalize them, referred to as a "conceptual muddle" by Forbat et al. (2009), are echoed by many, including Elwyn, Edwards, Mowle, and Wensing (2001), McCormack et al. (2011), and Staniszewska, Brett, and Mockford (2011), who stated:

> While there are some helpful definitions of involvement, the conceptualization or theorization of PPI [Patient and Public Involvement] has generally been poor. There have been some attempts to develop conceptual or theoretical frameworks, but there is no overall conceptual model of PPI impact that captures the essence of the concept and has been empirically tested. Such models can be very helpful because they can provide a blueprint for evaluation, identifying key areas for assessment. (p. 394)

Beyond the conceptual confusion, the irony is not lost in combining the passively connoted term "patient" (Latin *patiere*, to bear or endure) with actively connoted words "involvement" (Latin *involvere*, to enwrap), "activation" (Latin *activus, actus, agree*, to drive, to do), "engagement" (Old French *gager*, to guarantee, as in to do something), or "participation" (Latin *particip*, to take). Nonetheless, these terms are used, sometimes interchangeably, in studies encompassing disciplines of nursing, psychology, social work, medical practice, public health, medical anthropology, medical sociology, health service delivery, policy studies, patient advocacy, illness phenomenology, ethics, and post-modernity. Menichetti, Libreri, Lozza, and Graffigna (2014), in a review of some 60,000 articles using the terms "patient activation," "patient adherence," "patient compliance," "patient empowerment," "patient engagement," "patient involvement," and "patient participation," found that the majority of these papers (74%) were in the health domain (medicine and nursing). Higgins, Larson, and Schnall (2016) found a somewhat higher distribution of 96% in medicine and nursing among 722 articles they reviewed based on keyword "patient engagement."

In conducting a literature review for this dissertation study, two approaches were taken. First, a review of five most commonly used terms—"patient empowerment," "patient engagement," "patient involvement," "patient participation," and "patient activation"—is presented in this chapter. Second, through parsing the relevant literature by differentiating between disease and illness, the notion of patient involvement in one's care is scrutinized from these two perspectives. In addition, a review of adult learning literature is included, as learning is another focus of this study. Last, theories and models informing the conceptual framework of this study are presented in more depth in a separate section, followed by a chapter summary. Thus, this literature review chapter is structured in seven parts:

1. The various conceptualization of patients' active roles
2. Disease versus Illness
3. Involvement as attending to disease in clinical interactions
4. Involvement as attending to illness in lifeworld
5. Theoretical Context: Adult learning theories
6. Conceptual Framework
7. Literature review summary

Various Conceptualizations of Patients' Active Roles

Menichetti et al. (2014) reviewed 58,987 studies from 92,771 articles retrieved using the search terms "patient empowerment," "patient involvement," "patient participation," "patient engagement," "patient adherence," "patient compliance," and "patient activation" for the years 2002-2013. Studies focusing on the concept of patient compliance constituted 80% (47,042) of the articles, followed by those using the concept of patient participation (18%). The remaining concepts ranged from 1% to 3% of the total studies. A temporal analysis for the period between 2002 and 2013 showed a drop of 10%

in the frequency of using the term "patient compliance," while the usage for "patient engagement" and "patient activation" increased 18-fold and 3-fold, respectively. Interpreting these changes from a historical perspective, Menichetti et al. viewed the period of 2002-2004 as being concerned with compliance and adherence. This was interpreted as an indication of the passive behavioral expectation for patients by providers. The period of 2006-2009 was interpreted as focusing on clinical encounters and patients' involvement and participation in clinical settings. For the period 2010-2012, patient empowerment and notions of autonomy and self-determination took center-stage. Menichetti et al. contend that from 2013 onward concepts of activation and engagement have been the foci, suggesting a "consumer behavioural perspective" (p. 523) that goes beyond the subjective experience of illness and into relational and organizational realms. In particular, Menichetti et al. claim that the concept of patient engagement subsumes the other concepts (activation, involvement, participation, adherence, and compliance), which were "traditionally used to generally denote the role of patients in their care" (p. 524).

Fumagalli et al. (2015) reviewed 286 studies from an initial list of 3,088 articles published between 1990 and 2013 addressing patient empowerment, patient involvement, patient participation, patient enablement, patient engagement, and patient activation. They found an explicit definition for these concepts was given only in "17% of studies about 'patient involvement', 29% about 'patient engagement', 30% about 'patient enablement', 42% about 'patient empowerment'" (p. 385). Only studies about activation, adapting Hibbard et al.'s (2004) activation measure, reached a 72% level of definitional reporting.

These five concepts—empowerment, engagement, involvement, participation, and activation—are delved into separately next.

Patient Empowerment

Fumagalli et al. (2015) argued that patient empowerment was conceptualized in three ways in the literature: (a) as an emergent state (having knowledge, skills, self-awareness, motivation, etc. that enabled patients to have an active role); (b) as a process leading to such a state (e.g., education, patient-centered care, etc. that facilitated active patient behavior); or (c) as patients' behavior (the actual behavior that exploited their power in a given context). Considering patient activation, enablement, engagement, involvement, participation, and empowerment, Fumagalli et al. devised a concept map to indicate the relationships among these. Subsequently, they suggested a definition of empowerment based on this concept map:

> Patient empowerment is the acquisition of motivation (self-awareness and attitude through engagement) and ability (skill and knowledge through enablement) that patient might use to be involved or participate in decision-making, thus creating an opportunity for higher levels of power in their relationship with professionals. (p. 390)

In this definition, an engaged patient "deploys a strong motivation to become more knowledgeable (e.g. by preparing in advance question, gathering additional expert opinions) and more 'powerful' (e.g. by fortifying the relationship with professionals, by seeking more appropriate settings and providers" (p. 389). Enabled patients are those who "are able to participate in self-care or shared-decision making" (p. 389) through interventions accorded to them by their healthcare providers. An involved patient participates in decision making, or shared-decision making with his/her provider. The proposed definition of empowerment subsumes engagement, enablement, participation, and involvement.

Patient Involvement and Patient Participation

The terms "patient involvement" and "participation" are shortened phrases for the concepts of patient involvement in health delivery and participation in health care consultation. As such, they are associated with patient-physician interactions, provision

of information, and level of decision making. Thompson (2007) using patient interviews and focus groups arrived at a taxonomy of patient involvement and participation consisting of three levels of patient-determined, co-determined, and professional-determined active roles. Within the patient-determined level, patient-desired degrees of involvement included non-involvement, information-seeking, information-giving, or autonomous decision-making. Thompson differentiated between involvement and participation by labeling the co-determined shared decision-making where providers surrendered some of their power as "participation." In this definition, patient participation is a subset of patient involvement, although in many studies these two terms are used interchangeably.

In Thompson's conceptualization of involvement and participation, power is negotiated between patients and providers. At the lowest level of non-involvement, the provider assumes full power, while at the autonomous decision-making level, the patient enjoys full autonomy in decision making. Fumagalli et al. (2015) reported other interpretations of involvement in studies they reviewed, including: (a) involvement as a combination of participation in decision making and in self-care and treatment; and (b) involvement as activities helping patients to have a more active role regarding choices. They further concluded that these varied interpretations make patient involvement and participation either the antecedents or consequences of patient empowerment.

Patient Activation

The term "patient activation" is generally associated with Hibbard et al.'s (2004, 2005) work in operationalizing the concept of "person's ability to manage their health and health care" (Hibbard & Cunningham, 2008). This work has been influenced by Von Korff, Gruman, Schaefer, Curry, and Wagner (1997), who enumerated a set of behavioral principles involved in collaborative management of chronic illnesses. These included

self-directed behavior and learned skills to manage the illness; motivation and self-efficacy in self-care; and monitoring and adaptation to emotions, symptoms, and disease states. They further asserted that in collaborative management of chronic illness, "providers as well as patients will benefit from active participation in this process" (p. 1099). Taking these assertions, Hibbard et al. (2004) inquired into the question of "what skills, knowledge, beliefs, and motivation do they [patients] need to become more effectual heath care actors?" (p. 1006). Relying on a literature review, expert consensus, and patient focus groups, they arrived at a definition of activation based on these elements:

> Those who are activated believe patients have important roles to play in self-managing care, collaborating with providers, and maintaining their health. They know how to manage their condition and maintain functioning and prevent health declines; and they have the skills and behavioral repertoire to manage their condition, collaborate with their health providers, maintain their health functioning, and access appropriate and high-quality care. (p. 1010)

Based on this definition, Hibbard et al. developed a measure of activation, PAM (Patient Activation Measure), which measures a patient's belief in the role they need to play, knowledge of the disease, and skills and confidence in enacting behavioral changes to manage the illness.

Fumagalli et al. (2015) compared patient activation to patient empowerment and noted that both concepts are related to "an increased ability and motivation, and growing patient awareness of having an important role in the management of own healthcare" (p. 388). Given the overlap, they argued that empowerment has a broader scope, while activation "is more focused on precise and specific improvement goals" (p. 388) in the context of a disease.

Graffigna, Barello, and Triberti (2015), on the other hand, drew a comparison between patient activation and patient engagement, asserting that "the concept of activation features numerous elements that resemble the characteristics of the patient

engagement process. Indeed, the two terms are often used synonymously in the scientific debate" (p. 19). Despite these similarities, they argued that the focus of patient activation is on "the conative dimension of the behavior of the patient, and assumes that the main driver of activation is the level of knowledge of the patient about the disease … [and] focuses on the behavior of patients as a reactive response to the healthcare system and to its organizational practices" (p. 20). Graffigna et al. argued that patient engagement, on the other hand, attends to non-institutional context of patients' experience of an illness.

Patient Engagement

Fumagalli et al. (2015), in a review of related literature, noted that "patient engagement has rarely received an explicit and precise definition" (p. 389) and pointed to two interpretations in literature that often coexist in the same study: "patient engagement as the participation of patients in self or shared management," and "the behavior that patient preform to improved their role in healthcare" (p. 389).

Barello et al. (2014) sifted through 1,020 potential articles published between 2002 and 2013, ultimately retaining 259 abstracts that "adjectively defined and contextualized" patient engagement. Using occurrence and co-occurrence of words and by employing chi-square tests, Barello et al. attempted to determine how the meaning of patient engagement (semantic patterns) was constructed in these abstracts. The definitions assigned to patient engagement were reported within five fields of inquiry. In the biomedical field, engagement was conceptualized as "'learnable' and 'malleable' patient attitude" and a top-down strategy to help patients self-manage their disease. In nursing, engagement was viewed as patients' self-awareness in expressing their physical and emotional needs and providers' role in facilitating support, choice, and the desired interventions. In mental health, engagement was portrayed as fostering a collaborative approach and the alliance between patients and providers in the service of obtaining better outcomes. In the public health domain, engagement was seen as empowering

patients, orienting health policies, and promoting public health. In multidisciplinary health research, engagement was equated to disease self-management.

Barello et al. (2014) concluded that "despite the widespread use of the term 'patient engagement' our study showed significant variations in its conceptualization, testifying that the debate on this issue is still in its infancy" (p. 4). Furthermore, they observed that "the voice of patients in defining what patient engagement is and what may favour it is still under-represented in the literature, thus suggesting the need for more research on patients' perspectives" (p. 4).

Higgins (2016) reviewed 722 published articles through mid-2014, over 90% of which were in medicine and nursing, to arrive at a definition of engagement through concept analysis. Out of 722 articles, ultimately 96 articles that defined or measured engagement or had it as their main subject were retained for further analysis. A large number of attributes (446), as characteristics of the engagement that were either directly or indirectly inferred, were identified and grouped into three general domains of "process" containing 43% of the attributes, "behavior" encompassing 33% of the attributes, and "environment" entailing 24% of the attributes. These domains, then, were organized thematically into six categories of time, activity, process, cognition/emotion, ethics, and communication, and finally were reduced to four overall themes of "personalization of the approach to care," "access to necessary resources," "commitment to pursuing quality care," and "nurturing the relationships between actors in the encounter" (p. 57). Higgins contends that these themes are "the defining attributes of patient engagement as they describe the nature of the behaviors, processes and environments that support engagement" (p. 57). Separately, 34 antecedents to engagement (which were reduced to 12) and 21 consequences of engagement were collected from the 96 articles. These antecedents and consequences, together with the four overall themes, formed the basis of a definition of patient engagement as:

> the desire and capability to actively choose to participate in care in a way uniquely appropriate to the individual in cooperation with a healthcare provider or institution for the purpose of maximizing outcomes or experiences of care. Patient engagement on the part of the provider is the effort to foster that desire and capability. (p. 62)

To elaborate on this definition, Higgins defined the terms "desire," "capability," "active," "choice," "unique," and "cooperation" used in the above definition of patient engagement as follows:

- Desire: "emotional and psychological factors such as past experiences or social support structures that shape their interest in exploiting the healthcare options that may be made available" (p. 63).
- Capability: "cognitive factors such as functional literacy, as well as social economic factors, that enable the patient to access those resources that support self-care" (p. 63).
- Active: "the belief or self-efficacy to participate in care, which is a central aspect of the concept of patient activation" (p. 63).
- Choice: "patient's freedom to decided how and to what extent he/she wants to participate in shared decision making and the process of care' (p. 63).
- Unique: "the unique circumstances of each patient such as patient's personal preferences and health literacy status" (p. 63)
- Cooperation: "elements of the provider-caregiver-patient therapeutic alliance as a form of the human connection that provides psychological and social support to patient efforts to participate in care" (p. 63).

These elaborations make the patient engagement definition proposed by Higgins a complex and compound construct defined based on six other compound constructs, which, in turn, include concepts such as experience, social support, functional literacy, self-care, self-efficacy, freedom to decide, health literacy, personal preference, and alliance with providers.

Whereas in the domain of nursing, Higgins's (2016) concept of engagement is defined in terms of patients' "desire and capability to actively choose to participate" and cooperate with health providers in order to "maximize[e] outcomes or experiences of care" (p. 62), Graffigna, Barello, and Triberti (2015) conceptualize engagement as a "psychosocial process" (p. 29) of decision making and negotiation with healthcare providers based on consumer psychology theories. These theories are concerned with "processes that individuals, groups, or organizations enact to select, secure, use, and dispose of products, services, experiences, or ideas, in order to satisfy their needs and to fulfill their goals and values" (p. 29). Thus, engagement may be seen as what patients are able to exhibit in terms of decision making, sense making, and negotiation with providers, given the particular confluences of patients' feelings, thinking, and acting. Formally, engagement is defined as a conjoint cognitive, emotional, and conative orientation that patients take toward the management of their health.

This orientation unravels sequentially as a process in four phases. Each dimension of cognition, emotion, and conation contributes, synergistically, in each phase. In the initial phase, patients are in a "blackout" state, and everything seems to be out of their control; they are passive recipients of care. In the second stage, although better-informed, patients are overwhelmed by emotions and "unable to self-manage their diseases and treatment prescriptions" (p. 31). In the third phase, patients have acquired disease knowledge and skills to follow medical interventions and cope with their condition, but are doing so passively and not as collaborators. In the last stage, patients come to the acceptance of their illness, are fully engaged, and can form partnerships with their providers. Arrival at this stage is dependent on a successful movement from one stage to the next. This movement is viewed as an "elaboration and reframing" (p. 36) of health experiences. To facilitate this movement, "the healthcare system needs to educate patients and improve their understanding of their health and related conditions" (p. 36) and "provide patients with occasions to improve their sense of self-efficacy and

confidence" (p. 37). In later stages, patients need to take up active social roles and incorporate their patient identity into their lives. These positive steps need "to be legitimized by the healthcare system" (p. 38). This model of engagement is operationalized in a Patient Health Engagement Scale (PHE) as a measure of patient engagement.

Graffigna, Barello, and Triberti's (2015) four stages of engagement are reminiscent of Hibbard et al.'s (2004, 2005) four levels of activations, although Graffigna et al. argue that these stages encompass more dimensions than Hibbard et al. behavioral focus. Given Graffigna et al.'s claim of a model designed to "see engagement from patients' eyes" (p. 28), there is a great amount of reliance on health providers and the healthcare system to help move patients from one stage to the next by "educating" and "legitimizing" their efforts.

In concluding this section, several observations may be made given the above definitions for the five terms used to capture patients' active roles: (a) these definitions and conceptualizations are defined, by the authors of these studies, as either overlapping or subsuming the other concepts, making for compound constructs that are difficult to tease out, measure, and operationalize; (b) in these definitions varying levels of agency and knowledge are afforded to patients; similarly, to varying degrees healthcare system and providers are seen as enablers of patients' active roles; (c) the discipline within which a particular conceptualization is situated (e.g., psychology vs. nursing) exerts an influence in the definitional focus presented; and (d) some of the observed differences may be attributed to the underlying paradigmatic views of disorder either as a biomedical disease or an illness experience.

Among these, the perspective of disease versus illness deemed to be the most important and relevant to this study, as one denotes the perspective of the professionals and, on the other hand, that of the patients. This distinction sets the stage for the expected roles for each. To represent patients' voice in this discussion, therefore, it is important to

understand how approaches to patients' active role are grounded on such a distinction. This is explored in the next section. Given the lack of clear definitions, the terms patients' "active role," "involvement," and "engagement" are used interchangeably.

Disease versus Illness

The biomedical sciences adopt a positivist posture characterizing disease as an aberration or deviation from statistically determined biological, physiological, and pathological norms observed in aggregate of the human species. Larsen and Lubkin (2009) echo the prevailing sentiment that disease is an "alteration in structure and function" (p. 4). Campbell, Scadding, and Roberts (1979) identify disease as referring to "(a) a described and recognizable combination of symptoms and signs; (b) a phenomenon associated with a specified disorder of structure or function; or (c) a phenomenon due to a specific cause or causes" (p. 757). These definitions coexist with at least seven other views in biomedical literature identified by Boorse (1979). From these views, the body, or more precisely its functioning, is the main focus, and patient involvement may be viewed as compliance with the interventions that would restore normal conditions, or at least prevent deterioration of functions. The roles assigned to patients include cooperation in taking medication and doing home monitoring, undergoing procedures, keeping doctors' and labs' appointments for professional monitoring, and making lifestyle changes deemed to improve outcomes.

On the other hand, social and psychological considerations prompt a distinction between disease (pathophysiologic) and illness (symptomatic human experience and suffering), extending the notion of an ailment from a purely biological deviation or abnormality to a psychologically and socially constructed one. A common thread among many perspectives on illness is the premise that the self has a social nature and the meaning given to illness is not inherent within it, but comes about through interactions

and relations of individuals within their social contexts. The notion of "sick role" coined by Talcott Parsons (1951), American sociologist, captures this idea that "our repertoire of responses to illness are governed by a set of social expectations and responsibilities" (Bissell, Travlsen, & Haugbolle, 2002, p. 60). These social norms allow, for example, exemption from work and an obligation to seek medical help when sick, and resumption of normal work, when better. Over time, attention to patient's role turned to "illness behavior" and "health behavior" (Burnham, 2013). A major component in viewing illness became the nature of the altered relationship with one's body when sick. This altered relationship occurs in the context of one's social world giving way to a "loss of self" (Charmaz, 2003), or a "biographical disruption" (Bury, 1982, 1991) affecting a person's sense of identity and the ability to fulfill roles in everyday life (Thorpe, 2009). Illness poses as a stressor, disrupting life's steady state (Cohen & Lazarus, 1979; Moos & Tsu, 1977), requiring patients' proactive engagement with tasks as a means of adjustment. Coping strategies and self-regulating behavior based on one's illness representation (Leventhal, Meyer, & Nerenz, 1980), and integration of illness into one's life through biographical, illness, and everyday life work (Corbin & Strauss, 1988), among others, gave impetus to the notion of self-management behaviors and self-care activities as a more formalized approach to view and promote what patients engage with in between doctors' visits.

These two general strands reflecting the primacy given to the disease or the illness when considering the patients' roles are reviewed next.

Patients' Active Role: Attending to Disease in Clinical Interactions

In medical studies, engagement stems from the perspective of attending to disease condition and improving the physiological symptoms of a disorder. Therefore, patient engagement is viewed to take shape in the context of the patient-physician dyad in the

confines of clinical consultations and interactions. Two major components of such a construction are: (a) provisions of information, and (b) preference for control in decision making. Involvement is promoted by providing information to patients and encouraging their participation in decision making in treatments.

Provision for Information

In clinical settings with a focus on patient-physician interactions, patients' need for information in order to make decisions regarding their illness is considered essential for involvement, yet studies examining the informational needs of patients are limited. Among studies in this area are those exploring the nature of informational materials given to patients and their impact, factors affecting information seeking, the nature of patient and physician exchanges of information, challenges in conveying risk information to patients, and the role of technology in patients' information-gathering activities.

In a discourse analysis of 1,000 patient-leaflets by Dixon-Woods (2001), only a small subset of patient-leaflets was characterized as having a patient empowerment discourse, while the majority were seen as having the traditional approach of treating patients as "passive" (p. 1417) recipients of information. Wetzels, Wensing, Van Weel, and Grol (2005) provided educational leaflets to patients prior to doctors' visits as an intervention, and found no difference in terms of patients' involvement, satisfaction, or enablement between intervention and control groups. Pegg (2003), in studying the difference between the impact of personalized information and general information, found that patients receiving personalized information on physical therapy had higher levels of participation and reported greater satisfaction. Aldoory (2001) looked for factors impacting patients' engagement with health messages, specifically those influencing women's involvement with public health messages (e.g., harm of smoking; caffeine in pregnancy, etc.). Receptivity to health messages was related to life roles and situations (motherhood, pregnancy, etc.), one's sense of identity (racial, ethnic, economic, etc.),

source of information (media, celebrities, authority figures, etc.), and women's own conceptions of health status.

The face-to-face information transfer between doctors and patients was studied by Lee and Garvin (2003). They found that the "one-way model of information transfer" (p. 449) from professionals to patients was pervasive and relied on three problematic assumptions that privileged expert perspective, considered as appropriate in a one-way flow of information from doctor to patient, and focused on individual risk and behaviors without the consideration of structural barriers. Beisecker and Beisecker (1990), in a study of 106 patients, attributed patients' information-seeking behavior to variables such as length of interaction, diagnosis, and reason for visit, and found that interactions with a duration of 19 minutes or longer were required for patients to engage with information-seeking behavior.

Increasingly, technology plays a role in acquisition of information and is altering the traditional information exchanges with doctors. Lee and Hawkins (2010) reported that women with breast cancer who perceived unmet informational and emotional support were more likely to engage with online sources. Synnot et al. (2016) found that MS patients perceiving "too much information" and "too little that applied to them" in their online searches, while Ginnossar (2016) reported age, education, ethnicity, and prevention orientation as predictors of information-seeking behavior in cancer prevention. Chung (2013) reported White, middle age, highly educated women as being more likely to go online for health information. In that study, only 30% of those searching online discussed the search results with their health providers, confirming other reports that only 20% to 30% of patients disclosed or discussed their online findings. Chung listed men and those concerned with quality of online information as being more likely to discuss their findings with doctors. In a review of 18 studies on the online information-seeking behavior of patients, Tan and Goonawardene (2017) found that patients used various strategies in sharing or verifying information they obtained online,

including directly disclosing findings, verifying silently, and bringing printouts to doctors' offices. Having a family member present, doctors initiating questions, and advertisement seen in media facilitated office conversation on online health information, while fear of doctors perceiving online information as a challenge or insult, resistance and discouragement of doctors in use of online sources, embarrassment in patients' abilities to relate the information to their own cases, and lack of time were seen as barriers to information sharing.

Lastly, imparting information regarding risk and uncertainty of treatments increasingly is seen as essential for patients to make informed decisions (Smith, Street, Volk, & Fordis, 2013; Zikmund-Fisher, 2013). Han (2013) studied the challenges in conveying concepts such as meaning of probability (risk), ambiguity (reliability & credibility of information), and complexity (multiplicity of risk factors) related to treatments and procedures. He identified the key areas in need of future research as: conceptualizing uncertainty, identifying what and how much to communicate, devising a standardized language and methods of communication, and determining ways to take on a patient-centered approach.

Among the above issues, the online information gathering and conveyance of risk and complicated medical concepts, exacerbated by limited time available at visits, have taken patients' informational needs to uncharted territory. Providers and patients alike are not quite sure how to manage these situations. Inconsistent expectations from patients, variously conveyed by providers to be independently informed or not, are confusing to the patients. Similarly, unreliable online content and demands for interventions read on the Internet are frustrating to the providers. Additionally, the access to a wealth of information has transformed the notion of informational need to a need for discussion to evaluate and discriminate the relevancy and validity of information. This is occurring at a time when health providers are constrained by the amount of time they can devote to each

patient, and compensation models (fee-for-service, pay-for-performance, etc.) that discourage such engagement.

Thus, access to online information has made the traditional notion of provision for information, as it has been explored in literature, outdated and moot. The nature of information exchange between patients and providers is, at best, a fluid one, as new norms are being established.

Preference for Decision-Making

Exerting influence in decision-making is the second notion most equated to involvement in the health domain literature. Storm and Edwards (2013) assert that "user involvement is intended to increase the actual and 'real' influence of patients on decisions about their treatments, to ensure that services are provided in accordance with patients' needs and to enhance patients' control over their health care" (p. 314). Arnstein's (1969) ladder of control is frequently adopted to devise a hierarchy of control and influence in decision-making to describe patients' sphere of influence. The best-known hierarchy is that of Emanuel and Emanuel (1992), consisting of levels of *paternalistic, informative, interpretive,* and *deliberative models*. In the paternalistic model, physicians act as guardians articulating, persuading, and implementing what is best for patients. In the informative model, physicians are the experts providing facts patients need to decide. In the third interpretive model, physicians act as counselors in an advisory capacity by not only providing medical facts but also helping patients sort through their values and determine which treatments realize those values. Lastly, in the deliberative model, physicians are teachers who engage the patients in dialogue. Charles, Whelan, and Gafni (1999) suggest another configuration of similar levels consisting of three elements: the unilateral extremes of paternalistic and informed models, and a shared decision-making model where doctors and patients jointly arrive at decisions.

Thompson (2007) divides the hierarchy of control into two strands of "professional-determined patient involvement" and "patient-desired involvement." The former includes a range consisting of "being excluded," "paternalistic consultation and information giving," "shared-decision making with professional-as-agent," and "informed decision making" The patient-desired involvement, on the other hand, has five levels of "non-involvement," "information seeking," "information giving," "shared decision-making," and "autonomous decision-making." Epstein and Gramling (2012) collapse these hierarchies into a collaborative emergent model of "shared mind," where complexity and uncertainty of decisions in the context of a serious and life-threatening disease are tackled through interactions of all stakeholders (patients, family, and doctors). An array of instruments for measuring patients' preferences for decision-making have been proposed. For the period of 1980-2005, Dy (2007) identified 11 instruments, including CPS—Control Preferences Scale (Degner, Sloan, & Venkatesh, 1997), PSDM—Problem-solving Decision Making Scale (Deber, Kraetschmer, & Irvine, 1996), API—Autonomy Preference Index (Ende, Kazis, Ash, & Moskowitz, 1989), and HOS—Health Opinion Survey (Krantz, Baum, & Wideman, 1980). The focus of these instruments is on patients' perceptions of role (how passive or active they want to be) and their preferences for information, and the amount and type of information desired using assessments such as scenarios and vignettes.

Dy (2007) found two challenges in reviewing these instruments: the heterogeneity of domains in which the instruments were originally designed and tested, and a general lack of validity testing. In spite of these limitations, there is an abundance of studies in this area. Say, Murtagh, and Thomson (2006) reviewed 25 quantitative studies using the above measures, seven qualitative, and one mixed methods covering years 1975-2003. Seventeen out of 20 studies found associations between younger age and more active decision-making preferences. Five out of eight studies found that women were more likely to express a preference in having an active role. Sixteen out of twenty associated

higher education with involvement. Also, White patients were more likely to be more involved. Associations between involvement and marital status, class, occupation and income were more problematic to interpret. The severity of disease and the decision-making involvement were inversely associated (the more severe a condition, and more serious a decision, less involvement was displayed).

In a literature review of studies done between 1966 and 2009, Tariman, Berry, Cochrane, Doorenbos, and Schepp (2010) examined preferred and actual/perceived decision-making roles (autonomous, shared, passive) in cancer patients. Twenty out of 22 studies used the CPS measure. These studies showed the level of control patients preferred, initially, was more than what they perceived as ultimately having occurred. Only three studies (all of prostate cancer patients) reported the reverse. Furthermore, the majority of patients wanted information, but only 2/3 desired active participation in decision-making. Gaston and Mitchell (2005), looking at studies between 1966 and 2003 in late-stage cancer patients where palliative care rather than cure was the goal, found that advanced cancer patients had the same desire for information as early cancer patients, and about 2/3 of these patients wanted to be a part of the decision-making in some form. Furthermore, those whose conditions deteriorated wanted to surrender control; those who improved wanted more control.

Chewning et al. (2012) conducted a systematic review of 115 studies done between 1980 and 2007. These studies utilized measures of CPS in 44 studies, API in 16, HOS in 5, PSDM in 6, and other measures in the remaining 49 studies. Across all studies, 63% found that most patients wanted to be a part of the decision-making process, and 22% found that the majority of patients wanted to delegate decision-making to their physicians. These included 75% of cancer and invasive-procedures patients, who indicated preference for decision-making, as opposed to 50% of those with other chronic conditions and in the general population. The authors concluded, “The choice of the study population contributes to contradictory findings in the literature” (p. 14). In

addition, “clear differences occurred in patients’ reported decision role preferences depending on the measure used” (p. 14). In particular, they found that instruments using hypothetical vignettes (such as API) had inconsistent results, and instruments that explicitly provided an option for shared or autonomous decision roles (CPS or PSDM) had a higher percentage of patients opting for them. Chewning et al. concluded that there was an important methodological issue in interpreting mean scores used by many measures, and pointed out the lack of psychometric data to assess reliability and validity of some of these tools.

Frongillo, Feibelmann, Belkora, Lee, and Sepucha (2011), arguing a lack of “consensus over how to measure shared decision making or patient involvement” (p. 70), operationalized the perception of involvement for early stage breast cancer patients, by a score based on seven items in four categories measuring patients’ perceptions of whether a discussion of options (mastectomy or radiation) took place; the levels of “pro discussions” and “against discussions” for each option; and the extent of discussion concerning patients’ own preferences. Similarly, Ashraf et al. (2013) determined the nature of involvement by mapping survey questions, similar to the CPS measure, ranging from “I made the decision with little or no input from my doctors” to “My doctors made the decision with little or no input from me” (p. 666) to three levels of paternalistic, informed, and shared decision-making.

A limited number of qualitative studies have examined how patients interpret decision-making. Ziebland, Evans, and McPherson (2006) interviewed 43 women between ages 33 and 80 with ovarian cancer in the U.K. regarding their involvement in treatment choices. Responses ranged from perceptions of not having any ‘real’ decisions to make to being confused about options and worrying about going against a doctor’s recommendations. Ziebland et al. concluded that giving patients choices could be interpreted as abandonment by some patients. In another qualitative study of 17 patients, Edwards and Elwyn (2006) found that, contrary to the emphasis placed on decision-

making in the literature, it is of little importance to the patients; "it is the process of involvement that appeared to deliver benefits for patients, not the action of making the decision" (p. 317). Entwistle et al. (2008) found that diabetic patients' involvement in decision-making was related to three aspects of "ethos and feel of healthcare encounters" (respectful and non-judgmental), "communication about health problems" (clear explanations, listened to), and "communication about treatments" (rationale for treatment and having a say) (p. 362). Based on these findings, they point to the "insufficiency of models and practices that focus narrowly on the exchange of information about treatment options and the activation of patients as choosers" (p. 373). Lastly, Joseph-Williams, Elwyn, and Edwards (2013), studying patients' perceptions of enablers and barriers to involvement in decision-making, found two key factors of knowledge (of treatment options and personal preference) and power ("perceived capacity to influence the decision-making encounter," p. 15) as instrumental for participation.

Overall, these studies point to the role and power of providers in setting the tone for patients' participation in decision-making. The severity of patients' conditions, age, race, gender, and educational attainment were also implicated as factors impacting the desire to engage with decision-making. The diversity of disorders and contextual factors, such as confusion about choices and concerns for doctors' reactions, make generalization of these studies challenging. Furthermore, the notion of involvement as a dynamic process, which may vary at different times during the course of illness, is missing from these studies.

Patients' Active Role: Attending to Illness in Lifeworld

Whereas involvement in clinical settings and interactions is concerned with patient-physician dyads, a second body of literature looks at patients' active role as engagement with one's care, occurring in between doctors' visits, through management of symptoms

in everyday routines. This type of involvement is generally labeled as self-management or self-care. Again, neither concepts nor terminologies are precise. Jones, MacGillivray, Kroll, Zohor, and Connaghan (2011), in a review of studies between four decades of the 1970s to the 2000s, found 139 definitions of self-care. Terms such as *self-management*, *lay self-management*, *illness-management*, *self-care*, *participation*, *coping*, and *self-regulation* are used to discuss similar or overlapping ideas. Lorig and Holman (2000) write that "one of the first uses of the term self-management appeared in an article on asthma self-care written by Thomas Creer in the mid-1970's" (p. 4), with self-care and self-management appearing synonymous in the same sentence. Godfrey et al. (2011), in a thematic analysis of conceptualization of self-care, self-management, and self-management support, concluded that "these concepts can be differentially conceptualized according to the nature and level of networks involved, the level of imperative for action, and the types of goals to be achieved" (p. 183). Kralik et al. (2004) distinguish between *coping,* a "state of tolerating, minimizing, accepting, or ignoring," and self-management as "activities people undertake to create order, discipline and control" (p. 260), and further contend that self-management may be taken to be a structure or a process.

The frameworks looking at chronic conditions through the lens of patient experience espouse self-management as a *way of life* and a *socially negotiated process* that occurs within life world and the social contexts of family, work, and community, organically. Patients choose to be *self-managing* (Kendall, Ehrlich, Sunderland, Muenchberger, & Rushton, 2011) around "'mundane' reality of living with chronic disease" (Morden, Jinks, & Ong, 2011, p. 87).

Motivations for such an engagement are attributed to outcomes. The personally important outcomes to patients are listed as: establishing normality, happiness, and well-being (Vassilev et al., 2011), bringing order to life (Kralik et al., 2004), being able to discharge roles and social obligations (Morden et al., 2011), and creating balance in personal life (Hinder & Greenhalgh, 2012), while dealing with the burdens of disease.

These burdens include psychological ones such as lack of control over the disease, uncertain course of illness, and difficulty in maintaining a positive outlook (Cudney, Sullivan, Winters, Paul, & Oriet, 2005); and physical burdens like side effects of medication, time spent travelling to obtain care, attending to and monitoring symptoms, financial strains and impact on employment, and health services issues like lack of continuity of care (Sav et al., 2013).

At the same time, the desired outcomes could be professionally determined, bio-medical, normative, and disease-specific. They might include objectives such as regular symptoms' monitoring, medication adherence, behavioral changes, and increasing self-efficacy and psychological empowerment. Some contend the successful attainment of these objectives is predicated on acquiring disease knowledge and "learning and practicing the skills necessary" (Hibbard, Stockard, Mahoney, & Tusler, 2004; Lorig, 1993) through programmatic and formal interventions.

Looking for processes that are involved in self-management, Schulman-Green et al. (2012) conducted a meta-synthesis of 101 qualitative studies on self-management between 2000 and 2011. They identified tasks and skills associated with three categories of "focusing on illness needs," "activating resources," and "living with the chronic illness" (p. 5). In total 76 different skills were identified to accomplish 20 discrete tasks. These skills ranged from simple ones such as keeping appointments and taking medications to complex skills of communicating effectively and dealing with stigma.

The various self-management/self-care models, in spite of overlaps, may be arranged broadly in a number of categories based on the core elements of their conceptualizations.

Self-Management as Developing Skills

Lorig and Holman (2000) conceptualize self-management based on the Corbin and Strauss (1989) model in combination with Bandura's (1977) self-efficacy framework.

"Five core self-management skills: problem-solving, decision-making, resource utilization, forming of a patient/healthcare provider partnership, and taking actions" (p. 2) form the core of this model. Based on these concepts, a self-management intervention program was developed for arthritis patients at Stanford University, which since then has been applied to other conditions (chronic pain, HIV) and further refined as a generic model called the Chronic Disease Self-Management Program (CDSMP).

Self-Management as Problem-Solving

Hill-Briggs (2003) combines three problem-solving theories from cognitive psychology, educational research, and social problem-solving to provide a theoretical framework for self-management as a problem-solving process. The four components of having a "problem-solving orientation," acquiring the "problem-solving skills," learning the "disease-specific knowledge," and the "transfer of past experience" (p. 190) to new situations are deemed to lead to an effective outcome in self-management.

Self-Management as Decision-Making and Learning

Thorne, Paterson, and Russell (2003) have looked at the process of decision-making as a means of gaining and assuming control over the management of illness. By decision-making they meant "all behaviors undertaken by affected individuals for the purpose of promoting or restoring their health" (p. 1339). Being in control was identified as "being able to mediate the effects of the disease so that they could live as 'normally as possible'" (p. 1341). Three common processes were identified: (a) learning how to take charge of own life through learning about the disease, treatment, and body's responses; *body listening* ("monitoring of the body's sensations and functioning," p. 1242), and reflecting on priorities and life meanings; (b) fine-tuning to the disease-specific context by managing social context, lifestyle changes, treatment, and healthcare interactions, all within the context of one's assumptions about the future; and (c) evaluating learning and fine-tuning processes.

Self-Management as a Progressive Process with Distinct Phases

Audulv (2013) argued that self-management behaviors do not develop as a simple chronological process of increasing knowledge and skills, or as fluctuating phases in response to life changes and illness experiences' ebbs and flows. Rather, in her longitudinal study (2½ years), she observed four patterns of emerging self-management behaviors. These behaviors, labeled as *consistent, episodic, on demand,* and *transitional,* developed over time, each with its own goal and timeline. Consistent self-management behavior, such as taking medications, remained unchanged during the study period. Episodic behavior, characterized by periods of actions and inactions, consisted of behaviors that were difficult to sustain, such as smoking cessation or exercise, in spite of good intentions to do so. On demand behaviors were those addressing acute and short-term crisis. Lastly, transitional self-management behaviors were those stemming from a change in one's values, focus, or acceptance of a new situation.

The progression in Whittemore and Dixon's (2008) work is in the form of integration and assimilation through "a complex person-environment interaction whereby new life experiences are assimilated into the self and activities of daily living resulting in an overall life balance" (p. 179). They identified five phases of integration: "shifting sand," "staying afloat," "weathering the storms," "rescuing oneself," and "navigating a life" (pp. 181-182). These phases corresponded to the initial emotional response to the diagnosis, coming to terms with disease and understanding one's self-management needs, dealing with barriers and setbacks, engaging with a meaningful life to weather the storm, and managing the oscillation between "living a life" and "living an illness" (p. 177). They further asserted that in addition to *behavioral work* necessary to manage a disease and the *psychological work* needed to cope with an illness, *social/vocational work* associated with one's roles and *existential work* required in living a meaningful life were essential. Townsend, Wyke, and Hunt (2006) observed a similar concept to *existential work* in their study, identifying it as the core *moral work* of self-management.

Self-Management as Work

Hinder and Greenhalgh (2012) noted that patients viewed their efforts in self-management as work and used metaphors such as *hassle*, *grief,* and *ballache* to express the nature of physical, cognitive, and emotional work needed. Corbin and Strauss (1988), in their highly influential work, saw patients as engaging in three categories of "work"—"illness work," "biographical work," and "everyday life work"—in managing their illness. Corbin and Strauss's study has been influential in subsequent research and development of a number of self-management models. Their work forms a component of the conceptual framework of this study and is delved into in detail under "conceptual framework" later in this chapter.

The above studies provided a range of characteristics that described the nature of patients' involvement with their own care. These included development of skills, problem-solving, integration of life and illness, decision-making, and learning along a multi-dimensional process involving varying levels of engagement characterized as phases. There were overlaps in some articulations, among them the consideration of illness as a multi-faceted and dynamic entity. At the same time, each study accentuated particular features. The influence of the field of study within which the study was located accounted for this, coupled with study's population and characteristics of chronic condition considered. Most studies were of patients with a single chronic condition. Patients with multi-morbidity, in advanced stages of diseases or age, or certain chronic conditions pose a more complex situation. Bayliss et al. (2007) argued that "the person with complex care must integrate numerous, and potentially conflicting, self-management tasks "(p. 168). Among the issues faced by these patients were: getting multiple and possibly conflicting self-management recommendations due to care from multiple providers, suffering from symptomatic depression hampering self-management abilities, dealing with compounding effects of multiple conditions and medications, being seen by poorly trained physicians with little expertise in managing complex care needs, and

having to contend with lack of care coordination. Other studies confirm that patients with multi-morbid conditions have more extensive learning needs for self-management skills. (Noel et al., 2007). These more complex situations do not fit neatly into self-management models focusing on formal knowledge and skill development as main approaches to illness management.

Theoretical Context: Adult Learning Theories

An ongoing illness precipitates an avalanche of issues and thrusts them into one's life, often unexpectedly. These include existential threats, disabilities, and bodily dysfunctions; practical exigencies such as near-constant interactions with healthcare system, and vigilance with treatment regimens; interruptions in one's roles and responsibilities; and alterations in familial and social relationships. These changes, alterations, and disruptions to one's way of life bring with them enormous challenges and suffering, but also new experiences and potentials for learning and meaning-making. Many adult learning theories focus on experience and reflection on experience as the primary bases of learning. This view of learning is in contrast to learning theories that emphasize purely cognitive knowledge acquisition and recall or give primacy to behavior. This review will focus on experience, brought about by an illness, as the main impetus for learning. Among theories and models of learning from experience, informal and incidental learning, and somatic and embodied learning are highlighted.

Learning from Experience

Experience in the context of adult learning theories could mean: (a) a mental, physical, or emotional engagement in the moment; (b) a past, simulated, or introspective experience; or (c) one's sense making through collaboration in a community (Merriam, Caffarella, & Baumgartner, 2007). In Dewey's (1930) formulation, experience has

"peculiarly combined" active and passive elements: "On the active hand, experience is trying … on the passive, it is undergoing" (p. 163). By the peculiar combination, he meant there is a "backward and forward connection" where the "change made by action is reflected back into a change made in us … we learn something" (p. 163). Thus, in this view the experience of going through a chronic condition may be viewed as having the potential of undergoing a change that may be labeled as learning.

Drawing on the work of Dewey, Lewin, and Piaget, Kolb (1984) elaborates one of the well-known models of experiential learning in the field. This model suggests that learning is involved with the resolution of two conflicting or opposing ways of dealing with the world. Learning becomes an adaptation in the process of reconciling the dialectics of the experience in a transaction between the individual (subject) and the environment (object). For a patient, the object or the environment may include the disease itself, medical interventions and treatments, physical, emotional, everyday changes, as well as family, workplace, healthcare, and social interactions. This experiential model has been promoted and used in the design of patient educational programs (Arndt & Underwood, 1990; Deakin, 2011; Montez-Ray, 2011; Sorin-Peters, 2010), and in a study of dental patients' decision-making (Jayson, 2000). Kolb's model, having a reflective/constructivist perspective that focuses on the cognitive and rational processes within an autonomous agent, is criticized for its lack of adequate attention to the emotional, environmental, social, and historical contexts of experience, including issues of power (Fenwick, 2000; Heron, 1992; Jarvis, 2006).

Jarvis (2006) addresses some of these shortcomings by including a person's life history into his model and emphasizing the notion of a "disjuncture" (p. 9). Disjuncture occurs when our "biographical repertoire is no longer sufficient to cope automatically with our situation" and when "people are consciously aware that they do not know how to act. We have to think, to plan or to learn something new" (p. 9). For Jarvis, experience is not just a "segment of our life-world" that we happen to be focusing on, but also an

"episode in time." It is at these intersections of person, life, and time that we learn through emotions, reflection, and action in the world. Collins (2012) used Jarvis's model to frame an art-based HIV/AIDS adult education program, and Hakanson, Sahlberg-Blom, Ternestedt, and Nyhlin (2012) explored the experience of patients with irritable bowel syndrome participating in a group-based patient education program through Jarvis's model.

For Mezirow (2009), the incongruity and continuous negotiation of "contested meanings" (p. 3) of our experiences is our human condition. When there is a disjuncture and incongruity between our sensory experiences of an event or a phenomenon, and our internal processes try to make the event meaningful, transformative learning occurs. Learning is viewed as transformative since it alters (expands, modifies, replaces) our meaning-making processes such that we do not process our experience of the world the same way as prior to learning. Such a perspective transformation is usually prompted by a "disorienting dilemma," along an eleven-step process, and concluding with reintegration into one's social life (Mezirow, 1994, p. 224).

The mechanism through which we resolve or accommodate the incongruities in our experience is through critical reflection and dialogue. Critical reflection is differentiated from mere reflection and thinking. It is the kind of thinking that prompts examination and questioning of the underlying assumptions and suppositions that shape the way we process our experiences. By theorizing the underlying processes of change and meaning-making, Mezirow's theory is considered one of the most influential contributions to the field's understanding of human experience. It has been applied in studying the transformative learning in HIV/AIDS patients (Courtenay, Merriam, & Reeves, 1998), a follow-up study of a sub-sample of these patients (Courtenay, Merriam, Reeves, & Baumgartner, 2000), rheumatoid arthritis patients (Dubouloz, Laporte, Hall, Ashe, & Smith, 2004), stroke patients (Kessler, Dubouloz, Urbanowski, & Egan, 2009), multiple sclerosis patients (Lewis, 2009; Taylor, 2008), and Tarlov cyst patients (Reasoner, 2010).

In addition, based on Mezirow's theory, Brendel (2009) suggested a narrative-driven transformative framework to promote proactive and critical mindset in patients. Malkki (2012) has applied Mezirow's theory to the context of infertility, as an emotionally chaotic experience, to explore the connection between disorienting dilemma and initiation of reflection.

Informal and Incidental Learning

Much of adult learning is understood to occur in life and work interactions, and experiences outside formal structures and institutions (Fenwick, 2000). In a study of organ transplant patients, Plunkett (2011) found that 44% of learning came from formal sources such as medical care professionals and conventions/seminars, while patients attributed the majority of their learning to sources such as other transplant patients (28%), Internet (15%), books and videos (11%), and magazines and other publications (2%). Eraut (2004) characterized such informal learning as "implicit, unintended, opportunistic and unstructured learning and the absence of a teacher" (p. 250). He further differentiated such informal learning on two dimensions of "levels of intentionality," and the "focus on when the experience occurred" (i.e., past experience, present, or envisioned future). Three levels of intention were observed: *implicit* learning where the learner is not conscious of explicit knowledge, *reactive* or opportunistic learning where learning is more deliberate, but is near spontaneous in the middle of an action, and *deliberate* learning, either by setting definite learning goals, or through engagement of problem-solving and planning activities. In this type of deliberate learning, "learners are aware that they are learning; learning with a definite, specific goal rather than generalized learning" (Boud, Keogh, & Walker, 1985, p.18), albeit in informal settings. Such informal learning in patients with chronic conditions has been studied from the perspective of self-directedness by Rager (2004) in breast cancer patients, Rager (2006) in prostate cancer patients, and Holland (1992) in multiple sclerosis patients.

Inquiring into learning that is not deliberate, but tacit and incidental, Marsick and Watkins (1990) state that such learning is "a byproduct of some other activity, such as task accomplishment, interpersonal interactions, sensing the organizational culture, or trial-and-error experimentation" (p. 12). Their model of informal and incidental learning is part of the conceptual framework of this study and is explained in detail later in this chapter. The notion of incidental and tacit learning in chronic patients has been studied by Keeping and English (2001) in end-stage renal disease patients, and Jackson (2006) in the context of lifelong learning.

Somatic Learning

Most learning from experience theories relies on cognitive bases of learning through critical reflection, inference, creativity and innovative thinking, problem-solving, and analogical reasoning. Experience in these theories is presented as an interaction with entities in one's environment. Having a chronic disease, at one level, is the experience of a diseased body in the world. On another level, it is a somatic experience where the body is the object and subject of experience, all at once.

Literature on embodiment considers the role of body in experience by questioning the dichotomy of mind-body in Western epistemology. This disengagement of mind from body makes experience and learning "disembodied." To remedy this disembodiment, the body is recast as both "a lived, experiential structure" and "the context or milieu of cognitive mechanisms" (Varela, Thompson, & Kesch, 1993, p. xvi). Experiential learning is redefined as the "experience that re-members body and mind" (Michelson, 1998, p. 224), through a holistic process that takes into account somatic knowing (Crowdes, 2000). Embodiment becomes "a way to construct knowledge through direct engagement in bodily experiences and inhabiting one's body through being-in-the-world" (Freiler, 2007, p. 6).

Somewhat differently, somatic learning is "directly experienced through bodily awareness and sensation during body-centered (somatic) approaches and movements" (Freiler, 2007, p. 5; Bennett, 2012). According to Heron and Reason (1997), such expressions through movements and other artistic presentations are "metaphors of aesthetic creation" (p. 281) and indicative of presentational knowing. This paper defines presentational knowing as an "intuitive grasp of the significance of our resonance with and imaging of our world" (p. 281). This intuitive grasp is based on and derived from experiential knowing, which is the direct encounter with the world. The two other forms of knowing, propositional knowing (conceptual) and practical knowing (knowing how do things), complete the four ways of knowing in Heron's (1992) original epistemology. What Heron articulated was the existence of a perspective of knowing and learning that lies in between the knowing through a purely experiential encounter of physical reality on one hand, and knowing through conceptual and descriptive classification of things, where words are available to us to name things. Heron posited that these modes of knowing, in a hierarchical fashion, ground each other in one direction and consummate each other in the "up hierarchy" (Heron, 1992). This hierarchy provides for a more holistic and embodied way of knowing and learning.

Having an illness creates an awareness of body and one's state of health, which many patients find difficult to articulate in a propositional way. Somatic learning and Heron's holistic model provide a lens to look for this type of knowing and learning.

Conceptual Framework

The conceptual framework for analysis of this study's findings draws on the work of Corbin and Strauss (1988) in illness management, as well as Marsick and Watkins's (2001) model of informal and incidental learning. Corbin and Strauss's and Marsick and Watkins's models are described in this section. This is followed with a rationale for

choosing these theoretical lenses to aid in conceptualizing notions of commitment to illness management through involvement with care and learning in this study's sample.

Corbin and Strauss Model of Illness Management

Corbin and Strauss (1985, 1988) turn to theoretical perspectives on occupation and work, specifically work processes, to conceptualize management of a chronic illness in terms of the work and the task components that are performed by patients, spouses, partners, friends, etc. They define work "as a set of tasks performed by an individual or a couple, alone or in conjunction with others, to carry out a plan of action designed to manage one or more aspects of the illness and the lives of ill people and their partners" (Corbin & Strauss, 1988, p. 9). What differentiates this work from work done in professional settings (e.g., hospitals) is the nature of its inseparability from one's life. Due to this intimate interconnection with life, three types of work are identified:

- Illness work—the disease management works of controlling and monitoring symptoms, preventing/managing crises, implementing regimens, rehabilitation work, and dealing with social isolation.
- Biographical work—for "defining and maintain an identity" (p. 10). This is done by:
    - Contextualizing the illness into biography
    - Coming to terms with limitations caused, and the possibility of death
    - Restructuring new conceptions of self
- Work of everyday life—occupational, marital, domestic and child-rearing work.

On a daily basis, Corbin and Strauss note that there are great variations in type, degree of difficulty, duration, frequency, and consistency of the work. This dynamic nature is exacerbated further by the fluctuation in the disease and its movement along an illness trajectory, providing a constant threat of instability. Even small changes and

perturbations can cause chaos. In order to bring a sense of relative stability and equilibrium to this, at times, chaotic system, strategies are needed. Corbin and Strauss describe these "adjustable and changeable" strategies as "management in process."

Strategy is often thought of as plans or methods to achieve goals or desired outcomes. As such, it is involved with goal setting, planning, figuring out the needed actions and resources, and execution. Bakir and Todorovic (2010) argue that when the means-end relationship is ambiguous, while maintaining the notions of intentionality and goals, strategy takes on an interpretative characteristic. As "goals persistently shift and change as a result of interactions, and the outcomes are predominantly emergent," strategy formation becomes more of a "muddling through," "incrementality," "organized anarchy," "strategy-as-practice," "social action," and dealing in "complexity" (p. 1050). That is, strategies in complex and uncertain environments, become emergent and opportunistic, similar to the type of strategy that Corbin and Strauss see needed in illness management.

In addition to strategies, in Corbin and Strauss's formulation, motivation is needed to allow patients to continue engaging with three lines of work. Three types of motivation are listed: commitment to engage with the illness, hope to carry on, and having biographical and trajectory schemes. The latter refers to having the will to live and having an indication of the course of illness and what might lie ahead. However, they acknowledge that, if uncertainty surrounding one's condition is great, then maintaining motivation becomes challenging.

In summary, Corbin and Strauss's model has a number of features that can illuminate patients' commitment to their illness management and their involvement:

- Illness is treated as a dynamic trajectory where the only constant is change.
- What patients do is described as work, consisting of intertwined and interacting strands of activities, which simultaneously, interact with the changes in disease

providing for a complex system. Maintaining a relative equilibrium in this system through emergent strategies becomes a goal.

- Achieving this goal and engaging with lines of work hinge on motivation, which in turn is predicated on having hope, commitment, and biographical and trajectory schemes.

Marsick and Watkins Model of Informal and Incidental Learning

Marsick and Watkins (1990) contend that informal and incidental learning is the type of learning that "almost always takes place although people are not always conscious of it" (p. 12); it is often taken it for granted, because of its tacit nature. They further argue that incidental learning is often "the result of a significant unplanned or unexpected event" (Marsick & Watkins, 2001, p. 27), where people have to figure out solutions and be inventive in techniques or strategies to achieve desired results and outcomes. Observing that "learning grows out of everyday encounters while working and living in a given context" and that "a new life experience may offer a challenge, a problem to be resolved, or a vision of a future state" (p. 29), Marsick and Watkins (2001) proposed a non-linear model of informal and incidental learning. The components of the model, not intended to be sequential or in a particular order, are listed as:

1. Learning begins with an internal or external trigger signaling "dissatisfaction with current ways of thinking or being."
2. Interpretation of that experience and context by framing the experience and assessing what is problematic or challenging about it based on one's worldview.
3. Interpretation leading to examination of alternative solutions by comparing the new situation with prior experience and search for potential model of actions.

4. Employing learning strategies including acquisition of knowledge, acquiring resources and the needed skills, and motivating oneself to learn to achieve desired solution.
5. Proposing a solution and taking action.
6. Assessing outcome, the intended and unintended consequences.
7. Drawing lessons learned.
8. Using the lesson as new frame for future actions.

Three conditions that enhance this type of learning are:

- "Critical reflection to surface tacit knowledge and belief,
- Stimulation of proactivity on the part of the learner to actively identify options and to learn new skills for implementing those options and solutions,
- And creativity to encourage a wider range of options" (p. 30).

This model is suited to understand the type of unintended learning that occurs when attending to new and constantly changing experiences of dealing with an illness and resolving associated issues.

Rationale for Conceptual Framework

The Corbin and Strauss model of chronic illness management was chosen to encapsulate the notion of patients' commitment to their illness management and their involvement in their own care. First, this model is based on interviews of patients and their partners forming "a body of grounded information about the problems such couples are up against" (Corbin & Strauss, 1988, p. xi). Thus, this is a patient-derived rather than a professional-derived model. Second, the notion of work, in all its variations, as a series of purposeful activities "to carry out a plan of action designed to manage one or more aspects of the illness and the lives of ill people and their partners" (p. 9), describes how one may be involved with management of one's illness very concisely. Third, this model views a chronic illness and activities related to its management as dynamically changing,

resulting in a complex system that is more descriptive of the everyday experience of living with a chronic illness. This is an elegant model for a messy process that reflects the realities of a complex illness like scleroderma very well.

The inclusion of Marsick and Watkins's learning model is based on the following assumptions: First, being involved as a patient requires learning and working in multiple domains. This work changes in form, but is continuous. It involves doing (working and experiencing) and learning from it, and learning in order to do. One's involvement resides in this space. Second, the majority of this learning is informal and incidental. The unexpected and jolting experience of being diagnosed with a serious chronic condition may provide the need, motivation, and opportunity for a great deal of learning. In addition, the disabilities and host of problems resulting from a lack of appropriate and responsive care structure mean that patients have to figure out solutions to their problems. Along the way, they learn from their experiences and learn through their bodies and socially with other patients in venues such as support groups.

Last, many models and theories of informal and incidental learning are developed in the context of work as an unstructured environment where learning occurs. Eraut (2004) notes, "This (unstructured environment not designed for learning) is true of family and community contexts, which are even more difficult to research" (p. 247). Drawing similarities between the work of patients and those in a work environment, similar to the approach taken by Corbin and Strauss, makes the application of informal learning models to this study more appropriate. The alignment of Corbin and Strauss's and Marsick and Watkins's models also makes sense from this point of view.

Conceptual Framework Components

The Conceptual model consists of the following components:

- Illness Trajectory
- Management

    - Lines of Work (illness work, biographical work, everyday work)
    - Creating Relative Equilibrium
- Buttress
    - Strategies
    - Motivation
        - Commitment
        - Hope
        - Trajectory and Biographical Scheme
- Learning
    - Incidental Learning

These components are shown in Figure 2.1. In this representation, the dynamic and interacting illness trajectory and various forms of work are kept in an equilibrium by the buttress of strategy, learning, and motivation.

Summary of Chapter II

In this chapter, five concepts of patient empowerment, patient engagement, patient involvement and participation, and patient activation were reviewed. It was observed that these definitions and conceptualizations are either overlapping or subsuming the other concepts, making for compound constructs that are difficult to tease out, measure, and operationalize. The perspective of disease versus illness deemed to be an important distinction in defining these concepts, and it was suggested that these perspectives have shaped the way involvement is defined and explored, both in clinical interactions and patients' lifeworld. The medical view of involvement as patients' informational needs and preference for decision-making was presented. It was noted that patients' access to online information has changed the notion of the need for information to the need for discussion at a time when doctors are increasingly unable to fulfill that role.

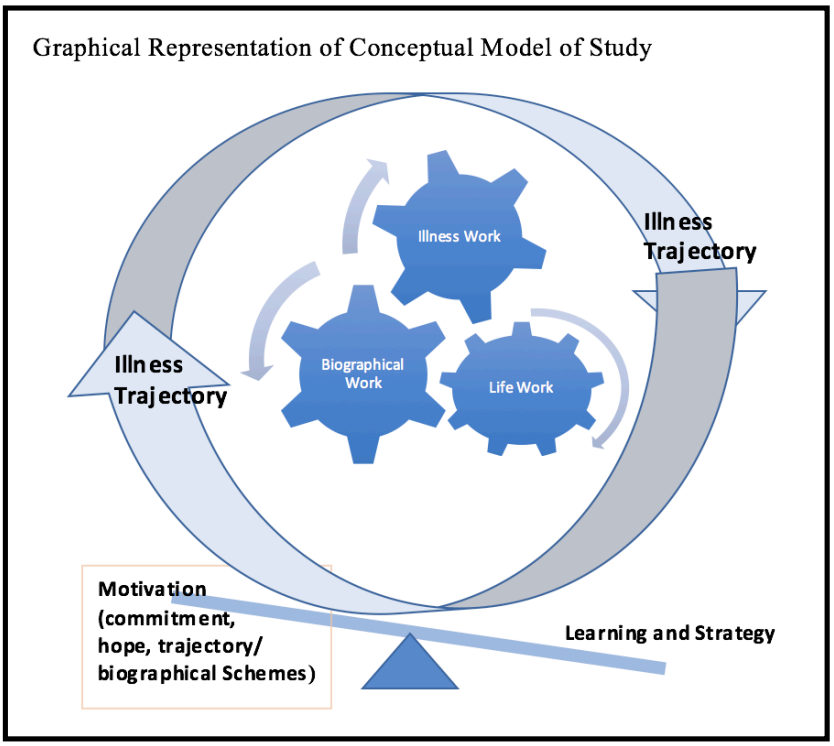

Figure 2.1. Conceptual model of study based on elements of Corbin and Strauss's and Marsick and Watkins's models. A relative equilibrium may be maintained, in this dynamic system, through motivation, learning and strategy.

Severity, age, race, gender, education, confusion about choices, and concerns for doctors' reactions impact decision-making preferences. The diversity of disorders and conflicting results made generalization of studies on decision-making preference challenging.

Patient involvement as engagement with one's care outside a clinical setting, sometimes framed as self-management and self-care, was presented as development of skills, problem-solving, integration of life and illness, decision-making and learning, work, and a multi-dimensional process involving varying levels of engagement occurring in phases. Some of these conceptualizations were deemed not sufficiently fitting patients with comorbidities and complex diseases.

A selection of adult learning theories was presented to foreground the concept of learning as central to this study. Learning was portrayed as closely tied to our experiences

and includes informal, incidental, and somatic learning. The conceptual framework of the study, based on work of Corbin and Strauss and Marsick and Watkins, was elaborated and the rationale behind the choices was given.

Chapter III

METHODOLOGY

This chapter outlines the details of the research design for inquiring into scleroderma patients' commitment to illness management, including how they are involved in dealing with their illness and how they learned to do so. The chapter is organized into the following sections:

- Rationale for a mixed-methods qualitative framework
- Rationale for a descriptive qualitative component
- Rationale for a non-experimental fixed design quantitative component
- The research sample and the selection process
- Information needed to conduct the study
- The research design, including data collection, analysis and synthesis, ethical considerations, issues of trustworthiness, validity, and limitations and delimitations of the study
- Chapter summary

Rationale for Mixed-Methods Qualitative Framework

A mixed-methods research combines quantitative and qualitative data within a single study to answer research questions. In doing so, a mixed-methods approach acknowledges the paradigmatic characteristics of quantitative and qualitative modes of inquiry, assumes that each approach provides only a partial understanding of our social

world, and advocates a respectful dialogue and integration between the two (Schiazza, 2013). These assumptions philosophically motivate the choice of a mixed-methods approach for inquiring into the complex phenomenon of patient involvement. On a practical level, scleroderma's complexity gives rise to an array of illness experiences that are difficult to catalogue. Furthermore, the rarity of the disease makes the population from which a sample may be drawn small and difficult to access. Hence, a general understanding of population characteristics as a whole is sparse, and validated instruments for this patient population are rare. Given these limitations, it was this researcher's contention that involvement of scleroderma patients was best understood when data from an individual perspective and data from a population perspective were brought together. Neither method, quantitative or qualitative, by itself would have adequately provided a clear picture of this population and the phenomenon under consideration. Therefore, a mixed-methods approach was deemed to be a more appropriate and fruitful strategy.

Issues of purpose, implementation, and integration are given consideration in a mixed-methods strategy. Purpose has to do with the reasons for mixing methods; while implementation is concerned with the order (sequential or parallel) in which qualitative and quantitative data are collected and analyzed. Integration attends to the points in the design process where qualitative and quantitative data are connected or mixed.

According to Greene, Caracelli, and Graham (1989), there are five purposes for conducting a mixed-methods analysis:

- Triangulation: to converge and corroborate results through different methods.
- Complementarity: to elaborate, clarify, enhance results from one method by the other.
- Development: using the results obtained from one method in the service of the other method, including sampling, implementation, and measurement decisions.

- Initiation: to discover paradoxes and contradictions.
- Expansion: to extend the breadth of inquiry, using different methods for different components.

The mixed-methods strategy in this study served the purposes of development, complementarity, and triangulation. First, the survey data were used to inform the selection of patients for the qualitative portion, and the answers to survey questions were used to guide interviews. Second, the survey results were elaborated and clarified by interview findings, contributing to triangulation. Specifically, the prevalence and distribution of certain quantitative data made more sense and had added plausibility, when viewed in light of the interview findings. Last, the convergence of findings corroborated the findings from each method of inquiry where integration and interpretation across all findings occurred. The overall mixed-methods design may be shown as an overall convergent design, depicted in Figure 3.1. The dashed arrows indicate where mixing of data or results occurred.

The overall convergent design of the study indicates separate collection and analysis steps for qualitative and quantitative data, followed by convergence of results in the last stage (Creswell, 2015).

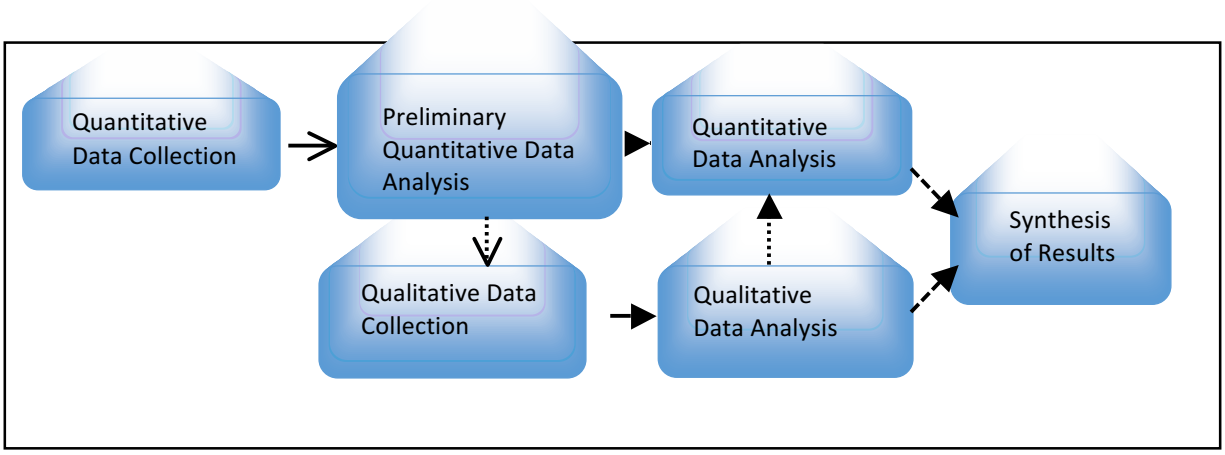

Figure 3.1. Mixed methods design of study. Solid arrow indicates the sequence of events. Dashed arrows, additionally, indicate exchange/mixing of findings or results.

The design deviated from a fully sequential, or fully parallel ordering due to practical constraints: a gap between the collection of survey data and their full analysis, followed by interviews of a geographically dispersed and qualitatively large sample of 25 people would have jeopardized access to the interview sample and would have made a fully sequential approach impractical. At the same time, lack of any information on the population's makeup and the need for a theoretical variation in the sample relative to involvement, coupled with need for a mechanism to identify the subset of participants willing to be interviewed would have made a fully parallel design unworkable. The modified design was a compromise given these constraints.

Rationale for a Descriptive Qualitative Component

Qualitative research with its inductive approach is well-suited to study perceptions and experiences of a sample. Ritchie and Lewis (2003) contend that a "major feature of qualitative methods is their facility to describe and display phenomena as experienced by the study population, in fine-tuned detail and in the study participants' own terms" (p. 27). In their view, this would allow for unpacking issues relative to dimensions, features, meaning, and typologies related to the phenomenon under consideration. These descriptive and exploratory functions provide for a mechanism to examine and "portray an accurate profile" (Robson, 1993, p. 59) of a phenomenon. A descriptive or exploratory approach is a common approach in qualitative health care related studies (Kim, Sefcik, & Bradway, 2017; Polit & Beck, 2009), where studying the phenomenon in a natural state (observations or interviews), using a maximum variation sampling, connecting to work of other researchers in the field (as opposed to just theory), and tying "nicely with quantitative data" are some of the features (Neergaard, Olsen, & Andersen, 2009, p. 4; Sandelowski, 2000, 2010).

In this study, the main focal point was the description of commitment to illness management and involvement and learning from patients' points of view, obtained by

interviewing adult patients with an official diagnosis of scleroderma living in New York, New Jersey, or Connecticut areas (as indicated by being on the Scleroderma Foundation's mailing list). A descriptive and exploratory approach was considered appropriate to obtain a direct portrayal. An initial assumption of the study was that heterogeneity in terms of involvement may be indicated by support group membership, giving the rationale for a maximum variation sampling (Patton, 2002, p. 243).

Rationale for a Non-experimental Fixed Design Quantitative Component

A non-experimental fixed design or a correlational study was chosen for the quantitative portion of the study. In such a design, the phenomenon under consideration is not manipulated by the researcher and is best suited for descriptive purposes (Robson, 2007). Since the interest in the phenomenon of involvement in the scleroderma patients was that of understanding and describing the phenomenon coupled with a one-time opportunity to collect data, this methodology was deemed well-suited and is in alignment with the purposes of the qualitative portion.

Furthermore, according to Robson (2007), a correlational design is useful in "establishing cause in the sense of providing supportive evidence for the operation mechanisms and for teasing out the particular situations and groups of people where enabling or disabling mechanisms have come into play" (p. 155). When associations are seen in the data, an opportunity is provided for integration of qualitative findings to shed light on the mechanisms involved. This made the choice of a correlational study design, as part of a mixed methodology, attractive.

The Research Sample

The Scleroderma Foundation is a 501(c)(3) nonprofit patient support, education, research, and advocacy organization for scleroderma. Twenty active chapters and

approximately 160 support groups nationwide function under this organization. The Scleroderma Foundation, Tri-State Chapter, based in Binghamton, NY is one of the semi-autonomous and larger chapters in the country covering New York, New Jersey, and Connecticut. Some 20 patient-led support groups are associated with this chapter. This chapter is not a HIPAA (Health Insurance Portability and Accountability Act of 1996)-covered entity and does not collect any health information. It has limited data on its membership, primarily consisting of contact information for the purpose of sending information on its activities (patient education, fund-raising, and legislative and policy advocacy). Scleroderma patients as the participants of this study were recruited through mail using a mailing list maintained by the Scleroderma Foundation, Tri-State Chapter. The inclusion in the mailing list might have occurred due to registration for patient educational programs sponsored by the Foundation, membership in a support group offered by the Foundation, participation in fund-raising walks and events and donations to the Foundation, or as a result of contact with the Foundation for information and support. Those on the mailing list receive the organization's newsletter, educational program promotions, and fund-raising and membership appeals. This mailing list of 1,000 names and addresses of potential participants was made available to the researcher in the form of mailing labels. Only names and addresses were available from this list. No other information regarding the population was shared. Information such as age, validity of addresses (use of non-profit bulk mail rate precludes knowledge of undelivered mail and invalid addresses), and whether the individual was a patient, caregiver, underage, or deceased were not reflected in the list, and had to be subsequently dealt with.

Recruitment procedures were negotiated with the Scleroderma Foundation, Tri-State Chapter, which provided mailing envelopes and labels. The agreement included the use of official Foundation envelopes, one-time mailing of recruitment materials, no duplication of names and addresses from the mailing labels, and the labeling of envelopes and their mailing done under observation of the Foundation's staff. Protecting the privacy

of participants and a one-time solicitation attempt were the two major considerations for allowing access to this mailing list.

The 1,000 mailed recruitment packets contained an invitation to take part in the study, along with an explanation of the study, the required IRB forms including a consent form, a demographic questionnaire, two instruments of PAM-13 and MOS-SSS, an interview interest form, and a return envelope with a dedicated P.O. Box address to which only the researcher had access. These were sent, using first class mail, to 678 patients in NY, 214 in NJ, and 108 in CT, adding up to 1,000 in total.

Simultaneously, an identical, password-protected online version of the above forms and questionnaires was provided through the Qualtrics survey tool, available from Teachers College. The link to the Qualtrics survey and the password to access it were provided in the recruitment packet. Thus, the participants were given the option of filling out the paper questionnaires and instruments and returning them to the researcher by mail to a dedicated P.O. Box address secured for this purpose, or going online and filling out the identical forms on the Qualtrics site.

Seventy-five packets were returned to sender as undeliverable [6 (5.6%) CT, 17 (7.9%) NJ, 52 (7.7%) NY)]. Presumably, 925 packets were successfully delivered. Twenty patients filled out the online version, and 192 mailed back the paper version. The online version had the completion date and time digitally recorded, and the paper versions were date-stamped upon arrival. A total of 205 (96.7%) responses were received within the 3 months after the mailing date of the recruitment materials. Another 7 (3.3%) trickled in several months later. This represented a 23% response rate for a sample of 212 respondents. Since the main focus of the study was involvement, the process of engaging with the survey and responding to it, even writing notes on the margins and including additional letters and notes by some, might have indicated this sample was the more involved sub-population that the study needed to focus on. Thus, these respondents may

be viewed as representative of more involved patients on account of their response to this study.

Among the 212 respondents, 86 patients expressed an interest to be contacted for an interview. These 86 patients were stratified by support group participation (leaders, members, non-members) and gender. Nine belonged to the "leaders" group, and seven were interviewed. From the 21 respondents in the "members" group, 4 females and 2 males were randomly selected. From the 59 respondents in the "non-members" group, 10 females and 2 males were randomly selected. In total, 7 leaders, 6 members, and 12 non-members, consisting of 20 females and 5 males, were selected through this systematic sampling and were interviewed.

The choice to include support group participation as part of the study design was based on the underlying rationale and logic for maximum variation sampling. The assumption was that support group membership status might have been indicative of higher levels of commitment and involvement, and having participants representing a theoretical variation including varying levels of participation could shed light on varied manifestations and perceptions of commitment and involvement. In addition, like most autoimmune diseases, scleroderma primarily afflicts women. A conscious choice was made to include at least a few men to increase the variation of the sample.

Briefly, the 25 interview-participants were patients with variations in support group involvement and gender who were distributed proportionally between members and non-members. The study respondents were a sample of 212 patients across NY, NJ, and CT. Details on sample characteristics for study respondents, survey participants, and interview participants are provided in Chapter IV.

Information Needed to Conduct the Study

Information required to conduct this study consisted of (a) information needed to determine inclusion criteria, (b) quantitative data, and (c) qualitative data. These are summarized in Table 3.1 and described following the table.

Table 3.1

Information or Data Needed to Conduct the Study

Data	Source	Procedure	Purpose
a) Inclusion Criteria			
Age Official SSc Diagnosis Desire to be interviewed	Patient	Survey	To identify study eligibility To identify interview eligibility & obtain permission & contact information for interviews
b) Quantitative Data			
Demographic			
Personal data Clinical data Support group participation	Patient	Survey, interview	To Identify characteristic of sample and context within which involvement is studied in this sample. To develop criteria for interview sample selection
Measurement Data			
Involvement measure Social Support measure	Patient	PAM-13, MOS-SSS instruments	To obtain macro understanding of involvement and social support in sample
c) Qualitative Data			
Perceptual			
Perceptions of: Illness & its changes Involvement per framework Learning and its changes Social support Support group	Patient	Interview	To obtain understanding of involvement and social support from individual perspective To understand connections among learning, involvement and social support
Theoretical			
Conceptions of: Involvement Learning Strategy Social Support	Journal articles, Books, Dissertations	Literature review, Web search	To determine the status of what is known on topic. To obtain theoretical basis for data collection, analysis and interpretation. To obtain an understanding of disease and its treatments To locate the sample of this study among similar studies To identify gaps, contribution of this study, and areas of future research

Inclusion Criteria Information

In order to determine the overall participation, two pieces of information: age (above 18 years of age) and an official diagnosis with scleroderma, were needed. Furthermore, to select interview participants, an indication of willingness to do an interview and contact information were required.

Quantitative Data Needed

In order to answer research questions 1 and 3, from a deductive perspective, demographic, clinical, support group status data, as well as measurement data operationalizing involvement and social support were needed. These categories were determined by review of other studies in this area and consideration of research questions.

Demographic and clinical information. Data needed included personal data, clinical/disease-related data, indicators of ease of access and extent of care (number of doctors, how far patients travel to obtain care), an indicator of patients' disease knowledge or illness status, and support group participation status.

Patient measurements of an active role. The review of existing literature indicated that patient involvement was a difficult construct to operationalize. As such, only two known instruments for assessing some conceptualization of involvement were available: the Patient Activation Measure (PAM) and the Patient Health Engagement (PHE) scale.

The Patient Activation Measure (PAM) (Hibbard et al., 2004) is a measure of patients' "ability or readiness to engage in health behaviors that will maintain or improve their health status" (Wong, Peterson, & Black, 2011). This measure is based on a construct consisting of belief in one's role in illness, knowledge of condition, and skills for obtaining high quality care (Hibbard et al., 2004).

The Patient Health Engagement (PHE) scale (Graffigna, Barello & Triberti, 2015) operationalizes a somewhat different concept of patient engagement. Engagement is

defined as a multi-dimensional, process-like experience involving emotional, cognitive, and conative dimensions. The median of 5 item scores is transformed to one of four patient engagement positions, indicative of the psychological phase a patient is in and ensuing engagement with one's health conditions (Graffigna, Barello, Bonanomi, & Lozza, 2015). This model was developed using a panel of Italian patients with chronic conditions, lacked validation with any U.S. patient population at the time of this dissertation's design stage, and suffered from an absence of independently published results. For these reasons, the Patient Activation Measure (PAM) was chosen for this study and is detailed later in this chapter.

Social support measures. In contrast to the paucity of patient involvement measures, many instruments to measure social support are available. Shumaker and Brownell (1984) define social support as "an exchange of resources between two individuals perceived by the provider or the recipient to be intended to enhance the wellbeing of the recipient" (p. 11). More nuanced conceptualizations have emerged through the years taking into consideration type of resources (material, emotional, informational, instrumental), the quantity of social connections (social network), the quality of ties between parties (formal, informal), along with notions of perceived and actual social supports indicating individuals' appraisal of availability and adequacy of support (Lopez & Cooper, 2011).

Social support instruments differ based on which of these aspects are the main focus. For example, the Multidimensional Scale of Perceived Social Support (MSPSS) (Zimet, Dahlem, Zimet, & Farley, 1988) assesses the source of support (family, friends, significant others), the Social Provisions Scale (SPS) (Cutrona & Russell, 1987) measures six dimensions of interpersonal relationship, such as guidance, reliable alliance, reassurance of worth, etc., while the Medical Outcomes Study: Social Support Survey, MOS-SSS (Sherbourne & Stewart, 1991) measures four dimensions of perceived support. The MOS-SSS measure was chosen for this study and is discussed later in this chapter.

Qualitative Data Needed

The qualitative data needed for this study included demographic, contextual, perceptual, and theoretical data (Bloomberg & Volpe, 2012). The demographic and contextual data needs were addressed above, as shared data with the quantitative portion. In addition, descriptive contextual data concerning diagnosis, receiving care, interactions with medical professionals and family, and emotional and psychological states were needed.

Perceptual data. Perceptual information in qualitative studies attends to the "participants' descriptions of their experience" (Bloomberg & Volpe, p. 106) relative to the phenomenon being studied. Perceptual information, such as how patients viewed and described their involvement, the perception of learning that occurred or needed to occur, its changes during the course of illness, and the perception of support and its role in involvement including their views of support groups, was needed.

Theoretical data. Theoretical information was obtained from research literature concerning the topic of inquiry. This included the state of knowledge about the topic, and theories utilized for conceptual framework and interpretation of results (Bloomberg & Volpe, 2012). In addition to the literature that was reviewed in Chapter II, ongoing review of other literature, specifically concerning studies of scleroderma patients, studies utilizing PAM-13 and MOS-SSS instruments, social support, and strategy, was conducted throughout the study process to guide the collection, analysis, and interpretation of data.

Research Design

The steps taken to conduct this research were as follows:

- A literature review of relevant literature was conducted and preliminary contacts with the Scleroderma National Foundation and the Scleroderma Foundation, Tri-State were made regarding access to study subjects.

- A pilot study consisting of in-person interviews of a convenience sample of three patients with various chronic diseases and disabilities, and a caregiver of a patient with a chronic disease was conducted to test out interview questions and the conceptual frameworks that were under consideration. The interview questions were shortened and re-phrased as a result.
- Research license for use of the PAM-13 measure was obtained from Insignia Health.
- Upon reaching agreement with the Scleroderma Foundation, Tri-State for access to participants, and completion of a proposal defense, the IRB approval process was initiated. This process entailed careful consideration given to standards for human subject studies, and provisions of informed consent and participants' confidentiality. IRB approved forms are attached in Appendices A-D.
- Upon IRB approval, recruitment materials were mailed to 1,000 potential participants. Over a period of 4 months, as responses were received electronically and through mail, the demographic data and instrument responses were captured in an Excel worksheet for a preliminary analysis.
- Based on the preliminary demographic data, an expressed desire to be interviewed by a subset of respondents, and through a stratified selection, potential interview participants were identified and contacted through email or phone and arrangements for conducting the interviews were made.
- Using an interview instrument, in-person interviews were conducted with 22 scleroderma patients from the northernmost parts of New York State to New York City, and from eastern Connecticut to southern New Jersey. Three additional interviews were conducted over the phone.

- MAXQDA 12 software and SPSS statistical package were obtained, and transcribed interview responses and instrument data were analyzed and subsequently integrated, synthesized, and reported.

The overall design of the study is depicted, graphically in Figure 3.2.

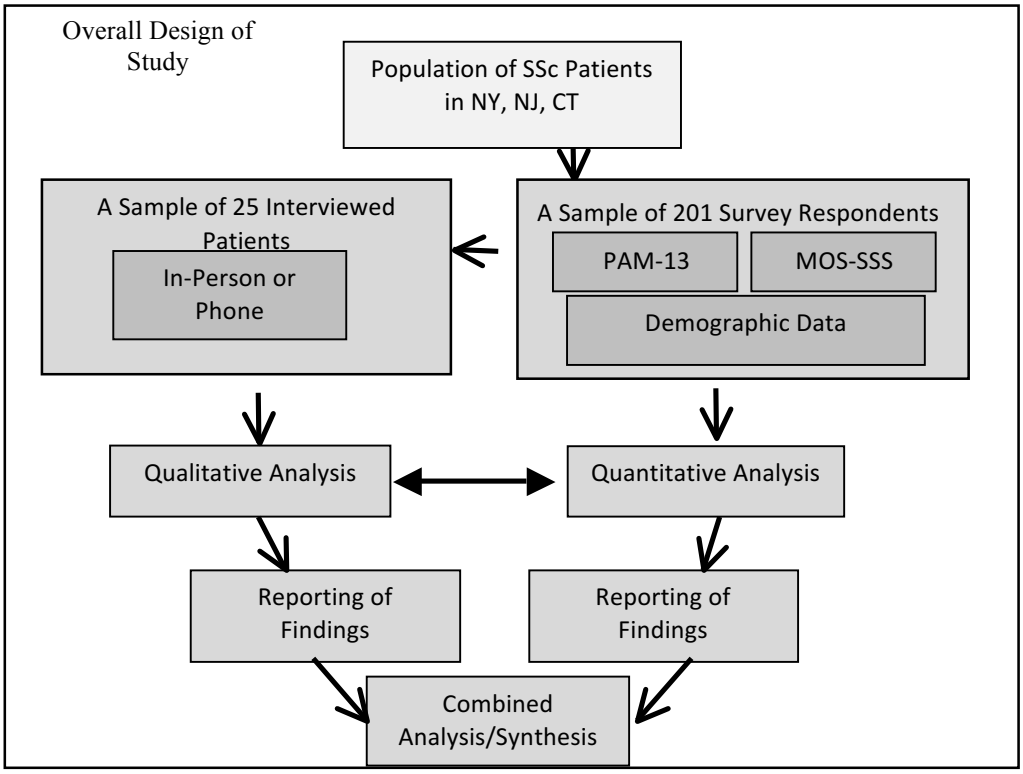

Figure 3.2. Overall design of study. Sequence and order of sample selection, data collection, analysis and mixing

Data Collection Methods

To collect data for this study, three methods of survey, scales, and interview were employed. Details of each method and the type of data collected are provided in this section.

Survey. Surveys are a common means of collecting "a small amount of data in standardized form from a relatively large number of individuals" (Robson, 2007, p. 230). They fit the non-experimental fixed design and are appropriate for descriptive and

explanatory purposes, while protecting the anonymity of respondents (Robson, 2007). As such, this was an appropriate fit for this study. To collect standardized demographic and a survey consisting of 13 questions encompassing the following demographic information was utilized:

- Personal data: gender, age, education, employment status
- Clinical data: SSc sub-type, disease duration, organs involved, co-morbidity, illness burden (fatigue, depression, pain, sleep disturbance, body image stress)
- Ease of access and extent of care: number of doctors seen, distance to travel to receive care
- Disease knowledge: knowledge of autoantibodies
- Support group participation: status as leader, member, non-member

In addition, two questions to assess inclusion criteria in the study and the desire to be interviewed were included. An affirmative answer to a survey statement "Yes, I have been diagnosed with scleroderma by a doctor" was taken as an indicator of official diagnosis. To be included in the interview portion, the respondents were asked to check if they were participating in the survey part only or, if in addition, they desired to be interviewed. Contact information for those who desired to be interviewed was collected in a separate interview-interest form. This allowed for a separate mailing of the interview-interest form if the participants wanted to keep their survey and scale data anonymous.

The Survey and the interview recruitment letters are included in Appendices D and E. All questions employed the standard ranges and categories for age, gender, education, employment, disease duration, and organ involvement, illness symptoms seen in similar types of studies, and the studies with scleroderma patients. The collected survey data were ordinal, nominal, and dichotomous (yes/no) in nature. Continuous type data (like exact age or disease duration) and certain categories (race/ethnicity,

geographical location) were deliberately not collected to safeguard against privacy violations, given the relative small size of the scleroderma community.

Scales. Measuring instruments or scales are means to assess quantitatively individuals' levels or standings relative to an attribute. Two common types of scales are the summated rating (Likert) and the cumulated (Guttman) scales. In the Likert scale, the instrument's items are independent from each other and may relate to different dimensions of an attribute. Scores are added up to obtain a total score. In the Guttman scale, there is a cumulative order to the instrument's items, so endorsing (agreement with) an item implies endorsement of all previous items, making for a unidimensional scale (Robson, 2007).

To measure involvement and the perception of social support, two instruments—the PAM-13 and the MOS-SSS—were used. The PAM-13 is a Guttman-like unidimensional scale with Likert scaling for each item (instead of just "agree" or "disagree" options, it has a range of five options), and the MOS-SSS is a Likert scale instrument. Each instrument is described, and the rationale for each choice is given next.

PAM-13 measure. The PAM (Patient Activation Measure) developed by Hibbard et al. (2004), originally as a 22-item instrument and later reduced to a short form of 13 items (Hibbard et al., 2005) (Appendix F), is intended to measure patient activation. In this model, patient activation is viewed as a construct, consisting of belief, knowledge, and skill domains, defined qualitatively through an expert panel and patient focus group study. It assumes that patients exhibit their activations through (a) the belief that they have a role to play, (b) the knowledge to manage their conditions and prevent decline, and (c) the skills to do so by obtaining high-quality care (Hibbard et al., 2004).

This construct is operationalized through a one-dimensional, Guttman-like (i.e., a probabilistic Guttman) scale. The scale's items are calibrated using a Rasch model and ordered accordingly by their difficulties. After item locations are determined, a person's location (proficiency) on the scale may be estimated. In a Rasch model, the probability of

an expected answer for a given item is a (logit) function of person's ability and the difficulty of that item on the scale. Thus, the higher the proficiency relative to an item's difficulty, the higher the probability of the expected answer for that item will be. The expected value of the observation is the sum of the expected probability for all items given the response pattern, indicative of the proficiency or trait under consideration.

Five possible response options of "strongly disagree," "disagree," "agree," "strongly agree," and "not applicable" are available for each item. The raw scores are normalized to a 0-100 range of theoretical activation score using a propriety algorithm, and are divided into four categories of activation according to pre-determined cut-off points. Table 3.2 depicts the levels, the cut-offs, the interpretation of activation levels, and each level's associated items (Hibbard, Mahoney, Stock, & Tusler, 2007). Data collected through this instrument were entered in an Excel workbook, which was provided by Insignia Health as part of their licensing to calculate scores and levels.

Table 3.2

PAM Activation Levels, Cut-offs, and Interpretations of Level

Level	PAM Score Cut-offs	Interpretation of Level	Instrument's items
1	Less than 47.0	"Passive recipient of care; not realizing importance of active role."	1-2
2	47.1-55.1	"Lacks basic knowledge about condition, or its relevance to treatment."	3-8
3	55.2-67.0	"Cognizant of key facts and beginning to take action, but lacks confidence and skills for new health behaviors."	9-11
4	Greater than 67.1	"Taken action and have adopted new behavior, but might falter when under stress /facing health crises."	12-13

This instrument has shown high internal consistency (Cronbach's $\alpha = .87$) and reliability (Skolasky et al., 2011), and has been validated with many chronic conditions, Multiple Sclerosis (MS), diabetes, Chronic Obstructive Pulmonary Disease (COPD),

chronic heart failure, chronic renal disease, spinal cord injuries, cancer, multi-morbid and mixed chronic and healthy general populations. It has also been translated and validated internationally in the U.K., the Netherlands, India, Germany, Norway, Finland, Turkey, Portugal, Australia, Italy, Israel, and South Africa (Insignia Health, 2017).

The PAM-13 instrument was chosen over the only other available option, PHE (Graffigna et al., 2015), due to validation of the PAM-13 with MS patients, another autoimmune disease causing disability. A lack of clarity in calculating scores for PHE, and the absence of independent studies with a U.S. patient population at the time the choice was made were the other considerations.

The PAM-13 measure was selected with the understanding that it addresses a narrower aspect of dealing with the physical impacts of the illness. Furthermore, it is assumed that in utilizing this measure, the physical impact of disease is considered to be the most direct and tangible consequence of having an illness, and an active involvement in physical aspects may be taken as an indicator of an overall involvement.

The MOS-SSS measure. The Medical Outcome Study-Social Support Survey (MOS-SSS) is a multidimensional 19-item instrument covering four dimensions: emotional/informal support (EMI), tangible support (TAN), positive social interaction (POS), and affectionate support (AFF) (Appendix G). It was developed by Sherbourne and Stewart (1991) in a 2-year longitudinal study of chronic illness patients (hypertension, diabetes, coronary heart disease, depression) in three metropolitan areas encompassing some 2,987 patients. It operationalized the concept of social support in terms of the functional support that patients perceived was available to them. Functional support refers to "the degree to which interpersonal relationships serve particular functions," (p. 705) such as the aforementioned dimensions of the instrument. The instrument has high construct validity overall, and for each dimension (Cronbach's α= .91-.97) (Sherbourne & Stewart, 1991).

The emotional/informational (EMI) support dimension, captured by items 1-8, is related to offering of positive affect, understanding and empathy, and availability of information and feedback when seeking solution to problems. Tangible (TAN) or instrumental support is measured by items 9-12 and relates to availability of material/behavioral assistance. Affectionate (AFF) support (items 13-15) assesses the expression of love/affection available to a person. Positive social (POS) interaction references availability of fun social interactions (items 16-18), and together with an additional item (item19), it relates to "having someone to help get one's mind off things." The dimensions are rated on a 5-point Likert scale of: "(1) None of the time," "(2) A little of the time," "(3) Some of the time," "(4) Most of the time," and "(5) All the time." The average of item scores in that subscale provides the raw score for that subscale. An overall index is calculated by taking the average of all items. These scores may be normalized to 0-100 (RAND Health Communications, 2002). The range of possible raw scores for each subscale are shown in Table 3.3.

Table 3.3

MOS-SSS Dimensions, Items, and the Range of Subscales' Raw Scores

Support Subscales	Raw Score Minimum	Raw Score Maximum
Emotional/Informational (EMI): items 1-8	8	40
Tangible (TAN): items 9-12	4	20
Affectionate (AFF): items 13-15	3	15
Positive Social Interaction (POS): items 16-18	3	15
Additional item 19	1	5

The MOS-SSS differs from other instruments that view the notion of support in terms of structure of social network and number of connections to others. As the main

focus of this study relative to social support was on support groups and learning in that context, structure and social networks were less important than the perceived types of support garnered from support groups. Teasing out social support through multiple dimensions was an attractive feature of this instrument, as support groups are particularly strong in being an all-in-one source of the type of support measured by this instrument. In addition to strong psychometric properties, this instrument is relatively short, easily worded, and can be self-administered.

Interview. Interviews are common data collection methods in social research where the meaning or perception of a phenomenon or its historical development is the focus of study (Robson, 2007). In addition to their primacy as a data collection tool in qualitative studies, they are well suited to mixed-methods research where insights and illustrations obtained through them can shed light on quantitative findings. The structure of an interview refers to the degree of formality, flexibility, and composition imposed on exchanges between researcher and subjects, resulting in a fully structured, semi-structured, or unstructured interview. Robson defines a semi-structured interview as an interview that:

> has predetermined questions, but the order can be modified based upon the interviewer's perception of what seems most appropriate. Question wording can be changed and explanations given; particular questions, which seem inappropriate with a particular interviewee can be omitted, or additional ones included. (p. 270)

Regardless of format, the researcher seeks to illicit facts, discover behavior, or uncover beliefs and attitudes. To that end, a semi-structured format was adopted to collect data on perceptions of the participants regarding their involvement; learning; their actions in dealing with their illness; support group participation; and facts surrounding the reality and context of living with this illness. An interview protocol was developed (Appendix H) based on the literature review on the topic and, in particular, concepts in Corbin and Strauss's (1998) and Thorne, Paterson, and Russell's (2003) models. It was

piloted with a convenience sample of three patients with chronic conditions or disabilities, and one caregiver of such a patient. Based on the pilot study's findings, the length of the protocol was reduced, and the initial multiple interview format was modified to one.

The semi-structure of the protocol included an open-ended first question: "To the extent you like, please tell me your story of being diagnosed with scleroderma." The open-ended format of this question was intentional to address a potential problem in the qualitative study of patients. Bury (2001) described the problem with illness narrative obtained from such interviews this way: "[Illness] narratives—which, by their very nature, require the presence of a listener (interviewer) and his/her questions and comments—may be more concerned with the repair and restoration of meaning than with conveying the 'mundane aspects of experience'" (p. 283). To ameliorate the possibility of participants' needs for telling their 'stories' taking precedence over the researcher's questions, the open-ended question was asked first. The subsequent questions were modified or eliminated depending on how much relevant information was shared by participants as part of the open-ended question. Additionally, strong responses to the instrument's items (e.g., a strong disagreement) were used as triggers for probing with follow-up questions.

Using this protocol, 22 in-person interviews and 3 phone interviews were conducted. The researcher traveled to the participants' locations. Eight interviews were conducted in the participant home, nine occurred in restaurants, and five took place at libraries, bookstores, or offices. On average, each interview took one hour and 20 minutes. With the written consent of interviewees, all interviews were recorded. Subsequently, about 32 hours of interviews were transcribed and loaded to MAXDQA software for analysis. Subsequent to each interview, field notes were written to include general observations and striking features of the interview. Table 3.4 illustrates the

connection between the research questions and the interview questions intended to secure related data.

Table 3.4

Research Questions and Corresponding Interview Questions in Interview Protocol

Research question	Interview Questions Providing Answer
1. How do SSc patients describe their experience of involvement? Did their involvement change overtime? If so when and how?	1, 2, 3, 7, 10, 11, 12, 14, 20, 22
2. What strategies do SSc patients use to be involved? What facilitated or impeded this? How did they overcome these?	4, 5, 6, 7, 8, 9, 10, 13, 15, 16, 17, 18, 19, 21
3. How are social interactions particularly, support groups' facilitations/participations perceived in terms of involvement and learning?	22, 23

As part of the interview, participants were asked about their support group involvement. All support groups in this study were affiliated with the Tri-State Chapter, encompassing states of the NY, NJ, and CT.

Methods for Data Analysis and Synthesis

This section describes the data set creation, cleaning, analysis, and synthesis for both quantitative and qualitative data.

Quantitative data. The quantitative data, initially captured in an Excel file from survey and PAM-13 and MOS-SSS measures for preliminary analysis, were reviewed for eligibility and completeness.

Data set creation and cleaning. Out of 212 responses, one online survey was determined to be a duplicate of a paper survey, as identical contact information was provided for both. The paper survey was retained. No other duplications were found. Five surveys were excluded for patients being either underage or deceased. In four cases, indicated by the response in the survey, someone other than the patient had filled out the surveys. These were eliminated due to a lack of clarity as to whose voices were reflected

in the questionnaires. One survey did not include demographic data and was eliminated. With the exclusion of these 11 responses, data from 201 participants, with complete demographic data, were retained for further processing to generate PAM-13 and MOS-SSS scores.

When PAM-13 questionnaires were scrutinized for completeness and validity, seven were found to be completely blank, and four had more than three items marked as "N/A not applicable." These were excluded. Seven additional responses were excluded by the PAM scoring algorithm, as it scrutinized the data for outliers (those marked as strongly agree/disagree for all items resulting in a score of 100) (Hibbard & Cunningham, 2008; Rademakers, Nijman, Hoek, Heijman & Rijken, 2012; Wong et al., 2011). The net number of valid PAM scores available for analysis was, therefore, 183. The raw scores were normalized to a 0-100 range of theoretical activation score using a proprietary algorithm, and were divided into four categories of activation, according to pre-determined cutoff points (see Table 3.2 for levels and cutoffs). This was done using an Excel worksheet containing PAM algorithm macros, which was provided by Insignia Health for this purpose.

For all respondents with valid PAM scores (n=183), MOS-SSS scores were calculated using the algorithm provided by RAND Health Communications (2002). Prior to the calculation of scores, individual MOS responses were scrutinized for missing data. A total of 10 MOS responses were missing on average two items. For these 10 responses, the missing values were replaced with the average of remaining items. Three respondents with valid PAM scores did not answer any or most MOS questions. These were given a score of 0 for MOS dimensions. The average of raw scores in each of four subscales (EMI, TAN, AFF, POS) constituted that dimension's score. The computed average of 19 items provided an overall index of support. The raw subscales and the overall scale were transformed to a 0-100 scale.

Data analysis. The demographic data and calculated PAM-13 and MOS-SSS scores were imported into the SPSS statistical package version 24 in preparation for statistical analysis. Univariate analyses were conducted on continuous and categorical variables. Descriptive statistics (mean and standard deviation, minimum, maximum, range, and skewness) for continuous variables, and frequency and percentage for categorical variables, and appropriate tabular representations were generated. Distributions of data were checked for normality, and non-parametric tests were employed where a normal distribution could not be shown.

In order to understand possible predictors for involvement, existence of relationships among the clinical variables and PAM levels, among MOS dimensions and PAM levels, and MOS dimensions and support group participation were explored. For these bivariate analyses, cross-tabulation and a Chi-square test of independence for significance associations among variables were performed, and appropriate tests to measure the strength of relationship, when one existed, were applied. These tests included Goodman and Kruskal's Gamma (γ) for clinical variables.

Clinical variables were dichotomous variables indicating the presence or absence of symptoms or organ involvement. Dichotomous variables can be treated at any measurement level: categorical or interval. To treat them as ordinal and the choice of Gamma as association measure were warranted by small table size (2X2) and the sample size of (n=183). Whereas Gamma overestimates the degree of ordinal association when sample size is fairly large (over 250), for smaller samples and table sizes it represents a closer estimate of the association than other estimates such as Pearson r, Spearman's rho, Kendall's Tau-b or Tau-c, and Somers' d (Goktas & Isci, 2011). In addition, Gamma is a symmetrical measure of association where its value is not based on identification of dependent or independent variables. Gamma has a range/direction of -1 to +1 and is defined as: (Ns- Nr)/(Ns+ Nr), where Ns is the number of same ordered pairs (concordant) and Nr (discordant) is the number of inverse ordered pairs. As a

Proportional Reduction of Error (PRE) measure, its value is interpreted as the improvement (reduction in prediction error) in predicting the dependent variable, knowing the values of the independent variable. The following ranges of magnitude were used for interpretation of results: .00-.24 “no relationship”; .25-.49 “weak relationship”; .50-.74 “moderate relationship”; .75-1.00 “strong relationship.”

To explore the association, if any, between PAM activation levels and age, educational attainment, employment, disease duration, disease subtypes, presence of certain symptoms, and other demographic and clinical variables, the Chi-square test of independence was performed. The nominal and ordinal nature of variables, the unequal groupings of categories, skewed distribution of data, and mutually exclusive and independent categories made this choice appropriate. In order to meet the expected cell values assumption of the Chi-square test, the following categories were collapsed: age category was collapsed into four groupings (18-49, 50–59, 60-69, >70), the employment category into two groupings (employed, not gainfully employed), education category into three groupings (some high school to some college, college graduate, postgraduate), disease duration into three groupings (1 to <5 years, 5-10 years, >10 years), number of doctors seen into three groupings (<=3, 4-5, >5), and four groupings for how far traveled to see a SSc expert (<24 miles, 25-99 miles, >100 miles, not seeing an expert).

The Chi-square test was followed by calculation of Somers’ d (d) as a measure of strength of association. The activation level was the dependent variable. The measure Somers’ d is given as d= (Ns –Nr) / (Ns+ Nr + TD), where Ns is the number of concordant pairs, Nr is the number of discordant pairs, and TD is the number of pairs with tied ranks on the dependent variable. Somers’ d is an asymmetric measure with the magnitude range of -1 to 1, and is an appropriate choice when a distinction between a dependent and an independent variable is made.

To test how the four PAM activation levels differed in degree of social support, as indicated by MOS variables, the Kruskal-Wallis H test was performed. The Kruskal-

Wallis test, also known as one-way ANOVA on ranks, is a non-parametric test for determining differences among groups of an independent variable and ordinal or continuous dependent variables. This test is used when assumptions of one-way ANOVA do not hold. ANOVA's assumptions include normality, no outliers, and balanced sample sizes.

To test for normality, the Shapiro-Wilk test was performed on MOS data, grouped by PAM levels. Only the distributions of overall support and EMI for level 1 and level 2 showed approximately normal distributions. Other dimensions, in particular TAN and POS scores in levels 3 and 4, were found to be skewed, with a larger number of participants claiming a high degree of support. The sample size for each activation level was also not balanced. Since the normality assumption did not hold across the board, and the sample size of level 3 was almost 3 times that of level 1, the Kruskal-Wallis H test was chosen instead. In this test, PAM activation was treated as the independent variable (grouping variable) and MOS variables as dependent for the purpose of determining if there was a difference in medians of MOS variables among four activation levels.

The Kruskal-Wallis (1952) H test ranks each score of the dependent variable based on its value, without regard to its grouping. After this ranking is done for all scores, scores are summed and averaged within the groups they belong to, and a one-way ANOVA is applied to ranks (as opposed to the original observations). Formally the H statistic is calculated as:

$$H=\frac{12}{N(N+1)} * \left(\sum_i^n T_i^2/_n\right) - 3(N+1)$$

where T is sum of each group's rank, n is the sample size in each group, and N is the total sample size. For number of groups (k) greater that three, and for five or more observations in each group, H approximates a Chi-Square distribution with degree of freedom (d = k-1). The acceptance or rejection of the null hypothesis is reported based on Chi-square critical value. The Kruskal-Wallis tests the similarity of distributions among

groups. If we can assume the distributions among groups are the same (i.e., groups have the same shape and dispersion and are only shifted up or down relative to a center point, i.e., median), then the null and alternative hypotheses may be stated as follows:

H0: The population median among groups are equal.

Ha: The population median among groups are not equal.

Using boxplot and frequency histograms, distributions of MOS variables, among four PAM levels, were visually compared. While the level 3 group showed the highest frequencies, the shape and dispersions for all MOS variables were similar for all groups, as assessed by this visual inspection. Based on this determination, comparison of median scores was considered. When the Kruskal-Wallis test is significant and there are more than two groupings in the independent variable, a follow-up test to compare pairwise differences in the groups is usually conducted to determine which two groups are different from the others. Subsequent to the Kruskal-Wallis test, pairwise comparisons were conducted in SPSS. SPSS uses Dunn's (1964) procedure, which includes Bonferroni correction.

Qualitative data. Qualitative data consist of unstructured text. The data from interviews were converted to textual format through a transcription process. In qualitative analysis, each transcript represents one data source, and it is examined individually by the identification and classification of texts of interest therein (words, sentences, or paragraphs). Additionally, categories and concepts across all data sources are pulled together for comparison. One way to approach this process is through thematic analysis, defined as an approach to "identify commonalities and differences in qualitative data, before focusing on relationships between different parts of the data, thereby seeking to draw descriptive and/or explanatory conclusions clustered around themes" (Gale, Heath, Cameron, Rashid, & Redwood, 2013, p. 2). A strategy to make this process more methodical was developed by Ritchie and Spencer at the National Centre for Social Research in U.K. in the 1980s, known as the Framework Method. This method allows for

management of data and its structured representations and organizing, so data reduction and analysis may be performed in a more systemic way.

The proposed analytical structure of Framework Method provides for three hierarchical stages or "viewing platforms" (Ritchie & Lewis, 2003, p. 213) to view and make sense of data, in progressively abstract terms. The first stage deals with development of a coding matrix, and assignment of data to themes and categories. The second stage is about refining themes and categories, and finding associations among themes; the third stage is associated with developing associations or patterns among concepts (Smith & Firth, 2011). These stages are by no means linear. Rather they are bi-directional, allowing for movements up and down to refine and link concepts among levels of abstraction. Notwithstanding these backward and forward iterative movements among stages, the essence of each stage is delineated below.

- Stage 1: Data management
    - Creating a coding index: A subset of transcripts is carefully reviewed (open coding) to generate an index to identify initial labels or codes for texts of interest in transcripts, for the purpose of devising a coding index. Pre-defined codes based on a conceptual framework may be used instead, or in conjunction with open coding.
    - Creating a working analytical framework: Sorting and grouping of codes into broader categories and main themes to create a working analytical framework in order to index (code based on framework) all data sets.
- Stage 2: Descriptive Accounts
    - Indexing of data set: The entire data set is indexed by using the working analytical framework. The framework is refined with each transcript coded in an iterative process, and is a fluid work in progress until the last transcript is indexed.

    - Creation of framework matrix: A framework matrix (a worksheet made of sample as rows, and categories as columns) is created; data is summarized (vs. verbatim text) by category for each transcript. References to illustrative quotations are retained.
    - Detection of typologies: Association and typologies may be detected by examining non-overlapping categories and identifying discriminant factors or dimensions
- Stage 3: Explanatory Accounts
    - Explanatory account: Explanatory accounts are developed based on patterns of associations or clustering of themes.

The above approach was adopted in this study.

Data set creation and reduction. All recorded interviews were transcribed and uploaded to MAXDQA software. Each interview was read several times in order for the researcher to become fully familiar with the transcripts. A handful of transcripts representing variations in transcripts were selected for coding. Starting with an initial and provisional coding index based on literature, these transcripts were coded. Codes related to contextual and background information surrounding diagnosis and each individual, absent in the initial coding index which relied on literature, were added as emergent codes. Subsequently, the resulting working analytical framework was applied to the entire data set of 25 interviews in an iterative process, going back and forth between transcripts, as new categories emerged and categories collapsed. Along the way, analytic memos ("the theorizing write-up of ideas about codes and their relationships as they strike the analyst while coding," Glaser, 1978, pp. 83-84) were written. These included references to other transcripts, outlines of emerging typologies, and nuances or emerging features that seemed to define or characterize a code/category and its variations. MAXDQA provides for recording of memos per transcript and per category, allowing

observations and insights to be saved and associated with categories and transcripts in a systematic and convenient manner.

Once the iterative process of indexing was deemed to have reached a satisfactory saturation and ran its course, the framework matrix was created. This was achieved conveniently in MAXDQA through the Summary Grid feature, providing a mechanism for summaries to be written for each coded segment. The wordings of summaries were kept as close to the original text as possible. At this stage, the data were reduced considerably and categorized, allowing for patterns and associations to become apparent, and typologies discovered, leading into an explanatory and interpretive process. These typologies are presented in Chapter IV. One such example is the modes of doctors' interaction as perceived by patients, graphically shown in Figure 3.3. Additionally, a sample index and a sample framework matrix, resulting from the first two stages of the framework method, are provided in Appendices I and J.

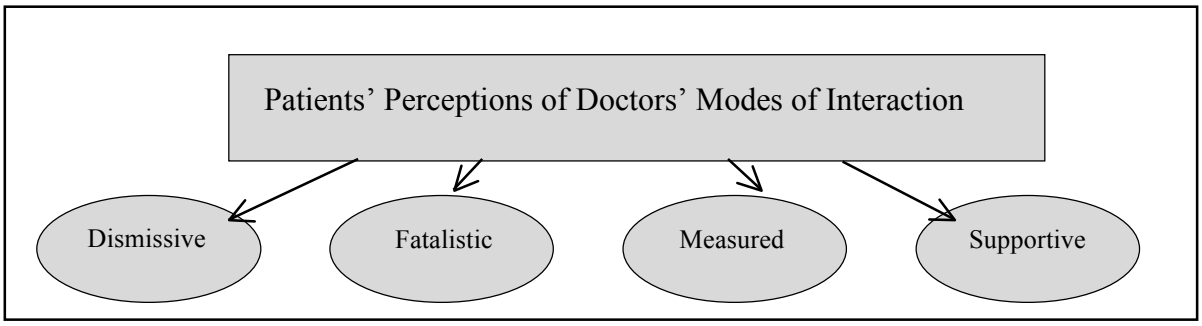

Figure 3.3. Example of a typology seen in data.

Illustrative texts and participants' own words from transcripts were included to support the reporting of findings. In order to safeguard the anonymity of participants, demographics and descriptive data were presented in aggregate; data tables were ordered by different variables to remove the possibility of linking them to each other. In addition, to protect the identities of interview participants, pseudonyms were used, and identifiable information was removed whenever participants were quoted.

Analysis and synthesis. The categories, themes, and typologies obtained from stage two provided the descriptive accounts of the phenomenon under consideration, by making explicit the constituent components. To arrive at explanatory accounts, as Miles and Huberman (1994) quote Bernard, requires showing how these "component parts fit together according to some rule" (p. 90). These "rules" may come from participants' explicitly stated logic and reasoning; from the researcher's inferences based on evidence from data, common sense, or hunches; from other empirical research, or more formally from theoretical frameworks (Ritchie & Lewis, 2003). The goal is to decide on general findings that hold true for the data and examine them in light of the literature (Miles & Huberman, 1994). The problem of how components that were found fit together was tackled through all these means, "presentation of evidence," and "consideration of alternative interpretations" (Yin, 2009, p. 127). Bloomberg and Volpe (2012) suggest to make this process explicit by:

- Providing answers to research questions through findings
- The synthesis of quantitative and qualitative data
- Showing the relevance of findings in the context of literature
- The appraisal of findings relative to researcher's initial assumptions.

These recommendations were followed and are presented in discussions and analysis, in Chapter V.

Synthesis of quantitative results and qualitative findings. In a convergent mixed-methods design, the integration of the qualitative and quantitative data occurs when results from the respective methods are brought together and compared (Creswell, 2015). One approach consists of discussing the results of each method, in sequence, in the discussion section of the study. This approach was taken in this study. Additionally, quantitative findings were elaborated and clarified by interview findings and vice versa, whenever such a mixing was appropriate.

Ethical Considerations

As part of the institutional Review Boards (IRB) approval requirements, safeguards regarding informed consent and the protection of the rights of the subjects were implemented. Statements of participant's rights and informed consent for survey respondents were developed and included in the recruitment packet. The personally identifiable information and certain demographics related to race/ethnicity and geography were not collected from the survey participants. The completed surveys were collected through a dedicated post office box accessible only to the researcher, and the results of the surveys were reported as aggregates.

Those interested in being interviewed were given the option of keeping their survey anonymous by mailing in their contact information separately. Statements of participants' rights and informed consent for interview respondents were shared with interviewees, and written consent for conducting the interviews and audio recording was obtained prior to the interviews. Each participant in the study was assigned a pseudonym for confidentiality purposes and was identified as such in the research analysis and reporting. Any identifying references to the individuals disclosed during interviews was masked in reporting. Transcripts, coding, and data material were stored in a private, secure space, and digital files were password protected. Confidentiality was maintained by reports of aggregate data. Consent forms and statements of participants' rights are included in Appendices A-C.

Issues of Trustworthiness

Trustworthiness of a study concerns the validity or credibility and dependability or reliability of results. Quantitative studies deal with threats to validity through controls built into the design before research commences, while in qualitative designs, most threats are addressed after the start of fieldwork and when tentative accounts are formed (Maxwell, 2005). Determination of validity is obtained through presentation of evidence and theory to support the interpretations of results or findings.

Validity and reliability related to results and assertions in this study are presented separately under quantitative and qualitative headings.

Validity and reliability of the quantitatively obtained results. Trustworthiness in quantitative fixed designs encompasses construct validity (measuring of what is intended), reliability of measure (measuring the same consistently), and external validity (extent of generalizability).

Three categories of data were collected in the quantitative portion of this study; two categories were collected through the use of PAM-13, MOS-SSS instruments, and for the third category, the demographic data including disease state and illness perceptions (depression, pain, fatigue, etc.) were self-reported.

Construct validity and criterion validity analysis, conducted by the developers of the PAM measure, based on hypothesis related to the expected behavior consistent with high activation (e.g., engaging in self-care, seeking information, comparing medical options, having better functioning, less health fatalism, etc.) indicated considerable construct validity (Hibbard et al., 2004). Packer et al. (2016), using measures of health status and lifestyle (diet, activity, etc.), reported both a strong internal consistency, and a strong construct validity through comparison with these measures.

On the other hand, Graffigna et al. (2017) showed a correlation between the PAM-13 and her PHE (Patient Health Engagement) scale, indicating an overlap of activation and engagement constructs. Rademakers et al. (2012) reported a weak to moderate correlation between PAM and health literacy, a suspected factor in high activation, and concluded that more research in PAM validity with respect to health literacy is warranted. These highlight the ambiguities in drawing boundaries around concepts of activation, engagement, and involvement, bringing to mind Forbat et al.'s (2009) characterization of "the conceptual muddle with which involvement is articulated, understood and actioned" (p. 2553). In this dissertation, PAM is used as an approximation to patients' commitment and involvement to manage their illness, while

acknowledging PAM covers a specific subset of such a concept. The inherent difficulty in defining and operationalizing concepts such as activation, involvement, and engagement was explored previously and was at the heart of the rationale for conducting this study. The difficulty in establishing construct validity in a clear and all-encompassing way is viewed as giving credence to the argument and purpose of this study.

A source of validity evidence in use of scales is the "Response Process" to the instrument. Response process is defined as "actions and thought processes of test takers" (Cook & Beckman, 2006, p. e10) while taking the test, and is looked into through a "think aloud" approach. A number of participants in this study had written notes and comments on the margins of this instrument, indicative of their thought processes and their ambivalence with some items. Specifically, items 3 and 12 were more difficult to endorse, while items 7 and 6 were easier than expected to endorse. Other studies (Packer et al., 2016; Staerk, 2015; Stepleman, et al., 2010) have reported similar variations in scaling. In spite of such variations, these studies concluded that PAM is a reliable and valid instrument in assessing patients' activation. The notion of the response process of this instrument was taken into consideration in interpreting the results.

PAM-13 has shown to have high reliability across different levels of self-reported health status, age, race, education, household income, and nine chronic conditions (Hibbard et al., 2005). A high internal consistency with Cronbach's α of above .84 has been reported by others in a wide range of chronic illnesses (Brenk-Franz, Hibbard, Herrmann, Freund, & Szecsenyi, 2013, in German patients; Moljord et al., 2015 in mental health; Packer et al., 2016 in neurological illnesses; Rademakers et al., 2012, in Dutch patients; Skolasky et al., 2011, in multi-morbid older adults; Stepleman et al., 2010, in MS).

The second instrument used in this study, MOS-SSS, has shown strong validity through confirmatory factor analysis of four dimensions (Sherbourne & Steward, 1991). The four dimensions were shown to be highly correlated (.69-.82), while number of

family or friends indicated low/moderate correlations (.19-.24) with these dimensions. Other factors such as family/marital functioning, mental/physical health, and social activity were positively correlated, while loneliness had a negative correlation. Other studies in patient populations (Kettman & Altmaier, 2008, in bone-marrow transplant patients; Rodin et al., 2007, in cancer) have shown a negative relationship between social support and depression. Sherbourne and Stewart (1991) have reported reliability coefficients of .97, .96, .92, .91, and .94 for overall support, EMI, TAN, AFF, and POS dimensions, respectively.

The third category of data collected was self-reported information as to the existence of pain, depression, fatigue, sleep disturbance, body image stress, existence of comorbidity, and involvement of organs for the purpose of assessing disease severity. The self-reported nature of these data makes them susceptible to inaccuracies. At the same time, the anonymous design of the survey makes an intentional misreporting unlikely. The validity threats of this type may be avoided by drawing the sample from a medical setting, where access to clinical findings and official health status are available. Additionally, the presence and levels of many illness complaints, including those listed above (e.g., pain, depression, fatigue, etc.), may be measured by related instruments.

Access to a sample from a medical setting was not available to this researcher, and inclusion of more instruments in the survey packets was deemed impractical and counter-productive, given the degree of hand-disabilities in this population, and the increased risk of non-participation due to the length of survey. To mitigate some of the uncertainty about these data, an extensive review of studies reporting on these variables using scales in scleroderma patients was conducted to assess the presence of possible deviations and anomalies. The reported percentages in this dissertation fell within the ranges reported in other studies, with the exception of a slightly higher lung involvement reporting. Hypothesis as to this occurrence, as well as details related to other variables, are provided in Chapter IV.

In a cross-sectional study, causal inferences are not supported, and the results are considered descriptive of the sample. However, given the shared characteristic among chronic disease patients discussed previously, it is expected the results to be applicable to involvement in other illnesses as well.

Credibility and dependability of the qualitatively obtained assertions. Maxwell (2005) contends that two main validity threats in qualitative studies are the selection of a sample that fits the researcher's bias and preconceptions, and the influence of the researcher on study subjects (reactivity), making alternative hypotheses or rival explanations plausible. Therefore, the main task in dealing with validity issues in qualitative studies is to account for these threats to reduce the plausibility of other explanations, in order to increase confidence in interpretations, and lend credence to the reasonableness of the conclusions drawn. Among suggested strategies (Maxwell, 2005; Miles & Huberman, 1994), the following were utilized to reduce validity threats:

- Triangulation—collecting a diverse range of data by "data source, by method, by research, by theory, by data type" (Miles & Huberman, 1994, p. 267): Triangulation was incorporated by inclusion of a large number of participants (25), two types of data (qualitative, quantitative), considerations of theories from various fields of study, and obtaining inter-rater reliability through coding of transcripts by another colleague.
- Collection of rich data ("data that are detailed and varied enough that they provide a full and revealing picture of what is going on," Maxwell, 2005, p. 110): Inclusion of open-ended questions, allowing for participants to tell their "stories" and particularly the recounting of specific instances and examples resulted in a rich set of data, and providing safeguards against a "uniform mistaken conclusion" (p. 110).
- Checking for representativeness by looking for outliers, observing the sample in other venues; increasing the number of the sample, including negative and

extreme cases, and stratifying the cases: Stratifying the sample based on support group membership was a strategy to increase the diversity of sample selection. Characteristics of outliers were noted in drawing interpretations. Scleroderma patients were observed in two national conferences and multiple educational forums. The number of participants for this interview (25) was twice as many as in most qualitatively conducted studies.

- Getting feedback from informants/member check: Interview-participants were given an opportunity to comment on findings, and those comments were considered in the interpretation of results. The summary of the interview findings along with the cumulative trajectory was emailed to all interview-participants. Eight participants acknowledged the receipt of the email, and four included substantial feedback. These comments are included in the reflections section in Chapter V.
- Checking for researcher's effects: "Researcher threatens or disrupts ongoing social and institutional relationships, insiders do not want outsiders in on most troubling/negative aspects" (Miles & Huberman, 1994, p. 265). The researcher was sensitive to the relationships between Scleroderma the Foundation, Tri-States and its members and leaders. Use of the Foundation's envelopes for mailing provided for some level of official sanction for the study. Disclosing the researcher's interest in scleroderma due to having a relative with the disease facilitated the sharing of information.
- Bracketing researcher's assumptions: When it came to the researcher's assumptions and beliefs, having a relative with the disease was a double-edged sword. On the one hand, familiarity with the disease and an "insider" status provided advantages. On the other hand, assumptions of the researcher were formed by witnessing the lived experiences of one patient, most likely not typical of other patients. Journaling thoughts and assumptions, and discussing

these assumptions with the relative, observing other patients in educational forums, and hearing the staff at the Scleroderma Foundation, Tri-State, made these assumptions explicit; the researcher was cognizant of their impacts on the interpretations of results and checked against alternative interpretations.

Reliability in qualitative studies has to do with reproducibility of the study's results, and existence of an audit trail. To increase the reliability of this study, procedures, including sampling methods, interview protocol, and framework method, were delineated in detail to allow for duplication of study.

External validity in qualitative studies, in the sense that it exists in quantitative studies, does not apply. However, Miles and Huberman (1994) suggest a number of features that may be considered to assess a broader application of the findings, including the full description of the sample to allow comparison to other samples, a theoretically diverse sampling to allow for broader applicability, congruency of results with prior theory and experiences of a range of readers, and generic descriptions of processes and outcomes to provide applicability to other settings. Attempts were made to provide this level of detail in writing this dissertation.

Limitations and Delimitations

The delimitations of this study extend to a number of areas. Among the six broad categories that patients take an active role and are involved in, this study focuses only on the concept of patients' involvement in their own care, such as engagement with health information, participation in health care decisions, self-monitoring, etc. Furthermore, this study is an inquiry into involvement of a select group of patients, mostly middle-aged women with a rare disease living in three states in the northeast U.S. The rarity of the disease and certain unique characteristics of the disease may make the experience of living with this disease and the nature of involvement different, at some levels, from other chronic diseases.

A number of potential limitations exist for this study: Within the population of scleroderma patients, there is a wide variation in disease presentation, duration, and severity. It is possible that the full range of such presentations was not reflected in the study sample. In addition, no information regarding race/ethnicity or income was collected. The effect of these factors on the study's results are unknown. Additionally, this study, in part, relies on the recollection of events and reporting of symptoms. Such self-reporting is liable to inaccuracies. The two instruments used for data collection have been validated with patients with some autoimmune diseases, but not specifically with scleroderma patients, posing validity and reliability threats. Furthermore, the correlational design of the quantitative portion does not allow for interpretation of associations among variables as causal relationships, and generalizability of results beyond the study sample might be problematic. Lastly, the researcher witnessing a relative with the disease might have introduced certain assumptions and biases into study's design and interpretation of results. Efforts to minimize the effects of some of these limitations are described in the trustworthiness section in this chapter.

Summary of Chapter III

It is the researcher's belief that patient involvement in their own care is best understood when data from the perspectives of individuals and those from collective beliefs and behaviors of the population are brought together; a mixed-methods approach best serves this purpose and was an appropriate choice, given that scleroderma has varying manifestations, the population is small and difficult to access and draw from, and the availability of validated scales is limited. A modified parallel design was adopted, and points at which the integration of data and results occurred were noted. A descriptive/ exploratory approach for the qualitative portion and a non-experimental fixed

correlational design for the quantitative portion were chosen, and the rationales for both were given.

The sampling processes were delineated, and the study participants were described as 212 survey respondents, 86 of whom expressed an interest to be interviewed. A stratified random selection of 25 participants from this group constituted the interview sample. The information needed to conduct the study was outlined, and descriptions of data collection methods, including scales, survey, and interviews, were given. Methods for analysis of data were described for quantitative and qualitative data, and strategies for synthesis of data were outlined. Lastly, issues of trustworthiness, ethical considerations, and limitations and delimitations of the study were addressed.

Chapter IV

RESULTS AND FINDINGS

The quantitative results and the qualitative findings of this study are presented in this chapter. The quantitative results concerning survey data, including those of MOS and PAM measures, are reported first. The primary utilization of these results is in elaborating answers to research questions 1 and 3. These results obtained from a larger pool of 201 survey respondents impart a more expansive view of the population of this study, provide a quantitative measure of their levels of activation and engagement, and make explicit associations among activation and social support, and demographic and clinical attributes. Subsequent to presentation of the quantitative results, the qualitative findings are reported. These findings provide answers to research questions from the perspectives of 25 interview participants.

This chapter is organized in two main sections and a number of sub-sections.

- Quantitative results
    - Demographic, clinical characteristics, and support group participation
    - PAM-13 and MOS-SSS results
    - Bivariate analysis
    - Summary of quantitative findings
- Qualitative findings
    - Contextual description
    - Patients' work

- Strategies
- Learning
- Motivation
- Social interactions
- Summary of qualitative findings

Quantitative Results

A total of 211 respondents (23%) out of 925 scleroderma patients with presumed valid addresses responded to the mailed survey packets. After elimination of disqualified returns, 201 responses with complete demographic information were retained and, henceforth, are referred to as respondents (n=201). Additionally, from among the 88 (43.8%) of these patients who expressed an interest to be interviewed, 25 (28.4%) were chosen through a stratified random selection for interview, and henceforth are called interview-participants (n=25). The remaining 176 participants who were not interviewed are labeled as survey-participants (n=176).

Demographics and Clinical Characteristics

The demographic and clinical characteristics for all three categorizations of respondents (n=201), survey-participants (n=176), interview-participants (n=25) are tabulated in Table 4.1.

Table 4.1

Demographic and Clinical Characteristics

Variable	Respondents (n=201)	Survey-Participants (n=176)	Interview-Participants (n=25)
Age Range, years			
18-29	4 (2%)	4 (2%)	0 (0%)
30-39	10 (5%)	9 (5.1%)	1 (4%)
40-49	25 (12.4%)	20 (11.4 %)	5 (20%)
50-59	59 (29.4%)	53 (30.1%)	6 (24%)
60-69	52 (25.9%)	43 (24.4%)	9 (36%)
70-79	37 (18.4%)	34 (19.3%)	3 (12%)
>=80	14 (7.0%)	13 (7.4%)	1 (4%)
Gender			
Female	180 (89.6%)	160 (90.9%)	20 (80%)
Male	21 (10.5%)	16 (9.1%)	5 (20%)
Education			
Some H.S.	2 (1%)	2 (1%)	0 (0%)
H.S. Grad	37 (18.4%)	33 (18.8%)	4 (16%)
Some College	27 (13.4%)	22 (12.5%)	5 (20%)
College Graduate	69 (34.3%)	63 (35.8%)	6 (24%)
Some Post Graduate	13 (6.5%)	12 (6.8%)	1 (4%)
Postgraduate Degree	53 (26.4%)	44 (25%)	9 (36%)
Employment			
Full-time	54 (26.9%)	47 (26%)	7 (28%)
Part-time	17 (8.5%)	16 (9.1%)	1 (4%)
Homemaker	11 (5.5%)	11 (6.3%)	0
Student	0	0	0
Retired	70 (34.8%)	64 (36.4%)	6 (24%)
Disabled/On Sick Leave	44 (22.0%)	36 (20.5%)	8 (32%)
Unemployed	3 (1.5%)	1 (0.6%)	2 (8%)
Others	2 (1.0%)	1 (0.6%)	1 (4%)
Patient-Reported Disease Subtype			
Localized	19 (9.5%)	19 (10.8%)	0
Limited (lcSSc)	87 (43.3%)	76 (43.2%)	11 (44%)
Diffuse (dcSSc)	69 (34.3%)	60 (34.1%)	9 (36%)
Sine SSc (ssSSc)	10 (5.0%)	7 (4.0%)	3 (12%)
Do not know	16 (8.0%)	14 (8.0%)	2 (8%)

Table 4.1 (continued)

Variable	Respondents (n=201)	Survey-Participants (n=176)	Interview-Participants (n=25)
Time since diagnosis of SSc			
<= 12 months	0	0	0
1 to < 2 years	8 (4.0%)	8 (4.5%)	0
2 to < 3 years	10 (5.0%)	8 (4.5%)	2 (8%)
3 to < 5 years	23 (11.4%)	19 (10.8%)	4 (16%)
5 to < 8 years	21 (10.4%)	19 (10.8%)	2 (8%)
8 to 10 years	10 (5.0%)	8 (4.5%)	2 (8%)
> 10 years	129 (64.2%)	114 (64.8%)	15 (60%)
Number of doctors			
1	13 (6.5%)	13 (7.4%)	0
2-3	68 (33.8%)	59 (33.5%)	9 (36%)
4-5	69 (34.3%)	59 (33.5%)	10 (40%)
>5	51 (25.4%)	45 (25.6%)	6 (24%)
Organ Involvement*			
Skin	152 (75.6%)	134 (76.1%)	18 (72%)
Gastrointestinal	142 (70.7%)	125 (71.0%)	17 (68%)
Joints	119 (59.2%)	103 (58.5%)	16 (64%)
Lung	105 (52.2%)	90 (51.1%)	15 (60%)
Heart	55 (27.4%)	50 (28.4%)	5 (20%)
Kidney	21 (10.5%)	19 (10.8%)	2 (8%)
Others	26 (12.9%)	22 (12.5%)	4 (16%)
Symptoms & Impact*			
Raynaud	167 (83.1%)	146 (83%)	21(84%)
Fatigue	160(79.6%)	138 (78.4%)	22(88%)
Pain	139 (69.2%)	119 (67.6%)	20 (80%)
Sleep Disturbance	119 (59.2%)	104 (59.1%)	15 (60%)
Body Image Stress	77 (38.3%)	67 (38.1%)	10 (40%)
Depression	68 (33.8%)	56 (31.8%)	12 (48%)
Co-morbidity			
Yes	69 (34.3%)	61 (34.7%)	8 (32%)
No	120 (59.7%)	103 (58.5%)	17 (68%)
Not sure	12 (6.0%)	12 (6.8%)	0

* - Participants could select multiple organs and symptoms. Responses do not add up to 100%.

Among all respondents (n=201), 7% were below the age of 40 and another 7% over the age of 80, totaling 28 patients. Nearly one-third of all patients were 50-59 year-olds, followed by another one-fourth in the 60 to 69 age range. Thus, the 50-69 age group, together, constituted over half (55.3%) of the respondents. The respondents were overwhelmingly female, 180 (89.6%) vs. 21 (10.5%) male. They had an above-average educational attainment, with 67.2% having a college degree or above, and only 19.4% having a high school diploma or less. In terms of employment, 56.8% were either retired

or on disability or sick leave; about one-third were gainfully employed (26.9% full-time; 8.5% part-time); and the rest were homemakers, unemployed, or "others."

The prevalence of self-reported disease subtypes among respondents included 9.5% localized; 43.3% limited cutaneous scleroderma (lcSSc); 34.3% diffuse cutaneous scleroderma (dcSSc); 5.0% scleroderma sine scleroderma (ssSSc); and 8.0% did not know their disease subtype. About 9% of them were diagnosed less than 3 years prior to the time of survey, 26.8% between 3 and 10 years, while 64.2% had lived with the disease for over 10 years. Some 34% of respondents indicated co-morbidity, 60% indicated none, while 6% did not know.

About one-third of respondents, 33.8% (68), saw 2 to 3 doctors; another one-third, 34.3% (69), saw 4 to 5 doctors; and a quarter of patients, 25.4% (51), saw more than 5 doctors for their medical needs. Of these, 35.8% travelled more than 25 miles to see an SSc expert, 44.3% saw doctors locally, and 19.9% did not see a SSc expert at all (most likely saw a general rheumatologist). Of all the respondents, 30.8% (62) knew their SSc related antibodies, 45.3% (91) did not know, and 23.9% (48) were not sure.

Eighty-three percent of all respondents suffered from Raynaud's phenomenon (RP); over 79% had fatigue. Other reported symptoms included: skin issues (76%), gastrointestinal issues (71%), and joints/muscle problems (59%). The lung and heart involvements were reported at 52% and 27%, respectively, while kidney involvement was at 10.5%. Other disease impacts reported were: pain (69%), sleep disturbance (59%), body image stress (38%), and depression (34%). Respondents with the disease duration of less than 2 years reported the highest percentage of having pain (88%) and fatigue (100%), whereas from the second year onward, the pain was lower, with a mean of 66.4% $\pm$ 4.6. At the same time, fatigue fluctuated with a mean of 84.8% $\pm$ 11.6. Similarly, those who indicated their employment status as "disabled or on sick-leave," compared to those who were "employed" or "retired," had substantially more pain (92.5% vs. 67.7%, 50%), and fatigue (95% vs. 75.4%, 75%), and were markedly more

depressed (75% vs. 32.3%, 28%), suffered from a considerable degree of lung (70% vs. 49.2%, 46.7%), heart (47.5% vs.,15.4%, 28.3%), gastrointestinal (82.5% vs., 67.7%, 70%), and joint/muscle (77.5% vs., 50.8%, 56.7%) issues.

When grouped as survey-participants and interview-participants, the latter was slightly younger (20% in 40-49 age group vs. 11.4% of survey group, and 12% in 70-79 age range vs. 19.3% of survey group); more educated (36% post grad degrees vs. 25% of survey group), with a higher number of patients on disability or sick leave (32% vs. 20.5% of survey group). The interview participants' disease duration, number of treating doctors, and major disease subtypes (limited and diffuse) were comparable to the survey participants'. However, the interview participants differed from survey participants in number of patients travelling over 100 miles (presumably to scleroderma specialty health centers) (20% vs. 12.5%), and the number of patients knowing their antibodies (40% vs. 29.5%). In addition, interview participants indicated more fatigue, pain, depression, joint and lung issues, but slightly fewer heart problems and comorbidities than survey participants.

Support Group Participation

When asked about their support group involvements, 67.7% (136) of respondents did not participate in a support group, while 23.9% (48) were support group members and 8.5% (17) were support group leaders. Among survey participants, 23.9% (42) were support group members, 70.5% (124) did not participate in support groups, and 5.7% (10) acted as support group leaders. Among the interview participants, 32% (8) have not participated in any support groups beyond a few trial visits, if any; 16% (4) were associated with support groups in some capacity in the past, but have not been active or participating for a while; 28% (7) acted as support group leaders or co-leaders; and 24% (6) considered themselves members who attended meetings with some regularity. The interview participants were, therefore, considerably more active in support groups, both

as leaders and members, in comparison to survey participants. Details are given in Table 4.2.

Table 4.2

Support Group Participation

Support Group Participation	Respondents (n=201)	Survey-Participants (n=176)	Interview-Participants (n=25)
Not a Member	136 (67.7%)	124 (70.5%)	8(32%) never participated/ 4(16%) did previously
Member	48 (23.9%)	42 (23.9%)	6 (24%)
Leader/Co-leader	17 (8.5%)	10 (5.7%)	7 (28%)

When viewed by employment status, among those who were employed, 4.6% were leaders, 18.5% were members, and 76.9% were non-members. The participation (as leaders, members, and non-members, in that order) for those not-working or retired (6.8%, 25.7%, 67.6%) and those "disabled/on sick-leave" (15%, 30%, 55.8%) were progressively higher. Among all leaders, 42.9% were "disabled/on sick leave," and 35.7% were those who did not work.

PAM-13 Results

As was delineated in Chapter III, after detection and elimination of incomplete and invalid entries, the net number of valid PAM scores available for analysis was reduced to 183. The calculated PAM scores had a range of 66.29 (24.40 to 90.69), with a mean of 59.76 ($\pm$ 11.27) and a variance of 127.11, and subsequently were converted to four PAM activation levels.

The distribution of levels in three sample groups is shown in Table 4.3 and includes 15.8% in level 1, 20.2% in level 2, 48.6% in level 3, and 15.3% in level 4, with a mean of 2.63, median of 3.0, and mode of 3 for respondents. Interview participants' activation consisted of 12% at level 1 and level 4, and 32% and 44% at middle levels of 2

and 3, respectively. In comparison, almost 50% of survey-participants were in level 3, with a nearly equal distribution among the remaining levels.

Table 4.3

Distribution of PAM-13 Activation Levels for Sample Groupings

	Respondents (n=183)		Survey-Participants (n=158)		Interview-Participants (n=25)	
PAM Levels	Frequency	%	Frequency	%	Frequency	%
1	29	15.8%	26	16.5%	3	12%
2	37	20.2%	29	18.4%	8	32%
3	89	48.6%	78	49.4%	11	44%
4	28	15.3%	25	15.8%	3	12%
Total	183	100%	158	100%	25	100%

Excluding those with unknown disease subtype, the majority of respondents fell into level three activation, irrespective of disease subtypes. Those with systemic-diffuse subtype had the highest percentage in level 3 and level 4, combined, as shown in Table 4.4.

Table 4.4

Distribution of PAM-13 Activation Levels by Disease Subtypes

Disease Subtype	Level 1	Level 2	Level 3	Level 4	Total (subtype)
Localized	6.7% (1)	20.0% (3)	60.0% (9)	13.3% (2)	8.2% (15)
Systemic-Limited	19.5% (16)	20.7% (17)	42.7% (35)	17.1% (14)	44.8% (82)
Systemic-Diffuse	11.1% (7)	14.3% (9)	58.7% (37)	17.1% (10)	34.4% (63)
Systemic-Sine	22.2% (2)	11.1% (1)	55.6% (5)	11.1% (1)	4.9% (9)
Do not Know	21.4% (3)	50.0% (7)	21.4% (3)	7.1% (1)	7.7% (14)
Total	15.8% (29)	20.2% (37)	48.6% (89)	15.3% (28)	100.0% (183)

MOS-SSS Results

For all dimensions of MOS-SSS, the following mean scores were observed for respondents: Overall (67.56), TAN (67.01), EMI (62.86), AFF (76.05), and POS (72.68),

with median ranging from 65.6 to 91.7 among all dimensions. All four dimensions showed negative skewness: EMI (-.351), TAN (-.486), POS (-.747), and AFF (-1.073), where 7.7% of EMI, 26.8% of TAN, 35.5% of POS, and 48.6% of AFF displayed the maximum score. Using the median as a cut-off, the reported high support consisted of 50.3% for the overall support, 53% for TAN, 40.4% for EMI, 72.1% for AFF, and 61.7% for POS. Table 4.5 depicts MOS-SSS statistics by the sample's groupings. The perceived overall, TAN, and POS supports for survey-participants and interview-participants were comparable. Interview participants, however, reported higher mean EMI and AFF supports (66.2 vs 62.4, and 79.7 vs 75.5), respectively.

Table 4.5

MOS-SSS Statistics for Sample Groupings

	Respondents (n=183)			Survey-Participants (n=158)			Interview-Participants (n=25)		
MOS Dimensions	M	SD	Median	M	SD	Median	M	SD	Median
Overall	67.6	24.9	72.0	67.2	25.4	71.5	69.5	21.2	74
TAN	67.1	30.3	75.0	67.1	30.7	75	67.3	27.3	75
EMI	62.9	25.6	66.0	62.4	26.3	66	66.2	20.8	63
AFF	76.1	30.5	92.0	75.5	31.2	96	79.7	25.0	92
POS	72.7	27.3	75.0	72.8	27.4	75	72.0	26.8	75

Bivariate Analysis

The bivariate analysis findings are presented by clinical variables, and PAM levels and MOS scores and their predictor variables.

Clinical variables. Among clinical variables, a moderate association between pain and joint/muscle involvement, χ^2 (1, N=183) = 17.418, p < .000 (λ = .591, p < .000); a moderately high association between fatigue and joint/muscle involvement, χ^2 (1, N=183) = 19.706, p < .000 (λ = .708, p < .000); a moderate association between fatigue and lung involvement, χ^2 (1, N=183) = 9.332, p < .002 (λ = .542, p < .002); and a

moderate association between fatigue and heart involvement, χ^2 (1, N=183) = 6.538, $p < .011$ ($\lambda = .634$, $p < .001$) were detected.

Among pain, fatigue, depression, body image distress, and sleep disturbance, the strongest associations were between fatigue and depression, χ^2 (1, N=183) = 15.071, $p < .000$ ($\lambda = .835$, $p < .000$); and fatigue and pain, χ^2 (1, N=183) = 21.021, $p < .000$ ($\lambda = .704$, $p < .000$). In general, pain, fatigue, and depression seemed deeply intertwined and associated with each other. Sleep disturbance was moderately associated with pain, χ^2 (1, N=183) = 16.497, $p < .000$ ($\lambda = .579$, $p < .000$). Body image distress had a moderate association with depression, χ^2 (1, N=183) = 18.737, $p < .000$ ($\lambda = .601$, $p < .000$), and no association with any other physical symptoms and organ involvements. Also of note were lung and heart involvements, which strongly associated with each other (χ^2 (1, N=183) = 34.948, $p < .000$ ($\lambda = .856$, $p < .000$), as PAH (pulmonary arterial hypertension) is a heart and lung combined issue, which many patients identify as a heart problem.

Overall, respondents who had joint/muscle issues had to deal with both pain and fatigue; those with lung and heart issues suffered from fatigue. Fatigue was associated with both pain and depression. In turn, pain was associated with sleep disturbance, and depression with body image distress. The significant associations among clinical characteristics based on Chi-square and Gamma calculations are depicted in Table 4.6.

Table 4.6

Significant Associations among Clinical Variables

	χ^2	df	Significance	Strength Measure λ/ (significance)
Pain				
Fatigue	21.021	1	.000	.704 (.000)
Sleep Disturbance	16.497	1	.000	.579 (.000)
Depression	13.449	1	.000	.611 (.000)
Body Image	5.984	1	.014	.399 (.010)
Joint/Muscle	17.418	1	.000	.591 (.000)
Fatigue				
Depression	15.071	1	.000	.835 (.000)
Lung	9.332	1	.002	.542 (.002)
Heart	6.538	1	.011	.634 (.001)
Joint/Muscle	19.706	1	.000	.708 (.000)
Sleep Disturbance	7.886	1	.005	.491 (.007)
Sleep Disturbance				
Depression	11.028	1	.001	.512 (.000)
Gastro-Intestinal	6.142	1	.013	.389 (.015)
Joint/Muscle	8.505	1	.003	.422 (.003)
Depression				
Body image	18.737	1	.000	.601 (.000)
Joint/Muscle	5.134	1	.023	.358 (.019)
Lung				
Heart	34.948	1	.000	.856 (.000)
Gastro-Intestinal	13.283	1	.000	.553 (.000)
Raynaud	6.678	1	.010	.495 (.010)
Heart				
Gastro-Intestinal	9.859	1	.002	.633 (.000)
Joint/Muscle	12.128	1	.000	.592 (.000)
Kidney	6.557	1	.010	.534 (.035)
Raynaud				
Gastro-Intestinal	20.051	1	.000	.714 (.000)
Joint/Muscle	7.069	1	.008	.493 (.012)
Gastro-Intestinal		3		
Joint/Muscle	15.402	1	.000	.578 (.000)

PAM levels and predictors. The association between PAM activation levels and demographic data (age, education, employment) and clinical data (disease duration and presence of symptoms) was sought. An association between activation and disease duration (years of having the disease), χ^2 (6, N=183) = 16.438, $p < .012$ was seen.

However, the strength of this relationship was not statistically significant. When disease duration of less than 10 years, where more granular data for number of years having the disease were available, was scrutinized, an association, χ^2 (3, N=68) = 11.601, p < .009, with the strength of the relationship given by Somers' d (d =.371, p < .004) was detected. This statistically significant relationship between independent variable years of having the disease and dependent variable PAM activation levels indicates that within the first decade of disease, the longer disease duration was moderately associated with higher PAM activation levels.

A weak association was seen between activation and educational attainment, χ^2 (6, N=183) = 15.664, p < .016 (d = .123, p < .074). No significant associations were found between activation levels and other demographic variables (gender, age, employment), nor the clinical variables (disease subtypes, number of doctors seen, or knowledge of antibody, or any of symptoms and organs involved).

MOS dimensions and predictors. When MOS dimensions were dichotomized, using the median as the cutoff for high and low values, only the association between PAM activation levels and TAN did not reach the level of significance: Overall support: χ^2 (3, N=183) = 14.268, p < .003 (λ = .405, p < .000) ; EMI: χ^2 (3, N=183) = 8.137, p < .043 (λ = .336, p < .003); POS: χ^2 (3, N=183) = 9.368, p < .025 (λ = .235, p < .044); AFF: χ^2 (3, N=183) = 12.002, p < .007 (λ = .328, p < .003).

To test how respondents in the four PAM activation levels (level1, n=29; level 2, n=37; level 3, n=89; and level 4, n=28) differed in degrees of social support, as indicated by MOS variables, the Kruskal-Wallis H test (one-way ANOVA on ranks) was performed based on the following hypothesis:

H_0: The population median among groups (PAM levels) are equal.

H_a: The population median among groups (PAM levels) are not equal.

For all MOS measures, the median scores were significantly different across activation levels: Overall support: χ^2 (3) =15.809, p =.001; EMI: χ^2 (3) =14.511, p =.002;

TAN: χ^2 (3) =8.216, p =.042; AFF: χ^2 (3) =11.900, p =.008; POS: χ^2 (3) =13.174, p =.004. Therefore, the null hypotheses were rejected.

When the Kruskal-Wallis test is significant and there are more than two groupings in the independent variable, a follow-up test to compare pairwise differences in the groups is usually conducted to determine which two groups are different from the others. Subsequent to the Kruskal-Wallis test, pairwise comparisons were conducted using SPSS. Statistically significant differences were observed as a result. The overall MOS median score of level 4 was significantly higher than level 1 and level 2. The EMI score of level 4 activation was significantly higher than level 1 and level 2, and the POS median score of level 4 was significantly higher than level 2: Overall MOS median scores between level 1 (73.62) and level 4 (118.52), p (.008); Overall MOS median scores between level 2 (73.70) and level 4 (118.52), p (.004); EMI median scores between level 1 (71.05) and level 4 (117.1), p (.006); EMI median scores between level 2 (77.35) and level 4 (117.18), p (.016); POS median scores between level 2 (72.07) and level 4 (111.7), p (.013). No other significant differences were detected among any other level pairings.

When MOS dimensions were tested against depression, pain, fatigue, body image, stress, or sleep disturbance, all MOS dimensions were negatively associated with only depression: Overall: χ^2 (1, N=183) =15.549, p < .000 (λ = -.565, p < .000); TAN: χ^2 (1, N=183) =8.575, p < .003 (λ = -.432, p < .003); EMI: χ^2 (1, N=183) =9.023, p < .003 (λ = -.466, p < .001); POS: χ^2 (1, N=183) =10.048, p < .002 (λ = -.465, p < .002); AFF: χ^2 (1, N=183) =7.232, p < .007 (λ = -.404, p < .006).

When MOS dimensions were analyzed within three categories of "support group leader," "support group member," and "support group non-member," the means of all MOS dimensions were higher for leaders, followed by members, and then non-members, implying the perceived support received by patients in this sample was distinctly different when grouped by membership status. However, the Kruskal-Wallis H test failed to

determine a statistically significant association between support group participation and perceived social support.

Summary of Quantitative Results

Overall, the survey respondents of this study may be described as predominantly female, middle-aged, with an above average educational attainment, retired, or on disability/sick-leave. A majority of the respondents have lived with the disease for over a decade, and more than half did not have the systemic form of scleroderma. More than half were under the care of four or more doctors, and one-third traveled more than 25 miles to see an SSc expert. Only a third was sure about their SSc related autoantibodies.

Clinically, almost two-thirds suffered from RP, fatigue, and skin and gastrointestinal issues, while joint/muscle and lung problems were reported by over half of the sample. Those disabled or on sick leave had more pain, were more depressed, and had more lung, heart, gastrointestinal, and joint/muscle issues than employed respondents.

The interview-participants indicated more fatigue, pain, depression, joint and lung issues, but slightly fewer heart problems and comorbidities than the survey-participants. The interview-participants were also slightly younger and more educated with a higher number on disability or sick leave. Clinical characteristics of both groups were comparable, but interview participants were more likely to travel over 100 miles, presumably to scleroderma specialty health centers, to obtain care, and were more likely to know their autoantibodies.

Among the clinical variables, fatigue and joint/muscle issues, and fatigue and lung and heart involvement had high and moderate associations, respectively. Fatigue and depression, and fatigue and pain each had high association with each other, while pain and sleep disturbance and depression and body image distress, each were moderately associated.

Using the PAM-13 activation measure to describe involvement, 15.8% of respondents were classified as passive recipients of care (level 1), 20.2% of them were lacking knowledge of the disease (level 2), 48.6% of them were knowledgeable but lacked the necessary skills (level 3), and 15.3% of them were active (level 4). Overall, 64% of respondents, in levels 3 and 4, may be classified as having high activation. Those with systemic diffuse subtype had the highest percentage of respondents in level 3 and level 4. A weak association between educational attainment and activation was discerned. A moderate positive association between activation and years of having the disease duration the first decade of disease was also found.

The dichotomized MOS social support scores, with the median as the cutoff for high and low values, indicated 50.3% of respondents perceived having high overall support, 53% had high tangible (TAN) support, 72.1% had high affectionate (AFF) support, 61.7% had high positive (POS) support, but only 40.4% had high emotional/information (EMI) support. Except for TAN scores, the dichotomized MOS scores of all other dimensions had statistically significant associations with PAM activation levels. Furthermore, the perceived overall support, and EMI support for those in level 4 were significantly higher than those in level 1 and level 2. All MOS dimensions were negatively associated with depression.

Almost 70% of respondents did not participate in a support group. Being employed was a barrier to support group participation, and being “disabled/on sick leave” was more conducive to active participation in support groups. No statistically significant association between support group participation and perceived social support was detected.

Qualitative Findings

The aggregate demographic and clinical characteristics of respondents, survey participants, along with those of interview-participants were reported in Table 4.1. Anonymized data for interview-participants, including additional demographic and clinical information gathered at the time of interview, are reported in this section in order to provide an overall context for the interview group and for each participant.

Contextual Descriptions

The 20 female and 5 male interview participants ranged in age from the mid-30s to 85 years old, with 64% having a college degree or above. Those who worked full-time or part-time constituted 32% of the sample, while 56% were retired or on disability or sick leave. Except for two persons not disclosing, all held white-collar jobs such as teaching, accounting, sales, executive/managerial, administrative assistance, or research and health-related work. Ethnically, 72% (18) were White American, 12% (3) White European, 4% (1) African American, 4% (1) Asian American, 4% (1) South American, and 4% (1) Asian. Table 4.7, ordered by age, captures these data.

Interview participants reported their scleroderma subtypes as: 11 (44%) having limited cutaneous scleroderma (lcSSc), including 2 with overlap syndrome (i.e., other autoimmune diseases concurrently); 9 (36%) indicated diffuse cutaneous scleroderma (dcSSc), including 1 with overlap syndrome; 3 (12%) scleroderma sine scleroderma (ssSSc), with one overlap syndrome; and 2 (8%) who did not know the subtype of their disease. Five (20%) of participants were diagnosed when they were young, defined as being in early 20s to mid-30s. Six (24%) were diagnosed in their mid-30s to mid-40s.

Table 4.7

Interview Participants' Demographics, Ordered by Age

Gender	Age	Race/Ethnicity	Education	Employment Status
F	30-39	White European	Post grad	Working full-time
M	40-49	White American	College grad	Working full-time
F	40-49	White American	College grad	Disabled-Sick leave
F	40-49	White American	College grad	Disabled-Sick leave
F	40-49	White American	Some college	Disabled-Sick leave
M	50-59	White European	Post grad	Disabled-Sick leave
M	50-59	South American	Some college	Working full-time
F	50-59	White American	Post grad	Working full-time
F	50-59	White American	College grad	Working full-time
F	50-59	Asian American	College grad	Working full-time
M	50-59	Asian Caucasian	Post grad	Working full-time
F	50-59	White American	Some college	Disabled-Sick leave
F	60-69	White American	Some college	Not working
M	60-69	White American	HS grad	Retired
F	60-69	White American	HS grad	Retired
F	60-69	White American	HS grad	Other
F	60-69	White European	College grad	Not working
F	60-69	White American	Post grad	Disabled-Sick leave
F	60-69	White American	Some post	Disabled-Sick leave
F	60-69	White American	Some college	Disabled-Sick leave
F	60-69	White American	Post grad	Working part-time
F	60-69	White American	Post grad	Retired
F	70-79	White American	Post grad	Retired
F	70-79	White American	HS grad	Retired
F	80 or greate	African American	Post grad	Retired

Ten (40%) were in their mid-40s to mid-50s, and four (16%) were older than 56 when diagnosed. Eleven (44%) participants were diagnosed within six months of seeing their doctors, two (8%) in a year, five (20%) in one and half years, two (8%) in two years, one (4%) in three years, and one (4%) was diagnosed in five years. Two participants were diagnosed due to other health issues unrelated to SSc, and one was self-diagnosed. On

average, these participants have lived with the disease for approximately 14.5 years, with the range given as of 2½ to 49 years (± 10.2). Eliminating the 30 and 49 years of longevity as outliers, the average disease longevity was 12.2 years. Ordered by disease duration, the SSc subtypes and the time taken to be diagnosed are depicted in Table 4.8.

Table 4.8

Interview Participants' Disease Subtypes and Diagnosis Time, Ordered by Disease Duration

Disease Duration	SSc Subtype	Time took to be diagnosed
2.5	ssSSc	Self-Diagnosed
3	dcSSc	1-6 months
3	ssSSc	1-6 months
4	dcSSc	1 year
5	dcSSc	1-6 months
7	lcSSc	1-6 months
7	dcSSc	1 ½ years
10	ssSSc	1-6 months
10	dcSSc	2 years
10	lcSSc	1-6 months
12	lcSSc	1 ½ years
12	dcSSc	1-6 months
12	lcSSc	1-6 months
13	lcSSc	1-6 months
15	ssSSc	1 ½ years
15	lcSSc	Coincidentally diagnosed due to other health issues
16	lcSSc	5 years
19	dcSSc	1-6 months
19	dcSSc	1 year
20	dcSSc	Coincidentally diagnosed due to other health issues
21	lcSSc	1-6 months
22	lcSSc	3 years
25	dcSSc	1 ½ years
30	Don't know	1-6 months
49	Don't know	2 years

As to previous health conditions, 16 (64%) of the participants did not suffer from any chronic health condition prior to their SSc diagnosis, while 4 (16%) participants had other prior health issues. Nine (36%) had developed other conditions subsequent to SSc. Eleven participants (44%) made note of their excellent health status prior to their illness. Five were highly trained athletes at the level of marathon runners and sports trainers, 3

were highly active in sports and outdoor activities, and 3 considered themselves physically active with no prior history of health problems.

Among the 25 interview participants, only 1 indicated ongoing medical issues since childhood. Growing up, 1 had a sibling with an autoimmune chronic condition (not SSc), 1 had a sibling with diabetes, and 3 had a parent with autoimmune diseases (Lupus, Graves, SSc). Two had children who had major chronic conditions (not SSc) from early childhood. The other 16 (64%) patients did not indicate any major or long-term illnesses that they had to face personally, or had witnessed a close family member undergo. As to the familiarity with SSc, 3 patients mentioned a parent, an in-law, and a coworker with SSc. Thus, 88% of interviewees had never seen or heard of this disease prior to their own diagnosis.

One patient had worked in the health field in clinical settings not related to chronic or immune diseases; one was in an administrative role in a health facility, and one had worked in a home care aid capacity in private settings dealing with chronic patients. Two people had expertise in the health field at policy and structural levels, and one patient had a spouse as a medical assistant working in a physician's office. Four patients mentioned having family members or close friends in the medical field, as doctors and nurses, who provided some guidance related to medical decisions or navigating the healthcare system.

Overall, this sample's first-hand exposure to chronic conditions and the workings of health system, as indicated by their experiences of a prolonged disease or occupations within the industry, was limited.

Patients' Work

The interview participants engaged with their illness experience through various types of work (Corbin & Strauss, 1998). These included what patients did in order to mediate the primary physical impact of the disease on their bodies, and the secondary tiers of impact encompassing the emotional, psychological, relational, and financial

aspects in their lives. Initially, this engagement was set in motion by efforts to find an explanation for baffling symptoms they were encountering.

Obtaining a diagnosis. Initially, patients were taken off-guard, describing their symptoms as "strange" and "crazy," and they did not know where to turn to other than their primary care doctors. For some, these doctors propelled them forward through the healthcare system. Others had to find the way through their own efforts. Accordingly, the starting point for patients' involvement was their efforts to find a diagnosis, and this involvement was qualitatively different among the sample, indicating a difference in the intensity, duration, and persistence of effort that patients had to put forward to get a diagnosis. The diagnostic experiences of all patients were formed by a confluence of four elements, whch came together uniquely for each person. These elements consisted of the initial symptoms, the initial medical encounters, the rate of appearance of subsequent symptoms, and experiences with doctors early on.

Initial symptoms. The early symptoms prompting participants to seek medical attention varied widely among the sample. Participants recalled feelings of general malaise, exhaustion, and fatigue; strange skin sensations and changes such as pins and needles, burning or being on fire, itchiness, dry and cracking skin, skin thickening/hardening, pigment changes (white spots), and red dots. Other complaints included swelling of fingers, joint tightening, pain, shortness of breath, and visual disturbance. Initially, some participants dismissed these complaints, instead attributing the symptoms such as dyspnea to being out of shape or being overweight, fatigue to overworking or aging, swelling and skin sensations as too minor. Two participants (8%) were diagnosed without them seeking a diagnosis when they were being evaluated for another condition/procedure. Due to abnormal blood work, they were referred to rheumatologists, who diagnosed them. The remaining 23 patients sought medical attention on their own.

Initial medical encounters. Six patients (24%) saw their primary doctors and were referred to rheumatologists because their doctors suspected some rheumatologic illness (arthritis, lupus, or even SSc). Another seven (28%) patients presented symptoms that were considered alarming, such as shortness of breath, very high or low blood pressures, or lack of motility. These were referred to hospitals or had lab work and tests done, expeditiously. Together these two groups of patients (52% in total) had official diagnoses that came fairly quickly within weeks or months (mean of 4 months), depending on how quickly lab work and appointments with rheumatologists were secured. For these patients, the initial experience of diagnosis, more or less, paralleled the general perception of how known chronic conditions (diabetes, heart disease, etc.) might be diagnosed by seeing a primary doctor who directs the patient to a specialist. They saw on average two doctors for diagnostic purposes; some sought a second or third opinion; and some were referred to other specialists (pulmonologist, cardiologist) to determine the extent of their disease, post diagnosis.

Rate of appearance of symptoms. The remaining ten patients (40%) had a much more prolonged diagnostic process, due to non-specificity of their symptoms, or seeing doctors who did not have the expertise. Initially, some were told to take Tums for heartburn, iron supplement for anemia, cream for dry skin, diuretics or limited salt for hand/leg swellings. One person was told his/her symptoms were psychosomatic, and another was labeled as a “hysterical female” and was put on a tranquilizer. Not counting repeated visits to their primary physicians, they saw on average 4.6 doctors, and it took anywhere between 1 and 5 years before they were diagnosed. Being healthy up to that point and not having any prior experiences with a serious illness, one patient expressed that she did not even know what a rheumatologist did, let alone that it was the medical specialty she needed to seek. At some point in this process, either due to additional symptoms presenting themselves, or through luck, they came across a physician, a friend,

or a family member who guided them toward a rheumatologist. Table 4.9 summarizes the diagnostic experiences of these interview participants.

Table 4.9

Interview Participants' Diagnostic Experiences

How Diagnosed	By chance	Expedited Due to Alarming Symptoms	Thru a primary doctor in short order	A prolonged Process
(n)% of Patients	2 (8%)	7 (28%)	6 (24%)	10 (40%)
How Long	A few weeks	<= 1 month	Mean of 4 months	1-5 years
Number of doctors seen	2 doctors for diagnosis + more to check extent of disease			4.6

Experience with doctors early on. The symptoms and their rate of appearance dictated the person's initial disease state and the extent of medical attention they received. One of the most salient parts of interview participants' experiences of the early days of the illness, in the pre-diagnostic and the immediate post-diagnostic period, was their encounters with doctors. These initial encounters may be said to have been mostly of a convenient nature, in the sense that doctors might or might not have had the expertise to diagnose or treat such a complicated and rare disease; these doctors were the primary care physicians or general rheumatologists who were available locally. Later on, patients searched for doctors in a more discriminating way, which was influenced by these early encounters. The vivid descriptions of these early encounters conveyed four distinct perceptions of doctors' responses: dismissive, fatalistic, measured, and supportive.

Dismissive. Twelve patients (52%) perceived a dismissive attitude on the part of physicians they saw, in at least one of their encounters. Their perceptions were that doctors were not hearing them out, were refusing to do tests, were not taking seriously the extent of their suffering, and were dismissive of the evidence they were presenting. These patients needed to bring their doctors on board by showing their symptoms were

real and by being persistent. More often, they had to find other doctors who listened to them. This group of patients with a contested illness had to put forth the most efforts and persistence to get diagnosed, and their diagnoses generally took the longest. One patient who had travelled far and wide to well-known medical centers in search of a diagnosis and every time was told she was just anemic summed it up this way:

> I wasn't happy that he was telling me this [diagnosing me with SSc] but I was happy that somebody was saying something other than "go home and take your iron." (Rene)

Fatalistic. Six (24%) of interview-participants saw doctors who diagnosed them correctly, but presented a pessimistic view of their prognoses, indicating there was nothing that could be done and predicted their imminent demise. These patients were given a life expectancy of 5 or 6 years and were told to get their lives in order. Two of these patients were diagnosed as early as 4 and 10 years ago, and the remaining four had an average of 17.5 years' disease duration.

Not succumbing to their doctors' fatalistic pronouncements and finding other doctors who were willing to work with them in a more hopeful and responsive manner were approaches taken by these patients. The most dramatic story was that of Francis, who for a year and half had been going to various specialists with severe skin tightening and other internal symptoms. Nobody paid any attention to his skin. Finally, a doctor diagnosed him, but considering his case hopeless, refused to treat him and declined to refer him to another physician. Francis and another patient in this group, among all interviewees, showed the most determination and effort in finding doctors that treated them.

Measured. In contrast to a fatalistic response, five patients (20%) described a measured approach, which left them unclear or confused as to the nature of their illness. These patients' perceptions were that explanation of illness, indication of its severity, what can be done, or how it might progress were lacking. The main symptom for this

group of patients was related to their skin. With no or few other symptoms and no internal impact, doctors took a wait-and-see approach. This measured approach, intended not to alarm the patients, nevertheless left them uninformed about a progressive disease. For these patients, their understanding of the scope and nature of their illnesses came from living with the disease, reading on their own, and going to support groups.

Supportive. Patients who saw SSc specialists and in a few instances other doctors reported the most positive experience of physicians' encounters and the diagnostic process. Twenty-two patients (88%) have seen a scleroderma expert either initially or at some point. With one exception, patients described these encounters as affirming. Patients appreciated the knowledge and experience of their doctors; they felt heard, and given some clarity on the nature of disease and its progression. They also felt they were given hope and a sense that the doctor was accessible to them, so they were not alone in this journey; a number of patients were given doctors' email addresses and could call them at any time.

The sense of relief expressed by patients when they finally found such a doctor, who diagnosed them or gave them hope, answers, and a plan of action, was very distinct and palpable:

> I am lucky to have Dr. ___; again, thank God, I found her. I see Dr. ___ probably twice or three times a year, but every time I go there she has the right answers, the right questions. (Francis)

Overall, 16 (64%) patients reported experiencing two or more of these four types of responses early in their disease in pursuit of the "right" doctor. For about half the patients, the rheumatologist who diagnosed them became their main doctor, and the rest moved on to others.

These early experiences continued to exert an influence and were manifested in an ongoing search and alertness for finding doctors. Not being content with the nature of their interactions was one factor precipitating such a search. Other factors included

logistical issues such as relocation, doctors moving out of a practice/retirement/death; a need to add other specialists due to new organ involvement; the need to supplement their local doctors with an SSc expert or going to an SSc center due to the progression of the disease; and looking for doctors offering alternative therapies or clinical trials. No matter the reasons, finding and interacting with doctors took a considerable amount of time and effort in these patients' lives. See Appendix L, Table L1 for patients' accounts on obtaining a diagnosis.

Managing relationship with doctors. One hundred percent of interview samples saw two or more doctors: nine (36%) patients saw 2-3 doctors, ten (40%) saw 4-5 doctors, and six (24%) indicated seeing more than 5 doctors in various specialties. Many articulated a need to develop a relationship with doctors and their teams. Practically, all patients predicated the development of these relationships on finding doctors with medical expertise, communication skills, and respectful demeanors, but also their own willingness to express and advocate for their expectations, wants, and needs. Thus, interview-participants had developed a list of criteria as to what to look for in their doctors and clinical encounters, as well as a perception of their own role in this exchange.

Expertise. Doctors' expertise was the number one concern of all patients. Even the three who did not seek an SSc expert had made the determination that their rheumatologists were very knowledgeable about scleroderma. For many, the diagnostic experience had created a keen awareness that knowledge and experience in treating SSc was wanting, and they needed to find doctors who had the right credentials. Most were not hesitant to travel long distances to see one. They found these doctors through doctor referrals, their own research, but also through the experiences of other patients.

> Bedside manner is not so much, if I felt that they were really the person who had the most knowledge about it. Bedside manner is nice but I don't think it's something that would be the number one thing I'd look for in my doctor. I would look for the expertise first. The thing that bothered me about the doctor that I changed is that I didn't have confidence in him. (Maddison)

> I have a lot of people that thought I was crazy to travel to California to see a doctor. They're like, "There are good doctors here." That's a special one. So, I run against what everybody else said, and I'm really glad I did. (Jenelle)

Trustworthiness and competence. Patients did not merely seek knowledge in their doctors, but also a type of competency that engendered confidence and trust; a meticulousness and thoroughness that safeguarded against things falling through the cracks invoking a sense of safety for them. A number of patients noted that they do not even consider their doctors "nice," but rather competent and trusted.

> This doctor is not even nice. I don't even want to say nice. She's just a smart and a good doctor and looks for everything … she just gets right through the crap, stops all the bullshit.… This is a crazy disease. You have to be comfortable with the doctor.... I feel so much safer with Dr. ____. It's a good word. I feel safe where I feel like I'm going to be okay and if I'm not, she's going to always take care of it. (Mackenzie)

> She is very, very meticulous. (Francis)

Interview participants mostly spoke of one doctor that they had this trusting relationship with. It seems it was not necessary for all their doctors' relationships to be of this nature, as long as there was one trusted doctor. Patients used terminology attributing a "North Star" quality to their trusted doctors. Not only were these doctors seen as showing the path, but they also acted as a reliable resource for keeping in check patients' own pushbacks, for checking the soundness of other doctors' recommendations, and for finding other good doctors.

> I got to [trust doctors]. I go crazy otherwise.… I trust them; you got to trust them. It is like, you know, getting here; you got to trust your GPS. (Ralph)

Communication style. Interview participants wanted doctors whose communication style matched their expectations. By this, they meant finding a doctor who had the ability and willingness to convey information to them, who really listened to what they were saying, and answered their questions truthfully in a straightforward manner without being falsely hopeful, evasive, patronizing, or demeaning.

> So, to me, I need to know if the doctor is listening to what I'm saying. (Jenelle)

> I expect him not to candy coat it; I just want to know what it is now and next and that is it. Tell me what it is, do not give me false hopes and as far as that goes you know, I know you are working on a cure and give me a call when you got it. Do not just tell me you are working on it. (Ralph)

Non-verbal communication style. One non-verbal communication cue that patients took as belying trust was eye contact. "Looking them in the eyes when spoken to" had a significant meaning to many of these patients. As doctors increasingly look at their computer screens and tablets while attending to their patients, this was interpreted as either not seeing the patient as an individual, or as an inability to grapple with uncertainty and ambivalence about the prognosis or course of action to be taken.

> This pulmonologist who always said, "There's nothing you can do." He wouldn't look at me. He never examined me. He would just sit and type and stare at his computer and I'm over here and he would give me bad news and not look at me. And I had to stop and then say, "Look at me and tell me that. I need you to stop typing and look at me." (Jenelle)

Even when patients did not take it negatively, they made a note of it:

> Sometimes, he'll close his eyes but because he's thinking. I can tell he's thinking about what you're saying. (Maddison)

Patients' role. Interview-participants' perceptions of their own role included compliance with therapies and expression of their preferences. All patients claimed a very high level of compliance with medications and vigilance in keeping appointments for regular tests and checkups. Given the seriousness of their symptoms, patients felt compliance was necessary and expressed that they did not modify their regimen without consultation with their doctors. Within the boundaries of conventional medicine, however, they argued for choices. This included delaying or refusing certain treatments, taking minimal amount of medication, asking for certain medications, requesting inclusions in clinical trials, or refusing to participate in one. Patients expressed an awareness of voicing their questions or dissatisfaction, and asserting their wants and

expectations, and saw themselves as playing their part by having these conversations with their doctors:

> I said, "I'll work with you but you got to work with me."… I didn't jump and all of a sudden voice all of this. First, I let him come in and ask and check me and do everything and then when it was time for me to talk, I explained about myself and my own feeling of how I want to wait taking medicine for this disease until I feel that I can't function rather than the belief that "Catch it early. Go on all these." (Lauren)

> My primary care used to say, "Let's try this. Let's try this. Take these drugs." And every time we would do it, I would say, "Do I really have to do that? I don't think I really have to do that?" He would say, "I think you should. I think you should start on this kind of a regimen and see where it goes." And I would try to do that and I would come back and say, "I don't want to do it. Let's try something else." (Kristeen)

Patients attributed their assertiveness in expressing their preferences to their experiences of living with the disease and having confidence in their own knowledge, holding a non-intimidated view of doctors because of knowing relatives who were doctors, but also a belief that a good doctor would not be offended by their pushbacks. Such confidence in expressing wants and needs, and the expectations of mutuality, were more often articulated by patients with longer duration of the disease. They spoke of their relationship with their doctors in terms of friendship or partnership, or lamented the loss of one such relationship due to relocation, retirement, or death. In a similar vein, those with more complex cases were more likely to verbalize the greatest sense of trust and confidence in, and satisfaction with their doctors, irrespective of the length of illness. See Appendix L, Table L2 for patients' accounts on managing doctors' relationships.

Pursuing effective therapies. Therapies for these patients, regardless of the subtype of scleroderma, were not a simple matter of taking proven pills or injections. Rather, it was an individual trial-and-error process where choices were dependent not just on the professional judgments of physicians, but also how much efforts patients were putting in to find the medications that worked for them. Based on their symptoms,

responses to the drug, rate of progression, organs involved, and the length of illness, patients described different experiences related to therapeutic interventions.

The most drastic experience was that of Nicole with a rapidly deteriorating lung function, who went from six months of chemotherapy to autologous stem cell transplantation (ASCT) within a year of diagnosis. She was one of the handfuls of patients in a trial that was stopped shortly thereafter. A cohort of patients went through this trial in a serial fashion, each patient working closely with the one next in line, preparing her/him as to the sequence of events, procedures, reactions, and generally what to expect. She was coached by the person immediately preceding her, and coached the one succeeding her, both of whom passed away within a year.

Unlike Nicole, the majority of patients went through therapies in isolation. In effect, they were a "medical trial of one," all by themselves trying to assess their body's responses to powerful medications over many months. When an effective medication was found, they needed to be persistent and continue their vigilance, looking for side effects, or diminished effectiveness over time as the disease progressed. Some had to try a cocktail of three or four drugs for maximum effectiveness.

Patients spoke of onerous delivery mechanisms (e.g., pumps, or heavy oxygen tanks), complicated dosages and regiments (e.g., given high doses, weaned off and starting the cycle again, or having to maintain a precise concentration of drug in their bodies), compounded by physical changes, such as esophageal issues that made swallowing pills difficult, or hardening of skin that made injection needles penetrating skin an ordeal. These required a considerable degree of compliance commitment not only in the form of dedication and discipline in taking the medications as prescribed, but also by enacting changes in their lives to accommodate the therapies, such as modifying activities and changing eating and sleep patterns, etc.

> They have put me on Remodulin [a vasodilator for PAH in form of continuous subcutaneous infusion], which looked like I had an implant

> hooked up to me. And so, I had to reevaluate how and when I did things because it was very sore so I was not as active as I could. It made that difficult in changing it and I was on it for about two years but now, I'm finally off the pump and they put me on the pill now. So now I feel free. I can take baths. I can go running. I can do a lot of activities that I was restricted to before. (Winola)

Of note were two patients with exceptional disease longevity of 45 and 30 years. Very few medical options were available to them at the time. However, one continued seeing doctors and eventually was treated and, according to her, has been in remission; the other gave up on the medical profession and decided to "live her life." She was the only exception in the sample that did not actively seek medical interventions early on, with detrimental effects.

> There wasn't very much to do, not at all if you're talking 30 years ago. It's not like today. Today, they're more aggressive…. Back then, it was strictly—they treat you with the high blood pressure medications.… So why am I going to go and get the same drill?… So, I just said I'm tired of it. I'll just live my life, whatever befalls me- because you could spend your life at a doctor's office getting picked and poked and I didn't want. (Vanessa)

One complication that some patients needed to manage was insurance drug coverage. Since many of medications for SSc were prescribed off-label, insurance companies generally did not cover them. Furthermore, they were subjected to formularies on a list of eligible drugs for a given plan. A handful of patients spoke of having to continue taking drugs that were not effective and/or had side effects for months in order to establish their inefficacy/harm for insurance purposes. Only then could they try other drugs, hoping, having built a case, the insurance would cover the cost of new medications. This put these patients in the difficult position of having to make decisions as to what therapies they could afford or were willing to take, in spite of the professional recommendation of their doctors.

Patients' engagement with therapies was most intense in the early and active phases of disease, but was also ongoing for many patients due to loss of effectiveness, toxicity, and progression needing to alter medications. Being alert about and engaged

with their medications was a constant endeavor throughout the course of illness. See Appendix L, Table L3 for patients' accounts on pursuing effective therapies.

Attending to emotional and psychological impact. Interview data indicated a range of ongoing emotional and psychological burdens in the course of illness, initiated by the fear of dying, and followed by fear of progression of disease, depression, and suicidal thoughts, frustration related to their disabilities, and distress over body image.

Fear of dying. Whether through their own research, having known an SSc patient, or having information imparted to them by their doctors, the primary emotion shared by the overwhelming majority of patients was fear, in particular fear of dying. Out of 25 patients, 1 person explicitly stated that she was not scared, 3 did not speak of their affective reactions, whereas the remaining 21 (84%) used one or more of the following words and phrases to describe their emotional states: scared, fearful, terrified, panicked, afraid, frightened, freaked out, petrified, upset, shocked, devastated, going to die, needing a bucket list, never will see my kids grow up/get married, sobbing, crying, tears pouring down my face.

> Fear. Fearful, just—I don't think it was anger because I was just—everything I read was just so devastating to me…. I didn't know what it was. I couldn't even say it [scleroderma], and then I saw pictures of people very disfigured. It's hard not to be upset about it. (Brenda)

> I thought I am going to die. I had young children … I started to cry … I never heard of [SSc] and when I looked it up and said 80% of people diagnosed with it will die within 10 years. So, talk about a mid-life crisis, I had one. I had a bucket list a mile long. (Paula)

> First I was scared. Then I did the Internet thing, which is really stupid, because then you get a wide variety of information and people initially think it is a death sentence. (Stacee)

Fear of progression. After the perception of an immediate death subsided, the fear of progression of the disease moved to the forefront. Patients took certain indicators as foreboding, signaling the worsening of their diseases or portending a decline in the near

future. These included appearances of new symptoms and lab and test results falling outside normal ranges, but also those they harnessed through body surveillances.

> I did not cry through a lot of it at the beginning. I was sort of surprised. But when I got the diminished lung function, I was really devastated.… I thought that was sort of 'oh gush it could be the beginning of the downturn'. (Willa)

> One of my biggest fears is that I'm going to end up having to go back on O2 [oxygen]. (Laurie)

> I really get worried when these nodules, they start scraping against each other and I'm worried they're going to open up. I do know somebody that has all the open sores here and that's another thing I'm scared of. I don't want to look like that. (Mackenzie)

Whereas the above measures indicated the short-term changes in the disease, the patients in more advanced stages whom one could see in support groups and other venues embodied the progression of the disease in the long term. Seeing these patients, in support groups or patient educational forums, was particularly difficult for many interview participants. Another way patients revealed their angst about progression was the uneasiness and ambivalence they had in differentiating between possible new symptoms and those related to aging, genetics, or other illnesses. Not knowing the attribution was particularly difficult for older patients, who were dealing with aging at the same time.

Body image distress. Three patients in the sample had identified their subtype as scleroderma sine scleroderma, meaning they had no skin manifestations. The remaining 22 (88%) had various degrees of skin thickening characterized by tightness, hardness, or leathery appearance of skin. Other visible changes included telangiectasia (red dots), discoloration of skin, changes to the shape of mouth and nose, rupturing of skin and ulcers on fingertips, and contraction of fingers. Out of 22 patients, seven (31.8%) considered their skin issues minimal and not concerning image-wise. For ten (45.5%) patients, there were sufficient physical changes to cause body image distress; this was

irrespective of gender. Among these were patients who deemed the changes so drastic that they were not recognizable from their old selves, prior to having the illness. A few characterized their concerns as vanity; others alluded to social expectations of women being pretty and men being athletic. Other signifiers of illness, including publicly visible therapies like oxygen tanks or loss of hair due to chemo, were also mentioned as sources of social anxiety. Presentation of self, altered by skin deformities, was particularly disconcerting for younger patients and was brought up in the context of dating by two patients.

Ironically, the patients who did not have any noticeable disfigurements had a problem of their own. The phrase "but you do not look sick" was a detested statement that was repeated by several patients. This was especially true early in the disease process when the inflammation in skin, before it became hardened, eliminated the wrinkles and patients looked younger. The misalignment between how they felt and how they looked was perceived as needing to prove or legitimize their illness. This was a source of stress for a number of patients.

Frustration, anger, and depression. Seventeen patients (68%) used words such as "frustration," "angry," and "upset" in describing issues they have faced. Twelve (48%) interview participants indicated in their survey that they had experienced depression. Among the patients with depression, seven spoke openly about their anxiety and suicidal thoughts during interviews.

> I was fearful and I just—it was weird because I actually got depressed and I suffered from depression and I don't think that's uncommon. (Brenda)

> I'd be driving home and I'd be in such pain…. I swear to God, I saw this building, a brick building, was like a cleaner, I came to this fork and I thought if I just kept going and crashing that building I could just end it all, and you would never think that you would think that way, but I did. It was torture. I loved my family and they needed me and I needed them, but I was in such pain that I actually had these thoughts … every time I saw the building. (Brittany)

Interview participants attributed disabilities, fatigue, and alteration or loss of roles as major contributing factors to their sense of frustration and depression.

Disabilities and fatigue. The inability to be active as before or to do mundane chores was especially pronounced for patients with joint issues and contracted fingers. Ordinary tasks like opening a jar or a water bottle, turning a rotating light switch, a round doorknob, or faucet, turning on the car, or closing the trunk door became ordeals on a recurrent and daily basis. Even though the disability and physical limitations that patients were facing varied widely in type and severity, they implicated all patients in one form or another, contributing to their daily irritation. Complaints about Raynaud's were universal. Stiff fingers and joints, pain, shortness of breath, and digestive and bowel issues, variously, affected patients. All of these complaints curtailed their activities and contributed to their frustrations. Kurt's detailed description of cascading frustrations is instructive to understand the daily struggles of these patients:

> I told you about GI issues, you have SSc hand issues. You have [finger] contractions. Try being all bundled up in dead cold of winter here in _____ as it was and has been the last few years. First imagine that, then, imagine you're outside in public area, you're not at home and you got to go to the bathroom because your stomach is giving you a problem. Now, only have a short time to find a place—your fingers and hands aren't working—you got to get yourself unbuttoned. You're in a public restroom. What are you going to do with your clothes?… Your hands really don't work. They're frozen from the Raynaud's. Try reaching for the toilet paper…. Think about those thin pieces of garbage that you're given for toilet paper in the restroom. Think about trying to pull on that big roll. Think about trying to fold it. Now, think about reaching back when nothing is working in trying to clean yourself. (Kurt)

Another universal complaint was fatigue. Both the disease itself and the use of immunosuppressant and chemotherapy drugs plagued the patients with constant weariness.

> Feeling crappy all the time. Being tired all the time. Before I got really sick, at work they used to call me the energizer bunny. I always kept going and I cannot do that anymore. I feel I lost my batteries. (Patricia)

These forced patients to give up on activities like doing housework, gardening, traveling, and the isolation that resulted from giving up their social lives and staying at home, because getting out was just too difficult in light of the disabilities they were facing. Other major sources of frustration for patients were the lack of understanding on the part of others regarding their illness, and the difficulties in dealing with the healthcare system and insurance coverage. Four patients used the word "anger" to indicate the intensity of their frustrations. These patients tended to be living with the disease for a shorter period of time, and their frustrations were related to dealing with issues outside their direct control like the healthcare system.

Disruptions of roles and identities. Many patients had to give up their work, to take on less demanding jobs for which they were over-qualified, or to relinquish plans for advancing their education and moving up the career ladder. Two male patients openly lamented the loss of their identity as the provider, fixer, and protector of the family, while a third expressed anxiety in not being able to provide for his family financially. Disruptions in discharging parental roles weighed heavily on those with younger children; the unexpected physical and emotional unavailability due to fatigue and depression, failure to provide financially, and the possibility of not being there to rear their children to adulthood was devastating to all participants who were parents.

A number of interviewees were athletes. The high levels of athletic ability developed over years of training being taken away and replaced by extensive physical limitations were heartbreaking to them. Other patients reported the loss of fine motor skills: one person spoke of her inability to play the piano, another talked about limitations in doing artwork, yet a third could not type on a computer keyboard, and a fourth had lost tactile ability to handle small instruments needed for work. Table 4.10 summarizes the extent of the main psychological stressors on patients. See Appendix L, Table L4 for patients' accounts on the psychological burden of illness.

Table 4.10

Interview Participants' Psychological Burden of Illness

Psychological Burden	Fear of Dying or Progression	Depression	Frustration & Anger	Body Image Stress
n (%) patients	21 (84%)	12 (48%)	17 (68%)	10 (40%)

Approaches to coping. The interview participants presented various approaches to cope with and mediate the emotional and psychological burdens of disease, and to reframe their lives in light of the disabilities and disfigurements that precipitated some of their angsts and disquietudes.

Seeking professional help. Seven patients (28%) indicated that they sought professional help and attributed their coping skills to years of therapy and medications.

Keeping busy and being productive. Some interview participants found working and keeping their employment very helpful.

> Even the doctors recommended I stop working. My doctor, Dr. ___, told me even if it is only 2 or 3 months, I give you the paperwork whatever you need. I said, Dr. first of all financially I cannot afford and I want to keep active. I think that is what has helped me the most. Being active and going to work. (Francis)

Doing for others and keeping busy in the process was another major theme in many patients' lives. Seven patients (28%), in spite of their own conditions, were caregivers to aging or ailing spouses, parents, or children; three (12%) were helping raise grandchildren; and at least nine (36%) had cared for children at home while going through this illness. Seven (28%) had initiated a support group or taken on a leadership role, and a few volunteered for other causes and neighborhood organizations.

Reducing daily irritations. On a practical level, many patients had found creative ways to ameliorate some of their disabilities in order to reduce daily irritations and frustration. One patient with joint issues in his hands had put his young children in charge of unbuttoning the shirts he was to wear for work; another had crafted a warming

beanbag for her steering wheels; yet another inserted cork stoppers in rotating light switches, and few put rubber bands around pens, brushes, and doorknobs, and used stemware for a better grasp. Others mentioned keeping a blanket in the car, a pair of cold weather gloves in every jacket, hand warmers, and rubber gloves in their bags (to grasp or turn things outside the home). Some have found modifying the type of clothing they wear (pants with elastics not zippers, wide-toe shoes, using insoles) as helpful. Through such small solutions, many had ushered a sense of control and reduced the constant irritations over mundane tasks in their daily lives.

Controlling what they could control. Some patients found the secret in controlling what they could control like doing things that were helpful to their bodies, such as exercise and eating well, reducing their stress, and by not dwelling over things that have not happened: "Why borrow trouble?"

> It's just managing how the disease makes you feel…. There's a lot of SSc issues that we can't do a thing about, but I think again, lifestyle changes. You're just accepting that you can't live your life the way that you would like to. There are limitations and just try to make the best when you do have a good day. You get out there, smell the roses, breathe the fresh air. (Laurie)

> I wake up every morning and say OK, put one foot in front of the other and keep going. Regardless of how you feel, everybody has those days that you feel horrible, you know, the aches and pains, my skin hurts and my lungs hurt, but then you make up your mind: am I going to make it through this day or am I going be miserable all day? I have a job to do; I have kids. So, I can either let it overtake everything I am, or I can plow on and hope for the best and keep going and that is what I do…. It is just a decision. It's just a decision that you make. Am I going to be miserable? I am not. (Stacee)

A handful of patients also acknowledged that not putting their disease front and center in their lives in terms of a constant preoccupation has been helpful.

Reducing fear. For some patients, learning about the disease took away the fear and the hopelessness; for others, finding out about patients who successfully have managed their illness, or seeing patients who were worse off than them but were pushing through,

provided the courage. Yet, for others having someone to talk to and vent out, and drawing on faith and spiritual beliefs calmed their nerves and allayed their anxieties.

The early critical years demanded the most from patients in mediating the emotional and psychological burdens of the disease. Notwithstanding that, the psychological burdens, like fear of progression, continued to be ongoing for patients requiring their full engagement throughout the course of illness. See Appendix L, Table L5 for patients' accounts on approaches to coping.

Dealing with employment and financial impact. The most consequential impact of the disabilities experienced by these patients was their inability to perform at work. Employment had a significant meaning for patients, not only as a means to make a living, but also instrumental in having medical insurance coverage and affording auxiliary services (physical therapy, home assistance, etc.). They and/or their spouses needed to work as long as possible, delay retirement to maintain medical insurance coverage, or earn enough credits to qualify for social security.

Giving up work also was tantamount to a loss in one form or another. Some indicated it was a loss of identity tied to professional or expected roles. For others, it was a loss of a passion—giving up something they loved to do and were good at. For yet another group who had reached high levels in their careers, it was a premature loss of achievement. For a younger patient who was planning to further her education in her chosen field, having to give up those plans and find a less demanding job that she could do was a loss of dreams and hopes.

The interview participants dealt with job and ensuing financial impact by continuing to work; retiring, or going on sick leave or disability; receiving assistance from family, including spouses delaying retirement, or financial assistance; and becoming a discriminating medical consumer.

Continue working. Remaining productive as long as they could, both for financial and insurance reasons, was the approach taken by seven (28%) interviewees who had

continued with full-time work, and one with a part-time job. A handful of patients felt that keeping active and working not only was necessary financially, but also helped them psychologically, even when it was against their doctors' recommendation. In order to continue working full-time, a number of patients had to switch jobs, finding work they could do, often at a great economic cost. Others had managed to keep their positions by asking for accommodations. Some employers were sympathetic and accommodating, and some were not.

> I need to do a little bit more work because of social security—I'm four credits short of getting my social security benefits when I'm 65. I'm 62 now … that's my big concern at the moment is trying to earn these credits. (Laurie)

> I actually changed jobs because … I could not handle the instruments anymore. So, I had to change jobs to do something that did not require such a fine tactile movement…. So, I got a job doing something in an office setting rather than the _____. You know it is easier. I am not doing what I am trained to do; it is way downscale and I took a humongous pay cut. (Stacee)

> I was in the [this] business so they are not very giving on giving you extra time away and it was high stress, long hours and very physical work. So that made it very difficult. So, I was also working with a company that was not very sympathetic at all. So, I told them about my situation and I tried to get some accommodations which they wouldn't do and then I went out on medical leave and then they wouldn't let me back. (Jenelle)

Retiring, sick-leave, disability insurance. Among the remaining patients, eight (32%) retired, or stopped working before retirement age, and another eight (32%) went on disability and sick leave. Debilitating fatigue, pain, hand deformities, and impacted tactile and fine motor skills were major culprits for these patients.

> I felt like I was made out of wood and the pain was excruciating … the pain was constant.… I couldn't sleep. I needed to [work], because my husband was a teacher just to afford my three kids…. I kept working as long as I could and then I couldn't anymore. I had to stop. It was just too difficult. (Brittany)

Those patients who went on disability insurance described it as a very difficult decision and undertaking symbolically, and logistically. Logistically, they found the process complicated; symbolically, it was the acceptance of their disability.

> If you don't put the right things down, you'll get denied. You just need to be very descriptive … chronically ill patients don't talk about it that much. They either over talk about it or they don't talk about what they really go through, and that's really what has to be on this form. (Jenelle)

> It just did not seem, you know, and when I think of somebody young who is on disability [insurance], I have a friend who has an MS and she is in a wheelchair; that to me makes sense. It was not the image of me that I had in my head. (Sandy)

Accepting family assistance. Patients with a partner had to rely on their spouses to continue to work and maintain their medical insurance for the foreseeable future. Single, or younger patients faced more challenging circumstances: working until their symptoms deteriorated enough to be eligible for disability coverage, or pushing through to the retirement age of 65 were the only viable paths for them. These were the patients who expressed the greatest financial uncertainty looking forward. A few patients had to accept financial assistance from extended families.

Becoming discriminating medical consumers. Given these financial difficulties, patients had to become more aware and discriminating medical consumers and had to decide how, where, and what medical treatments they would seek or agree to in order to manage costs.

> I had that discussion [with doctor]. I said, "Well, I don't want to take it [Methotrexate] in pill form. I have enough GI problems and [my local hospital] doesn't do the injections. I get to pay a fortune, go out of state." So, we discussed it and decided no Methotrexate type treatment. (Lauren)

Five patients (20%) saw doctors in SSc specialty centers more than 100 miles away from home, while three (12%) had to travel 25-50 miles to see one. For most, the closest centers were located out-of-state and were out-of-network. At least three patients had learned that by splitting the medical encounters—seeing the expert at the centers, but

doing lab work or procedures locally—they could be partially covered by insurance. Sometimes this had put patients in an awkward position of asking doctors for special accommodation, and to push back on insurance companies and hospitals' bureaucracy.

Dealing with the financial fallout of their disease was a grave burden and concern of all patients. It required mental energy, as well as vigilance to deal, manage, and plan, regardless of patients' financial situations. Overall, the younger and single patients had the most financial issues and concerns, as they had to face the brunt of this impact earlier in their careers and without the safety net of another wage-earner in their households. See Appendix L, Table L6 for patients' accounts on dealing with employment and financial impacts.

Handling familial relationships. The interview data revealed the impact of this constantly present illness on family members in at least three distinct arenas: (a) financial, which has already been touched upon, such as the loss of income, and when spouses were impacted by having to delay retirement; (b) psychological and emotional reactions of the family members to the illness itself; and (c) the role family members took on in relation to the patients.

The interview participants expressed the psychological/emotional impact of their illness on their family members as those of fear, worry, concern, denial, and disbelief. Many patients not only had to deal with their own emotional state, but also needed to find ways to ameliorate the fear of their loved ones.

> They were very scared. They were really scared, and my kids saw that I was pretty upset. So that was upsetting to them. I never said to them, "Mommy is going to die." My husband didn't know what to do.… I think he was very afraid, but we worked through it, but it was hard. It was hard. (Brenda)

The levels of engagement of family members, including their levels of knowledge of disease, varied greatly. Some took the lead in researching the disease, interventions, and doctors, exceeding those of the patients themselves. Others made no attempt to know

or understand, perhaps due to their own fears. Some family members took on an active role in supporting and engaging with the needs of patients, while others were more oblivious and removed. A handful of patients mentioned coming from large families (>5) or having close extended families. These patients expressed the most positive views of perceived support. Conversely, there were a small number of patients who spoke of stress and a lack of familial support that went beyond obliviousness. This, according to many patients in the sample who attended support groups, was not unheard of and was quite common. In supportive families, the roles members took on included:

- Information gathering, therapeutic suggestion, and finding physicians
- Accompanying patient to doctors, tests, and infusion sessions, support group, and educational forums
- Mental/emotional support, including decision making and reality checks
- Help with tasks/housework/driving/preparing healthy meals
- Monitoring for symptoms and changes
- Financial help (from parents, grown children, or siblings)

Patients spoke appreciatively of these supports as expressions of love and commitment, especially coming from their spouses, daughters, and extended families, using phrases such as "being lucky," "blessed," "wonderful family," "amazing support," "always being there." This was intertwined with feelings of guilt and being a burden for at least four patients. The most salient sentiment, however, was that of a disconnection. The overwhelming majority of patients felt that their illness experience was theirs alone.

> My sister doesn't [understand], for however much she wants to try, because it doesn't make sense unless you're experiencing it, or seeing it every day or have another chronic illness that other people don't understand. (Jenelle)

Not only strangers, but also closest loved ones could not have possibly comprehended it. Patients' descriptions indicated that life had dramatically changed for them, while it stood at a standstill for their families. This disconnection had created murky waters in which

notions of expectation, support, limits and boundaries, and dependence and independence were all submerged. Kristeen captured the essence of this most vividly. Her articulation is extensively presented below because it so critically and candidly reveals the intricacies of the familial relationships when a chronic illness is present.

> I've come from a very big family and it's tough and so when I say, "I really can't do this because I'm hurting so bad today, I can't do it." They think, "Oh, you're wimping out" but they don't understand. They don't understand what it's like … they don't get it. Really, unless you have it, you don't get it. I think my husband—I've been with him for 30 years—he doesn't get it … they don't get that every single day, you have it. It's not something that you have in three days and then it goes away and life goes back. Life never goes back. They have that saying where the patient goes to the doctor and the doctor finds out that you have a chronic illness and another patient says, "What kind of chronic illness is this?" And you say, "Well, it's good news and bad news. Good news is you're going to live, bad news is you're going to live." I don't know how else to [describe it]—but it's every day. It's every single day and they don't understand it. It's like, "you couldn't come last week, how about today?" No, I still can't. It's like, "Can you come and help me paint?" I still can't. Even on a good day, I can't hold the brush. I can't. Even when you mean to do, you can't get through it. They want to understand. They want to, but it's—I don't know … I feel sorry for caregivers. I do because on one hand, they don't understand, but on the other hand, I don't want your help unless I really need your help…. How can a family member help you? I can say, Well, they can listen to me when I'm sad, they can try and go with me to the doctor, take me for a test but when I'm trying to get a scoop of ice cream out and it's hard, leave me alone for a minute. Let me at least try, because if you start taking it out of my hand, I'm going to let you but then I lose that ability to even try anymore. So, it's a really a fine line and I feel bad for caregivers. I haven't been nasty to anybody but I've tried to say, you have to let me try and do it. Even with trips with family, they'll say, "Can you do it?" I'm going to try. I really don't want to miss it. I have bad knees and it hurts, but yes, I don't want to give it up so let me try. Let me try and if today, I don't feel like going on to the beach, I can sit on the deck and watch everybody. It's a fine line, I understand and it's hard because they have a life, too, and they want you part of their life and sometimes you can, sometimes, you can't. (Kristeen)

Being in and out of family activities, as the pendulum of good days and bad days swung, upended routines, schedules, plans, expectations, and commitments. This inability to predicate how and when they could be involved with family and social circles was a clear stressor on patients. In addition, many patients struggled with family's and

caregivers' suggestions, interventions, and information overloads as these contrasted with their own ideas of expectations, needs, and support. Patients confronted these in different ways:

- Putting the efforts into informing and educating the family
- Keeping the stress level on family members low by not complaining, maintaining independence, and being self-reliant
- Accepting their own limits, and clarifying the boundaries and their responses to others' demands
- Stating clearly their expectations, and guarding against assumptions of support
- Limiting how much information they were willing to receive from family and friends
- Planning ahead and resting so they had energy to participate in activities and to keep their commitments; making it clear that spontaneous participation was difficult and they needed ample notice to prepare
- Not committing to one-on-one activities, giving them a way out: a group activity could go on without them, if they needed to excuse themselves from it without causing too much disappointment
- Modifying their lifestyles in terms of becoming "choosy" in their social circles, not getting entangled in family problems, and in some cases cutting off from their families and/or friends altogether

Many of the issues that patients and their families were facing were not fundamentally different in nature from those most families face. However, the levels of stress, physical limitations, and uncertainty due to illness accentuated them. Furthermore, the assumption that patients had the benefit of a supportive home environment did not hold across the board for these patients. See Appendix L, Table L7 for patients' accounts on managing familial relationships.

Motivation

The motivation of patients to engage with their illness through various types of work was related to their acceptance of disease, and commitment and resolve to take ownership of the illness, which temporally took different forms for patients. This commitment was sustained by hope, a will to live, and imagining a future.

Commitment. Participants described the difficulties of accepting the diagnosis. The disease was so scary and beyond anything they had known that it simply could not be true. All the intensity of seeking a diagnosis in a short period of time became a force of resistance.

> When he first diagnosed me, I told him he was crazy because I said I have a friend that has SSc, I do not have what he has. That is not true. So, I stayed away for a while, suffered and finally called him back one day and said I am sorry I was upset and I need this taken care of. (Ralph)

> It really took me two or three years to really [accept]. "This is it. This is what I have." (Maddison)

Those early days and months, post-diagnosis, were characterized as being shell-shocked. Some clearly defined a period of withdrawal, lasting several years, to wrap their heads around what had happened, to accept it, and to decide to move on. For others, equally shell-shocked, the decision came much earlier, within weeks or a few months.

> One statistic was 50% die within three to five years so that terrified me and had stuck in my heart. So, I think I stuck there, kind of like out of life, for a couple of years- not weeks but years.... I cried all the time. I didn't want to do anything. I was terrified I was going to die tomorrow in my sleep and it was just awful. (Kristeen)

In order to attend to their failing-bodies, they needed to pull themselves up by the bootstraps and get on with the business of tending to their illness. This was described as a commitment or resolve to take ownership of and to engage with the predicament they found themselves in.

> You need to be able to say, 'I want to survive this. What can I do to survive this?' (Kurt)

> I went through a period of time where I was like, "Poor me. Poor me." I had to snap out of that…. Somewhere around four or five [years into illness], I started saying, "I got to start living this. I got to live with this disease. I got to figure that out. (Mackenzie)

Each patient had a reasoning of his/her own for this commitment. Some attributed it to their personality traits and characteristic such as proactivity, pragmatism, feistiness, and optimism and positivity.

> It was difficult in the beginning and then I realized either I do something about it and succeed, or I crawl into a ball, go to a corner, wait to die…. I knew, almost from the beginning, I had to take [charge of] it. I knew from the beginning because that's who I am. I am an A-type and I hate that expression but that's what people think of me. I always did take charge. (Kurt)

> I'm sort of an optimist about taking control of myself. So, personality has a lot to say. I didn't get scared by it. (Lauren)

> I am not a quitter. I have an aggressive personality and I think that serves me well to some degree. I do not mean aggressive in a bad way—I am not afraid; I am not a scary cat. I think that serves me well because it would be very easy to sit in a corner and cry. Because it is weird; it is just weird. (Stacee)

Other reasons included having children and families; the realization that life went on and their death was not immanent; the possibility that prognosis would not be as grave; stabilization of disease for some, and the worsening of symptoms for others; and the realization that confronting the disease was solely up to them. This perspective was expressed by three patients who believed no one would or could rescue them—not family or friend, nor medical professionals or pharmaceuticals.

> I just started to realize I was the only one. Nobody was going to come and save me from this. Nobody was going to figure out anything for me. I had to do it myself and I had to take responsibility. I'm not responsible for the disease but—Yeah, nobody was going to figure it out. I would have loved if they did and just told me what to do, but— (Kristeen)

> People expect the doctors to cure them. They expect the pharmaceuticals to come out with a magic capsule that will make their entire life better. Life doesn't work that way … unlike most people, I don't believe they're going to find a cure in my lifetime.... (Kurt)

Medically, there could have been other contributing factors on how quickly patients needed or were compelled to take charge of their situations. Unlike the patients with a slower progression of disease, those with a rapid deterioration subsequent to initial symptoms had to race against time in terms of finding effective treatments to halt the progression of the disease. According to one study (Domsic, Lucas & Medsger, 2010), over 90% of organ involvements, in particular gastrointestinal, lung, heart, and kidney, experienced within the first 5 years of disease (as opposed to those that occur much later) occur during the first 2 years. Eleven participants (44%) reported this type of experience. Among these, five patients had rapid decreases in their lung functions, two patients faced heart issues, another was subject to a speedy and painful skin hardening, and the rest dealt with a combination of symptoms occurring in rapid succession.

Thus, temporally, the experience of the disease placed the patients in two different camps. For 13 patients, the disease experience was characterized by a more drawn out phenomenon: being diagnosed with a serious and uncertain illness, some medical interventions, and a gradual involvement of organs overtime. On the other hand, about 12 patients described a rapid onset of new symptoms within the first 2-3 years of the disease, needed to find SSc experts, had to make decisions on aggressive medical interventions such as chemotherapy, had to weigh experimental or unconventional treatments as options, saw more doctors to consult with, were subjected to more frequent procedures (CT scans, echocardiograms, right heart cauterization, pulmonary function tests, etc.) to check the status of their various organs, and overall had a greater sense of urgency. These differences might have been the other contributing factors propelling patients to take a position on how quickly they would engage with their illness. See Appendix L, Table L8 for patients' accounts on temporal dimensions of making a commitment.

Sustaining this commitment was predicated on hope, finding a will to live, and imagining the future.

Hope. A few patients attributed their religious and spiritual beliefs as the source of hope; others to possibility of effective treatments and a cure happening soon. Most interview -participants described hope as given by their doctors' assurance and explanations.

> "Yes, you have this and we can treat the symptoms. There's really no one medication to take for the whole thing so we're going to give you a great team. The team is going to work with you and we're going to address the symptoms as they come up…." [He] was always there for whenever I needed him. (Winola)

> He wrote down so much and he asked so many questions and he did explain a lot to me, "Right now, you're alright. You're going to progress and we're going to try and start things but for the moment, you're okay." And I think it helped me to hear somebody tell me that. (Kristeen)

> My current doctor, when I ask her about it [prognosis], she just gives me a hug and says, "I'm going to keep you alive until you're in your 90s." (Lauren)

A will to live. The sustaining power to continue their commitment to illness was illustrated by the way participants looked for reasons to live. Having children that they needed to care and live for continued to be one of the most powerful reasons for interview participants. Being there for their children and seeing them grow up weighed heavily on patients, as 76% (19) of interview participants were parents. The ages of their children at the time of their diagnoses ranged from 4 years old to the early 20s.

> My priority was my two children and how this is going to affect their lives. (Brenda)

> My kids are the reason I keep going, pretty much, because I love my kids. (Stacee)

Obligations to spouses and parents, and entertaining possibilities for a cure, remission, or arresting the progression of disease kept many participants going. The prospect of a shortened life was another impetus to live fully in the time they had. Patients tended to present "not dying," "cure and remission," and concerns about children

in their descriptions of earlier years with the disease. When speaking in later years, expressions of "slowing the progression" and "living their lives" were more prominent.

> I am not going to make myself crazy. There are too many other things in my life: we've got __ children and 8 grandchildren and we are very involved in our church and I need my energy for that. (Ralph)

> I have a job to do; I have kids. So, I can either let it overtake everything I am, or I can plow on and hope for the best and keep going and that is what I do. (Stacee)

> Make my money and then do what I like to do in life because I might not have a lot of time to do that. I might, I might not. You never know. I just do stuff even though it is a work night, I do not care. (Patricia)

> I'm looking for to be in remission again. (Vanessa)

Imagining a future. The most difficult part of sustaining a commitment was the ambiguity and uncertainty surrounding the course of illness, and the interview-participants' expressed inability to visualize how the disease will progress and what lies ahead—in short, imagining a future.

> [The hardest part is] not knowing what to expect, what is going to happen next, what is going to be taken from you as a person. (Vanessa)

> I was told it could go two ways, limited or diffused.… She [Dr.] told me it might be years before I would be able to, probably know where it was going. One or two years later, she said, "I think it's going to stay limited." (Mackenzie)

> I relied on the doctors, but I did go to three different doctors … just to confirm if this [diagnosis] was true and my long-term prognosis was pretty uncertain. (Bryce)

> I do a lot but I'm not out of the woods. You're never really cured from it and damage that it's done doesn't go away ... but you just never know what's around the corner. (Rene)

A number of participants had arrived at a point where they had accepted uncertainty as a feature of disease and had tried to live with it, in spite of the unease they felt. Others tried to learn as much as they could about the disease and other patients to help them chart a possible course for their illness.

> Well, what are you going to do about it? You're going to sit there, you're going to take it and you're going to live with it and you're going to learn to deal with it. You sit like crying over spilled milk? Lighten up and move on…. It wasn't my attitude in the first month. I can honestly say that. I had a very negative attitude the first month but then after that, I just learned when I started researching it more and finding people who are living longer with it then I had a very positive attitude and I still have a very positive attitude about this. (Winola)

Strategies

In order to cull the data on what strategies patients employed, a general definition of strategy as a guide to action was adopted. The following general strategies were identified: taking on a problem-solving/problem-avoidance orientation, utilizing resources and advocating for themselves, being transactional in evaluating care, being open to experimentation.

Taking on a problem-avoidance/problem-solving orientation. A main strategy for many patients was a focus on problem solving and problem avoidance. Problems might have occurred in encounters to obtain care, brought about due to their disabilities, or manifested in social situations. This strategy included anticipating problems, thinking creatively, being persistent, and treating difficulties as challenges that needed reframing or a work-around:

> This does not work anymore, what do I have to do to make it work. It is just re-prioritizing, re-evaluating. I have not changed my priorities. I just changed the way I do things. Change the way you look at things. (Stacee)

Interview participants used a problem-avoidance strategy in dealing with the healthcare system by anticipating what problems might occur during their doctor's visits, such as previous records and lab results not being available to doctors, making their visits wasteful. Many patients spoke of unknowledgeable emergency room doctors and nurses, and how they had to preemptively be prepared for that. Practically all patients (80%) kept a medical file of lab reports and health records that they would carry along to their visits with doctors and hospitals.

> I have found that having my own records is the best thing because if you go to a doctor and they, for some reason, they didn't get your lab results or test results then sometimes, your whole visit is in vain. (Rene)

Anticipating problems with their energy levels, access to medications, and difficulties outside of home were other areas in which patients showed watchfulness. For example, a strategy to deal with limited energy levels was to partition their days into good or bad days, which dictated how they regulated their activities. Those with a more sever fatigue used the spoon theory to allocate energy within a given day. Spoon theory quantifies the amount of energy a person has as the number of spoons that can be used in a given day, forcing patients to pace themselves and put a limit on what they planned to do. This strategy, in particular, minimized their withdrawals from family and social events and assured that they could take on more arduous tasks.

Utilizing resources and advocating for themselves. Utilizing resources and harnessing the power of others to get what they needed was a strategy that was especially helpful to patients in navigating the healthcare system and other systemic bureaucracies.

> My insurance, there is a program connected with Blue Cross/Blue Shield called health advocate. I love it.… I called the university medical referral and they have kids there who just look at a book and give you the names of doctors and I wasn't successful in finding a doctor then; they give you names but that's all. Then you call the office, "We're no longer taking new patients." So anyway, this health advocate—…They gave me the names of three doctors and they said, "All three of them are taking new patients. Here are their names, their numbers. Call them and interview them." And I did. (Marcia)

Being transactional and evaluative in obtaining care. By taking on a transactional posture and considering metrics such as harm, benefit, futility, trade-off, positive return, cost, reliability, trustworthiness, availability, convenience, and personal preferences, patients were able to check the value propositions of choices in front of them. As such, they expected their doctors' recommendations to produce results and reduce their symptoms; the tests and procedures they were subjected to have an actionable objective; for the side effects to not outweigh benefits; and the medical care

provided to them to meet certain cost, quality, convenience, and preference criteria. If they did not, patients would not assent to doing them.

> If I do not feel any positive return from anything that my doctor is doing, then, something is not right. (Sandy)

> [The test] is uncomfortable and then if they found out there is a problem what were they going to do about it? Could they do anything about it? No, not really. So, I did not do it. (Pricilla)

> My rheumatologist, I asked him, "Well, what's the side effect?" And he'll tell me and I'll say, "I don't want to do that yet" (Sharyl)

Five patients (20%) saw doctors in SSc specialty centers more than 100 miles away from home, while three (12%) had to travel 25-50 miles to see one. For most, the closest centers were located out-of-state and were out-of-network. At least three patients had learned that by splitting the medical encounters—seeing the expert at the centers, but doing lab work or procedures locally—they could be partially covered by insurance. All patients, when asked if they evaluated the care they received, answered empathically yes, and proceeded to provide other examples. The criteria that many patients had developed to evaluate the quality of their doctors was one example. In addition, interview participants showed a high level of vigilance monitoring their doctors and were aware of factors such as preoccupation with research and travel to conferences, busy practices or family lives, retirement mode, or suboptimal office staffs and were ready to react either by expressing dismay or looking for another doctor.

> I did switch up from a very good primary to another primary. I finally found one that I liked a lot and the only reason I left the other primary is I had had him for so many years and I saw a change. He was in retirement mode in his focus and I knew I had to get a new, young doctor. (Lauren)

> I said I had some questions and I pull out my list…. She did take the sheet and look at it. She glanced and saw the amount of writing that was on it [half a page] and she said something about doctors are limited in the amount of time that they're supposed to spend on you depending on whatever the insurance company pays or whatever and she said something about my time was up, and I should ask her the most important question. I

> said, "They're all important." Anyway, I saw another doctor. I got another doctor. It wasn't just that one incident. (Marcia)

Being open to experimentation. Many patients expressed that they did not feel they had any control over the disease itself, but could control what they engage with. Patients viewed experimentation as an option wholly within their powers. They were acutely aware that little was known about the disease, making any treatment experimental, including the conventional medications they were given.

> To some extent, I mean, I've recognized that putting me on medication is not necessarily going to be the answer, because it's trial and error and I recognize that. (Marcia)

All patients claimed they were compliant with medications, meaning they followed doctors' directions in determining the efficacy of prescribed drugs. At the same time, some patients would experiment with dosages of medications that they felt they had some leeway with, such as gastro-intestinal or pain medications.

> A lot of these medicines for stomach, I take them and I wait a couple of days and see how I feel about it. My stomach is easier to find out, if it is working or not, because I kind of know how I felt the day before and the day after. So, what I have done with these medicine, especially the stomach medicines, if I see a difference with me, I stop it for one day or two days, and if I am feeling better, I take it the next day—if something is wrong then I kind of come to the conclusion that it is the medicine. (Francis)

Almost a third of patients, 7 (28%), have participated in nine clinical trials, and 3 patients had actively pursued trials without success, due to strict inclusion criteria or a lack of insurance coverage through which some of the cost could be covered.

> He had tried the Remodulin on me. It just became a pump … I was the first one to get it and it was experimental and I said, "I'll try it if it helps, if it's going to help other people." (Winola)

As to experimenting with complementary and unconventional approaches, 18 (72%) patients spoke of a range of treatments they have experimented with, including acupuncture, occupational and physical therapy, chiropractic medicine, massage therapy, yoga, meditation, nutritional approaches, high dose vitamin therapy, hot paraffin, stone

therapy, Reiki, collagen injection, hot tub, holistic medicine, herbal medicine, homeopathic, and Minocycline therapy (a controversial therapy not embraced by mainstream medicine). Acupuncture, yoga, and massage therapy were the top three choices, followed by nutritional and physical therapy. The main reasons for discontinuing were cost and ineligibility for insurance coverage.

A third of the patients, 7 (28%), who were wary of alternative therapies and have never tried them, perceived the practices as having elements of "quackery" and "voodoo witch," not research-driven, risky, or ineffective. On the other hand, the patients experimenting with alternative therapies believed therapies were not harmful and might have been helpful to varying degrees. Those who have done yoga or nutritional therapy were most enthusiastic about the efficacy of these interventions and a belief that they saw a difference. The sole patient who had done Minocycline therapy firmly believed it was the only intervention that worked in her case. Conversely, patients were less enthusiastic about results from some other choices, like Reiki and acupuncture. Acupuncture, in particular, did not work for many patients due to their skin hardening.

One concern that patients repeatedly voiced was their aversion to taking medications and their desire to minimize the amounts they were prescribed. The willingness to experiment outside traditional Western medicine had availed some patients with relief from a purely pharmaceutical approach, an alternative to ineffective medications, and a sense of doing something natural for their bodies.

> If you do not want to do big pharmaceutical stuff because it makes you sick, what else can you do? I do chiropractic, which I was on meds for headaches that really did not do anything and I do not take it anymore, because I do not need them, I have my chiropractor. It works. (Patricia)

Overall, when patients spoke of experimentation, their efforts were directed at medications, nutrition, exercise, and other lifestyle changes. These were the areas that patients had the most control over. Differentiating the controllable from uncontrollable,

patients were open to investigate alternative and potentially helpful ideas, wherever they could find them. See Appendix L, Table L9 for patients' accounts on strategy.

Learning

Irrespective of the diagnostic path taken and the initial responses they got from doctors, once patients were diagnosed, they all arrived at the same spot, facing the reality of having a disease they had not heard of, an unknown.

> I went in and the doctor really said, "I think you have SSc." And I said, "Okay," because I had no idea what it was. (Nicole)

Twenty-two (88%) patients had never heard of the disease. They had been handed an unfamiliar word—scleroderma—and they had to figure out how to make sense of the unknown, what it was, and more importantly what to do about it.

> In the beginning, it's an unknown and you're constantly searching and reading and looking and wondering and anything and everything, more doctors, more people [to talk to].… My first thought was I had to learn as much as I could when I knew that's what I had. (Brittany)

Patients, by and large, did not grasp the gravity of such a diagnosis until later when they were able to research it on their own. With no prior knowledge of disease and the limited experience with chronic diseases of any kind, most patients wanted as much information as they could find. These information-gathering activities were their first foray into learning. The key areas of learning in interview participants are detailed in this section.

Informally. In their first attempts to gather as much information as possible, patients spoke of utilizing a variety of resources from the Internet in the more recent past, going to libraries, and seeking answers from social workers, nurses, or family or friends in the medical field. Some read brochures they were given, academic papers, and medical journals, and wrote to research scientists. Others contacted random SSc patients, miles away, that somebody who knew somebody had put them in touch with. A few reached out to various support groups (SSc Federation, SSc Foundation, Road Back Foundation,

Pulmonary Fibrosis Foundation), a handful went to educational gatherings, and some tried to learn by going from doctor to doctor. Regardless, most found more questions than answers, confusion, uncertainty, and fright in these efforts.

> Well, of course the first thing they say is don't go on the Internet and that was the first thing I did. That night, I went and I looked it up on the internet and I started reading where it says the life expectancy and so on with SSc is only five years and then I started crying and I had to have my husband finish reading it…. The first initial reaction was I was going to die. (Winola

> When I was reading about SSc, I thought, "No way. I'm not even going to read about this." It was scary. (Brittany)

Two patients took the advice of their doctors and stayed away from the Internet and support groups altogether, leaving information gathering to other family members or none at all. For some, the search was empowering, never-ending, and became an ongoing feature of their lives. For a handful, it subsided after learning the basics, or as one patient put it, "I really believed I was living it. I didn't need to read about it." For a third group, the initial stages of search for information plagued them with debilitating emotions forcing them to pause; their search for information resumed once their symptoms became more stable. Thus, each group curated their informational needs in a particular way. The initial search for the most part was not informative and was the cause of panic and confusion. Over the course of the illness, the character of these searches, sources, and learning would change, best described as curating information.

The word "curate" is used here to highlight the patients' discriminating nature of information seeking, carefully managing and/or limiting the amount, type, or the sources of information. After the traumatic experience of going to the Internet, subsequent to their diagnosis and being flooded with statistics and tales that were difficult to sort through, patients became more discriminating about how they accessed information and how they got what they needed without getting overwhelmed. Having multiple sources of information was how most patients approached this task.

> I speak to other people. I see what other people go through. I look at myself, what I have or have not gone through, and what I'm able to research. (Kurt)

> I would talk to [technicians], for example, during the pulmonary function tests; "Do other people have challenges with this one? Is this as hard for most patients as this one?" Just to try and find out how I stood compared to some other people. (Jenelle)

There were four main sources of information for patients: (a) their doctors, nurses, nurse advocates at insurance companies, technicians, social workers, and researchers in the field; (b) websites, publications, patient brochures, conferences and educational forums of organizations such as the Scleroderma Foundation, Pulmonary Fibrosis Foundation, and well-known research institutions like the Mayo Clinic, the Cleveland Clinic, the National Health Institute, Johns Hopkins Medicine, and PubMed; (c) other patients, known either through participation in support groups or otherwise; (d) certain documents such as their lab reports and doctors' notes.

These sources not only allowed for triangulation of data, but satisfied disparate informational needs: from medical professionals and their medical records, they would get individualized and personally specific data, as well as hints of where they might fit relative to the other patients seen by the same doctors and technicians. From websites and organizations, they would obtain general knowledge, the latest research news, and reports of advances in the field. Other patients served as good informants for how the disease might progress, how it was managed, which doctors they saw and their opinions of them, tips on helpful gadgets, and experiences with taking medications and applying for disabilities.

The majority of patients reported getting their lab reports and doctors' notes; some had set up elaborate filing systems. By asking their doctors for explanations or looking up online, many had learned to read and understand these reports and would follow the trends and changes closely.

Interview participants provided different rationales for their information-seeking. Many equated having information with having knowledge as a means to:

- Understand what disease they had and what could be done about it
- Overcome their fears and feel in control
- Be a "good" patient and better manage their disease, especially as to medications' side-effects
- Make informed decisions and negotiate treatments with doctors
- Face reality and prepare for what will be coming
- Keep tabs on their progression and monitor their health status
- Keep tabs on the quality of care they were getting and catch oversights
- Be aware of new research, medications, and trials for which they might qualify
- Be prepared when facing new doctors or emergencies to educate doctors and nurses
- Distinguish between changes that are SSc-related and those due to other causes

These were summed in patients' statements below that suggested that information seeking in part was to compensate for the inadequacy of a healthcare system that made dealing with this disease daunting:

> To me, information is power. Knowledge is power and I feel, as a patient, the more knowledge I can have, the better I feel about it and dealing with the disease. I think if you just go to the doctor, you're given your pills, you take your pills and that's the end of it, and life could be quite daunting having a disease like this … as I said, it's up to the patient to really educate themselves and knowledge is power and it takes away the fear. (Laurie)

> And I think, over time, you hear about the mistakes and you experience the mistakes and then you say, "You know, I need to educate myself." So, when I'm entering to this [situation], I know what I'm talking about, what I want the outcome to be, and how we got to get there. (Kristeen)

Knowledge was a way to take some control over some aspect of their disease. At least one patient considered being knowledgeable an effective strategy in dealing with doctors, as she could speak their language.

Patients with vital internal organs affected or those at earlier stages of disease tended to utilize all or most of the resources above and expressed more vigilance and determination in keeping up with the advances and latest information in the field.

> I do research on the Internet. I read a lot of SSc research. I read medical journals and I talk to informed patients, which is very different than talking to—not all patients are informed. So, I make sure to talk to other patients that read research and might know something different than I do…. I'm online all the time looking for new treatments and touching base with doctors that either I've worked with in the past who have been my physicians in the past or doctors I worked with through the foundation that I'm still in touch with to see what research they're doing that may be information that hasn't been published, but anecdotal information they might have found. I have a little Google alert. Anytime SSc is in the news that day that comes to me. (Jenelle)

Patients with a limited form of the disease or those who had the disease for a long time and were stable were more content with information their doctors provided at visits and less watchful for new developments; they considered it their doctors' job to be on the lookout, and a few admitted that one becomes complacent overtime.

Six patients (24%) indicated that they intentionally did not seek any information or limited what they read. In particular, they did not want to read the grim news and believed they were in good hands with trusted doctors. Some had other family members who had taken on the task. Having these watchful eyes has relieved the patients from the burden of keeping informed. Not wanting to project into the future and having a constant reminder of the disease were the most stated reasons for these patients:

> My husband filled a binder with stuff he printed for me to read, and I would never read it. I did get a couple of pamphlets about SSc form the doctor…. So, I had enough to know what is there and what it was but I did not want to read everybody's story. (Nicole)

> No, I really don't. I don't get online a lot. Everything I read, sometimes, I find it's worse to read it. I just don't want to project into the future. (Mackenzie)

Concerned about the traumatizing impact and relevancy of online information, doctors and patient organizations warned patients about indiscriminate Internet searches.

This had an impact on how some patients sought information, restricting themselves to websites sanctioned by the medical establishments and written for patients by professionals.

> Dr. ____'s first words to me were, "Stay off the Internet because what you read there, you'll only hear the bad things." So, I never once looked. (Nicole)

> [Using] Internet mainly, but very carefully. I was warned not to read everything. I was guided by the rheumatologist, and I was guided by him and her team. He had a wonderful, wonderful assistant who would give me links and then I would attend patient information days on a regular basis, which I found very, very helpful. (Laurie)

A number of patients had access to both patient materials and academic technical journals, or to researchers in the field who were willing to communicate with them. The latter was not looked upon favorably for one patient:

> I wrote letters to different research scientists, because I was not getting anything from my doctors … and one day I went into his office and I started asking him about different things and he said, "How is it that you know about these things?" I said because I am interested in my own well-being and I did research on it. He said, "Well that is not for public consumption." (Stacee)

Contrary to this assumption of a lack of discernment on the part of patients in dealing with medical information, the majority of patients exhibited an awareness of the need to be discriminating about, contextualizing the information they were getting, and evaluating the trustworthiness of websites.

Socially. The 13 (52%) interview participants who were involved with support groups, and the 4 (16%) who previously participated in some form described learning through and with other patients in these encounters. These learnings included exchanges of information on interventions and doctors. Recommendations on doctors and medical facilities from other patients themselves were considered valuable first-hand information. Other areas of vicarious learning from experiences of other patients were related to description of symptoms, progression, and reaction to medications. Obtaining practical

tips and solutions to deal with disabilities, including applying for disability insurance and helpful gadgets and apparel and where to shop for these, were other areas. Most importantly, through observing others, patients could learn where they might fit in the spectrum of disease. This learning occurred not only in support groups but also educational forums where patients could come in contact with other patients.

Although not part of this study, a number of patients mentioned their learning through online exchanges. The Scleroderma Foundation's portal was the primary site for patients to learn from each other. These online exchanges largely consisted of educating new patients by recommending doctors, discussing medications and nutrition, and giving and receiving hope and encouragement.

Formally. Two major sources of formal education for patients were expert talks provided in support groups and various formal educational forums and national conferences with specific topics and programs. During the initial years of illness, when patients' knowledge was limited and most subjects were of interest, these venues for learning were often attended. Many activities described by support group leaders could be characterized as problem anticipation and avoidance training. For example, patients learned about spoon theory and how to keep medical records in support groups. Patients' awareness and uptake of a problem-avoidance/problem-solving orientation might be partially attributed to the formal learning in support groups.

As patients' level of general knowledge increased and their intentional learning took on a more focused approach, patients reported that these venues ceased to meet their needs and became repetitive or "pedantic." For a number of interview participants, exposure to formal programs about the progression of disease, immediately after their diagnosis and before they had gained a footing, was traumatic and scared them away. These mismatches alluded to difficulties in providing formal learning programs for such a diverse patient population.

Experientially. For the most part, the interview participants learned experientially and incidentally. When asked about their learning, the majority would point to their information-gathering activities and formal learning, unaware of any incidental learning. However, in specific examples, they referenced experiences in which they had learned through trial-and-error, after facing upsetting mishaps, or facing limits imposed on them by bureaucratic systems These experiences had mostly to do with interactions with the healthcare system and dealing with disabilities, and seem to have given rise to the problem-solving and anticipation, and advocacy strategies that were covered under the Strategies section. Experiences through mishaps such as medical tests and procedures falling through the cracks, and dealing with inexperienced healthcare providers and staff were especially poignant.

> I had to learn it and I learned it the hard way by what didn't work. So now, it's 15 years in, I know…. I learned through seeing poor doctors … they had no idea what to do with me. And I know for those two years, I wasn't getting the care that I needed. (Jenelle)

> I've learned, too, over the years that if they can't get it [draw blood] on the first two tries, I have them get somebody else to draw the blood. (Winola)

> When you leave the doctor's office, they give you this nice printout with all of this information on it. You're finished with the doctor so you can't ask any questions about it. So now, I started—the next doctor I see, the form that the previous doctor gave me, I'll ask, "Explain this to me. What does this mean?" (Marcia)

These experiences led to patients learning to advocate for their needs. Maddison, who had a pacemaker that was not working right, could not make any headway with her electrophysiologist and had to resign herself to "it's as good as it is going to be, so let's just leave it at that." Talking to the doctor's nurse, she found out "sometimes, the technician knows more about the device itself than the doctor." Through a technician, she got the device adjusted. Learning from this experience, she got her SSc specialist to assign an intern to research pacemakers, "so when mine is due for the new battery in a

couple of years, he's going to have a list of pacemakers to suggest to my electrophysiologist."

Learning from prior experiences unrelated to their illness was also instrumental in voicing their needs. Advocacy came easier to a handful of patients whose occupational skills transferred well for this purpose.

> Navigating the healthcare system—I guess I just know what to do, because of my work probably…. I know how to get people to help me if I need help like getting to and from endoscopy or getting information I know how – where to call, who reach out to. (Willa)

> I'm a [a client advocacy type of work], and I think I know how to advocate for myself or at least research and go and look for things, go to find me a decent doctor. I have no problem asking people … I just pick up the phone or I just advocate for—I'll just do what I have to do. I think it's because I have a good handle on what's going on with the healthcare system now. (Mackenzie)

Other patients expressed having learned from advocating for their children, and maturing and aging, in general, as helpful experiences.

Another area where patients' everyday experiences led to learning was in dealing with their disabilities and in trying to make their daily lives easier. Patients, in aggregate, had accumulated a host of knowledge in practical solutions, some of which were listed under problem-solving strategies.

Somatically. A majority of patients characterized scleroderma as an illness with a unique combination of symptoms, a varying susceptibility to treatments, and differing from patient to patient. Thus, patients saw themselves as unique.

> Because our diseases have the same name does not mean it is the same disease. (Stacee)

> I do not have the skin issue but have a lot of the other symptoms. Most of the people when I went to the support group they had skin issues. They would not understand joint.… I think that is what makes me unique, you just have a few and, that makes it hard to have a blanket approach how to fix things. (Patricia)

> I have different pain than anybody I've ever spoken to, and then right now, I'm having GI and lung issues that is different from what some other people I've talked to have experienced, but it's nothing unusual. But to have the certain combination is weird and not to have— … I think the challenge with this disease is that it hits every single person differently. Nobody has the same symptoms. (Jenelle)

Patients not only reported on the specific set of symptoms afflicting them, but also the changes (additions, reduction) of symptoms during the course of the disease, and waxes and wanes in shorter intervals, known as flares, making for a dynamic phenomenon. Given the perception of a unique and individualized illness, and the experience of a dynamically changing disease, many patients exhibited a high degree of somatic knowledge and a focused attention on their bodies as unique and different from those of other patients. In short, they "individualized" it.

> You need to individualize it, because our disease manifests individually in each person. (Kurt)

> I think this disease is really an individual disease and that it affects individuals very differently. (Maddison)

Many, in particular, patients who were athletes prior to their illness, claimed they were always body-aware. One third of patients used the phrase "I know my body" in their interviews.

> I know my body and know what's normal and what's not normal and even with all the craziness that goes on, you know what isn't right. (Jenelle)

> I'm a very attentive and intuitive for my own changes in my body and that's a huge thing to recognize.… I probably do that too much. I do pay attention. The biggest thing for me is seeing how far I can stretch (physically) because that's telling me whether I'm losing ground or not. (Lauren)

Many patients knew that the onset of a flare-up (exasperation of symptoms), the sequence of symptoms occurring, could differentiate between pain and ache, or fatigue and being tired, but were not readily able to verbalize how. These were conveyed through imagery such as: flare-ups occurring like a cascade; the intensity of pain forcing one into a fetal position for days; being made out of wood; feeling my body is angry; and every

molecule of your body being tired. Sandy, being aware of her inability to express her somatic perception of fatigue, put it this way:

> I do not necessarily describe it [fatigue] in the medical format but as far as feeling: I was walking through the door and somebody smash my whole body, just overwhelming you are tired from inside out.… I think that for me it is the biggest indicator [of disease], because like my body is not tired like oh I had a crazy day I am really beat. It is like a tired from full encompassing like your every molecule of your body. (Sandy)

To some degree or another, all patients, through inexplicable internal mechanisms, or external observations and instruments, were gauging the changes in their bodies. Skin alterations entailed significant physical changes for many patients. These changes were visibly and tangibly available for examination in a more direct manner. Other variations, like changes in breathing or fatigue, were measured indirectly. Many had developed assessment methodologies, including using instruments such as blood pressure monitors, oximeters (oxygen saturation monitors), wearable health gadgets; taking pictures of skin changes overtime for comparison; tactile feel of their skin for degree of softness; using walks and exercises to appraise stamina and energy levels; and quantifying the amount of pain, skin discolorations, and extent of body stretch. Patients were particularly sensitive in regard to their breathing, using exercise and climbing stairs and general exertion as tools to detect changes.

> If you get winded and you exercise regularly, then there might be an issue with your lungs. But if you do not exercise regularly and you get winded you get confused whether it is being out of shape or it is PAH…. So that way I could distinguish. (Willa)

Another marker was the quality of fatigue. Patients could make a qualitative distinction between fatigue from the illness and tiredness due to overexerting. Physical responses to medications, from Advil to chemo, were used to gauge the state of bodies. Those who fluctuated between flare-ups and periods of calm used the contrast between the two phases and how their bodies felt ramping up to a flare as a rubric.

A few patients expressed that they were bodily-aware even before their illness, but the majority attributed their somatic focus to the illness itself. Through body monitoring, they had learned and developed a niche expertise, based on which they could monitor the progression of the disease, determine the efficacy of medications, check against errors and mishaps with their medical care, and advocate for themselves when faced with unknowledgeable healthcare workers.

> I think he [Dr.] was on vacation as well, but I went to the emergency room. They didn't help me at all, and because I know my body, I know what's going on and I know what helps with the pain and they just looked at me like, "Oh, someone is here for just pain meds or something." (Winola)

Being attuned to their bodies and learning somatically, in addition to comparing themselves to other patients, were the mechanisms with which they individualize their illness.

Learning informally, formally, socially, experientially, and somatically, patients made sense of their illness and developed high levels of knowledge and expertise. Empowered by their own knowledge, at least ten patients alluded to a possible shift in how they have come to view the nature of received knowledge.

> I think at the beginning, I just believed whatever they told me.... I don't know when it just started to change, but I began to realize they get up in the morning and just like me every day and they may have gone to school for something but they don't know everything and I'm living it—so maybe I know, too. (Kristeen)

In addition, many patients became purveyors of information to other patients, friends, relatives, and medical professionals. The kind of expertise a number of patients had developed was in the area of tests and procedures, assessment of severity, and an ability to recognize undiagnosed individuals based on visible symptoms. Patients also reported educating emergency room doctors and medical students. Emergency rooms and unexpected hospital visits were particularly problematic for these patients, as healthcare professionals dealing with acute diseases and accidents were reported to be among the

least informed about SSc. All patients who had such a visit considered the experience painful and identified the occasion as an instant when they had to educate the doctors. In addition, their own rheumatologists at teaching hospitals had asked a number of patients to speak to, or be examined by, medical students. To these requests, they had happily responded. Their motivation was their annoyance with the lack of knowledge among healthcare workers, and their attempts to do something about it.

Social Interactions in Support Groups

Among the interviewees, eight (32%) have not participated in any support groups beyond a few trial visits, if any, four (16%) were associated with support groups in some capacity in the past, but have not been active or participating for a while, seven (28%) acted as support group leaders or co-leaders, and six (24%) considered themselves members who attended meetings with some regularity. Two of the leaders had served on the Chapter's boards in two different states.

Format. The leaders and members described the format of support groups in various ways, indicating there was not a pre-determined format followed by all groups. The number of attendees seemed to have determined the level of formality of format. In small groups, members updated each other of new problems and developments in informal settings, while larger groups had public meeting spaces and more formal programs with guest speakers and themes. Dealing with the psychological burden, making social connections, having positive interactions, and sharing information were the major themes.

Members' interests in topics and their suggestions shaped the programs, but leaders brought their own visions, perceptions of needs, personalities, strengths, and resources to how they conducted the meetings. For one leader, helping new patients was the priority; a second leader felt patients wanted to connect and needed opportunities to speak and get positive hope from others. A third leader believed in positive meetings based on fun and

laughing, and covered topics such as coping with holidays and stress, depression, getting your doctor to listen to you, and the best way to prepare for appointments. A fourth leader was against guest speakers, believing such information was available online. Instead, calling the approach psychological/emotional, that leader facilitated discussions around topics such as acceptance and guilt. Access to resources and educational background influenced some of these choices. For one leader who had worked in a health-related field and had contacts in the medical community, invited guest speakers were a regular feature; two others with psychology backgrounds had their focus on emotional aspects, while an outgoing leader had the focus on social aspects and inclusivity. Leaders made the point that they took feedback from their memberships in choosing topics and activities. A training manual was available from the Chapter's office, and leaders spoke of an annual weekend retreat where group dynamics and facilitation skills training were offered. However, the leaders were volunteers and patients themselves, and they did what they could with the limited formal training available.

Leaders' involvement. Five of the eight leaders (7 current, 1 previous) had initiated a group, or took over one when no one was available to lead. The motivation for leading a group was expressed as sharing their experiences and knowledge with other patients in order to support them in this journey. Due to the length of their illness and the experiences they had accrued, a relevant educational background or skills, certain traits or personality, or a vision for a more effective group, they could contribute something of value to other patients.

> The first time I started a group was because I thought, "There are two people. There's got to be more." And they were scared and so I was helping them through it. (Leader 1)

> I do it because when I was first diagnosed with it, I wished there was one here for me to have someone to talk to that could understand what I was going through, someone that knew. (Leader 2)

> I realized with all my qualms and all my frailties I am stronger than a lot of people. So that's why I'm doing this. (Leader 4)

> I think the main reason is because so little was known about it. I was looking for other people to share with. (Leader 7)

> There are support groups, [the leader] starts every group pretty much, "We're all going to die from this so let's just talk about it. Let's have a bitch session," that's not my way of doing it. I was, more, "You know what, we can talk about that and we can grumble about it but before we walk out the door, we're going to be laughing and we're going to find something that we can do today to make something better." (Leader 8)

Except for one person, all the leaders got involved with support groups in later stages of their disease when they felt stable, both physically and emotionally, to do so. Leaders viewed involvement of group members in managing their illness as a spectrum ranging from very involved to none. Involvement was described as being very informed, active, proactive, and showing strength and resilience, while the lack of involvement was characterized as being negatively impacted by fear and denial, trying things indiscriminately, inability to work with their doctors, and not being open to proven suggestions and guidance available at support groups.

> I think the majority of SSc patients are amazingly informed … and then you got the other patients that really are just, "I'm not dealing with it. I'm just going to go on with my life." (Leader 8)

> The people that tend to feel sorry for themselves are really the ones that don't end up doing that well, because they kind of are in that mode of, "Poor me. I can't do this. I can't do that." (Leader 7)

> There are those who are afraid to talk to their doctors about their symptoms—to those who think they know more than the doctors. (Leader 5)

> Some people become very strong all of a sudden. All of a sudden, they find their inner strength. I've seen people who you'd think nothing in the world would stop them. (Leader 4)

Members' involvement. The patients who had joined a support group characterized their involvements as an opportunity to (a) have social interactions with people who understood, (b) learn; and (c) join forces with others for a greater cause.

Social interactions were seen as an opportunity to converse with people who knew what one was going through; to make friends with people suffering from the same illness; to commiserate and give and get moral and emotional support; and to laugh and uplift one's spirit. Learning included finding out about doctors and medications from other patients; observing other patients for symptoms, disease progression, and how others handle it; learning best practices from other patients; sharing of news and materials from conferences; accessing books and materials provided by the group; and partaking in discussions and talks given by guest speakers. To join forces with others for a greater cause or purpose included organizing events such as walks, fund-raising, public awareness to improve research and funding, and outreach to new patients.

Non-members' non-involvement. Interview participants presented a number of reasons for not joining a group or participating in educational programs organized by groups or the Chapter. These included logistical issues such as other commitments (children, working) and lack of time; fatigue and lack of energy to participate; distance and lack of transportation; and inconvenient meeting times or inconsistent meeting schedules. Personal issues consisted of not being ready for group interactions; being frightened by seeing members having severe disabilities or dying, or speakers talking about progression of disease; not wanting to know the worst, or projecting into future; not wanting to look at oneself as being sick (belonging to a patient community); not needing a constant reminder of illness through monthly group meetings; not wanting SSc to be all-consuming by doing yet another SSc-related activity; doctor recommended against it; and the belief that did not need it, or it is good for new patients or severe cases. Issues related to group dynamics consisted of not feeling the group is a good fit; whining and defeated attitudes of other patients; or too few people attending. Lastly, the content and programming issues encompassed meetings being perceived as social events, or not having enough opportunity to socialize; exchanges of unscientific or non-factual

information among members; the focus of materials on new patients; topics not of interest or repetitive.

Among these, two reasons stood out as major impediments:

- Six (24%) patients stated the fear invoked by seeing other patients with severe disability, or hearing the worst from other patients or speakers, caused them to back away from participating:

  > I was scared of what I might hear so I kind of stayed away. (Sharyl)

  > Just the appearance of people, how you're going to look, you picture yourself in there, it was too much for me to bear. (Vanessa)

- Eight (32%) patients considered negativity or the complaining demeanors of other members in the group as off-putting:

  > A lot of patients, I could say this to you, whine too much and because I don't feel that that helps us, I don't want to hear it and I think it brings me down. (Lauren)

  > I got into the support group and I really hated the whole thing. Because all they do is whine to each other about how bad they feel, and I do not find that to be helpful either. I am not going to let this disease control me. This is what I have, not who I am. (Stacee)

Summary of Qualitative Findings

From among respondents wanting to be interviewed, 25 SSc patients were selected for interview. These included 7 leaders of support groups, 6 members of support groups, and 12 non-members, consisting of 20 females, and 5 males. The sample was predominantly female (80%), White American (72%) who tended to be middle-aged (50-55), more educated, held white-collar jobs, and were more likely not to be gainfully employed at the time of interview. The average disease longevity was 14.5 years. The major self-reported SSc subtypes consisted of 11 (44%) limited cutaneous scleroderma (lcSSc) and 9 (36%) diffuse cutaneous scleroderma (dcSSc). Fourteen (56%) had major

internal organ (lung, heart, and kidney) involvements, while 11 (46%) had symptoms limited to skin, joint, gastro-intestinal, and musculo-skeletal systems.

Patients engaged with various types of work or efforts to mitigate the physical and emotional/psychological, relational, and financial impacts of disease in their lives, including obtaining a diagnosis, managing relationships with doctors, pursuing effective therapies, attending to emotional and psychological impacts of the disease, dealing with employment and financial impacts, and handing familial relationships. The motivation to engage with these activities and work was predicated on a commitment to engage with the illness, and sustaining that commitment through hope, a will to live, and envisioning a future.

Participants described four strategies as a guide to action: taking a problem-solving/problem avoidance orientation; utilizing resources and advocating for themselves; being transactional and evaluative in obtaining care; and being open to experimentation. Participants learned informally by looking for information on the disease; socially in support groups; formally by attending talks and conferences; experientially through problem solving; and somatically by body-listening.

Support groups' format was described as informal and seen as an opportunity to not only facilitate voicing of experiences and concerns by members, but also allow leaders to share their own experiences, skills, and visions with other patients. Members saw support groups as providing opportunities to interact, learn, and join forces with other patients. Two major impediments to membership were described as fear invoked by seeing other patients, and the negative and complaining attitudes of other patients.

Chapter V

DISCUSSION, INTERPRETATION, SYNTHESIS, AND CONCLUSIONS

This chapter presents discussions, interpretations, and synthesis of results and findings in relation to the three research questions that this study had sought to answer. The discussion of quantitative findings consisting of the interpretations of results in the context of literature are presented first. This is followed by the interpretation of qualitative findings, mainly through the conceptual framework of the study and the related studies that were presented in Chapter II. The quantitative results and qualitative findings are integrated in answering research questions 1 and 3.

The purpose of this study was to explore and describe, through scleroderma patients' own perceptions and understanding, their commitment to illness management, including how they were involved in dealing with their illness and how they learned to do so. Additionally, an understanding of the role of social interactions, in particular, support group participation in their involvement, was sought. The two main discussion sections, quantitative results and qualitative findings, are organized by the three research questions of the study.

Discussion and Interpretation of Quantitative Results

The discussion and interpretation of the quantitative findings related to levels of activation, perceived social support, and any association with demographic and clinical attributes are presented in this section to elaborate on answers to research questions 1 and

3, and accordingly are organized by these two questions. These are prefaced by a discussion of the participants' characteristics.

Participants' Characteristics

The demographic and clinical variables of the study were compared to other studies of scleroderma patients to detect any differences that potentially might impact the discussion of results.

Thirteen recent studies on scleroderma patients (Achaval, Kallen, & Mayes, 2013; Bauer, 2013; Elhai, Meune, & Avouac, 2012; Leavens, Patten, & Hudson, 2012; Merz et al., 2013; Muangchan, Canadian Scleroderma Research Group, Baron, & Pope, 2013; Muller, Rehberger, Gunther, & Schmitt, 2011; Radvanski, 2015; Richards et al., 2003; Sandusky, McGuire, & Smith, 2009; Singh, Clements, Furst, Maranian, & Khanna, 2012; Taillefer et al., 2010; Thombs et al., 2015) were considered. In addition, following scleroderma registries (EUSTAR—European Union SSc Trial & Research Group, the German Registry, and University of Pittsburg SSc database) were consulted indirectly through other studies (Meier, Frommer, & Dinser, 2012; Hunzelmann, Genth, & Krieg, 2008; Steen, Domsic, & Lucas, 2012, respectively), and the Canadian SSc Research Group (CSRG, 2014) through its website, due to lack of a direct access. The demographic and clinical data from these studies are tabulated in three tables in Appendix K.

Among studies reviewed, sample size and recruitment procedures encompassed a wide range, such as recruiting participants at SSc centers during doctor visits, patients in database registries during their annual visits, patients in a given geographic area served by certain clinics, patients attending support groups, conferences, and through mail, emails, magazines, and newsletter ads, as well as cross-sectional, retrospective meta-studies and historical records, resulting in samplings that ranged from random to convenience sampling. In spite of variations in each study's purpose and non-conformity in collected data and reporting formats, this broad spectrum of studies provided an

overview of the SSc population and helped locate this dissertation's sample characteristics within this larger pool.

Gender, age, general education, and employment characteristics of this dissertation study were well within the ranges presented in the 13 studies considered. The only exception was the percentage of college graduates, which was higher, at 34% vs. 27% of the reported upper range. Although within range, the mean age of this study's participants was on the higher end of the spectrum, indicating that the population of this study was somewhat older.

The distributions of disease subtypes were comparable to other studies, which did not recruit their samples from a medical setting. Among the 13 studies reviewed, those that recruited their participants at clinics, medical settings, or during doctors' visits tended to have a larger diffuse subtype in the sample. This reflects the fact that, rheumatologically (hence medically), the interest in scleroderma is in its systemic form, not the localized subtype (Mays, 2008). On the other hand, studies using mail, ads, non-clinical venues (conferences, support group meetings), or historical data had higher percentages of patients with limited subtype. This dissertation study, did not recruit through a medical setting and was prone to the sampling bias with more limited and localized respondents. This self-selection bias may be attributed to limited and localized patients having fewer internal organ involvements and presumably being more willing or able to participate in a study or survey outside a medical setting. On the other hand, those with diffuse have more frequent medical visits and are targeted for more recruitment during these visits.

Among studies reviewed, a study by Radvanski (2015) conducted in the general geographical area of this research shared many similarities in its demographic compositions. In particular, Radvanski's study of a random sample of 68 patients recruited from a scleroderma clinic at Rutgers University/Robert Wood Johnson Medical School reported a high educational attainment: 77% with "some college or above,"

similar to 80.6% of this study (53% having "some college or a college degree" similar to 47.7% of this study, and 24% attended graduate school vs. 32.9% of this study). The higher educational attainment in both Radvanski's study and this dissertation study may be attributed to the geographic area from which these two samples were drawn and whose residences rank among the top ten states in having college and advanced degrees. According to data from the United States Census Bureau (2009, 2011), 36.5% of adults 25 years or older in Connecticut have a bachelor's degree, followed by 35.8% in New Jersey and 33.2% in New York. Another 4.5% of Connecticut adults have an advanced degree, with New Jersey and New York at 4.1% and 3.8%, respectively. These statistics are as high as 50-60% for a bachelor's degree in some counties within these states. Thus, the region of country from which this sample was drawn may partially account for the high educational attainment seen.

In terms of disease subtype, Radvanski's (2015) study had a 57.4% to 42.6% split between diffuse and limited, whereas in this study the distribution consisted of 34.3% diffuse; 43.3% limited; 5.0% sine; 9.5% were localized; and 8.0% did not know their disease subtype. The difference may be attributed to Radvanski's recruitment at a scleroderma clinic where verifiable diagnoses were available through medical records, and where localized subtypes were excluded, versus the self-reported diagnosis for this study. During the interview portion of this study, some level of discrepancy between the disease subtypes reported and the descriptions of symptoms and progression of disease were observed for a handful of participants. It was apparent from interview participants that not all doctors made a clear distinction between disease subtypes to the patients. This is mainly due to the dynamic and uncertain progression of the disease early on (the first 3-5 years), which made the classification difficult, and perhaps also due to a desire not to alarm the patients by typecasting them. In addition, not knowing the subtype was more prevalent among older participants or those who had the disease for a long time

(>20 years). Self-reporting of disease subtype, therefore, should be viewed with caution, especially in older patients and those with relatively long disease duration.

This study's mean age of 60.0 $\pm$ 13.6, even though on the high side when compared to other studies, aligned well with Radvanski's study mean age of 60.3 $\pm$ 12.2. Scleroderma is mostly diagnosed in middle-age patients (40s to 50s age groups). The study sample was left-skewed by participants who had the disease for more than 10 years and hence the higher mean age of 60.0 at the time of survey.

The employment status of Radvanski's sample and this study was very similar (full-time 25.0% vs. 26.9, part-time 8.8% vs. 8.5%, homemaker 4.4% vs. 5.5%, retired 36.8% vs. 34.8%, disabled 17.6% vs. 22%, and unemployed 5.9% vs. 2.5%). Over half the participants (56.8%) in this dissertation study were retired or on disability, and another 35.4% were employed full-time or part-time. While the percentage of those having a "college degree or above" was 67.2% of the sample, they constituted 72.2% of those working full-time or part-time. The higher educational attainment of this sample may imply a higher probability of holding white collar jobs with better benefits (retirement/pensions, insurance coverage, etc.). Such benefits might explain the employment status seen, where many could continue working due to less physical demands of their jobs and wanted to work for retaining the needed benefits. Sandqvist and Eklund (2008) reported that greater working ability was correlated to less physically demanding jobs in a study of SSc women. A number of interview participants holding white collar jobs reported accommodations (switching to less demanding positions, flexible hours, time off, etc.), which allowed them to continue to be employed. Nguyen, Poiraudeau, Mestre-Stanislas, and Rannou (2010), in a study of employment status and socio-economic burden of SSc in a French sample, suggested that employment status in SSc "could be related more to perceived health status and disability than to specific organ involvement" (p. 987). However, in this dissertation, those on disability or sick leave had more significant organ involvement than those holding employment: three times heart,

1.5 times lung and joint/muscle involvements. Age and disease duration distributions were similar for both groups.

The findings from the interview portion of this dissertation study suggested other confounding factors, such as level of accommodation afforded by employers, the availability of insurance coverage and income through a spouse, and an impending retirement age factored in patients' decisions to continue to work or not. Those who sought disability insurance found the process to be a difficult and complicated task requiring certain amount of savvy, knowhow, or financial resources to successfully navigate the program's eligibility requirements. Having had a relationship with physicians who worked with patients to provide documentation for disability insurance application or having had access to legal resources were crucial in this process for a number of interview participants. Patients seeing scleroderma experts are more likely to get this type of support from their doctors, as these doctors are keenly aware of the burden faced by patients and are experienced with providing documentation for off-label use of medications to insurance companies. All these present a more nuanced picture of the employment status of SSc patients in the U.S. than was suggested by Nguyen et al.'s (2010) study.

As to organ involvement, the reporting of lung and heart involvement was higher than in the 13 studies reviewed. The higher levels of lung and heart reporting may be explained by the larger number of older participants and the substantially larger number of participants with longer disease duration. The lungs and heart in SSc patients will be impacted with passage of time, even for those with the limited form. It is estimated that up to 90% of SSc patients develop some degree of lung disease detectable by high resolution CT scans, while in some 40-75% of patients, the changes will be seen in their pulmonary function tests (i.e., breathing tests) (Herzog et al., 2014; Solomon et al., 2013). Since lung disease and pulmonary arterial hypertension (PAH) are considered the leading causes of death in SSc (Solomon et al., 2013, Steen & Medsger, 2007), patients

are screened carefully and regularly. By and large, patients have a clear understanding of the degree of impact of SSc on their lungs and heart and can accurately self-report them in surveys and questionnaires. Therefore, reporting of these organs' involvement in this study may be viewed as reliable.

The moderate association between fatigue and heart and lung problems found in this study, in light of higher reporting of lung and heart involvement, may explain the higher fatigue reporting. Another contributing factor may have been treatment with immunosuppressant drugs, which are known to cause fatigue. In lung and heart patients, due to the criticality of these organs, such treatments are prescribed more aggressively, which might have contributed to the higher reporting of fatigue in these participants.

For this study, the numbers of participants reporting fatigue, pain, and depression were at 80%, 70%, and 34%, respectively; all three were associated with each other in this sample. This finding is corroborated by findings in a number of other studies, such as the correlation of fatigue, pain, and depression in a cross-sectional study of 659 Canadian patients (Thombs, Hudson, Bassel, & Taillefer, 2009), and correlation of pain and depressive symptoms in a study of 121 patients from Johns Hopkins and the University of Maryland Scleroderma Center (Benrud-Larson, Haythornthwaite, Heinberg, & Boling, 2002).

Another finding of this dissertation study was the moderate association between pain and joint/muscle and the moderately high association of fatigue and joint/muscle. Joint/muscle issues were reported by 77.3% of those who were "disabled/on sick leave," who also reported suffering from pain (90.9%) and fatigue (93.2%). A study by Muller et al. (2012) of 84 patients in Germany found that those with musculoskeletal system involvement who were in pain had the greatest disability in performing everyday activities. In another study, pain was reported as "the single strongest predictor of physical functioning" (Benrud-Larson et al., 2002, p. 272). These reports seem to explain

the association of pain and joint/muscle issues and the levels of physical disability seen in this dissertation.

Thombs, Bassel, McGurie, and Smith (2008) compared the fatigue levels in 106 SSc patients with those levels reported in the general population, cancer, and rheumatic diseases. SSc fatigue levels were significantly higher than levels found in the general population, but were on par with levels seen in patients with rheumatic diseases or patients in cancer treatment. Lankveld, Vonk, Teunissen, and van den Hoogen (2007) found fatigue to be the "most annoying" stressor among 138 SSc patients in a study in the Netherlands, indicated on a 4-point scale ranging from "not annoying at all" to "very annoying." In the interview portion of this dissertation study, almost all interview participants expressed a similar sentiment that fatigue and not pain was the most bothersome symptom they battled. In this dissertation study, fatigue was associated with both pain and depression. In turn, pain was associated with sleep disturbance

In a meta-study of eight articles on depression in SSc, Thombs, Taillefer, Hudson, and Baron (2007) found consistently high levels of depression (36-65%) reported. The reporting of depression level in this dissertation study was within the above reported range (34%) overall, but reached as high as 56.8% in participants who had pain and were on sick leave. Lankveld et al. (2007) did not find any significant correlations between disease duration, education, employment, and depression in scleroderma patients, nor any difference in depression between limited and diffuse subtypes. However, they found a high correlation between depression and pain, and depression and fatigue. In this dissertation study, depression was moderately associated with body image distress and pain, and strongly with fatigue.

In summary, this study's participants were more educated and, although within range, were older. Higher educational attainment may be partially explained by the geographical location from where sample is drawn. Other disease characteristics and demographic make-up of this study fell within the general characteristic of SSc samples

of other studies, such as gender divisions and employment. The disease subtype distribution of this study was in line with studies recruiting in non-medical settings. Furthermore, the self-reports of fatigue, pain, and depression were comparable with a number of studies using measurement scales for these variables. In particular, self-reports of lung and heart issues, and prevalence of fatigue in these patients, may be considered reliable. Overall, the impact of higher educational attainment needs to be considered in the interpretations of results.

Research Question 1: Description of Patients' Commitment to Illness Management

Research question 1 sought an answer to the following research question and sub-questions: How do scleroderma patients describe/understand their commitment to illness management? Did commitment to be involved change overtime? If so when and how?

Using the PAM scores of the sample as a proxy for commitment and involvement of patients in their own care, the distribution of PAM activation levels provided a quantitative description and an overall sense of activation of the sample. The PAM activation construct is based on an individual's belief, knowledge, motivation, and actions regarding their self-care. It assumes that activation is manifested by the belief that one has a role to play in one's own care, and has the knowledge and skills to manage the condition and obtain quality care (Hibbard et al., 2004, 2005). Interpretation imposed on PAM levels placed 15.8% of the participants at the lowest level, indicating that these patients were passive recipients of care and were not cognizant of having an active role. At the next level, 20.2% of the participants were lacking fundamental knowledge of their disease and treatments. The third level had the highest proportions of participations (48.6%). These patients were knowledgeable about the key facts related to their condition and behaviorally showed signs of action, but lacked the skills and confidence to bring about new health behavior. The top 15.3% of the participants were those who took action

and adopted new health behaviors for the most part, perhaps finding the behaviors unsustainable only during a crisis or stress.

PAM has been validated for a number of chronic diseases, including multiple sclerosis, which is also considered an autoimmune mediated disease. However, PAM has not been validated specifically for SSc, and no other study of scleroderma patients with this measure existed. Studies using PAM in chronic diseases and conditions, including MS (Staerk, 2015; Stepleman et al., 2010), diabetes (Remmers, 2008; Tabrizi, Wilson, & O'Rouke, 2010), COPD (Halding & Grov, 2017; Korpeshoek, Bos-touwen, de Man-Van Ginkel, & Lammers, 2016), chronic heart failure (Do, Young, Barnason, & Tran, 2015; Witt, Benson, Campbell, & Sillah, 2016), chronic renal disease (Bos-Touwen et al., 2015), spinal cord injuries (Houlihan, Brody, Everhart-Skeels, Pernigotti, & Burnett, 2017), cancer (Mazanec, Sattr, Delaney, & Daly, 2016; Salgado, Mackler, Severson, & Batra, 2017), as well as mixed chronic and healthy general populations (Hibbard & Cunningham, 2008; Skolasky et al., 2011; Wong et al., 2011) reported a range of PAM levels distributions that varied widely across studies.

This dissertation study's distribution levels of 15.8%, 20.2%, 48.6%, 15.1% were in the general range (64%-70% at active levels 3 and 4) of two studies with certain similarities in the sample populations: Blackmore et al. (2016), in a large sample of 3,636 older patients over age 64 (mean age 75 $\pm$ 6.8) found a distribution of 15%, 16%, 45%, 25%). Witt et al. (2016), in a study of 157 women with heart conditions (mean age of 63 $\pm$ 10.8), reported a distribution of 14.6%, 12.7%, 38.9%, 28.7%. Similarities with Witt et al.'s study included age, gender, nature of issues faced with the illness, sample size, and inclusion of support group members, while older age and one or more chronic conditions and employment status were shared features with Blackmore et al.'s study.

A number of studies indicated an association of education with higher activation (Salgado et al., 2017; Skolasky et al., 2011; Smith et al., 2013; Stepleman et al., 2010). In this dissertation, higher educational attainments had a weak association with higher

activation. The general high educational attainment of participants might have precluded the detection of a stronger association. In 16 PAM studies reviewed, gender was not reported as correlating with PAM levels, and reports of association of activations and age, race, disease duration, and comorbidity were mixed and inconclusive. Hibbard and Cunningham (2008) reported higher activations in patients with multiple chronic conditions or more serious illnesses like cancer.

In this study, a moderate association between disease duration within the first decade of illness and activation was detected. PAM is intended to measure "an individual's self-concept as a self-manager" (Hibbard, 2014) and "reflects the degree to which one feels 'in charge' of one's own health" (Hibbard & Cunningham, 2008, p. 3). It is shown to have a developmental and hieratical nature, and "patients need to sequentially pass through each of these stages on the way to becoming effective self-managers" (Hibbard et al., 2005, p. 1928), indicative of learning. This developmental aspect presumably, occurring over time and as a result of experience of dealing with the disease and learning, may explain the association found in this dissertation study between higher activation and longer disease duration within the first decade of the disease. Alternatively, the quantitative findings indicated the existence of a period of shock post-diagnosis and lasting up to five years for some patients. During this period, patients were not able to engage with their illness in a meaningful way, as they needed to "wrap their heads around" the situation they found themselves in. Once patients got past this period, they would engage with their care in a more intense manner.

Within a broader definition of patients taking an active role in their own care, PAM addresses aspects of engagement relating to the physical impacts of the disease. One assumption of this study was that physical impacts were the primary, most direct, and tangible consequences of having a disease, an active involvement in physical aspects was an indicator of an overall involvement, and the PAM instrument would capture the construct of active involvement. However, scleroderma is a complex disease, and some

items in the PAM-13 measure posed problems for patients, as indicated by their notes on the margins.

Interview findings shed light on some of these difficulties. For example, item 8 ("I understand the nature and causes of my health conditions") garnered comments such as "Doctors do not even know," "Hard to answer," or "Experts cannot be 100% confident." For item 10 ("I know how to prevent further problems with my health condition"), the remarks indicated that the respondents knew what would be helpful (exercise, walk) but found it difficult to implement due to fatigue and/or pain. Therefore, non-adherence was caused by physical impediments. Item 3 ("I know what each of my prescribed medications does") was more difficult to endorse by some patients than was expected. This might have been partially due to the experimental nature of treatments where medications are used off-label, making it difficult for patients to connect them to symptoms being addressed, even if they researched them. It also may be due to the systemic and overlapping nature of this disease entailing the vascular, connective, and immune systems that are difficult to disentangle for patients, either through their own research or doctors' explanations. Patients saw the disease as one entity, whereas it was treated at different organ levels and body systems, pharmacologically. Item 12 ("I am confident that I can figure out solutions when new situations or problems arise with my health condition") was harder to endorse than item 13 ("I am confident that I can maintain lifestyle changes, like diet and exercise, even during times of stress") for this study's sample, because the interview data revealed the ambivalence in differentiating between possible new symptoms and those related to aging, genetics, or other illnesses. This was particularly difficult for older patients, who were dealing with the effects of aging at the same time, and newly diagnosed patients, who perceived every new symptom as signaling the worsening of their conditions. Lastly, item 13 ("I am confident that I can maintain lifestyle changes, like diet and exercise, even during times of stress") was problematic for some SSc patients to endorse because many had disabilities that

made adapting and adhering to lifestyle changes difficult. For others, since their disease waxes and wanes, the "times of stress" can easily be interpreted as flare-ups. Flare-ups can last for many months, could consist of debilitating cascades of symptoms, and may entail profound fatigue. Obviously, patients won't be able to maintain lifestyle changes during such stressful periods of flares, and for many SSc patients, the "right" answer that more accurately reflected the realities of the disease might have indeed been to disagree with this item.

On the other hand, some items were easier to endorse than the instrument's scaling was predicting, such as items 6 and 7 ("I am confident I can tell when I need to go get medical care and when I can handle a health problem myself" and "I am confident that I can take actions that will help prevent or minimize some symptoms or problems associated with my health condition"). By and large, the interview sample had an awareness of the criticality of their conditions and the understanding that they could not ignore symptoms and problems. This, coupled with a similar understanding on the part of doctors, meant that many participants felt confident and were able to reach their doctors when symptoms or problems occurred. Furthermore, the reported treatment compliance by interview participants was at 100%. Participants may have interpreted their compliance with medication and doctor's recommendations as taking actions to minimize and prevent symptoms.

A number of other studies noted similar variations in scaling (Packer et al., 2016; Staerk, 2015; Stepleman et al., 2010), but concluded that PAM is a reliable and valid instrument in assessing patients' activation. Nonetheless, these variations point to nuances that were difficult for respondents to resolve, which might have pushed some participants from higher activation levels to lower ones, resulting in some degree of under-reporting of activation.

A different type of under-reporting was observed in interview patients. Six participants who exhibited high degrees of disease knowledge, awareness of and

utilization of expert and quality medical care, and displayed high levels of vigilance and initiative in care for themselves were scored at level 2 and level 3. These participants avoided the emphatic endorsements of items in general, and items 12 or 13 in particular. The expertise of these patients and awareness of the nuances of disease might have worked against endorsing items in the general sense that was intended in the instrument. These observations lead to a suspicion that activation levels of patients were higher than what was captured by the PAM instrument.

The quantitative findings do not provide answers to question 2 of the study. Findings related to the third research question are discussed next.

Research Question 3: Impact of Social Interactions on Involvement and Learning

The third research question was concerned with the question: "How are social interactions, particularly, support group facilitation/participation, perceived in terms of involvement and learning?"

The findings indicated that the mean and median for all dimensions of social support ranged from 67.01 to 76.68, and 65.6 to 91.7, respectively. The dichotomized MOS scores, with the median as the cut-off for high and low values, indicated that 50.3% of the respondents perceived having high overall support, 53% had high tangible (TAN) support, 72.1% had high affectionate (AFF) support, and 61.7% had high positive (POS) support, but only 40.4% had emotional/information (EMI) support.

PAM activation levels had a significant association with all dimensions of support except for TAN. The Overall MOS and EMI median scores of level 4 were significantly higher than levels 1 and 2, while a POS median score of level 4 was significantly higher than level 2. All MOS dimensions were negatively associated with depression.

Only about one-third (32.3%) of the respondents participated in support groups. Among non-members, 76.9% were employed, and 55.8% were on disability/sick leave.

No statistically significant association between support group participation and perceived social support was detected.

The above-reported levels of support indicate that the majority of respondents benefited from friendship and personal and intimate relationships that afforded them expressions of love and affection (AFF), and fun social interactions (POS). In comparison, a considerably fewer respondents perceived practical (TAN), as well as emotional and informational (EMI) supports. The TAN refers to assistance and instrumental support, while EMI is related to understanding and empathy, and availability of information or feedback when seeking solutions to problems. (Sherbourne & Stewart, 1991).

Given the level of physical disability and fatigue in this population, the Tangible support (TAN) entailing actual material aid and behavioral assistance is of great significance for patients. Interview data indicated that this type of support was generally provided by family and friends, but could have also been institutionally provided. One interview participant lived not in a nursing home, but in a housing development that was specially designed for seniors, with an array of programs including onsite nurse and doctor visits, a peers checking-on-each-other system, shopping trips, and household help. In spite of living alone and having extensive disabilities, she had one of the highest perceived social support scores. Had the researcher not interviewed her at her residence, such a high score might have been viewed as an anomaly. Notwithstanding such arrangements, among interview participants, patients living alone were those with the most difficulties in obtaining assistive support of this kind.

The interview data revealed four factors contributed to the perceived lack of tangible supports (TAN) by patients: (a) the lack of coverage for supportive services by insurance and social support programs; (b) the inability of patients to afford help such as house-cleaning services; (c) living arrangements not conducive to their disabilities; and (d) a general obliviousness on the part of family members to their needs. A number of

patients spoke of how their adult children and siblings, and even spouses, do not realize the extent of their disabilities and needs, and unless they ask for very specific assistance with tasks, they do not generally receive help as a matter of course. In addition, living on multiple floors and housing not designed for people with disabilities made their awareness of their need for assistance more pronounced.

The high perception of TAN support was the only social support dimension not associated with higher activation levels. This may indicate that higher TAN support may make patients' lives physically easier, but does not have the same significance as non-physical support in terms of their involvement. Another interpretation based on the interview data may be made. For many patients, the need for help and assistance was intertwined with feelings of guilt, being a burden, and an ambivalence about taking help and expecting help. As such, TAN support is a complex concept in need of unpacking. Even when patients express that they have high support, there might be a substantial emotional undercurrent, such as guilt, involved.

Emotional and Informational support (EMI) measures the perceived emotional and empathic interactions and responses to informational needs. The perceived lack of this type of support may be explained through the sentiments expressed by interview participants. The general sense of ambiguity, uncertainty, and lack of information to demystifying their illnesses may account for the perceived absence of informational support. The inability of the health profession to provide definite answers might have played a role in reinforcing this impression. This was expressed succinctly by one patient: "It is the most frustrating thing in the world for someone to say: 'You have this. Up until this point it is pretty much fatal, there is no cure for it, and we cannot really answer your questions.'"

The second aspect of this support, the affordance of understanding and empathy, relates to the general belief expressed by many interview-participants that unless one has experienced the disease personally, one cannot understand what patients go through and

be truly empathic. This could be the connection between EMI and TAN, as a true empathic understanding is needed for an offering of meaningful and consistent material assistance.

One place that an authentic understanding might be offered is in support groups. The means of all MOS dimensions, although not reaching statistical significance, were higher for leaders, followed by members, and then non-members. However, 67.7% of respondents did not avail themselves of this resource for a host of reasons. Quantitative findings point to employment and level of disability as factors, as non-members included 76.9% of employed and 55.8% of those on disability/sick leave. Other factors are detailed under qualitative findings. The low perception of EMI and POS in this sample may be partially attributed to absence of a connection to a community of patients.

Health professionals are another source of support for patients. EMI support consists of "expression of positive affect, empathetic understanding, and the encouragement of expressions of feeling/offering of advice, information, guidance, or feedback" (Sherbourne & Stewart, 1991, p. 707). Positive patient-physician interactions entail such characteristics and could provide a particularly powerful form of EMI support for patients. Graffigna et al. (2017) found that "the perceived quality of patient/doctor relationship" played a role in "sustaining patient activation" (p. 12). Using the PAM measure, Alexander, Hearld, Mittler, and Harvey (2012) found that three variables of higher perception of interpersonal exchange with doctors, frequency of out-of-office contact, and reports of fair and respectful treatments free of bias (racial/ethnic, socio-economical, gender, disease-specific misjudgments) were associated with higher patient activation. These factors were not measured in the quantitative portion, but are covered under the discussions of qualitative findings.

When social support was viewed in light of activation, statistically significant associations between PAM activation levels and EMI, AFF, and POS were observed. Specifically, the overall, MOS, EMI, and POS median scores were significantly higher

for those in the highest level of activation, level 4, than those in the lowest levels (1 and 2). Solberg et al. (2015) found that severely obese patients with high social support were more likely to have high activation and enjoyed high emotional well-being. Association of high social support and high activation has been reported in other chronic conditions. Witt et al. (2016) found that cardiac patients with high level of social support were more likely to have high activation levels (level 3 and 4). In Blackmore et al.'s (2016) study, those patients with multi-morbid conditions who had better social support had higher activation. Both these studies had used the ENRICHD Social Support instrument (ESSI) (Vaglio et al., 2004), which measures similar emotional, instrumental, and informational dimensions as MOS-SSS.

Among study respondents, self-reported depression was negatively correlated with MOS dimensions. In a sample of autoimmune disease patients that included scleroderma patients, Taylor (2008) found that only depression and social support, and not disease severity, predicted healthy behaviors measured by the HPLP (Health Promoting Life Style Profile). Other studies (Blackmore et al., 2016; Mazanec et al., 2016; Smith et al., 2013; Stepleman et al., 2010) using depression scales found depression to be strongly associated with lower activation. In this study, depression and lower activation were indirectly connected, through social support. The moderate association of depression and body image distress and pain and the strong association of depression with fatigue implicate pain, fatigue, body image concerns and depression as impacting patients' activation.

Summary of Quantitative Findings

In summary, the answer to the first question, drawn from the quantitative findings, may be given as follows. Patients' commitment to their illness management and their involvement in their own care, as represented by the activation construct, manifested in a range from passivity to high activation, with the majority of respondents being in the

upper range of activation. Increase in activation was related to disease duration in the first decade of the disease and was hypothesized to be attributed to the developmental nature of the illness experience. Other factors included the perception of social support and, indirectly through pain, fatigue, body image concerns, and depression. Disease severity, as it may be discerned by physical symptoms or disease subtypes, did not have an impact.

To answer the third research question, higher activation was associated with higher perception of support; however, with almost 70% of respondent not participating in support groups, the role of a support group in respondents' involvement was not clear. With 60% of respondents having a perception of low informational/emotional (EMI) support and almost half perceiving low tangible (TAN) support, there is a substantial void in the type of social support and interactions needed by these patients.

Discussion and Interpretation of Qualitative Findings

The qualitative findings of the study are discussed in light of the conceptual framework of the study and the related literature. The discussion of the findings is organized by the three research questions of the study and is presented after a brief contextual description of the participants.

Contextual Description

The participants of this study included 25 scleroderma patients (5 male and 20 females), majority White -American, between the ages of mid-0s to 85, with 64% having a college degree or above, and 56% retired or on disability/sick-leave. Disease sub-types represented include 44% limited, 36% diffuse, 12% sine, and 8% unknown. Participants' familiarity with prior chronic conditions and the healthcare system was limited, as the majority of participants (64%) enjoyed excellent health. For 40% of the participants, diagnoses occurred in their mid-40s to mid-50s. Forty-four percent of diagnoses occurred

within six months, while 56% took between a year and 5 years. The average disease longevity was approximately 12 years, excluding the two outliers with 30 and 49 years' disease duration.

In the next section, a summary of qualitative findings is followed by the discussion of the findings by the three research questions and the associated sub-questions of the study.

Research Question 1: Description of Patients' Commitment to Illness Management

Research question 1 sought an answer to the following research question and sub-questions: How do scleroderma patients describe/understand their commitment to illness management? Did commitment to be involved change over time? If so, when and how?

The findings related to question 1 described patients' commitment to managing their illness in terms of work that patients did and the motivational factors that were needed to sustain this work. This included the work and the efforts put forth to mitigate the physical and emotional/psychological, relational, and financial impact of the disease in their lives. These efforts for the interview participants spanned across six categories of obtaining a diagnosis, managing relationships with doctors, pursuing effective therapies, attending to emotional and psychological impact, dealing with employment and financial impact, and handling familial relationships.

To the extent that participants were motivated to do the above work, they were committed and involved. Their motivation was predicated on accepting the illness, and was sustained through hope, a will to live, and imagining a future. Patients spoke of needing time to accept their illness and certain periods when they were not fully engaged.

In interpreting these findings, the starting point was to abstract a description of patients' commitment and involvement from the interview-participants' depictions of how they manage their illness. The notion of *work* suggested by Corbin and Strauss

(1988) was utilized for this purpose. Corbin and Strauss define work as the tasks that patients undertake in order to manage some aspects of their illness. Participants also used this terminology in referencing their activities as the full-time work of tending to the demands of the disease. Identifying the tasks that made up this work was one way of approaching the description of their commitment and involvement. Altogether, six major areas to which participants devoted their energies were singled out, and detailed themes regarding the nature of each engagement were listed. The salient features of these engagements are discussed in this section, using Corbin and Strauss's model, which constitutes one part of the conceptual framework of this study.

The six areas of tasks engaged in by participants aligned with the three categories of illness work, biographical work and everyday work of Corbin and Strauss's (1988) model. The first three (obtaining a diagnosis, managing doctor's relationship, and pursuing effective therapies) related to illness work; the fourth area (attending to emotional/psychological aspect of disease) consisted of biographical work; and the last two areas (managing employment and finances and familial relationships) fell under the everyday work of living one's life.

Participants' descriptions indicated that the level of energy, effort, and attention given to these areas fluctuated along the illness course. Corbin and Strauss (1988) studied this temporal aspect and suggested the concept of an "involuntary illness trajectory" (p. 72) in order to describe the dynamic interactions between the three types of work and the fluctuating chronic disease progression. To create an equilibrium in this unpredictable system with interacting and moving parts, the model suggests that patients needed to come to terms with having a disease. Specifically, this was defined as a "movement toward an understanding and acceptance of the irrevocable quality of chronic illness, of the performance limitations accompanying it, of death, and of the biographical consequences it brings about such as lost jobs, failed marriages, and dependency" (p. 76). In this view, the imposed trajectory impels patients to take a position along a continuum,

ranging from non-acceptance to acceptance of disease, resulting in degrees of commitment to engage with the illness. Thorne et al. (2003) identified this commitment to engage as an act of taking control. They further reported that arrival at such a point is prompted by a realization that disease is chronic and ongoing; interventions may be limited and ineffective; one could prevent complications or improve outcomes; considerations of responsibilities toward others; and being the only one who can assume responsibility. Reaching such a mindset was characterized as a "philosophical shift" (p. 1341) in patients' relationship with the disease.

Looking among the sample for either signs of a gradual movement or a shift revealed mixed results, as detailed in the findings. For some patients, a shift occurred early on in the form of a response, which was articulated as a mobilization to fight. The intensity and rapidity of symptoms creating an urgency, and a sense that their bodies were assaulted and needed a forceful response may be added to the precipitating factors mentioned by Thorne et al. (2003). On the other hand, for most patients, the process appeared to have been a gradual movement toward acceptance of the illness, especially in cases where progression of symptoms occurred more slowly. A few recalled a vivid and sudden shift, located within a prolonged struggle of resistance. The realization of the steady deterioration of their condition seemed to have been the main motivating factor in accepting and taking a stand regarding their disease. The quantitative findings regarding distribution of activation levels and their associations with disease duration in the first decade of disease may be given a different interpretation in light of notions of a philosophical shift and commitment: different levels of activation may be viewed as different degrees of commitment, and the increase in activation levels as a gradual shift in commitment as disease progressed in the first ten years of illness.

In the early parts of the trajectory, biographical work, described in this study as what patients did to attend to the emotional and psychological impact of the illness, was particularly intense and demanding. Corbin and Strauss (1988) described the process as a

series of closures, breaking away from the past and what has been to take on what is here and will come, which might include grieving and depression. Many patients, in this study, described a period of grieving when they needed time to "feel sorry" for themselves, and "wrap their heads around" their diagnoses. Nearly 50% of the interview sample spoke of depression. Far from being episodic occurrences, depression was an ongoing feature for many, and patients toiled, irrespective of it, throughout their illness.

At the root of these sufferings, Charmaz (1983) contends, is the "loss of self" (p. 168). From this view, self is socially constituted. The functional limitations, disabilities, and disfigurements for these patients may be interpreted as having caused a disruption in their concept of self (Charmaz, 1983; Corbin & Strauss, 1988). This disruption separated the past, present, and future, making unavailable the "experiences and meanings" (Charmaz, 1983, p. 168), or the "biography" (Corbin & Strauss, 1988, p. 50) upon which the former self-image was built, leaving the patients at a *loss of self*. Accordingly, many activities and work that patients engaged with in this area were attempts in appraising the extent of such a loss, contextualizing and accommodating disease, and reconstituting and repairing the shattered self. For some patients, the extent of the efforts was commensurate with the extent of the perceived loss and their determination to reconstitute it. For others, the discontinuity between the past (who one was), now (who one has become), and the future (who one imagined to be, but has to forgo) was parallelizing for various lengths of time.

Given the variations in interview sample, the perceptions of such a loss spanned a wide range. Some of these were described under the theme of managing employment. The concept of loss expands this theme from a purely work and financial concern to an existential one. Those who were diagnosed later in life and closer to retirement age expressed less angst about loss of identity in terms of their professional roles. On the other hand, the sense of loss was very palpable for younger patients who had to give up their work, take on less demanding jobs for which they were over-qualified, or relinquish

plans for advancing their education and moving up the career ladder. The ability to continue to work, more or less as before, or retire in an expected timeframe provided for the continuity of professional self-image. On the other hand, for those having to go on disability, the perceived gap between the image of someone on disability and themselves needed to be bridged. As one patient put it, "When I think of somebody young who is on disability [like] I have a friend who has MS and she is on wheelchair, that to me makes sense. It was not the image of me that I had in my head."

Gender identity was another area where a disruption seemed to have occurred. Two male patients openly lamented the loss of their identity as the provider, fixer, and protector of family, while a third expressed anxiety in not being able to provide for his family financially. A few patients with visible skin and facial changes spoke of their appearance prior to the disease. One patient showed pictures of an extremely attractive person who was barely visible any longer; another patient spoke of bags of beautiful party attire that had to be given away, as "the previously known public, sociable presentation of self" (Charmaz, 1983, p. 179) was no longer available to her. Presentation of self, altered by skin deformities, was particularly disconcerting for two younger patients and was brought up in the context of dating. These findings corroborate the quantitative finding of the moderate association between body image stress and depression.

Disruptions in discharging parental roles weighed heavily on those with younger children; the unexpected physical and emotional unavailability due to fatigue and depression, failure to provide financially, and the possibility of not being there to rear their children to adulthood were devastating to all participants who were parents. Again, the strong association of fatigue and depression in the quantitative findings may be, partially, explained by participants' inability to fulfill their roles.

The impact of pain and fatigue and associations with depression and joint issues were also on full display for many with joint and muscle issues. The high levels of

athletic ability developed over years of training being taken away and replaced by extensive physical limitations were spoken of wistfully. Other patients reported the loss of fine motor skills: one person spoke of her inability to play the piano, another talked about limitation in doing artwork, yet a third could not type on a computer keyboard, and a fourth had lost tactile ability to handle small instruments needed for work. Such losses, where the decline could be objectively measured, inflicted a harder blow on patients in two ways: not only a cherished part of who they were was taken away, but also the continued deterioration provided a constant reminder and measure of the downward trajectory of their disease.

According to Corbin and Strauss (1988), one moves forward by letting go of what was lost and the past, and by integrating limitations imposed by the disease and reconstituting a new identity. Some of these were observed as participants described letting go of jobs they could not manage and the associated role identities they held dear; distinguishing between what could and could not be salvaged from the past like hobbies and pastimes that could still be performed; integrating their illness by discovering new ways they could feel productive and active, such as volunteering, tending to grandchildren, and working with other scleroderma patients; finding ways to lessen the intrusiveness of symptoms through creative inventions and gadgets, and the use of modified clothing; and lastly, respecting the limitations of their bodies by pacing themselves and altering social expectations and demands. Attempts to deal with these losses went across the six categories of tasks outlined earlier, highlighting the overlap and inter-relatedness of the tasks and work patients engaged with.

In addition to biographical work, the vigilance in illness work—such as watching for new symptoms, medications' side-effects and maintaining a cadre of competent specialists knowing that the threats of new organ involvement were looming—never let up for most patients in one form or another. Challenges in dealing with everyday work, in

light of the perceived lack of tangible support reported under quantitative findings, and the management and stress of familial relationships and finances, were not trivial.

Given all these challenges, sustaining one's commitment and engagement with work, according to the model, was predicated on hope, finding a will to live, and imagining a future.

Hope was fundamental to this process: "hope for a better, albeit altered future… without hope, there is no incentive to move" (Corbin & Strauss, 1988, p. 77). By hope was meant the "perception of an exit—a way out of the present situation" (p. 77). "A way out" for the study sample included finding effective medications, seeing reduction of symptoms, going into remission, or believing a cure was possible in the near future. These perceptions were shifting ones. A number of patients stated that they no longer believed a cure was possible in their lifetime. A few believed they were in remission or have been, at some point, and could regain it. For most, managing symptoms, keeping stable, and arresting further deterioration were considered realistic and hopeful. At the same time, many acknowledged that a total resolution was out of reach, their time was limited, and at least one person saw the disease as "winning" the race.

The need for signs of hope was detectable early on and did not seem to have abated in the course of illness for these patients. The vivid descriptions of interactions with diagnosing doctors may be interpreted in light of this need. Doctors exhibiting empathy and understanding, offering information and a plan of action, and engaging patients by experimenting with interventions, enrolling in clinical trials, or guiding them to adopt lifestyle changes were perceived as reassuring and hopeful, propelling patients to move forward with their illness. For those paralyzed by diagnosis, like a patient who had struggled for years being "stuck there, kind of like out of life," and hearing her doctor explain that she was alright for the time being and there were interventions to try to dislodge her from the "stuck" position of being shell-shocked. Similar assurances by doctors, like the patient who asked about her prognosis and was told, "I'm going to keep

you alive until you are in your 90s," provided for the possibility of an exit, and motivations to move on. This need to find reassuring doctors to sustain their engagement with their illness seems to have had a disproportionate role in their involvement. The needed characteristics of expertise, trustworthiness, and compatible communication styles listed by patients seem to speak to this aspect of patients' relationship with their doctors, more than purely a rational list of criteria in getting the best medical care.

Among the four motivational factors of commitment, hope, finding reasons to live, and imagining the future, the last one was the most difficult to address for the interview-participants. Corbin and Strauss (1985) label the last two as biographical and trajectory schemes, and state, "Visualizing the illness course and some of its attendant medical work is usually the physician's task. But the patient and spouse will have to discover all that is really entailed in carrying it out" (p. 238). In the slow progressing subtype with no major internal impact, a wait-and-see approach may be taken by doctors; the difficulty in classification of disease and a desire not to alarm the patients, combined with the unexpected trajectory and different responses to treatment, make visualizing the illness course for patients out of reach. Coupled with the unavailability of information or patients' inability to sort through it, this creates a great amount of uncertainty and confusion. Corbin and Strauss assert:

> If there is a great deal of uncertainty about conditions, then the maintenance of biographical projection and updated schemes is problematic. How can you plan when you have only vague definitions of what the future will bring?… How much longer do I have to live? How disabled will I become? Will there be a flare up of the disease, complications? If so, when? What will bring it on? Why continue with the work when there seems to be no gain? (p. 239)

While a few patients seemed to have learned to be at ease with uncertainty in a state of what the poet John Keats (1899) called *negative capabilities:* "when a man is capable of being in uncertainties, mysteries, doubts, without any irritable reaching after fact and reason" (p. 277), most patients described it as the most difficult aspect of living

with this disease. Mishel (1988) defines uncertainty in the context of an illness as "the inability to determine the meaning of illness-related events" (p. 225).

The uncertainty management literature attributes uncertainty appraisal as the impetus for psychological and behavioral actions that are employed to manage uncertainty (Barbour, Rintamaki, Ramsey, & Brashers, 2012; Brashers, Hsieh, Neidig, & Reynolds, 2006; Mishel, 1990). These include actions such as seeking information when ignorance is perceived as harmful, and avoidance of information when one needs to deny or forestall undesirable outcomes, to avoid overexposure, or when knowing would not lead to any solution (Barbour et al., 2012). Both behaviors were observed, in an uneven proportion, across participants. The majority of patients looked for information, even if intermittently, throughout the illness trajectory. Those who did not perceived their doctors or knowledgeable relatives (in the medical field) as conduits for information.

Whether seeking information or ignoring it, patients were taking an intentional approach in engaging with their situation in a way that allowed them to move on. Many patients took a similar stance toward information as they took toward patients in advanced stages of disease. For some, seeing such patients and observing the extent of disabilities and its management was useful information in anticipation of finding themselves in a similar situation in the future; for others, limiting exposure and forestalling the thoughts of such possibilities to a later time was the prudent way to deal with the enormity of the here and now. Information avoidance, far from being a passive posture, might have indeed been the appropriate behavior at a given moment in the illness trajectory. Barbour et al. (2012) argued that, in situations where information avoidance can reduce stress and limit anxiety, when circumstances may be made more manageable by waiting for the right time to act, or when one suspects the existence of flawed information or unreliable sources, information avoidance might be a sensible act. To this list may be added at least two other situations observed in this study. One has to do with the quality of information as being either inaccessible due to academic language and

terminology, or being overly simplified and basic, as in patient education materials or programs designed for new patients. Specific to scleroderma and other illnesses with phenotype plasticity is the variation in characteristics and presentation of the disease and its subtypes, which makes it nearly impossible for patients to locate themselves in the spectrum of the disease and cull relevant information specific to their disease presentation without a tremendous amount of work.

Some of these factors contributed to some patients seeking information through their doctors. Doctors as "credible authorities" (Mishel, 1988, p. 228) could reduce uncertainty by explaining the disease and its course, creating a sense of stability, and controlling treatments decisions and providing a course of action (Brashers et al., 2006). At the same time, with a disease like scleroderma where medical knowledge is limited and treatments are experimental, a number of threats to the credible authority of health providers existed, undermining the stabilizing role it might have played in patients' lives. Brashers et al. list threats such as invalidating patient's experience by a lack of understanding and absence of empathy, a perceived lack of knowledge of the field, and violation of expectations in terms of objectivity of medical technologies and protocols that were used. For scleroderma patients, lack of knowledge, expertise, or competence was the most major threat to the credible authority of doctors, prompting a continued vigilance in finding the "right" doctors and maintaining complex relationships, at times, among many doctors they saw locally and in specialty centers.

Particularly evident in descriptions of patients who were initially misdiagnosed or those whose symptoms were contested were their attempts to do their own independent research to hedge against this threat and to reduce uncertainty. Using a variety of resources, but mostly through the Internet and other patients, they sought information, which took some of the mystery of the disease away. The unedited and acontextual information thus obtained, however, was a double-edge sword. Mortality statistics and reported progression rates for scleroderma were devastating and overwhelming to most

patients, moving some to avoid or limit information seeking for a while, and come back to it at a later time, or to relegate the task to loved ones and doctors until they were able to do it themselves.

These struggles with uncertainty and biographical interruptions, in tandem with fluctuation of the disease, seemed to have staged the commitment of patients to their illness management in the form of a trajectory.

A cumulative commitment to illness management trajectory. An illness trajectory, incorporating the course of the disease and patients' responses to it, may cumulatively and across all participants be discerned:

1. *The "deer in the headlight" period* in the first months up to years, when patients were stunned by their diagnosis and the poor prognostic assessments.
2. *The immediate post-diagnostic phase*, characterized as either a period of withdrawal for patients with slower progression lasting 2 to 5 years, or a period of aggressive treatment and frenzy for patients with rapid progression.
3. *The familiar and more stable phase*, where some patients described a "calming" of their symptoms, some 5-6 years into illness.
4. *The late stage* of the disease where organ involvement was most extensive and the symptoms most debilitating.

This proposed illness trajectory is used to provide a descriptive account of patients' commitment to their illness management and its changes. This is done by highlighting the type of work patients were more focused on at various phases of this trajectory.

The "deer in the headlight" period. The focus of early involvement was on allowing oneself to grieve, overcoming fear, researching the disease, making sense of available options, sorting out recommendations for doctors, and finding scleroderma experts. Patients had to deal with their own fears and anxiety, as well as their loved ones. Searching on the Internet, halting it due to the shock and fear, and being propelled to

resume it were common. Concerns for children and the impact on them were overwhelming.

The immediate post-diagnostic phase. After a doctor or a team of doctors was secured, those facing rapid progression had to undergo many tests and treatments. Perceptions of pain and fatigue were most intense and bothersome in this period. Finding their way through the healthcare system and getting to know their body's reactions to treatments and tests were new learning experiences. Discovering how to evaluate the progression of the disease through body-listening and research, evaluating treatments, and finding alternative treatments like nutrition and exercise were among the work patients engaged with. If the initial medical providers were deemed not optimal, finding a trusted and competent doctor became a major focus of energy. Search for new treatments and clinical trials was ongoing. Vigilance for new and emerging symptoms was taxing. Patients spoke freely of their continued fears, anxiety, and depression. Dealing with the familial relationships and partners' response to their illness was most intense, and attending to the psychological and emotional impact was considerable during this time. Participating in support groups and educational programs provided the needed support and information for some, while it frightened and overwhelmed others. For many, seeing patients in more advanced stages was unsettling and was avoided. With emerging disabilities, finding solutions to reduce the impact on their daily lives became important, and considerations of life-style changes including taking time off from work and switching to less demanding jobs took on an urgency.

The familiar and more stable phase. With a shift to the middle stages of illness, the initial frenzy and confusion subsided, effective interventions were narrowed, some symptoms stabilized, patients' understanding grew of the ebb and flow of their symptoms, the flares and calmer periods became clearer, their disease and somatic knowledge became more reliable, and medical visits and interventions became familiar, routine, and accepted. The focus of many shifted to improving the quality of life, the

management of symptoms, and the business of living with the disease beyond medical interventions, such as those related to solving daily problems, employment, increased disabilities, and family and social relations. Executing strategies to accomplish these goals, and researching and learning about the disease continued for many as regular features of their lives.

As patients moved along, the focus of those with fewer disabilities became more outwardly. Whereas earlier on, contact with other patients felt overwhelming and avoided, in this stable period patients were reaching out to others. Many became active and involved in support groups and advocacy work, mostly to share their experiences and knowledge with other patients and the public, and also to learn from those in more progressive stages in anticipation of facing similar disabilities. The need for information and educational forums decreased, as patients felt these venues were geared toward new patients, and as they became more confident in their own knowledge. Some admitted to becoming complacent in keeping up with the latest information. For others, learning interests shifted from focusing on the disease to learning about new research and the management of disease. Attending venues, where patients with more severe cases could be seen, ceased to be jolting. Searching for what might have caused their illness lost importance; living and enjoying activities, being with loved ones that brought them joy and satisfactions, and eliminating situations and associations with people who put them or their bodies in stressful conditions took priority.

The late stage. Distinguishing between new emerging symptoms and those related to aging or other causes became more confusing for some, as the threat of new organ involvement in later stages of disease increased. For some, new medical tests became part of their routines. Those with more progressive symptoms found themselves more limited and socially isolated and had their focus on attending to the increasing physical impact of the disease and managing the day-to-day difficulties. Some with slower progression

continued to be outwardly active by helping other patients through initiating support groups and mentoring them to take on more active roles.

Throughout this illness trajectory, having access to competent and expert doctors was a paramount concern, and as doctors retired or moved, the effort to find new ones was ongoing. Body image distress and stresses associated with disabilities in public, at work, and in social situations were greatly burdensome; the economic issues and handling of employment, finances, medical expenses, and health insurance coverage weighed heavily on patients; and the negotiation of social interactions and management of familial stress were the added layers of strain in an already complex situation for many. Efforts to manage these complexities through problem solving, engaging creatively, and educating others were considerable.

In the next section, a discussion on findings related to strategies and learning to answer the second research question is presented.

Research Question 2: Patients' Strategies and Their Learning

The second research questions consisted of the following components: "What strategies do scleroderma patients use to be involved, and how did they learn them? What do they perceive as impeding or facilitating their learning? How do they overcome the barriers?"

Four strategies utilized by the interview participants were identified: taking a problem-solving/problem avoidance orientation; utilizing resources and advocating for themselves; being transactional and evaluative in obtaining care; and being open to experimentation. The findings indicated that participants learned informally on their own; learned socially in support groups; learned formally in educational programs and support groups; learned somatically through their bodies; but mostly learned incidentally through experiences and trial-and-error processes when solving problems. The participants attributed trial-and-error; previous experiences; availability of sources like support

groups, the Scleroderma Foundation's newsletters and website; doctors; other patients; and their own ability to research online as facilitating their learning.

As the site of illness, the body is a complex biological system made up of subsystems of interacting components through biochemical, neural, and thermal feedback loops and pathways. When an autoimmune chronic disease, with unknown etiology, throws this complex system into disarray, patients find themselves in the surreal position of being at once the chaotic system at war with itself and the one who interacts with it, and the other complex systems (medical and social), in the hope of taming it. Complex systems exhibit highly variable behaviors, behaviors that are non-linear or chaotic (Brugnach et al., 2008). Examples include the wax and wane of symptoms, fluctuating intervals of stability and deterioration, and unpredictable responses to medication compounded by a multiplicity of interventions with unknown interactions. These account for some of the complexity seen in scleroderma. Corbin and Strauss (1985) observe another level of complexity that exists in the interactions of a dynamic disease course and enactments of various work. To create a relative equilibrium in this system, their model suggests, the management of illness needs "strategies and techniques for control which are adjustable and changeable, in response to various contingencies that arise … with an emphasis on adaption to change" (p. 240). This "management in process" is similar to "muddling through," "incrementality," "organized anarchy," and "strategy-as-practice" argued by Bakir and Todorovic (2010) in situations where a means-end relationship is ambiguous. McCool, Freimund, and Breen (2015) suggest a number of approaches to engage with complexity. These include scenario planning to predict potential future problems and situations; using connections and networks and leveraging them to obtain desired outcomes; using models and simplifications (models could be as primitive as deferring to experts, or establishing routines); accepting and understanding complexity through experimentation; employing different forms of knowledge; and learning continuously. Using these as possible approaches, four strategies employed by the

interview participants were identified as: taking a problem-solving/problem avoidance orientation; utilizing resources and advocating for themselves; being transactional and evaluative in obtaining care; and being open to experimentation. These strategies are collectively discussed in the next section.

Strategies. The four strategies identified as interview participants' primary strategies had similarities with what McCool et al. (2015) suggested as approaches to deal with complexity. However, patients' strategies lacked the intentionality and thought-out formalities of McCool et al.'s list. Participants' strategies were themes identified in approaches that participants took when facing problems. As such they were by no means pre-meditated and consciously planned courses of action, but seemed to have emerged incidentally through trial and error and in response to specific problems. These may be viewed as learning processes and are discussed in that context.

Five types of learning were reported in these patients (informal, formal, social, experiential, and somatic). While all these learning types played a role in patients' involvement, learning experientially was most relevant in answering research question 2. Learning from experience, specifically Marsick and Watkins's (1990, 2001) incidental learning lens, is utilized to explore the learning that might have precipitated the above strategies in order to answer this research question.

Learning. For patients in this study, the burden of disease provided plenty of uncertainty, doubt, and problematic situations, and disjuncture when patients were "consciously aware that they do not know how to act" (Jarvis, 2006, p. 9). From the perspective of learning, a chronic illness provides an opportune venue for learning. This is so because not only it is "a context of experience in which problems naturally suggest themselves" (Dewey, 1930, p. 183), but also the "quality of problem it involves" (p. 183) is "a personal thing of such a nature as inherently to stimulate and direct observation of the connection involved, and to lead to inference and its testing" (p. 183).

At one level, the findings showed that almost all patients took an intentional approach to learning. This intentionality could be discerned through phrases such as "knowledge is power." Generally, however, in daily encounters, learning was informal and incidental. The inability of patients to pinpoint how they learned hinted at the incidental nature of learning that went unnoticed. Marsick and Watkins (1990) defined incidental learning "as a byproduct of some other activity, such as task accomplishment, interpersonal interaction, sensing the organizational culture, trial-and-error experimentation, or even formal learning" (p. 12). They further added, such learning "almost always takes place although people are not always conscious of it" (p. 12).

Marsick and Watkins's (1990) informal and incidental leaning model relies on the problematics of a given situation and the ensuing cycles of trial and error as the impetus for learning that ultimately would bring about a solution. These problematics, starting with a trigger causing dissatisfaction, lead to reframing of experience by interpretation of the experience one is facing and consideration of alternative solutions through comparison to previous experiences. Other steps, not necessarily in a sequential manner, include the possible need for acquiring new knowledge, resources, or skills; taking action on the proposed solution; and assessing the consequences and drawing lessons, resulting in learning that would help in the reframing of new situations.

The constant barrage of triggers from unsatisfactory responses of their own bodies, to inadequacies of healthcare system, inhospitable environments to their disabilities, including the weather, inaccessibility of public places, and social stigma due to their appearance were sources of dissatisfaction for participants. Many of these triggers did not go beyond frustration, anger, and feelings of helplessness. However, there were notable areas where patients were able to interpret their experiences by reframing the problem, finding solutions, and learning. The most prominent was in regard to receiving medical care. The frequency of doctors' visits and lab work provided for repeated experiences that afforded patients opportunities to encounter problems and ultimately reframe their views

in the process of finding a solution. The criteria for the type of doctors they looked for, and what treatments and tests they were willing to undertake, for example, were arrived at from their assessments in problem-solving situations they encountered in their doctor visits and when subjected to medical procedures. Devising assessment criteria as learned lessons, in a tacit way, was a striking feature of learning in this context.

As patients learned and gained experiences, established a reliable medical team, and got to know the staff, technicians, and procedures, the levels of dissatisfaction and problems that were shared by participants reduced considerably, only occurring when established medical routines were interrupted, such as when a doctor moved out of a practice, a need for a new specialist arose, patients found their doctors in retirement mode, or changes in insurance coverage occurred. Thus, learning in this area was most intense in the early years of the disease.

On the other hand, with the passage of time and increased disabilities, participants were in more problematic situations in dealing with their disabilities and found ways to reduce or circumvent problems through practical solutions, gadgets, and approaches. Experimenting with unconventional therapies was one area in which they looked for alternative solutions outside what was offered by their medical team.

Marsick, Watkins, Callahan, and Volpe (2008) posit that if the learner is stimulated to proactively look for options and acquire the skills to implement solutions, use creativity to exploit more options, and reflect critically to make the tacit beliefs and knowledge explicit, incidental learning would be enhanced. The varying levels of proactivity and efforts in finding creative solutions were a differentiating factor among interview participants. Proactivity relates to "a readiness to take initiative, an alertness to the environment and to opportunities it might afford learning" (p. 576). Creativity is what "enables people to imagine alternatives and think beyond their current circumstance or point of view" (p. 576). Nicolaides and Yorks (2008) attribute such a creativity to the *interactivity* aspect of experience, and its role of being "the catalysis for creativity and

innovation" (p. 2), especially in complex and dynamically changing environments. The proactive investigation of options and skills to implement solutions and use of creativity often went hand in hand. These were manifested, for example, in how patients found the right person to solve their problems, who in turn facilitated a creative solution.

The ability to enlist others, including those in formal positions (nurses, insurance company representatives, etc.), to find solutions repeatedly showed up as a major strategy to solve problems for many participants, and it seems it was learned by replicating their experiences through assessing "similarities or differences and use interpretation to make sense of the new situation" (Marsick et al., 2008, p. 577). This is using analogies in the service of solving a problem. The strategy to advocate for their needs and leverage resources was one area in which this type of analogical reasoning could be seen.

Gavetti and Rivkin (2005) stated that when people "reach back to an earlier experience for a solution, they are using analogy" (p. 56) and argued that analogical reasoning was the middle ground between the two extremes of trial and error (in ambiguous settings) and deduction (in information-rich settings). Yorks and Nicolaidas (2012) considered using analogies as a strategic learning tool. A number of patients used their experience in advocating for clients in their professional life as models to advocate for themselves. Some patients noted the experiences of having to advocate for their children as templates. Yet, many who were unable to pinpoint specific situations spoke of a cumulative learning in acquiring the skills and mindset needed for advocacy through living, or vicariously through watching family members operating in certain roles and positions, such as in the political arena. More generally, transferring knowledge and skills from their professional lives and looking for similarities in their illness situation to utilizing these skills were common practices. Another area where patients drew on their previous experiences to frame a new situation was in anticipating problems. Again, this approach was used in the context of medical problems, but also in regard to the

functioning of their own bodies and levels of fatigue in order to manage familial and social obligations.

The ability to engage others to solve problems was not only about proactivity and reaching out, but also about a willingness to accept help. Generally, the perception of vulnerability by others accorded the patients legitimacy in asking for help, and such requests elicited more positive responses by others. At the same time, the perception of vulnerability by patients themselves and beliefs about how help should be given or taken was limiting to their proactivity. Some of these beliefs were alluded to in Chapter IV under relationships with family and friends.

These types of unexamined assumptions and beliefs are major barriers to adult learning (Mezirow, 2009). Reflecting on the underlying tacit and taken-for-granted assumptions "may help learners rethink situation in which they find themselves and re-frame their understanding for the kind of learning they might need to undertake" (Marsick et al., 2008, p. 571).

Verbalizing the tacit assumptions that one currently holds is a difficult task. It was much easier for patients to speak about the tacit assumptions they had previously held. Charmaz (1983) states, "Ill persons often become highly aware of previously taken-for-granted aspects of self because they are altered or gone" (p. 170). This alteration in taken-for-granted assumptions displayed itself in a few participants' attitudes and beliefs regarding received medical knowledge. This was discussed previously under the threat to the credible authority of doctors. Such critical examination of one's beliefs provides personal insight, and insight, Yorks and Nicolaidas (2012) argue, is "an essential foundation of strategic learning" (p. 186). Alhadeff-Jones (2016) suggests that life is not just a series of events; there exists a rhythm or pattern to our life experiences. A rhythm suggests continuity, discontinuity, and dimensions like "patterns," "periodicity," and "movement." He contends that our strategies and responses, whether conscious or not, "tend to follow existing behaviors that repeat themselves," (p. 274) and many times "so

much suffering comes indeed from the incapacity of individuals to change the strategies they mobilized in order to assert their own autonomy" (p. 275). The attention to and consciousness of these rhythmic patterns may frame one's learning, specifically if it is transformative learning. In this light, utilization of some of the strategies seen in participants may fit a model of unconscious repetition of strategies that worked previously, not entailing substantial learning. Neither Alhadeff-Jones's framework of rhythmic learning nor the transformative learning lens was utilized to collect or examine data; however, it was apparent that, for some participants, this illness was a disruption and discontinuity of a magnitude like no other in their lives, prompting a conscious reexamination of strategies and the adaptation of new ones. For most, this may be characterized as a situational awareness in certain context. One patient stood out, however, who repeatedly made contrast to her patterns of behavior and beliefs prior to her illness. The mere ability to draw these contrasts and to differentiate how her views and actions have changed in a systemic way hinted at transformational learning.

Impediments and facilitators of learning. The incidental nature of learning in these patients and a conception of formal learning as the real learning held by the participants precluded arriving at a direct answer to what they perceived as impeding or facilitating their learning and a means to overcome the impediments directly. Outside general comments as to the availability of information, the usefulness of the Scleroderma Foundation's newsletters and publications, helpfulness of support groups in providing materials, their own ability to research and find information on the Internet and through more formal means, the notion of learning and impediments, and facilitators to it were not available from these data.

The inferences made using Marsick and Watkins's model pointed to unsurfaced beliefs and assumptions, and the lack of proactivity and creativity as major impediments. As was illustrated previously, beliefs about the nature of help, how it was asked, and given were a major barrier for some patients. The inability of patient to place themselves

in the illness spectrum and the mindset of not exposing themselves to patients in more advanced stages as an avoidance strategy were another barrier. This was true in newly diagnosed patients, but also was seen in patients with more than a decade of living with the disease. Lack of proactivity, by relying on their doctors and assuming a passive role regarding information and experimentation with interventions, was another observed barrier. Some patients were more creative in devising tangible solutions for their disabilities through gadgets, while others were more creative socially in advocating for themselves. To the degree one was lacking the creativity in one area or another, their ability to deal with problems and learn through them was limited.

In terms of structural barriers, doctors' interactions played a substantial role. The more collaborative doctors provided for experimentation and mutual learning. Patients who reported that their health providers made time for conversation and discussion around symptoms and therapies and did not assume the mantle of total authority displayed engagement in the process of learning and figuring out their illness through these collaborations. On the other hand, the health providers who told patients they had to educate themselves but viewed that learning as something patients did outside their offices and on their own frustrated their patients' attempts to learn. Providers who disregarded or trivialized patients' information-seeking attempts by making them feel ill-equipped to understand such information took away one means patients had to validate their understanding of their illness. This highlights the divide between the medical perception of care, and the illness care that patients of chronic illness expect from their providers.

The structure of support groups and educational forums where patients were mixed regardless of their disease subtypes or other variables such as age and disease duration posed some challenges for patients, leading to their avoidance of such venues, where dialogue and sharing of experiences could have occurred and learning promoted. The nature of educational materials that address the needs of new patients, but have little to

offer to patients in later stages, is difficult to address and poses a barrier to learning. Another structural barrier is a lack of creativity in utilizing the expertise and learned lessons of experienced patients for the mutual benefit of newer patients in a format that goes beyond local interactions.

In summary, through the incidental learning lens of the Marsick and Watkins's model, the four strategies listed for the interview participants may be viewed as steps along a problem-solving process. These steps occurred in response to triggers and the appraisal of unsatisfactory situations leading to learning in the process of finding a solution to encountered problems. Learning occurred in some key areas important to managing the illness, particularly in interactions with the healthcare system. Surfacing tacit assumptions, creativity, and proactivity deemed to facilitate learning, while structural barriers, mostly outside patients' direct control, played a role in impeding learning.

Research Question 3: Impact of Social Interactions on Involvement and Learning

The third research question was concerned with the question, "How are social interactions, particularly, support group facilitation/participation, perceived in terms of involvement and learning?"

Participants discussed their involvement in support groups in terms of their contributions to and benefits from such participation. Leaders saw their involvement as sharing their experiences and vision, and utilizing their skills and resources for the benefit of other patients. Members described the involvement as an opportunity for learning, interacting with others, and joining forces for a greater cause, such as fundraising or helping other patients. Non-members described their non-involvement in support groups as being caused mainly by a fear invoked in seeing patients in more advanced stages, and the negativity and complaining demeanors of other members.

In interpreting these findings, two opportunities for significant interactions for patients were found to be those with doctors in and out of clinical settings and other patients in the context of support groups and educational forums. A third opportunity, electronically through patient portals, was not considered in this study.

A total of 64% of the interview participants saw four or more doctors. Interaction with medical professional for scleroderma patients of this study, therefore, was extensive and made up a sizable portion of their illness experience. Using the PAM measure, Alexander et al. (2012) found that three variables—higher perception of interpersonal exchange with doctors, frequency of out-of-office contact, and reports of fair and respectful treatment free of bias (racial/ethnic, socio-economical, gender, disease-specific misjudgments) —were associated with higher patient activation. Interview patients indicated that characteristics such as listening, explaining, considering patient's preferences, looking the patient in the eye, and spending adequate time in visits were among the most important metrics by which they judged the quality of their interactions.

Some interview patients spoke of having cell phone numbers and email addresses of their doctors for out-of-office access, and reported instances when they included their doctors in fundraising events and walks, had them as speakers in support group meetings, or had lunch with them. This researcher, having attended two national scleroderma patient conferences and a number of educational forums, observed doctors as invited speakers in these venues to be accessible to patients after their talks and to be mingling informally with them throughout the events. A number of patients spoke of these doctors as their friends and partners in fighting the disease, and many considered scleroderma experts as "special" and uncommonly compassionate. Such out-of-office interactions are hypothesized to increase patients' involvement by reducing the power differential and the levels of passive and paternalistic roles that patients and doctors may take on otherwise (Alexander et al., 2012; Roter, Larson, Sands, Ford, & Houston, 2008).

On the other hand, referencing doctors as "they," expressions such as "they talk down to you, like you are stupid," or "[saying] Trust me, that is not good enough [reason]" were indicative of patients' negative perceptions of not being treated respectfully or without bias. Alexander et al. (2012) reported a 5.55-unit increase in patients' activation per 1 unit increase in respectful treatment level, where a 4-6-unit differential was considered significant. As such, positive doctor interactions may be viewed as an important form of support, impacting patients' activation and involvement.

In this dissertation study, the perceived EMI (Emotional and Informational) support was associated with high activation. This type of support provides "expression of positive affect, empathetic understanding, and the encouragement of expressions of feeling/offering of advice, information, guidance, or feedback" (Sherbourne & Stewart, 1991, p. 707). Positive patient-physician interactions entail such offerings and could provide a particularly powerful form of EMI support for patients. The role of doctors as credible authorities in providing hope and reducing the uncertainty of disease was previously discussed, and providing positive interaction is argued to be linked to patients' activation.

In addition to potential support by doctors, support groups can provide EMI and POS support. Fifty-two percent (13) of interview-participants were either leaders (7) or members (6) of support groups. These participants provided descriptions indicating that interacting with other patients who truly and empathically knew the experience of the illness was one of the most import reasons to join. POS support provided in support groups could be like no other, as expressed by a leader: "If you make jokes about being sick and debilitated, they [families] do not find it funny, but we do, and what a release [that is]." In addition, informational support in the form of practical information and guidance, in particular those related to doctors' referral, medication side effects, and solutions to everyday difficulties through the subject-matter expertise of other patients, was deemed very valuable by most interview participants.

Specifically, leaders in this study indicated that sharing of experiences and utilizing their skills, knowledge, vision, and resources for the benefit of other patients were the main motivation for their involvement with support groups. For members to have social interactions with people who understood and were empathic, to learn, and join forces for a greater cause was the main reason. These findings were in agreement with Embuldeniya et al. (2013), who undertook a meta-study of 25 articles in chronic illness support group. In this study, mentors' (peer leaders') perceived impact of support group participation included the sharing of their experiential knowledge, sense of reciprocity, helping, role satisfaction, and emotional entanglement. The sense of connection, finding meaning, and reducing isolation were commonalities with members.

These reasons, expressed by those connected to support groups, indicate their perceptions of their involvement as getting or giving support, or both, and learning through others' experiences. A more nuanced understanding may be obtained by inquiring into reasons for not joining support groups, as the above reasons did not seem to have been motivating factors for 48% of the interview participants and nearly 68% of the survey participants.

Reasons cited by the interview participants for not joining were grouped into four categories: logistical, personal, group dynamics, and programming. These categories were similar to three themes found by Gumuchian et al. (2017) in a three-factor model describing data obtained from 242 scleroderma respondents to a 21-item non-attendance survey: personal, practical (similar to logistical), and beliefs about support groups (e.g., too negative, similar to the group dynamics category). In that study, the two items with the most ratings of "important" or "very important" were "already having enough support from family" and "not knowing any scleroderma support groups offered in my area" (p. 4). Given that AFF (Affectionate) support was the highest form of support reported by survey-respondents (mean of 76.1 and median of 92.0) in this dissertation study, one may infer that Gumchian et al.'s sample may have enjoyed the same level of support and

equated this type of support received from family with what was offered in support groups. In contrast to Gumchian et al.'s findings, the two major reasons expressed by interview participants were the fear invoked by seeing patients in more advanced stages (expressed by 24% of participants) and the negativity of groups (32%).

Mazanderani, Lock, and Powell (2012) observed the phenomenon of fear or stress invoked by seeing other patients among Parkinson's and motor neurone disease (MND) patients. Similar to findings of this dissertation study, patients were seeing a deteriorated version of themselves in other patients, but simultaneously considered themselves different and unique. Mazanderani et al. argue that this creates an epistemic tension that patients need to resolve in order to value other patients' experiences and have an empathic relationship with them. These conditions may be viewed as prerequisites for support group participation. Mazanderani et al. suggest that patients primarily negotiate this tension through the metaphor of being *differently the same*. This "cultivation of sense of difference" labeled in this dissertation as individualizing was previously reported as part of somatic knowing of the participants and is in agreement with Mazanderani et al.'s assertions.

The findings of this dissertation study based on those who participated in support groups or educational forums, however, suggest that, ironically, one mechanism to determine one's "difference" is through observations of other patients afforded in support group meetings and educational forums. These venues, by allowing patients to gauge the extent of their symptoms in comparison to other patients, provide for a mechanism to locate themselves in the spectrum of disease and, by extension, imagine a future. In this sense, interview participants who got involved with support groups were in two camps: those who could see themselves as different now but wanted to observe and learn how their illness might become, and those who knew they were different because they had lived with disease long enough to have developed a solid footing and wanted to help others get there.

Overall Synthesis

This study started off by inquiring into out what scleroderma patients' commitment to their illness management looked like. In answering this question, the PAM-13 measure, as one of the few available scales to measure patients' active role, was utilized. At the outset, it was assumed that this measure would not capture all dimensions of patients' commitment to their illness management. This assumption was proven correct through participants' comments on the margins of questionnaires, the patterns of response to the measure's items, and through the qualitative findings based on patients' interviews. These findings indicated that patients' commitment to the management of their illness entails attending to many concerns beyond those related to the physical impact of the disease, as discussed in this chapter.

The existence of levels and stages of activation and engagement, as suggested by Hibbard et al. (2004, 2005) and Graffigna, Barello, Bonanomi, and Lozza (2015) was also detected in this study. However, whereas Hibbard et al. attributed these variations to differences in knowledge, skills, and beliefs among patients and Graffigna et al. saw psychosocial processes as the culprit, the findings of this study suggest a different framing. Specifically, the illness trajectory entwined with uncertainty and patients' experiential learning in dealing with the illness-related problems and the use of strategies to surmount them shaped the patients' commitment to their illness management. These elements are shown in Figure 5.1.

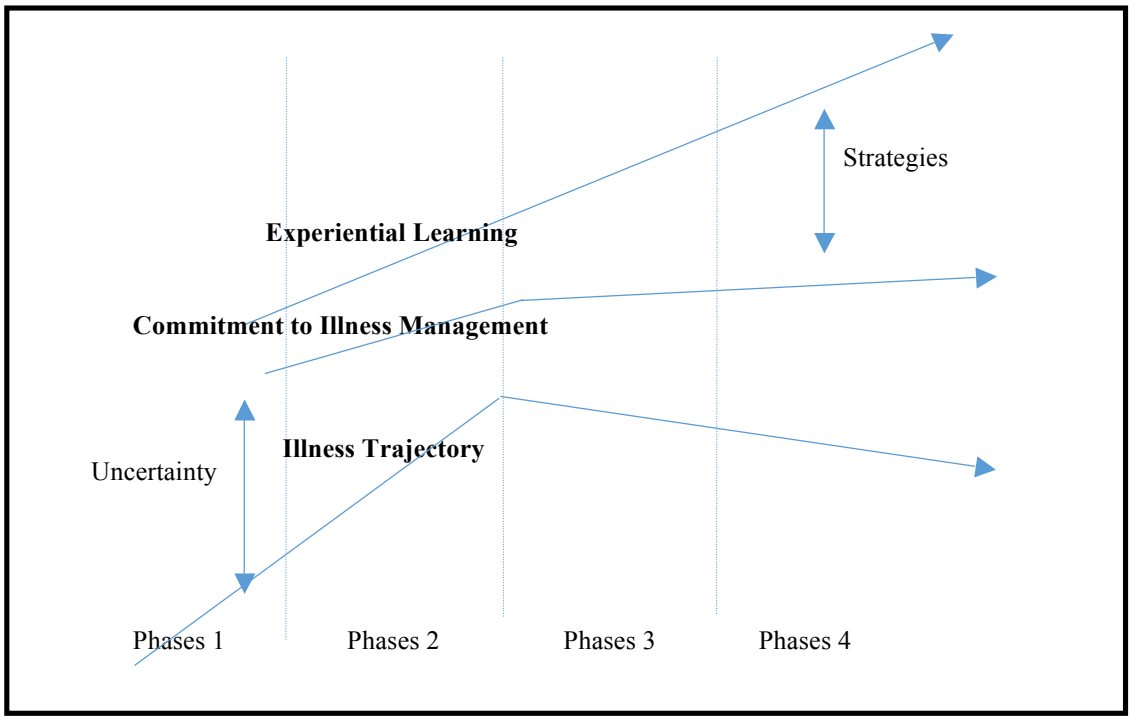

Figure 5.1. Commitment to illness management

In this sketch, commitment to illness management moves in tandem with illness trajectory and phases of the disease and the experiential learning of living with the illness. The mediating factors are strategies and uncertainty. Level of uncertainty is greater early on and may subside somewhat when patients feel more stable; it increases with deterioration of symptoms and fluctuates with the wax and wane of disease. As patients learn experientially to deal with their problems, their repertoire of strategies increases, helping them to better manage their illness in spite of increasing uncertainty due to the downward trend of disease and aging. The full development of a model was beyond the scope of this exploratory and descriptive study. In future work, this model will be fully developed, and a measure to assess patients' commitment to the management of their illness under uncertain and ambiguous conditions will be designed.

This model does not merely propose a different semantics (learning vs. knowledge and skills, or uncertainty and strategies vs. psychosocial processes), but puts the patients' experiences of illness front and center and frames commitment to illness management as a process of experiential learning in tandem with the needs of the disease as it is lived day in and day out, organically and holistically. Therefore, it confirms the agency of patients through their own learning processes as central to their engagement.

This sample, by the virtue of having an illness with little known sanctioned medical knowledge and solutions and a lack of formal self-management program, was shown to have traversed different phases of their illness, organically and through their own learning. However, uncertainty and lack of knowledge about a disease are not unique to scleroderma. In their book on lupus patients, Price and Walker (2015) state: "Lupus is a condition that challenges the certainty for which contemporary medicine strives and throws into sharp relief the difficulties associated with diagnosing, treating and managing conditions that have uncertain and contested aetiologies, treatment approaches and prognoses" (p. 35). Other autoimmune diseases, like fibromyalgia (Johnson, Zautra, & Davis, 2006), also put patients under uncertain and ambiguous conditions. The fact that many of these patients are able to engage with and manage their illness does not deny the importance of programs and structures that can facilitate this process. A number of recommendations are provided in the next section that are aligned with a belief in the benefits of such instrumental and short-term interventions. However, the findings of this study questioned the fundamental issue at the heart of patient education programs and the care delivery systems that ignore the contribution of patients' experiential learning and fail to incorporate patients' illness experience into models of care. This includes models assessing patients' engagement, activation, and involvement marketed to healthcare providers with the intention of promoting patient-centered interventions. Weeks and Weinstein (2016), in their book, *Unraveled*, quoted a cancer patient whose definition of patient-centered care was that of "patient better be in the center of and in charge of his

care, because no one is" (p. 42). Sadly, this indicates the cynicism of patients and how far-removed their experiences are from healthcare practices that claim they want them involved. English (2012) echoes the same, observing:

> Ideologies such as individual-focused health education, professional as all-knowing purveyors of health, and citizens as consumers of expert medicine have come to dominate the Western world. These ideologies have become hegemonized so that some citizens see themselves subject to the machinery of hospitals and evidence-based medicine in which their health needs are not always met. A revised theoretical approach, rooted in health promotion, works with learners in participatory ways to reclaim health. (p. 17)

It was argued in the introductory chapter of this dissertation that the real-world problem of understanding how patients should or choose to have an active role in their own care is critical in addressing the mounting chronic disease problem faced by healthcare systems. Viewing patients as learners and engaging with them in collaborative and participatory ways that bring in their experiential knowing, as exemplified by some health providers of patients interviewed in this study, will be an important step in addressing this problem and reframing the nature of health delivery and health education.

In the next section, reflection on research, and the conclusions and recommendations based on study's findings and discussion are presented.

Reflection on the Research

When interview-participants were asked to comment on the findings of this study, the comments of one participant, who had extensive contact with patients in various capacities, seemed to indicate that the variations in the sample covered the breadth of patients in terms of their illness manifestations: "Thank you for sharing your findings. They are what I would expect and seem to run the gamut, which is typical because no two patients are alike." Another participant expressed his views through a phone call. His critique of the study included the lack of educational and, by extension, socio-economic,

and racial/cultural diversity of the sample, and a want of attention to the suffering of scleroderma patients in terms of understanding and support within families.

Another patient's comments confirmed the critical role that trusted health providers play in encouraging and giving hope to patients to carry on, and also point to varying levels of comfort patients have with ambiguity. She saw the "wait and see approach" of her doctor

> as a positive because I had never thought in terms of "nothing could be done about it." Now, as well as then, there still is no clear understanding of what the disease is or what can be done about it. Nevertheless, I do not recall experiencing any anxieties associated with lack of a well-defined protocol of treatment. I felt confidence in my doctor. In spite of the fact that my disease progressed to total disability, what I remember most is my doctor encouraging me to continue my life activities to the fullest extent of my ability. The result is that I have been able to have a full life, and now … am regarded by some doctors as possibly never having had scleroderma. Of course, they never saw me when I displayed the classic symptoms. Nor did they have access to my medical records when those symptoms were present.

She concluded, "Your incisive summary makes me feel that the ways in which I have been involved with my 'illness' have been positive and appropriate."

Lastly, another patient's comments indicated that the findings had resonated with her experience of illness:

> I thoroughly enjoyed reading your findings! I am interested in your future summaries and information.... I most often conclude that non-Scleroderma affected people (those who are not sufferers of the disease) do not, and possibly never will, understand the way Scleroderma has changed us. Your conclusions have proven that I need to rethink my opinions! You seem to understand quite well!… You have successfully captured and shared the information given to you- good work and many thanks!

These commentaries, although limited in number, provide some measure of assurance that the study has been on the right track and has captured the salient points of involvement with these patients. During the course of this study, this researcher has come to view scleroderma as a mighty river that patients found themselves thrown into, navigating its meandering and murky waters. Through this process of navigation, they

showed their commitment and involvement. The perseverance, resilience, ingenuity, and determination that were displayed by the 25 participants of this study were overwhelming, humbling and a testament to the human spirit. I feel privileged to have been welcomed to their homes, offices, and neighborhoods and to have been trusted with accounts of their struggles and experiences.

This study started off with four general assumptions: Patients want to be involved in their own care; patients want to know about their diseases and treatment options; whereas in acute illnesses a passive role is a welcome option, with chronic conditions, most patients are compelled to become engaged with their illness; and commitment to illness management and becoming involved in one's care are not easy, but people learn through the process of doing it. In conclusion of the study, this researcher has come to the realization that re-phrasing some of these assumptions is in order. Committing to be involved or not is not a binary choice. Patients are involved and involvement is a process that, like most processes in life, includes a good deal of learning in different forms, consciously or unconsciously, to discover the most effective ways of going about living and managing an illness. Learning about the disease constitutes a part of this learning, but by no means may it be equated to involvement; neither the unwillingness to learn at a given time nor the delegating of that task to others may be viewed as a lack of involvement. Involvement demands support by all those who can provide it to patients. Through family, health professionals, other patients, support groups, and educational entities, involvement takes shape.

What sets scleroderma apart from more common illnesses is the degree to which there is a paucity of knowledge about it. From the perspective of the day-to-day living with the illness and its impact, however, there are commonalities with other immune diseases and chronic conditions. Fatigue, pain, disability, uncertainty, fear, stress, and depression are all the warp and weft of most illness fabrics. As such, many findings of this study do not stray far from what most patients with chronic diseases experience.

Conclusions and Recommendations

Twenty-five scleroderma patients and 201 survey patients painted descriptions of a devastating and debilitating disease and their struggles to live with it and carry on. Their perceptions and descriptions informed this study in understanding their commitment to illness management, including how they were involved in dealing with their illness, how they learned to do so, and how social interactions and support groups impacted their involvement. Based on these findings and analysis, four conclusions are drawn.

Conclusion One: Patient Commitment to Managing Their Illness is a Complex Process

Scleroderma impacts the patients on three fronts: physically through an assault on the body, socially by altering roles and identities, and psychologically by engendering uncertainty. Patients' commitment to manage their illness and strategies they use are concurrent attempts to mitigate these impacts and create a relative equilibrium in a complex situation. The degree of these impacts fluctuates throughout the illness trajectory. Far from a simplistic view that patients are either involved or not involved, involvement is a spectrum where at each point a patient may find him/herself in complex negotiation with these multiple needs in attending to his/her illness. Patients may become stuck at certain points or intervals, but overall forced to move, if for no other reason but the deterioration of their symptoms. Understanding this trajectory with its physical, social and psychological elements, the larger temporal view of where patients are at a given time and the appreciation of existence of a movement along this path will allow patient commitment and involvement to be viewed as a process and not a static patient characteristic. As such, it is rich in learning potential: learning in dealing with disease, learning in living with illness, and learning to develop and transform.

Conclusion Two: Importance of Managing Uncertainty

The perception of uncertainty and challenges in dealing with this uncertainty are at the core of patients' ability to engage with their illness. Patients have different levels of tolerance for ambiguity. For most patients, decreasing uncertainty is important. This study found that the patients' own attempts to chart the progression of disease through information-seeking and drawing similarity or dissimilarity to other patients; passage of time, the learned patterns of wax and wane of symptoms, and occurrence of periods of stability; and health-providers' willingness to provide assurances or share an assessment are the main means through which patients reduce their perceived levels of uncertainty. The first two put the onus on patients who have differing levels of skill and resources to manage these. Health professionals have a fundamental role to play by understanding the importance of uncertainty management in the care of their patients, and the ongoing need for hope and assurance, so patients can carry on and move forward.

Conclusion Three: Role of Support Groups Needs Further Study

Higher social support enhances patients' commitment and involvement, but is undermined by the underutilization of support groups. Survey studies by others suggest that this underutilization is due to a perception of not needing support or a lack of knowledge of and access to these groups. The interview findings of this study point to an undercurrent that seems to have been overlooked: support groups are paradoxical for some scleroderma patients in negotiating notions of *being different* and *being the same.* Further understanding in this area is needed, as support groups are venues for social and experiential learning and the promotion of involvement for patients.

Conclusion Four: Learning Occurs Through Problem-solving

Patients learn informally, formally, socially, experientially, and somatically. However, they learn mostly informally and incidentally through solving problems. This learning occurs early in the illness trajectory facing dissatisfaction with the healthcare

system, and later in dealing with problems related to disability. Examining tacit assumptions and patterns of strategies that one utilizes can enhance patients' learning. There were no indications that support and scaffolding to promote this type of examinations exist for this study's participants.

Based on these conclusions, the following recommendations for practice and further research are provided.

Recommendations for Practice

The following recommendations are applicable to patient education and advocacy practices, and professional development in dealing with scleroderma patients and other chronic conditions. The notion of illness trajectory and where a patient is located within this trajectory is an important concept to assess the needs of a patients, reduce uncertainty, integrate the illness, and learn to live with the physical impacts of the disease. Differentiating these needs, in addition to considerations of disease subtype, severity, age, and disease duration, may reveal programming content and presentation formats that are more conducive to meeting the needs and the learning of scleroderma patients. Facilitating critical thinking and examination of beliefs should be a part of these formats.

Patients learn in multiple ways throughout their illness. The experience of a diseased body is an embodied experience like no other. Through the arts, movements, and other non-verbal methods, opportunities to express this embodied experience should be provided. These types of opportunities generally do not exist for patients, and efforts in creating programs to allow for patients' participation in this manner should be advocated and encouraged.

Given the extent of disabilities in these patients, helping patients integrate the disease into their lives by considering new roles and activities needs to be a part of patient education programs. Being active and feeling useful to others was one of the most

cited coping mechanisms mentioned by patients. Peer educators who successfully have done so and can share their own experiences, like many remarkable patients who were interviewed in this study, would be invaluable in this regard.

Inclusion of opportunities in support group meetings to surface tacit beliefs and assumptions that are holding patients back would be more difficult to implement, but necessary to assist patients to enhance their learning and become more involved. Implementing training programs for group leaders to facilitate such discussions would be a first step in this process.

Although having a chronic and life-threating disease at some level is a solitary process, the role that family, friends, doctors, other patients play in providing the types of support that engender more involvement cannot be separated from the patients' journey. A difficult disease like scleroderma impacts the entire family. Family and caregivers' education needs to go hand in hand with patient education. Facilitating discussions among patients and caregivers and family members, as well as providing specifically designed programs for caregivers, will go a long way in promoting the involvement of all stakeholders, in clarifying expectations and boundaries in familial relationships, and in allowing patients to learn without overly helpful caregivers taking away such opportunities.

Professionals, from doctors and nurses to technicians and office staff, must also learn that patients' needs for reassurance and hope, clarity and information, being heard and treated as individuals and collaborators, being believed, and being looked in the eyes are as real as their physical symptoms. Appropriate responses to these needs will provide for far more involved patients.

Recommendations for Policy

The Centers for Disease Control and Prevention (2015) have a four-pronged approach toward efforts in chronic diseases prevention and management:

(1) epidemiology and surveillance, (2) environmental approaches, (3) healthcare system interventions, and (4) community programs connected to clinical services. Based on this study's findings, the following recommendations, aligned with the above approaches, are made:

Fundamental health care system policy changes, specific to chronic conditions, are sorely needed. These include exemption from formulary restrictions, and deference given to physicians' judgment and flexibility in the use of off-label medications when no tried and true intervention exists. Policies and regulations to prevent putting patients in the position of using ineffective drugs for months to prove inefficacy while dealing with onerous side effects need to be implemented. Removal of out-of-network limitations when patients have to cross state lines to specialty centers and to where clinical expertise exists will provide for more appropriate care and will remove major barriers to early diagnosis in cases that are difficult to diagnose. Furthermore, such policies would facilitate participation in clinical trials, as some clinical trials are not entirely free to patients and partial costs are expected to be passed to patients' insurance. The out-of-network coverage limits or eliminates access to these trials for many patients, not only by curtailing patients' only viable option, but also by impeding the development of effective interventions in the long run.

Coordination of care among multiple specialties is a major problem for patients with chronic illnesses and those with multi-morbidities. The current incompatibility of medical electronic systems put the burdens of dealing with systems' inadequacies on patients themselves. Until technological advances and stakeholders' cooperation allow for a seamless sharing of medical records, other intermediate measures to remove the onus from patients are needed. These measures may include more incentives for timely and automatic release and transfer of records to all care providers of a patient upon a visit or a major intervention; a system of regular training and education of office staff as to the existence of these systemic incompatibilities and their duties and procedures to remediate

the problems; and support and promotion of initiatives such as OpenNotes (www.opennotes.gov) and Blue Button (www.healthIT.gov), which provide for patients' easy access to electronic health records.

As was seen with the population of this study, the prevalent medical model of chronic care, with its focus on biological disease, leaves patients to themselves in dealing with the psychological impact of illness. An integrated approach, especially early on and in the form of teams of multi-specialty clinicians, social workers, and spiritual advisors working alongside physicians to heal the body and soothe the mind and soul, is desperately needed for all chronic illnesses. Examples of such multidisciplinary models need to be highlighted and promoted.

Community interventions, such as self-management programs, have greater availability for certain chronic conditions like diabetes through local hospitals and health providers. By taking into consideration the commonalities in living a chronic disease and devising general programs to address common issues like disability, nutrition, exercise, fatigue, pain, and emotional and psychological issues, the isolation faced by many chronic patients may be reduced and educational resources may be shared more equitably. Local health entities need to be made aware of such needs and encouraged and enticed to implement such programs and outreach to patients whom they may not be fully aware of in their communities due to the smallness of their numbers and their needs for specialists outside local communities.

As part of community design and interventions promoted by policies such as more walking/biking areas, physical education in schools, full-service groceries and farmers' markets in underserved areas, attention to innovative designs in housing developments (outside nursing homes and care facilities) that take into account chronic illnesses as part of life and the aging process of majority of people need to be given consideration. Such innovative designs can facilitate elements of tangible support and self-sufficiency, which, in turn, could encourage a more active and involved posture toward one's care.

Removing some of the daily agitations by providing more hospitable environments for living with disability may reduce the cost of care in the long term, as more patients opt to continue living in their own homes.

Recommendations for Future Research

This study inquired into the notion of patient commitment to managing their illness, primarily from the perspective of 25 patients with mixed scleroderma disease subtypes. Survey data of 201 patients provided additional insights into the role of support in involvement, and possible influences of factors such as educational attainment and disease duration in involvement. First, this study may be expanded in a number of ways by focusing on particular subgroups in the study such as those within a subtype of disease, a given age category, racial/ethnic make-up, or gender. The interview sample of this study, overall, had above-average education. The influence of education and income on patient engagement and involvement warrants further exploration in this population.

The quantitative portion of this study collected categorical data and limited demographic information that generally are collected in this type of study. A more detailed questionnaire based on insights gained from this study may be developed, specifically for scleroderma patients. As an example, data related to disease duration need to go beyond the notions of early and late stages of disease perceived in biomedical research to what makes sense in the context of living with an illness. A nationwide outreach to have an adequate sample size is recommended.

When this study was initiated, only one widely used instrument to measure some aspects of patients' active role was available. Since then, a number of studies around a second instrument measuring patient engagement (Graffigna, Barello, & Triberti, 2015) have been published. Inquiry into the appropriateness of this instrument with this population of patients may prove to be a worthwhile effort.

Two male interview participants of this study had their spouses present while being interviewed. At times, these spouses offered unsolicited comments. Such participation was insightful in a number of ways, including the consideration of ownership of disease and involvement in its management as a shared project. A study to include patients and their partners or caregivers may provide not only more accurate and nuanced data, but additional insights into the role of caregivers and family members, and development of an expanded definition of involvement as a shared or familial enterprise.

The notion of learning in this sample was explored mainly through the lens of informal and incidental, and experiential learning. A cursory look at somatic learning indicated that other ways of knowing were also involved in these patients' encounters with their illness. This type of learning and knowing deserves further study, and this population seems to be a good candidate for such an exploration. Additionally, it was apparent that a number of interview participants had a transformative learning experience dealing with their illness, which had led to a sense of empowerment not seen in other participants. Utilizing the lens of Mezirow's (1994) transformative learning theory to study these patients could provide another window into how such learning may occur in the context of a rare illness. In spite of studies in patient populations using transformative theory, the characteristic of this illness provides a unique situation unlike others previously explored. In addition, Alhadeff-Jones's (2016) work in the rhythmic nature of learning in time might be an interesting framework to study this population, given the temporal nature of disease trajectory and its changing needs for learning.

Similar to other autoimmune diseases, the majority of scleroderma patients are middle-aged women. Five men were included in this study, and the initial reaction of four of these men regarding their diagnosis consisted of doubt and disbelief of the expert opinion they were given. None of the women in the study expressed such a reaction. Belenky, Clinchy, Goldberger, and Tarule (1997) suggested five perspectives on women's ways of knowing in developing a voice to be heard, and their choices as to

whom to hear. Women's ways of knowing perspectives included silence, received, subjective, procedural, and constructed knowing. These perspectives collectively could provide another lens through which to explore learning as a developmental process, particularly as it relates to the notion of received knowledge in a disease where formal and expert knowledge is limited.

Further research into various aspects of support groups is needed to discover factors inhibiting attendance, programming needs, and training of peer group leaders. The phenomenon of *being different* and *being the same* merits further investigation to unpack the nuances involved. Further research in understanding emotional factors in perception of tangible (TAN) support may also be helpful for support groups.

Lastly, it was argued that living with scleroderma, even though a rare disease, shares many commonalities with other chronic illnesses. A formal study of these similarities can give general policy recommendations for chronic diseases more validity.

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Appendix A

Participant's Rights

TEACHERS COLLEGE
COLUMBIA UNIVERSITY

525 West 120th Street New York NY 10027 212 678 3000

www.tc.edu

PARTICIPANT'S RIGHTS

Principal Investigator: Shohreh V. Anand

Research Title: Learning to be involved: What does it mean for scleroderma patients?

I have read and discussed the Research Description with the researcher. I have had the opportunity to ask questions about the purposes and procedures regarding this study.

My participation in research is voluntary. I may refuse to participate or withdraw from participation at any time without jeopardy to future medical care, employment, student status or other entitlements.

The researcher may withdraw me from the research at his/her professional discretion.

If, during the course of the study, significant new information that has been developed becomes available which may relate to my willingness to continue to participate, the investigator will provide this information to me.

Any information derived from the research project that personally identifies me will not be voluntarily released or disclosed without my separate consent, except as specifically required by law.

If at any time, I have any questions regarding the research or my participation, I can contact the investigator, who will answer my questions. The investigator's phone number is (914) xxx-xxxx.

If at any time I have comments, or concerns regarding the conduct of the research or questions about my rights as a research subject, I should contact the Teachers College, Columbia University Institutional Review Board /IRB. The phone number for the IRB is (212) 678-4105. Or, I can write to the IRB at

Teachers College, Columbia University, 525 W. 120th Street, New York, NY, 10027, Box 151.

I should receive a copy of the Research Description and this Participant's Rights document.

If video and/or audio taping is part of this research, I ( ) consent to be audio/video taped. I ( ) do NOT consent to being video/audio taped. The written, video and/or audio taped materials will be viewed only by the principal investigator and members of the research team.

Written, video and/or audio taped materials ( ) may be viewed in an educational setting outside the research ( ) may NOT be viewed in an educational setting outside the research.

My signature means that I agree to participate in this study.

Participant's signature:_____ Date:___/___/____

Name: _____

Appendix B1

Research Description and Informed Consent for Interviews

TEACHERS COLLEGE
COLUMBIA UNIVERSITY
525 West 120th Street New York NY 10027 212 678 3000
www.tc.edu

RESEARCH DESCRIPTION and INFORMED CONSENT FOR INTERVIEWS

DESCRIPTION OF THE RESEARCH: Many experts agree that patient involvement and activation in their own care and management of their illness are important in chronic conditions. Our understanding of what patient involvement is and how patients learn to be involved, and what helps or hinders such learning are limited, especially when it comes to patients suffering from rare and complex conditions. You are invited to participate in a research survey and interviews that will explore the views of scleroderma patients on how they are involved in managing their illness, and how they learned to do so. Based on your experiences as a scleroderma patient, the researcher is interested in obtaining your points of view on: a) how you describe your experience of involvement; b) what you have learned; and c) if you are part of a support group how you view your support group engagements in terms of involvement and learning; The research will be conducted by a doctoral candidate. Two interviews lasting approximately 60-90 minutes each, will be scheduled based on your availability at a location of your choice. Interviews may be conducted over the phone or Skype, if an in-person interview cannot be arranged. These interviews will be informal and conversational in nature. At any point in time you can decline to answer a question, request what you have said be deleted, or withdraw from the study. If you have not completed the survey portion of this study, the researcher may ask you to fill out the survey prior to the interviews.

RISKS AND BENEFITS: The interviews will be conversational in nature and will concern your personal experiences related to your involvement in managing your condition. The interviews will center on topics that you most likely have discussed or thought about before. Therefore, the researcher anticipates the risk of engaging in these conversations to be minimal. However, in the event of emotional distress related to these interviews, appropriate psychological counseling referrals will be provided. There are no direct benefits for participating in this study.

PAYMENTS: There are no payments or other compensation for participation in the study.

DATA STORAGE TO PROTECT CONFIDENTIALITY. You will be assigned a pseudonym and will not be identified. Your interview and your survey data if you mailed it together with the pink sheet or have taken in person before the interviews will only be known to the researcher.

For purposes of accuracy, the researcher requests that the interviews be audio recorded. Transcriptions and recordings, along with any handwritten notes, will be kept in a locked cabinet in the researcher's office. All electronic files will be password protected. The researcher will destroy the tapes and surveys after she defends her dissertation.

TIME INVOLVEMENT: Your interview participation will take approximately 2-3 hours, to be distributed over two 60-90 minute interviews. Interviews will be scheduled based on your availability, and will be held in a convenient location to you, or may be conducted over the phone or Skype. If you have to take the survey again, it will take an additional 10-20 minutes.

HOW WILL RESULTS BE USED: The results of the study will be used to inform the
researcher's proposed dissertation research for completion of the doctoral degree. The research data may also be published in professional journals and/or articles, or be presented in professional conferences. The findings and aggregate data (but not the individual data) may be shared with the Scleroderma Tri-State Chapter. Since all study participants will be given pseudonyms, and all identifying information will be masked, there will be no way to identify the subjects in the publication of the research data.

Appendix B2

Informed Consent for Survey

TEACHERS COLLEGE
COLUMBIA UNIVERSITY

525 West 120th Street New York NY 10027 212 678 3000

www.tc.edu

INFORMED CONSENT FOR SURVEY

DESCRIPTION OF THE RESEARCH: Many experts agree that patient involvement and activation in their own care and management of their illness are important in chronic conditions. Our understanding of what patient involvement is and how patients learn to be involved, and what helps or hinders such learning are limited, especially when it comes to patients suffering from rare and complex conditions. You are invited to participate in a research survey. The research will be conducted by a doctoral candidate and will involve a two-part survey where you indicate your level of agreement/disagreement with a list of 13 statements, and the type of social support available to you when you need it. This survey is a part of a larger study that explores the involvement and learning of scleroderma patients. Your completing the survey and mailing it, or completing it online, indicates your consent in taking the survey and conveying your opinions.

RISKS AND BENEFITS: The risks of participating in this study are minimal. If there is discomfort in answering a question, you may skip the question or abandon the survey altogether. There are no direct benefits for participating in this study.

PAYMENTS: There are no payments or other compensation for participation in the study.

DATA STORAGE TO PROTECT CONFIDENTIALITY: Your survey will be anonymous: there are no personally identifiable information collected; the researcher does not have access to your name or contact information, and there is no way to link your survey and demographic information back to you, either by me or the Scleroderma Foundation. All mailed surveys go to a dedicated P.O. box and only researcher has access to this mailbox. The online surveys will be password protected and is not accessible by anyone except the researcher. The results of the survey will be presented in a combined and aggregate form, not individually. Paper surveys and saved electronic surveys will be kept in a locked cabinet in the researcher's office. All electronic files will be password protected. The researcher will destroy survey papers/files after she defends her dissertation.

TIME INVOLVEMENT: The survey will take approximately 10 to 20 minutes to complete.

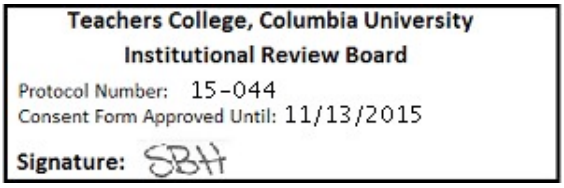
HOW WILL RESULTS BE USED: The results of the study will be used to inform the researcher's proposed dissertation research for completion of her doctoral degree. The research data may also be published in professional journals and/or articles, or be presented in professional conferences. The findings and aggregate data (but not the individual data) may be shared with the Scleroderma Tri-State Chapter.

Since the surveys are anonymous, there will be no way to identify the subjects in the publication of the research data.

CONTACT INFORMATION: If you have any questions about this study or your rights as a participant in this research, you can contact the Principal Investigator, Shohreh Anand, or the Institutional Review Board at Teachers College, Columbia University at the contact information listed below.

Principal Investigator: Shohreh
Anand Phone: 914-xxx-xxxx

Teachers College Institutional Review
Board 525 West 120th St. Box 151
NY,10027 Phone: 212-678-4105



Appendix C

Interview Contact Form

INTERVIEW CONTACT FORM (PINK FORM)

If in addition to this survey, you are willing to be interviewed about your views on how you are involved in managing your illness and learned to do so, and what helps or hinders this process, please read this page. Otherwise ignore this page.

The interview part of this research project will explore the views of scleroderma patients on how they are involved in managing their illness and how they learned to do so.

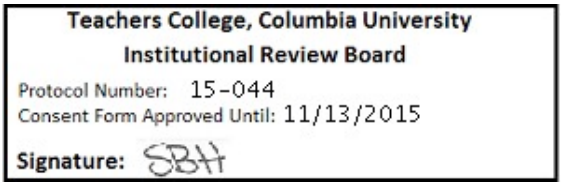
I am very interested in your perspectives and experience. Your participation in this study is completely voluntary. If you decide to participate you will be asked to have two interviews between 60 to 90 minutes each, depending on your schedule and availability at a time and location convenient to you. These interviews will be informal and conversational in nature, and may also be conducted over the phone or Skype. Your interview comments will be completely confidential.

If you are interested in participating in interviews, please return this page with your survey, or email me at sva2111@tc.columbia.edu, or call me at 914-xxx-xxxx. *Please note if you return this form with you survey, your survey will not be anonymous, but it will be totally confidential (no one other than the researcher will know about your survey answers).* I prefer to get your survey with this form to better tailor my interview questions and prepare for our conversation. However, if anonymity is a concern for your, please either mail this form separately, call or email me instead, or fill out the survey online.

Your name: _____
Your email: _____
Phone number: (____) _____
Address: _____
_____

The best way to contact me is through:

Email
Phone, best time to call is: _____
Mail



Appendix D

Recruitment Letter

Dear Scleroderma Patient,

My name is Shohreh Anand and I am a scleroderma caregiver. My husband is a scleroderma patient; you might have read his story in the Scleroderma Foundation Tri-State newsletter, *Scleroderma Exchange,* recently. I am reaching out to you for assistance on a study that I am conducting as part of my doctoral dissertation on the experiences of scleroderma patients in managing their illness and learning to do so. I became interested in this subject after my husband's diagnosis and my own involvement in learning about this illness.

Enclosed is a two-part survey that will ask about your agreement or disagreement with 13 statements that people make when they talk about taking care of their health, and the type of assistance and support that is available to them when they need it. I will be grateful if you take a few minutes to fill out this survey and the demographic information sheet that goes with it, and return them in the envelope that is provided. Alternatively, you may fill out the survey online: Please use the password**:** colorteal and the link to survey: **http://tinyurl.com/SclerodermaSurvey** (please email me at sva2111@tc.columbia.edu and I will email you a link to click on, if you have problems with typing this in.) Taking part in this survey is totally voluntary and anonymous: there are no personally identifiable information collected; I do not have any access to your name or contact information, and there is no way to link your survey and demographic information back to you. In addition, the demographic information and answers you provide will be combined with everyone else's and will be presented in an aggregate form, not individually.

In addition to the survey, I would appreciate the opportunity to interview a few patients and support group leaders about the journey you have been on. My interest is in understanding how adults learn when faced with a challenging situation like the one many scleroderma patients face. In dealing with this disease, you have learned and have become experts in managing your condition. I would love to hear your views on what has been your learning. The interviews that I have in mind will be informal and conversational in nature, and totally confidential. If you decide to participate, I ask you to allow me two occasions for our conversation, between 60 to 90 minutes each, at a time and location convenient to you. Interviews may be conducted over the phone or Skype, if an in-person interview cannot be arranged. If you are willing to share your insights about your learning, and what helps or hinders this process, please contact me or provide your contact information on the enclosed pink page, and I will get in touch with you.

Regardless of whether you want to be interviewed or not, I hope you take a few minutes to fill out the survey. I truly appreciate your time and willingness to assist me in this project. Please feel free to contact me if I can

provide additional information or answer any questions. Thank you again for your help.

Sincerely,

Shohreh V. Anand
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Appendix E

Survey Demographic Information

Please choose the best answer for each question and mark your choice with an X in the box with that choice.

1. Are you a scleroderma patient?
   - ☐ a) Yes, I have been diagnosed with scleroderma by a doctor.
   - ☐ b) No, I am not a scleroderma patient.
     - o I am a family caregiver
     - o I am a friend
     - o Other

2. How long ago were you diagnosed with scleroderma?
   - ☐ a) 12 months or less
   - ☐ b) 1 to < 2 years ago
   - ☐ c) 2 to < 3 years ago
   - ☐ d) 3 to < 5 years ago
   - ☐ e) 5 to < 8 years ago
   - ☐ f) 8 to 10 years ago
   - ☐ e) More than 10 years ago.

3. Which of the following describes your type of scleroderma?
   - ☐ a) Localized (Morphea, Linear, En coup de sabre)
   - ☐ b) Systemic- Limited or CREST
   - ☐ c) Systemic-Diffuse
   - ☐ d) Systemic- Sine Scleroderma
   - ☐ e) I do not know.

4. Do you know your auto-antibodies?
   - ☐ a) Yes
   - ☐ b) No
   - ☐ C) I am not sure.

5. Please indicate your gender:
   - ☐ a) Female
   - ☐ b) Male

6. Please indicate your age range:
   - ☐ a) 18-29 years old
   - ☐ b) 30-39 years old
   - ☐ c) 40-49 years old
   - ☐ d) 50-59 years old
   - ☐ e) 60-69 years old
   - ☐ f) 70-79 years old
   - ☐ g) 80 years and over

7. Please indicate the highest level of education completed?
   - ☐ a) Some High School
   - ☐ b) High School graduate
   - ☐ c) Some College
   - ☐ d) College graduate
   - ☐ e) Some postgraduate work
   - ☐ f) Postgraduate degree

8. What is your employment status?
   - ☐ a) I am working full-time
   - ☐ b) I am working part-time
   - ☐ c) I am a non-working homemaker
   - ☐ d) I am a student
   - ☐ e) I am retired
   - ☐ f) I am disabled or on sick leave
   - ☐ g) I am unemployed
   - ☐ h) Other

9. How many doctors do you see for your medical care?
   - ☐ a) 1
   - ☐ b) 2-3
   - ☐ c) 4-5
   - ☐ d) More than 5.

10. If you see a scleroderma expert, how far do you have to travel to see this doctor?
    - ☐ a) Less than 24 miles
    - ☐ b) 25 to 49 miles
    - ☐ c) 50 to 99 miles
    - ☐ d) 100 miles or more
    - ☐ e) I do not see a scleroderma expert.

11. Which of these symptoms do you experience? Please check all that apply.
    - ☐ a) Pain
    - ☐ b) Fatigue
    - ☐ c) Sleep disturbance
    - ☐ d) Depression
    - ☐ e) Body image distress

12. Which of these organs are involved in your illness? Please check all that apply.
    - ☐ a) Lung
    - ☐ b) Heart
    - ☐ c) Digital Ulcer/Raynaud
    - ☐ d) Gastro-intestinal
    - ☐ e) Musculo-skeletal, joints
    - ☐ f) Kidney
    - ☐ g) Skin
    - ☐ h) Other

13. Do you have other medical conditions unrelated to scleroderma such as cancer, diabetes, etc.?
    - ☐ a) Yes
    - ☐ b) No
    - ☐ c) I am not sure

14. Are you part of a support group?
    - ☐ a) Yes, I am a Support Group Leader or Co-Leader
    - ☐ b) Yes, I am a Support Group Member
    - ☐ c) No, I am NOT part of a support group.

Please check one:

- ☐ **A) I am just participating in the survey part of this study.**

- ☐ **B) I want to be contacted for interviews and I am including the pink form (interview interest form) with my survey. I understand that my information will be kept confidential and only researcher will have access to my survey, and my demographic and contact information.**

- ☐ **C) I want to be contacted for interviews, but I want my survey to be anonymous. Therefore, I will contact the researcher by email or phone to be included in the interviews, or will send my pink form in a separate envelope.**

Appendix F

PAM-13 Measure

Below are some statements that people sometimes make when they talk about their health. Please indicate how much you agree or disagree with each statement as it applies to you personally by circling your answer. Your answers should be what is true for you and not just what you think others want you to say. If the statement does not apply to you, circle N/A

1. When all is said and done, I am the person who is responsible for taking care of my health	Disagree Strongly	Disagree	Agree	Agree Strongly	N/A
2. Taking an active role in my own health care is the most important thing that affects my health	Disagree Strongly	Disagree	Agree	Agree Strongly	N/A
3. I am confident I can help prevent or reduce problems associated with my health	Disagree Strongly	Disagree	Agree	Agree Strongly	N/A
4. I know what each of my prescribed medications do	Disagree Strongly	Disagree	Agree	Agree Strongly	N/A
5. I am confident that I can tell whether I need to go to the doctor or whether I can take care of a health problem myself	Disagree Strongly	Disagree	Agree	Agree Strongly	N/A
6. I am confident that I can tell a doctor concerns I have even when he or she does not ask	Disagree Strongly	Disagree	Agree	Agree Strongly	N/A
7. I am confident that I can follow through on medical treatments I may need to do at home	Disagree Strongly	Disagree	Agree	Agree Strongly	N/A
8. I understand my health problems and what causes them	Disagree Strongly	Disagree	Agree	Agree Strongly	N/A
9. I know what treatments are available for my health problems	Disagree Strongly	Disagree	Agree	Agree Strongly	N/A
10. I have been able to maintain (keep up with) lifestyle changes, like eating right or exercising	Disagree Strongly	Disagree	Agree	Agree Strongly	N/A
11. I know how to prevent problems with my health	Disagree Strongly	Disagree	Agree	Agree Strongly	N/A
12. I am confident I can figure out solutions when new problems arise with my health	Disagree Strongly	Disagree	Agree	Agree Strongly	N/A
13. I am confident that I can maintain lifestyle changes, like eating right and exercising, even during times of stress	Disagree Strongly	Disagree	Agree	Agree Strongly	N/A

Appendix G

MOS-SSS Measure

People sometimes look to others for companionship, assistance, or other types of support. How often is each of the following kinds of support available to you if you need it? Circle one number in each line.

	None of the time	A little of the time	Some of the time	Most of the time	All of the time
Someone you can count on to listen to you when you need to talk	1	2	3	4	5
Someone to give you information to help you understand a situation	1	2	3	4	5
Someone to give you good advice about a crisis	1	2	3	4	5
Someone to confide in or talk to about yourself or your problems	1	2	3	4	5
Someone whose advice you really want	1	2	3	4	5
Someone to share your most private worries and fears with	1	2	3	4	5
Someone to turn to for suggestions about how to deal with a personal problem	1	2	3	4	5
Someone who understands your problems	1	2	3	4	5
Someone to help you if you were confined to bed	1	2	3	4	5
Someone to take you to the doctor if you needed it	1	2	3	4	5
Someone to prepare your meals if you were unable to do it yourself	1	2	3	4	5
Someone to help with daily chores if you were sick	1	2	3	4	5
Someone who shows you love and affection	1	2	3	4	5
Someone to love and make you feel wanted	1	2	3	4	5
Someone who hugs you	1	2	3	4	5
Someone to have a good time with	1	2	3	4	5
Someone to get together with for relaxation	1	2	3	4	5
Someone to do something enjoyable with	1	2	3	4	5
Someone to do things with to help you get your mind off things	1	2	3	4	5

Appendix H

Interview Protocol

1) Please tell me, to the extent you want to share, your story of being diagnosed and living with scleroderma.
2) Given your story, what did you think, then, about what you needed to do to take care of yourself?
3) Do you recall making a conscious decision to ___ (make a plan, have a role, be involved, be active, be engage, take responsibility, be in charge, have control, or other words that may have been used by interviewee) in order to take care of yourself? Describe the situation & your thoughts then.
    a. (If yes to #3) What (incidents, realization, thoughts, events, encounters, etc.) did prompt you to make this decision? Describe what happened. Who else was involved and in what way?
    b. (If yes to #3) So you said you wanted to (have a role, be involved, to have control, to be active, to be in charge, to take responsibility, or other words used by interviewee). What does _____ (having a role, being in control, etc.) mean to you? Or how does it look like for you?
4) When you first started on this journey, how did you get information and learned about your illness and treatments? Who/what did you use as sources of information?
5) How did you make sense of information you got? Were there other people who you consulted or were helpful in making sense of it?
6) Has the way you go about getting information, stayed the same, or changed in the course of your illness? How?
7) Here I have a list of types of things that people say they do when they are involved in managing an illness (list at the end of the protocol). Please pick the top 5 that stand out for you, or are especially important for you to do, and describe in details what you do and why these are important.
8) Were there times when you had to change course and do things that you had not done before- maybe some of the items on the list? Thinking back over this time, were you in situations where you felt you did not have all the knowledge and skills that you needed or did not know what to do? What did you do then to gain what was missing? What did make it easy or difficult for you to learn what you needed to learn?
9) How do you describe the familiarity with scleroderma of health providers (nurses, therapists, pharmacist, etc.) that you have encountered? What did you do when you felt they were not familiar with your condition?
10) How did you choose your main doctor when you were diagnosed with this illness? Were you ever had to change doctors?
11) Can you describe your expectations about your main doctor's role, and your doctor's visits?
12) When your doctor recommended a treatment, how did you decide what to do? Who else helped you decide? What kinds of things you look for to decide if a recommended treatment is appropriate for you?
13) Do think you have a different awareness of your body? How do you validate the

things that you notice or feel? (Do you wait to check with the doctor? ask other patients, etc.)?

14) When you notice something unfamiliar, or a new health situation occurs, how do you decide what to do?
15) Some say each scleroderma patient is unique in the symptoms he/she has and the response to treatments. Do you agree with that? How do you know if something is unique to you? Can you give some examples of the criteria that you use?
16) Do you modify treatments in any way to suit your individual situation?
17) If you have made life style changes (nutrition, exercise, stress reduction, etc.), or have tried alternative medicine, what are your thoughts as why you needed to experiment with these?
18) Do priorities in life change because of such an illness? How?
19) I am interested in your thoughts about some of the strategies, some of the steps you have or could have taken along the way, or the lessons learned. If you have to write an advice book for other patients what are some of the strategies and tips that you would give them?
20) What are some of the qualities that have helped you in this journey? What other qualities do you think a patient should have? What do you think is the best way to develop these?
21) Do you enlist others, family, friends, co-workers, and professionals (your doctor, therapist, etc.), in the work of your care? How?
22) Do you participate in advocacy activities (support groups, research fundraising, etc)? If yes why is that important to you?
23) Are you part of a support group? Why or why not?

If yes:

a) Tell me, what does being a support group leader/member mean to you? Why do you do it?
b) In your observation, what does involvement mean to the people in the support group? Does it come up where you might discuss how they have been involved in their illness or how they are thinking about it? If it has come up, can you give an example?

24) Is there anything else that you would like to share?

What Some Patients Do

1. I seek Information about my medical condition and possible treatments.
2. I attend patient educational programs.
3. I ask questions and consult with others (doctors, patients, etc.) about my illness.
4. I keep up with the latest research.
5. I comply with medical treatments my doctor prescribed.
6. Sometimes I decide not to comply with medical treatments.
7. I evaluate if a treatment is working for me.
8. I evaluate the expertise of my doctor and change doctors if needed.
9. I evaluate the quality of care I am getting from doctors, nurses, clinic, or the hospital I go to, and do something about it if not satisfied.

10. I pay attention to my body and observe and monitor it.
11. I experiment with alternative therapy.
12. I hypothesize about the causes of my illness and what might make it better or worse and talk to my doctor or other patients about these hypotheses.
13. I make lifestyle changes.
14. I develop strategies to manage my care and navigate the healthcare system, and to get what I need.
15. I do advocacy work.
16. I know how to mobilize the resources I need, and ask for help.
17. I think and reflect about my medical condition and what I can do about it.
18. I have found ways to live with my limitations.
19. I think differently about my roles and my priorities in life after my illness.
20. I do things to bring a sense of control and balance to my life.
21. I Feel empowered that there are variety of ways of dealing with my health condition outside the conventional boundaries and expectations.
22. I expect my experiences and knowledge of my illness are listened to by health professional.
23. I am involved in making decisions about my treatments.
24. I use coping skills to manage, and minimize the physical and psychological burdens of my condition.
25. I participate in self-management activities to take responsibility for my own care.
26. I plan for future and think long-term about my medical needs.

Appendix I

Sample Framework Analysis

Participant Words from Transcript	Initial Thoughts	Initial Category
I do what I am told by my doctor because I have no one else to trust. And I wake up every morning and say ok, put one foot in front of the other and keep going… I can either let it overtake everything I am or I can plow on and hope for the best and keep going and that is what I do.	Trusts doctor, no choice; Pushes through; Hopes for the best	Has to trust doctor. Has to push through. Has to have hope.
Those will be the kind of things that will be really helpful in educational programs to give people standards by which they can sort of measure … he [Dr]is pinching my skin and he is looking at stuff and I asked what is he doing. He said I am just checking your skin. What are you checking for tell me. Tell me, it should not be a secret from me. I want to know what signs you are looking for, or what are your measurements.	Helpful to have measures. Wants to know signs to look for	Wants measure, and signs of disease
I think that is really interesting that you have the same symptoms that I do, yet you react differently to the drug than I do. It makes you mind go, ok, if this is the case, if we look at all the same things why can't we figure out why you are reacting differently than me if we are having the same symptoms going into it…Cancer used to be the kiss of death and they found ways to treat that. I do not understand what is so elusive about the etiology of this disease.	Same symptoms, but react differently to drug; Etiology puzzling; frustrating	The same but different from other patients. Unknown etiology frustrating.
I mean I suck up information. I belong to the page on Facebook on National SSc foundation and lot of time I post academic articles and I read all of them and I print them out. I have a binder of stuff and keep track of different - there is a, trying to remember the name of it, it is an international clearinghouse of research trials for different drugs and I am constantly on that looking for that I can jump on board for.	Sucks up information; Active online; reads research articles	Information seeking. Sources: online, research articles.
That the day is going to look like based on when I wake up. Generally, pretty much I am on the mark. I have gotten good at predicting. Again, altering what you do makes a huge difference. If my knees hurt in the morning I am not wearing heels, because it is going to hurt worst.	Predicts the day. Has become good at predicating; What she does makes a big difference. It is going to hurt	Anticipating; Experience; Doing makes a difference; Learning from mistake

Appendix J

Sample Themes and Codes

Initial Themes	Initial Codes
Assessing care	*Not a good feeling about the medication that was prescribed.* *The only thing that really, really worked.* *didn't like side effects; had it under control already.* *Getting team of doctors in a SSc center*
Anticipating-Planning	*Attacking problems differently* *Noting where can sit when walks* *prepare for cold weather with RP*
Learning from mistake	*knees hurting wearing wrong shoe* *Mistake going to Mayo Clinic* *Doctor giving wrong medication.* *Wasted visit with lab results not there* *Nurses not knowing how to draw blood.* *Doctor not monitoring drugs - dangerous blood levels* *Ignoring infection and having to have surgery* *Blood tests not accurate.*
Overcoming fear	*no longer frightens by symptoms* *less frightened, better knowing than wondering.* *less scared with device*
Individualizing	*Asking technician how compared to others.* *asked doctor where fitted compared to others.*

Appendix K

Comparison to Other Scleroderma Studies

Table K1. Comparison Data: Gender/Education/Employment Status

	Sample Size (n)	Gender & Age			Education				Employment					
		% Female (Range)	% Male (Range)	Mean Age (Range)	% HS or Lower	% Some College	% College Grad	% Grad School	% Full-time	% Part-time	% Unemployed	% Retired	% Homemaker	% Other
14 Studies	<100 (5 studies)	81-89%	7-18%	53.0 – 60.3 +/- 12	24 - 68%	27%	27%	24%	22-54%	9%	3-46%	37-75%	4%	1.5%
	100 -500 (4 studies)	81-90%	10-19%	49.05-51.8 +/- 14.2	15-67%	10-35%	14-23%	8-47%	25-42%		27-29%	20-28%	12%	6%
	> 500 (5 studies)	83-87%	13-17%	55.4-56.1+/-12	39-51%	24%	11%	27%	24-43%	9%	33%	30%	12%	8.4%
This study	202 (All cases)	90%	10%	60.0+/- 13.6	19.5%	13.4 %	34.3 %	32.9 %	26.9 %	8.5 %	23.5 %	34.8 %	5.5 %	1%
	183 (PAM valid cases	90.2	9.8	59.9+/- 13.5	19.7 %	13.1 %	34.4 %	32.8 %	27.3 %	9.3 %	23.0 %	33.9 %	5.5 %	1%

Table K2. Comparison Data: Disease Duration and Subtype

	Sample Size (n)	Disease Durati	Disease Subtype			
			% Limited	% Diffuse	% Sine	% Localized/ Do not know
14 Studies	<100 (5 studies)	11+/-8	37-67%	9-63%	13%	
	100 -500 (4 studies	8.7+/- 8.3	42-75%	25-58%		
	> 500 (5 studies)	11.25+/-9.2	62-83%	26-42%		
EUSTAR as of 6/2011	7655		57.7%	36.9%		
CSRG as of 3/2007	1465		63%	37%		
German as of 2/2007	1483		45.5-48.2%	31.6-32.7%		
Univ. of Pittsburg 1972-2007 Steen (2012)	3148		48.45%	43%		
This study	202	11.4+/-10	43.3%	34.3%	5.0%	9.5/8.0
	183	12.5 +/-6	44.8	34.4	4.9	8.2/7.7

Table K3. Comparison Data, Symptoms, and Organ Involvement

	Sample Size (n)	Symptoms and Organ Involvement											
		% Pain	% Fatigue	% Sleep Disturbance	% Depression	% Lung	% PAH	% Heart	% Raynaud's	% Gastro/ Intestinal	% Joint/Muscle	% Skin	% Kidney
14 Studies	<100 (5 studies)					30	22		100	75	13		33
	100 -500 (4 studies)		76										
	> 500 (5 studies)	56-75	56-76	53	51								
EUSTAR As of 6/2011	7655												
CSRG As of 3/2007	1465					34.5							
German As of 2/2007	1483							15.8	94.4	79.9	47.5	87.8	10.5
Univ. of Pittsburgh 1972-2007 Steen (2012)	3148					45		18	98	62.5	79.5	95	9.75
This study	202	69	80	59	34	52		27	83	71	59	76	11
	183	68	81	60	34	53		26	84	72	60	76	11

Appendix L

Themes and Patients' Accounts

Table L1. Obtaining a Diagnosis: Perception of Initial Encounters with Doctors

Patient's Perceptions	Selected Patients' Words
Dismissive • Patients not being heard • Symptoms not taken seriously • Unwillingness to order lab work • Discarding patients' evidences or hunches	*Ralph: [complained about extreme pain all the time] It "kind of bothered me [that doctor did not believe him,] because you think I am lying to you."* *Sharyl: [saw an article in NY Times with pictures identical to what happened to her hands. Took the article to her doctor.] "My doctor, he threw the clipping on the side and he said, 'Don't tell me. I'm the doctor. You have dry hands.'*
Fatalistic • Nothing can be done • You will die in 5-6 years • Get your life in order • Will have lung/heart failure • Invoked fear	*Harper: "He [doctor] said, I have SSc and there is not much we can do at this time. We usually have five years and we will lose you."* *Jenelle: 'You're going to die,' but I just wasn't listening … doctors were still telling people you had at most, six years."* *Francis: [After 1 ½ years of doctor hopping finally a doctor diagnosed him, but refused to treat him] saying, "these are very complicated medicines …they have side effects". "I said you know I am dying. If I am going to die, I am going to die trying."*
Measured • No explanation of disease was given • No indication how disease might progress • Patients felt confused as what they had and what could be done.	*"Marcia: [Doctor] calmly, blithely announced to me—he didn't say anything about it but then he said, 'You have SSc.' Well, I had never heard of it before and he gave me no indication that it was anything to be especially concerned about or alarmed about. So as far as I was concerned, he could have been telling me that I had a bad cold. "* *Bryce: I didn't even know what to make of it. I was like, "Okay, can I get a cream that I can put on my hands? What is it?" He's like, "No, it's a little bit more involved than that. It's a disease. It's tough to say," and there were no answers as far as, "Okay. Is this going to shorten my life?" "Well, we can't really say." Really, everybody is different and they really couldn't say what it would mean for me as far as my life experience from that point on. So, I was just a little confused.*
Supportive • Doctors were attentive and listening taking note of all symptoms • A confirmatory diagnosis was given, with no ambiguity • Nature of disease was made clear. • Patient was informed of symptomatic treatment, but no cure-all • The need for a team of doctors was explained • Previous damages cannot be fixed • Patients felt hopeful	Winola: [Dr. said]: *"yes, you have this and we can treat the symptoms. There's really no one medication to take for the whole thing so we're going to give you a great team. The team is going to work with you and we're going to address the symptoms as they come up…. "[He] was always there for whenever I needed him. If I had any kind of question, he'd call me back personally himself. If it was seven or eight o'clock at night, he'd call me back himself, so I always felt comfortable if I had a problem because I knew I could get a hold of him.* Kristeen: *He wrote down so much and he asked so many questions and he did explain a lot to me, "Right now, you're alright. You're going to progress and we're going to try and start things but for the moment, you're okay." And I think it helped me to hear somebody tell me that.*

Table L2. Managing Relationships with Doctors

Issues	***Select Patients' Words***
Expertise	*Nicole: I would say she's not a warm and fuzzy person. However, she is very thorough. You walk in she goes through a list of things that she has checked. As far as my progress, she explains why she wants me to do the next pieces that she always has for me. You leave from there feeling like she's keeping up and she wants to make sure you are too.* *Lauren: A lot of people don't like my rheumatologist but I like her. I think she's just busy and honest mainly because my rheumatologist will say, "Well, go to the SSc center and get looked at." She recognizes the fact she doesn't have all the answers and I sort of like that. That's fine if the doctor says, "I don't know. Can I get back to you?" Or, "I don't know. I got to think about that." That's fine. I'm not afraid of a doctor saying that to me. I actually think that's probably good advice.* *Francis: She[Dr.] is very, very meticulous.*
Communication (listen, convey information, honest)	*Kristeen: I think that they being patience with me helps and then I can also ask them questions and they don't act like they think it's a stupid question even if they do think it's a stupid question. If they at least treat me with respect, I think that I will do the same… So, I think sometimes, asking questions rattles them but yet, if you have a good physician, they will drop the façade and talk to you.* *Ralph: I expect him not to candy coat it; I just want to know what it is now and next and that is it. Tell me what it is, do not give me false hopes and as far as that goes you know, I know you are working on a cure and give me a call when you got it. Do not just tell me you are working on it.* *Lauren: So, what I want him to do—and many doctors do this very, very well- is they can give me an example of what treatment, how they think it's going to help me. I want my doctor to tell me, "How do you think it's going to help me, how it's going to help the skin or this and that? Why do you think this? Not because your computer said this is the protocol for SSc.*
Changing doctors	*Mackenzie: That's why I think I changed my doctor, the rheumatologist, because she really wasn't giving me a lot of information…. [The new doctor,] She's straightforward and she really cares. …She makes me understand things.* *Willa: Most important thing to me is practical expertise. Second, would be personal rapport, the ability to communicate. If the doctor is a poor communicator I would switch.*
Looking in the eyes	*Lauren: Do they look me in the eye?... It's okay if they don't but in my mind, it's like, Okay, this guy is not seeing me as a person and his eye is on his tablet and he's so focused that he's doing the standard protocol and he's not seeing the individuality.* Kristeen: *Well, when I went to see the primary care that I see now—I've seen him for all these years now—he didn't look me in the eye but he wrote down so much and he asked so many questions and he did explain a lot to me.*
Patients' role- Expressive of their wants & needs	*Ralph: Listen to me I am the patient. I need to not have this pain and I rather have bleeding …these are the side effects, but they are side effects I live with; I choose to live with. To me it was easier to buy Band-Aid and keep them in my backpack than to go off of Prednisone … He kind of finally one day said you are right, kind of milestone, a marker so to speak where he said you know your own body, there are no two ways about it and that was his reaction.* *Lauren: I said, "I'll work with you but you got to work with me… I didn't jump and all of a sudden voice all of this. First, I let him come in and ask and check me and do everything and then when it was time for me to talk, I explained about myself and my own feeling of how I want to wait taking medicine for this disease until I feel that I can't function rather than the belief that "Catch it early. Go on all these."*
Patient's role- Monitoring level of care	*Laurie: At the moment, I feel that my doctor has slipped a little bit. Now, I know it's because of the pressures of work. I know what's happening at that hospital and I also know that she was away teaching … but I do expect a level of response and speed, one, and in depth. When I have queries, especially when it comes to being on the new drug and to do the side effects, they need to get back to you ASAP… At the moment, I'm a little upset with my doctor because I feel that she's being a little, not dismissive, but just overly busy and hasn't responded to me the way I would have liked last week. So, when I see her, I will bring it up.*

Patients' role-Not get intimidated	*Kristeen: Yeah, I think at the beginning, I just did whatever they told me. They used to scare me. The doctors would scare me but like I said, I don't know when it just started to change but I began to realize they get up in the morning and just like me every day and they may have gone to school for something but they don't know everything and I'm really like—so maybe I know, too, and maybe we have to work together instead of me just listening to you.* *Laurie: I know some people are intimidated, but I am not intimidated by medical experts, not in the least… Sometimes, they're surprised at the number of questions and the depth of questions and sometimes, they're not. Again, as I said, it's up to the patient to really educate themselves and knowledge is power and it takes away the fear.* *Rene: some people are afraid that they're going to—they're afraid of the feelings that the rheumatologist may have if they go elsewhere [for a 2nd opinion or treatment], but my experience is anybody that is worth their salt—or however they say it—is not going to mind. In fact, they welcome it.*

Table L3. Pursuing Effective Therapies

Engagement with Finding Therapies	**Selected Patients' Words**
• Tittering drug, looking for right dosage with least side-effect	*Marcia: I work with the doctor in tittering, for example. The doctor doesn't know what the dose is for me so, "Take this and let's see how that goes. If there's a wrong reaction, if you're taking too much, we'll reduce it, or if it's not working, we'll increase it*
• Determining efficacy over an extended period of time	*Winola: He had me on Cellcept (an immunosuppressant) and I was taking that and taking it and I did not notice anything different, nothing changed, nothing improved, nothing got worse or better so after six months, I finally told him, 'This is not for me. I don't notice any kind of change. I still feel the same way. Nothing is improving.' So, we moved on and tried something else.*
• Experimenting with combination/cocktails of drugs	*Laurie: There were five, maybe six medications for PAH (pulmonary arterial hypertension secondary to SSc) when I was first diagnosed in ___ and it was a matter of the doctor finding which cocktail of medications were going to help you most, because everyone reacts so differently. So, there was a trial period where I was on the Viagra, … then she changed me to the Cialis, … and then this other new drug came on the market, Letairis. It was available and she said, 'Let's throw Letairis into the mix and see how you do.'*
• Serially trying drugs	Willa: *I remember trying different drugs [for Raynaud's]. I tried everything from Norvasc, Prozac, and nothing worked, really. I think the last one I tried was Revatio and I finally told my doctor I do not want to do anything … Revatio gave me a headache. It opened up all blood vessels not just the ones in your hands. So, my face gets flushed, my head starts hurting. I said I do not want to suffer through this for my fingers.*
•Insurance drug coverage complications	*Sandy: He [doctor] gives me choices, like Methotrexate. He said we can try that with something else, and I said no I do not want to stay on Methotrexate. It is not working for me. It is making me sick. It makes me feel like crap. I understand, like for me Rituxan (a chemo drug that destroys B cells) is insanely expensive. To get the approval, he said we have to go through the steps. To say we tried this, we tried that for the insurance to approve it. Ok, fine I understand the hoops you have to jump through, but beyond that I do not want to just test everything. Let's try that one. Let's just go and try it… we had to try x, y, z. If we can try the minimum of what we have to try to get that approved, I am not just going to say ok [to every drug], because it is my life.* *Stacee: I have done 6 months of chemo, because one of the drugs that he prescribed which was Cellcept (a pill), all of a sudden, they stopped covering it on my insurance. I said, I am not paying $400 a month; I have insurance; it should be covered. So, he*

	started me on a course of Cytoxan (chemo) intravenously. So, it was 10 hours at a time. It was terrible. So, I go in the morning and strapped up to the thing and infused over 10 hours…I did it every 2 weeks for six month. It was bad. Then my hair started falling off and then they stopped that and then my insurance changed and they covered the Cellcept, so I went back on that and now, of course, it is not covered again and I do not know who are the people who make all these decisions. I just got a letter last week that said that it is no longer on the formulary.

Table L4. Attending to Emotional and Psychological Impact

Sample Psychological Burden	**Select Patients' Words**
Fear of Progression	Paula**:** *One educational forum I went to in __, it was like an eye opener to me: these people with cripple hands, mouths that could not close. I was upset, you know what, that could be me in a couple of years.* Sandy**:** *I went to a research seminar … I was in a room filled with other SSc patients and seeing the vast difference and seeing people with that, I felt bad saying this, but I feel like it is like you were like bird people because the skin so, the mouth is smaller and makes their nose look elongated like [a beak] … I was freaked out and I was like I do not want that to happen to me.*
Body Image Distress	*Rene: I think one of the toughest things that—this shouldn't be one of the toughest things- but the physical changes that SSc does to you and if you saw a picture of me before and a picture of me now, pretty significantly different.* *Francis: When I first got sick, I had people that I have not seen every day, a month or two they had not seen me; they have come to me face-to-face and they would not recognize me. Even now, I have people that I have not seen in 2 or 3 years. People come to me. I had a customer that came to me and asked me face to face saying I am looking for ____. Here I am.*
Don't look Sick	*Lauren: I think they all think I look good. I'm an invisible health issue so they don't know. I mean, they know but they don't know.* *Brittany: When people looked at me, they did not know what I was feeling inside. They really couldn't tell. It wasn't obvious. I didn't have wrinkles. I actually looked good for a while…they didn't know the pain because it was an internal pain. Even now, people do not realize—if they don't see my hands, nobody would know.* *Sandy: I find one of the biggest difficulties is that because I look normal that people do not get what it is that I am dealing with…. So, when people usually like peoples' reactions when I do have that conversation with them is like "but you look so healthy" and I am like, well I guess I should be thankful for that, you know.*

Table L5. Sample Approaches to Coping

Coping Approaches	**Select Patients' Words**
Attitudinal tactics: control what you can, determination To carry on, being inspired by people worse off	*Laurie: It's just managing how the disease makes you feel… There's a lot of SSc issues that we can't do a thing about, but I think again, lifestyle changes. You're just accepting that you can't live your life the way that you would like to. There are limitations and just try to make the best when you do have a good day. You get out there, smell the roses, breathe the fresh air.* *Stacee: I wake up every morning and say ok, put one foot in front of the other and keep going. Regardless of how you feel, everybody has those days that you feel horrible, you know, the aches and pains, my skin hurts and my lungs hurt, but then you make up your mind: am I going to make it through this day or am I going be miserable all day? I have a job to do; I have kids. So, I can either let it overtake everything I am, or I can plow on and hope for the best and keep going and that is what I do… It is just a decision. It's just a decision that you make. Am I going to be miserable? I am not.* *Rene: There were always people worse than I was. I mean, my hands are bad but not anywhere near as bad as people … they couldn't open their hands at all… it doesn't serve its purpose to feel sorry for yourself. Maybe that's just my nature. I don't think I've ever really done that. You just figure out to do what we have to do. Do your best doing that. I don't know*
Doing for others	***Sharyl:*** *But I think the secret, too, when you're not feeling well is that you try to still do for others … you don't feel so sorry for yourself if you have something to do…. So, I think—one of the things would just be—to help others and get out of your own feeling sorry for yourself. That's what it is.*
Not dwelling on illness	***Willa:*** *I kind of accepted it at this point. It is part of what I have to do. It is just like what I mentioned before: I have to go to the doctor; I have more doctors than people who do not have this condition … I guess I think about it, but I do not dwell on it.* ***Ralph****: I am not going to make myself crazy. There are too many other things in my life: we've got __ children and__ grandchildren and we are very involved in our church and I need my energy for that.*

Table L6. Dealing with Employment and Financial Impact

Job/Financial Related Impact	**Select Patients' Words**
Forced to change jobs	*Stacee: I actually changed jobs because I could not, I was in the ____, and I could not handle the instruments anymore. So, I had to change jobs to do something that did not require such a fine tactile movement… So, I got a job doing something in an office setting rather than the _____. You know it is easier. I am not doing what I am trained to do; it is way downscale and I took a humongous pay cut.*
Having to continue working	*Laurie: I need to do a little bit more work because of social security- I'm four credits short of getting my social security benefits when I'm 65 … that's my big concern at the moment is trying to earn these credits. People say, "Well, why don't you go on disability?" I don't feel I'm sick enough to go on disability.*
Needing accommodation at work	*Patricia: I have 2 heaters at work and a heated mouse. I used to share an office with other people and they complained that it was too hot. So, they made me move. In the [workplace], there are all these machines, so it was like 68 degrees, and I am like I cannot do this. So, they made me move to the other side of the hallway, which is nice, but it is temporary for 3 months. I do not know where they're going to make me move then, so I can run my heaters, so nobody complains. I wear hoodie, with the hood up at work every day because it is cold. I don't want to turn purple, and people just give me the stares.* *Jenelle: I was in the [this] business so they are not very giving on giving you extra time away*

	and it was high stress, long hours and very physical work. So that made it very difficult. So, I was also working with a company that was not very sympathetic at all. So, I told them about my situation and I tried to get some accommodations which they wouldn't do and then I went out on medical leave and then they wouldn't let me back.
Difficulty of giving up working	*Rene: It wasn't easy because I finally got my Master's degree. I was the first woman promoted to be a supervisor. I finally reached that level and then kaboom, you know. So, it was hard, but it is what it is.* *Winola: It was very hard. It's so hard. Since July ___ and it's so hard. I used to be a workaholic.* *Sandy: I remember the day: I almost dropped a ___. It was like I hurt really bad that day. My legs were killing me, my hips were killing me, I just was in so much pain and I could not focus mentally. I almost drop this ____ and something just clicked. It was like, I cannot do this anymore, or at least for now. .... When I realized that I had to stop working that was definitely difficult and I think I ignored it for a while.*
Becoming savvy consumers of health services	*Patricia: Pittsburg [Univ. of Pittsburg's SSc Center] is hard. I really avoid going there because it is out of network. So, I have to save up my money to go there. The last time I went, I think, I spent like $400-$500 just to see the doctor and then the prescription of course it did not cover. So, I had to try to get that prescription from my other [local] rheumatologist and then you do not want to piss him off- to be like "oh I went to Pittsburg, oops", you know. So that is hard. Then I had the surgery done for my finger; they referred me to a guy in [my town] that does it. Well, the guy was covered, but the surgery facility was not covered. So that was another 3 grand. Just hard. It bothers me that insurance companies just do not understand that you do not fit the cookie-cutter view and they just don't- I mean I had like appeals and everything and they are like "no, because you are going to go back there and we have to pay again." I am like this is my finger, don't you understand?* *Stacee: I actually found and made an appointment with an [SSc] specialist [out of town] …they called me back and said you do realize that we do not take your insurance? It costs $600 to walk through the door and any additional tests you have to pay out-of-pocket for it; it is usually ranges around $2500 for the first visit". I said thank you very much you can have your appointment.*

Table L7. Handling Familial Relationships

Familial Issues	**Select Patients' Words**
Family's Emotional Reaction to Illness	*Laurie: It was very upsetting especially for my husband. He went to pieces when I was diagnosed.* *Stacee: My husband does a lot of research, but he gets scared. He gets scared by the information, that is why I say do not read everything … he is naturally frightened. We have been married a long time, we have kids, and naturally he is frightened.*
Family's Involvement	*Paula: My daughter definitely [is involved], my husband 85% of the time, my son is oblivious to the world, so I do not think so.* *Mackenzie: My son is a mess but my daughter is very in tuned to my situation* *Maddison: I have a very, very supportive family. I have five siblings and so we're quite close. So, I get a lot of support from them, my children, and my husband.*
Family's lack of understanding or support	*Patricia: I am married but like [he]does not understand., and just gets upset about it… I mean there is not too much support* *Sandy: I think I did have a situation with my stepmother and she just could not wrap her head around the fact, like how can she be sick, like she looks normal.* *Brittany: I found that my parents—my father ... he never really acknowledged my condition, never, which was very annoying. I hated it .... He would say things; "You could open up your*

	hands if you tried, if you wanted to." That hurts because he didn't know the seriousness. He never looked up my disease. He never learned about it..
Tactics used with family	*Laurie: With regard in the family, it's to bring the family and make sure that they understand what's going on, that they understand the medications you're on and possible side effects and how they make you feel and how the disease makes you feel so that they are not—they don't have disappointments. Say, you make a plan to do a family activity and then that day you're just feeling lousy, just want to go to bed, that you can say freely, "I can't do this today."* *Stacee: Some days I do want to be miserable, some days I am just cranky. But again, I do not feel that the disease I have should affect everyone around me. Yes, it does affect them and as a family we deal with that stuff.* *Brittany: I just have learned that I do what I want to do and if I can't do it, don't feel bad about it. Don't apologize; just say it's too much for me. I have to just listen to myself… Don't expect help. Say what you need. Like I'll say to my son, "my garbage pail is really heavy. Can you bring it down Monday night for me? I don't think I can manage it." Then yes, he will do it. Or, "can you pick me up this?" …You have to be exactly specific but to just sit back and think they're going to offer and say, "Hey, Ma! Do you need anything? You want me to come help you do that. I'll give you an hour of my time," which is what I'd love them to say…. I don't want to beg. So that's a hard thing when you don't get help.* *Nicole: I would tell them I don't want to hear about it. He [husband] would try, continually, try. My sister and my husband focused on what was out there, what people were doing. So [husband] could name at the time every doctor in the United States and everywhere and what studies they were doing and he would try to tell me that. My sister, like I said, she was doing some of the same things, my aunt …*

Table L8. Temporal Dimensions of Commitment

Patients' Reactions	**Selected Patients' Words**
Period of Withdrawal lasting several years: The statistics indicating a high mortality rate within 5-10 years, was paralyzing to many creating a scary, confusing time, requiring time to "wrap one's head around it", and accept it.	*Kristeen: One statistic was 50% die within three to five years so that terrified me and had stuck in my heart. So, I think I stuck there, kind of like out of life, for a <u>couple of</u> years- not weeks but years ... I cried all the time. I didn't want to do anything. I was terrified I was going to die tomorrow in my sleep and it was just awful…And then I don't know exactly what clicked in my brain. One day, I woke up and I said, "you know what, it's Monday and my life is wasting away sitting here. Tuesday is going to come tomorrow, and then Wednesday and let's just get on with it." And I think it was my own personal turning point when I just started to say, 'Let's do this.'* Mackenzie*: I went through a period of time where I was like, "Poor me. Poor me." I had to snap out of that… Just by talking to myself, and my friends that said, "Stop living like that. Start living with the solution, not the problem." … Somewhere <u>around four or five</u> [years into illness], I started saying, "I got to start living this. I got to live with this disease. I got to figure that out."* *Brenda: I guess you have to find that new normal. So, I think it took me a <u>couple of years</u> … I think I was quite confused… After I found that stabilization, it fell into place* *Maddison: It really took me <u>two or three years</u> to really [accept] this is it. This is what I have…I decided that I was going to seek the best care that was out there."*

More Immediate resolve to fight on	***Kurt**: it was difficult in the beginning and then I realized either I do something about it and succeed, or I crawl into a ball, go to a corner, wait to die … I knew, almost from the beginning, I had to take [charge of] it. I knew from the beginning because that's who I am. … it means to me that I just need to find ways to cope and survive. That's what it boils down to.* ***Winola:** Well, what are you going to do about it? You're going to sit there, you're going to take it and you're going to live with it and you're going to learn to deal with it. You sit like crying over spilled milk? Lighten up and move on…. It wasn't my attitude in the first month. I can honestly say that. I had a very negative attitude the first month but then after that, I just learned when I started researching it more and finding people who are living longer with it then I had a very positive attitude and I still have a very positive attitude about this.*

Table L9. Strategies

Strategy	Select Patients' Words
Problem-avoidance/ Solving Orientation	***Rene:** I have found that having my own records is the best thing because if you go to a doctor and they, for some reason, they didn't get your lab results or test results then sometimes, your whole visit is in vain.* ***Lauren:** I've always been a problem solver. Some thing's online that I look up and say, "I can make that." I make heat sacks. I put in flax seed and I sew the heat sacks and heat them up in the winter. I have one for the leg, two for the hands.* ***Patricia**: It [her academic training] definitely helps me thinking about things analytically. Like it does not work, let's try a different move or something. I think that helps.*
Utilizing resources & advocating for themselves	*Marcia: My insurance, there is a program connected with Blue Cross/Blue Shield called health advocate. I love it … I called the university medical referral and they have kids there who just look at a book and give you the names of doctors and I wasn't successful in finding a doctor then; they give you names but that's all. Then you call the office, "We're no longer taking new patients." So anyway, this health advocate—…They gave me the names of three doctors and they said, "All three of them are taking new patients. Here are their names, their numbers. Call them and interview them." And I did.*
Transactional Posture	*Patricia: I guess if that [treatment] is working for me, it is good, if not we have to do something else… it makes symptoms better and there is no side effect. So, minimal side effects, the side effects I can tolerate and it do not diminish my life.* *Brittany: The podiatrist told me—because I asked if he would recommend surgery for my feet and he said no … Once I learned that he didn't recommend surgery, even the inserts I buy are better than the ones that he had specially made. S, I'm kind of my own judge.*
Open to Experimentation	*Rene: I did participate in two clinical research trials after that. One of them was for [name of treatment] …. That one, I don't know how much that helped. The other one has a big deal study for relapsing, which they had high hope for. I did it through the University of* *Patricia: So, I think a lot of it is diet, and trying to get the correct nutrition & vitamins and everything. I mean U.S. has a lot of – like a lot of stuff that are ok to eat here, to sell here, eat here like back home- I am from [another country] - back home you could not do that. Back home I can eat wheat. I can eat stuff with eggs in it and I do not get sick. Here I cannot do it.*