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Caring for the Patient With Limited Systemic Scleroderma

Kelly Denise Lachner

Systemic scleroderma (systemic sclerosis) is a rare, autoimmune, collagen-vascular disease of unknown etiology that affects the connective tissues of the skin, internal organs, as well as the small blood vessels. There are 3 subclasses of systemic scleroderma: limited cutaneous, diffuse cutaneous, and sine scleroderma. Prognosis depends on the extent of organ involvement. Complications of systemic scleroderma can involve the cardiovascular, pulmonary, gastrointestinal, renal, integumentary, and the skeletal–muscular systems. Because systemic scleroderma is not common, many orthopaedic nurses may be unfamiliar with how to best provide care. This article provides information about the complexity of the different types of this disease and the basic nursing care of the patient with the most common subclass of systemic scleroderma, limited cutaneous systemic scleroderma.

Introduction

Ann is a 52-year-old woman admitted to your orthopaedic unit with nonhealing digital ulcers of her fingertips that have become infected. She will require intravenous (IV) prostacyclin to preserve skin integrity and IV antibiotics to treat the infection. While completing the admission assessment, Ann tells you that she has a history of limited (cutaneous) systemic scleroderma and is currently being treated by her rheumatologist. Her complaints related to this disease includes fatigue, tight itchy skin, achy joints and muscles, exertional dyspnea, problems with constipation, gastroesophageal reflux disease (GERD), the feeling of food being stuck in her throat, and a poor appetite. She has had Raynaud's phenomenon for the past 10 years and has difficulty with manual dexterity tasks, such as opening jars, buttoning buttons, and zipping zippers. Clinically, Ann looks frail and is underweight. The skin on her hands appears puffy, tight, and shiny with patchy areas of darkened skin. Her fingertips and knuckles have open sores and some are reddened in appearance. As the orthopaedic nurse caring for Ann, you recognize that your unit does not typically see patients with systemic scleroderma. This article discusses the different types of systemic scleroderma and focuses on the nursing care of the patient who has the most common type of systemic scleroderma, limited systemic scleroderma.

Who Develops Scleroderma?

Approximately 300,000 Americans have scleroderma and about one-third of them have the systemic form (Scleroderma Foundation, 2014). However, the early signs are often not identified as scleroderma, so these numbers are believed to be underreported (Uwe-Frithjof, 2011). Scleroderma can develop at any age. Typically, systemic scleroderma is seen in adults, whereas localized, particularly linear, scleroderma is more likely to occur in children (Scleroderma Foundation, 2014). With the exception of linear scleroderma, the most common age to develop scleroderma is between 25 and 55 years, with women being four times more likely to develop this illness (Jimenez, Cronin, Koenig, & Spevak O-Brien, 2012).

What Is Scleroderma?

The term *scleroderma* is interpreted as "sclerosis" meaning hardening and "derma" referring to the skin. There are two classifications of scleroderma: localized and systemic. Each class of scleroderma has subtypes (see Figure 1). Morphea and Linear scleroderma are the two major subtypes of localized scleroderma. Systemic scleroderma can be further categorized into three subtypes: limited cutaneous, diffuse cutaneous, and sine scleroderma. Sine scleroderma typically resembles limited cutaneous scleroderma with the exception of the skin hardening (Chattergee, 2010).

Localized Scleroderma

Localized scleroderma affects approximately 70% of people with scleroderma (Scleroderma Foundation, 2014). Generally, it affects only the skin but can extend to the subcutaneous, deep tissues, and muscles. Localized scleroderma rarely develops into systemic scleroderma (Scleroderma Foundation, 2014). Approximately 10% of individuals affected with localized scleroderma will suffer from scar formation leading to muscle spasms, deformities, and disfiguration (Ravelli, Andriolo, Vasconcellos, Lyddiatt, & Trevisani,

Kelly Denise Lachner, RN, MSN, Mentor Avenue, Painesville, OH.

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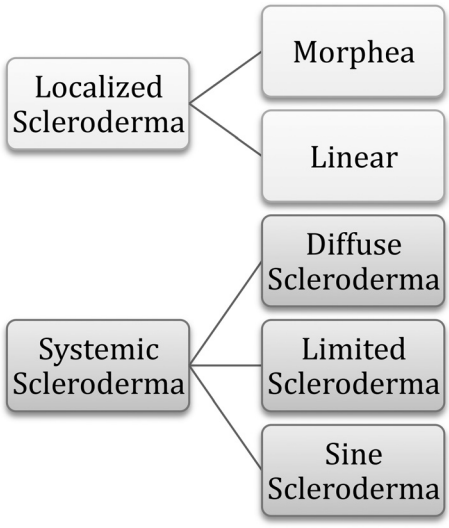

Figure 1. Classifications with subclasses of scleroderma.

2014). Localized scleroderma can be classified into two groups: morphea and linear scleroderma.

Morphea scleroderma can develop during childhood or adulthood and is characterized by patches of fibrotic ivory colored plaque on the skin that may involve the underlying tissues (Ravelli et al., 2014). Linear scleroderma occurs in children and is generally seen as a band-like thickening (linear streaks) that can affect underlying subcutaneous tissues, bone, and muscles (Ravelli et al., 2014). If linear scleroderma crosses a joint, it can cause contractures and growth impairment of the affected limb. Because of the physical disability associated with linear scleroderma, a more aggressive treatment is often used. Treatment for localized scleroderma may include phototherapy, vitamin D derivatives, immunomodulators, and oral and topical steroids as well as physical therapy to prevent any deformity or contractures (Ravelli et al., 2014).

Systemic Scleroderma

Systemic scleroderma can be life-threatening and may affect internal organs, including the gastrointestinal, pulmonary, cardiac, and renal systems (Jimenez et al., 2012). Raynaud's phenomenon is usually one of the earliest signs of scleroderma and is present in more than 90%–95% of people with systemic scleroderma (Hughes, 2012; Parker, 2013). Raynaud's phenomenon affects the small blood vessels of the hands and feet causing vasoconstriction and loss of blood flow to the areas. Symptoms occur with exposure to cold or emotional distress and can cause numbness and pain (Parker, 2013). It is important for the orthopaedic nurse to recognize that most people with primary Raynaud's phenomenon do not develop scleroderma. Moreover, scleroderma-associated (secondary) Raynaud's phenomenon tends to be more severe than primary Raynaud's phenomenon and is more likely to progress to tissue damage including ulcerations and ischemia of the fingertips (Hughes, 2012; Parker, 2013).

Early intervention of systemic scleroderma may help improve quality of life and may extend the patient's life (Hughes, 2012; Uwe-Frithjof, 2011). There is no cure for systemic scleroderma. The treatment is aimed at slowing the progress of the disease by controlling the symptoms and complications associated with it (Chattergee, 2010). The average survival rate is 12 years but is determined by the specific subclass of systemic scleroderma and the organ involvement (Jimenez et al., 2012). As the name suggests, diffuse systemic scleroderma tends to involve more systems in the body, and people suffering from it fare worse than people with limited scleroderma.

Historically, scleroderma renal crisis was considered the most fatal complication of systemic scleroderma (Hughes, 2012). But, because of early intervention and use of angiotensin-converting enzymes inhibitors, deaths from scleroderma renal crisis have decreased significantly (Uwe-Frithjof, 2011). In addition, the treatment of pulmonary hypertension and esophageal involvement has also decreased the morbidity and mortality rates of systemic scleroderma (Uwe-Frithjof, 2011). In spite of this, deaths related to systemic scleroderma continue to be related to pulmonary complications, particularly interstitial lung disease including pulmonary fibrosis and pulmonary hypertension (Hughes, 2012). There are three subclassifications of systemic scleroderma: limited cutaneous, diffuse cutaneous, and sine scleroderma.

Limited Cutaneous Systemic Scleroderma

Limited cutaneous systemic scleroderma, commonly called limited scleroderma, tends to have a slower progression than diffuse cutaneous scleroderma. This is the most common subtype of the systemic sclerodermas (Brown, 2010). More than 90% of the individuals with limited scleroderma will have Raynaud's phenomenon for years before thickening of the skin (Parker, 2013). Skin thickening (fibrosis) is usually limited to below the elbows and knees, on the hands and feet, and sometimes on the face (Chattergee, 2010). Symptoms can range from mild to severe. Mild symptoms can progress quickly into severe disease. The 10-year survival rate for limited systemic scleroderma is 71% (Jimenez et al., 2012). People with limited scleroderma generally suffer from chronic pain, fatigue, and disfigurement (Sierakowska et al., 2007).

Interstitial lung disease, primarily pulmonary arterial hypertension (PAH), develops later in the progression of limited scleroderma and is a key prognostic factor for survival (Jimenez et al., 2012; Varga, 2014). The classic symptoms of pulmonary hypertension include shortness of breath and decreased exercise tolerance (Hughes, 2012).

Limited scleroderma is often referred to as CREST syndrome (Chattergee, 2010; Varga, 2014). CREST syndrome is an acronym for calcinosis, Raynaud's phenomenon, esophageal dysfunction, sclerodactyly, and telangiectasia (see Table 1). However, the acronym, CREST, is being used less often because people suffering with the diffuse type of scleroderma may also have the symptoms of CREST syndrome (Hughes, 2012).

Diffuse Cutaneous Systemic Scleroderma

Diffuse cutaneous systemic scleroderma, commonly known as diffuse scleroderma, tends to have a rapid onset. Raynaud's phenomenon occurs simultaneously

Table 1. Description of the Acronym of CREST Syndrome for Limited Scleroderma

Calcinosis	Raynaud's Phenomenon	Esophageal Dysfunction	Sclerodactyly	Telangiectasia
Accumulation of calcium salt under the skin Causes hard painful raised areas Areas can open and cause ulcerations of the skin	Vasoconstriction with intermittent loss of blood flow to fingers, toes, and nose Occurs with exposure to cold and stress Causes numbness, tingling, and pain	Weakened lower esophagus causing reflux Esophageal strictures cause swallowing difficulties	Excess collagen in the skin layers Thin, tight, shiny skin on fingers/toes Difficulty moving fingers	Dilation of tiny blood vessels causing small blanchable red spots On hands, face, chest, and inside mouth Harmless, may cause disfigurement

with arm and trunk skin hardening (Chattergee, 2010; Uwe-Frithjof, 2011). Organ involvement, including interstitial lung disease, diffuse gastrointestinal disease, oliguric renal failure, and myocardial involvement, generally occurs early in diffuse scleroderma (Chattergee, 2010; Uwe-Frithjof, 2011). The amount of skin thickening (fibrosis) is often associated with the severity of the organ involvement. Recognition and management of symptoms are essential to extend life. The serious symptoms of diffuse scleroderma are typically seen within the first 3 years after diagnosis (Parker, 2013). After the initial first 3 years, the disease may enter a remission stage. However, the damage that has occurred to the lungs, kidney, heart, and digestive system will remain. Diffuse scleroderma has a 10-year survival rate of 21% (Jimenez et al., 2012).

Sine Systemic Scleroderma

Sine systemic scleroderma is a rare form of scleroderma (Chattergee, 2010; Scleroderma Foundation, 2014). Sine means without skin involvement. Most often, the esophagus is affected followed by pulmonary involvement. Sine scleroderma closely resembles limited systemic cutaneous scleroderma with the exception of the skin hardening (Chattergee, 2010). Because of the lack of skin changes, sine scleroderma is often undiagnosed.

Pathogenesis of Systemic Scleroderma

Scleroderma is a connective tissue disease and the etiology is poorly understood. Estrogen, environmental triggers, and genetic links have all been suspected as the cause of scleroderma, but research has been inconclusive. The pathogenesis involves a disruption of the vascular and immune systems, resulting in the increased production of collagen (Uwe-Frithjof, 2011). In greater than 90% of the cases, the vascular symptoms of Raynaud's phenomenon occur years, sometimes decades, before fibrosis occurs (Chattergee, 2010; Jimenez et al., 2012). Regardless of this, it is unclear whether it is the vascular system or the immune system that initiates the progression of the disease (Chattergee, 2010).

Changes in the vascular system involve a disruption of the small blood vessels (microvasculature) in the lining of the blood vessels (vascular endothelium). This causes inflammation and vasoconstriction of the blood vessels sending out a message that it is damaged, thus inciting or advancing the autoimmune response. To further insult the blood vessels, individuals with scleroderma have been found to be 11 times more likely to have a moderate to high incidence of calcium deposition in their blood vessels (Mok et al., 2011). As the body attempts to repair itself (autoimmunity), collagen is overproduced. Collagen is a connective tissue found in the skin, tendons, joints, and other major organs. This overproduction causes fibrosis, or hardening, of the skin and other organs and tissues where collagen is normally found in the body.

Caring for the Patient With Limited Systemic Scleroderma

General Care

Caring for a patient with systemic scleroderma requires a holistic interdisciplinary healthcare approach that includes both physical and emotional support. The nursing plan of care will need to be individualized to each patient because of the heterogeneous nature of the symptoms and should focus on alleviating symptoms, preventing infections, and monitoring for life-threatening conditions (Table 2) (Sierakowska et al., 2007).

Dermatologic Manifestations

Skin care is a priority for patients with scleroderma. These patients suffer from a multitude of dermatologic issues. Often, they suffer from dry itchy skin that can become tight and hardened. It may be difficult to start an IV, draw blood, or obtain a blood pressure because of the thickened or tight skin. These skin changes can lead to a reduction of range of motion and potential tissue damage (Hughes, 2012). Telangiectasia are small red spots that blanche with pressure and are usually seen on the face or hands. Benign in nature, these red spots are dilated blood vessels that can cause cosmetic concerns for the patient. A frequent complication of scleroderma, calcinosis occurs when calcium deposits become localized and create nodules in the subcutaneous tissues. These deposits generally occur in the fingertips or knuckles affecting the patient's ability to perform activities of daily living that require the function of their hand. Calcinosis presents as painful ulcerations as the calcium deposit nodule breaks through the skin (Brown, 2010). Because this calcium deposit is not related to the amount of calcium that one ingests, no nutritional restrictions of calcium is necessary (Scleroderma Foundation, 2014).

Skin changes, damage to the small blood vessels, calcinosis, and tissue damage associated with Raynaud's phenomenon can all lead to skin complications (Brown, 2010). Digital ulcers or injuries to the skin are often difficult to heal and prone to infection. This may be due to damage to the blood vessels that occurs with systemic scleroderma. Avoiding vasoconstriction of the blood

Table 2. Nursing Interventions for the Symptoms of CREST Syndrome

Calcinosis	Raynaud's Phenomenon	Esophageal Dysfunction	Sclerodactyly	Telangiectasia
Care to avoid breakage of calcium deposits Dry sterile dressings are often used May require narcotic pain medication If infected, may require vasodilator to increase circulation to the area for healing No need to restrict calcium in diet, these calcium deposits are of diet related	Must control vasoconstriction of the blood vessels to preserve skin integrity Keep patient warm Avoid drafts/air conditioning Use warm blankets Special considerations pre-/postoperation with warmth Avoid smoking/stress Calcium channel blockers Other vasodilators may be used if severe and patient has calcinosis	Offer small frequent meals Moist foods are often easier to swallow Avoid aggravating foods and alcohol Have patient sit up during and after meals Administer proton pump inhibitors	Protect from injury or bumping Frequent repositioning using pillows May need assistance with activities of daily living especially fine motor movements Exercise ball can be used to maintain mobility of hands and increase circulation	Make-up may help conceal cosmetic concerns May require laser treatments

Note. Data from Chattergee, S. (2010).

vessels is the most important measure to prevent and promote the healing of digital ulcers (Brown, 2010). To achieve this, the patient should be instructed to avoid air conditioning, wear gloves if it is cold outside, quit smoking, and minimize stress. In addition, the patient may be placed on a calcium channel blocker. It is important for the nurse to recognize that calcium channel blockers are only effective preventatively and is not an effective treatment for preexisting digital ulcers (Brown, 2010). Care of noninfected digital ulcers typically includes keeping the area clean and dry with a dry sterile dressing. The goal is to prevent colonization of bacteria. When infection does occur, IV antibiotics and IV prostacyclin may be used (Brown, 2010).

Cardiopulmonary Manifestations

More than 70% of all people with systemic scleroderma will have some loss of lung function (Varga, 2014). Pulmonary arterial hypertension (PAH) can occur as a later manifestation and is the leading cause of death in people with limited scleroderma (Varga, 2014). Pulmonary arterial hypertension results from the scarring (fibrosis) of the small pulmonary arteries, resulting in a progressively increasing pulmonary vascular resistance (Duquette, 2015). Right-sided heart failure may eventually develop as the right side of the heart works harder to move blood into the lungs. The classic symptoms of PAH include shortness of breath and decreased exercise tolerance (Hughes, 2012). Other symptoms may include fatigue, syncope, dependent edema, tachycardia, and elevations in blood pressure. When assessing a patient with PAH, lung sounds may be clear to auscultation (Duquette, 2015).

Diuretics and supplemental oxygen, at night or with activity, are commonly used for patients with PAH. A combination of medications specific for scleroderma patients with PAH includes diuretics, endothelin receptor antagonist (bosentan), phosphodiesterase inhibitors (sildenafil), and inhaled or IV prostandoids (Hughes, 2012). These drugs induce vasodilation of blood vessels and provide an antiproliferative effects.

Nursing interventions for patients with PAH include providing education and monitoring fluid balance, vital signs, oxygen saturation levels, and activity tolerance (Duquette, 2015). Patients need to be advised on compliancy with the complicated medication regimen, diagnostic testing, follow-up appointments, and smoking cessation.

To monitor fluid balance, daily weights and accurate intake and output may be necessary especially in patients with dependent edema. Any increases in weight or changes in serum electrolytes, particularly the potassium and magnesium, should be brought to the physician's attention immediately (Duquette, 2015). Abrupt changes in blood pressure can cause decompensation in a patient with PAH (Duquette, 2015). Therefore, it is important for the orthopaedic nurse to consult with the physician before withholding blood pressure medications despite lower than normal blood pressure readings. Syncope or near syncope may occur as a result of decreased cardiac output, so the nurse should take measures to maintain patient safety.

Oxygen saturation levels should be maintained at 90% or greater when a patient has PAH (Duquette, 2015). If the patient has severe Raynaud's phenomenon, it may be difficult to obtain a pulse oximeter on the finger. Pulse oximeter monitors for the forehead should be available. Supplemental oxygen, providing for rest periods, and elevating the head of the bed can be used to sustain oxygen levels. Gastroesophageal reflux, a common problem in patients with scleroderma, will need to be controlled to avoid further pulmonary complications such as aspiration.

Gastrointestinal Manifestations

Gastrointestinal symptoms occur in more than 90% of people with scleroderma (Varga, 2014). This occurs because of a hypomobility, or a slow movement, of the food in the gastrointestinal tract due to collagen overproduction and can include GERD, dry mouth, difficulty swallowing, poor appetite, and the feeling of food being stuck in the throat (Chattergee, 2010; Nagaraja, McMahan, Getzup, & Dhanna, 2015). Constipation, pseudo-obstructions, and actual obstructions of the bowel can occur because of the slowed peristalsis (Chattergee, 2010).

Poorly controlled GERD can lead to esophageal strictures. Stricture formation can be found in up to 29% of

patients with scleroderma (Nagaraja et al., 2015). Strictures form as healing occurs from the acid burning the esophagus. As this area heals, it causes excessive collagen formation, fibrosis, and narrowing of the esophagus (Nagaraja et al., 2015). To avoid stricture reoccurrences, it is necessary to keep a tight control on the reflux, and if the strictures cause dysphagia, esophageal dilatation is sometimes used (Nagaraja et al., 2015). Nursing interventions need to be implemented to prevent esophageal erosions and strictures and include administering proton pump inhibitors, elevating the head of bed, sitting the patient in a chair while and after eating, and avoidance of aggravating foods and alcohol (Nagaraja et al., 2015).

Approximately half of all patients with systemic scleroderma have gastroparesis (Nagaraja et al., 2015). This delayed gastric emptying can cause nausea, vomiting, early satiety, abdominal bloating, and weight loss. Prokinetics, such as metoclopramide, and anti-emetics are commonly used for the symptoms of gastroparesis (Nagaraja et al., 2015). Patients should be offered small frequent meals and meals should be moist foods, as dry foods seem to be difficult for patients with scleroderma to swallow. Fluids, fiber, and stool softeners as well as increasing activity can help ease problems with constipation.

Renal Manifestations

Scleroderma renal crisis is characterized by a sudden decline in kidney function and an increase in blood pressure. Scleroderma renal crisis used to be considered as the most fatal complication of systemic scleroderma and still occurs in 5%–10% of people (Hughes, 2012). Early intervention and the use of angiotensin-converting enzymes inhibitors have significantly decreased deaths related to scleroderma renal crisis (Uwe-Frithjof, 2011). The use of high doses of corticosteroids is contraindicated in patients with scleroderma because it can lead to renal crisis although, on occasion, low dose of steroids may be used for muscle inflammation or as a conjunctive treatment for interstitial lung disease (Chattergee, 2010; Hughes, 2012).

Musculoskeletal and Mobility Manifestations

Muscle weakness and pain and joint pain and swelling occur during the early stages of systemic scleroderma (Varga, 2014). Often, the hands are affective first. Early morning stiffness affects almost half of the people early in the progression of this disease (Sierakowska et al., 2007). Swelling and achy fingers generally occur before the skin thickens. Sclerodactyly occurs as the skin thickens on the fingers causing the fingertips to contract or curl under. This is often characterized by tight shiny skin. Skin thickening and fibrosis around tendon can cause contractures of different joints. The most commonly affected joints include wrists, elbows, and ankles (Varga, 2014). This can cause difficulty for the person when performing his or her own daily living tasks and she will most likely need assistance with personal care. To maintain mobility of hand function, occupational therapy should be working with the individual. Hand exercises may include using a stress ball that will help with the mobility of the hands as well as facilitating circulation to aid in problems such as Raynaud's phenomenon and digital ulcers.

Mobility safety is an issue for a patient with systemic scleroderma for a variety of reasons. Pulmonary artery hypertension may cause syncope or near syncope. Generalized fatigue, malnourishment, and a decrease in range of motion may cause weakness resulting in fall risks. This patient would be a high fall risk. Nursing interventions to prevent falls should be implemented. Physical therapy should be consulted and include low-impact strength training exercises. Home care posthospitalization may be necessary to evaluate home safety, provide physical and occupational therapy, and evaluate the need for assistance with daily living tasks.

Psychosocial Support

Systemic scleroderma is a chronic, incurable disease that causes fatigue, functional disability, and disfigurement. High morbidity and mortality is associated with this disease. In a study conducted by Sierakowska et al. (2007), it was concluded that patients with systemic scleroderma had increased anxiety and depression related to the fear of negative evaluation from others and suffer from bouts of low spirits, anxiety, and fear. These patients may feel embarrassed by the distorted way they look. When caring for the patient with scleroderma, the nurse should provide care in an empathetic nonjudgmental way allowing time for a patient to verbalize their needs or fears to promote a trusting relationship.

There are two scleroderma support groups that may help an individual with emotional support. The Scleroderma Foundation (www.sclerodermafoundation.org) has local support groups throughout the United States, and the International Scleroderma Organization (www.scler.org/forums/) has online scleroderma support groups and chats.

Caring for Ann's Infected Digital Ulcers

Remember Ann from the introduction with the infected digital ulcers on her fingertips? Her infected nonhealing digital ulcers may be considered a medical emergency because they can quickly progress to gangrene possibly resulting in amputations (Brown, 2010). Ann was admitted to your unit for IV prostacyclin to preserve her skin integrity and antibiotics to treat the infections of her digital ulcers. In scleroderma, digital ulcers can result from a variety of skin issues associated with scleroderma, but because of the inflammation and vasoconstriction of the blood vessels associated with Raynaud's phenomenon, nonhealing digital ulcers are difficult to treat. Prostacyclin drugs are used to increase circulation to the fingertips, thus promoting the healing of the ulcers. According to the Cochrane Review, iloprost is an effective vasodilator and platelet-aggregation inhibitor often used in the treatment of Raynaud's phenomenon secondary to systemic scleroderma, therefore promoting the healing of digital ulcers (Pope et al., 1998). Pain will often be a concern for patients with digital ulcers, so opioid medications should be given prior to any dressing changes or debridement.

Conclusion

Individuals are living longer with systemic scleroderma, thereby increasing the likelihood that the orthopaedic nurse may encounter a patient with this devastating disease. Affecting nearly every aspect of the person's life, scleroderma is a complex and progressive disease. Although there is no cure, treatment options may improve the quality of life and extend the life of the patient with scleroderma. Treatment options are generally related to the symptoms or organs involved. Diffuse scleroderma tends to have a more rapid onset, but limited scleroderma can be just as devastating. Respiratory complications occur in more than 70% of scleroderma patients and is the major reason for mortality. The characteristic signs of scleroderma are Raynaud's phenomenon and hardening of the skin (fibrosis). Generally speaking, the extent of the fibrosis relates to the extent of damage to the internal organs.

Because of the heterogeneous nature of scleroderma, the nursing plan of care must be modified to meet the individual patient's needs. The nurse is the perfect advocate for this patient as she or he is placed nicely in the middle of the multidisciplinary team taking care of the patient with scleroderma. It is important to involve the patient in all aspects of their medical care to ascertain compliancy, trust, and cooperation. Alleviating symptoms, preventing infection, and observing for life-threatening conditions are the goals of the nursing care for this patient.

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