FEATURE ARTICLE
Continuing Education

Scleroderma

Living with Unpredictability

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Many individuals with chronic health conditions are living longer and coping with their conditions while attempting to work and enjoying active and productive lives. Some of these conditions, such as diabetes, stroke, spinal cord injuries, and traumatic brain injuries, are well documented in the literature and generally understood by nurses. Other conditions, such as scleroderma or systemic sclerosis, are less understood because of their rarity. Nurses may encounter individuals with scleroderma in a variety of settings, including adult inpatient units, home health care, rehabilitation care, and the work setting. The purposes of this article are to provide a review of scleroderma, to identify implications for occupational health nurses in understanding and managing the condition, and to assist workers in coping with this disease successfully.

WHAT IS SCLERODERMA?

Scleroderma is classified as a connective tissue or collagen disorder, and leads to progressive fibrosis. Scleroderma is characterized by fibrosis of the skin, but can also involve the muscles, joints, blood vessels, and internal organs. Problems with fine motor control, difficulty manipulating buttons and opening jars, shortness of breath, and coughing are common. The physical symptoms can range from shiny skin to an unusually small mouth or grossly deformed fingers. Because of esophageal problems, the individual may be unable to swallow effectively and may resort to a diet of soft foods or liquids. Individuals report choking problems and often choke on their own saliva. Involvement of the small and large bowel results in constipation or diarrhea.

CAUSATION

The cause of scleroderma is unknown, although environmental factors have been associated with the development of some of the clinical forms. Environmental agents implicated in scleroderma can be divided into three categories: silica, organic solvents, and drugs (Silman, 1996). Silica dust was first associated with scleroderma in a group of gold miners in South Africa in the 1950s. Organic solvents associated with scleroderma include toluene, benzene, and vinyl chloride. Drugs associated with scleroderma include bleomycin, pentazocine, and cocaine. Additional environmental agents have been associated with scleroderma or scleroderma-like conditions. In 1981 in Spain, scleroderma-like symptoms occurred with a foodborne disease known as toxic oil syndrome (TOS) (Silver, 1996). The suspected cause was rapeseed oil denatured with aniline, which had been sold as pure virgin oil. Some of the cases of scleroderma related to environmental factors may differ from classic clinical scleroderma, and are termed pseudoscleroderma (Silver, 1996).

HISTORY OF THE DISEASE

Because of the rarity of the condition and the diverse nature and unpredictability of its symptoms, scleroderma is a condition little understood by both the general public and health care providers. Both Hippocrates (460 to 370

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Table

Subtypes of Scleroderma

	Localized (limited) Scleroderma	Systemic (diffuse) Scleroderma
Incidence rate per 100,000	2.7	1.9
Prevalence rate per 100,000	50 to 220	.4 to 29
% of total	80%	20%
Female:male ratio	3:1	5:1
Onset	Slower onset, usually following Raynaud's	More rapid onset
Skin	Skin sclerosis limited to hands, feet, face, and forearms, or absent	Truncal and acral skin involvement
Organ involvement	Low risk and later involvement of major organs	Early and significant major organ involvement of the lung, heart, gastrointestinal tract, or kidney
Average age at diagnosis	33 years	49 years
Survival rate	> 70% at 10 years	40% to 60% at 10 years
Raynaud's phenomenon	May precede by 10 to 15 years	Onset within 1 year or at time of first symptoms

Gulin, 1999; Haustein, 2002; Hawk, 2001; Humes, 2001; Silman, 1996.

BC) and Galen (AD 130 to 299) describe cases compatible with scleroderma (Barnett, 1996). However, the first diagnosed case of a scleroderma-like condition was described by Curzio of Naples in 1753. The client was a young woman suffering from excessive hardness of the skin. Scleroderma was first recognized as a clinical condition in the mid-nineteenth century, and was given the name "sclerodermie" by the French physician Gintrac. In the second half of the 20th century, the involvement of internal organs was recognized.

SYMPTOMS

Scleroderma presents a variety of physical symptoms that come and go, symptoms that may change within the same individual as the disease progresses. The course varies with the individual, is unpredictable, and often progresses slowly (Casale, 1997). Common symptoms of scleroderma include:

- Generalized fatigue.
- Weight loss.
- Pain.
- Swelling of fingers.
- Arthritic joints.
- Hard skin.
- Tight mask-like facial skin.
- Thick shiny skin on arms and legs.
- Esophageal reflux.
- Dysphagia.
- Skin discoloration.

Difficulty coping with cold temperatures is also common.

Living with scleroderma is a life of unpredictability—individuals never know how they will feel at any given time. One day they might experience leg pain, the following day chest pain. The symptoms vary within an individual and from person to person. Fatigue is present most days. Coughing is not only a physical problem, but also a social one because it is noisy and disruptive to daily living. The changes in physical appearance brought on by scleroderma cause anxiety in most affected individuals. A sense of loss is also present in individuals who are not able to take part in family activities they previously enjoyed. The unpredictable nature of the symptoms and the future brings considerable anxiety.

Most people with scleroderma have Raynaud's phenomenon, which occurs in only 20% of the general population (Sule, 2000). Raynaud's is characterized by intermittent attacks of color changes in the digits in response to cold, and in some situations, to stress. Numbness, tingling, and burning sensations may occur during the attacks, and Raynaud's phenomenon can ultimately result in loss of fingers and toes. For some, the symptoms include involvement of the toes, nose, ears, and tongue. Clients should be encouraged to maintain body heat by keeping the torso covered with layers of clothing, in addition to keeping the hands and feet well covered. Whenever possible, clients should avoid extreme cold, or prolonged periods of cold, either outdoors or in air conditioning. Workers need to maintain a warm work environment because the most effective method of preventing Raynaud's phenomenon is to avoid exposure to cold.

DISEASE PROGRESSION

The disease manifestations are highly individualized and depend upon the part of the body affected. Clinically, the condition may be divided into two main sub types: sys-

temic and local scleroderma (see Table). Systemic (diffuse) scleroderma usually involves a more rapid progression of internal organ involvement, resulting in fibrosis atrophy of the heart, lungs, kidneys, and gastrointestinal tract (Black, 1997; Sule, 2000). Complications can include cardiac, lung, or renal failure. Although no outward signs of the scleroderma may be present, the manifestation of the systemic form of the condition can be life threatening. In diffuse scleroderma, there is rapid skin involvement occurs during the first 2 to 3 years of the disease, with a high risk of internal organ involvement during this time (Humes, 2001). The disease then appears to plateau.

When the condition is local (limited), the progression is slower and internal organ involvement is delayed. The manifestations are skin and underlying tissue involvement that can result in serious facial disfiguration. A thickening and tightening of the skin occurs, accompanied by swelling and edema. Morning stiffness and pain in the joints are common. The skin of the face and neck become involved, and the lips may become pursed. Mouth opening becomes limited and causes problems with eating, drinking, dental hygiene, and the fitting of dentures. Limited scleroderma is also termed the CREST syndrome (i.e., subcutaneous calcinosis, Raynaud's phenomenon, esophageal dysmotilily, sclerodactyly, telangiectasia) (Gulin, 1999; Humes, 2001). Individuals with limited scleroderma show a continuing progression of skin and visceral involvement. For the most part, morbidity and mortality rates for scleroderma are related to the amount of visceral involvement.

Diagnosis and Treatment

The diagnosis is based on history and examination, with the extent of scleroderma skin involvement a key determinant. The assessment includes measures of subjective symptoms, degree of disability, and physical and psychological functioning. It also includes barium swallow, skin biopsy, electrocardiogram, renal function tests, endoscopy, and studies of the small and large bowel. Sclerodermatous changes occur in a variety of conditions and a differential diagnosis must be made to distinguish these pseudosclerodermatous conditions from clinical scleroderma. Sclerodermatous changes have been reported in endocrine conditions such as diabetes, and in malignancies such as myeloma (Jablonska, 1996).

The treatment is individualized and determined by the degree of skin involvement and the presence or absence of internal organ involvement. Treatment is aimed at modification of the disease to slow its progression and to manage symptoms. Although no curative therapy is available, the survival rate has increased during the past 20 years, largely because of better management of renal crisis in systemic scleroderma. Vasodilator therapy may assist with symptom relief. During the early edema and swelling phase of the skin, corticosteroid therapy may be effective.

CASE STUDY

Ms. C is 53 years old and was diagnosed 9 years ago as having scleroderma. Divorced, with a 12 year old child, Ms. C lives in a two bedroom town house in a large urban area. She works as a secretary or receptionist in the student records department of a community college. Nine months after diagnosis, she had kidney failure, followed by a kidney transplant. After the transplant, she broke a hip, which left her with difficulty walking. She feels that being so ill and near death (i.e., at the time of the transplant) has made her a better person. She said, "I'm more humane, a better person, ...I appreciate life, I work harder, I achieve more...I recognize everyone's potential."

Currently, Ms. C has difficulty swallowing, and has a small appetite. She jokes about the size of her meals when compared to the size of her growing child's meals. She finds it difficult to sustain her energy level, but despite her fatigue, Ms. C acknowledges the importance of keeping active and of daily exercise, no matter how minimal. She has swollen hands, is sensitive to the cold, wears clothes to bed, and wears socks no matter what the weather. The cold affects her ability to speak, which she finds very frustrating.

Despite the many limitations of her condition, Ms. C describes her health as 9 on a scale of 1 (poor) to 10 (excellent). In thinking of the future, Ms. C hates the thought of deteriorating visually and becoming disfigured. Ms. C wants to work, yet fatigues easily. She is proud of the quality of her work and does not wish to be viewed by her coworkers as ill. When she uses her computer, her hands feel very cold and stiff. She is worried about being able to remain in her job and support her child. The occupational nurse can be of assistance to Ms. C and others with her condition by following management protocols.

MANAGEMENT PROTOCOLS

Mobility

Stiffness, tenderness, pain, and swelling frequently accompany scleroderma. Daily range of motion exercise is encouraged to prevent stiffness. The facial muscles need to be exercised as well. A moisturizing cream should be applied prior to facial exercises. Occupational health nurses should remind individuals with scleroderma of the importance of daily range of motion exercises in preventing and slowing joint contractures. At home and in the work setting, individuals should be encouraged to move around frequently to prevent stiffness and discomfort. Depending on the situation, referral to a physiotherapist may be required.

Mouth Care

Scleroderma may cause the mouth opening to be reduced in size, making lip movements and oral hygiene difficult. Mouth stretching exercises may delay tightening around the mouth, but must be performed with care to prevent further damage. Individuals with scleroderma need to be encouraged to chew their food thoroughly. Occupational health nurses can promote the importance of preventive dental care, including regular flossing and brushing of the teeth and gums. Regular visits to the dentist should be encouraged as infections and dental caries are a risk.

Nutrition

Esophageal dysfunction can lead to difficulties in swallowing and may lead to acid buildup with resulting heartburn. Small, regular meals with plenty of fluids are

IN SUMMARY

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Living with Unpredictability

Acorn, S., & Joachim, G.

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1 Scleroderma, or systemic sclerosis, is a little understood chronic condition, largely because of its rarity. It is a connective tissue or collagen disorder characterized by fibrosis of the skin, but can also involve the muscles, joints, blood vessels, and internal organs.

2 Occupational health nurses must be aware that individuals with scleroderma require assistance in areas such as mobility, mouth care, nutrition, depression, information, and support and accommodation in the workplace.

3 Scleroderma frequently occurs during the productive years, with many individuals remaining in the work force for years after diagnosis. Alternative work options can assist in retaining individuals with chronic conditions, such as scleroderma.

recommended. Food should be high in nutritional value and low in volume. It is preferable to remain upright during eating and for 2 to 3 hours after eating, to avoid problems with swallowing and to aid in digestion. Some individuals need to sleep with the head of the bed elevated and to use antacids for comfort. In the workplace, the occupational health nurse can facilitate a schedule to include more frequent breaks and educate other workers about the reason for this.

Depression

As with many individuals living with a chronic condition, workers with scleroderma are more likely to suffer from depression. Angelopoulos (2001) reported that almost half of the clients with scleroderma in the study presented with mild depressive symptoms and an additional 17% with moderate to severe depression. The occupational health nurse must be aware of the potential for depression and observant for signs of depression. Attention to symptom management and pain relief is essential. Antidepressants may be required, and continuous familial and collegial support from the work group is equally important.

Information and Support

As with many individuals who have a chronic condition, those with scleroderma need strong support from families, coworkers, and support networks. Frequently, diseases that are little understood can provoke negative reaction among others. The history of AIDS, for example, shows a disease by which individuals are stigmatized and feared. Living with a chronic disease, especially when the condition includes physical deformities, may subject a person to stigmatization. By becoming knowledgeable about scleroderma, the occupational health can do much to promote understanding and lessen stigma.

In the workplace, the nurse can facilitate the understanding of coworkers about the condition and provide ongoing support to the individual with scleroderma. The nurse should first encourage the individual with scleroderma to discuss the condition with others, and then provide additional information as needed. Community scleroderma and arthritis support groups offer useful information and management tips as well as emotional support. Mayes (1999) offers a well written review of the disease and offers many helpful hints for clients and families living with and managing scleroderma. Online information sources include:

- http://www.arthritis.co.za
- http://www.merck.com/pubs/mmanual/section5/chapter50/50g.htm

Accommodation in the Workplace

Contrary to the assumption that individuals with a chronic condition are unable or unwilling to work, many wish to remain productive and maintain as active a life as possible. Scleroderma frequently occurs during the productive years, and because of its unpredictability and chronicity, many of those affected remain in the work force for years after diagnosis. Individuals with scleroderma may require alternative work options or retraining to accommodate them in the workplace. Alternative work options for the older worker are outlined by Healy (2001), and many are relevant to individuals with chronic conditions and include:

- Compressed work week.
- Flexible work schedule.
- Job reassignment.
- Part time work, temporary assignments, or job sharing.
- Telecommuting part or all of the time.
- Retraining for new careers.

The occupational health nurse, working with the human resources department and other team members within the organization (e.g., physicians, management), can develop an alternative work plan to meet the needs of the individual worker. There is a need to keep the work space warm and to explore the work schedule to facilitate more frequent breaks of shorter duration.

A view from the client's perspective promotes an understanding of the scleroderma experience. Clients with scleroderma frequently experience a variety of nonspecific symptoms including fatigue; generalized weakness; and vague aching of muscles, joints, or bones. Occupational health nurses must work with clients to understand their symptoms and limitations, and to assist them in developing strategies to manage the symptoms, and thus, maintain as high a level of independence as possible. The occupational health nurse should not make assumptions because individuals differ in their symptoms and care needs. Only by asking clients about their symptoms and preferences about care can needs be met.

SUMMARY

Every individual with scleroderma is unique. Symptoms vary in severity, duration, and effect on the individual. Many with scleroderma live an unpredictable life, as they struggle to understand the condition and to manage the unpredictable symptoms. Because of rarity of the condition, many nurses know little about scleroderma, its symptoms, and how to manage them. Nurses should equip themselves with information about rare conditions and not shy away from asking individuals about their condition and how best to help them.

It is essential for occupational health nurses to appreciate the individuality of the condition and discuss workers' unique symptoms and care needs with them. Equipped with knowledge, caring for clients with scleroderma can be a rewarding experience for both the nurses and the client. In the work setting, the occupational health nurse can provide leadership to assist the individual with scleroderma to live as productive a life as possible. Nurses in all settings, hospital, community, and work, can do much to assist individuals with the management of chronic conditions such as scleroderma.

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Correction

On page 292 of the July issue of the *AAOHN Journal* (Vol. 51, No. 7), the following name in the acknowledgement section is incorrect:

Patricia Klingemann

The correct name is:

Pamela Klingemann

We apologize for any inconvenience or confusion this may have caused.

CE Answers

Tularemia: A Pathogen in Nature and A Biological Weapon

August 2002

1. D
2. B
3. A
4. A
5. C
6. B
7. D
8. B
9. A
10. C

Scleroderma: Living with Unpredictability

This issue of the AAOHN JOURNAL contains a Continuing Education Module on "Scleroderma: Living with Unpredictability." 1.1 contact hours of continuing education credit will be awarded by AAOHN upon successful completion of the posttest and evaluation.

A certificate will be awarded and the scored test will be returned when the following requirements are met by the participant: 1) The completed answer sheet is received at AAOHN on or before July 31, 2004; (2) A score of 70% (7 correct answers) is achieved by the participant; (3) The answer sheet is accompanied by a $10.00 processing fee. Expect up to 6 weeks for delivery of the certificate.

Upon completion of this lesson, the occupational health nurse will be able to:

1. List the symptoms of scleroderma.
2. Differentiate systemic (diffuse) scleroderma from local (limited) scleroderma.
3. Identify occupational health nurse interventions for mobility, mouth care, nutrition, and depression.
4. Describe workplace accommodations for the employee with scleroderma.

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Contact hour credits received for successful completion of the posttest and evaluation may be used for relicensure, certification, or re-certification.

Directions: Circle the letter of the best answer on the answer sheet provided. (Note: You may submit a photocopy for processing.)

1. All of the following are common problems associated with scleroderma except:
A. Difficulty with gross motor control.
B. Shortness of breath.
C. Difficulty swallowing.
D. Coughing.

2. Drugs associated with the development of scleroderma include:
A. Tetracycline.
B. Sertraline.
C. Cocaine.
D. Erythromycin.

3. The occupational health nurse counsels an employee recently diagnosed with scleroderma. The occupational health nurse includes which of the following in relation to symptomatology?
A. The course of the disease is predictable.
B. Symptoms vary within a person and from person to person.
C. Scleroderma usually progresses quickly.
D. Difficulty coping with heat is common.

4. A characteristic which distinguishes localized from systemic scleroderma is:
A. Early and major organ involvement of lungs, heart, gastrointestinal tract, or kidneys.
B. More rapid onset.
C. Survival rate at 10 years is 40% to 60%.
D. Average age at diagnosis is 33 years.

5. An employee with systemic scleroderma is most likely to experience which of the following?
A. Serious facial disfiguration.
B. Pursed lips.
C. Cardiac, lung, or renal failure.
D. Morning stiffness and joint pain.

6. In the acronym "CREST" for local scleroderma, the "T" stands for:
A. Telangiectasis.
B. Tight, mask-like facial skin.
C. Thick, shiny skin on arms and legs.
D. Tingling sensations in digits.

7. During the past 20 years, the survival rate for individuals with systemic scleroderma has increased largely from:
A. Vasodilator therapy.
B. Better management of renal crisis.
C. Corticosteroid therapy.
D. Earlier diagnosis of the disease.

8. When providing nutrition information for an employee with scleroderma, the occupational health nurse includes:
A. Restrict fluids because of edema.
B. Increase foods high in volume.
C. Remain upright for 4 hours after eating.
D. Remain upright during eating.

9. In the study by Angelopoulos (2001), __________% of patients with scleroderma had moderate to severe depression?
A. 5%.
B. 17%.
C. 34%.
D. 50%.

10. The occupational health nurse recommends all but which of the following for support of the employee with scleroderma?
A. Community arthritis support groups.
B. Work schedule with frequent breaks.
C. Online information sources.
D. Workspace with cool temperature.

ANSWER SHEET
Continuing Education Module
Scleroderma: Living with Unpredictability
August 2003

(Goal: To gain ideas and strategies to enhance personal and professional growth in occupational health nursing.)

Mark one answer only!
(You may submit a photocopy of the answer sheet for processing.)

1. A B C D
2. A B C D
3. A B C D
4. A B C D
5. A B C D
6. A B C D
7. A B C D
8. A B C D
9. A B C D
10. A B C D

EVALUATION (must be completed to obtain credit)

Please use the scale below to evaluate this continuing education module.

	4 - To a great extent	3 - To some extent	2 - To little extent	1 - To no extent
1. As a result of completing this module, I am able to:				
A. List the symptoms of scleroderma.	4	3	2	1
B. Differentiate systemic (diffuse) scleroderma from local (limited) scleroderma.	4	3	2	1
C. Identify occupational health nurse interventions for mobility, mouth care, nutrition, and depression.	4	3	2	1
D. Describe workplace accommodations for the employee with scleroderma.	4	3	2	1
2. The objectives were relevant to the overall goal of this independent study module.	4	3	2	1
3. The teaching/learning resources were effective for the content.	4	3	2	1
4. How much time (in minutes) was required to read this module and take the test?	50	60	70	80

Please print or type: (this information will be used to prepare your certificate of completion for the module). DEADLINE: JULY 31, 2004. Allow up to 4 weeks for processing.

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