Accordant
A CVS Caremark Company

Clinical Practice Guidelines

Scleroderma

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Scleroderma

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INTRODUCTION

The AccordantCare™ program works with health plans to assess, monitor, and support those with certain complex, chronic conditions. The focus of the program is to improve health outcomes and prevent or limit disease-related complications. AccordantCare offers unique services at no additional charge to the patient, putting them in a strong position to adhere to their treatment plan.

There are several ways AccordantCare augments physicians' efforts. Through regular telephone contact, AccordantCare nurses:

- Keep patients informed about the disease process
- Coach patients in self-motivation and self-care skills
- Encourage patients to alert their physician when new symptoms arise
- Direct patients to resources that help pay for medication, transportation, home modifications, etc.
- Ensure preventive and screening measures are accomplished
- Provide emotional support to patients and caregivers
- Screen for depression
- Find local support groups

We invite physicians to make use of the services offered by AccordantCare and to suggest ways we can further patients' treatment goals. To offer feedback, get more information, ask questions, or voice concerns, call toll-free 1-800-948-2497 to speak with a program representative from 8 a.m. to 9 p.m., Monday through Thursday, and from 8 a.m. to 5 p.m. on Friday, Eastern Time. Messages left after hours will be returned the next business day.

Intent of Guidelines

The purpose of this Clinical Practice Guideline is to describe current patterns of practice where there is no fully established national guideline for diagnosis and management. It is not meant to dictate care of patients. Decisions about care are made by the physician and the patient based on the individual needs of that patient.

A patient's health plan may or may not pay for the all medicines, tests, equipment, or services mentioned in this document. Benefits should be checked with the individual's health plan to assure payment.

DISEASE OVERVIEW

The word *scleroderma*, from the Greek *skleros* (hard) and *derma* (skin) is a word used to describe a number of minimally related clinical disorders, and the term can be quite confusing. Scleroderma is in fact not a single disease but a set of rheumatic, connective tissue diseases that share the basic symptoms of fibrosis

and inflammation. More specifically, scleroderma is marked by an abnormal build-up of tough, scarlike tissue in the skin and damage to the cells lining the walls of the blood vessels.[1] Scleroderma is also called systemic sclerosis (SSc).

Prevalence of Disease

It is estimated that 49,000 adults in the US have systemic sclerosis (SSc).[2] The estimated prevalence of SSc is 50 to 300 cases per 1 million persons.[3] Prevalence has increased in recent years because of an increased survival rate, better detection, and improved diagnostic tools. The prevalence of SSc in women between the ages of 35 years and 65 years is estimated to be as high as 400 cases per million.

Cost of Disease

Among scleroderma patients, work disability is prevalent and occurs early. Associated costs of the disease are presumed high. Correlates of disability include comorbidities, diffuse disease, disease severity, pain, fatigue, and physical function.[4]

In a study covering the period from 2003 to 2008, the average annual healthcare costs of patients with systemic sclerosis ($17, 365) were more than 3-fold higher than the average annual costs of matched controls unaffected by SSc. For these SSc patients, outpatient costs were the biggest driver (39% of yearly costs), with inpatient costs (31%) next, followed by pharmacy costs (22%).[5]

Types of Disease

Many different ways to classify scleroderma exist, but most experts agree that there are two major types: (A) localized and (B) systemic.

(A) Localized scleroderma typically affects only the skin on the hands and face and, in extreme cases, the muscle right below the skin.[6] Accordant does not enroll members with localized scleroderma, but healthcare professionals should be aware of the symptoms. Because it does not affect the internal organs, localized scleroderma is generally less destructive than systemic scleroderma, is not life-threatening, and frequently the symptoms improve or go away over time.[6] Localized scleroderma never progresses to the systemic form of the disease.[2]

(B) Systemic scleroderma (also called systemic sclerosis) is characterized by vascular, inflammatory, and fibrotic dysfunction and affects multiple organs and systems including the[7]:

- skin;
- underlying blood vessels;
- muscles and joints;
- lungs and respiratory system;
- heart and cardiovascular system;
- kidneys and genitourinary system; and

- gastrointestinal tract and digestive system.

This most dangerous form of the disease, systemic scleroderma, includes three subcategories, limited SSc, diffuse SSc, and sine sclerosis. All three forms are considered to be progressive diseases.

(B1) Limited systemic scleroderma (formerly called CREST) tends to gradually affect the skin only in certain areas: the fingers, hands, lower arms, legs, and face.[2] Chronic pain, loss of mobility, and disfigurement are possible, and esophageal and lung dysfunction can lead to serious complications. Other characteristics of the disorder include[2]:

- calcinosis—formation of calcium deposits in the connective tissues typically on the fingers, hands, face, and trunk and on the skin above the elbows and knees. When the deposits break through the skin, painful ulcers can result.
- Raynaud's phenomenon—condition in which the small blood vessels of the hands and/or feet are very sensitive and contract in response to cold or anxiety.
- esophageal dysfunction—impaired function that occurs when smooth muscles in the esophagus are scarred and lose normal movement, resulting in swallowing difficulties, chronic heartburn, inflammation, and other symptoms of gastroesophageal reflux disorder (GERD).
- sclerodactyly (also called acrosclerosis)—condition resulting from deposits of excess collagen within skin layers on the fingers causing thick, tightened, often darkened skin with hair loss.
- telangiectasia—condition caused by the swelling of tiny blood vessels, in which small red spots appear on the hands, forearms, face, lips, and tongue.

(B2) Diffuse systemic scleroderma (also called diffuse cutaneous scleroderma) is the most serious form of the disease; it typically has a rapid and severe onset with an early inflammatory phase.[6] Diffuse scleroderma typically involves more proximal areas like the upper arms and chest.[8] Diffuse SSc may overlap with other autoimmune diseases, in which case the disorder is called mixed connective tissue disease. Early diagnosis, monitoring, and treatment are essential.

(B3) Sine systemic scleroderma (or sine sclerosis) is a term used to describe systemic scleroderma without (Latin “sine”) the usual skin involvement. Many of the other complications of SSc may be present (e.g., damage to internal organs), but because the external signs are missing, the disease can be difficult to diagnose.

In addition to these classifications, many doctors recognize overlap syndrome, in which the patient experiences any of the three systemic scleroderma subsets

along with systemic lupus erythematosus, polymyositis, Sjögren's syndrome, or rheumatoid arthritis.[3] Although most patients with SSc can be classified as having limited or diffuse SSc, there is considerable overlap in the clinical features of the two variations. All patients are at risk for most of the complications.[7]

DIAGNOSIS OF DISEASE

Diagnosis of SSc is based primarily on patient history and a clinical examination. While laboratory tests can help to make a diagnosis of scleroderma or to discover another disease, no one test conclusively confirms the diagnosis. In some cases a diagnosis may require months or years while the doctor eliminates other possible causes of the symptoms.

In 2013, the American College of Rheumatology (ACR) and the European League against Rheumatism (EULAR) proposed new classification criteria for SSc.[9] The aim is to be more inclusive that previous criteria.[10] Patients with earlier disease are better represented by making use of nail fold capillary pattern and SSc-specific autoantibodies.

Classification criteria are not the same as diagnostic criteria. Classification criteria tend to be more standardized and less inclusive than physician diagnosis. They are officially used to group patients with similar clinical features together for research purposes. Nevertheless, classification criteria typically mirror diagnostic criteria. The 2013 criteria below are considered by many to be consistent with current clinical practice and feasible to use in daily clinical settings. They should allow for more patients to be classified correctly as having SSc.

The ACR-EULAR Criteria for the Classification of Systemic Sclerosis*

Items	Sub-Items	Weight/Score†
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints *(sufficient criterion)*		9
Skin thickening of the fingers† (only count the highest score)	Puffy fingers	2
	Sclerodactyly of the fingers (distal to MCP but proximal to the PIPs)	4
Finger tip lesions† (only count the highest score)	Digital tip ulcers	2
	Finger tip pitting scars	3
Telangiectasia		2
Abnormal nail fold capillaries		2

Pulmonary arterial hypertension and/or interstitial lung disease (maximum score is 2)	PAH ILD	2
Raynaud's phenomenon		3
Scleroderma related antibodies** (anti-centromere, anti-topoisomerase 1 [anti-Scl 70], anti-RNA polymerase III) **maximum score is 3	Anti-centromere Anti-topoisomerase 1 Anti-RNA polymerase III	3

* These criteria are applicable to any patient considered for inclusion in an SSc study. The criteria are not applicable to patients with skin thickening sparing the fingers or to patients who have a scleroderma-like disorder that better explains their manifestations (e.g., nephrogenic sclerosing fibrosis, generalized morphea, eosinophilic fasciitis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft-versus-host disease, and diabetic cheiroarthropathy).
† The total score is determined by adding the maximum weight (score) in each category. Patients with a total score of 9 are classified as having definite SSc.

Definitions of the SSc Classification Criteria Items

Item	Definition
Skin thickening	Skin thickening or hardening not due to scarring after injury, trauma, etc.
Puffy fingers	Swollen digits - a diffuse, usually non-pitting increase in soft tissue mass of the digits extending beyond the normal confines of the joint capsule. Normal digits are tapered distally with the tissues following the contours of the digital bone and joint structures. Swelling of the digits obliterates these contours. Not due to other reasons such as inflammatory dactylitis.
Finger tip ulcers or pitting scars	Ulcers or scars distal to or at the PIP joint not thought to be due to trauma. Digital pitting scars are depressed areas at digital tips as a result of ischemia, rather than trauma or exogenous causes.
Telangiectasia	Telangiectasia in a scleroderma like pattern are round and well demarcated and found on hands, lips, inside of the mouth, and/or large matt-like telangiectasia(e). Telangiectasia are visible macular dilated superficial blood vessels; which collapse upon pressure and fill slowly when pressure is released; distinguishable from rapidly filling spider angiomas with central arteriole and from dilated superficial vessels.
Abnormal nailfold capillary pattern consistant with SSc	Enlarged capillaries and/or capillary loss with or without peri-capillary hemorrhages at the nailfold and may be seen on the cuticle
Pulmonary arterial hypertension	Pulmonary arterial hypertension diagnosed by right heart catheterization according to standard definitions.

Interstitial lung disease	Pulmonary fibrosis on HRCT or chest radiograph, most pronounced in the basilar portions of the lungs, or presence of `Velcro' crackles on auscultation not due to another cause such as congestive heart failure.
Raynaud's phenomenon	Self report or reported by a physician with at least a two-phase color change in finger(s) and often toe(s) consisting of pallor, cyanosis and/or reactive hyperemia in response to cold exposure or emotion; usually one phase is pallor.
Scleroderma specific antibodies	Anti-centromere antibody or centromere pattern on antinuclear antibody (ANA) testing; antitopoisomerase I antibody (also known as anti-Scl70 antibody); or anti-RNA polymerase III antibody. Positive according to local laboratory standards.

APPROACH TO MANAGEMENT OF PRIMARY CONDITION

Once SSc is diagnosed, initial screening for major organ manifestations—especially interstitial lung disease, pulmonary hypertension, renal involvement, and cardiac disease—is a priority because early intervention with aggressive therapy might prevent disease progression. Involvement of these organs is associated with shorter life expectancies. Screening for risk factors for internal organ manifestations should continue on a regular basis thereafter.[11]

Currently no cure or truly disease-modifying therapy for SSc exists.[12] The goal of treatment is to improve the prognosis by reducing the development of complications. Until a disease-modifying agent is found, and in an effort to halt the progress of this serious disease, many agents or therapies are being tried that are believed to possess potential disease-modifying power.[12]

Corticosteroids have little role in the treatment of scleroderma. In fact, their use is implicated in precipitating renal crisis.[13] They are only used cautiously in low doses in carefully screened patients (e.g., patients with puffy hands, mild arthritis, or myositis).[14]

The following interventions are currently being used to treat SSc, but these agents are unproven and require further study in patients with scleroderma.[15]

Immunomodulatory Therapies

- Methotrexate (Rheumatrex®)
- Cyclophosphamide (Cytoxan®)
- Mycophenolate mofetil (CellCept®)
- Antithymocyte globulin (Atgam®, Thymoglobulin®)
- Stem cell therapy

Antifibrotic Therapies

- D-penicillamine (Cuprimine®, Depen®)

- Interferon γ (Actimmune®)

Because SSc can affect so many bodily organs, many different drugs are used to treat the multiple symptoms. The table below from the Scleroderma Foundation displays the classes of drugs most commonly used to manage the manifestations of SSc.[16]

Raynaud's Phenomenon

Calcium Channel Blockers
nifedipine (Procardia®, Adalat®)
amlodopine (Norvasc®)
isradipine (Dynacirc®)
diltiazem (Cardizem®, Dilacor XR®)
nicardipine (Cardene®)
nisoldipine (Sular®)
felodipine (Plendil®)

Angiotensin Receptor Blockers
losartan (Cozaar®)
valsartan (Diovan®)

Phosphodiesterase 5 (PDE5) inhibitors[17]
sildenafil (Revatio®)
tadalafil (Adcirca®)

Others
prazosin (Minipress®)
doxazosin (Cardura®)
pentoxifylline (Trental®)

Gastrointestinal Symptoms

Gastroesophageal Reflux Disease (GERD)

Antacids
Gaviscon, Maalox, Mylanta, Rolaids, Tums

H-2 Blockers
cimetidine (Tagamet®)
ranitidine (Zantac®)
famotidine (Pepcid®)
nizatidine (Axid®)

Proton Pump Inhibitors
omeprazole (Prilosec®)
lansoprazole (Prevacid®)

esomeprazole (Nexium®)
rabeprazole (Aciphex®)
pantoprazole (Protonix®)

Others
sucralfate (Carafate®)

Gastrointestinal Symptoms
Improve Motility

GI Stimulants
bethanecol (Urecholine®)
metoclopramide (Reglan®)
erythromycin
octreotide acetate (Sandostatin®)

Bulking Agents
calcium polycarbophil (FiberCon®, Fiberall®)
psyllium (Metamucil®)

Softening Agents
docusate calcium (Surfak®, Sulfolax®)
docusate sodium (Colace®)

Others
lactulose (Cephulac®, Cholac®)
polyethylene glycol (MiraLax®)

Gastrointestinal Symptoms
Small Intestine Dysfunction: Bacterial Overgrowth/Diarrhea

Broad Spectrum Antibiotics
tetracycline
ampicillin
metronidazole (Flagyl®)
vancomycin, ciprofloxacin (Cipro®)
amoxicillin/clavulanate (Augmentin®)
clarithromycin (Biaxin®)
azithromycin (Zithromax®)

Joint & Tendon Pain

Nonsteroidal Anti-Inflammatory Drugs (NSAIDs)
flurbiprofen (Ansaid®)
fenoprofen (Nalfon®)
nabumeton (Relafen®)

diclofenac (Voltaren®, Cataflam®)
diclofenac & misoprostol (Arthrotec®)
diclofenac sodium (Voltaren®)
etodolac (Lodine®)
ibuprofen (Motrin®, Advil®, Nuprin®)
indomethacin (Indocin®)
ketoprofen (Orudis®)
ketorolac (Toradol®)
meloxicam (Mobic®)
naproxen (Naprosyn®, Anaprox®, Aleve®)
oxaprozin (Daypro®)
piroxicam (Feldene®)
suldinac (Clinoril®)

COX-2 Inhibitors
celecoxib (Celebrex®)

Analgesics
acetaminophen (Tylenol®)
tramadol (Ultram®)

Narcotics
acetaminophen/hydrocodone (Vicodin®)
acetaminophen/oxycodone (Percocet®)
oxycodone hydrochloride (OxyContin®, OxyIR®)

Pulmonary Fibrosis/Alveolitis

Immunosuppressants
(none proven in controlled trials; the following have been used)
cyclophosphamide (Cytoxan®)
azathioprine (Imuran®)
cyclosporine (Neoral®, Sandimmune®)
mycophenolate mofetil (Cellcept®)

Pulmonary Arterial Hypertension

Patients diagnosed with pulmonary arterial hypertension should be referred to a pulmonary hypertension center for experienced evaluation and management of this serious scleroderma complication.

Endothelin Receptor Antagonists
ambrisentan (Letairis®)
bosentan (Tracleer®)
macitentan (Opsumit®)

Prostaglandin Derivatives

epoprostenol (Flolan®)
iloprost (Ventavis®) inhalation solution
trepostinil (Remodulin®)
treprostinil extended-release (Orenitram™); oral

Calcium Channel Blockers
See Raynaud's section

Phosphodiesterase Type 5 (PDE5) Inhibitor
sildenafil citrate (Revatio®)
tadalafil (Adcirca®)[18]

Soluble Guanylate Cyclase Stimulators
riociguat (Adempas®)

Scleroderma Renal Crisis

ACE Inhibitors
captopril (Capoten®)
enalapril (Vasotec®)
lisinipril (Prinivil®, Zestril®)
quinapril (Accupril®)
ramipril (Altace®)
fosinopril (Monopril®)
benazopril (Lotensin®)
moexipril (Univasc®)
trandolapril (Mavik®)

Warning About Corticosteroids and Renal Crisis
Use of corticosteroids in SSc patients can precipitate a renal crisis.[13] They should be used cautiously, in a carefully screened patient population. Anyone taking corticosteroids should monitor blood pressure frequently, and report changes to the healthcare provider immediately.

Skin Fibrosis

Immunosuppressants
(none proven in controlled trials; the following have been used)
cyclophosphamide (Cytoxan®)
cyclosporine (Neoral®, Sandimmune®)
D-penicillamine (Cuprimine®, Depen®)
methotrexate (Rheumatrex®, Trexall®)
mycophenolate mofetil (Cellcept®)

Sjögren's Syndrome

Over-the-Counter Products
Dry Mouth:
Salivart®
Biotene Oral Balance
Moisturizing Gel®
Biotene Dry Mouth Gum®
Biotene Dry Mouth Toothpaste®
Biotene Dry Mouth Mouthwash®
Optimoist®
Salix Lozenges®
Dry Eyes:
Artificial tears®

Prescription Drugs
pilocarpine hydrochloride (Salagen®)
cevimeline hydrochloride (Evoxac®)

Reactive Depression

Selective Serotonin Reuptake Inhibitors (SSRIs)
sertraline (Zoloft®)
paroxetine (Paxil®)
fluoxetine (Prozac®)
fluvoxamine (Luvox®)

Tricyclic Antidepressants
amitriptyline (Elavil®)
nortriptyline (Pamelor®)
imipramine (Tofranil®)
clomipramine (Anafranil®)

Other Treatments
bupropion (Wellbutrin®)
venlafaxine (Effexor®)
nefazodone (Serzone®)
trazadone (Desyrel®)

Skin Itching (Pruritis)/Dryness

Over-the-Counter Skin Lotions
KeriCream®
Lanalor®
Lubriderm®
Eucerin Moisturizing® creams and lotions

Nivea Moisturizing®
UltraDerm®
Alpha Keri Bath Oil®
Penederm alpha hydroxy cream®
Aveeno bath oil & moisturizer®

Antihistamines (Prescription)
diphenhydramine (Benadryl®)
hydroxyzine (Atarax®)
Colchicine

Vaccine Accordant follows the immunization recommendations and guidelines published by the Advisory Committee on Immunization Practices (ACIP) from the Centers for Disease Control and Prevention.[19] Beyond that, a more aggressive, scleroderma-specific immunization schedule has also been proposed[20]		
Inactivated/component vaccines		
Diphteria-tetanus-acellular pertussis*		
Inactivated polio vaccine	Booster every 10 years	Booster every 10 years**
Flu shot	Recommended annually	Recommended annually***
Initial pneumococcal conjugate vaccine (PCV13) followed by pneumococcal polysaccharide vaccine (PPSV23) at least 8 weeks later[19]	Routine use	Revaccination with PPSV23after 5 years
Hepatitis B	Routine use	Recommended****
Live Vaccines***		
Measles-mumps-rubella		
Varicella[21]	Routine use	Without evidence of varicella immunity: consider if treated with low-dose immunosuppression (methotrexate ≤0.4 mg/kg/week; azathioprine ≤3.0 mg/kg/day; corticosteroid <20 mg/day)
Tuberculosis (BCG)	Use if indicated	Contraindicated
Live attenuated influenza	Contraindicated	Contraindicated

vaccine		
Zoster[21]	Routine use	Consider if ≥60 years and treated with low-dose immunosuppression (methotrexate ≤0.4 mg/kg/week; azathioprine ≤3.0 mg/kg/day; corticosteroid <20 mg/day)

*Only one booster containing acellular pertussis vaccine.
**Antibody concentrations should be checked before travel to endemic areas, and a booster with the inactivated polio vaccine should be given in cases of low titers.
***Live vaccine contraindicated.
****Vaccination and/or revaccination recommended with verification of serologic response; if anti-hepatitis B (HB) levels <10 UI/L, 1-3 additional doses recommended; after successful immunization, booster doses are necessary if anti-HB levels tend to fall <10 UI/L.
*****Live vaccines are contraindicated in patients receiving immunosuppressive therapy. When indicated, the vaccine should be given at least 3 weeks and preferably 4 weeks before immunosuppressive therapy initiation or after at least 3 months off immunosuppressive therapy.

PREVENTION AND MANAGEMENT OF COMPLICATIONS

Accordant helps patients prevent and manage complications by teaching early warning signs, encouraging adherence to treatment plans, offering supportive care, and recommending physician contact where needed. The list of goals and cooperative interventions listed below does not represent a comprehensive list of complications but reflects some of the more common clinical situations specific to SSc. General health topics (e.g., age-appropriate cancer screening) are beyond the scope of this document.

Goal: Improve Self-management Skills
Cooperative interventions: Teach patients to:

- strengthen their personal motivation skills;
- develop prevention-focused, self-management skills;
- develop purposeful communication skills and maintain open, ongoing communications with their physician; and
- work with Accordant for education, information, and self-care needs.

Goal: Prevent and Manage Skin Problems
Cooperative interventions: Teach patients to[2]:

- apply oil-based lotions frequently, and especially right after bathing;
- avoid hot baths and showers;
- use sunscreen before going into sunlight;
- use humidifiers to moisten indoor air in cold climates, and clean humidifiers frequently to prevent bacterial growth;
- protect susceptible skin areas from stress or trauma;

- take all medications as prescribed; and
- inform a nurse or doctor about skin infections or ulcerations as soon as possible.

Goal: Detect and Treat Cardiac Issues Early
Cooperative interventions: Teach patients to:

- take their blood pressure regularly (as recommended by a doctor) and report elevated blood pressure as soon as possible;
- understand the risk of atherosclerosis[1] and cardiac disease[22];
- recognize the signs and symptoms of cardiac disease and report them to a nurse or doctor;
- perform regular physical activity as directed and learn to set a reasonable pace;
- understand the importance of stopping smoking;
- take all medicines as prescribed; and
- report all adverse side effects of medicines to a nurse or doctor.

Goal: Prevent and Manage Renal Dysfunction
Cooperative interventions: Teach patients to:

- understand the importance of early detection and early, aggressive treatment of high blood pressure, usually with angiotensin converting enzyme (ACE) inhibitors[3,23];
- understand the signs and symptoms of kidney dysfunction and report them to a nurse or doctor as soon as possible;
- understand the life-threatening nature of renal crisis;
- monitor their blood pressure three times a week for high-risk patients; otherwise, one to two times a week[11] (or as recommended by their doctor) and report any persistent increase to a doctor or nurse immediately;
- keep blood pressure at treatment goal by taking all medicines as prescribed;
- comply with all blood and urine tests, as ordered by their physician; and
- understand that corticosteroids can contribute to kidney dysfunction[13] and discuss the risks and benefits of corticosteroids with their doctor.

Goal: Detect and Treat Lung Involvement Early
Cooperative interventions: Teach patients to:

- understand the symptoms of pulmonary disease but know that often it is asymptomatic[24];
- immediately report to their doctor shortness of breath that cannot be explained by other causes (eg, flu, ILD), that has become a problem for the patient, or that has worsened over the last 3 months[25];
- discuss regular screening with their doctor (e.g., PFTs, diffusing capacity of the lung for carbon monoxide [DL_{CO}])[26], transthoracic Doppler echocardiography[24,27]);

- understand that pregnancy in the context of pulmonary hypertension is life-threatening;
- understand that pregnancy should be avoided using effective methods of birth control (barrier methods and surgical sterilization are preferable options if taking bosentan (Tracleer®) or ambrisentan (Latairis®);understand treatment options and potential side effects;
- comply with treatment regimen;
- understand the importance of flu and pneumonia vaccines;
- realize the importance of stopping smoking; and
- understand their disease status and plan advance directives as necessary.

Goal: Prevent and Manage Osteoporosis
Cooperative interventions: Teach patients to[28]:

- know the importance of bone density scanning (DEXA)[29];
- discuss treatment options to prevent and/or repair bone loss with a doctor;
- recognize the importance of calcium and vitamin D supplements;
- understand the importance of smoking cessation;
- learn the importance of reducing alcohol consumption;
- learn fall prevention strategies (e.g., correct vision problems, talk to doctor about medications that might affect balance)[30];
- improve home safety[31] (e.g., install grab bars in bathrooms, ensure proper lighting); and
- participate in exercise that improves balance, agility, strength, and posture (e.g., Tai Chi, weight resistance exercises[30]). Encourage members to get doctor approval of new exercise programs.

Goal: Prevent Complications Associated with Pregnancy
Cooperative interventions: Teach female patients of child-bearing age to:

- seek care at a high-risk obstetric clinic if she is pregnant or considering pregnancy[32];
- understand that limited evidence suggests that an increased number of miscarriages are associated with SSc, but the frequency of miscarriage is only slightly higher than in healthy people;
- understand that the birth of premature infants is significantly more frequent in patients with scleroderma than in healthy women, but that neonatal outcomes are generally good[32];
- understand that pregnancy in the context of renal complications, pulmonary hypertension, or rapidly developing diffuse SSc is life-threatening and should be avoided[27,32];
- understand that some treatments for pulmonary hypertension lessen the effectiveness of hormonal contraceptives and/or are contraindicated during pregnancy[27];
- understand that those with diffuse SSc run a higher risk for developing serious cardiopulmonary and renal complications[33];

- take their blood pressure regularly (as recommended by a doctor); and
- perform respiratory muscle exercises and work on general strength training.

Goal: Prevent and Manage Gastrointestinal (GI)/Nutritional Problems
GERD Cooperative interventions: Teach patients to[34]:

- understand the symptoms and medication treatments of upper and lower GI problems;
- weigh themselves monthly and report any unplanned weight loss to their doctor or Accordant nurse;
- alleviate GERD by using a wedge pillow, blocks under the head of the bed, or an electric bed;
- understand the importance of strict adherence to dosing schedules for proton pump inhibitors;
- watch for the side effects of proton pump inhibitors, including headache, dizziness, and diarrhea[16];
- eliminate GERD triggers like chocolate, alcohol, fat, coffee, and tobacco from their diet[12];
- wear loose-fitting clothing to minimize gastric pressure;
- avoid food intake for two hours before going to bed[12]; and
- eat more frequently (5 to 6 times per day) with less quantity at each feeding during the day, with the largest meal occurring around noon.

Stomach and Swallowing Cooperative interventions: Teach patients to:

- take antisecretory and promotility medicines regularly as prescribed;
- avoid certain medicines associated with causing gastroparesis;
- discuss food preferences, swallowing issues, and any unplanned weight loss[35] with a doctor, nurse, or dietician to ensure appropriate nutrition[36];avoid smoking and alcohol;
- minimize the use of NSAIDs;
- chew food thoroughly before swallowing;
- have one or more family members learn the Heimlich maneuver; and
- know what foods irritate their GI system and avoid those foods.

Small Intestine Cooperative interventions: Teach patients to:

- take broad-spectrum antibiotics as prescribed; and
- eat yogurt that contains bowel-friendly bacteria for symptomatic relief from bacterial overgrowth syndrome.

Total Parenteral Nutrition (TPN) Cooperative interventions:
Teach patients to:

- understand aseptic techniques necessary for safe home TPN;
- understand home TPN equipment and procedures;
- know the symptoms of TPN complications;
- know how to prevent complications of home TPN; and

- know when to call physician to report possible complications of home TPN.

Goal: Prevent and Manage Raynaud's phenomenon
Cooperative interventions: Teach patients to:

- understand that Raynaud's is common in SSc;
- understand that lack of preventive measures and/or proper treatment can lead to digital ulcers, which may progress to gangrene and possible amputation;
- protect skin ulcers from further injury or infection by applying nitroglycerine paste or antibiotic cream.[2]
- inform their doctor if they experience persistent cyanosis, pallor, or pain from digital ulcers[11];
- try to stay inside on cold, wet days;
- avoid putting hands in freezers or refrigerators unless wearing insulated gloves or oven mits;
- avoid drafty areas and refrigerated/frozen foods areas of supermarkets;
- avoid areas of heavy air conditioning;
- avoid swimming and other activities that expose the body to cold, wet conditions;
- stop smoking;
- avoid stress (e.g., learn and use biofeedback);
- wear warm clothing, especially over the hands and feet but also layered clothing on the trunk to minimize the contribution of reflex responses to cold central body temperature;
- avoid certain medicines that might aggravate Raynaud's, such as nonselective beta blockers like propranolol, some cold medicines, and narcotics; and

Goal: Mange the Symptoms of Sjögren's syndrome
Cooperative interventions: Teach patients to[37,38]:

- keep water available and take frequent sips of water and other fluids;
- let chips of ice dissolve in the mouth;
- use saliva substitutes, such as Biotene® Oral Balance Moisturizing Gel and Dry Mouth Liquid, especially prior to sleep to reduce the need to urinate;
- avoid sugary snacks and chew sugar-free gum or eat sugar-free candy to stimulate the flow of saliva;
- use products containing xylitol, a natural sweetener that interferes with bacterial growth and tooth decay, such as Xponent® Xylitol Gum, Carefree Koolerz®, Xylichew® Mints, and other brands;
- use alcohol-free mouthwashes, such as Biotene® Mouthwash, Natural Anticavity Fluoride Mouthwash for a Dry Mouth, and others;
- avoid caffeine, which is a mild diuretic;
- take extra care with dental hygiene and have regular dental checkups;

- practice facial exercises to help keep the mouth and face more flexible[2];
- recognize the symptoms of candidiasis and report them to a doctor or nurse;
- watch for signs of eye infections and report them to a doctor or nurse;
- watch for signs of infected parotid glands and report them to a doctor or nurse; and
- use vaginal lubricants as necessary.

PATIENT FOLLOW-UP

Frequency of appointments is based on individual patient needs and will vary according to type of SSc, level of disease activity, and medication issues. Many patients require regularly scheduled appointments that may include[39]:

- History and physical exam every three to six months to monitor disease progression;
- Laboratory testing every 3 to 12 months to monitor disease progression and complications
  - Serum chemistry
  - CBC
  - Urinalysis
  - PFTs
  - Doppler echocardiography
- Monitor for drug side effects every one to three months

PATIENT EDUCATION

The Accordant Health Communities website at:
https://www.accordant.com offers resources for patients with SSc.

Other approved and informative websites for patient education include the following:

Scleroderma Foundation
https://www.scleroderma.org

Scleroderma Research Foundation
http://www.srfcure.org

Arthritis Foundation
http://www.arthritis.org

American College of Rheumatology
http://www.rheumatology.org/index.asp

National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS)

http://www.niams.nih.gov

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