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Quality indicator set for systemic sclerosis

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Abstract

Background—Systemic sclerosis (SSc) is associated with a marked economic burden, high treatment costs and decreased productivity. Although treatment strategies for SSc can have a substantial effect on patients' outcomes, it is not known whether patients with SSc consistently receive such care. Evaluation of process-of-care quality requires specification of quality indicators (QIs), clinically detailed statements of the eligible patients and the care they should receive to achieve a minimal level of quality of care. Our objective was to develop QIs for patients with SSc.

Methods—We performed a comprehensive literature review of diagnosis and treatment of SSc and proposed QIs that were evaluated by a national Expert Panel (n=9) who were asked to review the supporting literature and individually rank the validity of each QI. These rankings formed the basis of discussion at a face-to-face meeting following the RAND/UCLA method to integrate expert opinion with literature review to identify a set of final QIs. We then presented these QIs to members of the Scleroderma Clinical Trials Consortium (SCTC).

Results—Thirty-two QIs for SSc care were judged valid by the Expert Panel. The QI set includes 9 QIs for newly diagnosed with SSc, 12 follow-up QIs for management of SSc, and 11 treatment QIs. The SCTC experts agreed with the validity of each of the 32 QI and agreed that for all but one QI the specified tests, procedures and treatments recommended in the QI were generally available.

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Disclosure of interest

Dr D. Khanna has received consultancy fees from Actelion, Gilead, United Therapeutics, MedImmune and Pfizer, and grant support from Actelion, Gilead and NIH;

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Conclusion—We have developed 32 QIs for SSc using a rigorous methodology that can be employed to evaluate and improve care for patients with SSc, as well as inform policy decisions supporting appropriate care for SSc patients.

Keywords

Systemic sclerosis; quality indicator

Introduction

Systemic sclerosis (scleroderma, SSc) is a rheumatic disease with substantial morbidity and mortality (1) and many detrimental effects on health-related quality of life. In addition, SSc is associated with a marked economic burden, with high treatment costs and decreased productivity (2). Although early treatment for SSc can have a substantial effect on patients outcomes (3-5), no studies of the quality of care provided to patients with SSc have been performed. One well-established method of evaluating the care provided for a specific condition is to develop and apply indicators of care quality. MacLean and colleagues (6) developed an Arthritis Foundation set of quality indicators (QI) to assess quality of health care in arthritis, especially for rheumatoid arthritis, osteoarthritis, and analgesic use, and QIs exist concerning gout (7), safety in rheumatologic prescribing (8), and systemic lupus erythematosus (9). However, no QIs exist for the treatment of SSc.

The quality of health care can be assessed in many ways, and is most commonly evaluated by measuring health outcomes or processes of care (6, 10). *Process* of care describes what health care providers do for patients and includes taking a health history, performing a physical examination, ordering diagnostic tests, prescribing medications, and performing procedures. We chose to develop measures of process because processes of care tend to be under the control of the health care provider or health system and are more efficiently measured than outcomes. Furthermore, performance on process measures can identify specific areas of care that are deficient and hence can be targeted for quality improvement. We chose not to develop outcome measures because clinically important outcomes in SSc may take years to develop, and may be affected by factors outside the control of the health care provider or health care system.

A process-of-care QI is a specific statement that describes care necessary to achieve a minimal level of quality of care. A QI must be measurable; clinically detailed QIs are often measured using information contained in the medical record. QIs are applicable to any physician providing care and not just limited to subspecialists providing the care. Like any other measurement, QIs will have acceptable ranges of misclassification of care (*i.e.* false positives and negatives with regard to true quality). They are thus most beneficially applied where misclassifications, if random with respect to variables of interest, will cancel each other out. In contrast, clinical guidelines are meant to guide individual clinicians in the care of individual patients. As such they describe a flexible range of diagnostic and therapeutic processes that might be considered for different groups of patients and often advocate best practices. Given their flexibility, guidelines may advocate higher performance than that required by a corresponding a QI. The indicators are not intended to replace existing guidelines, but rather to provide a means of assessing a minimum standard of care.

Methodology

Preparation of the preliminary set of Quality Indicators

A comprehensive search was performed to identify published recommendations and guidelines in SSc and SSc-specific organ involvements (process detailed in Fig. 1). We

excluded procedures for the diagnosis and management of other rheumatic diseases, even if these overlapped with SSc, localised scleroderma, or juvenile SSc. The QIs were constructed using an “IF, THEN, BECAUSE” format where “IF” defined the eligible patient for whom the care should be provided, “THEN” described the process of care that should occur, and “BECAUSE” described the relationship between the process and a clinical outcome.

Based on the literature search results and clinical experience, 69 preliminary QIs were developed. These QIs were sent to 9 international experts (2 of them were part of the Expert Panel) to provide their comments/ suggestions and to eliminate/edit/add new quality QIs. Based on their comments, 23 QIs were eliminated, leaving 46 preliminary QIs.

Comprehensive literature review

A comprehensive literature review for each of the areas covered by the QIs was performed by three members of the Steering Committee (OKB, PPK, AL). For each procedure (*e.g.* echocardiography, pulmonary function tests) or treatment, a structured literature search was performed in the PubMed database (1966 to June 2009) using predefined key words which included combination of “systemic sclerosis” OR “scleroderma” OR “CREST” and terms specific for a particular procedure (*e.g.* forced vital capacity). This search was combined with the recent systematic review of PubMed, EMBASE and Cochrane databases used for developing recommendations for treatment of SSc (11) [available through OKB]. In this systematic review, the majority of articles in the Cochrane database were also captured by PubMed and no relevant non-English article was found. Therefore, our search was limited to PubMed and articles written in English which included adult humans only. The domains/ systems and procedures/tools for which a literature search was performed are listed in Table I. To identify other potentially relevant articles, reference lists of recent reviews were examined. In addition, web pages of medical societies and international and national organisations (EULAR, EUSTAR, American College of Rheumatology, American Heart Association, American College of Cardiology, World Gastroenterology Organisation, American Gastroenterological Association, and British Thoracic Society) were screened for particular recommendations and/or guidelines that might apply. Initial selection was done based on screening of the titles and/or abstracts of the identified publications. Then, full-text articles were retrieved for detailed review. The literature search team members (PK, OKB or AL) held weekly teleconferences to reach consensus in 8 domains grouped primarily according to body systems (Table I). Each section consisted of baseline, follow-up, and treatment supportive literature review and references. QIs for osteoporosis and drug safety were excluded as they have recently been developed by the American College of Rheumatology, (ACR) (<http://www.rheumatology.org/practice/qmc/drug-safety.asp>).

RAND/ UCLA Appropriateness Panel meeting

We used the RAND/UCLA appropriateness method to quantify expert opinion regarding the proposed indicators. This well-established method allows panelists to extend the scope of the indicators where the supporting evidence is not completely clear. It structures consideration of the literature and efficiently brings all experts’ points of view to attention without forcing consensus. In other applications, it has been shown to predict future randomised controlled trials (12).

The 46 draft QIs with the supporting detailed literature review (available on request from 1st author) were sent to a national Expert Panel (5 rheumatologists, 1 cardiologist, 1 gastroenterologist, 1 general internist, and 1 pulmonologist). Each panelist spends 20–100% of their time in patient care that ranges from outpatient clinics to inpatient consults. All physicians have an interest in management of SSc but also manage other patients in their subspecialties.

Before the panel meeting, each panel member was asked to review the supporting literature and individually rank the validity each QI on a 1–9 scale (1 = completely invalid to 9 = completely valid). A QI was considered valid if 1) there was adequate scientific evidence or professional consensus to support a link between the performance of care specified by the QI and subsequent accrual of health benefit to the patient, and 2) physician or health plan performance of the care processes contained in the QI indicated higher quality care and 3) the care process in the QI was under the control of the physician or health plan. In considering the link between process and outcome, panelists were instructed to use both their clinical experience and expert guidelines as well as more rigorous published scientific evidence like randomised controlled trials and/or observational data. The 9 member panel was invited to suggest changes to the QIs and provide general comments; the panel suggested 9 additional QIs.

The expert panelists were explicitly asked to consider the process to outcome link in rating the indicators. Their judgment about the link was informed by both their clinical experience and expert guidelines as well as more rigorous published scientific evidence like randomised controlled trials, or if these were unavailable/ observational data.

A one-day face-to-face Expert Panel meeting was held in Los Angeles, CA, led by an experienced moderator (SA) to discuss the proposed 55 QIs. Each QI was considered by the group after a brief presentation of the supporting evidence by one of the literature search team members. After reviewing the first set of ratings and having a detailed discussion, the Expert Panel again rated the validity of each QI. QIs with a median rating of 7 and no statistical disagreement were accepted as final QIs for SSc. Disagreement was defined as one-third or more panelists rating the QI in the lowest tertile (1-3) and one-third or more rating the same QI in the highest tertile (7-9) (13).

Assessing agreement among other Scleroderma Experts

In order to assess whether there was agreement concerning validity and feasibility of the QIs recommended by the Expert Panel, we surveyed physicians who were US members of Scleroderma Clinical Trials Consortium (SCTC). We only included US members as QIs were developed for US healthcare although is applicable for any country. The members of SCTC include private practitioners and academic clinicians who have a special interest in SSc. However, majority of members also see patients with other rheumatic diseases. The survey asked about validity using the same 1 to 9 scale completed by the Expert Panel and also asked about the availability/feasibility of obtaining the tests/ procedures contained in the care processes in their geographic region. This group did not get the literature review. SCTC raters responded on a 1-9 Likert scale, where “1” was “totally unavailable” and “9” was “routinely available.” The survey was conducted using the internet and was sent to 35 members (all rheumatologists); 20 (57%) returned the survey.

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Results

Thirty-two of the 55 QIs were judged to be valid by the Expert Panel. Table II presents the final QIs. The QI set includes 9 baseline QIs that apply to patients with newly diagnosed SSc. There are 12 follow-up QIs for treatment of the patient with prevalent SSc and 11

treatment QIs. The QIs are further divided into general measures and by organ system. As an example, for initial assessment of cardio-pulmonary status, a resting echocardiogram with Doppler should be offered within 12 months of diagnosis since echocardiogram with Doppler screens for pulmonary arterial hypertension, diastolic dysfunction, pericardial effusion, and cardiomyopathy (QI no.2). On other hand, for a follow-up visit, echocardiogram with Doppler should be offered within 3 months of a new complaint of dyspnea on exertion and/or a new finding of a DLCO of <65% of predicted (QI no.11).

SCTC experts agreed with the validity of each of the 32 QI with median rating 7. They also rated the tests, procedures and treatments with median rating 7 with the exception of one QI (QI no.32). This QI assesses availability of treatment (*e.g.* calcium channel blockers, prostacyclin therapy, topical nitrate therapy, PDE-5 inhibitor) within 3 months of the occurrence of digital tip ulcers.

Discussion

We have developed a new set of QIs for SSc using rigorous and well-established methodology (6). These QIs address important issues in the diagnosis and management of SSc, a multi-system disease.

QIs can be used for public accountability, quality improvement, accreditation, and research (6). For example, the National Committee for Quality Assessment has adopted one of the Arthritis Foundation's QIs for rheumatoid arthritis – requiring as a minimal standard of care that rheumatoid arthritis patients followed in the outpatient setting be dispensed at least one prescription for a disease modifying anti-rheumatic drug. Physicians and health plans are rated based on the performance of these measures (14). Unlike guidelines or recommendations, QIs are minimum standards of care – they are also measurable actions. As an example, QI no.16 assesses the adherence of spirometry with DLCO in an SSc patient with new onset dyspnea. In real practice, one would not wait for 6 months to investigate new onset dyspnea, but if spirometry was not offered within 6 months this would be considered poor care. Similarly, for new onset SSc, one would not wait for 12 months to initiate work for internal organ involvement but if these are not offered within 12 months, this would be considered suboptimum care. Commonly, clinically-detailed QIs are abstracted on chart review by independent auditors and are presented as the proportion of eligible patients who received the recommended QI at the level of the physician or health plan. This translates into documentation of pertinent history (such as ability to perform ADLs), discussion of adverse events due to medications, or refusal of a recommended procedure or treatment by the patient. For example, in a single-centre study to assess adherence of QIs for RA and drug safety endorsed by the American College of Rheumatology (ACR), 99% of patients with RA were receiving disease-modifying agents (DMARDs), but discussion of potential risks for a new DMARD or glucocorticoids was documented in only 35% of patient records (15). This finding is explained by the requirement to document an action. Although rheumatologists and their staff members routinely counsel patients concerning the risks associated with these therapies and many patients are provided with relevant pamphlets from the Arthritis Foundation, adherence to QIs is based strictly on medical record documentation.

Quality measurement with explicit QIs can be used as a trigger for quality improvement. If performance for a QI is low, this can stimulate a search for the source of the deficit in care, including provider factors, resource constraints or other barriers to access. For example, one of the QIs recommends that a patient with newly diagnosed SSc have a baseline echocardiogram with Doppler performed. Patients with SSc have a high prevalence of pulmonary hypertension (3, 16). The American College of Chest Physicians (ACCP) guidelines recommend baseline echocardiogram with Doppler in high-risk populations (17).

However, in a prospective study of 669 patients with SSc and “mixed connective tissue disease” in community rheumatology practices, only 27% had ever had an evaluation for PH with Doppler echocardiogram. Since ordering an echocardiogram with Doppler is a measurable action, it is hoped that the adherence to this QI in newly diagnosed SSc will lead to earlier diagnosis and treatment of cardiopulmonary involvement in SSc and thereby improve health outcomes in these patients. Poor scores on QIs should lead to clinical reminders or the use of other informatic tools (available at Veterans Affairs hospitals), the development of clinical registries and provider education, payment guidelines and other initiatives.

During the development of QIs, it was decided not to divide patients into limited and diffuse subtypes since this condition involves skin examination. Although skin examination (and skin score) is routinely done in scleroderma centers, it is rarely performed in private rheumatology practices. In addition, skin tends to soften over time and patients may have normal texture of skin later in their disease course. Furthermore, since QIs are generalisable to any physician, it was considered that distinguishing between limited *vs.* diffuse SSc may not be feasible in clinical practice. Therefore, QI no.9 states that patients with early systemic sclerosis (<5 years from first signs or symptoms), should be counseled to perform at least weekly blood pressure measurements. We also want to emphasise that QIs are meant to be achieved by a single physician. In other words, if any physician (not necessarily the one seeing the patient at the time) involved in the care of a patient performs Doppler echocardiogram at baseline visit (no.2) or serum haemoglobin annually (no.11), then the QIs are met for the particular patient.

The EULAR/EUSTAR have recently made 14 recommendations for the treatment of SSc involving 6 organ systems (18). Our panel proposed 6 QIs that are similar to these recommendations. These belong to the gastrointestinal tract (n=3), digital ulcers, cardiopulmonary, pulmonary, and renal systems (n=1 each). The difference is that our QIs are measurable. As an example, EULAR/EUSTAR stated that “In view of the results from two high-quality RCTs and despite its known toxicity, cyclophosphamide should be considered for treatment of SSc-ILD”, whereas our QI (no.30) provides measurable parameters: “IF a patient has systemic sclerosis-associated interstitial lung disease and documented a >10% decline in FVC during the past 12 months, THEN immunosuppressive treatment (*e.g.* cyclophosphamide, methotrexate, azathioprine, cyclosporine, mycophenolate mofetil) should be offered within 3 months.”

Certain recommendations from the panelists (such as QI no.1 to test for SSc-specific autoantibodies) concerned us because of questions regarding validity and availability of these tests/ procedures. Therefore, we took the unusual step of externally validating the QI set by presenting them to a separate set of SSc clinical experts using a Delphi exercise performed by US members of the Scleroderma Clinical Trial Consortium (SCTC). The members of the SCTC include academic and practicing rheumatologists with an interest in SSc. Among the rheumatologists who responded, the median ratings were very similar to the ratings of the Expert Panel, thus providing strong external validity for the QIs. In addition, we asked the SCTC investigators about the availability of tests/ procedures in their local area or whether their health plan would cover the costs of the proposed QIs. We felt that this inquiry was important in determining if practicing physicians would have access to the proposed tests and procedures. The median ratings availability was 7 (agreement that the tests were routinely available) for all QIs except 1 (QI no.32). This QI, assessing the availability of therapies for digital ulcers, received a median score of 6. This result is probably because expensive therapies (prostacyclin analogues, PDE-5 inhibitors) proposed in this QI are not yet approved for the treatment of digital ulcers and thus are not widely available.

In summary, we developed QIs for SSc using rigorous well-established methodologies. The users of the indicator set are free to include or exclude those indicators at their discretion. It is our hope that these QIs will be employed to improve care and in turn improve health outcomes in patients with SSc, as well as inform policy decisions supporting appropriate care for SSc patients.

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Key messages

- We have developed quality indicators for systemic sclerosis that can be employed to evaluate and improve care for patients with SSc
- Quality indicators can also be used to inform policy decisions supporting appropriate care for SSc patients

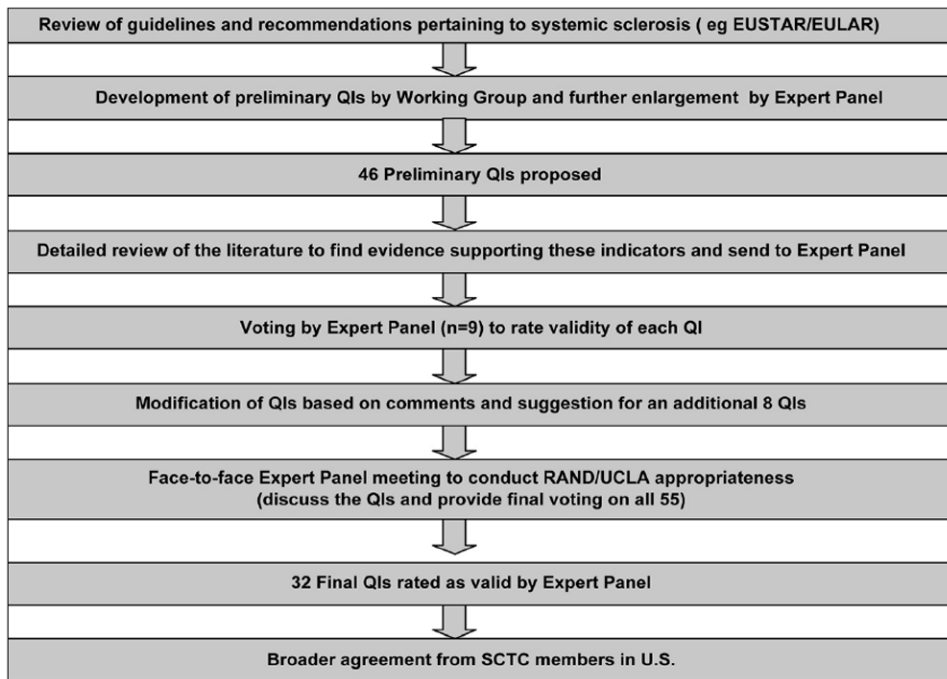


Fig. 1.
Methodology used to develop the SSc QI set.

Table I

Domain/Organ System	Tools/procedures
I Cardiopulmonary	<ul style="list-style-type: none"> • Echocardiography with Doppler • Six minute walk test • Right heart catheterisation • Laboratory markers (BNP, pro-BNP) • Measures of dyspnea • Electrocardiogram • Blood pressure • Treatment
II Pulmonary	<ul style="list-style-type: none"> • Spirometry and diffusing capacity • Chest radiograph • High-resolution computed tomography (HRCT) of lungs • Treatment
III Gastrointestinal	<ul style="list-style-type: none"> • Weight and Body mass index (BMI) • Laboratory markers (serum albumin, etc.) • Test for gastroparesis • Test for esophageal dysmotility • Test for malabsorption • Treatment
IV Renal	<ul style="list-style-type: none"> • Blood pressure • Laboratory markers (serum creatinine, creatinine clearance, urine protein, etc.) • Treatment
V Musculoskeletal	<ul style="list-style-type: none"> • Assessment of muscle weakness on physical exam • Measure of joint involvement (<i>e.g.</i> number of tender joints) • Laboratory markers (serum creatine phosphokinase) • Treatment
VI Cutaneous	<ul style="list-style-type: none"> • Physical exam to determine skin involvement • Treatment
VII Health-Related Quality of Life	<ul style="list-style-type: none"> • Measure of function
VIII Serologies	<ul style="list-style-type: none"> • Test for anti-topoisomerase I, anti centomere and anti-RNA polymerase III antibodies
IX Prevention and Drug Monitoring	

Table II

Quality indicators for systemic sclerosis.

BASELINE	
General	
1.	IF a patient has newly diagnosed systemic sclerosis, THEN anti-topoisomerase I, anti-centromere, and anti-RNA-polymerase III antibody tests should be offered* within 12 months of diagnosis BECAUSE these tests can help determine prognosis.
Cardio-Pulmonary	
2	IF a patient has newly diagnosed systemic sclerosis, THEN a resting echocardiogram with Doppler should be offered within 12 months of diagnosis BECAUSE this screens for pulmonary arterial hypertension, diastolic dysfunction, pericardial effusion, and cardiomyopathy.
Physical Function	
3	IF a patient has newly diagnosed systemic sclerosis, THEN the medical record should document a measure of functional status (<i>e.g.</i> activities of daily living, health assessment questionnaire-disability index or self-report) within 12 months of diagnosis BECAUSE scores on these measures predict morbidity and mortality associated with SSc.
Musculoskeletal	
4	IF a patient has newly diagnosed systemic sclerosis, THEN a serum creatine phosphokinase (CPK) should be offered within 12 months of diagnosis BECAUSE this test helps define the presence of muscular involvement.
5	IF a patient has newly diagnosed systemic sclerosis and has one or more palpable tendon friction rubs, THEN a follow-up visit should be offered within 3 months BECAUSE tendon friction rubs indicate active disease and predict increasing skin thickness and new internal organ involvement during subsequent months.
Pulmonary	
6	IF a patient has newly diagnosed systemic sclerosis, THEN spirometry and diffusion capacity should be offered within 12 months of diagnosis BECAUSE these tests can identify and define the patient's degree of pulmonary involvement and also define treatment.
7	IF a patient has newly diagnosed systemic sclerosis and has a FVC or DLCO below 80% of predicted THEN a high-resolution computed tomography scan of the lung should be offered within 12 months of diagnosis BECAUSE this can define the presence of pulmonary fibrosis.
Renal	
8	IF a patient has newly diagnosed has systemic sclerosis, THEN a serum creatinine should be offered within 6 months of diagnosis BECAUSE it can help define the presence and degree of renal involvement by systemic sclerosis.
9	IF a patient has early systemic sclerosis (<5 years from first signs or symptoms), THEN the medical record should document counseling to perform at least weekly blood pressure measurements BECAUSE blood pressure self monitoring can lead to early detection of scleroderma renal crisis. This in turn leads to change in therapy.
FOLLOW-UP	
General	
10	IF a patient has systemic sclerosis, THEN a haemoglobin test should be offered at least annually BECAUSE anemia is associated with increased mortality and it may result in a change in therapy.
Cardio-Pulmonary	
11	IF a patient has systemic sclerosis and complains of new dyspnea on exertion and/or has new DLCO of <65% of predicted, THEN an echocardiogram with Doppler should be offered within 3 months BECAUSE echocardiography can help define the degree of pulmonary or cardiovascular involvement (including diastolic dysfunction).
Gastrointestinal	
12	IF a patient has systemic sclerosis, THEN the medical record should document weight or body mass index at least annually BECAUSE this can help define general nutrition and can be used as a measure to follow response to therapy for malabsorption.
13	IF a patient has systemic sclerosis, THEN the medical record should document the presence or absence of symptoms of gastro-esophageal reflux disease (GERD) (<i>e.g.</i> heartburn, nocturnal cough, dysphonia, acid taste, chest pain) at least annually BECAUSE these symptoms can help define upper gastrointestinal involvement and therapy.
Musculoskeletal	

- 14** IF a patient with systemic sclerosis has proximal muscle weakness on examination and a CPK level 3 upper limit of normal, THEN an electromyogram (EMG), muscle biopsy or magnetic resonance imaging (MRI) should be offered because an appropriate diagnosis will help define treatment.

Pulmonary

- 15** IF a patient has early systemic sclerosis (<5 years from first signs or symptoms), THEN spirometry and DLCO should be offered at least annually for the first 5 years BECAUSE this can help detect decline in lung function to identify patients with progressive interstitial lung disease.
- 16** IF a patient has systemic sclerosis and new onset dyspnea on exertion, THEN spirometry with DLCO should be offered within 6 months BECAUSE presence of dyspnea and spirometry with diffusion capacity can help define the degree of pulmonary or cardiovascular involvement.
- 17** IF a patient has systemic sclerosis and is diagnosed with interstitial lung disease (defined by chest x-ray, HRCT of the chest, or spirometry), THEN spirometry and diffusion capacity should be offered at least every 12 months until stabilisation of the FVC (within 10% over 1 year) BECAUSE these tests can predict decline lung function in interstitial lung disease and response to therapy.
- 18** IF a patient has systemic sclerosis and complains of new onset dyspnea on exertion or has a newly abnormal FVC or DLCO <80% of predicted, THEN HRCT of lungs should be offered within 6 months BECAUSE HRCT of the lungs can help define the degree of pulmonary or cardiovascular involvement. This, in turn, can help define the treatment.
- 19** IF a patient has systemic sclerosis with new onset dyspnea on exertion and a resting echocardiogram with Doppler suggestive of new pulmonary hypertension (estimated right ventricular systolic pressure >50 mm Hg or tricuspid regurgitation velocity >3.5 mm/sec), THEN she/he should be referred for consideration of right heart catheterisation within 3 months BECAUSE right heart catheterisation is the definitive test for the diagnosis of pulmonary arterial hypertension.

Renal

- 20** IF a patient has systemic sclerosis, THEN the medical record should document a blood pressure measurement at every clinic visit BECAUSE it can help define the presence and degree of renal involvement by systemic sclerosis. This, in turn, can lead to a change in therapy.
- 21** IF the patient has systemic sclerosis and new onset hypertension (systolic BP> 140 or diastolic BP> 90 mmHg confirmed on 2 separate occasions), THEN serum creatinine, CBC with platelets, and urinalysis should be offered within 72 hours BECAUSE they can identify renal dysfunction. This, in turn leads to change in therapy.

TREATMENT

General

- 22** IF a patient has systemic sclerosis, THEN annual inactive influenza vaccine should be offered unless contraindications are documented, BECAUSE this can prevent or decrease the severity of influenza infection.
- 23** IF a patient has systemic sclerosis, THEN pneumococcal vaccine should be offered every 5 years unless contraindications are documented, BECAUSE this can prevent or decrease the severity of pneumococcal infection.

Cardiac

- 24** IF a patient has systemic sclerosis and is diagnosed with clinical symptoms of diastolic dysfunction and symptomatic heart failure, THEN a treatment (*e.g.* ACE inhibitor, diuretic, beta-blocker) or a referral to a cardiologist should be offered within 3 months BECAUSE treatment can improve morbidity associated with diastolic dysfunction.

Cardio-Pulmonary

- 25** IF a patient has systemic sclerosis and has NYHA/WHO functional class II-IV due to pulmonary arterial hypertension diagnosed by right heart catheterisation (RHC) *, THEN treatment (endothelin blockers, prostacyclin analogs and /or PDE-5 inhibitors) should be initiated within 3 months BECAUSE these therapies improve morbidity associated with pulmonary hypertension.

Gastrointestinal

- 26** IF a patient has systemic sclerosis and is diagnosed with GERD, THEN anti-acid therapy with a proton pump inhibitor (PPI) or H2 blocker should be offered within 3 months of the GERD diagnosis BECAUSE these therapies can improve symptoms and quality of life and decrease long-term complications associated with GERD.
- 27** IF a patient has systemic sclerosis and has symptoms of early satiety, post-prandial abdominal bloating, post-prandial vomiting or regurgitation for at least 1 month, THEN a test for impaired gastric emptying (*e.g.* upper endoscopy, gastric emptying study, upper GI series) or an empiric trial of therapy (*e.g.* prokinetics, PPI) should be offered within 6 months BECAUSE this can lead to earlier diagnosis and initiation of treatment which can improve symptoms and quality of life.
- 28** IF a patient has systemic sclerosis and unintentional weight loss (< 5%) over 3 months with symptoms of nausea or vomiting, bloating, or diarrhoea for 4 weeks, THEN a test for malabsorption or bacterial overgrowth (*e.g.* lactulose breath test, glucose breath test, xylose test, jejunal culture, serum carotene, faecal fat determination) or

an empiric trial of therapy (*e.g.* antibiotics, prokinetics, octreotide) should be offered within 3 months BECAUSE malabsorption can lead to malnutrition and can be treated.

Musculoskeletal

- 29** IF a patient has early systemic sclerosis (<5 years from first signs or symptoms) and presents with decreased range of motion or function of the hands, THEN a range of motion exercise program should be offered within 6 months BECAUSE it may improve hand joint range of motion or hand function.

Pulmonary

- 30** IF a patient has systemic sclerosis-associated interstitial lung disease and documented a >10% decline in FVC during the past 12 months, THEN immunosuppressive treatment (*e.g.* cyclophosphamide, methotrexate, azathioprine, cyclosporine, mycophenolate mofetil) should be offered within 3 months BECAUSE this therapy improves lung function and quality of life associated with scleroderma lung disease.

Renal

- 31** IF a patient with systemic sclerosis presents with scleroderma renal crisis (defined as accelerated arterial hypertension [at least SBP 140 and a rise of SBP 30mmHg from baseline] or rapidly progressive renal failure), THEN s/he should be prescribed an ACE inhibitor within 72 hours BECAUSE ACE inhibitors improve survival.

Peripheral Vascular

- 32** IF a patient with systemic sclerosis has digital tip ulcer(s), THEN treatment (*e.g.* calcium channel blockers, prostacyclin therapy, topical nitrate therapy, PDE-5 inhibitor) should be prescribed within 3 months of diagnosis BECAUSE treatment improves healing of digital ulcers and hand function.

* Offered: offered or performed or reason for non-performance documented

Note: The care process in a quality indicator is considered to have been passed if the care is documented in the medical record or if the care is recommended, even if it is refused by the patient.

ACE: Angiotensin converting enzyme; CPK: creatine phosphokinase; DLCO: diffusion capacity for carbon monoxide; FVC: forced vital capacity; NYHA/WHO: New York Heart Association / World Health Organisation; PDE-5: Phosphodiesterase-5