A. Goals

The goals of the examination are to:

1. Assess whether the participant has signs of an undiagnosed connective tissue disease (CTD), other than SS.

2. Catalog the signs of CTD in patients who ultimately may be diagnosed with primary or secondary SS.

3. Provide disease markers to help identify subsets of patients for diagnostic referral or future studies of extraglandular components of SS such as thyroid, liver or kidney diseases, or lymphoma.

B. Definitions

**Graves’ ophthalmopathy**
Evidence of exophthalmos (proptosis), lid retraction, or lid lag (eyelids lag behind the globe during downward or upward gaze).

**Scleral icterus**
Bilateral diffuse yellow discoloration of the scleral conjunctivae.

**Enlargement of thyroid gland**
On palpation: unilateral or bilateral enlargement of the thyroid gland; single or multiple nodules.

**Synovitis**
Swelling, thickening, or tenderness in the tissues surrounding the proximal interphalangeal (PIP), metacarpophalangeal (MCP), wrist, or elbow joints.

**Bouchard and Heberden nodes**
On examination of the hand joints, presence of bony enlargement at the PIP joint (Bouchard nodes) or DIP joint (Heberden nodes) would be indicative of nodal osteoarthritis.

**Sclerodactyly**
Loss of normal mobility of skin (with or without atrophy of soft tissues) of the fingers.
**Dactylitis**
Diffuse swelling in the joints of the digits, giving the appearance of a sausage.

**Raynaud’s phenomenon**
Prolonged blanching of one or more digits.

**Dilated capillary loops**
Erythematous blush or visible capillaries in and around the nail folds of the fingers.

**Nail fold infarcts**
Fixed punctate erythema, ulceration, or eschar in and around the nail folds of the fingers.

**Rheumatoid nodules**
Fixed or mobile nodular swellings in the subcutaneous or periosteal areas.

**Hepatomegaly**
Screen for hepatomegaly by palpating the right upper quadrant of the abdomen in the midclavicular line. If the liver is palpable on deep inspiration, percuss its span from upper to lower border. A span of 8cm or greater constitutes hepatomegaly.

**Splenomegaly**
A palpable spleen tip under the left costal margin on deep inspiration constitutes splenomegaly.

**Lymphadenopathy**
Palpable lymph nodes greater than 1cm in diameter.

**Malar rash**
Fixed erythema, flat or raised, centered over the malar eminence and sparing the nasolabial fold. Tends to extend over the bridge of the nose.

**Discoid lesions**
 Raised erythematous patches with adherent keratotic scale, telangiectasia, scarring, and follicular plugging. Hyperpigmentation or depigmentation may occur.

**Mat telangiectasias**
Blanching, square-appearing clusters of dilated vessels.

**Psoriasis:**
Erythematous, well demarcated, raised plaques with silver scale. Common affected areas are the elbows, knees and scalp.
C. Conditions Requiring Additional Evaluation

If the examining physician detects an abnormality that requires additional clinical and laboratory evaluation, the patient should be referred to his or her primary health care provider for further diagnosis and treatment. All SICCA evaluations planned for the patient on that day should be completed, but continuation in the Registry would depend on the results of the subsequent diagnosis and treatment.

D. Autoimmune or Interstitial lung disease”

1. Given that lung disease is very common, this section requires the examiner to ask the patient questions to determine if they have a specific autoimmune or interstitial lung disease such as NSIP (non-specific interstitial pneumonia) or UIP (usual interstitial pneumonia). It may be helpful to ask if they have ever had an abnormal breathing test, chest x-ray or CT scan. If dry crackles are heard on lung auscultation with a stethoscope, there may be evidence for an interstitial lung disease.

2. If the patient states that they have been diagnosed by a physician with an autoimmune or interstitial lung disease, a physician confirmation of diagnosis form must be completed and sent to the diagnosing physician for confirmation and details of this diagnosis. Please refer to the end of this chapter for lung disease confirmation form SOP for details.
Autoimmune or Interstitial Lung Disease Diagnosis Confirmation
STANDARD OPERATING PROCEDURE (SOP):

A. When to Obtain Diagnosis Confirmation

When completing the rheumatologic exam form, if a respondent reports that he/she has been given a diagnosis of autoimmune or interstitial lung disease by a physician (Question 23), a diagnosis confirmation must be obtained from the respondent’s physician.

B. How to Proceed

Obtain from the respondent all contact information (name of diagnosing physician(s) phone and fax numbers; address if no fax number is available. Use this information to complete the top portion of the Physician Confirmation of Diagnosis form (with participant name and physician contact information). Have the subject sign the Authorization for Release of Medical Record/Medical Record Information. Explain that we need to confirm the diagnosis of an autoimmune or interstitial lung disease with their physician.

At the earliest opportunity, complete a Diagnosis Confirmation Fax Cover form fax the cover form, Physician Confirmation of Diagnosis form(s) and signed Authorization for Release of Records form to the diagnosing physician(s).

C. Following up with the Diagnosing Physician’s Office

If the completed Physician Confirmation of Diagnosis form is not returned by the physician’s office two weeks after sending the initial fax, contact the physician’s office to follow up. Try to establish an appropriate person at the physician’s office to consistently follow up with. Continue follow up calls on a monthly basis until the Physician Confirmation of Diagnosis form is returned. If the form has not been returned after six months and there seems to be little hope of it being completed, mark “unable to obtain” on the Baseline Systemic Diagnosis Form (BSD) next to item 10. Initial, sign and date the BSD. Mark the box for the BSD form on the participant’s Baseline Data Checklist (BD) and initial and date next to the box. Fax both the updated BD and the BSD to SICCA Data Coordinating Center.

D. Transcribing information onto the Baseline Systemic Diagnosis Confirmation Form

When the completed Physician Confirmation of Diagnosis form is returned, the information on the form must be transcribed onto the Baseline Systemic Diagnosis Confirmation form (BSD). (The BSD is a Case Report Form while the Physician Confirmation of Diagnosis form is not.) Mark the “yes” box on the BSD for any disease the physician has checked on the Physician Confirmation of Diagnosis form. On the
BSD mark whether the diagnosing physician is a GP (General Practitioner) or a Specialist. (For more detailed information on completing the BSD see the BSD Question by Question document under Chapter 15 of the Manual of Procedures.) If you have questions about the information the physician has included on the Physician Confirmation of Diagnosis Form, consult the appropriate SICCA clinician or Research Director at your site.)

Enter your initials at the bottom of the BSD and sign and date the form. Update the Baseline Data Checklist (BD) by marking the box for the BSD form and initial and date next to the box. Fax both the completed BSD and the updated BD to the SICCA Data Coordinating Center at datafax@df1.ucsf.edu.

Do NOT fax the Physician Confirmation of Diagnosis form to the SICCA Data Coordinating Center. This form should be filed with the participant’s signed Informed Consent Form in a locked file cabinet separate from all completed SICCA case report forms for the participant.