

SF-DCT INFORMATION FOR PRIMARY SJOGREN'S SYNDROME (PSS) CLAIMS

OPTION 1

(PSS claims are not eligible for Disease Option 2)

Primary Sjogren's Syndrome (PSS)

Sjogren's Syndrome (SHOW-grins) is an autoimmune disease characterized by inflammation in certain glands of the body:

- Inflammation of the glands that produce tears (lacrimal glands) leads to decreased water production for tears and eye dryness. The resulting eye dryness can progressively lead to eye irritation, decreased tear production, “gritty” sensation in the eyes, infection, and serious abrasion of the dome of the eye (cornea).
- Inflammation of the glands that produce the saliva in the mouth (salivary glands, including the parotid glands) leads to mouth dryness, swallowing difficulties, dental decay, gum disease, mouth sores and swelling, stones and/or infection of parotid gland inside of the cheeks.

Not all of the symptoms listed above are eligible symptoms for PSS in the Plan. Read the Disease Claimant Information Guide and this document carefully to make sure that you document eligible symptoms.

Primary Sjogren's Syndrome (PSS) vs. Secondary Sjogren's Syndrome

- **Primary Sjogren's Syndrome** is when the Sjogren's syndrome occurs without an accompanying inflammatory arthritis. PSS is an eligible condition in the Plan in Disease Option 1.
- **Secondary Sjogren's Syndrome** is when the Sjogren's syndrome is accompanied with connective tissue disease (arthritis). Secondary Sjogren's Syndrome is not an eligible condition in the Plan.

Checklist of documents to submit for a PSS claim:

- ❑ An evaluation by a Qualified Medical Doctor (QMD) (see page 5)
- or
- ❑ Medical records supporting the symptoms of PSS (see pages 6-16)
- AND
- ❑ Documentation of the severity level of the disease (see pages 20-28)

Acceptable QMD Certification for PSS

One way to qualify for PSS is to submit a QMD statement or diagnosis. A QMD is a physician who writes a letter for purposes of the settlement and is/or became board certified in one or more of the following specialties before he or she wrote the letter:

1. Internal Medicine; or
2. Rheumatology; or
3. Allergy/Immunology; or
4. Doctor of Osteopathy with similar specialty certifications; or
5. Foreign doctor with equivalent specialty certification.

A physician with a status of “Board Eligible” does not qualify as a QMD. A physician can be Board Certified in more than one of these specialties. A QMD may also be your treating physician.

What are the eligible symptoms of PSS in the Plan?

In order to be approved for PSS, the file must contain ALL 3 symptoms below:

1. Dry Eyes confirmed by either an abnormal Rose Bengal, Fluorescein stain, or Schirmer's test (see pages 8-10); and
2. Dry Mouth confirmed by a decreased parotid flow rate, or by an abnormal biopsy of the salivary glands (see page12); and
3. One of the following Autoantibodies listed on (see page14):
 - a. elevated Rheumatoid Factor (RF) or
 - b. elevated Antinuclear antibody (ANA) or
 - c. the presence of Anti-SS-A (RO) antibodies or
 - d. the presence Anti-SS-B (LA) antibodies.

SYMPTOM 1 – DRY EYES

The medical records must document an abnormal result from **ONE** of the following tests listed below:

- Rose Bengal Staining (see page 8); **or**
- Fluorescein Staining (see page 9); **or**
- Schirmer's Test (see page 10)

DRY EYES: acceptable records for Rose Bengal Staining

Rose Bengal Staining – Rose Bengal is a dye that, when applied to the cornea and conjunctiva of the eye, is taken up by sick epithelial cells. An abnormal result is noted as a “Positive” Rose Bengal test.

- The test can be performed by any medical doctor but is generally performed by an Ophthalmologist. The records must state or show that the staining was performed on both the cornea and conjunctiva of the eye.

DRY EYES – acceptable records for Fluorescein Staining

Fluorescein Staining – This test uses orange dye (Fluorescein) and a blue light to detect foreign bodies in the eye. A piece of blotting paper containing the dye is touched to the surface of the eye, and you will be asked to blink. Blinking spreads the dye around and coats the “tear film” covering the surface of the cornea. A blue light is then directed at your eye. Any problems on the surface of the cornea will be stained by the dye and appear green under the blue light. If the test is normal, the dye remains in the tear film on the surface of the eye and does not adhere to the eye itself. The test is also known as the Tear Break-Up Time (TBUT).

- The test can be performed by any medical doctor but is generally performed by an ophthalmologist. The records must state or show that the staining was performed on both the cornea and conjunctiva of the eye.

DRY EYES – acceptable records for Schirmer's Test

Schirmer's Test – This test involves placing a thin tear strip (paper) inside the lower eyelid for 5 minutes. The tear strip is then removed and the length of the strip that is wet from tears is measured and compared to a standard. Individuals with dry eyes will have less wetting of the tear strip than normal. For this symptom to be credited by the SF-DCT, the Schirmer's Test must show a result of less than 10 millimeters of wetting in 5 minutes.

- The test can be performed by any medical doctor but is generally performed by an ophthalmologist. The test results must include the millimeters of wetting and the time frame in minutes.

DRY EYES – unacceptable records

Common reasons why claimants receive a deficiency notice about the symptom of dry eyes:

- The file does not contain an acceptable test
- The file reflects a complaint of dry eyes only
- The Rose Bengal and/or the Fluorescein staining is not performed on both the cornea and the conjunctiva

SYMPTOM 2: DRY MOUTH, acceptable records

Your records must document an abnormal result from ONE of the following tests:

- Decreased parotid flow rate using Lashley cups or any other method that allows objective measurement of saliva production including a cotton ball test, salometry test or the lemon drop test; or
- Biopsy of a minor salivary gland. Minor salivary glands are located in the lips, tongue and cheek. The most common type of minor gland biopsy is the removal of a small piece of lip tissue. This is a minor, outpatient procedure using local anesthesia of the lip; or
- The pathology report must reflect a focus score of >2 (greater than 2) based on at least 4 evaluable lobules.

DRY EYES – unacceptable records

Common reasons why claimants receive a deficiency notice about the symptom of dry mouth:

- The biopsy report does not reflect at least 4 lobules
- The file does not contain any of the required tests
- The file reflects a complaint of dry mouth only

SYMPTOM 3 – AUTOANTIBODIES, acceptable records

Your records must document elevated or positive results from one of the following blood tests listed below:

1. An elevated Rheumatoid Factor (RF) > 1:160; or
2. Elevated Antinuclear Antibody (ANA) > 1:160; or
3. Positive Anti-SS-A (RO) antibodies. (Note: the test must show the presence of the antibody.); or
4. Positive Anti-SS-B (LA) antibodies. (Note: the test must show the presence of the antibody.)

Autoantibodies – unacceptable records

Common reasons why claimants receive a deficiency notice about the symptom of Autoantibodies:

- The file does not contain any of the required laboratory tests
- The ANA and Rheumatoid factor results are less than 1:160
- The file does not reflect a positive Anti-SSA or Anti-SSB antibody

Reasons for deficiencies in PSS claims:

If a physician states that a symptom is caused by a source other than PSS, then that symptom will not be credited or considered eligible by the SF-DCT.

For example, if your DRY EYES and/or DRY MOUTH are related to any of the following conditions, then the SF-DCT will not credit the symptom of DRY EYES and/or DRY MOUTH:

- Sarcoidosis
- Pre-existing Lymphoma
- Acquired Immunodeficiency Disease
- Neuropathies
- Tumors
- Tuberculosis
- Excessive alcohol consumption
- Dry eyes or dry mouth that is a side effect of certain medications
- Infections
- Allergic reactions

If your claim does not qualify for PSS, the SF-DCT will review your claim for “ACTD”.

If your medical records document clinical symptoms or laboratory findings atypical of PSS, the SF-DCT will review the claim for ACTD.

If the symptoms more closely resemble Atypical Connective Tissue Disease (ACTD), then the SF-DCT will review and possibly approve the claim in ACTD, and not PSS. (1)

The compensation amounts for approved PSS claims and ACTD claims are the same. Compensation amounts in Disease Option 1 are not based on the disease or condition that a claimant is approved for; it is based solely on the severity of the disease or the claimant’s level of disability.

(1) The eligible symptoms for ACTD are listed in the Disease Claimant Information Guide at Tab 1.

What are the levels of compensation for PSS?

- Severity or Disability Level A – \$50,000 – Death or Total Disability
- Severity Level B – \$20,000 – PSS with associated central nervous system or severe cardiopulmonary involvement or PSS with pseudo-lymphoma or associated lymphoma
- Severity Level C – \$10,000 – All other PSS claims

If Premium Payments are approved by the District Court, approved PSS claimants could receive an additional payment of up to 20% of their approved compensation amount.

The compensation amounts for approved PSS and ACTD claims are the same. Compensation amounts in Disease Option 1 are not based on the disease or condition that a claimant is approved for; it is based solely on the severity of the disease or the claimant's level of disability.

Level A – Death, \$50,000 (U.S.)

To qualify for Level A based on a claimant's death, you must submit ONE of the following:

- A death certificate that indicates that the primary or secondary cause of death is PSS or one of the approved symptoms of PSS; or
- An autopsy report that indicates that the cause of death is related to PSS; or
- A letter from a QMD or the claimant's medical records that directly relate the primary or secondary cause of death to PSS or one of the approved symptoms.

Level A can be approved without a death certificate or autopsy report.

The claimant's death cannot be caused by any other disease or condition.

Functional Disability Level A Claims

- The Claims Resolution Procedures document defines Disability A as: "Death or total disability resulting from the compensable condition. An individual will be considered totally disabled if she demonstrates a functional capacity adequate to consistently perform none or only a few of the usual duties or activities of vocation or self-care."
- The SF-DCT's current standard for disability Level A claims requires claimants to submit proof that you are 100% disabled in both vocation **and** self-care. The CAC has a motion pending before the Court on this issue. Until the court rules, you may accept a lower payment for a Level B disability claim (if you qualify) and, if the Court rules in favor of the CAC, the SF-DCT will identify claims affected by the ruling, re-review them and notify you accordingly.

Level A-Total Disability (100%), \$50,000 (U.S.)

Examples of a PSS Total Disability A Claim:

- Medical records from 1974 reflect that the claimant had severe dry eyes resulting in multiple eye surgeries including the insertion of plugs. Records from 1984 reflect that the claimant's vision deteriorated to the point of partial loss of vision in both eyes. In 1994, she was approved for SSI based solely on her diagnosis of Primary Sjogren's Syndrome (PSS) because of dry eye syndrome. Records dated from 1995 through 2000 reflect that she required assistance from friends and family members to cook, clean, vacuum and grocery shop due to her loss of vision from dry eye syndrome. Her daughter must assist with dressing and undressing, must take her to and from the toilet and assist her with getting on and off the toilet because of the loss of the patient's vision. All of the claimant's grooming is done by her daughter such as combing, brushing and blow drying her hair because the claimant can no longer see to do so.
- Because of dry eye syndrome, the claimant quit her job in 1994 because her limited vision made her unable to read documents, answer the phones or perform filing duties. She requires help from her husband with combing, brushing and washing her hair due to her partial loss of vision. Her husband must assist with feeding because she is unable to see and focus on small objects due to the deterioration in her vision as a result of dry eye syndrome.

Level A Deficiencies:

The following are some of the common reasons why the Level A disability claim may be found deficient:

- The file does not provide any details or descriptions about your inability to perform both vocation and self-care.
- The file reflects detailed descriptions about your inability to perform your vocation (job or homemaking) because of an approved symptom, but it does not contain information about your self-care limitations.
- The file reflects detailed descriptions about your inability to work and perform your self-care activities because of an approved symptom, but it indicates that you are able to perform homemaking duties without difficulty.
- The file reflects conflicting information about either your vocation and/or self-care limitations. (Example: the file states that you are either working or that you are able to perform all or most of your self-care activities.)

Level A Deficiencies (continued):

- The file reflects that your vocation is affected or limited by an ineligible symptom or condition. (Example: work injury, car accident, heart attack, etc.)
- The treating doctor or QMD bases your Level A total disability rank on symptoms that were not eligible or approved. (Example: The QMD states that you are unable to work because of fatigue; chronic fatigue is not an eligible symptom for PSS.)
- The treating doctor or QMD increases your disability rank to Level A total disability but he or she does not perform a new examination or provide current medical records to support the new disability rank. The new disability rank cannot be based solely on a phone call with you or a review of a questionnaire that you completed.
- The treating doctor or QMD states that you are totally disabled, but your medical records dated within the same time frame indicate that you are actively exercising and/or socializing.

Level B – \$20,000 (U.S.)

To qualify for Severity Level B, your QMD statement and/or your medical records must document ONE of the following conditions:

- Central Nervous System Involvement; or
- Severe Cardiopulmonary Involvement; or
- Pseudo-lymphoma or Lymphoma.

Level B - Central Nervous System Involvement

Central Nervous System (CNS) involvement includes both the central and peripheral nervous system. Acceptable ways to document this are:

- Records showing Multiple Sclerosis-like symptoms with abnormal cerebral spinal fluid (fluid that surrounds the brain and spinal cord); or
- Multiple Sclerosis-like symptoms with an abnormal MRI; or
- Peripheral Neuropathy symptoms: numbness, tingling, burning pain, paresthesias, loss or decreased sensation in the extremities; or
- Cutaneous Vasculitis – Inflammation of blood vessels affecting the skin often with involvement of other organs.

Level B – Cardiopulmonary Involvement

Cardiopulmonary involvement includes both the heart and the lungs. The physician must state that he or she believes that ONE of the following conditions are directly related to PSS:

- Chronic Bronchitis secondary to dryness of the upper and lower airway with mucous plugging; or
- Lymphocytic Interstitial Pneumonitis; or
- Pseudo-lymphoma with nodular infiltrates; or
- Pleural Effusions; or
- Pulmonary Hypertension; or
- Mild to moderate obstructive airway disease; or
- Interstitial Fibrosis; or
- Desiccation of tracheo-broncheal mucous membrane; or
- Lymphoid Interstitial disease.

Level B – Pseudo-lymphoma or Lymphoma

Pseudo-Lymphoma or Lymphocytoma is a disorder that exhibits clinical and histological features suggestive of malignant lymphoma.

Severity Level C – \$10,000 (U.S.)

If you are approved for PSS but you do not meet the criteria for Severity/Disability Level A or Severity Level B, then the SF-DCT will automatically approve you for Severity Level C – \$10,000 (U.S.)

Other types of diseases or conditions eligible for Disease compensation in the Plan:

A very small percentage of the PSS claims (1%) processed by the SF-DCT are approved.

Most claims that do not qualify for PSS are reviewed by the SF-DCT for another condition in the Plan called “Atypical Connective Tissue Disease” or “ACTD.” Information about ACTD is in your Disease Claimant Information Guide at Tab 1 and at Annex A-97. Pursuant to Court Order, additional information about ACTD will be released by the SF-DCT on or before January 7, 2008.

The compensation amounts for approved PSS and ACTD claims are the same