

SF-DCT INFORMATION FOR PM AND DM CLAIMS

OPTION 2

Polymyositis (PM) and Dermatomyositis (DM)

Polymyositis (pol-E-my-OH-sigh-tiss) is a rare disease in which the immune system chronically inflames and weakens the body's muscle tissue in many parts of the body, especially those closest to the trunk, e.g., shoulders, hips, and/or thigh areas. It is a chronic illness with periods of increased symptoms, called flares or relapses and minimal or no symptoms, known as remissions. (1)

Dermatomyositis (der-MAT-o-my-OH-sigh-tiss) is also a rare disease with inflammation that damages both muscle fibers and skin. Like PM, a person develops muscle weakness. In addition, there is a distinctive patchy, reddish rash on the eyelids, cheeks, bridge of the nose, back or upper chest, elbows, knees and knuckles. In some cases, hardened bumps under the skin may develop. (1)

Other symptoms that may be associated with PM and DM are dysphagia, fatigue and pain. These symptoms are not eligible symptoms to qualify for PM or DM in the Dow Corning Settlement.

(1) Source: Website: www.MedicineNet.com

Checklist of documents to submit for PM and DM Disease Option 2 claims:

Your records must document ALL of the following to qualify for PM or DM under Disease Option 2:

- ❑ Diagnosis of PM or DM by a Board Certified Rheumatologist (BCR); and
- ❑ All medical records – including the laboratory tests, muscle biopsy and EMG test results – that support the symptoms of PM or DM (p. 4-25); and
- ❑ The medical records should include the actual office notes of your exam by the BCR; and
- ❑ The symptoms of PM/DM must have occurred within a single 24 month period within 5 years prior to the time the claim was submitted; and
- ❑ A statement or letter from the BCR that none of the symptoms of PM/DM existed prior to the date of your first breast implantation.

What are the symptoms of PM and DM Disease Option 2?

PM – in addition to the diagnosis of PM or DM, your records must document that you have ALL FOUR symptoms below (1-4):

- 1. Symmetrical proximal muscle weakness (p. 5-7); **and**
- 2. EMG changes characteristic of Myositis (p. 8-11); **and**
- 3. Elevated serum muscle enzymes (p. 12-18); **and**
- 4. Abnormal muscle biopsy (p. 19-23).

DM – in addition to the diagnosis of PM or DM, your records must document that you have 3 of the 4 symptoms above PLUS the skin rash below:

- Dermatological (Skin Rashes) (p. 24-25).

CITE: Annex A-105 and
Disease Claimant Information Guide at Tab 1

SYMPTOM 1: SYMMETRICAL PROXIMAL MUSCLE WEAKNESS

This means that you must have muscle weakness on both sides of your body at the primary muscle attachment or those nearest the body, e.g. shoulders, hips and/or thighs.

- ✓ Symmetrical means on both sides.
- ✓ Proximal refers to muscles nearest to the body.

This symptom must be documented in writing in your medical records.

SYMMETRICAL PROXIMAL MUSCLE WEAKNESS - acceptable records

You can document this symptom in writing if the exam by the BCR shows one of the following:

- ✓ Bilateral shoulder girdle weakness; or
- ✓ Decrease muscle strength in the hips; or
- ✓ Bilateral lower extremity weakness (thighs or hip areas); or
- ✓ Weakness of the supraspinatous bilaterally (these muscles help elevate the arms); or
- ✓ Hip flexor strength as R 3/5 and L 3/5 (or lower) (this means strength rating of 4 or less on a scale of 5).

SYMMETRICAL PROXIMAL MUSCLE WEAKNESS - unacceptable records

Common reasons why claimants receive a deficiency notice about the symptom of symmetrical proximal muscle weakness:

- Your file does not contain a medical record that shows muscle weakness found on exam.
- The exam shows muscle weakness only on one side of the body; this is not symmetrical.
- The exam shows muscle weakness of the distal muscles, such as foot, wrists or ankles. It must be from the proximal muscles (shoulders, hip, thighs).

SYMPTOM 2: EMG changes characteristic of myositis

EMG stands for Electromyography.

It is the insertion of a needle electrode into the skeletal muscle. It measures and records the electrical activity of the muscle at rest and during contraction. It is used in the evaluation of patients with diffuse or localized muscle weakness. An EMG is usually performed in the early stages of diagnosing PM or DM. It is a very invasive procedure and is rarely ever repeated after a diagnosis is established.

The EMG report must be submitted .

EMG - acceptable records

The EMG report must be abnormal and show characteristics of myositis (at least ONE of the following descriptions must be contained in the EMG report):

- ✓ short duration, small, low amplitude polyphasic potential; or
- ✓ fibrillation potentials; or
- ✓ bizarre high-frequency repetitive discharges.

What if my EMG is not in the same 5 year/24 month time frame as my other symptoms?

The Settlement Facility will not require you to repeat an invasive test just to meet the 24 month time frame. We will allow the test to be credited outside the required 5 year/24 month time frame.

EMG – unacceptable records

We must have the actual EMG report.

A physician's statement that you have abnormal EMG results that are diagnostic of myositis, or that you have at least one of the confirming findings of PM/DM on the EMG report is not acceptable.

SYMPTOM 3: ELEVATED SERUM MUSCLE ENZYMES

Certain tissue cells contain characteristic enzymes which enter the blood only when the cells to which they are confined are damaged or destroyed. The presence in the blood of significant quantities of these specific enzymes indicates the probable site of tissue damage.

The actual laboratory report of the muscle enzyme test must be submitted.

Elevated serum muscle enzymes – acceptable records

Serum muscle enzymes are measured by a blood test. This must be documented by a lab report that shows at least one of the following enzymes is elevated:

- **CPK** – (creatine kinase, creatinine phosphokinase); or
- **Aldolase** – (no abbreviation); or
- **SGOT** – (serum glutamic transaminase); or
- **SGPT** – (serum glutamic pyruvic transaminase); or
- **LDH** – (lactate dehydrogenase).

CPK - Creatine Phosphokinase

- Most of the CPK resides in skeletal muscle, heart muscle, and in the gastrointestinal tract. CPK enters the blood rapidly following damage to muscle cells. CPK can be an excellent marker for skeletal muscle damage.

Aldolase

- Aldolase is present most significantly in skeletal and heart muscle. Damage to skeletal muscle produces high serum levels of aldolase, particularly in the case of progressive muscular dystrophy.

SGOT and SGPT

- Serum Glutamic-Oxaloacetic Transaminase (SGOT) occurs in moderate amounts in skeletal muscle. Elevated SGOT levels can be used to diagnose PM or DM.
- Serum Glutamic-Pyruvic Transaminase (SGPT) is found in significant quantities in skeletal muscle. Elevated SGOT levels can be used to diagnose PM or DM.

LDH – Lactic Dehydrogenase

LDH is an enzyme that catalyzes the reversible reaction between pyruvic and lactic acids.

- LDH is present in nearly all types of metabolizing cells. The enzyme is especially concentrated in the muscles. As muscle cells die, their LDH is released and the enzyme enters the blood, causing the elevation in blood levels.
- The total LDH in the blood can be further divided into five components or fractions labeled by number: LDH-1, LDH-2, LDH-3, LDH-4, and LDH-5. Each of these fractions are called isoenzymes and help determine the location of the muscle injury, for example differentiate between heart muscle (heart attack) or skeletal muscle (polymyositis).

Elevated Serum Muscle Enzymes – unacceptable records

1. A physician's statement alone reporting the laboratory abnormalities is not acceptable. We must have the report from the performing laboratory, including the reference values used by the lab.
2. If medical records show that the enzymes are not elevated, this is not acceptable. Abnormally low values are not acceptable.
3. If medical records indicate elevated levels of enzymes but they are related to another disease or condition and not PM/DM, this is not acceptable.

SYMPTOM 4: ABNORMAL MUSCLE BIOPSY

A muscle biopsy is the removal of muscle tissue for microscopic examination and chemical analysis. It is usually performed in the early stages of diagnosing PM or DM and is a very invasive procedure. It is rarely ever repeated after a diagnosis is established.

The actual muscle biopsy report must be submitted.

Abnormal Muscle Biopsy – acceptable records

The muscle biopsy report must show ALL of the following elements:

- ✓ necrosis of type 1 and 2 muscle fibers; and
- ✓ areas of degeneration and regeneration of fibers; and
- ✓ Phagocytosis; and
- ✓ Interstitial or perivascular inflammatory response.

What if my Muscle Biopsy is not in the same 5 year/24 month time frame as my other symptoms?

The Settlement Facility will not require you to repeat an invasive test just to meet the 24 month time frame. We will allow the test to be credited outside the required 5 year/24 month time frame.

What if all of the information is not in the Muscle Biopsy report?

If all elements of the muscle biopsy are not present, there must be other evidence that the findings present are consistent with a diagnosis of Polymyositis or Dermatomyositis.

Abnormal Muscle Biopsy – unacceptable records

We must have the histology/pathology report with interpretation of the findings.

A physician's statement that describes the presence of the elements of the muscle biopsy is not acceptable.

DERMATOMYOSITIS – Skin Rash

To document the skin rash necessary for Dermatomyositis, the medical records must show ONE of the following rashes:

1. heliotrope (lilac) discoloration of the eyelids; or
2. erythematous rash-scaly involvement of the face, neck, shawl area (upper torso area) and extensor surfaces of the knees and medial malleoli (ankle area); or
3. Gottron's papules- pink to violaceous scaling areas typically found over the elbows, knuckles, and knees.

Skin Rash – unacceptable records

The following rashes are not acceptable rashes for DM:

- eczema
- contact dermatitis
- rash caused by a medication
- rash caused by any other disease or condition
- a rash other than the types of rashes listed on p. 24.

What is the payment for PM or DM in Disease Option 2?

All Class 5 disease claims that are approved for PM or DM in Disease Option 2 will be paid a Base Payment of \$110,000 (U.S.) (subject to the multiple manufacturer reduction).

If Premium Payments are approved by the District Court, approved PM-DM claimants could receive an additional payment of \$22,000 (U.S.).

SF-DCT INFORMATION FOR PM AND DM CLAIMS

OPTION 1

Polymyositis (PM) and Dermatomyositis (DM)

Polymyositis (pol-E-my-OH-sigh-tiss) is a rare disease in which the immune system chronically inflames and weakens the body's muscle tissue in many parts of the body, especially those closest to the trunk, e.g., shoulders, hips, and/or thigh areas. It is a chronic illness with periods of increased symptoms, called flares or relapses and minimal or no symptoms, known as remissions. (1)

Dermatomyositis (der-MAT-o-my-OH-sigh-tiss) is also a rare disease with inflammation that damages both muscle fibers and skin. Like PM, a person develops muscle weakness. In addition, there is a distinctive patchy, reddish rash on the eyelids, cheeks, bridge of the nose, back or upper chest, elbows, knees and knuckles. In some cases, hardened bumps under the skin may develop. (1)

Other symptoms that may be associated with PM and DM are dysphagia, fatigue and pain. These symptoms are not eligible symptoms to qualify for PM or DM in the Dow Corning Settlement.

(1) Source: Website: www.MedicineNet.com

Checklist of documents to submit for PM – DM Disease Option 1 claims:

- ❑ An evaluation by a Qualified Medical Doctor (QMD) (p. 30-31); or
 - ❑ Medical records supporting the symptoms of PM/DM (p. 32).
- AND**
- ❑ Documentation of the severity level of the disease (p. 35-38).

How can you document a PM or DM Claim in Disease Option 1?

There are two ways to document a claim for PM or DM in Option 1:

1. A signed statement or diagnosis from a Qualified Medical Doctor (QMD), or
2. Submit your medical records for review by the SF-DCT.

Acceptable QMD's for PM or DM – Disease Option 1

One way to qualify for PM or DM in Disease Option 1 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.

Note: 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.

What are the symptoms of PM or DM – Disease Option 1?

PM – requires all 4 symptoms below:

- 1. Symmetrical Proximal Muscle Weakness (p. 5 -7); and
- 2. EMG Changes Characteristic of Myositis (p. 8-11); and
- 3. Elevated Serum Muscle Enzymes (p. 12-18); and
- 4. Abnormal Muscle Biopsy (p. 19-23).

DM – requires 3 of the 4 symptoms above plus:

- Dermatological (Skin Rashes) (p. 24-25).

NOTE: You do not need to submit the laboratory tests, muscle biopsy report or EMG report, if the test results are adequately described in the QMD letter. However, it is best to submit the actual reports, because a QMD seldom writes all of the required elements of a report in his/her letter.

CITE: Annex A-95 – 96 and
Disease Claimant Information Guide at Tab 1

If your claim does not qualify for PM or DM in Disease Option 1, the SF-DCT may review your claim for “ACTD.”

FOR DISEASE OPTION 1 CLAIMS ONLY: If a claimant’s records document clinical symptoms or laboratory findings atypical of PM or DM, but who nonetheless have a PM-like or DM-like disease, the SF-DCT will review the claim for ACTD. If the symptoms more closely resemble Atypical Connective Tissue Disease (ACTD), then the SF-DCT shall review and possibly approve the claim in ACTD, and not PM or DM. (1)

The compensation amounts for approved Option 1 PM/DM 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 is PM-like and DM-like (or atypical PM/DM) disease?

A diagnosis of Polymyositis-like disease requires the presence of 3 out of the 4 symptoms listed on p. 32 **without** the presence of the skin rash.

A diagnosis of Dermatomyositis-like disease requires the presence of 2 out of the 4 symptoms listed on p. 32 **with** the presence of one of the skin rashes on p. 24.

What are the levels of compensation for PM and DM – Disease Option 1?

- Level A - \$50,000 - Death or Total Disability.
- Level B – \$20,000 - PM or DM with associated malignancy and/or respiratory involvement.
- Level C - \$10,000 - Other, including PM or DM with muscle strength of Grade III (3) or less.

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

The compensation amounts for approved Option 1 PM/DM 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.

Examples of acceptable ways to document Severity Level A disability for PM or DM- \$50,000 (U.S.)

- Medical records from 1974 reflect that the patient had severe muscle weakness. Records from 1984 reflect that her muscle weakness deteriorated so that she is in a wheelchair. In 1994, she was approved for SSI based solely on her diagnosis of Polymyositis due to muscle weakness. 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 muscle strength. 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 since she is unable to stand or walk alone due to muscle weakness in her legs. All of her grooming is done by her daughter such as combing, brushing and blow drying her hair because she can no longer do so as a result of arm weakness.
- Due to muscle weakness, the claimant quit her job in 1994 because of her limited mobility and the inability to perform filing duties due to muscle arm weakness. She requires help from her husband with combing, brushing and washing her hair due to weakness in her arms. Her husband must assist with feeding because she is unable to hold eating utensils due to muscle arm weakness.

Severity Level B for PM or DM- \$20,000 (U.S)

Polymyositis or Dermatomyositis: with associated malignancy (cancer) and/or respiratory muscle involvement.

Medical records must support PM or DM with a malignancy (cancer). The cancer can occur before, at the same time or after the diagnosis of PM or DM has been made. Ovarian and gastric cancers are common types of cancer associated with PM.

Medical records must support PM or DM with respiratory muscle involvement. Respiratory problems can be before, at the same time or after the diagnosis of PM or DM has been made. Respiratory involvement may include: Velcro like crackles on auscultation of the chest and interstitial fibrosis on the chest x-ray. Aspiration pneumonia can occur if swallowing difficulties are present.

Severity Level C for PM – DM- \$10,000 (U.S.)

To qualify for Severity Level C, you may have PM or DM with muscle strength of Grade III (3) or less.

If you do not qualify for Severity/Disability Levels "A" or "B", you will automatically qualify for Option I PM/DM severity level "C" by default, if you qualify for disease.

In summary, what is the difference between Disease Option 1 and 2 claims for PM - DM?

Listed below are some of the differences:

DISEASE OPTION 2	DISEASE OPTION 1
You must be examined by a Board Certified Rheumatologist (BCR).	You can rely on the evaluation of a “Qualified Medical Doctor” or a review of your medical records by the SFDCT.
You must have a diagnosis of PM or DM by the BCR.	You can qualify if you do not have a diagnosis of PM or DM if the symptoms support the claim; atypical presentations and PM-like or DM-like descriptions are sufficient.
You must submit the physician’s office notes <u>and</u> the laboratory report, EMG report and muscle biopsy report that support your symptoms.	You must submit the medical records or a QMD evaluation that support your symptoms. You are not required to submit the laboratory report, EMG report or muscle biopsy report if these symptoms are documented in the QMD letter or medical records.
Your symptoms must have occurred within a single 24 month within 5 years of submitting your claim	There is no 24 month/5 year time frame requirement in Option I.
Pre-existing PM or DM symptoms will not be credited as eligible symptoms. You must submit a letter or statement from your BCR stating that none of your symptoms existed prior to the date of your first breast implantation.	Pre-existing PM or DM symptoms can be credited as eligible symptoms; however, the amount of compensation will depend on whether the severity or disability of the disease became worse after implantation.

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

A very small percentage of the PM or DM claims (less than 10%) processed by the SF-DCT are approved.

Most claims that do not qualify for PM or DM 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 amount of compensation paid for Option I PM/DM and ACTD claims is the same.