

SF-DCT INFORMATION FOR MIXED CONNECTIVE TISSUE DISEASE (MCTD) CLAIMS

OPTION 1

(MCTD claims are not eligible for Disease Option 2)

Mixed Connective Tissue Disease (MCTD)

Mixed Connective Tissue Disease or MCTD is an autoimmune disease in which the immune system attacks the body. It commonly causes joint pain/swelling, Raynaud's Phenomenon, muscle inflammation, and scarring of the skin of the hand and has features of Lupus, Scleroderma, Myositis and Rheumatoid Arthritis, together with a large quantity of antibodies against one specific antigen, U1RNP. It is considered a distinct clinical disorder despite its overlap with other diseases and syndromes.

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

Checklist of documents to submit for a MCTD claim:

- ❑ An evaluation by a Qualified Medical Doctor (QMD) (see page 4)
- or
- ❑ Medical records supporting the symptoms of MCTD (see pages 5-21)
- AND
- ❑ Documentation of the severity level of the disease (see pages 22-36)

Acceptable QMD Certification for MCTD

One way to qualify for MCTD 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 can also be your treating physician.

What are the eligible criteria for MCTD in the Plan?

To be eligible for MCTD in the Plan, you must document:

1. Clinical symptoms characteristic of two or more of the following rheumatic diseases: Systemic Sclerosis, Systemic Lupus Erythematosus (SLE), Myositis and/or Rheumatoid Arthritis (RA) (see pages 6-15);

and

2. Positive RNP antibodies (see pages 19-20).

1st Criteria: Clinical Symptoms of Eligible Rheumatic Diseases

You must document that you have clinical symptoms characteristic of 2 or more of the following rheumatic diseases:

- **Systemic Sclerosis** (Scleroderma) (skler-a-DER-ma) – Scleroderma is a chronic and progressive disease that causes inflammation and thickening of the skin and the formation of scar tissue in organs (see pages 13-18 for symptoms related to Systemic Sclerosis); or
- **Systemic Lupus Erythematosus** (SLE) – Lupus is a condition of chronic inflammation when the body's tissues are attacked by its own immune system or
- **Myositis** (my-OH-sigh-tis) – Myositis is inflammation of the skeletal muscles, which are also called the voluntary muscles. These are the muscles you consciously control that help you move your body (see pages 11-12); or
- **Rheumatoid Arthritis** (RA) – Rheumatoid arthritis is an autoimmune disease that causes chronic inflammation of the joints. Rheumatoid arthritis can also cause inflammation of the tissue around the joints, as well as other organs in the body (see pages 9-10).

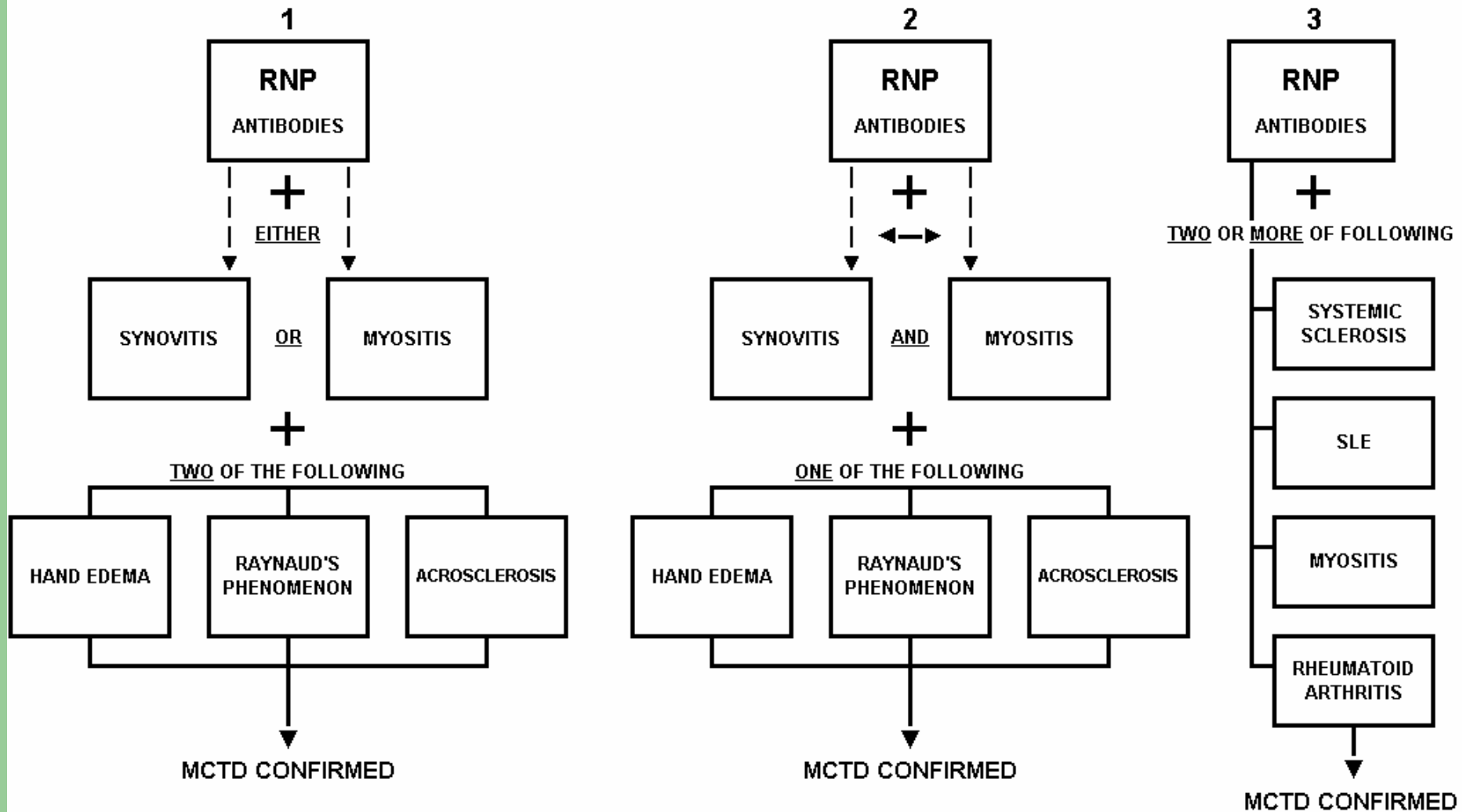
1st Criteria: Clinical Symptoms of Eligible Rheumatic Diseases

You can also document that you have clinical symptoms of 2 or more rheumatic diseases by submitting either a QMD letter or medical records documenting positive RNP antibodies **plus** one of the following:

1. Synovitis **OR** Myositis **AND** any two of the following: hand edema, Raynaud's phenomenon, or Acrosclerosis (see pages 13-18); **or**
2. Synovitis **AND** Myositis **AND** any one of the following: hand edema, Raynaud's phenomenon, or Acrosclerosis (see pages 13-18).

For additional ways to approve MCTD, see chart on page 8.

WAYS TO APPROVE MCTD



1st Criteria: Synovitis and how it is credited

Synovitis (sin" o-vi'tis) is the swelling of a joint. You can document this by either:

- A QMD letter that recites a history of synovitis or joint **swelling** in one or more joints; or
- Your medical records reflect either a complaint or history of synovitis or joint **swelling** in one or more joints.

NOTE: Complaints of joint **pain** (arthralgias) alone are not sufficient. There must be a complaint of joint **swelling** for the symptom of Synovitis to be credited.

Synovitis - unacceptable records

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

- Medical records do not reflect a complaint or a history of joint swelling.
- Medical records do not reflect a history or a complaint of synovitis.
- Medical records reflect complaints of arthralgias (joint pains) **not** joint swelling.

1st Criteria: Myositis and how it is credited

Myositis (my-OH-si'-tis) is an inflammation of the muscle. You can document this by either:

- A positive or abnormal muscle biopsy report; or
- A laboratory report reflecting an elevated CK or CPK.

CK or CPK is a blood test that measures creatine phosphokinase, an enzyme found mainly in the heart, brain and skeletal muscle. When the total CPK level is very high, it usually means there has been injury or stress to the heart, the brain, or muscle tissue. For example, when a muscle is damaged, CPK leaks into the bloodstream. Determining which specific form of CPK is high helps doctors determine which exact tissue has been damaged.

NOTE: If the file reflects a positive/abnormal biopsy result or an elevated CK or CPK, the actual biopsy or lab report is not required.

Myositis - unacceptable records

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

- Medical records do not contain a muscle biopsy report.
- Muscle biopsy results do not confirm a diagnosis of Myositis.
- CK or CPK laboratory results are decreased rather than elevated.

1st Criteria: Hand Edema and how it is credited

Hand Edema is defined as an excessive accumulation of fluid in the tissue spaces of the hand. You can document this by either:

- A QMD letter that recites either a complaint or a history of Hand Edema or swelling, “puffy edema” or “Scleroderma-like puffy edema”; or
- Your medical records reflect either a complaint or history of Hand Edema, swelling, “puffy edema” or “Scleroderma-like puffy edema.”

NOTE: If Hand Edema is related to any condition or illness other than early stages of Scleroderma, the symptom cannot be credited.

Hand Edema – unacceptable records

Common reasons why claimants receive a deficiency notice about the symptom of Hand Edema

- Medical records do not reflect a complaint or a history of hand edema.
- Medical records directly relate hand edema to ineligible medical conditions or illnesses such as diabetes, injuries, arthritis etc.

1st Criteria: Raynaud's Phenomenon and how it is credited

Raynaud's phenomenon (RP) is a condition resulting in a particular series of discolorations of the fingers and/or toes after exposure to changes in temperature (cold or hot). Initially, the digit(s) involved turn white because of the diminished blood supply. The digit(s) then turn blue because of prolonged lack of oxygen. Finally, the blood vessels reopen causing a local “flushing” phenomenon which turns the digit(s) red. This three-phase color sequence (white to blue to red), most often upon exposure to cold temperatures, is characteristic of RP.

Raynaud's Phenomenon (RP) can be credited if it the file reflects any ONE of the following:

- A diagnosis of, or notes a history of Raynaud's phenomenon; or
- Complaints of color changes on the hands when exposed to the cold; or
- Color changes or ulcerations found on physical exam.

NOTE: If the file reflects a diagnosis of Raynaud's, it is not required that the color changes or ulcerations (sores) be described.

Raynaud's – unacceptable records

Common reasons why claimants receive a deficiency notice about the symptom of Raynaud's Phenomenon:

- Medical records do not reflect a diagnosis of Raynaud's.
- Medical records reflect a history of only one color change.
- Medical records reflect only one color change found on physical exam.

1st Criteria: Acrosclerosis and how it is credited

Acrosclerosis (AK-ro-skla-RO-sis) is a special form of Scleroderma characterized by skin thickening and stiffness affecting the extremities, head and face and is associated with Raynaud's Phenomenon.

Acrosclerosis can be credited if the file reflects:

- Evidence of skin thickening found on physical examination.

The symptom cannot be credited based on a complaint alone by you to a doctor. The doctor must examine you and find evidence of skin thickening.

Acrosclerosis - unacceptable records

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

- Medical records do not reflect evidence of skin thickening found on physical examination.
- Medical records reflect only complaints or a history of skin thickening.

2nd Criteria – RNP Antibodies

In addition to documenting that you have clinical symptoms characteristic of 2 or more rheumatic diseases, you must document that you have positive RNP antibodies. RNP stands for Ribonucleoprotein and is a substance comprised of both protein and ribonucleic acid. (Dorland's Medical Dictionary, page 1463)

RNP can be credited if the file reflects ONE of the following:

- A laboratory test that reflects positive RNP antibodies greater than or equal to 1:600; or
- Your medical records or QMD letter refers to your laboratory results as positive for RNP antibodies and notes that the results are greater than or equal to 1:600.

NOTE: If the file does not contain a notation of positive RNP antibodies, the SF-DCT will not review the claim for MCTD. The claim will be reviewed for Overlap Syndrome (OS) and Atypical Connective Tissue Disease (ACTD).

RNP Antibodies – unacceptable records

Common reasons why claimants receive a deficiency notice about the symptom of RNP Antibodies

MCTD claims are denied because the files do not contain positive RNP antibodies.

If your claim does not qualify for MCTD, the SF-DCT will review your claim for Overlap Syndrome and ACTD.

If MCTD is not approved, the SF-DCT will review your file to see if you qualify for “Overlap Syndrome”(1) as that term is defined in the Plan. If Overlap Syndrome is not approved, the SF-DCT will then review your file for Atypical Connective Tissue Disease (ACTD)”(1) as that condition is defined in the Plan.

The compensation amounts for approved MCTD, Overlap Syndrome 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 Overlap Syndrome and ACTD are listed in the Disease Claimant Information Guide at Tab 1.

What are the levels of compensation for MCTD?

- Severity or Disability Level A – \$50,000 – Death or Total Disability
- Severity Level B – \$20,000 – Major organ involvement or major disease activity including central nervous system, cardio-pulmonary, vasculitic or renal involvement or Hemolytic Anemia (marked) or Thrombocytopenic Purpura or Severe Granulocytopenia
- Severity Level C – \$10,000 – Other

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

The compensation amounts for approved MCTD and ACTD claims are the same. Compensation amounts in Disease Option 1 are not based solely 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 the primary or secondary cause of death is related to MCTD or one of the approved conditions; or
- An autopsy report that indicates the cause of death is related to MCTD; or
- A letter from a QMD or the claimant's medical records that directly relate the primary or secondary cause of death to MCTD or one of the approved conditions.

Level A based on a claimant's death 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 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 an MCTD Total Disability A Claim:

- Medical records from 1974 reflect that the claimant's Hand Edema (swelling) and pain from Synovitis resulted in multiple treatment modalities, including but not limited to pain medication, physical therapy and steroid injections. Records dated from 1992 show that the claimant had to take early retirement because of her Synovitis and Hand Edema. In 1994, the claimant was approved for SSI based solely on the diagnosis of Mixed Connective Tissue Disease (MCTD). 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 function in her hands. Because of the pain from her Synovitis, the claimant's daughter must take her to and from the toilet and assist her with getting on and off the toilet because of the Hand Edema and Synovitis. All of the claimant's grooming is done by her daughter such as combing, brushing and blow drying her hair.
- In 1994, the claimant had to quit her job because of muscle weakness in her arms due to Myositis. She lost function in her arms and was unable to answer the phones or perform filing or typing duties due to muscle weakness and hand pain. Because of arm weakness, her husband must help her with combing, brushing and washing her hair. Her husband must also assist with feeding because of her inability to grasp small objects due to Synovitis and Hand Edema.

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 MCTD.)
- 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
- Cardiopulmonary Involvement; or
- Vasculitis; or
- Renal Involvement; or
- Hemolytic Anemia (marked); or
- Thrombocytopenic Purpura; or
- Severe Granulocytopenia.

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
- 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 related to MCTD:

- Heart – This may include Congestive Heart Failure, pericardial friction rub observed on Echocardiogram, pericardial thickening or effusion, EKG abnormalities (may include atrial and ventricular arrhythmias, ventricular gallops and sinus tachycardia), conduction disturbances, pericarditis and tamponade.
- Lungs – This may include dyspnea on exertion, limited effort tolerance, non-productive cough, chest pain, pleuritic symptoms, i.e., inspiratory crackles, rales, pulmonary hypertension-resulting in exaggerated second heart sound, right ventricular gallops, murmurs of pulmonic and tricuspid insufficiency, jugular venous distention, hepatojugular reflux, pedal edema and increased sputum.

Level B – Vasculitis

Vasculitis – inflammation of the blood or lymph vessels documented by lab reports, angiogram or biopsy.

The file must document **ONE** of the following:

- A diagnosis of Vasculitis; **or**
- An abnormal angiogram (x-ray) confirming Vasculitis; **or**
- An abnormal biopsy report confirming Vasculitis.

Level B – Renal Involvement

To establish severe renal involvement (failure) – the medical records must document **ONE** of the following:

- An abnormal 24 hour urine, creatine clearance, proteinuria >0.5 grams/day or 3+, possible albumin or casts **and** the medical records supporting ongoing renal dysfunction; **or**
- A diagnosis of nephrotic syndrome; **or**
- Kidney transplant or pending kidney transplant; **or**
- Dialysis treatments.

Level B – Hemolytic Anemia (marked)

Hemolytic Anemia (marked) – Hemolytic Anemia is a condition where there are not enough red blood cells in the blood. It is caused by premature destruction of red blood cells.

The Laboratory reports or medical records must document ONE of the following:

1. Decreased Hemoglobin and Hematocrit (H&H) – Hemoglobin is a protein in red blood cells that carries oxygen. Hematocrit is a blood test that measures the number of red blood cells and the size of red blood cells; **or**
2. Increased Mean Cell Volume (MCV) – The mean cell volume (MCV) is the average volume of a red blood cell; **or**
3. Increased Reticulocytes – A blood test that measures the body's production of immature red blood cells (reticulocytes); **or**
4. Hematuria – Hematuria is the presence of red blood cells (RBCs) in the urine; **or**
5. Jaundice – Jaundice is a yellow color in the skin, the mucous membranes, or the eyes.

Level B – Thrombocytopenic Purpura

Thrombocytopenic Purpura – "Thrombocytopenic" means the blood doesn't have enough platelets. "Purpura" means a person has excessive bruising. You can document this condition by a blood test that shows an abnormal decrease in the number of blood platelets or by physical examination.

In order to receive credit for this symptom, you must submit **ONE** of the following:

- Laboratory test reflects a decrease platelet count **and** a diagnosis of Thrombocytopenic Purpura; **or**
- A bone marrow test confirming a diagnosis of Thrombocytopenic Purpura; **or**
- Medical records reflect a history of abnormal bleeding or bruising **and** a diagnosis of Thrombocytopenic Purpura; **or**
- Medical records reflect bruises found on exam **and** a diagnosis of Thrombocytopenic Purpura.

Level B – Severe Granulocytopenia

Severe Granulocytopenia – Granulocytopenia is an abnormal reduction in the number of white blood cells. White blood cells fight infection and tissue damage.

In order to receive credit for this symptom, you must submit **ONE** of the following:

- Laboratory test reflects a decrease in granulocytes or neutrophils (white blood cells) **and** a diagnosis of Granulocytopenia; **or**
- A diagnosis of Granulocytopenia **and** signs and symptoms noted by history or found on physical examination. Symptoms may include: fever, sore throat, anal ulcers, painful mouth ulcers, poor healing wounds and/or repeated infections.

Level C – Other – \$10,000 (U.S.)

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

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

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

Most claims that do not qualify for MCTD 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 MCTD and ACTD claims are the same.