

EXHIBIT E1 — Revised Disease/Symptomology Definitions and Compensation Levels

I. General

- A. A claimant must file with the Claims Office all medical records establishing the required findings or laboratory abnormalities. Qualifying findings must have occurred within a single 24-month period within the five years immediately preceding the submission of the claim. (Findings supplemented in response to a deficiency letter sent by the Claims Office do not have to fall within the 24-month period outlined above.)
- B. If exclusions are noted for a required finding, the physician making the finding or ordering the test must affirmatively state that those listed exclusions are not present. The physician recording a GCTS finding or making a disease diagnosis must also affirmatively state that the qualifying symptoms did not exist before the date of first implantation. (This statement can be based upon patient history so long as consistent with medical records in the physician's possession.) Failure to make these affirmative statements will result in a deficiency letter. All underlying office charts, radiology/pathology reports, and test results must be supplied to the Claims Office.

II. Scleroderma (SS)

A claim for scleroderma must include a diagnosis of systemic sclerosis/scleroderma made by a board-certified rheumatologist based upon personal examination of the patient. [Exclusion: localized scleroderma] Supporting medical documentation must affirmatively reveal that the major or at least two of the minor criteria listed below are present:

- A. Major criterion: Proximal scleroderma -- symmetric thickening, tightening, and induration of the skin of the fingers and the skin proximal to the metacarpophalangeal or metatarsophalangeal joints. The changes may affect the entire extremity, face, neck, and trunk (thorax and abdomen). Description of this criterion is adequate if the board-certified rheumatologist records that physical examination of the patient revealed scleroderma skin thickening, and adequately describes the parts of the body where that thickened skin was found.
- B. Minor Criteria:
 1. Sclerodactyly: Above-indicated skin changes limited to the fingers.
 2. Digital pitting scars or loss of substance from the finger pad: Depressed areas at tips of fingers or loss of digital pad tissue as a result of ischemia.
 3. Bibasilar pulmonary fibrosis: Bilateral reticular pattern of linear or lineonodular densities most pronounced in basilar portions of the lungs on standard chest roentgenogram; may assume appearance of diffuse mottling or "honeycomb lung." These changes should not be attributable to primary lung disease.

Compensation Levels:

- A. Death resulting from SS, or severe chronic renal involvement manifested by a glomerular filtration rate of less than 50% of the age- and gender-adjusted norm, as measured by an adequate 24-hour urine specimen collection.
- B. Clinically significant cardio-pulmonary manifestations of scleroderma^{1/} or proximal scleroderma on the trunk (thorax and abdomen).
- C. A diagnosis of scleroderma in accordance with the above criteria that does not involve the findings in A or B above.

1. As manifested by interstitial fibrosis (based upon physical examination findings and abnormalities seen on chest x-ray or chest CT) or pulmonary hypertension (based upon physical examination findings and 2-D Echo doppler or angiography with hemodynamic measurements showing pulmonary artery pressures of greater than 25 TORR).

III. Lupus (SLE)

A claim for SLE must include a diagnosis of SLE (lupus) made by a board-certified rheumatologist based upon personal examination of the patient. [Exclusion: mild lupus (SLE not requiring regular medical attention including doctor visits and regular prescription medications)] Supporting medical documentation must affirmatively reveal that at least four of the following 11 criteria are present:

Criterion	Definition
1. Malar rash	Fixed erythema, flat or raised, over the malar eminences, tending to spare the nasolabial folds
2. Discoid rash	Erythematous raised patches with adherent keratotic scaling and follicular plugging; atrophic scarring may occur in older lesions
3. Photosensitivity	Skin rash as a result of unusual reaction to sunlight, by patient history or physician observation
4. Oral ulcers	Oral or nasopharyngeal ulceration, usually painless, observed by a physician
5. Arthritis	Nonerosive arthritis involving two or more peripheral joints, characterized by tenderness, swelling, or effusion [exclusion: erosive arthritis]
6. Serositis	a) Pleuritis -- convincing history of pleuritic pain or rub heard by a physician or evidence of pleural effusion, or b) Pericarditis -- documented by ECG or rub or evidence of pericardial effusion
7. Renal disorder	a) Persistent proteinuria greater than 0.5 grams per day or greater than 3+ if quantitation not performed, or b) Cellular casts -- may be red cell, hemoglobin, granular, tubular, or mixed
8. Neurologic disorder	Seizures -- in the absence of offending drugs or known metabolic derangements, <i>e.g.</i> , uremia, ketoacidosis, or electrolyte imbalance
9. Hematologic disorder	a) Hemolytic anemia -- with reticulocytosis, or b) Leukopenia -- less than 4,000/mm total on two or more occasions, or c) Lymphopenia -- less than 1,500/mm on two or more occasions, or d) Thrombocytopenia -- less than 100,000/mm in the absence of offending drugs
10. Immunologic disorder	a) Positive LE cell preparation, or b) Anti-DNA: antibody to native DNA in abnormal titer, or c) Anti-Sm: presence of antibody to Sm nuclear antigen, or d) False positive serologic test for syphilis known to be positive for at least 6 months and confirmed by Treponema pallidum immobilization or fluorescent treponemal antibody absorption test
11. Antinuclear antibody	An abnormal titer or antinuclear antibody by immunofluorescence or an equivalent assay at any point in time and in the absence of drugs known to be associated with "drug-induced lupus" syndrome

Compensation Levels:

- A. Death resulting from SLE, or severe chronic renal involvement manifested by a glomerular filtration rate of less than 50% of the age- and gender-adjusted norm, as measured by an adequate 24-hour urine specimen collection.
- B. SLE with involvement of one or more of the following: glomerulonephritis, seizures in the absence of offending drugs or known metabolic derangements, Lupus Psychosis, myocarditis, pneumonitis, thrombocytopenic purpura, hemolytic anemia (with hemoglobin of 10 grams or less), severe granulocytopenia (with a total white cell count less than 2000), or mesenteric vasculitis.
- C. A diagnosis of lupus in accordance with the above criteria that does not involve the findings in A or B above. (Default compensation level.)

IV. Polymyositis (PM) /Dermatomyositis (DM)

A claim for polymyositis or dermatomyositis must include a diagnosis of the disease made by a board-certified rheumatologist based upon personal examination of the patient. Supporting medical documentation must affirmatively reveal that the following criteria are present:

- for polymyositis, the first four criteria without the rash;
- for dermatomyositis, three of the first four criteria, plus the rash (#5).

Criteria:

1. symmetrical proximal muscle weakness;
2. EMG changes characteristic of myositis including (a) short duration, small, low-amplitude polyphasic potential, (b) fibrillation potentials, (c) bizarre high-frequency repetitive discharges;
3. elevated serum muscle enzymes (CPK, aldolase, SGOT, SGPT, and LDH);
4. muscle biopsy showing evidence of necrosis of type I and II muscle fibers areas of degeneration and regeneration of fibers, phagocytosis, and an interstitial or perivascular inflammatory response;
5. dermatologic features including a lilac (heliotrope), erythematous, scaly involvement of the face, neck, shawl area and extensor surfaces of the knees, elbows and medial malleoli, and Gotton's papules.

Compensation Level:

All confirmed PM/DM diagnoses will be compensated at the GCTS/PM/DM--A level.

V. General Connective Tissue Symptoms (GCTS):

A claim for GCTS does not have to include a diagnosis for "General Connective Tissue Symptoms," but the medical documentation must establish that the combination of findings listed below are present. [Exclusion: classical rheumatoid arthritis diagnosed in accordance with the revised 1982 ACR classification criteria.]

For compensation at Level A:

- (1) any two findings from Group I; or
- (2) any three non-duplicative findings from Group I or Group II.

For compensation at Level B:

- (1) one finding from Group I plus any four non-duplicative findings from Group II or Group III; or
- (2) two findings from Group II plus one non-duplicative finding from Group III.

The following duplications exist on the list of findings:

- rashes (#3 and #8)
- sicca (#2 and #12)
- serological abnormalities (#4 and #9)

In addition to the medical verification of the required findings, a claim for GCTS must include the affirmative physician statements outlined in General Guidelines above.

GROUP I FINDINGS

1. Polyarthritis, defined as synovial swelling and tenderness in three or more joints in at least two different joint groups observed on more than one physical examination by a board-certified physician and persisting for more than six weeks. [Exclusion: osteoarthritis.]
2. Keratoconjunctivitis Sicca, defined as subjective complaints of dry eyes and/or dry mouth, accompanied (a) in the case of dry eyes, by either (i) a Schirmer's test less than 8 mm wetting per five minutes or (ii) a positive Rose-Bengal or fluorescein staining of cornea and conjunctiva; or (b) in the case of dry mouth, by an abnormal biopsy of the minor salivary gland (focus score of greater than or equal to two based upon average of four evaluable lobules). [Exclusions: drugs known to cause dry eyes and/or dry mouth, and dry eyes caused by contact lenses.]

3. Any of the following immune-mediated skin changes or rashes, observed by a board-certified rheumatologist or board-certified dermatologist: (a) biopsy-proven discoid lupus; (b) biopsy-proven subacute cutaneous lupus; (c) malar rash -- fixed erythema, flat or raised, over the malar eminences, tending to spare the nasolabial folds [exclusion: rosacea or redness caused by sunburn]; or (d) biopsy-proven vasculitic skin rash.

GROUP II FINDINGS

4. Positive ANA greater than or equal to 1:40 (using Hep2), on two separate occasions separated by at least two months and accompanied by at least one test showing decreased complement levels of C3 and C4; or a positive ANA greater than or equal to 1:80 (using Hep2) on two separate occasions separated by at least two months. All such findings must be outside of the performing laboratory's reference ranges.
5. Abnormal cardiopulmonary symptoms, defined as (a) pericarditis documented by pericardial friction rub and characteristic echocardiogram findings (as reported by a board-certified radiologist or cardiologist); (b) pleuritic chest pain documented by pleural friction rub on exam and chest x-ray diagnostic of pleural effusion (as reported by a board-certified radiologist); or (c) interstitial lung disease in a non-smoker diagnosed by a board-certified internist or pulmonologist, confirmed by (i) chest x-ray or CT evidence (as reported by a board-certified radiologist) and (ii) pulmonary function testing abnormalities defined as decreased DLCO less than 80% of predicted.
6. Myositis or myopathy, defined as any two of the following: (a) EMG changes characteristic of myositis: short duration, small, low amplitude polyphasic potential; fibrillation potentials; and bizarre high-frequency repetitive discharges; (b) abnormally elevated CPK or adolase from the muscle (outside of the performing laboratory's reference ranges) on two separate occasions at least six weeks apart. (If the level of the initial test is three times normal or greater, one test would be sufficient.) [Exclusions: injections, trauma, hypothyroidism, prolonged exercise, or drugs known to cause abnormal CPK or aldolase]; or (c) muscle biopsy (at a site that has not undergone EMG testing) showing evidence of necrosis of type 1 and 2 muscle fibers, phagocytosis, and an interstitial or perivascular inflammatory response interpreted as characteristic of myositis or myopathy by a pathologist.
7. Peripheral neuropathy or polyneuropathy, diagnosed by a board-certified neurologist, confirmed by (a) objective loss of sensation to pinprick, vibration, touch, or position; (b) symmetrical distal muscle weakness; (c) tingling and/or burning pain in the extremities; or (d) loss of tendon reflex, plus nerve conduction testing abnormality diagnostic of peripheral neuropathy or polyneuropathy recorded from a site that has not undergone neural or muscular biopsy. [Exclusions: thyroid disease, antineoplastic treatment, alcoholism or other drug dependencies, diabetes, or infectious disease within the last three months preceding the diagnosis.]

GROUP III FINDINGS

8. Other immune-mediated skin changes or rashes, observed by a board-certified rheumatologist or board-certified dermatologist: (a) livedo reticularis; (b) lilac (heliotrope), erythematous scaly involvement of the face, neck, shawl area and extensor surfaces of the knees, elbows and medial malleoli; (c) Gotton's sign, pink to violaceous scaling areas typically found over the knuckles, elbows, and knees; or (d) diffuse petechiae.
9. Any of the following serologic abnormalities: (a) ANA greater than or equal to 1:40 (using Hep2) on two separate occasions separated by at least two months; (b) one or more positive ANA profile: Anti-DNA, SSA SSB, RNP, SM, Scl-70, centromere, Jo-1 PM-Scl, or double-stranded DNA (using ELISA with standard cutoffs); (c) anti-microsomal, anti-cardiolipin, or RF greater than or equal to 1:80.
10. Raynaud's phenomenon, evidenced by a physician-observed two (cold-related) color change as a progression, or by physician observation of evidence of cold-related vasospasm, or by physician observation of digital ulceration resulting from Raynaud's phenomenon.
11. Myalgias, defined as tenderness to palpation, performed by a physician, in at least three muscles, each persisting for at least six months.
12. Dry mouth, subjective complaints of dry mouth accompanied by decreased parotid flow rate using Lashley cups with less than 6 ml per five minutes. [Exclusion: drugs known to cause dry mouth]