Section K. Hematologic and Lymphatic Systems

Overview

In This Section This section contains the following topics:

Topic	Topic Name
1	Sickle Cell Disease
2	Other Hematologic and Lymphatic Conditions

1. Sickle Cell Disease

Introduction

This topic contains information about hemic and lymphatic conditions, including

- definition of
  – sickle cell disease, and
  – sickle cell anemia
- inheritance of
  – sickle cell trait, and
  – sickle cell anemia
- characteristics of sickle cell anemia, and
- mechanism of inheritance of sickle hemoglobin.

Change Date

a. Definition: Sickle Cell Disease

Sickle cell disease is a generic term for all disorders characterized by the presence of sickle hemoglobin in the red blood cells and includes

- sickle cell anemia
- sickle cell trait, and
- other hemoglobinopathies such as
  – sickle cell thalassemia, and
  – sickle-hemoglobin C disease.

Note: The phenomenon of sickling of red blood cells is a hereditary abnormality that of itself usually produces few ill effects.

b. Definition: Sickle Cell Anemia

Sickle cell anemia is a hereditary and familial disorder characterized clinically by symptoms of

- anemia
- arthritis
- leg ulcers, and
- acute attacks of pain.

Note: The age of onset is generally early childhood.

c. Inheritance of Sickle Cell Trait

Inheritance of sickle cell trait may be from one or both parents.

If sickle hemoglobin is inherited from one parent and normal hemoglobin from the other, the combination is referred to as sickle cell trait.

Note: Except for unusual circumstances, this is a benign asymptomatic condition and is not associated with increased morbidity.

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d. Inheritance of Sickle Cell Anemia

The inheritance of sickle hemoglobin from each parent results in sickle cell anemia.

Sickle cell anemia is usually accompanied by

- moderate to severe anemia, and
- appropriate clinical signs and symptoms, such as
  – enlargement of the heart
  – abnormalities of the musculoskeletal system
  – bone and joint pain, and/or
  – fever.

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e. Characteristics of Sickle Cell Anemia

Sickle cell anemia is a morbid state characterized by hemolytic anemia and the following manifestations

- the presence of peculiar sickle-shaped, or oat-shaped, red blood cells
- signs of excessive blood destruction and active blood formation, and
- repeated vaso-occlusive episodes.

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f. Mechanism of Inheritance of Sickle Hemoglobin

The presence of sickle hemoglobin, a variant of the normal hemoglobin in human red blood cells, is subject to the usual mechanisms of biologic inheritance.

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2. Other Hematologic and Lymphatic Conditions

Introduction

This topic contains information about other hemic and lymphatic conditions, including

- assigning a permanent and total (P&T) evaluation for amyloid light chains (AL) amyloidosis (primary amyloidosis)
- review examinations of non-Hodgkin's Lymphoma (NHL) and other persistent cancers
- pyramiding of NHL and chronic lymphocytic leukemia (CLL)
- definitions of bone marrow and stem cell transplant
- rating schedule update, and
- historical P&T evaluations.

Change Date

a. Assigning a P&T Evaluation for AL Amyloidosis

Assign a permanent and total (P&T) evaluation for amyloid light chains (AL) amyloidosis (primary amyloidosis). AL amyloidosis is considered incurable and progressive.

Notes:

- Evaluate AL amyloidosis under 38 CFR 4.117, diagnostic code (DC) 7717.
- Consider ancillary benefits associated with the award of P&T disability evaluations.
- AL amyloidosis is a disability that is presumptively associated with herbicide exposure.

References: For more information on

- rating disabilities associated with herbicide exposure, see M21-1, Part IV, Subpart ii, 2.C.3, and
- addressing claims for P&T malignancy, see M21-1, Part III, Subpart iv, 5.B.3.b.

b. Review Examinations of NHL and Other Persistent Cancers

When evaluating the need for a review examination at the two-year period prescribed in the rating schedule under 38 CFR 4.117, DC 7715, following discontinuance of the treatment phase for non-Hodgkin's lymphoma (NHL) or any other persistent cancer with a high mortality rate, consider that the various therapeutic treatment modalities may continue at intervals greater than the review period indicated in the rating schedule.

If the disease has actively persisted for several years, thoroughly examine the medical record to determine whether the disease is

- actually in remission, or
- still active and being regularly treated over extended periods of time.

Do ***not*** schedule a review examination unless the record clearly shows a long-term and stable remission.

Important: Consider assigning a P&T evaluation when a provision under 38 CFR 3.327(b)(2) applies or as otherwise warranted under the provisions of M21-1, Part III, Subpart iv, 5.B.3.b.

Reference: For more information about when not to schedule a review examination, see M21-1, Part III, Subpart iv, 3.B.2.d.

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c. Pyramiding of CLL and NHL

Do not assign separate evaluations for chronic lymphocytic leukemia (CLL) (38 CFR 4.117, DC 7703) and NHL (38 CFR 4.117, DC 7715). They are cancers of the same body system and assignment of multiple evaluations would be pyramiding.

In cases where both cancers are diagnosed, assign the appropriate evaluation using a hyphenated DC.

References: For more information on

- pyramiding, see M21-1, Part III, Subpart iv, 5.B.2.b, and
- presumptive service connection for CLL and NHL, see M21-1, Part IV, Subpart ii, 2.C.

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d. Definitions: Bone Marrow and Stem Cell Transplant

A ***bone marrow transplant***, also called a ***stem cell transplant***, is a procedure used to infuse healthy cells, called stem cells, into the body to replace damaged or diseased bone marrow. The process of infusing healthy cells into the body to replace diseased tissue may be referred to in a variety of manners based on the donor source and the type of tissue used. The following procedures all satisfy the criteria for the 100-percent rating under 38 CFR 4.117:

- stem cell transplant
- bone marrow transplant
- bone marrow stem cell transplant
- peripheral blood transplant, and
- peripheral blood stem cell transplant.

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e. Rating Schedule Update

The criteria for rating disabilities of the hematologic and lymphatic systems in 38 CFR 4.117 were updated effective December 9, 2018. The purpose of this update was to

- incorporate medical advances
- update medical terminology
- add disabilities not previously included, and
- refine rating criteria.

Note: This update was not a liberalizing change in the rating criteria.

Reference: For more information on historical changes to 38 CFR 4.117, see

- 38 CFR 4, Appendix A, and
- 38 CFR 4.117 (Historical).

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f. Historical P&T Evaluations

Historical policy guidance in existence prior to the December 9, 2018, rating schedule update directed the assignment of P&T evaluations for CLL and multiple myeloma. P&T evaluations assigned under the prior version of the rating schedule with application of those policies are protected under 38 CFR 3.951(a).

P&T evaluations were directed

- from November 6, 2003, to December 9, 2018, for CLL, with diagnosis being the sole requirement unless the condition was considered cured via treatment with a bone marrow transplant, and
- from January 28, 2003, to December 9, 2018, for multiple myeloma unless an exceptional situation was present in which inactive multiple myeloma was documented.

Important: In all situations, the evidence of record must be analyzed to determine the treatment plan and prognosis for active malignancy. As directed in M21-1, Part III, Subpart iv, 5.B.3.a-c, assign a P&T evaluation for active malignancy when warranted, whether the disability is evaluated under historical or current rating criteria.
