

# Medicaid Disability Manual

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## 107.00 Hematological Disorders

### A. What hematological disorders do we evaluate under these listings?

1. We evaluate non-malignant (non-cancerous) hematological disorders, such as hemolytic anemias (107.05), disorders of thrombosis and hemostasis (107.08), and disorders of bone marrow failure (107.10). These disorders disrupt the normal development and function of white blood cells, red blood cells, platelets, and clotting-factor proteins (factors).
2. We evaluate malignant (cancerous) hematological disorders, such as lymphoma, leukemia, and multiple myeloma, under the appropriate listings in 113.00, except for two lymphomas associated with human immunodeficiency virus (HIV) infection. We evaluate primary central nervous system lymphoma associated with HIV infection under 114.11B, and primary effusion lymphoma associated with HIV infection under 114.11C.

### B. What evidence do we need to document that you have a hematological disorder?

We need the following evidence to document that you have a hematological disorder:

1. A laboratory report of a definitive test that establishes a hematological disorder, signed by a physician; or
2. A laboratory report of a definitive test that establishes a hematological disorder that is not signed by a physician and a report from a physician that states you have the disorder; or
3. When we do not have a laboratory report of a definitive test, a persuasive report from a physician that a diagnosis of your hematological disorder was confirmed by appropriate laboratory analysis or other diagnostic method(s). To be persuasive, this report must state that you had the appropriate definitive laboratory test or tests for diagnosing your disorder and provide the results, or explain how your diagnosis was established by other diagnostic method(s) consistent with the prevailing state of medical knowledge and clinical practice.
4. We will make every reasonable effort to obtain the results of appropriate laboratory testing you have had. We will not purchase complex, costly, or invasive tests, such as tests of clotting-factor proteins, and bone marrow aspirations.

### C. What are hemolytic anemias, and how do we evaluate them under 107.05?

1. Hemolytic anemias, both congenital and acquired, are disorders that result in premature destruction of red blood cells (RBCs). Hemolytic anemias include abnormalities of hemoglobin structure (hemoglobinopathies), abnormal RBC enzyme content and function, and RBC membrane (envelope) defects that are congenital or acquired. The diagnosis of hemolytic anemia is based on hemoglobin electrophoresis or analysis of the contents of the RBC (enzymes) and membrane. Examples of congenital hemolytic anemias include sickle cell disease, thalassemia, and their variants, and hereditary spherocytosis. Acquired hemolytic anemias may result from autoimmune disease (for example, systemic lupus erythematosus) or mechanical devices (for example, heart valves, intravascular patches).

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2. The hospitalizations in 107.05B do not all have to be for the same complication of the hemolytic anemia. They may be for three different complications of the disorder. Examples of complications of hemolytic anemia that may result in hospitalization include dactylitis, osteomyelitis, painful (vaso-occlusive) crisis, pulmonary infections or infarctions, acute chest syndrome, pulmonary hypertension, chronic heart failure, gallbladder disease, hepatic (liver) failure, renal (kidney) failure, nephrotic syndrome, aplastic crisis, and strokes. We will count the hours you receive emergency treatment in a comprehensive sickle cell disease center immediately before the hospitalization if this treatment is comparable to the treatment provided in a hospital emergency department.

3. For 107.05C, we do not require hemoglobin to be measured during a period in which you are free of pain or other symptoms of your disorder. We will accept hemoglobin measurements made while you are experiencing complications of your hemolytic anemia.

4. 107.05D refers to the most serious type of beta thalassemia major in which the bone marrow cannot produce sufficient numbers of normal RBCs to maintain life. The only available treatments for beta thalassemia major are life-long RBC transfusions (sometimes called hypertransfusion) or bone marrow transplantation. For purposes of 107.05D, we do not consider prophylactic RBC transfusions to prevent strokes or other complications in sickle cell disease and its variants to be of equal significance to life-saving RBC transfusions for beta thalassemia major. However, we will consider the functional limitations associated with prophylactic RBC transfusions and any associated side effects (for example, iron overload) under functional equivalence and any affected body system(s). We will also evaluate strokes and resulting complications under 111.00 and 112.00.

### **D. What are disorders of thrombosis and hemostasis, and how do we evaluate them under 107.08?**

1. Disorders of thrombosis and hemostasis include both clotting and bleeding disorders, and may be congenital or acquired. These disorders are characterized by abnormalities in blood clotting that result in hypercoagulation (excessive blood clotting) or hypocoagulation (inadequate blood clotting). The diagnosis of a thrombosis or hemostasis disorder is based on evaluation of plasma clotting-factor proteins (factors) and platelets. Protein C or protein S deficiency and Factor V Leiden are examples of hypercoagulation disorders. Hemophilia, von Willebrand disease, and thrombocytopenia are examples of hypocoagulation disorders. Acquired excessive blood clotting may result from blood protein defects and acquired inadequate blood clotting (for example, acquired hemophilia A) may be associated with inhibitor autoantibodies.

2. The hospitalizations in 107.08 do not all have to be for the same complication of a disorder of thrombosis and hemostasis. They may be for three different complications of the disorder. Examples of complications that may result in hospitalization include anemias, thromboses, embolisms, and uncontrolled bleeding requiring multiple factor concentrate infusions or platelet transfusions. We will also consider any surgery that you have, even if it is not related to your hematological disorder, to be a complication of your disorder of thrombosis and hemostasis if you require treatment with clotting-factor proteins (for example, factor VIII or IX) or anticoagulant medication to control bleeding or coagulation in connection with your surgery. We will count the hours you receive emergency treatment in a comprehensive hemophilia treatment center immediately before the hospitalization if this treatment is comparable to the treatment provided in a hospital emergency department.

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## **E. What are disorders of bone marrow failure, and how do we evaluate them under 107.10?**

1. Disorders of bone marrow failure may be congenital or acquired, characterized by bone marrow that does not make enough healthy RBCs, platelets, or granulocytes (specialized types of white blood cells); there may also be a combined failure of these bone marrow-producing cells. The diagnosis is based on peripheral blood smears and bone marrow aspiration or bone marrow biopsy, but not peripheral blood smears alone. Examples of these disorders are myelodysplastic syndromes, aplastic anemia, granulocytopenia, and myelofibrosis. Acquired disorders of bone marrow failure may result from viral infections, chemical exposure, or immunologic disorders.
2. The hospitalizations in 107.10A do not all have to be for the same complication of bone marrow failure. They may be for three different complications of the disorder. Examples of complications that may result in hospitalization include uncontrolled bleeding, anemia, and systemic bacterial, viral, or fungal infections.
3. For 107.10B, the requirement of life-long RBC transfusions to maintain life in myelodysplastic syndromes or aplastic anemias has the same meaning as it does for beta thalassemia major. (See 107.00C4.)

## **F. How do we evaluate bone marrow or stem cell transplantation under 107.17?**

We will consider you to be disabled for 12 months from the date of bone marrow or stem cell transplantation, or we may consider you to be disabled for a longer period if you are experiencing any serious post-transplantation complications, such as graft-versus-host (GVH) disease, frequent infections after immunosuppressive therapy, or significant deterioration of organ systems. We do not restrict our determination of the onset of disability to the date of the transplantation in 107.17. We may establish an earlier onset of disability due to your transplantation if evidence in your case record supports such a finding.

## **G. How do we consider your symptoms, including your pain, severe fatigue, and malaise?**

Your symptoms, including pain, severe fatigue, and malaise, may be important factors in our determination whether your hematological disorder meets or medically equals a listing, or in our determination whether you otherwise have marked and severe functional limitations. We cannot consider your symptoms unless you have medical signs or laboratory findings showing the existence of a medically determinable impairment(s) that could reasonably be expected to produce the symptoms. If you have such an impairment(s), we will evaluate the intensity, persistence, and functional effects of your symptoms using the rules throughout 107.00 and in our other regulations. (See sections 416.928 and 416.929 of this chapter.) Additionally, when we assess the credibility of your complaints about your symptoms and their functional effects, we will not draw any inferences from the fact that you do not receive treatment or that you are not following treatment without considering all of the relevant evidence in your case record, including any explanations you provide on why you are not receiving or following treatment.

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## **H. How do we evaluate episodic events in hematological disorders?**

Some of the listings in this body system require a specific number of events within a consecutive 12-month period. (See 107.05, 107.08, and 107.10A.) When we use such criteria, a consecutive 12-month period means a period of 12 consecutive months, all or part of which must occur within the period we are considering in connection with your application or continuing disability review. These events must occur at least 30 days apart to ensure that we are evaluating separate events.

## **I. How do we evaluate hematological disorders that do not meet one of these listings?**

1. These listings are only common examples of hematological disorders that we consider severe enough to result in marked and severe functional limitations. If your disorder does not meet the criteria of any of these listings, we must consider whether you have a disorder that satisfies the criteria of a listing in another body system. For example, we will evaluate hemophilic joint deformity under 101.00; polycythemia vera under 103.00, 104.00, or 111.00; chronic iron overload resulting from repeated RBC transfusion (transfusion hemosiderosis) under 103.00, 104.00, or 105.00; and the effects of intracranial bleeding or stroke under 111.00 or 112.00.

2. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing. (See section 416.926 of this chapter.) Hematological disorders may be associated with disorders in other body systems, and we consider the combined effects of multiple impairments when we determine whether they medically equal a listing. If your impairment(s) does not medically equal a listing, we will also consider whether it functionally equals the listings. (See section 416.926a of this chapter.) We use the rules in §416.994a of this chapter when we decide whether you continue to be disabled.

## **107.01 Category of Impairments, Hematological Disorders**

**107.05 Hemolytic anemias**, including sickle cell disease, thalassemia, and their variants (see 107.00C), with:

A. Documented painful (vaso-occlusive) crises requiring parenteral (intravenous or intramuscular) narcotic medication, occurring at least six times within a 12-month period with at least 30 days between crises.

OR

B. Complications of hemolytic anemia requiring at least three hospitalizations within a 12-month period and occurring at least 30 days apart. Each hospitalization must last at least 48 hours, which can include hours in a hospital emergency department or comprehensive sickle cell disease center immediately before the hospitalization (see 107.00C2).

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OR

C. Hemoglobin measurements of 7.0 grams per deciliter (g/dL) or less, occurring at least three times within a 12 month period with at least 30 days between measurements.

OR

D. Beta thalassemia major requiring life-long RBC transfusions at least once every 6 weeks to maintain life (see 107.00C4).

**107.08 *Disorders of thrombosis and hemostasis***, including hemophilia and thrombocytopenia (see 107.00D), with complications requiring at least three hospitalizations within a 12-month period and occurring at least 30 days apart. Each hospitalization must last at least 48 hours, which can include hours in a hospital emergency department or comprehensive hemophilia treatment center immediately before the hospitalization (see 107.00D2).

**107.10 *Disorders of bone marrow failure***, including myelodysplastic syndromes, aplastic anemia, granulocytopenia, and myelofibrosis (see 107.00E), with:

A. Complications of bone marrow failure requiring at least three hospitalizations within a 12-month period and occurring at least 30 days apart. Each hospitalization must last at least 48 hours, which can include hours in a hospital emergency department immediately before the hospitalization (see 107.00E2).

OR

B. Myelodysplastic syndromes or aplastic anemias requiring life-long RBC transfusions at least once every 6 weeks to maintain life (see 107.00E3).

**107.17 *Hematological disorders treated by bone marrow or stem cell transplantation*** (see 107.00F). Consider under a disability for at least 12 consecutive months from the date of transplantation. After that, evaluate any residual impairment(s) under the criteria for the affected body system.