CD Part 7

Blood Diseases

A. Definitions

The following definitions are for words used in this chapter and during the SSA disability process. If you need additional definitions, consult a good medical dictionary, available in most bookstores and libraries. You can also look at online medical dictionaries like the one at www.medlineplus.gov.

Allergen. Substances that trigger an allergic immune reaction, such as pollen, molds, certain drugs, insect venom, and dust.

Anemia. Low red blood cell count, usually determined by a decrease in the *hematocrit*.

Antigens. Substances that trigger the body's immune system. Antigens are usually foreign substances such as allergens, bacteria, or viruses, but there are also autoimmune disorders in which the immune system mistakenly reacts to its own tissues as if they were antigens.

Aplastic anemia. Any anemia resulting from decreased red blood cell production by the bone marrow. There are many possible causes of aplastic anemia, such as drugs, infection, radiation, cancer, and toxic substances.

Autoimmune disorders. Disorders in which a person's immune system forms antibodies against their own tissues.

Biopsy. The process of taking a sample of tissue for detailed analysis of various kinds. Biopsy specimens are observed grossly with the eyes, microscopically with a variety of possible tissue stains, and in some cases may involve specific chemical and DNA analysis.

Blast crisis. See *leukemia*.

Bone marrow. Material inside of some bones that produces white blood cells, red blood cells, and platelets.

Bone marrow aspiration. A procedure involving suction of a sample of liquid *bone marrow* cells into a syringe, after penetrating the overlying bone with a special large needle. Bone marrow aspiration is often done in association with *bone marrow biopsy*.

Bone marrow biopsy. Procedure involving taking a small core of solid bone and underlying marrow so that the structure remains intact.

Cerebrospinal fluid (CSF). Fluid that flows around and within the spinal cord and brain, providing moisture and cushioning.

Cerebrovascular accident (CVA). Stroke. There are basically two kinds of stroke: hemorrhagic strokes caused by bleeding in the brain and thrombotic strokes caused by blockage of blood flow to a part of the brain.

Chronic. Persistent.

Coagulation. Clotting of blood.

Coagulation factors. Proteins that are necessary for the clotting of blood.

Complete blood count (CBC). Test providing information about the numbers and types of white blood cells, the numbers of red blood cells and how much hemoglobin they contain, the hematocrit, and the platelet count.

Electrophoresis. Method of identifying substances by the different distances they move when subjected to an electric field. Electrophoresis is usually performed on serum or urine samples. If the reactions of antibodies are combined with electrophoretic testing, the test is said to be immunoelectrophoresis.

Erythrocytosis. Increased red blood cell count.

Fibrosis. Degenerative process involving the replacement of normal tissue with fiber-like tissue.

Granulocytopenia. Decreased numbers of granulocytes. Granulocytes are neutrophils, basophils, and eosinophil types of white blood cells.

Heavy chain disease. A disorder of plasma cells (plasma cell dyscrasia) associated with the production of fragments of antibodies. There is more than one type of heavy chain disease, depending on the type of antibody involved.

Hemarthrosis. Bleeding into a joint space.

Hematocrit (Hct). The percentage of red blood cells in a volume of blood. For example, a hematocrit of 50% means that half of the blood volume is made up of red cells. In men, a normal Hct is about 42–48% and in women about 38–44% at sea level. At high altitudes, normal values are higher. Also known as the volume of packed red cells (VPRC).

Hemoglobin (Hb). The oxygen-carrying chemical in red blood cells.

Hemoglobinopathy. Any disorder involving the production of an abnormal type of *hemoglobin*. For example, sickle cell anemia is a hemoglobinopathy.

Hemolysis. Condition characterized by the destruction of red blood cells. Hemolysis can be caused by a number of disorders, such as immune diseases, drugs, poisons, and genetic disorders.

Hemolytic anemia. Anemia caused by hemolysis.

Hemophilia. Any of the genetic disorders characterized by insufficient production of coagulation factors.

Hereditary telangiectasia. A genetic disorder associated with areas of malformation and dilation (enlargement) of small blood vessels. Hereditary telangiectasias can affect any organ or other living tissue.

Hypercalcemia. Increased blood calcium. Normal serum calcium is about 8–10 mg/dl.

Idiopathic. Of unknown cause.

Immunoglobulin (Ig). A chemical produced by plasma cells that is part of the body's immune response to antigens. Immunoglobulins perform many specialized functions. The various types immunoglobulins are G, M, A, D, and E. These are abbreviated as IgG, IgM, IgA, IgD, and IgE. Also known as *antibodies*.

Infarction. Death of a piece of tissue as a result of insufficient blood flow.

Intracranial bleeding. Hemorrhage occurring within the skull. Intracranial bleeding inside the brain is called intracerebral bleeding.

Lesion. Abnormality.

Leukemia. Any of the white blood cell cancers arising in the bone marrow or lymph nodes. Specific leukemias are named according to which type of white cell is involved, such as lymphocytic leukemia and myelocytic leukemia. Leukemia is also classified as acute or chronic. Acute leukemias are those with the most cancerous cells, while chronic leukemias have more normal cells.

Leukocytes. See *white blood cells*.

Leukocytosis. Increased white blood cells, not specific in regard to what type of white cell is involved.

Lymph. A usually transparent liquid that comes from tissue fluids and also contains lymphocytes originating in lymph nodes.

Lymphangiography. X-rays of the lymphatic system following the injection of x-ray contrast material.

Lymphangitis. Inflammation of a lymphatic vessel as could occur with infection.

Lymphedema. Swelling of tissues caused by retention of lymph fluid.

Lymph nodes. Specialized collections of cells found in various locations along the system of lymph vessels.

Lymphoblasts. Immature *lymphocytes*.

Lymphocytes. Specialized white cells that play important roles in the function of the immune system involving both cells (cellular immunity) and antibodies (humoral immunity). Lymphocytes are found in the bone marrow, blood, and lymphatic system.

Lymphyocytopenia. Decreased numbers of lymphocytes in the blood.

Lymphocytosis. Increased numbers of *lymphocytes* in the blood or other fluid.

Lymphoma. Cancer of the lymph nodes and spleen that results in abnormal lymphocytes. Lymphoma can invade any organ of the body. Hodgkin's lymphoma and Non-Hodgkin's lymphoma are two important lymphomas.

Lymphoproliferative disorders. See *lymphomas*.

Lymphoscintigraphy. Method of visualizing lymph nodes by injection of a radioactive substance and making images of lymph nodes that concentrate the radioactivity. Lymphoscintigraphy is used for detecting spread of cancer to *lymph nodes*.

Macroglobulinemia. Disorders associated with the production of excessive amounts of immunoglobulin M antibody (IgM). Macroglobulinemia involves an abnormality of plasma cells (plasma cell dyscrasia).

Monocytes. A types of white blood cells with immune functions. For example, some monocytes change into macrophages. Macrophages engulf (eat) bacteria and other abnormal material.

Myelofibrosis. A disorder of several different causes associated with loss of normal bone marrow and replacement with fibrosis.

Myeloma. A form of cancer involving excess production of plasma cells by the bone marrow. Also known as *multiple myeloma*.

Myeloproliferative disorders. Disorders that involve the proliferation of bone marrow cells, in or outside of the marrow. For example, multiple myeloma and leukemia are myeloproliferative disorders.

Neutropenia. Decreased numbers of blood neutrophils.

Neutrophils. Type of white blood cell important in fighting infection.

Osteomyelitis. Infection of bone.

Osteosclerosis. Bone abnormality associated with myelofibrosis. Osteosclerosis may be seen on x-rays, as areas of increased bone density.

Packed red cells. Concentrated red blood cells for transfusion.

Peripheral blood. Blood that is circulating in the arteries and veins, outside of the bone marrow.

Petechiae. Pinpoint-sized spots of bleeding into the skin, which are red to purple in color. They may be a physical sign of thrombocytopenia.

Phlebotomy. The controlled removal of blood, usually from a vein.

Plasma. The clear liquid (noncell) part of blood before clotting.

Plasma cells. Type of white blood cell normally found in bone marrow and lymph nodes, but not circulating blood. Plasma cells make antibodies. (**Note:** plasma and plasma cells are not related.)

Plasma cell dyscrasias. Disorders involving abnormal plasma cell function, like excessive antibody production (such as macroglobulinemia) or abnormal pieces of antibodies (as in heavy chain disease).

Plasmapheresis. Method for removing substances from the blood, involving the following steps: (1) Blood is taken from the body; (2) the plasma is separated from the cells by spinning in a centrifuge; and (3) blood cells are reinjected along with fresh replacement plasma or albumin. Plasmapheresis is usually done on an outpatient basis. Plasmapheresis can either be used to remove unwanted substances from blood (such as elevated cholesterol) or to harvest plasma blood components for donation to patients who need them.

Platelet count. Normal platelet counts are about 150,000–300,000 per mm^3.

Polycythemia. Polycythemia rubra vera is a serious disease of the bone marrow resulting in erythrocytosis, leukocytosis, thrombocytosis, and increased blood volume. Also known as *primary* or *absolute polycythemia*. Far less serious is secondary polycythemia, where increased numbers of red cells in a body attempt to improve the oxygenation of tissues. Secondary polycythemia is frequently seen in people with advanced lung disease.

Purpura. Small spots of bleeding into the skin, red to purple in color. They are larger than a petechia, but not over about one centimeter in diameter. Purpura may be a sign of thrombocytopenia. Purpura may refer to only one spot or multiple spots. A purpura caused by trauma rather than decreased platelets is a bruise.

Red blood cells (RBCs). Cells made in the bone marrow that have the major function of oxygen transport by means of the hemoglobin they contain.

Reticulocytes. Immature forms of red blood cells.

Serum. The clear liquid part of blood after clotting.

Sickle cell anemia. An inherited red blood cell defect in which red cells are unusually susceptible to destruction. Sickle cell anemia is a type of hemoglobinopathy because it involves the production of an abnormal form of hemoglobin called sickle hemoglobin.

Sickle cell preparation. A fast screening test for sickle cell anemia.

Splenomegaly. Enlarged spleen.

Systemic. Affecting the whole body.

Thalassemia. Any of the disorders in which parts of the hemoglobin molecule are made at a decreased rate.

Thrombocytopenia. Decreased numbers of platelets.

Thrombocytosis. Increased numbers of blood platelets.

Thrombosis. Formation of a blood clot.

White blood cells (WBCs). White blood cells consist of neutrophils, lymphocytes, basophils, eosinophils, and monocytes. Also known as *leukocytes*.

Whole blood. Blood without alteration from its natural state.

B. General Information

Blood consists of red cells that carry oxygen; white cells, which are important in immunity and fighting infection; and platelets, which help prevent abnormal bleeding. In addition, the liquid part of blood (plasma) carries numerous substances, such as proteins (like albumin), vitamins, minerals, glucose, and fats. Blood is formed in the marrow of bone. Marrow is found in bone cavities, but is not present in all bones.

The lymphatic system circulates lymph fluid separately from the vascular system of blood vessels,

but lymph fluid does empty into the bloodstream in several locations. Lymph nodes stationed along lymphatic vessels function for the immune system and contain lymphocytes. Lymph nodes can trap and destroy bacteria. Lymph nodes can also catch cancerous cells that are being spread through the lymphatic system. Therefore, biopsy of lymph nodes is important in determining whether cancer has spread (metastasized) from the original tumor. Lymphangiography and lymphoscintigraphy are also useful tests in detecting lymphatic disease such as spreading cancer. Lymphangitis, cancer, and associated treatments and trauma can damage the lymphatic system and produce lymphedema. The swelling caused by lymphedema can result in functional limitations.

A prediction that a condition will last the required 12 months must be based on at least three months of medical documentation. Medical documentation of blood or lymphatic disorders always involves laboratory tests, and such tests must include the values reported on more than one examination over that three-month period.

C. Specific Listings and Residual Functional Capacity

The listings that follow are in the federal regulations. They have been interpreted and commented on for greater ease of understanding while explaining their requirements. It is impossible to discuss here all of the medical possibilities related to every kind of disorder, and you may need to seek help from your treating doctor to more fully understand how your particular impairment relates to these listings. The discussion of residual functional capacity does not apply to children.

1. Listing 7.02: Chronic Anemia (Adults)

The most frequent cause of anemia in the United States is iron deficiency in women. This disorder is seen frequently by the SSA, but is usually easily treated and is rarely a basis for disability. The many possible causes of anemia include infections, drugs, toxins, autoimmune diseases, vitamin deficiencies, genetic blood disorders such as sickle cell anemia, and cancer. This listing is applicable to any kind of anemia that is severe and lasts, or is expected to last, 12 months.

a. Listing Level Severity

For your condition to be severe enough to meet the listing, you must have chronic anemia due to any cause with a hematocrit persisting at 30% or less. Additionally, you must satisfy Ⓐ or Ⓑ, below.

Ⓐ You have had one or more blood transfusions on an average of at least once every two months. Whole blood or packed red cell transfusions qualify. To determine the frequency of the transfusions, divide the duration of your anemia in months by the number of incidents in which transfusion was required. Count one incident as one transfusion, even if more than one unit of blood was given. For example, say you were hospitalized twice in six months, requiring two units of blood during one hospitalization and one unit of blood during the other. Your anemia would not qualify under this listing because you cannot divide six months by your three blood transfusions and come up with an average of a transfusion every two months. Instead, you must count the two units during the first hospitalization as one transfusion, for an average of one transfusion every three months.

A need for transfusions to keep your hematocrit up to 30% implies that without the transfusions your anemia would be much worse. Also, your hematocrit will go down, reflecting a need for transfusion, and then rise up to a correct level after the transfusion. Because of the rise and fall, you will be considered more impaired than people with a steady hematocrit of 30%.

Ⓑ Other impairments associated with the anemia should be evaluated under the criteria of whatever listings are appropriate. For example, sickle cell anemia can cause strokes and should be evaluated under Listing 11.04 (CD Part 11).

b. Residual Functional Capacity

The major limiting factors in anemia are weakness and easy fatigability. This is because fewer red cells than needed are carrying oxygen to the body's

tissues. Also, the heart rate increases in an attempt to compensate for the anemia, meaning the heart has less reserve to handle exertion. For example, if your resting heart rate is increased to 100 beats per minute because of anemia, you've already used up a significant amount of your heart rate reserved for exercise. Still, anemia affects each person differently. A young person is less affected than an elderly one, as are people in better overall physical condition.

If your hematocrit is only a couple of percentage points below normal, you would not have any restrictions—assuming that is your only problem. On the other hand, if your hematocrit is only a couple of percentage points over that required by the listing—say 33%—but otherwise you qualify under Part Ⓐ, your RFC should be for no more than sedentary work. Of course, if you suffer from other impairments, the SSA should consider the effect of your anemia upon them. For example, the presence of significant anemia can markedly worsen the limitations imposed by heart or lung disease.

2. Listing 107.03: Hemolytic Anemia (Children)

The many possible causes of hemolytic anemia include drugs, autoimmune diseases, infection, and genetic disorders. Genetic disorders resulting in easily destructible red blood cells are a significant cause of hemolytic anemia in children. Such disorders may make the red cells more susceptible to rupture by affecting the outer membranes of red cells or by producing abnormal forms of hemoglobin within the red cells (hemoglobinopathies) or by causing deficiencies of important enzymes that red cells need to function. A common type of genetic hemolytic anemia resulting from hemoglobinopathy is sickle cell anemia; for children, that disorder is separately considered under Listing 107.05.

Reticulocytes, or immature red cells, are confined mostly to the bone marrow. When the bone marrow struggles to turn out new red cells to make up for those destroyed by hemolysis, however, reticulocytes are released into the blood stream in larger numbers. Normally, the reticulocyte count in peripheral blood is in the 0.5–2.5% range and averages about 1.5%. A percentage increase is a sign of continuing destruction of red cells and that the anemia has not been controlled. A reticulocyte count is part of a complete blood count (CBC).

Two important enzyme deficiencies in children causing hemolytic anemia are pyruvate kinase deficiency and glucose-6-phosphate dehydrogenase (G-6-PD) deficiency. You don't need to know the details; but if your doctor diagnoses your child with either disease, the disorder would be evaluated under this listing.

a. Listing Level Severity

For the child's condition to be severe enough to meet the listing, the child must have hemolytic anemia due to any cause, manifested by the persistence of a hematocrit of 26% or less despite prescribed therapy. Additionally, the child's reticulocyte count must be 4% or greater.

3. Listing 7.05: Sickle Cell Disease (Adults)

The SSA often sees sickle cell disease in both children and adults. All of the sickle cell diseases are hemoglobinopathies because they involve abnormal sickle hemoglobin (Hb S) in red blood cells. Sickle cell disease is also a type of hemolytic anemia because the Hb S makes red cells delicate and susceptible to easy destruction.

Normal adult hemoglobin is known as hemoglobin A. If you inherit genes for normal hemoglobin from both parents, all of your hemoglobin will be normal (Hb AA). Infants have fetal hemoglobin (Hb F), but this is normally rapidly replaced with adult hemoglobin.

If you inherit genes to make sickle hemoglobin from both parents, all of your hemoglobin will be abnormal (Hb SS) and you have sickle cell anemia. If one parent gives you a gene for normal hemoglobin (Hb A) and the other gives you a gene for sickle hemoglobin (Hb S), you will end up with half of each (Hb AS) and you will have sickle cell trait. The distinction between these two disorders is important, because sickle cell trait does not cause significant symptoms or limitations. Sickle cell anemia, on the other hand, can cause numerous and serious problems, which are considered by this listing.

There are other sickle cell diseases besides sickle cell anemia and sickle cell trait. A person could inherit Hb S from one parent and a different kind of abnormal hemoglobin from the other parent—for example, hemoglobin S and hemoglobin C (Hb SC). Hemoglobin SC disease results in abnormalities like sickle cell anemia, but generally less severe.

There is also a disorder called thalassemia. In thalassemia, the normal hemoglobin is made at a decreased rate. When a sickle hemoglobin gene is inherited form one parent and a thalassemia gene from the other, the result is fairly common disorder known as sickle thalassemia. Abnormalities are similar to sickle cell anemia, but generally milder.

A reliable way of diagnosing any type of sickle cell disease is by means of a test called hemoglobin electrophoresis. Hemoglobin electrophoresis separates and measures the types of hemoglobin present, using a blood sample. The SSA must have the results of hemoglobin electrophoresis to make an accurate diagnosis of sickle cell disease. A screening test called a sickle cell preparation is not acceptable as a means of diagnosing sickle cell disease.

a. Listing Level Severity

For your condition to be severe enough to meet the listing, you must have sickle cell disease or one of its variants. Additionally, you must satisfy Ⓐ, Ⓑ, Ⓒ, or Ⓓ, below.

Ⓐ Documented painful thrombotic crises (also known as vaso-occlusive crises) occurring at least three times during the five months before disability determination. The sickle shape of the abnormal red blood cells makes it difficult for them to move through blood vessels and blood flow may be further worsened by clotting in the affected vessel. Complete blockage of blood flow results in death of the tissue served by that vessel. The associated symptoms depend on the organ involved and the size of the infarction. Bone infarctions can be quite painful; infarctions of the brain produce strokes; infarctions of the heart may result in heart failure; infarctions of the lung can cause chest pain and pneumonia; and infarctions of the kidneys can result in kidney failure.

Bone infarction can be triggered by a number of things—most frequently exposure to cold—but also infection, dehydration, and pregnancy. Painful bone infarctions can involve the spine, breastbone, long bones in the arms and legs, the ribs, and pelvic bones. It is important that your treating doctor document your pain in your medical records.

Ⓑ Sickle cell disease requiring extended hospitalization (beyond emergency care) at least three times during the 12 months before disability determination. Hospitalization may be required due to aplastic episodes, in which the hematocrit falls rapidly because of the bone marrow's inability to produce enough red cells. Hospitalization might also be the result of hyperhemolytic crises, in which the unusually rapid destruction of red cells causes a sudden fall in hematocrit. Other possible complications to sickle cell disease that could result in hospitalization include strokes, heart attacks, heart failure, kidney failure, and pneumonia.

Ⓒ Chronic, severe anemia with persistence of hematocrit of 26% or less. The SSA should apply this requirement reasonably. For example, if your hematocrit values are consistently low with a single value of 27%, you should not be denied benefits. Remember that if your hematocrit is as high as 30%, you might still be granted benefits under Listing 7.02Ⓐ when you receive blood transfusions.

Ⓓ Evaluate the resulting impairment under the criteria for the affected body system. For example, a stroke would be evaluated under Listing 11.04 (CD Part 11) and a heart attack under the listings for heart disease (CD Part 4).

b. Residual Functional Capacity

The major limiting factors in anemia are weakness and easy fatigability. This is because fewer red cells than needed are carrying oxygen to the body's tissues. Also, the heart rate increases in an attempt to compensate for the anemia, meaning the heart has less reserve to handle exertion. For example, if your resting heart rate is increased to 100 beats per minute because of anemia, you've already used up a significant amount of your heart's reserve for exercise. Still, anemia affects each person differently. A young person is less affected than an elderly one, as are people in better overall physical condition.

If you have a hematocrit less than 30%, it is doubtful you could do more than sedentary work. If your sickle cell disease is sickle cell anemia, you should always avoid strenuous exertion and exposure to cold and your RFC should be no higher than light work. The SSA has no definite rules, however, and each case must be evaluated on an individual basis.

4. Listing 107.05: Sickle Cell Disease (Children)

The comments under Listing 7.05 apply here.

a. Listing Level Severity

For the child's condition to be severe enough to meet this listing, the child must have sickle cell disease or one of its variants. Additionally, the child's condition must satisfy Ⓐ, Ⓑ, Ⓒ, Ⓓ, or Ⓔ, below.

Ⓐ Recent, recurrent, severe vaso-occlusive crises involving the musculoskeletal system (including the spine) or abdominal organs (liver, spleen, etc.). This is similar to part Ⓐ under Listing 7.05. The SSA does not define recent or recurrent. Medical judgment must be applied individually, depending on how much the disorder affects the child's ability to function. The more severe and prolonged the crises, the fewer episodes would be required to be disabling. Using adult Listing 7.05 as a guide, the child would have to have at least three episodes during the five months before disability determination.

Ⓑ A major complication involving a large organ during the 12 months before application for disability benefits. Complications might include meningitis, lung infection (pneumonia) or infarction, osteomyelitis, heart failure, kidney infarction, liver infarction, intestinal infarction, or cerebrovascular accident. A major complication would ordinarily be one that required hospitalization, although the listing includes no such requirement. The fact that the child may have improved by the time you apply for benefits does not prevent qualification. For example, the child may be doing well with a major complication six months in the past. But the child would still be entitled to disability because the 12-month allowance window is still in force.

Ⓒ A hyperhemolytic or aplastic crisis within 12 months before the application for disability benefits. The fact that the child may have improved by the time you apply for disability does not prevent qualification.

Ⓓ Chronic, severe anemia with persistence of hematocrit of 26% or less. This is the same as part Ⓒ under adult Listing 7.05.

Ⓔ Congestive heart failure, strokes, or emotional disorders as described under the criteria in the child listing for heart failure (104.02, CD Part 4), the appropriate child neurological listings (CD Part 11), or the appropriate child mental disorder listings (CD Part 12). This is essentially the same as part Ⓓ under adult Listing 7.05.

5. Listing 7.06: Chronic Thrombocytopenia (Adults)

Several disorders, including leukemia, myelofibrosis, drugs, toxic substances, immune diseases, and genetic disorders, can cause decreased platelets. This lowered platelet count (thrombocytopenia) can result in repeated episodes of serious, even life-threatening, bleeding. Furthermore, idiopathic thrombocytopenia is of unknown cause. Platelets circulating in the blood help prevent serious bleeding by sticking together at sites where bleeding tries to start; they are also important in starting the coagulation process. Platelet counts are easy to make with a sample of blood and are routinely done with a complete blood count (CBC). Spontaneous bleeding starts without any traumatic cause.

a. Listing Level Severity

For your condition to be severe enough to meet the listing, you must have chronic thrombocytopenia due to any cause, with platelet counts repeatedly below 40,000 per mm^3. Additionally, you must satisfy Ⓐ or Ⓑ, below.

Ⓐ At least one episode of spontaneous bleeding within five months of the date of disability determination requiring transfusion. The transfusion could be either packed red blood cells or whole blood. That you have improved after the transfusion does not prevent you from qualifying, as long as the transfusion was within

five months of your application for disability benefits. Spontaneous bleeding is especially likely if the platelet count falls below 40,000 per mm^3. A platelet count this low is life-threatening and should receive immediate treatment.

Ⓑ Intracranial (within the head) bleeding within 12 months before disability determination. The severity of the intracranial bleeding or the degree to which you may have recovered when applying for disability does not matter, as long as the event occurred within the prior 12 months.

b. Residual Functional Capacity

The danger with thrombocytopenia is bleeding. Even though you may have the strength to do heavy work, to do so is probably too dangerous. Doing heavy lifting or working around heavy equipment occasionally results in physical trauma. What might ordinarily result in only a bruise in a normal person can cause severe internal bleeding if your platelet count is low. This is particularly true for blows to the head. Even if a job does not involve heavy work, you should not perform work that has a significant danger of physical trauma such as falling or being cut. If your platelet count is only modestly decreased—say 100,000 mm^3—you wouldn't need any restrictions because your chance of bleeding is not increased.

6. Listing 107.06: Chronic Idiopathic Thrombocytopenic Purpura (Children)

The comments about thrombocytopenia under Listing 7.06 are relevant to child applicants, although they are not exactly the same. Unlike the adult listing, this child listing concerns only one disorder involving a low platelet count—idiopathic thrombocytopenic purpura (ITP). ITP is of unknown cause, and like other types of thrombocytopenia, may involve bleeding into the skin to produce purpura. In most cases of child ITP, the child responds to treatment with steroid drugs.

a. Listing Level Severity

For the child's condition to be severe enough to meet the listing, the child must have chronic idiopathic thrombocytopenic purpura and platelet counts of 40,000 per mm^3 or less despite prescribed therapy or recurrent upon withdrawal of treatment.

7. Listing 7.07: Hereditary Telangiectasia (Adults)

Hereditary telangiectasia is a disorder associated with a tendency to form abnormal blood vessels that can affect any organ or other living tissue. The danger of this disorder is that life-threatening bleeding may occur, such as hemorrhaging in the lungs, intestine, or brain. The severity of bleeding episodes is established for disability purposes by the need of transfusion of blood. Transfusion could be either whole blood or packed red blood cells.

a. Listing Level Severity

For your condition to be severe enough to meet the listing, you must have hereditary telangiectasia with bleeding requiring transfusion at least three times during the five months before your disability determination.

b. Residual Functional Capacity

Your RFC must be evaluated on a case-by-case basis, applying medical judgment to the number of bleeding episodes, their severity, and the types of activities associated with the bleeding, as well as the size, location, and number of telangiectasias. For example, if lifting 20 pounds triggers an episode of bleeding from the intestine, your RFC should not be higher than sedentary work. In general, if you have significant telangiectasia in any major organ, you'll want an RFC for no more than medium work because the increased blood pressure associated with strenuous exertion might cause serious bleeding.

8. Listing 7.08: Hemophilia and Other Coagulation Disorders (Adults)

Coagulation is the ability of the blood to form clots, and is caused by a long complex chain of chemical reactions involving a number of proteins in the blood called coagulation factors. Coagulation factors are made in the liver.

The various types of hemophilia are caused by a deficient production of some type of coagulation

factor. For example, hemophilia A is caused by a coagulation factor VIII deficiency, hemophilia B by a factor IX deficiency, and hemophilia C by a factor XI deficiency. About 80% of hemophiliacs have hemophilia A, as do most disability applicants.

The treatment for hemophilia is periodic administration of the deficient coagulation factor. Periodic transfusion of the missing coagulation factor to prevent bleeding is not the same as a transfusion to replace blood loss as required by the listing. Transfusions to replace blood loss from bleeding may be either in the form of whole blood or packed red blood cells.

A spontaneous hemorrhage as required by the listing is one in which there is bleeding without any traumatic cause. The listing concerns only the severity of the bleeding itself caused by a coagulation disorder. If your bleeding damages an organ, such as the brain, evaluation would also involve other appropriate listings. Bleeding into a joint space is known as hemarthrosis and can result in severe arthritis and functional loss of a limb.

Hemophilias are not the only type of coagulation disorders that could potentially qualify under this listing. Some coagulation disorders are not caused by a deficiency of a coagulation factor, but by an abnormality of the coagulation system. For example, some hemophiliacs develop excessive amounts of an antibody that acts as an inhibitor to the needed coagulation factor and causes an increased tendency to bleed. Advanced liver disease interferes with the production of coagulation factors and such individuals may have a tendency to bleed excessively, but these cases can be allowed under Listing 5.05 for chronic liver disease (CD Part 5).

The presence of a coagulation disorder must be proven by the measurement of coagulation factors (coagulation factor assay), abnormal coagulation times, or other appropriate tests. If you have a coagulation disorder, you would already have had the needed diagnostic work-up and the SSA will require that information from your treating doctor.

a. Listing Level Severity

For your condition to be severe enough to meet the listing, you must have coagulation defects (hemophilia or a similar disorder) with spontaneous hemorrhage requiring transfusion at least three times during the five months before disability determination.

b. Residual Functional Capacity

Evaluation must be done on an individual basis, taking into account not only bleeding but any other permanent damage done to the body by previous episodes of bleeding. For example, bleeding into a knee joint can result in arthritis that markedly limits ability to stand and walk, especially with repeated episodes of bleeding into the joint. Inability to stand and walk at least six to eight hours daily limits an RFC to no more than sedentary work with further limitation on the use of leg controls. Even if you have the strength to do heavy work, to do so is probably too dangerous: it is difficult to do heavy lifting or work around heavy equipment without occasional physical trauma. What might ordinarily result in only a bruise in a normal person can result in severe bleeding with a coagulation disorder like hemophilia. This is particularly true for blows to the head. Even if a particular job is not heavy work, you should not be performing it if there is significant danger of physical trauma such as falling or being cut.

9. Listing 107.08: Inherited Coagulation Disorders (Children)

The comments under Listing 7.08 apply here.

a. Listing Level Severity

For the child's condition to be severe enough to meet the listing, the child must have an inherited coagulation disorder. Additionally, the child's condition must satisfy Ⓐ or Ⓑ, below.

Ⓐ Repeated spontaneous or inappropriate bleeding. Inappropriate bleeding means bleeding starting as a result of trauma (such as a mild bump or fall) that would not cause bleeding in a normal person or that results in excessive bleeding. The SSA does not define repeated. Medical judgment must be applied case by case, taking into consideration the frequency and severity of bleeding episodes and whether severity is likely to fulfill the 12- month requirement for disability.

Ⓑ Bleeding into a joint with resulting joint deformity. Marked deformities would obviously qualify—such

as fixation of the joint in a particular position (ankylosis), destruction of the joint, determined by x-ray, or instability of the joint requiring a brace to walk; however, this listing does not say that a marked joint deformity is necessary. It is reasonable to expect that significant—more than slight—joint damage is required to produce a deformity, and that the child would have some difficulty walking. But the SSA should not assert that the child needs a brace or crutch to walk or that the joint be unable to move—the listing does not specify that degree of required severity.

10. Listing 7.09: Polycythemia Vera (Adults)

Polycythemia vera is a serious disease of the bone marrow resulting in abnormally increased numbers of red blood cells, white blood cells, platelets, and blood volume. Also, the patient usually has an enlarged spleen.

Because of the increased number of red blood cells, the hematocrit is abnormally high in polycythemia. This isn't much of a problem until the hematocrit reaches about 60%. At that point the viscosity (self-stickiness) of the blood greatly increases and you are in danger of blood vessel blockage somewhere in the body. If blood vessel blockage happened in the brain, for example, you'd suffer a stroke. Similarly, you might experience a heart attack from blockage of a coronary artery.

Even without blockage of a blood vessel, a high red cell count indicated by hematocrits of 60% or more and high blood volume can stress any organ, including the heart and vascular system. Several neurological symptoms can result from abnormal blood circulation through the brain.

The most frequent treatment for polycythemia vera is phlebotomy. Such blood removal keeps the blood volume and hematocrit from getting too high. If the red cell count and blood volume can be kept in normal ranges, symptoms and dangers are minimized. Measurement of the hematocrit is an easy way to monitor the need for phlebotomy.

If phlebotomy fails, treatments that intentionally damage part of the bone marrow so it won't produce excessive blood cells may be administered. Radioactive phosphorus has been used for this purpose, but carries an increased risk of cancer.

Just having polycythemia vera does not result in a granting of disability. The severity of the impairment depends on the kind of damage done to the body. The diagnosis of polycythemia vera requires measurements of the blood volume. These measurements are exacting and require the use of radioactive substances (radionuclides) to tag red blood cells and plasma. Your treating doctor should have done such tests if you've been diagnosed with polycythemia vera.

Far less serious than polycythemia vera is the secondary polycythemia of people with increased numbers of red cells in the body's attempt to improve the oxygenation of tissues. Secondary polycythemia is frequently seen in people with advanced lung disease; it is not considered under this listing, however, and is much more common than polycythemia vera. Do not confuse the two disorders.

a. Listing Level Severity

For your condition to be severe enough to meet the listing, you must have polycythemia vera with erythrocytosis, splenomegaly, and leukocytosis or thrombocytosis. The SSA will evaluate any resulting impairment under the listings dealing with the affected body system.

b. Residual Functional Capacity

Medical judgment must be applied to individual cases, taking into account the response to treatment and the nature and extent of major organ damage. Even without organ damage, the presence of increased blood volume (indicated by a high hematocrit, especially of 60% or more) can produce headaches, weakness, and easy fatigability. It is important that you have your symptoms clearly documented, especially regarding your functional limitations and preferably in your treating doctor's medical records.

11. Listing 7.10: Myelofibrosis (Adults)

Myelofibrosis (myeloproliferative syndrome) means a change of normal bone marrow into fibrotic tissue. Normally, bone marrow consists of large numbers of cells—red cells, white cells, plasma cells, and platelets. The bone marrow supplies the cells for the blood stream. Myelofibrosis can be caused by

toxic substances, cancer, malnutrition, infection, radiation, or may be of unknown cause. Fortunately, myelofibrosis is not a common disorder.

The three most serious complications of myelofibrosis are:

- anemia secondary to the loss of red blood cells
- recurrent bacterial infections secondary to decreased white blood cells, and
- bone pain created by a bone abnormality called osteosclerosis.

Myelofibrosis must be diagnosed by a bone marrow biopsy. Your treating doctor must perform the biopsy; the SSA will not order this test because it is invasive.

a. Listing Level Severity

For your condition to be severe enough to meet the listing, you must have Ⓐ, Ⓑ, or Ⓒ, below,

Ⓐ Chronic anemia. Evaluate according to Listing 7.02.

Ⓑ Documented recurrent systemic bacterial infections occurring at least three times during the five months before disability determination.

Ⓒ Severe and intractable bone pain with x-ray evidence of osteosclerosis.

b. Residual Functional Capacity

No absolute rules can be given. Early myelofibrosis might produce no symptoms or limitations, but at a severe stage you could be restricted to as low as sedentary work. Your symptoms and resulting functional limitations need to be documented in your treating doctor's medical records. If you have anemia, see the RFC discussion under Listing 7.02.

12. Listing 7.15: Granulocytopenia (Adults)

Granulocytopenia refers to low levels of certain types of white blood cells called granulocytes, especially neutrophils. Neutrophils are important in fighting infection, and a low neutrophil blood count is specifically referred to as neutropenia. Granulocytopenia might be caused by infection, toxic substances, or drugs, or be of unknown cause.

a. Listing Level Severity

For your condition to be severe enough to meet the listing, you must have chronic granulocytopenia (due to any cause). Additionally, you must satisfy Ⓐ and Ⓑ, below.

Ⓐ Absolute neutrophil counts repeatedly below 1,000 cells/cubic millimeter of blood (1,000 neutrophils per mm^3). This number is significant, because when the neutrophil count falls that low, you are in great danger of life-threatening systemic infections. Although neutrophils are a type of white blood cell (WBC), the neutrophil count should not be confused with the total white blood cell count. If the number of neutrophils in laboratory reports are expressed as a percentage of the total white cell count, the absolute neutrophil count is simply that percentage multiplied by the total white cell count. For example, if your total WBC count is 5,000 per mm^3 and your neutrophil count is 10% of the total, then your absolute neutrophil count is 500 per mm^3. "Absolute" merely emphasizes that an actual count of neutrophil cells is considered, rather than neutrophils as a percentage of the total white cells. Knowing this, it is easy for you to determine your neutrophil count by obtaining your laboratory work from your hospital or treating doctor. The absolute neutrophil count is easily done on a blood sample.

Ⓑ Documented recurrent systemic bacterial infections occurring at least three times during the five months before disability determination. Such systemic infections would require treatment in a hospital with intravenous antibiotics. Cultures of blood that show the presence of bacteria are irrefutable evidence of systemic infection. Such blood cultures would be standard practice to treat systemic bacterial infection, so that information should be available to the SSA.

b. Residual Functional Capacity

Cases must be evaluated on an individual basis, taking into account the effects of any residual impairment including weakness, fatigue, and side effects of treatment. During any time you are not suffering the effects of infection or treatment, you might not have any limitations, given that granulocytopenia does not, in itself, produce symptoms. It is quite possible, however, to have a granulocytopenia in which there is

also associated anemia. If so, such anemia would be evaluated under Listing 7.02 and the RFC discussion under that listing would also apply here.

13. Listing 7.17: Aplastic Anemia With Bone Marrow or Stem Cell Transplant (Adults)

Bone marrow transplants are sometimes necessary to treat myelofibrosis and other causes of aplastic anemia that result in loss of bone marrow. Leukemias or other cancers that destroy the bone marrow may also require a transplant, but are considered under the appropriate cancer listings discussed in CD Part 13. The SSA has no bone marrow transplant listing for children, but a child who has a transplant can be considered under this listing. Even if you do very well after a marrow or stem cell transplant, you are still entitled to the full year of disability.

a. Listing Level Severity

For your condition to be severe enough to meet the listing, you must have aplastic anemia with bone marrow or stem cell transplantation. The SSA will consider you to be under a disability for 12 months following transplantation. Thereafter, you will be evaluated according to the type of impairment remaining.

b. Residual Functional Capacity

See discussion of RFC under whatever listings are used to evaluate residual impairment after transplantation. ■