ICD-10-CM Documentation and Coding Best Practices
Hematological Disorders

Myeloproliferative Neoplasms

A group of neoplasms in which the bone marrow makes too many RBCs, platelets or certain WBCs.

Myeloproliferative disease, NOS *(D47.1)*

Polycythemia vera *(D45)* – a slow-growing blood cancer in which the bone marrow makes too many RBCs. Excess RBCs thicken the blood, slowing its flow. Complications, such as blood clots, can lead to heart attack or stroke. There is also a risk of progressing to more serious blood cancers including myelofibrosis or acute leukemia.

Etiology

Occurs due to a mutation in the protein Janus kinase 2 (JAK2). The cause of this mutation is unknown, but it generally is not inherited.

Treatment

- **Phlebotomy** is the first treatment option. This reduces the number of blood cells and decreases blood volume.
- **Low dose aspirin** reduces the risk of blood clots
- **Hydroxyurea (Droxia, Hydrea)** suppresses bone marrow's ability to produce blood cells
- **Interferon alpha** stimulates immune system to fight the overproduction of RBCs
- **Ruxolitinib (Jakafi)** helps immune system destroy cancer cells

Sideroblastic anemias *(D64.0-D64.3)*

While anemia is commonly caused by iron deficiencies, in sideroblastic anemia the body has enough iron but is unable to use it to make hemoglobin. Iron accumulates in the mitochondria of RBCs, giving a ringed appearance to the nucleus (ringed sideroblast).

Etiology

Can be inherited or acquired. Acquired forms may be caused by myelodysplastic syndrome, nutritional deficiencies (copper, vitamin B6), zinc overdose, lead poisoning, drugs (antibiotics, hormones, and chemotherapy agents), alcohol, and hypothermia.

Treatment

- **Reversible causes** – In cases of drug and toxin-induced sideroblastic anemia, avoidance of the drug or removal of the toxin can lead to recovery. In hypothermia-induced cases, normalization of temperature reverses the sideroblastic changes.
- **Vitamin B6 (pyridoxine)** – can be beneficial in both congenital and acquired cases.
- **Zinc and Alcohol Avoidance** – All individuals with sideroblastic anemia should avoid zinc-containing supplements and the use of alcohol.
- **Blood Transfusion** – can be useful when vitamin B6 therapy is ineffective. However, since it has been known to worsen iron overload, this option should be carefully considered.
- **Bone Marrow Transplant** – may be utilized when all other options have been exhausted

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Hereditary Hemolytic Anemias

Hemolytic anemias occur when RBCs are destroyed faster than they can be made. These can be inherited or acquired. In hereditary forms, mutations in the genes that control RBC production lead to problems with the *hemoglobin*, *cell membrane* or *enzymes* that maintain healthy RBCs. Abnormal cells may be fragile and break down while moving through the bloodstream.

Thalassemia *(D56.-)* – the body produces abnormal *hemoglobin* molecules resulting in abnormal RBCs with a reduced ability to carry oxygen to the tissues. Sign and symptoms vary depending on disease type and severity. In minor cases, patients may be asymptomatic, or they may experience a general fatigue and weakness.

Complications

- **Iron overload –** Too much iron in the blood can be a result of the disease itself, or frequent blood transfusions. Iron overload results in damage to the heart, liver and endocrine system.
- **Infection –** Thalassemia results in an increased risk of infection. This is especially true if the spleen has been removed.
- **Splenomegaly –** The spleen fights infection and filters old and damaged blood cells. Because thalassemia involves the destruction of large numbers of RBCs, it can cause the spleen to work harder and enlarge. Splenomegaly can make anemia worse, and it can reduce the life of transfused RBCs.

Treatment

- **Blood transfusions –** Severe forms often require frequent blood transfusions, possibly every few weeks. Since blood transfusions cause a buildup of iron, *deferasirox (Exjade, Jadenu)* may be required to rid of the body of extra iron.
- **Splenectomy –** may be required if the spleen becomes too large.

Anemia due to G6PD deficiency *(D55.0)* – Also known as *Favism*. The absence of an important *enzyme* called G6PD causes RBCs to rupture and die when they are exposed to certain substances in the bloodstream. Factors that can trigger the breakdown of RBCs include:

- fava beans
- sulfa drugs
- exposure to naphthalene (found in mothballs)
- antimalarial medications
- viral or bacterial infections

Treatment

- **Avoidance –** the main treatment is avoidance of those factors that would contribute to episodes of hemolysis.
- **Transfusions –** Episodes of hemolysis that occur after exposure are usually short-lived, and anemia is rarely severe enough to warrant blood transfusions.

Coding and Documentation Guidance

In order to code to the highest level of specificity, please include details such as:

- Acute or Chronic
- For anemias, specify the type: nutritional, hemolytic, aplastic or as a result of blood loss
- For hemolytic anemia, specify whether hereditary, acquired, enzyme disorder, autoimmune or non-autoimmune
- Underlying cause of “unknown cause”
- Link laboratory findings to the anemia diagnosis if known
- Specify if anemia is associated with a malignancy