

Guidance on Documentation and Coding for Aplastic Anemia

Overview

Aplastic/Aplasia is characterized by the failure of an organ or tissue to develop or to function normally. Anemia is a condition in which you lack enough healthy red blood cells to carry adequate oxygen to your body's tissues.

Types of aplastic anemia and other bone marrow failure syndromes

Aplastic Anemia, also known as idiopathic aplastic anemia, is bone marrow failure characterized by underproduction of red blood cells, white blood cells, and platelets.

Pure Red Cell Aplasia (PRCA) is a rare bone marrow disorder characterized by an isolated decline of red blood cells (erythrocytes) produced by the bone marrow.

Acquired Pure Red Cell Aplasia is bone marrow failure characterized by underproduction of red blood cells while white blood cell and platelet production remains normal. This disorder can have multiple causes including:

- A parvovirus B19 infection, which may be toxic to the red blood cells starters (or precursors).
- Certain lymphocytic leukemias (blood cancers) such as large granular lymphocytic leukemia (LGLL).
- Thymoma (a tumor of the thymus, which is a lymphoid organ).
- Autoimmune diseases such as Rheumatoid Arthritis or Lupus.
- Common variable immunodeficiency (CVID).

Blackfan-Diamond Anemia (or Diamond-Blackfan Anemia) is a rare blood disorder that occurs when the bone marrow fails to make red blood cells, which are essential for carrying oxygen from the lungs to all the other parts of the body. DBA is a genetic disease that affects the body's ribosomes, which are small cellular structures that play an important role in building proteins in the body.

Fanconi's Anemia (FA) is a rare disease passed down through families (inherited). Mutations in at least 15 genes can cause Fanconi anemia, but 80-90% of cases are due to mutations in one of three genes—FANCA, FANCC, and FANCG. It is most often inherited in an autosomal recessive pattern, which means both copies of the gene in each cell have mutations.

Drug-Induced Aplastic Anemia is one of the few life-threatening reactions to drugs. Although most reported cases have been associated with Chloramphenicol, many drugs have the potential to be toxic to the bone marrow. There are two distinct types of toxicity with differing pathogenic mechanisms — a dose-related reversible marrow aplasia and a dose independent idiosyncratic aplasia with a high mortality.

Myelophthisis is a condition that occurs when normal hematopoietic tissue in the bone marrow is replaced with abnormal tissue, such as fibrous tissue or tumors.

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Signs and symptoms may include:

- Damage to vital organs as a result of iron overload from transfused red blood cells.
- Severe viral, bacterial, and fungal infections.
- Heart arrhythmia and heart failure.
- Disease progression to myelodysplastic syndrome or acute leukemia.

Complications of:

- Nosebleeds and bleeding gums.
- Fatigue/lethargy and abnormal paleness of the skin.
- Head, face, and neck abnormalities.
- Sensitivity to cold temperatures.

Treatment options of:

- Corticosteroids, immunoglobins, immunosuppressive therapy, chelations therapy.
- Blood transfusions.
- Removal of the thymus.
- Bone marrow transplants.
- Stem cell transplants to rebuild the bone marrow.

ICD-10-CM Code information

D60.0 Chronic acquired pure red cell aplasia

D60.8 Other acquired pure red cell aplasias

D60.9 Acquired pure red cell aplasia, unspecified

D61.01 Constitutional (pure) red blood cell aplasia

D61.09 Other constitutional aplastic anemia

D61.2 Aplastic anemia due to other external agents

- Coding Guideline: Code first, if applicable, toxic effects of substances chiefly nonmedicinal as to source (T51-T65).

D61.3 Idiopathic aplastic anemia

D61.82 Myelophthisis

- Coding Guideline: Code also the underlying disorder, such as: Malignant neoplasm of breast (C50.-); Tuberculosis (A15.-)

D61.89 Other specified aplastic anemias and other bone marrow failure syndromes

Documentation guidance for metabolic syndrome

Provider documentation should include elements of:

- Patient questions/answers such as:
 - Have you had recent infections?
 - Have you bled unexpectedly?
 - Are you more tired than usual?
- Detailed and through physical exam.
- Ordering of tests and results such as:
 - CBC with differential.
 - A reticulocyte and lymphocyte count.
 - Measuring the monoclonal proteins in the blood,
 - Bone marrow aspiration.
- Detailed PMH, including any prior infections that could be precursors.
- Note if patient has any autoimmune diseases that can cause these disorders (i.e. Rheumatoid Arthritis or Lupus).

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References

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- “Pure Red Cell Aplasia.” Cleveland Clinic, Cleveland Clinic, 15 April 2019, <https://my.clevelandclinic.org/health/diseases/14475-pure-red-cell-aplasia-prca>
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- “Drug-Induced Aplastic Anemia: Pathogenesis and Clinical Aspects.” Pub Med, National Library of Medicine, <https://pubmed.ncbi.nlm.nih.gov/2285121/>
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