

ICCS Guidelines identify patient populations at high risk for PNH

ICCS Clinical Indications for PNH Testing

Intravascular hemolysis as evidenced by hemoglobinuria or elevated plasma hemoglobin

Evidence of unexplained hemolysis with accompanying:

- Iron deficiency, OR
- Abdominal pain or esophageal spasm, OR
- Thrombosis (see below), OR
- Granulocytopenia and/or thrombocytopenia

Other acquired Coombs'-negative, non-schistocytic, non-infectious hemolytic anemia

Thrombosis with unusual features:

- Unusual sites
 - Hepatic veins (*Budd-Chiari syndrome*)
 - Other intra-abdominal veins (*portal, splenic, splanchnic*)
 - Cerebral sinuses
 - Dermal veins
- With signs of accompanying hemolytic anemia (see above)
- With unexplained cytopenia

Evidence of bone marrow failure:

- Suspected or proven aplastic or hypoplastic anemia
- Refractory cytopenia with unilineage dysplasia
- Other cytopenias of unknown etiology after adequate workup

Relevant ICD-10 Codes for high-risk patients

Cytopenia

- D61.818 Other pancytopenia
- D69.6 Thrombocytopenia, unspecified
- D70.9 Neutropenia, unspecified (D70.9 is assigned for unspecified granulocytopenia)

Aplastic anemia

- D60.1 - D60.9 Acquired pure red cell aplasia
- D61.01 - D61.9 Other aplastic anemias and other bone marrow failure syndromes

Myelodysplastic Syndromes

- D46.C Myelodysplastic syndrome with isolated del(5q) chromosomal abnormality
- D46.9 Myelodysplastic syndrome, unspecified
- D46.Z Other myelodysplastic syndromes

Hemolytic anemia

- D59.8 Other acquired hemolytic anemias
- D59.9 Acquired hemolytic anemia, unspecified

Thrombosis

- I67.6 Nonpyogenic thrombosis of intracranial venous system (I67.6 is assigned for thrombosis of the cerebral sinuses)
- I74.8 Embolism and thrombosis of other arteries (I74.8 is assigned for less common sites of arterial thrombosis and excludes the aorta, arteries of the extremities, and the iliac artery)
- I81 Portal vein thrombosis (I81 is also assigned for mesenteric thrombosis) (I81 is also assigned for hepatic vein thrombosis)
- I82.0 Budd-Chiari syndrome
- I82.890 Acute embolism and thrombosis of other specified veins (I82.890 is assigned for splenic and dermal vein thrombosis)

Hemoglobinuria

- D59.6 Hemoglobinuria due to hemolysis from other external causes (D59.6 is assigned for acquired hemolytic anemia with hemoglobinuria)
- R82.3 Hemoglobinuria

The listing of ICD 10 codes are intended as educational information for health care providers. It does not replace a health care provider's professional judgment or clinical diagnosis.

Reference: Borowitz MJ, et al. *Cytometry B Clin Cytom.* 2010;78(4):211-230.

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