CANADIAN PNH NETWORK

Partners in Care for PNH

PNH Destroys

To C.A.T.C.H. PNH, know who's at risk.

Paroxysmal nocturnal hemoglobinuria:
A fatal blood disorder that results in the destruction of red blood cells.

Screen your patient. Early intervention is critical.[1]

International Clinical Cytometry Society (ICCS) Guidelines and the International PNH Interest Group (I-PIG) recommend evaluation of the following higher-risk patient populations:[2,3]

References: **1.** Santarone S, et al. Hematopoietic stem cell transplantation for paroxysmal nocturnal hemoglobinuria: long-term results of a retrospective study on behalf of the Gruppo Italiano Trapianto Midollo Osseo (GITMO). Haematologica 2010;95:983–988. **2**. Borowitz MJ, et al. Guidelines for the diagnosis and monitoring of paroxysmal nocturnal hemoglobinuria and related disorders by fl ow cytometry. Cytometry B Clin Cytom 2010;78:211–230. **3.**Parker C, et al. Diagnosis and management of paroxysmal nocturnal hemoglobinuria. Blood 2005;106:3699–3709. **4.** Movalia M, et al. Poster presented at the 53rd Annual Meeting of the American Society of Hematology; December 10-13, 2011; San Diego, CA. Abstract 1033.

Cytopenia, unexplained	AA/RA-MDS	Thrombosis, unexplained	Coombs -negative hemolytic anemia	Hemoglobinuria
Incidence of PNH clone[4]*				
5.7%	26.3% / 5.5%†	1.4%	22.7%	18.9%

Rule PNH in or out using high-sensitivity flow cytometry‡ and comprehensive clinical assessment

AA = aplastic anemia; LDH = lactate dehydrogenase; LLN = lower limit of normal; RA-MDS = refractory anemia-myelodysplastic syndromes; ULN = upper limit of normal.
* Study description: An analysis of the incidence of PNH clones in 6897 patients recommended for testing according to guidelines from the ICCS and the I-PIG.
† Includes all MDS subtypes.
‡ 0.01% PNH cell threshold.

C Cytopenia, unexplained

Unexplained cytopenias
Where a bone marrow test would be performed

... With evidence of hemolysis

- LDH >ULN

OR

- Haptoglobin <LLN

OR

- Elevated reticulocyte count (with or without anemia)

OR

... With coexisting findings

- Thrombosis
- Anemia
- Coombs-negative hemolytic anemia
- Bone marrow failure disorder
- Hemoglobinuria

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Cytopenia, unexplained	AA/RA-MDS	Thrombosis, unexplained	Coombs -negative hemolytic anemia	Hemoglobinuria

A Aplastic anemia (AA) or refractory anemia-myelodysplastic syndromes (RA-MDS)

Aplastic anemia

All patients who have ever had aplastic anemia – ideally at diagnosis and then annually thereafter

Low-risk or intermediate-1 MDS

- Hypoplastic bone marrow
- High serum erythropoietin (>500)

OR

RA-MDS with evidence of hemolysis

- LDH >ULN

OR

- Haptoglobin <LLN (with or without anemia)

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T Thrombosis, unexplained

Unexplained thrombosis

- Despite anticoagulation
- In young patients (<50 years)
- In unusual sites (e.g., Budd-Chiari, cerebral, mesenteric, dermal, subclavian)

... With evidence of hemolysis

OR

- LDH >ULN

OR

- Haptoglobin <LLN

OR

- Elevated reticulocyte count (with or without anemia)

... With coexisting findings

OR

- Anemia
- Cytopenia
- Coombs-negative hemolytic anemia
- Bone marrow failure disorder
- Iron deficiency
- Hemoglobinuria

... With other clinical manifestations

- Abdominal pain
- Chest pain
- Dyspnea
- Dysphagia
- Severe fatigue

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C Coombs-negative hemolytic anemia

- Screen all patients with otherwise unexplained Coombs-negative hemolytic anemia

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H Hemoglobinuria

- Screen all patients with otherwise unexplained hemoglobinuria
- All patients with hematuria should be tested for hemoglobinuria

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EE-2753-11-25-RARE