CANADIAN PNH NETWORK
Partners in Care for PNH

PNH Destroys

Here's who can help.

The PNH Network is a nationwide network of experts dedicated to the diagnosis, treatment, management, education and investigation of Paroxysmal Nocturnal Hemoglobinuria (PNH), for the benefit of Canadian patients and the Canadian healthcare system.

Learn more about us at **www.PNHnetwork.ca**

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 flow 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.

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

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)

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
- LDH >ULN

OR
- Haptoglobin <LLN

OR
- Elevated reticulocyte count (with or without anemia)

OR

... With coexisting findings
- Anemia
- Cytopenia
- Coombs-negative hemolytic anemia
- Bone marrow failure disorder
- Iron deficiency
- Hemoglobinuria

OR

... With other clinical manifestations
- Abdominal pain
- Chest pain
- Dyspnea
- Dysphagia
- Severe fatigue

C Coombs-negative hemolytic anemia

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

H Hemoglobinuria

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

Cytopenia, unexplained	AA/RA-MDS	Thrombosis, unexplained	Coombs-negative hemolytic anemia	Hemoglobinuria
Incidence of PNH clone4*				
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.