CANADIAN PNH NETWORK
Partners in Care for PNH

STANDARDIZED REPORTING TEMPLATE FOR PNH FLOW CYTOMETRY

INTERPRETATION

Cytometric analysis demonstrates no (undetectable) **populations of White/Red Blood Cells with deficiency of GPI-linked proteins** (i.e. populations of cells with an immuno-phenotype associated with the pathophysiology of paroxysmal nocturnal hemoglobinuria [PNH]).

Additional comments

GPI-Deficient Populations	Current Assessment	Previous Assessment Input Accession Date (DD-MM-YY)	Previous Assessment Input Accession Date (DD-MM-YY)
Type III (GPI-Deficient) RBCs (%) (CD235a+CD59-)			
Type II (Partial GPI-Deficient) RBCs (%) (CD235a+CD59-intermediate)			
Total GPI-Deficient RBCs (%) (Type III plus Type II)			
GPI-Deficient Neutrophils (%) (CD15+FLAER-CD157-)			
GPI-Deficient Monocytes (%) (CD64+FLAER-CD157-)			

Technologist 1:		Technologist 2:	

RBC stained with CD235aFITC and CD59PE. WBC stained with FLAER, CD157PE, CD64ECD, CD15PC5, CD45PC7. The Lower Limit of Quantification (LLOQ)* for the RBC assay is better than 0.01% and that for the WBC assay is typically better than 0.1%.[1-3] Below this level, PNH cells are reported as "rare cells with PNH phenotype detected below the level of quantification." For severely pan-cytopenic patients, WBC assay sensitivity will be much lower.

Further physician and patient information is available from the Canadian PNH Network at **www.PNHnetwork.ca**

Patient Re-testing Recommendations

Classical PNH: A clinical diagnosis of PNH is dependent on the presence of GPI-deficient red and white blood cell populations with evidence of DAT-negative hemolysis and/or thrombotic events which may be life threatening.[4] The frequency of testing is dictated by clinical and hematological parameters: repeat testing is indicated upon any significant change in clinical or laboratory parameters, and is suggested at least annually for routine monitoring.[4]

Aplastic Anemia-PNH: GPI-deficient populations can be detected in 40-57% of patients with aplastic anemia. PNH clone size is determined by the size of the GPI-deficient population in the WBC lineages (the larger of that detected in neutrophils or monocytes) and may evolve over time, occasionally progressing to clinical PNH.[4-6] In the setting of aplastic anemia, international guidelines recommend screening for PNH at diagnosis, and every 3 to 6 months initially, reducing the frequency of testing if the proportion of GPI-deficient cells has remained stable over an initial two year period.[6]

Myelodysplastic Syndrome-PNH: In the setting of MDS, GPI-deficient populations can be detected in approximately 2% of patients with myelodysplastic syndrome[7], and may evolve over time. Consider screening for PNH at diagnosis in hypoplastic MDS or if evidence of DAT-negative hemolysis is present.

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*LLOQ for the RBC assay is based on 50 Type III PNH RBCs in 500,000 cells while that for neutrophils is based on 50 PNH neutrophils in 50,000 cells. The Lower Limit of Detection (LLOD) for the RBC assay is 0.002% and that for the neutrophil assay is 0.05-0.1%. [1-3]

1. Sutherland DR, *et al.* Cytometry B Clin Cytom 2012; 82(4):195-208. **2.** Sutherland DR, *et al.* Cytometry Protoc Cytom 2015; 72:6.37.1-29. **3.** Sutherland DR, *et al.* Cytometry B Clin Cytom 2014; 86(1):44-55. **4.** Borowitz MJ, *et al.* Cytomtey B Clin Cytom 2010; 78(4):211-30. **5.** Scheinberg P, *et al.* Haematologica 2010; 95(7):1075-80. **6.** Killick SB, *et al.* Br J Haematol 2016; 172(2):187-207. **7.** Raza A, *et al.* Cytometry B Clin Cytom 2014; 86(3):175-82.