HOTLINE: Effective August 15, 2022

2005006 Paroxysmal Nocturnal Hemoglobinuria (PNH), High Sensitivity, RBC and WBC PNH PAN

Specimen Required: Collect: Lavender (EDTA), pink (K_2EDTA), or green (sodium or lithium heparin).
Specimen Preparation: Transport 4 mL whole blood. (Min: 4 mL)
Storage/Transport Temperature: Refrigerated.
Remarks: Specimens must be analyzed within stability times provided.
Unacceptable Conditions: Clotted or hemolyzed specimens.
Stability (collection to initiation of testing): Ambient: 24 hours; Refrigerated: 72 hours; Frozen: Unacceptable
New York State Clients: Ambient: 24 hours; Refrigerated: 48 hours. Frozen: Unacceptable

Interpretive Data:
This test is preferred for the initial diagnosis of PNH, and was developed according to published guidelines (Cytometry B Clin. Cytom. 2010; 78:211) and as updated in 2018 (Cytometry B Clin. Cytom. 2018; 94B:49). The test includes high-sensitivity WBC and RBC analysis with a lower limit of quantification of 0.02 percent for PNH RBCs and PMNs (based on 250,000 cells analyzed) and 0.5 percent for PNH monocytes (based on 10,000 cells analyzed). The lower limit of detection for PNH RBCs and PMNs is 0.008 percent and for PNH monocytes 0.2 percent. For severely pancytopenic patients, the WBC assay sensitivity will be much lower.

WBC analysis is the most accurate measurement of the PNH clone size. FLAER and CD157 are used as GPI-linked markers; CD15 (PMNs) and CD64 (monocytes) are used as lineage-specific markers. RBC analysis quantifies Type II and Type III RBC clones when the percentage of PNH RBCs is greater than 1 percent. Glycophorin A (CD235a) is used to gate the RBC population, and CD59 is the GPI-linked antigen. Recent RBC transfusions may decrease the percentage of PNH cells measured in RBCs (Cytometry 2000; 42:223). The presence of a subclinical PNH population in myelodysplastic bone marrow disorders, such as aplastic anemia or refractory anemia, may correlate with a positive immunotherapeutic response (Blood 2006; 107, 1308-1314).

Patient Retesting Recommendations: The frequency of testing is dictated by clinical and hematologic parameters; repeat testing is indicated upon any significant change in clinical or laboratory parameters and is suggested at least annually for routine monitoring. 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 (Int J Lab Hematol 2019;41 Suppl 1:73-81).

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the U.S. Food and Drug Administration. This test was performed in a CLIA-certified laboratory and is intended for clinical purposes.

Note: If ≥1% PNH RBCs are detected, then PNH RBC TYPE reflex will be added at no additional charge

HOTLINE NOTE: There is a reflexive pattern change associated with this test.
Add reflex to 3005006, PNH RBC TYPE
There is a component change associated with this test.
Add component 3005033, RBC PNH Phenotype
Add component 3005034, Neutrophil PNH Phenotype
Add component 3005035, Monocyte PNH Phenotype
There is a clinically significant charting name change associated with this test.
Change the charting name for component 2004367, % PNH RBC from % PNH RBC to Total (II and III) CD59-deficient RBC.
Change the charting name for component 2005005, % PNH PMN from % PNH PMN to FLAER and CD157-deficient neutrophils.
Change the charting name for component 2005004, % PNH Monocytes from % PNH Monocytes to FLAER and CD157-deficient monocytes.