HOTLINE: Effective August 15, 2022

2005003 Paroxysmal Nocturnal Hemoglobinuria, High Sensitivity, WBC PNH WBC

Specimen Required: Patient Prep: **New York State Clients: Testing is only approved for the Paroxysmal Nocturnal Hemoglobinuria Panel (ARUP test code 2005006) on whole blood specimens.**
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

Interpretive Data:
WBC analysis is the most accurate measurement of the PNH clone size. In this high-sensitivity assay, FLAER and CD157 are used as GPI-linked markers; CD15 (PMNs) and CD64 (monocytes) are used as lineage-specific markers. The assay 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 lower limit of quantification is 0.02 percent for PNH 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 PMNs is 0.008 percent and for PNH monocytes 0.2 percent. For severely pan-cytopenic patients, the WBC assay sensitivity will be much lower.

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).

For initial diagnosis of PNH, order High Sensitivity RBC and WBC Panel (ARUP test code 2005006).

For delineation of RBC Types II and III populations when the RBC clone size is greater than 1 percent, order PNH, High Sensitivity, RBC (ARUP test code 2004366).

Patient Retesting Recommendations: 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. 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.

HOTLINE NOTE: There is a clinically significant charting name change associated with this test.
Change the charting name for component 2005004, % PNH Monocytes from % PNH Monocytes to FLAER and CD157-deficient monocytes.
Change the charting name for component 2005005, % PNH PMN from % PNH PMN to FLAER and CD157-deficient neutrophils.
There is a component change associated with this test.
Add component 3005034, Neutrophil PNH Phenotype
Add component 3005035, Monocyte PNH Phenotype