HOTLINE: Effective August 15, 2022

2004366 Paroxysmal Nocturnal Hemoglobinuria, High Sensitivity, RBC PNH RBC

Specimen Required: Patient Prep: **New York State Clients: Testing is only approved for the Paroxysmal Nocturnal Hemoglobinuria (PNH), High Sensitivity, RBC and WBC (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: 0.5 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: 4 days; Refrigerated: 4 days; Frozen: Unacceptable

Interpretive Data:
This high-sensitivity RBC assay tests for CD59 expression on erythrocytes using flow cytometry. It 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 RBCs (based on 250,000 cells analyzed). The lower limit of detection for PNH RBCs is 0.008 percent.

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).

For the most accurate measurement of the PNH clone size, order Paroxysmal Nocturnal Hemoglobinuria, High Sensitivity, WBC (ARUP test code 2005003) to assist with therapeutic decisions in conventional PNH.

For initial diagnosis of PNH and analysis of both RBCs and WBCs, order Paroxysmal Nocturnal Hemoglobinuria (PNH), High Sensitivity, RBC and WBC (ARUP test code 2005006).

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 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.
There is a component change associated with this test.
Add component 2005033, RBC PNH Phenotype