

## PNH

### Paroxysmal Nocturnal Hemoglobinuria Kit

Reference	Size
PNH (V50)	50 test

#### PRODUCT DESCRIPTION

**Reagent provided:** monoclonal antibodies combination: CD157/CD45/CD64

**Tested application:** flow cytometry

**Storage buffer:** aqueous buffered solution containing protein stabilizer and 0.09% sodium azide (NaN<sub>3</sub>).

**Recommended usage:** Immunostep's PNH Kit, is intended for the identification and enumeration of Paroxysmal Nocturnal Hemoglobinuria (PNH) to evaluate the possible loss of expression of the glycosylphosphatidylinositol (GPI) anchor molecule on neutrophils and monocytes by flow cytometry.

This reagent is effective for direct immunofluorescence staining for flow cytometric analysis using  $\leq 1 \mu\text{g}/10^6$  cells.

**Presentation:** liquid

**The vial contains:**

- PE Anti-Human CD157, clone SY11B5, isotype IgG1
- PerCP Anti-Human CD45, clone D3/9, isotype IgG1, 4th International Workshops on Human Leucocyte Differentiation, WS Code 825.
- APC Anti-Human CD64, clone 10.1, isotype IgG1, 6th International Workshops on Human Leucocyte Differentiation, WS Code MA36

#### CLINICAL RELEVANCE

Although a rare disease<sup>(2)</sup>, the PNH assay is frequently requested since the screening of PNH should be performed in patients with hemoglobinuria, patients with coombs-negative intravascular hemolysis, especially patients with concurrent iron deficiency, patients with venous thrombosis involving unusual sites, patients with aplastic anemia, patients with refractory anemia-MDS and patients with episodic dysphagia or abdominal pain with evidence of intravascular hemolysis.<sup>(1,3,4)</sup>

#### ANTIGEN DETAILS

**Large description:** CD157 is expressed by macrophages, neutrophils, bone marrow stromal cells, endothelial cells, follicular dendritic cells, and T and B cell progenitors prior to the rearrangement of the antigen receptors. It also plays a role in neutrophil migration and adhesion.

CD45 recognizes a human leucocyte antigen that is a member of the leucocyte common antigen (LCA) family. The CD45 antigen is present on all human leucocytes and is weakly expressed on hematopoietic progenitor cells.

The CD64 molecule is a single chain, heavily N-glycosylated type I transmembrane protein. CD64 is also known as the high-affinity receptor for IgG (FcγRI).

#### APPROPRIATE STORAGE AND HANDLING CONDITIONS

Store in the dark, refrigerated between 2 °C and 8 °C. DO NOT FREEZE. The antibody is stable until the expiry date stated on the vial label if kept at 2°C-8°C. Do not use after the date indicated.

Once the vial is open, the product is stable for 90 days.

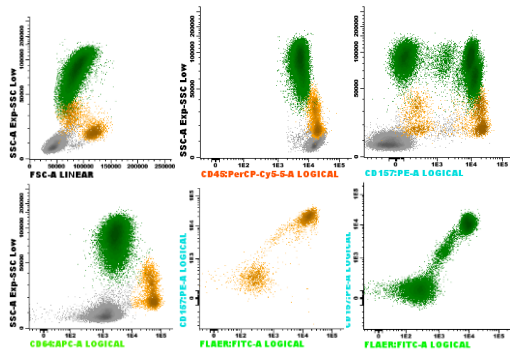
#### EVIDENCE OF DETERIORATION

Reagents should not be used if any evidence of deterioration is observed. For more information, please contact our technical service: [tech@immunostep.com](mailto:tech@immunostep.com)

The product's normal appearance is a semi-transparent, colourless liquid. It should not be used if liquid medium is cloudy or contains precipitate. It should be odourless.

#### RECOMMENDATIONS AND WARNINGS

- The reagents contain sodium azide. In acid conditions, it is transformed into hydrazoic acid, a highly toxic compound. Azide compounds must be diluted in running water before being discarded. These conditions are recommended so as to avoid deposits in plumbing, where explosive conditions could develop. The safety data sheet (SDS) is available online at [www.immunostep.com](http://www.immunostep.com)
- Avoid microbial contamination of the reagent.
- Protect from light. Use dim light during handling, incubation with cells and prior to analysis.
- Never mouth pipette.
- In the case of contact with skin, wash in plenty of water.
- The samples should be handled in the same way as those capable of transmitting infection. Appropriate handling procedures should be guaranteed.
- Do not use after the expiry date indicated on the vial.
- Deviations from the recommended procedure could invalidate the analysis results.
- FOR RESEARCH USE ONLY.
- For professional use only.
- Before acquiring the samples, it is necessary to make sure that the flow cytometer is calibrated and compensated.



The histograms are biparametric representations (Side Scatter versus Fluorescence Intensity) of a lysate stabilized whole-blood PNH sample gated on Leukocytes. Human peripheral blood was stained with PNH kit. Monocytes are represented in yellow and Neutrophils in green.

Please, refer to [www.immunostep.com](http://www.immunostep.com) for technical information.

#### WARRANTY

Warranted only to conform to the quantity and contents stated on the label or in the product labelling at the time of delivery to the customer. Immunostep disclaims hereby other warranties. Immunostep's sole liability is limited to either the replacement of the products or refund of the purchase price.

#### REFERENCES

1. Hernández-Campos PM et al. Hemoglobinuria paroxística nocturna. Med Clin 2008; 131 (16): 617-630.
2. Richards ST, Hillmenen P. Advances in the laboratory diagnosis of paroxysmal nocturnal hemoglobinuria. Clin Appl Immunol Rev 1 2001; 1:315-330.
3. Takeda J et al. Deficiency of the GPI anchor caused by a somatic mutation of the PIG-A gene in paroxysmal nocturnal hemoglobinuria. Cell 1993; 73:703-711.
4. Hernández-Campos PM et al. Detailed immunophenotypic characterization of different major and minor subsets of peripheral blood cells in patients with paroxysmal nocturnal hemoglobinuria. Transfusion 2008; 48:1403-1414.

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