PNH

Paroxysmal Nocturnal Hemoglobinuria

Rapid. Sensitive. Reliable.

PNH results from the loss of GPI-anchored proteins, leading to complement-mediated destruction of blood cells that can be accurately detected by high-sensitivity flow cytometry.

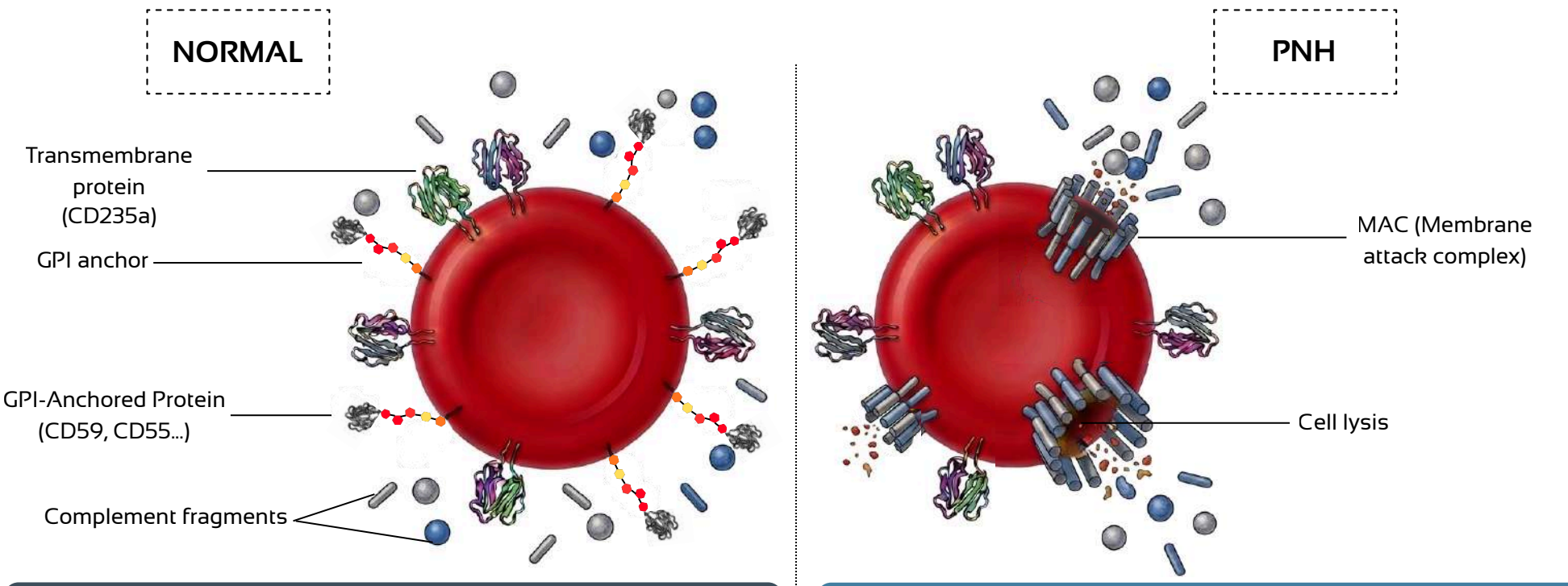
Multiparametric analysis
Single-tube workflow for neutrophils and monocytes

High sensitivity detection
Reliable identification of GPI-anchor deficient populations

Optimized workflow
Ready-to-use liquid and lyophilized formats

Research focused
Designed for advanced flow cytometry applications

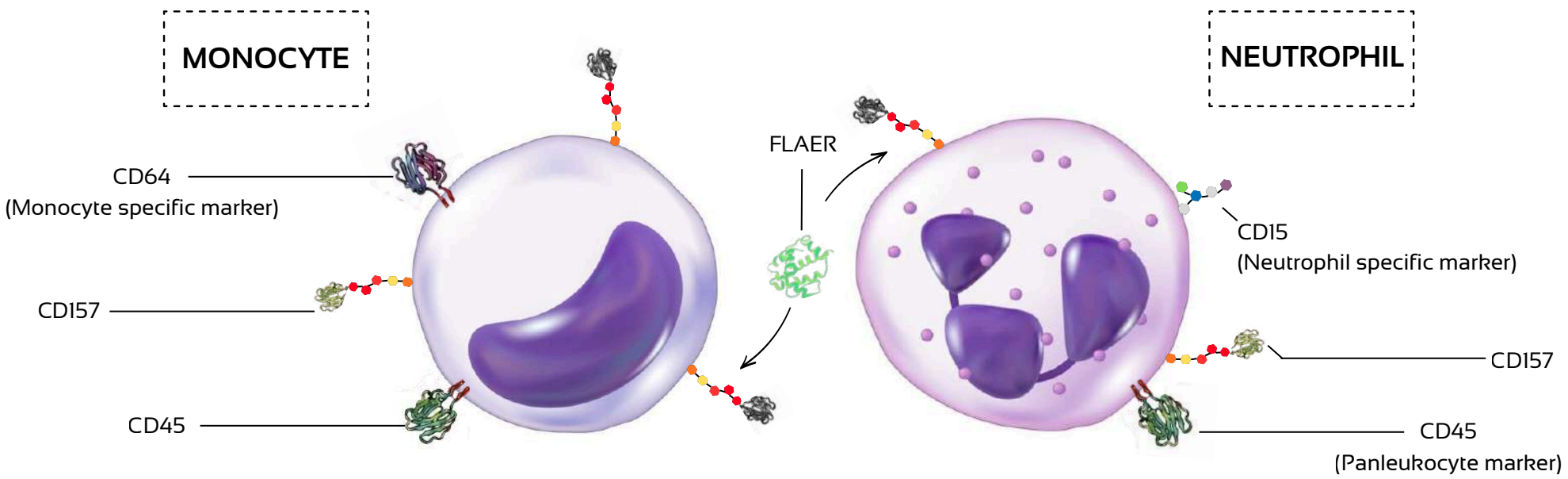
PNH Pathology



The presence of CD59 and CD55, GPI-Anchored Proteins (GPI-AP), prevents the complement system from attacking the cells.

Without GPI (due to a mutation in the PIGA gene), these protective proteins are absent and the cell is lysed by the complement system, leading to paroxysmal nocturnal hemoglobinuria.

Detection of PNH clones in leukocytes



CD157 is a GPI-anchored protein expressed on both granulocytes and monocytes, enabling simultaneous high-sensitivity identification of PNH clones, in which CD157 is lost, in a simplified workflow.

FLAER directly binds the GPI anchor, enabling highly sensitive detection of GPI-deficient populations.

RUO **For Research Use Only. Not for use in diagnostic procedures.**

Advanced PNH Detection by Flow Cytometry

Product references

REFERENCE	FORMAT	SIZE
PNH-25T	Liquid	25 test
PNH-50T	Liquid	50 test
PNHWBC-25T	Liquid	25 test
PNHRBC-25T	Liquid	25 test
LYOPNH-25T	Lyophilized tube	25 test
LYOPNHWBC-25T	Lyophilized tube	25 test
LYOPNHRBC-25T	Lyophilized tube	25 test

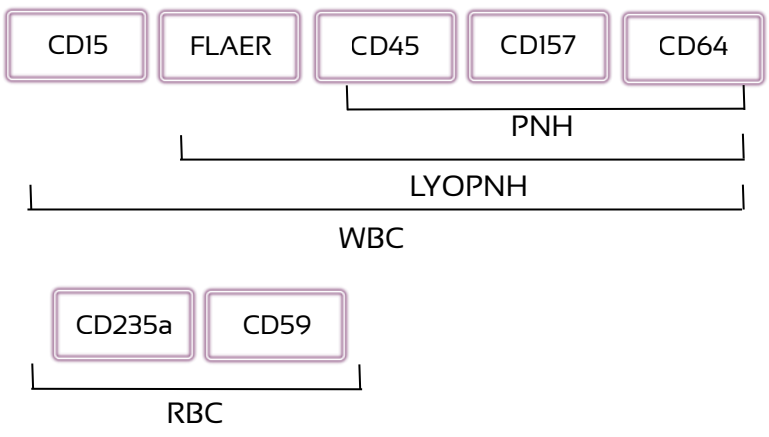
Recommended in:

- Hemoglobinuria
- Coombs-negative hemolysis
- Aplastic anemia
- MDS evaluation
- Unusual venous thrombosis

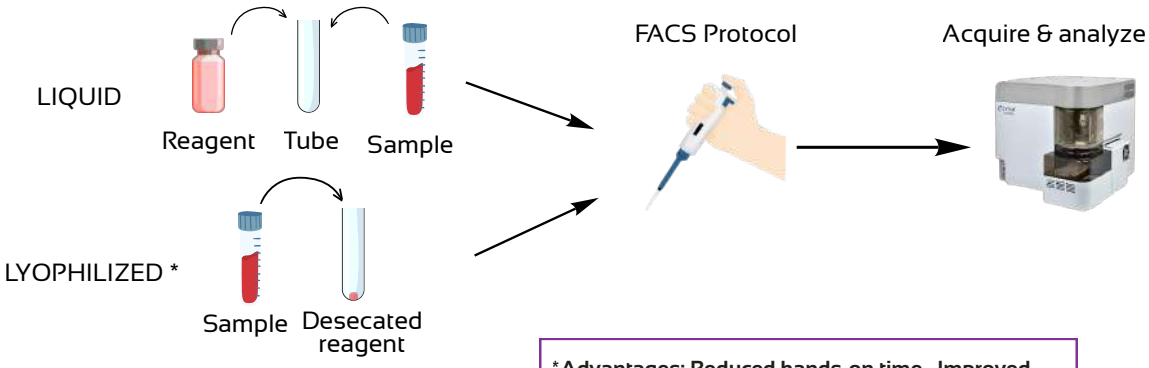
Clinical relevance

Paroxysmal Nocturnal Hemoglobinuria (PNH) is a rare hematological disorder characterized by loss of GPI-anchored proteins.
High-sensitivity flow cytomerty enables rapid and quantitative identification of abnormal neutrophil and monocyte populations.

Reagents

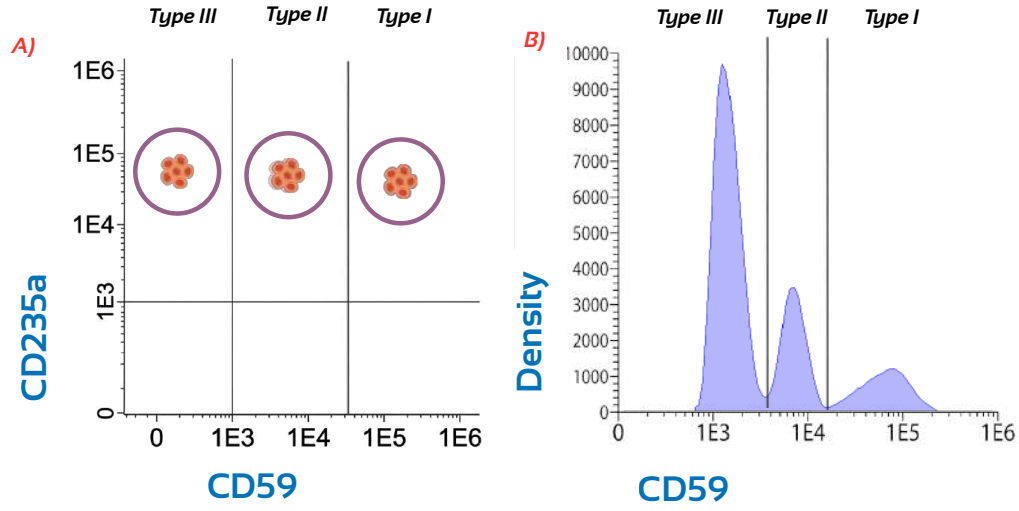


Simple workflow



*Advantages: Reduced hands-on time, Improved inter-assay reproducibility, Lower operator variability.

Analysis strategy in erythrocytes

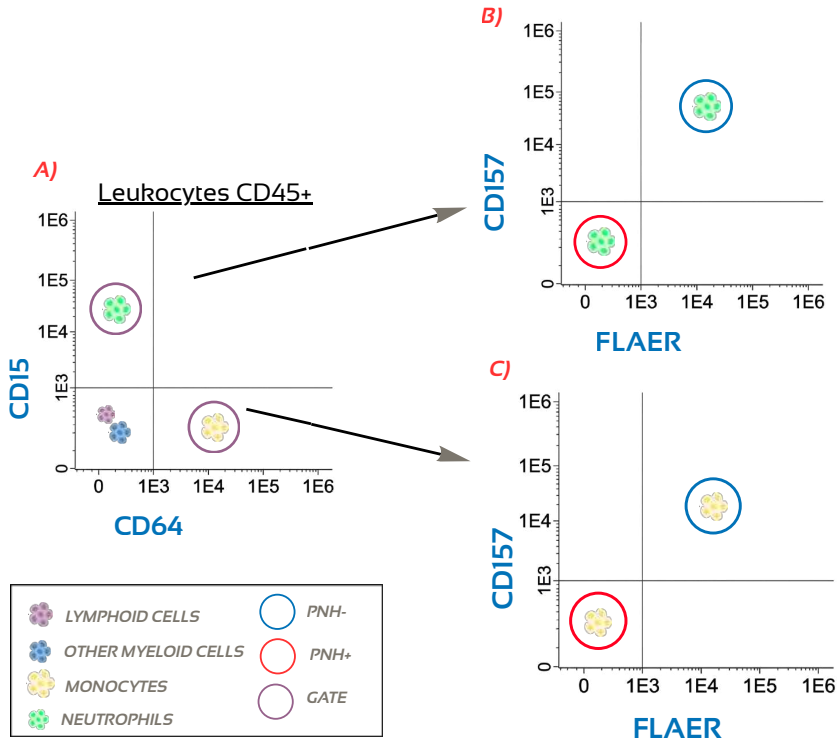


(A) Erythrocytes are identified using CD235a, and CD59 is used to identify the type associated with PNH:

CLASSIFICATION	DESCRIPTION	DOT PLOT/HISTOGRAM
Type I	Healthy cell, total GPI expression	CD59++
Type II	Partial loss of GPI	CD59+
Type III	Complete absence of GPI	CD59-

(B) Erythrocyte types can be quantified using a CD59 histogram.

Analysis strategy in leukocytes



CD45+ leukocytes are separated into monocytes and neutrophils using CD64 and CD15, respectively. **(A)**
Each of the populations, monocytes **(B)** and neutrophils **(C),** is analysed using CD157 and FLAER; this may reveal double-positive (DP) populations, which indicate a healthy cell, and double-negative (DN) populations, which indicate a PNH clone.

CLASSIFICATION	DESCRIPTION	DOT PLOT
PNH -	Healthy cell, GPI expression	A DP population
Partial PNH +	Absence of GPI in some populations	Two populations, DP and DN
Complete PNH +	Absence of GPI in all populations	A DN population

Note 1: The term 'partial' in leukocyte analysis refers to the size of the PNH clone and not to the level of expression of GPI and anchored proteins (as identified in erythrocytes). When there is no clear trend towards 100% healthy or 100% PNH, one part is positive and the other negative; the size of the PNH clone is expressed as 'partial' as a detected percentage. However, reductions in GPI expression may also be observed.

Note 2: An FLAER+/CD157- population may be observed when there is partial loss of GPI expression. CD157 expression becomes undetectable (or reduced), but FLAER is sensitive enough to detect the total GPI that is expressed, albeit at a lower level than in a healthy cell.

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