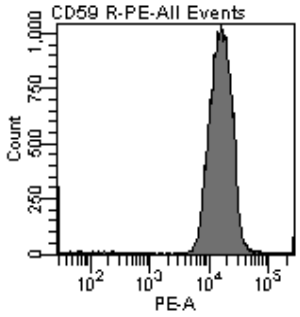
Product Flyer

Monoclonal antibodies against

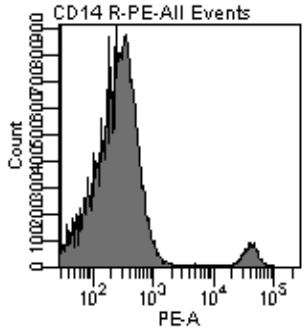
GPI-anchored proteins

Erythrocyte analysis



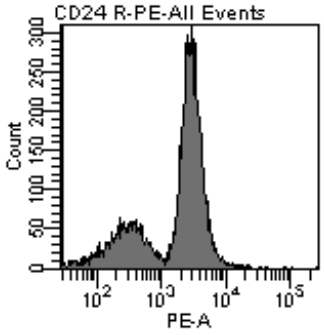
CD59

Leukocyte analysis



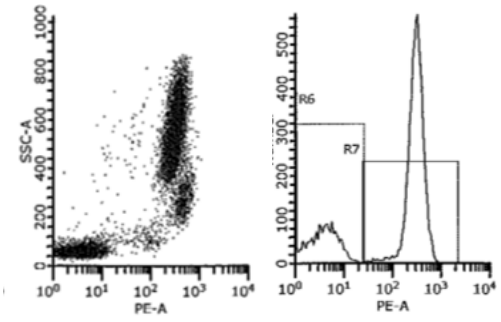
CD14

GPI-anchored
monocyte marker



CD24

GPI-anchored
granulocyte marker



CD157

GPI-anchored **monocyte** AND
granulocyte marker

Features

- Antibodies which specifically detect GPI-anchored proteins on erythrocytes, monocytes and granulocytes

Rozenburglaan 13a • 9727 DL Groningen • The Netherlands
Tel. +31 (0)50 57 57 000 • *Fax* +31 (0)50 57 57 002 • *Email* marketing@iqproducts.nl • *Web* **www.iqproducts.nl**

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Background information

Paroxysmal Nocturnal Hemoglobinuria (PNH) is a clonal hematopoietic stem cell (HSC) disease characterized by the continuous destruction of red blood cells [1]. PNH is caused by somatic mutations in the phosphatidylinositol N-acetylglucosaminyl-transferase subunit A (*PIGA*) gene, which is essential in the biosynthesis of glycosylphosphatidylinositol (GPI) anchors [2]. PIGA mutations lead to a deficiency in GPI-anchored proteins including the complement inhibitory proteins CD55 and CD59 that result in chronic complement-mediated hemolysis of GPI-deficient erythrocytes, as well as activation of platelets, monocytes and granulocytes [3]. Consequently, when left untreated this may lead to life-threatening thrombosis [4].

PNH is a very rare disease with an estimated incidence of 1.3 per million inhabitants [5]. The clinical manifestation of PNH includes hemolytic anemia, thrombosis, and smooth muscle dystonias, as well as bone marrow failure in some cases [6]. However, these symptoms are not solely related to PNH. Therefore, PNH testing is not only recommended for patients that develop PNH, but also for patients with PNH-like symptoms in disorders such as Aplastic Anemia (AA) and Myelodysplastic syndrome (MDS). Both AA and MDS are characterized by diminished production of red- and white blood cells and thrombocytes in the bone marrow.

PNH is a clinical diagnosis that should be confirmed with flow cytometry to detect the absence or severe deficiency of GPI-anchored proteins on ≥ 2 lineages (e.g. monocytes and granulocytes) [7]. The loss of GPI-anchored proteins can be detected using a combination of monoclonal antibodies directed against monocyte and granulocyte lineages and FLAER (fluorescent aerolysin) that binds *ALL* GPI-anchored proteins expressed on the surface of leukocytes [8].

Available antibodies and conjugates

Item	Clone	FITC	R-PE	CyQ(1)	APC	PerCP	PerCP-Cy5.5	OC515
Erythrocytes								
CD235a	NaM10-6G4	IQP-145F						
CD59	MEM-43		IQP-561R					
Leukocytes (granulocytes and monocytes)								
CD14	UCHM1		IQP-143R(2)		IQP-143A(2)			
CD24	SN3		IQP-559R		IQP-559A			
CD157	SY11/B5		IQP-563R					
CD15	MEM-158			IQP-564C	IQP-564A		IQP-564PCC	
CD64	22			IQP-568C	IQP-568A			
CD45	ML2					IQP-124PC		IQP-124OC

(1) CyQ, tandem conjugate of Cy5.18 and R-PE
(2) IVD CE in vitro diagnostic medical device. The products are registered as IVD in the countries belonging to the European Union.

References

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2. *Takeda, J. et al. Deficiency of the GPI anchor caused by a somatic mutation of the PIG-A gene in paroxysmal nocturnal hemoglobinuria. Cell 73, 703–711 (1993).*
3. *Brodsky, R. A. Paroxysmal nocturnal hemoglobinuria. Blood 124, 2804–2811 (2014).3.*
4. *Hill A, et al. Thrombosis in paroxysmal nocturnal hemoglobinuria. Blood. 2013;121(25):4985-4996.*
5. *Hill A, et al. The incidence and prevalence of paroxysmal nocturnal hemoglobinuria (PNH) and survival of patients in Yorkshire. Blood. 2006;108:985.*
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7. *Sutherland, DR, et al. ICCS/ESCCA Consensus Guidelines to detect GPI-deficient cells in Paroxysmal Nocturnal Hemoglobinuria (PNH) and Related Disorders: Part 2; Reagent Selection and Assay Optimization for High Sensitivity Testing. Cytometry B Clin Cytom 2018;94B:23–48.*
8. *Brodsky, R. A. et al. Improved detection and characterization of paroxysmal nocturnal hemoglobinuria using fluorescent aerolysin. Am. J. Clin. Pathol. 114, 459–466 (2000).This is the first description of FLAER reagent for the diagnosis of PNH.*

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