# Anti-human Gp1b alpha VHH (clone GP1b-17)

#### **Properties**

Product type:	VHH				
Catalog number:	G-001				
Clone number:	Gplb-17				
Immunogen:	Purified recombinant human glycoprotein lb alpha (GPlbα)				
Reacts with:	Human glycoprotein lb alpha (GPlbα)				
Tested applications:	ELISA, Flow cytometry (FC)				
Source:	Recombinant monoclonal VHH ( <i>Llama glama</i> ), purified from HEK293-E 253 cells using affinity chromatography				
Clonality:	Monoclonal				
Purity:	IMAC purified with Nickel excel Sepharose, >98%				
Storage buffer:	PBS				
Form:	Liquid				
Concentration:	4.3 mg/ml				
Storage:	Store at -80°C				

#### **Products**

Cat. No.	Target	Clone	Form	<b>Applications</b>	Size
G-001	Human GPIb alpha	Gplb-17	Purified	ELISA, FC	250 μg

## **Description**

Glycoprotein Ib alpha (GPIbα), also known as CD42, is a transmembrane protein of 135 kDa. Together with GPIbβ, GPIX and GPV, it forms the non-covalent GPIb-V-IX complex on megakaryocytes and platelets. GPIbα is present at 25.000 copies per platelet. Platelet activation is accompanied by a transient clearance of GPIb from the platelet surface, which is followed by a slow reappearance to a normal surface expression level within 30 to 60 min. Also, deficiency of a single subunit dramatically decreases the surface expression of the whole complex.

The GPIb-V-IX complex functions as a receptor for von Willebrand factor, allowing platelet adhesion and platelet plug formation at sites of vascular injury. Additionally, GPIb contains a binding site for P-selectin, Mac-1, coagulation factor XI and XII, thrombin and high molecular-weight kininogen. Hence, GPIb is an omnivalent receptor that links primary and secondary hemostasis.

Defects in the gene encoding for  $\mathsf{GPIb}\alpha$ , in addition to the genes for  $\mathsf{GPIb}\beta$  and  $\mathsf{GPIX}$ , give rise to a serious bleeding diathesis, which is accompanied by morphological platelet anomalies, including giant platelets. Collectively, this is defined as Bernard-Soulier Syndrome (BSS), a rare hereditary thrombocytopathy. A gain-of-function mutation causes platelet-type von Willebrand disease.

# Use, storage and stability

Anti-human GPIb $\alpha$  (GpIb-17) VHH is suitable for use in ELISA and flow cytometry studies. GpIb-17 is directed against the N-terminal leucine-rich repeats within GPIb alpha, and as a result interferes with the binding of vWF to GPIb $\alpha$ .

After thawing, Gplb-17 should be stored in appropriate small aliquots at -20°C or -80°C or at4°C for short time storage.

Product Datasheet PODI-VHH G-001

## PRODUCT USE LIMITATIONS, WARRANTY, DISCLAIMER

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