

## Anti-human GPIb alpha (Gplb-17) VHH



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## Properties

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

## Products

Cat. No.	Target	Clone	Form	Applications	Size
G-001	Human GPIb alpha	Gplb-17	Purified	ELISA, FC	250 $\mu$ g

## Description

Glycoprotein Ib alpha (GPIb $\alpha$ ), also known as CD42, is a transmembrane protein of 135 kDa. Together with GPIb $\beta$ , GPIX and GPV, it forms the non-covalent GPIb-V-IX complex on megakaryocytes and platelets.<sup>1-3</sup> GPIb $\alpha$  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.<sup>4</sup> 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.<sup>2,5-8</sup>

Defects in the gene encoding for GPIb $\alpha$ , in addition to the genes for GPIb $\beta$  and 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.<sup>4,8</sup> A gain-of-function mutation causes platelet-type von Willebrand disease.<sup>9</sup>

## Use, storage and stability

Anti-human GPIb $\alpha$  (Gplb-17) VHH is suitable for use in ELISA and flow cytometry studies. Gplb-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 (stable for at least 6 months) or at 4°C for short time storage.

## References

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