Product name:	CD42b Rabbit Polyclonal Antibody
Cat number:	ABN08396
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from the Internal region of human GP1BA. AA range:271-320
Reactivity:	Human,Rat,Mouse
Applications:	WB 1:500-1:2000,IHC 1:100-1:300,ICC/IF 1:50-1:200,ELISA 1:10000-1:20000
Molecular Weight:	69kDa
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

Background:

Glycoprotein Ib (GP Ib) is a platelet surface membrane glycoprotein composed of a heterodimer, an alpha chain and a beta chain, that is linked by disulfide bonds. The Gp Ib functions as a receptor for von Willebrand factor (VWF). The complete receptor complex includes noncovalent association of the alpha and beta subunits with platelet glycoprotein IX and platelet glycoprotein V. The binding of the GP Ib-IX-V complex to VWF facilitates initial platelet adhesion to vascular subendothelium after vascular injury, and also initiates signaling events within the platelet that lead to enhanced platelet activation, thrombosis, and hemostasis. This gene encodes the alpha subunit. Mutations in this gene result in Bernard-Soulier syndromes and platelet-type von Willebrand disease. The coding region of this gene is known to contain a polymophic variable number tandem repeat (VNTR) domain that isdisease:Defects in GP1BA are a cause of Bernard-Soulier syndrome (BSS) [MIM:231200]; also known as giant platelet disease (GPD). BSS patients have unusually large platelets and have a clinical bleeding tendency.,disease:Defects in GP1BA are a cause of von Willebrand disease (vWD) [MIM:177820]; also known as platelet-type von Willebrand disease or pseudo-von Willebrand disease (pseudo-vWD). This autosomal dominant bleeding disorder is caused by an increased affinity of GP-Ib for soluble vWF resulting in impaired hemostatic function due to the removal of vWF from the circulation.,disease:Defects in GP1BA are the cause of benign mediterranean macrothrombocytopenia [MIM:153670]; also known as autosomal dominant benign Bernard-Soulier syndrome. Benign mediterranean macrothrombocytopenia is characterized by mild or no clinical symptoms, normal platelet function, and normal megakaryocyte count.,disease:Genetic variations in GP1BA may be a cause of susceptibility to nonarteritic anterior ischemic optic neuropathy (NAION) [MIM:258660]; also known as susceptibility to anterior ishcemic optic neuropathy (AION). AION involves loss of vision due to damage to the optic nerve from insufficient blood supply. AION is generally divided into two types: arteritic AION and NAION. NAION probably results from minute infarctions of the optic nerve caused by occlusion of the posterior ciliary arteries. Hypercholesterolemia, diabetes mellitus, ischemic heart disease, hyperhomocysteinemia, hypertension, and crowded disk have been implicated as predisposing conditions.,function:GP-Ib, a surface membrane protein of platelets, participates in the formation of platelet plugs by binding to the A1 domain of vWF, which is already bound to the subendothelium.,miscellaneous:Binding sites for vWF and thrombin (the latter site with unknown function) are in the N-terminal part of the molecule.,miscellaneous:Platelet activation apparently involves disruption of the macromolecular complex of GP-Ib with the platelet glycoprotein IX (GP-IX) and dissociation of GP-Ib from the actin-binding protein.,polymorphism:Polymorphisms arise from a variable number of tandem 13-amino acid repeats of S-E-P-A-P-S-P-T-T-P-E-P-T in the mucin-like macroglycopeptide (Pro/Thr-rich) domain. Allele D (shown here) contains one repeat starting at position 415, allele C contains two repeats, allele B contains three repeats and allele A contains four repeats. Allele B is associated with susceptibility to nonarteritic anterior ischemic optic neuropathy.,polymorphism:Position 161 is associated with platelet-specific alloantigen Siba. Siba(-) has Thr-161 and Siba(+) has Met-161. Siba is involved in neonatal alloimmune thrombocytopenia (NATP).,PTM:Glycocalicin, which is approximately coextensive with the extracellular part of the molecule, is cleaved off by calpain during platelet lysis.,similarity:Contains 6 LRR (leucine-rich) repeats.,subunit:Heterodimer composed of GP-Ib alpha and beta; disulfide linked. GP-IX is complexed with the GP-Ib heterodimer via a non covalent linkage. Interacts with FLNB.,