

Murine Anti-Factor VIII

Clone GMA-8001

Factor VIII (FVIII) is a heterodimer consisting of a heavy chain (ranging in mass from 90 to 200 kDa) bound via metal ions to a light chain (80 kDa). In plasma, FVIII circulates in an inactive form bound to von Willebrand factor. Following activation by factor Xa or thrombin, factor VIIIa can function as cofactor for the enzyme factor IXa in the activation of factor X in the presence of phospholipid and Ca²⁺. Absent or defective FVIII is the cause of the X-linked recessive bleeding disorder hemophilia A. GMA-8001 (also known as 2-113)¹ recognizes the A3 domain of FVIII, is suitable for ELISA and bio-layer interferometry pairing applications.

Description

Antibody Source: mouse monoclonal, IgG₁

Antigen Species Bound: human

Specificity: FVIII A3 domain

Immunogen: B-domain deleted recombinant human FVIII

Formulation and Storage

Purity: Purified by protein G affinity chromatography from serum-free cell culture supernatant.

Product Formulation: Lyophilized from a ≥1 mg/ml solution in 20 mM NaH₂PO₄ 0.15 M NaCl, 1.0% (w/v) mannitol, pH 7.4. Concentration determined by absorbance measurement at 280 nm and using an extinction coefficient of 1.4 (ε_{0.1%}).

Reconstitution: Reconstitute with deionized water.

Storage: Store lyophilized or reconstituted and aliquoted material at -20° C for prolonged periods. Avoid freeze-thaw cycles. Alternatively, add 0.02% (w/v) sodium azide to reconstituted solution and store at 4° C.

Country of Origin: USA

Size Options: 0.1 mg or 0.5 mg

Applications

Working Concentration: Approximately 1-5 µg/ml. Researcher should titer antibody in specific assay.

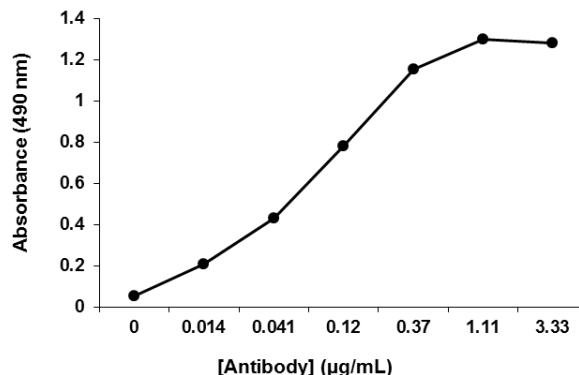
ELISA: Binds immobilized human FVIII.

Immunoblotting: Not recommended.

Inhibition: Moderately inhibitory in aPTT clotting assay.

Bio-layer Interferometry: Can be used in conjunction with GMA-8002, -8004, -8005, -8013, and -8020 for detection of FVIII.

GMA-8001 binding in ELISA



References

[1] R.J. Summers, S.L. Meeks, J.F. Healey, H.C. Brown, E.T. Parker, C.L. Kempton, C.B. Doering, P. Lollar. Factor VIII A3 domain substitution N1922S results in hemophilia A due to domain-specific misfolding and hyposecretion of functional protein. (2011). *Blood*. 117(11):3190-3198.

[2] M.A. Zimmermann, J. Oldenburg, C.R. Muller, S. Rost. Expression studies of mutant factor VIII alleles with premature termination codons with regard to inhibitor formation. (2014). *Haemophilia*. 20(3) e215–e221.

[3] M. Elnaggar, A. Al-Mohannadi, D. Kizhakayil, C. M. Raynaud, S. Al-Mannai, G. Gentilcore, I. Pavlovski, A. Sathappan, N. Van Panhuys, C. Borsotti, A. Follenzi, J-C. Grivel, S. Deola. Flow-Cytometry Platform for Intracellular Detection of FVIII in Blood Cells: A New Tool to Assess Gene Therapy Efficiency for Hemophilia A. (2020). *Molecular Therapy: Methods & Clinical Development*. 17:1-12.