

Product Information

MONOCLONAL ANTI HUMAN FACTOR IX CLONE HIX-1 Purified Mouse Immunoglobulin

Product No. **F2645**

Product Description

Monoclonal Anti-Human Factor IX (mouse IgG1 isotype) is derived from the HIX-1 hybridoma produced by the fusion of mouse Sp2/0-Ag14 myeloma cells and splenocytes from BALB/c mice immunized with factor IX purified from human plasma (developed by J.P. Miletich and colleagues). The isotype is determined using the Sigma ImmunoType Kit (Product Code ISO-1) and by a double diffusion immunoassay using Mouse Monoclonal Antibody Isotyping Reagents (Product Code ISO-2).

Monoclonal Anti-Factor IX, a divalent cation-independent antibody, recognizes factor IX when used on immunoblots of non-denatured, non-reduced human plasma.

Assays of factor IX antigen levels are useful for:

1. Initial characterization of the genetic defect in patients affected by hemophilia B.
2. Detection of female carriers of hemophilia B in families affected by mutant genes that are expressed by dysfunctional factor IX.
3. Prenatal diagnosis by fetal blood sampling when molecular genetic techniques cannot be used.
4. In-vitro studies of the role of factor IX in the intrinsic or extrinsic pathways of blood coagulation.

The antibody is useful for the preparation of factor IX depleted plasma and for purification of factor IX. It is also useful as paired labeled antibody in sandwich-type immunoassays with Monoclonal anti-Factor IX, clone HIX-5 (Product No. F1020).

Factor IX is a 55 kDa, single chain vitamin K-dependent plasma zymogen which plays a key role in the intrinsic and extrinsic blood coagulation systems. Hereditary deficiencies or dysfunctions of factor IX cause hemophilia B or "Christmas Disease" (the surname of the first family described). A disulfide bond in factor IX connects the N-terminal sequence (light chain) to the C-terminal sequence (heavy chain). Upon activation of factor IX to

factor IXa by factor XIa in the intrinsic system, an 11 kDa activation peptide is removed from the factor IX molecule by cleavage of two peptide bonds. These changes allow the exposure of the serine protease site on the heavy chain, which can then activate factor X in the presence of factor VIII, Ca⁺⁺ and phospholipid. Factor IX can be similarly activated by the extrinsic system, i.e. the tissue factor-factor VII complex.¹ Factor IX is synthesized in liver parenchymal cells and requires a post-translational vitamin K-dependent modification in order to become a mature plasma zymogen. When patients lack vitamin-K or take oral anticoagulants that interfere with the metabolism of vitamin-K, a hypocoagulable or antithrombotic state is induced. This state stems from the diminished ability of factor IX to bind to phospholipids. Factor IX concentration in human plasma ranges between 2.5-5 µg/ml and its half life is approximately 24 hr. The human factor IX gene is about 40 Kb in size and is localized at the distal end of the X-chromosome. This gene has been completely sequenced² and so far more than 50 gross or subtle mutations have been discovered.³

Reagents

The product is provided as purified antibody in 10 mM HEPES, 140 mM NaCl, pH 7.4, containing 0.05% sodium azide as a preservative.

Precautions and Disclaimer

Due to the sodium azide content a material safety sheet (MSDS) for this product has been sent to the attention of the safety officer of your institution. Consult the MSDS for information regarding hazardous and safe handling practices.

Product Profile

Protein Concentration: 4.0 mg/ml

Working Dilution

A concentration of 2-5 µg/ml specifically recognizes factor IX in human plasma using indirect immunoblotting.

Storage

For continuous use, store at 2-8 °C for up to one month.

For extended storage, freeze in working aliquots. Repeated freezing and thawing is **not** recommended. Storage in "frost-free" freezers is **not** recommended. If slight turbidity occurs upon prolonged storage, clarify the solution by centrifugation before use.

References

1. Osterud, B. and S.I. Rapaport, Proc. Natl. Acad. Sci. (USA), **74**, 5260 (1977).
2. Yoshitake, S., et al., Biochemistry, **24**, 3736 (1985).
3. Thompson, A.R., Progress in Hemostasis and Thrombosis, (Ed. Collier B.S.) **10**, 175 (1990).

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