

**BM5131****Monoclonal Antibody to Coagulation factor VIII (F8) - Purified**

<b>Alternate names:</b>	AHF, Antihemophilic factor, F8C, Procoagulant component
<b>Quantity:</b>	0.5 mg
<b>Background:</b>	Factor VIII, along with calcium and phospholipid, acts as a cofactor for factor IXa when it converts factor X to the activated form, factor Xa. It is an extracellular factor. Defects in F8 are the cause of hemophilia A (HEMA). HEMA is a common recessive X linked coagulation disorder. The frequency of hemophilia A is 1-2 in 10,000 male births in all ethnic groups. About 50% of patients have severe hemophilia A with F8C activity less than 1% of normal; they have frequent spontaneous bleeding into joints, muscles and internal organs. Moderately severe hemophilia A occurs in about 10% of patients; F8C activity is 2-5% of normal, and there is bleeding after minor trauma. Mild hemophilia A, which occurs in 30-40% of patients, is associated with F8C activity of 5-30% and bleeding occurs only after significant trauma or surgery. Of particular interest for the understanding of the function of F8C is the category of CRM (cross-reacting material) positive patients (approximately 5%) that have considerable amount of F8C in their plasma (at least 30% of normal), but the protein is nonfunctional; i.e., the F8C activity is much less than the plasma protein level. CRM reduced is another category of patients in which the F8C antigen and activity are reduced to approximately the same level. Most mutations are CRM negative, and probably affect the folding and stability of the protein.
<b>Uniprot ID:</b>	<a href="#">P00451</a>
<b>NCBI:</b>	<a href="#">NP_000123.1</a>
<b>GeneID:</b>	<a href="#">2157</a>
<b>Host / Isotype:</b>	Mouse / IgG2a
<b>Recommended Isotype</b>	AM03096PU-N
<b>Controls:</b>	
<b>Clone:</b>	MH104
<b>Immunogen:</b>	Human factor VIII antigen
<b>Format:</b>	<b>State:</b> Lyophilized purified IgG <b>Purification:</b> Protein A affinity chromatography <b>Buffer System:</b> PBS, pH 7.4 containing 0.09 % NaN <sub>3</sub> , 0.5% BSA <b>Reconstitution:</b> Restore in 1 ml dist. water
<b>Applications:</b>	ELISA: 1:5,000; detection and quantitation of human factor VIII. Immunoblotting. Immunohistochemistry on frozen sections. Other applications not tested. Optimal dilutions are dependent on conditions and should be determined by the user.

**Specificity:**

MH104 is specific for human factor VIIIc, a 300 kD protein present in plasma in a complex with von Willebrandt factor. Involved in the clotting cascade (activated by thrombin) by forming a complex with factor IXa, calcium, and phospholipids. Elevated levels of factor VIII have been associated with acute and chronic liver diseases, vascular disorders, diabetes and with acute phase reactions.

**Species:** Human.

Other species not tested.

**Storage:**

Prior to reconstitution store at 2-8°C.

Following reconstitution store the antibody at -20°C.

Avoid repeated freezing and thawing.

Shelf life: one year from despatch.

**General Readings:**

1. Stel HV, van der Kwast TH, Veerman EC: Detection of factor VIII / coagulant antigen in human liver tissue. Nature 303, 530-532 (1983)