

**\*\*REPRESENTATIVE DATASHEET\*\***

## Sheep anti-human Factor VIII

Affinity-Purified IgG

0.5 mg

**Product #:** SAF8C-AP

**Lot #:** XXXX

**Expiry date:** XXXX

Store at -10 to -20°C

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For Research Use Only.

Not for use in diagnostic procedures.

### Description of Factor VIII (FVIII)

Factor VIII (formerly referred to as antihemophilic globulin and Factor VIII:C) is a large glycoprotein (320 kDa) that circulates in plasma at approximately 200 ng/ml. Synthesized in the liver, the majority of Factor VIII is cleaved during expression, resulting in a heterogeneous mixture of partially cleaved forms of FVIII ranging in size from 200-280 kDa. The FVIII is stabilized by association with von Willebrand Factor to form a FVIII-vWF complex required for the normal survival of FVIII *in vivo* ( $t_{1/2}$  of 8-12 hours).

FVIII is a pro-cofactor that is activated through limited proteolysis by thrombin. In this process FVIIIa dissociates from vWF to combine with activated Factor IX, calcium and a phospholipid surface where it is an essential cofactor in the assembly of the Factor X activator complex. Once dissociated from vWF, FVIIIa is susceptible to inactivation by activated Protein C and by non-enzymatic decay.

Hemophilia A is a congenital bleeding disorder resulting from an X-chromosome-linked deficiency of FVIII. The severity of the deficiency generally correlates with the severity of the disease. Some Hemophiliacs (~10%) produce a FVIII protein that is partially or totally inactive. The production of neutralizing antibodies to FVIII also occurs in 5-20% of Hemophiliacs<sup>1-3</sup>.

### REFERENCES and REVIEWS

- Lollar P, Fay PJ, Fass DN; Factor VIII and Factor VIIIa. *Methods in Enzymology*, 222, pg 122, 1993.
- Hoyer, LW, Wyshock EG, Colman RW, in Hemostasis and Thrombosis, 3<sup>rd</sup> Edition, eds. RW Colman, J Hirsh, VJ Marder and EW Salzman, pp. 109-133, J.B. Lippincott Co., Philadelphia, 1994.
- Pittman DD, Kaufman RJ. Structure-Function Relationships of Factor VIII Elucidated through Recombinant DNA Technology. *Thromb. Haemostas.* 61:161-165, 1989.

## Product Specifications

### Description:

Vial containing XXXX ml of IgG purified by affinity-chromatography on immobilized FVIII. Total protein is 0.5 mg.

### Format:

Affinity-Purified IgG, clear liquid.

### Host Animal:

Sheep

### Immunogen:

Human FVIII (FVIII:C) purified from concentrate.

### Concentration:

APIgG concentration is XXXX mg/ml, determined by absorbance using an extinction coefficient ( $E^{1\%}_{280}$ ) of 13.4.

### Buffer:

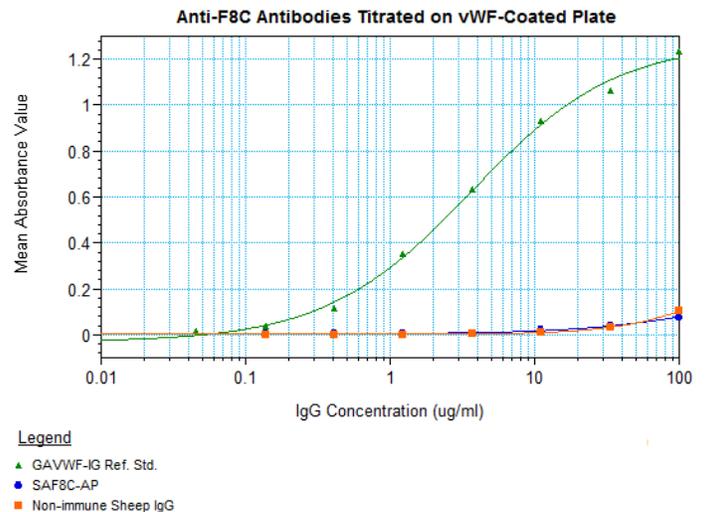
10 mM HEPES, pH 7.4, 150 mM NaCl, 50% (v/v) glycerol.

### Storage:

Store between -10 and -20°C. Product will become viscous but will not freeze. Avoid storage in frost-free freezers. Keep vial tightly capped. Allow product to warm to room temperature and gently mix before use.

### Specificity:

This antibody is specific for FVIII as demonstrated by immunoelectrophoresis and ELISA. When titrated on vWF-coated plates, SAF8C-AP does not demonstrate any reactivity above the non-immune sheep negative control.



### Applications:

Suitable as a source of antibodies to Factor VIII.

### Neutralizing activity:

XXXX Bethesda Units/ml IgG against normal plasma (Kasper CK *et al*, *Thromb Diath Haemorrh* **34**:869, 1975). One Bethesda unit/ml is defined as the amount of inhibitor that resulted in 50% residual FVIII activity after 2 hours at 37°C.

**Species Cross Reactivity:**

Not determined.

**Related Products:**

Cat #: **SAF8C-IG** Sheep anti-human Factor VIII, whole IgG from serum  
Cat #: **SAF8C-HRP** Sheep anti-human Factor VIII, whole IgG-peroxidase  
Cat #: **F8C-EIA** Paired antibodies for Factor VIII ELISA, 4 x 96 wells  
Cat #: **FVIII-DP** Human plasma deficient in Factor VIII, immune depleted

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