

Recombinant Human Factor VIII protein ab158403

1 References 画像数 1

製品の詳細

|             |   |
|-------------|---|
| 製品名         | Recombinant Human Factor VIII protein   |
| 発現系         | Wheat germ  |
| アクセッション番号   | <b>P00451-2</b>   |
| タンパク質長      | Full length protein   |
| Animal free | No  |
| 由来          | Recombinant   |
| 生物種         | Human   |
| 配列          | MRIQDPGKVFFGNVDSSGIKHNI FNPPIIARYIRLHPHTHYSI<br>RSTLRMEL<br>MGCDLNSCSMPLGMESKAISDAQITASSYFTNMFATWSPSKA<br>RLHLQGRS<br>NAWRPQVNNPKEWLQVDFQKTMKVTVGTTQGVKSLLTSMYVK<br>EFLISSSQ<br>DGHQWTLFFQNGKVKVFQGNQDSFTPVVNSLDPPLLTRYLRI<br>HPQSWVHQ IALRMEVLGCEAQDLY |
| 予測される分子量    | 51 kDa including tags   |
| 領域          | 1 to 216  |
| タグ          | GST tag N-Terminus  |
| 配列の追加情報     | Full length protein for Isoform 2 of Factor VIII. Also known as: F8B.   |

特性

|  |                       |
|--|-----------------------|
| Our <b>Abpromise guarantee</b> covers the use of <b>ab158403</b> in the following tested applications.                               |                       |
| The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user. |                       |
| アプリケーション   | Western blot<br>ELISA |
| 製品の状態  | Liquid                |
| 備考   |                       |

## 別処理および保存

### 保存方法および安定性

Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles.

pH: 8.00

Constituents: 0.31% Glutathione, 0.79% Tris HCl

## 関連情報

### 機能

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.

### 関連疾患

Defects in F8 are the cause of hemophilia A (HEMA) [MIM:306700]. A disorder of blood coagulation characterized by a permanent tendency to hemorrhage. About 50% of patients have severe hemophilia resulting in frequent spontaneous bleeding into joints, muscles and internal organs. Less severe forms are characterized by bleeding after trauma or surgery. Note=Of particular interest for the understanding of the function of F8 is the category of CRM (cross-reacting material) positive patients (approximately 5%) that have considerable amount of F8 in their plasma (at least 30% of normal), but the protein is non-functional; i.e., the F8 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.

### 配列類似性

Belongs to the multicopper oxidase family.

Contains 3 F5/8 type A domains.

Contains 2 F5/8 type C domains.

Contains 6 plastocyanin-like domains.

### ドメイン

Domain F5/8 type C 2 is responsible for phospholipid-binding and essential for factor VIII activity.

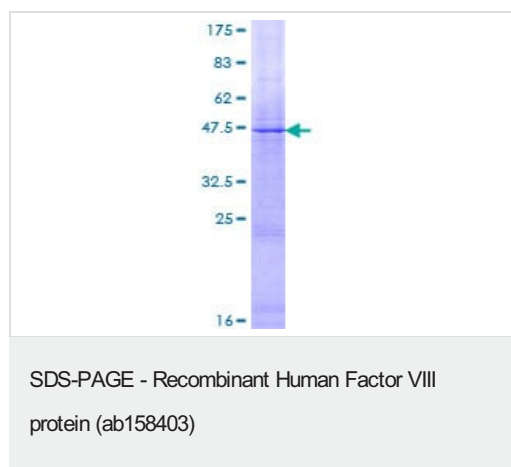
### 翻訳後修飾

Sulfation on Tyr-1699 is essential for binding vWF.

### 細胞内局在

Secreted > extracellular space.

## 画像



ab158403 on a 12.5% SDS-PAGE stained with Coomassie Blue.

**Please note:** All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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