abcam

Product datasheet

# Native Human Fibrinogen protein ab62253

## 製品の詳細

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| **製品名** | Native Human Fibrinogen protein |
| **精製度** | > 95 % Ion Exchange Chromatography.<br>Isolated by salt fractionation, gel filtration and ion exchange chromatography. |
| **発現系** | Native |
| **アクセッション番号** | P02671<br>P02675<br>P02679 |
| **タンパク質長** | Protein fragment |
| **Animal free** | No |
| **由来** | Native |
| **アミノ酸配列 1** | |
| **生物種** | Human |
| **アミノ酸配列 2** | |
| **生物種** | Human |
| **アミノ酸配列 3** | |
| **生物種** | Human |

## 特性

Our **Abpromise guarantee** covers the use of **ab62253** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

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| **製品の状態** | Lyophilized |

## 前処理および保存

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| **保存方法および安定性** | Shipped at 4°C. Upon delivery aliquot. Store at +4°C. Avoid freeze / thaw cycle.<br>Constituents: 0.9% Sodium chloride, 3% Glycine |
| **再構成** | Add 1 ml of water for a final concentration of 0.1 mg/ml. |

## 関連情報

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| **機能** | Fibrinogen has a double function: yielding monomers that polymerize into fibrin and acting as a cofactor in platelet aggregation. |
| **組織特異性** | Plasma. |
| **関連疾患** | Defects in FGA are a cause of congenital afibrinogenemia (CAFBN) [MIM:202400]. This is a rare autosomal recessive disorder characterized by bleeding that varies from mild to severe and by complete absence or extremely low levels of plasma and platelet fibrinogen. Note=The majority of cases of afibrinogenemia are due to truncating mutations. Variations in position Arg-35 (the site of cleavage of fibrinopeptide a by thrombin) leads to alpha-dysfibrinogenemias.<br>Defects in FGA are a cause of amyloidosis type 8 (AMYL8) [MIM:105200]; also known as systemic non-neuropathic amyloidosis or Ostertag-type amyloidosis. AMYL8 is a hereditary generalized amyloidosis due to deposition of apolipoprotein A1, fibrinogen and lysozyme amyloids. Viscera are particularly affected. There is no involvement of the nervous system. Clinical features include renal amyloidosis resulting in nephrotic syndrome, arterial hypertension, hepatosplenomegaly, cholestasis, petechial skin rash. |
| **配列類似性** | Contains 1 fibrinogen C-terminal domain. |
| **ドメイン** | A long coiled coil structure formed by 3 polypeptide chains connects the central nodule to the C-terminal domains (distal nodules). The long C-terminal ends of the alpha chains fold back, contributing a fourth strand to the coiled coil structure. |
| **翻訳後修飾** | The alpha chain is not glycosylated.<br>Forms F13A-mediated cross-links between a glutamine and the epsilon-amino group of a lysine residue, forming fibronectin-fibrinogen heteropolymers.<br>About one-third of the alpha chains in the molecules in blood were found to be phosphorylated.<br>Conversion of fibrinogen to fibrin is triggered by thrombin, which cleaves fibrinopeptides A and B from alpha and beta chains, and thus exposes the N-terminal polymerization sites responsible for the formation of the soft clot. The soft clot is converted into the hard clot by factor XIIIA which catalyzes the epsilon-(gamma-glutamyl)lysine cross-linking between gamma chains (stronger) and between alpha chains (weaker) of different monomers.<br>Phosphorylation sites are present in the extracellular medium. |
| **細胞内局在** | Secreted. |

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