abcam

Product datasheet

Native Human Fibrinogen protein ab62253

製品の詳細

製品名 Native Human Fibrinogen protein

精製度 > 95 % Ion Exchange Chromatography.

Isolated by salt fractionation, gel filtration and ion exchange chromatography.

発現系 Native

アクセッション番号 P02671

P02675 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.

製品の状態 Lyophilized

前処理および保存

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.

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

1

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.

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 fibringen C-terminal domain.

トメイン 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.

Forms F13A-mediated cross-links between a glutamine and the epsilon-amino group of a lysine

residue, forming fibronectin-fibrinogen heteropolymers.

About one-third of the alpha chains in the molecules in blood were found to be phosphorylated. 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.

Phosphorylation sites are present in the extracellular medium.

細胞内局在 Secreted.

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		3