abcam

Product datasheet

Natural Cow Elastin protein (FITC) ab123533

製品の概要

製品名	Natural Cow Elastin protein (FITC)
タンパク質長	Full length protein

製品の詳細

由来	Native
由来	Native

アミノ酸配列

アクセッション番号	P04985
生物種	Cow
分子量	61 kDa
領域	27 to 747
配列の追加情報	Source = bovine neck ligament elastin
標識	FITC. Ex: 493nm, Em: 528nm

特性

Our Abpromise guarantee covers the use of **ab123533** 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	Lyophilised
備考	Protect from light.

前処理および保存

保存方法および安定性	Shipped at 4°C. Store at -20°C. Store under desiccating conditions.

関連情報

機能	Major structural protein of tissues such as aorta and nuchal ligament, which must expand rapidly and recover completely. Molecular determinant of the late arterial morphogenesis, stabilizing arterial structure by regulating proliferation and organization of vascular smooth muscle.

組織特異性	Expressed within the outer myometrial smooth muscle and throughout the arteriolar tree of uterus (at protein level). Also expressed in the large arteries, lung and skin.
関連疾患	Defects in ELN are a cause of autosomal dominant cutis laxa (ADCL) [MIM:123700]. Cutis laxa is a rare connective tissue disorder characterized by loose, hyperextensible skin with decreased resilience and elasticity leading to a premature aged appearance. The skin changes are often accompanied by extracutaneous manifestations, including pulmonary emphysema, bladder diverticula, pulmonary artery stenosis and pyloric stenosis. Defects in ELN are the cause of supravalvular aortic stenosis (SVAS) [MIM:185500]. SVAS is a congenital narrowing of the ascending aorta which can occur sporadically, as an autosomal dominant condition, or as one component of Williams-Beuren syndrome. Note=ELN is located in the Williams-Beuren syndrome (WBS) critical region. WBS results from a hemizygous deletion of several genes on chromosome 7q11.23, thought to arise as a consequence of unequal crossing over between highly homologous low-copy repeat sequences flanking the deleted region. Haploinsufficiency of ELN may be the cause of certain cardiovascular and musculo-skeletal abnormalities observed in the disease.
配列類似性	Belongs to the elastin family.
翻訳後修飾	Elastin is formed through the cross-linking of its soluble precursor tropoelastin. Cross-linking is initiated through the action of lysyl oxidase on exposed lysines to form allysine. Subsequent spontaneous condensation reactions with other allysine or unmodified lysine residues result in various bi-, tri-, and tetrafunctional cross-links. The most abundant cross-links in mature elastin fibers are lysinonorleucine, allysine aldol, desmosine, and isodesmosine. Hydroxylation on proline residues within the sequence motif, GXPG, is most likely 4-hydroxy as this fits the requirement for 4-hydroxylation in vertebrates.
細胞内局在	Secreted > extracellular space > extracellular matrix. Extracellular matrix of elastic fibers.