abcam

Product datasheet

Natural Cow Elastin protein (FITC) ab123533

概述

产品名称	Natural牛Elastin蛋白(FITC)
蛋白长度	Full length protein

描述

性质	Native
来源	Native

氨基酸序列

Accession	P04985
种属	Cow
分子量	61 kDa
氨基酸	27 to 747
额外的序列信息	Source = bovine neck ligament elastin
偶联物	FITC. Ex: 493nm, Em: 528nm

技术指标

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.

形式	Lyophilised
补充说明	Protect from light.

制备和贮存

稳定性和存储	Shipped at 4°C. Store at -20°C. Store under desiccating conditions.

常规信息

功能	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.

组织特异性	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.
疾病相关	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.
序列相似性	Belongs to the elastin family.
翻译后修饰	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.
细胞定位	Secreted > extracellular space > extracellular matrix. Extracellular matrix of elastic fibers.

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