III 型胶原单克隆抗体

产品货号： mlR33129

英文名称： Collagen III

中文名称： III 型胶原单克隆抗体

别　　名： COL 3A1; COL3A1; Collagen alpha 1(III) chain; Collagen III alpha 1 chain precursor; Collagen III alpha 1 polypeptide; Collagen type III alpha 1 (Ehlers Danlos syndrome type IV autosomal dominant); Collagen type III alpha 1; Collagen type III alpha; EDS4A; Ehlers Danlos syndrome type IV, autosomal dominant; Fetal collagen; Type III collagen; CO3A1_HUMAN; Collagen alpha-1(III) chain; Type III collagen; type III preprocollagen alpha 1 chain.

研究领域： 细胞生物　免疫学

抗体来源： Mouse

克隆类型： Monoclonal

克 隆 号： 7B6

交叉反应： Human, Mouse, Rat,

产品应用： WB=1:500-1000

not yet tested in other applications.

optimal dilutions/concentrations should be determined by the end user.

分 子 量： 117kDa

细胞定位： 细胞外基质

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human Collagen III:

亚　　型： IgG

纯化方法： affinity purified by Protein G

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： Store at -20 ° C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20° C. When

reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 ° C.

PubMed : PubMed

产品介绍 : The This gene encodes the pro-alpha1 chains of type III collagen, a fibrillar collagen that is found in extensible connective tissues such as skin, lung, uterus, intestine and the vascular system, frequently in association with type I collagen. Mutations in this gene are associated with Ehlers-Danlos syndrome types IV, and with aortic and arterial aneurysms. Two transcripts, resulting from the use of alternate polyadenylation signals, have been identified for this gene. [provided by R. Dalgleish, Feb 2008]

Function:

Collagen type III occurs in most soft connective tissues along with type I collagen.

Subunit:

Trimers of identical alpha 1(III) chains. The chains are linked to each other by interchain disulfide bonds. Trimers are also cross-linked via hydroxylysines.

Subcellular Location:

Secreted, extracellular space, extracellular matrix.

Post-translational modifications:

Proline residues at the third position of the tripeptide repeating unit (G-X-Y) are hydroxylated in some or all of the chains.

O-linked glycan consists of a Glc-Gal disaccharide bound to the oxygen atom of a post-translationally added hydroxyl group.

DISEASE:

Defects in COL3A1 are a cause of Ehlers-Danlos syndrome type 3 (EDS3) [MIM:130020]; also known as benign hypermobility syndrome. EDS is a connective tissue disorder characterized by hyperextensible skin, atrophic cutaneous scars due to tissue fragility and joint hyperlaxity. EDS3 is a form of Ehlers-Danlos syndrome characterized by marked joint hyperextensibility without skeletal deformity.

Defects in COL3A1 are the cause of Ehlers-Danlos syndrome type 4 (EDS4) [MIM:130050]. EDS is a connective tissue disorder characterized by hyperextensible skin, atrophic cutaneous scars due to tissue fragility and joint hyperlaxity. EDS4 is the most severe form of the disease. It is characterized by the joint and dermal manifestations as in other forms of the syndrome, characteristic facial features (acrogeria) in most patients, and by proneness to spontaneous rupture of bowel and large arteries. The vascular complications may affect all anatomical areas.

Defects in COL3A1 are a cause of susceptibility to aortic aneurysm abdominal (AAA) [MIM:100070]. AAA is a common multifactorial disorder characterized by permanent dilation of the abdominal aorta, usually due to degenerative changes in the aortic wall. Histologically, AAA is characterized by signs of chronic inflammation, destructive remodeling of the extracellular matrix, and depletion of vascular smooth muscle cells.

Similarity:

Belongs to the fibrillar collagen family.

Contains 1 fibrillar collagen NC1 domain.

Contains 1 VWFC domain.

SWISS:

P02461

Gene ID:

1281

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

产品图片