

Recombinant Human TGF beta Receptor I protein ab70837

1 图像

描述

产品名称	重组人TGF beta Receptor I蛋白
纯度	> 90 % SDS-PAGE. Purity was determined to be >90% by densitometry. Affinity purified.
表达系统	Insect cells
蛋白长度	Protein fragment
无动物成分	No
性质	Recombinant
种属	Human
氨基酸	80 to 426

技术指标

Our **Abpromise guarantee** covers the use of **ab70837** in the following tested applications.

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

应用	SDS-PAGE
形式	Liquid

制备和贮存

稳定性和存储	Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles. pH: 7.50 Constituents: 0.00174% PMSF, 0.00385% DTT, 0.79% Tris HCl, 25% Glycerol (glycerin, glycerine), 0.29% Sodium chloride
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常规信息

功能	On ligand binding, forms a receptor complex consisting of two type II and two type I transmembrane serine/threonine kinases. Type II receptors phosphorylate and activate type I receptors which autophosphorylate, then bind and activate SMAD transcriptional regulators. Receptor for TGF-beta.
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组织特异性

Found in all tissues examined, most abundant in placenta and least abundant in brain and heart.

疾病相关

Defects in TGFBR1 are the cause of Loeys-Dietz syndrome type 1A (LDS1A) [MIM:609192]; also known as Furlong syndrome or Loeys-Dietz aortic aneurysm syndrome (LDAS). LDS1 is an aortic aneurysm syndrome with widespread systemic involvement. The disorder is characterized by arterial tortuosity and aneurysms, craniosynostosis, hypertelorism, and bifid uvula or cleft palate. Other findings include exotropia, micrognathia and retrognathia, structural brain abnormalities, intellectual deficit, congenital heart disease, translucent skin, joint hyperlaxity and aneurysm with dissection throughout the arterial tree.

Defects in TGFBR1 are the cause of Loeys-Dietz syndrome type 2A (LDS2A) [MIM:608967]. LDS2 is an aortic aneurysm syndrome with widespread systemic involvement. Physical findings include prominent joint laxity, easy bruising, wide and atrophic scars, velvety and translucent skin with easily visible veins, spontaneous rupture of the spleen or bowel, diffuse arterial aneurysms and dissections, and catastrophic complications of pregnancy, including rupture of the gravid uterus and the arteries, either during pregnancy or in the immediate postpartum period. LDS2 is characterized by the absence of craniofacial abnormalities with the exception of bifid uvula that can be present in some patients.

Defects in TGFBR1 are the cause of aortic aneurysm familial thoracic type 5 (AAT5) [MIM:608967]. Aneurysms and dissections of the aorta usually result from degenerative changes in the aortic wall. Thoracic aortic aneurysms and dissections are primarily associated with a characteristic histologic appearance known as 'medial necrosis' in which there is degeneration and fragmentation of elastic fibers, loss of smooth muscle cells, and an accumulation of basophilic ground substance.

序列相似性

Belongs to the protein kinase superfamily. TKL Ser/Thr protein kinase family. TGFBR receptor subfamily.

Contains 1 GS domain.

Contains 1 protein kinase domain.

翻译后修饰

Phosphorylated at basal levels in the absence of ligand binding. Activated by multiple phosphorylation, mainly in the GS region.

细胞定位

Membrane.

图片



SDS-PAGE showing ab70837 at approximately 65kDa.

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