

# Recombinant Human TGF beta Receptor I protein ab70837

画像数 1

### 製品の詳細

|             |  |
|-------------|--|
| 製品名         | Recombinant Human TGF beta Receptor I protein  |
| 精製度         | > 90 % SDS-PAGE.<br>Purity was determined to be >90% by densitometry. Affinity purified. |
| 発現系         | Insect cells   |
| タンパク質長      | Protein fragment   |
| Animal free | No   |
| 由来          | Recombinant  |
| 生物種         | Human  |
| 領域          | 80 to 426  |

### 特性

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.

|          |          |
|----------|----------|
| アプリケーション | SDS-PAGE |
| 製品の状態    | Liquid   |

### 前処理および保存

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| 保存方法および安定性 | Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles.<br>pH: 7.50<br>Constituents: 0.00174% PMSF, 0.00385% DTT, 0.79% Tris HCl, 25% Glycerol (glycerin, glycerine), 0.29% Sodium chloride |
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### 関連情報

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| 機能 | 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.<br>Receptor for TGF-beta. |
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## 組織特異性

## 関連疾患

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

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.

## 配列類似性

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

Contains 1 GS domain.

Contains 1 protein kinase domain.

## 翻訳後修飾

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

## 細胞内局在

Membrane.

## 画像



SDS-PAGE showing ab70837 at approximately 65kDa.

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