

LTBP4 Rabbit pAb

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Product Information

Application WB, IHC-P, IHC-F, IF, E

Primary Accession Q8N2S1

Reactivity Rat, Pig, Mouse, Dog

Host Rabbit
Clonality Polyclonal
Calculated MW 173435
Physical State Liquid

Immunogen KLH conjugated synthetic peptide derived from human LTBP4

Epitope Specificity 151-250/1624

Isotype IgG

Purity affinity purified by Protein A

Buffer 0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.

SUBCELLULAR LOCATION Secreted, extracellular space, extracellular matrix.

SIMILARITY Belongs to the LTBP family.Contains 16 EGF-like domains.Contains 4 TB

(TGF-beta binding) domains.

SUBUNIT Forms part of the large latent transforming growth factor beta precursor

complex; removal is essential for activation of complex. Interacts with LTBP1

and TGFB1. Binds to FBN1 (By similarity).

Post-translational modifications DISEASE

Contains hydroxylated asparagine residues (By similarity).

Defects in LTBP4 are the cause of Urban-Rifkin-Davis syndrome (URDS)

[MIM:613177]; also known as Cutis laxa with severe pulmonary

gastrointestinal and urinary abnormalities. URDS is a syndrome characterized

by disrupted pulmonary, gastrointestinal, urinary, musculoskeletal,

craniofacial and dermal development. Clinical features include cutis laxa, mild cardiovascular lesions, respiratory distress with cystic and atelectatic changes in the lungs, and diverticulosis, tortuosity and stenosis at various levels of the intestinal tract. Craniofacial features include microretrognathia, flat midface,

receding forehead and wide fontanelles.

Important Note This product as supplied is intended for research use only, not for use in

human, therapeutic or diagnostic applications.

Background Descriptions May be involved in the assembly, secretion and targeting of TGFB1 to sites at

which it is stored and/or activated. May play critical roles in controlling and directing the activity of TGFB1. May have a structural role in the extra cellular

matrix (ECM).

Additional Information

Gene ID 8425

Other Names Latent-transforming growth factor beta-binding protein 4, LTBP-4, LTBP4

Target/Specificity Highly expressed in heart, skeletal muscle, pancreas, uterus, and small

intestine. Weakly expressed in placenta and lung.

Dilution WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,ELISA=1:5000

-10000

Storage Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name LTBP4

Function Key regulator of transforming growth factor beta (TGFB1, TGFB2 and TGFB3)

that controls TGF-beta activation by maintaining it in a latent state during storage in extracellular space. Associates specifically via disulfide bonds with the Latency-associated peptide (LAP), which is the regulatory chain of TGF-beta, and regulates integrin-dependent activation of TGF-beta.

Cellular Location Secreted, extracellular space, extracellular matrix

Tissue Location Highly expressed in heart, skeletal muscle, pancreas, uterus, and small

intestine. Weakly expressed in placenta and lung.

Background

May be involved in the assembly, secretion and targeting of TGFB1 to sites at which it is stored and/or activated. May play critical roles in controlling and directing the activity of TGFB1. May have a structural role in the extra cellular matrix (ECM).

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.