

LTBP2/C14orf141 Rabbit pAb

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Catalog # AP57083

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q14767
Predicted	Rat, Pig, Human, Mouse, Rabbit, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	195052
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human LTBP2/C14orf141
Epitope Specificity	701-800/1821
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. Localized in nuchal ligament and aorta to the fibrillin-containing, microfibrillar component of elastic fibers.
DISEASE	Defects in LTBP2 are the cause of primary congenital glaucoma type 3D (GLC3D) [MIM:613086]. An autosomal recessive form of primary congenital glaucoma (PCG). PCG is characterized by marked increase of intraocular pressure at birth or early childhood, large ocular globes (buphthalmos) and corneal edema. It results from developmental defects of the trabecular meshwork and anterior chamber angle of the eye that prevent adequate drainage of aqueous humor. Defects in LTBP2 are the cause of microspherophakia (MCSPH) [MIM:251750]. It is a rare disease characterized by smaller and more spherical lenses than normal bilaterally, an increased anteroposterior thickness of the lens and highly myopic eyes. Lens dislocation or subluxation may occur, leading to defective accommodation.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The protein encoded by this gene belongs to the family of latent transforming growth factor (TGF)-beta binding proteins (LTBP), which are extracellular matrix proteins with multi-domain structure. This protein is the largest member of the LTBP family possessing unique regions and with most similarity to the fibrillins. It has thus been suggested that it may have multiple functions: as a member of the TGF-beta latent complex, as a structural component of microfibrils, and a role in cell adhesion. [provided by RefSeq, Jul 2008]

Additional Information

Gene ID	4053
Other Names	Latent-transforming growth factor beta-binding protein 2, LTBP-2, LTBP2,

C14orf141, LTBP3

Dilution	IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:500 0-10000
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	LTBP2
Synonyms	C14orf141, LTBP3
Function	May play an integral structural role in elastic-fiber architectural organization and/or assembly.
Cellular Location	Secreted, extracellular space, extracellular matrix
Tissue Location	Expressed in the aorta (at protein level). Expressed in lung, weakly expressed in heart, placenta, liver and skeletal muscle.

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