

ITM2B Rabbit pAb

Catalog # AP98096

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q9Y287
Other Accession	Q9Y287
Reactivity	Rat
Predicted	Human, Mouse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	30338
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human ITM2B
Epitope Specificity	10-90/266
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the ITM2 family. Contains 1 BRICHOS domain.
SUBUNIT	Homodimer; disulfide-linked. Interacts with SPPL2A and SPPL2B. Interacts with APP. Mature BRI2 (mBRI2) interacts with the APP amyloid beta A4 protein; the interaction occurs at the cell surface and in the endocytic compartments and enable alpha- and beta-secretase-induced APP cleavage inhibition. Mature BRI2 (mBRI2) interacts with the APP C99; the interaction occurs in the endocytic compartments and enable gamma-secretase-induced C99 cleavage inhibition. May form heterodimers with Bri23 peptide and APP beta-amyloid protein 40.
Post-translational modifications	The C-terminal part of the ectodomain is processed by furin and related proteases producing a secreted peptide of 4 to 5 kDa. For the ABRI and ADAN variants the C-terminal secreted peptide is larger and may produce amyloid fibrils responsible for neuronal dysfunction and dementia. The remaining part of the ectodomain containing the BRICHOS domain is cleaved by ADAM10 and is secreted as a peptide of 25 kDa. The membrane-bound N-terminal fragment (NTF) of 22 kDa is further proteolytically processed by SPPL2A and SPPL2B through regulated intramembrane proteolysis producing a secreted peptide (BRI2C) and an intracellular domain (ICD) released in the cytosol.
DISEASE	Defects in ITM2B are a cause of cerebral amyloid angiopathy ITM2B-related type 1 (CAA-ITM2B1) [MIM:176500]. A disorder characterized by amyloid deposition in the walls of cerebral blood vessels and neurodegeneration in the central nervous system. Cerebral amyloid angiopathy, non-neuritic and perivascular plaques and neurofibrillary tangles are the predominant pathological lesions. Clinical features include progressive mental deterioration, spasticity and muscular rigidity. Defects in ITM2B are a cause of cerebral amyloid angiopathy ITM2B-related type 2 (CAA-ITM2B2) [MIM:117300]; also known as hereditary ophthalmic-oto-encephalic. A disorder characterized by amyloid deposition in the walls of the blood vessels of the cerebrum, choroid plexus, cerebellum, spinal cord and retina. Plaques and neurofibrillary tangles are observed in the hippocampus. Clinical features include progressive ataxia, dementia, cataracts and deafness.

Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The type II integral membrane (ITM2) protein family consists of three members: ITM2A (also designated E25), ITM2B and ITM2C. ITM2A expression is high in osteogenic and lymphoid tissues, while both ITM2B and ITM2C are expressed in brain. ITM2B is a 266 amino acid protein that contains a potential N-glycosylation site, a potential single transmembrane-spanning domain between amino acids 52 and 74 and an extracellular C-terminal domain. Mutations in the ITM2B gene can lead to familial British dementia (FBD), and autosomal dominant disease with an onset around the fifth decade of life that is characterized by progressive dementia, spasticity and cerebellar ataxia. Familial Danish dementia (FDD), also designated heredopathia ophthalmol-oto-encephalica, is also associated with mutations in the ITM2B gene. FDD is an autosomal dominant disorder characterized by cataracts, deafness, progressive ataxia and dementia.

Additional Information

Gene ID	9445
Other Names	Bri; imBRI2; ABRI; BRI2; BRICD2B; E25B; E3-16; FBD; RDGCA; D14Sel6; E25BMM; ITM2B_BOVIN; ITM2B; Immature BRI2 (imBRI2); Transmembrane protein BRI (Bri); ITM2B_HUMAN; Protein E25B; ITM2B_MOUSE; ITM2B_PIG; ITM2B_RAT; integral membrane protein 2B; BRICHOS domain containing 2B
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
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	ITM2B
Synonyms	BRI, BRI2
Function	Plays a regulatory role in the processing of the amyloid-beta A4 precursor protein (APP) and acts as an inhibitor of the amyloid-beta peptide aggregation and fibrils deposition. Plays a role in the induction of neurite outgrowth. Functions as a protease inhibitor by blocking access of secretases to APP cleavage sites. Bri23 peptide prevents aggregation of APP amyloid-beta protein 42 into toxic oligomers.
Cellular Location	[Integral membrane protein 2B]: Golgi apparatus membrane; Single-pass type II membrane protein Note=Immature BRI2 (imBRI2) is cleaved by furin in the Golgi into mBRI2 and a Bri23 peptide. mBRI2 is transported to the plasma membrane and Bri23 peptide is secreted [Bri23 peptide]: Secreted. Note=Detected in the cerebral spinal fluid (CSF).
Tissue Location	Ubiquitous. Expressed in brain.

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