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Rabbit Anti-ITM2B antibody

SL11858R

Product Name:	ITM2B
Chinese Name:	Transmembrane proteinBRI抗体
Alias:	ABRI; ABri/ADan amyloid peptide; BRI 2; BRI; BRI2; BRICD 2B; BRICD2B; BRICHOS domain containing 2B; E25B; E3 16; E3-16; FBD; Integral membrane protein 2B; ITM 2B; ITM2B ; ITM2B_HUMAN; Protein E25B; Transmembrane protein BRI.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	30kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human ITM2B:10-90/266
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	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 ophthalmo-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.

Function:
Functions as a protease inhibitor. Plays a role in APP processing regulating the physiological production of the beta amyloid peptide. Restricts docking of gamma-secretase to APP and access of alpha- and beta-secretase to their cleavage APP sequence.

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.

Subcellular Location:
Golgi apparatus membrane. Cell membrane.

Tissue Specificity:
Expressed in brain and in other tissues.

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 heredopathia ophthalmo-oto-encephalica. 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.

Similarity:
Belongs to the ITM2 family.
Contains 1 BRICHOS domain.

SWISS:
Q9Y287

Gene ID:
9445

Database links:

Entrez Gene: 510575Cow

Entrez Gene: 476916Dog

Entrez Gene: 9445Human

Entrez Gene: 16432Mouse

Entrez Gene: 595120Rabbit

Entrez Gene: 290364Rat

Omim: 603904Human

SwissProt: Q3T0P7Cow

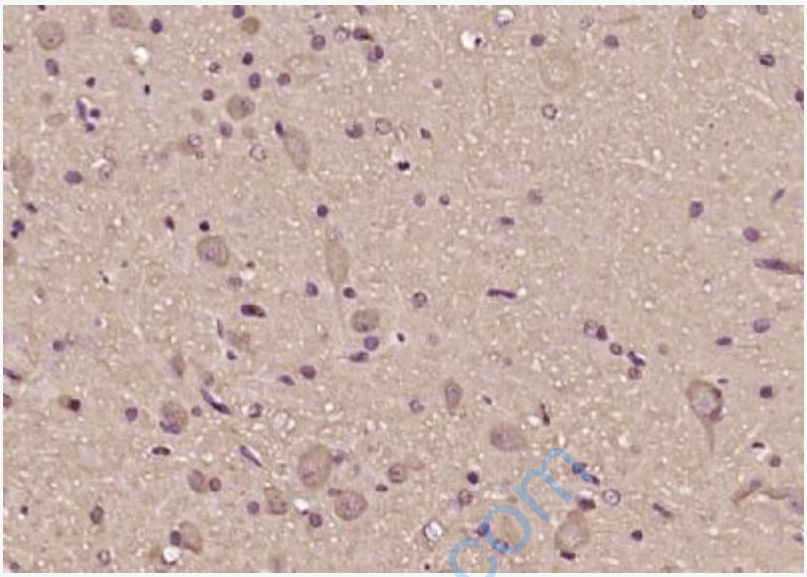
SwissProt: Q9Y287Human

SwissProt: O89051Mouse

SwissProt: Q52N47Pig

SwissProt: Q5XIE8Rat

Important Note:
This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Picture:	 Paraformaldehyde-fixed, paraffin embedded (rat brain); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (ITM2B) Polyclonal Antibody, Unconjugated (SL11858R) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.