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TBCE Rabbit pAb

Catalog # AP97632

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	Q15813
Other Accession	Q15813
Reactivity	Rat, Mouse
Predicted	Human, Rabbit, Dog
Host	Rabbit
Clonality	Polyclonal
Calculated MW	59346
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human TBCE
Epitope Specificity	1-100/527
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	Preservative: 0.02% Proclin300, Constituents: 1% BSA, 0.01M PBS, pH7.4.
SIMILARITY	Belongs to the TBCE family. Contains 1 CAP-Gly domain. Contains 7 LRR (leucine-rich) repeats. Contains 1 LRRCT domain.
SUBUNIT	Supercomplex made of cofactors A to E. Cofactors A and D function by capturing and stabilizing tubulin in a quasi-native conformation. Cofactor E binds to the cofactor D-tubulin complex; interaction with cofactor C then causes the release of tubulin polypeptides that are committed to the native state. Cofactors B and E can form a heterodimer which binds to alpha-tubulin and enhances their ability to dissociate tubulin heterodimers.
DISEASE	Defects in TBCE are a cause of hypoparathyroidism-retardation-dysmorphism syndrome (HRD) [MIM:241410]; also known as hypoparathyroidism with short stature, mental retardation, and seizures or Sanjad-Sakati syndrome. HRD is an autosomal recessive disorder reported almost exclusively in Middle Eastern populations. Defects in TBCE are the cause of Kenny-Caffey syndrome type 1 (KCS1) [MIM:244460]. KCS1 is similar to HRD with the additional features of osteosclerosis and recurrent bacterial infections.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Microtubules, the primary component of the cytoskeletal network, are highly dynamic structures composed of Alpha/Beta Tubulin heterodimers. Biosynthesis of functional microtubules involve the participation of several chaperones, termed Tubulin folding cofactors A (TBCA), D (TBCD), E (TBCE) and C (TBCC), that act on folding intermediates downstream of the cytosolic chaperon, alternatively named TCP. TBCE (tubulin folding cofactor E), also known as HRD, KCS, KCS1 or pac2, is a 527 amino acid cytoplasmic protein containing one CAP-Gly domain and seven LRR (leucine-rich) repeats. TBCE is involved in the second step of the Tubulin folding pathway and is implicated in the maintenance of the neuronal microtubule network. TBCE associates with microtubules and proteasomes, and protects against misfolded protein stress. Mutations in the gene encoding TBCE are the cause of hypoparathyroidism-retardation-dysmorphism syndrome and Kenny-Caffey syndrome type 1.

Additional Information

Gene ID	6905
Other Names	HRD; KCS; KCS1; PEAMO; pac2; 2610206D02Rik; C530005D02Rik; pmn; TBCE_HUMAN; TBCE; Tubulin-folding cofactor E; TBCE_MOUSE; TBCE_RAT; tubulin folding cofactor E; tubulin-specific chaperone e; Kenny-Caffey syndrome; hypoparathyroidism, growth and mental retardation, and dysmorphism
Dilution	WB=1:500-2000,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	TBCE
Function	Tubulin-folding protein; involved in the second step of the tubulin folding pathway and in the regulation of tubulin heterodimer dissociation. Required for correct organization of microtubule cytoskeleton and mitotic splindle, and maintenance of the neuronal microtubule network.
Cellular Location	Cytoplasm. Cytoplasm, cytoskeleton

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