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Rabbit Anti-TBCE antibody

SL11767R

Product Name:	TBCE
Chinese Name:	微管蛋白特定伴侣蛋白E抗体
Alias:	HRD; KCS; KCS1; Pac2; tbce; TBCE_HUMAN; Tubulin specific chaperone e; Tubulin-folding cofactor E; Tubulin-specific chaperone E.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Cow,Horse,Rabbit,Sheep,
Applications:	ELISA=1:500-1000 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	59kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human TBCE:442-527/527
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:	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.

Function:
Tubulin-folding protein; involved in the second step of the tubulin folding pathway. Seems to be implicated in the maintenance of the neuronal microtubule network. Involved in regulation of tubulin heterodimer dissociation.

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.

Subcellular Location:
Cytoplasm (By similarity). Cytoplasm, cytoskeleton (By similarity).

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.

Similarity:
Belongs to the TBCE family. Contains 1 CAP-Gly domain.
Contains 7 LRR (leucine-rich) repeats.
Contains 1 LRRCT domain.

SWISS:
Q15813

Gene ID:
6905

Database links:

Entrez Gene: 6905Human

	Entrez Gene: 70430Mouse Entrez Gene: 361255Rat Omim: 604934Human SwissProt: Q15813Human SwissProt: Q8CIV8Mouse SwissProt: Q5FVQ9Rat Unigene: 727621Human Unigene: 260209Mouse Unigene: 3547Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.