

TBCE (D-11): sc-398209

BACKGROUND

Microtubules, the primary component of the cytoskeletal network, are highly dynamic structures composed of α/β 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.

REFERENCES

1. Tian, G., et al. 1996. Pathway leading to correctly folded β -Tubulin. *Cell* 86: 287-296.
2. Parvari, R., et al. 2002. Mutation of TBCE causes hypoparathyroidism-retardation-dysmorphism and autosomal recessive Kenny-Caffey syndrome. *Nat. Genet.* 32: 448-452.
3. Grynberg, M., et al. 2003. Domain analysis of the Tubulin cofactor system: a model for Tubulin folding and dimerization. *BMC Bioinformatics* 4: 46.
4. Tian, G., et al. 2006. Cryptic out-of-frame translational initiation of TBCE rescues Tubulin formation in compound heterozygous HRD. *Proc. Natl. Acad. Sci. USA* 103: 13491-13496.
5. Naguib, K., et al. 2007. Hypoparathyroidism[corrected]-retardation-dysmorphism (HRD): is there a new variant not caused by a TBCE mutation? *Am. J. Med. Genet. A* 143A: 301-302.
6. Kortazar, D., et al. 2007. Role of cofactors B (TBCB) and E (TBCE) in Tubulin heterodimer dissociation. *Exp. Cell Res.* 313: 425-436.
7. Schaefer, M.K., et al. 2007. Progressive motor neuronopathy: a critical role of the Tubulin chaperone TBCE in axonal Tubulin routing from the Golgi apparatus. *J. Neurosci.* 27: 8779-8789.
8. Padidela, R., et al. 2009. Mutation in the TBCE gene is associated with hypoparathyroidism-retardation-dysmorphism syndrome featuring pituitary hormone deficiencies and hypoplasia of the anterior pituitary and the corpus callosum. *J. Clin. Endocrinol. Metab.* 94: 2686-2691.

CHROMOSOMAL LOCATION

Genetic locus: TBCE (human) mapping to 1q42.3; Tbcce (mouse) mapping to 13 A1.

SOURCE

TBCE (D-11) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 422-451 within an internal region of TBCE of human origin.

PRODUCT

Each vial contains 200 μ g IgG₁ in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

TBCE (D-11) is available conjugated to agarose (sc-398209 AC), 500 μ g/0.25 ml agarose in 1 ml, for IP; to HRP (sc-398209 HRP), 200 μ g/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-398209 PE), fluorescein (sc-398209 FITC), Alexa Fluor[®] 488 (sc-398209 AF488), Alexa Fluor[®] 546 (sc-398209 AF546), Alexa Fluor[®] 594 (sc-398209 AF594) or Alexa Fluor[®] 647 (sc-398209 AF647), 200 μ g/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor[®] 680 (sc-398209 AF680) or Alexa Fluor[®] 790 (sc-398209 AF790), 200 μ g/ml, for Near-Infrared (NIR) WB, IF and FCM.

Blocking peptide available for competition studies, sc-398209 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

Alexa Fluor[®] is a trademark of Molecular Probes, Inc., Oregon, USA

APPLICATIONS

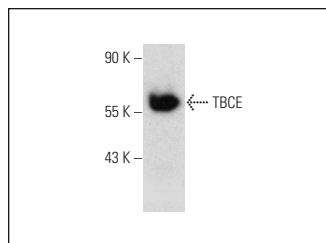
TBCE (D-11) is recommended for detection of TBCE of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for TBCE siRNA (h): sc-78922, TBCE siRNA (m): sc-106600, TBCE shRNA Plasmid (h): sc-78922-SH, TBCE shRNA Plasmid (m): sc-106600-SH, TBCE shRNA (h) Lentiviral Particles: sc-78922-V and TBCE shRNA (m) Lentiviral Particles: sc-106600-V.

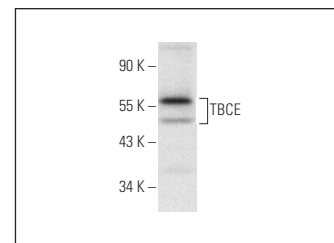
Molecular Weight of TBCE: 59 kDa.

Positive Controls: IMR-32 cell lysate: sc-2409 or Raji whole cell lysate: sc-364236.

DATA



TBCE (D-11): sc-398209. Western blot analysis of TBCE expression in Raji whole cell lysate.



TBCE (D-11): sc-398209. Western blot analysis of TBCE expression in IMR-32 whole cell lysate.

STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

For research use only, not for use in diagnostic procedures.