

# CTC1 (E-13): sc-136584

## BACKGROUND

Telomeres are DNA-protein structures that protect the ends of linear chromosomes and help maintain genomic stability and cell phenotype. CST complex plays an essential role in protecting telomeres from degradation. CTC1 (CTS telomere maintenance complex component 1), also known as CRMCC, AAF132, AAF-132, C17orf68 or tmp494178, is a 1,217 amino acid nuclear protein belonging to the CTC1 family. CTC1 along with OBFC1, also known as STN1, and TEN1 form the trimeric CST complex and is considered critical for telomere replication. Mutations in the gene encoding CST1 leads to an autosomal recessive pleiomorphic disorder that is characterized by intracranial calcifications, leukodystrophy, and brain cysts, which results in spasticity, ataxia, dystonia, seizures, and cognitive decline. Individuals afflicted with CTS1 gene mutation also may develop Coats disease, extraneurologic manifestations, osteopenia with poor bone healing and is at high risk of gastrointestinal bleeding and portal hypertension caused by vasculature ectasias in the stomach, small intestine, and liver. CTC1 exists as two alternatively spliced isoforms and is encoded by a gene located on human chromosome 17p13.1.

## REFERENCES

1. Surovtseva, Y.V., et al. 2009. Conserved telomere maintenance component 1 interacts with STN1 and maintains chromosome ends in higher eukaryotes. *Mol. Cell* 36: 207-218.
2. Polvi, A., et al. 2012. Mutations in CTC1, encoding the CTS telomere maintenance complex component 1, cause cerebrotelomeric microangiopathy with calcifications and cysts. *Am. J. Hum. Genet.* 90: 540-549.
3. Stewart, J.A., et al. 2012. Human CST promotes telomere duplex replication and general replication restart after fork stalling. *EMBO J.* 31: 3537-3549.
4. Anderson, B.H., et al. 2012. Mutations in CTC1, encoding conserved telomere maintenance component 1, cause Coats plus. *Nat. Genet.* 44: 338-342.
5. Chen, L.Y., et al. 2012. The human CST complex is a terminator of telomerase activity. *Nature* 488: 540-544.
6. Keller, R.B., et al. 2012. CTC1 Mutations in a patient with dyskeratosis congenita. *Pediatr. Blood Cancer* 59: 311-314.
7. Gu, P., et al. S. 2013. Functional characterization of human CTC1 mutations reveals novel mechanisms responsible for the pathogenesis of the telomere disease Coats plus. *Aging Cell* 12: 1100-1109.
8. Chen, L.Y., et al. 2013. Molecular basis of telomere syndrome caused by CTC1 mutations. *Genes Dev.* 27: 2099-2108.
9. Chen, L.Y., et al. 2013. CST for the grand finale of telomere replication. *Nucleus* 4: 277-282.

## CHROMOSOMAL LOCATION

Genetic locus: CTC1 (human) mapping to 17p13.1; Ctc1 (mouse) mapping to 11 B3.

## SOURCE

CTC1 (E-13) is an affinity purified rabbit polyclonal antibody raised against a peptide mapping within an internal region of CTC1 of human origin.

## PRODUCT

Each vial contains 100 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-136584 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

CTC1 (E-13) is recommended for detection of CTC1 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:50-1:500), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:25, dilution range 1:25-1:250) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000); non cross-reactive with other C17orf family members .

CTC1 (E-13) is also recommended for detection of CTC1 in additional species, including equine and canine.

Suitable for use as control antibody for CTC1 siRNA (h): sc-93645, CTC1 siRNA (m): sc-108239, CTC1 shRNA Plasmid (h): sc-93645-SH, CTC1 shRNA (m) sc-108239-SH, CTC1 shRNA (h) Lentiviral Particles: sc-93645-V and CTC1 shRNA (m) Lentiviral Particles sc-108239-V.

Molecular Weight of CTC1: 135 kDa.

## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

## 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.

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.