SANTA CRUZ BIOTECHNOLOGY, INC.

TSEN54 (h): 293T Lysate: sc-116179

BACKGROUND

The tRNA-splicing endonuclease complex is responsible for identifying and cleaving pre-tRNA at both 5' and 3' splice sites, thereby releasing introns and free tRNA molecules with 2',3' cyclic phosphates and 5'-OH termini. In addition to its role in pre-tRNA splicing, the heterotetrameric endonuclease complex participates in mRNA processing and, via its association with pre-mRNA processing factors, is thought to link pre-tRNA and pre-mRNA splicing events. TSEN54 (tRNA splicing endonuclease 54 homolog), also known as HsSEN54 (SEN54 homolog) or tRNA-intron endonuclease Sen54, is a 526 amino acid protein belonging to the SEN54 family. Localizing to nucleus, TSEN54 is a member of a complex which identifies and cleaves the splice sites in pre-tRNA, and may also be involved in mRNA processing. Defects in TSEN54 may result in pontocerebellar hypoplasia (PCH) type 4 and 2A, characterized by structural abnormalities to the cerebellum, inferior olive, and ventral pons. TSEN54 exists as two alternatively spliced isoforms.

REFERENCES

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2. Zody, M.C., et al. 2006. DNA sequence of human chromosome 17 and analysis of rearrangement in the human lineage. Nature 440: 1045-1049.
3. Budde, B.S., et al. 2008. tRNA splicing endonuclease mutations cause pontocerebellar hypoplasia. Nat. Genet. 40: 1113-1118.
4. Cassandrini, D., et al. 2010. Pontocerebellar hypoplasia: clinical, pathologic, and genetic studies. Neurology 75: 1459-1464.
5. Namavar, Y., et al. 2011. Clinical, neuroradiological and genetic findings in pontocerebellar hypoplasia. Brain 134: 143-156.
6. Namavar, Y., et al. 2011. TSEN54 mutations cause pontocerebellar hypoplasia type 5. Eur. J. Hum. Genet. 19: 724-726.
7. Kasher, P.R., et al. 2011. Impairment of the tRNA-splicing endonuclease subunit 54 (TSEN54) gene causes neurological abnormalities and larval death in zebrafish models of pontocerebellar hypoplasia. Hum. Mol. Genet. 20: 1574-1584.
8. Maricich, S.M., et al. 2011. Pontocerebellar hypoplasia: review of classification and genetics, and exclusion of several genes known to be important for cerebellar development. J. Child Neurol. 26: 288-294.

CHROMOSOMAL LOCATION

Genetic locus: TSEN54 (human) mapping to 17q25.1.

PRODUCT

TSEN54 (h): 293T Lysate represents a lysate of human TSEN54 transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

APPLICATIONS

TSEN54 (h): 293T Lysate is suitable as a Western Blotting positive control for human reactive TSEN54 antibodies. Recommended use: 10-20 µl per lane.

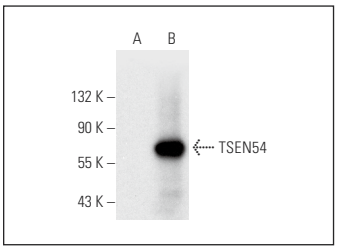
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

TSEN54 (A-9): sc-398327 is recommended as a positive control antibody for Western Blot analysis of enhanced human TSEN54 expression in TSEN54 transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048.

DATA



TSEN54 (A-9): sc-398327. Western blot analysis of TSEN54 expression in non-transfected: sc-117752 (**A**) and human TSEN54 transfected: sc-116179 (**B**) 293T whole cell lysates.

RESEARCH USE

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

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.