

# TSEN54 (A-9): sc-398327

## 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

1. Paushkin, S.V., et al. 2004. Identification of a human endonuclease complex reveals a link between tRNA splicing and pre-mRNA 3' end formation. *Cell* 117: 311-321.
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.

## CHROMOSOMAL LOCATION

Genetic locus: TSEN54 (human) mapping to 17q25.1; Tsen54 (mouse) mapping to 11 E2.

## SOURCE

TSEN54 (A-9) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 111-128 within an internal region of TSEN54 of human origin.

## PRODUCT

Each vial contains 200 µg IgG<sub>2b</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-398327 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

## RESEARCH USE

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

## APPLICATIONS

TSEN54 (A-9) is recommended for detection of TSEN54 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg 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).

TSEN54 (A-9) is also recommended for detection of TSEN54 in additional species, including equine, canine and bovine.

Suitable for use as control antibody for TSEN54 siRNA (h): sc-93849, TSEN54 siRNA (m): sc-154715, TSEN54 shRNA Plasmid (h): sc-93849-SH, TSEN54 shRNA Plasmid (m): sc-154715-SH, TSEN54 shRNA (h) Lentiviral Particles: sc-93849-V and TSEN54 shRNA (m) Lentiviral Particles: sc-154715-V.

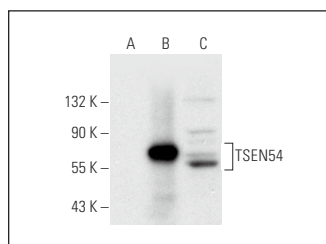
Molecular Weight of TSEN54 isoform 1/2: 59/20 kDa.

Positive Controls: TSEN54 (h): 293T Lysate: sc-116179 or Y79 cell lysate: sc-2240.

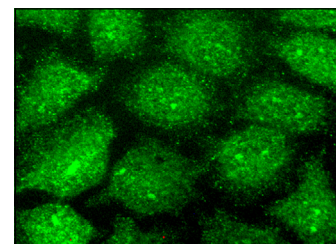
## RECOMMENDED SUPPORT REAGENTS

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. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

## DATA



TSEN54 (A-9): sc-398327. Western blot analysis of TSEN54 expression in non-transfected 293T: sc-117752 (A), human TSEN54 transfected 293T: sc-116179 (B) and Y79 (C) whole cell lysates.



TSEN54 (A-9): sc-398327. Immunofluorescence staining of methanol-fixed HeLa cells showing nucleolar and nuclear localization.

## SELECT PRODUCT CITATIONS

1. Störk, T., et al. 2019. TSEN54 missense variant in Standard Schnauzers with leukodystrophy. *PLoS Genet.* 15: e1008411.

## STORAGE

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