

Product name:	TSEN54 Rabbit Polyclonal Antibody
Cat number:	ABN19355
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human TSEN54. AA range:261-310
Reactivity:	Human,Rat,Mouse
Applications:	WB 1:500-1:2000,IHC 1:100-1:300,ICC/IF 1:50-1:200,ELISA 1:10000-1:20000
Molecular Weight:	60kDa
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

Background:

This gene encodes a subunit of the tRNA splicing endonuclease complex, which catalyzes the removal of introns from precursor tRNAs. The complex is also implicated in pre-mRNA 3'-prime end processing. Mutations in this gene result in pontocerebellar hypoplasia type 2.[provided by RefSeq, Oct 2009],disease:Defects in TSEN54 are the cause of pontocerebellar hypoplasia type 2A (PCH2A) [MIM:277470]. PCH type 2 is characterized by progressive microcephaly from birth combined with extrapyramidal dyskinesia and chorea, epilepsy, and normal spinal cord findings.,disease:Defects in TSEN54 are the cause of pontocerebellar hypoplasia type 4 (PCH4) [MIM:225753]. Pontocerebellar hypoplasia (PCH) is a heterogeneous group of disorders characterized by an abnormally small cerebellum and brainstem. PCH4 is characterized by severe course and early lethality.,function:Non-catalytic subunit of the tRNA-splicing endonuclease complex, a complex responsible for identification and cleavage of the splice sites in pre-tRNA. It cleaves pre-tRNA at the 5' and 3' splice sites to release the intron. The products are an intron and two tRNA half-molecules bearing 2',3' cyclic phosphate and 5'-OH termini. There are no conserved sequences at the splice sites, but the intron is invariably located at the same site in the gene, placing the splice sites an invariant distance from the constant structural features of the tRNA body. The tRNA splicing endonuclease is also involved in mRNA processing via its association with pre-mRNA 3' end processing factors, establishing a link between pre-tRNA splicing and pre-mRNA 3' end formation, suggesting that the endonuclease subunits function in multiple RNA-processing events.,PTM:Phosphorylated upon DNA damage, probably by ATM or ATR.,similarity:Belongs to the SEN54 family.,subcellular location:May be transiently localized in the nucleolus.,subunit:tRNA splicing endonuclease is a heterotetramer composed of SEN2, SEN15, SEN34/LENG5 and SEN54. tRNA splicing endonuclease complex also contains proteins of the Pre-mRNA 3' end processing machinery such as CLP1, CPSF1, CPSF4 and CSTF2. Also belongs to a complex containing isoform 2 of SEN2.,