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Rabbit Anti-SNRNP200/AF350 Conjugated antibody

SL12657R-AF350

Product Name	Anti-SNRNP200/AF350
Chinese Name	AF350 标记的 SNRNP200 蛋白抗体
Alias	Activating signal cointegrator 1 complex subunit 3 like 1; ASCC3L1; BRR2; BRR2 homolog; HELIC2; RP33; SNRNP200; U5 200KD; U5 small nuclear ribonucleoprotein 200 kDa helicase; U5 snRNP specific 200 kDa protein.
Research Area	Cell biology Epigenetics
Immunogen Species	Rabbit
Clonality	Polyclonal
React Species	Mouse,Rat(predicted:Human)
Applications	IF=1:100-500 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight	244kDa
Form	Lyophilized or Liquid
Concentration	1mg/ml
immunogen	KLH conjugated synthetic peptide derived from human SNRNP200
Lsotype	IgG
Purification	affinity purified by Protein A
Storage Buffer	1M TBS(pH7.4) with 1% BSA, 3% Proclin300 and 50% Glycerol
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 1M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
Product Detail	**background:** Pre-mRNA splicing is catalyzed by the spliceosome, a complex of specialized RNA and protein subunits that removes introns from a transcribed pre-mRNA segment. The spliceosome consists of small nuclear RNA proteins (snRNPs) U1, U2, U4, U5 and U6, together with approximately 80 conserved proteins.

U5 snRNP contains nine specific proteins. This gene encodes one of the U5 snRNP-specific proteins. This protein belongs to the DEXH-box family of putative RNA helicases. It is a core component of U4/U6-U5 snRNPs and appears to catalyze an ATP-dependent unwinding of U4/U6 RNA duplices. Mutations in this gene cause autosomal-dominant retinitis pigmentosa. Alternatively spliced transcript variants encoding different isoforms have been found, but the full-length nature of these variants has not been determined. [provided by RefSeq, Mar 2010]

Function:
Putative RNA helicase involved in the second step of RNA splicing. May promote one or more conformational changes in the dynamic network of RNA-RNA interactions in the spliceosome. Appears to catalyze an ATP-dependent unwinding of U4/U6 RNA duplices.

Subcellular Location:
Nucleus.

Tissue Specificity:
Widely expressed.

DISEASE:
Defects in SNRNP200 are the cause of retinitis pigmentosa type 33 (RP33) [MIM:610359]. It is a retinal dystrophy belonging to the group of pigmentary retinopathies. Retinitis pigmentosa is characterized by retinal pigment deposits visible on fundus examination and primary loss of rod photoreceptor cells followed by secondary loss of cone photoreceptors. Patients typically have night vision blindness and loss of midperipheral visual field. As their condition progresses, they lose their far peripheral visual field and eventually central vision as well.

Similarity:
Belongs to the helicase family. SKI2 subfamily.
Contains 2 helicase ATP-binding domains.
Contains 2 helicase C-terminal domains.
Contains 2 SEC63 domains.

Database links:

Entrez Gene: 23020 Human

Entrez Gene: 320632 Mouse

Entrez Gene: 296126 Rat

Omim: 601664 Human

SwissProt: O75643 Human

SwissProt: Q6P4T2 Mouse

SwissProt: F1LNJ2 Rat

Unigene: 246112 Human

Important Note:
This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.