

EXOSC3 Rabbit pAb

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Catalog # AP57992

Product Information

Application	WB
Primary Accession	Q9NQT5
Reactivity	Human, Mouse
Predicted	Rat, Rabbit, Chicken, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	29572
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human EXOSC3
Epitope Specificity	101-200/275
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Cytoplasm. Nucleus, nucleolus.
DISEASE	Defects in EXOSC3 are the cause of pontocerebellar hypoplasia type 1B (PCH1B) [MIM:614678]. A severe autosomal recessive neurologic disorder characterized by a combination of cerebellar and spinal motor neuron degeneration beginning at birth. There is diffuse muscle weakness, progressive microcephaly, global developmental delay, and brainstem involvement.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The exosome is a multisubunit complex of 3' to 5' exoribonucleases. It is involved in a variety of cellular processes and is responsible for degrading unstable mRNAs that contain AU-rich elements in their untranslated 3' region. EXOSC3 (exosome component 3), also known as p10, CGI-102, RRP40 (Ribosomal RNA-processing protein 40), Rrp40p or hRrp40p, is a component of the exosome multienzyme ribonuclease complex. Localizing to the cytoplasm and nucleolus, EXOSC3 contains a putative S1 RNA-binding domain and is capable of binding RNA. EXOSC3 is a component of the top cap of the exosome and is essential for exosome stability. In addition, EXOSC3 is required for the processing of the 7S pre-RNA to the mature 5.8S rRNA.

Additional Information

Gene ID	51010
Other Names	Exosome complex component RRP40, Exosome component 3, Ribosomal RNA-processing protein 40, p10, EXOSC3, RRP40
Dilution	WB=1:500-2000

Storage

Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information**Name**

EXOSC3

Synonyms

RRP40

Function

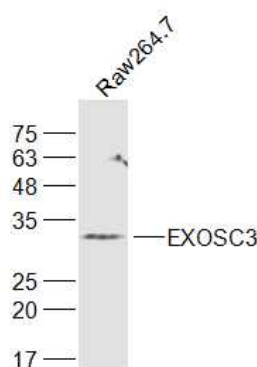
Non-catalytic component of the RNA exosome complex which has 3'→5' exoribonuclease activity and participates in a multitude of cellular RNA processing and degradation events. In the nucleus, the RNA exosome complex is involved in proper maturation of stable RNA species such as rRNA, snRNA and snoRNA, in the elimination of RNA processing by-products and non-coding 'pervasive' transcripts, such as antisense RNA species and promoter-upstream transcripts (PROMPTs), and of mRNAs with processing defects, thereby limiting or excluding their export to the cytoplasm. The RNA exosome may be involved in Ig class switch recombination (CSR) and/or Ig variable region somatic hypermutation (SHM) by targeting AICDA deamination activity to transcribed dsDNA substrates. In the cytoplasm, the RNA exosome complex is involved in general mRNA turnover and specifically degrades inherently unstable mRNAs containing AU-rich elements (AREs) within their 3' untranslated regions, and in RNA surveillance pathways, preventing translation of aberrant mRNAs. It seems to be involved in degradation of histone mRNA. The catalytic inactive RNA exosome core complex of 9 subunits (Exo-9) is proposed to play a pivotal role in the binding and presentation of RNA for ribonucleolysis, and to serve as a scaffold for the association with catalytic subunits and accessory proteins or complexes. EXOSC3 as peripheral part of the Exo-9 complex stabilizes the hexameric ring of RNase PH-domain subunits through contacts with EXOSC9 and EXOSC5.

Cellular Location

Cytoplasm. Nucleus, nucleolus. Nucleus

Background

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Images**Sample:**

Raw264.7(Human) Cell Lysate at 30 ug

Primary: Anti-EXOSC3 (AP57992) at 1/300 dilution

Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

Predicted band size: 29 kD

Observed band size: 29 kD