10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

DIS3L2 Rabbit pAb

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Catalog # AP59123

Product Information

Primary Accession	Q8IYB7
Reactivity	Mouse
Predicted	Rat, Pig, Dog, Human, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	99279
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human DIS3L2
Epitope Specificity	1-100/885
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.
DISEASE	Defects in DIS3L2 are the cause of Perlman syndrome (PRLMNS) [MIM:267000]. An autosomal recessive congenital overgrowth syndrome. Affected children are large at birth, are hypotonic, and show organomegaly, characteristic facial dysmorphisms (inverted V-shaped upper lip, prominent forehead, deep-set eyes, broad and flat nasal bridge, and low-set ears), renal anomalies (nephromegaly and hydronephrosis), frequent neurodevelopmental delay, and high neonatal mortality. Perlman syndrome is associated with a high risk of Wilms tumor. Histologic examination of the kidneys in affected children shows frequent nephroblastomatosis, which is a precursor lesion for Wilms tumor.
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 composed of several highly conserved subunits, some of which are 3′ to 5′ exoribonucleases. The complex is involved in a variety of cellular processes and is responsible for degrading unstable mRNAs that contain AU-rich (ARE) elements in their untranslated 3′ region. DIS3L2 (DIS3-like exonuclease 2) is an 885 amino acid protein that is thought to function as an exonuclease and may be required for the 3′ processing of pre-mRNA into mature mRNA. Defects or chromosomal translocations involving the gene encoding DIS3L2 may be associated with Marfanoid habitus, a genetic disorder characterized by abnormalities in the skeleton, eyes and cardiovascular system. DIS3L2 is expressed as five isoforms due to alternative splicing events.

Additional Information

Gene ID	129563

Other Names	DIS3-like exonuclease 2 {ECO:0000255|HAMAP-Rule:MF_03045}, hDIS3L2, 3.1.13.- {ECO:0000255|HAMAP-Rule:MF_03045}, DIS3L2 {ECO:0000255|HAMAP-Rule:MF_03045}, FAM6A
Dilution	Flow-Cyt=2ug/Test
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.

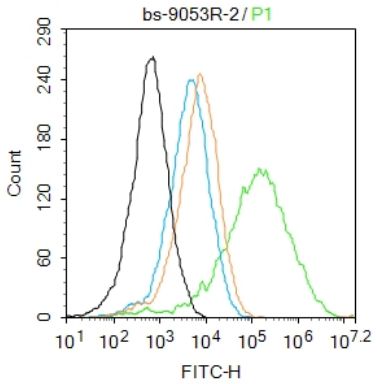
Protein Information

Name	DIS3L2 {ECO:0000255|HAMAP-Rule:MF_03045}
Synonyms	FAM6A
Function	3'-5'-exoribonuclease that specifically recognizes RNAs polyuridylated at their 3' end and mediates their degradation. Component of an exosome-independent RNA degradation pathway that mediates degradation of both mRNAs and miRNAs that have been polyuridylated by a terminal uridylyltransferase, such as ZCCHC11/TUT4. Mediates degradation of cytoplasmic mRNAs that have been deadenylated and subsequently uridylated at their 3'. Mediates degradation of uridylated pre-let-7 miRNAs, contributing to the maintenance of embryonic stem (ES) cells. Essential for correct mitosis, and negatively regulates cell proliferation.
Cellular Location	Cytoplasm. Cytoplasm, P-body

Background

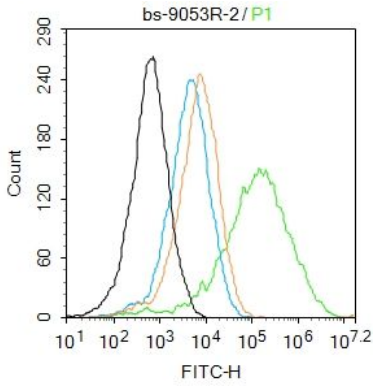
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Images



Blank control:Mouse kidney.
Primary Antibody (green line): Rabbit Anti-DIS3L2 antibody (AP59123)
Dilution: 2 µg /10^6 cells;
Isotype Control Antibody (orange line): Rabbit IgG .
Secondary Antibody : Goat anti-rabbit IgG-AF488
Dilution: 1 µg /test.
Protocol
The cells were fixed with 4% PFA (10min at room temperature)and then permeabilized with 0.1% PBST for 20 min at room temperature. The cells were then incubated in 5%BSA to block non-specific protein-protein interactions for 30 min at room temperature .Cells stained with Primary Antibody for 30 min at room temperature. The secondary antibody used for 40 min at room temperature. Acquisition of 20,000 events was performed.

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Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.