

C7orf27 Rabbit pAb

Catalog # AP96796

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	Q6PJG6
Other Accession	Q6PJG6
Reactivity	Human, Mouse
Predicted	Rat
Host	Rabbit
Clonality	Polyclonal
Calculated MW	88119
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human BAAT1/C7orf27
Epitope Specificity	21-120/821
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Contains 2 HEAT repeats.
SUBUNIT	Interacts with BRCA1 and ATM.
DISEASE	Defects in BRAT1 are the cause of rigidity and multifocal seizure syndrome, lethal neonatal (RMFSL) [MIM:614498]. A lethal, neonatal, neurologic disorder characterized by episodic jerking that is apparent in utero, lack of psychomotor development, axial and limb rigidity, frequent multifocal seizures, and dysautonomia. At birth, affected individuals have small heads, overlapping cranial sutures, small or absent fontanel, and depressed frontal bones. Infants show poorly responsive focal jerks of the tongue, face and arms in a nearly continuous sequence throughout life.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Additional Information

Gene ID	221927
Other Names	BRAT1_HUMAN; BRAT1; BRCA1-associated ATM activator 1; BRCA1-associated protein required for ATM activation protein 1; BAAT1; C7orf27; BRAT1_MOUSE; D3ZUM1_RAT; BRCA1 associated ATM activator 1; chromosome 7 open reading frame 27
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
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	BRAT1 {ECO:0000303 PubMed:25657994, ECO:0000312 HGNC:HGNC:21701}
Function	Component of a multiprotein complex required for the assembly of the RNA endonuclease module of the integrator complex (PubMed: 39032489 , PubMed: 39032490). Associates with INTS9 and INTS11 in the cytoplasm and blocks the active site of INTS11 to inhibit the endonuclease activity of INTS11 before formation of the full integrator complex (PubMed: 39032489 , PubMed: 39032490). Following dissociation of WDR73 of the complex, BRAT1 facilitates the nuclear import of the INTS9-INTS11 heterodimer (PubMed: 39032489). In the nucleus, INTS4 is integrated to the INTS9-INTS11 heterodimer and BRAT1 is released from the mature RNA endonuclease module by inositol hexakisphosphate (InsP6) (PubMed: 39032489). BRAT1 is also involved in DNA damage response; activates kinases ATM, SMC1A and PRKDC by modulating their phosphorylation status following ionizing radiation (IR) stress (PubMed: 16452482 , PubMed: 22977523). Plays a role in regulating mitochondrial function and cell proliferation (PubMed: 25070371). Required for protein stability of MTOR and MTOR-related proteins, and cell cycle progress by growth factors (PubMed: 25657994).
Cellular Location	Nucleus. Cytoplasm Note=Present at double strand breaks (DSBs)following ionizing radiation treatment. The ubiquitinated form localizes in the nucleus in a NDFIP1- dependent manner.
Tissue Location	Ubiquitously expressed.

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