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UBE3A Rabbit pAb

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Catalog # AP94180

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q05086
Reactivity	Human
Predicted	Rat, Pig, Dog, Mouse, Rabbit, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	100688
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human UBE3A/E6-AP
Epitope Specificity	701-800/875
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	Nucleus.
DISEASE	Defects in UBE3A are a cause of Angelman syndrome (AS) [MIM:105830]; also known as 'happy puppet syndrome'. AS is characterized by features of severe motor and intellectual retardation, microcephaly, ataxia, frequent jerky limb movements and flapping of the arms and hands, hypotonia, hyperactivity, hypopigmentation, seizures, absence of speech, frequent smiling and episodes of paroxysmal laughter, and an unusual facies characterized by macrostomia, a large mandible and open-mouthed expression, a great propensity for protruding the tongue ('tongue thrusting'), and an occipital groove.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	E6-associating protein is a component of the ubiquitin-mediated proteolytic pathway, which selectively targets proteins for degradation by the 26S proteasome. Ubiquitin (Ub) is directly conjugated to protein substrates by the transfer of Ub from an E2 ubiquitin conjugating enzyme to the target protein. This conjugation is facilitated by the enzymatic activity of E3 ubiquitin ligase family members such as E6-AP. Several substrates of E6-AP have been identified and include the tumor suppressor protein p53 and the mammalian homolog of Rad23, HHR23A. Previous studies have indicated that E6-AP associates with the human papillomavirus E6 oncogene, which complexes with p53 and thereby potentiates E6-AP mediated ubiquitination of p53. Genetic mutations that impair E6-AP activity result in the accumulation of p53 in the cytoplasm, and, in many instances, these mutations are associated with the development of the rare neurodevelopmental disorder Angelman syndrome (AS), which is characterized by severe motor dysfunction and mental retardation.

Additional Information

Gene ID	7337
Other Names	Ubiquitin-protein ligase E3A, 2.3.2.26, E6AP ubiquitin-protein ligase, HECT-type ubiquitin transferase E3A, Human papillomavirus E6-associated protein, Oncogenic protein-associated protein E6-AP, Renal carcinoma antigen NY-REN-54, UBE3A (HGNC:12496)
Dilution	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

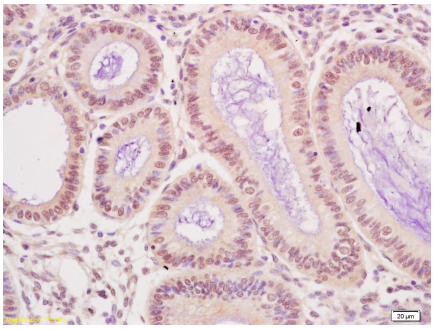
Name	UBE3A (HGNC:12496)
Function	E3 ubiquitin-protein ligase which accepts ubiquitin from an E2 ubiquitin-conjugating enzyme in the form of a thioester and transfers it to its substrates (PubMed:10373495, PubMed:16772533, PubMed:19204938, PubMed:19233847, PubMed:19325566, PubMed:19591933, PubMed:22645313, PubMed:24273172, PubMed:24728990, PubMed:30020076). Several substrates have been identified including the BMAL1, ARC, LAMTOR1, RAD23A and RAD23B, MCM7 (which is involved in DNA replication), annexin A1, the PML tumor suppressor, and the cell cycle regulator CDKN1B (PubMed:10373495, PubMed:19204938, PubMed:19325566, PubMed:19591933, PubMed:22645313, PubMed:24728990, PubMed:30020076). Additionally, may function as a cellular quality control ubiquitin ligase by helping the degradation of the cytoplasmic misfolded proteins (PubMed:19233847). Finally, UBE3A also promotes its own degradation in vivo. Plays an important role in the regulation of the circadian clock: involved in the ubiquitination of the core clock component BMAL1, leading to its proteasomal degradation (PubMed:24728990). Acts as transcriptional coactivator of progesterone receptor PGR upon progesterone hormone activation (PubMed:16772533). Acts as a regulator of synaptic development by mediating ubiquitination and degradation of ARC (By similarity). Required for synaptic remodeling in neurons by mediating ubiquitination and degradation of LAMTOR1, thereby limiting mTORC1 signaling and activity-dependent synaptic remodeling (By similarity). Synergizes with WBP2 in enhancing PGR activity (PubMed:16772533).
Cellular Location	Cytoplasm {ECO:0000250|UniProtKB:O08759}. Nucleus {ECO:0000250|UniProtKB:O08759}

Background

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

Images

Tissue/cell: human breast carcinoma; 4%



Paraformaldehyde-fixed and paraffin-embedded;
Antigen retrieval: citrate buffer (0.01M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37℃ for 20 min;
Incubation: Anti-UBE3A Polyclonal Antibody, Unconjugated(AP94180) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining

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