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# UBE3A Rabbit pAb

UBE3A Rabbit pAb
Catalog # AP94180

## Product Information

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| **Application** | IHC-P, IHC-F, IF |
| **Primary Accession** | Q96GR7 |
| **Reactivity** | Human |
| **Host** | Rabbit |
| **Clonality** | Polyclonal |
| **Calculated MW** | 105 KDa |
| **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. |
| **SIMILARITY** | Contains 1 HECT (E6AP-type E3 ubiquitin-protein ligase) domain. |
| **Post-translational modifications** | Phosphorylated upon DNA damage, probably by ATM or ATR. |
| **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

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| **Dilution** | IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500 |
| **Format** | 0.01M TBS(pH7.4) with 1% BSA, 0.09% (W/V) sodium azide and 50% Glyce |
| **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

## Background

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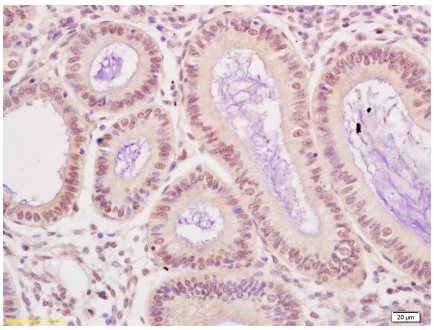
## Images



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