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# Kindlin Rabbit pAb

Kindlin Rabbit pAb
Catalog # AP56543

## Product Information

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| **Application** | IHC-P, IHC-F, IF |
| **Primary Accession** | Q9BQL6 |
| **Reactivity** | Human, Mouse |
| **Predicted** | Rat, Dog, Pig, Horse, Rabbit, Sheep |
| **Host** | Rabbit |
| **Clonality** | Polyclonal |
| **Calculated MW** | 77437 |
| **Physical State** | Liquid |
| **Immunogen** | KLH conjugated synthetic peptide derived from human Kindlin |
| **Epitope Specificity** | 601-677/677 |
| **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 > cytoskeleton. Cell junction > focal adhesion. Cell projection > ruffle membrane. Constituent of focal adhesions. Localized at the basal aspect of skin keratinocytes, close to the cell membrane. Colocalizes with filamentous actin. Upon TGFB1 treatment, it localizes to membrane ruffles. |
| **SIMILARITY** | Belongs to the kindlin family. Contains 1 FERM domain. Contains 1 PH domain. |
| **DISEASE** | Defects in FERMT1 are the cause of Kindler syndrome (KINDS) [MIM:173650]. An autosomal recessive skin disorder characterized by skin blistering, photosensitivity, progressive poikiloderma, and extensive skin atrophy. Additional clinical features include gingival erosions, ocular, esophageal, gastrointestinal and urogenital involvement, and an increased risk of mucocutaneous malignancy. Note=Although most FERMT1 mutations are predicted to lead to premature termination of translation, and to loss of FERMT1 function, significant clinical variability is observed among patients. There is an association of FERMT1 missense and in-frame deletion mutations with milder disease phenotypes, and later onset of complications (PubMed:21936020). |
| **Important Note** | This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications. |
| **Background Descriptions** | This gene encodes a member of the fermitin family, and contains a FERM domain and a pleckstrin homology domain. The encoded protein is involved in integrin signaling and linkage of the actin cytoskeleton to the extracellular matrix. Mutations in this gene have been linked to Kindler syndrome. [provided by RefSeq, Dec 2009] |

## Additional Information

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| **Gene ID** | 55612 |

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| **Other Names** | Fermitin family homolog 1, Kindlerin, Kindlin syndrome protein, Kindlin-1, Unc-112-related protein 1, FERMT1, C20orf42, KIND1, URP1 |
| **Target/Specificity** | Expressed in brain, skeletal muscle, kidney, colon, adrenal gland, prostate, and placenta. Weakly or not expressed in heart, thymus, spleen, liver, small intestine, bone marrow, lung and peripheral blood leukocytes. Overexpressed in some colon and lung tumors. In skin, it is localized within the epidermis and particularly in basal keratocytes. Not detected in epidermal melanocytes and dermal fibroblasts. |
| **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

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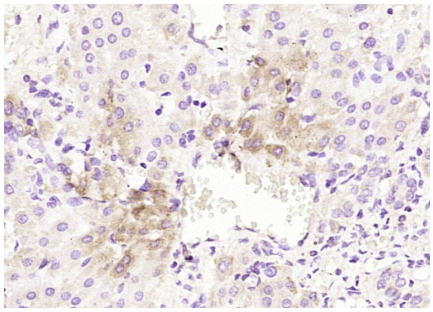
| **Name** | FERMT1 |
| **Synonyms** | C20orf42, KIND1, URP1 |
| **Function** | Involved in cell adhesion. Contributes to integrin activation. When coexpressed with talin, potentiates activation of ITGA2B. Required for normal keratinocyte proliferation. Required for normal polarization of basal keratinocytes in skin, and for normal cell shape. Required for normal adhesion of keratinocytes to fibronectin and laminin, and for normal keratinocyte migration to wound sites. May mediate TGF-beta 1 signaling in tumor progression. |
| **Cellular Location** | Cytoplasm, cytoskeleton. Cell junction, focal adhesion. Cell projection, ruffle membrane; Peripheral membrane protein; Cytoplasmic side. Note=Constituent of focal adhesions Localized at the basal aspect of skin keratinocytes, close to the cell membrane. Colocalizes with filamentous actin. Upon TGFB1 treatment, it localizes to membrane ruffles |
| **Tissue Location** | Expressed in brain, skeletal muscle, kidney, colon, adrenal gland, prostate, and placenta. Weakly or not expressed in heart, thymus, spleen, liver, small intestine, bone marrow, lung and peripheral blood leukocytes. Overexpressed in some colon and lung tumors. In skin, it is localized within the epidermis and particularly in basal keratocytes. Not detected in epidermal melanocytes and dermal fibroblasts. |

## Background

This gene encodes a member of the fermitin family, and contains a FERM domain and a pleckstrin homology domain. The encoded protein is involved in integrin signaling and linkage of the actin cytoskeleton to the extracellular matrix. Mutations in this gene have been linked to Kindler syndrome. [provided by RefSeq, Dec 2009]

## Images

Paraformaldehyde-fixed, paraffin embedded (human liver); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3%



hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Kindlin) Polyclonal Antibody, Unconjugated (AP56543) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.