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C4d-A Rabbit pAb

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Catalog # AP54268

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P0C0L4
Host	Rabbit
Clonality	Polyclonal
Calculated MW	192785
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human C4d-A
Epitope Specificity	1001-1100/1744
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	Secreted.
SIMILARITY	Contains 1 anaphylatoxin-like domain.Contains 1 NTR domain.
SUBUNIT	This protein is synthesized as a single-chain precursor and, prior to secretion, is enzymatically cleaved to form a trimer of non-identical chains (alpha, beta and gamma).
Post-translational modifications	Prior to secretion, the single-chain precursor is enzymatically cleaved to yield non-identical chains alpha, beta and gamma. During activation, the alpha chain is cleaved by C1 into C4a and C4b, and C4b stays linked to the beta and gamma chains. Further degradation of C4b by C1 into the inactive fragments C4c and C4d blocks the generation of C3 convertase. The proteolytic cleavages often are incomplete so that many structural forms can be found in plasma. N- and O-glycosylated. O-glycosylated with a core 1 or possibly core 8 glycan.
DISEASE	Defects in C4A are the cause of complement component 4A deficiency (C4AD). A rare defect of the complement classical pathway associated with the development of autoimmune disorders, mainly systemic lupus with or without associated glomerulonephritis.
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 the acidic form of complement factor 4, part of the classical activation pathway. The protein is expressed as a single chain precursor which is proteolytically cleaved into a trimer of alpha, beta, and gamma chains prior to secretion. The trimer provides a surface for interaction between the antigen-antibody complex and other complement components. The alpha chain may be cleaved to release C4 anaphylatoxin, a mediator of local inflammation. Deficiency of this protein is associated with systemic lupus erythematosus and type I diabetes mellitus. This gene localizes to the major histocompatibility complex (MHC) class III region on chromosome 6. Varying haplotypes of this gene cluster exist, such that individuals may have 1, 2, or 3 copies of this gene. Two transcript variants encoding different isoforms have been found for this gene. [provided by RefSeq, Nov 2011].

Additional Information

Gene ID	720;721
Other Names	Complement C4-A, Acidic complement C4, C3 and PZP-like alpha-2-macroglobulin domain-containing protein 2, Complement C4 beta chain, Complement C4-A alpha chain, C4a anaphylatoxin, Complement C4b-A, Complement C4b-alpha' chain, Complement C4d-A, Complement C4 gamma chain, C4A {ECO:0000303|PubMed:6546707, ECO:0000312|HGNC:HGNC:1323}
Target/Specificity	Complement component C4 is expressed at highest levels in the liver, at moderate levels in the adrenal cortex, adrenal medulla, thyroid gland,and the kidney, and at lowest levels in the heart, ovary, small intestine, thymus, pancreas and spleen. The extra-hepatic sites of expression may be important for the local protection and inflammatory response.
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.

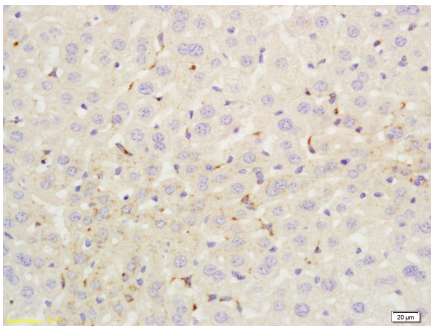
Protein Information

Name	C4A {ECO:0000303|PubMed:6546707, ECO:0000312|HGNC:HGNC:1323}
Function	Precursor of non-enzymatic components of the classical, lectin and GZMK complement pathways, which consist in a cascade of proteins that leads to phagocytosis and breakdown of pathogens and signaling that strengthens the adaptive immune system.
Cellular Location	Secreted. Synapse Cell projection, axon. Cell projection, dendrite [Complement C4b-A]: Secreted. Cell surface. Note=Covalently associated with the surface of pathogens: the internal thioester bond reacts with carbohydrate antigens on the target surface to form amide or ester bonds.
Tissue Location	Complement component C4 is expressed at highest levels in the liver, at moderate levels in the adrenal cortex, adrenal medulla, thyroid gland, and the kidney, and at lowest levels in the heart, ovary, small intestine, thymus, pancreas and spleen (PubMed:11367523). The extra-hepatic sites of expression may be important for the local protection and inflammatory response (PubMed:11367523).

Background

This gene encodes the acidic form of complement factor 4, part of the classical activation pathway. The protein is expressed as a single chain precursor which is proteolytically cleaved into a trimer of alpha, beta, and gamma chains prior to secretion. The trimer provides a surface for interaction between the antigen-antibody complex and other complement components. The alpha chain may be cleaved to release C4 anaphylatoxin, a mediator of local inflammation. Deficiency of this protein is associated with systemic lupus erythematosus and type I diabetes mellitus. This gene localizes to the major histocompatibility complex (MHC) class III region on chromosome 6. Varying haplotypes of this gene cluster exist, such that individuals may have 1, 2, or 3 copies of this gene. Two transcript variants encoding different isoforms have been found for this gene. [provided by RefSeq, Nov 2011].

Images



Tissue/cell: mouse liver tissue; 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-C4d-A Polyclonal Antibody, Unconjugated(AP54268) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining