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C4a+C4b Rabbit pAb

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Catalog # AP55856

Product Information

Application	E
Primary Accession	P0C0L5
Predicted	Human
Host	Rabbit
Clonality	Polyclonal
Calculated MW	192751
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human C4a and C4b Complement C4 beta chain
Epitope Specificity	21-120/1744
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	Circulates in blood as a disulfide-linked trimer of an alpha, beta and gamma chain.
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) [MIM:614380]. A rare defect of the complement classical pathway associated with the development of autoimmune disorders, mainly systemic lupus with or without associated glomerulonephritis. Defects in C4A are a cause of susceptibility to systemic lupus erythematosus (SLE) [MIM:152700]. A chronic, inflammatory and often febrile multisystemic disorder of connective tissue. It affects principally the skin, joints, kidneys and serosal membranes. It is thought to represent a failure of the regulatory mechanisms of the autoimmune system. Note=Interindividual copy-number variation (CNV) of complement component C4 and associated polymorphisms result in different susceptibilities to SLE. The risk of SLE susceptibility has been shown to be significantly increased among subjects with only two copies of total C4. A high copy number is a protective factor against SLE.
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	100293534;721
Other Names	Complement C4-B, Basic complement C4, C3 and PZP-like alpha-2-macroglobulin domain-containing protein 3, Complement C4 beta chain, Complement C4-B alpha chain, C4a anaphylatoxin, Complement C4b-B, C4d-B, Complement C4 gamma chain, C4B {ECO:0000303|PubMed:6546707, ECO:0000312|HGNC:HGNC:1324}
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	ELISA=1:5000-10000
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	C4B {ECO:0000303|PubMed:6546707, ECO:0000312|HGNC:HGNC:1324}
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 {ECO:0000250|UniProtKB:P0C0L4}. Synapse. Cell projection, axon. Cell projection, dendrite [Complement C4b-B]: 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. The extra- hepatic sites of expression may be important for the local protection and inflammatory response.

Background

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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].