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Rabbit Anti-Complement C4 gamma chain antibody

SL10454R

Product Name:	Complement C4 gamma chain
Chinese Name:	补体C4γ链蛋白抗体
Alias:	Complement C4 gamma chain; complement C4-A proprotein; Acidic complement C4; Basic complement C4; CH; Chido blood group; Complement component 4A; Complement component 4B; RG; Rodgers blood group; CO4A_HUMAN; CO4B_HUMAN; C4A; C4; C4A2; C4A3; C4A4; C4A6; C4AD; C4S; CO4; CPAMD2; RG.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	32/190kDa
Cellular localization:	Secretory protein
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Complement C4 gamma chain:1454-1550/1744
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed

Product Detail:	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]. **Function:** Non-enzymatic component of C3 and C5 convertases and thus essential for the propagation of the classical complement pathway. Covalently binds to immunoglobulins and immune complexes and enhances the solubilization of immune aggregates and the clearance of IC through CR1 on erythrocytes. C4A isotype is responsible for effective binding to form amide bonds with immune aggregates or protein antigens, while C4B isotype catalyzes the transacylation of the thioester carbonyl group to form ester bonds with carbohydrate antigens. Derived from proteolytic degradation of complement C4, C4a anaphylatoxin is a mediator of local inflammatory process. It induces the contraction of smooth muscle, increases vascular permeability and causes histamine release from mast cells and basophilic leukocytes. **Subunit:** Circulates in blood as a disulfide-linked trimer of an alpha, beta and gamma chain. **Subcellular Location:** Secreted. **Tissue 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. **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.

Similarity:
Contains 1 anaphylatoxin-like domain.
Contains 1 NTR domain.

SWISS:
P0C0L4

Gene ID:
720

Database links:

Entrez Gene: 720Human

Entrez Gene: 721Human

Omim: 120810Human

Omim: 120820Human

SwissProt: P0C0L4Human

SwissProt: P0C0L5Human

Unigene: 534847Human

Unigene: 720022Human

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