

Product name:	CABC1 Rabbit Polyclonal Antibody
Cat number:	ABN07802
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human ADCK3. AA range:301-350
Reactivity:	Human,Mouse,Rat
Applications:	WB 1:500-1:2000,IHC 1:100-1:300,ICC/IF 1:200-1:1000,ELISA 1:5000-1:20000
Molecular Weight:	70kDa
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

Background:

This gene encodes a mitochondrial protein similar to yeast ABC1, which functions in an electron-transferring membrane protein complex in the respiratory chain. It is not related to the family of ABC transporter proteins. Expression of this gene is induced by the tumor suppressor p53 and in response to DNA damage, and inhibiting its expression partially suppresses p53-induced apoptosis. Alternatively spliced transcript variants have been found; however, their full-length nature has not been determined. [provided by RefSeq, Jul 2008],disease:Defects in CABC1 are a cause of coenzyme Q10 deficiency [MIM:607426]; also known as primary CoQ10 deficiency. Coenzyme Q10 deficiency patients present a progressive neurological disorder with cerebellar atrophy, developmental delay, and hyperlactatemia.,disease:Defects in CABC1 are the cause of spinocerebellar ataxia autosomal recessive type 9 (SCAR9) [MIM:612016]; also known as autosomal recessive cerebellar ataxia type 2 (ARCA2). Spinocerebellar ataxia is a clinically and genetically heterogeneous group of cerebellar disorders. Patients show progressive incoordination of gait and often poor coordination of hands, speech and eye movements, due to degeneration of the cerebellum with variable involvement of the brainstem and spinal cord. SCAR9 is an autosomal recessive form characterized by gait ataxia and cerebellar atrophy with slow progression and few associated features. Patients can manifest brisk tendon reflexes and Hoffmann sign, mild psychomotor retardation, mild axonal degeneration of the sural nerve, exercise intolerance and elevated serum lactate.,function:May be a chaperone-like protein essential for the proper conformation and functioning of protein complexes in the respiratory chain.,induction:By p53.,similarity:Belongs to the protein kinase superfamily. ADCK protein kinase family.,similarity:Contains 1 protein kinase domain.,tissue specificity:Ubiquitously expressed with a relatively greater abundance in heart and skeletal muscle.,