Datasheet for ABIN881438

anti-COQ8A antibody (AA 501-647) (Alexa Fluor 647)

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Overview

Quantity:	100 µL
Target:	COQ8A
Binding Specificity:	AA 501-647
Reactivity:	Human, Rat
Host:	Rabbit
Clonality:	Polyclonal
Conjugate:	This COQ8A antibody is conjugated to Alexa Fluor 647
Application:	Western Blotting (WB), Immunofluorescence (Cultured Cells) (IF (cc)), Immunofluorescence (Paraffin-embedded Sections) (IF (p))

Product Details

Immunogen:	KLH conjugated synthetic peptide derived from human ADCK3/CABC1
Isotype:	IgG
Cross-Reactivity:	Human, Rat
Predicted Reactivity:	Mouse,Dog,Cow,Sheep,Pig,Horse,Chicken,Rabbit
Purification:	Purified by Protein A.

Target Details

Target:	COQ8A
Alternative Name:	CABC1 (COQ8A Products)

Target Details

Background:	Synonyms: mitochondrial, aarF domain containing protein kinase 3, aarF domain-containing protein kinase 3, ADCK 3, ADCK3, ADCK3_HUMAN, CABC 1, Chaperone ABC1 activity of bc1 complex S.pombe like, Chaperone ABC1 activity of bc1 complex homolog, Chaperone ABC1 like, Chaperone activity of bc1 complex like, Chaperone activity of bc1 complex like mitochondrial, Chaperone activity of bc1 complex-like, Chaperone-ABC1-like, Coenzyme Q8 homolog, COQ 8, COQ8. Background: May be a chaperone-like protein essential for the proper conformation and functioning of protein complexes in the respiratory chain.Tissue specificity:Ubiquitously expressed with a relatively greater abundance in heart and skeletal muscle.Involvement in disease:Defects in ADCK3 are a cause of coenzyme Q10 deficiency (CoQ10 deficiency). CoQ10 deficiency is an autosomal recessive disorder with variable manifestations. It can be associated with three main clinical phenotypes: a predominantly myopathic form with central nervous system involvement, an infantile encephalomyopathy with renal dysfunction and an ataxic form with cerebellar atrophy.Defects in ADCK3 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.

Application Details

Application Notes:	IF(IHC-P) 1:50-200
	IF(IHC-F) 1:50-200
	IF(ICC) 1:50-200

Restrictions:	For Research Use only

Handling

Format:	Liquid
Concentration:	1 μg/μL
Buffer:	Aqueous buffered solution containing 0.01M TBS (pH 7.4) with 1 % BSA, 0.03 % Proclin300 and

Handling

	50 % Glycerol.
Preservative:	ProClin
Precaution of Use:	This product contains ProClin: a POISONOUS AND HAZARDOUS SUBSTANCE, which should be handled by trained staff only.
Storage:	-20 °C
Storage Comment:	Store at -20°C. Aliquot into multiple vials to avoid repeated freeze-thaw cycles.
Expiry Date:	12 months