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Produktinformation

- Forschungsprodukte & Biochemikalien
- Zellkultur & Verbrauchsmaterial
- Diagnostik & molekulare Diagnostik
- Laborgeräte & Service

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See the following pages for more information!

Lieferung & Zahlungsart

siehe unsere Liefer- und Versandbedingungen

Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

SZABO-SCANDIC HandelsgmbH

Quellenstraße 110, A-1100 Wien

T. +43(0)1 489 3961-0

F. +43(0)1 489 3961-7

mail@szabo-scandic.com

www.szabo-scandic.com

linkedin.com/company/szaboscandic

Aprataxin. Rabbit Antigen Immunoaffinity Purified Polyclonal

Forkhead-associated domain histidine triad-like protein; FHA-HIT; APTX; AXA1

Background

DNA-binding protein involved in single-strand DNA break repair, double-strand DNA break repair and base excision repair. Resolves abortive DNA ligation intermediates formed either at base excision sites, or when DNA ligases attempt to repair non-ligatable breaks induced by reactive oxygen species. Catalyzes the release of adenylate groups covalently linked to 5'-phosphate termini, resulting in the production of 5'-phosphate termini that can be efficiently rejoined. Also able to hydrolyze adenosine 5'-monophosphoramidate (AMP-NH2) and diadenosine tetraphosphate (AppppA), but with lower catalytic activity. Protein is widely expressed. Defects in APTX are the cause of ataxia-oculomotor apraxia syndrome, an autosomal recessive syndrome characterized by early-onset cerebellar ataxia, oculomotor apraxia, early areflexia and late peripheral neuropathy. Also a cause of coenzyme Q10 deficiency. Coenzyme Q10 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. Coenzyme Q10 deficiency due to APTX mutations is typically associated with cerebellar ataxia.

Ordering Information

Catalog Number
X2725P

Size
10 Miniblots

Form
Affinity Purified

Host/Clone
Rabbit

Formulation
Provided as solution in phosphate buffered saline with 0.08% sodium azide

Concentration
See vial for concentration

Isotype

Applications
Western blotting, ELISA

Species Reactivity
Human, Rat, Mouse

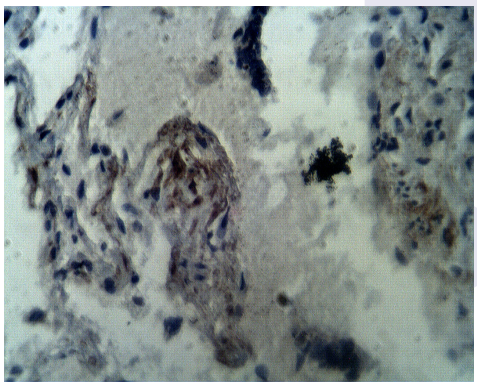
Accession Number

Human	Q7Z2E3
Rat	Q8K4H4
Mouse	Q7TQC5

Immunogen

Synthetic peptide derived from the human aprataxin protein.

Immunohistochemical staining of normal human lung tissue using Aprataxin antibody (Cat. No. X2725P) at 10 μg/ml.



Exalpha Biologicals, Inc. 2 Shaker Road, Bldg. B101 Shirley, MA 01464
Tel: 800.395.1137 Fax: 866.924.5100 www.exalpha.com info@exalpha.com

Positive Control/Tissue Expression

Human lung tissue

Comments

Antibody can be used for Western blotting (1:400 dilution). Optimal concentration should be evaluated by serial dilutions.

Purification

Antigen Immunoaffiinity Purification

Ship Conditions

Ship at ambient temperature, freeze upon arrival

Storage Customer

Product should be stored at -20ºC. Aliquot to avoid freeze/thaw cycles

Stability

Products are stable for one year from purchase when stored properly

References

1. Ahel, I., et al. 'The neurodegenerative disease protein aprataxin resolves abortive DNA ligation intermediates.' Nature 2006, 443, 713-716.

2. Kijas, A.W., et al. 'Aprataxin forms a discrete branch in the HIT (histidine triad) superfamily of proteins with both DNA/RNA binding and nucleotide hydrolase activities.' J. Biol. Chem. 2006, 281, 13939-13948.

3. Rass, U., et al. 'Actions of aprataxin in multiple DNA repair pathways.' J. Biol. Chem. 2007, 282, 9469-9474.

4. Rass, U., et al. 'Molecular mechanism of DNA deadenylation by the neurological disease protein aprataxin.' J. Biol. Chem. 2008, 283, 33994-34001.

Product Specific References

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