

Product datasheet

Recombinant Human PCCB protein ab127093

Description

Product name	Recombinant Human PCCB protein
Purity	= 90 % SDS-PAGE. Purified via His tag
Expression system	Escherichia coli
Accession	<u>P05166</u>
Protein length	Protein fragment
Animal free	No
Nature	Recombinant
Species	Human
Predicted molecular weight	28 kDa
Amino acids	7 to 266

Specifications

Our **Abpromise guarantee** covers the use of **ab127093** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Applications	SDS-PAGE
Form	Lyophilized

Preparation and Storage

Stability and Storage	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles. Constituents: 0.32% Tris HCl, 0.58% Sodium chloride
Reconstitution	Reconstitute with water to desired concentration.

General Info

Pathway	Metabolic intermediate metabolism; propanoyl-CoA degradation; succinyl-CoA from propanoyl-CoA: step 1/3.
Involvement in disease	Defects in PCCB are the cause of propionic acidemia type II (PA-2) [MIM:606054]. PA-2 is a life-threatening disease characterized by episodic vomiting, lethargy and ketosis, neutropenia,

periodic thrombocytopenia, hypogammaglobulinemia, developmental retardation, and intolerance to protein.

Sequence similarities

Belongs to the AccD/PCCB family.
Contains 1 carboxyltransferase domain.

Cellular localization

Mitochondrion matrix.

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

Our Abpromise to you: Quality guaranteed and expert technical support

- Replacement or refund for products not performing as stated on the datasheet
- Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours

- We provide support in Chinese, English, French, German, Japanese and Spanish
- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

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