abcam

Product datasheet

Recombinant Human QDPR/DHPR protein ab113148

1 Image

Description

Product name	Recombinant Human QDPR/DHPR protein
Purity	> 90 % SDS-PAGE. ab113148 was purified using conventional chromatography techniques.
Expression system	Escherichia coli
Accession	**P09417**
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Predicted molecular weight	28 kDa including tags
Amino acids	1 to 244
Tags	His tag N-Terminus

Specifications

Our **Abpromise guarantee** covers the use of **ab113148** 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 Mass Spectrometry
Mass spectrometry	MALDI-TOF
Form	Liquid
Additional notes	Previously labelled as QDPR.

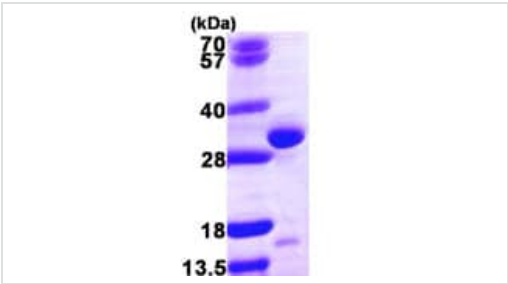
Preparation and Storage

Stability and Storage	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles. pH: 8.5 Constituents: 0.04% DTT, 0.32% Tris HCl, 10% Glycerol (glycerin, glycerine)

General Info

Function	The product of this enzyme, tetrahydrobiopterin (BH-4), is an essential cofactor for phenylalanine, tyrosine, and tryptophan hydroxylases.
Involvement in disease	Defects in QDPR are the cause of BH4-deficient hyperphenylalaninemia type C (HPABH4C) [MIM:261630]; also called dihydropteridine reductase deficiency (DHPR deficiency) or hyperphenylalaninemia tetrahydrobiopterin-deficient due to DHPR deficiency or quinoid dihydropteridine reductase deficiency (QDPR deficiency). HPABH4C is a rare autosomal recessive disorder characterized by hyperphenylalaninemia and severe neurologic symptoms (malignant hyperphenylalaninemia) including axial hypotonia and truncal hypertonia, abnormal thermogenesis, and microcephaly. These signs are attributable to depletion of the neurotransmitters dopamine and serotonin, whose syntheses are controlled by tryptophan and tyrosine hydroxylases that use BH-4 as cofactor. These patients do not respond to phenylalanine-restricted diet. HPABH4C is lethal if untreated.
Sequence similarities	Belongs to the short-chain dehydrogenases/reductases (SDR) family.

Images



SDS-PAGE - Recombinant Human QDPR/DHPR protein (ab113148)

15% SDS-PAGE showing ab113148 (3 µg) at approximately 28.2 kDa.

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- Replacement or refund for products not performing as stated on the datasheet
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- 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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