

Recombinant Human PDHB protein ab152601

1 图像

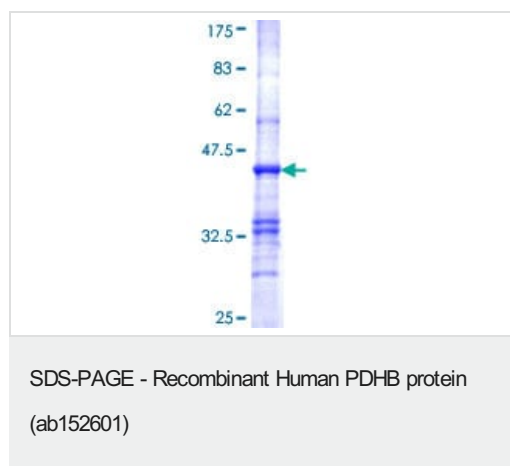
描述		
产品名称	重组人PDHB蛋白	
表达系统	Wheat germ	
蛋白长度	Protein fragment	
无动物成分	No	
性质	Recombinant	
种属	Human	
序列	LEAAAVLSKEGVECEVINMRTIRPMDMETIEASVMKTNHLVT VEGGWPQF GVGAEICARIMEGPAFNFLDAPAVRVTGADVMPYAKILEDN SIPQVKDI IFAIKKTLNI	
氨基酸	250 to 359	
标签	GST tag N-Terminus	

技术指标	
Our Abpromise guarantee covers the use of ab152601 in the following tested applications.	
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.	
应用	Western blot ELISA
形式	Liquid
补充说明	This product was previously labelled as Pyruvate Dehydrogenase E1 beta subunit.

制备和贮存	
稳定性和存储	Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.31% Glutathione, 0.79% Tris HCl

功能	The pyruvate dehydrogenase complex catalyzes the overall conversion of pyruvate to acetyl-CoA and CO ₂ . It contains multiple copies of three enzymatic components: pyruvate dehydrogenase (E1), dihydrolipoamide acetyltransferase (E2) and lipoamide dehydrogenase (E3).
疾病相关	Defects in PDHB are a cause of pyruvate dehydrogenase E1 component deficiency (PDHE1 deficiency) [MIM:312170]. PDHE1 deficiency is the most common enzyme defect in patients with primary lactic acidosis. It is associated with variable clinical phenotypes ranging from neonatal death to prolonged survival complicated by developmental delay, seizures, ataxia, apnea, and in some cases to an X-linked form of Leigh syndrome (LS) (Leigh encephalomyelopathy).
细胞定位	Mitochondrion matrix.

图片



ab152601 on a 12.5% SDS-PAGE stained with Coomassie Blue.

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

If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit <https://www.abcam.cn/abpromise> or contact our technical team.

Terms and conditions

- Guarantee only valid for products bought direct from Abcam or one of our authorized distributors