abcam

Product datasheet

Recombinant Human CPS1 protein ab152296

1 Image

Description

Product name	Recombinant Human CPS1 protein
Expression system	Wheat germ
Accession	P31327
Protein length	Protein fragment
Animal free	No
Nature	Recombinant
Species	Human
Sequence	ANNVPATPVAWPSQEGQNPSLSSIRKLIRDGSIDLVINLPN NNTKFVHDN YVIRRTAVDSGIPLLTNFQVTKLFAEAVQKSRKVDSKSLFH YRQYSAGKA A
Predicted molecular weight	37 kDa including tags
Amino acids	1400 to 1500

Specifications

Our **Abpromise guarantee** covers the use of **ab152296** in the following tested applications.

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

Applications	Western blot SDS-PAGE ELISA
Form	Liquid
Additional notes	

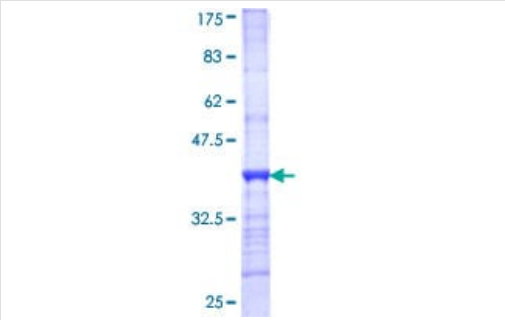
Preparation and Storage

Stability and Storage	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.31% Glutathione, 0.79% Tris HCl

General Info

Function	Involved in the urea cycle of ureotelic animals where the enzyme plays an important role in removing excess ammonia from the cell.
Tissue specificity	Primarily in the liver and small intestine.
Involvement in disease	Defects in CPS1 are the cause of carbamoyl phosphate synthetase 1 deficiency (CPS1D) [MIM:237300]. CPS1D is an autosomal recessive disorder of the urea cycle causing hyperammonemia. Clinical features include protein intolerance, intermittent ataxia, seizures, lethargy, developmental delay and mental retardation. Note=Genetic variations in CPS1 influence the availability of precursors for nitric oxide (NO) synthesis and play a role in clinical situations where endogenous NO production is critically important, such as neonatal pulmonary hypertension, increased pulmonary artery pressure following surgical repair of congenital heart defects or hepatovenocclusive disease following bone marrow transplantation. Infants with neonatal pulmonary hypertension homozygous for Thr-1406 have lower L-arginine concentrations than neonates homozygous for Asn-1406.
Sequence similarities	Contains 2 ATP-grasp domains. Contains 1 glutamine amidotransferase type-1 domain.
Domain	The type-1 glutamine amidotransferase domain is defective.
Cellular localization	Mitochondrion.

Images



SDS-PAGE - Recombinant Human CPS1 protein (ab152296)

12.5% SDS-PAGE showing ab152296 stained with Coomassie Blue.

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