# abcam

## Product datasheet

## Recombinant Human CPS1 protein ab152296

### 1 图像

描述

产品名称 重组人CPS1蛋白

表达系统 Wheat germ

Accession <u>P31327</u>

**蛋白长度** Protein fragment

无动物成分 No

性质 Recombinant

**种属** Human

序列 ANNVPATPVAWPSQEGQNPSLSSIRKLIRDGSIDLVINLPNN

NTKFVHDN

YVIRRTAVDSGIPLLTNFQVTKLFAEAVQKSRKVDSKSLFHY

RQYSAGKA A

预测分子量 37 kDa including tags

**氨基酸** 1400 to 1500

技术指标

Our <u>Abpromise guarantee</u> 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.

应**用** Western blot

SDS-PAGE

**ELISA** 

形式 Liquid

补充说明

制备和贮存

稳定性和存储 Shipped on dry ice. Upon delivery aliquot and store at -80℃. Avoid freeze / thaw cycles.

pH: 8.00

Constituents: 0.31% Glutathione, 0.79% Tris HCI

1

#### 常规信息

功能 Involved in the urea cycle of ureotelic animals where the enzyme plays an important role in

removing excess ammonia from the cell.

组织**特异性** Primarily in the liver and small intestine.

疾病相关 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.

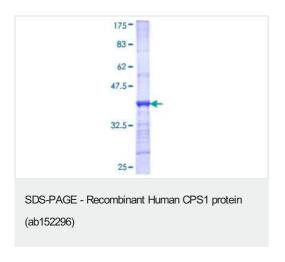
序列相似性 Contains 2 ATP-grasp domains.

Contains 1 glutamine amidotransferase type-1 domain.

结**构域** The type-1 glutamine amidotransferase domain is defective.

细胞定位 Mitochondrion.

#### 图片



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

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