Recombinant human HMBS protein

Catalog Number: ATGP1433

PRODUCT INFORMATION

Expression system
E.coli

Domain
1-361aa

UniProt No.
P08397

NCBI Accession No.
NP_000181

Alternative Names
Porphobilinogen deaminase, PBG-D, PBGD, PORC, uPS

PRODUCT SPECIFICATION

Molecular Weight
41.9 kDa (385aa) confirmed by MALDI-TOF

Concentration
1mg/ml (determined by Bradford assay)

Formulation
Liquid in. 20mM Tris-HCl buffer (pH 8.0) containing 1mM DTT, 10% glycerol, 0.1M NaCl

Purity
> 95% by SDS-PAGE

Tag
His-Tag

Application
SDS-PAGE

Storage Condition
Can be stored at +2C to +8C for 1 week. For long term storage, aliquot and store at -20C to -80C. Avoid repeated freezing and thawing cycles.

BACKGROUND

Description
Porphobilinogen deaminase, also known as HMBS, is a member of the hydroxymethylbilane synthase superfamily. It is a cytoplasmic enzyme found in the heme synthesis pathway. Deficiency of HMBS causes errors in pyrrole metabolism which in turn leads to an inherited autosomal disorder called acute intermittent porphyria (AIP) which is characterized by acute attacks of neurological dysfunctions with hypertension, tachycardia, peripheral neurologic disturbances, abdominal pain and excessive amounts of aminolevulinic acid and porphobilinogen in the urine. Recombinant human HMBS protein, fused to His-tag at N-terminus, was expressed

in E. coli and purified by using conventional chromatography techniques.

Amino acid Sequence

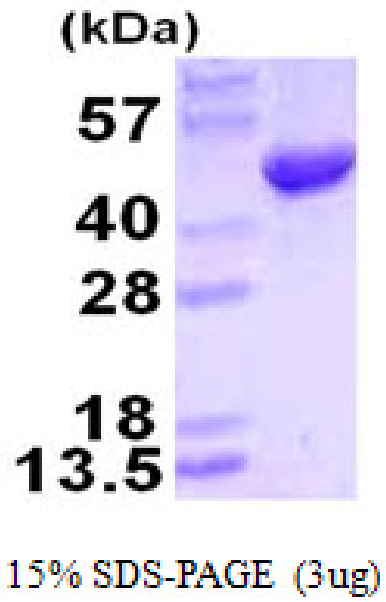
MGSSHHHHHH SSGLVPRGSH MGSHMSGNGN AAATAEENSP KMRVIRVGTR KSQLARIQTD SVVATLKASY PGLQFEIIAM STTGDKILDT ALSKIGEKSL FTKELEHALE KNEVDLVVHS LKDLPTVLPP GFTIGAICKR ENPHDAVVFH PKFVGKTLET LPEKSVVGTS SLRRAAQLQR KFPHLEFRSI RGNLNTRLRK LDEQQEFSAI ILATAGLQRM GWHNRVGQIL HPEECMYAVG QGALGVEVRA KDQDILDLVG VLHDPETLLR CIAERAFLRH LEGGCSVPVA VHTAMKDGQL YLTGGVWSLD GSDSIQETMQ ATIHVPAQHE DGPEDDPQLV GITARNIPRG PQLAAQNLGI SLANLLLSKG AKNILDVARQ LNDAH

General References

Schneider Yin X., et al. (2004) J Inherit Metab Dis. 625-631:471-474.
Sheppard L., et al. (1995) Paediatr Anaesth. 15:426-428.

DATA

SDS-PAGE



15% SDS-PAGE (3ug)

3ug by SDS-PAGE under reducing condition and visualized by coomassie blue stain.