

# Goat Anti-HMBS Antibody (internal region)

Purified Goat Polyclonal Antibody Catalog # AF4265a

#### Specification

# Goat Anti-HMBS Antibody (internal region) - Product Information

Application Primary Accession Other Accession

Reactivity Predicted Host Clonality Concentration Calculated MW WB, E <u>P08397</u> <u>NP\_000181.2</u>, <u>NP\_001019553.1</u>, <u>NP\_001245137.1</u>, <u>NP\_001245138.1</u> Human Human Goat Polyclonal 0.5 39330

# Goat Anti-HMBS Antibody (internal region) - Additional Information

Gene ID 3145

**Other Names** 

HMBS; hydroxymethylbilane synthase; PBG-D; PBGD; PORC; UPS; porphobilinogen deaminase; porphyria, acute; Chester type; pre-uroporphyrinogen synthase; uroporphyrinogen I synthase; uroporphyrinogen I synthetase

Dilution WB~~1:1000 E~~N/A

Format

Supplied at 0.5 mg/ml in Tris saline, 0.02% sodium azide, pH7.3 with 0.5% bovine serum albumin. Aliquot and store at -20°C. Minimize freezing and thawing.

Immunogen Peptide with sequence C-HLEFRSIRGNLNTR, from the internal region of the protein sequence according to NP\_000181.2; NP\_001019553.1; NP\_001245137.1; NP\_001245138.1.

Storage

Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

Precautions

Goat Anti-HMBS Antibody (internal region) is for research use only and not for use in diagnostic or therapeutic procedures.

#### Goat Anti-HMBS Antibody (internal region) - Protein Information



### Name HMBS

Synonyms PBGD, UPS

### Function

As part of the heme biosynthetic pathway, catalyzes the sequential polymerization of four molecules of porphobilinogen to form hydroxymethylbilane, also known as preuroporphyrinogen (PubMed:<a href="http://www.uniprot.org/citations/18004775" target="\_blank">18004775</a>, PubMed:<a href="http://www.uniprot.org/citations/18036296" target="\_blank">18936296</a>, PubMed:<a href="http://www.uniprot.org/citations/18936296" target="\_blank">19138865</a>, PubMed:<a href="http://www.uniprot.org/citations/19138865" target="\_blank">19138865</a>, PubMed:<a href="http://www.uniprot.org/citations/23815679" target="\_blank">23815679</a>). Catalysis begins with the assembly of the dipyrromethane cofactor by the apoenzyme from two molecules of porphobilinogen or from preuroporphyrinogen. The covalently linked cofactor acts as a primer, around which the tetrapyrrole product is assembled (PubMed:<a href="http://www.uniprot.org/citations/18936296" target="\_blank">18936296</a>). In the last step of catalysis, the product, preuroporphyrinogen, is released, leaving the cofactor bound to the holodeaminase intact (PubMed:<a href="http://www.uniprot.org/citations/18936296" target="\_blank">18936296" target="\_blank">18936296" target="\_blank">18936296" target="\_blank">18936296</a>

Cellular Location Cytoplasm, cytosol {ECO:0000250|UniProtKB:P22907}

**Tissue Location** [Isoform 1]: Is ubiquitously expressed.

# Goat Anti-HMBS Antibody (internal region) - Protocols

Provided below are standard protocols that you may find useful for product applications.

- <u>Western Blot</u>
- <u>Blocking Peptides</u>
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- <u>Cell Culture</u>

Goat Anti-HMBS Antibody (internal region) - Images



AF4265a (0.1  $\mu$ g/ml) staining of K562 lysate (35  $\mu$ g protein in RIPA buffer). Primary incubation was 1 hour. Detected by chemiluminescence.

# Goat Anti-HMBS Antibody (internal region) - References

Seven novel genetic mutations within the 5'UTR and the housekeeping promoter of HMBS gene responsible for the non-erythroid form of acute intermittent porphyria. Brancaleoni V, Granata F, Colancecco A, Tavazzi D, Cappellini MD, Di Pierro E. Blood cells, molecules & diseases 49 (3-4): 147-51.