bs-2283R **[Primary Antibody]**

EMC1 Rabbit pAb

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DATASHEET

Host: Rabbit **Isotype:** IgG

Clonality: Polyclonal

GeneID: 23065 **SWISS:** Q8N766

Target: EMC1

Immunogen: KLH conjugated synthetic peptide derived from human PPO: 901-993/993. < Cytoplasmic >

Purification: affinity purified by Protein A

Concentration: 1mg/ml

Storage: 0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
Shipped at 4°C. Store at -20°C for one year. Avoid repeated freeze/thaw cycles.

Background: Protoporphyrinogen oxidase, the penultimate enzyme in the heme biosynthetic pathway, catalyzes the 6-electron oxidation of protoporphyrinogen IX to form protoporphyrin IX. The PPOX protein localizes to the inner membrane of mitochondria from various tissues, including heart, brain, placenta, lung, liver, skeletal muscle, kidney and pancreas. Genetic deficiency of PPOX results in variegate porphyria, a low penetrance, autosomal dominant disorder characterized by cutaneous photosensitivity and/or various neurological manifestations. The rare homozygous variant of VP is characterized by severe PPOX deficiency, and results in the onset of photosensitization by porphyrins in early childhood, skeletal abnormalities of the hand, and, less constantly, short stature, mental retardation and convulsions.

Applications: **IHC-P** (1:100-500)
IHC-F (1:100-500)
IF (1:100-500)
ELISA (1:5000-10000)

Reactivity: (predicted: Human, Mouse, Rat, Rabbit, Cow, Chicken, Dog)

Predicted MW.: 107 kDa

Subcellular Location: Cell membrane