bs-15483R **[Primary Antibody]**

HIBADH Rabbit pAb

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DATASHEET

Host: Rabbit **Isotype:** IgG

Clonality: Polyclonal

GeneID: 11112 **SWISS:** P31937

Target: HIBADH

Immunogen: KLH conjugated synthetic peptide derived from human HIBADH: 251-336/336.

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: HIBADH is a 336 amino acid mitochondrial enzyme that catalyzes the NAD+-dependent, reversible oxidization of 3-Hydroxyisobutyrate to methylmalonate semialdehyde, an intermediate of valine catabolism. The enzyme functions as a homodimer between a pH of 7.0 and 10.0, with optimal activity between 8.8 and 9.0. It was previously hypothesized that defects in the gene encoding HIBADH may be the cause of 3-Hydroxyisobutyric aciduria, a rare disorder that is characterized by a variety of clinical manifestations such as neurodevelopmental problems and dysmorphic features. However, it was shown that HIBADH activity was equal in patients with 3-Hydroxyisobutyric aciduria as compared with controls.

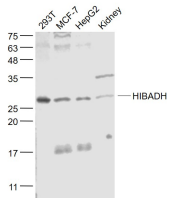
Applications: **WB** (1:500-2000)

Reactivity: Human, Mouse (predicted: Rat, Sheep, Cow, Horse)

Predicted MW.: 32 kDa

Subcellular Location: Cytoplasm

VALIDATION IMAGES



Sample: 193T(Human) Cell Lysate at 30 ug MCF-7(Human) Cell Lysate at 30 ug HepG2(Human) Cell Lysate at 30 ug Kidney (Mouse) Lysate at 40 ug Primary: Anti- HIBADH (bs-15483R) at 1/1000 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 32 kD Observed band size: 30 kD