

bs-15483R**[Primary Antibody]****HIBADH Rabbit pAb****Bioss**
ANTIBODIES

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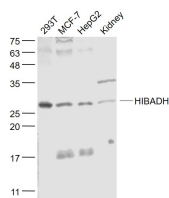
sales@bioss.com.cn

techsupport@bioss.com.cn

400-901-9800

— DATASHEET —

Host: Rabbit	Isotype: IgG	Applications: WB (1:500-2000)
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.		Reactivity: Human, Mouse (predicted: Rat, Sheep, Cow, Horse)
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.		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