

## Product datasheet for **TA344275**

### **BCKDHB Rabbit Polyclonal Antibody**

#### **Product data:**

<b>Product Type:</b>	Primary Antibodies
<b>Applications:</b>	WB
<b>Recommended Dilution:</b>	WB
<b>Reactivity:</b>	Human
<b>Host:</b>	Rabbit
<b>Isotype:</b>	IgG
<b>Clonality:</b>	Polyclonal
<b>Immunogen:</b>	The immunogen for Anti-BCKDHB antibody is: synthetic peptide directed towards the N-terminal region of Human BCKDHB. Synthetic peptide located within the following region: GFLHPAATVEDAAQRRQVAHFTFQPDPEPREYGQTQKMNLFQSVTSALDN
<b>Formulation:</b>	Liquid. Purified antibody supplied in 1x PBS buffer with 0.09% (w/v) sodium azide and 2% sucrose. <i>Note that this product is shipped as lyophilized powder to China customers.</i>
<b>Purification:</b>	Affinity Purified
<b>Conjugation:</b>	Unconjugated
<b>Storage:</b>	Store at -20°C as received.
<b>Stability:</b>	Stable for 12 months from date of receipt.
<b>Predicted Protein Size:</b>	23 kDa
<b>Gene Name:</b>	branched chain keto acid dehydrogenase E1, beta polypeptide
<b>Database Link:</b>	<a href="#">NP_000047</a> <a href="#">Entrez Gene 594 Human</a> <a href="#">P21953</a>



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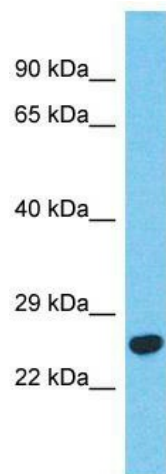
**Background:** Branched-chain keto acid dehydrogenase is a multienzyme complex associated with the inner membrane of mitochondria, and functions in the catabolism of branched-chain amino acids. The complex consists of multiple copies of 3 components: branched-chain alpha-keto acid decarboxylase (E1), lipoamide acyltransferase (E2) and lipoamide dehydrogenase (E3). This gene encodes the E1 beta subunit, and mutations therein have been associated with maple syrup urine disease (MSUD), type 1B, a disease characterized by a maple syrup odor to the urine in addition to mental and physical retardation, and feeding problems. Alternative splicing at this locus results in transcript variants with different 3' non-coding regions, but encoding the same isoform.

**Synonyms:** BCKDE1B; BCKDH E1-beta; E1B

**Note:** Immunogen Sequence Homology: Human: 100%; Dog: 93%; Horse: 93%; Bovine: 93%; Rabbit: 93%; Pig: 86%; Rat: 79%

**Protein Pathways:** Metabolic pathways, Valine, leucine and isoleucine degradation

### Product images:



Host: Rabbit; Target Name: BCKDHB; Sample Tissue: Thyroid tumor lysates; Antibody Dilution: 1.0 ug/ml