

## 内质网 A $\beta$ 相关结合蛋白抗体

产品货号： mlR0021

英文名称： ERAB/HSD17B10

中文名称： 内质网 A $\beta$  相关结合蛋白抗体

别名： HSD17B10; Mitochondrial L3 Hydroxyacyl CoA Dehydrogenase; 17 beta hydroxysteroid dehydrogenase 10; 17 beta hydroxysteroid dehydrogenase type 10; 17b HSD10; 3 hydroxy 2 methylbutyryl CoA dehydrogenase; 3 hydroxyacyl CoA dehydrogenase type 2; 3 hydroxyacyl CoA dehydrogenase type II; AB binding alcohol dehydrogenase; ABAD; Ads9; Amyloid beta binding polypeptide; Amyloid beta peptide binding alcohol dehydrogenase; Amyloid beta peptide binding protein; CAMR; DUPXp11.22; Endoplasmic Reticulum Amyloid Binding Protein; Endoplasmic reticulum associated amyloid beta peptide binding protein; ER associated amyloid beta-binding protein; ERAB; HADH 2; HADH2; HCD 2; HCD2; HSD17B10; Hydroxyacyl CoA Dehydrogenase type II; Hydroxyacyl Coenzyme A dehydrogenase type II; Hydroxysteroid (17 beta) dehydrogenase 10; Mental retardation X linked syndromic 11; MHBD; Mitochondrial L3 Hydroxyacyl CoA Dehydrogenase; Mitochondrial ribonuclease P protein 2; Mitochondrial RNase P protein 2; MRPP2; MRX17; SCHAD; SDR5C1; Short chain dehydrogenase/reductase family 5C member 1; Short chain L 3 hydroxyacyl CoA dehydrogenase type 2; Short chain type dehydrogenase/reductase XH98G2; Type 10 17b HSD; Type 10 17beta hydroxysteroid dehydrogenase; Type II HADH; XH98G2.

研究领域： 免疫学 神经生物学 线粒体

抗体来源： Mouse

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Dog, Pig, Cow, Horse, Sheep, Guinea Pig,

产品应用： ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 IF=1:100-500 （石蜡切片需做抗原修复）

not yet tested in other applications.

optimal dilutions/concentrations should be determined by the end user.

分子量： 21kDa

**细胞定位：** 细胞浆 线粒体

**性 状：** Lyophilized or Liquid

**浓 度：** 1mg/ml

**免 疫 原：** KLH conjugated synthetic peptide derived from human ERAB:51-130/196

**亚 型：** IgG

**纯化方法：** affinity purified by Protein A

**储 存 液：** 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

**保存条件：** Store at -20 ° C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20° C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 ° C.

**PubMed：** PubMed

**产品介绍：** This gene encodes 3-hydroxyacyl-CoA dehydrogenase type II, a member of the short-chain dehydrogenase/reductase superfamily. The gene product is a mitochondrial protein that catalyzes the oxidation of a wide variety of fatty acids, alcohols, and steroids. The protein has been implicated in the development of Alzheimer's disease, and mutations in the gene are the cause of 2-methyl-3-hydroxybutyryl-CoA dehydrogenase deficiency (MHBD). Several alternatively spliced transcript variants have been identified, but the full-length nature of only two transcript variants has been determined. [provided by RefSeq, Jul 2008].

**Function:**

Functions in mitochondrial tRNA maturation. Part of mitochondrial ribonuclease P, an enzyme composed of MRPP1/RG9MTD1, MRPP2/HSD17B10 and MRPP3/KIAA0391, which cleaves tRNA molecules in their 5'-ends. By interacting with intracellular amyloid-beta, it may contribute to the neuronal dysfunction associated with Alzheimer disease (AD).

**Subcellular Location:**

Mitochondrion

**Tissue Specificity:**

Expressed in normal tissues but is overexpressed in neurons affected in AD.

**DISEASE:**

2-methyl-3-hydroxybutyryl-CoA dehydrogenase deficiency (MHBD deficiency) [MIM:300438]: A disorder that leads to neurological abnormalities, including psychomotor retardation and, in virtually all patients, loss of mental and motor skills. Note=The disease is caused by mutations affecting the gene represented in this entry.

Mental retardation, X-linked, syndromic, 10 (MRXS10) [MIM:300220]: A disorder characterized by significantly below average general intellectual functioning associated with impairments in adaptative behavior and manifested during the developmental period. MRXS10 patients manifest mild mental retardation, choreoathetosis and abnormal behavior. Note=The disease is caused by mutations affecting the gene represented in this entry.

Mental retardation, X-linked 17 (MRX17) [MIM:300705]: A disorder characterized by significantly below average general intellectual functioning associated with impairments in adaptative behavior and manifested during the developmental period. Intellectual deficiency is the only primary symptom of non-syndromic X-linked mental retardation, while syndromic mental retardation presents with associated physical, neurological and/or psychiatric manifestations. Note=The gene represented in this entry is involved in disease pathogenesis. A chromosomal microduplication involving HSD17B10 and HUWE1 has been found in patients with mental retardation.

**Similarity:**

Belongs to the short-chain dehydrogenases/reductases (SDR) family.

**SWISS:**



Q99714

**Gene ID:**

3028

**Important Note:**

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.