
Product Information

Product SKU:	CAB8814	Gene ID:	1737	Size:	20uL, 100uL
Clone No:	ARC1314	Host Species:	Rabbit	Reactivity:	Human,Mouse,Rat

Additional Information

Observed MW:	70kDa	Conjugate:	Unconjugated
Calculated MW:	69kDa	Isotype:	IgG

Immunogen Information

Background: This gene encodes component E2 of the multi-enzyme pyruvate dehydrogenase complex (PDC). PDC resides in the inner mitochondrial membrane and catalyzes the conversion of pyruvate to acetyl coenzyme A. The protein product of this gene, dihydrolipoamide acetyltransferase, accepts acetyl groups formed by the oxidative decarboxylation of pyruvate and transfers them to coenzyme A. Dihydrolipoamide acetyltransferase is the antigen for antimitochondrial antibodies. These autoantibodies are present in nearly 95% of patients with the autoimmune liver disease primary biliary cirrhosis (PBC). In PBC, activated T lymphocytes attack and destroy epithelial cells in the bile duct where this protein is abnormally distributed and overexpressed. PBC eventually leads to cirrhosis and liver failure. Mutations in this gene are also a cause of pyruvate dehydrogenase E2 deficiency which causes primary lactic acidosis in infancy and early childhood.

Recommended Dilution: WB,1:500 - 1:2000

Synonyms: E2; PBC; DLTA; PDCE2; PDC-E2; DLAT

Purification Method: Affinity purification

Immunogen: A synthetic peptide corresponding to a sequence within amino acids 548-647 of human DLAT (P10515).

Storage: Store at -20°C. Avoid freeze / thaw cycles. Buffer: PBS with 0.02% sodium azide, 0.05% BSA, 50% glycerol, pH 7.3.