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Anti-Human/Mouse/Rat DLAT Polyclonal Antibody

多克隆抗体

产品货号: PA07479

第三版

**描述:** 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.

**抗原:** Fusion protein of human DLAT

**配方:**

**如何使用:** 加1ml超纯水重溶

**稳定性:** -20 ° C保存条件下，冻干粉,保质期为五年；液体，保质期为两年。

**稀释液:** PBS (pH7.4) ， 1% BSA

**应用:** WB 1 ~ 5  $\mu$ g/ml.

**特异性:**