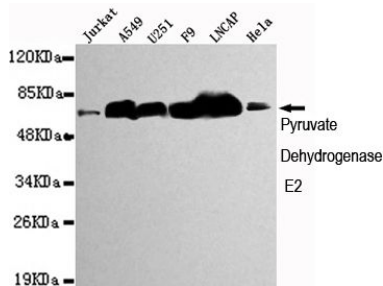




PDC E2

Mouse monoclonal Antibody

#53516

Catalog Number: 53516**Amount:** 100µg/100µl**Swiss-Prot No. :** P10515**Gene name:** dlat**Gene id:** 1737**Clone Number:** 4A4-B6-C10**Form of Antibody:** Purified mouse monoclonal in buffer containing 0.1M Tris-Glycine (pH 7.4, 150 mM NaCl) with 0.2% sodium azide, 50% glycerol**Storage/Stability:** Store at -20°C/1 year**Immunogen:** Purified recombinant human PDC E2 protein fragments expressed in E.coli**Purification:** affinity-chromatography**Specificity/Sensitivity:** This antibody detects endogenous levels of PDC E2 and does not cross-react with related proteins**Reactivity:** Human, Mouse**Applications:** Predicted MW: 69kd WB: 1:1000 ICC:1:300

Western blot detection of PDC E2 in Jurkat, A549, U251, F9, Lncap and HeLa cell lysates using PDC E2 mouse mAb (1:1000 diluted). Predicted band size: 69KDa. Observed band size: 69KDa.

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 anti-mitochondrial 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.