

LAL Monoclonal Antibody

YM0410 Catalog No:

Reactivity: Human

Applications: WB;ELISA

Target: LAL

Fields: >>Steroid biosynthesis;>>Lysosome;>>Cholesterol metabolism

Gene Name: LIPA

Protein Name: Lysosomal acid lipase/cholesteryl ester hydrolase

Q9Z0M5

Human Gene Id: 3988

Human Swiss Prot

P38571

No:

Mouse Swiss Prot

No:

Immunogen: Purified recombinant fragment of LAL expressed in E. Coli.

Specificity: LAL Monoclonal Antibody detects endogenous levels of LAL protein.

Formulation: Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.

Source: Monoclonal, Mouse

Dilution: WB 1:500 - 1:2000. ELISA: 1:10000. Not yet tested in other applications.

Purification: Affinity purification

Concentration: 1 mg/ml

Storage Stability: -15°C to -25°C/1 year(Do not lower than -25°C)

Molecularweight: 45kD

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Cell Pathway: Steroid biosynthesis;Lysosome;

P References: 1. Uta Drebber, Matthias Andersen, Hans U Kasper, et al, World J

Gastroenterol. 2005 Apr 21;11(15):2364-6.

2. Renata Boldrini, Rita Devito, R.Biselli, et al, Pathol Res Pract.

2004;200(3):231-40.

Background: This gene encodes lipase A, the lysosomal acid lipase (also known as

cholesterol ester hydrolase). This enzyme functions in the lysosome to catalyze the hydrolysis of cholesteryl esters and triglycerides. Mutations in this gene can result in Wolman disease and cholesteryl ester storage disease. Alternatively spliced transcript variants have been found for this gene. [provided by RefSeq,

Jan 2014],

Function: catalytic activity: A steryl ester + H(2)O = a sterol + a fatty acid., disease: Defects

in LIPA are the cause of cholesteryl ester storage disease (CESD) [MIM:278000]. CESD is a mild manifestation of LIPA deficiency, leading to the accumulation of cholesteryl esters and triglycerides in most tissues of the body. It is characterized by late-onset., disease: Defects in LIPA are the cause of Wolman disease (WOD) [MIM:278000]. WOD is a severe manifestation of LIPA deficiency, leading to the accumulation of cholesteryl esters and triglycerides in most tissues of the body.

WOD occurs in infancy and is nearly always fatal before the age of 1

year.,function:Crucial for the intracellular hydrolysis of cholesteryl esters and triglycerides that have been internalized via receptor-mediated endocytosis of lipoprotein particles. Important in mediating the effect of LDL (low density

lipoprotein) uptake on s

Subcellular Lysosome .

Location:

Expression: Most abundantly expressed in brain, lung, kidney and mammary gland, a

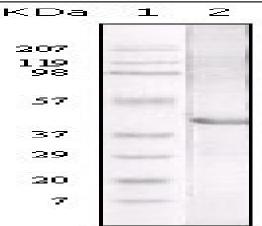
moderate expression seen in placenta and expressed at low levels in the liver and

heart.

Products Images

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Western Blot analysis using LAL Monoclonal Antibody against LAL recombinant protein.