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ACOX1 Rabbit mAb

Catalog # AP75028

Product Information

Application	WB, FC, IP
Primary Accession	Q9R0H0
Reactivity	Rat, Human, Mouse
Host	Rabbit
Clonality	Monoclonal Antibody
Isotype	IgG
Conjugate	Unconjugated
Purification	Affinity Purified
Calculated MW	74649

Additional Information

Gene ID	11430
Other Names	Acox1
Dilution	WB~~1:5000-1:50000 FC~~1 IP~~1:20-1:50
Format	1xPBS(pH 7.4), 150mM NaCl, 50% Glycerol, 0.02% Sodium azide and 0.05% BSA
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.

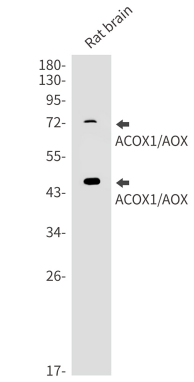
Protein Information

Name	Acox1 {ECO:0000312|MGI:MGI:1330812}
Synonyms	Acox, Paox
Function	Involved in the initial and rate-limiting step of peroxisomal beta-oxidation of straight-chain saturated and unsaturated very-long- chain fatty acids. Catalyzes the desaturation of fatty acyl-CoAs that have a saturated bond between C2 and C3 (2,3-saturated acyl-CoA) to 2- trans-enoyl-CoAs ((2E)-enoyl-CoAs), and donates electrons directly to molecular oxygen (O(2)), thereby producing hydrogen peroxide (H(2)O(2)).
Cellular Location	Peroxisome {ECO:0000250|UniProtKB:P07872}.
Tissue Location	Highest levels of isoform 1 are found in liver and kidney while highest levels of isoform 2 are found in white adipose tissue. Isoform 1 is expressed at higher levels than isoform 2 in liver and kidney while isoform 2 is expressed at higher levels in brain, heart, lung, muscle, white adipose tissue and testis

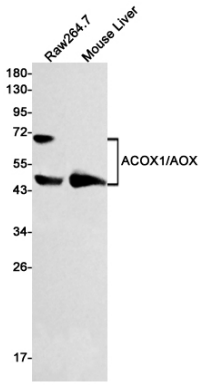
Background

Defects in ACOX1 are the cause of adrenoleukodystrophy pseudoneonatal (Pseudo-NALD); also known as peroxisomal acyl-CoA oxidase deficiency. Pseudo-NALD is a peroxisomal single-enzyme disorder. Clinical features include mental retardation, leukodystrophy, seizures, mild hepatomegaly, hearing deficit. Pseudo-NALD is characterized by increased plasma levels of very-long chain fatty cids, due to decreased or absent peroxisome acyl-CoA oxidase activity. Peroxisomes are intact and functioning.

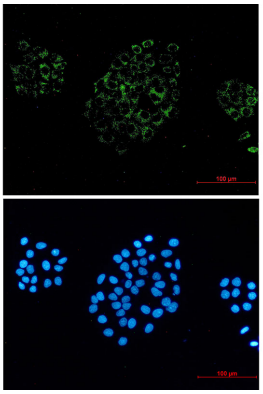
Images



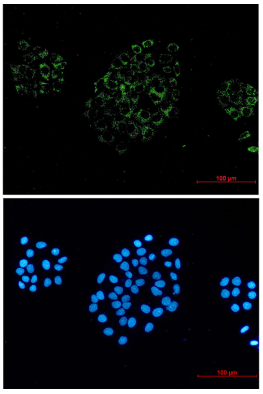
Western blot analysis of ACOX1/AOX in rat brain lysates using ACOX1 antibody.



Western blot analysis of ACOX1/AOX in Raw264.7, mouse Liver lysates using ACOX1/AOX antibody.



Immunocytochemistry analysis of ACOX1 (green) in hela using ACOX1 antibody,and DAPI(blue)



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