

ABHD5 (C-8): sc-393949

BACKGROUND

α/β -hydrolase domains are characterized by a catalytic triad composed of a histidine, an acid and a nucleophile. ABHD5 (abhydrolase domain containing 5), also known as CGI-58, NCIE2 or CDS, is a 349 amino acid protein that contains an α/β -hydrolase domain through which it conveys catalytic function. Localized to the surface of lipid droplets, ABHD5 is widely expressed and interacts with Perilipin on the surface of lipid droplets where it facilitates lipolysis, the breakdown of fat. Defects in the gene encoding ABHD5 are the cause of Chananin-Dorfman syndrome (CDS), an autosomal recessive inborn error of lipid metabolism with impaired long-chain fatty acid oxidation. CDS symptoms include congenital generalized ichthyosis, vacuolated leukocytes, hepatomegaly, myopathy, cataracts, neurosensory hearing loss and developmental delay.

REFERENCES

- Lefèvre, C., et al. 2001. Mutations in CGI-58, the gene encoding a new protein of the esterase/lipase/thioesterase subfamily, in Chananin-Dorfman syndrome. *Am. J. Hum. Genet.* 69: 1002-1012.
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- Schleinitz, N., et al. 2005. Two new mutations of the ABHD5 gene in a new adult case of Chananin-Dorfman syndrome: an uncommon lipid storage disease. *Arch. Dermatol.* 141: 798-800.
- Lass, A., et al. 2006. Adipose triglyceride lipase-mediated lipolysis of cellular fat stores is activated by CGI-58 and defective in Chananin-Dorfman syndrome. *Cell Metab.* 3: 309-319.
- Ben Selma, Z., et al. 2007. A novel S115G mutation of CGI-58 in a Turkish patient with Dorfman-Chananin syndrome. *J. Invest. Dermatol.* 127: 2273-2276.
- Fischer, J., et al. 2007. Neutral lipid storage diseases and ATGL (adipose triglyceride lipase) and CGI-58/ABHD5 (α - β hydrolase domain-containing 5) deficiency: myopathy, ichthyosis, but no obesity. *Med. Sci.* 23: 575-578.

CHROMOSOMAL LOCATION

Genetic locus: ABHD5 (human) mapping to 3p21.33.

SOURCE

ABHD5 (C-8) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 10-29 near the N-terminus of ABHD5 of human origin.

PRODUCT

Each vial contains 200 μ g IgG_{2b} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-393949 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

APPLICATIONS

ABHD5 (C-8) is recommended for detection of ABHD5 of human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for ABHD5 siRNA (h): sc-78146, ABHD5 shRNA Plasmid (h): sc-78146-SH and ABHD5 shRNA (h) Lentiviral Particles: sc-78146-V.

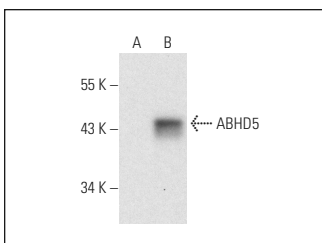
Molecular Weight of ABHD5: 39 kDa.

Positive Controls: ABHD5 (h): 293 Lysate: sc-112234 or A-431 whole cell lysate: sc-2201.

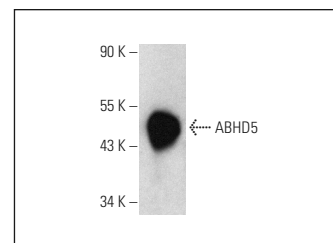
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgG κ BP-FITC: sc-516140 or m-IgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



ABHD5 (C-8): sc-393949. Western blot analysis of ABHD5 expression in non-transfected: sc-110760 (A) and human ABHD5 transfected: sc-112234 (B) 293 whole cell lysates.



ABHD5 (C-8): sc-393949. Western blot analysis of ABHD5 expression in A-431 whole cell lysate.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.