

NSDHL (G-14): sc-161965

BACKGROUND

NSDHL (NAD(P) dependent steroid dehydrogenase-like), also known as H105E3, XAP104 or SDR31E1, is a 373 amino acid single-pass membrane protein expressed in brain, heart, liver, lung, kidney, skin and placenta. Belonging to the 3- β -HSD family, NSDHL localizes to the endoplasmic reticulum and is involved in cholesterol biosynthesis. NSDHL is thought to be involved in the demethylation of sterol precursors in one of the later steps of cholesterol biosynthesis. Defects in the gene encoding NSDHL causes congenital hemidysplasia with ichthyosiform erythroderma and limb defects (CHILD), which is an X-linked dominant disorder of lipid metabolism with defective cholesterol biosynthesis that usually results in male lethality. CHILD is characterized by congenital hemidysplasia with ichthyosiform erythroderma and ipsilateral hypoplasia of limbs and other parts of the skeleton.

REFERENCES

- Hummel, M., et al. 2003. Left-sided CHILD syndrome caused by a nonsense mutation in the NSDHL gene. *Am. J. Med. Genet. A* 122A: 246-251.
- Caldas, H. and Herman, G.E. 2003. NSDHL, an enzyme involved in cholesterol biosynthesis, traffics through the Golgi and accumulates on ER membranes and on the surface of lipid droplets. *Hum. Mol. Genet.* 12: 2981-2991.
- Ohashi, M., et al. 2003. Localization of mammalian NAD(P)H steroid dehydrogenase-like protein on lipid droplets. *J. Biol. Chem.* 278: 36819-36829.
- Murata, K., et al. 2003. A unique point mutation in the NSDHL gene in a Japanese patient with CHILD syndrome. *J. Dermatol. Sci.* 33: 67-69.
- Mehra, S., et al. 2005. A novel somatic mutation of the 3 β -hydroxysteroid dehydrogenase gene in sporadic cutaneous verruciform xanthoma. *Arch. Dermatol.* 141: 1263-1267.
- Bittar, M., et al. 2006. CHILD syndrome in 3 generations: the importance of mild or minimal skin lesions. *Arch. Dermatol.* 142: 348-351.
- Guggenberger, C., et al. 2007. Functional analysis of cholesterol biosynthesis by RNA interference. *J. Steroid Biochem. Mol. Biol.* 104: 105-109.
- Saito, M. and Ishiko, A. 2008. A novel silent mutation in the NSDHL gene causing CHILD syndrome as a result of aberrant splicing. *Br. J. Dermatol.* 159: 1204-1206.

CHROMOSOMAL LOCATION

Genetic locus: *Nsdhl* (mouse) mapping to X A7.3.

SOURCE

NSDHL (G-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus of NSDHL of mouse origin.

STORAGE

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

PRODUCT

Each vial contains 200 μ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-161965 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

NSDHL (G-14) is recommended for detection of NSDHL of mouse origin by Western Blotting (starting dilution 1:200, 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 NSDHL siRNA (m): sc-150073, NSDHL shRNA Plasmid (m): sc-150073-SH and NSDHL shRNA (m) Lentiviral Particles: sc-150073-V.

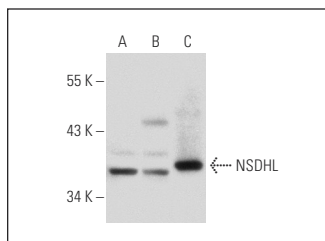
Molecular Weight of NSDHL: 42 kDa.

Positive Controls: F9 cell lysate: sc-2245 or mouse liver extract: sc-2256.

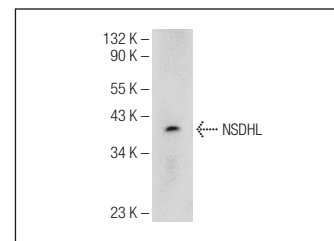
RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 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 donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



NSDHL (G-14): sc-161965. Western blot analysis of NSDHL expression in A-431 (A) and HeLa (B) whole cell lysates and mouse liver tissue extract (C).



NSDHL (G-14): sc-161965. Western blot analysis of NSDHL expression in F9 whole cell lysate.

RESEARCH USE

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

PROTOCOLS

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