

# LLH1 (H-40): sc-292455

## BACKGROUND

Lysyl hydroxylases (LLHs) 1-3 are hydroxyllysines that function as attachment sites for carbohydrates. In collagen, the LLHs form hydroxyllysine residues in -Xaa-Lys-Gly- sequences and are crucial for collagen cross-link stability. They form homodimers that localize to the endoplasmic reticulum. LLH1 is strongly expressed in liver, heart, lung, skeletal muscle and kidney tissue. LLH2 is highly expressed in heart, lung, kidney, eye, ovary and placenta, whereas LLH3 is expressed mainly in heart, lung, liver and testis. LLH1 preferentially hydroxylates triple helical lysine residues at the cross-link positions. Decreased levels of LLH1 expression may lead to Ehlers-Danlos syndrome type VI in skin fibroblasts. This syndrome refers to a heterogeneous group of inherited connective tissue disorders that are characterized by joint hypermobility, skin fragility and hyperextensibility.

## REFERENCES

1. Passoja, K., Myllyharju, J., Pirskanen, A. and Kivirikko, K.I. 1998. Identification of Arginine 700 as the residue that binds the C-5 carboxyl group of 2-oxoglutarate in human lysyl hydroxylase 1. *FEBS Lett.* 434: 145-148.
2. Ruotsalainen, H., Sipilä, L., Kerkelä, E., Pospiech, H. and Myllylä, R. 1999. Characterization of cDNAs for mouse lysyl hydroxylase 1, 2 and 3, their phylogenetic analysis and tissue-specific expression in the mouse. *Matrix Biol.* 18: 325-329.
3. Yeowell, H.N., Allen, J.D., Walker, L.C., Overstreet, M.A., Murad, S. and Thai, S.F. 2000. Deletion of Cysteine 369 in lysyl hydroxylase 1 eliminates enzyme activity and causes Ehlers-Danlos syndrome type VI. *Matrix Biol.* 19: 37-46.
4. Yeowell, H.N. and Walker, L.C. 2000. Mutations in the lysyl hydroxylase 1 gene that result in enzyme deficiency and the clinical phenotype of Ehlers-Danlos syndrome type VI. *Mol. Genet. Metab.* 71: 212-224.
5. Risteli, M., Niemitalo, O., Lankinen, H., Juffer, A.H. and Myllylä, R. 2004. Characterization of collagenous peptides bound to lysyl hydroxylase isoforms. *J. Biol. Chem.* 279: 37535-37543.

## CHROMOSOMAL LOCATION

Genetic locus: PLOD1 (human) mapping to 1p36.22; Plod1 (mouse) mapping to 4 E2.

## SOURCE

LLH1 (H-40) is a rabbit polyclonal antibody raised against amino acids 254-293 mapping within an internal region of LLH1 of human origin.

## PRODUCT

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

## STORAGE

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

## APPLICATIONS

LLH1 (H-40) is recommended for detection of LLH1 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg 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).

LLH1 (H-40) is also recommended for detection of LLH1 in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for LLH1 siRNA (h): sc-60948, LLH1 siRNA (m): sc-60949, LLH1 shRNA Plasmid (h): sc-60948-SH, LLH1 shRNA Plasmid (m): sc-60949-SH, LLH1 shRNA (h) Lentiviral Particles: sc-60948-V and LLH1 shRNA (m) Lentiviral Particles: sc-60949-V.

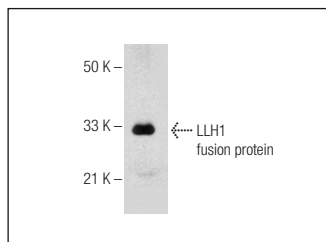
Molecular Weight of LLH1: 85 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200 or mouse ovary extract: sc-2404.

## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

## DATA



LLH1 (H-40): sc-292455. Western blot analysis of human recombinant LLH1 fusion protein.

## RESEARCH USE

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

**MONOS**  
Satisfaction  
Guaranteed

Try **LLH1 (B-5): sc-271640**, our highly recommended monoclonal alternative to LLH1 (H-40).