

LLH1 (C-19): sc-50062

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.
6. Takashi, M., Tsubaki, S., Tsuzuki, T., Duarte, W.R., Yamauchi, M. and Sato, H. 2005. Differential gene expression of collagen-binding small leucine-rich proteoglycans and lysyl hydroxylases, during mineralization by MC3T3-E1 cells cultured on titanium implant material. *Eur. J. Oral Sci.* 113: 225-231.
7. Walker, L.C., Overstreet, M.A., Siddiqui, A., De Paepe, A., Ceylaner, G., Malfait, F., Symoens, S., Atsawasawan, P., Yamauchi, M., Ceylaner, S., Bank, R.A. and Yeowell, H.N. 2005. A novel mutation in the lysyl hydroxylase 1 gene causes decreased activity in an Ehlers-Danlos VIA patient. *J. Invest. Dermatol.* 124: 914-918.
8. Zuurmond, A.M., van der Slot-Verhoeven, A.J., van Dura, E.A., De Groot, J. and Bank, R.A. 2005. Minoxidil exerts different inhibitory effects on gene expression of lysyl hydroxylase 1, 2, and 3: implications for collagen cross-linking and treatment of fibrosis. *Matrix Biol.* 24: 261-270.

SOURCE

LLH1 (C-19) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the C-terminus 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.

Blocking peptide available for competition studies, sc-50062 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

LLH1 (C-19) is recommended for detection of LLH1, LLH3 and, to a lesser extent, LLH2 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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 (C-19) is also recommended for detection of LLH1, LLH3 and, to a lesser extent, LLH2 in additional species, including equine, canine, bovine, porcine and avian.

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 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) 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.

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 or our catalog for detailed protocols and support products.



Try **LLH1 (B-5): sc-271640**, our highly recommended monoclonal alternative to LLH1 (C-19).