

CIRH1A siRNA (m): sc-142347

BACKGROUND

CIRH1A (cirrhosis, autosomal recessive 1A), also designated cirhin, NAIC or TEX292, is a 686 amino acid protein that, when mutated, causes a severe autosomal recessive intrahepatic cholestasis known as North American Indian childhood cirrhosis (NAIC). NAIC is found in aboriginal children from north-western Quebec, and is characterized by transient neonatal jaundice which progresses to biliary cirrhosis, portal hypertension and periportal fibrosis during childhood and adolescence. Localizing to nucleolus, CIRH1A contains eleven WD repeats, exists as three alternatively spliced isoforms and is encoded by a gene that maps to human chromosome 16q22.1. Chromosome 16 encodes over 900 genes in approximately 90 million base pairs, makes up nearly 3% of human cellular DNA and is associated with a variety of genetic disorders.

REFERENCES

1. Weber, A.M., et al. 1981. Severe familial cholestasis in North American Indian children: a clinical model of microfilament dysfunction? *Gastroenterology* 81: 653-662.
2. B  tard, C., et al. 2000. Localization of a recessive gene for North American Indian childhood cirrhosis to chromosome region 16q22-and identification of a shared haplotype. *Am. J. Hum. Genet.* 67: 222-228.
3. Drouin, E., et al. 2000. North American Indian cirrhosis in children: a review of 30 cases. *J. Pediatr. Gastroenterol. Nutr.* 31: 395-404.
4. Chagnon, P., et al. 2002. A missense mutation (R565W) in cirhin (FLJ14728) in North American Indian childhood cirrhosis. *Am. J. Hum. Genet.* 71: 1443-1449.
5. Online Mendelian Inheritance in Man, OMIM™. 2003. Johns Hopkins University, Baltimore, MD. MIM Number: 604901. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
6. Carlton, V.E., et al. 2004. Molecular basis of intrahepatic cholestasis. *Ann. Med.* 36: 606-617.
7. Yu, B., et al. 2005. Nucleolar localization of cirhin, the protein mutated in North American Indian childhood cirrhosis. *Exp. Cell Res.* 311: 218-228.
8. Yu, B., et al. 2009. Cirhin up-regulates a canonical NF  B element through strong interaction with Cirip/HIVEP1. *Exp. Cell Res.* 315: 3086-3098.

CHROMOSOMAL LOCATION

Genetic locus: Cirh1a (mouse) mapping to 8 D3.

PRODUCT

CIRH1A siRNA (m) is a pool of 3 target-specific 19-25 nt siRNAs designed to knock down gene expression. Each vial contains 3.3 nmol of lyophilized siRNA, sufficient for a 10 μ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see CIRH1A shRNA Plasmid (m): sc-142347-SH and CIRH1A shRNA (m) Lentiviral Particles: sc-142347-V as alternate gene silencing products.

For independent verification of CIRH1A (m) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-142347A, sc-142347B and sc-142347C.

STORAGE AND RESUSPENSION

Store lyophilized siRNA duplex at -20   C with desiccant. Stable for at least one year from the date of shipment. Once resuspended, store at -20   C, avoid contact with RNases and repeated freeze thaw cycles.

Resuspend lyophilized siRNA duplex in 330 μ l of the RNase-free water provided. Resuspension of the siRNA duplex in 330 μ l of RNase-free water makes a 10 μ M solution in a 10 μ M Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

APPLICATIONS

CIRH1A siRNA (m) is recommended for the inhibition of CIRH1A expression in mouse cells.

SUPPORT REAGENTS

For optimal siRNA transfection efficiency, Santa Cruz Biotechnology's siRNA Transfection Reagent: sc-29528 (0.3 ml), siRNA Transfection Medium: sc-36868 (20 ml) and siRNA Dilution Buffer: sc-29527 (1.5 ml) are recommended. Control siRNAs or Fluorescein Conjugated Control siRNAs are available as 10 μ M in 66 μ l. Each contain a scrambled sequence that will not lead to the specific degradation of any known cellular mRNA. Fluorescein Conjugated Control siRNAs include: sc-36869, sc-44239, sc-44240 and sc-44241. Control siRNAs include: sc-37007, sc-44230, sc-44231, sc-44232, sc-44233, sc-44234, sc-44235, sc-44236, sc-44237 and sc-44238.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor CIRH1A gene expression knockdown using RT-PCR Primer: CIRH1A (m)-PR: sc-142347-PR (20 μ l). Annealing temperature for the primers should be 55-60   C and the extension temperature should be 68-72   C.

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.