

## X123 siRNA (h): sc-92775

### BACKGROUND

Friedreich's ataxia is an inherited disease that is characterized by a progressive degeneration of the spinal cord and nerve tissue. Caused by a mutated gene region on chromosome 9 that results in mitochondrial malfunction, Friedreich's ataxia can lead to a variety of conditions including speech problems, vision impairment, muscle weakness, diabetes and scoliosis. X123, also known as C9orf61 (chromosome 9 open reading frame 61), is a 289 amino acid protein that is expressed at high levels in skeletal muscle and at lower levels in brain, heart and lung. The gene encoding X123 is located within the Friedreich's ataxia region on chromosome 9q21.11, suggesting a possible role for X123 in the pathogenesis of this disease.

### REFERENCES

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3. Dürr, A., Cossee, M., Agid, Y., Campuzano, V., Mignard, C., Penet, C., Mandel, J.L., Brice, A. and Koenig, M. 1996. Clinical and genetic abnormalities in patients with Friedreich's ataxia. *N. Engl. J. Med.* 335: 1169-1175.
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6. Hebert, M.D. 2007. Targeting the gene in Friedreich ataxia. *Biochimie* 90: 1131-1139.

### CHROMOSOMAL LOCATION

Genetic locus: FAM189A2 (human) mapping to 9q21.11.

### PRODUCT

X123 siRNA (h) 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  $\mu$ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see X123 shRNA Plasmid (h): sc-92775-SH and X123 shRNA (h) Lentiviral Particles: sc-92775-V as alternate gene silencing products.

For independent verification of X123 (h) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-92775A, sc-92775B and sc-92775C.

### PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) for detailed protocols and support products.

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

### APPLICATIONS

X123 siRNA (h) is recommended for the inhibition of X123 expression in human 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  $\mu$ M in 66  $\mu$ 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.

### GENE EXPRESSION MONITORING

X123 (3C7): sc-100741 is recommended as a control antibody for monitoring of X123 gene expression knockdown by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) or immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500).

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG $\kappa$  BP-HRP: sc-516102 or m-IgG $\kappa$  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) Immunofluorescence: use m-IgG $\kappa$  BP-FITC: sc-516140 or m-IgG $\kappa$  BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

### RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor X123 gene expression knockdown using RT-PCR Primer: X123 (h)-PR: sc-92775-PR (20  $\mu$ 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.