

# OPA3 siRNA (m): sc-75995

## BACKGROUND

OPA3 (optic atrophy 3), also known as MGA3, is a 179 amino acid mitochondrial protein that is highly expressed in kidney and skeletal muscle and is thought to play a role in a variety of mitochondrial processes. Defects in the gene encoding OPA3 are the cause of both 3-methylglutaconic aciduria type 3 (MGA3) and optic atrophy type 3 (OPA3). MGA3, also known as optic atrophy plus syndrome or Costeff optic atrophy syndrome, is an autosomal recessive disorder that is characterized by early-onset bilateral optic atrophy, spasticity, extrapyramidal dysfunction and cognitive deficit. OPA3, a neurological disorder, is also characterized by visual impairment, including severe loss of visual acuity, temporal optic disk pallor, color vision deficits and centrocecal scotoma of variable density. Multiple isoforms of OPA3 exist due to alternative splicing events.

## REFERENCES

1. Nystuen, A., et al. 1997. Iraqi-Jewish kindreds with optic atrophy plus (3-methylglutaconic aciduria type 3) demonstrate linkage disequilibrium with the CTG repeat in the 3' untranslated region of the myotonic dystrophy protein kinase gene. *Hum. Mol. Genet.* 6: 563-569.
2. Anikster, Y., et al. 2001. Type III 3-methylglutaconic aciduria (optic atrophy plus syndrome, or Costeff optic atrophy syndrome): identification of the OPA3 gene and its founder mutation in Iraqi Jews. *Am. J. Hum. Genet.* 69: 1218-1224.
3. Kleta, R., et al. 2002. 3-Methylglutaconic aciduria type III in a non-Iraqi-Jewish kindred: clinical and molecular findings. *Mol. Genet. Metab.* 76: 201-206.
4. Reynier, P., et al. 2004. OPA3 gene mutations responsible for autosomal dominant optic atrophy and cataract. *J. Med. Genet.* 41: e110.

## CHROMOSOMAL LOCATION

Genetic locus: Opa3 (mouse) mapping to 7 A3.

## PRODUCT

OPA3 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  $\mu$ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see OPA3 shRNA Plasmid (m): sc-75995-SH and OPA3 shRNA (m) Lentiviral Particles: sc-75995-V as alternate gene silencing products.

For independent verification of OPA3 (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-75995A, sc-75995B and sc-75995C.

## RESEARCH USE

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

## 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

OPA3 siRNA (m) is recommended for the inhibition of OPA3 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  $\mu$ 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.

## RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor OPA3 gene expression knockdown using RT-PCR Primer: OPA3 (m)-PR: sc-75995-PR (20  $\mu$ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.