

Product datasheet for **RC401126**

FH (NM_000143) Human Mutant ORF Clone

Product data:

Product Type:	Mutant ORF Clones
Product Name:	FH (NM_000143) Human Mutant ORF Clone
Mutation Description:	V394L
Affected Codon#:	394
Affected NT#:	1180
Nucleotide Mutation:	FH Mutant (V394L), Myc-DDK-tagged ORF clone of Homo sapiens fumarate hydratase (FH), nuclear gene encoding mitochondrial protein as transfection-ready DNA
Effect:	Muliple uneous nd uerine leiomyom syndrome
Symbol:	FH
Synonyms:	FMRD; HLRCC; HsFH; LRCC; MCL; MCUL1
E. coli Selection:	Kanamycin (25 ug/mL)
Mammalian Cell Selection:	Neomycin
Vector:	pCMV6-Entry (PS100001)
Tag:	Myc-DDK
ACCN:	NM_000143
ORF Size:	1530 bp
Restriction Sites:	Sgfi-MluI



[View online »](#)

ORF Nucleotide Sequence:

>RC401126 representing NM_000143
 Red=Cloning site Blue=ORF Green=Tags(s)

TTTTGTAATACGACTCACTATAGGGCGGCCGGGAATTCGTCGACTGGATCCGGTACCGAGGAGATCTGCC
 GCC**CGGATCGCC**

ATGTACCGAGCACTTCGGCTCCTCGCGCTCGCGTCCCCTCGTCCGGCTCCAGCCGACGCTTAGCTT
 CGGCTCCCGGCTTGGGTGGCGCGCCGTCGCCCTCGTTTTGGCCTCCGAACGCGGCTCGAATGGCAAGCCA
 AAATTCCTTCCGGATAGAATATGATACCTTTGGTGAACAAAGGTGCCAATGATAAGTATTATGGCGCC
 CAGACCGTGAGATCTACGATGAACCTTAAGATTGGAGGTGTGACAGAACGCATGCCAACCCAGTTATTA
 AAGCTTTTGGCATCTTGAAGCGAGCGCCGCTGAAGTAAACCAGGATTATGGTCTTGATCCAAAGATTGC
 TAATGCAATAATGAAGGCAGCAGATGAGGTAGCTGAAGTAAATTAATGATCATTTTCTCTCGTGTA
 TGGCAGACTGGATCAGGAACAGACAAATATGAATGTAATGAAGTCATTAGCAATAGACCAATTGAAA
 TGTTAGGAGGTGAACCTGGCAGCAAGATACCTGTGCATCCCAACGATCATGTTAATAAAGCCAGAGCTC
 AAATGATACTTTTCCACAGCAATGCACATTGCTGCTGCAATAGAAGTTCATGAAGTACTGTTACCAGGA
 CTACAGAAGTTACATGATGCTCTTGATGCAAAATCCAAAGAGTTTGCACAGATCATCAAGATTGGACGTA
 CTCATACTCAGGATGCTGTTCCACTTACTCTTGGGCAGGAATTTAGTGGTTATGTTCAACAAGTAAAATA
 TGCAATGACAAGAATAAAGCTGCCATGCCAAGAATCTATGAGCTCGCAGCTGGAGGCACTGCTGTTGGT
 ACAGGTTTAAATACTAGAATTGGCTTTGCAGAAAAGGTTGCTGCAAAAGTGGCTGCACCTACAGGCTTGC
 CTTTTGCTACTGCTCCGAATAAATTTGAAGCTCTGGCTGCTCATGACGCTCTGGTTGAGCTCAGTGGAGC
 CATGAACACTACTGCCTGCAGTCTGATGAAGATAGCAAATGATATTCGATTTTGGGTTCTGGTCTCGG
 TCAGGTCTGGGAGAATTGATCTTGCTGAAAATGAACAGCAAGCAGTATCATGCCAGGCAAGGTGAACC
 CTACTCAGTGAAGCAATGACCATGGTTGCAGCCCAAGTCATGGGGAACCATGTTGCTCAGTCTCGG
 AGGCAGCAATGGACATTTTGAAGTGAATGTTTTCAAGCCAATGATGATTAATAAATGTGTTACACTCAGCC
 AGGCTGCTGGGGATGCTTCAGTTTCTTTACAGAAAATGCGTGGTGGGAATCCAGGCAATACAGAAA
 GGATCAACAAGCTGATGAATGAGTCTCTAATGTTGGTGCAGCTCTCAATCCTCATATAGGGTATGACAA
 GGCAGCAAAGATTGCTAAGACAGCACACAAAAATGGATCAACCTTAAAGGAAACTGCTATCGAATTGGC
 TATCTCACAGCAGAGCAGTTTGACGAATGGGTAAAACCTAAGGACATGCTGGTCCAAAG

AG**CGGACCG**ACGCGTACGCGCCGCTCGAGCAGAAACTCATCTCAGAAGAGGATCTGGCAGCAAATGATATCC
 TGGATTACAAGGATGACGACGA TAAGGTTTAA

Protein Sequence:

>RC401126 representing NM_000143
 Red=Cloning site Green=Tags(s)

MYRALRLLARSRPLVRAPAAALASAPGLGGAAVPSFWPPNAARMASQNSFRIEYDTFGELKVPNDKYYGA
 QTVRSTMNFKIGGVTERMPVPIKAFGILKRAAAEVNQDYGLDPKIANAIMKAADEVAEGLNDHFPLVV
 WQTGSGTQTNMNVNEVISNRAIEMLGELGSKIPVHPNDHVNKSQSSNDTFPTAMHIAAAIEVHEVLLPG
 LQKLHDALDAKSKEFAQIIKIGRTHQTDAVPLTLGQEFSGYVQVQKYAMTRIKAAMPRIYELAAGGTAVG
 TGLNTRIGFAEKVAAKVAALGLPFVTAPNKFEALAAHDALVELSGAMNTTACSLMKIANDIRFLGSGPR
 SGLGELILPENEPGSSIMPQVNPQCEAMTMVAAQVMGNHVALTVGGSNGHFELNVFKPMMIKNLVHSA
 RLLGDASVSFTENCVVGIQANTERINKLMNESLMLVLTALNPHIGYDKAAKIAKTAHKNSTLKETAIELG
 YLTAEQFDEWVKPKDMLGPK

SGP**TRRRLEQKLI**SEEDLAANDILDYKDDDDKV

Restriction Sites:

Sgfl-MluI

Cloning Scheme:

OTI Disclaimer:

Due to the inherent nature of this plasmid, standard methods to replicate additional amounts of DNA in *E. coli* are highly likely to result in mutations and/or rearrangements. Therefore, OriGene does not guarantee the capability to replicate this plasmid DNA. Additional amounts of DNA can be purchased from OriGene with batch-specific, full-sequence verification at a reduced cost. Please contact our customer care team at custsupport@origene.com or by calling 301.340.3188 option 3 for pricing and delivery.

The molecular sequence of this clone aligns with the gene accession number as a point of reference only. However, individual transcript sequences of the same gene can differ through naturally occurring variations (e.g. polymorphisms), each with its own valid existence. This clone is substantially in agreement with the reference, but a complete review of all prevailing variants is recommended prior to use. [More info](#)

OTI Annotation:

This clone was engineered to express the complete ORF with an expression tag. Expression varies depending on the nature of the gene.

Components:

The ORF clone is ion-exchange column purified and shipped in a 2D barcoded Matrix tube containing 10ug of transfection-ready, dried plasmid DNA (reconstitute with 100 ul of water).

RefSeq:

[NP_000134](#)

RefSeq Size:

1530 bp

RefSeq ORF:

1533 bp

Locus ID:

2271

Cytogenetics:

1q43

Domains:

lyase_1

Protein Families:

Druggable Genome

Protein Pathways:

Citrate cycle (TCA cycle), Metabolic pathways, Pathways in cancer, Renal cell carcinoma

MW: 56.1 kDa

Gene Summary: The protein encoded by this gene is an enzymatic component of the tricarboxylic acid (TCA) cycle, or Krebs cycle, and catalyzes the formation of L-malate from fumarate. It exists in both a cytosolic form and an N-terminal extended form, differing only in the translation start site used. The N-terminal extended form is targeted to the mitochondrion, where the removal of the extension generates the same form as in the cytoplasm. It is similar to some thermostable class II fumarases and functions as a homotetramer. Mutations in this gene can cause fumarase deficiency and lead to progressive encephalopathy. [provided by RefSeq, Jul 2008]