

## Product datasheet for **RC401135**

### **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:	W500X
Affected Codon#:	500
Affected NT#:	1499
Nucleotide Mutation:	FH Mutant (W500X), Myc-DDK-tagged ORF clone of Homo sapiens fumarate hydratase (FH), nuclear gene encoding mitochondrial protein as transfection-ready DNA
Effect:	Fumrse defiieny
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:	1497 bp
Restriction Sites:	Sgfi-Mlul



[View online »](#)

**ORF Nucleotide Sequence:**

>RC401135 representing NM\_000143  
 Red=Cloning site Blue=ORF Green=Tags(s)

TTTTGTAATACGACTCACTATAGGGCGGCCGGGAATTCGTCGACTGGATCCGGTACCGAGGAGATCTGCC  
 GCC**CGGATCGCC**

ATGTACCGAGCACTTCGGCTCCTCGCGCTCGCGTCCCCTCGTCCGGCTCCAGCCGACGCTTAGCTT  
 CGGCTCCCGGCTTGGGTGGCGCGCCGTCGCCCTCGTTTTGGCCTCCGAACGCGGCTCGAATGGCAAGCCA  
 AAATTCCTTCCGGATAGAATATGATACCTTTGGTGAACAAAGGTGCCAATGATAAGTATTATGGCGCC  
 CAGACCGTGAGATCTACGATGAACCTTAAGATTGGAGGTGTGACAGAACGCATGCCAACCCAGTTATTA  
 AAGCTTTTGGCATCTTGAAGCGAGCGCCGCTGAAGTAAACCAGGATTATGGTCTTGATCCAAAGATTGC  
 TAATGCAATAATGAAGGCAGCAGATGAGGTAGCTGAAGTAAATTAATGATCATTTTCTCTCGTGGA  
 TGGCAGACTGGATCAGGAACTCAGACAAATATGAATGTAATGAAGTCATTAGCAATAGAGCAATTGAAA  
 TGTTAGGAGGTGAACCTGGCAGCAAGATACCTGTGCATCCCAACGATCATGTTAATAAAGCCAGAGCTC  
 AAATGATACTTTCCACAGCAATGCACATTGCTGCTGCAATAGAAGTTCATGAAGTACTGTTACCAGGA  
 CTACAGAAGTTACATGATGCTCTTGATGCAAAATCCAAAGAGTTTGCACAGATCATCAAGATTGGACGTA  
 CTCATACTCAGGATGCTGTTCCACTTACTCTTGGGCAGGAATTTAGTGGTTATGTTCAACAAGTAAAATA  
 TGCAATGACAAGAATAAAGCTGCCATGCCAAGAATCTATGAGCTCGCAGCTGGAGGCACTGCTGTTGGT  
 ACAGGTTTAAATACTAGAATTGGCTTTGCAGAAAAGGTTGCTGCAAAAAGTGGCTGCACCTACAGGCTTGC  
 CTTTTGCTACTGCTCCGAATAAATTTGAAGCTCTGGCTGCTCATGACGCTCTGGTTGAGCTCAGTGGAGC  
 CATGAACACTACTGCCTGCAGTCTGATGAAGATAGCAAATGATATTCGATTTTGGGTTCTGGTCTCGG  
 TCAGGTCTGGGAGAATTGATCTTGCCTGAAAATGAACCAAGCAAGTATCATGCCAGGCAAGGTGAACC  
 CTACTCAGTGTGAAGCAATGACCATGGTTGCAGCCCAAGTCATGGGGAACCATGTTGCTGCTACTGTCGG  
 AGGCAGCAATGGACATTTTGAAGTGAATGTTTTCAAGCCAATGATGATTAATAAATGTGTTACACTCAGCC  
 AGGCTGCTGGGGATGCTTCAGTTTCTTTACAGAAAATGCGTGGTGGGAATCCAGGCCAATACAGAAA  
 GGATCAACAAGCTGATGAATGAGTCTCTAATGTTGGTGCAGCTCTCAATCCTCATATAGGGTATGACAA  
 GGCAGCAAAGATTGCTAAGACAGCACACAAAAATGGATCAACCTTAAAGGAACTGCTATCGAATTGGC  
 TATCTCACAGCAGAGCAGTTTGACGAA

AG**CGGACCG**ACGCGTACGCGGCCGCTCGAGCAGAACTCATCTCAGAAGAGGATCTGGCAGCAAATGATATCC  
 TGGATTACAAGGATGACGACGA TAAGGTTTAA

**Protein Sequence:**

>RC401135 representing NM\_000143  
 Red=Cloning site Green=Tags(s)

MYRALRLLARSRPLVRAPAAALASAPGLGGAAVPSFWPPNAARMASQNSFRIEYDTFGELKVPNDKYYGA  
 QTVRSTMNFKIGGVTERMPVPIKAFGILKRAAAEVNQDYGLDPKIANAIMKAADEVAEGKLNDFPLVV  
 WQTGSGTQTNMNVNEVISNRAIEMLGELGSKIPVHPNDHVNSQSSNDTFPTAMHIAAAIEVHEVLLPG  
 LQKLHDALDAKSKEFAQIIKIGRTHQTDAVPLTLGQEFSGYVQVQKYAMTRIKAAMPRIYELAAGGTAVG  
 TGLNTRIGFAEKVAAKVAALTGLPFVTPAPNKFEALAAHDALVELSGAMNTTACSLMKIANDIRFLGSGPR  
 SGLGELILPENEPGSSIMPVKVNPQCEAMTMVAAQVMGNHVAVTVGGSNGHFELNVFKPMMIKNLVHSA  
 RLLGDASVSFTENCVVGIQANTERINKLMNESLMLVLTALNPHIGYDKAAKIAKTAHKNSTLKETAIELG  
 YLTAEQFDE

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](mailto: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:**

1497 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:** 54.9 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]