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DUX4 Rabbit pAb

Catalog # AP98016

Product Information

Application	WB, IHC-P, IHC-F, IF, E
Primary Accession	Q9UBX2
Other Accession	Q9UBX2
Predicted	Human
Host	Rabbit
Clonality	Polyclonal
Calculated MW	44940
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human DUX4
Epitope Specificity	53-120/424
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the paired homeobox family. Contains 2 homeobox DNA-binding domains.
SUBUNIT	May exist as a monomer or a dimer.
DISEASE	Defects in DUX4 may be the cause of facioscapulohumeral muscular dystrophy (FSHD) [MIM:158900]. FSHD is characterized by weakness of the muscles of the face, upper-arm and shoulder girdle. Severity is highly variable. Weakness is slowly progressive and about 20% of affected individuals eventually require a wheelchair. Approximately 70-90% of individuals have inherited the disease-causing deletion from a parent, and approximately 10-30% of affected individuals have FSHD as the result of a de novo deletion. Offsprings of an affected individual have a 50% chance of inheriting the deletion.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	DUX4 is a homeodomain protein with a similar protein sequence to Pax3 and Pax7. Defects in DUX4 may be the cause of facioscapulohumeral muscular dystrophy (FSHD). FSHD is characterized by weakness of the muscles of the face, upper-arm and shoulder girdle. Severity is highly variable. Weakness is slowly progressive and about 20% of affected individuals eventually require a wheelchair. Approximately 70-90% of individuals have inherited the disease-causing deletion from a parent, and approximately 10-30% of affected individuals have FSHD as the result of a de novo deletion. Offsprings of an affected individual have a 50% chance of inheriting the deletion.

Additional Information

Gene ID	100288687
Other Names	DUX4L; AW822073; Dux4; Duxbl; EG664783; DUX4_HUMAN; Double homeobox protein 10; DUX10; double homeobox 4

Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	DUX4 (HGNC:50800)
Synonyms	DUX10
Function	[Isoform 1]: Transcription factor that is selectively and transiently expressed in cleavage-stage embryos (PubMed:28459457). Binds to double-stranded DNA elements with the consensus sequence 5'- TAATCTAATCA-3' (PubMed:28459454, PubMed:28459457, PubMed:29572508, PubMed:30315230, PubMed:30540931). Binds to chromatin containing histone H3 acetylated at 'Lys-27' (H3K27ac) and promotes deacetylation of H3K27ac. In parallel, binds to chromatin that lacks histone H3 acetylation at 'Lys-27' (H3K27ac) and recruits EP300 and CREBBP to promote acetylation of histone H3 at 'Lys-27' at new sites (PubMed:26951377). Involved in transcriptional regulation of numerous genes, primarily as transcriptional activator, but also mediates repression of a set of target genes (PubMed:17984056, PubMed:26951377, PubMed:27378237, PubMed:28459454, PubMed:28459457, PubMed:29572508, PubMed:29618456, PubMed:30540931). Promotes expression of ZSCAN4 and KDM4E, two proteins with essential roles during early embryogenesis (PubMed:26951377, PubMed:27378237, PubMed:28459457, PubMed:29618456). Promotes nuclear translocation of CTNNB1/beta-catenin and its subsequent activation of target genes (PubMed:36158201). Heterologous expression in cultured embryonic stem cells mediates transcription of HERVL retrotransposons and transcripts derived from ACRO1 and HSATII satellite repeats (PubMed:28459457). May activate expression of PITX1 (PubMed:17984056). May regulate microRNA (miRNA) expression (PubMed:24145033). Inappropriate expression can inhibit myogenesis and promote apoptosis (PubMed:26951377, PubMed:28935672, PubMed:29618456).
Cellular Location	[Isoform 1]: Nucleus {ECO:0000255|PROSITE- ProRule:PRU00108, ECO:0000269|PubMed:15709750, ECO:0000269|PubMed:17984056, ECO:0000269|PubMed:21060811, ECO:0000269|PubMed:26816005, ECO:0000269|PubMed:26951377, ECO:0000269|PubMed:27378237, ECO:0000269|PubMed:28459457, ECO:0000269|PubMed:29618456}. Cytoplasm Note=Actively transported through the nuclear pore complex (NPC)
Tissue Location	Isoform 1: Does not seem to be expressed in normal muscle, but is detected in muscle of individuals with FSHD, and also in testis (at protein level) (PubMed:17984056, PubMed:21060811). Isoform 1: Does not seem to be expressed in normal muscle, but in muscle of individuals with FSHD, where it may be toxic to cells (PubMed:17984056, PubMed:21060811). Isoform 2: Detected in skeletal muscle, fibroblasts and testis from healthy individuals (PubMed:21060811)

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.