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Rabbit Anti-DUX4 antibody

SL12369R

Product Name:	DUX4
Chinese Name:	双同源框蛋白4抗体
Alias:	Double homeobox protein 10; Double homeobox protein 4; Double homeobox protein 4/10; DUX10; DUX4_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	45kDa
Cellular localization:	The nucleus
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human DUX4:53-120/424
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	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.

Function:
May be involved in transcriptional regulation.

Subunit:
May exist as a monomer or a dimer.

Subcellular Location:
Nucleus. Note=Actively transported through the nuclear pore complex (NPC).

Tissue Specificity:
Does not seem to be expressed in normal muscle, but in muscle of individuals with FSHD, where it may be toxic to cells.

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.

Similarity:
Belongs to the paired homeobox family.
Contains 2 homeobox DNA-binding domains.

SWISS:
Q9UBX2

Gene ID:
100288687

Database links:

Entrez Gene: 100288687Human

Entrez Gene: 664783Mouse

Omim: 606009Human

SwissProt: Q9UBX2Human

	Unigene: 553518Human **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.