

Rabbit Anti-DUX4 Polyclonal: RC0330

Intended Use: For Research Use Only

Description: DUX4 protein encoded by the gene located within a D4Z4 repeat array in the subtelomeric region of chromosome 4q. Each D4Z4 repeat unit has an open reading frame (named DUX4) that encodes two homeoboxes; the repeat-array and ORF is conserved in other mammals. DUX4 may be involved in transcriptional regulation. 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.

Specifications:

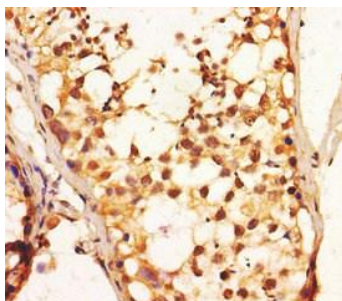
Clone:	Polyclonal
Source:	Rabbit
Isotype:	IgG
Reactivity:	Human
Immunogen:	Recombinant human Double homeobox protein 4 protein
Localization:	Nucleus
Formulation:	Antibody in PBS pH7.4, containing BSA and $\leq 0.09\%$ sodium azide (NaN ₃)
Storage:	Store at 2°- 8°C
Applications:	IHC
Package:	

Description	Catalog No.	Size
DUX4 Polyclonal Concentrated	RC0330	1 ml

IHC Procedure*:

Positive Control Tissue:	Testis, Ewing sarcoma
Concentrated Dilution:	10-100
Pretreatment:	Tris EDTA pH9.0, 15 minutes Pressure Cooker or 30-60 minutes water bath at 95°-99°C
Incubation Time and Temp:	30-60 minutes @ RT
Detection:	Refer to the detection system manual

* Result should be confirmed by an established diagnostic procedure.



FFPE human testis stained with anti-DUX4 using DAB

References:

1. Nuclear DUX4 immunohistochemistry is a highly sensitive and specific marker for the presence of CIC::DUX4 fusion in CIC-rearranged sarcomas: a study of 48 molecularly confirmed cases. Rodrigo T Macedo, et al. Histopathology. Feb;86(3):423-432, 2025.
2. N-terminus DUX4-immunohistochemistry is a reliable methodology for the diagnosis of DUX4-fused B-lymphoblastic leukemia/lymphoma (N-terminus DUX4 IHC for DUX4-fused B-ALL). Bradford J Siegele, et al. Genes Chromosomes Cancer. Aug;61(8):449-458, 2022.
3. Endogenous DUX4 expression in FSHD myotubes is sufficient to cause cell death and disrupts RNA splicing and cell migration pathways. Amanda M Rickard, et al. Hum Mol Genet. Oct 15;24(20):5901-14, 2015.

Doc. 100-RC0330

Rev. A