

#### DESCRIPTION

|                           |                                                                                                                                                                                                                                                                             |
|---------------------------|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <b>Species Reactivity</b> | Human                                                                                                                                                                                                                                                                       |
| <b>Specificity</b>        | Detects human DUX4/DUX4c in direct ELISAs.                                                                                                                                                                                                                                  |
| <b>Source</b>             | Monoclonal Rabbit IgG Clone # 2143C                                                                                                                                                                                                                                         |
| <b>Purification</b>       | Protein A or G purified from cell culture supernatant                                                                                                                                                                                                                       |
| <b>Immunogen</b>          | Human DUX4 synthetic peptide containing sequences within amino acids 200-300 of the human DUX4 protein<br>Accession # Q9UBX2                                                                                                                                                |
| <b>Conjugate</b>          | Alexa Fluor 350<br>Excitation Wavelength: 346 nm<br>Emission Wavelength: 442 nm                                                                                                                                                                                             |
| <b>Formulation</b>        | Supplied 0.2mg/ml in 1X PBS with RDF1 and 0.09% Sodium Azide<br><br>*Contains <0.1% Sodium Azide, which is not hazardous at this concentration according to GHS classifications. Refer to the Safety Data Sheet (SDS) for additional information and handling instructions. |

#### APPLICATIONS

**Please Note:** Optimal dilutions should be determined by each laboratory for each application. [General Protocols](#) are available in the Technical Information section on our website.

**Western Blot** Optimal dilution of this antibody should be experimentally determined.

#### PREPARATION AND STORAGE

**Shipping** The product is shipped with polar packs. Upon receipt, store it immediately at the temperature recommended below.

**Stability & Storage** Protect from light. Do not freeze. 12 months from date of receipt, 2 to 8 °C as supplied

#### BACKGROUND

Human Double homeobox 4 (aka DUX4) is a protein encoded by the DUX4 gene. This gene is located within a D4Z4 repeat array in chromosome 4q35. Each D4Z4 repeat unit has an open reading frame, DUX4, that contains two homeoboxes. Unregulated expression of DUX4 in muscle cells is the cause of facioscapulohumeral muscular dystrophy (FSHD), a common form of muscular dystrophy in adults that is one of the most prevalent genetic diseases of muscle.

#### PRODUCT SPECIFIC NOTICES

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