

**DESCRIPTION**

<b>Species Reactivity</b>	Human
<b>Specificity</b>	Detects human DUX4 in direct ELISAs and Western blots.
<b>Source</b>	Recombinant Monoclonal Rabbit IgG Clone # 2142A
<b>Purification</b>	Protein A or G purified from cell culture supernatant
<b>Immunogen</b>	Human DUX4 synthetic peptide
<b>Formulation</b>	Lyophilized from a 0.2 µm filtered solution in PBS with Trehalose. See Certificate of Analysis for details. *Small pack size (-SP) is supplied either lyophilized or as a 0.2 µm filtered solution in PBS.

**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.

	<b>Recommended Concentration</b>	<b>Sample</b>
<b>Western Blot</b>	0.1 µg/mL	See Below
<b>Immunohistochemistry</b>	3-25 µg/mL	See Below
<b>Simple Western</b>	20 µg/mL	See Below

**DATA**

**Western Blot**

**Detection of Human DUX4 by Western Blot.** Western blot shows lysates of C2C12 mouse myoblast cell line transfected with human DUX4 untreated (-) or treated (+) with Doxycycline. PVDF membrane was probed with 0.1 µg/mL of Rabbit Anti-Human DUX4 Monoclonal Antibody (Catalog # MAB9535) followed by HRP-conjugated Anti-Rabbit IgG Secondary Antibody (Catalog # HAF008). A specific band was detected for DUX4 at approximately 55 kDa (as indicated). This experiment was conducted under reducing conditions and using Immunoblot Buffer Group 1.

**Immunohistochemistry**

**DUX4 in Human Testis.** DUX4 was detected in immersion fixed paraffin-embedded sections of human testis using Rabbit Anti-Human DUX4 Monoclonal Antibody (Catalog # MAB9535) at 3 µg/mL for 1 hour at room temperature followed by incubation with the Anti-Rabbit IgG VisUCyte™ HRP Polymer Antibody (Catalog # VC003). Tissue was stained with hematoxylin (blue). Specific staining was localized to nuclei. View our protocol for IHC Staining with VisUCyte HRP Polymer Detection Reagents.

**Simple Western**

**Detection of Human DUX4 by Simple Western™.** Simple Western lane view shows lysates of HEK293 human embryonic kidney cell line either mock transfected or transfected with human DUX4 and untreated (-) or treated (+) with Doxycycline, loaded at 0.2 mg/mL. A specific band was detected for DUX4 at approximately 62 kDa (as indicated) using 20 µg/mL of Rabbit Anti-Human DUX4 Monoclonal Antibody (Catalog # MAB9535). This experiment was conducted under reducing conditions and using the 12-230 kDa separation system.

**PREPARATION AND STORAGE**

<b>Reconstitution</b>	Reconstitute at 0.5 mg/mL in sterile PBS.
<b>Shipping</b>	The product is shipped at ambient temperature. Upon receipt, store it immediately at the temperature recommended below. *Small pack size (-SP) is shipped with polar packs. Upon receipt, store it immediately at -20 to -70 °C
<b>Stability &amp; Storage</b>	<b>Use a manual defrost freezer and avoid repeated freeze-thaw cycles.</b> <ul style="list-style-type: none"> <li>• 12 months from date of receipt, -20 to -70 °C as supplied.</li> <li>• 1 month, 2 to 8 °C under sterile conditions after reconstitution.</li> <li>• 6 months, -20 to -70 °C under sterile conditions after reconstitution.</li> </ul>

**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.