

DESCRIPTION

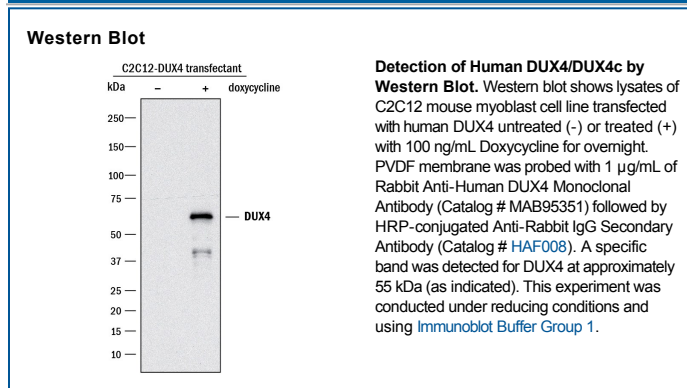
Species Reactivity	Human
Specificity	Detects human DUX4/DUX4c in direct ELISAs.
Source	Recombinant Monoclonal Rabbit IgG Clone # 2143C
Purification	Protein A or G purified from cell culture supernatant
Immunogen	Human DUX4 synthetic peptide containing sequences within amino acids 200-300 of the human DUX4 protein Accession # Q9UBX2
Formulation	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.

	Recommended Concentration	Sample
Western Blot	1 µg/mL	See Below

DATA



PREPARATION AND STORAGE

Reconstitution	Reconstitute at 0.5 mg/mL in sterile PBS.
Shipping	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
Stability & Storage	Use a manual defrost freezer and avoid repeated freeze-thaw cycles. <ul style="list-style-type: none"> ● 12 months from date of receipt, -20 to -70 °C as supplied. ● 1 month, 2 to 8 °C under sterile conditions after reconstitution. ● 6 months, -20 to -70 °C under sterile conditions after reconstitution.

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.