

Anti-Utrophin Antibody, clone MANCHO3 (8A4)

Art. ID	SAF-MABT1558-100UG
Unit	1 x 100 µg
Deliverydetails	No Dangerous Good

Description

Utrophin (UniProt: P46939, also known as Dystrophin-related protein 1, DRP-1) is encoded by the UTRN (also known as DMDL, DRP1) gene (Gene ID: 7402) in human. Utrophin is a dystrophin related protein that displays a sequence homology with dystrophin and shares many of the protein-binding properties. It plays a role in anchoring the cytoskeleton to the plasma membrane. It contains 2 calponin homology domains in its N-terminal region (aa 31-135 and 150-255) and has 22 spectrin homology repeats. The C-terminal sequence in utrophin is highly conserved between dystrophin and utrophin in most species. Utrophin is distributed throughout the sarcolemma in fetal and regenerating muscle, but is shown to be down-regulated in normal adult muscle and is restricted to the myotendinous and neuromuscular junctions. Four different isoforms of utrophin have been described that are produced by alternative promoter usage and they differ in their distribution pattern. Isoform 1 displays high expression in muscle tissue. Similar to dystrophin, utrophin can also reported to form an extensive lateral association with actin filaments and protect actin filaments from depolymerization in vitro. However, utrophin binds laterally along actin filaments through contribution of acidic spectrin-like repeats rather than the cluster of basic repeats used by dystrophin. It has been suggested that utrophin could functionally compensate for the lack of dystrophin and studies have shown that transgenic overexpression of utrophin in dystrophin-deficient mdx mice results in full recovery for all known parameters of the dystrophic phenotype. (Ref.: Sengupta, K., et al. (2020). Sci. Rep. 10, 21492, Porter, J.D., et al.(1998). J. Cell Sci. 111(13), 1801-1811, Rybakova, I.N., et al., (2002). Mol. Biol. Cell. 13(5), 1512-1521).