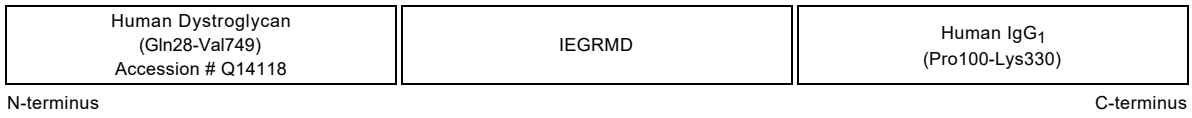
Recombinant Human Dystroglycan Fc Chimera

Catalog Number: 10223-DG

DESCRIPTION

Source

Human embryonic kidney cell, HEK293-derived human Dystroglycan protein



N-terminal Sequence Analysis	Gln28 inferred from AA sequence (alpha subunit) & Ser654 (beta subunit)
Structure / Form	Disulfide-linked homodimer
Predicted Molecular Mass	68 kDa (alpha subunit) & 37 kDa (beta subunit)

SPECIFICATIONS

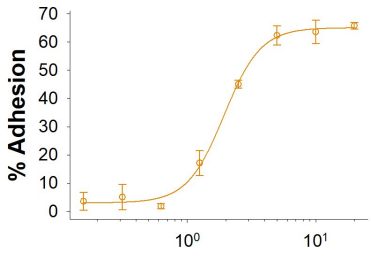
SDS-PAGE	70-100 kDa and 120-135 kDa (alpha subunit) & 45-55 kDa (beta subunit)
Activity	Measured by the ability of the immobilized protein to enhance the adhesion of H4 human neuroglioma cells. The ED_{50} for this effect is 1.5-9 µg/mL.
Endotoxin Level	<0.10 EU per 1 µg of the protein by the LAL method.
Purity	>95%, by SDS-PAGE visualized with Silver Staining and quantitative densitometry by Coomassie® Blue Staining.
Formulation	Lyophilized from a 0.2 µm filtered solution in PBS. See Certificate of Analysis for details.

PREPARATION AND STORAGE

Reconstitution	Reconstitute at 500 µg/mL in PBS.
Shipping	The product is shipped at ambient temperature. Upon receipt, store it immediately at the temperature recommended below.
Stability & Storage	**Use a manual defrost freezer and avoid repeated freeze-thaw cycles.** • 12 months from date of receipt, -20 to -70 °C as supplied. • 2 weeks, 2 to 8 °C under sterile conditions after reconstitution. • 3 months, -20 to -70 °C under sterile conditions after reconstitution.

DATA

Bioactivity



Immobilized Recombinant Human Dystroglycan Fc Chimera (Catalog # 10223-DG) enhances the adhesion of H4 human neuroglioma cells. The ED_{50} for this effect is 1.5-9 µg/mL.

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BACKGROUND

Dystroglycan, also called DAG1 (dystrophin-associated glycoprotein 1) or DG, is a heterodimeric adhesion molecule that links the extracellular matrix (ECM) to the cell cytoskeleton (1-4). Human DAG1 is a type I transmembrane protein that is initially expressed as a large prepro protein. Autocatalysis of the proform produces two fragments (an alpha and beta chain) that remain noncovalently-linked. The alpha chain (aa 28-653) contains a mucin-like region, while the beta chain (aa) consists of an extracellular domain, a transmembrane region, and a cytoplasmic domain (5). Over aa 28-749, human DAG1 shares 93% aa sequence identity with mouse DAG1. DAG1 is widely expressed but differentially O-glycosylated on skeletal muscle and epithelia (which contain a 160 kDa alpha -chain) as compared to cardiac muscle, smooth muscle, fibroblasts, keratinocytes, lymphocytes, and hematopoietic stem cells (which contain a 100-140 kDa alpha -chain) (1-3, 6-9). DAG1 binding of ECM molecules is influenced by its alpha -chain O-glycosylation (2, 6-10). In addition to skeletal muscle and neuromuscular junctions in which DAG1 binds several ECM molecules, DAG1 is important for neuronal migration (through neurexin interactions), keratinocyte attachment to the ECM (through laminin), and adhesion at the immunological synapse and in the hematopoietic stem cell niche (through agrin) (3, 6-11). In muscle, the beta -chain cytoplasmic domain connects with the cytoskeleton via formation of the dystrophin-glycoprotein complex with isoforms of dystrophin, sarcoglycan, syntrophin, and sarcospan (3). This complex is critical for skeletal muscle viability and regeneration (3, 4, 10, 11). MMP9 cleavage of the 44 kDa beta -chain creates a 30 kDa transmembrane form that causes dissociation of the heterodimer and a down-regulation of ECM interactions (6, 12). Dystroglycanopathies, a group of congenital muscular dystrophies affecting the brain, eye and skeletal muscle, are caused by either abnormalities in glycosyltransferases, or their accessory proteins, or rare DAG1 polymorphisms. All result in DAG1 hypoglycosylation, especially of O-mannosyl forms, and affect DAG1 binding to ECM proteins (2, 3, 10, 13, 14).

References:

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