

**DESCRIPTION**

<b>Source</b>	Mouse myeloma cell line, NS0-derived human Fibrillin-1/FBN1 protein		
	Human Fibrillin-1 (Ala25-Thr660) Accession # P35555	IEGRMD	Human IgG <sub>1</sub> (Pro100-Lys330)
	N-terminus		C-terminus
<b>N-terminal Sequence Analysis</b>	Ala25		
<b>Structure / Form</b>	Disulfide-linked homodimer		
<b>Predicted Molecular Mass</b>	95 kDa		

**SPECIFICATIONS**

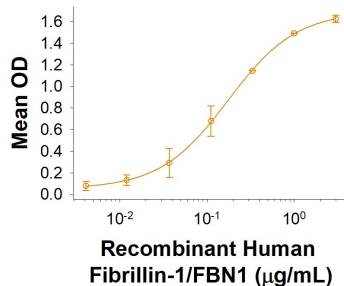
<b>SDS-PAGE</b>	97-108 kDa
<b>Activity</b>	Measured by its binding ability in a functional ELISA. When Recombinant Human MFAP4 (Catalog # 10230-MF) is immobilized at 0.5 µg/mL (100 µL/well), the concentration of Recombinant Human Fibrillin-1/FBN1 Fc Chimera (Catalog # 10224-FI) that produces 50% of the optimal binding response is 0.1-0.6 µg/mL.
<b>Endotoxin Level</b>	<0.10 EU per 1 µg of the protein by the LAL method.
<b>Purity</b>	>95%, by SDS-PAGE under reducing conditions and visualized by silver stain.
<b>Formulation</b>	Lyophilized from a 0.2 µm filtered solution in PBS with Trehalose. See Certificate of Analysis for details.

**PREPARATION AND STORAGE**

<b>Reconstitution</b>	Reconstitute at 500 µg/mL in PBS.
<b>Shipping</b>	The product is shipped at ambient temperature. Upon receipt, store it immediately at the temperature recommended below.
<b>Stability &amp; Storage</b>	<p><b>Use a manual defrost freezer and avoid repeated freeze-thaw cycles.</b></p> <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>• 3 months, -20 to -70 °C under sterile conditions after reconstitution.</li> </ul>

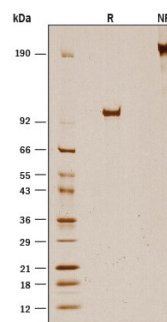
**DATA**

**Binding Activity**



When Recombinant Human MFAP4 (Catalog # 10230-MF) is coated at 0.5 µg/mL, 100 µL/well, Recombinant Human Fibrillin-1/FBN1 Fc Chimera (Catalog # 10224-FI) binds with an ED<sub>50</sub> of 0.1-0.6 µg/mL.

**SDS-PAGE**



1 µg/lane of Recombinant Human Fibrillin-1/FBN1 Fc Chimera (Catalog # 10224-FI) was resolved with SDS-PAGE under reducing (R) and non-reducing (NR) conditions and visualized by silver staining, showing bands at 97-108 kDa and 190-220 kDa, respectively.

**BACKGROUND**

Fibrillins are glycoproteins forming the backbone of microfibrils in elastic and non-elastic tissues. They interact with other components of the extracellular matrix (ECM) and play essential roles in tissue development, homeostasis and repair. Fibrillin-1 is a calcium-binding protein that assembles to form the structural component of the 10-12 nm microfibrils of the ECM. The human Fibrillin-1 has multiple domains, primarily consisting of epidermal growth factor (EGF)-like and other modules (1, 2). The calcium-binding modules in some of the EGF domains provide structural stability and the characteristic rod-like shape of the protein (3-8). Mature human Fibrillin-1 shares 97% amino acid (aa) sequence identity with mature mouse Fibrillin-1. Human Fibrillin-1 is synthesized as an approximately 350-kDa precursor molecule, which is then proteolytically processed by furin into its biologically active form (9-10). Fibrillin microfibrils are further engaged in a number of cell matrix interactions such as with integrins, bone morphogenetic proteins (BMPs) and the large latent complex of transforming growth factor-beta (11). Fibrillin-1 mutations are associated with a range of heritable connective disorders, including Marfan syndrome and acromelic dysplasias (11-12).

**References:**

1. Robertson, I. *et al.* (2011) *Biochem. J.* **433**:263.
2. Corson, G.M. *et al.* (1993) *Genomics* **17**:476.
3. Maslen, C.L. *et al.* (1991) *Nature* **352**:334.
4. Hanford, P.A. *et al.* (1991) *Nature* **353**:395.
5. Werner, J.M. *et al.* (2000) *J. Mol. Biol.* **296**:1065.
6. Downing, A.K. *et al.* (1996) *Cell* **85**:597.
7. Smallridge, R.S. *et al.* (2003) *J. Biol. Chem.* **278**:12199.
8. Reinhardt, D.P. *et al.* (1997) *J. Biol. Chem.* **272**:7368.
9. Raghunath, M. *et al.* (1999) *J. Cell. Sci.* **112**:1093.
10. Wallis, D.D. *et al.* (2003) *J. Cell. Sci.* **90**:641.
11. Jensen, S.A. *et al.* (2016) *Biochem. J.* **473**:827.
12. Sherratt, M.J. *et al.* (2001) *Micron.* **32**:185.