

# Product Datasheet

## FLNC Antibody - BSA Free NBP1-89300

Unit Size: 0.1 ml

Store at 4C short term. Aliquot and store at -20C long term. Avoid freeze-thaw cycles.

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Updated 2/21/2025 v.20.1

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**NBP1-89300**

FLNC Antibody - BSA Free

**Product Information**

<b>Unit Size</b>	0.1 ml
<b>Concentration</b>	Concentrations vary lot to lot. See vial label for concentration. If unlisted please contact technical services.
<b>Storage</b>	Store at 4C short term. Aliquot and store at -20C long term. Avoid freeze-thaw cycles.
<b>Clonality</b>	Polyclonal
<b>Preservative</b>	0.02% Sodium Azide
<b>Isotype</b>	IgG
<b>Purity</b>	Immunogen affinity purified
<b>Buffer</b>	PBS (pH 7.2) and 40% Glycerol

**Product Description**

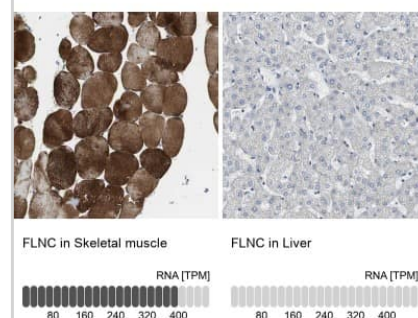
<b>Host</b>	Rabbit
<b>Gene ID</b>	2318
<b>Gene Symbol</b>	FLNC
<b>Species</b>	Human
<b>Immunogen</b>	This antibody was developed against Recombinant Protein corresponding to amino acids: HSLHETSTVLVETVTKSSSSRGSSYSSIPKFSSDASKVVTRGPGLSQAQFVGQK NSFTVDCSKAGTNNMMVGVHGPCKTPCEEVYVKHMGNRVYNVTYTVKEKGDY

**Product Application Details**

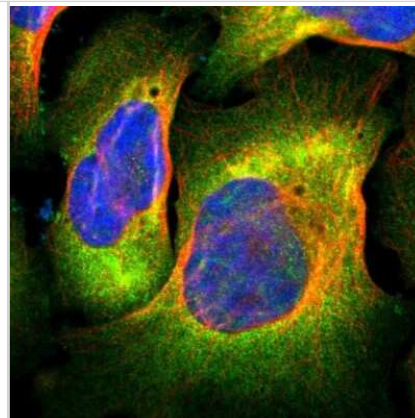
<b>Applications</b>	Western Blot, Immunocytochemistry/ Immunofluorescence, Immunohistochemistry, Immunohistochemistry-Frozen, Immunohistochemistry-Paraffin
<b>Recommended Dilutions</b>	Western Blot Reported in scientific literature (PMID: 33202721)., Immunohistochemistry 1:200 - 1:500, Immunocytochemistry/ Immunofluorescence 0.25 - 2 ug/mL, Immunohistochemistry-Paraffin 1:200 - 1:500, Immunohistochemistry-Frozen Validated from a verified customer review.
<b>Application Notes</b>	For IHC-Paraffin, HIER pH 6 retrieval is recommended. ICC/IF Fixation Permeabilization: Use PFA/Triton X-100.

**Images**

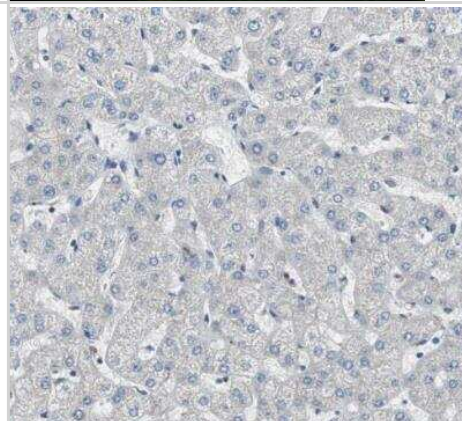
Immunohistochemistry-Paraffin: FLNC Antibody [NBP1-89300] - Analysis in human skeletal muscle and liver tissues. Corresponding FLNC RNA-seq data are presented for the same tissues.



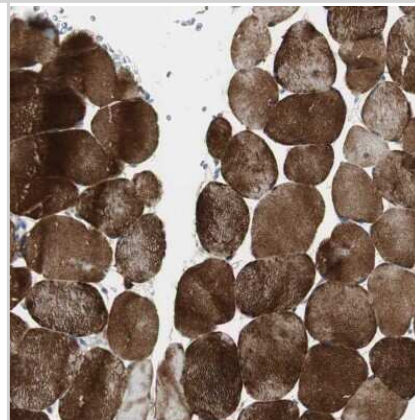
Immunocytochemistry/Immunofluorescence: FLNC Antibody [NBP1-89300] - Staining of human cell line U-2 OS shows localization to plasma membrane and cytosol. Antibody staining shown in green.



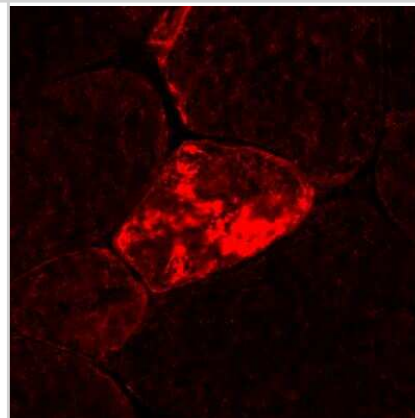
Immunohistochemistry-Paraffin: FLNC Antibody [NBP1-89300] - Staining of human liver shows no positivity in hepatocytes as expected.



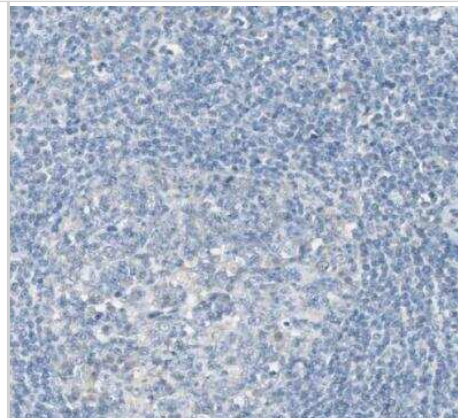
Immunohistochemistry-Paraffin: FLNC Antibody [NBP1-89300] - Staining of human skeletal muscle shows high expression.




Immunohistochemistry-Frozen: FLNC Antibody [NBP1-89300] - Mouse muscle vasus transverse cryosection stained with Filamin C antibody (NBP1-89300). IHC-Fr image submitted by a verified customer review.



Immunohistochemistry-Paraffin: FLNC Antibody [NBP1-89300] - Staining of human tonsil shows no positivity in non-germinal center cells as expected.



Immunohistochemistry-Paraffin: FLNC Antibody [NBP1-89300] - Staining of human prostate shows moderate cytoplasmic positivity in smooth muscle cells. 

## Publications

Marion Onnée, Audrey Bénézit, Sultan Bastu, Aleksandra Nadaj-Pakleza, Béatrice Lannes, Flavie Ader, Corinne Thèze, Pascal Cintas, Claude Cances, Robert-Yves Carlier, Corinne Metay, Mireille Cossée, Edoardo Malfatti, Paul Rösch The FLNC Ala1186Val Variant Linked to Cytoplasmic Body Myopathy and Cardiomyopathy Causes Protein Instability *Biomedicines* 2024-01-30 [PMID: 38397924]

Wu T, Xu Y, Zhang L et al. Filamin C is Essential for mammalian myocardial integrity *PLOS Genetics* 2023-01-27 [PMID: 36706168] (Western Blot)

Yerra VG, Batchu SN, Kaur H et al. Pressure overload induces ISG15 to facilitate adverse ventricular remodeling and promote heart failure *The Journal of clinical investigation* 2023-05-01 [PMID: 37115698] (WB, IHC-P, Mouse)

Knyazeva A, Khudiakov A, Vaz R et al. FLNC Expression Level Influences the Activity of TEAD-YAP/TAZ Signaling *Nat Commun* 2020-11-13 [PMID: 33202721] (WB, Human, Mouse)

Robertson R, Conte TC, Dicaire MJ et al. BAG3P215L/KO Mice as a Model of BAG3P209L Myofibrillar Myopathy *Am. J. Pathol.* 2020-01-14 [PMID: 31953038]

Kiselev A, Vaz R, Knyazeva A et al. De novo mutations in FLNC leading to early-onset restrictive cardiomyopathy and congenital myopathy *Hum. Mutat.* 2018-06-01 [PMID: 29858533] (Human)

Wu T, Mu Y, Bogomolovas J et al. HSPB7 is indispensable for heart development by modulating actin filament assembly. *Proc. Natl. Acad. Sci. U.S.A.* 2017-10-23 [PMID: 29078393] (Mouse)

Reinstein E, Gutierrez-Fernandez A, Tzur S et al. Congenital dilated cardiomyopathy caused by biallelic mutations in Filamin C. *Eur. J. Hum. Genet.* 2016-09-07 [PMID: 27601210] (IHC-P)

Valdes-Mas R, Gutierrez-Fernandez A, Gomez J et al. Mutations in filamin C cause a new form of familial hypertrophic cardiomyopathy *Nat Commun et al.* 2014-10-29 [PMID: 25351925] (IHC-P, Human)

### Details:

FLnC/Filamin C antibody used for IHC-P on heart tissue sections from human control subjects and three hypertrophic cardiomyopathy patient - one caused by a mutation in MYH7, and two without mutations in sarcomeric genes or FLnC, revealed large nuclei and larger fibre diameter, but not filamin C aggregates. IHC revealed the presence of FLnC in the protein aggregate occurring in cardiomyocytes of patients with FLnC mutations (Figure 3c).





### **Novus Biologicals USA**

10730 E. Briarwood Avenue  
Centennial, CO 80112  
USA  
Phone: 303.730.1950  
Toll Free: 1.888.506.6887  
Fax: 303.730.1966  
nb-customerservice@bio-techne.com

### **Bio-Techne Canada**

21 Canmotor Ave  
Toronto, ON M8Z 4E6  
Canada  
Phone: 905.827.6400  
Toll Free: 855.668.8722  
Fax: 905.827.6402  
canada.inquires@bio-techne.com

### **Bio-Techne Ltd**

19 Barton Lane  
Abingdon Science Park  
Abingdon, OX14 3NB, United Kingdom  
Phone: (44) (0) 1235 529449  
Free Phone: 0800 37 34 15  
Fax: (44) (0) 1235 533420  
info.EMEA@bio-techne.com

### **General Contact Information**

www.novusbio.com  
Technical Support: nb-technical@bio-techne.com  
Orders: nb-customerservice@bio-techne.com  
General: novus@novusbio.com

### **Products Related to NBP1-89300**

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NBP1-89300PEP	FLNC Recombinant Protein Antigen
HAF008	Goat anti-Rabbit IgG Secondary Antibody [HRP]
NB7160	Goat anti-Rabbit IgG (H+L) Secondary Antibody [HRP]
NBP2-24891	Rabbit IgG Isotype Control

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### **Limitations**

This product is for research use only and is not approved for use in humans or in clinical diagnosis. Primary Antibodies are guaranteed for 1 year from date of receipt.

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