Nanobody toolbox for your research

PRODUCT SPECIFICATION

Recombinant anti-human gelsolin FAF nanobody 1.

Catalogue number: sdAb-GSN-FAF Nb1

Background

The orphan disease known as familial amyloidosis of the Finnish type (FAF, AGel) is caused by a mutation in the gene encoding gelsolin. At least 4 mutations have been identified thus far, with D187N and D187Y being the first identified. The pathology (including but not limited to cranial neuropathy, cutis laxa, tongue atrophy) is caused by the secreted variant of gelsolin (plasma gelsolin). Two proteases, furin and the metalloprotease MT1-MMP, sequentially cleave off fragments of gelsolin, yielding 5kDa and 8kDa amyloidogenic peptides that assemble into fibrils.

Applications: WB, PD, ELISA. This product is for R&D use only, not for drug, diagnostic, therapeutic, household, or other uses.

Source and properties

Gelsolin FAF nanobody 1 was raised by immunizing a dromedary with the 8 kDa gelsolin amyloidogenic peptide derived from FAF (D187N) mutant gelsolin. The peptide originates by MT1-MMP cleavage of gelsolin. The nanobody binds to gelsolin with an approximate **affinity of 380 nM** (determined by ITC). On WB, the nanobody interacts with peptide monomers and oligomers. The nanobody interacts with the peptide precursor, known as C68 gelsolin, and protects partially against MT1-MMP degradation of C68. It promotes a therapeutic effect when injected in mice that recapitulate the disease. Labeling of the Nb with ^{99m}Tc allows imaging of amyloid fibrils in vivo.

Availability: Nanobody 11 comes with a COOH-terminal HA or Myc epitope tag. Available in 100 µg, 500 µg, 1000 µg quantities. For bulk amounts, please inquire.

Expression host: VHH single domain antibody purified from *E. coli*.

Cross reactivity: Reactivity of this nanobody with gelsolin from other species has not been tested.

Storage buffer: 20 mM Tris-HCl pH 8.0, 150 mM NaCl, 1mM DTT, 60 % glycerol. Store at -20°C. The sample will not freeze. Maintain sample in cold environment during transport to increase longevity.

Stability: Store at -20°C upon arrival. For long term storage, aliquot and store at -80°C. Avoid repeated freeze/thaw cycles.

Product citations:

1. Verhelle A, Nair N, Everaert I, Van Overbeke W, Supply L, et al. 2017. *Hum Mol Genet* 26: 1353-64
2. Verhelle A, Van Overbeke W, Peleman C, De Smet R, Zwaenepoel O, et al. 2016. *Mol Imaging Biol* 18: 887-97