CXCR4-specific Nanobodies as potential therapeutics for WHIM syndrome
Raymond H. de Wit, Raimond Heukers, Hendrik Brink, Angela Arsova, David Maussang, Pasquale Cutolo, Beatrijs Strubbe, Henry F. Vischer, Françoise Bachelerie and Martine J. Smit

Figure S1. Inhibition of CXCL12 binding in CXCL12-induced signaling in CXCR4 WHIM mutant S338X. A) Basal and CXCL12-induced (5 nM) [³H]-inositol phosphates accumulation of Mock (Gαq5-only transfection), CXCR4-WT, CXCR4-R334X or CXCR-R338X expressing HEK293T cells. B) Binding of ¹²⁵I-labeled CXCL12 (75 pM) to HEK293T membranes containing CXCR4 WHIM mutant S338X in the presence of a concentration range of AMD3100 (closed circles), 10A10 (open circles), 10A10-10A10 (open squares). C) CXCL12-induced (5 nM) accumulation of [³H]-inositol phosphates in HEK293T cells expressing CXCR4 WHIM mutant S338X by a concentration range of AMD3100 (filled circles) or 10A10-10A10 (open squares). CXCR4-mediated PLC activation was obtained by co-expression with Gαq5. D) CXCL12-induced CRE activation in HEK293T cells expressing CXCR4 wild type or the WHIM mutants R334X or S338X, as determined by CRE-luciferase reporter gene assay. E) Inhibition of CXCL12-induced CRE activation by a concentration range of AMD3100 (filled circles) or 10A10-10A10 (open squares). Plotted are mean with SEM of a representative graph, n = 3.