Shwachman-Diamond syndrome neutrophils have altered chemoattractant-induced F-actin polymerization and polarization characteristics

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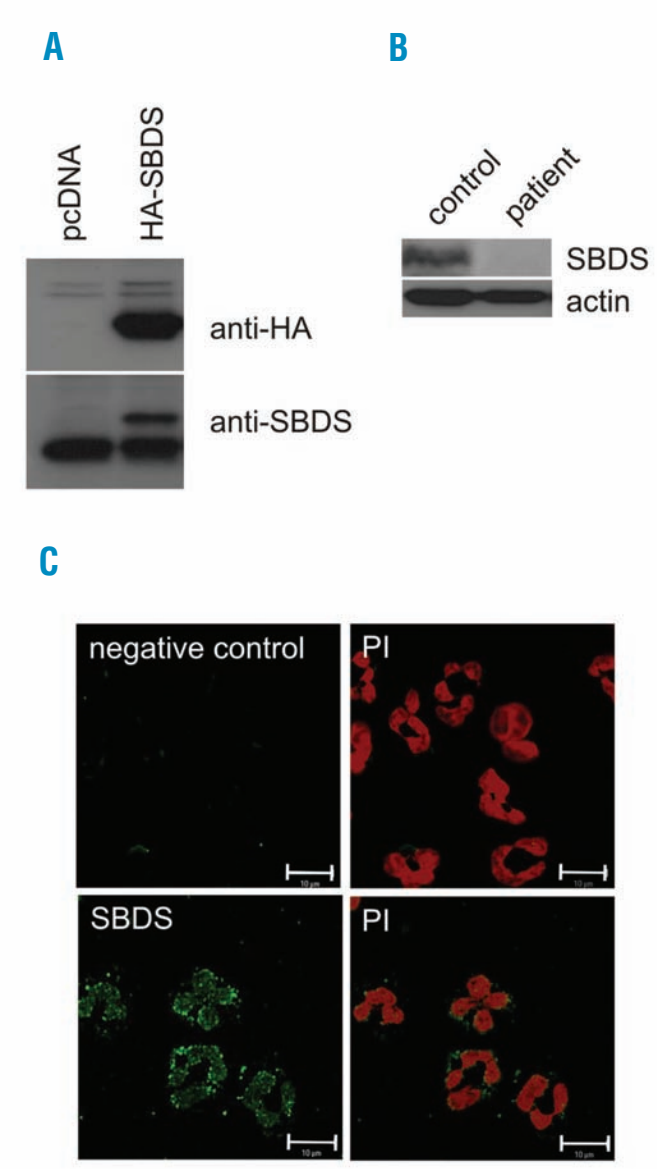

Supplementary Figure S1. SBDS antibody specificity test. (A) Western blot analysis with anti-SBDS and anti-HA antibodies for transiently expressed HA-SBDS in Cos-7 cells reveals that our generated anti-SBDS antibody (lower panel) recognizes an endogenous SBDS protein and a slightly larger HA-tagged SBDS protein. The anti-HA probed Western blot (upper panel) only shows a HA-SBDS protein in the transfected cells at a similar molecular weight as the HA-SBDS protein in the lower panel. (B) Western blot analysis for SBDS protein expression in peripheral blood monocytic cells from healthy controls and SDS patients. In SDS patients little or no SBDS protein is detected. Blot was reprobed for β-actin expression as a loading control. (C) Cytospins of freshly isolated peripheral blood neutrophils were fixed with paraformaldehyde and processed for immunofluorescence staining. The isotype control (negative control; top panel) reveals no aspecific antibody staining. The neutrophils stained for SBDS protein reveal prominent nuclear localization and to a lesser extent cytoplasmic SBDS protein localization.