Supplementary Data

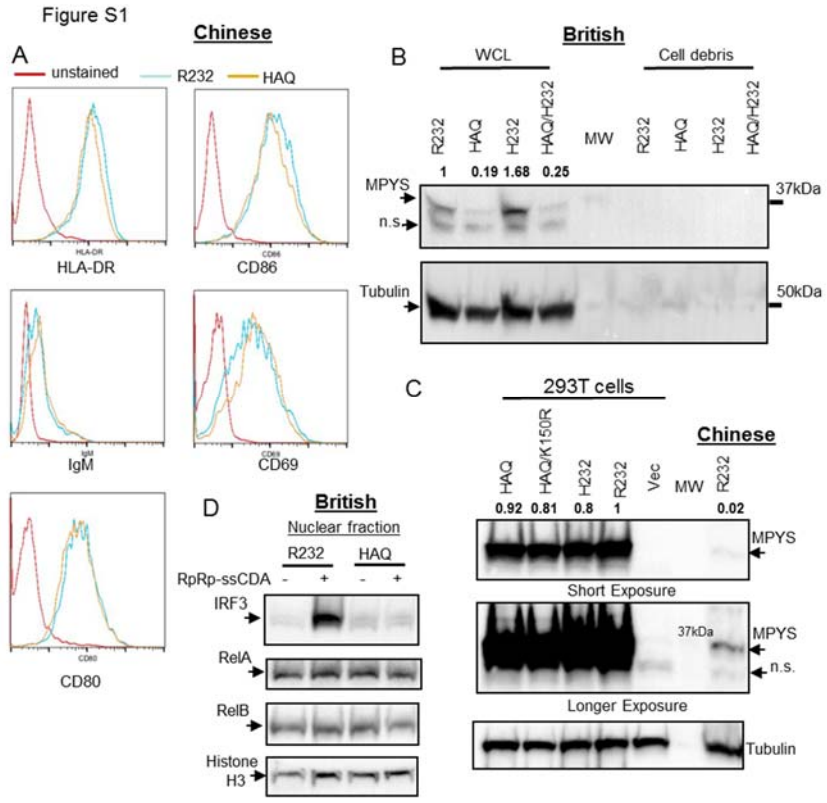

Figure S1. EBV-transformed human B cells express MPYS. A. *R232/R232* and *HAQ/HAQ* human B cells (Chinese) were stained with indicated Abs. Live cells were analyzed by flow cytometry (n = 2). **B**. *R232/R232, HAQ/HAQ, H232/H23, HAQ/H232* human B cells samples (British) were lysed in RIPA buffer. WCL and cell debris were separated by centrifuge (14,000g for 10mins), run on a SDS-PAGE gel, and probed for indicated proteins (n=2). **C.** Parent 293T cells, 293T cells stably transfected with indicated human *TMEM173* variants and *R232/R232* human B cells (Chinese) were lysed in RIPA buffer. WCL was run and probed for indicated proteins (n=3). **D**. *R232/R232, HAQ/HAQ* human B cells (British) were activated with RpRp-ssCDA (5μg/ml) for 5 h as described in Materials and Methods. Nuclear fractions were isolated. Samples were run on a SDS-PAGE gel and probed with the indicated Abs (n=3). n.s. nonspecific. M.W. molecular weight marker.

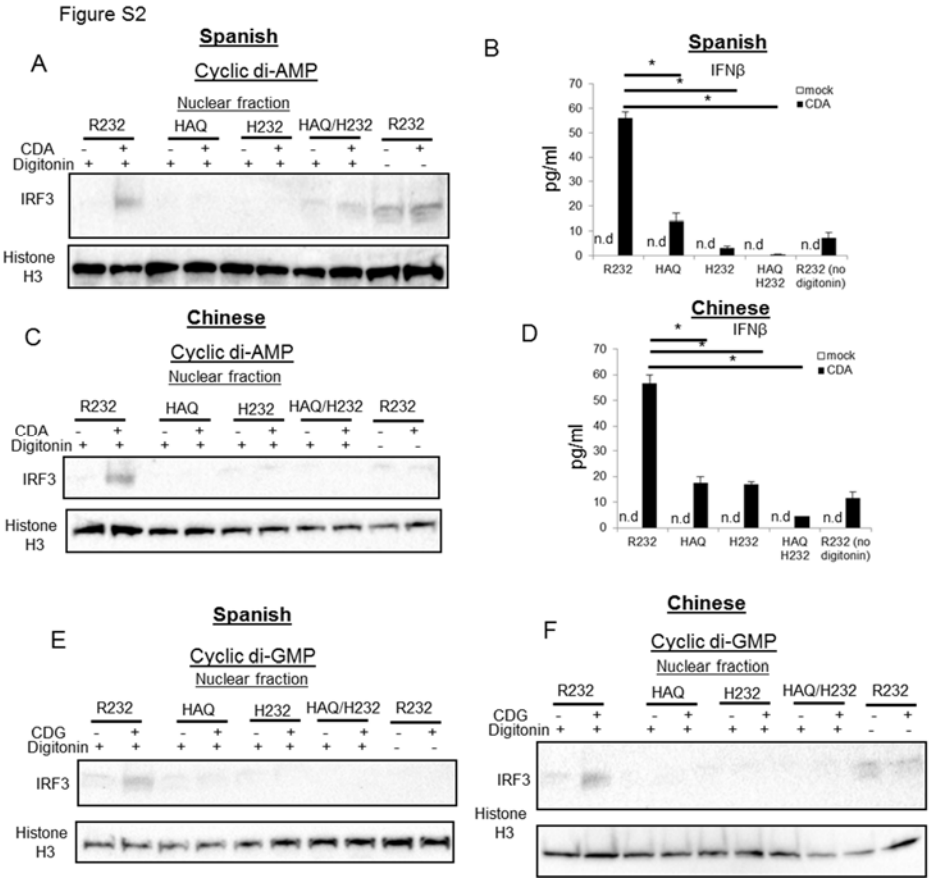

Figure S2. Homozygous *HAQ* B cells do not respond to CDNs. A&C. *R232/R232, HAQ/HAQ, H232/H232 and HAQ/H232* human B cells from indicated ethnic groups were activated with CDA (10 μg/ml) for 5 h as described in Materials and Methods. Nuclear fractions were isolated. Samples were run on a SDS-PAGE gel and probed with the indicated Abs (n=3). **B&D**. Human IFNβ was measured in cell supernatant from **A&C** by ELISA (n=3). **E&F**. *R232/R232, HAQ/HAQ, H232/H232 and HAQ/H232* human B cells from indicated ethnic groups were activated with CDG (10 μg/ml) for 5 h in culture. Nuclear fractions were isolated, run on a SDS-PAGE gel and probed with the indicated Abs (n=3). Graph present means ± SEM from three independent experiments. The significance is represented by an asterisk, where $p<0.05$. n.d. not detected.

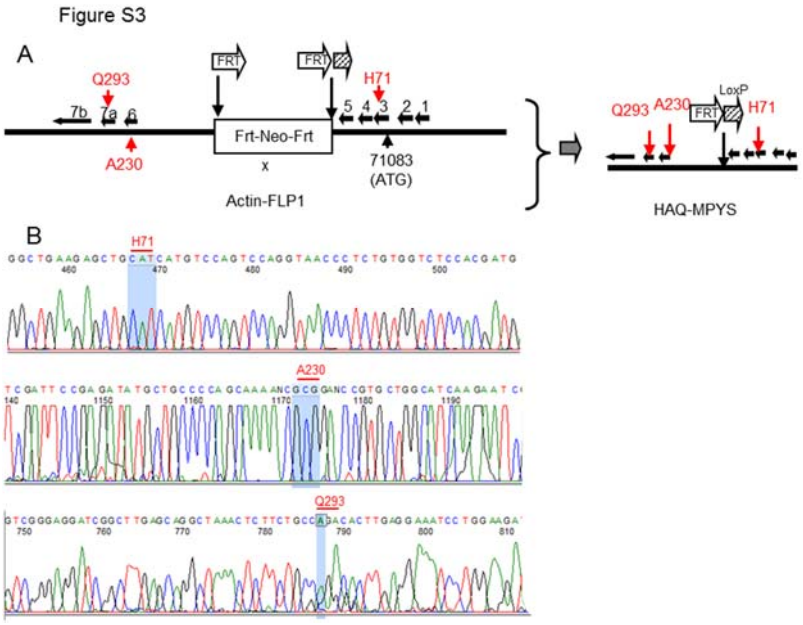

Figure S3. Making a HAQ knock-in mouse. **A.** Diagram illustrating the generation of an *HAQ* knock-in mouse. **B**. PCR sequencing of the H71-A230-Q293 mutations in the *HAQ* knock-in mouse.