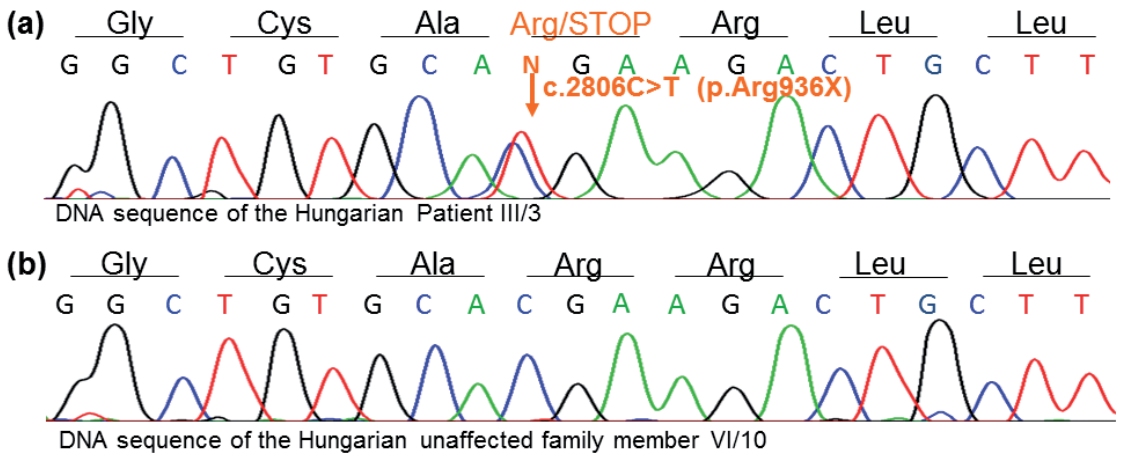

(c)

Haplotype analysis Polymorphisms		Hungarian BSS patient (V/2.)	Hungarian BSS patient (V/11.)	Hungarian BSS patient (V/12.)	Hungarian unaffected family member (VI/10.)	Anglo-Saxon BSS patient (IV/1.)	Anglo-Saxon BSS patient (IV/2.)
rs28705891	C/T	CC	CC	CC	CC	CC	CC
rs28654666	T/C	TT	TT	TT	TT	TT	TT
rs112993837	C/T	CC	CC	CC	CC	CC	CC
rs60077744	G/A	GG	GG	GG	GG	GG	GG
rs2160683	**T/A**	**TA**	**AA**	**TA**	**TT**	**TT**	**TT**
rs115042932	**C/G**	**CG**	**CG**	**CG**	**CC**	**CC**	**CC**
rs117998712	C/T	CC	CC	CC	CC	CC	CC
MUT CM001120	**C/T**	**CT**	**CT**	**CT**	**CC**	**CC**	**CC**
rs116979331	G/A	GG	GG	GG	GG	GG	GG
rs74822565	G/T	GG	GG	GG	GG	GG	GG
rs117713908	A/G	AA	AA	AA	AA	AA	AA
rs3743781	A/G	AA	AA	AA	AA	AA	AA
rs117537927	G/A	GA	GG	GG	GG	GG	GG

Fig. S1. Sequencing and haplotype analysis. (a) The DNA sequence of the affected Hungarian patient III/3 shows a heterozygous single nucleotide substitution c.2806C>T, which converts codon 936 CGA for arginine to TGA for stop codon; this mutation is designated p.Arg936X. (b) Wild-type sequence of genomic DNA spanning codons 933–939 of the *CYLD* gene. (c) Genotyping of the surrounding, common polymorphisms demonstrated that the same mutation carried by the 2 geographically distant pedigrees is surrounded by different haplotypes and was the result of 2 independent mutational events.