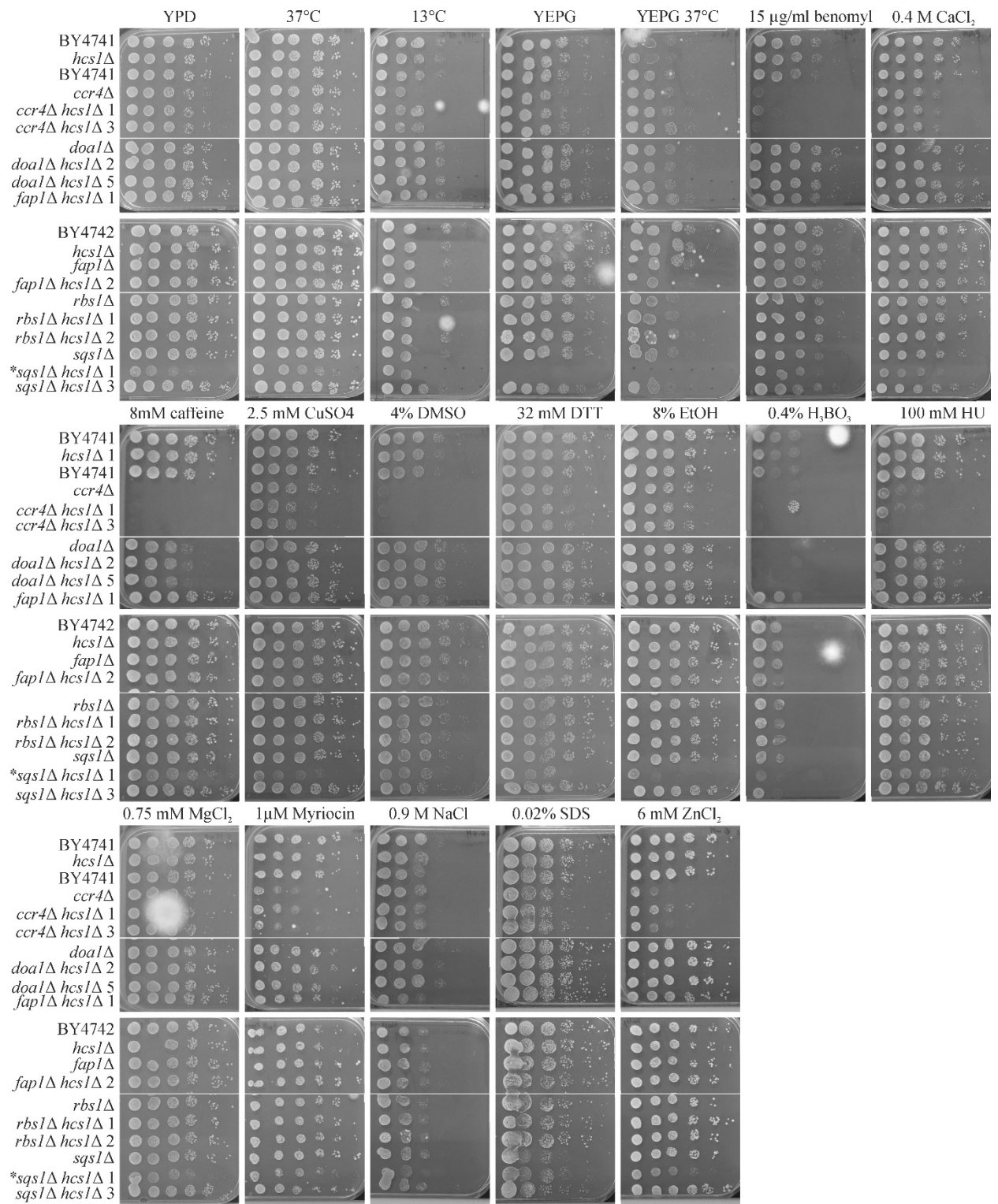


Table S1. The primer sequences for site directed mutagenesis.

Mutation	Primer Forward	Primer Reverse
<i>IGHMBP2</i>		
c.151C>G p.Gln51Glu	GTTTGCTGAAGCTGGAGGTATCCAGCC AG	CTGGCTGGATACCTCCAGCTTCAGCAA AC
c.163C>T p.Gln55*	CAGGTATCCAGCTAGCGCACTGGGC	GCCCAGTGCGCTAGCTGGATACCTG
c.223G>A p.Ala75Thr	CGATACGGGTCCACGGCAGCTCTTC	GAAGAGCTGCCGTGGACCCGTATCG
c.G595C p.Ala199Pro	CCTCCCAGAAAGAACCGGTTTTATTTG CGC	GCGCAAATAAAACCGGTTCTTTCTGGG AGG
c.734A>G p.Asn245Ser	GCGCCCCCTCCAGCATCGCCGTGGAC	GTCCACGGCGATGCTGGAGGGGGCGC
c.767C>G p.Ala256Gly	GTGGAGCGCCTGGGTCTGTGTAAGCA G	CTGCTTACACAGACCCAGGCGCTCCA C
c.823A>G p.Ile275Val	CTCCTGGAGTCCGTTTCAGCAGCACTC	GAGTGCTGCTGAACGGACTCCAGGAG
c.1082T>C p.Leu361Pro	CCGATGGCCCCCGAAGTTGCTGCC	GGCAGCAACTTCGGGGGGCCATCGG
c.C1478T p.Thr493Ile	CTCTTGGTGGACATCGCCGGCTGCGG	CCGCAGCCGGCGATGTCCACCAAGAG
c.G1738A p.Val580Ile	GTGATACTGTCCTTCATCAGATCCAAC AGG	CCTGTTGGATCTGATGAAGGACAGTAT CAC
c.C1794A p.Asn598Lys	CCGGAGGATCAAAGTGGCTGTCACC	GGTGACAGCCACTTTGATCCTCCGG
<i>HCS1</i>		
c.G643C p.Ala215Pro	GAATGACTCCCAGAAAACCTCCCATTA ATTTTGCCATTAAAC	GTAAATGGCAAAATTAATGGGAGTTTT CTGGGAGTCATTC
c.CG1574TC p.Thr525Ile	CAAAAATTCCTTTAATTTGGTATGATA TCCAAGGTGATGAATTTCAAGAGACT GC	GCAGTCTCTTGAAATTCATCACCTTGG ATATCATACCAAATTAAGGAATTTTT G
c.G1846A p.Val616Ile	GAAAAAGATGTTATCATATTGAGTTTA ATTCGTAGCAATGAAAAATTTGAAGTT G	CAACTTCAAATTTTTCATTGCTACGAA TTAAACTCAATATGATAACATCTTTTT C
c.C1902A p.Asn634Lys	GAAGAACGAAGACTGAAAGTCGCCAT GACAAGAC	GTCTTGTCATGGCGACTTTCAGTCTTCG TTCTTC



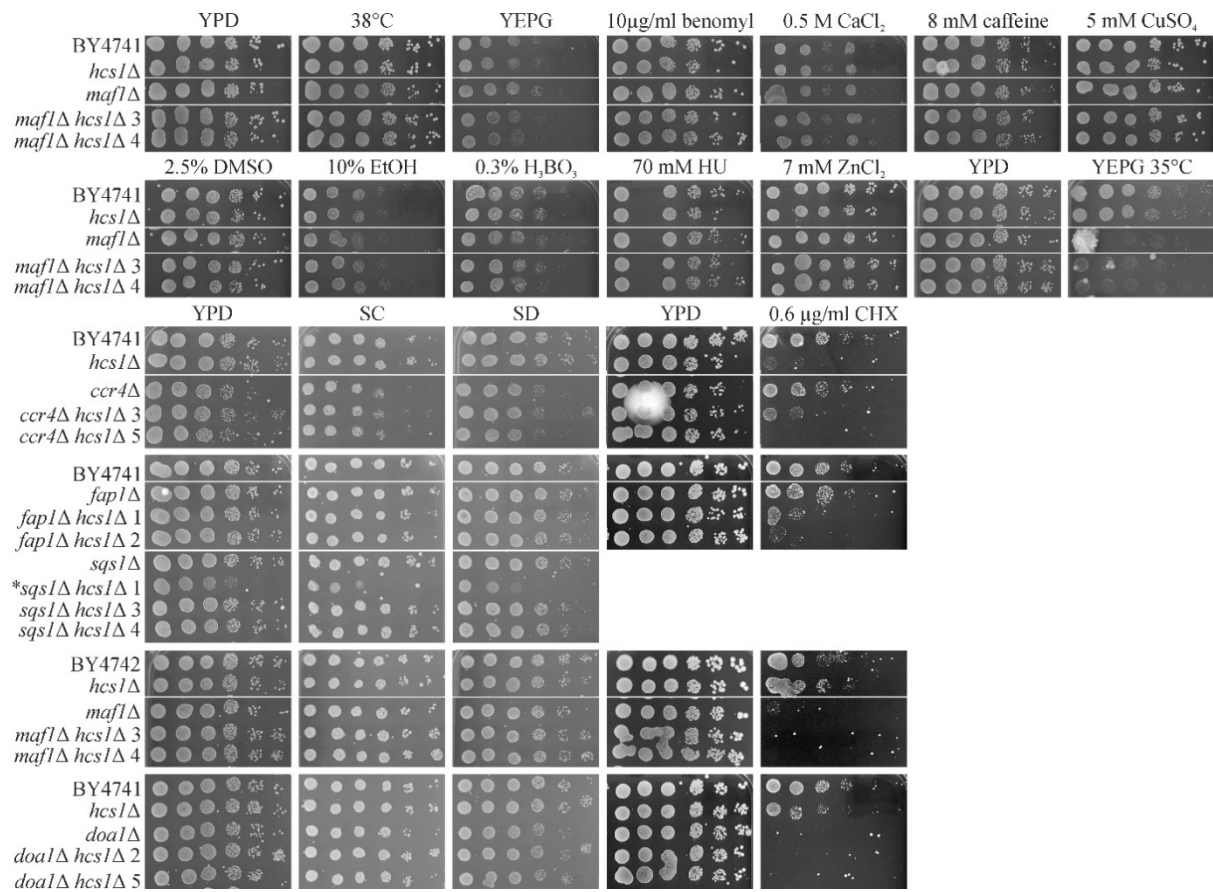


Figure S1. Growth of wild-type (BY4741 or BY4742 strain) and different mutant strains, in various conditions. The yeast cells were grown overnight in YPD medium, diluted to OD₆₀₀ ≈ 1 and ten-fold serial dilutions were spotted on YPD, YEPG, SD, SC or YPD medium with indicated supplements. Respiratory deficient strains are indicated by *.

		A199				T493				
IGHMBP2	192	LDTSQKE	A	VLFALSQ	206	486	VPLLLVD	T	AGCGLFE	500
Hcs1	208	LNDSQKT	A	INFAINN	222	518	IPLIWYD	T	QGDEFQE	532
		*:	***	*:	**::		::**:	**	*	: *
		A215				T525				
		V580				N598				
IGHMBP2	573	EAVILSF	V	RSNRKGE	587	591	LAEDRRIN	V	AVTRAR	605
Hcs1	609	DVIILSL	V	RSNEKFE	623	627	LKEERRLN	V	AMTRPR	641
		::**:	***	::***:	* *		* *::**:	***	::**	: *
		V616				N634				

Figure S2. The homology of regions containing the amino acids residues conserved between human IGHMBP2 and yeast Hcs1 proteins, which are substituted in patients (Ala199Pro, Thr493Ile, Val580Ile and Asn598Lys; in yeast respectively: Ala215Pro, Thr525Ile, Val616Ile and Asn634Lys). Alignment was prepared using Clustal Omega tool (EMBL-EBI).