Supplementary information to

Functional analysis and drug response to zinc and D-penicillamine in stable *ATP7B* mutant hepatic cell lines

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Supplementary tables

Table S1. Cell lines expressing *ATP7B* mutations

Cell line	Amino acid	Nucleotide	Type	Country
KO.L795F	p.Leu795Phe	c.2383C>T	Missense	Western India
KO.H1069Q	p.His1069Gln	c.3207C>A	Missense	Europe/United States[a]
KO.T977M	p.Thr977Met	c.2930C>T	Missense	Western India
KO.M573fs	p.Met573fs	c.1716delG	Deletion	Western India
KO.C271*	p.Cys271*	c.813C>A	Nonsense	India[a]
KO.E122fs	p.Glu122fs	c.365_366delinsTTCGAAGC	Deletion/Insertion	Western India
KO.R778L	p.Arg778Leu	c.2333G>T	Missense	Asia[a]

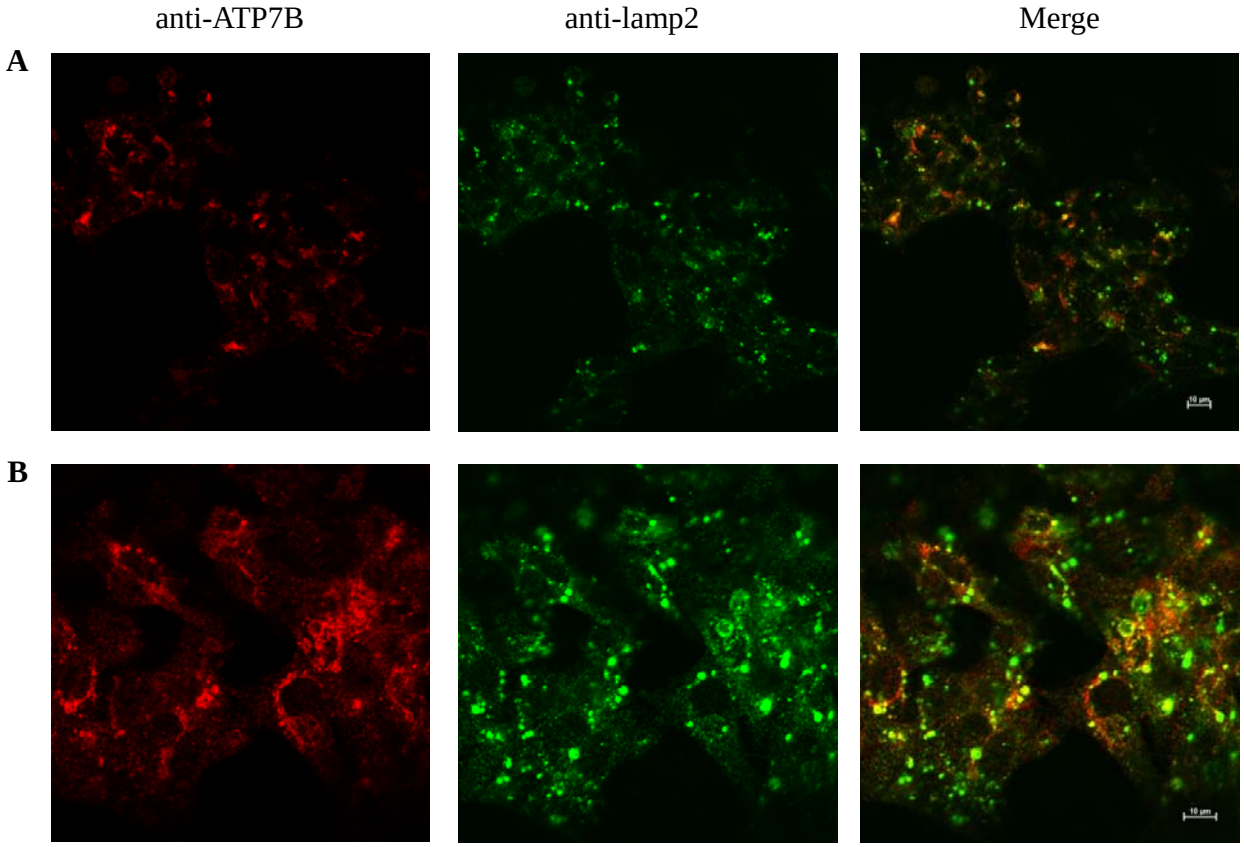
[a]Most frequent genotype of the country

Table S2. ATP7B protein expression and IC50

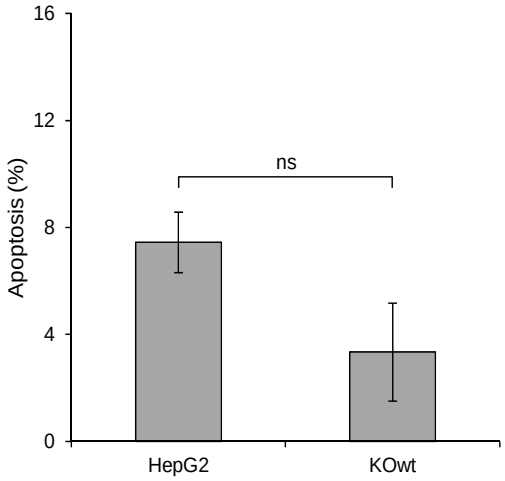
Cell line	ATP7B expression[a]		IC50
	37°C	30°C[b]	
KO.L795F	1.1±0.1	1.1±0.2 (1.1)	0.90±0.07
KO.H1069Q	0.6±0.0	0.9±0.1 (1.7)	0.62±0.02
KO.T977M	0.5±0.0	0.9±0.0 (1.8)	0.24±0.02
KO.M573fs	0.0±0.0	0.0±0.0 (0.0)	<0.20
KO.C271*	0.0±0.0	0.0±0.0 (0.0)	<0.20
KO.E122fs	0.0±0.0	0.0±0.0 (0.0)	<0.20
KO.R778L	0.2±0.0	0.4±0.0 (2.3)	<0.20
KO.wt	1.0±0.0	1.0±0.0 (1.0)	0.91±0.03
KO	0.0±0.0	0.0±0.0 (0.0)	<0.20

[a]relative protein expression versus KO.wt; [b]factor fold change versus 37°C (brackets)

Supplementary figures



Supplementary Figure S1 Localization of ATP7B protein in HepG2 cells. A representative photograph of co-localization with lamp2, a late endosome lysosome marker, at low copper (A) and in presence of 100 µM copper (B) is shown. Bar represents 10 µM. Note, that the staining pattern of HepG2 cells is similar to KO.wt cells.



Supplementary figure S2 Rate of apoptosis in HepG2 cells after copper exposure. Cells were exposed to 100 µM copper for 24 h. Induction of apoptosis was determined by Annexin-V staining followed by flow cytometric analysis. Mean ± SE of three independent experiments is shown. Note, that induction of apoptosis is at similar levels in both cell lines. ns, non-significant.