Table S1. Characterization of established cell lines

Cell	Mutation	Age	Gender	Clones
ATTR1	G47A	52	♂	2
ATTR2	V30M	34	♀	2
ATTR3	V30M	72	♀	2
ATTR4	A34T	60	♂	2
Healthy1	WT	29	♂	2
Healthy2	WT	25	♂	2
Healthy3	WT	52	♀	2
Healthy4	WT	57	♂	2

Table S2. Primers list

Click here to Download Table S2

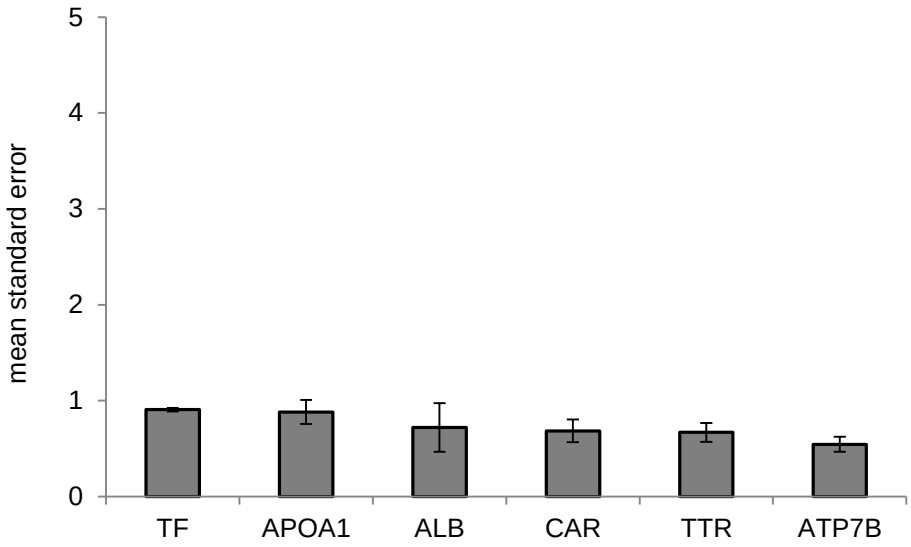

Fig S1. Variability of hepatic marker gene expression. First, hepatic marker variability (means of standard error) of ATTR and H-HLCs (ΔCt vs *GAPDH*) was determined. Then, means of both groups were established as depicted above. Standard deviation is given (data from three different experimental repeats).

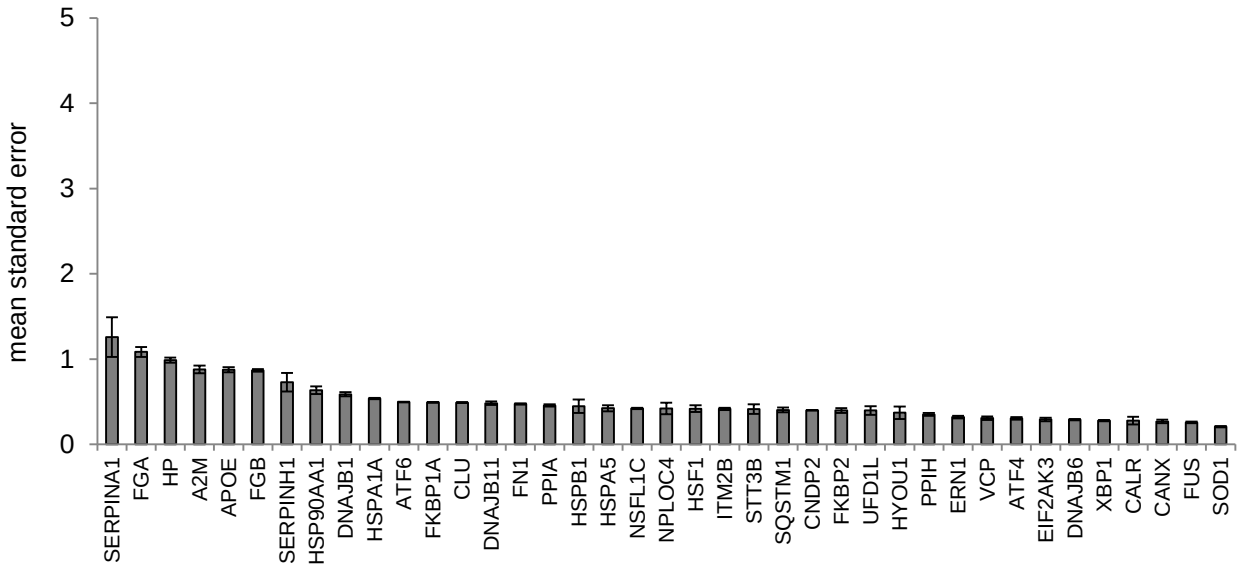

Fig S2. Variability of chaperone gene expression. Variability of chaperone gene expression of ATTR-HLCs and H-HLCs was determined. Mean of the variability and standard deviation obtained in the HLC groups is shown for at least two cell lines and several experiments (data from at least three different experimental repeats).

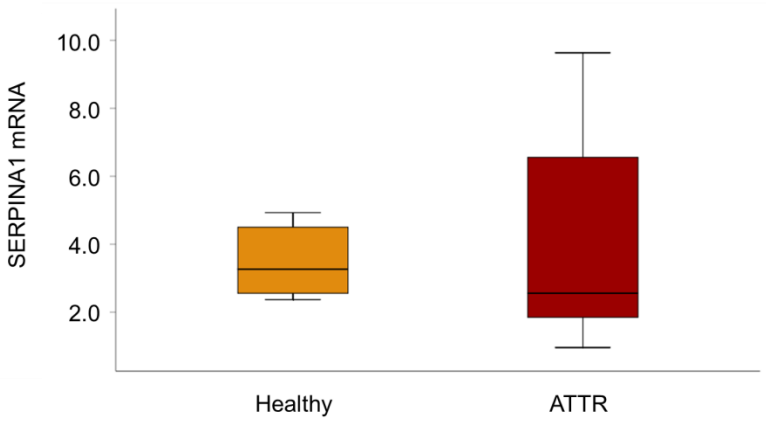

Fig. S3. SERPINA1 variability in ATTR-HLCs and H-HLCs. Boxplot representation of SERPINA1 mRNA expression in all four ATTR-HLCs and H-HLCs. ΔCt versus *GAPDH* was determined (data from three different experimental repeats).

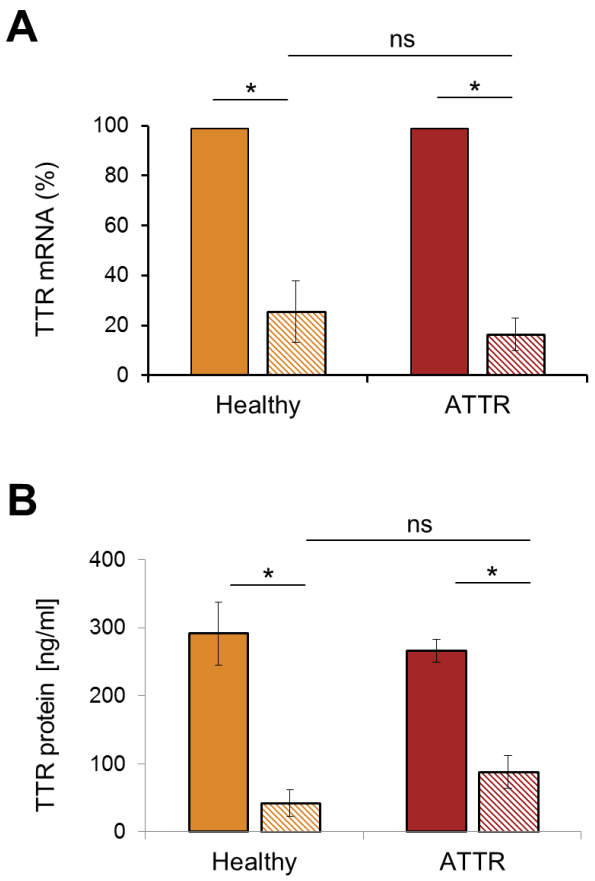

Fig. S4. TTR knockdown in ATTR-HLCs and H-HLCs. (A) ATTR-HLCs and H-HLCs were treated with oligonucleotides as previosuly described (Niemietz et al., 2016). mRNA knockdown (dashed) was determined by RT-qPCR relative to untreated. Means of four ATTR-HLCs and four H-HLCs are shown (data from three different experimental repeats). (B) TTR protein levels in cell culture supernatants of ATTR-HLCs and H-HLCs as determined by ELISA. TTR knockdown efficiency (dashed) was compared with untreated control. Means of four ATTR-HLCs and four H-HLCs are shown (data from three different experimental repeats). ns, not significant. *P<0.05.

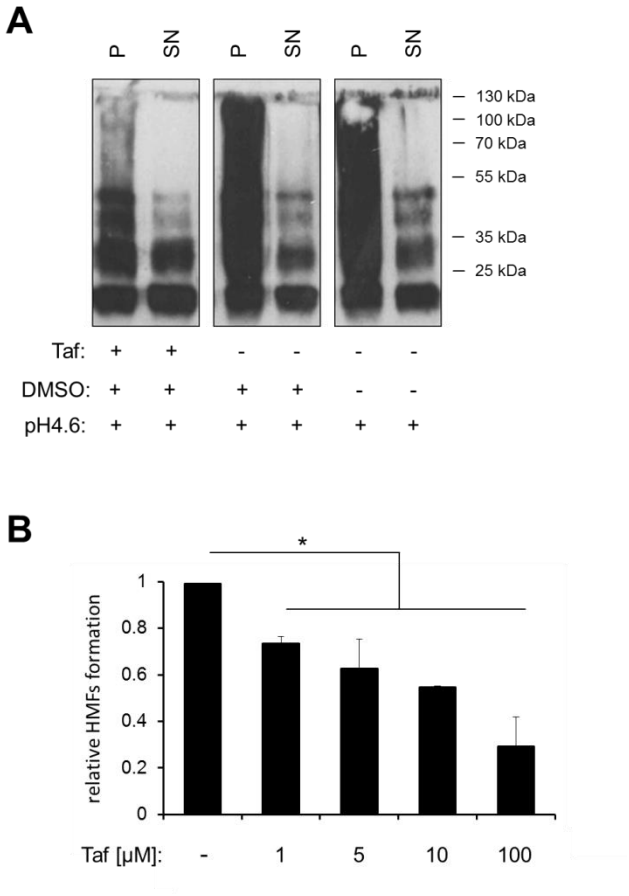

Fig. S5. Attenuation of TTR HMFs formation by tafamidis-meglumine. (A) Plasma-isolated TTR was pre-incubated with 100μM tafamdis-meglumine (Taf) and exposed to HMF formation using acetate buffer (pH4.6) and incubation for 72 hours. After centrifugation, insoluble pellet (P) and soluble supernatant (SN) fractions were subjected to SDS-PAGE. DMSO was used as control. (B) Dose-dependent inhibition of TTR HMFs formation by tafamidis-meglumine as determined by densitometric quantification (data from three different experimental repeats). **P*<0.05.

References

Niemietz, C. J., Sauer, V., Stella, J., Fleischhauer, L., Chandhok, G., Guttmann, S., Avsar, Y., Guo, S., Ackermann, E. J., Gollob, J. et al. (2016). Evaluation of Therapeutic Oligonucleotides for Familial Amyloid Polyneuropathy in Patient-Derived Hepatocyte-Like Cells. *PloS one*. **11**, e0161455.