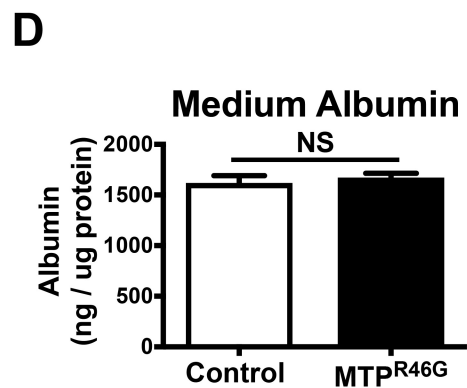
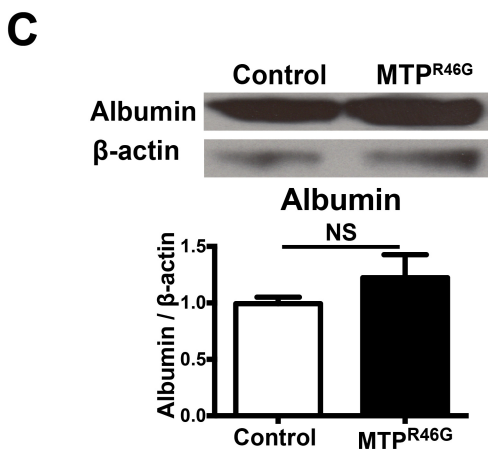
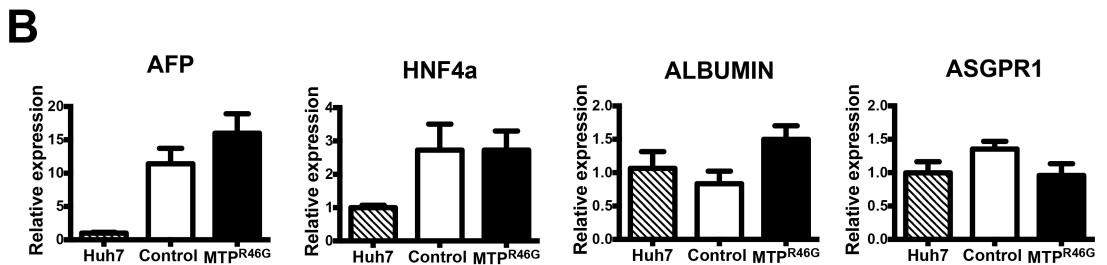
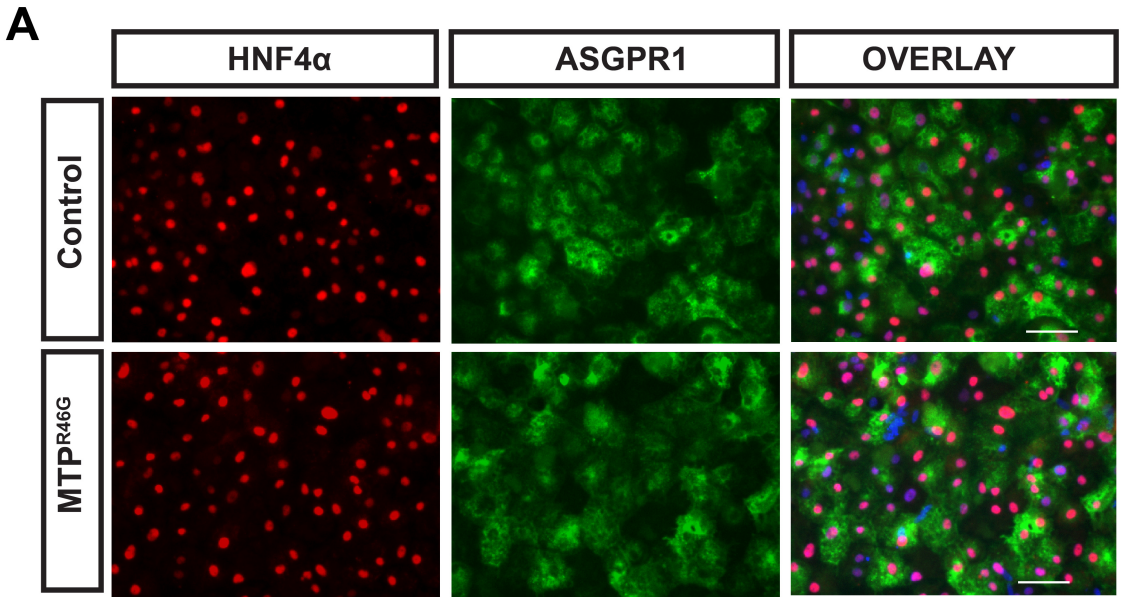


Supplement Figure 1: Generation of ABL-specific iPSCs and confirmation of mutation.

Related to Figure 1

(A) Sequence of MTP in iPSCs from the ABL patient showing a missense mutation c.136 C>G, p. Arg46Gly. (B) Predicted MTP protein structure and location of the mutation. Image used with permission from Walsh et al., 2015. (C) Representative images showing morphologies of iPSCs from control subjects. Scale bar: 200 μ m. (D) Quantification of SSEA4 and TRA1-60 by FACS analysis. Values are means and SEM for three or four independent experiments for all lipid analysis.



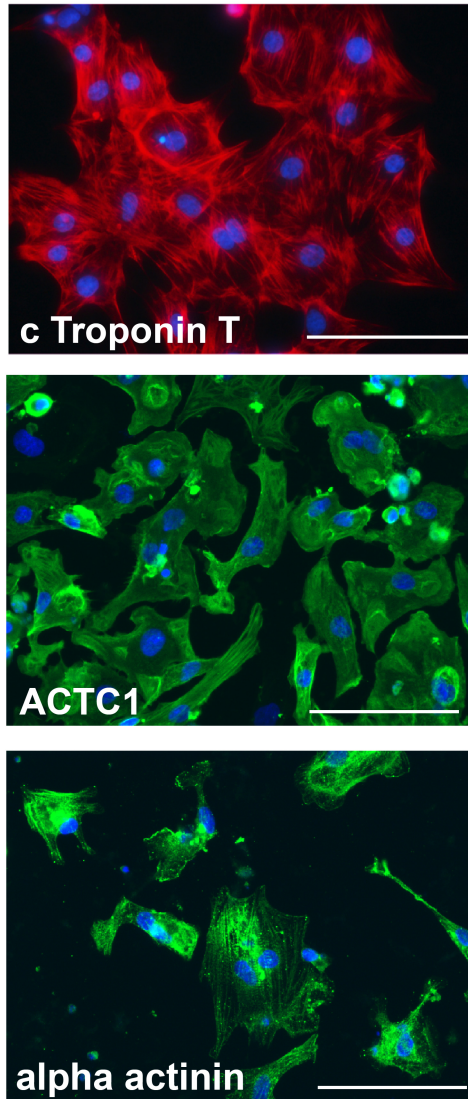
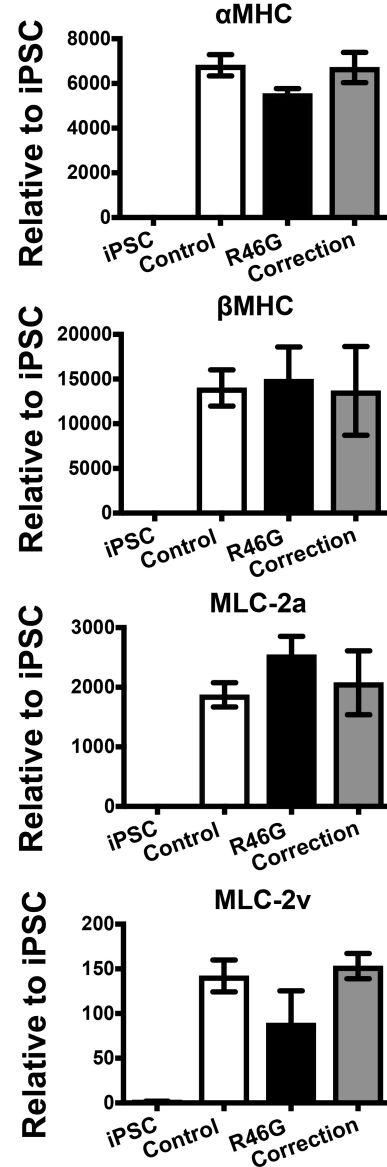
Supplement Figure 2: Differentiation of iPSCs into hepatocytes.

Related to Figure 1

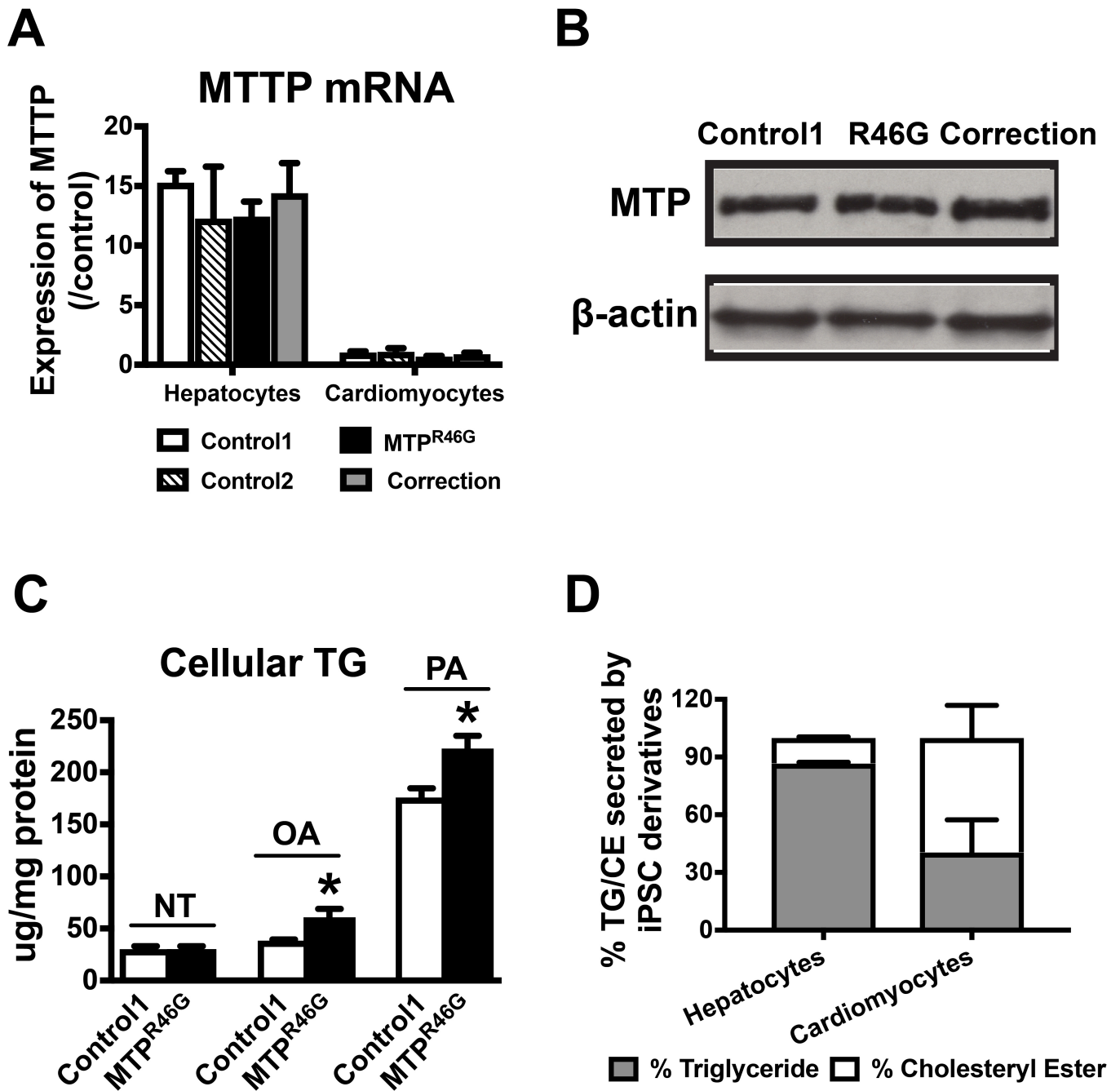
(A) Representative images showing positive immunostaining of hepatocyte markers HNF4 α (red) and ASGPR1 (green). Scale bar: 100 μ m.

(B) Hepatic genes, such as HNF4 α , ALBUMIN, AFP, and ASGPR1 were analyzed by real-time PCR.

(C-D) Cellular and medium albumin levels were measured by western blotting and ELISA respectively. β -actin was blotted as a loading control. \pm S.D. Values are means for three experiments for all lipid analysis.

A**B****Supplement Figure 3: Differentiation of iPSCs into cardiomyocytes****Related to Figure 4**

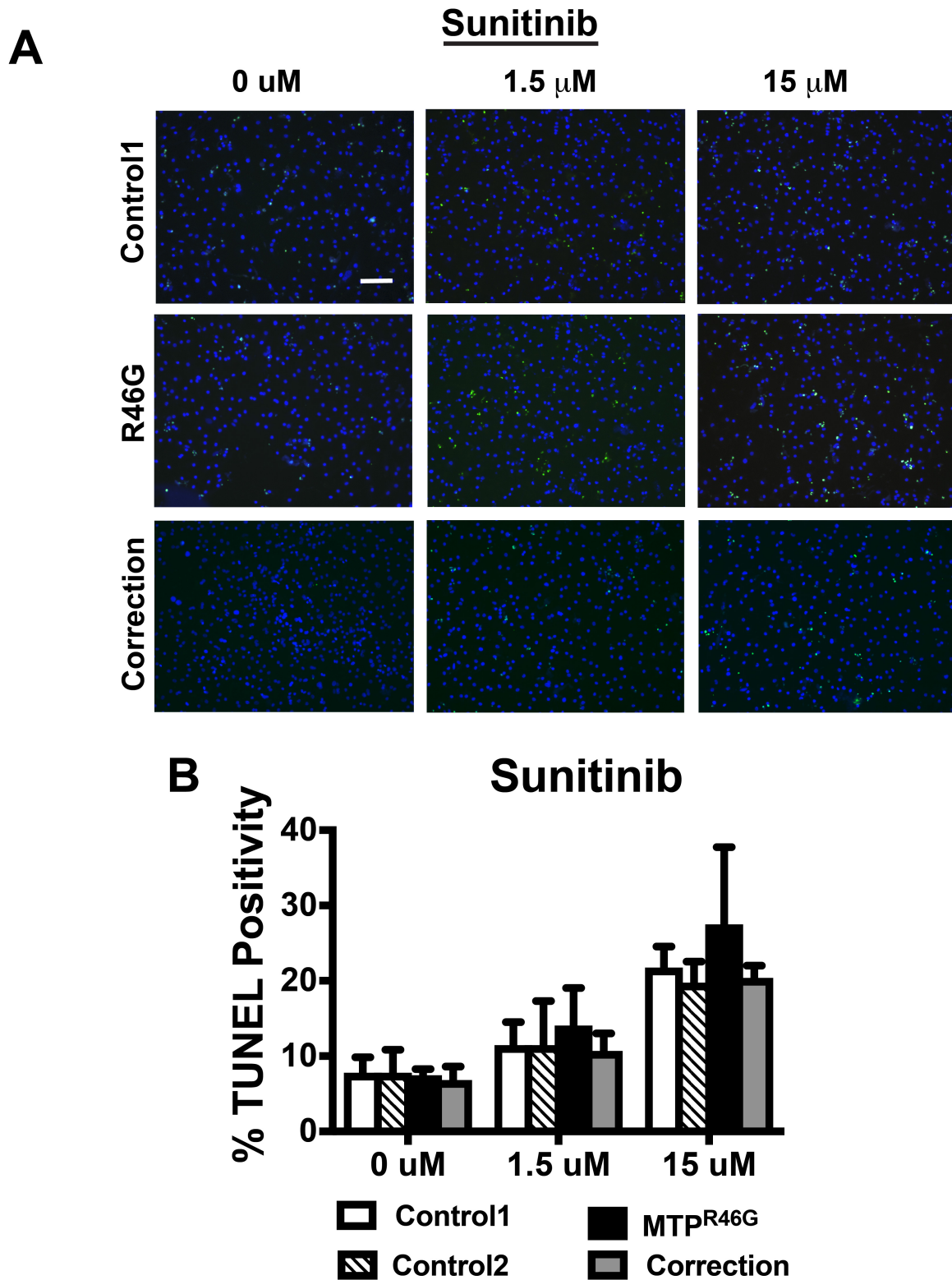
(A) Representative images of immunofluorescence staining of cardiomyocyte markers in iPSCs derived cardiomyocytes from control subjects. Scale bar: 200 μ m (B) Expression of cardiac-specific markers, such as, α MHC, β MHC, MLC2A, and MLC2V. \pm S.D. Values are means for three independent experiments.



Supplement Figure 4: Elevated cellular TG in MTP^{R46G} cardiomyocytes.

Related to Figure 4

(A-B) Quantification of messenger RNA MTTP and protein of MTP by real-time PCR and western blot. (C) Cellular TG contents were measured by enzymatic assays and normalized to total protein. (D) Percentage of secreted [¹⁴C] OA counts in either TG or CE as determined by thin layer chromatography separation in hepatocytes and cardiomyocytes. \pm S.D. * $P < 0.05$. Values are means for three independent experiments.



Supplement Figure 5: Sunitinib and staurosporine induced dose-dependent apoptosis in iPSC-derived cardiomyocytes.

Related to Figure 5

(A) Representative images of TUNEL staining in sunitinib (0, 1.5 μ M, 15 μ M) treated cardiomyocytes.

Scale bar: 150 μ m. (B) Percentage of apoptotic cells were counted by Image J (>1000 cells per condition).

(C) Percentage of TUNEL positivity in iPSC-derived cardiomyocytes induced by staurosporine (1 μ M) and PA (0.5mM).

(D) Expression of Caspase3 and Caspase9 upon staurosporine and PA treatment were analyzed by real-time PCR. \pm S.D.

*P<0.05. Values are means for three independent experiments.

**Table S1. Subject Lipid Panel Results
Related to Figure 1**

Subject	Race	Sex	Age	TC	VLDL	LDL	HDL	TG	ApoAI	ApoB
				mg/dl						
Control (M8)	White	Male	66	166	22	102	42	130	137	85
ABL (M7)	White	Male	60	52	-	13	40	10	61	10

TC: total cholesterol; VLDL: very low density lipoprotein; LDL: low density lipoprotein; HDL: high density lipoprotein; TG: triglyceride; ApoAI: apolipoprotein A-I; ApoB: apolipoprotein B; "-" indicates undetectable; Age reflects age at time of visit.

**Table S2. IPS clones from subjects
Related to Figure 1**

Subject	iPSC clones	WiCell ID
Control1	iPS-M8-SeV2	PENN156i-M8-2*
	iPS-M8-SeV3	N/A
	iPS-M8-SeV5	N/A
Control2	iPS-SV20	PENN123i-SV20*
	iPS-SV10	PENN078i-SV10*
ABL (R46G)	iPS-M7-SeV16	PENN144i-M7-16*
	iPS-M7-SeV14	N/A
	iPS-M7-SeV9	N/A

N/A: not available. *Detailed cell line information can be found at:
<http://www.wicell.org/home/stem-cell-lines/catalog-of-stem-cell-lines/collections/nhlbi-next-gen-rader.cmsx>