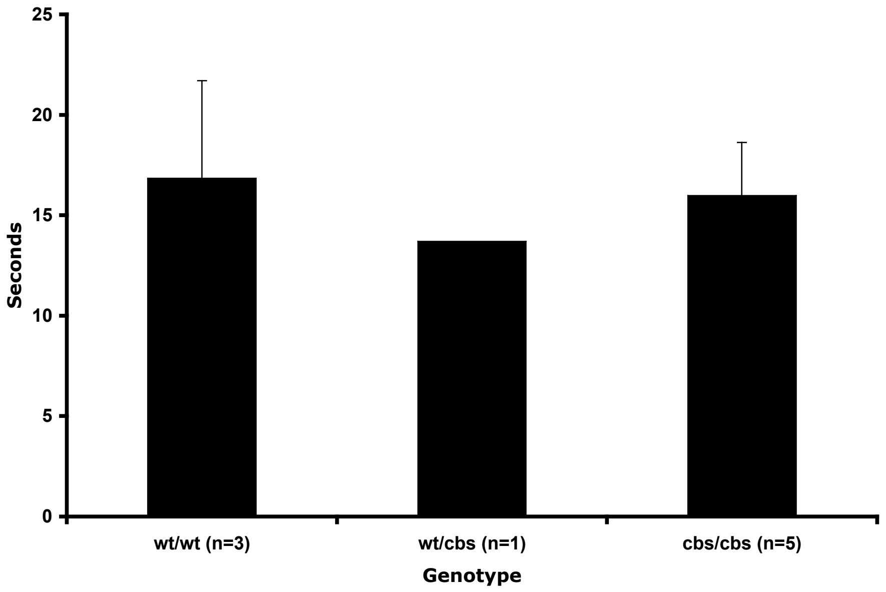
Supplemental Figure Legends

Supplemental Figure 1. Prothrombin and activated partial thromboplastin time in Tg-*I278T* *Cbs*$^{-/-}$ and control mice. (A) Prothrombin times. The *Cbs* genotype and the number of animals tested is shown on the X-axis. Error bars show standard deviation. (B). Activated partial thromboplastin time.

A.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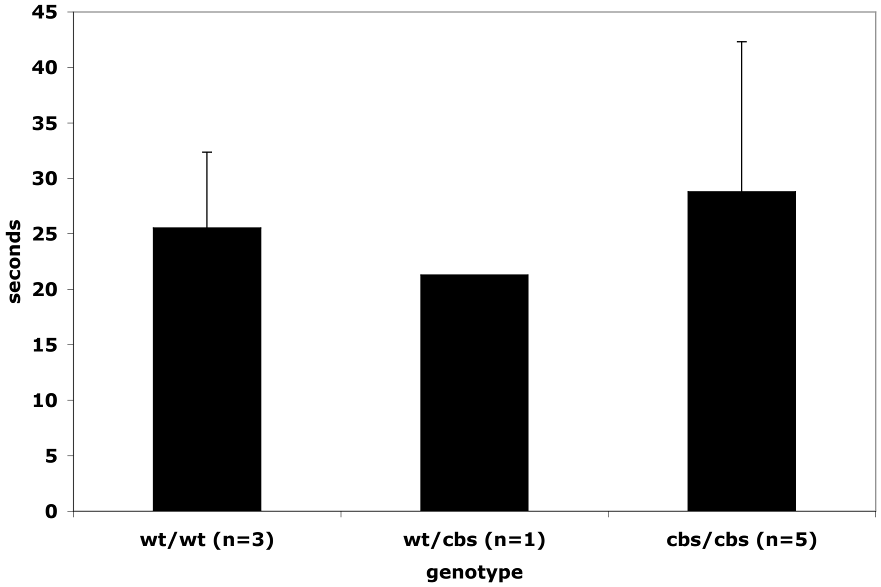
20
15
10
5
0
Seconds
wt/wt (n=3)
wt/cbs (n=1)
cbs/cbs (n=5)
Genotype

B.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40
35
30
25
20
15
10
5
0
seconds
wt/wt (n=3)
wt/cbs (n=1)
cbs/cbs (n=5)
genotype

Supplemental Table 1
Pathways upregulated in *Tg-I278T Cbs*$^{-/-}$ mice

KEGG pathway	**Genes upregulated**	**Enrichment**
Cytokine-cytokine receptor interaction^{A}	Acvr1b; Il8rb; Flt1; Prlr; Ccl25; Ccl6; Vegfa; Il28ra; Il17rb; Ccl24; Cxcl4	O=11; E=2.84; R=3.87; P=1.53e-4
Metabolism of xenobiotics by cytochrome P450	Adh1; Cyp2b10; Cyp2b13; Cyp2b9; Gstm1; Mgst3; Aldh3b1	O=7; E=0.69; R=10.2; P=5.65e-6
Focal adhesion^{A}	Itga9; Col5a2; Flt1; Igf1; Itgb6; Thbs1; Vegfa	O=7; E=2.26; R=3.09; P=8.05e-3
Arachidonic acid metabolism	Cbr3; Cbr1; Cyp2b10; Cyp2b13; Cyp2b9; Cyp4a14; Cyp4f16	O=7; E=0.81; R=8.62; P=1.71e-5
Fructose and mannose metabolism^{B}	Akr1b7; Fbp2; Pfkfb2; Gmds; Pfkp	O=5; E=0.59; R=8.46; P=3.06e-4
Pyrimidine metabolism	Aicda; Pnp; Rpo1-4; Umps; Dctd	O=5; E=1.01; R=4.95; P=3.47e-3
TGF-beta signaling pathway	Acvr1b; Smad3; Smad5; Myc; Thbs1	O=5; E=1.01; R=4.95; P=3.47e-3
Chronic myeloid leukemia	Acvr1b; Runx1; Cdkn1a; Smad3; Myc	O=5; E=0.91; R=5.49; P=2.22e-3
Glycolysis / Gluconeogenesis	Adh1; Fbp2; Pfkp; Aldh3b1	O=4; E=0.66; R=6.02; P=4.41e-3
Glycine, serine and threonine metabolism	Psat1; Chdh; Phgdh; Abp1	O=4; E=0.58; R=6.92; P=2.66e-3
Tyrosine metabolism	Adh1; Aldh3b1; Prmt3; Abp1	O=4; E=0.62; R=6.5; P=3.34e-3
mTOR signaling pathway	Igf1; Vegfa; Frap1; Ddit4	O=4; E=0.59; R=6.77; P=2.87e-3
Pentose phosphate pathway	Fbp2; G6pdx; Pfkp	O=3; E=0.30; R=10.2; P=3.09e-3
Histidine metabolism	Aldh3b1; Prmt3; Abp1	O=3; E=0.41; R=7.39; P=7.69e-3
Glycan structures - degradation	Arsb; Ids; Neu3	O=3; E=0.32; R=9.38; P=3.90e-3
C21-Steroid hormone metabolism^{C}	Akr1c18; Cyp17a1	O=2; E=0.15; R=13.5; P=9.19e-3
Biocarta pathway	**Genes upregulated**	**Enrichment**
CTCF: First Multivalent Nuclear Factor	Smad5; Myc; Frap1	O=3; E=0.22; R=13.5; P=1.32e-3

[A]pathways showing common behavior in both mice models; [B]pathways showing contrast behavior; [C]different genes showed upregulation and downregulation within the same pathway; O, observed gene number in particular pathway; E, expected number of genes in a specific pathway for an interesting gene set; R, ration of enrichment (O/E); P, significance of enrichment calculated from hypergeometric test.

Supplemental Table 2

Pathways downregulated in *Tg-I278T Cbs*$^{-/-}$ mice

KEGG pathway	**Genes downregulated**	**Enrichment**
PPAR signaling pathway[A]	Cyp8b1; Fabp2; Hmgcs2; Pltp; Scd1; Sorbs1; Acsl6; Fads2	O=8; E=0.65; R=12.3; P=2.93e-7
Neuroactive ligand-receptor interaction	Npffr2; Gabrb3; Ghr; Grid1; Grin3b; Mtnr1a; Hcrtr2; Gria3	O=8; E=2.77; R=2.89; P=7.12e-3
MAPK signaling pathway[A]	Bdnf; Cacna1d; Gadd45a; Fgf12; Fgf9; Ntrk2; Dusp14; Map3k7ip2	O=8; E=2.51; R=3.19; P=3.96e-3
Adipocytokine signaling pathway[A]	Acacb; G6pc; Acsl6; Adipor2	O=4; E=0.67; R=5.98; P=4.59e-3
Androgen and estrogen metabolism[B]	Hsd3b2; Hsd3b4; Hsd3b5; Bcdo2	O=4; E=0.33; R=12.1; P=3.23e-4
Phosphatidylinositol signaling system	Pik3c2g; Pip5k3; Fn3K; Plce1	O=4; E=0.65; R=6.15; P=4.14e-3
Nicotinate and nicotinamide metabolism	Aox1; Bcdo2; Fn3k; Nmnat3	O=4; E=0.35; R=11.5; P=4.01e-4
Methionine metabolism	Bhmt; Cbs; Mat2a	O=3; E=0.18; R=16.8; P=7.19e-4
C21-Steroid hormone metabolism[C]	Hsd3b2; Hsd3b4; Hsd3b5	O=3; E=0.11; R=26.5; P=1.71e-4
Benzoate degradation via CoA ligation	Pnpla3; Acot11; Fn3k	O=3; E=0.25; R=11.8; P=2.05e-3
Valine, leucine and isoleucine degradation	Pnpla3; Aox1; Hmgcs2	O=3; E=0.41; R=7.24; P=8.26e-3

[A] pathways showing common behavior in both mice models; [B] pathways showing contrast behavior; [C] different genes showed upregulation and downregulation within the same pathway; O, observed gene number in particular pathway; E, expected number of genes in a specific pathway for an interesting gene set; R, ration of enrichment (O/E); P, significance of enrichment calculated from hypergeometric test.

Supplemental Table 3
Pathways upregulated in *Tg-hCBS Cbs*$^{-/-}$ mice

KEGG pathway	**Genes upregulated**	**Enrichment**
Cytokine-cytokine receptor interaction[A]	Flt1; Ghr; Hgf; Cxcl10; Ifng; Cxcl13	O=6; E=0.94; R=6.39; P=3.83e-4
Calcium signaling pathway	Phka2; Pln; D830007F02Rik; Avpr1a; Phkg2	O=5; E=0.70; R=7.16; P=7.14e-4
Focal adhesion[A]	Birc4; Arhgap5; Flt1; Hgf; D830007F02Rik	O=5; E=0.75; R=6.69; P=9.66e-4
Leukocyte transendothelial migration	Arhgap5; Cldn1; Thy1; Cxcl13	O=4; E=0.45; R=8.87; P=1.12e-3
Huntington's disease	Ncor1; Cltc	O=2; E=0.10; R=18.9; P=5.00e-3
Androgen and estrogen metabolism[B]	Hsd3b4; Hsd3b5	O=2; E=0.14; R=14.1; P=8.94e-3
C21-Steroid hormone metabolism[B]	Hsd3b4; Hsd3b5	O=2; E=0.05; R=41.1; P=1.05e-3
Biocarta pathway	**Genes upregulated**	**Enrichment**
Rho cell motility signaling pathway	Arhgap5; D830007F02Rik	O=2; E=0.07; R=27.4; P=2.40e-3
Erk and PI-3 Kinase Are Necessary for Collagen Binding in Corneal Epithelia	Arhgap5; D830007F02Rik	O=2; E=0.08; R=25.9; P=2.68e-3

[A] pathways showing common behavior in both mice models; [B] pathways showing contrast behavior; O, observed gene number in particular pathway; E, expected number of genes in a specific pathway for an interesting gene set; R, ration of enrichment (O/E); P, significance of enrichment calculated from hypergeometric test.

Supplemental Table 4
Pathways downregulated in *Tg-hCBS* $Cbs^{-/-}$ mice

KEGG pathway	**Genes downregulated**	**Enrichment**
MAPK signaling pathway[A]	Cacna1d; Mknk2; Gadd45b; Ntrk1; Ntrk2; Dusp8; Hspa1a; Rasgrp2; Cacna2d2	O=9; E=1.83; R=4.90; P=1.07e-4
PPAR signaling pathway[A]	Cd36; Cpt1a; Cyp4a10; Cyp4a14; Mod1; Pltp; Pparg; Scd1	O=8; E=0.48; R=16.8; P=2.70e-8
Insulin signaling pathway	Acacb; Fasn; Mknk2; Slc2a4; Socs2	O=5; E=0.91; R=5.49; P=2.29e-3
Adipocytokine signaling pathway[A]	Acacb; Cd36; Cpt1a; Slc2a4	O=4; E=0.49; R=8.17; P=1.50e-3
Pyruvate metabolism	Acacb; Akr1b7; Mod1; Acss2	O=4; E=0.28; R=14.14; P=1.84e-4
Type II diabetes mellitus	Cacna1d; Slc2a4; Socs2	O=3; E=0.31; R=9.66; P=3.72e-3
Glycerolipid metabolism	Agpat6; Pnpla3; Akr1b7	O=3; E=0.31; R=9.66; P=3.72e-3
Fructose and mannose metabolism[B]	Akr1b7; Pfkfb3; Pmm1	O=3; E=0.33; R=9.06; P=4.47e-3
Fatty acid metabolism	Cpt1a; Cyp4a10; Cyp4a14	O=3; E=0.26; R=11.45; P=2.29e-3
Fatty acid biosynthesis	Acacb; Fasn	O=2; E=0.034; R=58; P=4.68e-4
Biocarta pathway	**Genes downregulated**	**Enrichment**
Nuclear Receptors in Lipid Metabolism and Toxicity	Abcb1a; Pparg; Abcd2	O=3; E=0.21; R=14 ; P=1.27e-3

[A] pathways showing common behavior in both mice models; [B]pathways showing contrast behavior; O, observed gene number in particular pathway; E, expected number of genes in a specific pathway for an interesting gene set; R, ration of enrichment (O/E); P, significance of enrichment calculated from hypergeometric test.

Supplemental Table 5
Commonly upregulated genes in *Tg-hCBS Cbs*$^{-/-}$ and *Tg-I278T Cbs*$^{-/-}$ mice

S. No	**Gene Symbol**	**Gene Name**	**Log FC (*Tg-hCBS Cbs*$^{-/-}$)**	**Log FC (*Tg-I278T Cbs*$^{-/-}$)**
1	Cdh1	cadherin 1	1.35	1.42
2	Cdh9	cadherin 9	1.17	1.40
3	Cldn1	claudin 1	1.03 ^{A}	1.60
4	Prnpip1	prion protein interacting protein 1	1.51	1.46
5	Flt1	FMS-like tyrosine kinase 1	1.63	1.35
6	H2-Bl	histocompatibility 2, blastocyst	2.73 ^{A}	1.85 ^{A}
7	Meox1	mesenchyme homeobox 1	1.65	1.55 ^{A}
8	Mep1b	meprin 1 beta	1.05	1.97
9	Mobp	myelin-associated oligodendrocytic basic protein	1.90	1.43
10	Pln	phospholamban	1.91	1.81
11	Sftpb	surfactant associated protein B	1.24	1.83
12	Syn1	synapsin I	1.02	1.32
13	AW456874 (Ptpdc1)	Protein tyrosine phosphatase domain containing 1	1.11	1.04
14	Zik1	zinc finger protein interacting with K protein 1	1.005 ^{A}	1.02
15	2610207I05Rik	RIKEN cDNA 2610207I05 gene	1.56	1.09
16	Asns	asparagine synthetase	2.45	3.86

17	Ablim3	actin binding LIM protein family, member 3	1.09	1.17
18	Ccdc66	coiled-coil domain containing 66	1.02	1.26
19	Flrt1	fibronectin leucine rich transmembrane protein 3	1.46	1.06
20	1810033M07Rik (Erp27)	Endoplasmic reticulum protein 27	2.16	1.11
21	Serpinb12	serine (or cysteine) peptidase inhibitor, clade B (ovalbumin), member 12	1.23	1.16
22	Ppp1r14c	protein phosphatase 1, regulatory (inhibitor) subunit 14c	1.69	1.41
23	Atp11b	ATPase, Class VI, type 11B	1.45	1.65

[A] Average of two probes

Supplemental Table 6

Commonly downregulated genes in *Tg-hCBS Cbs*$^{-/-}$ and *Tg-I278T Cbs*$^{-/-}$ mice

S. No	**Gene Symbol**	**Gene Name**	**Log FC (*Tg-hCBS Cbs*$^{-/-}$)**	**Log FC (*Tg-I278T Cbs*$^{-/-}$)**
1	Acacb	acetyl-Coenzyme A carboxylase beta	-1.64	-1.95
2	Tmem98	transmembrane protein 98	-1.30	-1.02
3	Lrrc39	leucine rich repeat containing 39	-1.68 ^{A}	-1.21
4	Gpr98	G protein-coupled receptor 98	-1.23	-1.91 ^{A}
5	Pnpla3	patatin-like phospholipase domain containing 3	-4.09	-1.67
6	Cacna1d	calcium channel, voltage-dependent, L type, alpha 1D subunit	-1.01	-1.37
7	Camk2b	calcium/calmodulin-dependent protein kinase II, beta	-1.13	-2.73
8	Cbs	cystathionine beta-synthase	-2.03	-2.24
9	Dnase1	deoxyribonuclease I	-1.16	-1.34
10	Lefty1	left right determination factor 1	-1.10	-1.33
11	H2-Q1	histocompatibility 2, Q region locus 1	-2.01	-3.24
12	Elovl6	ELOVL family member 6, elongation of long chain fatty acids (yeast)	-1.94	-1.45
13	Ntrk2	neurotrophic tyrosine kinase, receptor, type 2	-2.36 ^{A}	-2.33
14	Pltp	phospholipid transfer protein	-1.98	-1.09
15	Robo1	roundabout homolog 1 (Drosophila)	-1.52	-1.14
16	Scd1	stearoyl-Coenzyme A desaturase 1	-1.59	-2.81

17	Wdr67	WD repeat domain 67	-1.03	-1.08
18	Tpm2	tropomyosin 2, beta	-2.14[A]	-1.23[A]
19	Ccdc37	coiled-coil domain containing 37	-1.14	-2.1[A]
20	Olfr887	olfactory receptor 887	-1.02	-1.28
21	Glt25d2	glycosyltransferase 25 domain containing 2	-1.134	-1.65[A]
22	Omd	osteomodulin	-1.05	-1.84
23	Tnfrsf19	tumor necrosis factor receptor superfamily, member 19	-1.08	-1.61
24	Rgs3	regulator of G-protein signaling 3	-1.51[B]	-1.42[B]
25	Gria3	glutamate receptor, ionotropic, AMPA3 (alpha 3)	-1.42	-1.06
26	Acss2	acyl-CoA synthetase short-chain family member 2	-1.02	-1.00
27	Hamp2	hepcidin antimicrobial peptide 2	-1.87	-1.50
28	Wfdc2	WAP four-disulfide core domain 2	-1.099	-1.29
29	Mogat1	monoacylglycerol O-acyltransferase 1	-2.33[B]	-2.74[B]
30	Cgref1	cell growth regulator with EF hand domain 1; CGR11	-2.04	-1.45
31	Mllt3	myeloid/lymphoid or mixed lineage-leukemia translocation to 3 homolog (Drosophila)	-2.64	-1.10
32	Osbpl3	oxysterol binding protein-like 3	-1.44[A]	-1.62
33	Sulf2	sulfatase 2	-1.18[A]	-1.59[A]
34	Mpped2	metallophosphoesterase domain containing 2	-1.94[A]	-3.785[A]
35	Tchh	trichohyalin	-1.28	-1.09

[A] Average of two probes; [B] Average of three probes

Supplemental Table 7
Metabolic profiling of serum

	μM			One way ANOVA		Tukey's Multiple Comparison Test		
Metabolite	Control (n=7)	*Tg-hCBS Cbs*$^{-/-}$ (n=6)	*Tg-I278T Cbs*$^{-/-}$ (n=7)	p value	p value summary	Control vs *Tg-hCBS Cbs*$^{-/-}$	Control vs *Tg-I278T Cbs*$^{-/-}$	*Tg-hCBS Cbs*$^{-/-}$ vs *Tg-I278T Cbs*$^{-/-}$
Phosphoserine^{A}	4.18 ± 0.99	3.68 ± 1.36	5.35 ± 2.16	0.2234	ns	$p>0.05$	$p>0.05$	$p>0.05$
Taurine	422 ± 99.2	323 ± 150	558 ± 303	0.148	ns	$p>0.05$	$p>0.05$	$p>0.05$
Phosphoethanolamine	12.8 ± 4.18	8.71 ± 4.91	15.2 ±8.27	0.1929	ns	$p>0.05$	$p>0.05$	$p>0.05$
Urea	10620 ± 352	9300 ± 1190	9296 ± 1576	0.0789	ns	$p>0.05$	$p>0.05$	$p>0.05$
Asp^{B}	11.4 ± 1.94	ND	14.5 ± 2.72	0.1828	ns			
Thr	142 ± 20.5	156 ± 7.37	228 ± 48.4	0.0002	***	$p>0.05$	$\underline{p<0.001}$	$\underline{p<0.01}$
Ser	117 ± 32	104 ± 13.8	172 ± 39.3	0.0023	**	$p>0.05$	$\underline{p<0.05}$	$\underline{p<0.01}$
Asn	62 ± 17.5	53.1 ± 9.34	74.7 ± 35.2	0.2866	ns	$p>0.05$	$p>0.05$	$p>0.05$
Glu	33.9 ± 10.2	24.5 ± 11.2	41.7 ± 15.1	0.0703	ns	$p>0.05$	$p>0.05$	$p>0.05$
Gln	930 ± 86.2	835 ± 113	1105 ± 236	0.0229	*	$p>0.05$	$p>0.05$	$\underline{p<0.05}$
Sarcosine	14.9 ± 1.7	14 ± 1.41	14.9 ± 4.37	0.8321	ns	$p>0.05$	$p>0.05$	$p>0.05$
Gly	247 ± 44.6	210 ± 54.3	229 ± 55.1	0.4686	ns	$p>0.05$	$p>0.05$	$p>0.05$
Ala	264 ± 42.8	228.4 ± 31	436 ± 221	0.0259	*	$p>0.05$	$p>0.05$	$\underline{p<0.05}$

Citrulline	56.8 ± 8.82	46.7 ± 4.09	64.1 ± 11	0.0084	**	p>0.05	p>0.05	p<0.01
Val	196 ± 17.2	174 ± 28.9	214 ± 46.4	0.1376	ns	p>0.05	p>0.05	p>0.05
Met	56.8 ± 4.16	69 ± 5.18	105 ± 36	0.0018	**	p>0.05	p<0.01	p<0.05
Cystathionine[C]	0.93 ± 0.40	2.92 ± 0.63	0.43 ± 0.09	<0.0001	***	p<0.001	p>0.05	p<0.001
Ile	78.9 ± 7.44	72.3 ± 16	84.4 ± 19.2	0.3766	ns	p>0.05	p>0.05	p>0.05
Leu	140 ± 11.7	130 ± 18.7	174 ± 56.7	0.0863	ns	p>0.05	p>0.05	p>0.05
Tyr	74.5 ± 8.81	88.6 ± 21.6	93.6 ± 15.4	0.0921	ns	p>0.05	p>0.05	p>0.05
Phe	61.5 ± 3.68	62.2 ± 5.38	73.6 ± 21.7	0.2028	ns	p>0.05	p>0.05	p>0.05
Ammonia	245 ± 72.2	255 ± 57.1	262 ± 97	0.9224	ns	p>0.05	p>0.05	p>0.05
Ornithine	65.8 ± 16	53 ± 5.05	94.5 ± 20.8	0.0006	***	p>0.05	p<0.01	p<0.001
Lys	234 ± 22.9	192 ± 27.4	224 ± 73.8	0.3039	ns	p>0.05	p>0.05	p>0.05
1-Methyl-Histidine	9.37 ± 2.40	6.86 ± 1.67	11.2 ± 5.35	0.1316	ns	p>0.05	p>0.05	p>0.05
His	63.4 ± 5.37	51.6 ± 5.97	66.5 ± 22	0.1612	ns	p>0.05	p>0.05	p>0.05
Arg	93.1 ± 21.2	76 ± 4.72	99.4 ± 26.8	0.1392	ns	p>0.05	p>0.05	p>0.05
Pro	70.6 ± 17.6	69.3 ± 15.2	127 ± 49.2	0.0053	**	p>0.05	p<0.05	p<0.05

Values are represented in terms of Mean ± SD. ND, not done; ns, not significant. [A]Not detected in two controls and one *Tg-I278T Cbs*$^{-/-}$ *mice*. [B]Done only in three controls and three *Tg-I278T Cbs*$^{-/-}$ mice, unpaired t test was done instead of ANOVA. [C]Not detected in four *Tg-I278T Cbs*$^{-/-}$ mice. *p<0.05; **p<0.01; ***p<0.001.

Supplementary Table 8
Metabolic profiling of liver

	nmoles/ g wet weight			One way ANOVA		Tukey's Multiple Comparison Test		
Metabolite	Control (n=8)	*Tg-hCBS Cbs*$^{-/-}$ (n=4)	*Tg-I278T Cbs*$^{-/-}$ (n=4)	p value	p value summary	Control vs *Tg-hCBS Cbs*$^{-/-}$	Control vs *Tg-I278T Cbs*$^{-/-}$	*Tg-hCBS Cbs*$^{-/-}$ vs *Tg-I278T Cbs*$^{-/-}$
Phosphoserine[A]	72.5 ± 10.76	74.8 ± 13.5	57.9 ± 0.81	0.1198	ns	p>0.05	p>0.05	p>0.05
Taurine	8467 ± 1772	8220 ± 583	7519 ± 1139	0.572	ns	p>0.05	p>0.05	p>0.05
Urea	18690 ± 7046	11490 ± 3572	10050 ± 8574	0.1019	ns	p>0.05	p>0.05	p>0.05
Asp	194 ± 41.5	334 ± 37.7	388 ± 94.3	0.0002	***	p<0.01	p<0.001	p>0.05
Thr	205 ± 46.2	240 ± 33	332 ± 69.4	0.0043	**	p>0.05	p<0.01	p>0.05
Ser	119 ± 29.8	194 ± 28.4	310 ± 32.83	<0.0001	***	p<0.01	p<0.001	p<0.001
Asn	238 ± 75	215 ± 13.1	282 ± 146	0.5646	ns	p>0.05	p>0.05	p>0.05
Glu	497 ± 111	622 ± 162	1953 ± 860	0.0003	***	p>0.05	p<0.001	p<0.01
Gln	3864 ± 819	3991 ± 991	3396 ± 585	0.5541	ns	p>0.05	p>0.05	p>0.05
Gly	1459 ± 322	1302 ± 137	1193 ± 421	0.395	ns	p>0.05	p>0.05	p>0.05
Ala	3301 ± 506	3138 ± 340	3399 ± 766	0.7942	ns	p>0.05	p>0.05	p>0.05
Val	229 ± 88	183 ± 22	246 ± 74.9	0.4858	ns	p>0.05	p>0.05	p>0.05
Met	66.9 ± 15	62.6 ± 15.9	115 ± 12	0.0002	***	p>0.05	p<0.001	p<0.001
Cystathionine	8.82 ± 2.86	1.12 ± 0.53	BD	<0.0001	***	p<0.001	p<0.001	p>0.05

Ile	135 ± 54.9	102 ± 11.3	129 ± 69.7	0.6025	ns	p>0.05	p>0.05	p>0.05
Leu	339 ± 81.3	288 ± 21.2	375 ± 113	0.3441	ns	p>0.05	p>0.05	p>0.05
Tyr	110 ± 31.6	111 ± 17.5	138 ± 66.9	0.4882	ns	p>0.05	p>0.05	p>0.05
β-Ala	126 ± 62.7	89.8 ± 14.4	186 ± 51	0.0643	ns	p>0.05	p>0.05	p>0.05
Phe	135 ± 30.3	120 ± 8.75	134 ± 37.9	0.6788	ns	p>0.05	p>0.05	p>0.05
Ammonia	990 ± 256	693 ± 127	805 ± 205	0.109	ns	p>0.05	p>0.05	p>0.05
Ornithine	145 ± 34	202 ± 60	308 ± 73.2	0.0008	***	p>0.05	p<0.001	p<0.05
Lys	325 ± 82.1	280 ± 16.8	363 ± 75.6	0.2933	ns	p>0.05	p>0.05	p>0.05
His	288 ± 38.4	312 ± 27	333 ± 92.7	0.4143	ns	p>0.05	p>0.05	p>0.05
Pro	172 ± 39	168 ± 39.1	300 ± 140	0.036	*	p>0.05	P<0.05	p>0.05
AdoHcy	88.3 ± 25.2	370 ± 130	789 ± 186	<0.0001	***	P<0.01	P<0.001	P<0.001

Values are represented in terms of Means ± SD. BD, below detection; ns, not significant. ^{A}Not detected in one *Tg-I278T Cbs*$^{-/-}$ mice. *p<0.05; **p<0.01; ***p<0.001.