Supplemental Table 1.

All values reported are fold changes. Data highlighted in green are significantly reduced and data highlighted in red are significantly increased (P<0.05). Values in white boxes indicate no significant differences between the two groups. 22%-22% fat diet. M42%-Modified 42% fat diet.

		DIET COMP BY GENOTYPE		GENOTYPE COMP BY DIET		
PATHWAY	**CHEMICAL**	**MPSI M42% / MPSI 22%**	**WT M42% / WT 22%**	**MPSI M42% / WT M42%**	**MPSI 22% / WT 22%**	**MPSI M42% / WT 22%**
Alanine and aspartate metabolism	alanine	1.5	1.8	1.5	1.2	1.2
	asparagine	1.4	1.5	2.6	2.4	1.8
	aspartate	1.7	1.6	1.9	1.9	1.2
	beta-alanine	1.6	1.2	1.9	1.0	1.6
Butanoate metabolism	2,4-diaminobutyrate	1.2	0.8	1.2	0.8	0.9
	2-aminobutyrate	1.0	1.6	1.3	2.0	2.1
Creatine metabolism	creatine	1.3	1.1	1.1	1.3	1.0
Cysteine, methionine, SAM, taurine metabolism	cysteine	2.7	2.4	2.2	2.6	1.1
	hypotaurine	2.0	1.5	1.8	1.4	2.8
	methionine	1.6	1.3	1.8	2.1	1.3
	S-adenosylhomocysteine (SAH)	1.2	1.5	2.7	1.4	1.7
	taurine	2.1	1.8	1.6	1.8	1.2
Glutamate metabolism	gamma-aminobutyrate (GABA)	2.1	3.3	1.0	1.5	3.2
	glutamate	1.4	2.0	1.6	1.1	1.3
	glutamine	1.4	3.5	1.2	2.8	4.1
	N-acetylglutamate	1.2	1.8	1.1	1.4	1.7
	N-acetylglutamine	1.0	0.9	1.2	1.1	1.1
	N-methylglutamate	1.4	1.3	0.9	0.8	1.2
Glutathione metabolism	cysteine-glutathione disulfide	10.7	2.0	3.1	1.7	6.3
	glutathione, oxidized (GSSG)	1.0	1.2	1.1	1.4	1.3
	glutathione, reduced (GSH)	2.9	2.7	3.1	2.8	8.3
	ophthalmate	1.2	1.9	1.2	1.3	1.6
	pyroglutamate (5-oxoproline)	1.4	1.2	1.1	1.3	1.1
	S-lactoylglutathione	2.0	1.6	1.3	1.0	2.0
	S-methylglutathione	1.4	1.1	1.2	1.4	1.0
Glycine, serine and threonine metabolism	betaine	1.1	1.2	1.1	1.2	1.3
	betaine aldehyde	1.0	2.8	2.1	6.0	6.0
	butyrylglycine	1.6	1.1	1.4	1.1	1.5
	glycine	1.4	1.5	1.7	1.5	1.1
	hexanoylglycine	1.4	1.2	1.0	1.2	1.2
	serine	1.4	1.4	2.2	2.1	1.5
	threonine	1.6	1.2	1.7	2.1	1.4
Histidine metabolism	histidine	1.5	1.2	1.1	1.9	1.3
Lysine metabolism	2-aminoadipate	2.0	2.7	1.7	2.2	4.5
	lysine	1.3	1.7	2.0	1.5	1.2
	pipecolate	1.0	1.1	1.1	1.1	1.2

Phenylalanine & tyrosine metabolism	3-(4-hydroxyphenyl)lactate	2.2	1.9	2.0	1.7	3.7
	p-cresol sulfate	1.2	1.6	0.8	1.1	1.3
	phenylacetylglycine	0.9	0.8	0.9	0.8	0.7
	phenylalanine	1.3	1.3	1.8	1.8	1.4
	tyrosine	1.4	1.6	1.9	1.8	1.2
Polyamine metabolism	5-methylthioadenosine (MTA)	1.4	1.9	1.3	1.0	1.4
	spermidine	1.0	1.3	1.3	1.6	1.7
Tryptophan metabolism	3-indoxyl sulfate	0.8	0.8	1.0	1.0	0.8
	anthranilate	1.5	1.9	1.6	2.0	3.0
	kynurenine	2.3	1.5	1.1	1.3	1.7
	tryptophan	1.4	1.3	2.2	2.5	1.7
Urea cycle; arginine-, proline-, metabolism	arginine	1.0	1.1	1.2	1.3	1.3
	citrulline	1.4	1.2	1.1	1.8	1.3
	dimethylarginine	1.5	1.4	1.8	1.8	1.2
	ornithine	1.4	1.2	1.3	1.5	1.1
	proline	1.9	2.7	2.7	1.9	1.0
	urea	1.4	1.1	1.1	1.3	1.1
Valine, leucine and isoleucine metabolism	isoleucine	1.3	1.4	1.8	1.8	1.3
	leucine	1.2	1.2	1.4	1.4	1.1
	N-acetylalanine	1.3	1.2	1.5	1.6	1.3
	valine	1.6	1.4	1.7	2.0	1.3
Aminosugars metabolism	N-acetylneuraminate	0.7	0.8	1.1	1.3	0.9
Fructose, mannose, galactose, starch, and sucrose metabolism	fructose	1.3	2.6	1.3	2.5	2.0
	maltohexose	45.5	1.3	17.4	2.0	23.0
	maltose	1.8	2.8	4.4	1.2	1.5
	maltotetraose	5.1	1.8	6.3	1.4	3.5
	maltotriose	4.7	1.8	7.2	1.2	3.9
	mannitol	1.8	3.1	1.5	2.6	4.6
	mannose	1.4	2.4	1.9	1.6	1.2
	mannose-6-phosphate	3.7	1.4	2.3	1.1	3.3
Glycolysis, gluconeogenesis, pyruvate metabolism	1,5-anhydroglucitol (1,5-AG)	3.1	2.4	1.4	1.0	3.3
	3-phosphoglycerate	1.6	1.8	1.0	1.1	1.7
	fructose-6-phosphate	3.7	2.0	2.0	1.1	4.0
	glucose	1.0	2.0	1.3	1.5	1.5
	glucose-6-phosphate (G6P)	4.5	2.1	2.5	1.2	5.2
	glucuronate	1.4	1.3	1.4	1.3	1.1
	lactate	1.5	2.4	1.1	1.3	2.1
	phosphoenolpyruvate (PEP)	1.6	1.6	1.1	1.0	1.5
Nucleotide sugars, pentose metabolism	6-phosphogluconate	0.8	1.0	1.0	1.2	1.0
	gluconate	1.7	2.1	1.7	2.1	3.5
	maltopentaose	17.3	1.3	8.1	2.8	6.1
	ribitol (adonitol)	2.3	2.0	2.9	2.6	6.0
	ribonate	1.1	1.6	1.1	1.7	1.8
	ribose	1.5	1.8	1.1	1.3	1.9
	ribose 5-phosphate	1.0	1.2	1.7	1.5	1.5
	UDP-glucose	1.9	2.2	1.7	2.5	1.3
	xylitol	1.1	1.6	1.4	1.3	1.1
Ascorbate and aldarate metabolism	ascorbate (Vitamin C)	2.2	2.1	2.3	2.2	4.9
	gulono-1,4-lactone	1.6	1.3	2.2	1.1	1.7
	threonate	2.1	2.3	1.3	1.2	1.8
Folate metabolism	5-methyltetrahydrofolate (5MeTHF)	1.5	1.5	1.9	2.0	1.3

	biopterin	0.8	0.6	1.0	0.8	0.6
	dihydrobiopterin	1.1	0.7	1.8	1.1	1.3
Hemoglobin and porphyrin metabolism	heme*	1.5	42.2	3.0	9.6	14.2
	biliverdin	1.4	3.9	1.2	2.3	3.3
Nicotinate and nicotinamide metabolism	nicotinamide	1.1	1.3	1.3	1.5	1.7
	nicotinamide adenine dinucleotide (NAD+)	1.6	1.0	2.2	1.5	2.3
Pantothenate and CoA metabolism	3'-dephosphocoenzyme A	1.4	3.0	1.6	3.5	4.9
	pantothenate	1.7	1.7	1.3	1.3	2.2
	phosphopantetheine	1.6	4.0	1.3	2.0	3.1
Riboflavin metabolism	flavin adenine dinucleotide (FAD+)	1.2	1.0	1.5	1.2	1.4
	riboflavin (Vitamin B2)	1.2	1.7	1.1	1.3	1.5
Thiamine metabolism	thiamin (Vitamin B1)	1.5	1.0	2.4	1.6	2.4
Tocopherol metabolism	alpha-tocopherol	2.4	2.3	1.1	1.0	2.5
Vitamin B6 metabolism	pyridoxate	1.6	2.6	1.9	1.2	1.3
Krebs cycle	cis-aconitate	1.8	1.1	1.7	1.1	1.9
	citrate	2.9	1.9	2.1	1.4	4.0
	fumarate	1.2	1.3	1.1	1.1	1.3
	malate	1.4	1.4	1.1	1.1	1.5
	oxaloacetate	0.8	1.1	1.2	1.5	1.3
Oxidative phosphorylation	acetylphosphate	0.9	1.0	1.1	1.1	1.1
	phosphate	0.9	1.0	0.9	1.0	0.9
Bile acid metabolism	12-dehydrocholate	0.7	0.4	0.8	0.5	0.3
	alpha-muricholate	1.1	1.4	1.9	1.2	1.3
	beta-muricholate	1.0	2.7	2.1	1.2	1.3
	cholate	5.7	1.7	3.3	1.0	5.7
	deoxycholate	1.5	1.6	1.1	1.1	1.4
	tauro-beta-muricholate	3.9	1.0	3.1	1.2	3.2
	taurocholate	4.1	1.3	2.8	1.1	3.7
	taurodeoxycholate	1.4	1.8	1.4	1.1	1.3
	ursodeoxycholate	1.3	1.7	1.3	1.0	1.4
Carnitine metabolism	2-methylbutyroylcarnitine	1.0	0.7	1.4	0.9	0.9
	3-dehydrocarnitine	1.5	1.9	1.3	1.0	1.5
	acetylcarnitine	2.8	1.1	11.6	3.7	10.2
	carnitine	1.0	0.8	1.0	0.9	0.8
	glutaroyl carnitine	1.8	1.3	1.4	1.0	1.8
	isobutyrylcarnitine	1.3	1.0	2.2	1.6	2.1
	isovalerylcarnitine	1.4	2.0	1.1	1.3	1.8
	propionylcarnitine	1.5	1.2	1.1	2.1	1.3
Fatty acid, amide	oleamide	1.8	4.2	1.2	2.0	3.6
Fatty acid, monoene	5-dodecenoate (12:1n7)	1.1	1.4	1.6	1.1	1.2
	docosatrienoate (22:3n3)	7.2	5.5	1.2	1.1	6.5
	eicosenoate (20:1n9 or 11)	1.5	1.3	0.8	0.7	1.1
	myristoleate (14:1n5)	1.1	1.3	1.5	1.2	1.1
	oleate (18:1n9)	1.3	1.1	1.2	1.4	1.1
	palmitoleate (16:1n7)	1.5	1.1	1.3	1.8	1.2
Fatty acid, monoene, odd	10-heptadecenoate (17:1n7)	2.1	1.9	1.2	1.3	1.6
	10-nonadecenoate (19:1n9)	2.5	2.0	1.1	1.2	2.1
Fatty acid, polyene	adrenate (22:4n6)	0.8	1.2	0.9	1.3	1.1
	arachidonate (20:4n6)	1.2	1.1	1.9	1.5	1.8
	dihomo-alpha-linolenate (20:3n3)	1.6	1.1	2.5	1.4	2.2

	dihomo-linolenate (20:2n6)	0.7	0.9	0.7	0.9	0.6
	docosadienoate (22:2n6)	0.8	0.7	1.1	1.0	0.8
	docosahexaenoate (DHA; 22:6n3)	3.1	1.9	1.7	1.0	3.1
	docosapentaenoate (n3 DPA; 22:5n3)	2.2	2.5	1.4	1.2	1.9
	eicosapentaenoate (EPA; 20:5n3)	6.1	4.0	2.2	1.4	8.8
	linoleate (18:2n6)	1.5	1.5	1.1	1.1	1.6
	linolenate [alpha or gamma; (18:3n3 or 6)]	1.5	2.0	1.1	1.2	1.7
	stearidonate (18:4n3)	2.3	4.1	1.3	1.4	3.2
Fatty acid, saturated, even	arachidate (20:0)	1.0	2.0	1.7	1.1	1.2
	caprate (10:0)	1.2	1.2	1.7	1.1	1.4
	caproate (6:0)	0.8	1.0	0.8	1.0	0.8
	caprylate (8:0)	1.2	1.0	1.7	1.4	1.7
	laurate (12:0)	1.2	1.1	1.4	1.0	1.2
	myristate (14:0)	1.4	2.1	1.3	1.1	1.6
	palmitate (16:0)	1.1	1.2	1.1	1.2	1.3
Fatty acid, saturated, even	stearate (18:0)	1.0	1.2	1.2	1.5	1.5
Fatty acid, saturated, odd	heptanoate (7:0)	0.7	1.1	0.6	1.0	0.7
	margarate (17:0)	1.0	2.0	1.2	1.6	1.6
	nonadecanoate (19:0)	1.5	2.4	1.1	1.5	2.2
	pelargonate (9:0)	1.3	1.1	1.6	1.1	1.4
	pentadecanoate (15:0)	1.5	1.2	1.3	1.1	1.6
Glycerolipid metabolism	1-arachidonoylglycerophosphocholine	1.0	2.0	8.3	4.2	4.2
	1-linoleoylglycerophosphocholine	1.9	4.5	5.6	2.4	1.2
	1-oleoylglycerophosphocholine	2.0	4.5	3.4	1.5	1.3
	1-palmitoylglycerophosphocholine	0.8	2.1	0.4	1.1	0.9
	1-stearoylglycerophosphocholine	1.0	2.2	0.5	1.1	1.1
	1-stearoylglycerophosphoethanolamine	1.1	1.6	0.6	0.8	0.9
	2-arachidonoylglycerophosphocholine	0.8	3.7	0.5	2.4	2.0
	2-linoleoylglycerophosphocholine	0.8	1.8	0.5	1.1	0.9
	2-oleoylglycerophosphocholine	2.2	5.3	1.7	1.4	3.1
	2-palmitoylglycerophosphocholine	1.3	5.1	3.3	1.1	1.5
	2-stearoylglycerophosphocholine	1.2	2.9	0.5	1.1	1.3
	choline	1.4	1.5	1.2	1.1	1.3
	choline phosphate	1.2	1.1	1.4	1.3	1.5
	glycerol	1.0	2.0	1.3	1.4	1.5
	glycerol 3-phosphate (G3P)	1.1	1.4	3.0	2.1	2.2
	glycerophosphorylcholine (GPC)	1.1	1.6	3.0	1.8	1.9
	monoethanolamine	1.1	1.5	1.5	2.0	2.1
	o-phosphoethanolamine	1.1	1.2	1.2	1.3	1.4
Inositol metabolism	inositol 1-phosphate (I1P)	1.1	1.4	1.2	1.5	1.6
	myo-inositol	2.0	1.5	1.1	1.4	1.4
Ketone bodies	3-hydroxybutyrate (BHBA)	1.1	1.7	1.7	1.2	1.0
Monoacylglycerol	1-oleoylglycerol (1-monoolein)	4.6	1.4	3.6	1.1	5.0
	1-palmitoylglycerol (1-monopalmitin)	1.5	1.6	1.9	1.2	1.2
	1-stearoylglycerol (1-monostearin)	1.2	2.0	1.1	1.5	1.8
Monoglycerol	myristoylglycerol (monomyristin)	1.2	1.2	0.9	1.0	1.1
Phospholipid metabolism	cytidine 5'-diphosphocholine	0.9	1.8	0.7	1.5	1.3
Sphingolipid	sphinganine	1.7	8.8	1.1	5.5	9.6

	sphingosine	1.3	7.8	1.3	4.6	6.0
Sterol/Steroid	campesterol	1.8	2.8	5.3	1.0	1.9
	cholesterol	1.0	1.1	1.3	1.4	1.4
	corticosterone	1.1	1.4	0.8	1.1	1.2
Purine metabolism, (hypo)xanthine/inosine containing	hypoxanthine	1.1	1.0	1.4	1.3	1.3
	inosine	1.1	1.2	1.5	1.4	1.3
	inosine 5'-monophosphate (IMP)	1.0	2.0	2.0	1.0	1.0
	xanthine	1.2	1.2	1.2	1.2	1.5
	xanthosine	1.7	2.7	1.1	1.8	2.9
Purine metabolism, adenine containing	1-methyladenosine	1.2	0.8	1.6	1.0	1.2
	adenine	1.1	1.4	1.4	1.8	1.9
	adenosine	1.0	1.2	2.8	2.4	2.3
	adenosine 3',5'-diphosphate	1.0	3.4	4.4	1.2	1.3
	adenosine 3'-monophosphate	1.4	2.4	1.8	1.8	1.3
	adenosine 5'-diphosphate (ADP)	1.9	1.2	2.1	1.1	1.7
	adenosine 5'-monophosphate (AMP)	1.2	1.8	4.3	1.9	2.4
	adenylosuccinate	0.7	0.9	0.9	1.2	0.9
Purine metabolism, guanine containing	guanosine	1.4	1.9	5.2	3.8	2.8
	guanosine 5'- monophosphate (GMP)	2.4	6.9	9.2	3.2	1.3
Purine metabolism, urate metabolism	allantoin	1.6	1.8	1.2	1.3	2.1
	urate	2.5	6.3	1.1	2.7	6.8
Pyrimidine metabolism, cytidine containing	cytidine	1.1	1.2	1.5	1.9	1.7
	cytidine 5'-monophosphate	1.4	1.6	3.4	1.5	2.1
Pyrimidine metabolism, uracil containing	3-methyluridine	1.1	2.0	1.0	1.9	2.0
	uracil	1.3	1.1	1.4	1.2	1.6
	uridine	1.1	1.6	1.2	1.6	1.9
	uridine 5'-monophosphate (UMP)	2.1	4.1	1.6	1.2	2.6
Dipeptide	aspartylphenylalanine	1.6	1.1	2.6	1.8	2.8
	glutamylglutamate	1.3	1.0	2.2	2.8	2.2
	glutamylvaline	1.1	0.9	0.9	0.8	0.8
	glycylleucine	1.8	1.3	5.2	6.9	3.9
	glycylproline	2.1	1.0	1.2	2.6	1.2
	glycylvaline	1.4	1.3	4.5	4.9	3.6
	isoleucinylproline	2.5	1.4	2.4	4.2	1.7
g-glutamyl	gamma-glutamylisoleucine	1.0	1.1	1.0	1.2	1.2
	gamma-glutamylleucine	0.9	1.1	1.0	1.1	1.0
	gamma-glutamylphenylalanine	1.1	1.1	1.3	1.4	1.3
Chemical	glycerol 2-phosphate	1.4	1.9	1.0	1.3	1.9
	phenol sulfate	3.1	2.4	1.1	1.5	2.2
Drug	hydroquinone	1.2	1.2	1.6	1.0	1.3