

Table S1. Metabolite Levels in MUT Tissue Relative to WT- 2 Weeks of Age

Metabolite	Retina- Fold Change		Kidney- Fold Change		Eyecup- Fold Change	
	Mean	SEM	Mean	SEM	Mean	SEM
1-Methyladenosine	0.361	0.102	0.745	0.069	0.990	0.267
2-Hydroxyglutarate/Mevalonate	1.025	0.115	0.821	0.204	0.823	0.072
2-Methylbutyroylcarnitine	2.778	0.816	1.184	0.176	0.769	0.086
3-Aminoisobutanoic acid	1.791	0.488	1.051	0.064	1.154	0.188
3PG	0.908	0.163	2.165	0.746	0.945	0.104
4-Hydroxyproline	0.901	0.149	1.046	0.119	0.876	0.138
5-Oxoproline	0.920	0.062	0.998	0.113	0.881	0.077
α-Ketoglutarate	1.036	0.332	1.088	0.092	0.773	0.115
Acetyl CoA	0.325	0.155	0.547	0.124	0.681	0.194
Acetyl L Carnitine	1.398	0.233	1.120	0.051	1.123	0.018
AcetylCholine	2.431	0.773	0.905	0.124	0.795	0.222
Aconitate	0.601	0.232	0.717	0.219	0.699	0.128
Adenine	2.154	0.917	0.751	0.132	1.254	0.143
Adenosine	7.295	2.996	0.858	0.157	0.838	0.154
ADP	1.826	0.888	0.996	0.124	0.793	0.192
AICAR	0.401	0.198	1.206	0.200	1.039	0.139
Alanine	2.682	0.819	1.019	0.084	0.922	0.092
Alanine	0.998	0.080	1.135	0.092	1.136	0.099
Amino adipic acid	0.580	0.302	1.043	0.134	1.084	0.178
Arginosuccinate	1.743	0.398	1.309	0.197	1.532	0.173
Ascorbic acid	0.261	0.026	1.135	0.149	0.819	0.067
Asparagene	0.985	0.113	1.096	0.217	1.010	0.083
Aspartate	1.019	0.076	1.110	0.053	0.909	0.097
Aspartic acid	2.694	0.793	1.159	0.052	0.922	0.099
Betaine	2.227	0.679	1.023	0.008	0.962	0.081
Biotin	0.875	0.064	1.377	0.235	0.823	0.088
Butyrylcarnitine	1.572	0.510	0.928	0.130	0.855	0.109
cAMP	1.585	0.434	1.129	0.159	0.948	0.203
Carnitine	1.689	0.482	1.128	0.040	0.815	0.060
Carnosine	3.056	0.914	1.028	0.232	0.634	0.191
Cholesterol	0.812	0.110	0.882	0.211	0.510	0.109
Choline	2.383	0.572	1.018	0.037	1.010	0.077
Cis-aconitic acid	0.818	0.090	0.912	0.328	0.853	0.110
Citraconic Acid	0.679	0.321	0.747	0.196	0.765	0.165
Citrate	0.939	0.087	0.837	0.275	0.891	0.083
Citrulline	0.508	0.220	1.022	0.082	0.967	0.087
Creatine	1.506	0.308	0.956	0.038	1.007	0.091
Creatinine	0.886	0.066	1.075	0.059	0.753	0.130
Cyclic ADP-ribose	0.515	0.234	1.260	0.211	0.687	0.198
Cystathionine	1.113	0.186	1.436	0.284	0.735	0.182
Cystine	1.920	1.127	0.969	0.106	0.976	0.084
Cytidine	0.903	0.146	0.538	0.186	1.005	0.120
D-Ribulose 5-phosphate	0.613	0.223	1.876	1.040	1.052	0.207
Erythritol	0.610	0.152	1.105	0.040	0.879	0.055
Fumarate	0.956	0.105	1.115	0.072	0.834	0.089
Gamma-Aminobutyric acid (GABA)	1.040	0.101	1.209	0.134	0.785	0.142
Gamma-Aminobutyric acid (GABA)	1.738	0.344	1.191	0.144	2.084	1.222
GDP	0.879	0.210	0.854	0.110	1.271	0.330
Glucose	-	-	0.914	0.152	0.801	0.081
Glucose 1-phosphate	1.038	0.137	1.035	0.218	0.748	0.142

Glucose 6-phosphate	1.178	0.207	0.999	0.238	0.740	0.107
Glutamate	0.972	0.071	1.055	0.041	0.882	0.103
Glutamic acid	2.463	0.687	0.905	0.043	0.855	0.182
Glutamine	2.342	0.635	22.229	18.173	0.903	0.100
Glutamine	1.145	0.129	1.454	0.571	0.886	0.082
Glutathione	0.635	0.254	0.649	0.217	-	-
Glycerate	0.909	0.422	0.425	0.340	0.653	0.186
Glycine	1.818	0.508	1.066	0.030	0.807	0.086
Glycine	0.947	0.055	1.075	0.058	0.913	0.076
GMP	0.764	0.155	1.293	0.198	0.887	0.162
Guanine	1.516	0.565	0.885	0.117	1.350	0.185
Guanosine	0.895	0.254	0.749	0.142	1.144	0.238
Guanosine 5'-phosphate	0.653	0.202	0.837	0.112	0.895	0.185
Hexanoylcarnitine	0.887	0.224	0.799	0.126	0.976	0.110
Histidine	0.798	0.112	1.047	0.104	1.040	0.244
Histidine	1.009	0.177	1.095	0.122	1.033	0.217
Hypotaurine	0.529	0.251	1.140	0.236	0.750	0.502
Hypothanxine	6.794	3.111	1.746	0.636	1.498	0.648
Hypoxanthine	0.848	0.218	1.598	0.520	1.503	0.505
IMP	2.399	0.706	1.134	0.145	0.820	0.113
Inosine	11.666	5.160	0.913	0.053	1.058	0.155
Inosine	10.705	5.945	0.944	0.440	1.243	0.484
IPP	1.098	0.163	1.154	0.104	1.177	0.171
Isobutyrylcarnitine	1.824	0.484	0.916	0.131	0.876	0.105
Isocitrate	0.725	0.117	1.030	0.194	1.346	0.129
Isoleucine	0.937	0.090	1.191	0.092	0.894	0.122
L-Arginine	0.702	0.072	1.345	0.148	1.088	0.102
L-Asparagine	0.387	0.190	1.121	0.151	1.066	0.087
L-Homoserine	0.926	0.090	1.050	0.041	0.918	0.136
Lactate	2.390	0.639	1.131	0.035	1.160	0.090
Lactate	1.226	0.088	1.279	0.046	1.061	0.090
Leucine	2.192	0.680	1.179	0.061	1.049	0.131
Leucine	1.118	0.147	1.315	0.130	1.079	0.120
Lysine	1.568	0.420	1.183	0.095	0.785	0.123
Lysine	0.875	0.097	1.199	0.109	0.800	0.137
Malate	2.310	0.643	1.151	0.037	0.932	0.061
Malate	1.020	0.109	1.354	0.164	0.879	0.117
Maleic Acid	1.023	0.010	1.020	0.033	0.996	0.010
Methionine	2.495	0.798	1.048	0.188	1.035	0.129
Methionine	1.005	0.133	1.166	0.090	0.981	0.123
Myo inositol	2.399	0.711	1.224	0.112	1.114	0.125
Myristic acid-D27	0.997	0.045	1.106	0.054	0.956	0.035
Myristoylcarnitine	1.599	0.503	0.714	0.188	0.768	0.105
N-alpha-Acetyl-L-lysine	1.737	0.549	1.206	0.123	1.103	0.176
N-Asp	0.906	0.108	-	-	0.983	0.266
Ornithine	0.984	0.075	1.145	0.162	0.905	0.108
Oxalic acid	0.554	0.114	0.961	0.077	1.133	0.044
Oxalic acid	0.853	0.076	0.940	0.100	0.945	0.084
Oxidized glutathione	1.432	0.511	1.736	0.579	0.820	0.119
Palmitate	1.075	0.052	1.025	0.017	0.939	0.020
Palmitoyl CoA	0.377	0.130	1.069	0.193	0.873	0.204
Palmitoylcarnitine	1.687	0.142	0.758	0.144	0.808	0.092
Pantothenic acid	0.462	0.125	1.138	0.075	1.165	0.173

Pantothenic acid	1.007	0.125	1.270	0.096	0.734	0.110
PEP	0.866	0.328	2.169	0.687	0.815	0.114
Phenylalanine	0.933	0.123	1.173	0.123	1.003	0.078
Phenylalanine	1.974	0.660	1.037	0.114	1.046	0.090
Phosphocreatine	1.430	0.925	0.601	0.112	0.892	0.307
Propionylcarnitine	3.172	0.894	1.199	0.108	0.797	0.113
Pyroglutamic acid	1.903	0.651	1.140	0.080	0.957	0.171
Pyruvate	1.590	0.644	0.821	0.082	0.902	0.062
Riboflavin	0.359	0.179	0.837	0.169	0.937	0.128
Serine	1.168	0.067	0.927	0.112	0.915	0.096
Serine	0.970	0.086	1.094	0.062	0.839	0.090
Sphinganine	0.720	0.067	0.695	0.127	0.891	0.076
Stearic acid	1.077	0.039	1.031	0.017	0.986	0.017
Stearoylcarnitine	1.483	1.041	0.923	0.159	0.703	0.096
Succinate	1.843	0.265	1.049	0.042	0.905	0.060
Succinate	0.984	0.136	1.197	0.134	0.874	0.089
Succinyl CoEnzyme A	0.360	0.176	0.898	0.255	1.281	0.262
Taurine	1.177	0.118	0.996	0.052	1.017	0.133
Taurine	0.972	0.051	1.143	0.117	1.105	0.104
Thiamine	0.676	0.325	0.954	0.227	0.966	0.307
Thiamine Monophosphate	0.586	0.294	0.979	0.060	0.928	0.195
Thiamine Pyrophosphate	0.379	0.186	0.931	0.149	1.307	0.235
Threonine	0.480	0.213	0.961	0.095	0.942	0.152
Threonine	0.864	0.128	1.114	0.063	0.741	0.123
Trigonelline	1.373	0.348	1.833	0.618	1.516	0.525
Tryptophan	0.491	0.218	1.008	0.116	1.039	0.244
Tryptophan	0.945	0.129	1.069	0.086	0.958	0.139
Tyrosine	0.522	0.229	0.937	0.083	1.140	0.144
Tyrosine	1.032	0.139	1.257	0.111	1.150	0.105
UDP	1.458	0.691	0.962	0.125	0.865	0.232
UDP Glucosame	0.618	0.289	1.064	0.067	0.814	0.126
UDP-Glucose	0.447	0.220	0.965	0.106	0.998	0.181
Uracil	0.761	0.321	1.073	0.082	1.523	0.214
Uracil	1.025	0.063	1.044	0.151	0.768	0.102
Urate	1.222	0.223	1.367	0.302	1.260	0.498
Urea	1.020	0.107	0.826	0.123	0.869	0.158
Valine	1.241	0.133	1.042	0.075	1.060	0.146
Valine	1.014	0.123	1.287	0.167	0.954	0.155
Xanthine	0.381	0.155	1.173	0.239	1.193	0.229
Xanthine	3.943	1.199	1.270	0.460	0.948	0.232
XMP	1.848	0.389	1.287	0.206	0.774	0.103

Red bold indicates statistical significance $P < 0.05$. - indicates that measurable amounts of that metabolite were not detected. Repeated metabolites represent measurements by both LC/MS and GC/MS.

Table S2. Metabolite Levels in MUT Tissue Relative to WT- 3 Weeks of Age

Metabolite	Retina- Fold Change		Kidney- Fold Change		Eyecup- Fold Change	
	Mean	SEM	Mean	SEM	Mean	SEM
1-Methyladenosine	0.776	0.053	0.820	0.152	0.912	0.088
2-Hydroxyglutarate/Mevalonate	0.916	0.042	0.726	0.052	0.905	0.063
2-Methylbutyroylcarnitine	2.457	0.489	1.013	0.126	1.115	0.174
3-Aminoisobutanoic acid	0.981	0.025	0.947	0.022	1.061	0.089
3PG	0.992	0.097	2.713	1.435	1.677	0.478
5-Oxoproline	1.143	0.048	0.588	0.053	0.823	0.108
α -Ketoglutarate	1.112	0.088	-	-	0.930	0.035
α -Ketoglutarate	1.633	0.219	1.290	0.520	-	-
Acetyl L Carnitine	1.040	0.050	0.947	0.020	1.000	0.073
Acetyl-CoA	1.011	0.236	0.946	0.227	0.737	0.158
Aconitate	1.376	0.122	0.929	0.127	1.053	0.133
Adenine	0.976	0.031	0.956	0.079	1.087	0.085
Adenosine	1.114	0.072	1.437	0.161	1.183	0.142
ADP	1.259	0.127	1.017	0.181	0.911	0.114
AICAR	-	-	0.938	0.054	-	-
Alanine	1.077	0.074	0.892	0.078	0.977	0.058
Alanine	1.047	0.065	1.090	0.127	1.014	0.025
Amino adipic acid	0.922	0.146	0.456	0.096	0.824	0.046
Ammonia	1.524	0.292	1.454	0.054	0.995	0.411
AMP	0.984	0.074	1.173	0.118	1.535	0.185
Arginosuccinate	1.387	0.186	0.779	0.096	0.985	0.137
Asparagine	0.965	0.071	1.182	0.227	0.800	0.228
Aspartate	1.122	0.107	0.864	0.072	0.969	0.086
Aspartic acid	1.068	0.084	1.053	0.215	0.894	0.056
ATP	1.604	0.261	1.053	0.215	0.433	0.075
Beta-Alanine	1.035	0.037	0.622	0.094	0.689	0.185
Betaine	0.909	0.031	0.913	0.035	1.002	0.068
Biotin	1.150	0.111	1.029	0.039	1.253	0.294
Butyrylcarnitine	0.978	0.079	0.897	0.119	0.975	0.103
cAMP	1.181	0.117	-	-	1.166	0.129
Carnosine	1.035	0.098	0.807	0.064	0.723	0.086
Cholesterol	1.191	0.219	0.677	0.048	3.694	1.060
Cis-aconitic acid	1.221	0.161	0.929	0.163	0.794	0.157
Citrate	1.209	0.084	1.242	0.343	0.897	0.099
Citrate	1.225	0.127	1.029	0.398	0.964	0.150
Citrulline	0.932	0.083	0.785	0.104	1.038	0.104
Coenzyme A	-	-	1.321	0.300	-	-
Creatine	1.021	0.043	0.864	0.114	1.064	0.081
Creatinine	0.989	0.084	1.287	0.190	0.877	0.068
Cystathionine	1.010	0.087	0.797	0.094	0.516	0.173
Cystathionine	1.036	0.145	2.084	0.261	0.983	0.101
Cysteine	1.071	0.111	0.916	0.124	0.849	0.310
Cystine	-	-	0.677	0.078	-	-
Cytidine	0.931	0.087	0.585	0.311	0.929	0.108
D-2-Hydroxyglutaric acid	1.021	0.047	0.861	0.091	-	-
D-Erythrose 4-phosphate	0.961	0.052	0.881	0.080	1.221	0.071
D-Ribulose 5-phosphate	1.073	0.047	0.597	0.090	1.461	0.165
Decanoylcarnitine	1.075	0.167	1.083	0.216	1.002	0.116
Erythritol	0.989	0.050	0.887	0.057	0.979	0.098
FMN	-	-	0.692	0.038	-	-

Fructose 1,6-bisphosphate	1.071	0.085	-	-	-	-
Fumarate	1.169	0.057	1.805	0.285	0.987	0.116
GABA	0.948	0.055	1.092	0.120	0.865	0.426
Gamma-Aminobutyric acid	0.973	0.031	0.950	0.069	1.052	0.228
Glucose	-	-	0.871	0.115	1.027	0.050
Glucose 1-phosphate	1.015	0.051	1.147	0.136	1.137	0.053
Glucose 6-phosphate	0.994	0.054	1.180	0.133	1.116	0.050
Glutamate	0.998	0.046	1.039	0.042	0.953	0.110
Glutamic acid	1.011	0.034	0.899	0.024	0.858	0.040
Glutathione	1.600	0.325	1.126	0.656	-	-
Glyceraldehyde 3-phosphate	0.965	0.050	0.829	0.157	0.823	0.129
Glycine	1.080	0.070	0.881	0.033	1.024	0.117
Glycine	1.071	0.069	0.979	0.051	0.978	0.047
GMP	0.911	0.045	1.628	0.092	1.280	0.174
Guanine	1.060	0.083	0.883	0.060	0.856	0.082
Guanosine	1.066	0.088	0.910	0.081	1.178	0.127
Guanosine 5'-phosphate	1.669	0.292	0.852	0.169	0.534	0.074
Heptadecanoic acid	1.026	0.082	0.679	0.050	1.154	0.121
Hexanoylcarnitine	0.957	0.175	0.935	0.127	1.231	0.162
Hippurate	-	-	0.579	0.207	-	-
Histamine	-	-	0.680	0.250	0.647	0.082
Histidine	1.243	0.092	0.753	0.040	0.562	0.191
Histidine	1.041	0.086	1.876	0.146	0.636	0.045
Hypotaurine	0.915	0.132	0.803	0.059	-	-
Hypothanxine	0.998	0.067	0.783	0.057	1.112	0.262
Hypoxanthine	0.938	0.078	0.907	0.067	1.198	0.114
IMP	0.968	0.059	1.418	0.113	1.362	0.166
Inosine	1.255	0.151	0.843	0.049	1.710	0.247
Isobutyrylcarnitine	1.003	0.057	0.908	0.112	0.947	0.090
Isocitrate	1.211	0.110	-	-	0.700	0.082
Isoleucine	0.902	0.031	0.882	0.074	0.879	0.114
Isopentenyl pyrophosphate	0.922	0.082	1.045	0.245	1.069	0.054
L-Arginine	1.024	0.102	0.725	0.054	0.633	0.093
L-Asparagine	0.866	0.054	0.755	0.064	0.626	0.091
L-Homoserine	1.039	0.080	0.793	0.072	0.869	0.103
Lactate	1.039	0.017	1.067	0.041	0.878	0.075
Lactate	1.011	0.024	1.051	0.041	1.299	0.147
Leucine	0.964	0.064	0.919	0.051	0.918	0.128
Leucine	1.015	0.065	0.945	0.073	0.900	0.114
Lysine	1.188	0.046	0.752	0.025	0.548	0.181
Lysine	0.976	0.131	1.352	0.087	0.689	0.144
Malate	1.069	0.024	1.526	0.401	0.987	0.119
Malate	1.130	0.061	1.951	0.699	1.102	0.085
Methionine	0.957	0.086	0.731	0.074	0.636	0.056
Methionine	0.924	0.076	0.821	0.103	0.594	0.075
Myo inositol	1.201	0.133	1.100	0.035	1.012	0.071
Myristic acid-D27	0.877	0.040	0.819	0.066	1.237	0.142
Myristoylcarnitine	0.887	0.099	0.649	0.206	0.821	0.088
N-alpha-Acetyl-L-lysine	1.009	0.125	0.758	0.144	0.751	0.085
N-Asp	0.965	0.093	2.685	0.362	0.628	0.265
Ornithine	0.746	0.045	0.956	0.123	0.640	0.216
Ornithine	1.052	0.125	1.103	0.124	-	-
Oxalic acid	1.039	0.078	1.064	0.060	1.221	0.201

Oxalic acid	0.758	0.108	0.593	0.062	1.090	0.188
Oxidized glutathione	1.105	0.129	1.871	0.327	0.691	0.048
Palmitate	0.993	0.035	1.024	0.036	1.015	0.069
Palmitoylcarnitine	1.060	0.125	0.540	0.111	0.934	0.125
Palmitoyl-CoA	1.769	0.414	0.812	0.127	1.394	0.319
Pantothenic acid	0.960	0.092	1.060	0.141	0.810	0.132
Pantothenic acid	0.962	0.095	0.851	0.215	1.104	0.163
PEP	1.511	0.363	3.943	2.057	0.993	0.243
Phenylalanine	0.986	0.054	0.909	0.072	0.970	0.063
Phenylalanine	1.013	0.057	0.786	0.043	0.803	0.052
Phosphocreatine	1.102	0.078	0.897	0.214	0.987	0.158
Phosphoserine	-	-	1.247	0.116	-	-
Propionylcarnitine	1.044	0.141	0.821	0.067	0.804	0.127
Pyroglutamic acid	1.153	0.106	0.751	0.096	1.022	0.102
Pyruvate	1.698	0.196	0.592	0.140	1.016	0.080
Riboflavin	0.831	0.062	0.670	0.078	0.551	0.066
Ribose 5-phosphate	-	-	0.535	0.086	-	-
Serine	1.036	0.051	0.809	0.063	0.865	0.113
Serine	1.045	0.063	0.919	0.108	0.938	0.065
Stearic acid	0.993	0.033	1.009	0.023	0.994	0.056
Succinate	1.001	0.014	1.111	0.056	0.915	0.110
Succinate	0.978	0.027	1.109	0.137	0.998	0.050
Succinyl-CoA	1.305	0.193	0.930	0.185	1.004	0.344
Taurine	0.979	0.020	1.041	0.043	0.701	0.195
Taurine	0.791	0.129	1.325	0.071	1.009	0.019
Thiamine	1.041	0.264	1.022	0.121	0.615	0.186
Thiamine Monophosphate	-	-	0.889	0.045	-	-
Thiamine Pyrophosphate	-	-	1.047	0.114	-	-
Threonine	1.048	0.069	0.766	0.077	0.680	0.259
Threonine	1.218	0.221	0.955	0.154	0.846	0.095
Tyrosine	0.914	0.096	0.813	0.086	0.883	0.147
Tyrosine	0.849	0.103	0.872	0.111	0.762	0.052
UDP	1.256	0.158	1.156	0.154	0.878	0.117
UDP-Glucose	1.460	0.131	1.115	0.083	1.025	0.106
Uracil	0.893	0.096	1.017	0.075	0.962	0.081
Uracil	0.884	0.061	1.011	0.082	1.086	0.173
Urate	1.021	0.303	0.965	0.231	1.280	0.136
Urea	1.080	0.118	0.843	0.097	0.918	0.171
Valine	0.983	0.042	0.968	0.122	0.898	0.083
Valine	1.005	0.054	0.908	0.071	0.979	0.099
Xanthine	1.273	0.305	0.865	0.072	1.025	0.235
Xanthine	1.354	0.179	0.713	0.055	1.529	0.222

Red bold indicates statistical significance $P < 0.05$. - indicates that measurable amounts of that metabolite were not detected. Repeated metabolites represent measurements by both LC/MS and GC/MS.

Table S3. Metabolite Levels in MUT Tissue Relative to WT- 4 Weeks of Age

Metabolite	Retina- Fold Change		Kidney- Fold Change		Eyecup- Fold Change	
	Mean	SEM	Mean	SEM	Mean	SEM
1-Methyladenosine	0.669	0.148	0.830	0.139	1.758	0.342
2-hydroxyglutarate/Mevalonate	0.797	0.059	1.417	0.203	0.900	0.092
2-Methylbutyrylcarnitine	1.687	0.701	1.109	0.084	0.927	0.123
3-Aminoisobutanoic acid	1.327	0.604	1.004	0.035	1.095	0.164
3PG	1.694	0.134	1.454	0.412	1.129	0.106
4-Hydroxyproline	1.037	0.071	1.143	0.111	1.197	0.159
5-oxoproline	1.102	0.069	1.005	0.072	0.912	0.104
α -Ketoglutarate	1.341	0.165	1.068	0.283	0.881	0.143
Acetyl coa	0.741	0.218	2.637	0.440	0.558	0.086
Acetyl L carnitine	1.234	0.296	0.998	0.014	1.027	0.038
AcetylCholine	2.144	1.028	1.120	0.073	0.674	0.149
Acetylglutamic acid	0.666	0.179	0.953	0.059	-	-
Aconitate	0.849	0.196	0.758	0.152	0.662	0.101
Adenine	1.148	0.168	1.469	0.217	1.152	0.086
Adenosine	1.301	0.548	1.150	0.101	0.788	0.166
ADP	1.404	0.791	2.354	0.628	0.546	0.106
AICAR	0.965	0.302	0.915	0.072	0.693	0.079
Alanine	1.025	0.530	0.973	0.033	0.956	0.107
Alanine	1.144	0.067	0.972	0.065	0.994	0.099
Amino adipic acid	0.826	0.298	1.368	0.212	0.410	0.050
Arginosuccinate	1.964	0.573	1.589	0.115	0.943	0.137
Ascorbic acid	0.392	0.167	1.320	0.123	0.999	0.612
Asparagine	1.284	0.080	0.865	0.086	0.952	0.142
Aspartate	1.293	0.063	1.012	0.136	1.163	0.126
Aspartic acid	1.640	0.800	1.064	0.096	0.915	0.108
Betaine	1.363	0.663	0.988	0.021	1.064	0.077
Biotin	0.967	0.098	0.944	0.052	0.894	0.069
Butyrylcarnitine	1.748	0.894	1.133	0.109	0.793	0.107
cAMP	2.500	1.276	0.926	0.159	0.769	0.103
Carnosine	0.883	0.257	0.776	0.130	0.692	0.061
Cholesterol	1.263	0.172	0.771	0.123	0.932	0.328
Cis-aconitic acid	1.354	0.106	0.942	0.263	0.895	0.122
Citraconic acid	1.596	0.416	1.156	0.099	0.613	0.115
Citrate	1.359	0.073	1.157	0.387	0.846	0.096
Citrulline	0.826	0.228	0.945	0.043	0.795	0.058
Co Enzyme A	0.449	0.138	1.522	0.198	-	-
Creatine	1.150	0.335	1.062	0.061	0.947	0.070
Cyclic ADP-ribose	0.763	0.171	1.764	0.363	0.553	0.067
Cystathionine	1.498	0.152	1.105	0.116	0.925	0.129
Cystine	1.388	0.165	1.224	0.097	1.308	0.453
Cytidine	1.482	0.339	0.874	0.111	0.874	0.106
D-Ribulose 5-phosphate	0.917	0.251	0.841	0.096	0.838	0.100
Erythritol	0.882	0.189	0.989	0.023	0.861	0.126
Fumarate	1.321	0.089	1.021	0.185	1.105	0.206
Gamma-Aminobutyric acid (GABA)	1.130	0.046	0.975	0.123	1.718	0.709
Gamma-Aminobutyric acid (GABA)	1.082	0.255	1.046	0.156	1.145	0.381
GDP	1.137	0.246	2.027	0.529	0.522	0.000
Glucose	-	-	0.982	0.099	0.654	0.054
Glucose 1-phosphate	1.250	0.336	1.526	0.239	0.696	0.076
Glucose 6-phosphate	1.155	0.229	1.582	0.254	0.819	0.077
Glutamate	1.016	0.049	1.076	0.046	1.002	0.140
Glutamic acid	1.544	0.723	0.950	0.027	0.809	0.089

Glutathione	0.836	0.136	0.595	0.189	-	-
Glycerate	1.158	0.306	1.210	0.472	0.961	0.159
Glycine	1.282	0.589	1.046	0.042	0.872	0.089
Glycine	1.168	0.053	1.039	0.039	0.795	0.076
GMP	0.903	0.163	1.925	0.608	0.812	0.103
Guanine	0.939	0.129	1.471	0.264	1.042	0.077
Guanosine	0.813	0.181	1.675	0.332	0.855	0.103
Guanosine 5'-phosphate	1.063	0.166	2.087	0.573	0.630	0.068
Hexanoylcarnitine	0.891	0.146	0.795	0.081	0.911	0.139
Histidine	1.084	0.137	1.079	0.063	0.836	0.114
Histidine	1.359	0.137	1.078	0.058	0.690	0.127
Hypotaurine	0.854	0.259	1.120	0.109	0.410	0.078
Hypothanxine	1.239	0.405	0.874	0.075	0.905	0.121
Hypoxanthine	0.633	0.167	0.857	0.079	0.949	0.095
IMP	1.519	0.781	2.219	0.645	0.736	0.107
Inosine	1.725	1.086	0.728	0.110	0.802	0.177
Inosine	1.552	0.651	0.951	0.023	0.773	0.145
IPP	2.263	0.982	1.003	0.087	0.805	0.151
Isobutyrylcarnitine	1.342	0.530	1.149	0.104	0.794	0.117
Isocitrate	1.401	0.149	1.129	0.100	0.961	0.108
Isoleucine	1.085	0.064	1.041	0.018	0.863	0.101
L-Arginine	0.822	0.051	1.212	0.083	0.827	0.068
L-Asparagine	0.600	0.176	0.925	0.089	0.944	0.162
L-Homoserine	1.050	0.107	1.003	0.035	0.742	0.060
Lactate	1.115	0.207	1.083	0.037	0.828	0.142
Lactate	1.073	0.066	1.011	0.037	0.744	0.081
Leucine	1.380	0.648	1.013	0.036	0.864	0.108
Leucine	1.115	0.062	0.991	0.028	0.782	0.078
Lysine	1.455	0.586	1.079	0.050	0.818	0.081
Lysine	1.371	0.102	1.005	0.042	0.672	0.070
Malate	1.520	0.695	0.820	0.355	1.004	0.103
Malate	1.251	0.070	0.705	0.234	0.893	0.066
Maleic acid	0.991	0.009	1.007	0.010	0.981	0.010
Methionine	1.405	0.514	1.094	0.089	0.837	0.106
Methionine	1.116	0.061	1.054	0.115	0.777	0.100
Myo inositol	2.192	1.003	0.913	0.049	0.934	0.134
Myristic acid-D27	0.980	0.032	1.036	0.017	0.994	0.052
Myristoylcarnitine	1.323	0.590	0.891	0.235	0.699	0.128
N-alpha-acetyl-L-lysine	2.371	1.165	1.104	0.069	0.913	0.097
N-Asp	1.153	0.068	1.069	0.101	0.917	0.176
Ornithine	1.379	0.058	0.948	0.716	0.810	0.086
Oxalic acid	0.730	0.109	1.004	0.506	1.038	0.079
Oxalic acid	1.016	0.069	1.005	0.056	0.867	0.049
Oxidized glutathione	1.940	1.093	0.916	0.025	0.859	0.089
Palmitate	1.002	0.018	0.940	0.062	0.964	0.017
Palmitoyl CoA	0.872	0.170	1.075	0.706	0.913	0.165
Palmitoylcarnitine	1.335	0.421	0.529	0.024	0.660	0.079
Pantothenic acid	1.611	0.403	1.128	0.110	0.795	0.079
Pantothenic acid	0.968	0.102	1.063	0.173	0.702	0.079
PEP	2.137	0.413	1.492	0.047	0.903	0.096
Phenylalanine	1.201	0.074	1.018	0.047	0.907	0.102
phenylalanine	1.452	0.644	1.000	0.460	0.803	0.090
Phosphocreatine	1.835	0.942	1.247	0.029	1.079	0.238
Propionylcarnitine	1.650	0.757	1.172	0.053	0.893	0.219
Pyroglutamic acid	1.746	0.843	1.015	0.064	0.764	0.102

Pyruvate	1.541	0.238	1.314	0.075	0.975	0.197
Riboflavin	0.756	0.221	1.015	0.060	1.158	0.144
Serine	0.821	0.075	1.014	0.095	0.896	0.075
Serine	1.073	0.068	0.935	0.102	0.763	0.071
Sphinganine	0.746	0.139	1.296	0.034	0.971	0.169
Stearic acid	1.019	0.018	0.994	0.045	0.973	0.014
Stearyl carnitine	0.503	0.114	0.661	0.232	0.694	0.104
Succinate	1.405	0.372	1.298	0.011	0.926	0.102
Succinate	0.957	0.061	1.149	0.221	0.932	0.054
Succinyl CoA	0.780	0.230	1.584	0.081	0.497	0.087
Taurine	0.815	0.131	1.050	0.035	1.034	0.118
Taurine	0.849	0.062	1.202	0.185	0.747	0.083
Thiamine	0.741	0.233	0.917	0.097	-	-
Thiamine monophosphate	0.663	0.194	1.063	0.061	0.734	0.097
Thiamine pyrophosphate	0.684	0.204	1.122	0.101	1.057	0.261
Threonine	0.722	0.210	1.083	0.086	0.911	0.077
Threonine	1.106	0.108	0.994	0.229	0.712	0.058
Tryptophan	0.745	0.211	1.061	0.027	0.847	0.175
Tryptophan	1.519	0.290	1.027	0.105	0.670	0.099
Tyrosine	0.873	0.249	1.062	0.034	0.951	0.082
Tyrosine	1.205	0.071	1.014	0.044	0.688	0.052
UDP	1.373	0.662	1.848	0.040	0.575	0.076
UDP glucosamine	0.939	0.282	1.125	0.115	0.817	0.085
UDP-glucose	0.931	0.273	1.032	0.421	0.825	0.085
Uracil	1.704	0.295	0.930	0.111	0.850	0.075
Uracil	1.050	0.102	0.932	0.094	0.981	0.064
Urate	1.494	0.291	2.485	0.051	0.922	0.229
Urea	1.211	0.076	0.861	0.034	0.656	0.125
Valine	0.951	0.137	1.011	0.093	0.828	0.095
Valine	1.028	0.059	0.997	0.110	0.711	0.067
Xanthine	0.600	0.176	0.925	0.022	0.837	0.184
Xanthine	1.537	0.671	0.937	0.049	1.300	0.184
XMP	0.895	0.252	1.418	0.062	0.843	0.112

Red bold indicates statistical significance $P < 0.05$. - indicates that measurable amounts of that metabolite were not detected. Repeated metabolites represent measurements by both LC/MS and GC/MS.

Table S4. Altered Metabolites Longitudinal Analysis

Metabolite	Adjusted P Value
LC-MS Retina	
Adenosine	
2 Weeks-MUT vs. 2 Weeks-WT	<0.0001
2 Weeks-MUT vs. 4 Weeks-MUT	<0.0001
2 Weeks-MUT vs. 4 Weeks-WT	<0.0001
2 Weeks-MUT vs. 3 Weeks-MUT	<0.0001
2 Weeks-MUT vs. 3 weeks-WT	<0.0001
Carnosine	
2 Weeks-MUT vs. 2 Weeks-WT	0.0235
2 Weeks-MUT vs. 4 Weeks-MUT	0.0126
2 Weeks-MUT vs. 4 Weeks-WT	0.0235
Inosine	
2 Weeks-MUT vs. 2 Weeks-WT	<0.0001
2 Weeks-MUT vs. 4 Weeks-MUT	<0.0001
2 Weeks-MUT vs. 4 Weeks-WT	<0.0001
2 Weeks-MUT vs. 3 Weeks-MUT	<0.0001
2 Weeks-MUT vs. 3 weeks-WT	<0.0001
Propionyl Carnitine	
2 Weeks-MUT vs. 2 Weeks-WT	0.0127
2 Weeks-MUT vs. 4 Weeks-WT	0.0127
2 Weeks-MUT vs. 3 Weeks-MUT	0.0366
2 Weeks-MUT vs. 3 weeks-WT	0.0297
GC-MS Retina	
Hypothanxine	
2 Weeks-MUT vs. 2 Weeks-WT	<0.0001
2 Weeks-MUT vs. 4 Weeks-MUT	<0.0001
2 Weeks-MUT vs. 4 Weeks-WT	<0.0001
2 Weeks-MUT vs. 3 Weeks- WT	<0.0001
2 Weeks-MUT vs. 3 Weeks- MUT	<0.0001
PEP	
2 Weeks-MUT vs. 4 Weeks-MUT	0.0041
2 Weeks-WT vs. 4 Weeks-MUT	0.0156
4 Weeks-MUT vs. 4 Weeks-WT	0.0156
4 Weeks-MUT vs. 3 Weeks- WT	0.0331
Xanthine	
2 Weeks-MUT vs. 2 Weeks-WT	<0.0001
2 Weeks-MUT vs. 4 Weeks-MUT	<0.0001

2 Weeks-MUT vs. 4 Weeks-WT	<0.0001
2 Weeks-MUT vs. 3 Weeks- WT	<0.0001
2 Weeks-MUT vs. 3 Weeks- MUT	<0.0001
LC-MS Eyecup	
1-Methyladenosine	
2 Weeks-MUT vs. 4 Weeks-MUT	0.0043
2 Weeks-WT vs. 4 Weeks-MUT	0.0354
4 Weeks-MUT vs. 4 Weeks-WT	0.0128
4 Weeks-MUT vs. 3 Weeks-WT	0.0128
4 Weeks-MUT vs. 3 Weeks-MUT	0.0033
Aminoadipic acid	
2 Weeks-MUT vs. 4 Weeks-MUT	0.0197
2 Weeks-WT vs. 4 Weeks-MUT	0.0104
AMP	
2 Weeks-MUT vs. 3 Weeks-MUT	0.0175
4 Weeks-MUT vs. 3 Weeks-MUT	0.0066
Inosine	
4 Weeks-MUT vs. 3 Weeks-MUT	0.0007
4 Weeks-WT vs. 3 Weeks-MUT	0.0457
3 Weeks-WT vs. 3 Weeks-MUT	0.0457
Succinyl coenzyme A	
2 Weeks-MUT vs. 4 Weeks-MUT	0.0032
2 Weeks-WT vs. 4 Weeks-MUT	0.038
Thiamine2	
2 Weeks-WT vs. 4 Weeks-MUT	0.0262
LC-MS Kidney	
Glutamine	
2 Weeks-MUT vs. 2 Weeks-WT	<0.0001
2 Weeks-MUT vs. 4 Weeks-MUT	<0.0001
2 Weeks-MUT vs. 4 Weeks-WT	<0.0001
2 Weeks-MUT vs. 3 Weeks-WT	<0.0001
2 Weeks-MUT vs. 3 Weeks-MUT	<0.0001
GC-MS Kidney	
3PG	
4 Week-MUT vs. 3 Week-MUT	0.0036
4 Week-WT vs. 2 Week-MUT	0.0036
4 Week-WT vs. 3 Week-MUT	<0.0001

2 Week-MUT vs. 2 Week-WT	0.0036
2 Week-MUT vs. 3 Weeks-WT	0.0096
2 Week-WT vs. 3 Week-MUT	<0.0001
3 Weeks-WT vs. 3 Week-MUT	<0.0001
Cystathionine	
4 Week-WT vs. 3 Week-MUT	0.0207
2 Week-WT vs. 3 Week-MUT	0.0207
3 Weeks-WT vs. 3 Week-MUT	0.0384
PEP	
4 Week-MUT vs. 3 Week-MUT	<0.0001
4 Week-WT vs. 2 Week-MUT	0.0034
4 Week-WT vs. 3 Week-MUT	<0.0001
2 Week-MUT vs. 2 Week-WT	0.0034
2 Week-MUT vs. 3 Weeks-WT	0.0091
2 Week-MUT vs. 3 Week-MUT	<0.0001
2 Week-WT vs. 3 Week-MUT	<0.0001
3 Weeks-WT vs. 3 Week-MUT	<0.0001

Table S8. RNA Sequencing Biological Replicates and RIN Numbers

Mouse ID	Sample ID	Age	Tissue	Confirmed Nmnat1 Genotype	Sex	RIN Number
8001	10A	2 Weeks	Retina	WT	Male	9.8
8036	11A	2 Weeks	Retina	WT	Male	9.7
8141	18A	2 Weeks	Retina	WT	Female	9.7
7772	13A	2 Weeks	Retina	WT	Male	9.5
7997	14A	2 Weeks	Retina	WT	Male	9.6
8003	15A	2 Weeks	Retina	WT	Male	9.4
7774	1A	2 Weeks	Retina	MUT	Female	9.3
7778	2A	2 Weeks	Retina	MUT	Female	9.4
8039	3A	2 Weeks	Retina	MUT	Female	9.4
7761	8A	2 Weeks	Retina	MUT	Male	9.5
7994	5A	2 Weeks	Retina	MUT	Female	9.4
8084	6A	2 Weeks	Retina	MUT	Female	9.6
7995	9C	2 Weeks	Kidney	WT	Male	9.1
7772	13C	2 Weeks	Kidney	WT	Male	9.3
8001	10C	2 Weeks	Kidney	WT	Male	9.4
8036	11C	2 Weeks	Kidney	WT	Male	9.4
8141	18C	2 Weeks	Kidney	WT	Female	9.6
7997	14C	2 Weeks	Kidney	WT	Male	8.7
8003	15C	2 Weeks	Kidney	WT	Male	8.6
7778	2C	2 Weeks	Kidney	MUT	Female	9.4
7994	5C	2 Weeks	Kidney	MUT	Female	9.7
8039	3C	2 Weeks	Kidney	MUT	Female	9.2
8017	17C	2 Weeks	Kidney	MUT	Female	9.3
8084	6C	2 Weeks	Kidney	MUT	Female	8.6
7532	5A	3 weeks	Retina	WT	Female	9.2
7479	6A	3 weeks	Retina	WT	Female	8.7
7586	7A	3 weeks	Retina	WT	Female	9.1
7538	13A	3 weeks	Retina	WT	Male	9.2
7504	14A	3 weeks	Retina	WT	Male	8.8
7578	16A	3 weeks	Retina	WT	Male	9.3
7435	2A	3 weeks	Retina	MUT	Female	8.7
7587	3A	3 weeks	Retina	MUT	Female	8.8
7432	4A	3 weeks	Retina	MUT	Female	9.0
7487	10A	3 weeks	Retina	MUT	Male	8.9
7509	11A	3 weeks	Retina	MUT	Male	9.1
7503	9A	3 weeks	Retina	MUT	Male	9.4
7532	5C	3 weeks	Kidney	WT	Female	8.6
7479	6C	3 weeks	Kidney	WT	Female	9.3
7586	7C	3 weeks	Kidney	WT	Female	9.1
7585	8C	3 weeks	Kidney	WT	Female	9.7
7504	14C	3 weeks	Kidney	WT	Male	8.8
7507	15C	3 weeks	Kidney	WT	Male	9.5
7433	1C	3 weeks	Kidney	MUT	Female	9.4
7435	2C	3 weeks	Kidney	MUT	Female	9.1
7587	3C	3 weeks	Kidney	MUT	Female	9.6
7432	4C	3 weeks	Kidney	MUT	Female	8.8
7509	11C	3 weeks	Kidney	MUT	Male	8.9
7503	9C	3 weeks	Kidney	MUT	Male	9.2

Table S9. Primer Sequences and Efficiencies

Target	Forward (5'-->3')	Reverse (5'-->3')	Efficiency (%)	Ascession Number/Reference
<i>Gadd45b</i>	GCTCTGGGGATCTCCGTG	CTGTCGGGGTCCACATTCAT	107	NM_008655.1
<i>Gapdh</i>	CATCACTGCCACCCAGAAGACTG	ATGCCAGTGAGCTTCCCGTTCAG	98	NM_001289726.1 /PMID: 26491017
<i>Lad1</i>	CTAGCACCGTCAAGCTAGGG	ACGCAGATTCTCCTTTCGGA	96	NM_133664.3
<i>Lif</i>	AATGCCACCTGTGCCATACG	CAACTTGGTCTTCTCTGTCCCG	104	NM_008501.2 /PMID: 24008729

Table S10. Antibodies and Dilutions

Antibody	Manufacturer	Product ID	Host	WB Dilution	WB Molecular Weight (kDa)	IHC Dilution	Lot Number
Beta Actin	Abcam	ab8226	Mouse	1 to 5,000	42	-	GR3249122-16
Beta Actin	Abcam	ab8227	Rabbit	1 to 5,000	42	-	GR3244314-1
CD45	Invitrogen	14-0451-82	Rat	-	-	1 to 250	2269880
DAPI	Molecular Probes	D21490	-	-	-	1 to 7,500	1575898
Gfap	Abcam	ab68428	Rabbit	1 to 10,000	50	1 to 100	GR257920-32
HKII	Cell Signaling	C64G5 (2867S)	Rabbit	1 to 1,000	102	-	Lot 5
Iba1	Wako	019-19741	Rabbit	-	-	1 to 250	SKP3626
Phospho-PKM2	Cell Signaling	TYR105 (3827S)	Rabbit	1 to 1,000	60	-	Lot 3
PKM2	Cell Signaling	D78A4 (4053S)	Rabbit	1 to 1,000	60	1 to 400	Lot 6
PMK1	Cell Signaling	D30G6 (7067S)	Rabbit	1 to 1,000	60	1 to 100	Lot 4
yH2AX	Trevigen/ R and D Systems	4418-APC-020	Rabbit	-	-	1 to 100	CMFS0321012
Donkey Anti-Rat AlexaFluor 488	Invitrogen	A21208	Donkey	-	-	1 to 500	2180272
Goat anti-Rabbit AlexaFluor 594	Abcam	ab150080	Goat	-	-	1 to 500	GR3323881-1
Goat anti-mouse AlexaFluor 488	Invitrogen	A32723	Goat	-	-	1 to 500	VC300588
Goat anti-Rabbit Alexafluor 555	Invitrogen	A32732	Goat	-	-	1 to 500	VC297826