**S1 Table. Changed metabolites observed for *Pkd2-/-* and *Slc25a25-/-* genotypes.**

|  |  |  |  |
| --- | --- | --- | --- |
| Metabolite | HMDB ID | Function / Pathway | References |
| Ratio increased | | | |
| taurine | HMDB00251 | Non-proteinogenic, essential, sulphur containing amino acid. Implicated in development and degenerative processes of neuronal system. | [1-3] |
| choline phosphate | HMDB01565 | Intermediate in biosynthesis of amino acids glycine, serine and threonine, and glycerophospholipids. Involved in fetal lung development. | [4,5] |
| 2-methyl-butyrylcarnitine (C5) | HMDB00378 | Short-chain acyl carnitine involved in amino acid metabolism of valine, isoleucine and leucine. Elevation of 2-methylbutyrylcarnitine suggests a deficiency of a dehydrogenase specific for isobutyryl-CoA. | [6] |
| Ratio decreased | | | |
| 3-(4-hydroxyphenyl)-lactate | HMDB00755 | Tyrosine metabolite which can act as inhibitor of ROS generation in mitochondria. | [7] |
| proline | HMDB00162 | Proteinogenic amino acid essential for collagen structure. |  |
| N-acetylalanine | HMDB00766 | Substrate for Guanine nucleotide-binding proteins. |  |
| alanine | HMDB00161 | One of the most important amino acids released by muscle, functioning as a major energy source. |  |
| N-acetylserine | HMDB02931 | Mostly found as an N-terminal amino acid directing acetylated proteins to ATP-ubiquitin-dependent proteasome. |  |
| N-acetylmethionine | HMDB11745 | Metabolite of amino acid methionine. |  |
| gamma-glutamylalanine | HMDB29142 | Dipeptide, an intermediate of protein metabolism. |  |

HMDB ID is a unique identifier of a given metabolite [8]. Pathway assignment is derived from HMDB.

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