## Supplementary Material 1

### 1. Electromyography results of the proband.

| | | Slack status | | | | | With mild force | | | | | With heavy force | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Muscles | Side | Inserted potential | fibrillation potential | positive potential | Myotonic potential | Beam fibrillation potential | average time (ms) | average amplitude (mv) | polyphasic potential | giant potential | Myopathy potential | Interference phase | mixed phase | pure phase | largest amplitude | pathological interference phase | Other |
| tibialis anterior | left | 10 | 10 | 10 | | | ↑ | ↑ | | | | | | √ | | | |
| peroneus longus | left | 10 | 10 | 10 | | | ↑ | ↑ | | | | | | √ | | | |
| extensor digitorum brevis | left | 10 | 10 | 20 | | | ↑ | ↑ | | | | | | √ | | | |
| musculus gastrocnemius | left | 0 | | | | | 0 | 0 | | | | | | √ | | | |
| medial vastus muscle | left | 0 | | | | | ↑ | ↑ | | | | | | √ | | | |
| tibialis anterior | right | 10 | 10 | 20 | | | ↑ | ↑ | | | | | | √ | | | |
| peroneus longus | right | 10 | 10 | 10 | | | ↑ | ↑ | | | | | | √ | | | |
| extensor digitorum brevis | right | 10 | 10 | 20 | | | ↑ | ↑ | | | | | | √ | | | |
| musculus gastrocnemius | right | 0 | | | | | 0↑ | 0↑ | | | | | | √ | | | |
| medial vastus muscle | right | 10 | 10 | 10 | | | ↑ | ↑ | | | | | | √ | | | |
| paravertebral muscle T11 | right | - | | 10 | | | ↑ | ↑ | | | | | | | | | |
| paravertebral muscle T12 | left | 0 | | | | | 0 | 0 | | | | | | | | | |
| first dorsal interosseus | left | 0 | | | | | ↑ | ↑ | | | | | | √ | | | |
| musculi abductor pollicis brevis | left | 0 | | | | | ↑ | ↑ | | | | | √ | √ | | | |
| extensor digitorum | left | 0 | | | | | ↑ | ↑ | | | | | √ | | | | |
| deltoideus triangularis | left | 0 | | | | | 0 | 0 | | | | | √ | | | | |
| bicipital muscle of arm | left | 0 | | | | | 0 | 0 | | | | | √ | | | | |
| first dorsal interosseus | right | - | ± | ± | | | ↑ | ↑ | | | | | | √ | | | |
| musculi abductor pollicis brevis | right | 0 | | | | | 0↑ | 0↑ | | | | | √ | | | | |
| extensor digitorum | right | 0 | | | | | 0 | 0 | | | | | √ | | | | |
| deltoideus triangularis | right | 0 | | | | | 0 | 0 | | | | | √ | | | | |
| bicipital muscle of arm | right | 0 | | | | | 0 | 0 | | | | | √ | | | | |

Motor nerve conduction

| Nerve | Stimulating spot | Recording spot | Latency time (ms) | | Amplitude (mv) | | Conduction velocity or excitation rate (m/s) | | Other |
|---|---|---|---|---|---|---|---|---|---|
| | | | left | right | left | right | left | right | |
| deep peroneal nerve | Capitula fibula | tibialis anterior | 12.2 | 20.1 | 8.4 | 4.6 | 11 | 7 | |
| superficial peroneal nerve | Capitula fibula | peroneus longus | 14.4 | 13.8 | 5.6 | 7.4 | 9 | 11 | |
| total peroneal nerve | ankle | extensor digitorum brevis | 25.8 | 7.2 | 2.8 | 2.7 | 7 | 12 | |
| | Capitula fibula | | 87.0 | 32.9 | 2.5 | 2.7 | | | |
| nervus tibialis | popliteal fossa | musculus gastrocnemius | 16.0 | 30.3 | 8.6 | 3.2 | 10 | 7 | |
| nervus femoralis | | medial vastus muscle | 39.4 | 71.8 | 2.3 | 1.0 | 7 | 3 | |
| ulnar nerve | wrist | first dorsal interosseus | 15.1 | 10.9 | 7.9 | 11.4 | 5 | 6 | Waveform from several |
| | elbow | | 59.8 | 53.2 | 9.0 | 6.4 | | | Waveform from several |
| nervus medianus | wrist | musculi abductor pollicis brevis | 11.8 | 12.0 | 3.8 | 2.3 | 4 | 5 | Waveform from several |
| | elbow | | 58.5 | 54.6 | 4.1 | 1.4 | | | Waveform from several |
| musculospiralis | radial groove | extensor digitorum | 28.9 | 28.1 | 2.9 | 2.3 | 7 | 8 | Waveform from several |
| musculocutaneous nerve | ERB'S | deltoideus triangularis | 28.7 | 20.6 | 2.0 | 2.9 | 9 | 11 | Waveform from several |
| axillary nerve | ERB'S | bicipital muscle of arm | 12.1 | 26.2 | 6.1 | 6.7 | 12 | 6 | Waveform from several |

Sensory nerve conduction

| Nerve | Stimulating spot | Recording spot | Latency time (ms) | | Amplitude (mv) | | Excitation rate (m/s) | | Other |
|---|---|---|---|---|---|---|---|---|---|
| | | | left | right | left | right | left | right | |
| ulnar nerve | little finger | wrist | no SNAP | no SNAP | | | | | |
| nervus medianus | digitus medius | wrist | no SNAP | no SNAP | | | | | |
| superficial peroneal nerve | | | no SNAP | no SNAP | | | | | |
| nervus suralis | | | no SNAP | no SNAP | | | | | |

**Results description:** Electromyography shows fibrillar, positive phase potential, action potential amplitude, time-limit widening increased when light force; Motor nerve conduction amplitude is normal/mild decrease; nerve conduction velocity is about 10 m/s (which is unusual in CMT2A); sensory nerve conduction is seriously without SNAP.

**2. Detailed data of the vector construction and cell transfection.**

(1) cDNA sequences

No. pHS-AVC-LY059 (wild-type) MFN2 (human, NM_014874)

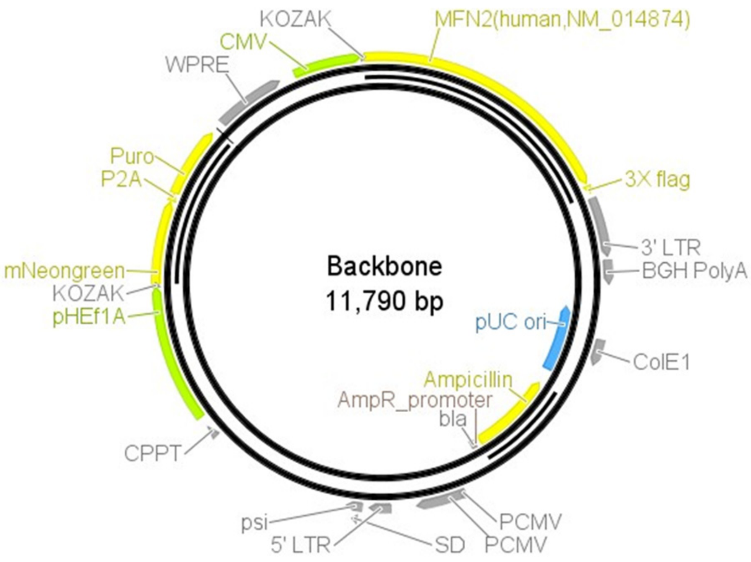
*5’-ATGTCCCTGCTCTTCTCTCGATGCAACTCTATCGTCACAGTCAAGAAAAATAAGAGACACATGGCTGAGGTGAATGCATCCCCACTTAAGCACTTTGTCACTGCCAAGAAGAAGATCAATGGCATTTTTGAGCAGCTGGGGGCCTACATCCAGGAGAGCGCCACCTTCCTTGAAGACACGTACAGGAATGCAGAACTGGACCCCGTTACCACAGAAGAACAGGTTCTGGACGTCAAAGGTTACCTATCCAAAGTGAGAGGCATCAGTGAGGTGCTGGCTCGGAGGCACATGAAAGTGGCTTTTTTTGGCCGGACGAGCAATGGGAAGAGCACCGTGATCAATGCCATGCTCTGGGACAAAGTTCTGCCCTCTGGGATTGGCCACACCACCAATTGCTTCCTGCGGGTAGAGGGCACAGATGGCCATGAGGCCTTTCTCCTTACCGAGGGCTCAGAGGAAAAGAGGAGTGCCAAGACTGTGAACCAGCTGGCCCATGCCCTCCACCAGGACAAGCAGCTCCATGCCGGCAGCCTAGTGAGTGTGATGTGGCCCAACTCTAAGTGCCCACTTCTGAAGGATGACCTCGTTTTGATGGACAGCCCTGGTATTGATGTCACCACAGAGCTGGACAGCTGGATTGACAAGTTTTGTCTGGATGCTGATGTGTTTGTGCTGGTGGCCAACTCAGAGTCCACCCTGATGCAGACGGAAAAGCACTTCTTCCACAAGGTGAGTGAGCGTCTCTCCCGGCCAAACATCTTCATCCTGAACAACCGCTGGGATGCATCTGCCTCAGAGCCCGAGTACATGGAGGAGGTGCGGCGGCAGCACATGGAGCGTTGTACCAGCTTCCTGGTGGATGAGCTGGGCGTGGTGGATCGATCCCAGGCCGGGGACCGCATCTTCTTTGTGTCTGCTAAGGAGGTGCTCAACGCCAGGATTCAGAAAGCCCAGGGCATGCCTGAAGGAGGGGGCGCTCTCGCAGAAGGCTTTCAAGTGAGGATGTTTGAGTTTCAGAATTTTGAGAGGAGATTTGAGGAGTGCATCTCCCAGTCTGCAGTGAAGACCAAGTTTGAGCAGCACACGGTCCGGGCCAAGCAGATTGCAGAGGCGGTTCGACTCATCATGGACTCCCTGCACATGGCGGCTCGGGAGCAGCAGGTTTACTGCGAGGAAATGCGTGAAGAGCGGCAAGACCGACTGAAATTTATTGACAAACAGCTGGAGCTCTTGGCTCAAGACTATAAGCTGCGAATTAAGCAGATTACGGAGGAAGTGGAGAGGCAGGTGTCGACTGCAATGGCCGAGGAGATCAGGCGCCTCTCTGTACTGGTGGACGATTACCAGATGGACTTCCACCCTTCTCCAGTAGTCCTCAAGGTTTATAAGAATGAGCTGCACCGCCACATAGAGGAAGGACTGGGTCGAAACATGTCTGACCGCTGCTCCACGGCCATCACCAACTCCCTGCAGACCATGCAGCAGGACATGATAGATGGCTTGAAACCCCTCCTTCCTGTGTCTGTGCGGAGTCAGATAGACATGCTGGTCCCACGCCAGTGCTTCTCCCTCAACTATGACCTAAACTGTGACAAGCTGTGTGCTGACTTCCAGGAAGACATTGAGTTCCATTTCTCTCTCGGATGGACCATGCTGGTGAATAGGTTCCTGGGCCCCAAGAACAGCCGTCGGGCCTTGATGGGCTACAATGACCAGGTCCAGCGTCCCATCCCTCTGACGCCAGCCAACCCCAGCATGCCCCCACTGCCACAGGGCTCGCTCACCCAGGAGGAGTTCATGGTTTCCATGGTTACCGGCCTGGCCTCCTTGACATCCAGGACCTCCATGGGCATTCTTGTTGTTGGAGGAGTGGTGTGGAAGGCAGTGGGCTGGCGGCTCATTGCCCTCTCCTTTGGGCTCTATGGCCTCCTCTACGTCTATGAGCGTCTGACCTGGACCACCAAGGCCAAGGAGAGGGCCTTCAAGCGCCAGTTTGTGGAGCATGCCAGCGAGAAGCTGCAGCTTGTCATCAGCTACACTGGCTCCAACTGCAGCCACCAAGTCCAGCAGGAACTGTCTGGGACCTTTGCTCATCTGTGTCAGCAAGTTGACGTCACCCGGGAGAACCTGGAGCAGGAAATTGCCGCCATGAACAAGAAAATTGAGGTTCTTGACTCACTTCAGAGCAAAGCAAAGCTGCTCAGGAATAAAGCCGGTTGGTTGGACAGTGAGCTCAACATGTTCACACACCAGTACCTGCAGCCCAGCAGA-3’*

No. pHS-AVC-LY060 (mutant) MFN2(human,NM_014874, 638T>C)

*5’-ATGTCCCTGCTCTTCTCTCGATGCAACTCTATCGTCACAGTCAAGAAAAATAAGAGACACATGGCTGAGGTGAATGCATCCCCACTTAAGCACTTTGTCACTGCCAAGAAGAAGATCAATGGCATTTTTGAGCAGCTGGGGGCCTACATCCAGGAGAGCGCCACCTTCCTTGAAGACACGTACAGGAATGCAGAACTGGACCCCGTTACCACAGAAGAACAGGTTCTGGACGTCAAAGGTTACCTATCCAAAGTGAGAGGCATCAGTGAGGTGCTGGCTCGGAGGCACATGAAAGTGGCTTTTTTTGGCCGGACGAGCAATGGGAAGAGCACCGTGATCAATGCCATGCTCTGGGACAAAGTTCTGCCCTCTGGGATTGGCCACACCACCAATTGCTTCCTGCGGGTAGAGGGCACAGATGGCCATGAGGCCTTTCTCCTTACCGAGGGCTCAGAGGAAAAGAGGAGTGCCAAGACTGTGAACCAGCTGGCCCATGCCCTCCACCAGGACAAGCAGCTCCATGCCGGCAGCCTAGTGAGTGTGATGTGGCCCAACTCTAAGTGCCCACTTCTGAAGGATGACCTCGTTTTGATGGACAGCCCTGGTATTGATGTCACCACAGAGCTGGACAGCTGGACTGACAAGTTTTGTCTGGATGCTGATGTGTTTGTGCTGGTGGCCAACTCAGAGTCCACCCTGATGCAGACGGAAAAGCACTTCTTCCACAAGGTGAGTGAGCGTCTCTCCCGGCCAAACATCTTCATCCTGAACAACCGCTGGGATGCATCTGCCTCAGAGCCCGAGTACATGGAGGAGGTGCGGCGGCAGCACATGGAGCGTTGTACCAGCTTCCTGGTGGATGAGCTGGGCGTGGTGGATCGATCCCAGGCCGGGGACCGCATCTTCTTTGTGTCTGCTAAGGAGGTGCTCAACGCCAGGATTCAGAAAGCCCAGGGCATGCCTGAAGGAGGGGGCGCTCTCGCAGAAGGCTTTCAAGTGAGGATGTTTGAGTTTCAGAATTTTGAGAGGAGATTTGAGGAGTGCATCTCCCAGTCTGCAGTGAAGACCAAGTTTGAGCAGCACACGGTCCGGGCCAAGCAGATTGCAGAGGCGGTTCGACTCATCATGGACTCCCTGCACATGGCGGCTCGGGAGCAG-*

*CAGGTTTACTGCGAGGAAATGCGTGAAGAGCGGCAAGACCGACTGAAATTTATTGACAAACAGCTGGAGCTCTTGGCTCAAGACTATAAGCTGCGAATTAAGCAGATTACGGAGGAAGTGGAGAGGCAGGTGTCGACTGCAATGGCCGAGGAGATCAGGCGCCTCTCTGTACTGGTGGACGATTACCAGATGGACTTCCACCCTTCTCCAGTAGTCCTCAAGGTTTATAAGAATGAGCTGCACCGCCACATAGAGGAAGGACTGGGTCGAAACATGTCTGACCGCTGCTCCACGGCCATCACCAACTCCCTGCAGACCATGCAGCAGGACATGATAGATGGCTTGAAACCCCTCCTTCCTGTGTCTGTGCGGAGTCAGATAGACATGCTGGTCCCACGCCAGTGCTTCTCCCTCAACTATGACCTAAACTGTGACAAGCTGTGTGCTGACTTCCAGGAAGACATTGAGTTCCATTTCTCTCTCGGATGGACCATGCTGGTGAATAGGTTCCTGGGCCCCAAGAACAGCCGTCGGGCCTTGATGGGCTACAATGACCAGGTCCAGCGTCCCATCCCTCTGACGCCAGCCAACCCCAGCATGCCCCCACTGCCACAGGGCTCGCTCACCCAGGAGGAGTTCATGGTTTCCATGGTTACCGGCCTGGCCTCCTTGACATCCAGGACCTCCATGGGCATTCTTGTTGTTGGAGGAGTGGTGTGGAAGGCAGTGGGCTGGCGGCTCATTGCCCTCTCCTTTGGGCTCTATGGCCTCCTCTACGTCTATGAGCGTCTGACCTGGACCACCAAGGCCAAGGAGAGGGCCTTCAAGCGCCAGTTTGTGGAGCATGCCAGCGAGAAGCTGCAGCTTGTCATCAGCTACACTGGCTCCAACTGCAGCCACCAAGTCCAGCAGGAACTGTCTGGGACCTTTGCTCATCTGTGTCAGCAAGTTGACGTCACCCGGGAGAACCTGGAGCAGGAAATTGCCGCCATGAACAAGAAAATTGAGGTTCTTGACTCACTTCAGAGCAAAGCAAAGCTGCTCAGGAATAAAGCCGGTTGGTTGGACAGTGAGCTCAACATGTTCACACACCAGTACCTGCAGCCCAGCAGA-3’*

(2) Vector backbone diagram



(3) Primers used in the vector construction and QF-PCR

| Target | Forward-primer | Reverse-primer | Segment length (bp) |
|---|---|---|---|
| MFN2 | GGACCCCGTTACCACAGAAG | TTGTCCCAGAGCATGGCATT | 162 |
| MFN2 (638T>C) | GGACCCCGTTACCACAGAAG | TTGTCCCAGAGCATGGCATT | 162 |
| GAPDH | TGACTTCAACAGCGACACCCA | CACCCTGTTGCTGTAGCCAAA | 121 |

(4) Data of QF-PCR (48 h after transfection)

| Gene | Group | Repetition 1 | Repetition 2 | Repetition 3 | Mean | SD | QC[1] |
|---|---|---|---|---|---|---|---|
| MFN2 | 293FT | 0.3 | 0.3 | 0.3 | 0.315 | 0.014 | 0.046 |
| | 293FT-pHS-AVC-LY059 | 2452.4 | 2435.5 | 2385.4 | 2424.436 | 34.872 | 0.014 |
| | 293FT-pHS-AVC-LY060 | 5673.4 | 5673.4 | 5442.3 | 5596.371 | 133.432 | 0.024 |
| | 293FT-pHS-BVC-LW345 | 1.0 | 1.0 | 1.0 | 1.000 | 0.012 | 0.012 |

[1] QC, the value of standard deviation/mean. The larger the value, the higher the volatility of the experimental data.

**Supplementary Table 1.** List of differentiated compounds

| m/z | FC (pHS-AVC-LY059/ pHS-AVC-LY060) | log2(FC) | raw. pval | vip | compound_id | compound_name | adduct | delta (ppm) | sub-class | Source | KEGG_compound | KEGG_pathway | KEGG_pathway_descriptionn |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| 89.9414 | 3.5093 | 1.8112 | 0.079369 | 1.7718 | HMDB0013667 | Yttrium | [M+H]+ | 315 | - | Endogenous | - | - | - |
| 116.9269 | 3.9056 | 1.9656 | 0.045241 | 1.9133 | HMDB0029596 | Chloroform | [M-H]- | 169 | Halomethanes | Endogenous\|Food\|Environmental\|Biological | C13827 | - | - |
| 122.0958 | 0.46894 | -1.0925 | 0.072381 | 1.798 | HMDB0001020 | N, N-Dimethylaniline | [M+H]+ | 5 | Amines | Endogenous\|Food\|Biological | C02846 | - | - |
| 128.0368 | 4.5875 | 2.1977 | 0.004952 | 2.1955 | HMDB0000267 | L-5-Oxoproline | [M-H]- | 12 | Amino acids, peptides, and analogues | Endogenous\|Food\|Biological | C01879 | map00480;<br>map01100 | Glutathione metabolism; Metabolic pathways |
| 137.0271 | 0.46029 | -1.1194 | 0.055543 | 1.8665 | HMDB0000500 | 4-Hydroxybenzoic acid | [M-H]- | 20 | Benzoic acids and derivatives | Endogenous\|Food\|Biological | C00156 | map00130;<br>map00362;<br>map00363;<br>map00623;<br>map00627;<br>map00790;<br>map01061;<br>map01100;<br>map01110;<br>map01120;<br>map01220;<br>map01240;<br>map07110 | Ubiquinone and other terpenoid-quinone biosynthesis; Benzoate degradation; Bisphenol degradation; Toluene degradation; Aminobenzoate degradation; Folate biosynthesis; Biosynthesis of phenylpropanoids; Metabolic pathways; Biosynthesis of secondary metabolites; Microbial metabolism in diverse environments; Degradation of aromatic compounds; Biosynthesis of cofactors; Benzoic acid family |
| 141.112 | 0.33401 | -1.582 | 0.003465 | 2.2178 | HMDB0029598 | Metenamine | [M+H]+ | 11 | 1,3,5-triazinanes | Endogenous\|Food | - | - | - |
| 167.0225 | 0.46796 | -1.0955 | 0.054323 | 1.8719 | HMDB0000289 | Uric acid | [M-H]- | 9 | Purines and purine derivatives | Endogenous\|Food\|Biological | C00366 | map00230;<br>map01100;<br>map01120;<br>map04976 | Purine metabolism; Metabolic pathways; Microbial metabolism in diverse environments; Bile secretion |

| | | | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| 191.0215 | 9.5827 | 3.2604 | 0.082728 | 1.7595 | HMDB0000094 | Citric acid | [M-H]- | 9 | Tricarboxylic acids and derivatives | Endogenous\|Food\|Synthetic\|Biological | C00158 | map00020; map00250; map00630; map00720; map00997; map01053; map01060; map01061; map01062; map01063; map01064; map01065; map01066; map01070; map01100; map01110; map01120; map01200; map01210; map01230; map01240; map02020; map04742; map04922; map05230 | Citrate cycle (TCA cycle); Alanine, aspartate and glutamate metabolism; Glyoxylate and dicarboxylate metabolism; Carbon fixation pathways in prokaryotes; Biosynthesis of various secondary metabolites - part 3; Biosynthesis of siderophore group nonribosomal peptides; Biosynthesis of plant secondary metabolites; Biosynthesis of phenylpropanoids; Biosynthesis of terpenoids and steroids; Biosynthesis of alkaloids derived from shikimate pathway; Biosynthesis of alkaloids derived from ornithine, lysine and nicotinic acid; Biosynthesis of alkaloids derived from histidine and purine; Biosynthesis of alkaloids derived from terpenoid and polyketide; Biosynthesis of plant hormones; Metabolic pathways; Biosynthesis of secondary metabolites; Microbial metabolism in diverse environments; Carbon metabolism; 2-Oxocarboxylic acid metabolism; Biosynthesis of amino acids; Biosynthesis of cofactors; Two-component system; Taste transduction; Glucagon signaling pathway; Central carbon metabolism in cancer |
| 212.0413 | 4.2951 | 2.1027 | 0.016832 | 2.0793 | HMDB0001511 | Phosphocreatine | [M+H]+ | 8 | Amino acids, peptides, and analogues | Endogenous | C02305 | map00330; map01100 | Arginine and proline metabolism; Metabolic pathways |
| 217.1039 | 0.3095 | -1.692 | 0.074128 | 1.7914 | HMDB0029027 | Prolyl-Threonine | [M+H]+ | 66 | Amino acids, peptides, and analogues | Endogenous | - | - | - |

| | | | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| 239.1487 | 3.0514 | 1.6095 | 0.002226 | 2.2403 | HMDB0139940 | N-(3-aminopropyl)-3-(3,4-dihydroxyphenyl)propanimidic acid | [M+H]+ | 41 | Benzenediols | Endogenous | - | - | - |
| 268.1042 | 2.1692 | 1.1172 | 0.079006 | 1.7731 | HMDB0000050 | Adenosine | [M+H]+ | 0 | - | Endogenous \| Food \| Synthetic \| Biological | C00212 | map00230; map01100; map02010; map04022; map04024; map04071; map04080; map04270; map04923; map04924; map05012; map05032; map05034 | Purine metabolism; Metabolic pathways; ABC transporters; cGMP-PKG signaling pathway; cAMP signaling pathway; Sphingolipid signaling pathway; Neuroactive ligand-receptor interaction; Vascular smooth muscle contraction; Regulation of lipolysis in adipocytes; Renin secretion; Parkinson disease; Morphine addiction; Alcoholism |
| 337.2046 | 0.42024 | -1.2507 | 0.055779 | 1.8655 | HMDB0040566 | [6]-Gingerdiol 3-acetate | [M-H]- | 8 | Fatty alcohol esters | Food \| Endogenous \| Biological | - | - | - |
| 341.3064 | 0.28176 | -1.8275 | 0.070702 | 1.8045 | HMDB0032476 | Polyoxyethylene (600) monoricinoleate | [M+H]+ | 4 | Fatty alcohols | Endogenous \| Food \| Biological | - | - | - |
| 400.345 | 0.23881 | -2.0661 | 0.021729 | 2.0443 | HMDB0000222 | L-Palmitoylcarnitine | [M+H]+ | 7 | Fatty acid esters | Endogenous \| Food \| Biological | C02990 | map00071; map01212 | Fatty acid degradation; Fatty acid metabolism |
| 417.2988 | 2.3256 | 1.2176 | 0.06897 | 1.8113 | HMDB0036886 | beta-Carotinal | [M+H]+ | 39 | Triterpenoids | Endogenous \| Food \| Biological | - | - | - |
| 425.125 | 2.3924 | 1.2585 | 0.090263 | 1.7328 | HMDB0003556 | Chitobiose | [M-H]- | 38 | Carbohydrates and carbohydrate conjugates | Endogenous | C01674 | map00520; map01100; map02010; map02060 | Amino sugar and nucleotide sugar metabolism; Metabolic pathways; ABC transporters; Phosphotransferase system (PTS) |
| 455.0973 | 2.9588 | 1.565 | 0.041847 | 1.9298 | HMDB0001520 | Flavin mononucleotide | [M-H]- | 0 | - | Endogenous \| Food \| Biological | C00061 | map00190; map00740; map00997; map01057; map01100; map01110; map01240; map04977 | Oxidative phosphorylation; Riboflavin metabolism; Biosynthesis of various secondary metabolites - part 3; Biosynthesis of type II polyketide products; Metabolic pathways; Biosynthesis of secondary metabolites; Biosynthesis of cofactors; Vitamin digestion and absorption |

| | | | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| 485.2707 | 0.24874 | -2.0073 | 0.068811 | 1.8119 | HMDB0011156 | PA (O-20:4 (5Z,8Z,11Z,14Z)/2:0) | [M-H]- | 7 | Glycerophosphates | Endogenous\|Food\|Biological | - | - | - |
| 494.3191 | 0.46812 | -1.095 | 0.074051 | 1.7917 | HMDB0010383 | LysoPC (16:1 (9Z)/0:0) | [M+H]+ | 10 | Glycerophosphocholines | Endogenous\|Food\|Biological | C04230 | map00564;<br>map05231 | Glycerophospholipid metabolism; Choline metabolism in cancer |
| 504.3039 | 0.40545 | -1.3024 | 0.093119 | 1.723 | HMDB0010381 | LysoPC (15:0/0:0) | [M+Na]+ | 4 | Glycerophosphocholines | Endogenous\|Food\|Biological | C04230 | map00564;<br>map05231 | Glycerophospholipid metabolism; Choline metabolism in cancer |
| 505.3484 | 21.317 | 4.4139 | 0.06939 | 1.8096 | HMDB0032111 | Adlupone | [M+Na]+ | 39 | Carbonyl compounds | Endogenous\|Food | - | - | - |
| 512.2956 | 0.43086 | -1.2147 | 0.088452 | 1.7392 | HMDB0002639 | Sulfolithocholylglycine | [M-H]- | 53 | Bile acids, alcohols and derivatives | Endogenous\|Food\|Biological | C11301 | - | - |
| 539.3611 | 0.19266 | -2.3759 | 0.061443 | 1.8416 | HMDB0002815 | LysoPC (18:1 (9Z)/0:0) | [M+NH4]+ | 39 | Glycerophosphocholines | Endogenous\|Food\|Biological | - | - | - |
| 539.5361 | 2.953 | 1.5622 | 0.050784 | 1.8876 | HMDB0041080 | 9-Hydroxytridecyl docosanoate | [M+H]+ | 7 | Fatty acid esters | Food\|Endogenous\|Biological | - | - | - |
| 540.329 | 0.48503 | -1.0439 | 0.076519 | 1.7824 | HMDB0010397 | LysoPC (20:5 (5Z,8Z,11Z,14Z,17Z)/0:0) | [M-H]- | 36 | Glycerophosphocholines | Endogenous\|Food\|Biological | C04230 | map00564;<br>map05231 | Glycerophospholipid metabolism; Choline metabolism in cancer |
| 544.3388 | 0.30709 | -1.7033 | 0.009198 | 2.1457 | HMDB0010395 | LysoPC (20:4 (5Z,8Z,11Z,14Z)/0:0) | [M+H]+ | 2 | Glycerophosphocholines | Endogenous\|Food\|Biological | C04230 | map00564;<br>map05231 | Glycerophospholipid metabolism; Choline metabolism in cancer |
| 553.3832 | 2.6147 | 1.3866 | 0.048019 | 1.9002 | HMDB0033632 | Furohyperforin | [M+H]+ | 10 | Monoterpenoids | Endogenous\|Food\|Biological | - | - | - |

| | | | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| 565.0451 | 4.2373 | 2.0831 | 0.090551 | 1.7318 | HMDB0000286 | Uridine diphosphate glucose | [M-H]- | 5 | Pyrimidine nucleotide sugars | Endogenous\|Food\|Biological | C00029 | map00040; map00052; map00053; map00240; map00500; map00520; map00524; map00541; map00561; map00908; map01051; map01060; map01100; map01110; map01240 | Pentose and glucuronate interconversions; Galactose metabolism; Ascorbate and aldarate metabolism; Pyrimidine metabolism; Starch and sucrose metabolism; Amino sugar and nucleotide sugar metabolism; Neomycin, kanamycin and gentamicin biosynthesis; O-Antigen nucleotide sugar biosynthesis; Glycerolipid metabolism; Zeatin biosynthesis; Biosynthesis of ansamycins; Biosynthesis of plant secondary metabolites; Metabolic pathways; Biosynthesis of secondary metabolites; Biosynthesis of cofactors |
| 570.3544 | 0.2986 | -1.7437 | 0.007826 | 2.1603 | HMDB0010402 | LysoPC (22:5 (4Z,7Z, 10Z,13Z, 16Z)/0:0) | [M+H]+ | 2 | Glycerophosphocholines | Endogenous\|Food\|Biological | C04230 | map00564; map05231 | Glycerophospholipid metabolism; Choline metabolism in cancer |
| 590.3433 | 0.23028 | -2.1185 | 0.044356 | 1.9175 | HMDB0029334 | Nummularine B | [M-H]- | 76 | Amino acids, peptides, and analogues | Endogenous\|Food\|Biological | - | - | - |
| 594.2841 | 0.47768 | -1.0659 | 0.054713 | 1.8701 | HMDB0002581 | Taurocholic acid 3-sulfate | [M-H]- | 72 | Bile acids, alcohols and derivatives | Endogenous\|Food\|Biological | - | - | - |
| 609.5544 | 2.1783 | 1.1232 | 0.034531 | 1.9675 | HMDB0060057 | CE (15:0) | [M-H]- | 12 | Steroid esters | Food\|Endogenous\|Biological | - | - | - |
| 623.5026 | 4.8138 | 2.2672 | 0.013259 | 2.108 | HMDB0029958 | Asitribin | [M+H]+ | 23 | Fatty alcohols | Food\|Endogenous\|Biological | - | - | - |
| 625.3451 | 5.122 | 2.3567 | 0.084958 | 1.7515 | HMDB0034091 | Ponasteroside A | [M-H]- | 23 | Steroidal glycosides | Endogenous\|Food\|Biological | - | - | - |
| 645.2995 | 0.47979 | -1.0595 | 0.082563 | 1.7601 | HMDB0030736 | Capsianoside IV | [M+H]+ | 75 | Fatty acyl glycosides | Endogenous\|Food\|Biological | - | - | - |
| 648.4355 | 2.8519 | 1.5119 | 0.023982 | 2.0294 | HMDB0008856 | PE (14:1 (9Z)/ 15:0) | [M+H]+ | 38 | Glycerophosphoethanolamines | Endogenous\|Food\|Biological | - | - | - |

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|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| 662.1056 | 15.383 | 3.9432 | 0.040327 | 1.9373 | HMDB0000902 | NAD | [M-H]- | 6 | - | Endogenous | C00003 | map00190;<br>map00730;<br>map00760;<br>map00983;<br>map01100;<br>map01240;<br>map04152;<br>map04211;<br>map04212;<br>map04925;<br>map04977 | Oxidative phosphorylation; Thiamine metabolism; Nicotinate and nicotinamide metabolism; Drug metabolism - other enzymes; Metabolic pathways; Biosynthesis of cofactors; AMPK signaling pathway; Longevity regulating pathway; Longevity regulating pathway - worm; Aldosterone synthesis and secretion; Vitamin digestion and absorption |
| 675.5414 | 0.37932 | -1.3985 | 0.085516 | 1.7495 | HMDB0012097 | SM (d18:1/14:0) | [M+H]+ | 3 | Phosphosphingolipids | Endogenous\|Food\|Biological | - | - | - |
| 688.5332 | 2.9021 | 1.5371 | 0.037444 | 1.9521 | HMDB0114836 | PA (16:0/18:3 (6Z,9Z,12Z)) | [M+NH4]+ | 61 | Glycerophosphates | Food\|Endogenous\|Biological | - | - | - |
| 694.5228 | 0.27354 | -1.8702 | 0.045556 | 1.9118 | HMDB0012321 | Galactosylceramide (d18:1/14:0) | [M+Na]+ | 0 | Glycosphingolipids | Endogenous\|Food\|Biological | C02686 | - | - |
| 696.5389 | 3.1372 | 1.6495 | 0.02101 | 2.0491 | HMDB0009213 | PE (18:4 (6Z,9Z,12Z,15Z)/P-16:0) | [M+H]+ | 61 | Glycerophosphoethanolamines | Endogenous\|Food\|Biological | - | - | - |
| 703.5717 | 2.5709 | 1.3623 | 0.027878 | 2.0053 | HMDB0010169 | SM (d18:1/16:0) | [M+H]+ | 5 | Phosphosphingolipids | Endogenous\|Food\|Biological | - | - | - |
| 722.5062 | 0.38759 | -1.3674 | 0.088715 | 1.7382 | HMDB0009215 | PE (18:4 (6Z,9Z,12Z,15Z)/P-18:1 (11Z)) | [M+H]+ | 8 | Glycerophosphoethanolamines | Endogenous\|Food\|Biological | - | - | - |
| 730.5422 | 0.25198 | -1.9886 | 0.002753 | 2.2301 | HMDB0007874 | PC (14:0/18:2 (9Z,12Z)) | [M+H]+ | 5 | Glycerophosphocholines | Endogenous\|Food\|Biological | - | - | - |
| 732.5532 | 0.25701 | -1.9601 | 0.014425 | 2.0983 | HMDB0007872 | PC (14:0/18:1 (11Z)) | [M+H]+ | 1 | Glycerophosphocholines | Endogenous\|Food\|Biological | - | - | - |

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|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| 736.4773 | 0.47898 | -1.0619 | 0.073823 | 1.7925 | HMDB0008847 | PE (14:0/22:6 (4Z,7Z,10Z,13Z,16Z,19Z)) | [M+H]+ | 19 | Glycerophosphoethanolamines | Endogenous\|Food\|Biological | C00350 | map00563; map00564; map01100; map01110; map04136; map04138; map04140; map04723; map05130; map05167 | Glycosylphosphatidylinositol (GPI)-anchor biosynthesis; Glycerophospholipid metabolism; Metabolic pathways; Biosynthesis of secondary metabolites; Autophagy - other; Autophagy - yeast; Autophagy - animal; Retrograde endocannabinoid signaling; Pathogenic Escherichia coli infection; Kaposi sarcoma-associated herpesvirus infection |
| 744.5588 | 0.21351 | -2.2276 | 0.041524 | 1.9314 | HMDB0007938 | PC (15:0/18:1 (11Z)) | [M-H]- | 5 | Glycerophosphocholines | Endogenous\|Food\|Biological | C00157 | map00564; map00590; map00591; map00592; map01100; map01110; map04723; map05231 | Glycerophospholipid metabolism; Arachidonic acid metabolism; Linoleic acid metabolism; alpha-Linolenic acid metabolism; Metabolic pathways; Biosynthesis of secondary metabolites; Retrograde endocannabinoid signaling; Choline metabolism in cancer |
| 746.5048 | 0.38267 | -1.3858 | 0.009179 | 2.1459 | HMDB0112425 | PS (18:2 (9Z,12Z)/15:0) | [M+H]+ | 11 | Glycerophosphoserines | Food\|Endogenous\|Biological | - | - | - |
| 748.5233 | 2.1143 | 1.0802 | 0.084229 | 1.7541 | HMDB0005780 | PE (P-16:0/22:6 (4Z,7Z,10Z,13Z,16Z,19Z)) | [M+H]+ | 6 | Glycerophosphoethanolamines | Endogenous\|Food\|Biological | C00350 | map00563; map00564; map01100; map01110; map04136; map04138; map04140; map04723; map05130; map05167 | Glycosylphosphatidylinositol (GPI)-anchor biosynthesis; Glycerophospholipid metabolism; Metabolic pathways; Biosynthesis of secondary metabolites; Autophagy - other; Autophagy - yeast; Autophagy - animal; Retrograde endocannabinoid signaling; Pathogenic Escherichia coli infection; Kaposi sarcoma-associated herpesvirus infection |
| 749.524 | 0.47217 | -1.0826 | 0.037025 | 1.9543 | HMDB0030449 | 20-Deoxynarasin | [M+H]+ | 6 | Terpene glycosides | Endogenous\|Food\|Biological | - | - | - |
| 769.5023 | 2.3254 | 1.2175 | 0.019433 | 2.0601 | HMDB0010575 | PG (16:0/18:2 (9Z,12Z)) | [M+Na]+ | 4 | Glycerophosphoglycerols | Endogenous\|Food\|Biological | - | - | - |
| 772.5854 | 0.27789 | -1.8474 | 0.082765 | 1.7594 | HMDB0007946 | PC (15:0/20:2 (11Z,14Z)) | [M+H]+ | 0 | Glycerophosphocholines | Endogenous\|Food\|Biological | - | - | - |
| 776.5609 | 2.6157 | 1.3872 | 0.034865 | 1.9657 | HMDB0009644 | PE (22:5 (4Z,7Z,10Z,13Z,16Z)/P-18:1 (11Z)) | [M+H]+ | 3 | Glycerophosphoethanolamines | Endogenous\|Food\|Biological | - | - | - |

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|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| 779.53 | 0.16474 | -2.6018 | 0.054714 | 1.8701 | HMDB0115112 | PA (20:1(11Z)/22:4 (7Z,10Z,13Z,16Z)) | [M+H]+ | 37 | Glycerophosphates | Food\|Endogenous\|Biological | - | - | - |
| 780.5078 | 0.43494 | -1.2011 | 0.026025 | 2.0166 | HMDB0012313 | 3-O-Sulfogalactosylceramide (d18:1/16:0) | [M+H]+ | 27 | Glycosphingolipids | Endogenous\|Food\|Biological | C06125 | map00600;<br>map01100 | Sphingolipid metabolism;<br>Metabolic pathways |
| 780.5469 | 2.9935 | 1.5819 | 0.01826 | 2.0686 | HMDB0007890 | PC (14:0/22:5 (4Z,7Z,10Z,13Z,16Z)) | [M+H]+ | 9 | Glycerophosphocholines | Endogenous\|Food\|Biological | - | - | - |
| 794.5675 | 0.40669 | -1.298 | 0.015275 | 2.0914 | HMDB0007956 | PC (15:0/22:5 (4Z,7Z,10Z,13Z,16Z)) | [M+H]+ | 3 | Glycerophosphocholines | Endogenous\|Food\|Biological | - | - | - |
| 797.525 | 0.27068 | -1.8854 | 0.033184 | 1.9749 | HMDB0010581 | PG (16:0/22:4 (7Z,10Z,13Z,16Z)) | [M-H]- | 11 | Glycerophosphoglycerols | Endogenous\|Food\|Biological | - | - | - |
| 804.548 | 0.41074 | -1.2837 | 0.010467 | 2.1332 | HMDB0007889 | PC (14:0/22:4 (7Z,10Z,13Z,16Z)) | [M+Na]+ | 4 | Glycerophosphocholines | Endogenous\|Food\|Biological | - | - | - |
| 806.5619 | 0.27614 | -1.8565 | 0.032865 | 1.9766 | HMDB0004866 | LacCer (d18:1/12:0) | [M+H]+ | 1 | - | Endogenous\|Food\|Biological | - | - | - |
| 825.5572 | 0.22222 | -2.1699 | 0.033226 | 1.9746 | HMDB0061422 | PC (DiMe (9,3)/MonoMe (11,3)) | [M+H]+ | 7 | Glycerophosphocholines | Endogenous\|Food\|Biological | - | - | - |
| 826.5584 | 8.1981 | 3.0353 | 0.001533 | 2.2558 | HMDB0008189 | PC (18:3(6Z,9Z,12Z)/22:6 (4Z,7Z,10Z,13Z,16Z,19Z)) | [M-H]- | 23 | Glycerophosphocholines | Endogenous\|Food\|Biological | - | - | - |
| 840.5239 | 0.29165 | -1.7777 | 0.057932 | 1.8563 | HMDB0061555 | PS (DiMe (11,3)/DiMe (9,3)) | [M+H]+ | 26 | Glycerophosphoserines | Endogenous\|Food\|Biological | - | - | - |
| 854.6698 | 0.33573 | -1.5746 | 0.068492 | 1.8131 | HMDB0009278 | PE (20:1 (11Z)/24:1 (15Z)) | [M-H]- | 6 | Glycerophosphoethanolamines | Endogenous\|Food\|Biological | - | - | - |

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|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| 865.5056 | 5.0623 | 2.3398 | 0.08721 | 1.7435 | HMDB0116605 | PG (22:6 (4Z,7Z,10Z,13Z,16Z,19Z)/22:6 (4Z,7Z,10Z,13Z,16Z,19Z)) | [M-H]- | 4 | Glycerophosphoglycerols | Food\|Endogenous\|Biological | - | - | - |
| 889.5481 | 0.46291 | -1.1112 | 0.05578 | 1.8655 | HMDB0009785 | PI (16:0/20:0) | [M+Na]+ | 33 | Glycerophosphoinositols | Endogenous\|Food\|Biological | - | - | - |
| 913.5269 | 0.40101 | -1.3183 | 0.078158 | 1.7763 | HMDB0034650 | Soyasaponin II | [M+H]+ | 12 | Terpene glycosides | Endogenous\|Food\|Biological | C12081 | - | - |
| 952.5933 | 0.2021 | -2.3068 | 0.037455 | 1.952 | HMDB0061563 | PS (DiMe (11,5)/DiMe (13,5)) | [M+H]+ | 36 | Glycerophosphoserines | Endogenous\|Food\|Biological | - | - | - |
| 1151.703 | 0.1284 | -2.9613 | 0.069028 | 1.811 | HMDB0004844 | Ganglioside GM3 (d18:1/16:0) | [M-H]- | 2 | Glycosphingolipids | Endogenous\|Food\|Biological | C04730 | - | - |