**Supplementary Table 2: Plasma exposure of nonconstituent AAs following multiple administrations of AXA1665 14.7 g TID and AXA1665 4.9 g TID.**

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| --- | --- | --- | --- | --- |
| Nonconstituent AA | Mean ratio (Day 15/Day 1) | | | |
| **AXAhigh+ SOCcontrol + SOC**  **(group 1)**  (n = 9) | | Control + SOCAXAlow + SOC  **(group 2)**  (n = 7) | |
| Control TID | AXA1665 14.7 g TID | Control TID | AXA1665 4.9 g TID |
| Alanine (α-alanine) | 1.21 | 1.16 | 1.31 | 0.98 |
| β-Alanine | 0.93 | 1.13 | 1.94 | 0.90 |
| Alloisoleucine | 1.07 | 0.99 | 1.13 | 0.77 |
| α-aminoadipic acid | 0.88 | 1.21 | 1.10 | 0.92 |
| γ-aminobutyric acid | 0.70 | NC | NC | NC |
| β-aminoisobutyric acid | 0.75 | 0.93 | 0.84 | 0.69 |
| α-amino-N-butyric acid | 0.97 | 1.16 | 1.00 | 0.80 |
| Arginine | 1.10 | 1.01 | 1.24 | 0.81 |
| Argininosuccinic acid | 0.80b | 0.50c | 1.00a | 0.40a |
| Asparagine | 1.00 | 1.16 | 1.35 | 0.91 |
| Citrulline | 0.94 | 1.15 | 0.98 | 0.86 |
| Cystathionine | 0.62 | 0.79d | 1.24a | 1.27 |
| Cystine | 1.07 | 0.93 | 0.92 | 1.00 |
| Glutamic acid | 1.04 | 1.05 | 1.06 | 0.62 |
| Glutamine | 1.01 | 1.17 | 1.23 | 0.88 |
| Glycine | 1.02 | 0.86 | 1.08 | 0.79 |
| Homocitrulline | 0.29e | 0.92f | 0.39c | 0.21a |
| Homocystine | NC | NC | NC | 0.00g |
| Hydroxylysine | 0.90 | 1.26 | 0.96 | 0.93 |
| Hydroxyproline | 0.81 | 1.04 | 0.84 | 0.92 |
| Methionine | 0.99 | 0.99 | 1.27 | 0.90 |
| Phenylalanine | 0.87 | 1.08 | 1.15 | 0.86 |
| Proline | 1.00 | 0.98 | 1.18 | 0.90 |
| Sarcosine | 0.90 | 0.74 | 0.96 | 0.75 |
| Serine | 0.91 | 0.94 | 1.07 | 0.76 |
| Taurine | 0.91 | 0.80 | 1.17 | 0.92 |
| Tryptophan | 1.00 | 1.08 | 1.27 | 0.83 |
| Tyrosine | 1.02 | 0.96 | 1.27 | 0.95 |
| Day 15 plasma exposures are normalized to Day 1 and expressed as arithmetic mean ratios.  SOC included 30–40 minutes of mandatory and supervised physical activity, standardized meals, and a daily late evening snack.  an = 6.  bn = 5.  cn = 4.  dn = 8.  en = 7.  fn = 2.  gn = 1.  AA, amino acid; AXAhigh, AXA1665 14.7 g TID; AXAlow, AXA1665 4.9 g TID; NC, not calculable; SOC, standard of care; TID, 3 times a day. | | | | |
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