**Supplementary Table 4: Plasma ammonia AUC0–5 h and Cmax on Day 1 and Day 15 of Part 2.**

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| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Plasma ammonia (µM)** | | | | | | | | | | | | |
|  | **AXAhigh+ SOCcontrol + SOC**  **(group 1)** | | | | | | **Control + SOCAXAlow + SOC**  **(group 2)** | | | | | |
|  | **AXA1665 14.7 g TID** | | | **Control TID** | | | **Control TID** | | | **AXA1665 4.9 g TID** | | |
| **Time** | **n** | **AUC0–5 h (μM\*h)** | **Cmax (μM)** | **n** | **AUC0–5 h (μM\*h)** | **Cmax (μM)** | **n** | **AUC0–5 h (μM\*h)** | **Cmax (μM)** | **n** | **AUC0–5 h (μM\*h)** | **Cmax (μM)** |
| Day 1 | 9 | 324 ± 203 | 81.2 ± 47.8 | 9 | 384 ± 218 | 100 ± 66.5 | 7 | 385 ± 258 | 91.3 ± 57.2 | 7 | 344 ± 112 | 82.4 ± 25.3 |
| Day 15 | 9 | 292 ± 246 | 77.4 ± 65.6 | 9 | 347 ± 225 | 90.1 ± 54.6 | 6 | 321 ± 75.1 | 78.3 ± 27.4 | 7 | 235 ± 45.5 | 59.7 ± 12.0 |
| Ratio:  Day 15 to  Day 1 |  | 0.90 | 0.95 |  | 0.90 | 0.90 |  | 0.83 | 0.85 |  | 0.68 | 0.72 |
| Data are presented as mean ± SD unless otherwise stated.  SOC included 30–40 minutes of mandatory and supervised physical activity, standardized meals, and a daily late evening snack.  AUC0–5 h, area under the plasma concentration-time curve from time 0 to 5 hours; AXAhigh, AXA1665 14.7 g TID; AXAlow, AXA1665 4.9 g TID; Cmax, peak plasma concentration; SD, standard deviation; SOC, standard of care; TID, 3 times a day. | | | | | | | | | | | | |