### Concentration (pmol/mg of protein)

| hiPSC Line | NT     | ELIGLUSTAT | IBIGLUSTAT |
|------------|--------|------------|------------|
| Control    | 0.16   | 0.42       | 0.35       | 0.12 | 0.25 | 0.14 | 0.13 | 0.23 | 0.13 |
| GD2a-1     | 27.83  | 20.33      | 45.23      | 4.24 | 0.86 | 8.27 | 1.90 | 5.47 | 13.47 |
| GD2a-2     | 29.69  | 36.62      | 42.17      | 8.15 | 2.94 | 6.26 | 13.08 | 4.55 | 3.20 |
| GD2b-1     | 5.56   | 4.70       | 10.52      | 0.46 | 0.49 | 2.74 | 0.59 | 0.38 | 2.12 |
| GD2b-2     | 5.13   | 2.37       | 3.46       | 0.66 | 0.56 | 0.10 | 0.76 | 1.18 | 0.49 |
| GD3a-1     | 3.35   | 1.76       | 3.54       | 0.35 | 0.00 | 1.18 | 0.06 | 0.00 | 0.74 |
| GD3a-2     | 4.62   | 3.17       | 3.08       | 1.29 | 0.64 | 1.37 | 0.39 | 0.66 | 0.30 |
| GD1        | 1.94   | 1.12       | 0.19       | 0.25 | 0.10 | 0.10 | 0.26 | 0.08 | 0.09 |

Average concentration of NT Control a = 0.31

### Fold change in Concentration

| hiPSC Line | NT  | ELIGLUSTAT | IBIGLUSTAT |
|------------|-----|------------|------------|
| Control    | 0.52| 1.35       | 1.13       | 0.39 | 0.81 | 0.45 | 0.42 | 0.74 | 0.42 |
| GD2a-1     | 89.77| 65.58      | 145.90     | 13.88 | 2.77 | 26.68 | 6.13 | 17.65 | 43.45 |
| GD2a-2     | 95.77| 118.13     | 136.03     | 26.29 | 9.48 | 20.19 | 42.19 | 14.88 | 10.32 |
| GD2b-1     | 17.94| 15.16      | 33.94      | 1.48 | 1.58 | 8.84 | 1.90 | 1.23 | 6.84 |
| GD2b-2     | 16.55| 7.65       | 11.16      | 2.13 | 1.81 | 0.32 | 2.45 | 3.81 | 1.58 |
| GD3a-1     | 10.81| 5.68       | 11.42      | 1.13 | 0.00 | 3.81 | 0.19 | 0.00 | 2.39 |
| GD3a-2     | 14.90| 10.23      | 9.94       | 4.16 | 2.06 | 4.42 | 1.26 | 2.13 | 0.97 |
| GD1        | 6.26 | 3.61       | 0.61       | 0.81 | 0.32 | 0.32 | 0.84 | 0.26 | 0.29 |

### Concentration (pmol/mg of protein)

| hiPSC Line | NT     | CARMOFUR  |
|------------|--------|-----------|
| Control    | 0.48   | 0.29      | 0.21      | 1.30 | 1.42 | 0.23 |
| GD2a-1     | 20.19  | 21.27     | 12.55     | 6.00 | 10.35 | 0.76 |
| GD2a-2     | 14.37  | 24.94     | 31.48     | 5.47 | 9.22 | 9.48 |
| GD2b-1     | 6.25   | 7.06      | 8.58      | 3.40 | 3.78 | 3.68 |
| GD2b-2     | 6.90   | 4.27      | 1.06      | 2.44 | 3.23 | 0.80 |
| GD3a-1     | 2.06   | 1.67      | 4.61      | 0.13 | 1.25 | 1.33 |

Average concentration of NT Control a = 0.33

### Fold change in Concentration

| hiPSC Line | NT  | CARMOFUR |
|------------|-----|----------|
| Control a  | 1.48| 0.89     | 0.64     | 3.97 | 4.33 | 0.71 |
| GD2a-1     | 61.66 | 64.98    | 38.34    | 18.33 | 31.63 | 2.32 |
| GD2a-2     | 43.90 | 76.19    | 96.17    | 16.71 | 26.17 | 28.96 |
| GD2b-1     | 19.09 | 21.57    | 26.21    | 10.39 | 11.55 | 11.24 |
| GD2b-2     | 21.08 | 9.34     | 3.24     | 7.45 | 9.87 | 2.44 |
| GD3a-1     | 6.29 | 5.10     | 14.08    | 0.38 | 3.82 | 4.06 |
## Concentration (pmol/mg of protein)

| hiPSC Line | NT  | ELIGLUSTAT | IBIGLUSTAT |
|------------|-----|------------|------------|
| Control    | 3.60| 5.62       | 3.71       | 4.01       | 7.76       | 3.69       | 4.72       | 9.13       | 3.56       |
| GD2a-1     | 4.96| 2.97       | 4.68       | 3.14       | 2.98       | 7.55       | 8.40       | 2.79       | 6.75       |
| GD2a-2     | 6.78| 6.06       | 6.48       | 4.49       | 5.16       | 8.32       | 8.31       | 3.83       | 7.72       |
| GD2b-1     | 7.91| 2.78       | 9.68       | 4.44       | 2.23       | 10.63      | 5.49       | 2.95       | 8.21       |
| GD2b-2     | 3.67| 3.27       | 3.36       | 3.09       | 2.19       | 4.29       | 3.66       | 3.05       | 6.56       |
| GD3a-1     | 3.28| 3.15       | 4.71       | 2.56       | 2.79       | 5.12       | 2.56       | 3.27       | 6.09       |
| GD3a-2     | 12.61| 10.17     | 8.66       | 6.29       | 7.35       | 8.75       | 5.12       | 10.47      | 8.94       |
| GD1        | 12.85| 5.89      | 4.07       | 4.71       | 3.32       | 3.96       | 6.62       | 5.08       | 4.02       |

Average concentration of NT Control = 4.31

## Fold change in Concentration

| hiPSC Line | NT  | ELIGLUSTAT | IBIGLUSTAT |
|------------|-----|------------|------------|
| Control    | 0.84| 1.30       | 0.86       | 0.93       | 1.80       | 0.86       | 1.09       | 2.12       | 0.83       |
| GD2a-1     | 1.15| 0.69       | 1.09       | 0.73       | 0.69       | 1.75       | 1.95       | 0.65       | 1.57       |
| GD2a-2     | 1.57| 1.41       | 1.50       | 1.04       | 1.20       | 1.93       | 1.93       | 0.89       | 1.79       |
| GD2b-1     | 1.83| 0.65       | 2.24       | 1.03       | 0.52       | 2.47       | 1.27       | 0.68       | 1.91       |
| GD2b-2     | 0.85| 0.76       | 0.78       | 0.72       | 0.51       | 1.00       | 0.85       | 0.71       | 1.52       |
| GD3a-1     | 0.76| 0.73       | 1.09       | 0.59       | 0.65       | 1.19       | 0.59       | 0.76       | 1.41       |
| GD3a-2     | 2.92| 2.36       | 2.01       | 1.46       | 1.71       | 2.03       | 1.19       | 2.43       | 2.07       |
| GD1        | 2.98| 1.37       | 0.94       | 1.09       | 0.77       | 0.92       | 1.54       | 1.18       | 0.93       |
### GcCer1t8 1/16:0

| NIPSC Line | Concentration (pmol of protein) | Fold change in Concentration |
|------------|---------------------------------|-----------------------------|
|            | NT | ELUGLSTAT | IBIGLSTAT | NT | ELUGLSTAT | IBIGLSTAT |
| Control    | 8.75 | 17.41 | 7.47 | 0.38 | 0.30 | 0.17 | 0.15 | 0.28 | 0.40 |
| GD2a-1     | 23.20 | 22.32 | 17.46 | 1.43 | 2.96 | 1.62 | 3.95 | 2.88 | 2.07 |
| GD2a-2     | 10.85 | 16.22 | 17.89 | 0.46 | 0.72 | 0.76 | 1.70 | 1.25 | 3.66 |
| GD2b-1     | 20.17 | 18.56 | 23.93 | 0.26 | 0.31 | 0.33 | 0.39 | 0.48 | 0.31 |
| GD2b-2     | 19.59 | 34.31 | 19.81 | 0.12 | 0.23 | 1.35 | 0.37 | 0.64 | 0.50 |
| GD3a-1     | 21.14 | 34.06 | 12.40 | 0.37 | 0.85 | 0.25 | 0.92 | 1.24 | 0.54 |
| GD3a-2     | 35.95 | 13.48 | 24.93 | 0.16 | 0.33 | 0.21 | 0.27 | 0.36 | 0.31 |
| GD1        | 25.73 | 6.64 | 16.48 | 0.38 | 0.21 | 0.14 | 0.83 | 0.62 | 0.52 |

**Average concentration of NT Control a = 11.21**

### GcCer1t8 1/16:0

| NIPSC Line | Concentration (pmol of protein) | Fold change in Concentration |
|------------|---------------------------------|-----------------------------|
|            | NT | ELUGLSTAT | IBIGLSTAT | NT | ELUGLSTAT | IBIGLSTAT |
| Control    | 2.18 | 4.50 | 1.84 | 0.20 | 0.30 | 0.17 | 0.15 | 0.28 | 0.16 |
| GD2a-1     | 4.15 | 23.2 | 4.93 | 0.72 | 1.09 | 0.55 | 2.38 | 0.94 | 0.89 |
| GD2a-2     | 6.21 | 13.34 | 10.77 | 0.40 | 1.09 | 1.11 | 0.90 | 1.90 | 3.91 |
| GD2b-1     | 5.24 | 3.68 | 6.17 | 0.13 | 0.07 | 0.87 | 0.14 | 0.19 | 0.25 |
| GD2b-2     | 3.57 | 0.73 | 3.31 | 0.12 | 0.09 | 0.55 | 0.13 | 0.37 | 0.35 |
| GD3a-1     | 2.56 | 3.35 | 5.14 | 0.07 | 0.12 | 0.30 | 0.21 | 0.33 | 0.26 |
| GD3a-2     | 9.28 | 4.48 | 4.88 | 0.18 | 0.49 | 0.34 | 0.12 | 0.31 | 0.36 |
| GD1        | 6.27 | 3.82 | 4.47 | 0.14 | 0.12 | 0.14 | 0.36 | 0.30 | 0.37 |
| GD1        | 2.19 | 3.13 | 1.56 | 0.05 | 0.04 | 0.05 | 0.13 | 0.10 | 0.13 |

**Average concentration of NT Control a = 2.87**

### GcCer1t8 1/12:4

| NIPSC Line | Concentration (pmol of protein) | Fold change in Concentration |
|------------|---------------------------------|-----------------------------|
|            | NT | ELUGLSTAT | IBIGLSTAT | NT | ELUGLSTAT | IBIGLSTAT |
| Control    | 2.04 | 4.32 | 2.03 | 0.15 | 0.30 | 0.17 | 0.15 | 0.28 | 0.16 |
| GD2a-1     | 6.62 | 5.25 | 4.49 | 0.74 | 1.24 | 0.30 | 1.79 | 1.21 | 0.74 |
| GD2a-2     | 7.65 | 12.21 | 11.23 | 0.18 | 0.51 | 0.57 | 0.84 | 1.04 | 2.20 |
| GD2b-1     | 4.69 | 3.52 | 9.60 | 0.13 | 0.07 | 0.33 | 0.14 | 0.19 | 0.25 |
| GD2b-2     | 5.15 | 10.41 | 4.43 | 0.12 | 0.09 | 0.45 | 0.13 | 0.38 | 0.12 |
| GD3a-1     | 3.37 | 3.14 | 9.88 | 0.07 | 0.12 | 0.48 | 0.22 | 0.10 | 0.62 |
| GD3a-2     | 8.66 | 4.47 | 4.80 | 0.16 | 0.21 | 0.34 | 0.12 | 0.31 | 0.36 |
| GD1        | 7.57 | 4.84 | 4.18 | 0.14 | 0.14 | 0.12 | 0.42 | 0.35 | 0.28 |

**Average concentration of NT Control a = 2.80**
## Table 1: Concentration (ppm) of NH3 vs. Effectiveness of Control

| Control | Concentration | ELIUG#LST | B#ll&LST |
|---------|---------------|-----------|-----------|
| Control | 10.54         | 15.88     | 8.05      |
| Control | 17.22         | 22.49     | 9.65      |
| Control | 14.70         | 15.18     | 9.54      |
| Control | 25.49         | 25.31     | 9.72      |
| Control | 24.32         | 33.75     | 9.68      |
| Control | 18.22         | 24.67     | 16.80     |
| Control | 30.14         | 14.01     | 10.85     |
| Control | 22.60         | 21.96     | 18.02     |

### Average Concentration of Control
- NH3: 12.92 ppm

## Table 2: Fold change in Concentration

| Control | Fold change in Concentration | ELIUG#LST | B#ll&LST |
|---------|------------------------------|-----------|-----------|
| Control | 0.85                        | 1.96      | 0.69      |
| Control | 0.92                        | 1.16      | 0.27      |
| Control | 1.21                        | 1.38      | 0.92      |
| Control | 0.99                        | 1.09      | 0.82      |
| Control | 0.14                        | 1.52      | 1.02      |
| Control | 0.48                        | 1.30      | 0.66      |

### Average Fold Change in Concentration
- 2.83 fold change

## Table 3: Concentration (ppm) of NH3 vs. Effectiveness of Control

| Control | Concentration | ELIUG#LST | B#ll&LST |
|---------|---------------|-----------|-----------|
| Control | 1.95          | 4.27      | 6.78      |
| Control | 2.92          | 3.09      | 1.16      |
| Control | 6.03          | 10.49     | 8.70      |
| Control | 3.70          | 4.23      | 14.01     |
| Control | 3.99          | 21.00     | 2.09      |
| Control | 2.93          | 4.02      | 2.87      |
| Control | 0.84          | 5.51      | 3.06      |
| Control | 4.08          | 3.35      | 3.72      |

### Average Concentration of Control
- 1.52 ppm

## Table 4: Fold change in Concentration

| Control | Fold change in Concentration | ELIUG#LST | B#ll&LST |
|---------|------------------------------|-----------|-----------|
| Control | 0.09                        | 1.81      | 0.80      |
| Control | 2.03                        | 1.03      | 0.49      |
| Control | 0.55                        | 0.77      | 0.79      |
| Control | 0.51                        | 0.22      | 0.35      |
| Control | 1.05                        | 0.39      | 1.01      |
| Control | 0.30                        | 0.09      | 0.38      |
| Control | 0.73                        | 0.73      | 0.63      |
| Control | 0.91                        | 0.91      | 0.82      |

### Average Fold Change in Concentration
- 0.43 fold change

## Table 5: Concentration (ppm) of NH3 vs. Effectiveness of Control

| Control | Concentration | ELIUG#LST | B#ll&LST |
|---------|---------------|-----------|-----------|
| Control | 4.05          | 9.37      | 4.13      |
| Control | 7.73          | 10.03     | 3.23      |
| Control | 15.22         | 18.65     | 18.57     |
| Control | 7.86          | 9.93      | 13.99     |
| Control | 9.36          | 35.73     | 9.60      |
| Control | 6.19          | 11.56     | 8.01      |
| Control | 17.16         | 9.48      | 9.08      |
| Control | 9.98          | 5.80      | 8.22      |

### Average Concentration of Control
- 5.85 ppm
| GlcSph(d18:1) | Sph(d18:1) | GlcCer(d18:1/16:0) | Cer(d18:1/16:0) |
|--------------|------------|-------------------|-----------------|
| **Concentration (pmol/mg of protein)** | **Concentration (pmol/mg of protein)** | **Concentration (pmol/mg of protein)** | **Concentration (pmol/mg of protein)** |
| Control a NT | Control a + CBE | Control a NT | Control a + CBE | Control a NT | Control a + CBE | Control a NT | Control a + CBE |
| 0.31 | 15.22 | 3.71 | 2.59 | 17.41 | 34.82 | 18.88 | 8.63 |
| 0.27 | 8.78 | 7.48 | 8.29 | 25.93 | 64.97 | 18.84 | 24.65 |
| 0.42 | 14.21 | 7.70 | 8.59 | 18.64 | 55.30 | 11.70 | 18.76 |
| **Average concentration of NT Control a** | 0.33 | **Average concentration of NT Control a** | 6.29 | **Average concentration of NT Control a** | 20.66 | **Average concentration of NT Control a** | 16.47 |
| **Fold change in Concentration** | **Fold change in Concentration** | **Fold change in Concentration** | **Fold change in Concentration** |
| Control a NT | Control a + CBE | Control a NT | Control a + CBE | Control a NT | Control a + CBE | Control a NT | Control a + CBE |
| 0.83 | 45.66 | 0.89 | 0.41 | 1.25 | 3.14 | 1.14 | 1.50 |
| 0.81 | 26.34 | 1.19 | 1.32 | 0.90 | 2.68 | 0.71 | 1.14 |
| **Average concentration of NT Control a** | 7.12 | **Average concentration of NT Control a** | 4.80 |
| **Fold change in Concentration** | **Fold change in Concentration** |
| Control a NT | Control a + CBE | Control a NT | Control a + CBE |
| 0.63 | 0.46 | 0.89 | 0.23 |
| 1.31 | 3.26 | 1.34 | 1.51 |
| 1.06 | 2.25 | 0.77 | 1.09 |
| **Average concentration of NT Control a** | 9.21 | **Average concentration of NT Control a** | 12.09 |
| **Fold change in Concentration** | **Fold change in Concentration** |
| Control a NT | Control a + CBE | Control a NT | Control a + CBE |
| 2.46 | 5.80 | 3.17 | 3.50 |
| 14.50 | 28.35 | 16.69 | 17.07 |
| 10.68 | 22.29 | 10.22 | 12.19 |
| **Average concentration of NT Control a** | 9.21 | **Average concentration of NT Control a** | 12.09 |
| **Fold change in Concentration** | **Fold change in Concentration** |
| Control a NT | Control a + CBE | Control a NT | Control a + CBE |
| 0.27 | 0.63 | 0.77 | 0.29 |
| 1.57 | 3.08 | 1.38 | 1.41 |
| 1.16 | 2.42 | 0.85 | 1.01 |
|                     | GlcSph(d18:1) | Sph(d18:1) | GlcCer(d18:1/16:0) | Cer(d18:1/16:0) |
|---------------------|--------------|------------|-------------------|----------------|
| **Concentration (pmol/mg of protein)** |              |            |                   |                |
| Control a NT        | 0.09         | 10.63      | 21.25             | 41.17          |
| Control a + CBE     | 0.38         | 0.62       | 34.84             | 34.22          |
|                     | 0.09         | 13.96      | 19.72             | 30.05          |
|                     | 0.38         | 2.39       | 18.55             | 30.62          |
|                     | 0.55         | 12.29      | 3.53              | 23.64          |
|                     | 0.96         | 18.85      | 3.84              | 34.22          |
| Average concentration of NT Control b = | 0.24         |            |                   |                |
|                     | 0.44         |            |                   |                |
| **Fold change in Concentration** |              |            |                   |                |
| Control a NT        | 3.77         | 43.84      | 1.08              | 1.31           |
| Control a + CBE     | 0.87         | 1.40       | 2.84              | 2.06           |
|                     | 0.37         | 57.60      | 0.95              | 0.95           |
|                     | 0.86         | 5.42       | 1.00              | 1.00           |
|                     | 2.26         | 50.72      | 0.96              | 0.75           |
|                     | 1.27         | 8.00       |                   |                |
| Average concentration of NT Control b = | 7.27         |            |                   |                |
|                     | 7.30         |            |                   |                |
| **GlCer(d18:1/18:0)** |              |            |                   |                |
| **Concentration (pmol/mg of protein)** |              |            |                   |                |
| Control a NT        | 8.49         | 17.14      | 9.51              | 5.36           |
| Control a + CBE     | 6.29         | 17.35      | 7.15              | 7.38           |
|                     | 7.92         | 22.69      | 5.22              | 7.82           |
| Average concentration of NT Control b = | 7.27         |            |                   |                |
|                     | 7.30         |            |                   |                |
| **Fold change in Concentration** |              |            |                   |                |
| Control a NT        | 1.17         | 2.36       | 1.33              | 1.01           |
| Control a + CBE     | 0.87         | 2.39       | 0.98              | 0.88           |
|                     | 0.97         | 3.12       | 0.72              | 1.07           |
| **GlCer(d18:1/24:1)** |              |            |                   |                |
| **Concentration (pmol/mg of protein)** |              |            |                   |                |
| Control a NT        | 6.97         | 14.08      | 12.60             | 8.84           |
| Control a + CBE     | 7.25         | 20.48      | 11.54             | 14.08          |
|                     | 9.60         | 27.96      | 10.96             | 14.54          |
| Average concentration of NT Control b = | 7.94         |            |                   |                |
|                     | 11.70        |            |                   |                |
| **Fold change in Concentration** |              |            |                   |                |
| Control a NT        | 0.88         | 1.77       | 1.08              | 0.58           |
| Control a + CBE     | 0.91         | 2.58       | 0.99              | 1.00           |
|                     | 0.91         | 3.52       | 0.94              | 1.24           |
| GlcSph(d18:1) | Sph(d18:1) | GlcCer(d18:1/16:0) | Cer(d18:1/16:0) |
|----------------|------------|-----------------|-----------------|
| **Concentration (pmol/mg of protein)** | **Concentration (pmol/mg of protein)** | **Concentration (pmol/mg of protein)** | **Concentration (pmol/mg of protein)** |
| Control a NT | Control a + CBE | Control a NT | Control a + CBE | Control a NT | Control a + CBE | Control a NT | Control a + CBE |
| 0.21 | 12.59 | 6.83 | 4.51 | 37.11 | 73.26 | 35.98 | 45.22 |
| 0.26 | 8.32 | 6.99 | 5.97 | 31.37 | 71.19 | 21.50 | 58.33 |
| **Average concentration of** | **NT Control c =** | **Average concentration of** | **NT Control c =** | **Average concentration of** | **NT Control c =** |
| 0.23 | 6.91 | 34.24 | 28.74 |

| GlcSph(d18:1) | Sph(d18:1) | GlcCer(d18:1/16:0) | Cer(d18:1/16:0) |
|----------------|------------|-----------------|-----------------|
| **Fold change in Concentration** | **Fold change in Concentration** | **Fold change in Concentration** | **Fold change in Concentration** |
| Control a NT | Control a + CBE | Control a NT | Control a + CBE | Control a NT | Control a + CBE | Control a NT | Control a + CBE |
| 0.89 | 54.68 | 0.99 | 0.65 | 1.08 | 2.14 | 1.25 | 1.57 |
| 1.11 | 36.14 | 1.01 | 0.86 | 0.92 | 2.08 | 0.75 | 2.03 |

| GlcCer(d18:1/16:0) | Cer(d18:1/16:0) |
|-------------------|-----------------|
| **Concentration (pmol/mg of protein)** | **Concentration (pmol/mg of protein)** |
| Control a NT | Control a + CBE | Control a NT | Control a + CBE | Control a NT | Control a + CBE |
| 18.64 | 29.20 | 10.62 | 16.01 |
| 20.47 | 47.35 | 19.95 | 23.58 |
| **Average concentration of** | **NT Control c =** | **Average concentration of** | **NT Control c =** |
| 19.56 | 15.28 |

| GlcCer(d18:1/16:0) | Cer(d18:1/16:0) |
|-------------------|-----------------|
| **Fold change in Concentration** | **Fold change in Concentration** |
| Control a NT | Control a + CBE | Control a NT | Control a + CBE | Control a NT | Control a + CBE |
| 0.95 | 1.49 | 0.69 | 1.05 |
| 1.05 | 2.42 | 1.31 | 1.54 |

| GlcCer(d18:1/24:1) | Cer(d18:1/24:1) |
|-------------------|-----------------|
| **Concentration (pmol/mg of protein)** | **Concentration (pmol/mg of protein)** |
| Control a NT | Control a + CBE | Control a NT | Control a + CBE | Control a NT | Control a + CBE |
| 18.16 | 28.08 | 21.55 | 26.94 |
| 16.45 | 32.55 | 26.40 | 31.22 |
| **Average concentration of** | **NT Control c =** | **Average concentration of** | **NT Control c =** |
| 17.31 | 23.97 |

| GlcCer(d18:1/24:1) | Cer(d18:1/24:1) |
|-------------------|-----------------|
| **Fold change in Concentration** | **Fold change in Concentration** |
| Control a NT | Control a + CBE | Control a NT | Control a + CBE | Control a NT | Control a + CBE |
| 1.05 | 1.62 | 0.90 | 1.12 |
| 0.95 | 1.88 | 1.10 | 1.30 |