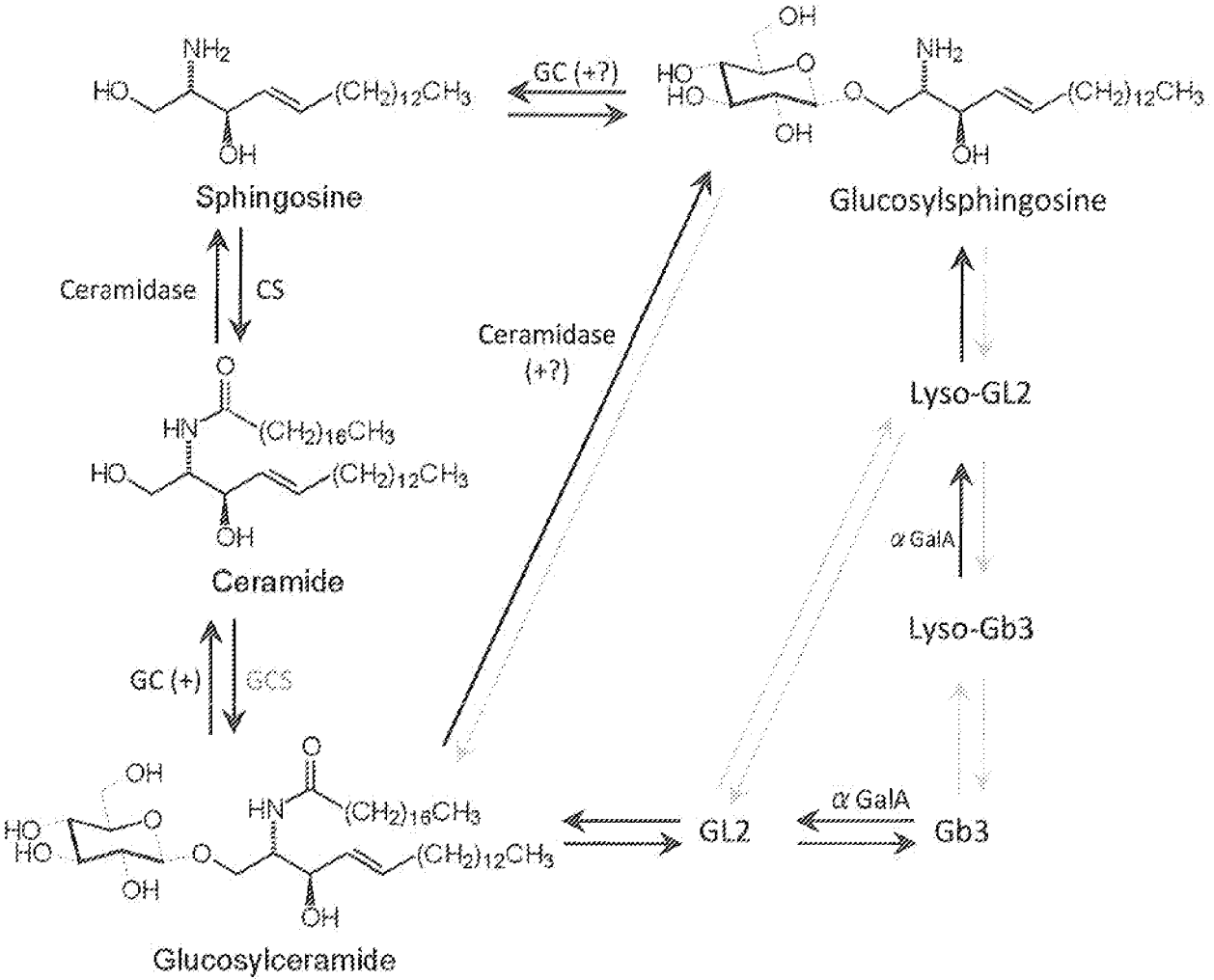
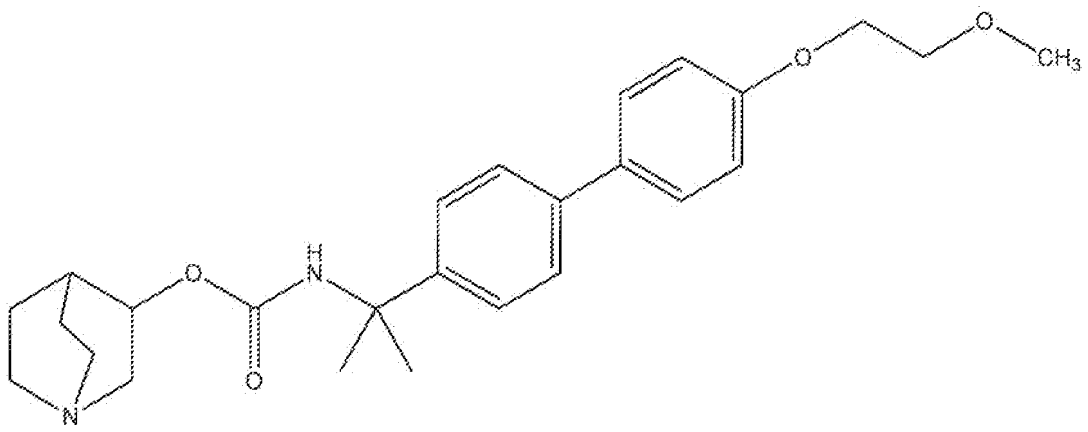

Sphingosine
GC (+?)
Glucosylsphingosine
Ceramidase
CS
Ceramidase
(+?)
Lyso-GL2
Ceramide
α GalA
Lyso-Gb3
GC (+)
GCS
GL2
α GalA
Gb3
Glucosylceramide

Figure 1



(*S*)-Quinuclidin-3-yl (2-(4'-(2-methoxyethoxy)-[1,1'-biphenyl]-4-yl)propan-2-yl)carbamate

Figure 2

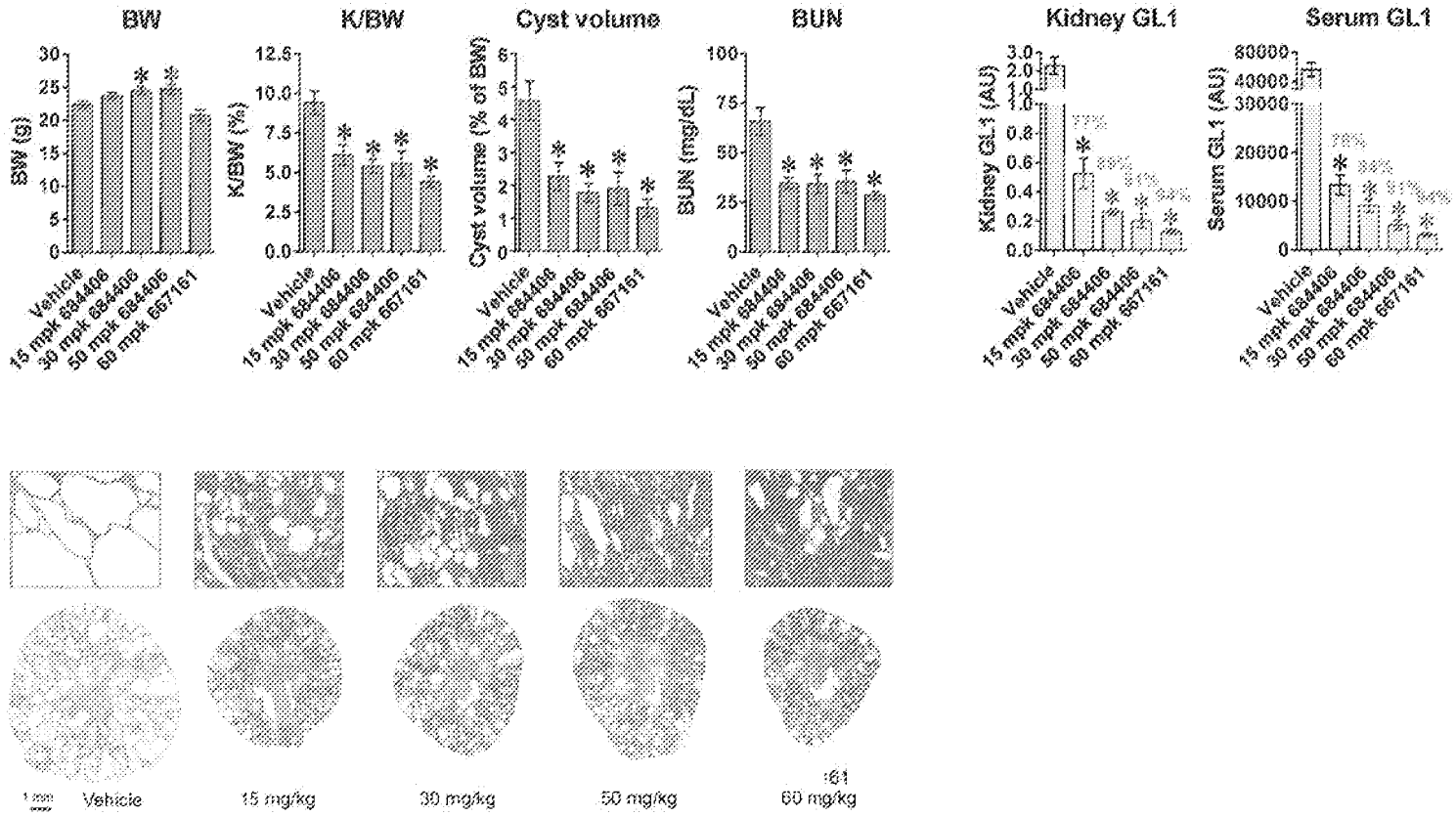

BW
BW (g)
K/BW
K/BW (%)
Cyst volume
Cyst volume (% of BW)
BUN
BUN (mg/dL)
Kidney GL1
Kidney GL1 (AU)
Serum GL1
Serum GL1 (AU)
Vehicle
15 mpk 684406
30 mpk 684406
50 mpk 684406
60 mpk 667161
1 cm
Vehicle
15 mg/kg
30 mg/kg
50 mg/kg
60 mg/kg

Figure 3