Figure 1

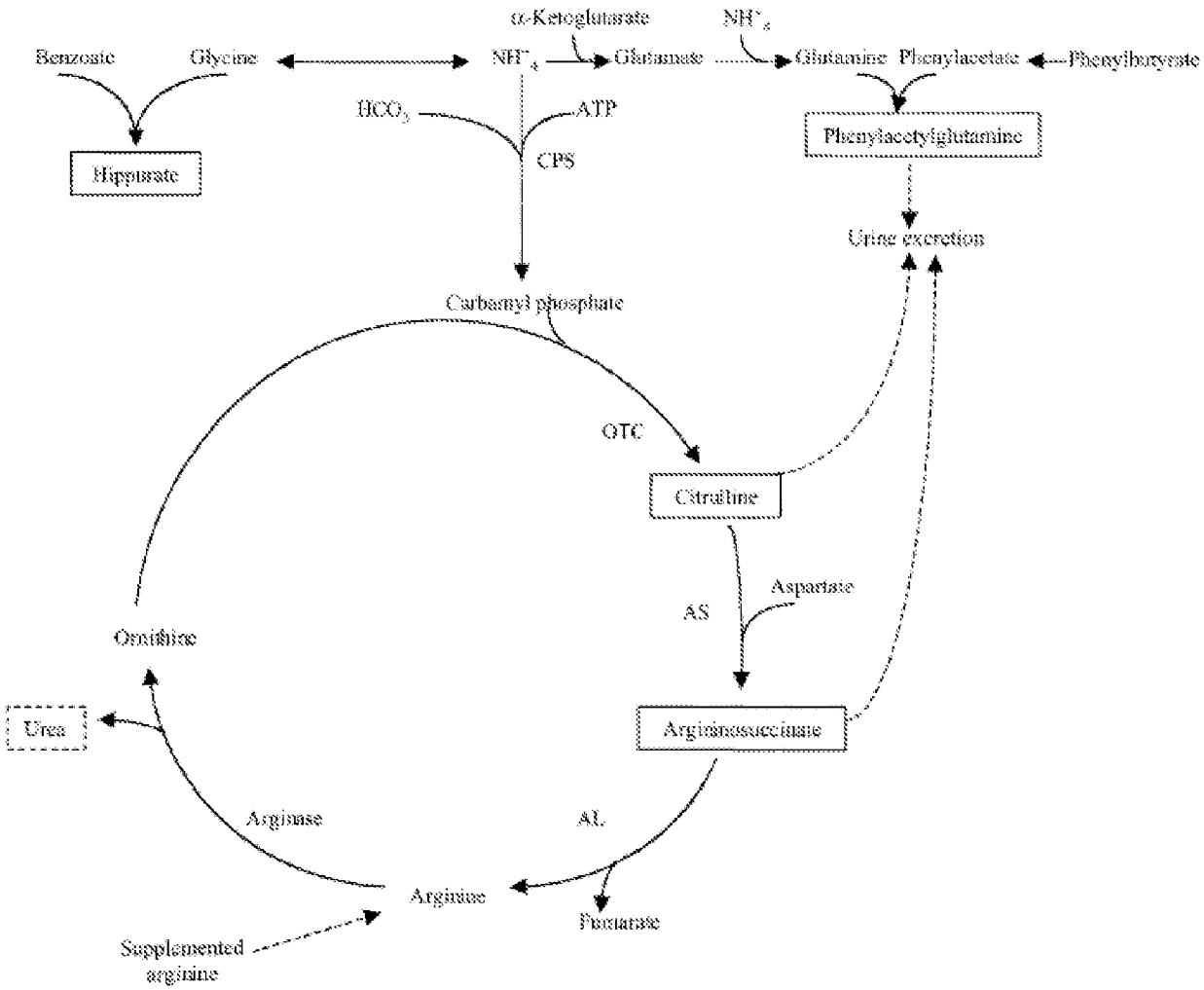

α-Ketoglutarate
NH^+_4
Benzoate
Glycine
NH^+_4
Glutamate
Glutamine
Phenylacetate
Phenylbutyrate
HCO_3
ATP
CPS
Hippurate
Phenylacetylglutamine
Urine excretion
Carbamyl phosphate
OTC
Citrulline
Aspartate
AS
Ornithine
Urea
Argininosuccinate
Arginase
AL
Arginine
Fumarate
Supplemented arginine

Figure 2A

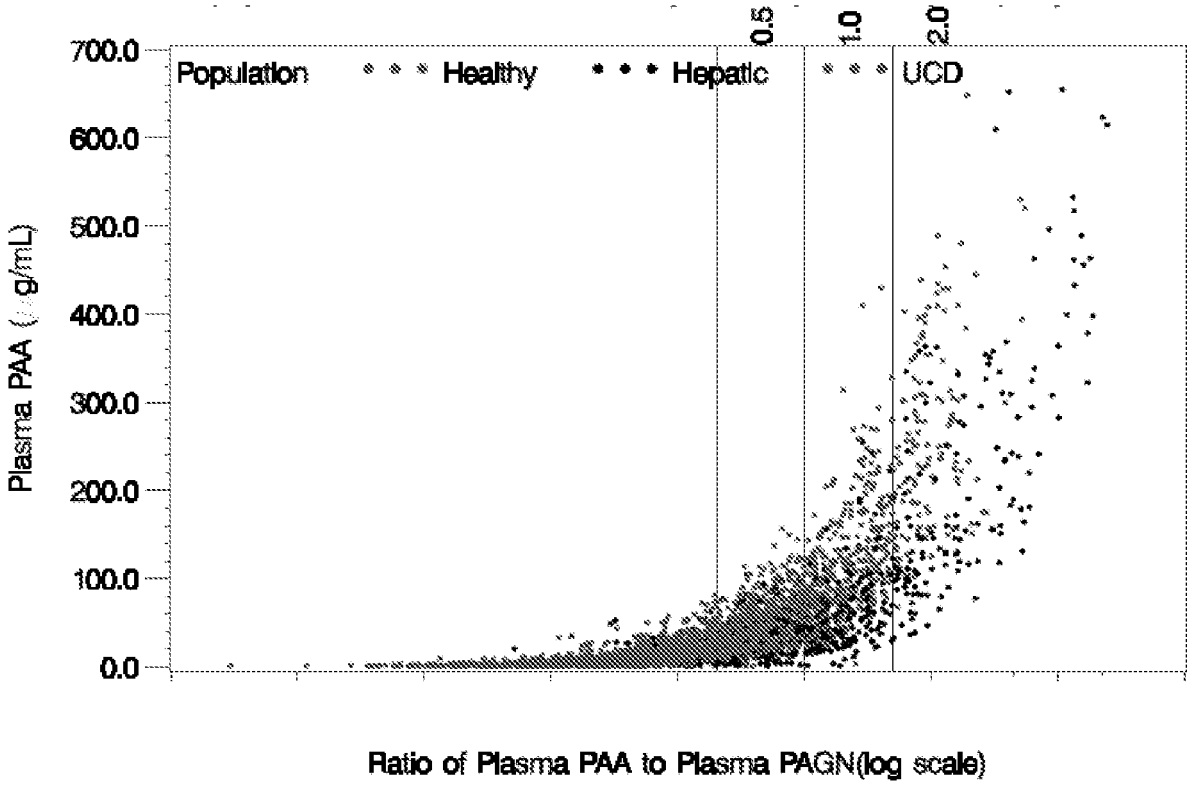

0.5
1.0
2.0
Population
Healthy
Hepatic
UCD
700.0
600.0
500.0
400.0
300.0
200.0
100.0
0.0
Plasma PAA (g/mL)
Ratio of Plasma PAA to Plasma PAGN(log scale)

Figure 2B

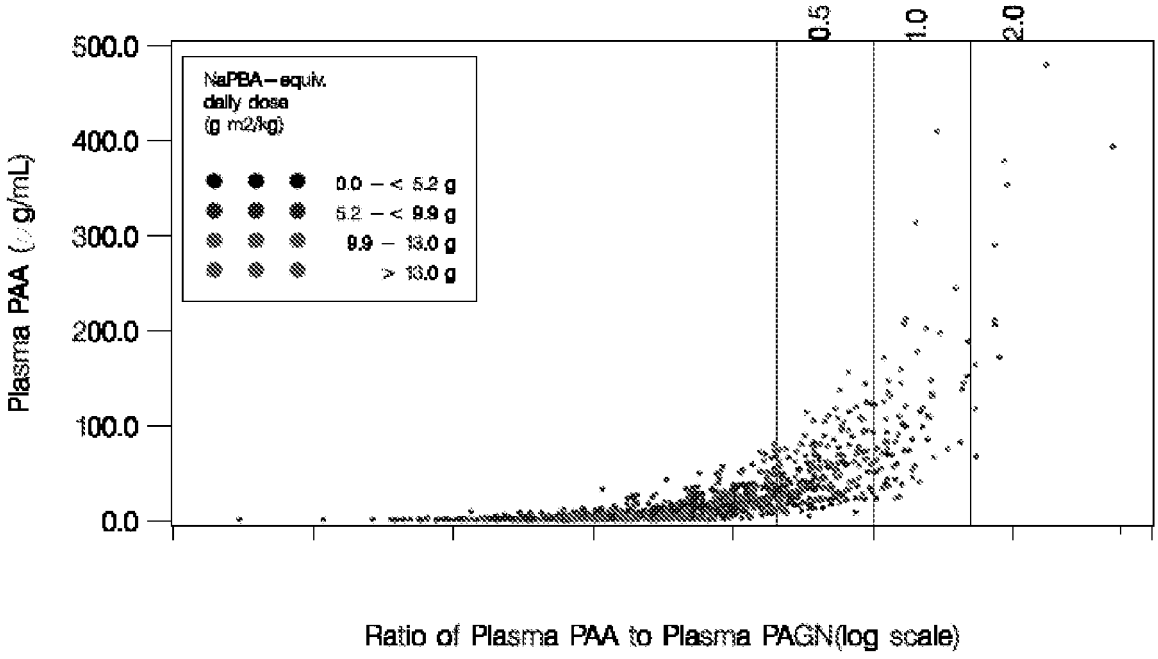

0.5
1.0
2.0
500.0
400.0
300.0
200.0
100.0
0.0
Plasma PAA (µg/mL)
NaPBA–equiv.
daily dose
(g m2/kg)
0.0 – < 5.2 g
5.2 – < 9.9 g
9.9 – 13.0 g
> 13.0 g
Ratio of Plasma PAA to Plasma PAGN(log scale)

Figure 2C

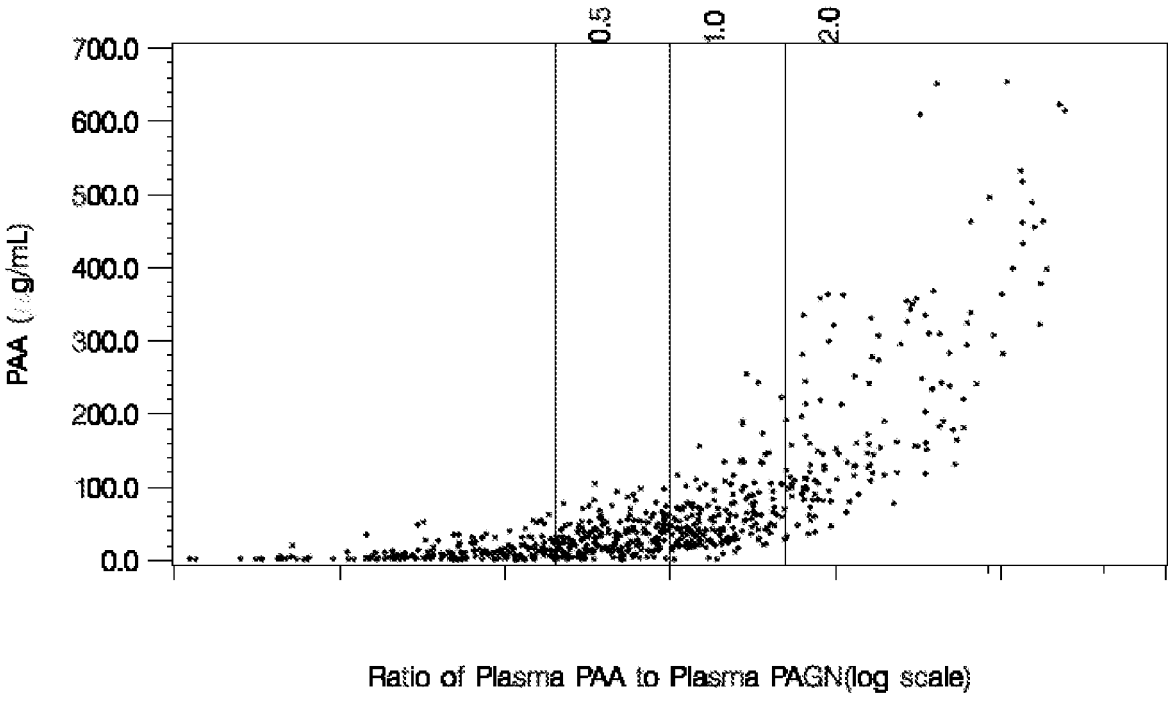
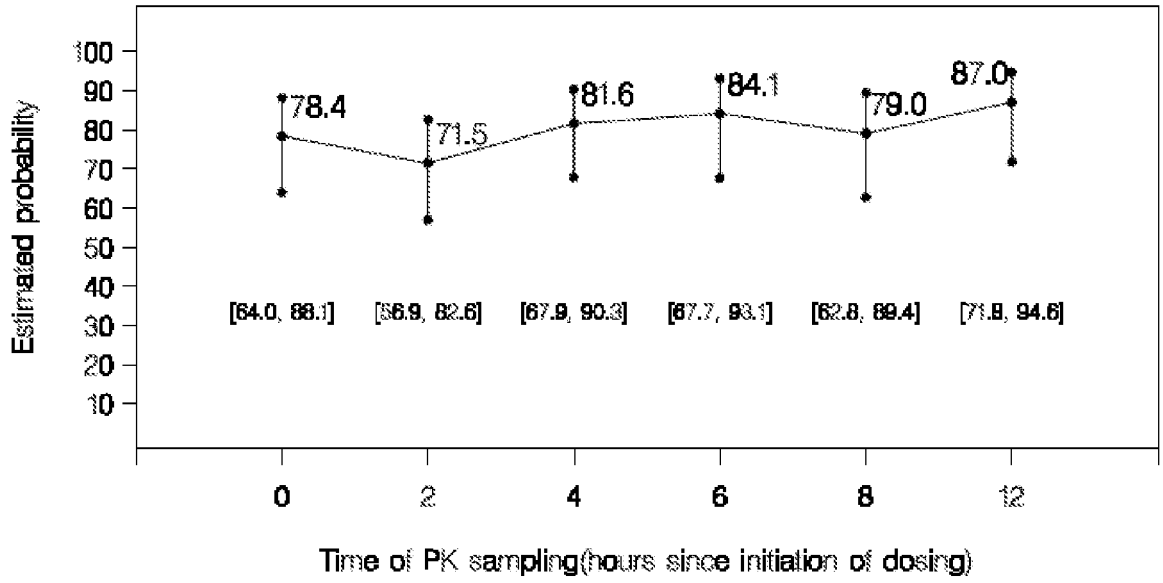

0.5
1.0
2.0
700.0
600.0
500.0
400.0
300.0
200.0
100.0
0.0
PAA (μg/mL)
Ratio of Plasma PAA to Plasma PAGN(log scale)

Figure 3



t=2 hrs signif. less than t=0(p=0.036), t=4(p=0.032), and t=6(p=0.017)
No other time differences statistically significant. Time=10 omitted due to too few observations

Figure 4

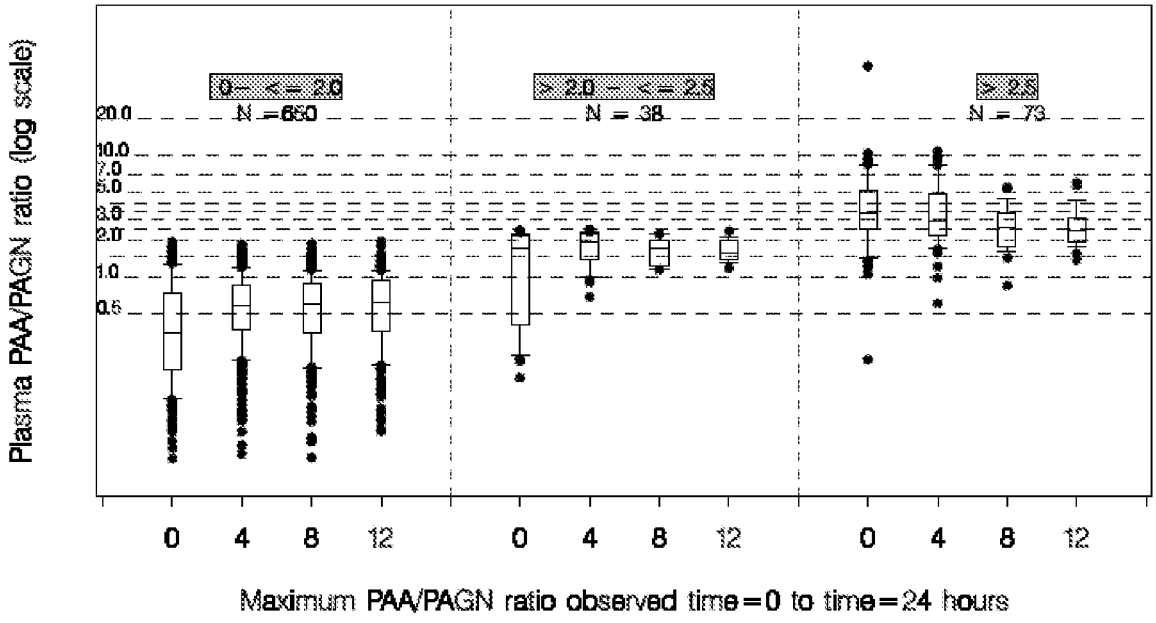

Plasma PAA/PAGN ratio (log scale)
0 - < = 2.0
> 2.0 - < = 2.5
> 2.5
N = 650
N = 38
N = 73
20.0
10.0
7.0
5.0
3.5
2.0
1.0
0.5
0
4
8
12
Maximum PAA/PAGN ratio observed time = 0 to time = 24 hours

Figure 5A

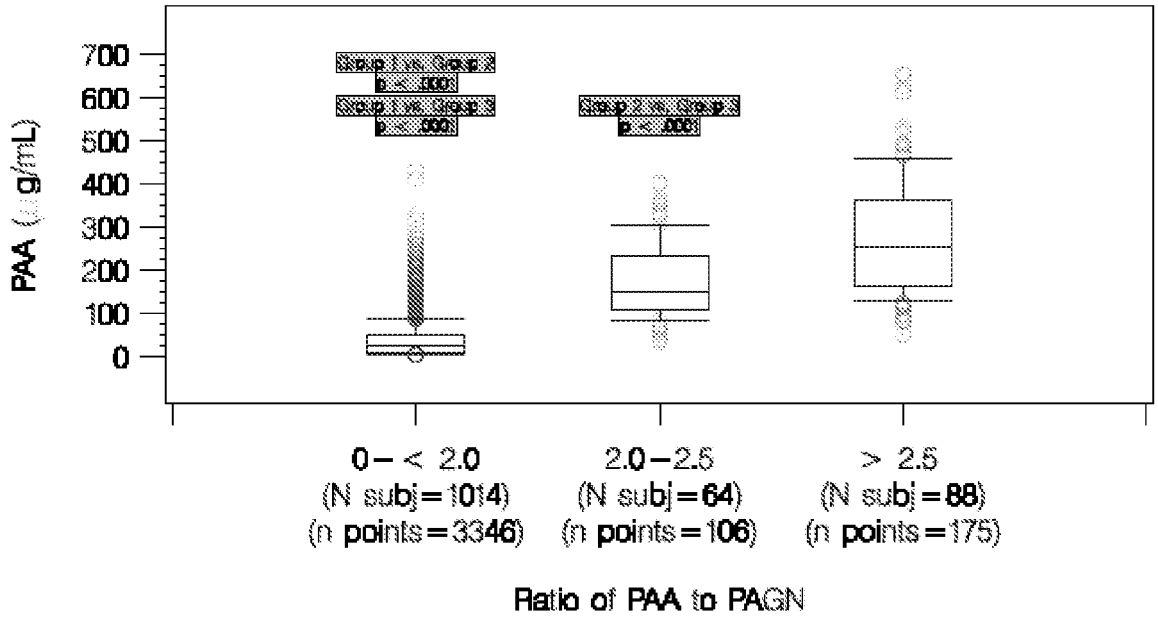


Figure 5B

