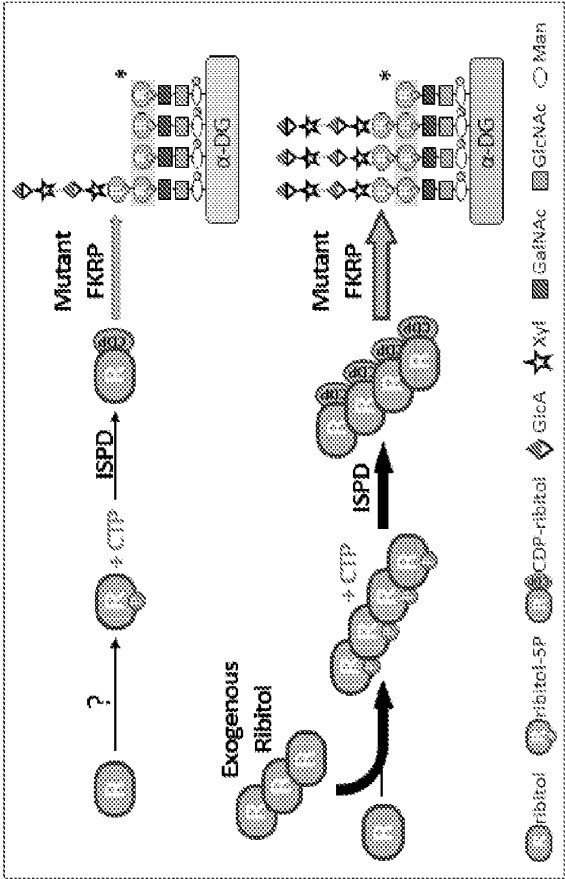


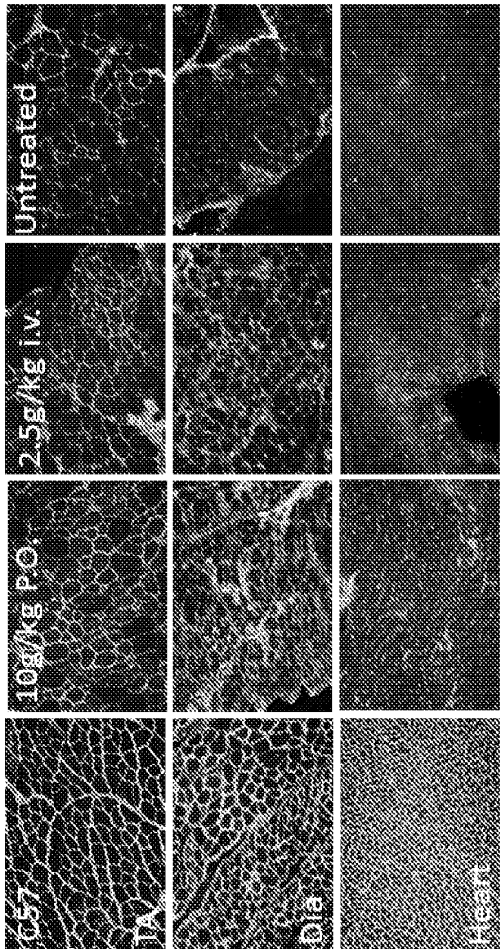
FIG. 1
Model for Ribitol-induced Functional Glycosylation
of α -DG in FKRP Mutant Cells



? = mechanism is not fully understood; "*" = the first ribitol-5-phosphate on the Core M3 of μ -DG is transferred by fukutin using CDP-ribitol as the donor substrate; CTP = Cytidine Triphosphate; D-Glucuronic acid (GlcA); Xylitol (Xyl); N-Acetyl-D-galactosamine (GalNAc); N-Acetyl-D-glucosamine (GlcNAc); D-Mannose (Man)
Source: [Cataldi et al. 2018](#)

FIG. 2

Detection of Matriglycan 1 Month after Ribitol
Treatment in P448L FKRP Mutant Mice



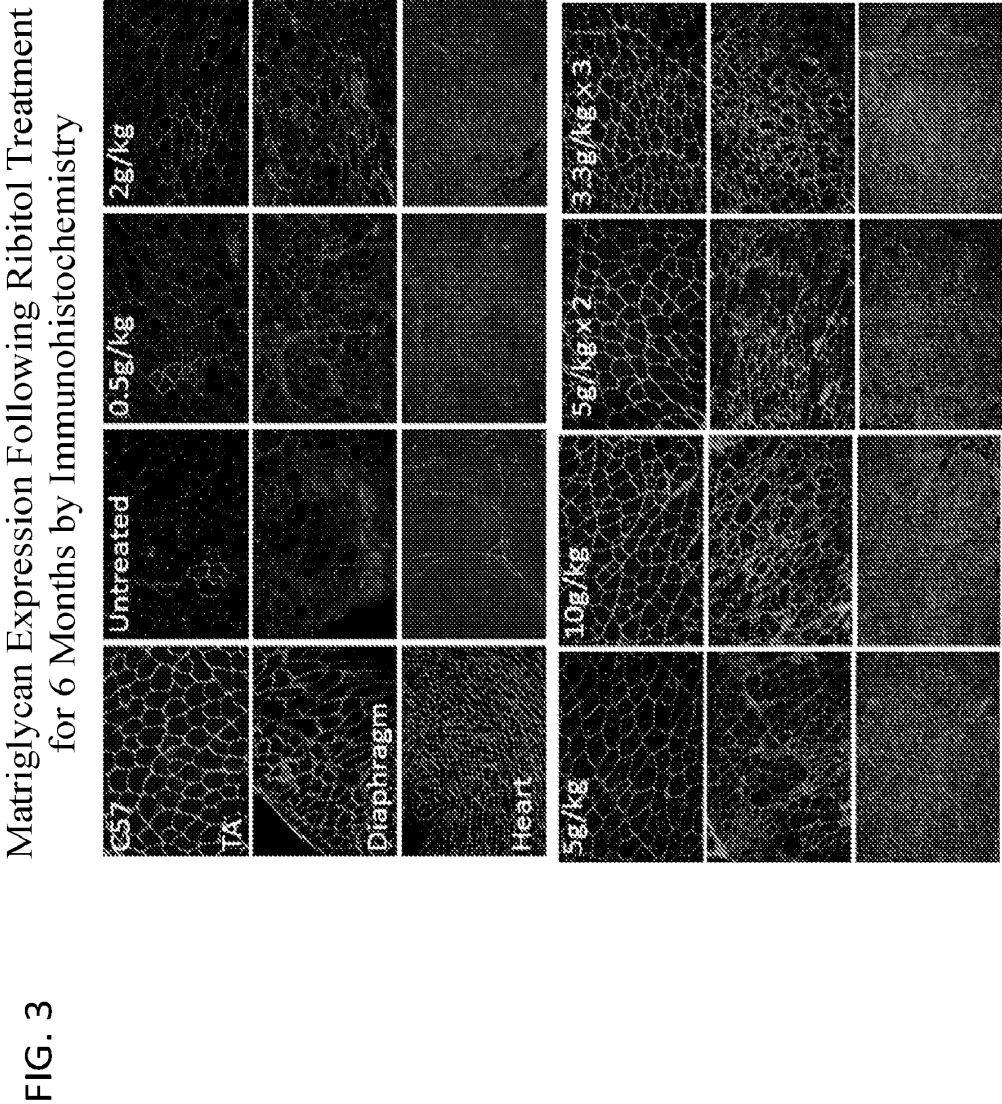
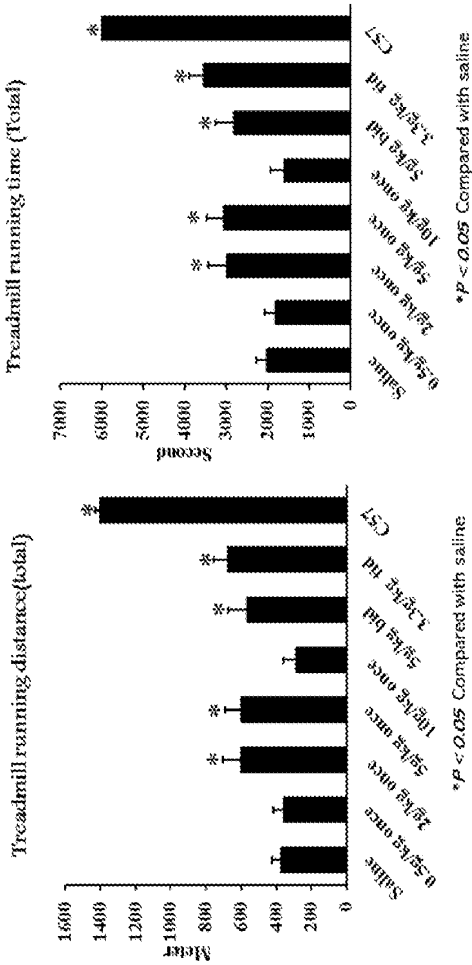


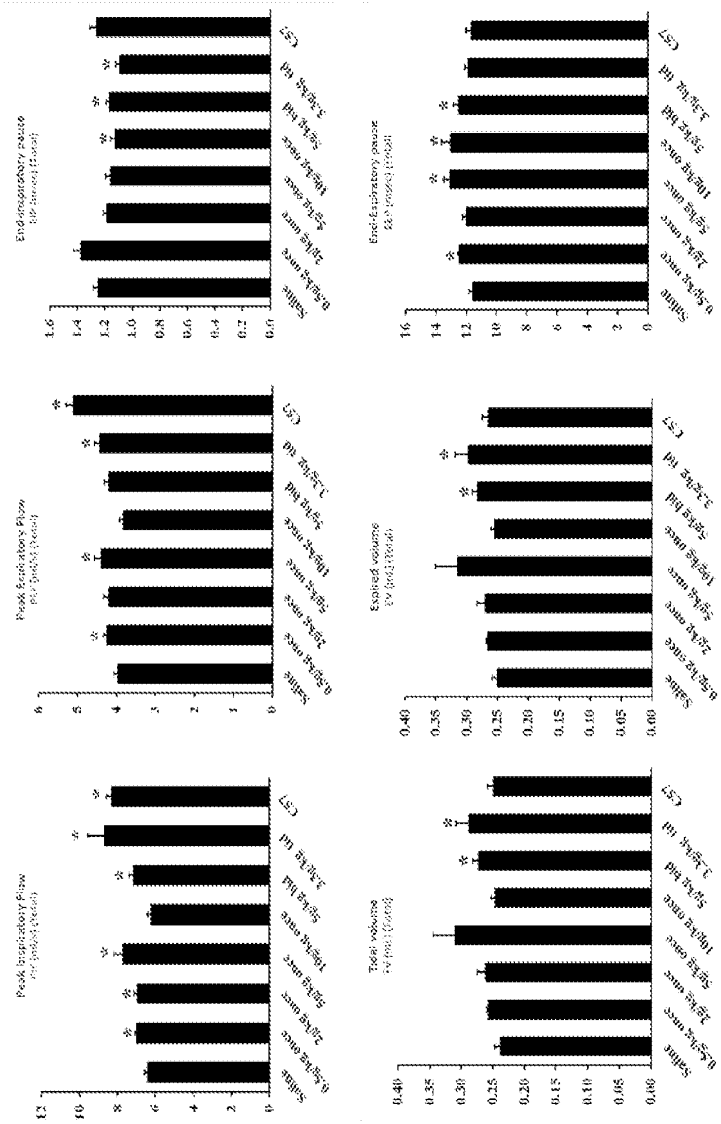
FIG. 4

Treadmill Exhaustion Test Results for P448L FKRP Mutant Mice with 6 Months of Ribitol Treatment



Statistical significance was assessed using an unpaired t test. * P≤0.05

FIG. 5 Whole-Body Plethysmography Parameters of P448K FKR^P Mutant Mice Following 6-months of Ribitol Treatment



Statistical significance was assessed using an unpaired t test. * P<0.05

FIG. 6

Serum Creatine Kinase Levels after Ribitol Treatment for 6 Months

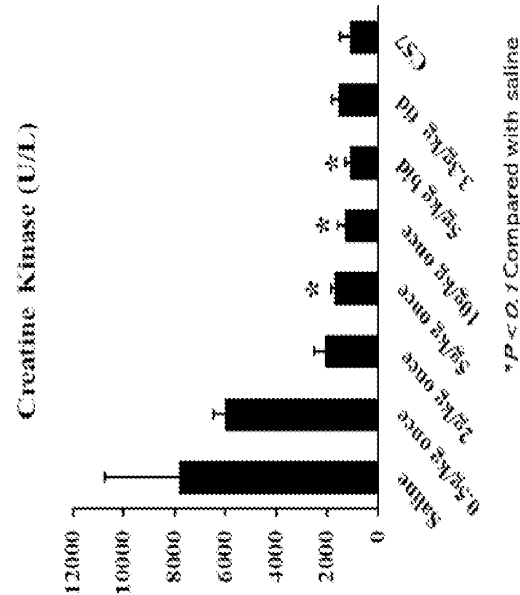


FIG. 7

Body Weight and Treadmill Exhaustion Test in L276I
FKRP Mutant Mice Treated with Ribitol or Saline

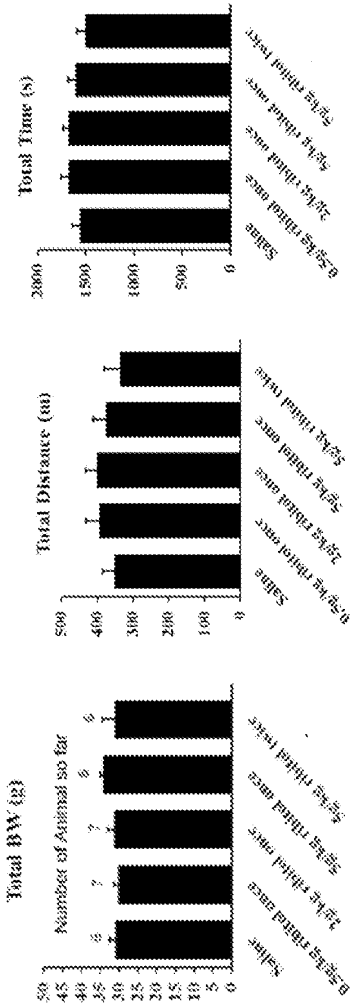


FIG. 8

Matriglycan Expression in L276I FKRP Mutant Mice
after 1 Year of Treatment with Ribitol or Saline

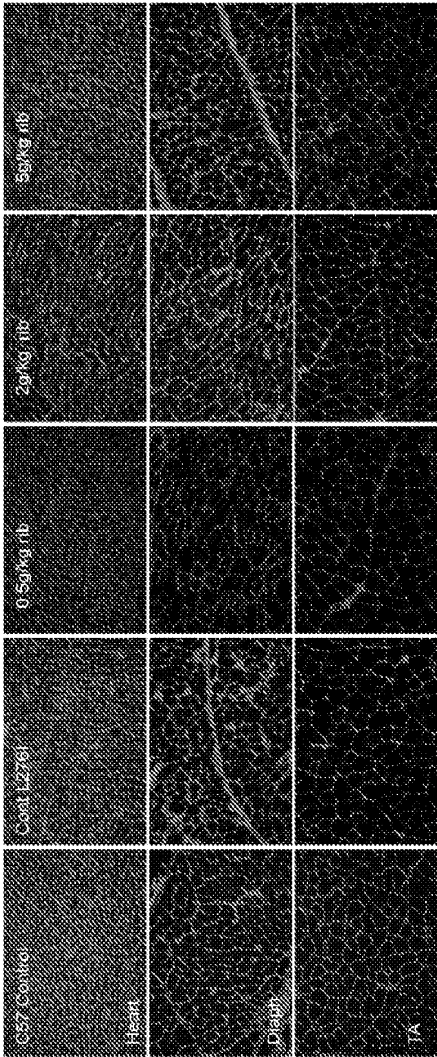


FIG. 9

Western Blot Analysis of Lysates from Heart, Diaphragm (Diaph.) and Tibialis Anterior (TA) of 2 Ribitol-treated (+) and 1 Untreated (-) C57/BL/6J Mice

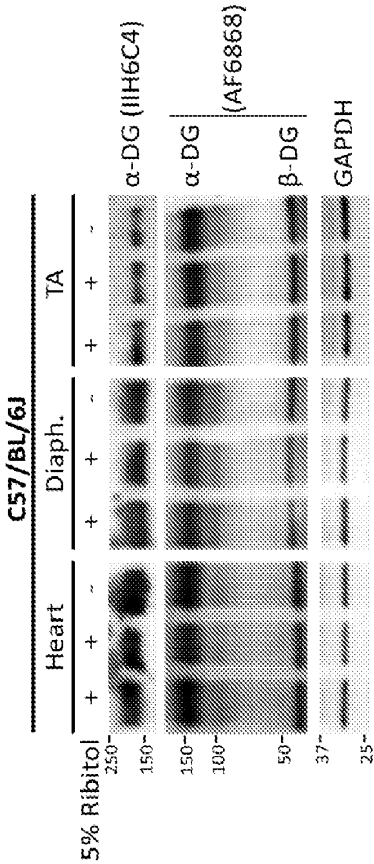


FIG. 10

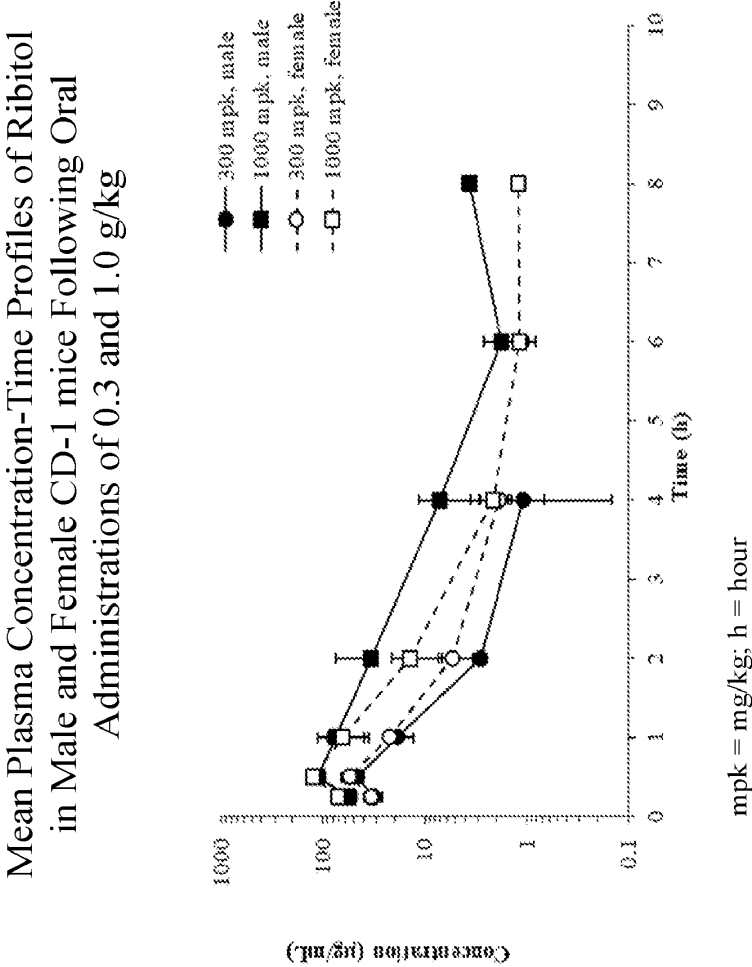
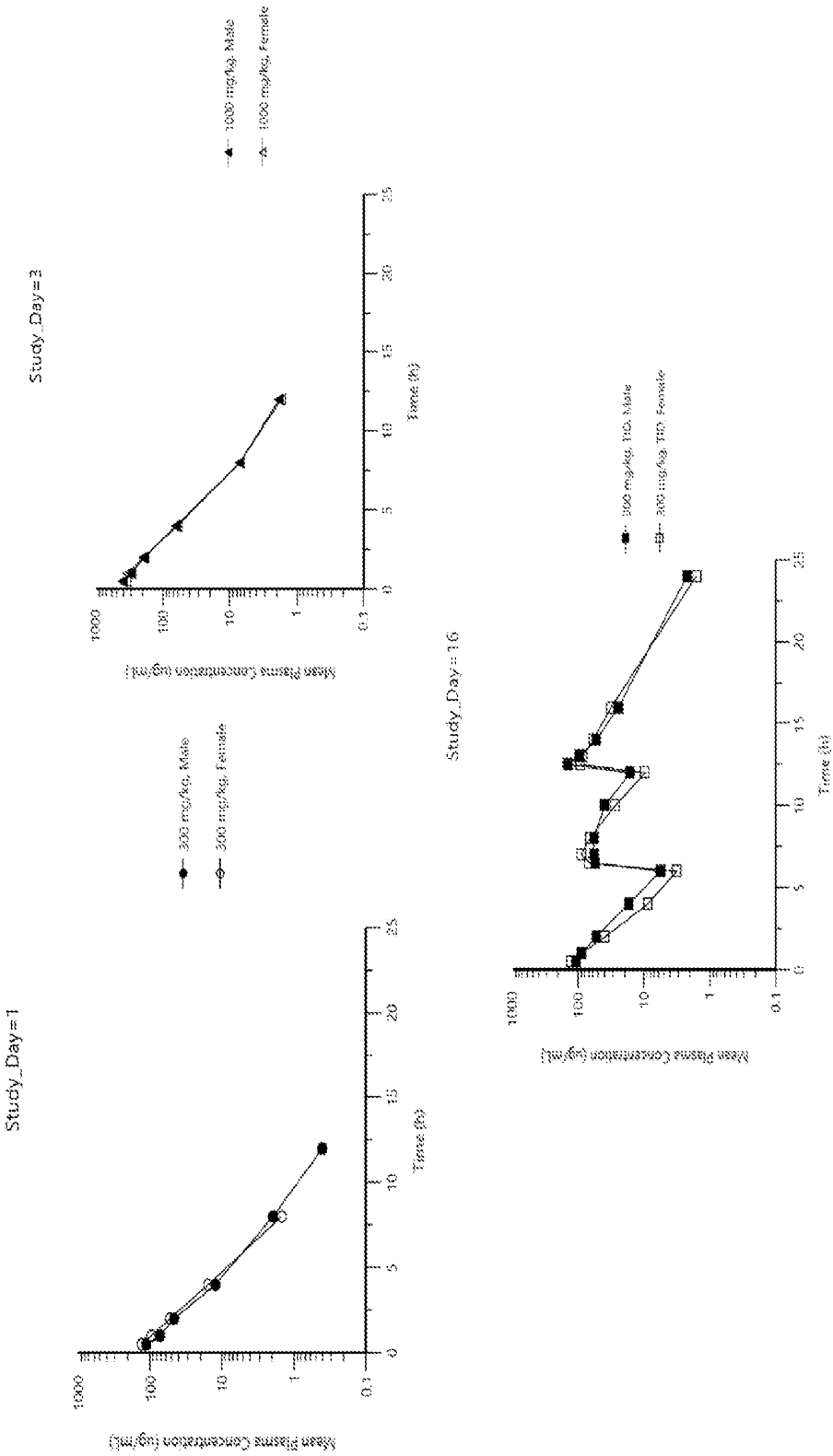


FIG. 11 Mean Plasma Concentration versus Time Profiles of Ribitol Following Oral Administration to Male and Female Bama Minipigs



INCORPORATED BY REFERENCE (RULE 20.6)

FIG. 12

Mean Plasma Concentration-Time Profiles of Ribitol
in Male and Female CD-1 mice Following IV
Administrations of 10, 30 and 100 mg/kg

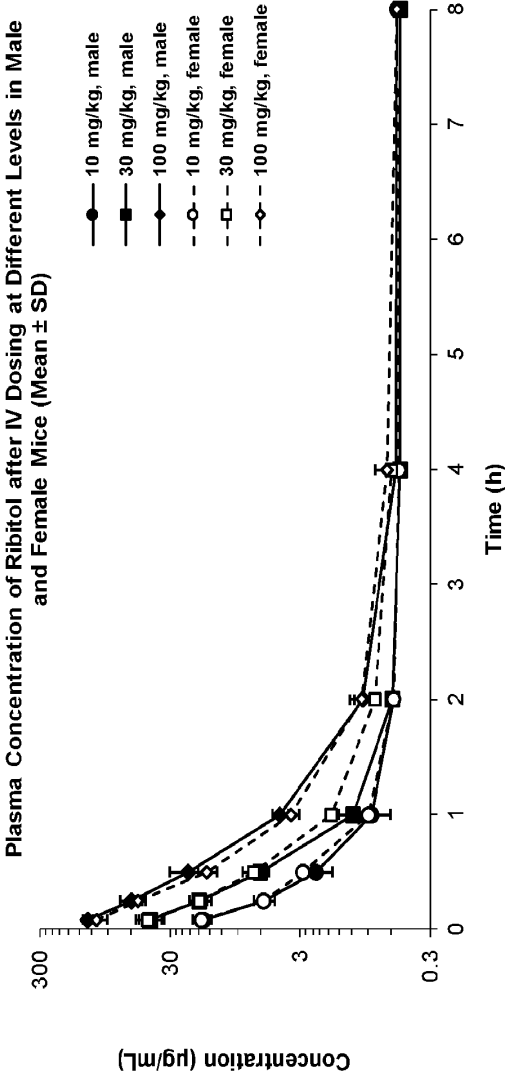


FIG. 13

