Claims

1. Glyceryl tri-[4-phenylbutyrate] (HPN-100) for use in:

A) a method of treating a nitrogen retention disorder selected from the group consisting of urea cycle disorders (UCD), hepatic encephalopathy (HE), and end-stage renal disease (ESRD) in a subject comprising:

(a) administering a first dosage of glyceryl tri-[4-phenylbutyrate] (HPN-100),

(b) measuring plasma phenylacetic acid (PAA) and phenylacetylglutamine (PAGN) levels,

(c) calculating a plasma PAA:PAGN ratio,

(d) determining whether the glyceryl tri-[4-phenylbutyrate] (HPN-100) dosage needs to be adjusted based on whether the PAA:PAGN ratio falls within a target range of 1 to 2.5, where a PAA:PAGN ratio below the target range indicates that the dosage needs to be increased and a PAA:PAGN ratio above the target range indicates that the dosage needs to be decreased, and

(e) administering a second dosage of glyceryl tri-[4-phenylbutyrate] (HPN-100) based on the determination in (d); or

B) a method of treating a nitrogen retention disorder selected from the group consisting of urea cycle disorders (UCD), hepatic encephalopathy (HE), and end-stage renal disease (ESRD) in a subject who has previously been administered a first dosage of glyceryl tri-[4-phenylbutyrate] (HPN-100) comprising:

(a) measuring plasma PAA and PAGN levels,

(b) calculating a plasma PAA:PAGN ratio,

(c) determining whether the first glyceryl tri-[4-phenylbutyrate] (HPN-100) dosage needs to be adjusted based on whether the PAA:PAGN ratio falls within a target range of 1 to 2.5, where a PAA:PAGN ratio below the target range indicates that the dosage needs to be increased and a PAA:PAGN ratio above the target range indicates that the dosage needs to be decreased, and

(d) administering a second dosage of glyceryl tri-[4-phenylbutyrate] (HPN-100) based on the determination in (c).

2. Glyceryl tri-[4-phenylbutyrate] (HPN-100) for use as in claim 1, wherein the target range is 1 to 2.

3. Glyceryl tri-[4-phenylbutyrate] (HPN-100) for use as in claim 1, wherein measurement of PAA and PAGN levels is carried out 48 hours to 1 week after the first dosage of glyceryl tri-[4-phenylbutyrate] (HPN-100) is administered.