REPLACEMENT SHEET

1. A method for adjusting the dosage of a nitrogen scavenging drug in a subject who has previously been administered an initial dosage of the nitrogen scavenging drug, comprising:

a) measuring a fasting blood ammonia level for the subject;

b) comparing the fasting blood ammonia level to the upper limit of normal for blood ammonia level; and

c) administering an adjusted dosage of the nitrogen scavenging drug, wherein the adjusted dosage is greater than the initial dosage if the fasting blood ammonia level is greater than half the upper limit of normal for blood ammonia level.

2. A method of administering a nitrogen scavenging drug to a subject having a nitrogen retention disorder comprising:

a) measuring a fasting blood ammonia level for the subject;

b) comparing the fasting blood ammonia level to the upper limit of normal for blood ammonia level; and

c) administering the nitrogen scavenging drug to the subject if the fasting blood ammonia level is greater than half the upper limit of normal for blood ammonia level.

3. A method of treating a subject with a nitrogen retention disorder who has previously been administered an initial dosage of a nitrogen scavenging drug comprising:

a) measuring a fasting blood ammonia level for the subject;

b) comparing the fasting blood ammonia level to the upper limit of normal for blood ammonia level; and

c) administering an adjusted dosage of the nitrogen scavenging drug that is greater than the initial dosage if the fasting blood ammonia level is greater than half the upper limit of normal for blood ammonia level.

REPLACEMENT SHEET

4. (canceled)

5. The method of any of claims 1-3, wherein the nitrogen retention disorder is selected from the group consisting of a urea cycle disorder and hepatic encephalopathy.

6. The method of any of claims 1-3, wherein the nitrogen scavenging drug is a PAA prodrug.

7. The method of claim 6, wherein the PAA prodrug is selected from the group consisting of glyceryl tri-[4-phenylbutyrate] (HPN-100), phenylbutyric acid (PBA), sodium PBA (NaPBA), and a combination of two or more of HPN-100, PBA, and NaPBA.

8. The method of any of claims 1-3, wherein the nitrogen scavenging drug is sodium benzoate.

9. The method of claim 3 or 4, wherein administering an increased dosage of the nitrogen scavenging drug produces a normal average daily ammonia level in the subject.

10. The method of any of claims 1-3, further comprising the step of determining an upper limit of normal for blood ammonia level for the subject prior to step (b).

11. The method of any of claims 1-3, wherein the upper limit of normal blood ammonia level is 35 µmol/L.

12. The method of claim 6, further comprising:

d) measuring urinary PAGN excretion; and

e) determining an effective dosage of the PAA prodrug based on a mean conversion of PAA prodrug to urinary PAGN of 60-75%.

CLAIMS

1. The use of glyceryl tri-[4-phenylbutyrate] for preparation of a medicament for use in therapy for a urea cycle disorder, wherein the therapy:

a) measuring a fasting blood ammonia level for a subject who has previously been administered an initial dosage of glyceryl tri-[4-phenylbutyrate];

b) comparing the fasting blood ammonia level to the upper limit of normal for blood ammonia level; and

c) administering an adjusted dosage of glyceryl tri-[4-phenylbutyrate], wherein the adjusted dosage is greater than the initial dosage if the fasting blood ammonia level is greater than half the upper limit of normal for blood ammonia level and less than the upper limit of normal for blood ammonia level.

2. The use of glyceryl tri-[4-phenylbutyrate] for preparation of a medicament for use in treating a subject having a urea cycle disorder, wherein glyceryl tri-[4-phenylbutyrate] is administered after:

a) measuring a fasting blood ammonia level for the subject;

b) comparing the fasting blood ammonia level to the upper limit of normal for blood ammonia level; and

c) administering an initial dosage of glyceryl tri-[4-phenylbutyrate] to the subject if the fasting blood ammonia level is greater than half the upper limit of normal for blood ammonia level and less than the upper limit of normal for blood ammonia level.

3. The use of glyceryl tri-[4-phenylbutyrate] for preparation of a medicament for use in treating a subject with a urea cycle disorder who has previously been administered an initial dosage of glyceryl tri-[4-phenylbutyrate], wherein the dose of the drug is adjusted by:

a) measuring a fasting blood ammonia level for the subject;

b) comparing the fasting blood ammonia level to the upper limit of normal for blood ammonia level; and

c) administering an adjusted dosage of glyceryl tri-[4-phenylbutyrate] that is greater than the initial dosage if the fasting blood ammonia level is greater than half the upper limit

of normal for blood ammonia level and less than the upper limit of normal for blood ammonia level.

4. The use of glyceryl tri-[4-phenylbutyrate] for preparation of a medicament for use as in claim 3, wherein administering an increased dosage of glyceryl tri-[4-phenylbutyrate] produces a normal average daily ammonia level in the subject.

5. The use of glyceryl tri-[4-phenylbutyrate] for preparation of a medicament for use as in any of claim 1-3, further comprising the step of determining an upper limit of normal for blood ammonia level for the subject prior to step (b).

6. The use of glyceryl tri-[4-phenylbutyrate] for preparation of a medicament for use as in any of claim 1-3, wherein the upper limit of normal blood ammonia level is 35 μmol/L.

7. The use of glyceryl tri-[4-phenylbutyrate] for preparation of a medicament for use as in any of claims 1-3, further comprising:

d) measuring urinary PAGN excretion; and

e) determining an effective dosage of glyceryl tri-[4-phenylbutyrate] based on a mean conversion of PAA prodrug to urinary PAGN of 60-75%.