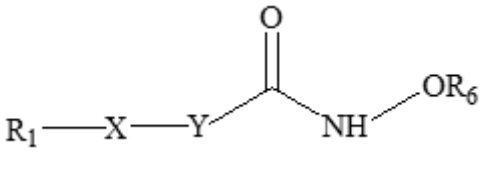
CLAIMS:

1. Enteral or urinary, preferably enteral, pharmaceutical formulation comprising (A) a compound of formula I,



(I),

wherein R_1 is a C1-C3 alkyl;

X is $-C(R_2)(R_3)-C(R_4)(R_5)-$ or $-C\equiv C-$,

wherein R_2, R_3, R_4, and R_5 are each independently H or methyl;

Y is $-CH=CH-CH_2-$, $-CH_2-CH=CH-$; $-C\equiv C-CH_2$; $-CH_2-C\equiv C-$; or $-C(R_7)(R_8)-C(R_9)(R_{10})-C(R_{11})(R_{12})-$,

wherein R_7, R_8, R_9, R_{10}, R_{11} and R_{12}, are each independently H or methyl;

provided that (1) when X is $-C\equiv C-$, Y is $-C(R_7)(R_8)-C(R_9)(R_{10})-C(R_{11})(R_{12})-$ and that (2) when Y is not $-C(R_7)(R_8)-C(R_9)(R_{10})-C(R_{11})(R_{12})-$, X is $-C(R_2)(R_3)-C(R_4)(R_5)-$; and

R_6 is H or CH_3,

or a stereoisomer or a mixture thereof, or a pharmaceutically acceptable salt, ester or solvate thereof; and (B) (a) at least one pharmaceutically acceptable excipient; (b) at least one other therapeutic agent; or (c) a combination of (a) and (b).

2. The formulation of claim 1, wherein R_1 is ethyl.

3. The formulation of claim 1, wherein

(i) X is $-C(R_2)(R_3)-C(R_4)(R_5)-$;

(ii) R_1 is ethyl;

(iii) R_2 is H;

(iv) R_3 is H;

(v) R_4 is H;

(vi) R_5 is H;

(vii) R_6 is H;

(viii) Y is $-CH=CH-CH_2-$, $-CH_2-CH=CH-$, $-C\equiv C-CH_2-$, or $-CH_2-C\equiv C-$; or

(ix) a combination of at least two of (i) to (viii).

4. The formulation of claim 1, wherein

(i) X is -C(R_2)(R_3)-C(R_4)(R_5)-;

(ii) R_1 is ethyl;

(iii) R_2 is H;

(iv) R_3 is H;

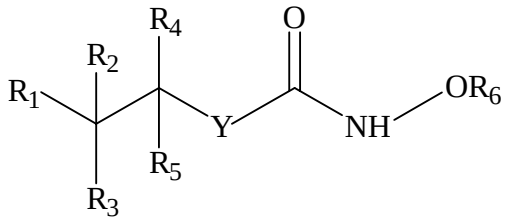
(v) R_4 is H;

(vi) R_5 is H;

(vii) R_6 is H; and

(viii) Y is -CH=CH-CH_2-, -CH_2-CH=CH-, -C≡C-CH_2-, or -CH_2-C≡C-.

5. The formulation of any one of claims 1 to 4, wherein the compound is of formula Ia:



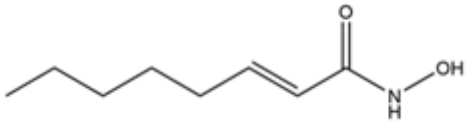
(Ia),

wherein R_1, R_2, R_3, R_4, R_5 and R_6 are as defined in any one of claims 1 to 4 and Y is as defined in claim 3 or 4, or a stereoisomer or a mixture thereof, or a pharmaceutically acceptable salt, ester or solvate thereof.

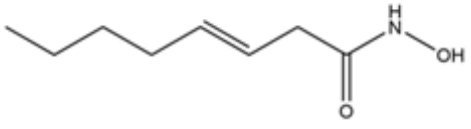
6. The formulation of any one of claims 1 to 5, wherein Y is CH=CH-CH_2-, -CH_2-CH=CH- or -CH_2-C≡C-, preferably Y is CH=CH-CH_2-, or -CH_2-C≡C-.

7. The formulation of any one of claims 1 to 5, wherein Y is -CH_2-C≡C-.

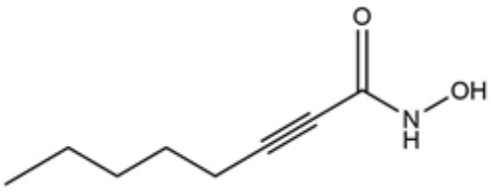
8. The formulation of claim 1, wherein the compound is:



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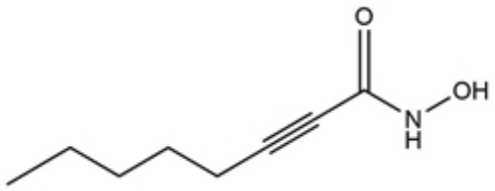
or



,

or a stereoisomer or a mixture thereof, or a pharmaceutically acceptable salt, ester or solvate thereof.

9. The formulation of claim 1, wherein the compound is



,

or a stereoisomer or a mixture thereof, or a pharmaceutically acceptable salt, ester or solvate thereof.

10. The formulation of any one of claims 1 to 9, for enteral or urinary administration, preferably enteral administration and most preferably through a delayed release formulation.

11. The formulation of claim 10, for delivery in the ileum or colon.

12. The formulation of any one of claims 1 to 11, comprising an enteric coating.

13. The compound, stereoisomer, mixture, salt, ester, solvate or formulation as defined in any one of claims 1 to 12, for use in the treatment or prevention of an ammonia-associated disease or disorder, or a symptom thereof in a subject in need thereof.

14. The compound, stereoisomer, mixture, salt, ester, solvate or formulation for use of claim 13, wherein the ammonia-associated disease or disorder is hyperammonemia, and the subject in need thereof preferably has hepatic encephalopathy or a urea cycle disorder.

15. The compound, stereoisomer, mixture, salt, ester, solvate or formulation for use of claim 13 or 14, for enteral or urinary administration, preferably enteral administration and most preferably wherein the enteral administration is administration to the ileum or the colon.

16. Use of the compound, stereoisomer, mixture, salt, ester, solvate or formulation as defined in any one of claims 1 to 12, for treating or preventing an ammonia-associated disease or disorder, or a symptom thereof in a subject in need thereof.

17. Use of the compound, stereoisomer, mixture, salt, ester, solvate or formulation as defined in any one of claims 1 to 12, in the manufacture of a medicament for treating or preventing an ammonia-associated disease or disorder, or a symptom thereof in a subject in need thereof.

18. The use of claim 16 or 17, wherein the ammonia-associated disease or disorder is hyperammonemia.

19. The use of any one of claims 16 to 18, for enteral or urinary administration, preferably enteral administration and most preferably wherein the enteral administration is administration to the ileum or the colon.

20. The use of any one of claims 16 to 19, wherein the subject in need thereof has hepatic encephalopathy or a urea cycle disorder.