WHAT IS CLAIMED IS:

1. An antisense oligonucleotide (ASO) targeting a regulatory RNA (regRNA) of carbamoyl phosphate synthase 1 (CPS1) comprising a nucleotide sequence complementary to at least 8 contiguous nucleotides of any one of SEQ ID NOs: 49, 50, 69-90.

2. The ASO of claim 1, wherein the ASO comprises a nucleotide sequence and optionally a chemical modification of any one of SEQ ID NOs: 1-48, 51-68, or 91-662.

3. An antisense oligonucleotide (ASO) comprising a nucleotide sequence of TGCAGGCACACACATCAGGC (SEQ ID NO: 1).

4. The ASO of claim 3, wherein the 3' end of the ASO is conjugated to a ligand moiety.

5. The ASO of claim 4, wherein the ligand moiety comprises N-acetylgalactosamine (GalNAc).

6. The ASO of claim 5, wherein the ligand moiety comprises a three-cluster GalNAc moiety (GalNAc3).

7. The ASO of claim 3, wherein one or more of the nucleotides of the ASO are chemically modified.

8. The ASO of claim 3, wherein the ASO comprises MT=MG=M5C-MA-MG=dG=d5C=dA=d5C=dA=d5C=dA=d5C=dA=dT=M5C-MA-MG=MG=M5C (SEQ ID NO: 404), wherein MT, MG, MA, and M5C are 2'-O-methoxyethyl thymidine, 2'-O-methoxyethyl guanosine, 2'-O-methoxyethyl adenosine, and 2'-O-methoxyethyl 5-methyl cytidine ribonucleosides, respectively; dA, dG, dT, and d5C are 2'-deoxy adenosine, 2'-deoxy guanosine, 2'-deoxy thymidine and 2'-deoxy 5-methyl cytidine ribonucleosides, respectively, "=" is a phosphorothioate linkage, and "-" is a phosphodiester linkage.

9. The ASO of claim 8, wherein the 3' end of the ASO is conjugated to a ligand moiety.

10. The ASO of claim 9, wherein the ligand moiety comprises N-acetylgalactosamine (GalNAc).

11. The ASO of claim 10, wherein the ligand moiety comprises a three-cluster GalNAc moiety (GalNAc3).

12. The ASO of claim 3, wherein the ASO comprises MT=MG=M5C-MA-MG=dG=d5C=dA=d5C=dA=d5C=dA=d5C=dA=dT=M5C-MA-MG=MG=M5C-[TEG] (SEQ ID NO: 409), wherein MT, MG, MA, and M5C are 2'-O-methoxyethyl thymidine, 2'-O-methoxyethyl guanosine, 2'-O-methoxyethyl adenosine, and 2'-O-methoxyethyl 5-methyl cytidine ribonucleosides; dA, dG, dT, and d5C are 2'-deoxy adenosine, 2'-deoxy guanosine, 2'-deoxy thymidine and 2'-deoxy 5-methyl cytidine ribonucleosides, "=" is a phosphorothioate linkage, "-" is a phosphodiester linkage, and [TEG] is a ligand moiety comprising a triethyleneglycol linked to N-acetylgalactosamine (GalNAc).

13. The ASO of claim 12, wherein the ligand moiety comprises a three-cluster GalNAc (GalNAc3).

14. An antisense oligonucleotide (ASO) comprising MT=MG=M5C-MA-MG=dG=d5C=dA=d5C=dA=d5C=dA=d5C=dA=d5C=dA=dT=M5C-MA-MG=MG=M5C-[TEG] (SEQ ID NO: 409), wherein MT, MG, MA, and M5C are 2'-O-methoxyethyl thymidine, 2'-O-methoxyethyl guanosine, 2'-O-methoxyethyl adenosine, and 2'-O-methoxyethyl 5-methyl cytidine ribonucleosides; dA, dG, dT, and d5C are 2'-deoxy adenosine, 2'-deoxy guanosine, 2'-deoxy thymidine and 2'-deoxy 5-methyl cytidine ribonucleosides, "=" is a phosphorothioate linkage, "-" is a phosphodiester linkage, and [TEG] is a ligand moiety comprising a triethyleneglycol linked to N-acetylgalactosamine (GalNAc).

15. The ASO of claim 14, wherein the ligand moiety comprises a three-cluster GalNAc moiety (GalNAc3).

16. An antisense oligonucleotide (ASO) comprising MT=MG=M5C-MA-MG=dG=d5C=dA=d5C=dA=d5C=dA=d5C=dA=d5C=dA=dT=M5C-MA-MG=MG=M5C (SEQ ID NO: 404), wherein MT, MG, MA, and M5C are 2'-O-methoxyethyl thymidine, 2'-O-methoxyethyl guanosine, 2'-O-methoxyethyl adenosine, and 2'-O-methoxyethyl 5-methyl cytidine ribonucleosides; dA, dG, dT, and d5C are 2'-deoxy adenosine, 2'-deoxy guanosine, 2'-deoxy thymidine and 2'-deoxy 5-methyl cytidine ribonucleosides, "=" is a phosphorothioate linkage, "-" is a phosphodiester linkage.

17. A pharmaceutical composition comprising the ASO of claim 1.

18. A pharmaceutical composition comprising the ASO of claim 3.

19. A pharmaceutical composition comprising the ASO of claim 14.

20. A pharmaceutical composition comprising the ASO of claim 16.

21. An *in vitro* or *ex vivo* method of modulating expression of CPS1 in a cell, comprising contacting the cell with the ASO of claim 1.

22. An *in vitro* or *ex vivo* method of increasing expression of CPS1 in a human cell, comprising contacting the cell with the ASO of claim 3.

23. Use of the ASO of claim 1 in the manufacture of a medicament for the treatment of a urea cycle disorder in a subject in need thereof, wherein an effective amount of the ASO is to be administered to the subject.

24. The use of claim 23, wherein the urea cycle disorder comprises carbamoylphosphate synthetase 1 (CPS1) deficiency, ornithine transcarbamylase deficiency (OTC) deficiency, argininosuccinate synthetase 1 (ASS1) deficiency, argininosuccinate lyase (ASL) deficiency, arginase 1 deficiency, ornithine translocase (ORNT1) deficiency, hyperornithinemia-hyperammonemia-homocitrullinuria (HHH) syndrome, or citrin deficiency.

25. Use of the ASO of claim 3 in the manufacture of a medicament for the treatment of a urea cycle disorder in a subject in need thereof, wherein an effective amount of the ASO is to be administered to the subject.

26. Use of the ASO of claim 1 in the manufacture of a medicament for the treatment or prevention of hyperammonemia in a subject in need thereof, wherein an effective amount of the ASO is to be administered to the subject.

27. The use of claim 26, wherein the subject comprises one or more of: a urea cycle disorder, hepatic encephalopathy, acute liver failure, liver cirrhosis, non-alcoholic fatty liver disease, renal dysfunction or failure, propionic acidemia, methylmalonic acidemia, isovaleric acidemia, a urinary tract infection, intestinal bacterial overgrowth, a fatty acid oxidation disorder, and a systemic infection, or wherein the subject has or is at risk of developing hyperammonemia associated with the intake of a drug.

28. The use of claim 27, wherein the drug is valproic acid, carbamazepine, fluorouracil, sulfadiazine, ribavirin, salicylates, or glycine.

29. Use of the ASO of claim 3 in the manufacture of a medicament for the treatment or prevention of hyperammonemia in a subject in need thereof, wherein an effective amount of the ASO is to be administered to the subject.

30. An *in vitro* or *ex vivo* method of increasing the level of CPS1 in a cell comprising contacting the cell with an ASO of claim 1.