We claim:

1. A double stranded ribonucleic acid (dsRNA) agent for inhibiting expression of C9orf72, wherein the dsRNA agent comprises a sense strand and an antisense strand forming a double stranded region,

wherein the sense strand comprises at least 15 contiguous nucleotides differing by no more than 3 nucleotides from any one of the antisense nucleotide sequences in any one of Tables 10A, 10B, 12-15, 19, 20, or 21.

2. A double stranded ribonucleic acid (dsRNA) agent for inhibiting expression of C9orf72, wherein the dsRNA agent comprises a sense strand and an antisense strand forming a double stranded region,

wherein the sense strand comprises at least 15 contiguous nucleotides differing by no more than three nucleotides from nucleotides 200-290 of SEQ ID NO: 133, and the antisense strand comprises at least 15 contiguous nucleotides from the corresponding nucleotide sequence of SEQ ID NO: 134, and

wherein the sense strand, the antisense strand, or both the sense strand and the antisense strand is conjugated to one or more lipophilic moieties.

3. A double stranded ribonucleic acid (dsRNA) agent for inhibiting expression of C9orf72, wherein the dsRNA agent comprises a sense strand and an antisense strand forming a double stranded region,

wherein the antisense strand comprises at least 15 contiguous nucleotides differing by no more than 3 nucleotides from the nucleotide sequence of SEQ ID NO:125, and

wherein the sense strand, the antisense strand, or both the sense strand and the antisense strand is conjugated to one or more lipophilic moieties.

4. The dsRNA agent of claim 1 or 2, wherein the nucleotide sequence of the sense and antisense strand comprise any one of the sense and antisense strand nucleotide sequences in any one of Tables 14, 15, 19, or 20.

5. The dsRNA agent of any one of claims 1, 2, and 4, wherein the antisense strand comprises at least 15 contiguous nucleotides differing by no more than three nucleotides from any one of the antisense strand nucleotide sequences of a duplex selected from the group consisting of AD-348904.1, AD-348136.1, AD-347612.1, AD-348639.1, AD-347606.1, AD-348905.1, AD-347613.1, AD-348842.1, AD-347610.1, AD-350329.1, AD-348140.1, AD-348930.1, AD-347863.1, AD-348500.1, AD-348597.1, AD-348578.1, AD-347923.1, AD-348588.1, and AD-347773.1.

6. A double stranded ribonucleic acid (dsRNA) agent for selectively inhibiting a C9orf72 target RNA comprising a hexanucleotide repeat comprising multiple contiguous copies of SEQ ID NO: 1, wherein the dsRNA agent comprises a sense strand and an antisense strand forming a double stranded region,

wherein the antisense strand comprises at least 15 contiguous nucleotides differing by no more than 3 nucleotides from any one of the antisense nucleotide sequences in any one of Tables 14, 15, and 21.

7. The dsRNA agent of any one of claims 1, 2, 4, 5, and 6, wherein the antisense strand comprises at least 15 contiguous nucleotides differing by no more than three nucleotides from any one of the antisense strand nucleotide sequences of a duplex selected from the group consisting of AD-463863.1, AD-463862.1, AD-463869.1, AD-463873.1, AD-463872.1, and AD-463860.1.

8. The dsRNA of claim 7, wherein the antisense strand comprises at least 15 contiguous nucleotides from any one of the antisense strand nucleotide sequences of a duplex selected from the group consisting of AD-463863.1, AD-463862.1, AD-463869.1, AD-463873.1, AD-463872.1, and AD-463860.1

9. The dsRNA agent claim 7, wherein the antisense strand comprises a nucleotide sequence differing by no more than three nucleotides from any one of the antisense strand nucleotide sequences of a duplex selected from the group consisting of AD-463863.1, AD-463862.1, AD-463869.1, AD-463873.1, AD-463872.1, and AD-463860.1.

10. The dsRNA agent of claim 7, wherein the antisense strand comprises a nucleotide sequence of any one of the antisense strand nucleotide sequences of a duplex selected from the group consisting of AD-463863.1, AD-463862.1, AD-463869.1, AD-463873.1, AD-463872.1, and AD-463860.1.

11. The dsRNA agent of claim 7, wherein the antisense strand consists of a nucleotide sequence of any one of the antisense strand nucleotide sequences of a duplex selected from the group consisting of AD-463863.1, AD-463862.1, AD-463869.1, AD-463873.1, AD-463872.1, and AD-463860.1.

12. The dsRNA agent of any one of claims 7-11, wherein the sense strand comprises at least 15 contiguous nucleotides differing by no more than three nucleotides from any one of the sense strand nucleotide sequences of a duplex selected from the group consisting of AD-463863.1, AD-463862.1, AD-463869.1, AD-463873.1, AD-463872.1, and AD-463860.1.

13 The dsRNA agent of any one of claims 7-12, wherein the sense strand comprises at least 15 contiguous nucleotides from any one of the sense strand nucleotide sequences of a duplex selected

from the group consisting of AD-463863.1, AD-463862.1, AD-463869.1, AD-463873.1, AD-463872.1, and AD-463860.1

14. The dsRNA agent of any one of claims 7-13, wherein the sense strand comprises a nucleotide sequence differing by no more than three nucleotides from any one of the sense strand nucleotide sequences of a duplex selected from the group consisting of AD-463863.1, AD-463862.1, AD-463869.1, AD-463873.1, AD-463872.1, and AD-463860.1.

15. The dsRNA agent of any one of claims 7-14, wherein the sense strand comprises a nucleotide sequence of any one of the sense strand nucleotide sequences of a duplex selected from the group consisting of AD-463863.1, AD-463862.1, AD-463869.1, AD-463873.1, AD-463872.1, and AD-463860.1.

16. The dsRNA agent of any one of claims 7-15, wherein the sense strand consists of a nucleotide sequence of any one of the sense strand nucleotide sequences of a duplex selected from the group consisting of AD-463863.1, AD-463862.1, AD-463869.1, AD-463873.1, AD-463872.1, and AD-463860.1.

17. The dsRNA agent of claim 1 or 3, wherein the nucleotide sequence of the sense and antisense strand comprise any one of the sense and antisense strand nucleotide sequences in any one of Tables 12 or 13.

18. The dsRNA agent of any one of claim 1, 3, and 17, wherein the sense strand comprises at least 15 contiguous nucleotides differing by no more than three nucleotides from any one of the nucleotide sequence of nucleotides 230-270, 233-262, 800-840, 800-830, 802-828, 1240-1290, 1240-1280, 1247-1288, 1590-1645, 1590-1620, and 1594-1642 of SEQ ID NO:121, and the antisense strand comprises at least 15 contiguous nucleotides from the corresponding nucleotide sequence of SEQ ID NO:125.

19. The dsRNA agent of any one of claims 1, 3, 17, and 18, wherein the sense strand comprises at least 15 contiguous nucleotides differing by no more than three nucleotides from any one of the nucleotide sequence of nucleotides 1594-1616, 802-824, 239-261, 1308-1330, 233-255, 1595-1617, 240-262, 1532-1554, 237-259, 3268-3290, 806-828, 1620-1642, 526-548, 1169-1191, 1266-1288, 1247-1269, 586-608, 1257-1279, and 400-422 of SEQ ID NO:121, and the antisense strand comprises at least 15 contiguous nucleotides from the corresponding nucleotide sequence of SEQ ID NO:125.

20. The dsRNA agent of any one of claims 1 and 4-19, wherein the sense strand, the antisense strand, or both the sense strand and the antisense strand is conjugated to one or more lipophilic moieties.

21. The dsRNA agent of claim 20, wherein the lipophilic moiety is conjugated to one or more internal positions in the double stranded region of the dsRNA agent.

22. The dsRNA agent of claim 20 or 21, wherein the lipophilic moiety is conjugated via a linker or carrier.

23. The dsRNA agent of any one of claims 20-22, wherein lipophilicity of the lipophilic moiety, measured by logKow, exceeds 0.

24. The dsRNA agent of any one of claims 1-23, wherein the hydrophobicity of the double-stranded RNAi agent, measured by the unbound fraction in a plasma protein binding assay of the double-stranded RNAi agent, exceeds 0.2.

25. The dsRNA agent of claim 24, wherein the plasma protein binding assay is an electrophoretic mobility shift assay using human serum albumin protein.

26. The dsRNA agent of any one of claims 1-25, wherein the dsRNA agent comprises at least one modified nucleotide.

27. The dsRNA agent of claim 26, wherein no more than five of the sense strand nucleotides and no more than five of the nucleotides of the antisense strand are unmodified nucleotides

28. The dsRNA agent of claim 26, wherein all of the nucleotides of the sense strand and all of the nucleotides of the antisense strand are modified nucleotides.

29. The dsRNA agent of any one of claims 26-28, wherein at least one of the modified nucleotides is selected from the group a deoxy-nucleotide, a 3’-terminal deoxy-thymine (dT) nucleotide, a 2'-O-methyl modified nucleotide, a 2'-fluoro modified nucleotide, a 2'-deoxy-modified nucleotide, a locked nucleotide, an unlocked nucleotide, a conformationally restricted nucleotide, a constrained ethyl nucleotide, an abasic nucleotide, a 2’-amino-modified nucleotide, a 2’-O-allyl-modified nucleotide, 2’-C-alkyl-modified nucleotide, 2’-hydroxly-modified nucleotide, a 2’-methoxyethyl modified nucleotide, a 2’-O-alkyl-modified nucleotide, a morpholino nucleotide, a phosphoramidate, a non-natural base comprising nucleotide, a tetrahydropyran modified nucleotide, a 1,5-anhydrohexitol modified nucleotide, a cyclohexenyl modified nucleotide, a nucleotide comprising a 5'-phosphorothioate group, a nucleotide comprising a 5'-methylphosphonate group, a nucleotide comprising a 5’ phosphate or 5’ phosphate mimic, a nucleotide comprising vinyl phosphonate, a nucleotide comprising adenosine-glycol nucleic acid (GNA), a nucleotide comprising thymidine-

glycol nucleic acid (GNA) S-Isomer, a nucleotide comprising 2-hydroxymethyl-tetrahydrofurane-5-phosphate, a nucleotide comprising 2'-deoxythymidine-3'phosphate, a nucleotide comprising 2'-deoxyguanosine-3'-phosphate, and a terminal nucleotide linked to a cholesteryl derivative and a dodecanoic acid bisdecylamide group; and combinations thereof.

30. The dsRNA agent of claim 29, wherein the modified nucleotide is selected from the group consisting of a 2'-deoxy-2'-fluoro modified nucleotide, a 2'-deoxy-modified nucleotide, 3'-terminal deoxy-thymine nucleotides (dT), a locked nucleotide, an abasic nucleotide, a 2'-amino-modified nucleotide, a 2'-alkyl-modified nucleotide, a morpholino nucleotide, a phosphoramidate, and a non-natural base comprising nucleotide.

31. The dsRNA agent of claim 29, wherein the modified nucleotide comprises a short sequence of 3'-terminal deoxy-thymine nucleotides (dT).

32. The dsRNA agent of claim 29, wherein the modifications on the nucleotides are 2'-O-methyl, GNA and 2'fluoro modifications.

33. The dsRNA agent of any one of claims 1-32, further comprising at least one phosphorothioate internucleotide linkage.

34. The dsRNA agent of claim 33, wherein the dsRNA agent comprises 6-8 phosphorothioate internucleotide linkages.

35. The dsRNA agent of any one of claims 1-34, wherein each strand is no more than 30 nucleotides in length.

36. The dsRNA agent of any one of claims 1-35, wherein at least one strand comprises a 3' overhang of at least 1 nucleotide.

37. The dsRNA agent of any one of claims 1-36, wherein at least one strand comprises a 3' overhang of at least 2 nucleotides.

38. The dsRNA agent of any one of claims 1-37, wherein the double stranded region is 15-30 nucleotide pairs in length.

39. The dsRNA agent of claim 38, wherein the double stranded region is 17-23 nucleotide pairs in length.

40. The dsRNA agent of claim 38, wherein the double stranded region is 17-25 nucleotide pairs in length.

41. The dsRNA agent of claim 38, wherein the double stranded region is 23-27 nucleotide pairs in length.

42. The dsRNA agent of claim 38, wherein the double stranded region is 19-21 nucleotide pairs in length.

43. The dsRNA agent of claim 38, wherein the double stranded region is 21-23 nucleotide pairs in length.

44. The dsRNA agent of any one of claims 1-43, wherein each strand is 19-30 nucleotides in length.

45. The dsRNA agent of any one of claims 1-43, wherein each strand is 19-23 nucleotides in length.

46. The dsRNA agent of any one of claims 1-43, wherein each strand is 21-23 nucleotides in length.

47. The dsRNA agent of any one of claims 21-46, wherein one or more lipophilic moieties are conjugated to one or more internal positions on at least one strand.

48. The dsRNA agent of claim 47, wherein the one or more lipophilic moieties are conjugated to one or more internal positions on at least one strand via a linker or carrier.

49. The dsRNA agent of claim 48, wherein the internal positions include all positions except the terminal two positions from each end of the at least one strand.

50. The dsRNA agent of claim 48, wherein the internal positions include all positions except the terminal three positions from each end of the at least one strand.

51. The dsRNA agent of claim 48-50, wherein the internal positions exclude a cleavage site region of the sense strand.

52. The dsRNA agent of claim 51, wherein the internal positions include all positions except positions 9-12, counting from the 5'-end of the sense strand.

53. The dsRNA agent of claim 51, wherein the internal positions include all positions except positions 11-13, counting from the 3'-end of the sense strand.

54. The dsRNA agent of claim 48-50, wherein the internal positions exclude a cleavage site region of the antisense strand.

55. The dsRNA agent of claim 54, wherein the internal positions include all positions except positions 12-14, counting from the 5'-end of the antisense strand.

56. The dsRNA agent of claim 48-50, wherein the internal positions include all positions except positions 11-13 on the sense strand, counting from the 3'-end, and positions 12-14 on the antisense strand, counting from the 5'-end.

57. The dsRNA agent of any one of claims 21-56, wherein the one or more lipophilic moieties are conjugated to one or more of the internal positions selected from the group consisting of positions 4-8 and 13-18 on the sense strand, and positions 6-10 and 15-18 on the antisense strand, counting from the 5'end of each strand.

58. The dsRNA agent of claim 57, wherein the one or more lipophilic moieties are conjugated to one or more of the internal positions selected from the group consisting of positions 5, 6, 7, 15, and 17 on the sense strand, and positions 15 and 17 on the antisense strand, counting from the 5'-end of each strand.

59. The dsRNA agent of claim 21, wherein the internal positions in the double stranded region exclude a cleavage site region of the sense strand.

60. The dsRNA agent of any one of claims 20-59, wherein the sense strand is 21 nucleotides in length, the antisense strand is 23 nucleotides in length, and the lipophilic moiety is conjugated to position 21, position 20, position 15, position 1, position 7, position 6, or position 2 of the sense strand or position 16 of the antisense strand.

61. The dsRNA agent of claim 60, wherein the lipophilic moiety is conjugated to position 21, position 20, position 15, position 1, or position 7 of the sense strand.

62. The dsRNA agent of claim 60, wherein the lipophilic moiety is conjugated to position 21, position 20, or position 15 of the sense strand.

63. The dsRNA agent of claim 60, wherein the lipophilic moiety is conjugated to position 20 or position 15 of the sense strand.

64. The dsRNA agent of claim 60, wherein the lipophilic moiety is conjugated to position 16 of the antisense strand.

65. The dsRNA agent of any one of claims 20-64, wherein the lipophilic moiety is an aliphatic, alicyclic, or polyalicyclic compound.

66. The dsRNA agent of claim 65, wherein the lipophilic moiety is selected from the group consisting of lipid, cholesterol, retinoic acid, cholic acid, adamantane acetic acid, 1-pyrene butyric acid, dihydrotestosterone, 1,3-bis-O(hexadecyl)glycerol, geranyloxyhexyanol, hexadecylglycerol, borneol, menthol, 1,3-propanediol, heptadecyl group, palmitic acid, myristic acid, O3-(oleoyl)lithocholic acid, O3-(oleoyl)cholenic acid, dimethoxytrityl, or phenoxazine.

67. The dsRNA agent of claim 65, wherein the lipophilic moiety contains a saturated or unsaturated C4-C30 hydrocarbon chain, and an optional functional group selected from the group consisting of hydroxyl, amine, carboxylic acid, sulfonate, phosphate, thiol, azide, and alkyne.

68. The dsRNA agent of claim 67, wherein the lipophilic moiety contains a saturated or unsaturated C6-C18 hydrocarbon chain.

69. The dsRNA agent of claim 67, wherein the lipophilic moiety contains a saturated or unsaturated C16 hydrocarbon chain.

70. The dsRNA agent of claim 69, wherein the saturated or unsaturated C16 hydrocarbon chain is conjugated to position 6, counting from the 5'-end of the strand.

71. The dsRNA agent of any one of claims 20-70, wherein the lipophilic moiety is conjugated via a carrier that replaces one or more nucleotide(s) in the internal position(s) or the double stranded region.

72. The dsRNA agent of claim 71, wherein the carrier is a cyclic group selected from the group consisting of pyrrolidinyl, pyrazolinyl, pyrazolidinyl, imidazolinyl, imidazolidinyl, piperidinyl,

piperazinyl, [1,3]dioxolanyl, oxazolidinyl, isoxazolidinyl, morpholinyl, thiazolidinyl, isothiazolidinyl, quinoxalinyl, pyridazinonyl, tetrahydrofuranyl, and decalinyl; or is an acyclic moiety based on a serinol backbone or a diethanolamine backbone.

73. The dsRNA agent of any one of claims 20-70, wherein the lipophilic moiety is conjugated to the double-stranded iRNA agent via a linker containing an ether, thioether, urea, carbonate, amine, amide, maleimide-thioether, disulfide, phosphodiester, sulfonamide linkage, a product of a click reaction, or carbamate.

74. The double-stranded iRNA agent of any one of claims 20-73, wherein the lipophilic moiety is conjugated to a nucleobase, sugar moiety, or internucleosidic linkage.

75. The dsRNA agent of any one of claims 20-74, wherein the lipophilic moeity or targeting ligand is conjugated via a bio-clevable linker selected from the group consisting of DNA, RNA, disulfide, amide, funtionalized monosaccharides or oligosaccharides of galactosamine, glucosamine, glucose, galactose, mannose, and combinations thereof.

76. The dsRNA agent of any one of claims 20-75, wherein the 3' end of the sense strand is protected via an end cap which is a cyclic group having an amine, said cyclic group being selected from the group consisting of pyrrolidinyl, pyrazolinyl, pyrazolidinyl, imidazolinyl, imidazolidinyl, piperidinyl, piperazinyl, [1,3]dioxolanyl, oxazolidinyl, isoxazolidinyl, morpholinyl, thiazolidinyl, isothiazolidinyl, quinoxalinyl, pyridazinonyl, tetrahydrofuranyl, and decalinyl.

77. The dsRNA agent of any one of claims 1-19, further comprising a targeting ligand that targets a liver tissue.

78. The dsRNA agent of claim 77, wherein the targeting ligand is a GalNAc conjugate.

79. The dsRNA agent of any one of claims 1-78 further comprising
a terminal, chiral modification occurring at the first internucleotide linkage at the 3' end of the antisense strand, having the linkage phosphorus atom in Sp configuration,
a terminal, chiral modification occurring at the first internucleotide linkage at the 5' end of the antisense strand, having the linkage phosphorus atom in Rp configuration, and
a terminal, chiral modification occurring at the first internucleotide linkage at the 5' end of the sense strand, having the linkage phosphorus atom in either Rp configuration or Sp configuration.

80. The dsRNA agent of any one of claims 1-78 further comprising
a terminal, chiral modification occurring at the first and second internucleotide linkages at the 3' end of the antisense strand, having the linkage phosphorus atom in Sp configuration,
a terminal, chiral modification occurring at the first internucleotide linkage at the 5' end of the antisense strand, having the linkage phosphorus atom in Rp configuration, and
a terminal, chiral modification occurring at the first internucleotide linkage at the 5' end of the sense strand, having the linkage phosphorus atom in either Rp or Sp configuration.

81. The dsRNA agent of any one of claims 1-78 further comprising
a terminal, chiral modification occurring at the first, second and third internucleotide linkages at the 3' end of the antisense strand, having the linkage phosphorus atom in Sp configuration,
a terminal, chiral modification occurring at the first internucleotide linkage at the 5' end of the antisense strand, having the linkage phosphorus atom in Rp configuration, and
a terminal, chiral modification occurring at the first internucleotide linkage at the 5' end of the sense strand, having the linkage phosphorus atom in either Rp or Sp configuration.

82. The dsRNA agent of any one of claims 1-78 further comprising
a terminal, chiral modification occurring at the first, and second internucleotide linkages at the 3' end of the antisense strand, having the linkage phosphorus atom in Sp configuration,
a terminal, chiral modification occurring at the third internucleotide linkages at the 3' end of the antisense strand, having the linkage phosphorus atom in Rp configuration,
a terminal, chiral modification occurring at the first internucleotide linkage at the 5' end of the antisense strand, having the linkage phosphorus atom in Rp configuration, and
a terminal, chiral modification occurring at the first internucleotide linkage at the 5' end of the sense strand, having the linkage phosphorus atom in either Rp or Sp configuration.

83. The dsRNA agent of any one of claims 1-78 further comprising
a terminal, chiral modification occurring at the first, and second internucleotide linkages at the 3' end of the antisense strand, having the linkage phosphorus atom in Sp configuration,
a terminal, chiral modification occurring at the first, and second internucleotide linkages at the 5' end of the antisense strand, having the linkage phosphorus atom in Rp configuration, and
a terminal, chiral modification occurring at the first internucleotide linkage at the 5' end of the sense strand, having the linkage phosphorus atom in either Rp or Sp configuration.

84. The dsRNA agent of any one of claims 1-83, further comprising a phosphate or phosphate mimic at the 5'-end of the antisense strand.

85. The dsRNA agent of claim 84, wherein the phosphate mimic is a 5'-vinyl phosphonate (VP).

86. The dsRNA agent of any one of claims 1-83, wherein the base pair at the 1 position of the 5′-end of the antisense strand of the duplex is an AU base pair.

87. The dsRNA agent of any one of claims 1-83, wherein the sense strand has a total of 21 nucleotides and the antisense strand has a total of 23 nucleotides.

88. A cell containing the dsRNA agent of any one of claims 1-87.

89. A pharmaceutical composition for inhibiting expression of a C9orf72, comprising the dsRNA agent of any one of claims 1-87.

90. A pharmaceutical composition comprising the dsRNA agent of any one of claims 1-87 and a lipid formulation.

91. The pharmaceutical composition of claim 89 or 90, wherein dsRNA agent is in an unbuffered solution.

92. The pharmaceutical composition of claim 88, wherein the unbuffered solution is saline or water.

93. The pharmaceutical composition of claim 89 or 90, wherein said dsRNA agent is in a buffer solution.

94. The pharmaceutical composition of claim 93, wherein the buffer solution comprises acetate, citrate, prolamine, carbonate, or phosphate or any combination thereof.

95. The pharmaceutical composition of claim 93, wherein the buffer solution is phosphate buffered saline (PBS).

96. Use of the dsRNA agent of any one of claims 1-87, or the pharmaceutical composition of any one of claims 89-95 in the manufacture of a medicament for inhibiting expression of C9orf72 in a cell.

97. The use of claim 96, wherein the cell is within a subject.

98. The use of claim 97, wherein the subject is a human.

99. The use of claim 98, wherein the subject has a C9orf72-associated disorder.

100. The use of claim 99, wherein the C9orf72-associated disorder is selected from the group consisting of C9orf72 amyotrophic lateral sclerosis/frontotemporal dementia or Huntington-Like Syndrome Due To C9orf72 Expansions.

101. The use of any one of claims 96-100, wherein contacting the cell with the dsRNA agent inhibits the expression of C9orf72 by no more than 50%.

102. The use of any one of claims 96-101, wherein inhibiting expression of C9orf72 decreases C9orf72 protein level in serum of the subject by no more than 50%.

103. Use of the dsRNA agent of any one of claims 1-87, or the pharmaceutical composition of any one of claims 89-95 in the manufacture of a medicament for treating a subject having a disorder that would benefit from reduction in C9orf72 expression.

104. Use of the dsRNA agent of any one of claims 1-87, or the pharmaceutical composition of any one of claims 89-95 in the manufacture of a medicament for preventing at least one symptom in a subject having a disorder that would benefit from reduction in C9orf72 expression.

105. The use of claim 103 or 104, wherein the disorder is a C9orf72-associated disorder.

106. The use of claim 105, wherein the C9orf723-associated disorder is selected from the group consisting of C9orf72 amyotrophic lateral sclerosis/frontotemporal dementia and Huntington-Like Syndrome Due To C9orf72 Expansions.

107. The use of claim 106, wherein the subject is human.

108. The use of claim 106 or 107, wherein the administration of the agent to the subject causes a decrease in a repeat-length-dependent formation of RNA foci, a sequestration of specific RNA-binding proteins, or an accumulation or aggregation or poly(glycine-alanine) peptides, poly(glycine-proline) peptides, poly(glycine-arginine) peptides, poly(alanine-proline) peptides, or poly(proline-arginine) peptides.

109. The use of claim 108, wherein the repeat-length-dependent formation of RNA foci, the sequestration of specific RNA-binding proteins, or the accumulation or aggregation or poly(glycine-alanine) peptides, poly(glycine-proline) peptides, poly(glycine-arginine) peptides, poly(alanine-proline) peptides, or poly(proline-arginine) peptides is decreased by more than 50% and the

expression of C9orf72 mature RNA is inhibited by less than 50%.

110. The use of any one of claims 103-109, wherein the dsRNA agent is to be administered to the subject at a dose of 0.01 mg/kg to 50 mg/kg.

111. The use of any one of claims 103-110, wherein the dsRNA agent is to be administered to the subject subcutaneously.

112. The use of any one of claims 93-110, wherein the dsRNA agent is to be administered to the subject intrathecally.

113. The use of any one of claims 93-112, further comprising determining the level of C9orf72 in a sample(s) from the subject.

114. The use of claim 113, wherein the level of C9orf72 in the subject sample(s) is a C9orf72 protein level in a blood, serum, or cerebrospinal fluid sample(s).

115. The use of any one of claims 103-114, further comprising that an additional therapeutic agent is to be administered to the subject.

116. A kit comprising the dsRNA agent of any one of claims 1-87 or the pharmaceutical composition of any one of claims 89-95.

117. A vial comprising the dsRNA agent of any one of claims 1-87 or the pharmaceutical composition of any one of claims 89-95.

118. A syringe comprising the dsRNA agent of any one of claims 1-87 or the pharmaceutical composition of any one of claims 89-95.