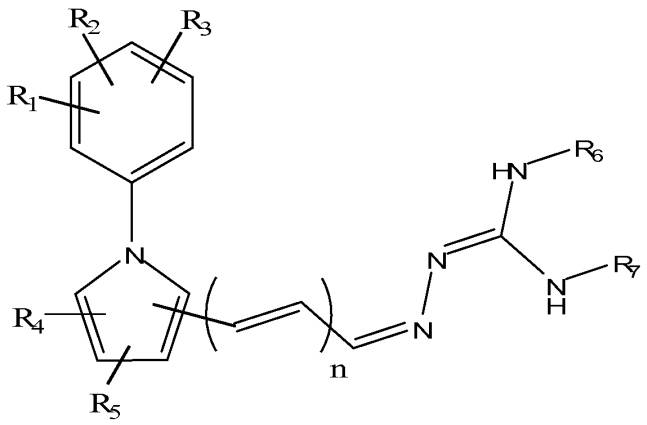
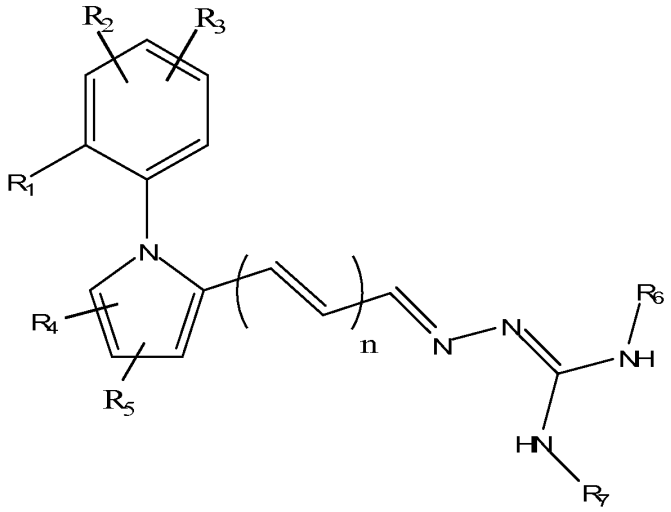
**Claims**

1. A composition comprising a compound of formula (I) or (Ia):



formula (I)



formula (Ia)

including tautomeric and isomeric forms, such as enantiomeric forms, diastereomeric forms and racemic forms, thereof;

wherein

n is 1, 2 or 3;

each $R_1$, $R_2$, $R_3$, $R_4$ and $R_5$ is independently selected from the group consisting of hydrogen, optionally substituted $C_{1-6}$-alkyl, optionally substituted $C_{3-6}$-cycloalkyl, optionally substituted $C_{2-6}$-alkenyl, optionally substituted $C_{4-6}$-alkadienyl, optionally substituted $C_{2-6}$- alkynyl, hydroxy, optionally substituted $C_{1-6}$-alkoxy, optionally substituted $C_{2-6}$-alkenyloxy, carboxy, optionally substituted $C_{1-6}$-alkoxycarbonyl, optionally substituted $C_{1-6}$- alkylcarbonyl, formyl, $C_{1-6}$-alkylsulphonylamino, optionally substituted aryl, optionally substituted aryloxycarbonyl, optionally substituted aryloxy, optionally substituted arylcarbonyl, optionally substituted arylamino, arylsulphonylamino, optionally substituted heteroaryl, optionally substituted heteroaryloxycarbonyl, optionally substituted heteroaryloxy, optionally substituted heteroarylcarbonyl, optionally substituted heteroarylamino, heteroarylsulphonylamino, optionally substituted heterocyclyl, optionally substituted heterocyclyloxycarbonyl, optionally substituted heterocyclyloxy, optionally substituted heterocyclylcarbonyl, optionally substituted heterocyclylamino, heterocyclylsulphonylamino, amino, mono- and di($C_{1-6}$-alkyl)amino, carbamoyl, mono- and di($C_{1-6}$-alkyl)aminocarbonyl, amino-$C_{1-6}$-alkyl-aminocarbonyl, mono- and di($C_{1-6}$-alkylamino-$C_{1-6}$-alkyl-aminocarbonyl, $C_{1-6}$-alkylcarbonylamino, amino-$C_{1-6}$-alkyl-carbonylamino, mono- and di($C_{1-6}$-alkyl)amino-$C_{1-6}$-alkyl-carbonylamino, cyano, guanidino, carbamido, $C_{1-6}$-alkanoyloxy, $C_{1-6}$-alkylsulphonyl, $C_{1-6}$-alkylsulphinyl, $C_{1-6}$-alkylsulphonyl- oxy, aminosulfonyl, mono- and di($C_{1-6}$-alkyl)aminosulfonyl, nitro, optionally substituted $C_{1-6}$-alkylthio and halogen, where any nitrogen-bound $C_{1-6}$-alkyl is optionally substituted with hydroxy, $C_{1-6}$-alkoxy, $C_{2-6}$-alkenyloxy, amino, mono- and di($C_{1-6}$-alkyl)amino, carboxy, $C_{1-6}$-alkylcarbonylamino, halogen, $C_{1-6}$-alkylthio, $C_{1-6}$-alkyl-sulphonyl-amino or guanidine;

each $R_6$ and $R_7$ is independently selected from the group consisting of hydrogen, optionally substituted $C_{1-6}$-alkyl, optionally substituted $C_{2-6}$-alkenyl, optionally substituted $C_{4-6}$- alkadienyl, optionally substituted $C_{2-6}$-alkynyl, optionally substituted $C_{1-6}$-alkoxycarbonyl, optionally substituted $C_{1-6}$-alkylcarbonyl, optionally substituted aryl, optionally substituted aryloxycarbonyl, optionally substituted arylcarbonyl, optionally substituted heteroaryl, optionally

substituted heteroaryloxycarbonyl, optionally substituted heteroarylcarbonyl, aminocarbonyl, mono- and di($C_{1-6}$-alkyl)aminocarbonyl, amino- $C_{1-6}$-alkyl-aminocarbonyl and mono- and di($C_{1-6}$-alkyl)amino- $C_{1-6}$-alkyl-aminocarbonyl; or $R_6$ and $R_7$ may together form a five- or six-membered nitrogen-containing ring;

or a pharmaceutically acceptable derivative thereof,

for use in the treatment of a kidney disease.

2. The composition for use according to claim 1, wherein said kidney disease presents with proteinuria.

3. The composition for use according to any of the preceding claims, wherein said kidney disease is a glomerular disease.

4. The composition for use according to any of the preceding claims, wherein said kidney disease is a disease affecting the podocytes of the glomeruli.

5. The composition for use according to any of the preceding claims, wherein said kidney disease is nephrotic syndrome (glomerulonephrosis).

6. The composition for use according to any of the preceding claims, wherein said nephrotic syndrome is primary nephrotic syndrome (primary glomerulonephrosis).

7. The composition for use according to claim 6, wherein said primary nephrotic syndrome is membranous glomerulonephritis (MGN) (or membranous nephropathy (MN)).

8. The composition for use according to claim 6, wherein said primary nephrotic syndrome is focal segmental glomerulosclerosis (FSGS).

9. The composition for use according to claim 6, wherein said primary nephrotic syndrome is membranoproliferative glomerulonephritis (MPGN) (mesangiocapillary glomerulonephritis).

10. The composition for use according to claim 9, wherein said membranoproliferative glomerulonephritis (MPGN) is selected from Type 1 MPGN and Type 2 MPGN.

11. The composition for use according to claim 6, wherein said primary nephrotic syndrome is rapidly progressive glomerulonephritis (RPGN) (crescentic GN).

12. The composition for use according to claim 6, wherein said primary nephrotic syndrome is minimal change disease (MCD).

13. The composition for use according to claim 5, wherein said nephrotic syndrome is secondary nephrotic syndrome (secondary glomerulonephrosis).

14. The composition for use according to claim 13, wherein said secondary nephrotic syndrome is caused by an underlying autoimmune disease.

15. The composition for use according to claim 13, wherein said secondary nephrotic syndrome is caused by an underlying disease selected from the group consisting of: Systemic lupus erythematosus (lupus nephritis), Diabetic nephropathy, Sarcoidosis, Sjögren's syndrome, Amyloidosis, Multiple myeloma, Vasculitis, Cancer and Genetic disorders (such as congenital nephrotic syndrome).

16. The composition for use according to claim 13, wherein said secondary nephrotic syndrome is caused by Diabetic nephropathy.

17. The composition for use according to claim 13, wherein said secondary nephrotic syndrome is caused by an infection.

18. The composition for use according to claim 13, wherein said secondary nephrotic syndrome is caused by a urinary tract infection.

19. The composition for use according to claim 17, wherein said infection is selected from the group consisting of HIV, syphilis, hepatitis such as hepatitis A, B and C, post-streptococcal infection, urinary schistosomiasis and Ebola.

20. The composition for use according to claim 13, wherein said secondary nephrotic syndrome is drug-induced, i.e. is drug-induced nephrotic syndrome.

21. The composition for use according to claim 20, wherein said drug-induced nephrotic syndrome is caused by a drug selected from the group consisting of antibiotics, nonsteroidal anti-inflammatory drugs (NSAID), radiocontrast media, anticancer drugs, antirheumatic drugs, antibody-based therapies, anti-TNF-α therapies, penicillamine, nicotine, lithium carbonate, gold and other heavy metals, ACE inhibitors and opiates (especially heroin).

22. The composition for use according to any of the preceding claims, wherein said kidney disease is an inflammatory kidney disease.

23. The composition for use according to any of the preceding claims, wherein said kidney disease is glomerulonephritis (GN).

24. The composition for use according to claim 23, wherein said glomerulonephritis is selected from the group consisting of IgA nephropathy (Berger's disease), IgM nephropathy, Post-infectious glomerulonephritis and Thin basement membrane disease.

25. The composition for use according to any of the preceding claims, wherein n = 1.

26. The composition for use according to any of the preceding claims, wherein each $R_1$, $R_2$ and $R_3$ is independently selected from the group consisting of -H, $-CH_3$, $-CF_3$, $-CCl_3$, -O-$C_{1-6}$ alkyl, -OH, $-OCH_3$, -NH-$C_{1-6}$ alkyl, -N($C_{1-6}$ alkyl)$_2$, -S-$C_{1-6}$ alkyl, -$^t$Bu, -CN, $-SO_3$-$C_{1-6}$ alkyl, $-NH_2$, $-NO_2$, $-CO_2H$, $-CO_2$-$C_{1-6}$ alkyl, -C(=O)-$C_{1-6}$ alkyl, -CHO, morpholine, pyrrolidine, pyrrole, or halogen.

27. The composition for use according to any of the preceding claims, wherein $R_1$ and/or $R_2$ is an electron-withdrawing group.

28. The composition for use according to any of the preceding claims, wherein $R_1$ is $CF_3$, $CCl_3$, F, Cl, CN or $NO_2$.

29. The composition for use according to any of the preceding claims, wherein $R_2$ and/or $R_3$ is hydrogen.

30. The composition for use according to any of the preceding claims, wherein each $R_4$ and $R_5$ is independently selected from the group consisting of -H, $C_{1-6}$ alkyl, halogen, -O($C_{1-6}$ alkyl), -NH($C_{1-6}$ alkyl) and -C(=O)$C_{1-6}$ alkyl.

31. The composition for use according to any of the preceding claims, wherein each $R_6$ and $R_7$ is independently selected from hydrogen, methyl, ethyl or propyl.

32. The composition for use according to any of the preceding claims, wherein $R_6$ and $R_7$ are hydrogen.

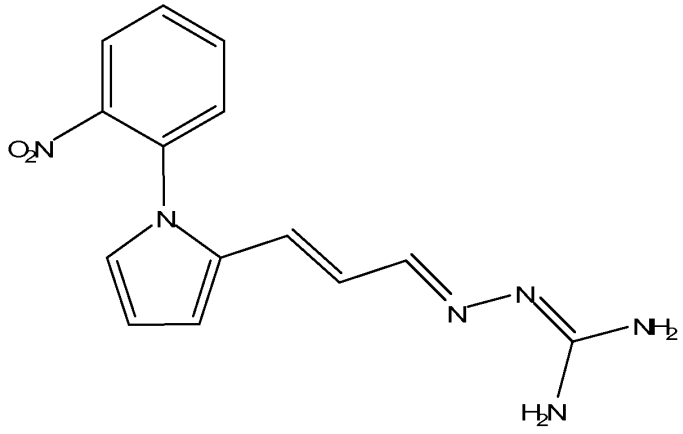
33. The composition for use according to any of the preceding claims, wherein $R_2$, $R_3$, $R_4$, $R_5$, $R_6$ and $R_7$ are hydrogen, and wherein $R_1$ is $CF_3$, $CCl_3$, F, Cl, $NO_2$ or CN.

34. The composition for use according to any of the preceding claims, wherein $R_2$, $R_3$, $R_4$, $R_5$, $R_6$ and $R_7$ are hydrogen, and wherein $R_1$ is $CF_3$, $CCl_3$, F, Cl, $NO_2$ or CN, and wherein n = 1.

35. The composition for use according to any of the preceding claims, wherein $R_3$ is located on the 4-position.

36. The composition for use according to any of the preceding claims, wherein $R_3$ is selected from F, Cl or Br.

37. The composition for use according to any of the preceding claims, wherein the compound is of formula (II)



formula (II)

including tautomeric and isomeric forms, such as enantiomeric forms, diastereomeric forms and racemic forms, thereof; or a pharmaceutically acceptable derivative thereof.

38. The composition for use according to any of the preceding claims, wherein said compound is selected from the group consisting of {3-[1-(2-nitrophenyl)-1H-pyrrol-2-yl]-allylidene}-aminoguanidine and (*E*)-*N*-*trans*-{3-[1-(2-nitrophenyl)-1H-pyrrol-2-yl]-allylidene}-aminoguanidine, or a pharmaceutically acceptable salt thereof.

39. The composition for use according to any of the preceding claims, wherein said pharmaceutically acceptable derivative thereof is a pharmaceutically acceptable salt of an inorganic acid or an organic acid.

40. The composition for use according to claim 39, wherein said organic acid is selected from the group consisting of: formic acid, acetic acid, trichloroacetic acid, trifluoroacetic acid, propionic acid, benzoic acid, cinnamic acid, citric acid, fumaric acid, glycolic acid, lactic acid, maleic acid, malic acid, malonic acid, mandelic acid, oxalic acid, picric acid, pyruvic acid, salicylic acid, succinic acid, methanesulfonic acid, ethanesulfonic acid, tartaric acid, ascorbic acid, pamoic acid, bismethylene salicylic acid, ethanedisulfonic acid, gluconic acid, citraconic acid, aspartic acid, stearic acid, palmitic acid, EDTA, glycolic acid, p-aminobenzoic acid, glutamic acid, benzenesulfonic acid and p-toluenesulfonic acid.

41. The composition for use according to any claims 39-40, wherein said organic acid is acetic acid, succinic acid, tartaric acid or propionic acid.

42. The composition for use according to claim 39, wherein said inorganic acid is selected from the group consisting of: hydrochloric acid, hydrobromic acid, hydroiodic acid, phosphoric acid, sulphuric acid and nitric acid.

43. The composition for use according to claim 41, wherein said organic acid is acetic acid.

44. The composition for use according to any of the preceding claims, wherein said compound is selected from the group consisting of {3-[1-(2-nitrophenyl)-1H-pyrrol-2-yl]-allylidene}- aminoguanidinium acetate and (*E*)-*N*-*trans*-{3-[1-(2-nitrophenyl)-1H-pyrrol-2-yl]-allylidene}- aminoguanidinium acetate.

45. The composition for use according to any of the preceding claims, wherein said compound:
    i) is an agonist of one or more MC receptors,
    ii) is an agonist of the MC1R and MC3R,
    iii) is an agonist of MC1R,
    iv) is an agonist of the MC1R and/or MC3R, but is not an agonist of MC2R and MC4R,
    v) is a biased agonist of the MC1R and MC3R,
    vi) is a biased agonist of the MC1R and MC3R, that mainly induces ERK1/2 phosphorylation, and insignificantly induces cAMP generation,
    vii) is a biased agonist of the MC1R and MC3R, that does not stimulate melanogenesis via the MC1R,
    viii) is an agonist of the MC1R and/or MC3R, with no effect on energy homeostasis such as food intake,
    ix) does not reduce food intake, such as does not reduce food intake via the MC3R and/or the MC4R, and/or
    x) does not induce melanogenesis.

46. The composition for use according to any of the preceding claims, wherein said composition comprises, separately or together, one or more additional active

pharmaceutical ingredients, such as one or more additional active pharmaceutical ingredients used for the treatment of kidney disease.

47. The composition for use according to any of the preceding claims, wherein said compound is administered in an amount of 1 mg to 1000 mg per day, such as 1 to 5 mg, 5 to 10 mg, 10 to 15 mg, 15 to 20 mg, 20 to 30 mg, 30 to 60 mg, 60 to 80 mg, 80 to 100 mg, 100 to 130 mg, 130 to 160 mg, 160 to 200 mg, 200 to 240 mg, 240 to 280 mg, 280 to 320 mg, 320 to 360 mg, 360 to 400 mg, 400 to 440 mg, 440 to 500 mg, 500 to 560 mg, 560 to 620 mg, 620 to 680 mg, 680 to 740 mg, 740 to 800 mg, 800 to 860 mg, 860 to 920 mg, 920 to 980 mg, 980 to 1000 mg, for example 500 to 1000 mg per day.

48. The composition for use according to any of the preceding claims, wherein said composition is pharmaceutically safe.