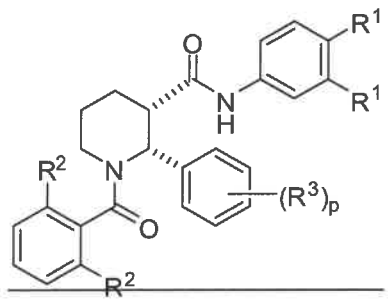
CLAIMS

1. A compound having the formula **(Ic)**, or a pharmaceutically acceptable salt thereof,



(Ic)

wherein

each R^1 is independently selected from the group consisting of

halogen, -CN, R^e, $-CO_2R^a$, $-CONR^aR^b$, $-C(O)R^a$, $-OC(O)NR^aR^b$, $-NR^bC(O)R^a$, $-NR^bC(O)_2R^e$, $-NR^aC(O)NR^aR^b$, $-NR^aR^b$, $-OR^a$, and $-S(O)_2NR^aR^b$; wherein each R^a and R^b is independently selected from hydrogen, C_{1-8} alkyl, and C_{1-8} haloalkyl, or when attached to the same nitrogen atom can be combined with the nitrogen atom to form a five or six-membered ring having from 0 to 2 additional heteroatoms as ring members selected from N, O or S; each R^e is independently selected from the group consisting of C_{1-8} alkyl, C_{1-8} haloalkyl, C_{3-6} cycloalkyl, heterocycloalkyl, aryl and heteroaryl, and wherein the aliphatic and cyclic portions of R^a, R^b and R^e are optionally further substituted with from one to three halogen, hydroxy, methyl, amino, alkylamino and dialkylamino groups; and optionally when two R^1 substituents are on adjacent atoms, are combined to form a fused five or six-membered carbocyclic ring;

each R^2 is independently selected from the group consisting of

halogen, -CN, R^f, $-CO_2R^d$, $-CONR^dR^e$, $-C(O)R^d$, $-OC(O)NR^dR^e$, $-NR^eC(O)R^d$, $-NR^eC(O)_2R^f$, $-NR^dC(O)NR^dR^e$, $-NR^dC(O)NR^dR^e$, $-NR^dR^e$, $-OR^d$, and $-S(O)_2NR^dR^e$; wherein each R^d and R^e is independently selected from hydrogen, C_{1-8} alkyl, and C_{1-8} haloalkyl, or when attached to the same nitrogen atom can be combined with the nitrogen atom to form a five or six-membered ring having from 0 to 2 additional heteroatoms as ring members selected from N, O or S; each R^f is independently selected from the group consisting of C_{1-8}

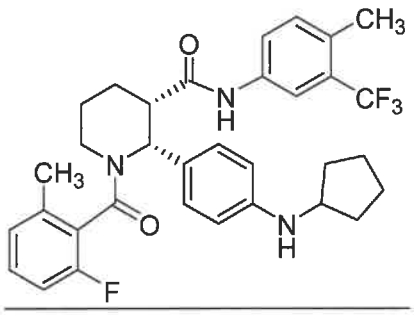
~~alkyl, C_{1-8} haloalkyl, C_{3-6} cycloalkyl, heterocycloalkyl, aryl and heteroaryl, and wherein the aliphatic and cyclic portions of R^d, R^e and R^f are optionally further substituted with from one to three halogen, hydroxy, methyl, amino, alkylamino and dialkylamino groups;~~

~~each R^3 is independently selected from the group consisting of~~

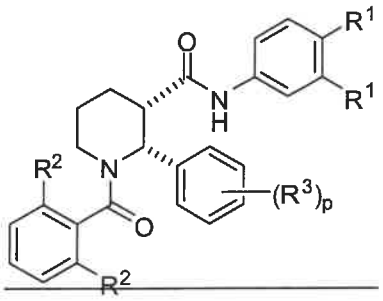
~~halogen, CN, R^i, CO_2R^g, $CONR^gR^h$, $C(O)R^g$, $OC(O)NR^gR^h$, $NR^hC(O)R^g$, $NR^hC(O)_2R^i$, $NR^gC(O)NR^gR^h$, NR^gR^h, OR^g, $S(O)_2NR^gR^h$, X^4-R^j, X^4-NR^gR^h, X^4-$CONR^gR^h$, X^4-$NR^hC(O)R^g$, NHR^j and $NHCH_2R^j$, wherein X^4 is a C_{1-4} alkylene; each R^g and R^h is independently selected from hydrogen, C_{1-8} alkyl, C_{3-6} cycloalkyl and C_{1-8} haloalkyl, or when attached to the same nitrogen atom can be combined with the nitrogen atom to form a five or six-membered ring having from 0 to 2 additional heteroatoms as ring members selected from N, O or S and is optionally substituted with one or two oxo; each R^i is independently selected from the group consisting of C_{1-8} alkyl, C_{1-8} haloalkyl, C_{3-6} cycloalkyl, heterocycloalkyl, aryl and heteroaryl; and each R^j is selected from the group consisting of C_{3-6} cycloalkyl, pyrrolinyl, piperidinyl, morpholinyl, tetrahydrofuranyl, and tetrahydropyranyl, and wherein the aliphatic and cyclic portions of R^g, R^h, R^i and R^j are optionally further substituted with from one to three halogen, methyl, CF_3, hydroxy, amino, alkylamino and dialkylamino groups; and~~

~~p is 0, 1 or 2; for use in therapy.~~

~~21~~. Use of a compound having the formula ~~(Ie),~~



or a pharmaceutically acceptable salt thereof.



~~(Ie)~~

~~wherein~~

~~each R^1 is independently selected from the group consisting of halogen, -CN, $-R^e$, $-CO_2R^a$, $-CONR^aR^b$, $-C(O)R^a$, $-OC(O)NR^aR^b$, $-NR^bC(O)R^a$, $-NR^bC(O)_2R^e$, $-NR^aC(O)NR^aR^b$, $-NR^aR^b$, $-OR^a$, and $-S(O)_2NR^aR^b$; wherein each R^a and R^b is independently selected from hydrogen, C_{1-8} alkyl, and C_{1-8} haloalkyl, or when attached to the same nitrogen atom can be combined with the nitrogen atom to form a five or six-membered ring having from 0 to 2 additional heteroatoms as ring members selected from N, O or S; each R^e is independently selected from the group consisting of C_{1-8} alkyl, C_{1-8} haloalkyl, C_{3-6} cycloalkyl, heterocycloalkyl, aryl and heteroaryl, and wherein the aliphatic and cyclic portions of R^a, R^b and R^e are optionally further substituted with from one to three halogen, hydroxy, methyl, amino, alkylamino and dialkylamino groups; and optionally when two R^1 substituents are on adjacent atoms, are combined to form a fused five or six-membered carbocyclic ring;~~

~~each R^2 is independently selected from the group consisting of halogen, -CN, $-R^f$, $-CO_2R^d$, $-CONR^dR^e$, $-C(O)R^d$, $-OC(O)NR^dR^e$, $-NR^eC(O)R^d$, $-NR^eC(O)_2R^f$, $-NR^dC(O)NR^dR^e$, $-NR^dC(O)NR^dR^e$, $-NR^dR^e$, $-OR^d$, and $-S(O)_2NR^dR^e$; wherein each R^d and R^e is independently selected from hydrogen, C_{1-8} alkyl, and C_{1-8} haloalkyl, or when attached to the same nitrogen atom can be combined with the nitrogen atom to form a five or six-membered ring having from 0 to 2 additional heteroatoms as ring members selected from N, O or S; each R^f is independently selected from the group consisting of C_{1-8} alkyl, C_{1-8} haloalkyl, C_{3-6} cycloalkyl, heterocycloalkyl, aryl and heteroaryl, and wherein the aliphatic and cyclic portions of R^d, R^e and R^f are optionally further substituted with from one to three halogen, hydroxy, methyl, amino, alkylamino and dialkylamino groups;~~

~~each R^3 is independently selected from the group consisting of halogen, -CN, $-R^i$, $-CO_2R^g$, $-CONR^gR^h$, $-C(O)R^g$, $-OC(O)NR^gR^h$, $-NR^hC(O)R^g$, $-NR^hC(O)_2R^i$, $-NR^gC(O)NR^gR^h$, $-NR^gR^h$, $-OR^g$, $-S(O)_2NR^gR^h$, $-X^4-R^j$, $-X^4-NR^gR^h$, $-X^4-CONR^gR^h$, $-X^4-NR^hC(O)R^g$, $-NHR^j$ and $-NHCH_2R^j$, wherein X^4 is a C_{1-4} alkylene; each R^g and R^h is independently selected from hydrogen, C_{1-8} alkyl, C_{3-6} cycloalkyl and C_{1-8} haloalkyl, or when attached to the same nitrogen atom can be combined with the nitrogen atom to form a five or six-membered ring having from 0 to 2 additional heteroatoms as ring members selected from N, O or S and is optionally substituted with one or two oxo; each R^i is independently selected from the group consisting of C_{1-8} alkyl, C_{1-8} haloalkyl, C_{3-6} cycloalkyl, heterocycloalkyl, aryl and heteroaryl; and each R^j is selected from the group consisting of C_{3-6} cycloalkyl, pyrrolinyl, piperidinyl, morpholinyl, tetrahydrofuranyl, and tetrahydropyranyl, and wherein the aliphatic and cyclic portions of R^g, R^h, R^i and R^j are optionally further substituted with from one to three halogen, methyl, CF_3, hydroxy, amino, alkylamino and dialkylamino groups; and~~

~~p is 0, 1 or 2~~ in the preparation of a medicament for treating ~~a human suffering from or susceptible to~~ complement 3 glomerulopathy in a human.

~~3. The use of claim 2, wherein~~

~~each R^1 is independently selected from the group consisting of halogen, -CN, $-R^e$, $-NR^aR^b$, and $-OR^a$; wherein each R^a and R^b is independently selected from hydrogen, C_{1-8} alkyl, and C_{1-8} haloalkyl, or when attached to the same nitrogen atom can be combined with the nitrogen atom to form a pyrrolidine ring; each R^e is independently selected from the group consisting of C_{1-8} alkyl, C_{1-8} haloalkyl, C_{3-6} cycloalkyl and wherein the aliphatic and cyclic portions of R^a, R^b and R^e are optionally further substituted with from one to three hydroxy, methyl, amino, alkylamino and dialkylamino groups; and optionally when two R^1 substituents are on adjacent atoms, are combined to form a fused five or six-membered carbocyclic ring;~~

~~each R^2 is independently selected from the group consisting of halogen, $-R^f$, and $-OR^d$; wherein each R^d is independently selected from hydrogen, C_{1-8} alkyl, and C_{1-8} haloalkyl, each R^f is independently selected from the group consisting of C_{1-8} alkyl, C_{1-8} haloalkyl, C_{3-6} cycloalkyl, heterocycloalkyl and heteroaryl, and wherein the aliphatic and cyclic portions of R^d and R^f~~

are optionally further substituted with from one to three halogen, hydroxy, methyl, amino, alkylamino and dialkylamino groups;

each R^3 is independently selected from the group consisting of halogen, R^i, $-CO_2R^g$, $-CONR^gR^h$, $-NR^hC(O)R^g$, $-NR^hC(O)_2R^i$, $-NR^gR^h$, $-OR^g$, $-X^4-R^j$, $-X^4-NR^gR^h$, $-X^4-CONR^gR^h$, $-X^4-NR^hC(O)R^g$, $-NHR^j$ and $-NHCH_2R^j$, wherein X^4 is a C_{1-4} alkylene; each R^g and R^h is independently selected from hydrogen, C_{1-8} alkyl, C_{3-6} cycloalkyl and C_{1-8} haloalkyl, or when attached to the same nitrogen atom can be combined with the nitrogen atom to form a five or six-membered ring having from 0 to 2 additional heteroatoms as ring members selected from N, O or S and is optionally substituted with one or two oxo; each R^i is independently selected from the group consisting of C_{1-8} alkyl, C_{1-8} haloalkyl, C_{3-6} cycloalkyl, heterocycloalkyl, aryl and heteroaryl; and each R^j is selected from the group consisting of C_{3-6} cycloalkyl, pyrrolinyl, piperidinyl, morpholinyl, tetrahydrofuranyl, and tetrahydropyranyl, and wherein the aliphatic and cyclic portions of R^g, R^h, R^i and R^j are optionally further substituted with from one to three halogen, methyl, CF_3, hydroxy, amino, alkylamino and dialkylamino groups; and

p is 1.

4. The use of claim 2, wherein

each R^1 is independently selected from the group consisting of C_{1-8} alkyl and C_{1-8} haloalkyl;

each R^2 is independently selected from the group consisting of halogen and C_{1-8} alkyl;

each R^3 is $-NR^gR^h$ wherein each R^g and R^h is independently selected from hydrogen and C_{3-6} cycloalkyl; and

p is 1.

5. The use of claim 2, wherein

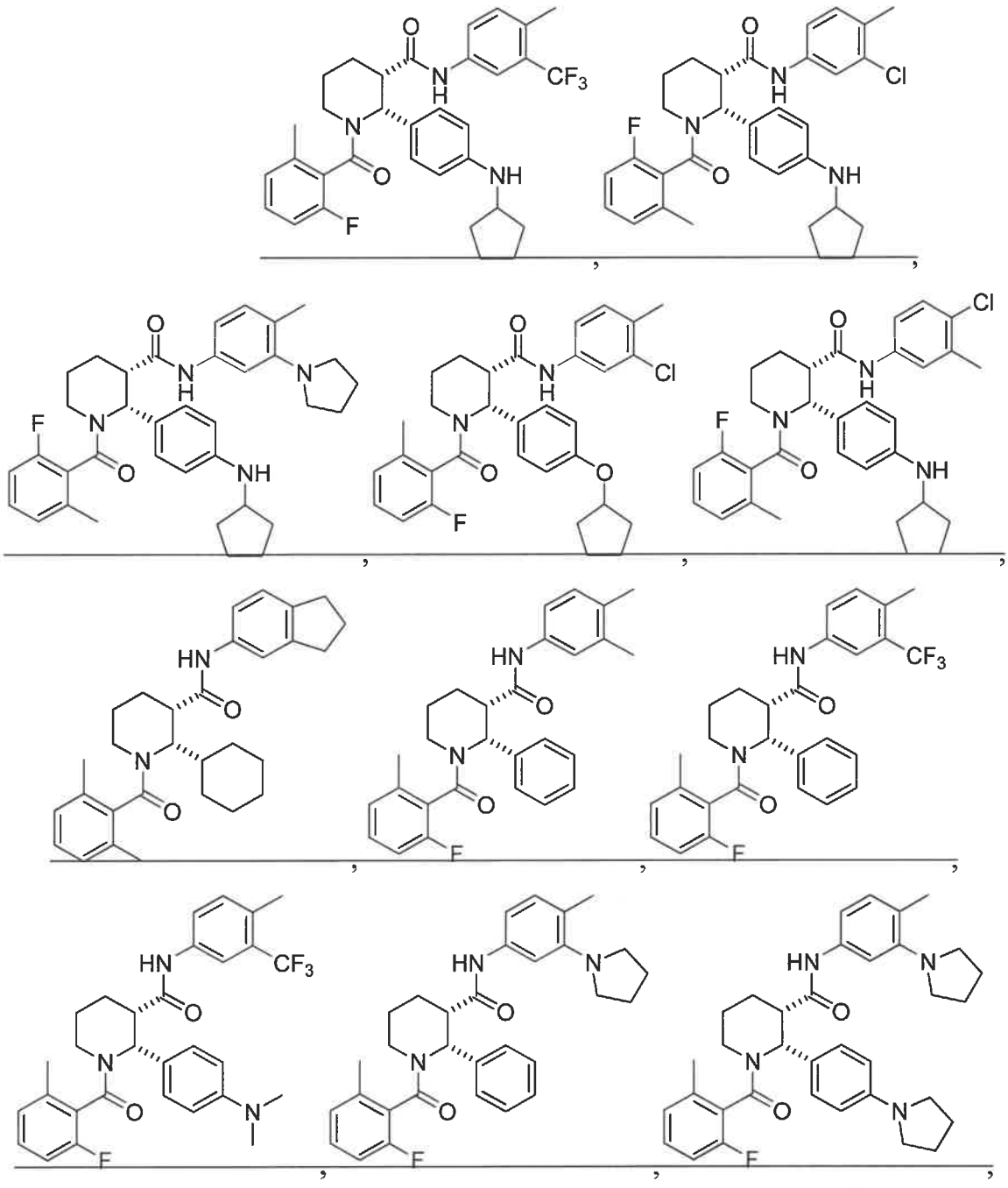
each R^1 is independently selected from the group consisting of C_{1-3} alkyl and C_{1-3} haloalkyl;

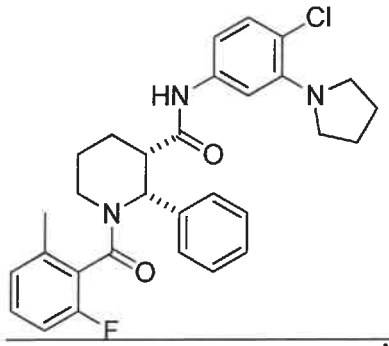
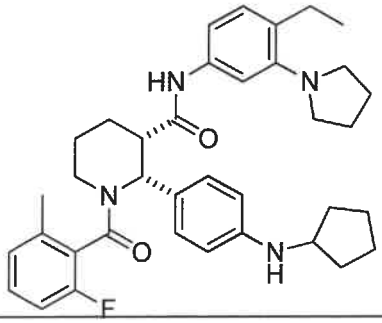
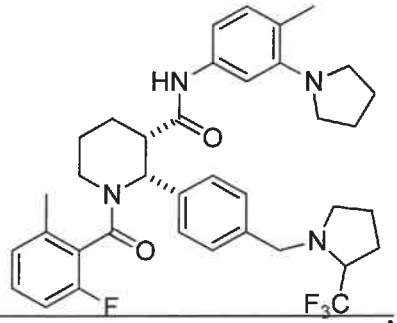
each R^2 is independently selected from the group consisting of halogen and C_{1-3} alkyl;

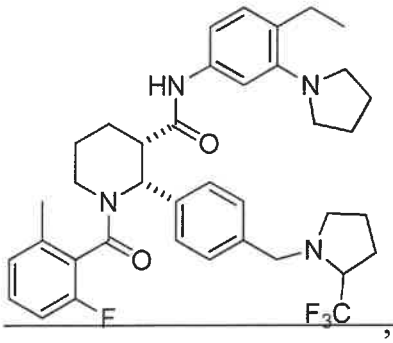
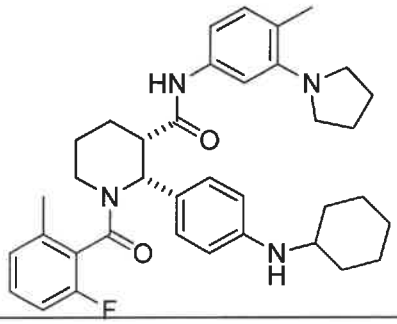
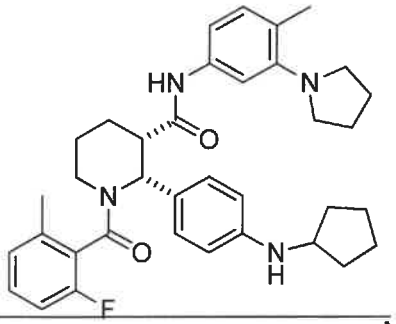
each R^3 is $-NR^gR^h$ wherein each R^g and R^h is independently selected from hydrogen and C_{4-6} cycloalkyl; and

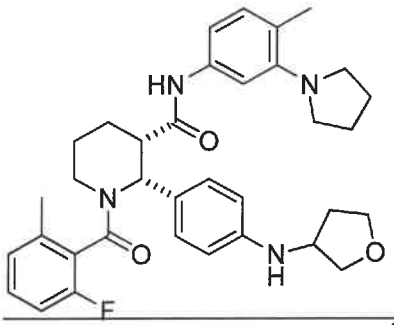
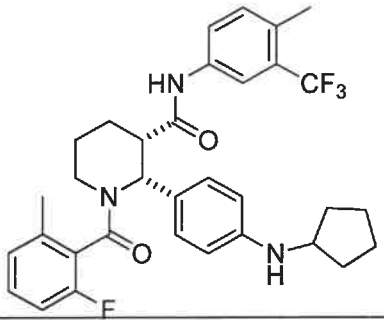
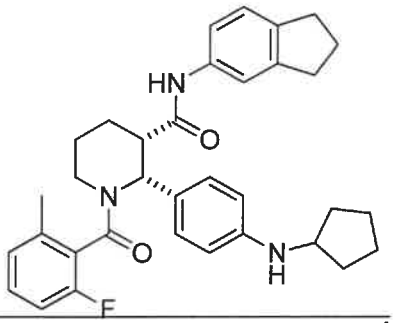
p is 1.

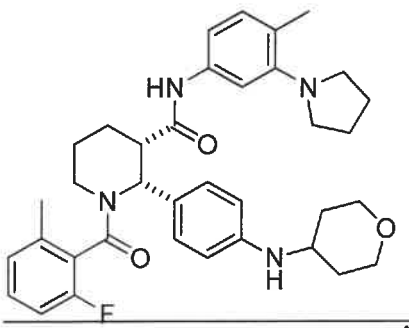
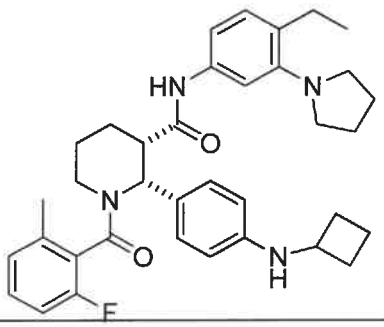
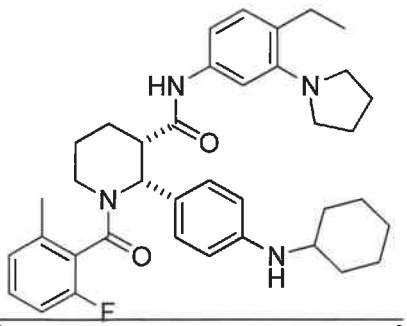
~~6. The use of claim 2, wherein the compound is selected from the group consisting of~~

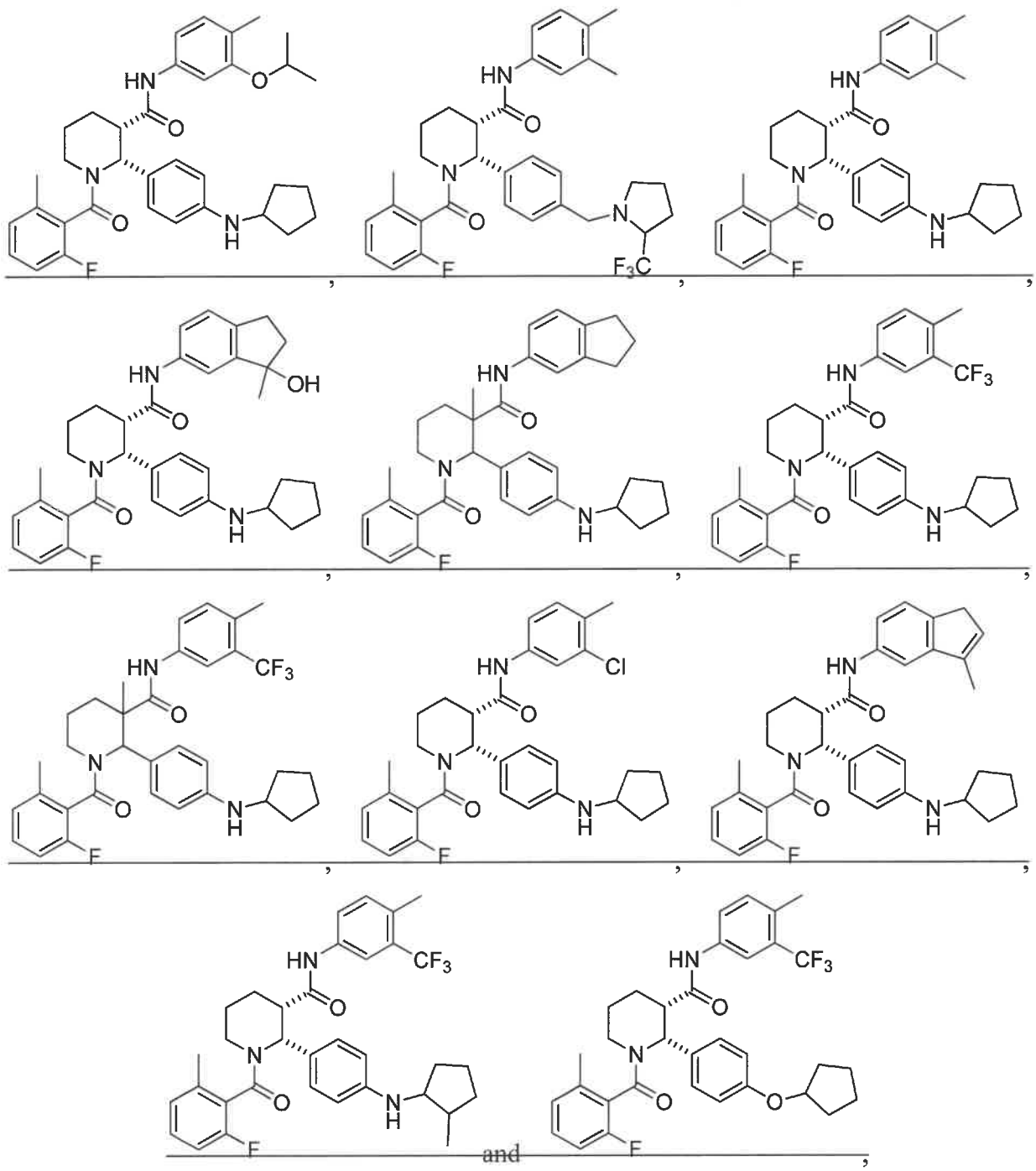


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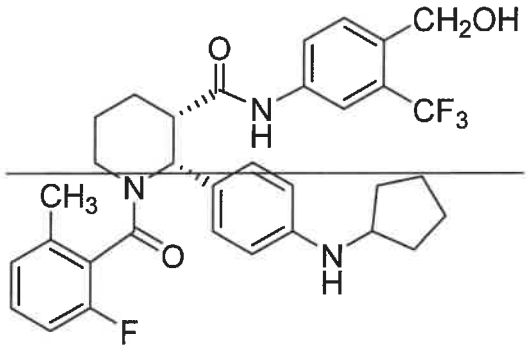
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and

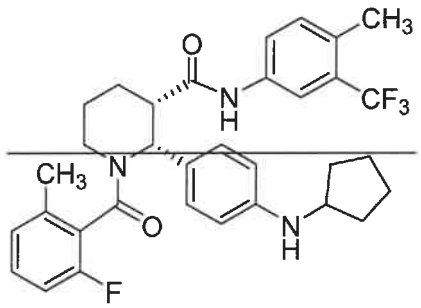
~~or a pharmaceutically acceptable salt thereof.~~

~~7. The use of claim 2, wherein the compound is~~



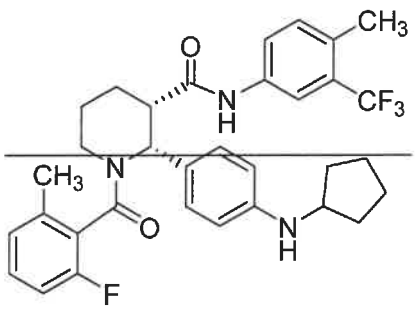
~~or a pharmaceutically acceptable salt thereof.~~

~~8. The use of any one of claims 2 to 6, wherein the compound is~~



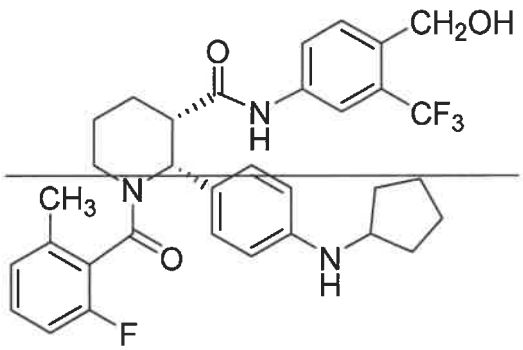
~~or a pharmaceutically acceptable salt thereof.~~

~~9~~2. The use of claim 1, wherein the medicament ~~A compound having the following formula:~~



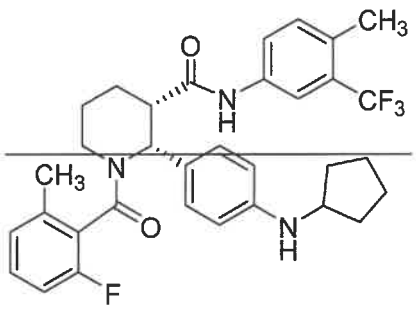
~~or a pharmaceutically acceptable salt thereof for use in therapy, for~~ slowings the rate of decline in Estimated Glomerular Filtration Rate (eGFR) in a human suffering from ~~or susceptible to~~ C3 glomerulopathy.

~~10. A compound having the following formula:~~



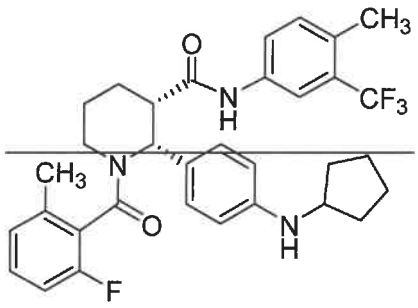
~~or a pharmaceutically acceptable salt thereof for use in therapy, for example slowing the rate of decline in Estimated Glomerular Filtration Rate (eGFR) in a human suffering from or susceptible to C3 glomerulopathy.~~

~~11~~3. The use of claim 1, wherein the medicament ~~A compound having the following formula:~~



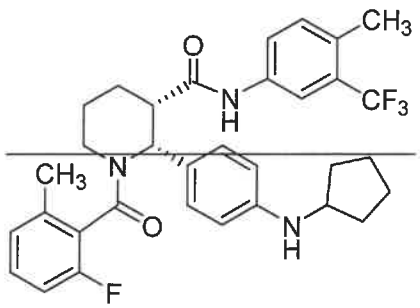
~~or a pharmaceutically acceptable salt thereof for use in therapy, for example~~ reduces~~ing~~ glomerular inflammation in a human suffering from ~~or susceptible to~~ C3 glomerulopathy.

~~12. A compound having the following formula:~~



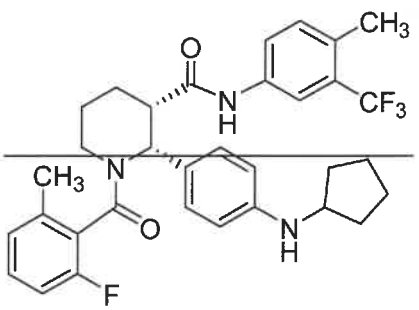
~~or a pharmaceutically acceptable salt thereof for use in therapy, for example clearing glomerular endocapillary proliferation in a human suffering from or susceptible to C3 glomerulopathy.~~

~~134~~. The use of claim 1, wherein the medicament ~~A compound having the following formula:~~



~~or a pharmaceutically acceptable salt thereof for use in therapy, for example~~ reduce~~sing~~ glomerular inflammatory macrophages in a human suffering from ~~or susceptible to C3~~ glomerulopathy ~~comprising administering to the human an effective amount~~.

~~145~~. The use of claim 1, wherein the medicament ~~A compound having the following formula:~~



~~or a pharmaceutically acceptable salt thereof for use in therapy, for example~~ reduce~~s~~ing proteinuria in a human suffering from ~~or susceptible to~~ C3 glomerulopathy.

~~15. The use or compound for use of any one of claims 2 to 14 wherein the human suffers from complement 3 glomerulonephritis.~~

~~16~~. The use ~~of compound for use~~ of any one of claims ~~2~~1 to ~~1~~45 wherein the human suffers from progressive complement 3 glomerulonephritis.

~~17~~. The use ~~or compound for use~~ of any one of claims ~~2~~1 to ~~1~~45 wherein the human suffers from recurrent complement 3 glomerulonephritis after a renal transplant.

~~18~~. The use ~~or compound for use~~ of any one of claims ~~2~~1 to ~~1~~45 wherein the human suffers from dense deposit disease.

~~19~~. The use ~~or compound for use~~ of any one of claims ~~2~~1 to ~~1~~45 wherein the complement 3 glomerulopathy is refractory to other treatment.

~~21~~0. The use ~~or compound for use~~ of any one of claims ~~2~~1 to ~~1~~9 wherein the human has refractory disease to immunosuppressive drugs.

~~21~~1. The use ~~or compound for use~~ of any one of claims ~~2~~1 to ~~2~~10 wherein the human has refractory disease to one or more of rituximab, cyclophosphamide, mycophenolate mofetil, tacrolimus, and steroids.

~~22. The use or compound for use of any one of claims 2 to 21 wherein the human shows improved health-related quality of life.~~

~~23. The use or compound for use of claim 22 wherein the health-related quality of life is based on Short Form-36 version 2 (SF-36 v2) or EuroQOL-5D-5L (EQ-5D-5L) assessment.~~

1~~24~~. The use ~~or compound for use~~ of any one of claims ~~2~~1 to ~~23~~11 wherein the ~~compound~~medicament is ~~f~~to~~r~~ be administe~~r~~ed~~ation~~ twice daily.

~~25~~13. The use ~~or compound for use~~ of any one of claims ~~2~~1 to ~~23~~11 wherein the ~~compound~~medicament is to be administered~~ation~~ once a day.

~~26~~14. The use ~~or compound for use~~ of any one of claims ~~2~~1 to ~~23~~11 wherein the ~~compound~~medicament is ~~f~~to be administration orally.

~~27~~15. The use ~~or compound for use~~ of any one of claims ~~2~~1 to ~~23~~11 wherein 30 mg of the compound is to be administered to the human twice daily.

~~28. The use or compound for use of any one of claims 2 to 23 wherein 20 mg of the compound is to be administered to the human twice daily.~~

~~29. The use or compound for use of any one of claims 2 to 23 wherein 10 mg of the compound is to for administration to the human twice daily.~~

~~30~~16. The use ~~or compound for use~~ of any one of claims ~~2~~1 to ~~29~~11, wherein the human has a Complement factor H related protein 5 (CFHR5) mutation.

~~31~~7. The use ~~or compound for use~~ of any one of claims ~~2~~1 to ~~30~~11 wherein the ~~compound~~medicament is ~~f~~to be administered~~ation~~ to the human for 12 weeks.

~~32. The use or compound for use of any one of claims 2 to 30 wherein the compound is for administration to the human for 26 weeks.~~

~~33. The use or compound for use of any one of claims 2 to 30 wherein the compound is for administration to the human for 52 weeks.~~

~~34. The use or compound for use of any one of claims 2 to 30 wherein the human is to receive chronic treatment.~~

~~35. The use or compound for use of any one of claims 2 to 34 wherein a therapeutically effective amount of one or more additional therapeutic agents is to be administered to the human.~~

~~36. The use or compound for use of claim 35, wherein the one or more additional therapeutic agents is selected from immunosuppressive drugs, angiotensin-converting enzyme (ACE) inhibitors, angiotensin II type-1 receptor blockers (ARBs) and corticosteroids.~~

~~37. The use of compound for use of claim 35, wherein the one or more additional therapeutic agents is selected from the group consisting of cyclophosphamide, mycophenolate mofetil, rituximab, eculizumab, tacrolimus, belimumab, OMS721, ACH-4471, AMY-101, Acthar Gel, SAND-5, corticotropin, CDX-1135, ramipril, perindopril, lisinopril, perindopril arginine, captopril, spirapril, quinapril, enalapril, imidapril, fosinopril, zofenopril, benazepril, trandolapril, verapamil, benazepril, amlodipine, trandolapril, P-003, cilazapril, delapril, moexipril, quinapril, fosinopril, temocapril, losartan, candesartan, irbesartan, telmisartan, olmesartan, valsartan, azilsartan, telmisartan, fimasartan, EMA-401, azilsartan medoxomil potassium, sparsentan, candesartan cilexetil, olmesartan medoxomil, TRV-027, losartan potassium, YH-22189, azilsartan trimethylethanolamine, allisartan isoproxil, and eprosartan.~~