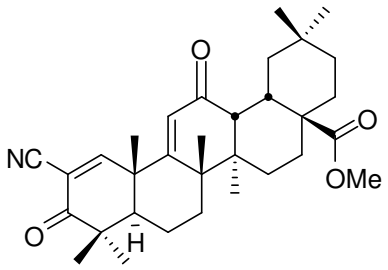
CLAIMS

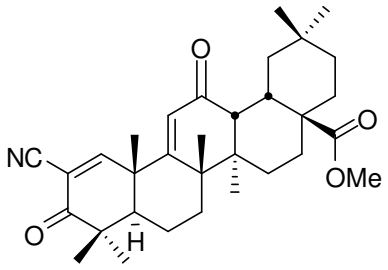
1. Use of a compound as shown by the following formula:



,

or a pharmaceutically acceptable salt thereof in the preparation of a medicament for treating or preventing Alport syndrome in a human patient.

2. Use of a compound as shown by the following formula:



,

or a pharmaceutically acceptable salt thereof in the preparation of a medicament for improving kidney function of a human patient who has been diagnosed with Alport syndrome.

3. The use of claim 2, further defined as for treating chronic kidney disease in a patient who has been diagnosed with Alport syndrome.

4. The use of any one of claims 1 to 3, wherein the patient does not have cardiovascular disease.

5. The use of any one of claims 1 to 4, wherein the cardiovascular disease is left-sided myocardial disease, atherosclerosis, restenosis, thrombosis, or pulmonary hypertension.

6. The use of any one of claims 1 to 5, wherein the patient does not have Stage 4 or higher chronic kidney disease.

7. The use of any one of claims 1 to 5, wherein the patient does not have an estimated glomerular filtration rate (eGFR) less than 45 mL/min/1.73 m^2.

8. The use of any one of claims 1 to 5, wherein the patient does not have an elevated albumin/creatinine ratio (ACR) is greater than 2000 mg/g.

9. The use of any one of claims 1 to 8, wherein the patient does not have diabetes.

10. The use of any one of claims 1 to 8, wherein the patient does not have a complication associated with diabetes.

11. The use of any one of claims 1 to 10, wherein the patient does not have insulin resistance.

12. The use of any one of claims 1 to 11, wherein the patient does not have fatty liver disease.

13. The use of any one of claims 1 to 12, wherein the patient does not have hepatic impairment.

14. The use of any one of claims 1 to 13, wherein the patient is not overweight.

15. The use of any one of claims 1 to 14, wherein the patient does not have cancer.

16. The use of any one of claims 1 to 15, wherein the patient does not have chronic obstructive pulmonary disease (COPD).

17. The use of any one of claims 1 to 16, wherein the patient is not a smoker.

18. The use of any one of claims 1 and 4 to 17, wherein the patient has impaired renal function.

19. The use of any one of claims 1 to 18, wherein the patient has elevated levels of at least one biomarker associated with renal disease, which is selected from serum creatinine, cystatin C and uric acid.

20. The use of any one of claims 1 to 19, wherein the Alport syndrome is X-linked, autosomal recessive, or autosomal dominant.

21. The use of any one of claims 1 to 20, wherein the patient has a deletion or nonsense mutation of *COL4A5,* a splicing mutation of *COL4A5,* a missense mutation of *COL4A5,* a mutation in *COL4A3,* or a mutation in *COL4A4*.

22. The use of any one of claims 1 to 21, wherein the patient exhibits microhematuria.

23. The use of any one of claims 1 to 22, wherein the patient further exhibits microalbuminuria.

24. The use of any one of claims 1 to 23, wherein the patient further exhibits proteinuria.

25. The use of any one of claims 1 to 24, wherein the patient is less than 75 years old.

26. The use of any one of claims 1 to 25, wherein the patient does not have the following characteristics:

(A) a cardiovascular disease;

(B) an elevated baseline B-type natriuretic peptide (BNP) level;

(C) an estimated glomerular filtration rate (eGFR) < 45 mL/min/1.73 m^2; and

(D) an elevated albumin/creatinine ratio (ACR) > 2000 mg/g.

27. The use of any one of claims 1 to 26, wherein at least a portion of the compound is present as a crystalline form having an X-ray diffraction pattern (CuKα) comprising significant diffraction peaks at about 8.8, 12.9, 13.4, 14.2 and 17.4 °2θ.

28. The use of claim 27, wherein the X-ray diffraction pattern (CuKα) is substantially as shown in FIG. 1A or FIG. 1B.

29. The use of any one of claims 1 to 26, wherein at least a portion of the compound is present as an amorphous form having an X-ray diffraction pattern (CuKα) with a peak at approximately 13.5 °2θ, substantially as shown in FIG. 1C, and a transition glass temperature (T_g).

30. The use of claim 29, wherein the T_g value is in the range of ~~about~~ 120°C to ~~about~~ 135°C.

31. The use of claim 30, wherein the T_g value is in the range of ~~about~~ 125 C to ~~about~~ 130°C.

32. The use of any one of claims 1 to 31, wherein the compound is to be used in an amount from ~~about~~ 0.1 mg to ~~about~~ 300 mg of the compound or a pharmaceutically acceptable salt thereof per day.

33. The use of any one of claims 1 to 32, wherein the medicament is to be administered orally, intraarterially or intravenously.

34. The use of any one of claims 1 to 33, wherein the medicament is formulated as a hard or soft capsule or a tablet.

35. The use of any one of claims 1 to 33, wherein the medicament is formulated as a solid dispersion comprising (i) the compound and (ii) an excipient.

36. The use of claim 35, wherein the excipient is a methacrylic acid – ethyl acrylate copolymer.

37. The use of any one of claims 1 to 36, which is characterized in that the medicament is to be used in combination with a second drug.

38. The use of claim 37, wherein the second drug is an angiotensin-converting enzyme inhibitor, an angiotensin receptor blocker, or an aldosterone antagonist.

39. The use of claim 38, wherein the angiotensin-converting enzyme inhibitor is Ramipril, enalapril, Lisinopril, benazepril, fosinopril, quinapril, cilazapril, or trandolapril.

40. The use of claim 38, wherein the angiotensin receptor blocker is losartan, candesartan, irbesartan, telmisartan, or valsartan.

41. The use of claim 38, wherein the aldosterone antagonist is spironolactone.