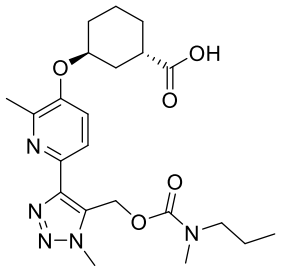
WHAT IS CLAIMED IS:

1. Use of Compound A:



Compound A;

or a pharmaceutically acceptable salt thereof in the manufacture of medicament for treating interstitial lung disease, wherein Compound A or a pharmaceutically acceptable salt is to be administered to a subject in need thereof at one or more daily dosages lower than the standard daily dosage during a dose titration period of treatment then increasing the daily dosage to the standard daily dosage of Compound A, wherein the standard daily dosage of is about 240 mg/day of Compound A or an equivalent amount of a pharmaceutically acceptable salt thereof.

2. The use of claim 1, wherein the dose titration period of treatment is completed within 21 days.

3. The use of claim 1 or 2, wherein the dose titration period of treatment is completed within 14 days.

4. The use of any one of claims 1 to 3, wherein the dose titration period of treatment is completed within 10 days.

5. The use of any one of claims 1 to 4, wherein the dose titration period of treatment is completed within 7 days.

6. The use of any one of claims 1 to 5, wherein the dose titration period of treatment is completed within 5 days.

7. The use of any one of claims 1 to 6, wherein the dose titration period of treatment comprises an initial period of treatment and a second period of treatment.

8. The use of claim 7, wherein the initial period of treatment is completed within 7 days.

9. The use of claim 7 or 8, wherein the initial period of treatment is completed within 4 days.

10. The use of any one of claims 7 to 9, wherein the initial period of treatment is completed within 3 days.

11. The use of any one of claims 7 to 10, wherein the initial period of treatment is completed within 2 days.

12. The use of any one of claims 7 to 11, wherein the initial period of treatment is completed within 1 day.

13. The use of any one of claims 7 to 12, wherein the second period of treatment is completed within 7 days.

14. The use of any one of claims 7 to 13, wherein the second period of treatment is completed within 4 days.

15. The use of any one of claims 7 to 14, wherein the second period of treatment is completed within 3 days.

16. The use of any one of claims 7 to 15, wherein the second period of treatment is completed within 2 days.

17. The use of any one of claims 7 to 16, wherein the second period of treatment is completed within 1 day.

18. The use of any one of claims 7 to 17, wherein the dose titration period of treatment further comprises a third period of treatment.

19. The use of claim 18, wherein the third period of treatment is completed within 7 days.

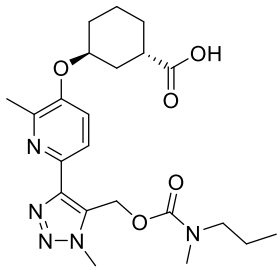
20. The use of claim 18 or 19, wherein the third period of treatment is completed within 4 days.

21. The use of any one of claims 18 to 20, wherein the third period of treatment is completed within 3 days.

22. The use of any one of claims 18 to 21, wherein the third period of treatment is completed within 2 days.

23. The use of any one of claims 18 to 22, wherein the third period of treatment is completed within 1 day.

24. Use of Compound A:



Compound A;

in the manufacture of a medicament for treating interstitial lung disease, wherein Compound A is to be administered to a subject in need thereof at a daily dosage lower than the standard daily dosage during an initial period of treatment, then a daily dosage higher than the initial period but lower than the standard daily dosage is to be administered for a second period of treatment, then increasing the dosage to the standard daily dosage, wherein the standard daily dosage is about 240 mg/day of Compound A or an equivalent amount of a pharmaceutically acceptable salt thereof.

25. The use of claim 24, wherein the initial period of treatment and the second period of treatment are each independently completed within 7 days.

26. The use of claim 24 or 25, wherein the initial period of treatment and the second period of treatment are each independently completed within 4 days.

27. The use of any of claims 24 to 26, wherein the initial period of treatment and the second period of treatment are each independently completed within 3 days.

28. The use of any of claims 24 to 27, wherein the initial period of treatment and the second period of treatment are each independently completed within 2 days.

29. The use of any of claims 24 to 28, wherein the initial period of treatment and the second period of treatment are each independently completed within 1 day.

30. The use of claim 24, wherein increasing the dosage comprises administration of the dosage at a daily dosage lower than the standard daily dosage for a third period of treatment, then administration of the standard daily dosage for a fourth period of treatment.

31. The use of claim 30, wherein the third period of treatment is completed within 7 days.

32. The use of claim 30 or 31, wherein the third period of treatment is completed within 4 days.

33. The use of any of claims 30 to 32, wherein the third period of treatment is completed within 3 days.

34. The use of any of claims 30 to 33, wherein the third period of treatment is completed within 2 days.

35. The use of any of claims 30 to 34, wherein the third period of treatment is completed within 1 day.

36. The use of any one of claims 1 to 35, wherein Compound A, or the pharmaceutically acceptable salt thereof, is to be administered once a day.

37. The use of any one of claims 1 to 36, wherein Compound A, or the pharmaceutically acceptable salt thereof, is to be administered twice a day.

38. The use of any one of claims 1 to 37, wherein Compound A, or the pharmaceutically acceptable salt thereof, is to be administered orally.

39. The use of any one of claims 1 to 38, wherein Compound A, or the pharmaceutically acceptable salt thereof, is to be administered as a tablet.

40. The use of any one of claims 1 to 39, wherein the subject is being treated with one or more additional therapies for interstitial lung disease.

41. The use of claim 40 wherein the one or more additional therapies is pirfenidone.

42. The use of claim 40, wherein the one or more additional therapies is ninedanib.

43. The use of any one of claims 1 to 42, wherein Compound A, or the pharmaceutically acceptable salt thereof, is to be administered with food.

44. The use of any one of claims 1 to 43, wherein Compound A, or the pharmaceutically acceptable salt thereof, is to be administered without food.

45. The use of any one of claims 1 to 44, wherein the interstitial lung disease is idiopathic pulmonary fibrosis (IPF).

46. The use of any one of claims 1 to 44, wherein the interstitial lung disease is progressive pulmonary fibrosis (PPF).

47. The use of any one of claims 1 to 46, wherein the standard daily dosage of Compound A is about 120 mg twice daily or an equivalent amount of a pharmaceutically acceptable salt thereof.

48. The use of any one of claims 7 to 47, wherein about 20 mg/day of Compound A, or an equivalent amount of a pharmaceutically acceptable salt thereof, is to be administered during the initial period of treatment.

49. The use of any one of claims 7 to 48, wherein about 10 mg twice daily of Compound A, or an equivalent amount of a pharmaceutically acceptable salt thereof, is to be administered during the initial period of treatment.

50. The use of any one of claims 7 to 49, about 60 mg/day of Compound A, or an equivalent amount of a pharmaceutically acceptable salt thereof, is to be administered during the second treatment period.

51. The use of any one of claims 7 to 50, wherein about 30 mg twice daily of Compound A, or an equivalent amount of a pharmaceutically acceptable salt thereof, is to be administered during the second treatment period.

52. The use of any one of claims 18 to 23 or 31 to 51, wherein about 120 mg/day of Compound A, or an equivalent amount of a pharmaceutically acceptable salt thereof, is to be administered during the third treatment period.

53. The use of any one of claims 18 to 23 or 31 to 52, wherein about 60 mg twice daily of Compound A, or an equivalent amount of a pharmaceutically acceptable salt thereof, is to be administered during the third treatment period.

54. The use of any one of claims 1 to 53, wherein Compound A comprises the crystal form characterized by at least one of the following:

a) single crystal structure having unit cell parameters substantially equal to

Crystal system, space group	Triclinic, P1	
Unit cell dimensions	a = 6.53 ± 0.10 Å	alpha = 92.8 ± 1.0°
	b = 13.06 ± 0.10 Å	beta = 95.5 ± 1.0°
	c = 14,04 ± 0.10 Å	gamma = 93.0 ±1.0°
Volume	1189(20) $Å^3$	
Density (calculated)	1.239 g/cm^3	

Temperature room temperature

wherein measurement of the single crystal structure is at room temperature;

b) a powder x-ray diffraction pattern substantially the same as shown in FIG. 1;

c) a powder x-ray diffraction pattern comprising 2 or more peaks at 2θ values selected from 6.4±0.2, 6.8±0.2, 9.6±0.2, 13.6±0.2, 15.7±0.2, 18.2±0.2, 19.9±0.2, 21.6±0.2, 24.8±0.2 and 26.8±0.2 (obtained at room temperature and CuKα λ=1.5418 Å);

d) a powder x-ray diffraction pattern comprising 3 or more peaks at 2θ values selected from 6.4±0.2, 6.8±0.2, 9.6±0.2, 13.6±0.2, 14.1±0.2, 14.5±0.2, 14.7±0.2, 15.7±0.2, 18.2±0.2 , 18.7±0.2, 19.2±0.2, 19.9±0.2, 20.5±0.2, 21.6±0.2, 22.5±0.2, 23.1±0.2, 24.1±0.2, 24.8±0.2, 25.6±0.2, 26.8±0.2, 27.1±0.2 and 27.8±0.2 (obtained at room temperature and CuKα λ=1.5418 Å);

e) a differential scanning calorimetry thermogram substantially similar to the one as shown in FIG. 2;

f) a differential scanning calorimetry thermogram with an endotherm having an onset at about 152 ºC; and/or

g) a thermal gravimetric analysis thermogram substantially similar to the one as shown in FIG. 3.

55. The use of any one of claims 1 to 54, wherein the subject experiences a smaller decline in forced vital capacity (FVC) after a treatment period compared to an untreated subject.

56. The use of any one of claims 1 to 55, wherein the subject experiences a greater time to first disease progression event after a treatment period than an untreated subject, wherein the first disease progression event is selected from:

absolute predicted forced vital capacity (ppFVC) decline of ≥ 10% from baseline;

acute exacerbation of lung fibrosis;

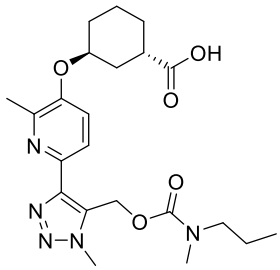
respiratory-related hospitalization; and

all-cause mortality.

57. The use of any one of claims 1 to 56, wherein the subject experiences a smaller increase in cough domain score as measured by the Living with Pulmonary Fibrosis (L-PF) questionnaire over a treatment period than an untreated subject.

58. The use of any one of claims 1 to 57, wherein the subject experiences a smaller increase in dyspnea score as measured by the Living with Pulmonary Fibrosis (L-PF) questionnaire over a treatment period than an untreated subject.

59. Compound A:



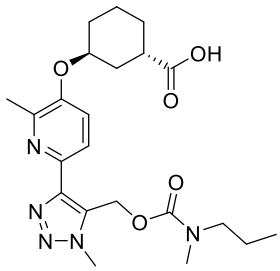
Compound A;

or a pharmaceutically acceptable salt thereof, for use in treating interstitial lung disease in a subject in need thereof, wherein Compound A is to be administered to the subject at a dosage lower than the standard daily dosage during an initial period of treatment then the dosage is increased to the standard daily dosage of Compound A, wherein the standard daily dosage of Compound A is about 240 mg/day.

60. The compound for use of claim 59, wherein the interstitial lung disease is idiopathic pulmonary fibrosis (IPF).

61. The compound for use of claim 59, wherein the interstitial lung disease is progressive pulmonary fibrosis (PPF).

62. Use of Compound A:



Compound A;

or an equivalent amount of a pharmaceutically acceptable salt thereof in the manufacture of medicament for treating interstitial lung disease, wherein Compound A is to be administered to a subject in need thereof about 240 mg/day.

63. The use of claim 62, wherein Compound A, or the pharmaceutically acceptable salt thereof, is to be administered once daily.

64. The use of claim 62, wherein Compound A, or the pharmaceutically acceptable salt thereof, is to be administered twice daily.

65. The use of claim 64, wherein about 120 mg of Compound A, or an equivalent amount of a pharmaceutically acceptable salt thereof, is to be administered twice daily.

66. The use of any one of claims 62 to 65, wherein Compound A, or the pharmaceutically acceptable salt thereof, is to be administered orally.

67. The use of claim 66, wherein Compound A, or the pharmaceutically acceptable salt thereof, is to be administered as a tablet.

68. The use of any one of claims 62 to 67, wherein the subject is concomitantly being treated with one or more therapies for interstitial lung disease.

69. The use claim 68, wherein the one or more pharmacological therapies is pirfenidone.

70. The use of claim 68, wherein the one or more pharmacological therapies is ninedanib.

71. The use of any one of claims 62 to 70, wherein Compound A, or the pharmaceutically acceptable salt thereof, is to be administered with food.

72. The use of any one of claims 62 to 71, wherein Compound A, or the pharmaceutically acceptable salt thereof, is to be administered without food.

73. The use of any one of claims 62 to 72, wherein the interstitial lung disease is idiopathic pulmonary fibrosis (IPF).

74. The use of any one of claims 62 to 72, wherein the interstitial lung disease is progressive pulmonary fibrosis (PPF).

75. The use of any one of claims 62 to 74, wherein Compound A comprises the crystal form characterized by at least one of the following:

a) single crystal structure having unit cell parameters substantially equal to

Crystal system, space group	Triclinic, P1	
Unit cell dimensions	a = 6.53 ± 0.10 Å	alpha = 92.8 ± 1.0°
	b = 13.06 ± 0.10 Å	beta = 95.5 ± 1.0°
	c = 14,04 ± 0.10 Å	gamma = 93.0 ±1.0°
Volume	1189(20) $Å^3$	
Density (calculated)	1.239 g/cm^3	
Temperature	room temperature	

wherein measurement of the single crystal structure is at room temperature;

b) a powder x-ray diffraction pattern substantially the same as shown in FIG. 1;

c) a powder x-ray diffraction pattern comprising 2 or more 2θ peaks selected from 6.4±0.2, 6.8±0.2, 9.6±0.2, 13.6±0.2, 15.7±0.2, 18.2±0.2 , 19.9±0.2, 21.6±0.2, 24.8±0.2 and 26.8±0.2 (obtained at room temperature and CuKα λ=1.5418 Å);

d) a powder x-ray diffraction pattern comprising 3 or more 2θ peaks selected from 6.4±0.2, 6.8±0.2, 9.6±0.2, 13.6±0.2, 14.1±0.2, 14.5±0.2, 14.7±0.2, 15.7±0.2, 18.2±0.2 , 18.7±0.2, 19.2±0.2, 19.9±0.2, 20.5±0.2, 21.6±0.2, 22.5±0.2, 23.1±0.2, 24.1±0.2, 24.8±0.2, 25.6±0.2, 26.8±0.2, 27.1±0.2 and 27.8±0.2 (obtained at room temperature and CuKα λ=1.5418 Å);

e) a differential scanning calorimetry thermogram substantially similar to the one as shown in FIG. 2;

f) a differential scanning calorimetry thermogram with an endotherm having an onset at about 152 ºC; and/or

g) a thermal gravimetric analysis thermogram substantially similar to the one as shown in FIG. 3.

76. The use of any one of claims 62 to 75, wherein the subject experiences a smaller decline in forced vital capacity (FVC) after a treatment period compared to an untreated subject.

77. The use of any one of claims 62 to 76, wherein the subject experiences a greater time to first disease progression event after a treatment period than an untreated subject, wherein the first disease progression event is selected from:

absolute predicted forced vital capacity (ppFVC) decline of ≥ 10% from baseline;

acute exacerbation of lung fibrosis;

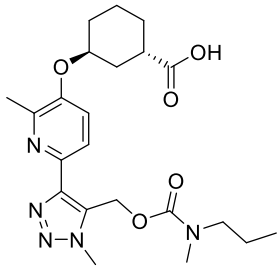
lung fibrosis-related hospitalization; and

all-cause mortality.

78. The use of any one of claims 62 to 77, wherein the subject experiences a smaller increase in cough domain score as measured by the Living with Pulmonary Fibrosis (L-PF) questionnaire over a treatment period than an untreated subject.

79. The use of any one of claims 62 to 78, wherein the subject experiences a smaller increase in dyspnea score as measured by the Living with Pulmonary Fibrosis (L-PF) questionnaire over a treatment period than an untreated subject.

80. Compound A:



Compound A;

or an equivalent amount of a pharmaceutically acceptable salt thereof, for use in treating interstitial lung disease, wherein about 240 mg/day Compound A is to be administered.

81. The compound for use of claim 80, wherein the interstitial lung disease is idiopathic pulmonary fibrosis (IPF).

82. The compound for use of claim 80, wherein the interstitial lung disease is progressive pulmonary fibrosis (PPF).