What is Claimed is:

1. Use of pirfenidone in preparation of a medicament for treating pulmonary fibrosis, optionally idiopathic pulmonary fibrosis (IPF), in a patient that exhibits (i) percent of predicted forced vital capacity volume (%FVC) ranging from about 50% to about 90%, and (ii) ratio of forced expiratory volume in one second (FEV1) to forced vital capacity volume (FVC) of about 0.80 or greater.

2. Use of a therapeutically effective amount of an agent in the manufacture of a medicament for treating pulmonary fibrosis, optionally idiopathic pulmonary fibrosis (IPF), in a patient that exhibits (i) percent of predicted forced vital capacity volume (%FVC) ranging from about 50% to about 90%, and (ii) ratio of forced expiratory volume in one second (FEV1) to forced vital capacity volume (FVC) of about 0.80 or greater, wherein the agent is a steroid, a cytotoxic agent, bardoxolone, a LPA agonist; Torisel (temsirolimus); a PI3K inhibitor; pentraxin or serum amyloid P; a MEK inhibitor; a p38 inhibitor; a PAI-1 inhibitor; an agent that reduces the activity of transforming growth factor-beta (TGF-β); lerdelimumab; metelimumab; an antibody that targets one or more TGF-β isoforms, an inhibitor of TGF-β receptor kinases TGFBR1 and TGFBR2; a modulator of post-receptor signaling pathways; a modulator of chemokine receptor signaling; an endothelin receptor antagonist; an agent that reduces the activity of connective tissue growth factor (CTGF); a matrix metalloproteinase (MMP) inhibitor; an agent that reduces the activity of epidermal growth factor receptor (EGFR); an antibody that targets EGF receptor; an inhibitor of EGF receptor kinase, a modulator of post-receptor signaling pathways; an agent that reduces the activity of platelet derived growth factor (PDGF); a PDGF neutralizing antibody; an antibody targeting PDGF receptor (PDGFR); an inhibitor of PDGFR kinase activity; an inhibitor of post-receptor signaling pathways; an agent that reduces the activity of vascular endothelial growth factor (VEGF); a VEGF-neutralizing antibody; an antibody targeting the VEGF receptor 1 (VEGFR1, Flt-1) and VEGF receptor 2 (VEGFR2, KDR); an inhibitor of VEGF receptor kinase activity; an inhibitor of multiple receptor kinases; an agent that interferes with integrin function; an integrin targeted antibody; an agent that interferes with the pro-fibrotic activity of IL-4, an agent that interferes with the pro-fibrotic activity of IL-13; a neutralizing antibody to IL-4 or IL-13, an antibody that targets IL-4 receptor or IL-13 receptor, or both; a chimeric protein including all or part of IL-13 and a toxin particularly pseudomonas endotoxin; an agent that interferes with epithelial mesenchymal transition; an inhibitor of mTor; an agent that reduces levels of copper; an agent that reduces oxidative stress; an agent that reduces interferon gamma; a PDE4 inhibitor; a PDE5 inhibitor; a modifier of the arachidonic acid pathway; a cyclooxygenase inhibitor; a 5-lipoxegenase inhibitor; an agent that reduces tissue remodeling or fibrosis; a prolyl hydrolase inhibitor; a peroxisome proliferator-activated receptor (PPAR)-gamma agonist, or a combination thereof.

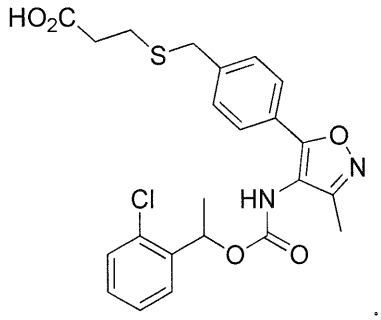
3. The use of claim 2, where in the agent is prednisolone, azathioprine, cyclophosphamide, bardoxolone, AM152, temsirolimus, PTX-2, PRM-151; ARRY-162; ARRY-300; Tiplaxtinin; GC-1008; lerdelimumab; CAT-152; Trabio; metelimumab; CAT-192; LY-

2157299; ACU-HTR-028 ; ALK5; ambrisentan; avosentan; bosentan; clazosentan; darusentan; BQ-153; FR-139317, L-744453; macitentan; PD-145065; PD-156252; PD163610;PS-433540; S-0139; sitaxentan sodium; TBC-3711; zibotentan; FG-3019; MMPI-12, PUP-1; tigapotide triflutate; erlotinib, gefitinib, BMS-690514, cetuximab; Imatinib mesylate; axitinib, bevacizumab, BIBF-1120, CDP-791, CT-322, IMC-18F1, PTC-299, ramucirumab; STX-100; IMGN-388; AER-001, AMG-317, APG-201, sIL-4Rα; anrukinzumab, CAT-354, cintredekin besudotox, MK-6105, QAX-576, SB-313, SL-102; AP-23573; rapamycin; tetrathiomolybdate; N-acetyl cysteine; Roflumilast; mirodenafil, PF-4480682, sildenafil citrate, SLx-2101, tadalafil, udenafil, UK-369003, vardenafil, zaprinast; Zileuton, 1016548, CG-0089, FG-2216, FG-4497, FG-5615, FG-6513, fibrostatin A, lufironil,P-1894B, safironil; pioglitazone; rosiglitazone, or a combination thereof.

4. Use of a therapeutically effective amount of an agent in the manufacture of a medicament for treating pulmonary fibrosis, optionally idiopathic pulmonary fibrosis (IPF), in a patient that exhibits (i) percent of predicted forced vital capacity volume (%FVC) ranging from about 50% to about 90%, and (ii) ratio of forced expiratory volume in one second (FEV1) to forced vital capacity volume (FVC) of about 0.80 or greater, wherein the agent is BG-12, a chemokine activity modulator, Lys1 oxidase inhibitor, a NOX4 inhibitor; an angiotensis II receptor antagonist; an inhibitor or Wnt-beta catenin signaling agent; a JNK inhibitor; a IL-4/IL-13 antibody/soluble receptor; an LPA1/LPA3 antagonist; a PG D2 antagonist, a pirfenidone analog, a deuterated pirfenidone, nitric oxide, a vitamin E and pentoxifylline combination, PXS25, desatinib, a PI3K/mTor dual inhibitor, a PI3K inhibitor; a 5-HT2A/B receptor antagonist; a telomerase activator; a modulator of chemokine activity; an anti αvβ6 integrin agent, pentraxin, or a combination thereof.

5. Use of a therapeutically effective amount of an agent in the manufacture of a medicament for treating pulmonary fibrosis, optionally idiopathic pulmonary fibrosis (IPF), in a patient that exhibits (i) percent of predicted forced vital capacity volume (%FVC) ranging from about 50% to about 90%, and (ii) ratio of forced expiratory volume in one second (FEV1) to forced vital capacity volume (FVC) of about 0.80 or greater, wherein the agent is an LPA1 receptor antagonist.

6. The use of claim 5, wherein the LPA1 receptor antagonist is



7. The use of any of claims 1-6, wherein the patient has been diagnosed with pulmonary fibrosis, optionally IPF, for at least six months, and optionally less than 48 months.

8. The use of any of claims 1-7, wherein the patient exhibits a diffusion capacity (%DLco) ranging from about 30% to about 90%.

9. The use of any one of claims 18, wherein the pirfenidone is to be administered at a total daily dosage of at least about 1800 mg.

10. The use of claim 9, wherein the pirfenidone is to be administered at a total daily dosage of about 2403 mg.

11. The use of any one of claims 1-10 wherein the pirfenidone is to be administered to the patient three times per day, with food.

12. The use of any one of claims 1-11, wherein the pirfenidone comprises a deuterated pirfenidone.

13. The use of any one of claims 2-4 and 7-8, further comprising administration of pirfenidone.

14. The use of claim 13, wherein the pirfenidone comprises a deuterated pirfenidone.

15. Use of an anti-IL-13 antibody in preparation of a medicament for the treatment of pulmonary fibrosis, optionally idiopathic pulmonary fibrosis (IPF), wherein said medicament is to be administered with pirfenidone.

16. Use of pirfenidone in preparation of a medicament for the treatment of pulmonary fibrosis, optionally idiopathic pulmonary fibrosis (IPF), wherein said medicament is to be administered with an anti-IL13 antibody.

17. The use of claim 15 or claim 16, wherein the medicament is for administration to a patient that exhibits (i) percent of predicted forced vital capacity volume (%FVC) ranging from about 50% to about 90%.

18. The use of any one of claims 15-17, wherein the medicament is for administration to a patient that further exhibits a ratio of forced expiratory volume in one second (FEV1) to forced vital capacity volume (FVC) of about 0.80 or greater.

19. The use of any one of claims 15-18, wherein the anti-IL-13 antibody is TNX-650.

20. Use of pirfenidone and an anti-IL-13 antibody in the manufacture of a medicament for the treatment of pulmonary fibrosis, optionally idiopathic pulmonary fibrosis (IPF).

21. The use of claim 20, wherein the medicament is for a patient that exhibits (i) percent of predicted forced vital capacity volume (%FVC) ranging from about 50% to about 90%.

22. The use of any claim 20 or claim 21, wherein the medicament is for administration to a patient that further exhibits a ratio of forced expiratory volume in one second (FEV1) to forced vital capacity volume (FVC) of about 0.80 or greater.

23. The use of any one of claims 20-22, wherein the anti-IL-13 antibody is TNX-650.

What is Claimed is:

1. Pirfenidone for use in treating pulmonary fibrosis, optionally IPF, in a patient that exhibits (i) percent of predicted forced vital capacity volume (%FVC) of about 90% or less or (ii) ratio of forced expiratory volume in one second (FEV1) to forced vital capacity volume (FVC) of about 0.80 or greater, or both.

2. Use of pirfenidone in preparation of a medicament for treating pulmonary fibrosis, optionally IPF, in a patient that exhibits (i) percent of predicted forced vital capacity volume (%FVC) of about 90% or less or (ii) ratio of forced expiratory volume in one second (FEV1) to forced vital capacity volume (FVC) of about 0.80 or greater, or both.

3. Use of a therapeutically effective amount of an agent in the manufacture of a medicament for treating pulmonary fibrosis, optionally IPF, in a patient that exhibits (i) percent of predicted forced vital capacity volume (%FVC) of about 90% or less, or (ii) ratio of forced expiratory volume in one second (FEV1) to forced vital capacity volume (FVC) of about 0.80 or greater, or both,
wherein the agent is a steroid, a cytotoxic agent, bardoxolone, a LPA agonist; Torisel (temsirolimus); a PI3K inhibitor; pentraxin or serum amyloid P; a MEK inhibitor; a p38 inhibitor; a PAI-1 inhibitor; an agent that reduces the activity of transforming growth factor-beta (TGF-β); lerdelimumab; metelimumab; an antibody that targets one or more TGF-β isoforms, an inhibitor of TGF-β receptor kinases TGFBR1 and TGFBR2; a modulator of post-receptor signaling pathways; a modulator of chemokine receptor signaling; an endothelin receptor antagonist; an agent that reduces the activity of connective tissue growth factor (CTGF); a matrix metalloproteinase (MMP) inhibitor; an agent that reduces the activity of epidermal growth factor receptor (EGFR); an antibody that targets EGF receptor; an inhibitor of EGF receptor kinase, a modulator of post-receptor signaling pathways; an agent that reduces the activity of platelet derived growth factor (PDGF); a PDGF neutralizing antibody; an antibody targeting PDGF receptor (PDGFR); an inhibitor of PDGFR kinase activity; an inhibitor of post-receptor signaling pathways; an agent that reduces the activity of

vascular endothelial growth factor (VEGF); a VEGF-neutralizing antibody; an antibody targeting the VEGF receptor 1 (VEGFR1, Flt-1) and VEGF receptor 2 (VEGFR2, KDR); an inhibitor of VEGF receptor kinase activity; an inhibitor of multiple receptor kinases; an agent that interferes with integrin function; an integrin targeted antibody; an agent that interferes with the pro-fibrotic activity of IL-4, an agent that interferes with the pro-fibrotic activity of IL-13; a neutralizing antibody to IL-4 or IL-13, an antibody that targets IL-4 receptor or IL-13 receptor, or both; a chimeric protein including all or part of IL-13 and a toxin particularly pseudomonas endotoxin; an agent that interferes with epithelial mesenchymal transition; an inhibitor of mTor; an agent that reduces levels of copper; an agent that reduces oxidative stress; an agent that reduces interferon gamma; a PDE4 inhibitor; a PDE5 inhibitor; a modifier of the arachidonic acid pathway; a cyclooxygenase inhibitor; a 5-lipoxegenase inhibitor; an agent that reduces tissue remodeling or fibrosis; a prolyl hydrolase inhibitor; a peroxisome proliferator-activated receptor (PPAR)-gamma agonist, or a combination thereof.

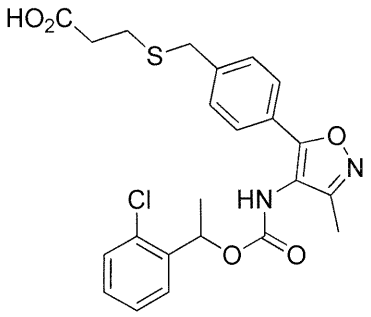
4. The use of claim 3, where in the agent is prednisolone, azathioprine, cyclophosphamide, bardoxolone, AM152, temsirolimus, PTX-2, PRM-151; ARRY-162; ARRY-300; Tiplaxtinin; GC-1008; lerdelimumab; CAT-152; Trabio; metelimumab; CAT-192; LY-2157299; ACU-HTR-028 ; ALK5; ambrisentan; avosentan; bosentan; clazosentan; darusentan; BQ-153; FR-139317, L-744453; macitentan; PD-145065; PD-156252; PD163610;PS-433540; S-0139; sitaxentan sodium; TBC-3711; zibotentan; FG-3019; MMPI-12, PUP-1; tigapotide triflutate; erlotinib, gefitinib, BMS-690514, cetuximab; Imatinib mesylate; axitinib, bevacizumab, BIBF-1120, CDP-791, CT-322, IMC-18F1, PTC-299, ramucirumab; BIBF-1120; STX-100; IMGN-388; AER-001, AMG-317, APG-201, sIL-4Rα; anrukinzumab, CAT-354, cintredekin besudotox, MK-6105, QAX-576, SB-313, SL-102; AP-23573; rapamycin; tetrathiomolybdate; N-acetyl cysteine; Roflumilast; mirodenafil, PF-4480682, sildenafil citrate, SLx-2101, tadalafil, udenafil, UK-369003, vardenafil, zaprinast; Zileuton, 1016548, CG-0089, FG-2216, FG-4497, FG-5615, FG-6513, fibrostatin A, lufironil,P-1894B, safironil; pioglitazone; rosiglitazone, or a combination thereof.

5. Use of a therapeutically effective amount of an agent in the manufacture of a medicament for treating pulmonary fibrosis, optionally IPF, in a patient that exhibits (i) percent of predicted forced vital capacity volume (%FVC) of about 90% or less, or (ii) ratio of forced expiratory volume in one second (FEV1) to forced vital capacity volume (FVC) of about 0.80 or greater, or both,
wherein the agent is BG-12, a chemokine activity modulator, Lys1 oxidase inhibitor, a NOX4

inhibitor; an angiotensis II receptor antagonist; an inhibitor or Wnt-beta catenin signaling agent; a JNK inhibitor; a IL-4/IL-13 antibody/soluble receptor; an LPA1/LPA3 antagonist; a PG D2 antagonist, a pirfenidone analog, a deuterated pirfenidone, nitric oxide, a vitamin E and pentoxifylline combination, PXS25, desatinib, a PI3K/mTor dual inhibitor, a PI3K inhibitor; a 5-HT2A/B receptor antagonist; a telomerase activator; a modulator of chemokine activity; an anti αvβ6 integrin agent, pentraxin, or a combination thereof.

6. Use of a therapeutically effective amount of an agent in the manufacture of a medicament for treating pulmonary fibrosis, optionally IPF, in a patient that exhibits (i) percent of predicted forced vital capacity volume (%FVC) of about 90% or less, or (ii) ratio of forced expiratory volume in one second (FEV1) to forced vital capacity volume (FVC) of about 0.80 or greater, or both,
wherein the agent is an LPA1 receptor antagonist.

7. The use of claim 6, wherein the LPA1 receptor antagonist is



8. The pirfenidone or use of any of claims 1-7, wherein %FVC ranges from about 50% to about 90%.

9. The pirfenidone or use of any of claims 1-8, wherein the patient has been diagnosed with pulmonary fibrosis, optionally IPF, for at least six months, and optionally less than 48 months.

10. The pirfenidone or use of any of claims 1-9, wherein the patient exhibits a diffusion capacity (%DLco) ranging from about 30% to about 90%.

11. The pirfenidone or use of any one of claims 1-2 and 8-10, wherein the pirfenidone is to be administered at a total daily dosage of at least about 1800 mg.

12. The pirfenidone or use of claim 11, wherein the pirfenidone is to be administered at a total daily dosage of about 2403 mg.

13. The pirfenidone or use of any one of claims 1-2 and 8-12 wherein the pirfenidone is to be administered to the patient three times per day, with food.

14. The pirfenidone or use of any one of claims 1-2 and 8-13, wherein the pirfenidone comprises a deuterated pirfenidone.

15. The use of any one of claims 3-5 and 8-10, further comprising administration of pirfenidone.

16. The use of claim 15, wherein the pirfenidone comprises a deuterated pirfenidone.