

WE CLAIM:

1. Use of an ActRII polypeptide in the manufacture of a medicament for treating pulmonary arterial hypertension (PAH) in a patient, wherein the polypeptide comprises an amino acid sequence that is at least 90% identical to an amino acid sequence that begins at amino acid 21 of SEQ ID NO: 1 and ends at amino acid 135 of SEQ ID NO: 1, and wherein the medicament is to be administered to the patient at a dosing regimen comprising a first dose of between 0.1 mg/kg and 1.0 mg/kg of said polypeptide for a first period of time, and a second dose of between 0.1 mg/kg and 1.0 mg/kg of said polypeptide is to be subsequently administered for a second period of time.
2. The use of claim 1, wherein administration of the medicament results in a change in one or more of the following hemodynamic or functional parameters:
 - a. increase in exercise capacity;
 - b. increase in 6-minute walk distance (6MWD);
 - c. provide clinical improvement;
 - d. improve World Health Organization (WHO) Functional Class; and
 - e. delay disease progression, including reducing the risk of death and hospitalization for PAH.
3. The use of claim 1 or claim 2, wherein the ActRII polypeptide comprises an amino acid sequence that is at least 90% identical to the amino acid sequence of SEQ ID NO: 2.
4. The use of any one of claims 1-3, wherein the ActRII polypeptide comprises an amino acid sequence that is at least 95% identical to the amino acid sequence of SEQ ID NO: 2.
5. The use of any one of claims 1-4, wherein the ActRII polypeptide comprises an amino acid sequence that is at least 97% identical to the amino acid sequence of SEQ ID NO: 2.
6. The use of any one of claims 1-5, wherein the ActRII polypeptide comprises an amino acid sequence that is at least 99% identical to the amino acid sequence of SEQ ID NO: 2.

7. The use of any one of claims 1-6, wherein the ActRII polypeptide comprises an amino acid sequence that is identical to the amino acid sequence of SEQ ID NO: 2.
8. The use of any one of claims 1-7, wherein the ActRII polypeptide is a fusion protein further comprising an Fc domain of an immunoglobulin.
9. The use of any one of claims 1-8, wherein the Fc fusion protein further comprises a linker domain positioned between the ActRII polypeptide domain and the Fc domain of the immunoglobulin.
10. The use of claim 9, wherein the linker domain is selected from the group consisting of: TGGG (SEQ ID NO: 20), TGGGG (SEQ ID NO: 18), SGGGG (SEQ ID NO: 22), GGGGS (SEQ ID NO: 22), GGG (SEQ ID NO: 16), GGGG (SEQ ID NO: 17), and SGGG (SEQ ID NO: 21).
11. The use of claim 10, wherein the linker domain is TGGG (SEQ ID NO: 20).
12. The use of any one of claims 1-11, wherein the polypeptide is to be administered to the patient every 3 weeks.
13. The use of any one of claims 1-11, wherein the polypeptide is to be administered to the patient every 4 weeks.
14. The use of any one of claims 1-13, wherein the ActRII polypeptide binds to one or more ligands selected from the group consisting of: activin A, activin B, BMP6, BMP10, GDF8, and GDF11.
15. The use of any one of claims 1-14, wherein the polypeptide is to be administered at a dose between 0.1 mg/kg and 1.0 mg/kg.
16. The use of any one of claims 1-15, wherein an additional active agent or supportive therapy is to be further administered to the patient, the additional active agent or supportive therapy is selected from the group consisting of: a beta-blocker, an angiotensin-converting enzyme inhibitor (ACE inhibitor), an angiotensin receptor blocker (ARB), a diuretic agent, a

lipid-lowering medication, an endothelin blocker, a PDE5 inhibitor, a prostacyclin, or a left ventricular assist device (LVAD).

17. The use of any one of claims 1-16, wherein one or more agents has been administered to the patient, the one or more agents is selected from the group consisting of: a phosphodiesterase type 5 inhibitor, a soluble guanylate cyclase stimulator, a prostacyclin receptor agonist, and an endothelin receptor antagonist.

18. The use of any one of claims 1-17, wherein the medicament is to be administered to the patient on a schedule selected from the group consisting of: every week, every two weeks, every three weeks, and every four weeks.

19. Use of an ActRII polypeptide in the manufacture of a medicament for treating or preventing cardiopulmonary remodeling associated with pulmonary arterial hypertension in a patient, wherein an effective amount of the medicament is to be administered to a patient in need thereof, wherein said medicament slows down cardiac remodeling and/or reverses cardiac remodeling.

20. The use of claim 19, wherein the reversal in cardiac remodeling is a sustained reversal.

21. The use of claim 19, wherein the cardiopulmonary remodeling is ventricle remodeling or ventricular dilation.

22. The use of any one of claims 1-18, wherein the first period of time is at least 3 weeks.

23. The use of any one of claims 1-18, wherein the second period of time is at least 3 weeks.

24. The use of any one of claims 1-18, wherein the second period of time exceeds the first period of time.

25. The use of any one of claims 1-18, wherein the second dose exceeds the first dose.

26. The use of any one of claims 1-18, wherein the first dose is in the range of about 0.2 mg/kg to about 0.4 mg/kg followed by a second dose in the range of about 0.5 mg/kg to about 0.8 mg/kg.

27. A kit comprising a lyophilized polypeptide and an injection device, wherein the polypeptide is an ActRII polypeptide comprising an amino acid sequence that is at least 90% identical to an amino acid sequence that begins at amino acid 21 of SEQ ID NO: 1 and ends at amino acid 135 of SEQ ID NO: 1.
28. The use of any one of claims 1-18, wherein the first dose is about 0.3 mg/kg followed by a second dose of about 0.7 mg/kg.
29. Use of an ActRII fusion protein in the manufacture of a medicament for treating pulmonary arterial hypertension (PAH) in a patient, wherein the fusion protein comprises an amino acid sequence of SEQ ID NO: 23, and wherein the medicament is to be administered to the patient at a dosing regimen comprising a first dose of between 0.1 mg/kg and 1.0 mg/kg of said fusion protein for a first period of time, and a second dose of between 0.1 mg/kg and 1.0 mg/kg of said fusion protein is to be subsequently administered for a second period of time.
30. Use of an ActRII fusion protein in the manufacture of a medicament for treating pulmonary arterial hypertension (PAH) in a patient, wherein the fusion protein comprises an amino acid sequence of SEQ ID NO: 23, and wherein the medicament is to be administered to the patient at a dosing regimen comprising a first dose of 0.3 mg/kg of said fusion protein for a first period of time, and a second dose of 0.7 mg/kg of said fusion protein is to be subsequently administered for a second period of time.