

CLAIMS

- ~~1. A method of treating or preventing mucocutaneous, musculoskeletal and/or renal disease in an systemic lupus erythematosus (SLE) patient in need thereof, the method comprising administering a therapeutically effective amount of a type I IFN receptor (IFNAR1) inhibitor to the patient, wherein the method treats mucocutaneous, musculoskeletal and/or renal disease in the patient.~~
- ~~2. The method of claim 1, wherein the method treats mucocutaneous, musculoskeletal and renal disease in the patient.~~
- ~~3. The method of claim 1 or 2, wherein the method reduces the mucocutaneous, musculoskeletal and/or renal flare rate in the patient relative to pre-treatment mucocutaneous, musculoskeletal and/or renal flare rate respectively.~~
- ~~4. The method of any preceding claim, wherein the method improves the patient's BILAG-2004 mucocutaneous, renal and/or musculoskeletal organ domain score.~~
- ~~5. The method of any preceding claim, wherein the method improves the patient's SLEDAI-2K mucocutaneous and/or musculoskeletal organ domain score.~~
- ~~6. The method of any preceding claim, wherein the method treats cardiorespiratory disease in the patient, optionally wherein the method improves the patient's BILAG-2004 cardiorespiratory organ domain score.~~
- ~~7. The method of any preceding claim, wherein the method treats constitutional disease in the patient, optionally wherein the method improves the patient's BILAG-2004 constitutional organ domain score.~~
- ~~8. The method of any preceding claim, wherein the method treats vascular, hematologic, renal and/or cardiorespiratory disease in the patient, optionally wherein the method improves the patient's SLEDAI-2K vascular, hematologic, renal and/or cardiorespiratory disease organ domain score.~~
- ~~9. The method of any preceding claim, wherein the method treats rash in the patient.~~
- ~~10. The method of claim 9, wherein there is a $\geq 50\%$ improvement in rash in the subject from pre-treatment levels of rash, optionally wherein the improvement is defined by Cutaneous Lupus Erythematosus Disease Area and Severity Index (CLASI).~~
- ~~11. The method of claim 10, wherein the method resolves rash in the patient, optionally wherein the method completely resolves SLEDAI-2K-defined rash in the patient.~~
- ~~12. The method of any preceding claim, wherein the method treats or prevent arthritis in the patient.~~
- ~~13. The method of claim 12, wherein the method completely resolves arthritis in the patient, optionally wherein the method complete resolves SLEDAI-2K-defined arthritis in the patient.~~

14. ~~The method of any preceding claim, wherein the method leads to a $\geq 50\%$ improvement in swollen and tender joint count in the patient compared to the pre-treatment swollen and tender joint count in the patient, optionally wherein the patient had ≥ 6 swollen and tender joint count pre-treatment.~~
15. ~~The method any preceding claim, wherein the method comprises a method of treating or preventing renal disease in the patient, wherein the method treats or prevents renal disease in the patient.~~
16. ~~The method of claim 15, wherein the patient has a 24-hour UPCR > 0.5 mg/mg pre-treatment, and wherein the method improves the subject's 24-hour UPCR to ≤ 0.5 mg/mg.~~
17. ~~A method of treating SLE in a patient thereof, the method comprising administering a therapeutically effective amount of a type I IFN receptor (IFNAR1) inhibitor to the patient, wherein the patient has a baseline CLASI-A ≥ 10 , wherein treatment reduces the patient's CLASI-A $\geq 50\%$.~~
18. ~~The method of claim 17, wherein the treatment reduces the patient's CLASI-A by at least week 12 of treatment.~~
19. ~~The method of claim 17 or 18, wherein the reduction in the patient's CLASI-A is maintained for at least 4, 8, 12, 16, 20, 24, 28, 32, 36 or 40 weeks.~~
20. ~~A method of treating a systemic lupus erythematosus (SLE) patient in need thereof, the method comprising administering a therapeutically effective amount of a type I IFN receptor (IFNAR1) inhibitor to the patient, wherein the subject has low complement at baseline compared to a healthy subject, wherein the method reduces SLE disease activity in the patient.~~
21. ~~The method of claim 20, where the patient has low C3 and/or C4 complement at baseline compared to a healthy subject.~~
- 22.1. Use A method of a type I IFN receptor (IFNAR1) inhibitor in the preparation of a medicament for the treatment of treatment-refractory ~~treating a systemic lupus erythematosus (SLE) in a patient in need thereof, the method comprising administering a therapeutically effective amount of a type I IFN receptor (IFNAR1) inhibitor to the patient, wherein the patient has treatment-refractory SLE, and wherein the method reduces SLE disease activity in the patient~~ IFNAR1 inhibitor is anifrolumab.
- 23.2. The use method ~~of claim 122~~, wherein the patient has previously received prior treatment with glucocorticoids, antimalarials and/or immunosuppressants.
- 24.3. The use method ~~of claim 122 or 223~~, wherein pre-treatment the patient has a SLEDAI-2K score of ≥ 6 , ≥ 1 A and/or a ≥ 2 B BILAG-2004 organ domain score, and/or a Physician's Global Assessment of ≥ 1 .

- ~~25.4. The use method of any of preceding claims 22 to 24, wherein the patient has received prior treatment with azathioprine, mizoribine, mycophenolate mofetil, mycophenolic acid, and/or methotrexate.~~
- ~~26. The method of any of claims 20 to 25, wherein reducing SLE disease activity in the patient comprises a BILAG Based Composite Lupus Assessment (BICLA) response.~~
- ~~27.5. The use method of any preceding claim, wherein the patient has moderate to severe SLE.~~
- ~~28. The method of any preceding claim, wherein the method has been demonstrated in a phase III clinical trial.~~
- ~~29. The method of any preceding claim, wherein the type I IFN receptor inhibitor is anifrolumab or a functional variant thereof.~~
- ~~30.6. The use method of any preceding claim 29, wherein the medicament method comprises administering a fixed dose of anifrolumab.~~
- ~~31.7. The use method of any preceding claim 30, wherein the medicament method comprises administering about 300 mg to about 1000 mg of anifrolumab.~~
- ~~32.8. The use method of any preceding claim 31, wherein the medicament comprises comprising administering about 300 mg anifrolumab.~~
- ~~33. The method of claim 31, comprising administering anifrolumab or the functional variant thereof at a dose of 300-1000 mg every four weeks (Q4W),~~
- ~~34. The method of claim 33, wherein anifrolumab or the functional variant thereof is administered intravenously.~~
- ~~35.9. The use method of any of claims 31 to 6, wherein the medicament comprises comprising administering anifrolumab or the functional variant thereof to the patient at a dose of 120 mg anifrolumab every week, optionally wherein anifrolumab or the functional variant thereof is administered subcutaneously.~~
10. The use of any of preceding claim, wherein the medicament is for subcutaneous administration to the patient.
11. The use of any of preceding claim, wherein the medicament is for subcutaneous administration to the patient once a week.
12. The use of any of preceding claim, wherein the medicament is for subcutaneous administration of 120 mg anifrolumab to the patient once a week.
13. The use of any of claims 1 to 8, wherein the medicament is for intravenous administration.

14. The use of any of claims 1 to 8, wherein the medicament is for intravenous administration to the patient once every four weeks (Q4W).
15. The use of any of claims 1 to 8, wherein the medicament is for intravenous administration of 300 mg anifrolumab to the patient Q4W.
- ~~36. The method of any of the preceding claims, the method comprising steroid sparing in the subject, wherein the dose of the steroid administered to the subject is tapered from a pre-sparing dose at baseline to a post-sparing dose.~~
- ~~37. The method of claim 36, wherein the post-sparing dose is ≤ 7.5 mg/day prednisone or prednisone equivalent dose.~~
- ~~38. The method of any of claim 36 or 37, wherein the pre-sparing dose is 10 mg/day or prednisone equivalent dose.~~
- ~~39. The method of any of claims 36-38, wherein the steroid comprises a glucocorticoid.~~
- ~~40. The method of claim 39, wherein the steroid comprises an oral glucocorticoid.~~
- ~~41. The method of any of claims 36-40, wherein the steroid is hydrocortisone, mometasone, fluticasone, flucinolone acetonide, flucinolone, flurandrenolone acetonide, ciclesonide, budesonide, beclomethasone, deflazacort, flunisolide, beclomethasone dipropionate, betamethasone, betamethasone valerate, methylprednisolone, dexamethasone, prednisolone, cortisol, triamcinolone, clobetasol, clobetasol propionate, clobetasol butyrate, cortisone, corticosterone, clocortolone, dihydroxycortisone, alclometasone, amcinonide, difluocortolone valerate, flucortolone, fluprednidene, fluandrenolone, fluorometholone, halcinonide, halobetasol, desonide, diflorasone, flurandrenolide, fluocinonide, prednicarbate, desoximetasone, fluprednisolone, prednisone, azelastine, dexamethasone 21-phosphate, fludrocortisone, flumethasone, fluocinonide, halopredone, hydrocortisone 17-valerate, hydrocortisone 17-butyrate, hydrocortisone 21-acetate, prednisolone, prednisolone 21-phosphate, clobetasol propionate, triamcinolone acetonide, or a mixture thereof.~~
- ~~42. The method of any of claims 36-40, wherein the steroid comprises prednisone.~~
- ~~43. The method of any preceding claim, wherein the patient is a type I interferon stimulated gene signature (IFNGS)-test high patient pre-treatment.~~
- ~~44. The method of any preceding claim, comprising identifying the subject as IFNGS-test high patient before administration of the IFNAR1-inhibitor.~~
- ~~45. A pharmaceutical composition for use in any of the methods of claims 1-44.~~
- ~~46. An injection device comprising the pharmaceutical composition of claim 45.~~
- ~~47. The injection device of claim 46, wherein the injection device is a pre-filled syringe (PFS).~~

- ~~48. The injection device of claim 47, wherein the injection device is an accessorized pre-filled syringe (AFPS).~~
- ~~49. The injection device of claim 47, wherein the injection device is an auto-injector.~~
- ~~50. A kit comprising the injection device of any of claims 46-49, and instructions for use.~~
- ~~51. The kit of claim 50, wherein the instructions for use comprise instructions for subcutaneous administration of the pharmaceutical composition or unit dose to the patient.~~
- ~~52. The kit of claim 50 or 51, wherein the instructions for use specify that the injection device, unit dose and/or pharmaceutical composition are for use in the treatment of SLE.~~
- ~~53. The kit of any of claims 50-52, comprising packaging, wherein the packaging is adapted to hold the injection device and the instructions for use.~~
- ~~54. The kit of any of claims 50-53, wherein the instructions for use are attached to the injection device.~~