

CLAIMS

1. Use of iptacopan or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for treating lupus nephritis (LN) in a subject in need thereof, wherein the medicament is to be orally administered to the subject at a dose of from about 50 mg to about 200 mg, e.g., from about 50 mg to from about 100 mg, from about 100 mg to about 200 mg, at a dose of about 50 mg, about 75 mg, about 100 mg, about 150 mg, or about 200 mg, each to be administered twice daily (b.i.d.), to thereby treat the subject.
2. The use of claim 1, wherein the dose is about 50 mg twice daily.
3. The use of claim 1, wherein the dose is about 100 mg twice daily.
4. The use of claim 1, wherein the dose is about 200 mg twice daily.
5. The use of any of claims 1 to 4, wherein the use further comprises an immunosuppressant to be administered to the subject.
6. The use of claim 5, wherein the immunosuppressant is a mycophenolic acid (e.g., mycophenolate mofetil (MMF), mycophenolate sodium (MPS)), a cyclophosphamide (CYC), an anti-B cell agent, or a calcineurin inhibitor (CNI).
7. The use of any of claims 1 to 6, wherein the use further comprises a corticosteroid to be administered to the subject.
8. The use of claim 7, wherein the corticosteroid is to be administered to the subject according to a tapering regimen.
9. The use of any of claims 1 to 8, wherein the subject achieves a daily corticosteroid dose of <10 mg/day following a corticosteroid tapering regimen.
10. The use of any of claims 1 to 8, wherein the subject achieves a daily corticosteroid dose of 0 mg/day following a corticosteroid tapering regimen.
11. The use of any of claims 1 to 6, wherein the immunosuppressant is to be administered without a corticosteroid.
12. The use of any of claims 1 to 11, wherein the subject has been vaccinated against *Neisseria meningitidis* and *Streptococcus pneumoniae* prior to iptacopan or a pharmaceutically acceptable salt thereof is to be administered.
13. The use of any of claims 1 to 12, wherein the subject has been vaccinated against *Haemophilus influenzae* prior to iptacopan or a pharmaceutically acceptable salt thereof is to be administered.
14. The use of any of claims 1 to 13, wherein the subject has been further treated with supportive care, wherein the supportive care is selected from the group consisting

of an antimalarial (e.g. hydroxychloroquine), an angiotensin-converting enzyme inhibitor and an angiotensin receptor blocker.

15. The use of any of claims 1 to 14, wherein treating lupus nephritis (LN) comprises achieving an UPCR < 0.5 g/g in a subject.
16. The use of any of claims 1 to 15, wherein treating lupus nephritis (LN) comprises achieving an UPCR in a subject that is reduced by 50% compared to prior to iptacopan or a pharmaceutically acceptable salt thereof is administered, *e.g.*, the UPCR value being measured by sampling from a first morning void or 24 hour urine collection.
17. The use of any of claims 1 to 16, wherein treating lupus nephritis (LN) comprises achieving an eGFR ≥ 90 ml/min/1.73 m².
18. The use of any of claims 1 to 17, wherein treating lupus nephritis (LN) comprises achieving an eGFR value of no less than 80% (*e.g.*, no less than 85%) compared to prior to iptacopan or a pharmaceutically acceptable salt thereof is administered.
19. Iptacopan or a pharmaceutically acceptable salt thereof for use in a treatment of lupus nephritis in a subject in need thereof, wherein the treatment comprises orally administering to the subject iptacopan or a pharmaceutically acceptable salt thereof, at a dose of from about 50 mg to about 200 mg, *e.g.*, from about 50 mg to from about 100 mg, from about 100 mg to about 200 mg, at a dose of about 50 mg, about 75 mg, about 100 mg, about 150 mg, or about 200 mg, each dose to be administered twice daily (*b.i.d.*), to thereby treat the subject.
20. A pharmaceutical composition comprising iptacopan or a pharmaceutically acceptable salt thereof, *e.g.*, iptacopan hydrochloride, for use in treating lupus nephritis, in a subject, *e.g.*, patient, in need thereof, wherein the pharmaceutical composition is to be administered orally at a dose of from about 50 mg to about 200 mg, *e.g.*, from about 50 mg to from about 100 mg, from about 100 mg to about 200 mg, about 50 mg, about 75 mg, about 100 mg, about 150 mg, or about 200 mg of iptacopan or a pharmaceutically acceptable salt thereof, *e.g.*, iptacopan hydrochloride, each dose to be administered twice daily (*b.i.d.*), *e.g.*, about every 12 hours, to thereby treat the subject, *e.g.*, patient.