Background and aims Preliminary data suggested efficacy of sirolimus in treatment of lupus nephritis (LN), but its long-term efficacy and tolerability data is lacking.

Methods We reviewed Class III/IV/V LN patients who received prednisolone and sirolimus either as initial or maintenance treatment during Jan 2007 to Jan 2016.

Results Sixteen patients were included (duration of sirolimus treatment: 27.2±19.6 months). Ten patients received sirolimus due to intolerance to standard immunosuppressive treatments and six patients because of a history of malignancy. Five patients received sirolimus during active LN, and showed improvement in proteinuria (2.8±1.9 g/day and 0.1±0.1 g/day at baseline and 36 months, p=0.011 compared to baseline), anti-dsDNA (107.7±91.9 IU/mL at baseline and 36 months, p=0.145) and C3 (54.8±26.1 mg/dL at baseline and 36 months, p=0.084). Eleven patients received sirolimus during disease quiescence, and showed significant improvement in C3 (90.4±18.1 mg/dL at baseline and 36 months, p=0.025) and stable renal function (58.5±25.2 ml/min and 56.7±29.0 ml/min at baseline and 36 months, p=0.263). One patient, whose serum creatinine was 244 µmol/L when sirolimus was started, developed renal failure after 27 months. Renal flare occurred in one patient after 36 months. Sirolimus was discontinued in five patients including one with leucopenia. Four patients showed lipid profile deterioration which was adequately controlled with statin.

Conclusions Sirolimus can be an alternative treatment option for LN and the long-term results do not suggest excessive adverse effects.