

Supplementary Table 1. Oxford Centre for Evidence-Based Medicine level of evidence and grading of recommendations

2011 Levels of Evidence		
LoE	Therapy/Prevention/Etiology/Harm	Risk factors/Prognosis
1a	Systematic reviews of RCT	Systematic review of inception cohort studies
1b	Individual, high-quality RCT	Individual inception cohort study (high quality)
2a	Systematic reviews of cohort studies	Systematic review of retrospective cohort studies or data from RCT
2b	Cohort study or low quality RCT	Retrospective cohort study or data from RCT
2c	"Outcomes" research studies	"Outcomes" research studies
3a	Systematic review of case-control studies	
3b	Case-control studies	
4	Case-series (and poor-quality cohort and case-control studies)	Case-series (and poor-quality prognostic cohort) studies)
5	Expert opinion	Expert opinion
Grades of recommendations, assessment, development and evaluations		
A	Consistent level 1 studies	
B	Consistent level 2 or 3 studies; or extrapolations from level 1 studies	
C	Level 4 studies; or extrapolations from level 2 or 3 studies	
D	Level 5 evidence; or very inconsistent or inconclusive studies of any level	

RCT: Randomized controlled trials

Supplementary Table 2. The 2003 International Society of Nephrology/Renal Pathology Society (ISN/RPS) histologic classification of LN, together with recent recommendations (2018)

Class I	<p>Minimal mesangial lupus nephritis</p> <p>Normal glomeruli by light microscopy, but mesangial immune deposits by immunofluorescence</p>
Class II	<p>Mesangial proliferative</p> <p>Purely mesangial hypercellularity of any degree or mesangial matrix expansion by light microscopy, with mesangial immune deposits</p> <p>A few isolated subepithelial or subendothelial deposits may be visible by immunofluorescence or electron microscopy, but not by light microscopy</p>
Class III	<p>Focal lupus nephritis</p> <p>Active or inactive focal, segmental, or global endocapillary or extracapillary glomerulonephritis involving <50% of all glomeruli, typically with focal subendothelial immune deposits, with or without mesangial alterations</p>
Class IV	<p>Diffuse lupus nephritis</p> <p>Active or inactive diffuse, segmental or global endocapillary or extracapillary glomerulonephritis involving $\geq 50\%$ of all glomeruli, typically with diffuse subendothelial immune deposits, with or without mesangial alterations. This class is divided to <i>diffuse segmental</i> (IV-S) lupus nephritis when $\geq 50\%$ of the involved glomeruli have segmental lesions and <i>diffuse global</i> (IV-G) when $\geq 50\%$ of the involved glomeruli have global lesions. Segmental is defined as a glomerular lesion that involves less than half of the glomerular tuft. This class includes cases with diffuse wire loop deposits but with little or no glomerular proliferation</p>
Class V	<p>Membranous lupus nephritis</p> <p>Global or segmental subepithelial immune deposits or their morphologic sequelae by light microscopy and by immunofluorescence or electron microscopy, with or without mesangial alterations</p> <p>Class V nephritis may occur in combination with class III or class IV, in which case both will be diagnosed</p> <p>Class V nephritis may show advanced sclerotic lesions</p>
Class VI	<p>Advanced sclerotic lupus nephritis</p> <p>$\geq 90\%$ of the glomeruli globally sclerosed without residual activity</p>

Revision of LN Classification (2018)	Activity/Chronicity Modification of the NIH lupus nephritis activity and chronicity scoring system, to be used instead of the currently used A, C, and A/C parameters Global and segmental lesions in class IV Elimination of segmental and global subdivisions of class IV in view of lack of clinical utility Endocapillary lesions of class III/IV The term endocapillary proliferation replaced by endocapillary hypercellularity Histopathological definitions adjusted Mesangial hypercellularity, cellular crescents, fibrous crescents were given new definitions. It is also advised to indicate whether interstitial inflammation occurs in presence or absence of interstitial fibrosis
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Supplementary Table 3. Paediatric doses of drugs used for the treatment of childhood-onset LN

Drug	Recommended dose	Dose adjustment needed in CKD
Glucocorticoids (<i>in prednisone equivalent</i>)	<i>Class II:</i> Starting dose 0.25–0.5 mg/kg/day (max. 30 mg/day) with tapering <i>Class III-IV:</i> Consider IV MP pulses 30 mg/kg/dose for three consecutive days (max. 1000 mg/dose), followed by 1–2 mg/kg/day (maximum 60 mg/day) with tapering <i>Class V:</i> ≤ 0.5 mg/Kg/day with tapering	No
Azathioprine	2–3 mg/kg/day, max. 150 mg/day	Yes
Mycophenolate mofetil	1200 mg/m ² /day (max. 2000 mg/day); in poor response, may be increased to max. 1800 mg/m ² /day (max. 3000 mg/day)	Yes
Cyclophosphamide	<i>Low-dose:</i> IV 500 mg on weeks 0, 2, 4, 6, 8 and 10 (Euro-Lupus regimen) <i>High-dose:</i> IV 500-750 mg/m ² BSA/month (max.1000-1200 mg) for 6 months	Yes
Rituximab	750 mg/m ² /dose (max. 1000 mg) at day 1 and day 15 <i>or</i> 375 mg/m ² /dose once a week for four doses	No
Cyclosporine A	1-3 mg/Kg/day or 100-400 mg/day in 2 doses	Avoid overall
Tacrolimus	0.05 to 0.1 mg/Kg/day or 2-4 mg/day in 2 doses - Titrate to target blood concentration 4-6 ng/ml 12 hours after dose	Yes

IV: Intravenous; MP: Methylprednisolone; BSA: Body surface area; CKD: Chronic kidney disease