Supplementary Table S1. Search strategies and flow data.

Topic	Key words	Date of the search	Records retrieved	Articles selected by title (n)	Articles selected by abstract (n)	Article evaluated full length (n)	Articles included	Additional articles retrieved	Final N of articles included in the SLR
Gastro-intestinal manifestations	"lupus erythematosus, systemic"[MeSH] AND "gastrointestinal" OR "Enteritis"[Mesh]OR "pseudo-obstruction" OR "protein-losing enteropathy" OR "Pancreatitis"[Mesh]	18/09/2021	978	126	72	72	33	4	37
Hepatic involvement in SLE	"lupus erythematosus, systemic"[MeSH] AND "Hepatitis"[Mesh]; "lupus erythematosus, systemic"[MeSH] AND "Liver"[Mesh]	3/9/2021	3660	59	41	19	6	11	6
Interstitial lung disease	lupus erythematosus, systemic"[MeSH] AND "interstitial lung disease" OR "ILD" OR "interstitial pneumonia" OR "Lung Diseases, Interstitial"[Mesh]	27/09/2021	175	56	25	22	13	0	12
Shrinking lung	"lupus erythematosus, systemic"[MeSH] AND "shrinking lung"	27/09/2021	320	51	28	23	10	0	10
Acute lupus pneumonitis	lupus erythematosus, systemic"[MeSH] AND "pneumonitis" OR "Pneumonia"[Mesh]	27/09/2021	1099	149	45	31	5	0	5
Lupus Myocarditis	"Mycocarditis" [MeSH]; AND "systemic lupus erythematosus" [MeSH]	29/9/2021	463	107	27	24	18	0	18
Pulmonary hypertension	"Hypertension, pulmonary" [MeSH]; AND "Lupus erythematosus, systemic "[MeSH;	5/10/2021	354	53	35	34	32	0	32
Ocular manifestations	"ophthalmological manifestations AND lupus erythematosus, systemic"[MeSH]; "retinal AND lupus erythematosus, systemic"[MeSH]; "Ocular AND lupus erythematosus, systemic"[MeSH]	18/9/2021	157	53	17	17	16	-	16
Neurological manifestation Aseptic meningitis (neuro)	"lupus erythematosus, systemic"[MeSH] AND "meningitis, aseptic"[MeSH])	01.09.21	34	20	7	7	3	2	5
Chorea/movement disorder (neuro)	"lupus erythematosus, systemic"[MeSH] AND "chorea"[MeSH) + ("lupus erythematosus, systemic"[MeSH] AND "movement disorders"[MeSH]	01.09.21	120	72	52	7	7	1	8

Supplementary Table S2. Gastrointestinal involvement.

Author, year Country	Study type	Number of SLE pts	Case definition and methods of ascertainment	Prevalence/incidence	Main findings
Pande *et al*. 1993, India	Retrospective	270	GI involvement included diarrhoea with no other specification	GI involvement: 17%	Presentation: similar prevalence in patients with adult vs childhood onset.
Mok *et al*. 2000, China	Case-series	6 IPO patients	Clinical features of intestinal obstruction without an identifiable organic obstructive lesion; presence of multiple fluid levels on abdominal X-rays	NA	Presentation: mean age 29 years; association with ureterohydronephrosis.
Lee *et al*. 2002, Korea	Retrospective	175	CT scan with typical findings.	Lupus enteritis: 9.7% Pancreatitis: 0.6%	Presentation: mean age 38 years; initial manifestation in 35.2%; no association with SLEDAI or laboratory abnormalities.
Pascual-Ramos *et al*. 2004, Mexico	Case-control	895	Typical clinical symptoms and elevation of serum amylase or lipase or evidence of image findings (CT scan or US)	Acute pancreatitis: 0.1%	Presentation: mean age 30 years; association with high disease activity. Prognosis: 20% mortality rate.
Mok *et al*. 2006, China	Prospective	16 PLGE (498 total SLE cases)	Protein leakage from the gastrointestinal tract demonstrated by technetium 99m-labelled human serum albumin scanning	PLGE: 3.2%	Presentation: Mean age 36.2 years, nitial manifestation in 75%; generalized ioedema and GI symptoms
Zonana-Nacach *et al*. 2007, Mexico	Prospective	41 (SLE patients with hospital admission)	Clinical, laboratory and radiologic	Lupus enteritis: 4.9% Acute pancreatitis: 2.4%	
Richer *et al*. 2007, France	Retrospective	201 (paediatric)	Enteritis: CT scan with typical findings Acute Pancreatitis: abdominal pain and increase in blood amylase or lipase activities; IPO: clinical features suggestive of bowel obstruction	GI involvement: 19.4% Enteritis: 1.5% Acute pancreatitis: 5.5% IPO: 0.5% AAC: 0.5%	Presentation: abdominal pain (87%), vomiting and diarrhoea (28%).
Kwok *et al*. 2007, South Korea	Retrospective	706	CT scan with bowel wall thickening, target sign, dilatation of intestinal segments, engorgement of mesenteric vessels and increased attenuation of mesenteric fat	GI involvement: 12.3% Lupus enteritis: 5.8% Acute pancreatitis: 1.7%	Presentation: Mean age 31.85 and mean disease duration 4.85 years.
Zheng *et al*. 2007, China	Retrospective	768	Protein leakage from the gastrointestinal tract demonstrated by technetium 99m-labelled human serum albumin scanning	PLGE: 2.0%	Presentation: mean age 40.1 years; initial manifestation in 53.3%.
Nazarinia *et al*. 2008, Iran	Prospective	410	GI involvement without specification	GI involvement: 8.3%	
To *et al*. 2009, China	Prospective cohort	1082	GI involvement included protein losing gastroenteropathy and enteritis/ colitis	GI involvement: 3.8%	Presentation: GI involvement was clustered with pulmonary, renal and hematologic involvement, but not with mucocutaneous expression.
Xu *et al*. 2010, China	Retrospective	177	Clinical	Enteritis: 10.7% PLGE: 4.5% IPO: 4.0% Pancreatitis: 1.1% Mesenteric venous thrombosis: 0.6%	Presentation: mean age 34.1 years, 30% at disease-onset Prognosis: higher mortality (10.3%) than patients without GI involvement (2.2%)
Mako *et al*. 2010, USA	Prospective cohort	1811	Abdominal pain elevation of pancreatic enzymes and confirmation by an imaging study (CT scan, MRI or US)	Acute pancreatitis: 3.5%	Presentation: association with higher damage index Prognosis: Recurrent episodes in 43%, chronic pancreatitis in 14% and death in 3%
Wang *et al*. 2011, Taiwan	Retrospective	2976	Typical clinical symptoms and elevation of serum amylase or lipase or evidence of image findings (CT scan or US)	Acute pancreatitis: 1.34%	Presentation: higher prevalence in patients with child (5.22%) vs adulthood (0.99%) onset. Prognosis: mortality of 53.8% in adults and 14.8% in children. Mortality was associated with SLE activity and pancreatitis severity.

Author, year Country	Study type	Number of SLE pts	Case definition and methods of ascertainment	Prevalence/incidence	Main findings
Tu *et al*. 2011, Taiwan	Retrospective	1542 (SLE patients with hospital admissions)	Clinical Typical CT scan	Enteritis: 1.8% Acute pancreatitis: 1.0% IPO: 0.6%	Presentation: enteritis was more frequent and recurring in children than adults.
Stefanidou *et al*. 2011, Greece	Retrospective cohort	594	GI involvement defined as hepatomegaly, increase of transaminases, ascites, vasculitis of the intestine, acute pancreatitis, as well as atypical symptoms such as persistent abdominal pain with nausea which could not be attributed to other causes	GI involvement at diagnosis: 9.9%	Presentation: GI involvement at diagnosis was more frequent in male (16.9%) than female (9.2%) patients (p=0.039).
Yang *et al*. 2012, China	Retrospective	4053	Typical clinical symptoms and elevation of serum amylase or lipase or evidence of image findings (CT scan or US)	Acute pancreatitis: 0.67%	Presentation: mean age 30 years; association with high disease activity Prognosis: 37% mortality rate
Law *et al*. 2012, China	Retrospective	48 PLGE (639 total SLE cases)	Protein leakage from the gastrointestinal tract demonstrated by technetium 99m-labelled human albumin scanning or faecal α1-antitrypsin clearance.	PLGE: 7.5%	Presentation: mean age 40.8 years; initial manifestation in 43.8%; peripheral oedema, malaise and weight loss.
García-Carrasco *et al*. 2013, Mexico	Cross-sectional	105	Rome III Adult Questionnaire	Irritable bowel syndrome-type symptoms: 48.6%	Presentation: Mean age 43.6 years, mean disease duration 10.5 years; associated with higher SLE activity and fibromyalgia.
Feng *et al*. 2013, China	Retrospective cohort	1898	GI involvement included: peritonitis, ascites, enteritis/colitis, malabsorption, protein-losing enteropathy, intestinal pseudo-obstruction, hepatitis, acute cholecystitis, acute pancreatitis	GI involvement: 10.3%	Presentation: Similar incidence in patients with juvenile (<18 years), early (18-45 years) or late onset (>45 years).
Xu *et al*. 2015, China	Retrospective	3840	Clinical and evidence of bowel obstruction on plain abdominal x-ray and CT images	IPO: 1.6%	Presentation: mean age 32 years; abdominal pain, nausea and vomiting, constipation, abdominal distension and diarrhoea.
Lim *et al*. 2015, Korea	Retrospective	34	Protein leakage from the gastrointestinal tract demonstrated by technetium 99m-labelled human albumin scanning or faecal α1-antitrypsin clearance.	14/34 PLGE cases due to SLE	Presentation: mean age 37 years; higher ESR and cholesterol levels than idiopathic PLE
Gormezano *et al*. 2015, Brazil	Retrospective	2192	Abdominal pain or vomiting associated increased serum pancreatic enzymes and/or pancreatic radiological abnormalities (CT scan or US)	Acute Pancreatitis: 1.5%	Presentation: higher prevalence in patients with child (3.3%) *vs*. adulthood (1.1%) onset
Wang *et al*. 2016, China	Retrospective	5665	Clinical, laboratory and imaging findings	Pancreatitis: 0.92%	Presentation: mean age 33 years; acute pancreatitis (0.8%) was more prevalent than chronic (0.1%) Prognosis: 30.8% mortality
Alves *et al*. 2016, UK(25)	Retrospective	675	Enteritis: CT scan with bowel wall thickening Pancreatitis: CT scan Celiac disease: biopsy	Lupus enteritis: 0.59% Pancreatitis: 0.44% Celiac disease: 0.15%	Presentation: Abdominal pain, diarrhoea, nausea and vomiting.
Zhang *et al*. 2016, China	Case-control	4331	Clinical symptoms, small bowel distension with air-fluid levels in X-ray or thickened gastric wall and dilated small or large bowels by CT scan	IPO: 1.96%	Presentation: abdominal distension and vomiting in the majority of patients. Initial manifestation 57.6%. Association with pyeloureterectasis, hypocomplementemia, and elevated C-reactive protein
Sönmez *et al*. 2017, Turkey	Retrospective	69 (paediatric)	CT scan with bowel wall thickening and dilated bowel	Lupus enteritis: 1.4%	Presentation: abdominal pain
Tahernia *et al*. 2017, Iran	Retrospective	138 (paediatric)	Typical clinical, laboratory and ultrasound features	Acute pancreatitis: 0.7%	Presentation: 1 case of pancreatitis which was the initial manifestation.
Yang *et al*. 2017, China	Retrospective	8411	AAC: Clinical manifestations and confirmed by radiologic findings including a distended gallbladder with thickened wall, pericholecystic fluid and absence of gallstones	AAC: 0.15%	Presentation: mean age 30.1 years; initial manifestation in 30.8%; abdominal pain and fever in 84.6%

Author, year Country	Study type	Number of SLE pts	Case definition and methods of ascertainment	Prevalence/incidence	Main findings
García-Carrasco *et al*. 2018, Mexico	Cross-sectional	86	Rome III Adult Questionnaire	Functional disorders: 76.7%	Presentation: functional dyspepsia (72.7%), functional heartburn (68.2%), belching disorders (62.1%) and functional bloating (62.1%). Higher prevalence in patients with longer disease duration.
Esmaeilzadeh *et al*. 2019, Iran	Cross-sectional	1322	Not specified	Lupus enteritis/mesenteric vasculitis: 0.2% IPO: 0.08% Acute pancreatitis/PLGE: 0%	Presentation: 2% of hospital admissions
Yoshida *et al*. 2021, Japan	Retrospective	13 lupus enteritis patients	Lupus enteritis: vasculitis or inflammation of the small or large bowel with supportive imaging or biopsy findings	NA	Presentation: median age 41 years; initial manifestation in 46%. Prognosis: low levels of CH50 associated with poor response to therapy.
Chen *et al*. 2021, China	Retrospective	985	Clinical features and radiologic evidence of bowel wall thickening, mesenteric vasodilation, or increased attenuation of mesenteric fat	Lupus enteritis: 4.4%	Presentation: mean age 40 years, mean disease duration 3 years; abdominal pain was the most common symptom (88.4%); ascites, hydronephrosis, leucopenia and reduced C3 were independently associated with lupus enteritis.
Soltani *et al*. 2021, Iran	Cross-sectional	130	CD: biopsy and response to gluten-free diet	GI symptoms: 40% CD: 3.1%	Presentation: mean age 31.5 years; mean disease duration 4.6 years.
Tejera-Segura *et al*. 2021, Spain	Retrospective	3654	BILAG definitions	GI involvement: 49.3% PLGE: 0.5 % CD: 0.1%	Prognosis: GI damage was present in 3.7% of patients and was associated with older age, longer disease duration, vasculitis, renal disease and serositis.
Liu *et al*. 2021, China	Retrospective	8505	Clinical features, typical CT scan and response to steroids	Lupus enteritis: 1.1%	A lupus enteritis risk assessment model was developed, consisting of 11 significantly associated variables, to predict development of lupus enteritis in SLE patients.
Muhammed *et al*. 2021, India	Retrospective	66 acute pancreatitis patients	Acute onset epigastric pain, elevation in serum lipase or amylase and characteristic imaging findings	NA	Presentation: mean age 27.1 years.

AAC: acute acalculous cholecystitis; CD: celiac disease; CsA: cyclosporine A; CT: computer tomography; CYC: cyclophosphamide; GI: gastrointestinal; IPO: intestinal pseudo-obstruction; MMF: mycophenolate mofetil; NA: not applicable; PLGE: protein-losing gastroenteropathy; SLE: systemic lupus erythematosus; US: ultrasound.

Supplementary Table S3. Liver involvement.

Author, year Country	Study type	Number of pts	Case definition and methods of ascertainment	Prevalence/incidence	Main findings
Takahashi *et al*. 2013, Japan	Retrospective study	206 SLE patients	Liver disfunction defined as elevated serum levels of alanine aspartate aminotransferase (AST), alanine aminotransferase (ALT) or alkaline phosphatase (ALP) and γ-glutamyl transpeptidase (γ-GTP). Patients were classified as having liver dysfunction when levels of at least two of these enzymes were outside the normal range.	Prevalence of liver disfunction: 59.7%	Liver dysfunction in the presence of SLE can be caused by many factors, but when extant at the time of SLE onset, either SLE itself or drugs can be the cause. Autoimmune hepatitis should be considered when liver dysfunction is relatively severe.
Piga *et al*. 2010, Italy	Retrospective study	283 SLE patients	Liver enzyme values were considered abnormal when a sustained increase in serum transaminase levels above the normal value was observed for a period of at least three months or when the increase was confirmed in two consecutive assessments.	Prevalence of liver enzyme abnormalities 18.6%; of these 5.8% had lupus hepatitis.	Lupus hepatitis is generally sub-clinical with a fluctuating course and responds well to moderate to high doses of prednisone without progression to end-stage liver disease.
Zheng *et al*. 2013, China	Retrospective/ Prospective study	504 SLE patients	Liver histopathological changes by haematoxylin and eosin (HE) staining and immunopathological changes by direct immunofluorescence test in lupus hepatitis were analysed and compared to those in patients with other liver diseases in a prospective study.	Prevalence of lupus hepatitis: 9.3%	Lupus hepatitis was not infrequent in active SLE patients. Positive intense deposit of complement 1q in liver may be a characteristic immunopathological feature of lupus hepatitis.
Ohira *et al*. 2004, Japan	Case-control study	61 SLE patients 20 patients with autoimmune hepatitis (AIH)	SLE was diagnosed on the basis of the ACR criteria. Diagnosis of AIH was based on the criteria established by the International Autoimmune Hepatitis Group (IAHG) in 1999. Liver biopsy samples were also obtained from all patients with SLE-associated hepatitis. SLE-associated hepatitis was reflecting SLE disease activity without showing histologically chronic active hepatitis, which excluded other causes of liver disease such as viral hepatitis, drug-induced hepatitis and fatty liver. All sera were assessed for reactivity against recombinant ribosomal P0 using commercially available ELISA kits.	Prevalence of liver dysfunction: 55.7%. Of 34 SLE patients with liver dysfunction, anti-ribosomal P antibody was detected in 15 (44.1%).	Anti-ribosomal P antibody may be a useful marker of SLE-associated hepatitis to differentiate it from AIH and other liver dysfunctions in SLE patients.
Calich *et al*. 2013, Brazil	Case-control study	96 AIH patients 82 healthy controls	The diagnosis of AIH was based on the AIH International Group criteria. Frozen sera samples were obtained at the time of diagnosis. The exclusion criteria were the presence of a definite SLE diagnosis and/or the presence of specific disease-related antibodies (anti-dsDNA and anti-Sm).	Moderate to high titers (>40 U) of anti-rib P antibody were found in 9.7% of the AIH patients	The demonstration of anti-rib P in AIH patients without clinical or laboratory evidence of SLE suggests a common underlying mechanism targeting the liver in these two diseases. In addition, this antibody appears to predict the patients with worse AIH prognoses.
Alves *et al*. 2016, UK	Retrospective study	675 SLE patients	Analysis of data from the SLE patient cohort at University College Hospital London, established in 1978, identifying patients with an associated autoimmune gastrointestinal disease (autoimmune hepatitis, primary biliary cirrhosis, ulcerative colitis, Crohn's disease, celiac disease, lupus enteritis and autoimmune pancreatitis.	4.3% of patients with an associated autoimmune GI disease	From the analysis of data the authors found 16 cases of SLE–AIH overlap syndrome. The dominant manifestation was elevation of liver enzymes. The association of PBC with SLE is extremely rare.

Supplementary Tables S4. Pulmonary involvement.

Supplementary Table S4.1. ILD

What is the incidence of ILD in SLE patients?

Author, year Country	Study type	Number of pts	SLE diagnosis/ Referral Cohort	Case definition and methods of ascertainment	Prevalence/ incidence	Main findings
Mosca 2019, international# (1)	retrospective	2/389	- ACR 1997 - SLICC 2012 - early diagnosed SLE (<1yr)	Pulmonary fibrosis	0.5%	
Frade-Sosa, 2020, Spain	retrospective cross-sectional	5/120	- ACR 1997 - SLE and overlap SLE+RA	Not reported	4.2%	• No differences between SLE and SLE+RA
Catoggio, 2015, Latin America	multicentric retrospective longitudinal	114/1480	- ACR 1982 - SLE patients <50yrs - SLE pts ≥50 yrs	Not reported	7.7%	• ILD more common in late-onset SLE ° SLE<50: 7.2% (99/1378) ° SLE ≥50: 14.7% (15/102)
Bankier, 1992, Austria§	Prospective	20/48	- ACR 1982 - SLE patients	- X-ray (3) - CT scan (17)	41.6%	• CT scan findings (n= 45) ° Septal thickening 33% (15) ° Interstitial thickening 33% (15) ° Air-space nodules 22% (10) ° Architectural distortion 22% (10) ° Bronchial wall thickening 20% (9) ° Bronchiectasis 18% (8) ° Pleural irregularity 13% (6) ° GGO 13% (6) ° Consolidation 7% (3)
Saito, 2002, Japan§	retrospective longitudinal	10/243	- Not reported - consecutive SLE patients	CT scan	4.1%	• CT findings (n= 5) ° GGO 60% (3) ° Consolidations 40% (2) ° Consolidations and bronchiectasis 20% (1) ° Septal thickening 80% (4) ° Pleural thickening 100% (5)
Medlin, 2018, USA	Meta-analysis	82/1562	- ACR 1997 - Early (<50) and late (≥50) onset-SLE	Not reported	- Total: 5.2% - early: 4.2% - late: 9.9%	• ILD is more common in late-onset SLE OR 2.56
Frodlund, 2019, USA	Cohort Longitudinal	17/520	- ACR 1982 or SLICC 2012	Clinical symptoms and radiology and/ or lung function test	3.3%	• ILD is more common in pts with severe organ damage
Narváez, 2018, Spain#	Registry	65/3679	- ACR 1997 - SLE (RESSLER-TRANS registry)	- SDI definition (Clinical and radiographical)	2%	• Mean SLE duration: 7.7 ±10.5yrs
Fidler, 2016, Canada§	Cross-sectional	11/110	- ACR 1982 and 1997 - consecutive SLE patients	- presence of reticular or interstitial opacities *Or* - honeycombing ± - GGO	10%	
Bertoli, 2007, international#	Longitudinal cohort (LUMINA)	25/626	- ACR 1982 and 1997 - disease duration ≤5y	Pulmonary fibrosis according to SDI (clinical or radiographic)	4%	
Mochizuki, 1999, Japan§	Retrospective case control	11/137	- ACR 1982 - consecutive SLE	X-rays and chest CT	8%	• Anti-Ro and sicca syndrome more common than in SLE without ILD • Malar rash less common
Haupt, 1981, USA	retrospective cross sectional	16/120	- ARA 1972 - autopsy of SLE from 1988 to 1980	Biopsy	13.3%	• Interstitial pneumonitis n=11 • Interstitial fibrosis n=5

Q2: What *are the clinical characteristics of SLE-related ILD?*

Author, year Country	Study type	Number of pts	SLE diagnosis/ Referral Cohort	Case definition and methods of ascertainment	Prevalence/ incidence	Main findings
Weinrib, 1990, USA#	Longitudinal	14	SLE-ILD	- X-rays infiltrates - symptoms - restrictive pattern at PFT	-	• Symptoms ° Dyspnoea on exertion 100% ° Chest pain 100% ° Cough 100% • X-rays onset ° Diffuse infiltrates 57% (8) ° Bilateral basal infiltrates 42.8% (6) ° Diaphragms elevation 57% (8) ° Pleural thickening 64.2% (9) • X-rays follow up (7.3yrs) ° Diffuse infiltrates 7% (1) ° Bilateral basal infiltrates 42.8% (6) ° Diaphragms elevation 64.2% (9) ° Pleural thickening 42.8% (6) • SLE patients with ILD are older • Deposits of IG and complement found in alveoli of 4 pts
Tanaka, 2018, Japan	Retrospective cross sectional	10	CTD-ILD	- CT scan	-	• CT features ° GGO 100% (10) ° HC 40% (4) • CT pattern ° NSIP 40% (4) ° UIP 30% (3) ° OP 10% (1) ° DAD 10% (1) ° LIP 10% (1) • Compared to other CTDs ° GGO is prevalently peri-BVB
Saito, 2002, Japan§	retrospective longitudinal	5	- Unknown - consecutive SLE patients	CT scan		• CT findings (n= 5) ° GGO 60% (3) ° Consolidations 40% (2) ° Consolidations and bronchiectasis 20% (1) ° Septal thickening 80% (4) ° Pleural thickening 100% (5)
Ooi, 1997, Hong Kong#	Retrospective longitudinal	6	- ACR 1982 - SLE patients with lung disease	CT scan showing honeycombing, septal thickening, GGO not isolated, pleural irregularity, distortion	-	• CT findings for ILD (n= 6) ° Pleural thickening 100% (6) ° Pleural irregularity 100% (6) ° GGO 67% (4) ° Honeycombing 83% (5)
Enomoto, 2019, Japan	Retrospective longitudinal	55	- ACR 1997 or SLICC 2012 - SLE patients with ILD	CT scan reviewed by expert radiologist	-	• Time of onset ° Before SLE diagnosis 12.7% (7) ° After SLE diagnosis 38.2% (21) ° Concomitant 49% (27) • At onset ° Chronic IP: 63.6% (35) ° Subacute IP: 20% (11) ° Acute IP: 12.7% (7) • ILD pattern ° Unclassifiable 29% (16) ° NSIP+OP 25% (14) ° OP 22% (12) ° NSIP 13% (7) ° UIP 9% (5) DAD 2% (1) • Prognosis ° Overall 5-yrs survival: 85.3% ° NSIP+OP is predictive of good prognosis (HR 0.089 vs NSIP)
Bankier, 1992, Austria§	Prospective	20	- ACR 1982 - SLE patients	- X-ray (3) - CT scan (17)		• CT scan findings (n= 45) ° Septal thickening 33% (15) ° Interstitial thickening 33% (15) ° Air-space nodules 22% (10) ° Architectural distortion 22% (10) ° Bronchial wall thickening 20% (9) ° Bronchiectasis 18% (8) ° Pleural irregularity 13% (6) ° GGO 13% (6) ° Consolidation 7% (3)

Author, year Country	Study type	Number of pts	SLE diagnosis/ Referral Cohort	Case definition and methods of ascertainment	Prevalence/ incidence	Main findings
Toyoda, 2019, Japan	Case-control	20	- ACR 1997 and SLICC 2012 - SLE patients with CT scan	- ATS/ERS/JRS/ ALAT statement 2011 - ATS/ERS 2013		• Time of onset ° At SLE diagnosis 50% (10) ° After SLE diagnosis 50% (10) • Clinical features ° Asymptomatic 70% (14) ° Cough 20% (4) ° Dyspnoea 15% (3) ° Lung crepitation 55% (11) • HRCT pattern ° NSIP 55% (11) ° UIP 25% (5) ° OP 10% (2) ° Unclass 10% (2) • Outcome ° Slow progression 60% (12) ° Stable 25% (5) ° Improvement 10% (2) ° Acute exacerbation5% (1) • Other findings ° More common in late onset SLE ° 5y survival not affected by ILD
Mochizuki, 1999, Japan§	Retrospective case control	11	- ACR 1982 - consecutive SLE	X-rays and chest CT	8%	• Anti-Ro and sicca syndrome more common than in SLE without ILD • Malar rash less common
Cha, 2006, USA	Retrospective cross sectional	1	- not reported - patients with lung biopsy of LIP	Biopsy		• LIP

Supplementary Table S4.2. shrinking lung

Author, year Country	Study type	Number of pts	SLE diagnosis/Referral Cohort	Case definition and methods of ascertainment	revalence/ Pincidence	Main findings
What is the incidence of shrinking lung syndrome in SLE patients?						
Narváez, 2018, Spain#	Registry	28/3679	- ACR 1997 - SLE (RESSLER-TRANS registry)	- BILAG 2004 definition (TLC<70%, normal KCO, dysfunctional diaphragm movements)	0.8%	• Mean SLE duration: 6 ±6.8yrs
Gheita, 2011, Egypt#	Prospective	27/200	- ACR 1997 - consecutive SLE patients	- dyspnoea on exertion - restrictive pattern - diaphragm elevation	13.5%	• Clinical findings ° Dyspnoea 100% (27) • CT scan ° Bilateral diaphragm elevation 29.6% (8) ° Right diaphragm elevation 70.4% (19) ° Atelectasis bands 22% (6) • PFT Restrictive pattern 100% (27)
Fidler, 2016, Canada§	Cross-sectional	10/110	- ACR 1982 and 1997 - consecutive SLE patients	- unexplained dyspnoea - FVC<80% *And* - no ILD on imaging ± diaphragm elevation	9.1%	
Deeb, 2018, Canada§	retrospective longitudinal	22/1439	- ACR 1997 - SLE patients from 1980 to 2018	- restrictive pattern with KCO>65% - no lung disease	1.5%	
Bertoli, 2007, international#	Longitudinal cohort (LUMINA)	4/626	- ACR 1982 and 1997 - disease duration ≤5y	- according to SDI (diaph elevation X-rays)	0.6%	
What ***are the clinical characteristics of shrinking lung syndrome?***						
Duron, 2016, France	retrospective longitudinal	15	- ACR 1997 - SLE patients	Restrictive pattern at FVC and exclusion of other causes		• CT/radiographic findings ° Elevated emidiaphragm 80% (12) ° Basal atelectasis 53% (8) ° Pleural thickening 27% (4) ° Pleural effusion 13% (2) • Symptoms ° Dyspnoea 100% (15) ° Chest pain 93% (14) ° Cough 13% (2) • Other findings ° EMG normal (4/4) ° MIP and MEP impaired (5/8) ° Impaired diaphragm mobility 4/4 • Outcome clinical ° Improvement 75% (9/12) ° Stability 25% (3/12) • Outcome PFT ° Improvement 70% (7/10) ° Stability 20% (2/10) • Outcome radiographic ° Improvement 60% (3/5) ° Stability 40% (2/5)
Weinrib, 1990, USA #	Longitudinal	9	SLE-ILD	- X-rays bilateral diaphragm elevation - symptoms - restrictive pattern at PFT		
Souza Neves, 2010, Brazil	Retrospective cross sectional	7	- ACR 1982 - SLE	- Dyspnoea *and* - decreased lung volume without evidence of ILD.		• Anti-Ro 6/7 • Serositis 7/7
Oud, 2005, Netherlands	Case series	5	- ACR 1982 - SLE	- dyspnoea *and* - restrictive pattern *and* - no ILD		• 5/5 pts improved with steroid
Karim, 2002, UK	Longitudinal	7	- ACR 1997 - SLE patients with SLS	- dyspnoea - restrictive pattern - diaphragm alteration (x-rays)		• Clinical features ° Dyspnoea 100% (7) ° Chest pain 86% (6) • Radiological findings ° Emidiaphragm elevation at X-rays 71.4% (5) ° FVC<70%: 100% (7) ° Normal CT scan 43% (3) ° Pleural thickening at CT scan 16% (1) ° Small atelectasis 28% (2) • Stable or improved 5/7

Author, year Country	Study type	Number of pts	SLE diagnosis/Referral Cohort	Case definition and methods of ascertainment	revalence/ Pincidence	Main findings
Henderson, 2013, USA	Retrospective longitudinal	4	- ACR 1997 - CTD patients	- dyspnoea - restrictive pattern w normal KCO - decreased lung volume on x-rays - no alterations at ILD		• Clinical features ° Pleuritic chest pain 75% (3) ° Dyspnoea 100% (4) • Radiographic findings ° Pleural effusion75% (3) ° Diaphragm dysmotility 50% (2) ° Elevated hemidiaphragm 25% (1) ° Low lung volume 75% (3) • PFT ° FVC<70%: 100% (4) ° DLCO >80%: 100% (4) • Reduced pulmonary compliance (oesophageal manometry): 100% (4) • Pleural inflammation may inhibit inspiration through neural reflex and pain and following parenchymal changes
Gheita, 2011, Egypt#	Prospective	27	- ACR 1997 - consecutive SLE patients	- dyspnoea on exertion - restrictive pattern - diaphragm elevation	13.5%	• Clinical findings ° Dyspnoea 100% (27) • CT scan ° Bilateral diaphragm elevation 29.6% (8) ° Right diaphragm elevation 70.4% (19) ° Atelectasis bands 22% (6) • PFT ° Restrictive pattern 100% (27)
Deeb, 2018, Canada§	retrospective longitudinal	22	- ACR 1997 - SLE patients from 1980 to 2018	- restrictive pattern with KCO>65% - no lung disease	1.5%	• Clinical features (n=22) ° Dyspnoea 95.5% (21) ° Chest pain 91% (20) • X-rays (n=20) ° hemidiaphragm elevation 60% (12) • PFTs (n=20) ° Decreased MIP or MEP 80% (16)
Borrell, 2016, Spain	Case series	9	- ACR 1997 - SLE and SLS	- dyspnoea ± chest pain - restrictive pattern - no lung disease		• Clinical features ° Chest pain 89% (8) ° Fever 33% (3) • Radiographic findings ° Bilateral diaph elevation 56% (5) ° Unilateral diaph elevation 44% (4) ° Pleural effusion 56% (5) ° Basal atelectasis 89% (8)

Supplementary Table S4.3. Acute pneumonitis.

Author, year Country	Study type	Number of pts	SLE diagnosis/ Referral Cohort	Case definition and methods of ascertainment	Prevalence/ incidence	Main findings
Mosca, 2019, international#	retrospective	6/389	- ACR 1997 - SLICC 2012 - early diagnosed SLE (<1 yr)	-	1.5%	-
Narváez, 2018, Spain#	Registry	118/3679	- ACR 1997 - SLE (RESSLER-TRANS registry)	- BILAG 2004 (radiological features of alveolar infiltration not due to infection or haemorrhage)	3.6%	• Mean SLE duration: 5.4 ±7.4yrs
What *are the clinical characteristics of SLE-related acute pneumonitis?*						
Ooi, 1997, Hong Kong	Retrospective longitudinal	2	- ACR 1982 - SLE patients with lung disease	CT scan with isolated GGO	3/10 patients	• CT findings ° Isolated GGO 2/2 ° Consolidations 1/10 ° Pleural thickening
Wan, 2016, Malaysia	Case series	5	- ACR 1997 - ALP as first manifestation	- clinical features - pulmonary infiltrates - negative cultures on admission		• Clinical features ° Fever 100% (5) ° Cough 80% (4) ° Lung crepitation 80% (4) ° Dyspnoea 60% (3) ° Hypoxia 60% (3) • Radiological findings ° X-ray infiltrates 100% (5) ° GGO 2/2

Supplementary Tables S5. Cardiac involvement.

Supplementary Table S5.1. Myocarditis.

Author, year Country	Study type	Number of pts	Case definition and methods of ascertainment	Prevalence/incidence	Main findings
Du Toit *et al*. 2021, South Africa	Prospective cohort	49	Clinical and subclinical lupus myocarditis (echocardiography and cardiac magnetic resonance)	-	Subclinical lupus myocarditis did not progress to clinical manifestations and had no significant prognostic implications over 12 months.
Du Toit *et al*. 2020, South Africa	Prospective cohort	49	All patients underwent cardiac magnetic resonance	23/49 (46.9%) patients had myocardial injury (60.9% subclinical)	Evidence of myocardial injury frequently occurs in lupus and is often subclinical
Gartshteyn, 2020, USA	Case series	11	Endomyocardial biopsies	-	Unspecific findings
Jia E, 2019, China	Retrospective	43	Clinical, biochemical and echocardiographic findings	5.7%	Myocarditis is a risk factor of mortality
Tanwani, 2018, Canada	Prospective cohort	30	Clinical, biochemical and echo-cardiographic findings	1.6%	40% mortality
Wang, 2018, China	Retrospective study	13	Clinical, biochemical echo-cardiographic and cardiac magnetic resonance findings	1.6%	Efficacy of rituximab in three cases
Thomas, 2017, France	Retrospective study	29	Clinical, biochemical echo-cardiographic and cardiac magnetic resonance findings	NA	Two deaths. Lupus myocarditis can be the first manifestation of the disease. The long-term prognosis was positive
du Toit, 2017, South Africa	Retrospective study	28	Clinical and echocardiographic findings	28/457 (6.1%)	Patients typically presented with high SLE disease activity and the majority had concomitant lupus nephritis. Lymphopenia and low LVEF at presentation were of prognostic significance, associated with lupus myocarditis related mortality or a persistent LVEF < 40%
Zhang, 2015, China	Retrospective case control study	25	Clinical and echocardiographic findings	NA	Lupus myocarditis could result in cardiac dysfunction and even sudden death. High disease activity might potentially predict the occurrence of lupus myocarditis at the early stage of lupus.
Zhang, 2015, China	Case control study	24	Cardiac magnetic resonance (1.5 Tesla)	NA	In inactive SLE with normal cardiac function, low grade myocardial inflammation can be detected by cardiac magnetic resonance (T2).
Gracia, 2014, GLADEL	Longitudinal inception cohort	7	Physician's judgment	0.4% (all cases of lupus); 3,5% (primary lupus cardiac involvement)	Primary cardiac disease had no impact on mortality (no information on myocarditis)
Mavrogeni, 2013, Greece	Cross-sectional controlled study	16	Clinical, biochemical, echo-cardiography, cardiac magnetic resonance and endomyocardial biopsy	NA	Due to the subclinical presentation of lupus myocarditis and the limitations of endomyocardial biopsy, magnetic resonance represents the best alternative for the diagnosis
Zawadowski GM, 2012. USA	Retrospective case series	24	Clinical, echocardiographic and cardiac magnetic resonance	NA	Association with anti-SSA and anti-RNP antibodies
Panchal, 2006, India	Case series	10	Autopsy	NA	Cardiovascular disease contributes significantly to the mortality in patients with lupus in India
Logar D, 1990, Slovenia	Retrospective	3	Clinical and electrocardiographic	3/36 anti-Ro positive patients	Myocarditis is reasonably common in adults with lupus and is associated with anti-Ro antibodies.
Badui, 1985	Longitudinal cohort	14	Electrocardiogram and echo-cardiogram	14%	Suggest to study routinely all patients with SLE through non-invasive cardiological methods.
Gobaira Maluf, 1982	Retrospective		Review of unselected 32 active lupus patients	16%	NA
Bulkley, 1975, USA	Case series	3	Autopsy	3/36	All patients also had endocarditis and pericarditis.

LVEF: left ventricular ejection fraction; GLADEL: Grupo Latino Americano De Estudio del Lupus Eritematoso.

Supplementary Table S5.2. PAH.

Author, year Country	Study type	Number of pts	Case definition and methods of ascertainment	Prevalence/incidence	Main findings
Guo *et al*. 2021, China	Prospective cohort	112	PAH by right heart catheterization	n/a	ratio between tricuspid annular plane systolic excursion (TAPSE) and pulmonary artery systolic pressure (PASP) measured by echocardiography as a simple surrogate of RV to pulmonary circulation coupling predicts the outcome of SLE-associated PAH.
Li *et al*. 2019, China	Retrospective cohort in lupus nephritis patients	352	Lupus nephritis by biopsy PAH by right heart catheterization and ultrasound	24/352 (6,8 %)	activation of the complement alternative pathway may be involved in the pathogenesis of pulmonary hypertension in lupus nephritis.
Che *et al*. 2019, Taiwan	Retrospective cohort	15.783	Diagnosis mentioned in database	336/15.783 (2.13 %)	PAH is a rare complication of SLE; majority of PAH cases occur within the first 5 years following diagnosis. Systemic hypertension may be a risk factor for PAH development. The overall 5-year survival rate after PAH diagnosis was 70.1%.
Zhang *et al*. 2019, China	Cross-sectional cohort	2278	PAH by right heart catheterization	292/2278 (14.7%)	serositis, anti-ribonucleoprotein (RNP) antibody positivity and diffusion capacity of carbon monoxide in the lung (DLCO)/%Pred <70% were independent predictors of PAH.
Donnarumma *et al*. 2019, Brasil	Retrospective case-control	65 (21 PAH, matched controls)	PAH by right heart catheterization	n/a	Scleroderma pattern with nailfold capillaroscopy associated with a 6.3-fold increased risk for PAH development
Qian *et al*. 2019, China	Multicentre prospective cohort	310	PAH by right heart catheterization	n/a	1-, 3- and 5-year survival rates were 92.1%, 84.8% and 72.9%, respectively.
Wang *et al*. 2018, China	Prospective study	60	PAH by right heart catheterization and ultrasound SLEDAI SF-36	n/a	tricuspid annular plane systolic excursion (TAPSE) showing strong correlation with pulmonary vascular resistance and cardiac index and is associated with health-related quality of life
Xu *et al*. 2018, China	Retrospective study	1734	PAH by right heart catheterization and ultrasound	95/1734 (5.5 %)	significant association of high fibrinogen, serositis, and thrombocytopenia with PAH
Sun *et al*. 2018, China	Prospective case control study	30 healthy controls 37 SLE-patients without PAH 34 SLE-patients with mild PAH 31 SLE-patients with severe PAH	PAH by right heart catheterization and ultrasound	n/a	Two-dimensional speckle-tracking echocardiography could effectively assess right atrium function in patients with SLE with different severities of PAH
Lv *et al*. 2018, World	Meta-analysis	7121	PAH by right heart catheterization and / or ultrasound	426/7121 (6.0%); weighed proportion 8%	Pooled prevalence of PAH 8%; significant differences in PAH prevalence in SLE patients of different gender, age, regions, year of publication, and diagnostic methods
Hachulla *et al*. 2018, France	Cross-sectional cohort	69 PAH 101 SLE without PAH	PAH by right heart catheterization	n/a	5-year survival rate of 83.9% after PAH diagnosis. Anti-SSA/SSB antibodies may be a risk factor for PAH; presence of anti-U1-RNP antibodies appears to be a protective factor regarding survival
Zuily *et al*. 2017, World	Meta-analysis	4480 SLE-patients 410 PAH patients	PAH by right heart catheterization and/or ultrasound	410/4480 (0.9 %)	Antiphospholipid antibodies can identify patients at risk for PAH
Wang *et al*. 2017, World	Meta-analysis	505	PAH by right heart catheterization and/or ultrasound	n/a	anti-RNP antibody and anti-Sm antibody are risk factors for SLE-associated pulmonary arterial hypertension
Huang *et al*. 2016, China	Case-control study	111	PAH by right heart catheterization	n/a	pericardial effusion and positive anti-RNP antibody are risk factors for SLE-associated PAH. Long SLE disease duration, the presence of interstitial lung disease, without acute skin rash, positive anti-SSA antibody, low SLEDAI and ESR, and high uric acid levels are also associated with PAH in SLE patients
Wang *et al*. 2016, China	Prospective cohort	60	PAH by right heart catheterization SF-36 SLEDAI	n/a	Self-reported HRQoL was impaired in patients with SLE-PAH. Higher CO was the most important predictor for better HRQoL in these patients
Perez-Renate *et al*. 2016, Spain	Prospective cohort	152	PAH by ultrasound and/or right heart catheterization	3/152 (2.0 %)	Low prevalence of PAH in SLE. Occult left ventricular diastolic dysfunction was a frequent diagnosis of unexplained dyspnoea. Dyspnoea, DLCO, and NT-proBNP could be predictors of pulmonary hypertension in patients with SLE

Author, year Country	Study type	Number of pts	Case definition and methods of ascertainment	Prevalence/incidence	Main findings
Qian *et al*. 2016, China	Meta-analysis	323	PAH by right heart catheterization	n/a	The pooled 1-, 3- and 5-year survival rates were 88%, 81% and 68%, respectively. WHO Functional class III/IV was an independent prognostic factor of mortality. Higher mean pulmonary arterial pressure (mPAP), higher pulmonary vascular resistance (PVR), lower six minutes walking distance (6MWD), higher brain natriuretic peptide (BNP) and higher N-terminal proBNP (NT-proBNP) level were related to poor survival.
Li *et al*. 2015, China	Prospective cross-sectional cohort study	1934	Resting transthoracic echo-cardiography	74/1934 (probable PAH, 3.8 %)	association of pericarditis, pleuritis and anti-RNP positivity with PAH suggests that higher disease activity and vasculopathy may contribute to the development of PAH
Hübbe-Tena *et al*. 2014, Mexico	Retrospective cohort	55	ultrasound	n/a	echocardiography-based definitions of PH are useful to predict 6-year mortality in SLE patients. A history of pulmonary thromboembolism and lung vasculitis/ haemorrhage, cumulated organ damage and long-lasting disease are associated with PAH in SLE
Kim *et al*. 2014, Korea	Prospective cross-sectional cohort	114	ultrasound	9/114 (6.8 %)	Serum uric acid level may be useful as a surrogate marker for screening of PH in patients with SLE
Xia *et al*. 2014, China	Meta-analysis	642	Transthoracic echocardio-graphy (TTE), X-ray, electrocardiogram and right heart catheterization	N/a	mean age 35.5 years, male to female ratio was 1:14, mean duration of SLE when PAH was diagnosed 10.7 years. Prevalence of PAH in SLE was 2.8-23.3%. Predictors include Raynaud's phenomenon (41.4%), serous effusion (27.7%), positive RNP (51.5%) and positive anticardiolipin antibodies (46.6%).
Chow *et al*. 2012, World	Systematic review	93	PAH by ultrasound and/or right heart catheterization	n/a	Elevated mean pulmonary artery pressure, Raynaud's phenomenon, thrombocytopenia, plexiform lesion, infection, thrombosis, pregnancy, pulmonary vasculitis and anticardiolipin antibodies were associated with decreased survival
Lian *et al*. 2012, China	Retrospective case-control study	106 SLE-patients 41 PAH/SLE patients	PAH by ultrasound and/or right heart catheterization	n/a	Raynaud's phenomenon, anticardiolipin antibodies, and anti-U1RNP were independent predictors of PAH in SLE
Foïs *et al*. 2010, France	Retrospective cohort	93	Ultrasound	12/93 (12.9 %)	PAH more common in Black subjects, in patients with longer disease duration and in patients with a history of peripheral nervous system involvement, pericarditis, anti-Sm, and anticardiolipin antibodies
Cefle *et al*. 2011, Turkey	Retrospective case-control study	97 SLE patients 10 PAH/SLE patients	ultrasound	n/a	Association PAH with antiphospholipid antibodies and Raynaud's phenomenon
Prabu *et al*. 2009, UK	prospective cross-sectional study	283	Resting transthoracic echo-cardiography	12/283 (4.2 %)	risk factor for PAH was presence of lupus anticoagulant
Gonzalez-Lopez *et al*. 2004, Mexico	Prospective study	34	Doppler echocardiography	n/a	IV cyclophosphamide decreased the median values of SPAP from 41 to 28 mmHg ($p < 0.001$), and enalapril from 35 to 27 mmHg ($p = 0.02$)
Tanaka *et al*. 2002, Japan	Case series	194 SLE patients 12 PAH/SLE patients	PAH by ultrasound and/or right heart catheterization	12/194 (6.2 %)	Treatment with corticosteroids +/- cyclophosphamide improved right ventricular systolic pressure
Horn *et al*. 2000, USA	Case series	6	Not reported	n/a	adverse effect of epoprostenol therapy (thrombocytopenia), may outweigh the potential benefits of epoprostenol treatment in some pulmonary hypertension patients with a history of SLE
Robbins *et al*. 2000, USA	Case series	6	PAH by right heart catheterization	n/a	Epoprostenol was effective for the treatment of pulmonary hypertension in this small group of patients with SLE
Li, Tam, 1999, Hong Kong	Retrospective cohort	419	2 dimensional echocardiogram and Doppler studies at rest	18/419 (4.3 %)	Association with Raynaud's phenomenon. Prognosis with mild/moderate PAH good (no changes after two years); 4 deaths (22.2 %) in two years
Winslow *et al*. 1995, USA	Longitudinal cohort	28	Doppler echocardiographic recordings of tricuspid insufficiency, with saline contrast enhancement when necessary	n/a	A significant increase in mean systolic pulmonary artery pressure was detected in the SLE patients during the 5y follow-up period: 23.4 vs 27.5 mm Hg ($p < 0.005$)

PAH: pulmonary arterial hypertension; SLE: systemic lupus erythematosus; SLEDAI: SLE-disease activity index; HRQoL: health-related quality of life

Supplementary Table S6. Ocular manifestations.

Author, year Country	Study type	Number of pts	Case definition and methods of ascertainment	Prevalence/incidence	Main findings
Lanham,1982, UK	Cross sectional, retrospective	52	clinical evaluation, FA	15 LR (28.8%): Disc vasculitis n.4, cotton wool spots, n.6 Focal leakage on FA, n.5	LR as feature of more active disease. Cotton-wool spots in particular are associated with disease activity.
Klinkhoff, 1986, Canada	Cross-sectional, prospective	43	clinical evaluation, visual field assessment, FA	7 LR (16.3%)	LR during SLE flare in 6/7 patients; in 5/6 LR improves alongside with general improvement.
Stafford-Brady, 1988, Canada	Cross-sectional, retrospective	550	clinical evaluation, FA	41 LR (7.4%) Microangiopathy, n.34 Transient papilledema, n.3 ION, n.2 CRAO, n.1 CRVO, n.1 Serious retina detachment, n.1	Patients with cotton wool or papilledema had not visual acuity loss; retinopathy was associated with active disease and active NPSLE
Montehermoso, 1999, Spain	Case-control, retrospective	82 SLE	clinical evaluation, FA	13 LR (15%) CRAO n.5 CRVO n.1 Microangiopathy n.8 ION n.1	Higher prevalence of aPL in patients with LR, also 5 cases of APS
Nguyen, 2000, USA	Case series	28	clinical evaluation, FA	28 coroidopathy	All cases associated with systemic disease activity
Ushiyama, 2000, Japan	Cross sectional	69	clinical evaluation, FA	6 LR (8.7%)	1 LR as SLE onset, retinopathy associated with higher prevalence of aCL IgG
Sitaula, 2011, Nepal	Cross sectional	89 SLE	clinical evaluation, FA, Schirmer	8 LR (9%) (CRAO, n.1) 1 nodular episcleritis	-
El Shereef, 2013, Egypt	Case control	52 SLE	clinical evaluation, FA, Schirmer	5 LR (9.6%): 4 bilateral vasculitis 1 unilateral	Higher SLEDAI and aCL prevalence in patients with LR
Gao, 2017, China	Retrospective case control	5298 SLE	clinical evaluation, FA	10 LR (0.66%)	NPSLE and leukopenia were independent risk factors for LR; anti-SSA antibody positivity was negatively correlated with LR
Kahwage PP, 2017, Brasil	Multicentre cohort study	852 childhood onset SLE	clinical evaluation, FA	7 uveitis (0.8%) posterior, n.4 anterior, n.2 panuveitis, n.1	Uveitis was the initial manifestation in 6/7 cases,
Seth, 2018, India	Hospital- based cross sectional	437	clinical evaluation, FA	45 LR (10.3%) cotton wool spots n. 41 hard essudates n.11 vasculitis n.3 CRAO n.1 CRVO n.1	lupus nephritis, anti-Sm and haemolytic anaemia were significantly associated with LR
Dammacco, 2018, Italy	Retrospective cohort study	98 SLE	clinical evaluation, FA	3 Scleritis-episcleritis (3.1%) 1 Purtscher-like retinopathy (1.0%) 1 CRAO (1.0%)	-
Braga, 2019, Portugal	cross-sectional case control study	15 SLE LN in remission 15 SLE no LN 15 HC	clinical evaluation, FA, OCT	NA	Choroid thickness higher in LN in remission than in SLE without LN or HC. LN is a risk factor for choroidopathy in these patients.
Hsu, 2020, Taiwan	Population-based data	521	ICD-9 code	44 LR (8.4%) 8 scleritis and episcleritis (1.7%)	SLE patients had a higher prevalence and frequency of outpatient ophthalmologist visits compared with HC
Dias-Santos, 2020, Portugal	Prospective cross sectional	161	clinical evaluation, FA, OCT	2 LR (1.2%) 1 Coroidopathy (0.6%) 1 Anterior uveitis (0.6%)	Reduction of events related to high systemic disease activity, with an increased drug and age-related complications
Uribe-Reina, 2021, Colombia	Observational cross-sectional study	79	Clinical charts	2 ON (2.5%)	-

LR: lupus retinopathy; FA: fluorescein angiography; ION: ischaemic optic neuropathy; CRAO: central retinal artery occlusion; CRVO: central retinal venous occlusion; aPL: antiphospholipid antibodies; APS: antiphospholipid antibodies; NPSLE: neuropsychiatric systemic lupus erythematosus; ON: optic neuritis; HC: healthy controls.

Supplementaty Table S7. Myelopathy and movement disorders.

Author, year Country	Study type	Number of pts	Case definition and methods of ascertainment	Prevalence/incidence	Main findings
Aseptic meningitis					
Sato *et al*. 2017, Japan	Cross sectional cohort study	5 pts with aseptic meningitis	Not specified	Out of 31 cases of NPSLE 5 had aseptic meningitis	Autoantibodies to triosephosphate isomerase (TPI) are elevated in SLE patients with aseptic meningitis
Hanly *et al*. 2007 SLICC (multiple nationalities)	Prospective inception cohort	4 SLE patients with aseptic meningitis, a total of 572 SLE patients included.	Clinical evaluation, neuropsychological testing (8 cognitive domains) NP events guided by the ACR glossary for NPSLE.	4/572(total SLE) (0.7%) 4/242(total NPSLE events in the cohort)	Article focus: NPSLE at time of diagnosis. 4 cases of aseptic meningitis in a total SLE inception cohort of 572 patients
Baizabal-Carvallo *et al*. 2009, Mexico	Retrospective cohort study	10 SLE patients with aseptic meningitis out of a total of 1411 SLE patients	Clinical assessment and lumbar puncture. CSF pleocytosis. Assessment for virus, bacteria and fungi negative.	10/1411 (0,7%)	Uncommon presentation of SLE. Associated with lymphopenia, steroid use, damage and active systemic disease.
Trysberg *et al*. 2004, Sweden	Retrospective cohort study	1 patient with aseptic meningitis	Clinical, MR-I, lumbar puncture.	1/122 (total SLE patients included) (0,8%) 1/43 (total NPSLE patients in the cohort)	High levels of matrix metalloproteinases in NPSLE particularly MMP-9 is associated with brain damage.
Kim *et al*. 2011, Korea	Retrospective cohort study	9 patients with aseptic meningitis (11 patients with septic meningitis) Total SLE patients 1420.	Clinical, lumbar puncture, CSF pleocytosis. Assessment for virus, bacteria and fungi negative.	9/1420 (0,6%)	Comparing infectious and aseptic meningitis in SLE patients, they found: altered mental status, plasma leukocytosis, neutrophilia, CSF pleocytose and hypoglycemia more prominent with infectious meningitis.
Movement disorders					
Reiner *et al*. 2011, France	Retrospective cohort study	32 patients with SLE, APS or positive aPL and a history of chorea (30 of the patients SLE or incomplete SLE)	Clinical, MRI	N/A	92% of the included patients had positive aPLs. Less severe than NPSLE in general. Increased risk of arterial thrombosis at long-term follow-up.
Dale *et al*. 2011, Australia	Case series	6 patients with chorea (2 SLE, 2 SLE+APS, 2APS)	Clinical, assessment of IgG binding to neuronal cells.	N/A	Chorea is associated with aPLs. Therapeutic benefits of RTX and plasma exchange support of humoral autoimmune pathogenesis.
Baizabal-Carvallo *et al*. 2011, Mexico	Retrospective cohort study, single centre (1989-2007)	5 patients with chorea	Clinical assessment, ACR classification, MRI	N/A	Chorea in patients with lupus may result from an immunologic-mediated mechanism or ischaemia
Dale *et al*. 2010, Australia	Prospective cohort study	52 children with movement disorders were included. 3 of these children had SLE.	Clinical assessment. Movement disorders were defined according to the Movement Disorders Society (http://www.movementdisorders.org/disorders)	N/A	Acute onset movement disorders in children may be treatable. Identifying underlying disease pivotal for correct treatment.
Hanly *et al*. 2007 SLICC (multiple nationalities)	Prospective inception cohort	2 SLE patients with movement disorders, a total of 572 SLE patients included.	Clinical evaluation, neuropsychological testing (8 cognitive domains) NP events guided by the ACR glossary for NPSLE.	2/572(total SLE) (0.3%) 2/242(total NPSLE events in the cohort)	Article focus: NPSLE at time of diagnosis. 2 cases of movement disorders in a total SLE inception cohort of 572 patients
Asherton *et al*. 1987, UK	Retrospective cohort study, single centre (1980-1985)	12 SLE patients with chorea	Clinical assessment	12/500 (2,4%)	Chorea appears early in most patients. Development of cerebral infarctions or TIAs occurred subsequently in seven of nine patients with aPLs.
Guttman *et al*. 1987, Canada.	Case series	4 SLE patients with chorea	Clinical assessment. Positron computed tomography.	N/A	Striatal hypometabolism, as seen in other disorders manifesting chorea, is not found in SLE related chorea
Bruyn *et al* 1984, the Netherlands	Retrospective case review of SLE-chorea cases published 1941-1982	51 SLE cases with chorea	Assessment of literature. Sufficient data needed to be available to confirm SLE diagnosis.	N/A	Choreatic movements occurred early in the course of the disease in most cases; the duration varied from several days to 3 years.