Results Of 169 children, 139 (82%) females. Median age at disease onset:11.4 years(3.4–18),median age at diagnosis:12 (3.5–19).

20% had history of autoimmune disease in first degree relative.

Therapy at disease onset(first 6 months):
Hydroxychloroquine: 100%,
Glucocorticoids: 98%,
Mycophenolate: 33%,
Methotrexate: 27%,
Azathioprine: 16%,
Cyclophosphamide: 9%,
Rituximab: 2%.

At last follow up: Glucocortocoids: 37%

Median follow up 48 months(1–195 months), Mortality:4%,lost to follow up: 19%, active disease at last follow up:25%

Conclusions Patients seen at our centre had a significant disease burden with a median SLEDAI score of >20 at presentation.

Upto 1/2 of the study population did not have a malar rash. 38% had renal disease. Fever was seen in 82% and often was the cause for seeking medical opinion. This is a small data set from a tertiary level centre and not representative of the community disease.

143 RITUXIMAB IN SLE MEMBRABOUS NEPHROPATHY

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Background and Aims We hereby report three cases of biopsy proven Type 5 SLE membranous nephropathy (T5: SLEMN) which responded to Rituximab after failed first line induction agents.

Methods Retrospective chart review

Results Case 1: Rituximab was used in this 14 year old girl as she failed to standard therapy of cyclophosphamide (750 mg/m^2) and high dose corticosteroid along with angiotensin convertase inhibitor (ACEI). 30 days post rituximab 24 hour proteinuria dropped (6790 to 876 mg) and albumin rose (1.8 mg/dl to 3 mg/dl). She is in remission at 14 month on low dose steroid and mycophenolate mofetil (MMF).

Case 2: 10 year old girl whose presentation was similar to Case 1 and failed to show any significant improvement tostandard therapy. 45 days post rituximab, 24 hour proteinuria dropped (4900 to 690 mg) and albumin rose (1.9 mg/dl to 3.3 mg/dl). At 12 month she is in remission on low dose steroid and MMF.

Case 3: 12 year old girl presented with features of nephrotic syndrome. At 30 days follow up there were no improvement despite standard therapy and she also started to have neuropsychiatric manifestation. 60 days post rituximab 24 hour proteinuria dropped (3548 to 300 mg) and albumin rose (1.8 mg/dl to 3.7 mg/d)l. Her neuro-psychiatric manifestation also improved.

Conclusions In all 3 cases 2 doses of rituximab at 375 mg/m^2 each achieved CD 19 count of zero (which normalised by 14, 9 and 11 month respectively) along with significant drop in proteinuria.

144 ROLE OF TUBULOINTERSTITIAL LESIONS IN PREDICTING RENAL OUTCOME AMONG PAEDIATRIC ONSET LUPUS NEPHRITIS – A RETROSPECTIVE COHORT STUDY

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Background and aims Raising evidence supported a prognostic utility of tubulointerstitial lesions in lupus nephritis (LN). The exact prevalence of tubulointerstitial abnormalities and its predictive value among paediatric onset systemic lupus erythematous (pSLE) cases, however, remained unknown.

Methods Sixty-seven pSLE subjects diagnosed with LN with initial renal samples available were enrolled and followed for an average of 6.43±3.06 years. Renal histology was evaluated according to the International Society of Nephrology/Renal Pathology Society classification, National Institute of Health classification and tubulointerstitial activity index (TIAI).

Results Tubulointerstitial injuries were observed in 38.81% of all LN cases, including 13.33% with non-proliferative lupus nephritis (nPLN) and 46.15% of with proliferative lupus nephritis (PLN). Tubulointerstitial injuries occurred solitarily in cases with nPLN(13.33%), but always associated glomerular changes and significantly impacted renal survival (p=0.032) among those with PLN. TIAI associated glomerular abnormalities (p=0.031) but did not correlate renal performance or subsequent outcome (p=0.445). Among the chronicity index, it was the chronic tubulointerstitial lesions which provided prognostic information (p=0.012). We observed a synergistic effect of all tubulointerstitial abnormalities rather than an individual factor attributed the prognostic utility (p=0.025 vs. p=0.083, 0.055, 0.354). Finally, considering tubulointerstitial injuries in PLN further discriminated subsequent renal outcome (p=0.006).

Conclusions The prevalence and clinical significance of tubulointerstitial abnormalities were similar among the pSLE and the adult population. With its importance in identifying those at risk of renal failure, histologic classification considering tubulointerstitial lesions may potentially assist outcome prediction.

Patient-submitted abstracts

145 A PATIENT'S FOUR DECADE JOURNEY TO WELLNESS : A MODEL OF CARE FOR LIVING WELL WITH LUPUS

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10.1136/lupus-2017-000215.145

Background and aims Mary Erceg is a former teacher and senior public servant who has lived with systemic lupus erythematosus for over 40 years since initial diagnosis.

This presentation explores her personal journey through initial diagnosis; medications; flares; acute relapses; and treatment errors which resulted in 6 days in a coma, four months

in hospital and profound long term health consequences. It charts a journey to a more effective model of treatment and self-management. This now sees her largely symptom free, on minimal lupus medication (blood pressure tablets) and leading an active personal and professional lifestyle.

Mary draws from her four decade experience to suggest a model of doctor - patient communication, collaboration and partnership that has implications for improving outcomes and quality of life for all lupus patients.

Methods

- Giving the patient a voice. Understanding the powerful psychological benefits with consequential direct and indirect physical benefits for the patient when they are regarded as an "equal partner", an active as opposed to a passive participant in the treatment journey.
- Importance of information sharing with the patient, the treating immunologist and the general practitioner.
- Understanding that the fluctuating and multi-organ nature of lupus symptoms means that treating obvious presenting symptoms without understanding and treating the underlying auto-immune causes can and does lead to treatment errors and adverse patient outcomes.
- Addressing the whole patient, their physical, mental and emotional wellbeing, and the general hormonal system can greatly reduce ongoing symptoms and acute flares.

Results Improved patient outcomes.
Conclusions A more effective model of care.

146 THE ITALIAN SLE SURVEY BY WEB: INVESTIGATING PATIENTS' UNMET NEEDS WITH ONLINE SURVEY TOOLS

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Background and aims Chronical illness perception and difficulties in everyday life due to disease and medications were explored by the first italian ONLINE SLE SURVEY, designed to assess chronic pain impact and health-care provision quality in Italy.
Methods Online questionnaire was created through Qualtrics, setting geographic data, disease duration, age at diagnosis, comorbidity, disability degree, care practices, treatments, subjective incidence and characteristics of pain. SLE patients were advertised by social media. Participation was voluntary and anonymous.
Results 550 SLE patients provided complete data; F 94.7%, M 5.3%; mean age 33 y. (14–82 y.); first SLE diagnosis at mean age 29 y.: 84% received SLE diagnosis between 18–42 y.; 36% comorbidity with other (1-6) autoimmune conditions. SLE impact on life is relevant, with specific problems and needs at different disease stage. Stress of life: relevant and worsening illness conditions; frequent relational problems. Need for psychological support: 54%. Osteoarticular pain is main symptom condition (83%), but only 54% use drugs for pain control. Physicians seem not responding to patients' request to take into account impact of pain. Women workers face many difficulties due to many combined factors which severely reduce access to proper care
Conclusions Health-care current model doesn't allow respect of SLE patients complex needs: most remain dissatisfied, affecting quality of life and doctor-patient concordance. SLE SURVEY highlights importance of competent clinical listening by physicians and capacity to hold patient's crisis. GRUPPO ITALIANO LES – a volunteer patients' organisation - established ONLINE SLE SURVEY practice to explore current facets, development of patients' needs, and set social-health policies

147 A CASE OF DERMATOMYOSITIS WITH ELEVATED SERUM KL-6 LEVEL ASSOCIATED WITH OVARIAN CANCER

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Background and aims Elevated serum Krebs von den Lungen-6 (KL-6) is a biomarker of interstitial pneumonia and some types of cancers. Because patients with dermatomyositis (DM) may accompany interstitial pneumonia and malignancies, serum KL-6 level has been commonly examined for patients with DM in Japan.
Methods A 62-year-old woman developed myalgia in bilateral thighs in November 2014. She was admitted to our hospital in the following month because of progressive bilateral proximal muscle weakness and myalgia, elevated serum creatine kinase (CK: 5569 U/L), and Gottron's papule. Although muscle biopsy had not been done, the diagnosis as DM was further supported by muscle MRI, electromyogram and skin biopsy. The body CT revealed pelvic mass with ascites, but not interstitial pneumonia.
Results Although prednisolone 50 mg/day and intravenous immunoglobulin therapy against progressive muscle weakness including severe dysphagia only showed a partial response, the surgical resection of ovarian mass, identified as ovarian serous adenocarcinoma stage II c, followed by chemotherapy resulted in clinical remission of DM. Serum KL-6 level, as well as CA-125 decreased below the upper normal limit.
Conclusions The present case suggests that the measurement of serum KL-6 may be useful in the evaluation of patients with DM.

148 THE ENGAGEMENT OF SLE PATIENTS IN THEIR HEALTH CARE

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Background and aims Patient engagement is recognised as a crucial component of high-quality healthcare services. Among rheumatic diseases, Systemic Lupus Erythematosus (SLE) appears particularly challenging for the engagement of patients in their own care. According to the Patient Health Engagement (PHE) model, patient engagement is a dynamic phenomenon that proceeds through four experiential positions (blackout, arousal, adhesion, and eudaimonic project). The aim of the present study was to describe the engagement process through the experiences of SLE patients.
Methods Ten in-depth interviews and four fous group were conducted with an international sample of SLE patients from different European countries. Interviews focused on several