Establishing the Jamaica Lupus Registry: Report of Patients with Systemic Lupus Erythematosus Attending a Major Referral Hospital in Jamaica

AK Soyibo, K DeCuelaer, RK Miller, R Smith, K Maloney, EN Barton

ABSTRACT

Background: *Systemic lupus erythematosus (SLE) is an autoimmune disorder characterized by multi-system microvascular inflammation with the generation of autoantibodies. There are reports on demographic data and clinical manifestation of lupus in the United States of America and some other developed countries. There is a single study that has reported on the clinical and immunological features of SLE patients in Jamaica and another that reported that the prevalence of SLE in Jamaica was 5–17/100 000 in 1979.*

Method: *A Jamaican lupus registry was established in 2008 at the Department of Medicine, The University of the West Indies. Data were collected using patient records and interview of patients fulfilling the American College of Rheumatology revised diagnostic criteria for SLE. Information on demographics, presence of diagnostic criteria for SLE, presence of complications and other clinical parameters were collected.*

Results: *There were a total of 107 patients that met the criteria for diagnosis of SLE at the referral centre, 96.3% of them female. Positive ANA (90.7%), arthritis (70.0%), malar rash (53.5%) and a positive dsDNA (40.1%) were the more frequent manifestations and diagnostic indices of the disease. Up to 41.7% of the SLE population suffered some form of complication.*

Conclusions: *The initiation of a lupus registry has allowed for reporting of preliminary demographic, clinical and serological data and identifying of disease burden.*

Keywords: Jamaica, lupus registry, systemic lupus erythematosus

Establecimiento del Registro de Lupus: Reporte de Pacientes con Lupus Sistémico Eritematoso que Asisten a uno de los Principales Hospitales de Remisión en Jamaica

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RESUMEN

Antecedentes: *El lupus sistémico eritematoso (LSE) es un trastorno autoimmune caracterizado por una inflamación microvascular multisistémica con generación de anticuerpos. Hay informes sobre datos demográficos y manifestaciones clínicas de lupus en los Estados Unidos de América y algunos otros países desarrollados. Solamente existen un estudio que ha reportado las características clínicas e inmunológicas de pacientes de LES en Jamaica, y otro que reportó la prevalencia del LES en Jamaica como de 5–17/100 000 en 1979 – un cálculo que ciertamente se queda por debajo.*

Método: *En el año 2008, se estableció un registro jamaicano de lupus. Se recopilaron datos del Departamento de Medicina de la Universidad de West Indies, usando historias clínicas de pacientes del hospital universitario HUWI, y entrevistas de pacientes que cumplían con los criterios diagnósticos de LES revisados del Colegio Americano de Reumatología. Asimismo se recopiló información sobre datos demográficos, presencia de criterios diagnósticos de LES, presencia de complicaciones y otros parámetros clínicos.*

Resultados: *Hubo un total de 107 pacientes que satisfacían los criterios para un diagnóstico de LES en el centro de remisión, 96.3% de ellos mujeres. La prueba ANA positiva (90.7%), artritis (70.0%), salpullido malar (53.5%) y resultado positivo en la prueba de ADN de doble cadena (40.1%) fueron*

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las más frecuentes manifestaciones e índices de diagnóstico de la enfermedad. Hasta un 41.7% de la población de LES sufrió alguna forma de complicación.
Conclusiones: *La iniciación de un registro de lupus ha permitido realizar informes de datos demográficos, determinar característicos preliminares, e identificar el peso de la enfermedad en la población de Jamaica.*

Palabras claves: Jamaica, registro de lupus, lupus eritematoso sistémico

West Indian Med J 2011; 60 (3): 250

INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic inflammatory systemic disease of unknown aetiology. Genetic, racial, hormonal and environmental factors are known to contribute to the development and clinical course of SLE (1–5). Both innate and acquired immunity play a pivotal role in the immune disturbance that occurs in SLE.

The prevalence of SLE varies between countries as well as within a particular country among various ethnic groups (6). There have been studies that have shown that ethnicity and sociodemographic factors may influence the clinical expression and outcomes of SLE (7–9). Systemic lupus erythematosus follows a relapsing and remitting course.

The island of Jamaica is made up of approximately 2.7 million people, with ninety per cent of the population being of African descent. There is only a single study that has reported the clinical and immunological features of SLE patients in Jamaica (10). The prevalence of SLE in Jamaica was reported to be 5–17/100 000 in 1979, but this is almost certainly an underestimation (10).

There are data available on lupus patients in the United States of America (USA), Europe and some other developed countries, and in the Caribbean from Martinique (12) and Jamaica (10, 11). There has been a report on lupus nephritis as a cause of chronic kidney disease (CKD) and end-stage renal disease (ESRD) in the Caribbean (13, 14) especially Jamaica. The report of lupus nephritis as a cause of CKD/ESRD in the Caribbean is thought to be under-reported. In a one-year and, later, three-year review of kidney biopsies performed in Jamaica, lupus nephritis was the commonest secondary glomerular disease in the series (15, 16).

The lupus registry was established in Jamaica in 2008 at the Department of Medicine, The University of the West Indies (UWI), as a result of collaboration between the nephrologists AK Soyibo, R Smith, EN Barton and rheumatologist K DeCuelaer. The lupus registry was established to record the prevalence and incidence of lupus within Jamaica; to record and determine the demographic, clinical and serological characteristics in Jamaican SLE patients. This will assist in determining the severity of the disease and its impact on the public healthcare system and hence enable better planning for improving social and medical services for patients with this disease and its complications. Additionally, the registry will allow for medical personnel to track cases of lupus, easy recall of patients for follow-up and identification of patients for enrolment in clinical studies/research.

The lupus registry focusses on different aspects of the disease, including epidemiology, ethnic diversity, genetic and environmental factors, clinical presentations and outcome. Development of the lupus registry has set the platform for lupus research in Jamaica. It will provide information on definition, causes, symptoms, diagnosing and treating lupus and its complications. It will identify matters concerning lupus and quality of life, pregnancy in women with lupus and current and future research on lupus.

The objective of this report is to give the initial findings of the lupus registry and to define the demographic, clinical and serological features of this cohort of SLE patients. Improving the epidemiological understanding and surveillance of lupus will ultimately enable a deeper scientific understanding of the disease and its impact on the population.

SUBJECTS AND METHODS

Data were collected using patient records and interviews of patients fulfilling the American College of Rheumatology revised diagnostic criteria, *via* questionnaire (Appendix) and later computed in an electronic programme for analysis. There was one rheumatologist in the island based at the University Hospital of the West Indies (UHWI) and hence, the majority of patients were referred to this centre. The questionnaire contained information on demographic data including age, gender, ethnicity, presence of diagnostic criteria for SLE (clinical and laboratory pre-sence of autoantibodies), presence of complications such as nephritis, cerebritis and pneumonitis, and other information important to public health surveillance. Data were collected on treatment options, drug effectiveness and reported side effects.

All information is treated confidentially and a committee deliberates on the release of data.

RESULTS

There were a total of 117 patients referred for suspected diagnosis of SLE, 107 patients met four of the criteria for diagnosis of SLE according to the American College of Rheumatology revised criteria (Fig. 1). The other ten fulfilled two or three diagnostic criteria. A significant propor-

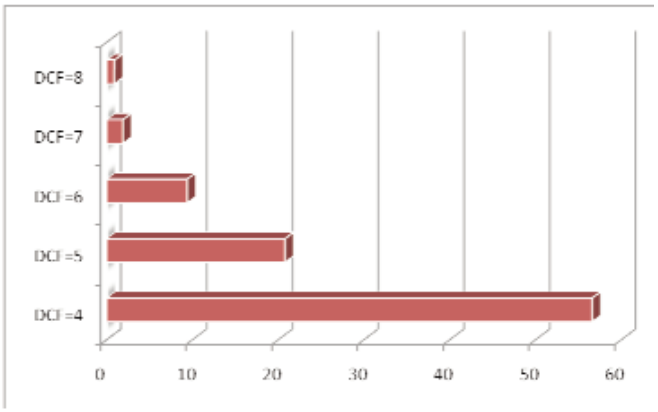

Fig. 1: Percentage of patients having four or more diagnostic criteria fulfilled for systemic lupus erythematosus.

tion of the population comprised females at 96.3% (Table). The demographic distribution of patients showed that the majority of the referrals to the UHWI was from the neighbouring parishes of St Catherine (34.6%) and St Andrew (26.2%), to the metropolitan city of Kingston where the centre is located (Fig. 2).

Evaluation of the patients' highest educational achievement showed that 55.8% had secondary education, while 16.3% achieved tertiary education. More than half (50.9%) had current employment with 25.5% and 11.3% currently unemployed and unemployed due to disease, respectively (Table).

The most prevalent clinical diagnostic criteria were arthritis (71.8%) and malar rash (52.0%) while positive ANA and dsDNA were the most prevalent serological markers, 91.0% and 38.7%, respectively. The characteristic and demographic distributions are outlined in the Table and Fig. 2, respectively.

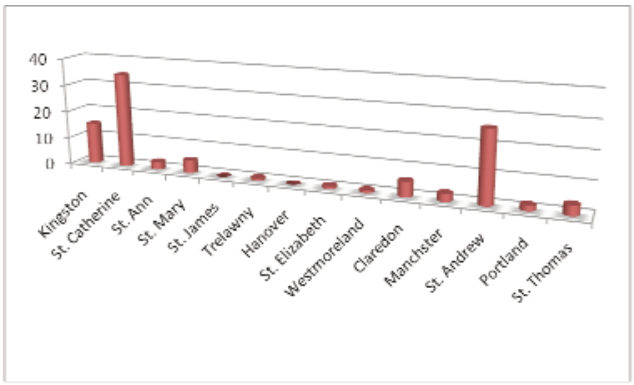

Fig. 2: Demographic distribution of systemic lupus erythematosus patients attending the referral centre.

DISCUSSION

This preliminary report from the lupus registry in Jamaica shows the distribution of lupus patients, determination of incidence and prevalence, control of disease population, and also allows for greater understanding of the disease. The public burden of the disease and implications for healthcare planning could be better understood.

The mean age of the population with SLE was 42.9 years (SD 13.1). However, this does not reflect the mean age at diagnosis of SLE in the Jamaican population. Such data will be obtained for future reference as the registry develops. The demographic distribution of patients showed that the majority of the referrals to the centre at the UHWI were from neighbouring parishes of St Catherine (34.6%), St Andrew (26.2%) and Clarendon (5.6%), to the metropolitan city of Kingston (15.0%) where the centre is located.

The prevalence of employment was 51.5% compared to 58.8% for the general population as reported in the Jamaica Health and Lifestyle Survey (JHLS) 2007–08 (17).

Table: Characteristic distribution, prevalence of diagnostic criteria and complications in SLE patients

Parameter (n = 107)	Percentage (%)
Gender	
Male	1.9
Female	96.3
Highest Education Achieved (n = 106)	
Primary education	6.7
Secondary education	55.8
Tertiary education	16.3
Not available	21.2
Employment Status	
Currently employed	50.9
Currently unemployed	25.5
Never employed	7.5
Unemployed due to illness	11.3
Retired	3.8
Diagnostic Index (present)	
Malar rash	52.0
Discoid rash	29.0
Photosensitivity	22.2
Oral ulcer	17.2
Arthritis	71.8
Serositis	27.0
Proteinuria (> 0.5 g/24 h)	39.0
Positive ANA	91.0
dsDNA positive	38.7
False-positive VDRL	8.8
Anti-Sm (Smith) positive	17.2
Blood dyscrasia	20.2
Seizures or psychosis	17.2
Any Complications	42.3
Patient Disease Perception	
Does not understand	6.5
Poor understanding	11.7
Fair understanding	55.8
Very good understanding	26.0
Family support	
Yes	97.4
No	2.6

There is a significant difference between the prevalence of unemployment between males and females (18.8% *vs* 43.3%) in the JHLS with a combined total unemployment rate of 31.3%. In the present report, the unemployment rate was 43.7%. Considering the fact that the study population is predominantly female, this figure is comparable to the female unemployment rate in the JHLS report. Employment status may affect the pattern of health-seeking behaviour and the associated ability to procure medication.

In a study evaluating the factors influencing medication adherence in Jamaican patients with SLE for more than a year (11), high cost and availability of medication were the main reasons for poor adherence. Other reasons included drug side effects and perception of the presence of mild disease. In the present report, when evaluating the presence of health insurance, which will help to subsidize medication cost, 54.5% had no insurance regardless of employment status. Evaluating the percentage of persons with current employment, it was revealed that 54.7% had secondary education while 22.6% had tertiary education. A higher level of formal education may positively affect the understanding of the disease and allow for better healthcare-seeking behaviour and improvement in general outcome.

Systemic lupus erythematosus may present with many different symptoms depending on the organ system(s) affected. The most common are arthritis, fatigue, skin rashes, photosensitivity and fever. In this report, arthritis (71.8%) and skin manifestation of malar rash (52.0%) and discoid rash (29.0%) were the most common clinical presentation. There was no report of fever but photosensitivity was reported in 22.2% of patients.

In a study of eighty-two Jamaican patients diagnosed with SLE (18), the prevalence of ANA was 100% while dsDNA was 49%. From the registry, ANA was positive in 91.0% and dsDNA in 38.7%. Haemolytic anaemia, thrombocytopenia or leucopenia was present in 26.6% of the SLE population; 47.1% had anaemia with 5.9% having pancytopenia.

Systemic lupus erythematosus can affect every organ system of the body, leading to complications. Complications were present in 42.3% of the SLE population. Approximately 65.5% of patients were started on prednisone at 20 mg or less at the time of presentation to the hospital. Other drugs, at presentation, were hydroxychloroquine, azathioprine and angiotensin converting enzyme inhibitor (ACEI). The most common drug side effects reported were gastrointestinal (GI) in origin.

Of the 37.1% with proteinuria above 0.5 g/day or with active urinary sediments, percutaneous kidney biopsy was performed in approximately half. Membranous lupus nephritis (class 5) was the most common histologied type (36.8%) seen in the biopsy. There is greater correlation with positive ANA and presence of renal disease either from urinary examination or biopsy proven lupus nephritis (Fig. 4).

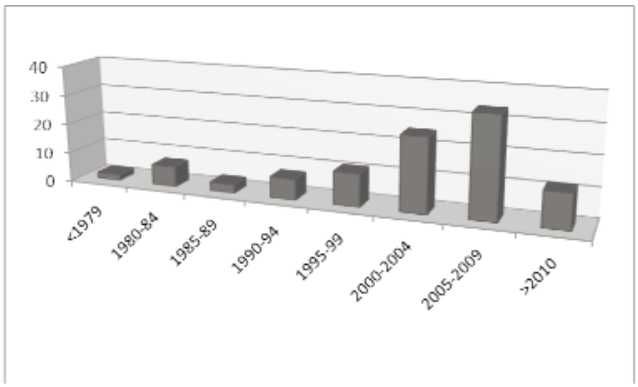

Fig. 3: Per cent distribution of patients fulfilling the diagnostic criteria for systemic lupus erythematosus over a 20-year period.

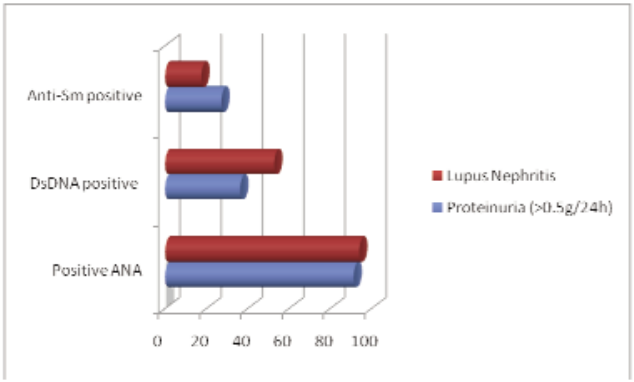

Fig. 4: Correlation of systemic lupus erythematosus diagnostic marker and presence of renal disease.

Complications of SLE as reported from the patient records were 42.3%. Frequently reported complications were: lupus nephritis (29.8%), central nervous system (CNS) involvement (15.4%) and pneumonitis (5.4%). There was a documented case of pancreatitis with bad outcome and three cases with positive lupus anticoagulant.

Lupus nephritis is a complication of SLE with increased morbidity and mortality. The histological type on kidney biopsy also affects the development of renal failure

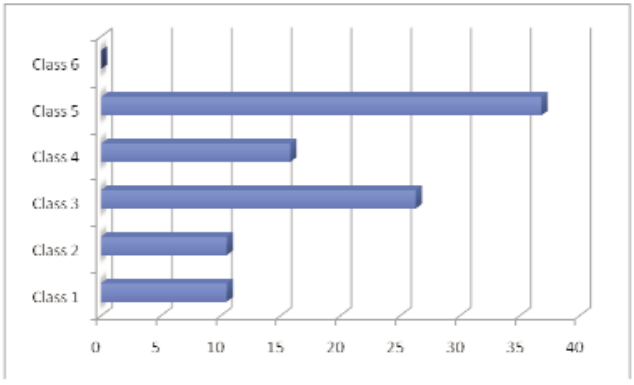

Fig. 5: Pattern of lupus nephritis on biopsy.

and ESRD (19). Based on histological diagnosis of lupus nephritis, class 4 was the most common histological type in a series by Williams *et al* (19). However, in a three-year review of renal biopsy performed in Jamaica (16), class 5 (membranous lupus nephritis) was the most prevalent. In this registry report, class 5 was also more prevalent (Fig. 5).

CONCLUSION

The initiation of a lupus registry has allowed for identifying disease burden and demographic and clinical parameters in the Jamaican population.

ACKNOWLEDGEMENTS

Special thanks to Heather Phillips (research nurse) and Anthony Myers (research assistant), Department of Medicine, The University of the West Indies, Kingston, Jamaica.

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APPENDIX

JAMAICA LUPUS REGISTRY QUESTIONNAIRE

ID number
DOB Gender
Country Parish
Highest level of education attained
Religion
Health insurance status
Employment status
Job description
Year diagnosed with SLE
Blood pressure at time of diagnosis
Diagnostic criteria (should have four out of eleven)
- Malar rash
- Discoid lupus
- Photosensitivity
- Oral ulcers
- Arthritis
- Proteinuria (> 0.5 g/day) or cellular cast in urine microscopy
- Seizure or psychosis
- Serositis
- Haemolytic anaemia or leucopenia or lymphopenia or thrombocytopenia
- Positive ANA
- Antibody to DNA or Sm or presence of LE cells or biologically false-positive serology to syphilis (VDRL)

Complication related to SLE
- Nephritis
- Cerebritis/psychosis
- Lupus anticoagulant or history of thromboembolism
- Arterial thrombosis
- Venous thrombosis
- Pancreatitis
- Pneumonitis

Prednisone dose at time of presentation
Current drugs use
Drug side effects
Urinary protein quantifications
Kidney biopsy result
Patient disease perception
Family/friend disease perception
Family support