



## BACKGROUND

SLE is a severe autoimmune disease that can present in childhood or adult life. Studies in children vary depending on the source of the study with incident rates of 0.22, 0.4, 0.47, 0.8 and 0.9 per 100 000 per year in The Czech Republic [1], Southern Sweden [2], Japan [3], United Kingdom (Midlands) [4] and Finland [5] respectively. Many studies have found a higher incidence or prevalence in those of African or Asian background [6] including the UK study [4] in children < 17 years which found that the incident rate in Asians was 5.6, Blacks 3.1 and Whites 0.4 per 100 000 per year. There are no data on incidence of SLE in Australia but it has been reported that there is a high prevalence of the disease in Australians of indigenous background (1: 1360) [7]. The magnitude of the problem of a severe disease such as SLE is not known in Australia and may well be considerable given the increase in the number of people of Asian background in Australia as well as the known increased prevalence in the indigenous population. Obtaining accurate data on our incidence and closely examining the ethnicity of the patients may indicate whether environmental factors in Australia play a role in the development of the disease. UV radiation has long been known to precipitate systemic and cutaneous lupus [8].

We are entering a new era in the management of lupus with many biological therapies being used/developed to treat SLE in a more targeted approach to control the inflammatory response [9]. Agents specifically targeting B cells (rituximab) and T/B cell interactions (abatacept) in particular show considerable promise. It still needs to be determined which type of patients will benefit from such therapies. This study, by determining the nature and severity of presentation of SLE children in Australia may allow some estimation of how many patients may benefit and require such biological therapies. The follow-up questionnaire may provide a clinical snapshot of current practice and how many have received these various types of therapies in the first year of diagnosis.

There is a paucity of information about the demographics and severity of presentation of SLE in children in Australia and this study will provide the first national dataset on SLE presentation, diagnosis and treatment.

The case definition for paediatric lupus is based upon the agreed international criteria for classification for SLE [10] Appendix. However we have also expanded our definition to a practical one where fewer manifestations are acceptable (with a positive ANA) as it is acknowledged that it may take time for these to develop. These patients will be analysed subsequently as 'likely' cases. By also adding patients with a tissue diagnosis of SLE we allow inclusion of patients recently diagnosed but yet to develop the standard criteria. Given a pathological diagnosis we plan to analyse these as 'definite' cases.

## STUDY OBJECTIVES

To estimate the incidence, geographical distribution and ethnic background of SLE in Australian children aged ≤15 years. This study will also describe the mode and severity of clinical presentation as well as 1 year outcome of SLE in children in Australia. The clinical specialty of the initial treating physician as well as types of medications and any medication related adverse outcomes used in the management of paediatric SLE in the first year after diagnosis will be described.

## CASE DEFINITION

Any child ≤ 15 years of age who meets any of the 3 definitions below:

1. Any child ≤ 15 years of age fulfilling the clinical diagnostic criteria for SLE ie. presenting with 4 or more of the 11 listed and defined in the table, namely:

1. Malar rash	2. Discoid rash	3. Photosensitivity
4. Oral ulcers	5. Arthritis	6. Serositis
7. Renal disorder	8. Neurological disorder	9. Haematological disorder
10. Immunological disorder	11. Antinuclear antibody	

**OR**

2. Any child ≤ 15 years of age who presents with 1 or more of the above clinical features **AND** a positive antinuclear antibody >1:320.

**OR**

3. Any child ≤ 15 years of age who presents with a tissue diagnosis of SLE renal biopsy diagnostic of SLE or skin biopsy consistent with SLE.

## REPORTING INSTRUCTIONS:

Please report ALL cases of children ≤ 15 years of age diagnosed with SLE in the last month. Please report all children in whom you suspect SLE even if results of tests are pending.

## FOLLOW-UP OF REPORTED CASES

A questionnaire requesting further details will be forwarded to practitioners who report a case.  
A follow-up questionnaire will be forwarded 12 months later.

**Table. Criteria for diagnosis of Systemic Lupus Erythema (SLE)**

Criterion	Definition
<b>1. Malar rash</b>	Fixed erythema, flat or raised, over the malar eminences, tending to spare the nasolabial folds
<b>2. Discoid rash</b>	Erythematous raised patches with adherent keratotic scaling and follicular plugging; atrophic scarring may occur in older lesions
<b>3. Photosensitivity</b>	Skin rash as a result of unusual reaction to sunlight, by patient history or physician observation
<b>4. Oral ulcers</b>	Oral or nasopharyngeal ulceration usually painless observed by a physician
<b>5. Arthritis</b>	Non-erosive arthritis involving 2 or more peripheral joints, characterised by tenderness, swelling or effusion
<b>6. Serositis</b>	a. Pleuritis-convincing history of pleuritic pain or rub heard by a physician or evidence of pleural effusion <b>OR</b> b. Pericarditis-documented by ECG or rub or evidence of pericardial effusion
<b>7. Renal disorder</b>	a. Persistent proteinuria greater than 0.5 grams per day or greater than 3+ if quantitation not performed <b>OR</b> b. Cellular casts- may be red cell, haemoglobin, granular, tubular or mixed
<b>8. Neurological disorder</b>	a. Seizures - in the absence of offending drugs or known metabolic derangements eg, uremia, ketoacidosis or electrolyte imbalance <b>OR</b> b. Psychosis - in the absence of offending drugs or known metabolic derangements eg, uremia, ketoacidosis or electrolyte imbalance
<b>9. Haematologic disorder</b>	a. Haemolytic anaemia – with reticulocytosis <b>OR</b> b. Leukopenia- less than 4000/mm <sup>3</sup> total on 2 or more occasions <b>OR</b> c. Lymphopenia- less than 1500/ mm <sup>3</sup> on 2 or more occasions <b>OR</b> d. Thrombocytopenia- less than 100 000/ mm <sup>3</sup> in the absence of offending drugs
<b>10. Immunologic disorder</b>	a. Anti-DNA: antibody to native DNA in abnormal titer <b>OR</b> b. Anti-Sm: presence of antibody to Sm nuclear antigen <b>OR</b> c. Positive finding of antiphospholipid antibody based on: 1. An abnormal serum level of IgG or IgM cardiolipin Antibody 2. Positive test result for lupus anticoagulant using a standard method or 3. False positive serologic test for syphilis known to be positive for at least 6 months and confirmed by Treponema pallidum immobilization or fluorescent treponemal antibody absorption test
<b>11. Antinuclear antibody</b>	An abnormal titer of antinuclear antibody by immunofluorescence or an equivalent assay at any point in time and in the absence of drugs known to be associated with “drug-induced lupus” syndrome

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