BRITISH PAEDIATRIC SURVEILLANCE UNIT
Royal College of Paediatrics and Child Health
5-11 Theobalds Road, London WC1 X 8SH
Tel: 020 7092 6173 Email: bpsu@rcpch.ac.uk Web: www.rcpch.ac.uk/bpsu/lupus

JUVENILE-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS (JSLE)

Abstract

JSLE or 'childhood lupus' is a rare multisystem illness where the immune system attacks many parts of the body. JSLE can be very variable in how it manifests, with some children and young people having a mild disease and others having a very severe disease (e.g. developing kidney failure or brain abnormalities). If it starts in childhood or adolescence it is generally much more severe, and requires more use of steroids and other potent medications which suppress the immune system (immune-suppressant medications) than in adults.[1]

Although we say around 15-20% of cases start in childhood[1], there is a great lack of strong studies examining the actual number of cases of JSLE in the UK and Republic of Ireland. This means that the overall burden of illness caused by JSLE is unknown.

This study aims to determine the incidence of JSLE in children and young people in the UK and Ireland. The data will be analysed to better understand the epidemiology and clinical features of the disease and how children and young people are accessing care.

Principal Investigators

Dr Hanna Lythgoe
Institute In The Park, Alder Hey Children's NHS Foundation Trust,
Eaton Road, Liverpool, L12 2AP
Email: bpsulupus@alderhey.nhs.uk **Tel:** (+44) (0)151 293 3566

Co-investigators

Prof M.W. Beresford, Dr Eve Smith, Dr Clare E. Pain, Alder Hey Children's NHS Foundation Trust, Liverpool, UK
Dr Orla Killeen, Our Lady's Children's Hospital, Crumlin, Ireland
Dr Clarissa Pilkington, Great Ormond Street Hospital, London
Dr Ruth Murphy, British Society for Paediatric Dermatology (BSPD)

Website

http://www.rcpch.ac.uk/bpsu/lupus

Background

How these children and young people present, are diagnosed and access initial and on-going care is not well described. New European guidelines (SHARE guidelines)[2] are due to be published which emphasise that patients with suspected JSLE should be referred to a paediatric rheumatologist and supported by a multi-disciplinary team including access to other paediatric specialists. Through experience of our current UK-wide collaboration (the UK JSLE Study Group), we do not believe that this European standard is currently being met throughout the UK. This study will establish the magnitude of this problem for UK patients, and where they are currently being managed.

The very varied presentation of JSLE means it is difficult to develop good criteria to define whether we can classify a child or young person as having JSLE or not. We know that the existing ACR classification criteria do not accurately identify all children and young people with JSLE. Some new classification criteria (SLICC classification criteria) have been suggested[3]. This study will help us to look at how well both these classification systems can classify children and young people with JSLE.

Coverage

United Kingdom and Republic of Ireland

Duration

September 2017 to September 2019 (25 months of surveillance) with a follow-up at 12 months.

Research Questions

- What is the incidence of JSLE in children and young people in the UK and ROI?
- What are the demographics of children/young people affected by JSLE (age, sex, ethnicity, geographical distribution)?
- What are the presenting features of JSLE in the UK and ROI?
- What is the delay between children and young people developing symptoms to receiving a diagnosis of JSLE?
- What damage is caused by JSLE by the time of diagnosis and what disease damage is present one year after diagnosis?
- Which clinicians look after children and young people with JSLE?
- How are children and young people with JSLE treated?
- What proportion of children and young people with probable JSLE (who do not initially meet existing classification criteria) go on to meet these criteria within one year of follow-up?
- What proportion of children meet newer SLICC classification criteria at the point of clinician diagnosis and after one year of follow-up?

Case definition

Any child / young person aged up to 18 years of age who:

1. Has a new, consultant diagnosis of suspected JSLE

AND

2. Fulfils 2 or more ACR criteria **AND / OR** has lupus nephritis on biopsy

AND

3. Has no alternative diagnosis for relevant disease features

Reporting instructions

Please report any child / young person aged up to 18 years of age who meets the case definition.

Methods

Each paediatrician reporting a child who meets the above case definition of JSLE will be sent a clinical questionnaire by the study team. A second questionnaire will be sent 12 months later to collect follow-up data.

Throughout the study, all patient data will be dealt with in strict confidence, and affected children and their families will not be contacted directly by the JSLE study team at any stage.

Funding

This study is being funded through the BPSU Sir Peter Tizard Bursary (http://www.rcpch.ac.uk/bpsu/bursary) and Lupus UK (http://www.lupusuk.org.uk)

Ethics approval

This study has been approved by North West Liverpool East REC (REC reference: 17/NW/0095) and has been granted Section 251 HRA-CAG permission (CAG Reference: 17/CAG/0075).

This study has been granted Public Benefit and Privacy Panel for Health and Social Care (PBPP) approval in Scotland (PBPP reference: 1516-0292).

Support group

Lupus UK (http://www.lupusuk.org.uk)

References

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For further information about the study, please contact:

Carla Roberts, Alder Hey Children's NHS Foundation Trust, 1st Floor, Institute in the Park, Eaton Road, Liverpool, L12 2AP
Email: bpsulupus@alderhey.nhs.uk **Tel:** +44 (0)151 293 3566