INCIDENCE AND PREVALENCE OF BEHÇET’S SYNDROME IN THE UK AND ROI
(Short Study Name: Behçet’s Syndrome)

Abstract
Behçet’s syndrome is a rare multi-system inflammatory condition characterised by recurrent oral ulceration, genital ulceration, eye and skin involvement. There is currently very little data on the incidence and prevalence of Behçet’s syndrome in children within the UK population. The purpose of this study is to establish the current UK and ROI incidence and prevalence of Behçet’s syndrome and to describe the burden of disease in children and young people under 16 years of age.

Because of the perceived rareness of the condition, the investigators plan to investigate both incidence AND prevalence.

Principal Investigator
Dr Clare Pain
Alder Hey Children’s NHS Foundation Trust
Eaton Road, Liverpool L12 2AP
Tel: 0151 282 4521 E-mail: clare.pain@alderhey.nhs.uk

Co-investigators
Prof Robert Moots, Aintree University Hospital, Longmoor Lane, Liverpool L9 7AL
Prof Michael Beresford, University of Liverpool, Crown Street, Liverpool L69 3BX
Prof Fárida Fortune, Queen Mary’s School of Medicine and Dentistry, London E1 4NS
Dr Paul Brogan, Great Ormond Street Hospital, London WC1N 3JH
Dr Ruth Murphy, Queen’s Medical Centre, Nottingham, NG7 2UH
Dr Clodagh Lowry, Children’s University Hospital, Dublin
Mr Chris Phillips, Behçet’s Syndrome Society, Worcester WR11 4SP

Website
http://www.rcpch.ac.uk/bpsu/BHT

Background
The prevalence of Behçet’s syndrome in the UK is actually unknown but estimates put it at 0.64 per 100,000 of the population [1]. Studies have shown that 4-26% of Behçet’s syndrome occurs in those under 16 years of age [2]. An international cohort study in children has suggested that children have less skin and eye involvement than adults [3].

Recent National Commissioning has led to the development of three National Centre’s of Excellence for Behçet’s syndrome. However, the disease burden of Behçet’s in patients under 16 years of age in the UK and ROI and how and by whom these patients are managed is not well described.

A 13-month study of the incidence and prevalence of paediatric Behçet’s syndrome will examine the current UK experience. By answering these questions, the study will provide vital data that will be of value in defining the need for additional supra-regional referral services and in designing future clinical trials. We aim to perform a one year follow-up study which would identify early disease progression and complications of Behçet’s syndrome in children.

Coverage
United Kingdom and the Republic of Ireland

Duration

Research Questions
- What is the incidence of Behçet’s syndrome in childhood?
- What is the prevalence of Behçet’s syndrome in childhood?
- What are the usual presenting symptoms of Behçet’s syndrome in children within the UK?
- What is the delay between symptom onset and formal diagnosis?
- What are the clinical features of Behçet’s syndrome in UK children?
- What is the pattern of care received by the affected children?
- What are the patterns of children affected by Behçet’s according to age, sex and ethnicity?
- What is the outcome of Behçet’s syndrome in children identified in initial study
- What is the outcome of Behçet’s syndrome in children followed over one year e.g. how many
children have new manifestations, damage or escalation of treatment?

**Case definition**
Children up to but not including the age of 16 who have 2 of more of the following features not explained by an alternative diagnosis:

- Oral aphthous ulceration
- Skin involvement defined as erythema nodosum, pustulosis, folliculitis or acneiform lesions
- Positive pathergy test (skin prick test)
- Eye involvement (defined as uveitis and/or retinal vasculitis)
- Genital ulceration
- Family history of Behçet’s syndrome in a biological parent or sibling
- Vascular involvement: arterial or venous thrombosis, thrombophlebitis and/or aneurysm
- Neurological involvement suggestive of Behçet’s syndrome

**Exclusion criteria**
Any child with above features due to an alternative confirmed diagnosis *e.g.* inflammatory bowel disease, coeliac disease, systemic lupus erythematosus, infection.

**Reporting instructions**
Please report any patients seen in the last month with possible Behçet’s syndrome (including new and follow-up cases) as defined in the case definition.

**Methods**
This surveillance study will be performed together with the British Paediatric Surveillance Unit (BPSU) and the British Society of Paediatric Dermatologists.

Members of the British Society of Paediatric Dermatologists will also receive monthly email notification forms. Some dermatologists may see children with Behçet's syndrome without a paediatrician being involved in the child’s care. To ensure we have a more complete picture of children affected with Behçet’s syndrome we feel it is important to include paediatric dermatologists in this study.

Both Paediatricians and Dermatologists that report a possible case will be sent a questionnaire requesting demographic, clinical information regarding disease manifestations and management.

Based on this information an expert panel (investigators named on this application) will decided whether the child has definite or probable Behçet’s syndrome based on pre-defined criteria.

One year later, all clinicians whom have reported a confirmed case of Behçet’s syndrome will be asked to complete a follow-up questionnaire.

The study team will analyse the information provided and produce a report of the results which we aim to share with doctors involved in the study (BPSU report), the wider medical community (via publishing report in a medical journal) and to patients and families of those affected with Behçet’s syndrome (via lay report on Behçet’s Syndrome Society website and newsletter).

**Funding**
Behcet’s Syndrome Society (from funds raised by the Worshipful Company of Horner’s), Alder Hey Children’s charity and the study investigators.

**Support Group**
The Behcet’s Syndrome Society - [http://behcets.org.uk/](http://behcets.org.uk/)

**Ethics approval**
This study has been approved by NRES Committee – North West – Liverpool East (REC reference: 15/NW/0035; IRAS project ID: 163430) and has been granted Section 251 HRA-CAG permission (CAG Reference: 15/CAG/0103).

**References**

For further information about the study, please contact:
Dr Clare Pain, Department of Rheumatology, Alder Hey Children’s NHS Foundation Trust, Eaton Road, Liverpool L12 2AP
Email: behcet@alderhey.nhs.uk

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