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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.

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Website

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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

May 2015 – May 2017 (25 months surveillance). Follow-up until May 2018 (12 month follow-up.

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/

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

1. Meeting report of the 13th International Conference on Behçet"s Disease in the Journal of Rheumatology 36:6;doi:10.3899/jrheum.081008.
2. Zouboulis, C.C., et al., Epidemiological features of Adamantiades-Behcet's disease in Germany and in Europe. Yonsei Med J, 1997. 38(6): p. 411-22.
3. Kone-Paut, I., et al., Registries in rheumatological and musculoskeletal conditions. Paediatric Behcet's disease: an international cohort study of 110 patients. One-year follow-up data. Rheumatology (Oxford), 2011. 50(1): p. 184-8.

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