

## BRITISH PAEDIATRIC SURVEILLANCE UNIT

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# Surveillance of ALL Kawasaki Disease (Incomplete and Complete) in the UK and Ireland

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<b>Abstract</b>	<p>Kawasaki disease is the commonest cause of acquired heart disease in children in the UK and USA. The serious sequelae of Kawasaki disease make it important to diagnose this condition early in order to treat it effectively and therefore minimise complications and long term ill health within the Paediatric population. The last BPSU survey of KD was in 1990 and since that time there has been increased awareness of the condition and treatment protocols.</p> <p><b>NOTE: this is now a new study taking in ALL cases of Kawasaki disease, incomplete (including atypical and partial) and complete forms. We no longer exclude streptococcal infection in the criteria.</b></p>
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<b>Website</b>	<p><a href="http://www.rcpch.ac.uk/bpsu/KD">www.rcpch.ac.uk/bpsu/KD</a></p>
<b>Background</b>	<p>The incidence of Kawasaki disease varies from 184 per 100,000 children under 5 years in Japan to about 8-9 per 100,000 in England. The high rates in North East Asians, which persists after migration to countries with low incidence, is strong evidence for a genetic factor, but the aetiology of Kawasaki disease is essentially unknown. Seasonal variation, with peaks in winter and spring, and reported epidemics have suggested that there are environmental factors, for example viral infections, which trigger the condition. The recent epidemiological study in the UK was limited to England and did not have access to the ethnicity of individual cases. Moreover this study which used Hospital Episode Statistics had no means of assessing the accuracy of the diagnosis and had to be limited to children under five year of age to avoid counting re-admissions in older children. Although 76% of children with Kawasaki disease are found to be under the age of five years, Kawasaki disease occurs in children over the age of 5 so the incidence, presentation and course of the disease in older children in Western Europe will be of scientific interest. The diagnosis is not straightforward, which has an impact on management, and it is important to assess how UK guidelines have affected treatment and outcomes.</p>
<b>Coverage</b>	<p>United Kingdom and the Republic of Ireland</p>
<b>Duration</b>	<p>February 2014 – February 2015 (13 months of surveillance).</p>
<b>Research Questions</b>	<p><b><u>Incidence:</u></b></p> <ul style="list-style-type: none"><li>• What are the demographics (sex, age, ethnicity, area of residence (partial postcode), history of sibling or parent with the disease) of those diagnosed with Kawasaki disease in the UK and Ireland under 16 years old?</li><li>• What is the incidence of Kawasaki disease in different parts of the UK and Ireland?</li><li>• Has the incidence changed since the last survey in 1990?</li><li>• Is there evidence of change in practise following the introduction of new guidelines in 2013?</li><li>• Are there more cases of incomplete Kawasaki disease being recognised?</li></ul> <p><b><u>Clinical Presentation:</u></b></p> <ul style="list-style-type: none"><li>• A description of the first presentation of Kawasaki disease and of the interval between presentation and diagnosis? This information will be available within the normal good history taking on presentation to the paediatric services. The data would be as recorded in the paediatric medical notes.</li><li>• What is the prevalence of early (within the initial echocardiograms) cardiac complications within 30 days following Kawasaki disease in the UK and Ireland and what are those complications? <sup>3</sup></li></ul>

### **Clinical Management:**

- What acute treatment is being given to patients during their initial hospital presentation with Kawasaki disease in the UK and Ireland?
- Are treatments other than aspirin and intravenous immunoglobulin being used, and if so, what is the outcome in patients with Kawasaki disease as measured in the acute setting?
- How is the introduction of steroids into the new guideline being utilised?

### **Outcome:**

- What is the prevalence of non-cardiac complications within 30 days following Kawasaki disease in the UK and Ireland?
- How are patients with diagnosed Kawasaki disease being followed up within the UK and Ireland?

### **Case Definition**

**Any infant or child up to the age of 16 years presenting for the first time with Fever of 5 or more days duration *plus* 4 of the following (complete) or *plus* any 2 of the following and coronary artery changes (atypical) or *plus* 2 or 3 of the following (incomplete):**

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|----------------------------------|---|
| <b>1. Conjunctivitis</b>         | Bilateral, bulbar, non-suppurative  |
| <b>2. Lymphadenopathy</b>        | Cervical > 1.5cm  |
| <b>3. Rash</b>                   | Widespread, polymorphous. <i>Not</i> vesicular.                                     |
| <b>4. Lips and mucosa</b>        | Red cracked lips, 'strawberry tongue', erythematous oral cavity                     |
| <b>5. Changes of extremities</b> | Erythema, oedema of palms and soles initially, then peeling of skin at later stage. |

#### **Exclusion:**

None. (No longer are we excluding streptococcal infections)

### **Methods**

#### *Surveillance*

Active national surveillance of all children who fulfil the case definition will be undertaken through the BPSU. Consultant Paediatricians, Consultant Paediatric Cardiologists and Paediatricians with a special interest in paediatric cardiology will be part of this surveillance.

#### *Questionnaires*

We will send questionnaires to collect demographic and clinical data, including NHS/CHI number, Hospital Number, sex, date of birth, ethnicity and clinical features. We wish to know treatments and outcomes within 30 days

### **Ethics approval**

This study has been approved by NRES Committee South West – (REC reference 11/SW/0310) and has been granted Section 251 permission under reference: ECC 6-02 (FT11)/2012

### **Funding**

Kawasaki disease parent support group and investigators  
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### **References**

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