Sponsors: Swiss Society of Paediatrics (SSP) and Federal Office of Public Health (FOPH)

KAWASAKI DISEASE IN CHILDREN

Studied disease:

Epidemiology of Kawasaki disease in children in Switzerland

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Protocol summary:

Kawasaki disease (KD) is the most frequent acquired heart disease in children in developed countries (1,2,3). It is an acute febrile illness with potentially serious sequelae as 25 % of children with Kawasaki disease will develop coronary artery lesions (CAL) if untreated (3,4,5). The use of single - dose intravenous immunoglobulins associated with Aspirine decreases this incidence significantly (6). The objectives of this study are to examine the epidemiology, clinical features, management and short and medium term outcome of children with Kawasaki disease in Switzerland.

Begin of the study:

March 2013

Expected cases per year in Switzerland (extrapolated from the data at our institution):

75 - 100 cases

Duration of the study:

Recruitment: March 2013 – March 2014
1 year follow-up: March 2014 – March 2015
5 year follow-up: March 2017 – March 2020

Aims of the study:

1.) Determine the epidemiology (incidence, age, sex, family history) of pediatric KD in Switzerland
2.) Describe the clinical presentation (complete, incomplete), the time between presentation and diagnosis of KD in children
3.) Establish the prevalence of cardiac involvement at the time of diagnosis
4.) Assess the type and duration of treatment (Immunoglobulins, Aspirine)
5.) Determine the follow-up in Switzerland for children with KD
6.) Evaluate the short term and medium- term (1 and 5 years) outcome of children after acute KD in Switzerland.

Background:

KD was first described in 1967 by Tomikasu Kawasaki. It is an acute febrile illness of unknown etiology, occurring mostly in children under 5 years of age (3-5,7). It has many characteristics of an infection mediated vasculitis or an autoimmune disorder (7,8,9). The diagnosis is based on internationally agreed clinical criteria (cf case definition) which are not specific of the disease (3-5,7). The lack of specific and sensitive diagnostic tests makes the diagnosis sometimes difficult and incomplete cases are frequent thus delaying treatment (4,8). If untreated 20-30% of patients with KD will develop CAL with potential long-term sequelae such as thrombosis and myocardial infarction (4, 8, 9). Cardiac complications can be decreased by treatment with intravenous immunoglobulin administration and oral Aspirine within 10 days of the onset of the disease (6, 10).
The incidence of KD varies depending on ethnicity, the highest incidence being in Japan and Asia (11-17). The incidence of KD is increasing globally especially in Asian countries (15,16). Genetic factors most certainly play a role in the epidemiology of the disease as illustrated by high rates of KD in the Asian population having migrated to countries with low incidence (13, 18).
The incidence of KD in Switzerland is unknown.

Methodology:

Epidemiological and clinical prospective study.
Data will be collected anonymously in all pediatric clinics of Switzerland on a voluntary basis.
It will include children until 16 years + 364 days of age.
It will give comprehensive information regarding the incidence and clinical presentation of KD in Switzerland, the time between first symptoms and diagnosis, the incidence of coronary aneurysms and the outcome at 1 year and 5 years after treatment. It may help to understand the etiology of the disease.
The study is proposed to the SPSU for their epidemiological surveillance scheme which covers all pediatric hospitals in Switzerland. As most likely all patients with KD in Switzerland are treated on an in-patient basis, the recruitment should be complete.
The benefit for treating physicians will be to have an increased knowledge of the situation in Switzerland and may help improve management of these patients. It will provide better information for parents and children.

Case definition:

Inclusion criteria:

Any infant or child up to the age of 16 years + 364 days for which the diagnosis “Kawasaki disease” was made by the treating physician, whether it is typical or atypical.

Complete Kawasaki will be defined as:

- Fever > 38.5 for 5 and more days

Plus 4 of the following symptoms:

- Conjunctivitis	bilateral, bulbar, non suppurative
- Lymphadenopathy	at least one, cervical, > 1.5 cm, non purulent
- Rash	polymorphous exanthema
- Lips and mucosa	red cracked lips, strawberry tongue, erythematous oral and pharyngeal mucosa
- Changes of extremities	initial stage: erythema, edema of palms and soles convalescent stage: desquamation of skin

Incomplete Kawasaki will be defined as:

- Fever of > 38.5 for 5 or more days
- And less than 4 of the above described symptoms
- And positive echocardiographic findings

Questionnaire:

See addendum

Reporting instruction:

Please report all new cases of Kawasaki disease satisfying case definition (typical and atypical) which have been diagnosed in the last month.
A follow-up questionnaire will be sent to the child’s pediatrician 1 and 5 years later.

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