### NATIONAL STUDY OF KAWASAKI DISEASE

### Background

Kawasaki disease (KD) is a disorder of acute onset and unknown aetiology but with many characteristics of an infection mediated vasculitis affecting mainly infants and young children. Severe cardiac manifestations, including coronary artery aneurysm formation and obliterative arteritis occur in up to 25% of untreated cases; myocardial infarction and death are recognised outcomes. It has been shown that both cardiac morbidity and mortality can be significantly reduced by the administration of intravenous gammaglobulin (IVIG) and aspirin within 10 days of the onset of the disease.

The incidence of KD varies within geographically and racially defined populations from 3.7 cases annually per 100,000 children aged under 5 years in Australia to 95 cases per 100,000 in Japan. There are no prospectively obtained data from New Zealand however in the Auckland hospitals 4.97 cases per 100,000 were recorded between 1979 and 1988. World wide mortality rates also vary between 0.08% in Japan to 3.7% in the UK. One of the highest recorded mortality rates was the 6% noted in the Auckland study. It is unclear whether mortality reflects intrinsic severity of disease and/or early recognition and delivery of effective treatment.

KD is a clinical diagnoses, there are many recorded manifestations and no diagnostic features exclusive to the disorder. There is also a considerable differential diagnosis. However, in most instances the diagnosis can be made fairly easily once the possibility of KD is considered.

## Objectives

- 1. To determine the incidence of KD in New Zealand.
- 2. To describe the epidemiology and clinical features of KD in New Zealand.
- 3. To describe the clinical management and outcome of KD in New Zealand.

### **CASE DEFINITION**

# (Based on Rowley and Shulman, Pediatric Clinics of North America, April 1999)

Fever persisting 5 days or more (in the presence of other diagnostic criteria the diagnosis may be made before the fifth day of fever)

### AND at least 4 of the following:

- 1. Changes of peripheral extremities:
  - (a) Initial stage: Reddening of palms and soles, indurative oedema.
  - (b) Convalescent stage: Membranous desquamation from fingertips.
- 2. Polymorphous exanthema
- 3. Bilateral conjunctival congestion
- 4. Changes of lips and oral cavity: Reddening of lips, strawberry tongue, diffuse injection of oral and pharyngeal mucosa
- 5 Acute non-purulent cervical lymphadenopathy.

#### <u>AND</u>

Disease not explained by other disease process (in particular streptococcal infection or measles)

Patients with fever and 3 other criteria may be diagnosed as Kawasaki disease if coronary abnormalities develop

### **Reporting Instructions**

Please report all new cases satisfying case definitions who have been diagnosed in the last month.

## Follow up of Positive Returns

A questionnaire requesting further details will be sent to the reporting doctor. A follow-up questionnaire will be sent 12 months later.

## **Principal Investigators**

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## **Any Questions**

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THANK YOU FOR YOUR HELP WITH THIS STUDY