

*societá*

*The UK  
Kawasaki  
Disease  
Foundation*

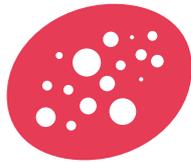




**Persistent fever**



**Cracked lips/tongue**



**Rash**



**Swollen fingers/toes**



**Bloodshot eyes**



**Swollen glands**

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## *Kawasaki Disease is the leading cause of acquired coronary artery disease in children in the UK*

### Why Societì?

**Kawasaki Disease affects hundreds of children in the UK each year and its effects can be devastating. It is the number one cause of acquired coronary artery disease in children in the UK.**

Societì Foundation was established in 2015 as an organisation with a specific focus to raise awareness of Kawasaki Disease. We also want to enable both urgently needed research and better support to families affected by Kawasaki Disease.

Not enough is known about Kawasaki Disease, its symptoms or its treatment. Support for families is limited. Many doctors and parents are unaware of the disease. We want to change this.

Perceived once as a rare illness, this is an outdated notion which we will work to change. This perception hampers research into Kawasaki Disease which is stifled by lack of funds. Finding the cause of Kawasaki Disease, developing better acute treatments and understanding the long term care needs for Kawasaki Disease are key priorities.

### Our Aims

Societì has four aims:

#### 1. Awareness raising

We will raise awareness of Kawasaki Disease to improve prompt diagnosis – through a dynamic multi media campaign, reaching parents, GPs, paediatric clinicians and health professionals. This is our primary aim.

#### 2. Clinical research

We will identify funds and lever efforts building on enhanced awareness of need, to undertake and urgently advance critical research to understand, diagnose and manage Kawasaki Disease

#### 3. Clinical supervision

Gaps exist in current provision for Kawasaki Disease children in their long term care. We will work to close these gaps, as well as connect long term care to long term research.

#### 4. Support for families

We recognise this as the lifeline when you're at the centre of the "flat spin" that is a Kawasaki Disease diagnosis – so securing resources to support groups which support families is our fourth important aim.

**Kawasaki Disease Support Group**

We are proud to be working in partnership with the Kawasaki Disease UK support group. For those who have been directly affected by Kawasaki Disease, the support group offer guidance and advice and a phone and email helpline for families and parents of children with Kawasaki Disease.

**About Kawasaki Disease**

Identified in 1967 in Japan by Tomisaku Kawasaki, the cause of Kawasaki Disease is still unknown and the illness – as Kawasaki Disease presents with some symptoms mirroring a range of other childhood diseases – is often misdiagnosed. Kawasaki Disease is a vascular disease which if untreated can cause coronary artery damage. If treated early however, the vast majority of children recover completely.

Kawasaki Disease has a range of symptoms including a persistent high fever for five days or more, rash, bloodshot eyes, swollen lymph nodes, irritability, “strawberry” tongue, cracked, dry lips and redness of the fingers and toes (sometimes skin peeling after ten days or more). Kawasaki Disease can be present with some (partial) or all (full) of these symptoms.

## UK Strategy

### Overview

**Kawasaki Disease is the leading cause of acquired coronary artery disease in children in the UK.** The most recent figures available evidence a dramatically increasing incidence of Kawasaki Disease, increasing threefold in a 25 year period to a currently recorded incidence of 9.1/100,000 children under five per annum. This pace of increase is of further concern when combined with indications of both under-diagnosis, and mis-diagnosis. No information exists which extrapolates these indications to achieve a comprehensive picture of the number of children in the UK and Ireland affected by Kawasaki Disease, but the current national survey records 300 children per year. Informed estimates suggest actual cases could exceed ten-fold this recorded number.

The national survey shows that of those diagnosed in the UK, 28% of children have coronary artery involvement - a rate much higher than children diagnosed in the US or Japan (where coronary involvement affects just 4% of treated children). This might reflect that only the most severe cases are diagnosed and these are those more likely to subsequently experience serious complications, including damage to the heart.

The detection rate in other countries is much higher than that in the UK. In Japan, 250 children per 100,000 are today being diagnosed with Kawasaki Disease. Assuming a constant rate of increase in the UK, as has been evidenced over the previous 25 year period, and by others countries in the world, by 2050 1 child in every 1000 in the UK will be affected by Kawasaki Disease.

Without immediate action to improve **diagnosis**, undertake urgently needed **research** and provide **long term care** to affected children, this represents the single **most significant coronary health risk to be faced by the UK today**. Moreover, the long term implications for those who have experienced Kawasaki Disease with adverse coronary involvement is leading to **an ever increasing population with a requirement for long term cardiac care**.

**What is known:**

There are many gaps in our knowledge in the UK, and globally, which exacerbate attempts to manage Kawasaki Disease proactively.

An early intervention window exists for treating Kawasaki Disease – and if diagnosis is made and treatment given during this initial window (ten days from onset) coronary complications should be avoided in 95% of cases. This early diagnosis and treatment window is the cornerstone of the rationale for a step change in awareness levels and diagnosis of Kawasaki Disease -- life long and life threatening coronary complications might be prevented.

What is also known is that there is a genetic predisposition which favours presentation of the disease. Acute treatments have been partially advanced using this knowledge.

The size of the UK Kawasaki Disease population as noted above, although recorded, represents significant under-diagnosis.

Ongoing and recently concluded research programmes in the UK include examination of a population of young adults presenting with coronary complications, now considered to have previously experienced Kawasaki Disease (Prof Tulloh, Bristol Heart Institute); and research investigating the long term coronary sequelae of Kawasaki Disease including those patients whose coronary involvement had apparently resolved (Prof Brogan, Great Ormond Street). These studies point strongly to the need for both ongoing clinical surveillance and consideration of ongoing preventative treatment.

Other ongoing research through the Kawasaki Disease Research Centre at the University of California School of Medicine, USA, present opportunities for collaborative research programmes and international partnerships, which Prof Levin (Imperial College London) is actively engaging within to build the knowledge base around Kawasaki Disease.

## *The key questions which remain around Kawasaki Disease*

**What is needed:**

Unlike many medical conditions of significance, no UK strategy is yet in place for improving the outcome of Kawasaki Disease; so an opportunity exists to collaboratively develop a national strategy. A strategy would allow the articulation of issues, a focus on key outcomes sought, assignment of responsibilities for investment and actions, next steps and the clear identification of the resources required to enable the outcomes.

The strategy will address the key questions which remain around Kawasaki Disease, including:

- how to routinely accurately diagnose and manage Kawasaki Disease
- why Kawasaki Disease disproportionately affects some groups over others
- what the trigger mechanism is for Kawasaki Disease
- linked to trigger identification, what prevention mechanisms can be put in place
- how to improve and standardise acute treatment
- what long term treatment provisions are needed, informing ongoing research and learning
- what is the strategy for the management of the previously undiagnosed young adult population
- how can we better resource the management and development of critical statistical datasets for Kawasaki Disease

## *Raising awareness and improving diagnosis of Kawasaki Disease*

### **The solutions:**

This strategy can explore a range of solutions aimed at creating a proactive environment around Kawasaki Disease; one which:

- places significant emphasis on awareness raising, improving prompt, accurate diagnosis
- is powerfully enabling for clinicians delivering life-saving and preventative research outcomes
- is world leading in clinical and pharmaceutical innovation
- is exceptional in its focus on enhancing levels of care for families experiencing Kawasaki Disease
- is proportionate in scale, recognising the significance of Kawasaki Disease and increasing incidence annually and cumulatively

Focused resources dedicated to addressing Kawasaki Disease and building on the centres of Kawasaki Disease excellence which exist in the UK will be essential to enable the step change needed to respond. Centres of excellence exist in Bristol (cardiology), Manchester (GOSH – vasculitis) and London (ICL – infectious diseases).

Providing enabling and targeted investment to these centres to support stronger, resourced collaborations around a portfolio of planned and systematic (rather than reactive) actions to address the range of Kawasaki Disease gaps and challenges will allow demonstrable progress. Practical illustrations of targeted

investment would include funded professorial posts, research posts and support staff, research budgets of competent scale, support for international collaborations and development of beneficial partnerships.

Within the medical arena, much greater levels of awareness are needed – amongst GPs, Paediatricians, A&E staff and all front line staff including those with responsibility for patient referrals (e.g. NHS Direct).

Additionally, resources need to be committed to raise awareness beyond professional staff, to the public at large – known to be unaware of both the existence of Kawasaki Disease and the very significant immediate and long term health threats it poses to children and young adults.

A strategic master plan for Kawasaki Disease with horizons at 1,2,5 and 10 years needs to be developed, building out from the strategy, which describes targets and outcomes, into which stakeholders and partners can mutually invest.

Only by working and acting together can we overcome the immediate threat that Kawasaki Disease presents.

## Global partnerships to collectively address Kawasaki Disease

### The opportunities:

- Cost effective to act now – An acute phase treatment window exists in Kawasaki Disease, which if capitalised on, can significantly reduce the likelihood of coronary complications (up to 95%). Whilst complex and costly measures are sought to tackle a range of causes of coronary artery disease, from lifetime obesity to poor diet across the UK, the coronary complications from Kawasaki Disease can be largely prevented with prompt diagnosis and treatment.

Whilst Kawasaki Disease presents a current and growing risk of scale to the UK's coronary health, early intervention preventative measures are possible and can be put in place cost effectively.

- Raising the profile of our nation's coronary health – almost uniquely affecting children and young adults, Kawasaki Disease presents a formidable opportunity to put a spotlight on both the critical issues of Kawasaki Disease and the imperative of a central focus on investing in preventative initiatives relating to coronary health.
- World leading health innovation – Kawasaki Disease is an international challenge with professionals across the globe seeking solutions to the growing threat of Kawasaki Disease. The UK, if it chooses, could mobilise, lead and participate in global partnerships to collectively address Kawasaki Disease.

### Immediate next steps:

#### 1 Assessing stakeholder appetite

As a new, small organisation, Societì must work in partnership with other key stakeholders already dedicated to improving the nation's coronary health, and child health – if the significant impact which is urgently needed is to be achieved. Understanding the appetite from partners to collaborate in addressing Kawasaki Disease is therefore a critical first step.

#### 2 Creating a partnership

As no national partnership exists to raise the profile of and target resources towards Kawasaki Disease, Societì has been created as an enabler and can lead a partnership effectively and strongly.

#### 3 Resourcing development of the UK strategy

Securing partner and stakeholder engagement into a definitive UK Strategy for Kawasaki Disease, embracing international partnerships will enable mutual ownership of the core principles within the strategy; thereafter continued engagement and investment. Initially, a commitment will be needed from partners to collectively resource strategy development.

# Kawasaki Disease Symptoms

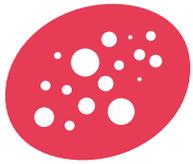
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