Kawasaki Disease

What is Kawasaki Disease?

Kawasaki Disease KD is the second most common paediatric vasculitis and it is the leading cause of acquired heart disease in children in the UK.

Identified in 1967 in Japan by Tomisaku Kawasaki, the cause of Kawasaki Disease is still unknown. The illness presents with several symptoms common to a variety of other childhood diseases and infections and is therefore often misdiagnosed, particularly as scarlet fever. Kawasaki Disease is a serious disease which if untreated can cause coronary artery damage. This complication is preventable with a timely diagnosis and appropriate treatment.

What are the symptoms of Kawasaki Disease?

Kawasaki Disease mainly affects young children under the age of 5 years but it can affect people of any age. It can be a serious illness as it can cause coronary artery damage – damage to the blood vessels in the heart. If treated early however, the majority of children recover well. Kawasaki Disease has a range of symptoms including:

- a characteristic and distinctively persistent high fever for five days or more
- rash
- bloodshot eyes
- “strawberry” tongue – and cracked, dry lips
- redness of the fingers and toes (sometimes skin peeling after ten days or more) and
- swollen glands in the neck – often only on one side

Kawasaki Disease can be present with some (partial) or all (full) of these symptoms. In particular, young children under the age of 12 months may only have a few of these features and yet still be at risk of coronary artery injury.

Children affected by Kawasaki Disease have much improved chances of a good
recovery with timely diagnosis and the correct treatment.

Who is affected?

At onset, Kawasaki Disease primarily affects young children with over 75% of those affected being under 5 – but it can affect any age group. And because the damage it can cause can be lifelong, thousands of children, young people and adults are affected in the UK today by Kawasaki Disease.

What is the aetiology (cause)?

There are lots of theories around the cause of Kawasaki Disease and much research ongoing across the globe to try and identify its cause. But at the moment, no-one is certain of the cause.

There is some evidence to suggest that some children have a genetic predisposition to being affected by Kawasaki Disease. Some researchers believe it could be a child’s response to an infection or a number of infections though no infectious cause has been found. There are theories too that suggest an environmental agent – perhaps something which is airborne or related to water bodies. Whilst there is much research ongoing, some of it appears contradictory – and the patterns of incidence of Kawasaki Disease (who gets it and where) are different in different places.

What we do know is that Kawasaki Disease is on the rise – possibly due to better diagnosis rates – and identifying the cause will be a major milestone in the fight against Kawasaki Disease.

Making a diagnosis

As yet there is no specific test to help doctors diagnose Kawasaki Disease. A number of tests may be performed by doctors as they work to establish a diagnosis for your child and these could include:

- blood tests – looking for signs of inflammation
- temperature checks – as Kawasaki Disease has a distinctively high and persistent fever
- an electrocardiogram (ECG) which allows doctors to check the rhythm of the heart
- a heart scan using ultrasound (echocardiogram or cardiac echo) from which doctors can check whether a child’s heart is being/has been affected by Kawasaki Disease

Doctors also look for the specific symptoms of Kawasaki Disease – and they may ask if patients (or parents) have noticed symptoms over the last few days – as Kawasaki Disease symptoms can appear one after the other over a number of days sometimes, not always all at once.

Treatment

Children with Kawasaki Disease are treated in hospital by specialist doctors. The
medication given (intravenous immunoglobulin (IVIG) and aspirin – often also with corticosteroids) is used with the aim of reducing fever and preventing heart damage including coronary artery aneurysms. Sometimes other drugs are used as well, in support of these three main treatment approaches. Which children will benefit most from corticosteroids (such as prednisolone) remains an area of controversy, so a major trial of this is being organised in the UK (KD-CAAP, in set up at the time of writing in Feb 2018): so don’t be surprised if you are asked to join in with this.

Some children may develop long-term coronary artery problems which require long-term care under a cardiologist.

**Drugs and Side effects**

For information on the main drugs prescribed for Kawasaki Disease see:

- **Immunoglobulin**

For information on other drugs used in the treatment of vasculitis see [Glossary of drugs and side effects](#).

More information on drugs used in Kawasaki Disease is available on the Societi Foundation website at [www.societi.org.uk](http://www.societi.org.uk) – within our Family Portal.

**Prognosis**

Outcomes for children in the UK are of concern, with 28% of treated children developing heart damage and 24% develop coronary artery aneurysms. Early diagnosis and early treatment are shown to reduce the likelihood of heart damage – but as Kawasaki Disease is not well-known, diagnosis is often delayed and treatments given may not be optimal – these are factors which are understood to be contributing to the poor outcomes we are seeing in patients in the UK.

**Key Points**

- Kawasaki Disease is increasingly common in the UK, currently affecting 4.5 per 100,000 children under 5 years of age.
- Early diagnosis and early treatment is key to improving outcomes including reducing the risk of heart damage
- 25% of patients affected are over 5 years of age – this is not just a disease of the very young
- Damage to the coronary arteries is the commonest serious problem of Kawasaki Disease

**Related Vasculitis Articles**

- [Vasculitis in Children](#) - Prof Paul Brogan
- [Fertility and Vasculitis](#) - Prof David Jayne
Further reading

- **Vasculitis Paediatric Guidelines 2012** - Dr Helen Foster & Dr Paul Brogan, Oxford University Press
- **Management of Kawasaki Disease, 2013** - Dr Paul Brogan, et al
- **Pathogenesis and management of Kawasaki disease** - Anne H Rowley, Stanford T Shulman
- **Medium-size Vessel Diseases** - Michael J Dillon, Despina Eletheriou, Paul A Brogan

Useful links

The UK Foundation for Kawasaki Disease – Societi – has a comprehensive website including a Family Portal with useful information for families who have been affected by Kawasaki Disease. [www.societi.org.uk](http://www.societi.org.uk)

The Kawasaki Support Group – a parent support group -UK have their [own website](http://www.societi.org.uk)

Our “Useful Vasculitis Links” page contains contact details for organisations offering help and support for patients with Kawasaki Disease and other vasculitis diseases.

**Kawasaki Disease experiences on camera** on the Societi Family Resource Portal