Kawasaki disease: information for families

Kawasaki disease is a disease that causes swelling of the blood vessels throughout the body. It causes a high fever and rash. It can also affect the blood vessels supplying the heart muscle (coronary arteries). This information sheet from Great Ormond Street Hospital (GOSH) explains the causes and symptoms of Kawasaki disease and how it can be treated.

Kawasaki disease is rare in the UK and only affects around eight in every 100,000 children under the age of five. It is much more common in Japan.

What causes Kawasaki disease?

Doctors do not yet know exactly what causes Kawasaki disease. It is probably due to a combination of factors.

Some children may be genetically more likely to develop the condition (predisposed) but no genetic change has been identified yet.

There may also be environmental factors such as infections and the way a child's body responds to that infection, could play a part too.

Kawasaki disease is not contagious or catching – you cannot pass it on to another person. No specific bug (bacterium or virus) has been identified. Links to coronavirus (COVID-19) have not been proven.

Doctors do know that children under the age of five are most often affected, and it is more common in boys than girls. It is more common in children of Japanese or Oriental background, but can affect anyone of any ethnicity.

What are the symptoms of Kawasaki disease?

The symptoms of Kawasaki disease are like a lot of other more common childhood illnesses. The symptoms can come and go over a period of days and weeks, with new ones appearing. Initial signs can include:

- feeling unwell
- fever (high temperature) over 38°C for five days or longer
- swollen lymph nodes (glands in the neck)
- rash (especially on the chest)
- palms of the hands and soles of the feet turn bright red, become puffy and the skin can peel
- bloodshot eyes
- redness on the inside of the lips and tongue and inside the mouth.

Some children can have painful joints, tummy ache, diarrhoea and vomiting as well.

For some children, the heart can be affected too. Around 20 to 40 per cent of children with Kawasaki disease develop problems in the blood vessels in the heart muscle. A bulge can develop in the wall of a heart blood vessel (coronary aneurysm). This can make it more likely for a
blood clot to develop inside the vessel or it may become narrower as it heals. The valves inside the heart can also become leaky, so blood can flow backwards (regurgitation). The bag or sac that surrounds the heart can also fill up with fluid, putting pressure on the heart (pericardial effusion). These heart problems can lead to symptoms like breathlessness and chest pain.

The risk of developing heart problems is lessened with prompt diagnosis and treatment. If heart problems have not developed within a few weeks of diagnosis, they are unlikely to occur.

Children can also have Kawasaki disease without showing all the symptoms above – this is often referred to as atypical Kawasaki disease. This is more common in younger children.

**How is Kawasaki disease diagnosed?**

In most cases, doctors can diagnose Kawasaki disease through the distinctive symptoms.

Children usually need a number of tests to find out whether the condition is affecting other organs of the body, in particular the heart. This will usually include an electrocardiogram (ECG) and echocardiogram (Echo). They will usually have some blood tests to rule out any other causes of the symptoms.

**How is Kawasaki disease treated?**

A child with Kawasaki disease is usually treated in hospital with high doses of immunoglobulin (IVIG) given directly into a vein through an infusion (drip) into a cannula; some children need steroids (such as prednisolone) as well. These treatments can reduce blood vessel inflammation and also treat the high temperature. As with most treatments, the earlier started, the less likely long term effects will occur.

Children may also be given aspirin liquid to swallow, which reduces the chance of blood clots developing. Even though children should not usually take aspirin due to the risk of Reye’s syndrome, the benefits of taking it outweigh the risk of developing other problems.

Some children may need treatment with corticosteroids as well as other medications, given intravenously or by mouth.

Most children start to feel better a few days after starting treatment. Children can get better without treatment but there is a higher risk of developing long term problems with the heart.

**What is the outlook for children and young people with Kawasaki disease?**

Children who do not have any heart problems usually recover fully and are back to normal within weeks.

For children whose coronary blood vessels have been affected, long term follow up with the heart specialists will be needed. If they continue to have heart problems, they may need to take blood thinning medicine longer term to stop clots forming in the blood vessels.

If a blood clot develops in a coronary artery, it could block blood flow (and therefore oxygen supply) to the heart muscle, so the muscle dies. They will need regular check-ups, including ECG and Echo tests to make sure their heart is working well. They may also need an exercise test or cardiac catheterisation to monitor the coronary arteries.

A second attack of the disease is rare.
Further information and support

The Kawasaki Support Group offers advice and support to anyone affected by condition. Call them on 02476 612178 or visit their website at www.kssg.org.uk

Societi – The UK Kawasaki Disease Foundation also offers advice and support. Visit their website at www.societi.org.uk/

All about Kawasaki disease

- Kawasaki disease is rare disease. It was first seen in Japan.
- It usually affects children under 5 years old.
- If you have Kawasaki disease, you will have a high temperature.
- The glands in your neck might be swollen.
- You might have a rash on your chest.
- Your hands and feet might look red and puffy. The skin might start to peel.
- The inside of your mouth and eyelids might look red.
- Sometimes, the blood vessels on your heart get swollen and sore.
- This means that your heart doesn’t get as much oxygen as it should.
- This could make breathing harder and your chest sore.
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<tr>
<th>The doctors will give you a check up to see if you have Kawasaki disease.</th>
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<tr>
<td>You might have a scan to look at your heart.</td>
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<td>You will usually have to stay in hospital so you can have medicine through a drip.</td>
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<td>You may also have medicine to swallow too.</td>
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<td>Most people start to feel better when they have treatment.</td>
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<td>You will need to have regular check-ups with the doctor after you have had Kawasaki disease.</td>
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<td>Please ask us if you have any questions.</td>
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