Follow up

During the acute illness, the heart should be examined at 2 and 6 weeks by echocardiography, and more regularly if an issue is seen. This monitoring informs initial treatment and helps determine the risk of future heart complications. Measurement of the blood vessels, patient height, age and weight together with comparison to a normal artery provide doctors with a 'z-score'. This score helps 'grade' the coronary artery aneurysm (CAA) and will inform treatment choices. If the score is high after 6 weeks, the patient is considered to have long-term heart damage. Z-scores which indicate "giant" coronary artery aneurysms (8mm+) are less likely to reduce in size and can mean a 50% chance of serious heart complications within 30 years.

Recommended follow up summary

Risk	Coronary artery involvement	Follow up	Imaging required	Specialist ongoing Kawasaki Disease care?
1	No involvement at any time (Z score<2)	2 weeks, 6 weeks, 6 mths, 12 mths. Discharge if normal at 12 mths	None	No - yearly heart and general health review with GP
2	Widening (dilation) only (2≤Z score<2.5), resolves within 1 year	2 weeks, 6 weeks, 6 mths, 12 mths. Discharge if normal at 12 mths	None	No - yearly heart and general health review with GP
3	Small CAA (2.5≤Z score<5) (even if resolved)	2 weeks, 6 weeks, 6 mths, 12 mths then yearly	Image heart muscle blood flow at 12 mths. Image heart annually and consider stress testing every 2 yrs.	Yes
4	Medium size CAA (5≤Z score<10) (even if resolved)	2 weeks, 6 weeks, 6 mths, 12 mths then yearly	Image heart muscle blood flow at 12 mths. Image heart annually and consider stress testing every 2 yrs.	Yes
5	Giant CAA in blood vessel (Z score≥10 or ≥8 mm) (even if reduced or resolved)	2 weeks, 6 weeks, 3 mths, 6 mths, 9 mths, 12 mths then every 6 mths	Image heart muscle blood flow at 12 mths. Image to look for clots 6 monthly. Consider yearly stress test.	Yes

About Kawasaki Disease

Kawasaki Disease is a vasculitis - a disease causing inflammation (swelling) of the blood vessels, including those that provide the heart muscle with its blood supply. It mostly affects children and cases are on the rise. Without rapid treatment it causes heart damage in 28% of all children affected, with 19% of children having lasting damage. It is the leading cause of acquired heart disease in children, in the UK.

The treatment for Kawasaki Disease is immunoglobulin given in hospital by a drip. Aspirin is also given. Treatment aims to stop the inflammation and reduce the risk of heart damage. **However, 19% of children overall and 39% of those under 1 year will have lasting heart problems,** often because they are not diagnosed and treated quickly. A delay in treatment is directly linked to increased risk of heart damage.

Children may develop abnormal swellings (aneurysms) in the blood vessels to the heart, the coronary arteries, which means there is a risk of complications. These include blood clots in the artery or narrowing of the artery later on, so blood cannot flow properly. Both can lead to a dangerous reduction in blood flow and oxygen supply to the heart, which could result in a major cardiac event.

The information in this leaflet has been directed by the Societi Scientific Advisory Board. It is intended for educational purposes only and does not constitute medical advice. For guidance, please access the full version of Lifetime cardiovascular management of patients with previous Kawasaki Disease here:

https://heart.bmj.com/content/106/6/411.full?fbclid=IwAR3aQENPy9eiIz7GNpb04ryC-eiaNbCerndqEAwXaYoKIweEPbt3KnlPTQw

Randal Charitable Foundation

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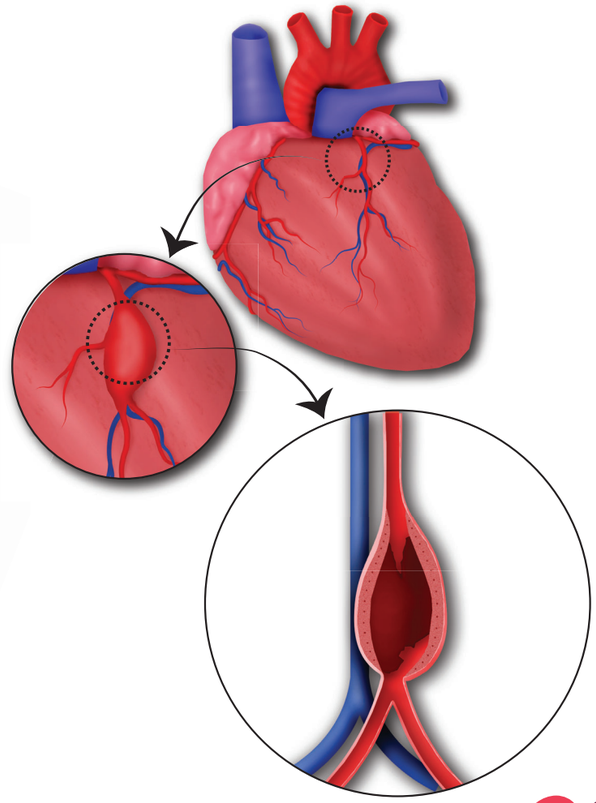
Societi is a registered charity in England & Wales (no.1173755)

societi.org.uk

societi *We are the UK Foundation for* **Kawasaki Disease**

Lifetime management of patients with heart damage due to **Kawasaki Disease**

A quick guide



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Medicines

Patients should be managed with the aims of preventing blood clots forming and the early detection of any blood clot, or narrowing of the blood vessel.

Anti-platelet medicines and anticoagulant medicines (medicines which work in different ways to prevent blood clots forming) are used. Aspirin is an anti-platelet medicine and is given to all patients with Kawasaki Disease initially. Patients who have swelling of the blood vessels at 6 weeks will stay on aspirin longer term. Patients with giant aneurysms also need an anticoagulant.

Person-specific protocol

All patients at risk levels 3, 4 and 5 need a person-specific protocol (PSP) with their Kawasaki Disease history. This should be held by the patient, parents, school (for a child), the Congenital Cardiac Surgical Centre (children) or Heart Attack Centre (adults) and their emergency medical services. This is so they can act quickly if needed.

The PSP should also include instructions to the local Ambulance Service on where the patient should be taken, and phone numbers (24 hours) for a Kawasaki Disease specialist.

A person-specific protocol template is available to download at heart.bmj.com

...access here!

Moving from child to adult services

All patients at risk level 3 or above will need to move to adult cardiology services at 16-18 years of age. To start with, a joint clinic with doctors from both child and adult services is recommended.

The adult service should be a specialist Kawasaki Disease clinic, or service led by a clinician knowledgeable in Kawasaki Disease with access to interventional cardiology. This is where diagnosis and treatment of heart and blood vessel conditions is carried out, often using small tubes (catheters). There should also be a heart CT or MRI available at all times.

All children moving to adult services should have heart imaging and tests to look for reduction of blood flow which currently has no symptoms, and tests to look at problems with valves of the heart, hardening or scarring of the heart tissue. A test to look at calcium levels in the blood vessels may also be helpful as an indicator of possible narrowing of the vessels.

mysocieti.org.uk *is a youth portal which includes information for young patients who are transitioning to adult care.*

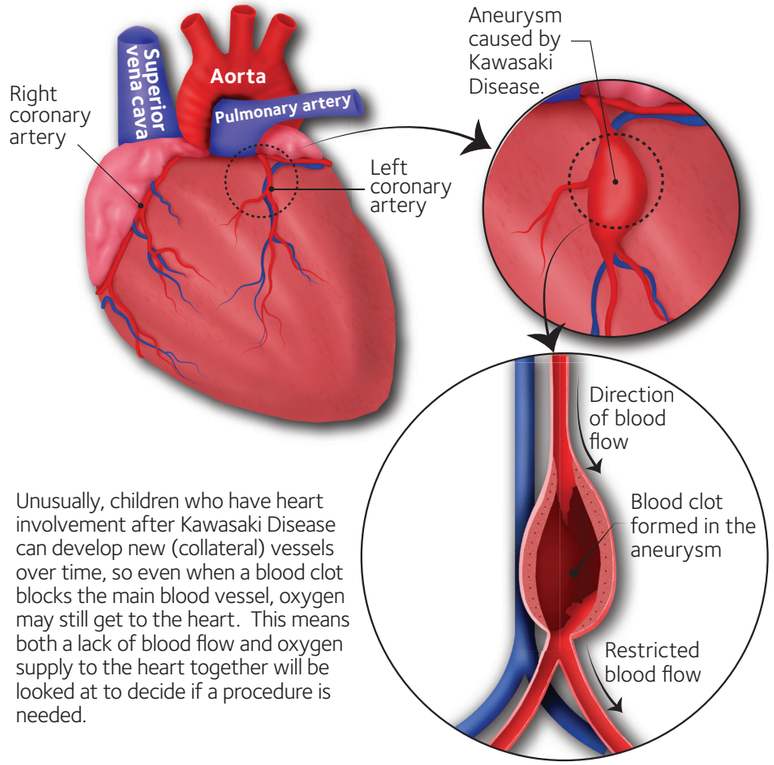
...access here!

Emergency care

Blood flow to the heart can be reduced at ANY age in patients with coronary artery aneurysms. Urgent tests should be carried out where doctors think oxygen flow to the heart is reduced as:

1. Typical presentation may NOT happen in children. Symptoms can include unexplained crying, being restless, unusual colour, sweating, or pain (not felt in one area)
2. Children and young adults may be better able to deal with the lack of oxygen so may not have typical symptoms, despite a large clot.
3. Initial tests (ECG and troponin) may not suggest a reduction in oxygen, even when blood supply is greatly reduced, due to the development of collateral vessels.
4. Blood clots and swellings in the blood vessels can occur even when taking medicines to prevent clots.

Coronary artery aneurysm



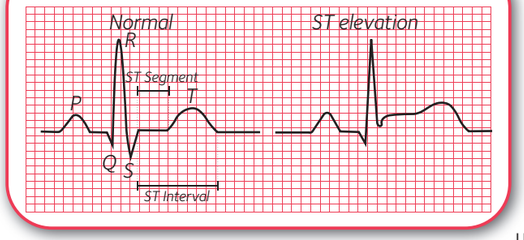
Unusually, children who have heart involvement after Kawasaki Disease can develop new (collateral) vessels over time, so even when a blood clot blocks the main blood vessel, oxygen may still get to the heart. This means both a lack of blood flow and oxygen supply to the heart together will be looked at to decide if a procedure is needed.

Initial emergency presentation

Unless symptoms are clearly due to a non-heart condition, such as trauma, patients with coronary artery aneurysms or past changes in the blood vessels should always be seen at a Heart Attack Centre (adults) or Congenital Cardiac Surgical Centre (children).

A high degree of suspicion must be maintained in a patient with a history of Kawasaki Disease.

ST elevation



ST elevation requires urgent contact with a heat specialist (interventionalist) who can carry out treatment to restore blood flow to the heart. Imaging should be undertaken where there is no obvious heart attack but reduction in blood flow is suspected. If ST elevation is seen or there is a sudden loss of blood flow and it will take more than 90 minutes to get to the specialist centre then medication to treat the clot should be given before the patient travels to the centre.

Emergency management

Where there is a blood clot, but blood can still flow, medicines to prevent clotting should be considered and medicines to reduce clots may need to be increased. In more than half of patients with coronary artery aneurysms, daily medication to break down the clot is effective. Child size will be part of the decision as to whether medication or a surgical intervention is used first.

Where a large blood clot is preventing blood flow, the preferred option is to insert a tube, through which a small balloon is inflated at the site of the blockage, widening the blocked blood vessel. The balloon is then deflated and removed. Removal of the clot is usually not needed but might be necessary to reopen the blood vessel.

For further information about lifetime cardiovascular management in patients with previous Kawasaki Disease, access the full guidance below:

To access the full version of the Lifetime cardiovascular management of patients with previous **Kawasaki Disease** *guidance*

...access here!

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