

Person Specific Protocol (suggested template for local/regional implementation)

KAWASAKI DISEASE PERSON SPECIFIC PROTOCOL

For all Kawasaki Disease patients with any prior history of coronary artery aneurysms (ongoing or remodelled coronary artery aneurysms)

Introduction:

This protocol has been specifically prepared for the patient named below and details the treatment to be given in specified circumstances. **Complete ALL sections highlighted YELLOW**

Name: [Patient name]
DOB: [Patient DOB]
NHS Number: [Patient NHS No]
Address: [Patient Address]

Reason for Protocol:

This patient previously had acute Kawasaki Disease and developed coronary artery aneurysms. These may have remodelled or persisted. The presence of coronary artery aneurysms predisposes this patient to heart problems, often many years after the initial illness. This is sometimes not recognised by the acute medical services and this protocol is to assist them in this process.

Children, young adults and adults who have had Kawasaki Disease and who develop later cardiac complications will require emergency specialist management, which is significantly different from the management of conventional paediatric or adult cardiac complications.

The signs and symptoms may be unusual, and the patient may be much younger than a 'typical' patient with heart problems. **ECG and Troponin may be normal even if there is acute coronary occlusion (ACS).**

Patients are at risk of a coronary event with **atypical presentation and due to a history of Kawasaki Disease, signs should be taken seriously.** Chest pain **may or may not be present**, the patient could complain of poorly localised pain, have unusual pallor, sweating, marked breathlessness, excessive tiredness, discolouration of the skin, abdominal pain, or unexplained vomiting – features which could be due to a potential coronary event including possible acute thrombus. **Serial troponin levels should be measured, and coronary intervention or imaging should be undertaken urgently.**

Local Emergency Response Plan (Kawasaki Disease):

A local emergency response plan should be established and documented below to include:

Destination centre - if a child – <u>Named Children's Cardiac Surgical centre:</u>	[Insert named hospital]
Destination centre - if an adult – <u>Named Heart Attack Centre:</u>	[Insert named hospital]

Local Emergency Response Team:

The following emergency response team should be in place and briefed at the destination centre, aware of the different circumstances and special requirements of a patient with prior Kawasaki Disease history and the actions needed should they present with suspected CAA thrombosis or myocardial ischemia:

- = Emergency medicine paediatric consultant / Emergency medicine consultant
- = Paediatric cardiology consultant and consultant interventional cardiologist
- = Paediatric or adult cardiac anaesthetist
- = Echocardiography and imaging staff (as age relevant)
- = Lead Clinician – responsible for long-term care - should be notified

The Local Emergency Response Plan (Kawasaki Disease) should include enabling urgent access to facilities which will be required in the event of a Kawasaki Disease emergency including cardiac catheter lab, MRI, CT etc.

Specific Instructions for Transportation, Investigation and Treatment**Ambulance Service:**

1. **If [Patient name] presents** with either chest pain of apparent cardiac origin or other symptoms indicative of a coronary event, such as abdominal pain, excess fatigue, poorly localised pain, unusual pallor, breathlessness, vomiting and restlessness, transport to [named specialist Children's Cardiac Surgical centre (Level 1) – or the nearest available Heart Attack Centre]* as appropriate for a suspected "High Risk Acute Coronary Syndrome (ACS)".

***[It is the responsibility of the Lead Clinician to maintain the PSP appropriate to the age of the patient assisting the Ambulance Service to direct the patient to the relevant age appropriate services]**

2. If the ECG shows ST elevation or LBBB, then transport to named or nearest available paediatric cardiac surgical centre OR Heart Attack Centre as "confirmed STEMI/new onset LBBB" call.
3. The Local Emergency Response Plan (Kawasaki Disease) should be activated and key staff notified at the destination (hospital named in this PSP).

At the Heart Attack Centre or Paediatric Cardiac Surgical Centre:

Even if ECG is normal urgent imaging is indicated to exclude thrombus / critical narrowing. Half of all adults with giant aneurysms due to KD present with thrombus with normal ECG/troponin

4. **If there is STEMI**
The working diagnosis must be of new thrombus in the coronary artery unless or until this can be excluded. Restoration of blood flow by urgent coronary intervention is the priority. If there is delay, administer intravenous thrombolysis. **NB See Algorithm overleaf [insert age relevant algorithm]**
5. **If there is NSTEMI**
The patient will require urgent consultant-led cardiological investigation, with Imaging to exclude Coronary aneurysm thrombosis and possibly including invasive angiography (available in each area 24 hours a day). **NB See Algorithm overleaf [insert age relevant algorithm]**
6. The attending consultant should be informed on admission and be directly involved in these investigations and in treatment.

7. Dr [Insert patient Lead Clinician (specialist Kawasaki Disease care) name] or their team, responsible for [Patient name] regular follow up, wishes to be contacted urgently to discuss treatment.
8. Dr [Insert Lead Clinician name] or their team can be contacted on telephone number [Insert details].

IMPORTANT NOTE: NON-CARDIAC EMERGENCY EVENT

If this patient is involved in an emergency event requiring urgent hospitalisation due to crisis, PLEASE CONTACT [Insert Name of Lead Clinician] for advice as this patient is under lifetime care for Kawasaki Disease and their emergency management may be affected.

[Signed]

Medical Director	[Insert Name]
NHS Trust(s)	[Insert Authorising Trust(s)]
Lead Clinician	[Insert Name]

[INSERT AGE SPECIFIC EMERGENCY MANAGEMENT ALGORITHM HERE]