



Patient Safety Alert

Stage One: Warning

Risk of death and serious harm from failure to recognise acute coronary syndromes in Kawasaki disease patients

11 May 2016

Alert reference number: NHS/PSA/W/2016/004

Alert stage: One - Warning

Around 10 to 20% of all children with Kawasaki disease (KD) develop aneurysms and are at long-term risk of thrombosis and/or stenosis of the coronary artery, which may result in coronary thrombosis and myocardial ischaemia or infarction in patients with KD related aneurysms. [1, 2]

More patients affected by KD in early childhood are now entering adult life and those with persistent or resolved coronary aneurysms are at lifelong risk of acute coronary syndromes (ACS) which could result in death or severe harm. [3, 4, 5]

NHS Improvement is aware of two cases where failure to recognise the likelihood of coronary thrombosis caused a delay in diagnosis and may have increased the risk of death or severe harm. Specialist clinicians are concerned that this risk may be more widespread because those managing such acute presentations lack awareness of the potential for coronary thrombosis.

Highlighting the concern related to one of these cases, a specialist wrote: "There remains a lack of awareness even amongst the cardiological community of the high risk that patients with giant aneurysms are at, of thrombosis and ischaemic events, and the need for urgent investigation of any new symptoms."

While no additional cases related to late diagnosis or delayed treatment of ACS in KD patients were located in the National Reporting and Learning System (NRLS), incident reports confirmed limited awareness of the cardiovascular risks associated with prior KD in many care settings including primary care, emergency departments, medical units, surgery and obstetric care.

This alert is to emphasise the high risk and atypical presentation of coronary artery aneurysms, coronary thrombosis and myocardial ischaemia or infarction in patients with KD, and to highlight the importance of specialist advice.

Increased awareness is required for prompt diagnosis and urgent appropriate investigation and treatment when symptoms of chest pain, pallor, excessive tiredness or breathlessness occur.

Adults and children previously diagnosed with coronary artery aneurysms following KD should be regularly reviewed by a specialist centre and supplied with a Person Specific Protocol (PSP) for use in emergencies.

Guidance and other sources of information are listed overleaf.





Actions

Who:

All providers of NHS-funded care

When:

To begin immediately and be completed by no later than 22 June 2016

-  1 Identify if missed diagnosis or delayed treatment of late complications of Kawasaki disease could occur in your organisation.
-  2 Consider if immediate action needs to be taken locally, and ensure that an action plan is underway if required, to reduce the risk of further incidents occurring.
-  3 Distribute this alert to all relevant staff in primary care, ambulance services, emergency care and inpatient care settings who care for children or adults with a past history of Kawasaki disease.
-  4 Share any learning from local investigations or locally developed good practice resources by emailing patientsafety.enquiries@nhs.net

See page 2 for technical notes and references

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Technical notes

NRLS search dates and terms

The two cases referred to where delay in diagnosis and treatment of ACS in children with a past history of KD had potential to cause death or severe harm were notified via clinical specialist networks. The National Reporting and Learning System (NRLS) was searched 01 January 2012 to 01 January 2016 for incidents containing the term %Kawasaki% in the free text. The search yielded 49 reports in total. STEIS was searched on 11 August 2015 and 01 February 2016 and yielded two reports both of which were also reported to the NRLS. The NRLS and STEIS searches did not identify any additional incidents directly related to harm in children or young adults from missed diagnosis or delayed treatment of late complications of acute coronary syndromes in those who have previously suffered from Kawasaki disease (KD), but confirmed limited awareness of the cardiovascular risks associated with prior KD in a range of settings, as described in the text of the alert.

Stakeholder engagement

- Medical Patient Safety Expert Group
- Primary Care Patient Safety Expert Group
- Infant Children & Young People's Patient Safety Expert Group

For details of the membership of the NHS Improvement patient safety expert groups and steering group see: www.england.nhs.uk/ourwork/patientsafety/patient-safety-groups/

References

1. Newburger JW, Takahashi M, Burns JC. Kawasaki Disease. *J Am Coll Cardiol* 2016; 67:1738–49.
2. Eleftheriou D, Levin M, Shingadia D, et al. Arch Dis Child Published Online First: [25 October 2013] doi:10.1136/archdischild-2012-302841.
3. Daniels LB, Gordon JB, Burns JC. Kawasaki disease: late cardiovascular sequelae. *Current Opinion in Cardiology* 2012; 27:572-7.
4. Gordon JB, Kahn AM, Burns JD, When children with Kawasaki disease grow up: Myocardial and vascular complications in adulthood. *J Am Coll Cardiol*. 17 November, 2009; 54(21): 1911–1920.
5. Shah V, Christov G, Mukasa T, Brogan KS, Wade A, Eleftheriou D, Levin M, Tulloh RM, Almeida B, Dillon MJ, Marek J, Klein N, Brogan PA. Cardiovascular status after Kawasaki disease in the UK. *Heart* 2015; 0:1–10. doi:10.1136/heartjnl-2015-307734.

Other sources of material

- Guidance from London Cardiac Strategic Clinical Network on acute cardiovascular management of patients with previous Kawasaki disease www.londonscn.nhs.uk/wp-content/uploads/2015/11/cvd-acute-kd-guidance-092015.pdf
- Letter to acute hospital trusts in London from London Cardiac Strategic Clinical Network www.londonscn.nhs.uk/wp-content/uploads/2015/11/cvd-acute-kd-letter-092015.pdf
- Newburger JW, Takahashi M, Gerber MA, et al. Diagnosis, treatment, and long-term management of Kawasaki disease: a statement for health professionals from the Committee on Rheumatic Fever, Endocarditis and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. *Circulation* 2004;110:2747–71 circ.ahajournals.org/content/110/17/2747.full.pdf