

Kawasaki Disease

(Also Known as Mucocutaneous Lymph Node Syndrome)

DISEASE REPORTABLE WITHIN 24 HOURS OF DIAGNOSIS

Per N.J.A.C. 8:57, healthcare providers and administrators shall report by mail or by electronic reporting within 24 hours of diagnosis, confirmed cases of Kawasaki disease to the health officer of the jurisdiction where the ill or infected person lives, or if unknown, wherein the diagnosis is made. A directory of local health departments in New Jersey is available at

<http://www.state.nj.us/health/lh/directory/lhdselectcounty.shtml>.

If the health officer is unavailable, the healthcare provider or administrator shall make the report to the Department by telephone to 609.826.5964, between 8:00 A.M. and 5:00 P.M. on non-holiday weekdays or to 609.392.2020 during all other days and hours.



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1 THE DISEASE AND ITS EPIDEMIOLOGY

A. Etiologic Agent

The cause of Kawasaki disease is unknown but is presumably an infectious or toxic agent.

B. Clinical Description

Kawasaki disease is a multisystem disease primarily affecting children under five years old. It is an acute, self-limited, systemic vasculitis characterized by an initial high spiking fever that can persist for one to two weeks. It does not respond to antibiotics and it is associated with extreme irritability and mood changes. The fever is accompanied by bilateral conjunctivitis (seen in about 85% of cases); changes to the lips and mouth including dry, red, or cracked lips; a sore red throat, and/or strawberry tongue (90%); changes to the extremities including peeling of skin, rashes, and/or swelling of the hands and feet (75%); a generalized red rash affecting the trunk or perineal region (80%); and an enlarged cervical lymph node (usually solitary, often unilateral) (70%). Other common symptoms include severe abdominal cramping, diarrhea, and vomiting. The convalescent (recovery) phase can be lengthy. Cardiac involvement is a major cause of morbidity and mortality associated with Kawasaki disease. Approximately 20% of untreated cases develop coronary artery aneurysms within six weeks. Prompt treatment with high-dose intravenous immune globulin and aspirin can reduce aneurysm formation to about 8% of cases, as well as reduce fever and inflammatory signs. The case-fatality ratio for Kawasaki disease is 0.1% to 1%. Other complications can involve any organ of the body.

C. Reservoirs

Reservoirs are unknown.

D. Modes of Transmission

Modes of transmission are unknown.

E. Incubation Period

The incubation period is unknown.

F. Period of Communicability or Infectious Period

The disease is not known to be communicable from person-to-person.

G. Epidemiology

Kawasaki disease occurs worldwide with most cases reported from Japan, where nationwide epidemics have been documented. The estimated number of new cases each year in the United States is approximately 2000. The peak age of occurrence in the United States is between 18 and 24 months, with 80% of cases reported in children younger than five. Males are affected more frequently than are females, and children of Asian descent have the highest incidence rate. Epidemics occur on a two- to three-year cycle, usually during the winter and spring. While the mode of transmission for Kawasaki disease has not been documented, siblings (especially twins) of cases are more likely to be affected than are the general population. An average of 23 cases of Kawasaki disease is reported annually to the New Jersey Department of Health and Senior Services.

2 CASE DEFINITION

There is no formal Centers for Disease Control and Prevention (CDC) surveillance case definition for Kawasaki syndrome.

A. New Jersey Department of Health and Senior Services (NJDHSS) Case Definition

1. Clinical Description

Kawasaki disease is a multisystem disease primarily affecting children under five years old. It is an acute, self-limited, systemic vasculitis characterized by an initial high spiking fever that can persist for one to two weeks. It does not respond to antibiotics and it is associated with extreme irritability and mood changes. The fever is accompanied by bilateral conjunctivitis (seen in about 85% of cases); changes to the lips and mouth including dry, red, or cracked lips; a sore red throat, and/or strawberry tongue (90%); changes to the extremities including peeling of skin, rashes, and/or swelling of the hands and feet (75%); a generalized red rash affecting the trunk or perineal region (80%); and an enlarged cervical lymph node (usually solitary, often unilateral) (70%). Other common symptoms include severe abdominal cramping, diarrhea, and vomiting. The convalescent (recovery) phase can be lengthy. Cardiac involvement is a major cause of morbidity and mortality associated with Kawasaki disease. Approximately 20% of untreated cases develop coronary artery aneurysms within six weeks. Prompt treatment with high-dose intravenous immune globulin and aspirin can reduce aneurysm formation to about 8% of cases, as well as reduce fever and inflammatory signs.

The case-fatality ratio for Kawasaki disease is 0.1% to 1%. Other complications can involve any organ of the body.

2. Laboratory Criteria for Diagnosis

There are no diagnostic laboratory tests for Kawasaki disease.

3. Case Classification

CONFIRMED

A febrile illness of greater than or equal to five days' duration, with at least four of the following physical findings and reasonable exclusion of other diseases (toxic shock, scalded skin syndrome, scarlet fever, rickettsial diseases, or drug reactions):

- Bilateral conjunctival injection
- Oral changes (erythema of lips or oropharynx, strawberry tongue, or fissuring of the lips)
- Peripheral extremity changes (edema, erythema of palms and soles, or generalized or periungual desquamation)
- Cervical lymphadenopathy
- Rash

PROBABLE

Not used.

POSSIBLE

Not used.

3 LABORATORY TESTING AVAILABLE

There are no specific laboratory tests to confirm or exclude Kawasaki disease. Diagnosis of Kawasaki disease is based on clinical presentation. Leukocytosis, thrombocytosis, and elevated sedimentation rate are common findings in the early stage of illness. Diagnostic decisions are made based on clinical presentations when there are no other reasonable explanations for the illness. The electrocardiogram changes and coronary artery aneurysms can be observed in the fifth or sixth week of illness.

4 PURPOSE OF SURVEILLANCE AND REPORTING AND REPORTING REQUIREMENTS

A. Purpose of Surveillance and Reporting

The purpose of surveillance and reporting is to identify disease clusters and demographic characteristics.

B. Healthcare Provider Reporting Requirements

The New Jersey Administrative Code (NJAC 8:57-1.8) stipulates that healthcare providers report (by telephone, confidential fax, over the Internet using the confidential and secure Communicable Disease Reporting and Surveillance System [CDRSS], or in writing) any case of Kawasaki disease to the local health officer having jurisdiction over the locality in which the patient lives or, if unknown, to the health officer in whose jurisdiction the healthcare provider requesting the laboratory examination is located.

C. Local Department of Health Reporting

NJAC 8:57-1.8 stipulates that each local health officer must report the occurrence of any case of Kawasaki disease, as defined by the reporting criteria in section 2A above.

Current requirements are that cases be reported to NJDHSS Infectious and Zoonotic Diseases Program (IZDP) using the Kawasaki Syndrome Report. A report may also be filed electronically using CDRSS.

5 CASE INVESTIGATION

It is the local health officer's responsibility to investigate a case and complete a Kawasaki Syndrome Report Form. Much of the information required on the forms can be obtained from the patient's healthcare provider or the medical record.

Use the following guidelines for assistance in completing the form:

1. Accurately record the demographic information, date of symptom onset, whether hospitalized (and associated dates), outcome of disease, and whether the patient has recurrent Kawasaki syndrome.
2. Collect the information requested in the "Clinical" section. Ask about duration of fever, presence of conjunctival injection, oral changes, peripheral extremity changes, cervical lymphadenopathy, and skin rash. This information is important in defining a case. Ask the healthcare provider to submit a copy of the medical record or enlist his/her aid in completing these sections of the Kawasaki Syndrome Report Form.

Communicable Disease Service Manual

3. Ask if toxic shock syndrome, scalded skin syndrome, scarlet fever, rickettsial diseases, and drug reaction were excluded.
4. If there have been several attempts to obtain patient information (e.g., the patient or healthcare provider does not return calls or respond to a letter, or the patient refuses to divulge information or is too ill to be interviewed), please fill out the form with as much information as possible. Please note on the form why it could not be filled out completely.

After completing the case report form, mail it in an envelope marked “Confidential” to NJDHSS IZDP, or file the report electronically using CDRSS. The mailing address is:

NJDHSS
Division of Epidemiology, Environmental and Occupational Health
Infectious and Zoonotic Diseases Program
PO Box 369
Trenton, NJ 08625-0369

A. Entry into CDRSS

The mandatory fields in CDRSS include: disease, last name, county, municipality, gender, race, ethnicity, case status, report status.

The following table can be used as a quick reference guide to determine which CDRSS fields need to be completed for accurate and complete reporting of Kawasaki disease cases. The “Tab” column includes the tabs which appear along the top of the CDRSS screen. The “Required Information” column provides detailed explanations of what data should be entered.

CDRSS Screen	Required Information
Patient Info	Enter the disease name (“KAWASAKI DISEASE”) in patient demographic information, illness onset date, and the date the case was reported to the local health department (LHD). There are no subgroups for Kawasaki Disease.
Addresses	Enter any alternate address (e.g., a daycare address). Use the Comments section in this screen to record any pertinent information about the alternate address (e.g., the times per week the case-patient attends daycare). Entering an alternate address will allow other disease investigators access to the case if the alternate address falls within their jurisdiction.

CDRSS Screen	Required Information
Clinical Status	Enter any treatment that the patient received and record the names of the medical facilities and physician(s) involved in the patient’s care. If the patient received care from two or more hospitals, be sure that all are entered so the case can be accessed by all infection control professionals (ICPs) covering these facilities. If immunization status is known, it should also be entered here. If the patient died, date of death should be recorded under the Mortality section.
Signs/Symptoms	Check appropriate boxes for signs and symptoms and indicate their onset. Please refer to the case definition for a complete description of the clinical criteria for Kawasaki disease. Make every effort to get complete information by interviewing the physician, family members, ICP, or others who might have knowledge of the patient’s illness. Also, information regarding the resolution of signs and symptoms should be entered.
Risk Factors	Enter complete information about risk factors to facilitate study of Kawasaki disease in New Jersey.
Laboratory Eval	Information regarding laboratory testing is not required for this disease.
Contact Tracing	Information regarding contacts is not required for this disease.
Case Comments	Enter general comments (i.e., information that is not discretely captured by a specific topic screen or drop-down menu) in the Comments section. NOTE: Select pieces of information entered in the Comments section CANNOT be automatically exported when generating reports. Therefore, whenever possible, record information about the case in the fields that have been designated to capture this information; information included in these fields CAN be automatically exported when generating reports.
Epidemiology	Under the Other Control Measures section, indicate if the patient falls into any of the categories listed under Patient Role(s)/Function(s) (e.g., “DAYCARE ATTENDEE,” “DAYCARE PROVIDER”). Record name of and contact information for case investigators from other agencies (e.g., CDC, out-of-state health departments). Document communication between investigators in the Comments section.

6 CONTROLLING FURTHER SPREAD

A. Isolation and Quarantine Requirements (NJAC 8:57-1.10)

None.

B. Protection of Contacts of a Case

None.

C. Managing Special Situations

1. Reported Incidence is Higher than Usual/Outbreak Suspected

If multiple cases of Kawasaki syndrome occur in an institutional setting or jurisdiction, or if an outbreak is suspected, investigate clustered cases. Identify common factors, such as age, school, workplace, or activities, to help elucidate risk factors. Consult with NJDHSS IZDP at 609.588.7500 to help determine a course of action and, if necessary, to perform surveillance for cases that may cross several jurisdictions and therefore be difficult to identify at a local level.

D. Preventive Measures

None.

Additional Information

A Kawasaki Syndrome Fact Sheet can be obtained at the NJDHSS Web site at www.state.nj.us/health.

References

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Leung DYM, Meissner HC. The Many Faces of Kawasaki Syndrome. January 15, 2000.

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Massachusetts Department of Public Health, Division of Epidemiology and Immunization. *Guide to Surveillance and Reporting*. Boston, MA: Massachusetts Department of Public Health, Division of Epidemiology and Immunization; January 2001.