Kawasaki disease, or mucocutaneous lymph node syndrome, is a disease of unknown etiology that most frequently (80% of the time) affects infants and children under 5 years of age. Accurate diagnosis and early therapeutic interventions such as aspirin and intravenous immunoglobulin can decrease the approximately 20% risk of developing coronary artery abnormalities. A specific diagnostic test does not exist. Thus, diagnosis of Kawasaki disease is based on characteristic clinical signs and symptoms, which are classified as principal clinical findings and other clinical and laboratory findings. The male-to-female ratio among patients with Kawasaki disease is 1.5:1. Children of nearly all racial backgrounds are affected. Recurrences and cases in siblings are seen only occasionally.

**Principal Clinical Findings***
**Fever persisting at least 5 days** and the presence of at least 4 of the following 5 principal features:

1. Changes in extremities:
   - Acute: Erythema and edema of hands and feet
   - Convalescent: Membranous desquamation of fingertips
2. Polymorphous exanthema
3. Bilateral, painless bulbar conjunctival injection without exudate
4. Changes in lips and oral cavity: Erythema and cracking of lips, strawberry tongue, diffuse injection of oral and pharyngeal mucosae
5. Cervical lymphadenopathy (≥1.5 cm in diameter), usually unilateral

*Patients with fever and fewer than 4 principal symptoms can be diagnosed as having Kawasaki disease when coronary artery disease is detected by 2-dimensional echocardiography or coronary angiography. Other diagnoses should be excluded. The physician should be aware that some children with illness not fulfilling these criteria have developed coronary artery aneurysms.

†Many experts believe that in the presence of classic features, the diagnosis of Kawasaki disease can be made by experienced observers before day 5 of fever.

**Other Significant Clinical and Laboratory Findings**

**Cardiovascular:** On auscultation, gallop rhythm or distant heart sounds; ECG changes (arrhythmias, abnormal Q waves, prolonged PR and/or QT intervals, occasionally low voltage, or ST–T–wave changes); chest x-ray abnormalities (cardiomegaly); echocardiographic changes (pericardial effusion, coronary aneurysms, or decreased contractility); mitral and/or aortic valvular insufficiency; and rarely, aneurysms of peripheral arteries (eg, axillary), angina pectoris, or myocardial infarction

**Gastrointestinal:** Diarrhea, vomiting, abdominal pain, hydrops of gallbladder, paralytic ileus, mild jaundice, and mild increase of serum transaminase levels

**Blood:** Increased erythrocyte sedimentation rate, leukocytosis with left shift, positive C-reactive protein, hypoalbuminemia, and mild anemia in acute phase of illness (thrombocytosis in subacute phase)

**Urinary:** Sterile pyuria of urethral origin and occasional proteinuria

**Skin:** Perineal rash and desquamation in subacute phase and transverse furrows of fingernails (Beau’s lines) during convalescence

**Respiratory:** Cough, rhinorrhea, and pulmonary infiltrate

**Joint:** Arthralgia and arthritis

**Neurological:** Mononuclear pleocytosis in cerebrospinal fluid, striking irritability, and rarely, facial palsy

**Selected Readings**


Figure 1. Rash of Kawasaki disease in a 7-month-old on the 4th day of illness.

Figure 2. Conjunctival injection, lip edema, and erythema in a 2-year-old boy on the 6th day of illness.

Figure 3. Erythematous and edematous hand of a 1½-year-old girl on the 6th day of illness.

Figure 4. Periungual desquamation in a 3-year-old on the 12th day of illness.

Figure 5. Two-dimensional echocardiogram. AO indicates aorta; PA, pulmonary artery; LAD, left anterior descending coronary; CX, circumflex coronary; and L Main, left main coronary.

Figure 6. Coronary angiogram demonstrating hugely dilated left anterior descending (LAD) artery with obstruction and very dilated right coronary artery (RCA) with an area of severe narrowing in a 6-year-old boy.
Diagnostic Guidelines for Kawasaki Disease
Council on Cardiovascular Disease in the Young Committee on Rheumatic Fever Endocarditis and Kawasaki Disease American Heart Association

_Circulation_. 2001;103:335-336
doi: 10.1161/01.CIR.103.2.335

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2001 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/103/2/335

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in _Circulation_ can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to _Circulation_ is online at:
http://circ.ahajournals.org/subscriptions/