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# At the Heart of the Fever: Kawasaki Disease

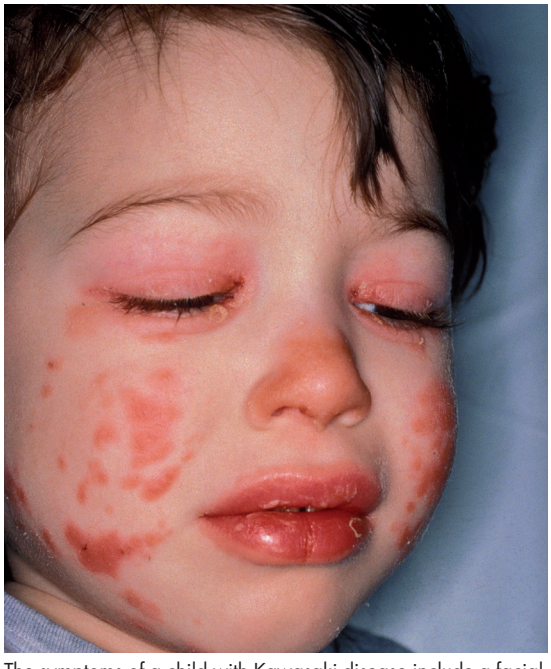
*Strategies for managing the most common cause of acquired pediatric heart disease in the United States.*

Jay Watts, a previously healthy, feisty four-year-old, was admitted through the ED to the hospital's general medicine unit with suspected Kawasaki disease. (This case is a composite based on several we've encountered in our practice.) Blood and urine samples were drawn, but the laboratory analysis was not yet complete. Jay had a five-day history of high fever and red, puffy hands and feet. His eyes were bloodshot and irritated, and his fiery lips were dry and cracked. Extremely irritable and fearful of staff, Jay initially batted away the nurse, who was trying to take his vital signs. To encourage Jay's cooperation, the nurse asked him to help examine a "sick" stuffed dinosaur. After that, the nursing evaluation proceeded smoothly, revealing that Jay had a strawberry tongue; a large, swollen lymph node on the right side of his neck; skin that was hot to the touch; and a fever of 104.5°F (40.3°C) that was unresponsive to antipyretics. Jay's trunk and groin were covered with a rash the color of his flaming red hair, and the skin around his groin was peeling.

Using a televised cartoon for distraction, the nurse had no trouble obtaining a baseline electrocardiogram, which showed Jay to have sinus tachycardia with nonspecific ST-segment and T-wave changes. The primary care provider ordered an echocardiogram to be performed as soon as possible. While awaiting laboratory and echocardiographic results, the care team supplemented Jay with half-maintenance IV fluids. They focused on monitoring his vital signs and keeping him comfortable, encouraging him to drink diluted apple juice to soothe his dry mouth and throat, dimming the lights in his room (because he said the bright lights hurt his eyes), and providing movies and games for diversion.

Because Jay's laboratory studies, like his presentation, were consistent with Kawasaki disease (also known as mucocutaneous lymph node syndrome), IV immunoglobulin (IV Ig) and aspirin treatment were initiated. In this acute stage, the primary treatment goals for Jay were fever reduction and to ensure his continued comfort.

**Kawasaki disease is the most common cause of acquired pediatric heart disease** in the United States.[1]



The symptoms of a child with Kawasaki disease include a facial rash with erythematous macules, cheilitis, swollen lymph nodes, and conjunctivitis associated with a prolonged fever. Photo © Dr. Jean Claude Amoric.

Coronary artery aneurysms develop in approximately 20% of untreated children.[2] Delayed diagnosis and treatment (more than 10 days after onset) substantially raise the risk of coronary artery abnormalities.[2,3] To ensure timely intervention, it's essential for clinicians to recognize the classic signs and symptoms of Kawasaki disease, be familiar with its incomplete presentation, understand its pathophysiology, and know the current standard of care and recommended follow-up for patients with and without cardiac involvement.

## EPIDEMIOLOGY AND ETIOLOGY

Kawasaki disease is best described as an acute, self-limiting vasculitis, with a total duration of six to eight weeks. Generally, patients are hospitalized only during the initial, febrile phase of the illness. The typical hospital length of stay is about three days.[4] Kawasaki

disease affects 50% to 70% more males than females, and in the United States, 76% of reported cases occur in children under the age of five, with two years being the median age of patients hospitalized with the disease.[5] Prevalence is greatest in Japan and among children of Japanese descent, regardless of where they live. In the United States, incidence is highest among those of Asian or Pacific Island descent, though the disease is known to occur in children of all races.[5]

Kawasaki disease was first described in 1967 by Dr. Tomisaki Kawasaki.[5] Despite many years of research devoted to its discovery, the cause of the disease is still unknown. Kawasaki disease peaks during winter and spring, is self-limiting, and affects primarily young children, suggesting an infectious etiology. Many experts believe that this illness may be an inflammatory response to an infectious trigger that occurs in genetically predisposed children.[5] Research into the genetic factors that may play a role in susceptibility is ongoing.

## Table 1. Clinical Features of Kawasaki Disease[5, 6]

| Classic clinical presentation |
|---|
| **Fever** of at least five days' duration accompanied by at least four of the following five clinical characteristics: |
| **Extremity changes** |
| • Erythema of palms and soles |
| • Edema of hands and feet |
| • Peeling of fingers and toes around the nails |
| **Polymorphic rash** |
| **Bilateral conjunctivitis** without exudates |
| **Oropharyngeal changes** |
| • Lip cracking and erythema |
| • Strawberry tongue |
| **Cervical lymphadenopathy** |
| **Associated clinical findings** |
| Pericarditis |
| Valvular regurgitation |
| Myocarditis |
| Coronary artery dilation |
| Joint pain |
| Diarrhea |
| Vomiting |
| Abdominal pain |
| Decreased oral intake |
| Hepatic dysfunction |
| Hydrops of the gallbladder |
| Extreme irritability |
| Aseptic meningitis |
| Sensorineural hearing loss |
| **Associated laboratory findings** |
| Elevated white blood cell count |
| Elevated erythrocyte sedimentation rate |
| Elevated C-reactive protein |
| Anemia |
| Low albumin levels |
| Low sodium levels |
| Thrombocytosis (usually peaks in the second week of illness) |
| Sterile pyuria |
| Elevated transaminases |

## PATHOPHYSIOLOGY

Kawasaki disease progresses through three phases—acute, subacute, and convalescent—which vary in duration (depending on whether and when the patient receives treatment) but have distinct pathophysiologic characteristics. The acute phase, defined as the period from the onset through the resolution of fever, is typified by widespread inflammation of blood vessels. Laboratory studies during this phase reveal such nonspecific inflammatory markers as an elevated erythrocyte sedimentation rate (ESR) and a high level of C-reactive protein (CRP). As the vascular inflammation progresses, it can affect medium-sized muscular vessels, particularly coronary arteries, destroying the integrity of vessel walls and, possibly, producing coronary artery aneurysms. During the subacute phase, which begins with the resolution of fever, inflammation is still evident. ESR and CRP level remain elevated, and anemia is common. This period is followed by the convalescent phase of the disease, in which laboratory studies continue to show evidence of inflammation, but signs and symptoms slowly dissipate, unless complications develop.

Cardiovascular complication is the leading cause of morbidity in Kawasaki disease. Although the disease mortality rate is low (less than 1.7% in the United States), when death occurs, it almost always has a cardiovascular cause.[5] In addition to coronary artery aneurysms, the cardiac sequelae of Kawasaki disease may include myocarditis, impaired left ventricular function, valvular regurgitation, arrhythmia, and pericardial effusion.

## DIAGNOSIS

In 2004, the American Heart Association (AHA) convened a multidisciplinary committee of experts to review and revise their recommendations for the diagnosis, treatment, and long-term management of Kawasaki disease; their report was published later that year.[5] The committee determined that the disease should be considered in the differential diagnosis of all pediatric patients presenting with characteristic signs and symptoms of Kawasaki disease with a prolonged, unexplained fever of five days or more, including adolescents and infants under the age of one in whom the diagnosis is too often overlooked. In addition to discussing classic presentations, these recommendations addressed incomplete Kawasaki disease, for which diagnosis is often delayed, putting patients at greater risk for aneurysms.

**Clinical manifestations.** There is no diagnostic test for Kawasaki disease; diagnosis is based on clinical and

laboratory findings (see Table 1[5,6]). The classic criteria for diagnosis are a fever of five or more days' duration, and at least four of the following five clinical characteristics (or fewer than four in the presence of coronary artery abnormalities detected by two-dimensional echocardiography or angiography):

- extremity changes, including erythema or edema of the hands and feet, with palms and soles appearing to be beefy and red (an acute sign, usually apparent within the first week of fever onset) and peeling of the fingertips and feet (usually a subacute sign)
- polymorphic, nonvesicular rash, often accentuated in the groin area and, possibly, peeling early in the illness
- bilateral, nonpurulent, limbal-sparing conjunctivitis
- oropharyngeal changes, such as a strawberry tongue; red, cracked lips; or nonexudative oropharyngeal erythema
- cervical lymphadenopathy (of 1.5 cm or more), which is generally unilateral, not painful, and often confined to the anterior cervical triangle

Patients like Jay, who present with more than four of the classic clinical characteristics, can be diagnosed after only four days of unresponsive high fever.[5] If there is echocardiographic or angiographic evidence of coronary artery abnormalities, Kawasaki disease may be diagnosed in patients with a five-day history of unresponsive fever and fewer than four clinical characteristics.

As Jay's case illustrates, fever associated with Kawasaki disease is unresponsive to antipyretics and remains high for at least five days. It tends to peak in the late afternoon or early evening. Without appropriate treatment, the average duration is 11 days, though fever may continue for four weeks or longer.[5]

A thorough history is essential in the successful treatment of Kawasaki disease. Several clinicians may see the patient during the acute phase, and acute symptoms don't always present simultaneously. The details of presentation are important in differentiating Kawasaki disease from other common pediatric illnesses that share some of the same signs and symptoms (see Table 2[5-9]).

Associated symptoms of Kawasaki disease may include irritability (for the six-to-eight-week duration of the illness); arthritis or arthralgia of multiple joints, generally starting in the small joints and sometimes progressing to the larger, weight-bearing joints; and gastrointestinal distress, including diarrhea, vomiting, and abdominal pain.[6] Hepatic inflammation, jaundice, and hydrops of the gallbladder may occur in the first two weeks of illness, but are usually self-limiting. Sensorineural hearing loss is a rare sequela.[5]

**Cardiac studies.** As it was done in Jay's case, a baseline electrocardiogram is usually obtained to detect arrhythmia, prolonged PR interval, or nonspecific ST-segment or T-wave changes that may occur with myocardial inflammation. Cardiac imaging, mainly echocardiography, is used to evaluate the cardiac status of patients suspected of having Kawasaki disease. The initial echocardiogram should be performed as soon as the disease is suspected, as it was with Jay. Its purpose is to establish the patient's baseline cardiac status with a focus on coronary artery dimensions, left ventricular size and function, presence of effusion, and valvular function. When no cardiovascular complications are evident, echocardiograms are repeated one to two weeks after treatment is initiated and again four to six weeks later.[5] More frequent echocardiographic evaluations are performed in patients who have coronary artery dilatation or fail to respond to treatment.

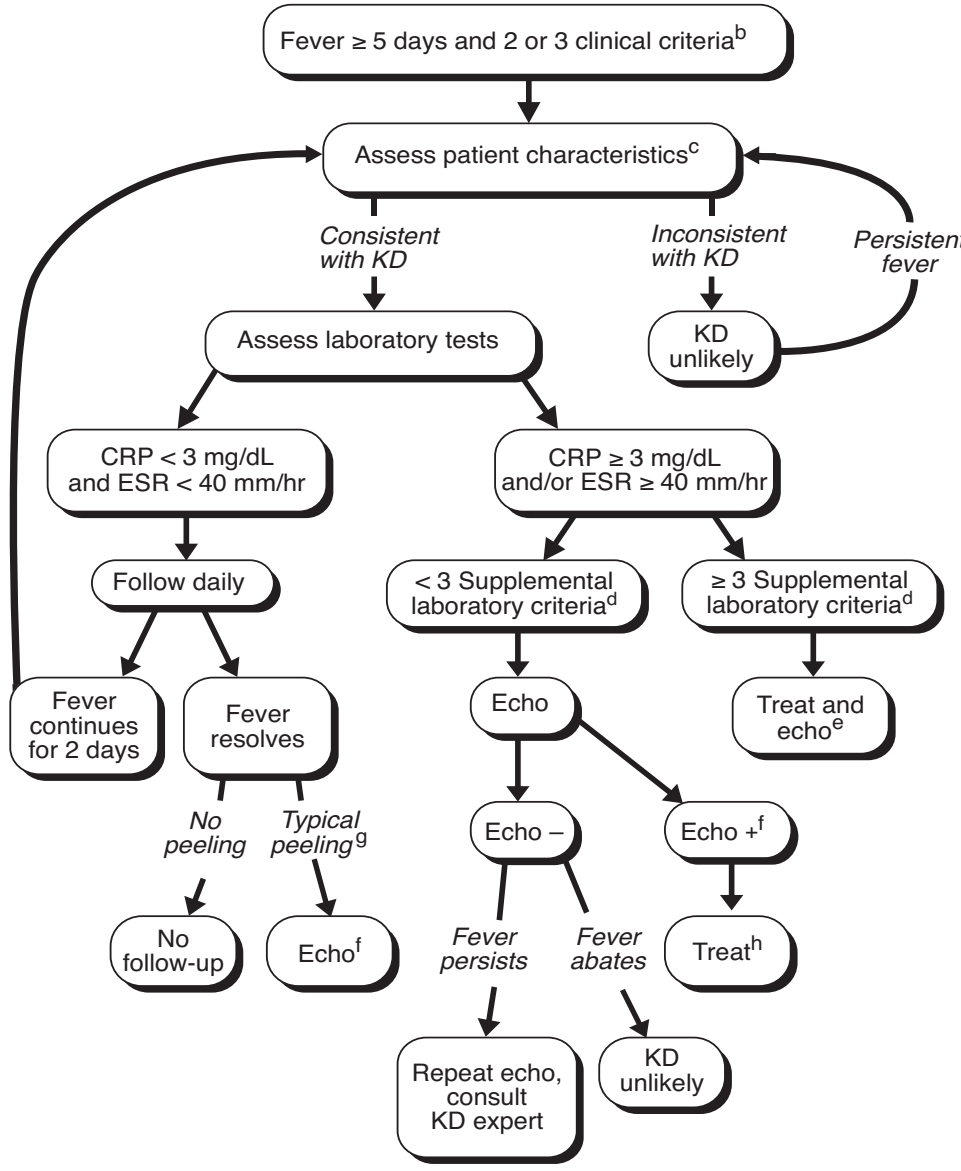
Initial echocardiograms may show prominent coronary arteries, with aneurysms developing over the first few weeks of illness. Other common acute and subacute signs of Kawasaki disease include echocardiographic evidence of pericardial effusion, valvular regurgitation, and (because many affected patients have some degree of myocarditis) decreased left ventricular function. Although ventricular function usually improves with IV Ig administration and returns to baseline, diminished function causes some children to become very ill during the initial phase of the illness.

Jay's baseline echocardiogram showed a small pericardial effusion and mild mitral valve regurgitation. The diameters of his coronary arteries were measured at the

## Table 2. Common Pediatric Illnesses that Share Clinical Features with Kawasaki Disease[5-9]

| Disease | Similar clinical symptoms |
|---|---|
| Staphylococcal infections<br>• Scalded skin syndrome<br>• Toxic shock syndrome | Fever, rash |
| Streptococcal infections<br>• Strep throat<br>• Scarlet fever | Fever, strawberry tongue, dry lips, rash, throat and mouth pain |
| Viral syndromes | Fever, rash, arthralgia, diarrhea, vomiting, throat pain, lymphadenopathy, conjunctivitis, photophobia, headache, muscle pain |
| Juvenile rheumatoid arthritis | Arthritis, arthralgia, fever |
| Steven-Johnson syndrome | Rash, fever |
| Drug reaction | Rash |
| Gastroenteritis | Vomiting, diarrhea, abdominal pain |
| Upper respiratory infection | Cough |
| Urinary tract infection | Sterile pyuria, fever, abdominal pain |
| *Rickettsiae rickettsii* (Rocky Mountain spotted fever) | Rash, fever, nausea, vomiting, muscle pain, headache |
| Leptospirosis | Fever, rash, headache, vomiting, diarrhea, abdominal pain, muscle pain, jaundice, reddened eyes |

## Figure 1. Algorithm for Evaluation of Suspected Incomplete Kawasaki Disease[a]




CRP = C-reactive protein; ESR = erythrocyte sedimentation rate; KD = Kawasaki disease.

[a] In the absence of a gold standard for diagnosis, this algorithm cannot be evidence based but rather represents the informed opinion of the expert committee (Newburger JW, et al[5]). Consultation with an expert should be sought any time assistance is needed; [b] Infants ≤ 6 months old on day ≥ 7 of fever without another explanation should undergo laboratory testing and, if evidence of systemic inflammation is found, an echocardiogram, even if they have no clinical criteria; [c] Clinical features of Kawasaki disease are listed in Table 1. Characteristics suggesting disease other than Kawasaki disease include exudative conjunctivitis, exudative pharyngitis, discrete intraoral lesions, bullous or vesicular rash, or generalized adenopathy. If any of these are present, consider alternative diagnoses (see Table 2); [d] Supplemental laboratory criteria include albumin ≤ 3 g/dL, anemia for age, elevation of alanine aminotransferase, platelets after 7 days ≥ 450,000/$mm^3$, white blood cell count ≥ 15,000/$mm^3$, and urine ≥ 10 white blood cells/high-power field; [e] Can treat before performing echocardiogram; [f] Echocardiogram is considered positive if any of three conditions are met: *z* score of left anterior descending (LAD) or right coronary artery (RCA) ≥ 2.5, coronary arteries meet Japanese Ministry of Health criteria for aneurysms, or ≥ 3 other suggestive features exist, including pervascular brightness, lack of tapering, decreased left ventricular function, mitral regurgitation, pericardial effusion, or *z* scores in LAD or RCA of 2–2.5; [g] Typical peeling begins under nail bed of fingers and then toes; [h] If echocardiogram is positive, treatment should be given within 10 days of fever onset or beyond day 10 in children with clinical and laboratory signs (high level of CRP or elevated ESR) of ongoing inflammation.

Source: American Heart Association, Inc.[5]

upper limits of normal, and his left ventricular function was slightly depressed. Since his left ventricular function was nearly normal, and he had no clinical signs of congestive heart failure, he was scheduled for a second echocardiogram in a week. (If his initial echocardiogram had showed enlarged coronary artery dimensions or clinically significant left ventricular dysfunction, the second test would have been scheduled sooner.) The cardiology team noted that indications for interim echocardiographic evaluation would include continued or recrudescent fever.

Coronary artery dilatation evolves over the first few weeks of illness.[10] The risk of coronary artery aneurysms rises with the duration of fever. Young infants (under the age of 12 months), especially the males, are at greatest risk for such aneurysms. Delayed diagnosis and treatment are associated with significant morbidity and mortality.

**Laboratory studies.** Initial laboratory studies generally include a complete blood count with differential and platelet count, along with ESR, CRP level, and alanine aminotransferase (ALT) and albumin tests. Elevation of the ESR, CRP level, and white blood cell counts, with a predominance of granulocytes, is characteristic of the acute phase of Kawasaki disease. Thrombocytosis occurs later in the disease process, with platelet counts of 500,000 to 1,000,000/mm$^3$ peaking in the second or third week of illness. Other laboratory findings include elevated serum transaminases, hyperbilirubinemia, sterile pyuria, and aseptic meningitis.

Jay's ESR was 96 mm/hr (normal is less than 20 mm/hr), his CRP level was 7.4 mg/L (normal is less than 0.5 mg/L), his white blood cell count was 15,000/mm$^3$ (normal is 4,300 to 10,800/mm$^3$) with an increase in band cells, and his ALT was twice the normal level. His microurinalysis revealed sterile pyuria, suggesting inflammation rather than infection.

## INCOMPLETE KAWASAKI DISEASE

Kawasaki disease may also present as incomplete Kawasaki disease; this possibility should be considered in all children with unexplained, persistent fever of more than five days' duration in the presence of two or three of the disease's classic clinical features.[2,5] Children under age one are more likely than older children to have incomplete Kawasaki disease.[1] The AHA recommendations include an algorithm for evaluating patients with suspected incomplete Kawasaki disease that details appropriate laboratory studies and cardiac imaging (see Figure 1[5]).

## TREATMENT

In the acute phase, treatment focuses on reducing inflammation with the goal of preventing coronary artery dilatation and aneurysm formation. Long-term therapy focuses on preventing thrombosis and myocardial ischemia or infarction in patients who have documented coronary artery aneurysms.

**Treatment in the acute phase.** The current standard of care is IV Ig treatment for all patients diagnosed with Kawasaki disease. This therapy shortens the duration of fever and reduces the incidence of coronary artery aneurysms to less than 5%, down from an estimated 15% to 25%.[5] High-dose IV Ig (2 g/kg) should be initiated as soon as the diagnosis is made (ideally, by day 7, but at least by day 10). Even when diagnosis is made after day 10, IV Ig should be administered in the presence of fever, coronary artery dilatation, aneurysms, or signs of inflammation (for example, an elevated ESR or CRP level).

Because IV Ig is a blood product, allergic reaction is possible. To reduce this risk, per hospital protocol, Jay was given IV diphenhydramine on the night of admission, before IV Ig was started. The eight-to-12-hour IV Ig infusion was initiated at a rate of 0.5 mL/kg/hr. After 30 minutes, it was increased to a rate of 1 mL/kg/hr, and after another 30 minutes, it was raised to a rate of 2 mL/kg/hr, where it was maintained for the remainder of the infusion. Nurses took Jay's vital signs at the start of the infusion, with each upward titration, every hour after the maximum infusion rate was achieved, and upon completion of the infusion. Jay's fever had subsided by the morning of day 2 of hospitalization, within six hours of IV Ig initiation. After Jay had been consistently afebrile for 24 hours, he was discharged to home on day 3.

Although aspirin hasn't been shown to reduce the incidence of aneurysm, it's administered routinely during the acute phase of the illness (at dosages of 20 to 25 mg/kg orally every six hours) because it helps to control fever and reduce discomfort. Practice varies across institutions regarding the duration of high-dose aspirin, but it's typically given until the patient has been afebrile for two to three days and is then replaced with an antiplatelet dose (3 to 5 mg/kg/day), which is maintained until laboratory values normalize and the patient's week-4-to-week-6 echocardiogram is completed. If the coronary artery dimensions are normal at that time, aspirin is discontinued.

**Treatment of refractory Kawasaki disease.** For about 10% to 20% of patients with Kawasaki disease, fever either persists or returns 36 hours or more after the initial IV Ig infusion is complete.[5] In such cases, a repeat dose of 2 g/kg IV Ig is recommended. Other antiinflammatory medications may be used in patients whose fever and inflammation persist after two or more IV Ig infusions.[5,11] Practice varies in the treatment of patients who don't respond to IV Ig. Clinical data do not currently support any particular additional therapy as optimal.

**Long-term management of Kawasaki disease.** In general, children without cardiac involvement return to their previous state of health and require infrequent follow-up, mainly for surveillance and to reduce coronary risk factors such as hypercholesterolemia and hypertension. Patients who develop coronary artery

aneurysms require more frequent follow-up, which is focused on the prevention and detection of ischemia.

About 50% to 67% of coronary artery aneurysms resolve within two years of onset of Kawasaki disease.[5] The larger the aneurysm, the less likely the vessel is to return to normal dimensions, and the more likely it is to become stenotic. Patients with giant aneurysms (larger than 8 mm in diameter) are at greatest risk for morbidity and mortality, with the prevalence of coronary stenosis rising over time.

Patients who develop coronary artery aneurysms are given ongoing therapy to prevent clots, which may include aspirin, dipyridamole, clopidogrel, warfarin, low-molecular-weight heparin, or any combination of these. The most common regimen for those with giant aneurysms is a combination of low-dose aspirin and warfarin to maintain an international normalized ratio of 2 to 2.5, though clinicians may prefer treating very young children with subcutaneous, low-molecular-weight heparin and aspirin, which is easier to regulate in this group.[5] If follow-up stress imaging suggests ischemia or stenosis, patients may undergo balloon angioplasty, vascular stenting, or coronary artery bypass surgery.[5]

their clinician if fever over 101°F (38.3°C) returns. Prolonged fever is strongly associated with coronary artery aneurysm, and a fever this high may signal the need for retreatment with IV Ig.

Emphasize the importance of follow-up cardiology appointments, which usually include a cardiac evaluation, laboratory work (complete blood count; ESR; and CRP, ALT, and albumin levels), and an echocardiogram. Infants and toddlers may require sedation for accurate echocardiography; review with parents the guidelines for presedation fasting and sedation recovery.

Kawasaki disease often causes children to develop a normocytic, normochromic anemia in the weeks after onset. As a result, children may be fatigued for several weeks. Although recovering children may need to take longer naps than they had in the past, encourage parents not to restrict activity during recovery but to let children regulate their own activity. They can often return to school a week or two after discharge, but they may benefit from a shorter school day initially.

Let parents know that irritability is often the last Kawasaki disease symptom to disappear, and that they may need to rely on other family members for support and respite. The child's behavior may be erratic and unpredictable for several weeks.

NURSES SHOULD MONITOR VITAL SIGNS CLOSELY DURING THE ACUTE PHASE OF KAWASAKI DISEASE, FREQUENTLY ASSESS HYDRATION, AND ENCOURAGE THE PATIENT TO DRINK FLUIDS, AVOIDING THOSE THAT ARE ACIDIC, SUGARY, OR CARBONATED.

## NURSING CONSIDERATIONS

Nurses should monitor vital signs closely during the acute phase of Kawasaki disease, recording temperatures immediately before administering aspirin. Frequently assess hydration and encourage the patient to drink fluids, avoiding those that are acidic, sugary, or carbonated and may exacerbate oral irritation. Initially, patients may prefer soft, bland foods. Decreased oral intake or excess fluid loss resulting from fever may require treatment with IV fluids.

Manage pain with age-appropriate distraction techniques and analgesics, other than ibuprofen, which has been shown to block aspirin's antiplatelet effects.[12] Child life specialists may be helpful in suggesting comfort measures.[13] For example, during the acute phase of illness, some children, like Jay, are photophobic and benefit from dim lighting. Topical, unscented creams or petroleum jelly may soothe peeling skin on the extremities; lip balms may help heal chapped lips.

**Patient teaching.** Before hospital discharge, teach parents what to expect over the next few weeks. Advise them to keep a log of their child's daily temperature for the first week or two after discharge and to call

Because IV Ig therapy may reduce the efficacy of live vaccines (such as those for measles, mumps, rubella, and varicella), the Centers for Disease Control and Prevention recommends delaying such immunizations for 11 months after IV Ig infusion.[14] Other immunizations may be resumed once the child has returned to health.

All children diagnosed with Kawasaki disease are prescribed low-dose aspirin for four to six weeks after hospital discharge. In the absence of aneurysms, aspirin therapy is discontinued after the week-4-to-week-6 echocardiogram. Because aspirin increases bleeding risk, children receiving aspirin therapy should avoid activities associated with physical injury (such as contact sports).

Children with coronary artery dilatation or aneurysms may be prescribed long-term antiplatelet or anticoagulation therapy. These children should continue to avoid contact sports and activities associated with injury. In addition, those using long-term aspirin therapy should be given an annual flu vaccine to reduce the risk of Reye's syndrome. Instruct parents of such

children to notify their pediatrician if their child is exposed to chickenpox or influenza or develops flulike symptoms while taking aspirin.[15]

Children who receive warfarin therapy should wear a MedicAlert bracelet denoting that fact. Parents and caregivers of these children should be taught about the medication's administration, requisite follow-up, and associated adverse effects (especially the elevated risk of bleeding). They should receive instruction in cardiopulmonary resuscitation techniques and be advised to report any new signs and symptoms or indications of myocardial ischemia to the child's cardiologist. In young children, signs and symptoms of myocardial ischemia are often nonspecific and may include nausea, vomiting, pallor, inconsolability, and chest pain or pressure.

## LONG-TERM OUTLOOK

Although some children with Kawasaki disease have such long-term cardiovascular sequelae as coronary artery aneurysms and stenosis, the vast majority return to full health.[16] Prompt diagnosis and treatment, along with appropriate follow-up, are critical in minimizing cardiovascular complications.

All children who have had Kawasaki disease should be evaluated for risk factors such as high cholesterol and hypertension. Patients without cardiovascular morbidity can be encouraged to live an active lifestyle without exercise restrictions; those who develop cardiovascular complications need to be tested more frequently. The AHA recommendations outline exercise restrictions and standard follow-up testing based on five levels of risk, ranging from "no coronary artery changes at any stage of illness" to "coronary artery obstruction."[5]

A week after discharge, Jay returned to the cardiology clinic. His fingertips were peeling, and his skin was still very dry. His parents reported that although he had remained afebrile over the week, he seemed to have tantrums at the least provocation and frequently awoke at night wanting to sleep with them. His movements tended to be stiff in the morning but improved as the day went on. The nurse reassured them that such behavior was not unusual in the early stages of recovery and encouraged them to continue gently setting limits, returning him to his own bed at night, and using distraction to quell tantrums. His first postdischarge echocardiogram was normal.

As Jay began to feel better over the next several weeks, his parents became more comfortable setting limits and Jay's behavior improved. Four weeks later, when the family returned for Jay's second follow-up visit, his parents felt that he was "90% back to normal." They asked about the horizontal grooves that had appeared across Jay's fingernails and toenails (Beau's lines) and were assured that this is a typical finding in the convalescent phase of Kawasaki disease (after the majority of clinical features have resolved but laboratory studies still show evidence of inflammation). Because Jay's echocardiogram was once again normal, aspirin therapy was discontinued six weeks after illness onset. ▼

**Keywords:** Kawasaki disease, Kawasaki syndrome, mucocutaneous lymph node syndrome

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