

# Recognizing Atypical Kawasaki Disease in Infants

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CAMBRIDGE, MASS. — Infants with prolonged, unexplained fever and laboratory evidence of systemic inflammation should undergo echocardiography to look for coronary changes suggestive of Kawasaki disease, even if there are no other clinical signs of the generalized vasculitis, according to Dr. Marisol Figueira. “The manifestations of Kawasaki dis-

ease in infants are often subtle, and many times infants with the condition do not meet the full diagnostic criteria,” Dr. Figueira said at a conference on pediatric infectious diseases.

Typically, a diagnosis of Kawasaki disease requires the presence of unexplained fever and the presence of at least four of the following five principal features: changes in the extremities such as redness, swelling, induration of the hands and feet, and, later, peeling of the fingers and toes;

polymorphic rash involving the trunk and extremities; nonexudative conjunctivitis; redness and swelling of the lips and oral cavity; and cervical adenitis.

Associated laboratory findings often include elevated erythrocyte sedimentation rate and C-reactive protein levels in the acute phase, and may include any of several nonspecific findings, such as neutrophilia, mild anemia, hypoalbuminemia, elevated serum immunoglobulin E levels, thrombocytosis, proteinuria, and sterile

pyuria, said Dr. Figueira of Boston University Medical Center.

Infants with Kawasaki disease—particularly those younger than 6 months—may present with only unexplained fever, or the fever may be accompanied by only two or three of the clinical features of classic Kawasaki disease. In fact, reported Dr. Figueira, “such cases of incomplete or atypical Kawasaki disease, in which patients have fewer than four of the five principal features, have been increasingly reported in infants.”

Recognizing incomplete Kawasaki disease in infants is particularly challenging, because the clinical features that do present in this population tend to be more subtle than those seen in older children and are often mistaken for symptoms of other conditions, Dr. Figueira said at the conference, which was sponsored by Boston University, PEDIATRIC NEWS, and FAMILY PRACTICE NEWS.

For example, “an infant with fever, rash, and cerebral spinal fluid pleocytosis may

be misdiagnosed with viral meningitis. The presence of rash and mucosal changes that follow often are mistaken for a reaction to antibiotics administered for presumed bacterial lymphadenitis, and sterile pyuria may be mistaken for a partially treated urinary tract infection,” she said.

To avoid misdiagnosis, physicians should make Kawasaki disease part of the differential diagnosis for all infants with unexplained prolonged fever and any of the characteristics consistent with the condition, Dr. Figueira said. In such patients, she noted, “laboratory values, particularly for the acute phase reactants, should be obtained.”

If these values are elevated—a C-reactive protein measure of 3 mg/dL or higher and an erythrocyte sedimentation rate greater than 40 mm/hour—an echocardiogram should be obtained to check for coronary changes that might indicate Kawasaki disease, Dr. Figueira said.

“Although aneurysms rarely form before day 10 of illness, perivascular brightness, ectasia, and lack of tapering of the coronary arteries can be seen in the acute stage of the disease,” she said. Other potential findings include decreased left ventricular contractility, mild valvular regurgitation, and pericardial effusion.

If an echocardiograph is positive for these changes, treatment for Kawasaki disease, which generally includes intravenous immunoglobulin and aspirin therapy, should begin immediately to preclude long-term, permanent coronary damage, Dr. Figueira said.

“If the echo is negative but the fever or any symptoms continue to persist, a repeat echo consult may be needed.” ■

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HOSPITALIZATIONS  
EVERY YEAR.<sup>1</sup>**

#### AN UNAVOIDABLE DISEASE...<sup>2</sup>

- The most common cause of severe gastroenteritis in infants and young children in the United States<sup>2</sup>
- Greatest risk for severe disease occurs primarily in young children between 6 and 24 months of age.<sup>3</sup>

#### WITH UNPREDICTABLE CONSEQUENCES<sup>2,4</sup>

- No way to predict which infants will suffer severe disease<sup>4</sup>
- Potential for rapid deterioration in cases in which severe vomiting occurs<sup>5</sup>
- Responsible for an estimated 500,000 physician visits,<sup>1</sup> 70,000 hospitalizations,<sup>1</sup> and 160,000 ER visits<sup>6</sup> among children <5 years of age every year in the United States<sup>1,6</sup>
- Responsible for an estimated 100 deaths per year among children <5 years of age in the United States—an average of nearly 2 deaths per week<sup>7</sup>

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Rotavirus hospitalizations