May 31, 2001

TO:                  ALL COUNTY CALIFORNIA CHILDREN’S SERVICES (CCS)
ADMINISTRATORS AND MEDICAL CONSULTANTS, AND STATE
CHILDREN’S MEDICAL SERVICES (CMS) STAFF

SUBJECT:    KAWASAKI DISEASE

Background

Kawasaki Disease is a generalized vasculitis of unknown etiology. It is a leading cause
of acquired heart disease in children in the United States. The majority of children
diagnosed with Kawasaki Disease is less than five years of age.

Diagnosis of Kawasaki Disease is made when there is:

1. Fever of at least five days duration AND

2. Presence of four of the following five principle features:
   - Bilateral conjunctival injection
   - Polymorphous, generalized rash
   - Changes of oral mucosa and lips (erythema, strawberry tongue, fissured lips)
   - Changes in peripheral extremities (erythema, swelling, induration)
   - Cervical lymphadenopathy (greater than 1.5 cm diameter); AND

3. Other known disease processes are excluded.

The vasculitis of Kawasaki Disease is most severe in medium-sized arteries, such as
coronary arteries. Arterioles, capillaries, veins, and large arteries are also affected.
Complications of the circulatory system can include coronary artery aneurysms,
valvulitis, myocarditis (present in greater than 50 percent of patients during the acute
phase, usually in absence of congestive heart failure), pericarditis with pericardial
effusion (present in 25 percent of patients during acute phase), aneurysms of peripheral
vasculature, or peripheral gangrene.
Complications are also seen in the musculoskeletal system (arthritis and arthralgias), gastrointestinal system (hepatitis and hydrops of gallbladder), respiratory system (pneumonia), and central nervous system (aseptic meningitis).

Patients who have evidence of coronary artery abnormalities may require more intensive monitoring by a cardiologist as well as continued treatment with aspirin.

**Policy**

Kawasaki Disease is a medically-eligible condition for the CCS program as per Title 22, California Code of Regulations, Section 41844 (Diseases of the Circulatory System) as it is a generalized vasculitis.

Children are **not** eligible for a diagnostic evaluation to determine the presence of Kawasaki Disease.

**Policy Guidelines**

1. A child who has been diagnosed with Kawasaki Disease, according to the criteria given above, shall be determined medically eligible for the CCS program.

2. As long as all other program eligibility requirements are met, the CCS program shall authorize the hospital stay for the treatment of the acute illness, starting the date the progress note confirms the diagnosis of Kawasaki Disease.

3. If at the time of hospital discharge, the child with Kawasaki Disease has coronary artery or other circulatory abnormalities, a CCS-approved Cardiac Special Care Center (SCC) shall be authorized, as well as a primary care physician in the child’s community to work in conjunction with the Cardiac SCC.

4. If at the time of hospital discharge, the child with Kawasaki Disease has no coronary artery or other circulatory abnormalities, a CCS-paneled cardiologist shall be authorized for treatment services for a period of one year. Authorization of continuing care shall be discontinued after this time period if there is still no evidence of cardiac abnormalities.

If you have any questions, please contact your Regional Office Medical Consultant.

Maridee A. Gregory, M.D., Chief
Children’s Medical Services Branch