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GOVERNOR

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TO: All County California Children's Services Program Administrators,
Medical Consultants, and Integrated Systems of Care Division Staff

SUBJECT: Kawasaki Disease – Updated

I. PURPOSE

The purpose of this Numbered Letter (N.L.) is to update the California Children's Services (CCS) Program medical eligibility guidelines for children who have been diagnosed with Kawasaki disease (KD) and incomplete Kawasaki disease (previously called atypical KD).

II. BACKGROUND

KD is a common form of vasculitis of childhood, which leads to coronary artery aneurysms in about 25% of untreated cases. It is a leading cause of acquired heart disease in children in developed countries.¹

KD is an acute, self-limited process characterized by systemic inflammation in medium-sized arteries and in multiple organs and tissues during the acute febrile phase, affecting the gastrointestinal system, lungs, meninges, lymph nodes, and urinary tract. Complications of the circulatory system include coronary artery aneurysms, myocarditis, pericarditis, valvulitis and valve dysfunction, aneurysms of peripheral vasculature, and peripheral gangrene.¹ The inflammation of the coronary arteries results in the most clinically significant outcomes.

III. POLICY

A. Diagnosis of classic KD is made when other known disease processes have been excluded and a patient exhibits:

¹ AHA Scientific Statement: Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease, Circulation, online March 29, 2017.

1. Fever which is typically high, spiking, and remittent, of at least five days duration with at least four of the following five principal clinical findings:
 - a. Erythema and cracking of lips, strawberry tongue, and/or erythema of oral and pharyngeal mucosa.
 - b. Bilateral bulbar conjunctival injection without exudate.
 - c. Maculopapular or diffuse erythematous rash.
 - d. Erythema and edema of the hands and feet in acute phase and/or periungual desquamation in subacute phase.
 - e. Cervical lymphadenopathy (greater than 1.5 cm diameter), usually unilateral.

OR

2. Fever of four days duration with at least four principal clinical findings, when redness and swelling of the hands and feet are present.

OR

3. Three days of fever and at least four principal clinical findings, if the diagnosis is made or confirmed by a subspecialist with experience treating patients with KD.

B. Diagnosis of incomplete KD, sometimes referred to as atypical KD, may be made by a specialist in the diagnosis and treatment of KD when the client has less than five days of fever and two or three compatible criteria, or in an infant, greater than seven days of unexplained fever and the diagnosis is supported by laboratory tests and/or echocardiographic findings, as outlined in the American Heart Association (AHA) Scientific Statements. Current guidance can be found in figure 3 of the 2017 AHA Scientific Statement, pages e936 – e938.¹

C. Clients are medically eligible for the CCS program when all of the following are met:

1. The diagnosis fits the criteria stated in the 2004 American Academy of Pediatrics (AAP) and 2017 AHA endorsed guidelines.²

² American Academy of Pediatrics and American Heart Association (AHA) endorsed guidelines, Pediatrics 2004:114,1708.

2. The diagnosis of classic KD is made by a CCS paneled physician or the diagnosis of incomplete KD is made by a CCS-paneled Cardiology, Infectious Disease, or Rheumatology subspecialist.
 3. The client has a CCS-eligible condition that is a sequela of KD complications including, but not limited to, myocarditis, pericarditis, and valvulitis.
- D. In County Organized Health System (COHS) counties that have implemented the Whole Child Model (WCM) program, clients with KD shall receive medically necessary specialty services through the Medi-Cal managed care plan contracted to provide services to CCS clients.
- E. Authorization of services:
1. The diagnostic evaluation to determine the presence of KD is not a benefit of CCS; requests for diagnostic services shall not be authorized.
 2. The initial treatment authorization shall be to any CCS approved hospital or special care center (SCC).
 3. Reauthorization for classic KD shall be to a CCS SCC or CCS-paneled provider with expertise in vasculitis for up to one year to evaluate for coronary artery aneurysms or dilatation and as long as necessary for management when these conditions are present.
 4. For incomplete KD, reauthorization shall be to a CCS-paneled Cardiology, Infectious Disease, or Rheumatology subspecialists for up to one year.
- F. For clients not in the WCM:
1. The county CCS program shall authorize the hospital stay during which the diagnosis of CCS-eligible KD is made.
 2. If at the time of hospital discharge, the child with KD has coronary artery or other circulatory abnormalities, the county CCS program shall authorize CCS approved cardiac SCC to provide evaluation and management. A primary care physician in the child's community may be authorized to work in conjunction with the cardiac SCC in the evaluation and management.
 3. If at the time of hospital discharge, the child with KD does not have coronary artery or other circulatory abnormalities, the county CCS program shall authorize a CCS-paneled cardiologist for treatment services for a

period of one year. Authorization of continuing care shall be discontinued at the end of this time if there is no evidence of coronary artery or other circulatory abnormalities.

If you have any questions regarding this Numbered Letter, please contact the ISCD Medical Director at ISCD-MedicalPolicy@dhcs.ca.gov.

Sincerely,

ORIGINAL SIGNED BY

Roy Schutzengel
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Integrated Systems of Care Division