DHCS
CALIFORNIA DEPARTMENT OF
HEALTH CARE SERVICES
Michelle Baass | Director

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TO: All County California Children's Services Program Administrators and Medical Directors

SUBJECT: CCS Program Coverage of Kawasaki Disease

I. PURPOSE

The purpose of this Numbered Letter (NL) 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 KD.

II. BACKGROUND

KD is a vasculitis syndrome that often presents in young children and, when untreated, often leads to coronary artery aneurysms and other cardiovascular sequelae.[1] It is a leading cause of acquired heart disease in children in developed countries.[2]

KD is characterized by fever and characteristic clinical signs during the acute phase. These signs, or principal findings, are bilateral conjunctival injection, erythema of lips, tongue, oral and pharyngeal mucosa, erythema and edema of hands and feet followed by desquamation, and cervical lymphadenopathy, usually unilateral. Individuals with a fever and at least four features have KD. Individuals with a fever and less than four of the principal clinical findings have atypical Kawasaki disease. Following the acute and subacute phases, affected individuals may develop long-term complications of the circulatory system including aneurysms in coronary and non-coronary arteries, myocarditis, pericarditis, valvulitis and valve dysfunction, and peripheral gangrene. Coronary artery aneurysms are the most clinically significant sequelae. Prompt treatment with intravenous gammaglobulin (IVIG) is strongly

[1] Pilania RK, et al. Cardiovascular Involvement in Kawasaki Disease Is Much More Than Mere Coronary Arteritis. Front Pediatr. 2020 Sep 24;8:526969. DOI: 10.3389/fped.2020.526969. PMID: 33072669; PMCID: PMC7542237.
[2] McCrindle BW, et al. Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease: A Scientific Statement for Health Professionals From the American Heart Association. Circulation. 2017 Apr 25;135(17):e927-e999. DOI: 10.1161/CIR.0000000000000484. Epub 2017 Mar 29. Erratum in: Circulation. 2019 Jul 30;140(5):e181-e184. DOI: 10.1161/CIR.0000000000000703. PMID: 28356445.

recommended for all individuals with KD.[3] A recent meta-analysis did not identify any CCS-eligible sequelae apart from those related to the cardiovascular system.[4] Guidelines for diagnosis and treatment of Kawasaki disease detail specific clinical and laboratory findings, as well as consensus-based recommendations. The guidelines were initially released in 2004 by the American Academy of Pediatrics and the American Heart Association (AHA) and recently updated in 2021 by the American College of Rheumatology (ACR) and the Vasculitis Foundation (VF).[5]

III. POLICY

A. KD as a CCS Benefit:

1. As a medium-vessel vasculitis, KD is a CCS medically eligible condition under Title 22, §41518.2(c) when the diagnosis fits the criteria stated in the clinical guidelines referenced above.[6]

2. Cardiovascular sequelae of KD, including coronary artery dilatation and aneurysms sequelae are CCS-eligible.

3. CCS shall authorize services related to the treatment of KD when the diagnosis of classic or incomplete KD is made by a CCS-paneled subspecialist in one of these subspecialties: Pediatric Cardiology, Pediatric Infectious Disease, Pediatric Rheumatology or Pediatric Immunology, or made by a paneled CCS physician and confirmed through consultation with a CCS-paneled specialist in one of the four subspecialties listed above.

B. Service Authorization:

1. The diagnostic evaluation to determine whether KD is present is only a CCS benefit if a diagnosis of KD or atypical KD is made; CCS Program must not authorize requests for diagnostic services or inpatient hospitalization until after KD or incomplete KD is diagnosed.

2. The treatment of the hospitalization for the acute phase of KD is a CCS

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[3] Gorelik, M., et al. 2021 American College of Rheumatology/Vasculitis Foundation Guideline for the Management of Kawasaki Disease. American College of Rheumatology. 2021. Vol. 0, No. 0, Month 2022, pp 1–11. DOI 10.1002/art.42041.

[4] Yee, J., et al. The Long-term Cardiac and Noncardiac Prognosis of Kawasaki Disease: A Systematic Review. 2022. *Pediatrics* (2022) 149 (3): e2021052567.DOI: https://doi.org/10.1542/peds.2021-052567

[5] Gorelik, M., et al. 2021 American College of Rheumatology/Vasculitis Foundation Guideline for the Management of Kawasaki Disease. American College of Rheumatology. 2021. Vol. 0, No. 0, Month 2022, pp 1–11. DOI 10.1002/art.42041.

[6] McCrindle BW, et al. Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease: A Scientific Statement for Health Professionals From the American Heart Association. Circulation. 2017 Apr 25;135(17):e927-e999. DOI: 10.1161/CIR.0000000000000484. Epub 2017 Mar 29. Erratum in: Circulation. 2019 Jul 30;140(5):e181-e184. DOI: 10.1161/CIR.0000000000000703. PMID: 28356445.

benefit.

3. CCS Program must authorize diagnosis and treatment of acute KD to any CCS-approved hospital or special care center (SCC) with available CCS-paneled physicians in one or more of the subspecialties named in III.A.3.

4. In cases of KD with acute vascular dilatation or other cardiovascular or circulatory abnormalities, the CCS Program shall authorize a CCS-paneled cardiologist and/or other CCS-paneled specialists with expertise in KD to evaluate and manage the client for coronary artery aneurysms or other cardiovascular or circulatory abnormalities and to manage significant sequelae in an ongoing manner as per the KD guidelines.

5. In cases of acute KD without acute vascular dilatation or other cardiovascular or circulatory abnormalities, the CCS Program shall authorize a CCS-paneled cardiologist and/or other CCS-paneled specialists with expertise in KD to evaluate the client for coronary artery aneurysms or other cardiovascular or circulatory abnormalities for up to twelve months following the acute KD, and to manage significant sequelae in an ongoing manner as per the KD guidelines.

6. For clients with KD and incomplete KD, reauthorization shall be to a CCS-paneled cardiology, infectious disease, or rheumatology subspecialist for up to one year.

7. Authorization of continuing monitoring of clients without evidence of coronary dilatation shall be discontinued twelve months after the acute phase of KD unless the client is determined to be high risk for late complications.

IV. POLICY IMPLEMENTATION

The CCS Program must process service authorization requests as follows:

A. For clients residing in Classic CCS independent counties, the county CCS Program reviews and adjudicates all requests.

B. For clients residing in Classic dependent counties, the Department of Health Care Services reviews and adjudicates all requests.

C. For clients residing in WCM counties, the Medi-Cal managed care plan is responsible for ensuring that clients receive medically necessary specialty services equivalent to those services available under ‘classic’ CCS.

If you have any questions regarding this NL, please contact ISCD-MedicalPolicy@dhcs.ca.gov.

Sincerely,

ORIGINAL SIGNED BY

Cortney Maslyn, Chief
Integrated Systems of Care Division
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