

KAWASAKI DISEASE CLINICAL PATHWAY

EXECUTIVE SUMMARY

Physician Owner(s): Aleisha Nabower, MD



Primary Objective

Increased diagnostic quality of echocardiograms performed in the evaluation of and clinical management of Kawasaki disease and outpatient follow-up.

Kawasaki Disease Diagnosis

Kawasaki Disease Clinical Features

- Mucositis-strawberry tongue
- Nonpurulent conjunctivitis
- Erythematous rash
- Extremity changes (swelling/peeling)
- Cervical lymphadenopathy ≥ 1.5 cm

Incomplete Kawasaki Disease Lab Criteria

- Anemia for age
- Platelet $\geq 450,000$ after 7th day of fever
- WBC $\geq 15,000/\text{mm}^3$
- Albumin ≤ 3.0 g/dL
- Elevated ALT
- Urine > 10 BC/hpf

Suspicion of Complete Kawasaki

- 5 days of fever + 4 clinical features
OR
- 4 days + 5 clinical features

Suspicion of Incomplete Kawasaki

- 5-10 days of fever + 2-3 clinical features
OR
- Patient < 6 months with > 7 days of fever, regardless of clinical features

Recommendations

Complete Kawasaki: Consult infectious disease and cardiology (Blaney, 2022). Order echocardiogram-discuss mild sedation. Obtain baseline ECG.

Suspected Incomplete Kawasaki: Obtain CRP and ESR, if above cut-offs obtain CBC, CMP, and UA; consult infectious disease and cardiology (Blaney, 2022), obtain echocardiogram-discuss sedation, obtain baseline ECG.

For diagnosis of KD: Give 2 g/kg IVIG (max 70 g) and medium-dosed aspirin 30-50mg/kg/day divided Q6H, monitor for at least 36 hours. Once afebrile x 12 hours may transition to low dose ASA 3-5mg/kg/day. While there are recommendations for the addition of steroids in patients at high risk for refractory disease or coronary artery aneurysms (largely based out of Japanese literature, however, the predictive models have

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not been found to be reliable in other populations (Blaney et al., 2022; Kuniyoshi et al., 2023)

For Refractory: No current standard (Buda, Friedman-Gruszczynska, & Ksiazzyk, 2021; Scherler et al., 2022).

Discharge: Cardiology follow-up with echo in 2 and 6 weeks, offer inactivated flu vaccine, live vaccine and fever monitoring education completed.

Rationale

Safety and Quality: In 2017 96% of echocardiograms done for Kawasaki disorder at our institution were not complete, with the majority being medically cleared without ever having a cardiology visit. Insufficient quality of echocardiograms causes a safety concern for our patients as providers may be reassured incorrectly. Additionally, an initial normal echocardiogram does not exclude the diagnosis of Kawasaki disease or the development of coronary aneurysms long term. 73% of patients discharged from our institution did not receive the recommended 2-week follow-up echocardiogram. The AHA has specific guidelines regarding counseling of patient's who have had Kawasaki disease that vary by echocardiogram results. Having a planned cardiology follow-up allows for individualized, specialized counseling. In 2017 30% of patients did not have a cardiology follow-up visit. With the implementation of the above pathway 98% of echocardiograms between 2019-2022 were complete, with 81% of patients having a timely 2-week echocardiogram obtained.

Patient/Family Satisfaction: If abnormalities are noted on follow-up echocardiograms there can be difficulty obtaining a same-day cardiology appointment for management if not scheduled ahead of time leading to parental anxiety and potentially a delay in treatment. Additionally, if echocardiograms are not qualitatively sufficient there is little intervention that can be planned the same-day requiring either an additional visit or admission if sedation is required to obtain a diagnostically sound echocardiogram.

Initial treatment of Kawasaki disease

Patients meeting complete or incomplete KD criteria should receive 2 g/kg IVIG (max 70 g) as a single infusion as soon as possible after diagnosis, ideally within 10 days of fever onset. Additionally, they should receive moderate to high-dose ASA 30-50 mg/kg/day divided Q6H until the patient is afebrile. The ESR is accelerated by IVIG therapy and should not be used to assess response to IVIG therapy. Once afebrile patients with no coronary artery involvement may transition to low-dose aspirin to be continued for 6-8 weeks (McCord et al., 2017; Scherler et al., 2022).

Laboratory evaluation for suspected incomplete Kawasaki:

CLINICAL



EFFECTIVENESS

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Updated 9/2023

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Patients with 5 or more days of fever and 2 or 3 clinical criteria of Kawasaki disease OR infants with fever 7 days or more without other explanation should have a screening CRP and ESR obtained. If negative, they should have serial re-evaluations if the fever persists. If positive (CRP ≥ 3.0 mg/dL or ESR ≥ 40 mm/hr) obtain CBC, CMP, UA and echocardiogram for further evaluation. While antibodies against SARs-CoV2 were initially helpful in distinguishing multisystem-inflammatory syndrome (MIS-C) from Kawasaki disease, they have become less useful now that a majority of the population has been infected with SARs-CoV2. Clinical features such as older patient age than is typical for Kawasaki (< 5 years), presentation in shock, or with respiratory and gastrointestinal symptoms, the presence of lymphopenia or thrombocytopenia, and evidence of myocardial involvement on echocardiogram or with elevated pro-BNP and troponin are more common in MIS-C. While coronary artery involvement and the presence of conjunctivitis are more common in Kawasaki disease (McCrindle et al., 2017; Tong et al., 2022).

Using sedation for Kawasaki echocardiogram

Detailed echocardiographic imaging is highly sensitive and specific for detecting abnormalities of the proximal coronary arteries. The initial echocardiogram should be performed as soon as the diagnosis is suspected. As imaging can be compromised in an uncooperative child, sedation is frequently needed for those less than 3 years of age and may also be required in older, irritable children. If a poor-quality initial echocardiogram is obtained because sedation was not administered, a sedated study should be repeated as soon as possible within 48 hours after diagnosis and initial treatment. Routine sedation (including oral chloral hydrate, oral or intranasal midazolam, IV ketamine, and intranasal dexmedetomidine) use has been associated with higher rates of visualization of all coronary arteries specifically with regards to the distal LAD and RCA (McCrindle et al., 2017).

An initial echocardiogram in the first week of illness is typically normal and does not rule-out the diagnosis. Coronary artery assessment should include quantitative assessment of the internal vessel diameters. Normalization of dimensions for BSA using Z scores allows for standardization and comparisons across time. According to the Pediatric Heart Network, there is no set Z score gold standard, but there should be a consistent formula (Lim et al., 2021; Lorenzoni et al., 2022). The high-risk formula for CAA is as follows: a Z score of greater than or equal to 2.5 (25–27) for the left anterior descending or right coronary artery at the time of the initial echocardiography and age <6 months.

For uncomplicated patients, echocardiography should be repeated both within 1-2 weeks and 4-6 weeks after treatment (Gu, Song, & Bai, 2019; Lim et al., 2021; Lorenzoni et al., 2022; Margossian et al., 2011; McCrindle et al., 2017).

Outpatient follow-up for Kawasaki disease:

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Patients with no evidence of coronary artery involvement at 1-2 weeks and 4-6 weeks post-treatment may discharge from cardiology assessment between 4 weeks and 12 months. They should receive cardiovascular risk factor assessment at 1 year. If coronary artery changes are noted, follow up with cardiology is recommended along varying timelines according to type of involvement which can be determined at the 4-6 week follow up visit (McCrindle et al., 2017).

Metrics

Outcome

- Monitor the proportion of patients that attend their scheduled cardiology appointment between 1-3 weeks of discharge
- Monitor the proportion of patients receiving correct IVIG dosing (2 g/kg or Max 70 g)
- Monitor the percentage of patients receiving appropriate dosing of aspirin at discharge (3-5 mg/kg/day)

Process

- Monitor the percentage of patients receiving an appropriate moderate dose (30-50 mg/kg/day) of aspirin while inpatient (day 0-2)
- Monitor the number of total non-diagnostic echocardiograms performed compared to total echocardiograms performed

Balancing

- Monitor the average length of stay
- Monitor the proportion of patients readmitted within 14 days for a fever

Team Members

Champion: Aleisha Nabower, MD (Pediatric Hospital Medicine)

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Evidence



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