Clinical Pathway
Executive Summary

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Children's HOSPITAL & MEDICAL CENTER

We know children.

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

Recommendations
Complete Kawasaki: Consult infectious disease and cardiology. 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, obtain echocardiogram-discuss sedation, obtain baseline ECG**. For diagnosis of KD:** Give 2 mg/kg IVIG and high dosed ASA (80-100 mg/kg/day divided every 6 hours) or medium dosed ASA (30-50 mg/kg/day divided every 6 hours), monitor for at least 36 hours. Once afebrile x 12 hours may transition to low dose ASA 3-5mg/kg/day. **Discharge:** Cardiology follow-up with echo in 2 and 6 weeks, offer inactivated flu vaccine, live vaccine and fever monitoring education completed

Rationale (Safety, Quality, Cost, Delivery, Engagement, & Satisfaction)

Safety and Quality: In 2017 96% of echocardiograms done for Kawasaki disorder at our institution are not complete, with the majority being medically cleared without ever having a cardiology visit**.** Insufficient quality of echocardiograms cause a safety concern for our patients as providers may be reassured incorrectly. Additionally, an initial normal echocardiogram does not exclude the diagnosis of Kawaski disease or the development of coronary aneurysms long term. 73% of patients discharged from our institution do 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.

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.

Implementation Items
1.Create Kawaski pathway including consult for cardiology prior to obtainment of echocardiogram to facilitate discussion for need for sedation and insurance that adequate images are obtained at the time of the initial exam.

Date Last Reviewed: 12/2015

2. Update KD orderset to match 2017 AHA guidelines.
3. Create/facilitate structured outpatient follow-up with cardiology and echocardiograms for patients diagnosed with KD.

Metrics Plan

1. Non-diagnostic echocardiograms compared to total echocardiograms performed 2. Percentage of patient's scheduled for cardiology follow-up with echocardiogram at discharge. 3. Percentage receiving appropriate aspirin dosing inpatient and at discharge. 4. Percentage receiving correct IVIG dosing within 10 days of fever.

Evidence

Laboratory evaluation for suspected incomplete Kawasaki:

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 ≥40mm/hr) obtain CBC, CMP, UA and echocardiogram for further evaluation.

McCrindle BW, Rowley AH, Newburger JW, et al. (2017) Diagnosis, treatment, and long-term management of Kawasaki disease: a scientific statement of health for professionals from the American Heart Association. Circulation. 135:e927–e999/10.1161/CIR.0000000000000484

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.

An initial echocardiogram in the first week of illness is typically normal and does not rule-out the diagnosis. Coronary artery assessment should included quantitative assessment of the internal vessel diameters. Normalization of dimensions for BSA using Z scores allows for standardization and comparisons across time. For uncomplicated patients, echocardiography should be repeated both within 1-2 weeks and 4-6 weeks after treatment.

Date Last Reviewed: 12/2015

Gu HB, Song YA, Bai J. (2019). Median effective dose of intranasal Dexmedetomidine for transthoracic echocardiography in children with Kawasaki Disease who have a history of repeated sedation. Medical Science Monitor. 25. 381-388. 10.12659/MSM.912517.

Lorenzoni R, Choi J, Choueiter, N, Munjal I, Katyal C, Stern K. (2018). Predictors of inadequate initial echocardiography in suspected Kawasaki disease: Criteria for sedation. Congenital Heart Disease. 13. 10.1111/chd.12598.

Margossian R, Minmin L, Minich L, Bradley T, Cohen M, Li J, Printz B, Shirali G, Sleeper L, Newburger J, Colan, S. (2011). Predictors of coronary artery visualization in Kawasaki Disease. Journal of the American Society of Echocardiography : official publication of the American Society of Echocardiography. 24. 53-9. 10.1016/j.echo.2010.10.015.

McCrindle BW, Rowley AH, Newburger JW, et al. (2017) Diagnosis, treatment, and long-term management of Kawasaki disease: a scientific statement of health for professionals from the American Heart Association. Circulation. 135:e927–e999/10.1161/CIR.0000000000000484

Initial treatment of Kawasaki disease:

Patients meeting complete or incomplete KD criteria should receive 2g/kg IVIG as a single infusion as soon as possible after diagnosis, ideally within 10 days of fever onset. Additionally they should receive moderate **(**30-50 mg/kg/day) or high dose (80-100 mg/kg/day) ASA divided every 6 hours until the patient is afebrile. There is not yet strong evidence to recommend high versus moderate dosing, although the evidence supports using either initially when compared to low dose. 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.

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Outpatient follow-up for Kawasaki disease:

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 time lines according to type of involvement which can be determined at the 4-6 week follow up visit.

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Supporting Documents (Pathway, inclusion/exclusion criteria, definitions, algorithm) Kawasaki Pathway

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