



# Using the Electronic Medical Record to Correlate Kawasaki Disease Phenotypes With Clinical Outcomes

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**Background.** We sought to systematically standardize the documentation of clinical and laboratory features in Kawasaki disease (KD) on the day of initial treatment and correlate the presentation with clinical outcomes.

**Methods.** Kawasaki disease features and classification were documented by the attending physician using a standardized documentation tool on the day of treatment for KD, including confidence in the KD diagnosis on a 4-point scale. Incomplete KD was further classified using American Heart Association (AHA) criteria (sufficient or insufficient) and baseline echocardiogram data. We prospectively recorded intravenous immunoglobulin (IVIG) resistance, coronary artery abnormalities (CAAs), periungual peeling, and retrospectively identified subsequent diagnoses of autoimmune/inflammatory disease.

**Results.** From November 2012 to October, 2015, 162 patients were treated for KD: 105 with complete KD (Group 1), 7 with incomplete KD based on CAAs on day of KD diagnosis (Group 2), 23 with incomplete KD meeting AHA criteria (Group 3), and 27 with incomplete KD and insufficient AHA criteria (Group 4). Group 4 patients had lower baseline median C-reactive protein levels (Group 4 median 4.65 mg/dL [interquartile range {IQR}, 2.3–13.6] vs Group 1 median 8.0 mg/dL [IQR, 4.5–17], Group 2 median 13.9 mg/dL [IQR, 1.4–18.2], Group 3 median 13.3 mg/dL [IQR, 4.9–20.2]), and no coronary abnormalities developed, although 11% had IVIG resistance. Group 4 had higher rates of subsequent autoimmune/inflammatory conditions diagnosed (11.1% in Group 4 vs <5% for all others,  $P = .02$ ).

**Conclusions.** Standardized documentation and classification of KD features may be useful to correlate with clinical outcomes, including subsequent diagnosis of autoimmune/autoinflammatory disease. Among patients with incomplete KD who did not meet AHA criteria and had a normal baseline echocardiogram, the IVIG resistance rate may have been related to a lower likelihood of an accurate diagnosis of KD.

**Keywords.** coronary artery; incomplete Kawasaki disease; Kawasaki disease; periodic fever syndrome.

Kawasaki disease (KD) requires timely diagnosis and treatment to prevent development of coronary artery abnormalities (CAA) [1]. The clinical diagnosis of KD can be somewhat subjective, because clinical features can be transient and vary in intensity with some children lacking complete features (incomplete KD) [2]. Kawasaki disease requires multidisciplinary care, and those who may diagnose KD (ie, hospitalist, rheumatology, or infectious diseases [ID] specialist) and cardiologists involved in the management of patients. Care may sometimes be fragmented with multiple providers and transfers of care at different stages of illness and/or follow up, particularly for those patients with refractory and recurrent disease. This may lead to lack of communication of pertinent history. To address this need for continuity of care, in 2012, we created a multidisciplinary team for

the care of all KD patients at our hospital, which comprised ID and cardiology clinicians with rheumatology care as needed. The attending ID physicians, who are typically the clinicians who first evaluate KD patients in our hospital, provided standardized clinical and laboratory data documentation of criteria for diagnosis on the day of treatment with intravenous immunoglobulin (IVIG). Data documentation was performed using an electronic data collection form specifically designed for this condition.

The objective of this study was to correlate documented clinical and laboratory findings and variables on the day of treatment with IVIG with subsequent clinical outcomes (IVIG resistance, CAA, periungual peeling, and subsequent diagnosis of autoimmune/autoinflammatory conditions including periodic fever syndromes) based on the documented type of KD presentation on day of treatment.

## METHODS

The multidisciplinary KD clinic (staffed by ID and cardiology physicians) was created in 2012. The custom online form was created at the same time within the electronic medical record (EPIC software, Madison, WI) to detail the clinical and

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laboratory KD data, which were completed by the attending ID physician on the day of treatment with IVIG (Supplementary Figure 1) and on the day of patient discharge. Due to the subjective nature of some KD features, the level of confidence in the diagnosis of KD by the attending physician on the day of treatment was also recorded on a 4-point scale: 1 was the highest confidence in the KD diagnosis and 4 was the least confidence in the KD diagnosis (“possible KD and benefits of treatment outweigh risks of not treating”).

Children diagnosed with KD were classified as complete or incomplete KD. For those treated for incomplete KD, cases were further classified as to whether sufficient or insufficient American Heart Association (AHA) clinical and laboratory criteria were present [2]. At minimum, all KD patients had an echocardiogram at baseline, 2 weeks, and 5–8 weeks after diagnosis. Coronary artery abnormalities were defined as those with a z score  $\geq 2.0$ , based on the maximal internal diameters of the proximal right coronary artery or left anterior descending coronary artery. The z score of  $\geq 2.0$  was chosen because it is included as part of the echocardiographic considerations of treatment for incomplete KD in the published AHA guidelines when additional echocardiographic changes are present.

All inpatients diagnosed with acute KD were scheduled for follow up in the multidisciplinary KD clinic, staffed by both ID and cardiology clinicians, at a minimum of 2 weeks and 6–8 weeks post-IVIG treatment, and more frequently if required by their clinical case and/or coronary artery evolution. After the 6- to 8-week visit, patients assessed as uncomplicated were transitioned for follow up by cardiology care only. We used the data collection tool to prospectively record the presence of arthralgia/arthritis, presence of periungual peeling, and CAA during follow up at both the 2- and 6- to 8-week follow-up visits.

Intravenous immunoglobulin resistance was defined as persistent or recrudescing fever from 36 hours to 7 days after the completion of the first IVIG. Diagnosis of autoinflammatory/autoimmune syndromes, which included periodic fever syndromes, were recorded as of May 1, 2016 by retrospective review of the electronic medical record for all included patients, with Institutional Review Board approval. Autoinflammatory/autoimmune conditions included juvenile idiopathic arthritis, Bechet’s disease, systemic lupus erythematosus, polyarteritis nodosa, ankylosing spondylitis, dermatomyositis, and inflammatory bowel disease. Macrophage activation syndrome within 1 month after the KD diagnosis alone was not included because this is a known complication of KD. Periodic fever syndromes required either a clinical diagnosis by an ID or rheumatology attending physician of periodic fever, aphthous stomatitis, pharyngitis, and adenopathy (PFAPA) syndrome or another recurrent fever syndrome [3].

Kruskal-Wallis tests were used for comparisons, and a  $\chi^2$  test was used for proportions. Two-tailed  $P < .05$  was considered

significant. All analyses were performed using GraphPad Prism (GraphPad Software Inc., San Diego, CA).

## RESULTS

### Kawasaki Disease Patients Treated With Intravenous Immunoglobulin

From November 2012 to October 2015, our institution provided initial IVIG treatment to 162 patients with acute KD. The clinical classification tool was used in 143 (88.3%) cases. For those patients in whom the tool was not used, we retrospectively classified the patients into the appropriate KD type based on available data. Respiratory viral testing was performed in 156 (96.2%) of patients and 52 were positive (33.3%), 4 of whom had coinfections with more than 1 respiratory virus. The most common viruses identified were rhinovirus/enterovirus ( $n = 36$ ), followed by human adenovirus ( $n = 12$ ), parainfluenza ( $n = 2$ ), human metapneumovirus ( $n = 2$ ), influenza ( $n = 2$ ), respiratory syncytial virus ( $n = 1$ ), and coronavirus ( $n = 1$ ).

### Clinical and Laboratory Characteristics by Type of Kawasaki Disease

Clinical and laboratory characteristics by type of KD are presented in Table 1. Group 1 presented with complete KD ( $n = 105$ ), Group 2 presented with incomplete KD and baseline echocardiographic abnormalities ( $n = 7$ ), Group 3 presented with incomplete KD and sufficient AHA-defined criteria ( $n = 23$ ), and Group 4 ( $n = 27$ ) presented with incomplete KD and insufficient AHA criteria. There were 24 patients (14.8%) with IVIG resistance in the defined time period, and there were no significant differences in IVIG resistance among groups. The median C-reactive protein (CRP) was significantly lower in Group 4 than all others (median 4.65 vs 8.0–13.9 mg/dL in other groups) (Table 1), and respiratory viral detection was most frequent in Group 4, but this was not statistically significantly different from the others. Subsequent autoimmune/autoinflammatory diagnoses were most common in Group 4 (11.1% vs 0%–4.1% in other Groups).

### Timing of Coronary Abnormalities

Coronary artery abnormalities had been identified in 17 of 162 patients (10.4% of the cohort). Of these, CAA were present at baseline in 13 patients (76.5%). Four patients with normal baseline coronary arteries progressed to CAA (time to progression on echocardiogram was 7, 11, 13, and 15 days after baseline). All 4 patients had either IVIG resistance or required intensive care. Three patients required intensive care for fluid refractory hypotension. Additional autoimmune/autoinflammatory diagnoses documented in patients who were treated for acute KD are described below and listed by type of KD presentation.

### Complete Kawasaki Disease

A 6-year-old African American boy was treated after 15 days of fever. He had a history of oropharyngeal changes, rash,

**Table 1. Kawasaki Disease Patients: Clinical, Laboratory, and Echocardiogram Characteristics**

	Complete KD (n = 105) Group 1	Incomplete KD-Coronary Artery Abnormalities (n = 7) Group 2	Incomplete KD-Sufficient AHA Criteria (n = 23) Group 3	Incomplete KD-Insufficient AHA Criteria (n = 27) Group 4	P Value
<b>Clinical Characteristics</b>					
Age (years), median (IQR)	2.6 (1.6–4.8)	5.0 (2.2–8.4)	3.6 (1.9–5.4)	2.2 (0.81–6.4)	.413
Days of fever on treatment, median (IQR)	5 (4–7)	11 (6–24)	7 (5–8)	8 (6–13)	<.0001
Infants <6 months of age	6 (5.7%)	1 (16.7%)	2 (8.6%)	3 (11.1%)	.68
Bilateral conjunctival injection, n (%) by history	88 (83.8%)	5 (71.4%)	13 (56.5%)	18 (66.6%)	.02
Bilateral conjunctival injection, n (%) by PE	94 (89.5%)	4 (57.1%)	15 (65.2%)	15 (55.5%)	.001
Oral mucosal changes, n (%) by history	81 (77.1%)	4 (57.1%)	9 (39.1%)	10 (37.0%)	.001
Oral mucosal changes, n (%) by PE	99 (94.3%)	3 (42.8%)	16 (69.5%)	16 (59.3%)	.001
Polymorphous rash, n (%) by history	94 (89.5%)	4 (57.1%)	16 (69.5%)	20 (74.0%)	.01
Polymorphous rash, n (%) by PE	85 (80.9%)	1 (14.2%)	17 (73.9%)	19 (70.3%)	.001
Changes of extremities, n (%) by history	72 (68.6%)	3 (42.8%)	9 (39.1%)	11 (40.7%)	.007
Changes of extremities by PE	93 (88.6%)	3 (42.8%)	10 (43.5%)	9 (33.3%)	.001
Unilateral neck swelling, n (%) by PE	43 (40.9%)	1 (14.2%)	5 (21.7%)	4 (14.8%)	.02
IVIG resistance, n (%)	17 (16.2%)	1 (14.2%)	3 (13.0%)	3 (11.1%)	.92
Treatment after 10 days of fever, %	5 (4.7%)	4 (57.1%)	3 (13.0%)	12 (44.0%)	.001
Periungual peeling by 2-week follow-up	66 (62.8%)	3 (42.8%)	14 (60.8%)	10 (37.0%)	.09
<b>Laboratory Characteristics: Pretreatment</b>					
Respiratory viral detection rate: number positive/total tested, percentage	31/101, 30.6%	0/6, 0.0%	9/22, 40.9%	12/27, 44.4%	.15
Erythrocyte sedimentation rate, median (IQR)	51 (43–55)	54 (50–58)	54 (44–59)	49 (36–57)	.33
C-reactive protein, median (IQR)	8.0 (4.5–17)	13.9 (1.4–18.2)	13.3 (4.9–20.2)	4.65 (2.3–13.6)	.009
WBC, median (IQR)	13.4 (10.2–15.7)	11.4 (8.0–12.2)	15.3 (10.7–20.0)	14.1 (9.0–18.6)	.078
Hemoglobin median, median (IQR)	11 (10.1–11.7)	11.0 (10.2–13.0)	10.0 (9.4–11.3)	10.8 (10.3–11.4)	.294
Platelet, thousands per, median (IQR)	342 (286–417)	372 (295–539)	361 (293–435)	400 (290–543)	.297
Albumin, median (IQR)	3.6 (3.2–3.9)	4.0 (3.6–4.1)	3.2 (2.9–3.6)	3.7 (3.5–4.0)	.020
Alanine aminotransferase, median (IQR)	60 (30–129)	26 (17–66)	54 (26–79)	30 (24–68)	.06
Pyuria: (of number tested) % of patients with >10 WBC/HPF	33 of 84 (39.3%)	1 of 4 (25%)	10 of 23 (43.4%)	1 of 21 (4.7%)	.02
<b>Maximum Coronary Measurements by 6–8 Weeks After Initial KD Diagnosis</b>					
Normal coronary arteries	96 (91.4%)	0	22 (95.6%)	27 (100%)	NA
Maximum z score 2–2.5	2	0	1	0	NA
Maximum z score ≥2.5	7	7	0	0	NA
Persistent disease at 6-week visit	3	3	0	0	NA
Coronary abnormalities present on initial echocardiogram	6	7	0	0	NA
Subsequent autoinflammatory/autoimmune diagnoses after KD diagnosis	1 (0.9%)	0	1 (4.1%)	3 (11.1%)	.02
Arthralgia/arthritides within 6 weeks after KD diagnosis	12 (11.4%)	1 (16.6%)	4 (16.6%)	3 (11.1%)	.96

Abbreviations: AHA, American Heart Association; HPF, high-powered field; IQR, interquartile range; IVIG, intravenous immunoglobulin; KD, Kawasaki disease; NA, not applicable; PE, pulmonary embolism; WBC, white blood cell count.

and hand/feet and lymph node swelling. On physical exam, perianal peeling was noted with oropharyngeal erythema; he lacked scleral injection and hand and feet changes on that day. He was treated for likely complete KD in subacute phase of illness. The level of confidence in the KD diagnosis was rated as 2 on the 4-point scale. He had previously experienced intermittent episodes of unexplained fever. He never developed CAA but did have periungual peeling. Two years after the KD diagnosis, he was seen in rheumatology clinic with a history of intermittent fevers and arthralgia. Genetic testing was positive (heterozygous) for a variant in the *MEFV* gene, and he was diagnosed with familial Mediterranean fever with cessation of symptoms after starting colchicine treatment.

#### **Incomplete Kawasaki Disease: Meeting American Heart Association Criteria**

As previously published in more detail [4], a 5-year-old white male presented with 7 days of fever and a 1-month history of rash. The treating clinician categorized the lowest confidence in the KD diagnosis before treatment. He was subsequently diagnosed with systemic onset juvenile idiopathic arthritis with macrophage activation syndrome at 7 days after the KD diagnosis. He did not demonstrate any periungual peeling or CAA.

#### **Incomplete Kawasaki Disease: Non-American Heart Association Criteria**

A 10-year-old white boy was treated for KD with fevers for 9 days at the time of treatment. The physician categorized the

**Table 2. Clinical Outcomes by Clinical Confidence in the Kawasaki Disease Diagnosis**

Clinical Outcome/Characteristic	Confidence in Kawasaki Disease Diagnosis				P Value
	1 (highest) n = 69	2 n = 42	3 n = 22	4 (lowest) n = 9	
Periungual peeling	44 (63.7%)	26 (61.9%)	11 (50%)	3 (33.3%)	.04
Coronary artery abnormalities	11 (15.2%)	3 (7.1%)	0	0	.09
Intravenous immunoglobulin resistance	13 (18%)	5 (11.9%)	2 (9%)	2 (22%)	.57
Subsequent autoimmune diagnosis	0	1 (2.3%) Familial Mediterranean fever	3 (13.6%) Crohn's disease, systemic onset juvenile idiopathic arthritis, PFAPA	1 (11.1%) Systemic onset juvenile idiopathic arthritis	.01
Percentage of cases with complete KD	59 (85%)	28 (66.6%)	6 (27.2%)	2 (22.2%)	<.001

Abbreviations: KD, Kawasaki disease; PFAPA, periodic fever, aphthous stomatitis, pharyngitis, and adenopathy.

lowest confidence in the KD diagnosis, documented that uveitis was found on ophthalmologic exam, and prompted treatment for peeling found on the pads of the fingers and palms (not periungual). The patient did not have subsequent CAA. Two years later, he developed bloody stools and mouth ulcers, and intestinal biopsies confirmed a diagnosis of Crohn's disease.

A 10-month-old white female was treated on day 8 of fever for conjunctival injection only. The treating clinician classified the confidence in KD diagnosis as "probably" (confidence level 3). Treatment was provided when the nasopharyngeal swab for respiratory viruses was negative and because the CRP was elevated at 13.6 mg/dL. She did not demonstrate any periungual peeling and had no CAA. The following year, the child was diagnosed with PFAPA syndrome in an ID clinic after she had repeated, intermittent monthly episodes of fever lasting 2–5 days while remaining well in between the episodes. No genetic testing was performed.

A 6-year-old white male presented with 6 days of fever, strawberry tongue, maculopapular rash by history (although absent on the day of presentation), unilateral cervical lymphadenopathy, and generalized myalgias. The treating clinician categorized the lowest confidence in the KD diagnosis before treatment. During initial hospitalization, he developed severe arthralgias. He received 2 doses of IVIG and corticosteroids, and joint symptoms progressed. He was subsequently diagnosed with systemic onset idiopathic juvenile arthritis. He did not demonstrate any periungual peeling and had no CAA.

#### Classification of Kawasaki Disease (KD) Phenotype by Clinical Confidence in the KD Diagnosis

There were 142 patients in whom the treating clinician provided data as to the confidence in the KD diagnosis (Table 2). No cases of CAA were found among those with 2 lowest clinical confidence ratings for the KD diagnosis. The groups that had the lower confidence in the KD diagnosis had higher frequency of subsequent autoimmune/autoinflammatory diagnosis. Those with the lowest confidence in the KD diagnosis had the highest frequency of IVIG resistance.

## DISCUSSION

Our study presents data linking the clinical features at the initial KD presentation to the clinical outcomes of periungual peeling, CAA, IVIG resistance, and subsequent diagnosis of autoimmune/autoinflammatory diagnoses. We added a rating scale to the overall confidence in the KD diagnosis, which allowed us to gauge the overall clinical impression on the day of treatment for KD. There are potential benefits to standardizing documentation of the clinical presentation of KD on the day of treatment. First, the standardization of data collection allows for reassessment of the diagnostic certainty before proceeding with subsequent treatments for those who fail to respond to initial IVIG. This is important before considering additional doses of IVIG, because adverse effects can be dose dependent, such as hemolytic anemia [5–7]. Second, more precise associations of clinical outcomes with the type of KD, especially incomplete KD, can be made using this approach. Utilization of the AHA algorithm for incomplete KD has been shown to improve the diagnosis of KD in children who developed CAA [8], but the accuracy of the diagnosis for those with incomplete KD who do not meet these criteria is less clear. Among children with incomplete KD who did not meet the AHA criteria and whose initial echocardiograms were normal in this cohort, the IVIG resistance rate remained relatively high at 11%, but none developed CAA, and they had a trend towards lower rate of periungual peeling than other groups. Finally, utilization of this approach can allow for more precise epidemiologic data for the incidence and characteristics of IVIG resistance. Intravenous immunoglobulin resistance rates may vary [9], but these rates may be related to a lower likelihood of an accurate diagnosis of KD initially in some patients.

Patients treated for incomplete KD without baseline coronary abnormalities and who did not meet the AHA-defined criteria were more likely to have a subsequent autoimmune/autoinflammatory diagnosis. We have previously reported the clinical presentation of systemic onset juvenile arthritis after the KD diagnosis and noted on literature review of prior reported cases that most were diagnosed as incomplete KD specifically

lacking scleral injection [4]. Mevalonate kinase deficiency and PFAPA diagnoses causing recurrent fevers have been previously described after initial treatment for incomplete KD [10, 11]. Whether children who were subsequently diagnosed with an autoimmune/autoinflammatory illness actually had KD initially or had an alternative diagnosis is unclear. It has been suggested that recurrent fever disorders and KD may share the same propensity for activation of innate immune responses causing elevations in cytokine profiles and that the diseases may be related in those predisposed [11]. In either case, clinicians should observe these children for development of other symptoms. Longer-term cardiology follow-up visits for KD patients may represent an ideal time to screen for autoimmune/autoinflammatory symptoms and referral when appropriate.

The generally accepted practice in our institution is to order viral testing for patients being evaluated for KD. We recognize that among patients diagnosed with KD, respiratory viral detection is fairly common [12] and should not impact the decision to treat for KD, particularly for those with complete KD. However, viral characterization of adenovirus in children may be useful in distinguishing the 2 illnesses when coupled with the clinical presentation, especially for those being evaluated for incomplete KD [13, 14]. In addition, viral testing is often obtained before an inpatient attending evaluation. We urge clinicians to use caution in interpretation of viral detection from the respiratory tract of patients being evaluated for KD; findings should be interpreted only in conjunction with the clinical and laboratory evaluation of KD.

Our findings are consistent with other recent US cohorts in that the majority of CAA in affected KD patients are present during the initial hospitalization, with prior reports ranging from 81% to 84% [15, 16]. Before discharge, we identified children as high or low risk for development of CAA based on known risk factors [9, 17]: ie, age (infants), those requiring intensive care, and those with nonresponse to initial IVIG treatment. For these high-risk patients and children with known CAA, follow up is 3–7 days after discharge. Among the 4 children with baseline normal echocardiograms who went on to develop CAA, all met at least 1 high-risk criterion.

## CONCLUSIONS

Limitations of this study include that it was from a single center, the subsequent diagnoses were retrospectively identified, and the numbers of patients were small. Moreover, the tool in the electronic medical record required continued education of providers to increase compliance. Nonetheless, these data suggest that further study correlating the clinical KD phenotype on the day of presentation with clinical outcomes, including

subsequent diagnoses of autoimmune/autoinflammatory disorders, should be considered. Prospective and standardized collection of KD criteria on the day of treatment together with clinical outcomes can provide useful information to further refine the KD diagnosis and the potential for risk of subsequent autoimmune/autoinflammatory conditions.

## Supplementary Data

Supplementary materials are available at the *Journal of The Pediatric Infectious Diseases Society* online.

## Note

**Potential conflicts of interest.** All authors: No reported conflicts.

All authors have submitted the ICMJE Form for Potential Conflicts of Interest. Conflicts that the editors consider relevant to the content of the manuscript have been disclosed.

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