SHORT COMMUNICATION

# Coronary Artery Dilatation in Incomplete Kawasaki Disease

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We conducted this study to compare the incidence of coronary artery dilatation in children with Incomplete and Classical Kawasaki disease, diagnosed as per AHA criteria. Subjects were included on a retrospective review of records (2002-2007); those with a discharge diagnosis of Kawasaki disease were enrolled. A total of 29 patients were identified (3.1 per 1000 pediatric admissions), out of which 22 were boys (median age: 4.8 years; range: 4 months-11 years). Seventeen (58.6%) had Classical KD and twelve (41.4%) children had Incomplete KD. All children received IVIG and underwent echocardiography. Coronary involvement was more in Incomplete KD (11/12 = 91.6 %) as compared to Classical KD (7/17= 41.1%). The sensitivity, specificity and predictive value of AHA criteria to detect coronary artery dilatation was 39%, 9%, and 41%, respectively. We conclude that children presenting with Incomplete Kawasaki disease are at a higher risk of coronary artery abnormalities.

**Key words**: *AHA criteria, Coronary artery abnormalities, Diagnosis, Kawasaki disease.*

Coronary artery dilatations or aneurysms develop in approximately 25% of untreated children with Kawasaki disease and may lead to myocardial infarction (MI), sudden death, or ischemic heart disease(1). Early diagnosis and treatment with intravenous immune globulin (IVIG) can reduce the incidence of coronary artery abnormalities (CAA) to <5%(2-4).

The diagnosis of Kawasaki disease is based on clinical criteria summarized by the American Heart Association (AHA) in 1993(2). These include fever for 5 or more days; four of the other five findings i.e. a polymorphous exanthem, nonpurulent conjunctivitis, changes in the lips or oral cavity, redness and edema with later desquamation of the extremities, and at least one cervical lymph node that is >1.5 cm in diameter and no evidence of another disease with similar clinical features. Incomplete Kawasaki disease is being increasingly diagnosed. According to AHA, incomplete Kawasaki disease should be considered in all children with unexplained fever for 5 days associated with 2 or 3 of the principal clinical features of Kawasaki disease. We sought to determine the incidence of cases of Incomplete Kawasaki disease; and whether there is a significant difference in the incidence of coronary artery abnormalities between Classical and Incomplete Kawasaki disease.

## METHODS

The study was conducted at Malabar Institute of Medical Sciences, a tertiary referral center. The records of all children between 2002 and 2007 with a discharge diagnosis of Kawasaki disease were reviewed retrospectively. The diagnosis of Classical or Incomplete Kawasaki disease was based on American Heart Association criteria(2). Hemoglobin, blood count, ESR, platelet count and routine urine examination were done in all

patients. Children in Classical and Incomplete Kawasaki disease groups were analyzed with respect to duration of fever, presence of the other AHA criteria, interval from symptom onset to initiation of treatment with IVIG, presence of coronary artery dilatation determined by echocardiography, and the results of all laboratory tests. Characteristics of the two groups were compared by Chi-square test.

All patients received IVIG in a dose of 2 g/kg bodyweight on day 1 of admission. One child failed to respond despite 2 doses of IVIG and needed steroids, in view of persisting fever and other symptoms.

## RESULTS

During the 5-year study period, we had 29 patients diagnosed with KD (3 cases per 1000 pediatric admissions). Of the 29 cases, 22 (76%) were males and 70 % of the cases were <5 years.

***Table* I** compares the characteristics of children with classical and Incomplete Kawasaki Disease. One child with Classical KD had sensorineural hearing loss. One patient in the Incomplete KD group was initially diagnosed with dengue shock syndrome. Majority of children presented late (average 10 days after onset of fever). Of 18 (62%) children with coronary involvement, 11 (61.1%) did not meet the AHA criteria.

Coronary involvement was seen in 7 of the 8 cases (87%) who presented late in the Incomplete group, while all the 4 children who had an early presentation of Incomplete KD also had coronary dilatation. In the Classical group, coronary involvement was seen only in 2 of the 8 cases who had fever >10 days (25%) while 5 of the remaining 9 patients (55%) had coronary abnormalities.

There was no significant difference in the mean age of those with or without coronary involvement. Giant axillary and coronary aneurysm was noted in one infant who had fever for more than one month.

## DISCUSSION

Our series data shows a high number of cases not satisfying AHA criteria. This might probably be due to increased awareness among the clinicians of a hitherto unknown disease with cardiac complications affecting otherwise normal children, and the fact that early treatment may prevent the risk of

**TABLE I** COMPARATIVE FEATURES OF CLASSICAL AND INCOMPLETE KAWASAKI DISEASE

| Features | Classical KD | Incomplete KD | *P* value |
|---|---|---|---|
| Total cases | 17 | 12 | |
| Males | 10 | 11 | |
| Age of presentation (y) Mean (SD) | 4.43 (2.54) | 5.25 (3.62) | >0.05 |
| Duration of fever (d) Mean (SD) | 9.76 (6.4) | 11.17 (7.1) | >0.05 |
| Rash | 13 (76.5%) | 7 (58.3 %) | >0.05 |
| Lymphadenopathy | 15 (88.2%) | 6 (50%) | <0.05 |
| Conjunctival congestion | 13 (76.4%) | 5 (41.7%) | <0.05 |
| Oral mucosal changes | 16 (94.1%) | 8 (66.7%) | <0.05 |
| Peeling | 10 (58.8%) | 2 (16.7%) | <0.05 |
| Arthritis | 6 (35.3%) | – | |
| Coronary dilatation | 7 (41.1%) | 11 (91.7%) | < 0.001 |
| Thrombocytosis | 14 (82.3%) | 6 (50%) | <0.05 |
| Pyuria | 8 (47 %) | 3 (25%) | >0.05 |
| Jaundice | 1 | – | |
| Giant peripheral aneurysm | 1 | – | |

**WHAT THIS STUDY ADDS?**

- Incidence of coronary artery dilatation is more in incomplete Kawasaki disease, as compared to classical KD.

developing coronary artery abnormalities. Literature review suggests that early treatment with IVIG within the first 10 days of onset of symptoms is effective in decreasing the incidence of coronary artery dilatation(1,2,4). We noted that coronary artery dilatation was more in patients who did not meet AHA criteria.

Is it probably because coronary artery dilatation may be developing slightly earlier in incomplete cases as compared to classical KD where we have the opportunity to start treatment early? We have not been able to find published reviews corroborating this and number of our cases is too small to demonstrate statistical significance. Or is it because the incomplete KD is basically a virulent subgroup of Kawasaki disease with more predilections for coronary arteries? In our series children with fever and single criteria like oral mucosal changes are not included. Do they have the risk of developing coronary artery dilatations as incomplete or complete KD? We have not studied their coronaries. And if so, are we missing out a major proportion of cases, which could be potential KD with dilated or normal coronaries?

This study points out the need for better diagnostic modalities to detect children susceptible for developing CAA. We agree that this study has limitations as it is a retrospective review. A prospective study with a large sample size could address some of the issues, clinicians will continue to face difficult situations until sensitive and specific diagnostic tests are developed for Kawasaki disease.

Our study has shown that the incidence of Incomplete Kawasaki disease is high, and the incidence of coronary artery dilatation is more in these children. We recommend that clinicians continue to maintain high levels of suspicion, even in the absence of the complete clinical picture of KD.

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**REFERENCES**

1. Brogan PA, Bose A, Burgner D, Shingadia D, Tulloh R, Michie C, *et al*. Kawasaki disease: an evidence based approach to diagnosis, treatment, and proposals for future research. Arch Dis Child 2002; 86: 286-290.
2. Newburger J , Takahashi M, Gerber MA, Gewitz MH, Tani LY, *et al*. Diagnosis, treatment, and long-term management of Kawasaki disease: A statement for health professionals from the committee on rheumatic fever, endocarditis, and Kawasaki disease, council on cardiovascular disease in the young, American Heart Association. Circulation 2004; 110: 2747-2751.
3. Maconochie IK. Kawasaki disease. Arch Dis Child Educ Pract Ed 2004 ; 89: ep3-8.
4. Oates-Whitehead RM, Baumer JH, Haines L, Love S, Maconochie IK, Gupta A, *et al*. Intravenous immunoglobulin for the treatment of Kawasaki disease in children. Cochrane Database System Rev 2003; 4: CD 004000.