

# Incomplete Kawasaki Disease: A Rising Trend

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#### Abstract

Kawasaki Disease is the most common cause of acquired heart disease in the developed countries. The diagnosis of typical Kawasaki Disease (KD) is not much of a problem. However incomplete Kawasaki Disease where only two to three clinical features are present, poses a diagnostic dilemma. Here we report our experience with the disease in the paediatric department of Vivekananda Institute of Medical Sciences (VIMS). In one year retrospective study from Jan 2015 to Jan 2016, 20 cases were diagnosed as KD, of which five were complete and 15 incomplete. Fifty percent of cases were less than one year's age and there was male preponderance. More than 90% showed echo changes and all except one responded to IVIG and aspirin who succumbed due to aneurysm rupture. So we conclude that high index of suspicion for diagnosing KD must be there and early treatment to prevent echocardiographic changes.

**Key words:** Incomplete KD, Vasculitis, IVIG

#### Introduction

Kawasaki disease is the most common cause of acquired heart disease in paediatric population in developed countries and is the 2<sup>nd</sup> common childhood vasculitis<sup>1</sup>. There is variation in the incidence of paediatric KD between 9.1-32.5/100,000 population in USA<sup>2</sup>, 82.8/100,000 in Taiwan, 134.4 in Korea and 265/100,000 in Japan<sup>3,4</sup>. It has a slight male preponderance. The most important risk associated with KD is coronary artery aneurysm which has an incidence of 20-25% and a mortality of 1-1.5%<sup>5</sup>. The peak incidence of KD is from 6 months-2 years which includes approximately 50% of all KD patients<sup>6</sup>. The incidence of KD is around 10% in infants <6m which is similar to 11.2% in Japan and 7.7% in Korea<sup>7</sup>. The 1<sup>st</sup> epidemiologic study on KD from India published in 2011 suggested incidence of KD in Chandigarh at least 4.54/100,000 children below 15 years<sup>8</sup>.

KD is a clinical diagnosis. The diagnosis of KD relies on the early identification of the key clinical components of prolonged fever > 5 days and any 4 of the 5 criteria given by AHA guidelines (American Heart Association)<sup>9</sup>. It is further supported by supplementary criteria and echo findings. The uncertainties occur when not all of the classical signs are present and there is incomplete or atypical presentation of

the disease. Large epidemiological studies estimated around 20% of all cases fall into incomplete presentation of the group<sup>8</sup>. It has been found that this group is at greatest risk of developing the more serious cardiac sequel of KD i.e. coronary artery aneurysm. This higher risk has been attributed to the delay in diagnosis & treatment.

The aim of the study was to see the trend of Kawasaki disease in our institute over a period of one year and the objectives were to analyse the clinical profile of KD patients and compare the echo findings of complete and incomplete KD cases and their response to treatment.

## Materials and Methods

This was a retrospective descriptive study done in the paediatric department of VIMS a tertiary care institute in Kolkata from Jan 2015 to Jan 2016. A total of 20 children were diagnosed as KD in the given time frame; of which five were complete KD with typical features (Table 1)<sup>9</sup>. However rest of the 15 cases were not fitting in typical KD. All these 15 cases presented with fever more than five days and rash at some point of illness. Most of them were referred cases. They were being treated with antibiotics. However fever still persisted during their hospital stay eight of them developed oral mucositis, five of them had non purulent conjunctivitis. All infective aetiology was ruled out. These were then suspected for atypical KD, all of them met the supplementary criteria (Fig 1)<sup>9</sup>.

Echo was done which had early changes suggestive of KD in all the atypical cases. They were

treated with IVIG and Aspirin and followed up in paediatric rheumatology clinic.

## Results

In our study we had 20 cases diagnosed as KD over a period of one year.

Among the varied clinical manifestations of KD all the children so diagnosed presented with fever and rash mainly diffuse maculopapular (Fig 4).

*Supplementary criteria:* Raised ESR & CRP in all cases. Mean ESR was 90.8 mm in 1<sup>st</sup> hr and mean CRP > 24 mg/dl. Anaemia, leucocytosis, thrombocytosis present in 28% cases after 1<sup>st</sup> week (Fig 5).

*Echo changes:* These were characterised by perivascular cuffing and loss of tapering and or Z score of LAD/RCA > 2.5 SD of BSA<sup>9</sup>. (Table 6&7) Of the incomplete KD all of them had loss of perivascular cuffing which is the earliest sign of coronary artery changes in KD, one had fully developed aneurysm which was diagnosed on the 14<sup>th</sup> day of illness.

*Response to treatment:* IVIG was given to all the cases as soon as it was diagnosed. All the cases responded to single dose IVIG (2g/kg) and high dose aspirin (80-100 mg/kg) given till afebrile for 3 days following which the dose was reduced to 5mg/kg/d.

Eight weeks follow up with repeat echo was done in all the cases (Fig 8). Five cases had complete resolution of echo changes. All the complete KD cases had no residual echo findings at eight weeks follow up.

**Table 1:** Diagnosis of Kawasaki disease

5 days of fever AND 4 of the 5 following criteria	
1.	Discrete bulbar conjunctival injection without exudate
2.	Erythematous mouth and pharynx, strawberry tongue, and red cracked lips
3.	A polymorphous generalised erythematous rash
4.	Changes in peripheral extremities consisting of induration of the hands and feet with erythematous palms and soles
5.	Cervical lymphadenopathy (>1.5 cm), usually unilateral

*Adapted from the American Academy of Pediatrics, Redbook-2000: Report of the Committee on Infectious Disease, 25<sup>th</sup> ed. (Elk Grove Village, Ill: American Academy of Pediatrics, 2000), pp. 360-61*

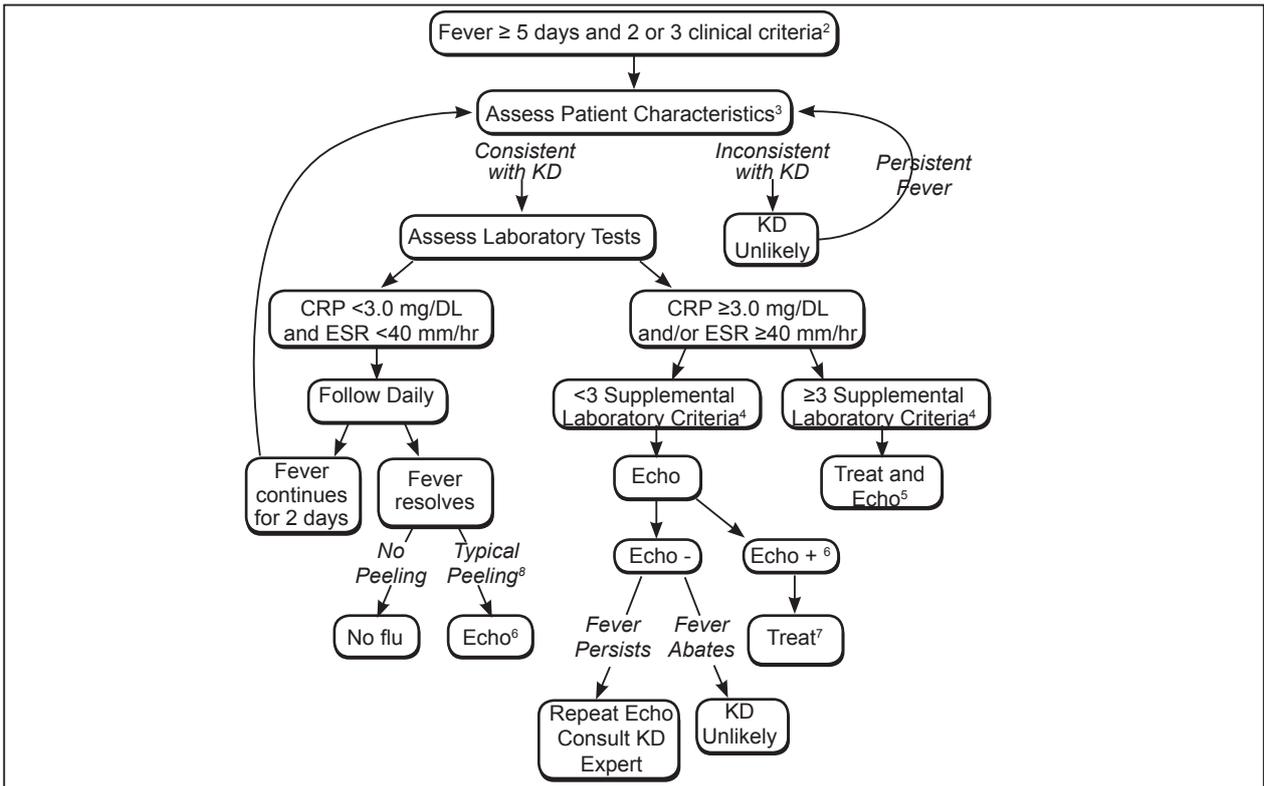


Fig 1: Diagnosis of incomplete KD

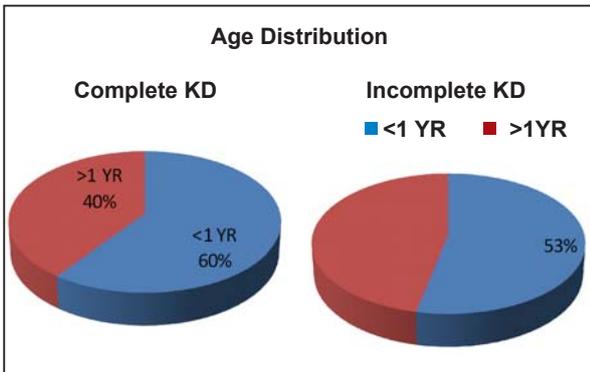


Fig 2: Showing complete and incomplete KD in <1 years age

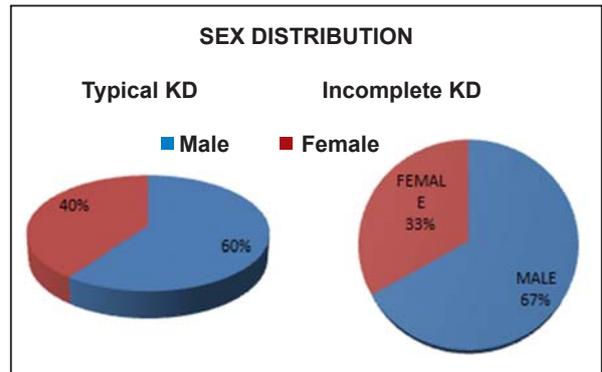


Fig 3: There was male preponderance both in the typical and incomplete KD groups.

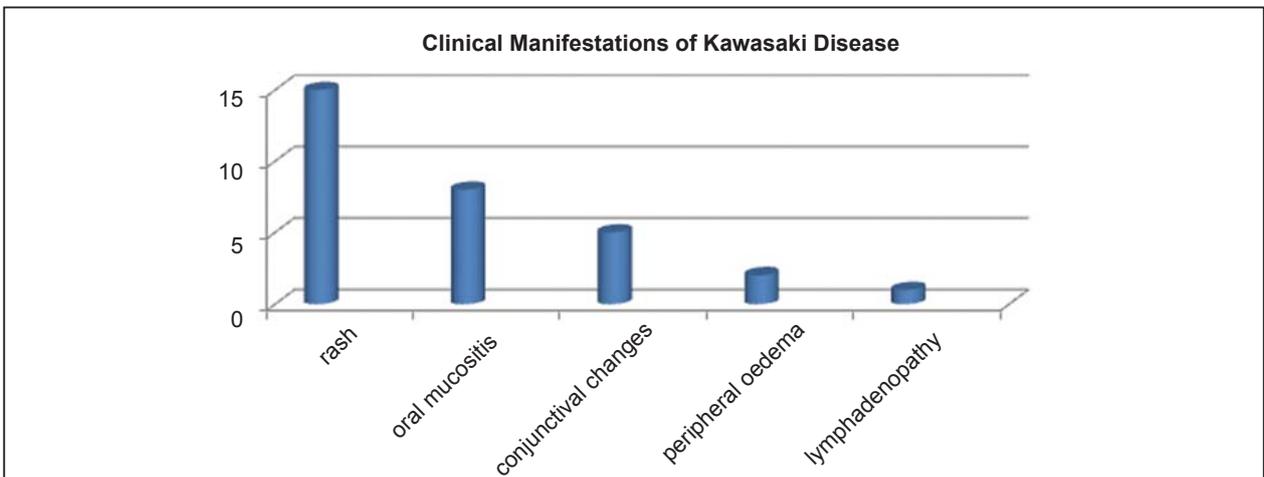
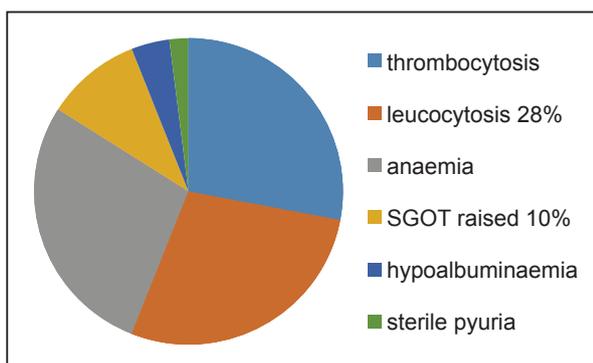
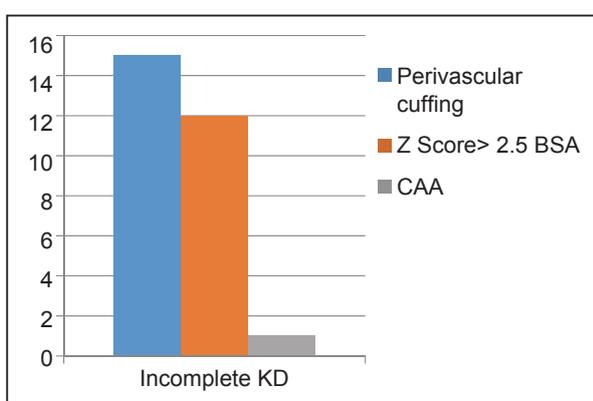


Fig 4: Showing rash as the predominant symptom in the cases followed by oral mucositis, conjunctival changes. Lymphadenopathy was however present in only 1 case.



**Fig 5:** in our study we found that anaemia, leucocytosis and thrombocytosis were present in 28% of cases. While raised SGOT was present in only 10% cases.



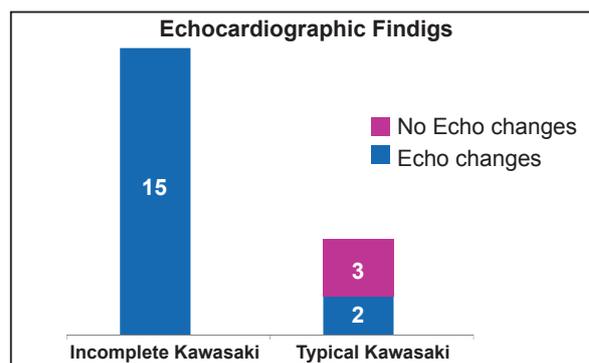
**Fig 7:** earliest echo changes in KD i.e. perivascular cuffing and loss of tapering were present in all the cases diagnosed as incomplete KD while coronary artery aneurysm developed in one case.

## Discussion

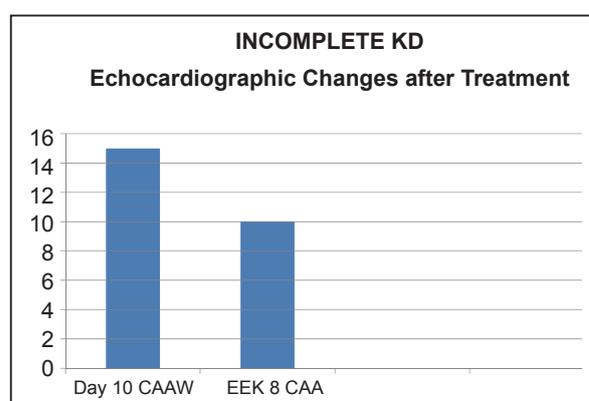
KD is a self limiting vasculitis occurring predominantly in children <5 year age. First Indian report was given in 1977 by Taneja *et al*<sup>10</sup>. 6 cases of Kawasaki disease (KD) diagnosed over a period of one year and review of all the cases reported from India<sup>11</sup>. The diagnosis of KD was based on clinical criteria. The mean age of patients was 6.83 years and mean duration of symptoms before diagnosis was 7.5 days.

In a study of patients referred because of possible KD, Burns and colleagues found that standard clinical criteria met in 46% of cases in whom other diagnoses was established<sup>12</sup>. Youngest patients are more likely to develop atypical features and aneurysm- 60% developed aneurysm in one series<sup>13</sup>.

KD is a disease of childhood. About 85% cases are less than 5 yrs age. In Japan<sup>4</sup>, the highest incidence occurs between 9 and 11 months old in boys, and between three and eight months old in girls. It is increasingly



**Fig 6:** Showing echo changes present in all the incomplete cases while only two of the complete KD cases. Three of the study population did not have echo changes at the time of diagnosis.



**Fig 8:** Eight weeks follow up showed resolution of echo changes in five of the 15 incomplete cases.

reported in infants and more common in males as also evidenced in our study. Baker *et al*<sup>14</sup> studied the symptoms in the 10 days prior to diagnosis of KD in 198 patients, and reported that irritability occurred in 50%, vomiting in 44%, decreased food intake in 37%, diarrhoea in 26%, and abdominal pain in 18%. Cough was reported in 28%, and 19% had rhinorrhoea. Over the next three to four days, cervical adenitis, conjunctivitis, changes in the buccal and oral mucosa, a pleomorphic rash, and erythema and edema in the hands and feet develop. In our study fever, rash, and irritability were present in all the cases. Conjunctival changes and lymphadenopathy occurred later in disease course.

Incomplete KD cases are more common in infants as seen in our study. 75% of the total cases had incomplete presentation and 60% of them were < 1yr age. Unfortunately all had echo changes as early as 10<sup>th</sup> day of fever and one had aneurysm which was diagnosed on 14<sup>th</sup> day of admission. This is in contrast to the study done by Karbuz *et al* in Turkey, which

showed CAA in 50% of incomplete KD<sup>16</sup>. The main reason could be delay in diagnosis in the absence of typical features, delay in referral to our institute thereby delay in treatment. A hospital based study done by Ajanthan R et al reported total 19 cases in one year of which 74% were incomplete and 26% were complete cases<sup>17</sup>. Coronary artery dilation was seen in incomplete criteria group during or after 2<sup>nd</sup> week due to delay in suspecting KD in the same study reported in Sri Lanka Journal<sup>17</sup>. In our study coronary changes were evident as early as 8<sup>th</sup> day of illness.

In Chandigarh cohort mortality rate over last 20 years is 0.8% as compared to 0.01-0.08% in developed countries<sup>18</sup>. Furusho and coworkers<sup>19</sup> first reported that high-dose IVIG appeared to decrease the incidence of coronary artery abnormalities. IVIG reduced the incidence of CAA by 78%, and no child suffered serious adverse effects from the therapy, confirming the remarkable therapeutic potential of IVIG.

In our study all patients responded to IVIG and high dose aspirin given till afebrile for 3 days thereafter low dose aspirin started. Follow up echo showed regression of echo changes in all typical KD cases while in eight of the incomplete ones echo changes persisted.

## Conclusion

Kawasaki disease is a medium vessel vasculitis with a high predilection for the coronary arteries. Incomplete KD is more common in infants and its incidence is on the rise. It is a medical urgency if not diagnosed and treated on time, child may develop coronary artery aneurysm the sequelae of which may be long lasting and devastating. Therefore high index of suspicion is required in any child with prolonged fever, rash and irritability. The diagnosis is mainly clinical and one need not delay administration of IVIG just because an echo has not been performed or echo changes not present at that time.

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