

also be provided if a case of AFP occurs. The outbreak also highlights the importance of administering the recommended doses of a potent TOPV vaccine to all the children at the earliest recommended age for prevention of such outbreaks in future.

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## Kawasaki Disease

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Kawasaki disease (KD), first reported in 50/ Japanese children by Tomisaku Kawasaki in 1967, is a syndrome of unknown etiology characterized by fever and mucocutaneous inflammation(1). Conjunc-

tivitis, redness of the tongue and buccal cavity, swelling of hands and feet and cervical lymphadenitis are the presenting signs. KD occurs most commonly below five years, boys being affected more fre-

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quently than girls. The diagnosis is essentially clinical, based on a consensus statement prepared by North American Participants at the third International Kawasaki Disease Symposium, Tokyo, Japan in December 1988(2) (*Table I*).

Since the first report, cases have been described world wide. However, there have been only a few reports from India(3). We present here the clinical data, laboratory tests, ECG and echocardiographic results of eight children with KD, admitted to the pediatric wards of the Medical College Hospital (S.A.T. Hospital) Trivandrum, between January 1994 and November 1995. The cases illustrate the presentation and outcome of KD in our setting.

### Case Reports

The important clinical features of the eight children who presented over a period of 20 months are listed in *Table II*. All the children were male and between one and five years of age. All patients presented with fever of five days or more, cervical lymphadenopathy, bilateral suffused conjunctivitis, deep red tongue with prominent papillae ("strawberry tongue") and redness and swelling of the hands and feet (*Figs 1-3*). A remarkable feature exhibited by all eight patients was extreme irritability at presentation. The cervical lymph nodes were 1-1.5 cm in diameter with at least one node in each group being significantly enlarged in all children. Seven of the eight children had a maculopapular rash at presentation mainly truncal. During the first week of hospitalization, three of the eight patients developed an apical systolic murmur with no chest pain or dysrhythmia. Two complained of abdominal pain and diarrhea. While only two boys had large joint (knees) arthritis, all eight had diffuse, tender edema of the feet with refusal to bear weight, possibly due to

**TABLE I—Diagnostic Criteria for Kawasaki Disease**

- |    |  |
|----|--|
| A. | Fever lasting for atleast 5 days*  |
| B. | Presence of four of the following five conditions: <ol style="list-style-type: none"> <li>1. Bilateral nonpurulent conjunctival injection</li> <li>2. Changes of the mucosa of the oropharynx, including infected pharynx, infected and/or dry fissured lips, strawberry tongue</li> <li>3. Changes of the peripheral extremities, such as edema and/or erythema of the hands or feet, desquamation, usually beginning periungually</li> <li>4. Rash, primarily truncal: polymorphous but nonvesicular</li> <li>5. Cervical lymphadenopathy</li> </ol> |
| C. | Illness not explained by other known disease process   |

\* Many experts believe that, in the presence of classic features, the diagnosis of Kawasaki disease can be made (and treatment instituted) before the 5th day of fever by experienced individuals.

**TABLE II—Clinical Findings in Kawasaki Disease.**

Clinical findings	No.	%
Fever more than 5 days	8	100.0
Cervical lymphadenopathy	8	100.0
Conjunctivitis	8	100.0
Oral changes	8	100.0
Polymorphous rash	7	87.5
Hands and feet changes	8	100.0
Severe irritability	8	100.0
Heart murmur	3	37.5
Arthritis	2	25.0
Diarrhea	2	25.0
Coronary arteritis	1	12.5

metatarsophalangeal involvement. No child had hypotension, Koplik's spots, jaundice or any neurological deficit. In the second or third week of hospitalization, all

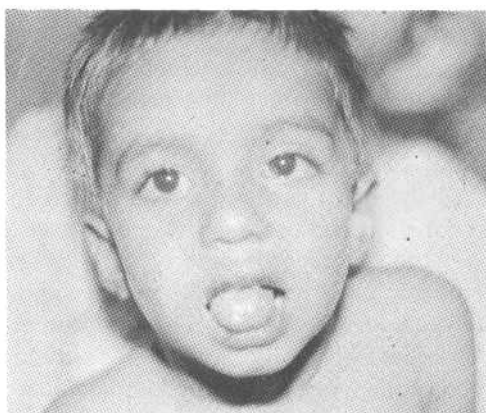


Fig. 1. Photograph showing dry fissured lips, strawberry tongue and conjunctivitis in one subject.

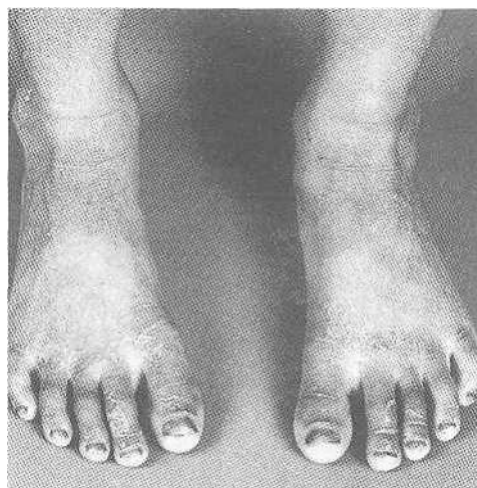


Fig. 3. Skin desquamation in the late stage.



Fig. 2. Edema dorsum of feet with early periungual desquamation of left foot in a case.

eight children developed periungual desquamation of the finger tips and toes.

The laboratory reports of the eight patients are shown in *Table III*. All cases showed a raised ESR and a mild polymorphonuclear leukocytosis. The mean platelet count of 3,14,000/cu mm was considered undoubtedly high for our laboratory

values. More importantly the peripheral blood picture in all cases showed thrombocytosis. C-reactive protein results were high in all cases (4). Blood culture was done in all cases and was sterile. The urine examination, liver function and renal function tests were normal in all cases. All the eight patients were seen by our pediatric cardiologist and were also referred to the neighboring Sri Chitra Thirunal Institute of Medical Science and Technology (SCTIMST) for cardiac evaluation. Open heart surgery, cardiac catheterization and angiography for adults as well as children

TABLE III—Results of Laboratory Investigations.

Investigation	Mean	Range
Hemoglobin (g/dl)	9.0	8-11
Total leukocyte count (per cu mm)	12,600	10,100-14,200
Polymorphs (%)	66.0	54-78
Platelet count (per cu mm)	314000	282000-396000
ESR (fall in 1st h)	87.5	45-130

are done here routinely. The decision regarding the nature of cardiac investigations to be done in each case was made by the team of cardiologists there. ECG and echocardiography were done in all cases and coronary angiography in two. All cardiac investigations were normal in seven children. However, one child showed evidence of dilatation and increased echogenicity of left main coronary artery, proximal (L) anterior descending and proximal (L) circumflex arteries.

A clinical diagnosis of Kawasaki disease was made in all cases based on the easily recognizable diagnostic criteria described and in the absence of evidence of other known disease process. All eight children were treated with aspirin and intravenous gamma globulin. Aspirin was given in a dose of 75 mg/kg/day until the ESR came down and then at 5 mg/kg/day for a further period of six months. Intravenous gammaglobulin was administered to all children on diagnosis in a dose of 200 mg/kg/day for three to five days except the child with coronary artery dilatation who received 400 mg/kg/day for 5 days.

In all the eight patients temperature returned to normal within 48 hours of administration of intravenous gammaglobulin. In 4 children conjunctivitis disappeared within 2 days and within a week in the other four. The ESR came down to normal by the second week in all the patients. Large joint arthritis also disappeared by second week. The most dramatic response was the disappearance of irritability within 24 to 48 hours of intravenous gammaglobulin in all eight subjects.

The patients were followed up clinically and by serial ECG and 2-dimensional echocardiography for 6-12 months. There was no recurrence of symptoms in any case. The cardiac murmur in 3 cases disappeared within a month of follow up. The

evidence of coronary artery dilatation in the lone case showed complete regression on echocardiography after 4 months.

### Discussion

All the eight cases in our study qualified for the diagnosis of Kawasaki disease as per the criteria laid down (*Table I*). The age of our patients was also in agreement with other reports(4,5). A male preponderance seems universal(5) and all the patients in our series were male. The fact that three of the eight cases showed clinical and one showed echocardiographic evidence of cardiac involvement also conforms to other reports of such involvement(6,7). In our patient it was the (L) coronary artery that was mainly involved. This has been described as the most common cardiac finding in other cases as well(8,9); moreover, the fact that the coronary artery involvement was present on admission agrees with the report(8) that coronary artery involvement can occur as early as the fifth day of illness.

The findings we thought unusual included the extreme, universal irritability and its dramatic disappearance within 24 hours of administration of intravenous immunoglobulin. Again while diarrhea has certainly been described, it is not a characteristic feature. Anemia, a feature that was present in various degrees in all our cases has not been commonly described.

Though no laboratory test in KD has been considered diagnostic, thrombocytosis has been consistent enough in reports to make it an important marker(9). While the range seen in this series cannot be described as very high, it was significantly higher than the normal for our hematology laboratory.

The decision to use a lower than recommended dose of intravenous immunoglobulin in our patients was influenced by two considerations, namely, economic constraint

(the medication is not provided free in the Government Medical College Hospital) and the fact that studies have shown that while a high dose (2 g/kg single dose or 400 mg/kg/day for 5 days) is recommended as the treatment of choice (2,10), the dose schedules of 200 mg/kg/day for 3 days as well as for 5 days have both been found effective<sup>1</sup>). We, therefore, acted on the premise that a lower effective dose of intravenous immunoglobulin would be beneficial than the higher optimal dose which could not be given for financial reasons. Moreover, it is possible that the lower dose is effective in our population.

Finally, the etiology of KD is still controversial. Is our experience of eight cases within 20 months, a clustering that argues in favor of an infective etiology which leads to an immune mediated syndrome in certain genetically susceptible children?

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