Controversies in diagnosis and management of Kawasaki disease

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Abstract

Kawasaki disease (KD) is a common medium vessel systemic vasculitis that usually occurs in small children. It has a predilection for the coronary arteries, but other medium sized arteries can also be involved. The etiology of this disorder remains a mystery. Though typical presentation of KD is quite characteristic, it may also present as incomplete or atypical disease in which case the diagnosis can be very challenging. As both incomplete and atypical forms of KD can be associated with serious coronary artery complications, the pediatrician can ill afford to miss these diagnoses. The American Heart Association has enunciated consensus guidelines to facilitate the clinical diagnosis and treatment of this condition. However, there are still several issues that remain controversial. Intravenous immunoglobulin remains the cornerstone of management but several other treatment modalities, especially glucocorticoids, are increasingly finding favour. We review here some of the contemporary issues, and the controversies thereon, pertaining to management of KD.

Key words: Kawasaki disease; Diagnosis; Intravenous immunoglobulin; Treatment; Controversies

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Core tip: The diagnosis of Kawasaki disease poses several challenges for the treating pediatricians as it is based on a set of criteria that are entirely clinical. To further complicate matters, several children present with incomplete and atypical forms of the disease. It is known that children with incomplete and atypical Kawasaki disease do not have milder form of the disease, rather the rate of coronary and non-coronary complications may even be higher in these subgroups as the diagnosis often gets delayed. While intravenous immunoglobulin...
remains the cornerstone of management, several children require additional form of therapy thereby further challenging the clinical skills and judgment of the pediatricians.

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WHAT IS KAWASAKI DISEASE?

Kawasaki disease (KD) is the most common medium vessel vasculitis and usually affects young children. It has a special predilection for coronary arteries[1,2]. KD is now the leading cause of pediatric acquired heart disease in developed countries like Japan, Korea and Taiwan as also in countries in North America and Europe. In several resource poor countries (e.g., India, China) as well, KD is now being increasingly reported[3]. However, anecdotal evidence suggests that many children still remain undiagnosed and untreated in such settings[4]. The etiology of KD remains an enigma[5,6].

This disease was first recognized in 1961 by Dr. Tomisaku Kawasaki based on a constellation of clinical signs and symptoms and he reported it in 1967 as “muco-cutaneous lymph node syndrome”[7]. Since then it has been reported from all continents. Even though the initial description of KD was given more than 50 years ago, the diagnosis still remains clinical and there is no laboratory test that can confirm a clinical diagnosis of KD[8].

DIAGNOSIS OF KD

Diagnosis of KD is essentially based on a constellation of clinical signs and symptoms and supported by laboratory investigations[8-12]. It cannot be overemphasized that there is no pathognomonic laboratory test for diagnosis of KD. A careful and meticulous history from parents, or documented clinical findings by a physician who has seen the child previously, may be useful in facilitating a diagnosis of KD[13].

The diagnostic criteria for KD have been modified from time to time. There are two sets of diagnostic criteria that have been used most frequently for diagnosis of KD. These included the Kawasaki Disease Research Committee guidelines [Japanese Ministry of Health (JMH) guidelines], 2002[14] and the American Heart Association (AHA) guidelines[1-13]. AHA guidelines were published in 2004 and have been widely used since then[14]. Mccrindle et al.[13] have recently published the AHA 2017 revised guidelines for diagnosis and management of KD. These criteria are based on clinical findings and do not differ significantly from the original descriptions of cases of KD given by Dr. Kawasaki himself in 1967[7].

Complete KD

The AHA 2017 have proposed a set of diagnostic criteria for complete KD (Table 1)[13]. Fever is the most common presenting clinical manifestation and is seen in nearly all patients. While fever is essential for the diagnosis of KD as per AHA criteria but according to the Japanese criteria, fever need not be present in all patients (Table 2)[14]. The duration of fever in KD is variable and is usually less than 2 wk but may persist for much longer periods of time. Clinical manifestations of KD evolve over days and many signs and symptoms may have disappeared by the time the patient seeks medical attention. This issue has been clearly highlighted in the recent AHA 2017 guidelines (Table 3)[13].

Incomplete KD and atypical KD

The diagnosis of KD can test the clinical acumen of even an astute physician. The signs and symptoms of KD are nonspecific and may overlap with those of infectious diseases seen in young children[2]. Adding to this challenge are patients with KD who do not fulfill the diagnostic criteria. A diagnosis of incomplete KD is usually made when there is ongoing fever but less than four clinical features[13]. In such cases the attending pediatrician has to do a thorough clinical assessment and look at supportive laboratory investigations. Incomplete KD is common in infants (especially in babies below 6 mo) and young children. On the other hand, atypical KD is said to be present when there are atypical manifestations, as for instance nephritis[15,16], pneumonia[17], arthritis[18], myositis[19,20], uveitis[21], retinal vasculitis[22,23] and CNS involvement[24,25]. Incomplete or atypical forms of KD should by no means be considered as mild KD because the risk of coronary abnormalities in these patients is comparable with, if not higher than, classic KD. This fact cannot be overemphasized[26-29].

Important clinical signs not included in the diagnostic criteria

There are several manifestations that are not included in the diagnostic criteria but may provide important clues towards diagnosis of KD. Perineal desquamation is one such clinical sign. It usually appears a few days prior to the appearance of periangual desquamation and may provide the initial clinical clue[26-29]. Similarly, reactivation of the Bacillus Calmette-Guérin (BCG) injection site is a pathognomonic clinical sign of KD and is almost exclusively observed in infants[33-36]. However, this has not been given enough consideration in the diagnostic criteria. Reason for this may be that many developed countries are not using BCG vaccine as a routine. Sterile pyuria[37,38], peripheral arthritis[39] and gall bladder hydrops[39,40] are other important indicators of KD. Extreme irritability, out of proportion to the fever, is often observed in young children with KD and may be a prominent clinical finding—but this too does not find a mention in the diagnostic criteria[41,42].
scarlet fever responds briskly to antimicrobials but not in scarlet fever. Further, the fever in children with conjunctival injection are features that are seen in KD. However, involvement of the lips and presence of a viral infection as is sometimes the case, therefore, not difficult to understand why KD can be confused with a viral illness. There are, however, some clinical features that can help in this differentiation. Children with KD usually do not have rhinorrhea or conjunctival discharge, in contrast to patients with viral infections. They are also often very irritable. Edema over dorsum of hands and feet and the characteristic desquamation (perianal in first few days and periungual over dorsum of hands and feet and the characteristic desquamation (perianal in first few days and periungual over dorsum of hands and feet) may then be ascribed to an adverse drug reaction to antimicrobials that are often given in such situations. Salgado et al. have reiterated these facts in their recent publication on KD in infants below 6 mo. Our experience is also similar. It is easy to understand why the diagnosis (and consequently the treatment) of KD gets delayed in these circumstances. Unfortunately, such delays can have disastrous consequences in the baby.

As per the recent AHA 2017 guidelines, the diagnosis of KD in infants may be considered in the following situations: (1) Infants < 6 mo old with prolonged fever and irritability; (2) infants with prolonged fever and unexplained aseptic meningitis; (3) infants or children with prolonged fever and unexplained or culture-negative shock; (4) infants or children with prolonged fever and cervical lymphadenitis unresponsive to antibiotic therapy; (5) infants or children with prolonged fever and retrophyaryngeal or parapharyngeal phlegmon unresponsive to antibiotic therapy.

In older children and adolescents
Diagnosis in older children and adolescents is difficult because KD is rarely kept as differential diagnosis by adult physicians. As the diagnosis usually gets delayed after days 10-12) is typical of KD but these findings are not there in all patients and can be easily missed if not looked for carefully. However, the picture gets further complicated when KD occurs concomitantly with a viral infection as is sometimes the case.

One of the closest mimics of KD is scarlet fever. However, involvement of the lips and presence of conjunctival infection are features that are seen in KD but not in scarlet fever. Further, the fever in children with scarlet fever responds briskly to antimicrobials.

### KD in infants

KD in infants is a challenging exercise for physicians and delays in diagnosis in this age group are not uncommon. KD in infants often does not fulfill the standard diagnostic criteria. KD may be incomplete in a large proportion of patients in this age group. Morbidity and mortality in this age group is highest compared to any other age group. Fever and excessively irritability may be the only clinical manifestations of KD in babies below 6 mo and such presentations can pose several difficult questions for the attending pediatrician.

Delays in diagnosis are common in such situations. Young infants are said to be at highest risk of developing coronary artery abnormalities. The presence of fever and pyuria in an infant can be mistakenly attributed to a urinary tract infection. This is not uncommon in our experience. Other clinical features of KD (e.g., rash, red eyes, and red lips) may then be ascribed to an adverse drug reaction to antimicrobials that are often given in such situations. Salgado et al. have reiterated these facts in their recent publication on KD in infants below 6 mo. Our experience is also similar. It is easy to understand why the diagnosis (and consequently the treatment) of KD gets delayed in these circumstances. Unfortunately, such delays can have disastrous consequences in the baby.

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### CONTROVERSIES IN DIAGNOSIS OF KD

#### Infections and KD

KD should be considered as a diagnostic possibility in all children with fever more than 5 d for which there is no discernable cause. KD is more common in young children and 80% of patients are under 5. This is the age group wherein viral infections are also common. Some of the clinical features of KD (e.g., conjunctival injection, rash and cervical lymphadenopathy) are also a common feature of viral illnesses like measles, rubella, adenovirus, enterovirus, Stevens-Johnson syndrome, toxic shock syndrome, drug hypersensitivity reactions, systemic juvenile idiopathic arthritis

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#### KD in older children and adolescents

Diagnosis in older children and adolescents is difficult because KD is rarely kept as differential diagnosis by adult physicians. As the diagnosis usually gets delayed...
Algorithm for incomplete KD has been simplified. The limitations of echocardiography and other diagnostic modalities have been highlighted. Z-score (by Manlhot) severe forms of the disease of coronary aneurysms and such patients often have count has also been found to correlate with development of macrophage activation syndrome. Thrombocytopenia in acute stage of KD can be a marker for diagnosis of KD, especially in children with incomplete or laboratory investigation may be used to support the conclusive evidence for diagnosis of KD. Clinical diagnosis of KD. Laboratory markers rarely provide conclusive evidence amongst adult cardiologists, such patients may never get recognized as having had late complications due to missed childhood KD.

**Table 3** Salient differences between American Heart Association 2004 and 2017 criteria

| Duration of fever | In the presence of ≥ 4 principal clinical features, particularly when redness and swelling of the hands and feet are present, KD can be diagnosed even within 4 days of fever |
| History | Presence of one or more principal clinical manifestations of disease that can be revealed on history but have disappeared by the time of presentation, have been considered important for diagnosis |
| KD shock syndrome | KDSS has been given special consideration in the 2017 revised guidelines because in the presence of shock the diagnosis of KD is often not considered |
| KD in infants | Clinicians should have a lower threshold for diagnosis of KD in this age group |
| Incomplete KD | Algorithm for incomplete KD has been simplified |
| KD and infections | The issue of infections and KD has been detailed at length. Diagnosis of KD must not be excluded even in the presence of a documented infection when typical clinical features of KD are present |
| Bacterial lymphadenitis | Ultrasonography and computed tomography findings in differentiating the 2 conditions- bacterial lymphadenitis is often single and has a hypoechoic core on ultrasonography, while lymphadenopathy in KD is usually multiple and is associated with retropharyngeal edema or phlegmon |
| 2D-echocardiography | The limitations of echocardiography and other diagnostic modalities have been highlighted. Z-score (by Manlhot et al) for severity classification of coronary artery abnormalities has been adapted |

KD: Kawasaki disease; KDSS: Kawasaki disease shock syndrome.

in these children, there is higher risk of coronary artery abnormalities[52,56]. Further, echocardiographic coronary artery assessment in this group of patients is difficult because of the thick chest wall[55].

Clinical consequences of missed KD can present as coronary ischemia in early adulthood[56,57]. Due to lack of adequate awareness amongst adult cardiologists, such patients may never get recognized as having had late complications due to missed childhood KD[57].

**KD shock syndrome**

Myocarditis is nearly universal in acute phase of KD and, at times, it can be severe and symptomatic[58,59]. These patients are usually admitted in intensive care units with cardiovascular collapse and may be mistakenly treated for bacterial sepsis and septic shock[13,60,61]. As a result, the diagnosis of KD gets delayed and this can have serious consequences. Such patients are at high risk of developing coronary artery abnormalities, intravenous immunoglobulin (IVIg) resistance and myocardial dysfunction[62]. It is, therefore, prudent to keep a differential diagnosis of KD in all children presenting with seemingly obscure myocardial dysfunction and shock. A presumptive diagnosis of viral myocarditis/ septic shock in the intensive care setting should have a differential diagnosis of KD. It is for these, and many other reasons that KD shock syndrome (KDSS) has been given special consideration in the AHA 2017 revised guidelines[13].

**Laboratory investigations may not always be corroborative**

There is no single laboratory test for confirmation of diagnosis of KD. Laboratory markers rarely provide conclusive evidence for diagnosis of KD. Clinical laboratory investigation may be used to support the diagnosis of KD, especially in children with incomplete or atypical KD and to assess the intensity of inflammation. Thrombocytopenia in acute stage of KD can be a marker of macrophage activation syndrome[13,63]. Low platelet count has also been found to correlate with development of coronary aneurysms and such patients often have severe forms of the disease[52].

N terminal pro-B-type natriuretic peptide (NT-pro-BNP) is a cardiac biomarker and has been found to be significantly elevated during acute stage of KD when compared to febrile controls[64]. There are age based Pro-BNP nomograms to help the treating physician in differentiating KD from other febrile illnesses[65]. The values are higher in patients who develop coronary artery abnormalities as compared to those with normal coronaries. Thus it has both diagnostic and prognostic implications. Level of ProBNP is also correlated with myocardial dysfunction in acute stage of KD[66].

**CONTROVERSIES IN IMAGING STUDIES IN KD**

**Role of 2D-echocardiography in KD**

2D-echocardiography is an essential component of the diagnostic work-up in children with KD. It is a useful tool to assess the status of coronary arteries and other cardiac structures during acute stage as well as on follow-up[13,55,67]. It is important to bear in mind, however, that a negative echocardiographic examination does not rule out KD. However, from the perspective of a developing country, there are several issues with regard to this investigation. It cannot be overemphasized that the quality of scans obtained on echocardiography is operator dependent. This investigation has significant inter-observer variability and needs expertise and patience, especially in infants and young children[10]. Artifacts pose an important problem and these can make the examination exceptionally difficult, especially when the left circumflex or right coronary artery is being scanned[56]. In developing countries like India there is a dearth of trained pediatric cardiologists. As a result, the investigation may be carried out by an adult cardiologist, who may not have the requisite expertise to assess the coronary arteries especially in young infants. It comes as no surprise that echocardiography reports are often incomplete and inaccurate in clinical practice, especially in developing countries[10].
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Table 4  Coronary artery abnormalities severity classification in different guidelines

| Criteria                          | Description                                                                 |
|----------------------------------|-----------------------------------------------------------------------------|
| JMH criteria[a]                  | Aneurysm definition                                                         |
|                                  | < 5 yr - ID > 3 mm                                                          |
|                                  | ≥ 5 yr - ID > 4 mm                                                          |
| Updated JMH (2008)[b]            | Small aneurysm (dilatation with ID < 4 mm or if child is ≥ 5 yr of age) |
|                                  | Medium aneurysm (dilatation with ID > 4 mm but ≤ 8 mm or if child is ≥ 5 yr of age) |
|                                  | Large aneurysm (dilatation with ID > 8 mm or if child is ≥ 5 yr of age) |
| AHA 2004 criteria[b]             | Aneurysm ID z score > 2.5 (as per body surface area adjusted z scores)    |
|                                  | Small: ≤ 5 mm                                                              |
|                                  | Medium: 5 to 8 mm                                                           |
| AHA 2017 criteria (Manlhiot et al)[c] | Giant aneurysm: > 8 mm based on absolute diameter                         |
|                                  | No involvement (Z score < 2)                                                |
|                                  | Dilation only (Z score 2 to < 2.5; or if initially < 2, a decrease in Z score during follow-up ≥ 1 thereby suggesting that coronary artery was dilated during acute stage though diameter was within normal standards and the diameter has regressed on follow-up) |
|                                  | Small aneurysm (Z score ≥ 2.5 to < 5)                                       |
|                                  | Medium aneurysm (Z score ≥ 5 to < 10, and absolute dimension < 8 mm)       |
|                                  | Large or giant aneurysm (≥ 10, or absolute dimension ≥ 8 mm)                |

ID: Internal diameter; AHA: American Heart Association; JMH: Japanese Ministry of Health.

The Japanese Ministry of Health has enunciated criteria for defining coronary involvement in KD on the basis of absolute dimension of internal diameter of coronary artery[b]. McCrindle et al[c] and Manlhiot et al[c] have proposed the classification scheme based on z score for severity of coronary artery abnormalities, which has been adapted and recommended by AHA 2017 guidelines (Table 4). It is mandatory that body surface area-adjusted ‘Z’ scores be used to grade the severity of coronary artery involvement so that objectivity can be maintained and results can be compared with other studies[c]. Echocardiography findings in KD other than coronary artery ectasia, dilatation and aneurysm, include lack of tapering of coronary arteries, myocardial dysfunction, pericardial effusion, aortic root dilatation and valvular regurgitation[13,55,59]. As myocarditis is almost universal, functional abnormalities are likely to be more in acute stage of KD[c]. An echocardiography examination should be done at diagnosis and, if normal, should be repeated on a daily basis for the next few days. Repeat echocardiography should be carried out 1-2 wk later and then at 4-6 wk. A normal echocardiography examination during the first week of illness does not rule out the development of coronary artery aneurysms later. Echocardiography should be repeated more frequently in children who have coronary artery z-scores > 2[13]. Recent literature suggests that follow-up echocardiography examination should include assessment of myocardial functions in addition to assessment of coronary arteries[c].

**Computerized tomography coronary angiography in KD**

While 2-dimensional echocardiography remains the imaging modality of choice to identify coronary artery abnormalities, it is subject to several fallacies and is operator dependent. Computerized tomography (CT) coronary angiography is rapidly emerging as a useful imaging modality for better characterization of dilatations, ectasia and aneurysms especially in the mid- and distal segments of coronary arteries. It provides precise details in terms of aneurysm size and morphology[70]. The limiting factor in more widespread use of this investigation hitherto was the high radiation exposure and therefore its application in children was rather limited. Over the last 5 years, with the advent of higher detector and dual-source CT scanners (DSCT), it is possible to delineate the coronary artery anatomy with higher temporal resolution and motion-free images at all heart rates with acceptable radiation risk[70]. CT coronary angiography can detect dilatations, ectasia and aneurysms in the mid and distal segments of coronary arteries with precise details in terms of aneurysm size and morphology. In the convalescent phase, it also can be used to delineate complications like segmental stenosis, intra-aneurysmal thrombus and mural calcifications.

**Magnetic resonance coronary angiography**

Magnetic resonance (MR) is useful in evaluation of coronary artery lesions and myocardial involvement in all stages of KD. The main advantage of MR is that there is no radiation exposure. However, young children would often need to be sedated and the procedure is time consuming. Interpretation of MR images requires a lot of expertise and skill[21,22].

**CONTROVERSIES IN MANAGEMENT OF KD**

Treatment of KD is yet another challenging and controversial issue. Prompt treatment of KD is absolutely
essential if one is to avoid the chances of development of CAA\textsuperscript{[11,13]}. Intravenous immunoglobulin (IVIG) remains the standard of care based on objective evidence collated from prospective studies and meta-analyses\textsuperscript{[13]}. However, there are still several controversies regarding management of children with KD.

**Acute phase management**

**IVIG:** For IVIG to be most effective, it should be given in the first few days of the illness\textsuperscript{[73]}. However, if the child presents after day 10 of fever, IVIG should still be given if the acute inflammatory parameters are high\textsuperscript{[73]}. Though there are recent meta-analyses stating similar outcomes of KD treatment at different doses of IVIg, a dose of 2 gm/kg administered intravenously is the preferred option\textsuperscript{[12,76]}. It has also been suggested that administration of IVIG before day 5 of fever may inadvertently increase the need for further IVIG therapy and also increase the chances of developing a refractory state\textsuperscript{[75]}.

**Aspirin:** Though aspirin is a widely used anti-inflammatory agent in KD and is given along with IVIG, its efficacy remains questionable as there is no proof that addition of aspirin in the acute phase significantly decreases the chances of development of coronary artery abnormalities\textsuperscript{[76]}.

Most clinicians prefer to use aspirin in doses of 30-50 mg/kg during the acute phase of KD. Duration of aspirin therapy is another controversial issue\textsuperscript{[77,78]}. Some centres prefer to continue it for 2 wk irrespective of fever status but consensus is rapidly evolving over continuing it only for febrile phase and then to change to a low dose (3 to 5 mg/kg per day) for its anti-platelet effect\textsuperscript{[79]}. This low dose is then continued for 6-8 wk and is stopped if follow-up echocardiographic examination is normal. Aspirin is continued indefinitely if there is persistence of CAA\textsuperscript{[13]}.

**Corticosteroid therapy in acute phase:** Kato et al\textsuperscript{[80]} reported that administration of steroids during the acute phase of KD resulted in increased incidence of CAA. But it is now argued that these results were due to the confounding factor of steroids having been given to children who were sicker than the other group\textsuperscript{[81]}. Kobayashi et al\textsuperscript{[82]} have recently published their study on use of steroids with IVIG and found that steroids may be useful in acute phase of KD.

Recent AHA guidelines do not support for administration of methylprednisolone pulses simultaneously with IVIG therapy but suggest possible benefit of 2-3 wk tapering steroid therapy along with IVIG and aspirin doses\textsuperscript{[13]}. Uptfront steroid therapy may be considered only for patients with KD who are proven to be IVIG resistant or presenting with significant CAA\textsuperscript{[10,46]}. The choice of steroid is usually intravenous methylprednisolone pulse followed by tapering dose of oral prednisolone\textsuperscript{[13]}.

**Refractory KD**

Almost one-tenth patients with KD may be refractory to primary IVIG therapy. In such conditions, fever will continue to appear even after more than 36 to 48 h of IVIG therapy. There is no consensus on management protocols to be followed in such patients. AHA guidelines emphasize use of a repeat dose of IVIG (2 mg/kg) in this subgroup. Alternatively, the guidelines reiterate the role of 3 doses of methylprednisolone with tapering prednisolone\textsuperscript{[13]}. Infliximab, given as a single dose of 5-6 mg/kg intravenously, is also very useful in treatment of refractory KD and appears to decrease the chances of developing CAA\textsuperscript{[83]}. Administration of infliximab often results in prompt reduction of fever\textsuperscript{[84,85]}. Tremoulet et al\textsuperscript{[86]} have shown that addition of infliximab in the primary treatment regimen did not reduce the incidence of IVIG resistant KD. However, fever duration, inflammatory markers and reaction rate were less in the infliximab group. There are various ongoing randomized trials to assess the efficacy of anti-TNF drugs. Plasma exchange has also been found to be helpful in patients with intractable KD\textsuperscript{[87,88]}. Other therapeutic options that are being considered includes interleukin-1 antagonist (e.g., anakinara), cyclosporine, and tacrolimus, etc\textsuperscript{[89-91]}.

For the preventing of thrombosis, low dose aspirin remains the first choice of therapy. If the patients show evidence of rapidly expanding CAA, heparin or warfarin anticoagulation along with aspirin can be given\textsuperscript{[13]}. Aspirin with another antiplatelet agent along with systemic anticoagulant agent like heparin or warfarin may be considered for patients with history of coronary thrombosis or giant aneurysm\textsuperscript{[79]}. Thrombolytic treatment or coronary recanalization procedures may be required for the minority of patients who develop coronary thrombosis in the context of KD. Aboximab is also useful in such patients\textsuperscript{[13]}.

**Management after acute phase**

Risk stratification of coronary artery abnormalities is of primary importance for the long term follow-up and management of patients with KD. It is our practice to keep all children with KD on long term follow-up, because there is some concern regarding development of premature atherosclerosis even in children who do not have overt CAA during the acute phases\textsuperscript{[92,93]}. Healthy lifestyle and an active physical regimen should be emphasized upon.

Patients with coronary dilatation that persists beyond 6 wk need to be kept on low dose aspirin for longer periods of time. For patients with large and giant aneurysms, frequent echocardiographic assessment should be continued. Such patients may also require CT coronary angiography at periodic (say 3-5 yearly) intervals. Statins have also been recommended in these situations. Thromboprophylaxis can be maintained with antiplatelet drugs (e.g., aspirin/dipyrimadole used singly or in combination) and anticoagulants (e.g., heparin/warfarin)\textsuperscript{[13]}.

To conclude KD is now one of the most common causes for acquired heart disease in children and all pediatricians need to be familiar with its varied clinical...
presentations. With some experience it is not difficult to pick up children with classic KD. However, the diagnosis of children with incomplete and atypical KD can pose significant issues for the attending pediatrician. The recent AHA 2017 guidelines have suggested a simplified management protocol for children with KD. Therapies other than IVIG are now being increasingly used in these patients.

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