

In some previous investigations, the similar sized aneurysms were defined as "giant" or "large" (1-5). So, we think there are subjective opinions about the definition of aneurysm size in the literature.

Thanks to author for their interest to our case.

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## Incomplete Kawasaki disease: a pediatric diagnostic conflict

### *İnkomplet Kawasaki hastalığı: Pediatrik tanısal zorluk*

Dear Editor,

We read with interest the article "Incomplete Kawasaki disease: a pediatric diagnostic conflict" by Çelik et al (1). They reported two children with incomplete Kawasaki disease (KD) who responded to 2 g/kg single dose intravenous immunoglobulins (IVIG) and 3 mg/kg aspirin (1).

Sonobe et al. (2) recently reported that the coronary artery abnormality (CAA) prevalence of incomplete KD (18.4%) was higher than that of complete KD (14.2%) among 15,857 cases with KD (83.9% complete KD and 16.1% incomplete KD) using the data from the 17th Japanese nationwide survey of KD. Because late diagnosis of KD increases the risk for coronary artery abnormalities, Minich et al. (3) also recently suggested that clinicians should maintain a high index of suspicion of KD in the infant who is younger than 6 months and has prolonged fever even with incomplete criteria. Nevertheless, it is not easy to suspect KD at an early stage of the disease.

Therefore, Sinha et al. reported that erythema at the site of BCG inoculation might be a useful diagnostic tool even for incomplete KD (4). Using this phenomenon, we previously diagnosed and reported a 9-week-old male infant with incomplete KD who was initially treated with high-dose IVIG (2 g/kg) and oral aspirin (100 mg/kg) (5). Initial echocardiography was normal in this patient, but a giant aneurysm of right coronary artery (RCA) was newly developed one week later. Intravenous dexamethasone and oral methotrexate were given due to rapidly progressive coronary artery aneurysm, but those treatments were not effective. On 38th hospital day, we performed coronary angiography, which demonstrated multiple giant aneurysms with sluggish blood flow on the entire RCA and a stenosis on the proximal anterior descending

branch of the left coronary artery. Because he had had a prolonged course of severe coronary involvement refractory to intensive medical therapies, surgical intervention, such as plication of dilated coronary artery, was tried. However, the patient died from acute cardiorespiratory failure shortly after weaning from cardiopulmonary bypass (5).

Although there has been no effective therapy in patients with incomplete KD resistant to IVIG and aspirin, one of our authors previously reported the beneficial effect of low-dose oral methotrexate on 4 patients with Kawasaki disease (age 8 months - 8 years) resistant to IVIG (6). However, methotrexate could not cease the rapid progression of coronary artery aneurysm associated with incomplete KD in our young patient (5).

The diagnosis of Kawasaki disease in very young infants is often difficult because of its rarity and atypical presentation. Although BCG reactivation may help us to suspect incomplete KD at an early stage of the disease, CAA can develop within a relatively short time in contrast to the patients of Çelik et al. (1). Therefore, not only early diagnosis but also more aggressive therapy will be important to prevent sudden cardiac death in incomplete KD and further studies should be performed to elucidate the epidemiology and natural course of incomplete KD in different ethnic populations.

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## Author's reply

Dear Editor,

In Kawasaki disease (KD) early diagnosis and specific treatment is essential to avoid mortality. As the authors emphasize, Kawasaki disease is a still diagnostic dilemma for pediatricians, especially with its atypical or incomplete presentations. Recently, the patients who does not fit the fulfill criteria were considered as incomplete or atypical Kawasaki by the specific signs and exclusion of other causes as we discussed before (1).

Independent predictors were well-defined in KD (2). They have included protracted fever, presumably reflecting worse vasculitis, anemia, elevated white blood count, low albumin, elevated C-reactive protein, male gender and age younger than 1 year (2). As we understood from the author's case, the patient had at least two risk factors (early age and male gender).

There are few randomized trials to guide therapy of the child in whom fever continues or recurs despite initial therapy with IVIG. In a retrospective review of coronary risk factors among KD patients treated with additional IVIG, the number of days of fever before initiation IVIG re-treatment was an independent predictor of coronary artery abnormality (3).

Although randomized trials are lacking most experts believe that children who are febrile, without other explanation, >36 hours after completion of first IVIG infusion should be retreated with IVIG. For children who defervesce with a second IVIG infusion but in whom fever recurs, a third IVIG infusion or alternatively intravenous methylprednisolone 30 mg/kg may be considered (4).

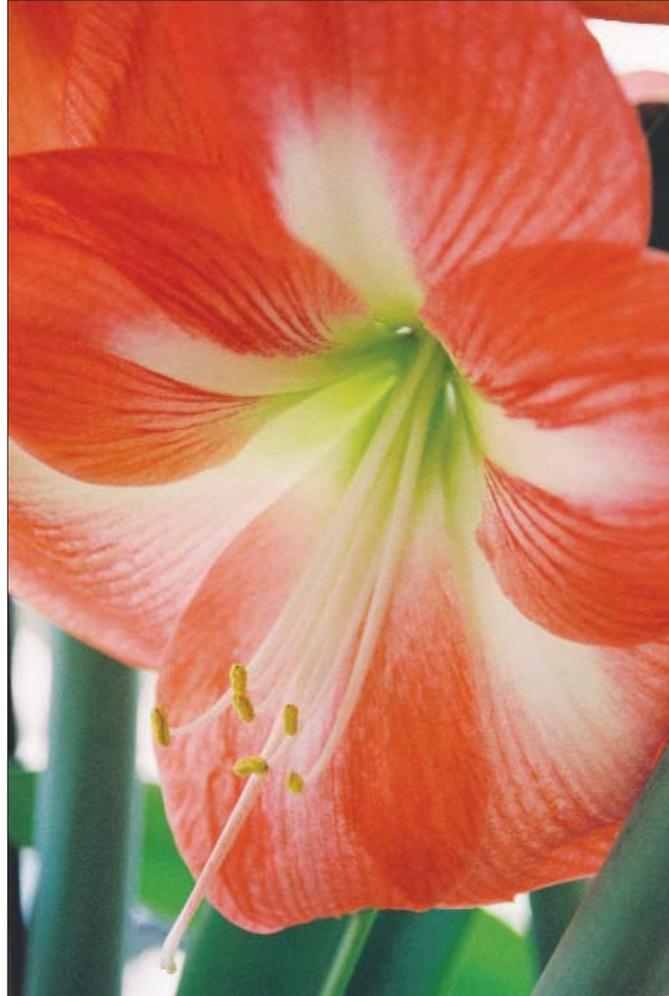
On the other hand, the authors discussed that erythema at the site of BCG inoculation might be a useful diagnostic tool even for incomplete KD. We agree with this opinion. Also, there are some reports about this issue in the literature (5). Clinicians should be aware of this clinical manifestation that could help diagnose incomplete Kawasaki disease.

We thank authors for attention to our report.

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