

Acute cardiac valvular involvement in Kawasaki disease

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Kawasaki disease (KD), a systemic childhood vasculitis, represents the leading cause of acquired heart disease in children in developed countries. Coronary aneurysms are the most known and severe complications, with a reported incidence of about 5% in timely treated patients (1). However, not only can coronary vessels be involved by inflammation, but also the whole heart tissue, determining myocarditis, pericarditis, and rarer valvular involvement (1), described in 1.1%–4.3% of patients (2-5).

In our study, we evaluated the incidence of heart involvement in a cohort of patients affected by KD, with particular attention to echocardiographic evaluation (performed between the second and the fourth week from onset) consistent with coronaritis, pericarditis, myocarditis and valvular involvement. Charts of all patients affected by KD and referred to our hospital (Institute of Child Health IRCCS Burlo Garofolo, Trieste) from 1988 to 2009 were reviewed. Medical records from onset to the end of follow-up were collected. Our population consisted of 45 patients (29 males, 16 females); the median age at diagnosis was 32 months (range 18–53) with 78% of patients <5 years old. Fourteen patients out of 45 (32%) presented atypical and/or incomplete clinical forms: 1 atypical, 4 incomplete, 9 incomplete forms with atypical manifestations, while in the literature the incidence of atypical/incomplete forms is about 20% (1). The mean follow-up time was 4.5 months (SD 5.7 months).

Eleven out of 45 patients (24%) developed cardiac involvement. These patients pre-

sented an overall amount of 17 cardiac manifestations which are as follows: 1 persistent giant coronary aneurysm (only one patient out of 45, 2% of the population) in association with 1 valvular insufficiency, 2 coronary transient thickening, 2 myocarditis (pancarditis) with 2 valvular insufficiency and 2 pericardial pouring, 2 pericarditis with 1 valvular insufficiency and 4 isolated valvular involvements. In our study, cardiac involvement was associated with older age at the onset, higher CRP levels at diagnosis, and lack of response to the first IVIG infusion, even though these findings did not reach statistical significance because of the small population. In four out of 8 patients, valvular insufficiency was associated with other cardiac complications: coronary aneurysm in one patient, myocarditis in two patients and pericarditis in one patient. We found an uncommonly high incidence of heart valves involvement (8 out of 45 patients; 18% of cases) consisting of a mild insufficiency of atrio-ventricular heart valves (5 mitral insufficiency, 1 tricuspidal insufficiency, 2 both mitral and tricuspidal). Only in patients with myocarditis, valves involvement presented itself as a systolic murmur in association with S3 gallop, whilst it was totally asymptomatic in all other patients.

As for the therapy, all these patients received IVIG within the first 10 days of fever, following the guideline recommendations; because of failure of the first IVIG infusion, three patients needed a second dose of IVIG, and in four patients corticosteroids were associated. Concerning the treatment, there was no significative difference between patients with and without valvular involvement. A complete recovery was achieved in all patients during the follow-up, within five weeks from onset.

In conclusion, despite the small number of patients, our study reports a relatively high frequency of valvular involvement in KD (18%), never found before. It was associated with other cardiac manifestations, or found isolated and completely asymptomatic. We would like to underline that valvular involvement in KD seems to be a benign condition, which resolves itself completely in all patients. These findings need to be confirmed by further studies.

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