INTRODUCTION

Kawasaki Disease (KD) is an acute febrile childhood disease capable of damaging the coronary arteries and causing aneurysms, which can then become complicated with thrombosis or coronary obstruction, leading to myocardial ischemia.

METHODS

Between October 1988 and April 2004, we assessed 150 children with KD. The mean age was 44.8 months (range, 3-114); 56 were girls and 94 were boys. Patients with “typical” American Heart Association (AHA) criteria were included: prolonged fever (≥ 5 days) plus 4 of the following criteria: a) changes in extremities, b) polymorphic exanthem, c) bilateral conjunctivitis without exudate, d) changes in oral cavity, and e) cervical lymphadenopathy. “Atypical or incomplete” cases with fever ≥ 5 days and fewer than 4 criteria, but with coronary artery complications on echocardiography, were also included.

Laboratory tests, including complete blood counts, erythrocyte sedimentation rate (ESR), and platelet count, were performed. The cardiac complications of KD were assessed by color Doppler echocardiography from the fifth day, except in three patients due to a late echocardiographic finding, a retrospective diagnosis of KD in one child with angina, and a postmortem diagnosis in 1 infant with prolonged fever. The presence of pancarditis was a predictor of a giant coronary artery aneurysm. Mortality was 3.7%. The presence of pancarditis was a predictor of a giant coronary artery aneurysm. The coronary abnormalities were classified according to severity as follows: diffuse ectasia in 40.7%; a small-to-medium-sized solitary coronary artery aneurysm in 33.3%; numerous small-to-medium-sized aneurysms in 11.1%; and coronary artery stenosis in 3.7%. The presence of pancarditis was a predictor of a giant coronary artery aneurysm.

The aim of this study was to determine risk factors for coronary lesions, the type of heart lesion present, and long-term outcome in Kawasaki disease. We studied 150 children, aged 3 months to 9.5 years, who met the criteria for a diagnosis of Kawasaki disease. Of the 18% who were diagnosed with heart disease, 100% had coronary artery abnormalities, 11.1% had pancarditis, and 3.7%, mitral insufficiency. The coronary abnormalities were classified according to severity as follows: diffuse ectasia in 40.7%; a small-to-medium-sized solitary coronary artery aneurysm in 33.3%; numerous small-to-medium-sized aneurysms in 11.1%; and coronary artery stenosis in 3.7%. The presence of pancarditis was a predictor of a giant coronary artery aneurysm.

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Enfermedad de Kawasaki: afección cardiaca durante la infancia

Con objeto de determinar los factores de riesgo para la coronariopatía, el tipo de lesión cardíaca y la evolución a largo plazo en la enfermedad de Kawasaki, evaluamos a 150 niños con criterios de la enfermedad, de 3 meses a 9,5 años de edad. El 18% desarrolló cardiopatía, en todos los casos lesiones coronarias: el 11,1%, pancarditis y el 3,7%, insuficiencia mitral. Las lesiones coronarias fueron clasificadas en: ectasia difusa en el 40,7%, aneurisma solitario en el 33,3%, aneurismas múltiples en el 11,1%, aneurismas gigantes en el 11,1% y estenosis coronaria en el 3,7%. La pancarditis fue prediccora de aneurismas gigantes. La mortalidad fue del 3,7%. Los aneurismas coronarios fueron las lesiones cardiacas predominantes. La edad menor de 27 meses, la febrícula durante más de 8 días, la velocidad de sedimentación globular > 70 mm y la pancarditis fueron factores de riesgo para aneurismas coronarios.

Palabras clave: Enfermedad de Kawasaki. Aneurismas coronarios.

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and degree of valvular, myocardial, and pericardial compromise were determined, as well as the presence and type of coronary lesions, which were classified as follows:

1. Diffuse ectasia or dilation (diameter greater than that expected for the body surface area). This was also indicated whether the aneurysm was single or multiple, small and/or medium-sized (up to 8 mm) or giant (over 8 mm).

2. Coronary aneurysm (segmental dilation >1.5 times the adjacent segment); it was also indicated whether the aneurysm was single or multiple, small and/or medium-sized (up to 8 mm) or giant (over 8 mm).

3. Coronary stenosis.

Echocardiography was repeated in 4-6 weeks and at 1 year from the onset of symptoms in patients with no heart disease, every 6 months in those who had small to moderately sized aneurysms, and every 1-3 months in children with giant aneurysms. From 5 years of age, the patients were evaluated by a yearly stress test. Patients with residual moderate or giant coronary aneurysms also underwent perfusion myocardial testing (single photon emission computed tomography [SPECT]) at rest and with pharmacologic challenge every two years after 10 years of age. Coronary angiography was indicated in a child who consulted for angina and experienced an acute myocardial infarction (AMI).

The comparison of quantitative variables between patients who developed an aneurysm and those who did not was done by one-way analysis of variance in the case of parametric distributions and the Mann-Whitney/Wilcoxon test for 2 samples in the case of nonparametric distributions. The association between the qualitative characteristics and the development of aneurysms was assessed by a $\chi^2$ test using the Yates correction or by Fisher’s exact test.

**RESULTS**

A total of 27 patients (18%) experienced cardiac complications. Mitral regurgitation was mild, transient, and uncommon (3.7%). Three male patients who developed an aneurysm presented pancarditis (Figure). There were no cases of aortic valvulitis.

All had coronary lesions, which were classified as follows:

- Group 1: transient dilation (ectasia) in 11 patients.
- Group 2: single aneurysm of small or moderate size in 9 patients (group 2a) and multiple aneurysms of small or moderate size in 3 patients (group 2b).
- Group 3: giant coronary aneurysms in 3 patients.
- Group 4: coronary stenosis in 1 patient (Table 1).

Follow-up was from 1 to 16 years (mean, 8.5 years). In total, 147 children received treatment following the diagnosis: oral acetylsalicylic acid (ASA) (80 mg/kg/day) and intravenous gamma globulin (400 mg/kg for 4

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**TABLE 1. Classification of Patients With Coronary Aneurysms According to Severity and Risk**

<table>
<thead>
<tr>
<th>Risk Group</th>
<th>Patients, n</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>11</td>
<td>40.7</td>
</tr>
<tr>
<td>2a</td>
<td>9</td>
<td>33.3</td>
</tr>
<tr>
<td>2b</td>
<td>3</td>
<td>11.1</td>
</tr>
<tr>
<td>3</td>
<td>3</td>
<td>11.1</td>
</tr>
<tr>
<td>4</td>
<td>1</td>
<td>3.7</td>
</tr>
</tbody>
</table>

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Figure. Echocardiographic image (short axis of large vessels) showing coronary aneurysms. Ao indicates aorta; LAD, left anterior descending artery; LCA, left coronary artery; RCA, right coronary artery.
If the patient
In our population,
9
as occurred in 1
and is predominantly
Our
In our series, mitral
P
P
389

Boys were affected more often in both groups, but only
under 2 years and in school-aged children (5-10 years).
coronary compromise.

We compared the clinical and laboratory data of the
patients without (group A) and with (group B)
coronary compromise.

In group B, coronary lesions predominated in children
under 2 years and in school-aged children (5-10 years).
Boys were affected more often in both groups, but only
boys had serious coronary lesions. In group B, 44%
presented typical KD and 56%, incomplete forms of the
disease (without statistical significance).

Group B had more prolonged fever than group A (8 vs
6 days; P<.05) and the average ESR was 70 mm in
group B and 48 mm in group A (P<.05) (Table 2).
There were no differences in the number of leukocytes
or platelets.

In the patients with coronary ectasia, diameters
returned to normal within the first 45 days of KD; the
aneurysm regressed in 100% of group 2a patients within
the first 3 years of follow-up and persisted in 66% of
group 2b patients, although without complications during
clinical follow-up and with stress and/or myocardial
perfusion testing.

Among the 4 patients with giant aneurysms, 1 died
undiagnosed; another had an AMI (which required
coronary bypass surgery) and the other 2 were
asymptomatic, with no ischemia detectable by noninvasive
tests (3 and 15 years of follow-up).

DISCUSSION
Kawasaki disease is an acute vasculitis of unknown
origin that occurs in childhood and is predominantly
observed among boys under 5 years of age. Our
patients under 27 months, but also those of school age
(5-10 years), had a higher incidence of cardiac
complications. Kawasaki disease can affect the valvular endocardium,
myocardium, and/or pericardium. In our series, mitral
valvulitis was mild and transient, although pancarditis
was always accompanied by giant aneurysms, which
determined an adverse prognosis.

Among children with coronary lesions, 56% presented
an incomplete form of the disease, an incidence similar
to that described in the literature. In our population,
males, age <27 months, prolonged fever, ESR>70
mm, and pancarditis were factors associated with
coronary abnormalities.

Generalized microvasculitis is expressed in 30%-50%
of the patients as coronary ectasia. In all of these patients
the condition resolved within the first 45 days of the
disease. Coronary aneurysms can regress or progress.
All aneurysms in group 2a involuted, compared with
only 66% in group 2b. Giant aneurysms accounted for
15% of the total. These aneurysms do not involute, but

can lead to obstruction.

Stress testing and myocardial perfusion (SPECT)
images were used to detect myocardial ischemia. If
Coronary angiography should be performed if the patient
is symptomatic or the noninvasive studies indicate
myocardial ischemia, in order to assess feasibility of
myocardial revascularization surgery, as occurred in 1
patient in our series.

CONCLUSIONS
Coronary aneurysms were the predominant cardiac
lesions in our series. The risk factors for coronary
aneurysms were age <27 months, prolonged fever of
more than 8 days, ESR>70 mm, and pancarditis.

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**TABLE 2. Clinical Characteristics of Patients With Kawasaki Disease According to Whether or Not Coronary Aneurysms Developed**

<table>
<thead>
<tr>
<th>Patient Type</th>
<th>Age, Median</th>
<th>Sex</th>
<th>Days of Fever, Average (P&lt;.05)</th>
<th>ESR (P&lt;.05)</th>
<th>Complete Clinical Evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>No aneurysms</td>
<td>43.5 months</td>
<td>Male, 60%</td>
<td>6 days</td>
<td>48 mm</td>
<td>28%</td>
</tr>
<tr>
<td>Aneurysms</td>
<td>27 months</td>
<td>Male, 66%</td>
<td>8 days</td>
<td>71 mm</td>
<td>44%</td>
</tr>
</tbody>
</table>

*ESR indicates erythrocyte sedimentation rate.*