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CORRESPONDENCE

Congenital heart disease, genetic syndromes, and major noncardiac malformations

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In a recent issue of your journal, we read the very interesting paper by Wren et al. [3] on mortality in infants with congenital heart disease (CHD). The authors reported that the deaths were not related to cardiovascular malformations in 33 % of all patients: in 57 % of infants with additional chromosomal or genetic abnormalities, in 76 % of infants with major noncardiac malformations, and in only 16 % of infants with isolated cardiac defects [3].

The decline of mortality occurred in all groups of patients but was marked in infants with isolated CHD [1]. Moreover, the proportion of postoperative deaths due to noncardiac cause increased from 9 % in the first 10 years of the study to 29 % in the second 10 years [3]. These data emphasize the role of additional noncardiac defects for the prognosis of children with CHD since these subjects with multiple and complex defects represent about one third of all CHD and can be considered one of the next challenges for pediatric cardiologists and cardiac surgeons [1, 2].

In analyzing mortality of patients with CHD associated with chromosomal and genetic syndromes, we must take into account not only the role of extracardiac malformations but also the specific cardiovascular patterns that in some occasions include peculiar additional cardiac defects that may influence cardiac surgery [1]. For this group of patients, an early and accurate multispecialistic approach is mandatory, and a careful assessment of cardiac and extracardiac risk factors can contribute to the preparation of genetic-specific diagnostic and treatment protocols [1]. Moreover, a specific follow-up targeted to cardiac and extracardiac problems is mandatory for these patients [1]. In the field of pediatric cardiology, accurate epidemiologic studies like this by Wren at al. [3] are very helpful to understand the significant role of additional genetic syndromes and major extracardiac defects in the mortality and the morbidity of children with CHD.

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