Aortic Coarctation. Important Considerations in Long-Term Follow-up After Correction

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It is well known that patients with unrepaired aortic coarctation have a poor clinical course and short life expectancy—an average of 30 years—and that this is basically caused by secondary disease arising from arterial hypertension. Repair of the anatomical defect in childhood or in adults is not curative. Many of these patients have abnormalities in cardiovascular regulation, significant long-term morbidity and, thus, shorter life expectancy. Early repair, even in the first months of life, has demonstrated a high incidence of hypertension, reaching 30% among patients during 10 years of follow-up, despite having achieved a good anatomical result.[1-3]

The current issue of the Journal presents a study by Balderrábano-Saucedo et al,[4] that included 40 patients undergoing aortic coarctation repair with a residual gradient of 15 mm Hg and normal blood pressure at rest. Patients with concomitant congenital heart disease were excluded from the study, some of whom had bicuspid valve disease. Various echocardiographic indices of left ventricular function were determined, among which the myocardial performance index stands out. This parameter was significantly greater in the patients than in the control group; the most relevant associated factors were being older than 4 years at the time of repair and having very abnormal left ventricular morphology due to hypertrophy or dilatation. As recognized by the authors, the limitations of the study are nonhomogeneous follow-up time and not having determined blood pressure during exercise or daily activity. It is essential to determine these in the future to better support the conclusions of this study. Despite this, the article reminds us of the need to explore more thoroughly the issue of patients with successfully repaired aortic coarctation and who should not be incorrectly considered "cured" such that appropriate long-term medical follow-up is ensured.

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It is well known that the leading causes of morbidity and mortality in aortic coarctation patients after repair are complications attributable to hypertension, early coronary disease, left ventricular failure, and complications of the aortic wall (aneurysms, dissection, and rupture).[1-3]

In 1989, Cohen et al[5] published a study which analyzed the long-term outcomes of aortic coarctation patients after repair, and found that age at the time of repair was an important variable in the prognosis of these patients. Thus, they established a limit of 14 years of age in order to reduce the incidence of persistent systemic hypertension and cardiovascular complications, and improve survival.

In their series, similar results were presented by Cervantes-Salazar et al[6] who found a cumulative survival rate at 120 months of 89% in patients who had undergone intervention at less than 10 years of age and of 80% in patients who were older at the time of intervention. When the incidence of hypertension was analyzed in relation to age at the time of repair, they found that this complication occurred in 20% of the patients who underwent repair at more than 10 years of age and only in 1.7% of those who underwent repair at a younger age.

These studies and others[7,8] support aortic coarctation repair at young ages, since this has a significant impact on morbidity. However, there is increasing evidence suggesting the need to classify this entity as a general disease of the cardiovascular system, in which persistent abnormalities in vascular regulation can explain the reduction in life expectancy among these patients.

The cause of late hypertension in patients who have undergone aortic coarctation repair by surgery or intervention remains unknown. Among others, the following have been suggested: poor response to vasoactive agents, aortic arch geometry abnormalities, and alterations in renal function with increased renin-angiotensin activity and in baroreflex function.[8-10]

The results of the study by Polson et al[11] indicated autonomic dysfunction in neonates who underwent

aortic coarctation repair. It is possible that the reduced sensitivity of the baroreceptor reflex is secondary to the circulatory changes produced by aortic coarctation, whose underlying mechanism would be renal hypoperfusion and increased renin-angiotensin activity, with the consequent abnormalities in the central nervous system. This hypothesis is based on clinical data that indicate that the baroreceptor response is abnormal in normotensive patients with a history of risk factors for hypertension.

Left ventricular mass is a well-established predictor of possible cardiovascular events, and can continue to increase in normotensive patients who have undergone aortic coarctation repair. After repair, aortic coarctation patients with normal resting blood pressure frequently have an abnormal pressure response on exercise and even during daily activities. This situation has clinical relevance, since it can also alter the cardiovascular pattern and lead to damage in the target organs.[12]

Abnormalities can be detected early using the new diagnostic methods to assess endothelial function and vascular structure (flow-mediated vasodilatation or arterial intima-media thickening). In aortic coarctation patients who have undergone successful repair, that is, with a minimum residual gradient, significant deterioration in endothelial function has been observed that could explain the development of late hypertension and atherosclerosis.[13] It has been shown that the composition of the arterial wall in the vascular bed differs before the coarctation site, with increased collagen content and less smooth muscle mass.[14] Changes have been found, many years after repair, in vascular function in children who underwent early aortic coarctation repair, indicating that such alterations may not be reversible. Although it seems that early aortic coarctation repair preserves the elastic properties of the artery, it does not seem to affect endothelial response. This suggests programming of vascular reactivity that could be determined by the underlying abnormal hemodynamics in the prenatal period or in the first days or week of life.[15]

Recent studies have aimed at evaluating the possible effect of various drugs to reverse the impaired endothelial function and inflammatory process in normotensive patients with successfully repaired aortic coarctation. Of special interest is the work of Brili et al,[16] in which 20 patients—age 27.3 (2.4) years who had undergone aortic coarctation repair 13.9 (2.2) years previously—received ramipril 5 mg/d for 4 weeks and that significantly improved endothelial function and decreased the expression of proinflammatory cytokines such as interleukin 6. These findings suggest that this treatment may modify the atherogenic process in patients with successfully repaired aortic coarctation, even in the absence of hypertension.

Taking the foregoing into account, it should be emphasized that it is not possible to approach the patient with aortic coarctation, whether a child or adult, as the carrier of a "simple" congenital malformation. Furthermore, strict follow-up protocols should be established to provide useful information and make it possible to eventually prescribe treatments that can modify or delay cardiovascular alterations and improve quality of life and life expectancy of these patients.

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