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Editorial Comment

A Long Look at Surgery for Coarctation of Aorta*

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Forty years after Crafoord and Nylin (1) in Stockholm and Gross and Hufnagel (2) in Boston independently reported on surgery to relieve coarctation of the aorta, it is appropriate to review what has been accomplished and what remains to be achieved. Early literature (3) had provided ample reasons for seeking surgical treatment. Although one man with coarctation lived 92 years and died of an unrelated cause, most of the deaths could be attributed to the malformation: sudden death and death from cardiac failure, hypertensive cardiovascular-renal disease, cerebral hemorrhage, ruptured aneurysm of the aorta or of the circle of Willis and endocarditis. Skills in preoperative diagnosis, surgical techniques and postoperative management quickly brought down operative risk of mortality and serious complications such as spinal cord injury. Early results of surgery were impressive for normalization of arm blood pressure and disappearance of symptoms. But what has happened long term?

The report from Johns Hopkins by Maron and colleagues (4) on patients operated on between 1945 and 1960 and reevaluated 11 to 25 years later was one of the first to suggest that all did not live healthily ever after. Premature disability (78%) and death (12%) related to cardiovascular disease, and in turn to the coarctation, still occurred. Some of those problems related to the fact that early surgical experience not only involved the learning curve but also included the most severe and symptomatic patients and those who had reached adulthood before surgery became possible. One might anticipate, therefore, that some deleterious effects had occurred which would be only partially reversible if at all.

The long-term follow-up (mean 15 to 29 years) from Stockholm (5) of patients operated on there from the first patient in 1945 indicated that age at operation was important. Among 11 reevaluated patients aged 5 to 15 years at operation, all were in New York Heart Association functional class I and only 1 had hypertension, whereas among 13 patients operated on between the ages of 16 and 31 years, 10 were in class I and 6 had hypertension. After age 31, only 6 of the 13 were in class I and 6 had hypertension.

Over the years we have learned that silent or insignificant abnormalities can develop and worsen. Though that is not the main thrust of the paper by Carpenter and colleagues (6) in this issue of the Journal, it is noteworthy that 10 of the 48 patients who responded to their invitation for follow-up study 2 to 27 years after the operation had significant aortic valve dysfunction or other cardiac defects while 5 of 32 who qualified for inclusion had "trivial" aortic regurgitation, 3 had mild systolic hypertension at rest, 7 had a significant, verifiable systolic pressure gradient between arm and leg, determined by repeated cuff measurements and 9 had a gradient by Doppler recording of 20 to 40 mm Hg. Three showed late progression from left ventricular hypertrophy to "strain." This selected "normotensive" group of 32 patients fared better with exercise than many of our postoperative patients, for they did not demonstrate a hypertensive response to exercise even though systolic pressure rose to over 200 mm Hg in some cases. Indeed, their response in heart rate and in blood pressure to the stress of supine bicycle exercise did not differ from that of adults in the control group.

Postoperative left ventricular function and hypertrophy. Though these 32 patients did not have complete correction of cardiovascular disease, they nonetheless represent a favored group with few obvious signs or symptoms of cardiac disability. It comes as a shock, therefore, to learn that on the basis of sophisticated study by radionuclide cineangiocardiography at rest and on exercise, they demonstrated left ventricular anatomic hypertrophy and physiologic hyperkinesia. Left ventricular mass averaged 120 ± 10 g/m^2 in contrast to 87 ± 10 g/m^2 for control subjects. Mean left ventricular ejection fraction was significantly elevated at rest and on exercise, while systolic ejection rate was significantly increased and end-systolic volume decreased. Indeed, 13 of 32 showed complete or nearly complete systolic cavity obliteration. The control group of 22 healthy adults, aged 23 to 49 years, did not show these abnormalities. Neither age at operation nor age on follow-up correlated with the results.

What caused this left ventricular abnormality in mass and function? The authors speculated that once the left ventricle hypertrophied in response to outflow obstruction, altered geometry did not completely normalize, so function was inefficient and this led to the need for greater than normal muscle mass. That in turn is associated with increased ejection fraction and shortened systolic ejection period as well as increased oxygen consumption. A mismatch of increasing

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muscle mass and decreasing coronary supply could bode ill for these patients in middle or late adult life, especially if coronary artery disease were to be superimposed.

Role of sympathetic nervous system. Additionally, the authors speculated on the role of the sympathetic nervous system in the postoperative hyperkinesia. Autonomic imbalance before surgery and continuing after surgery could, through chronic elevation of catecholamines, particularly norepinephrine, maintain high sympathetic tone, increase ejection fraction and sustain myocardial hypertrophy. Since chronic marked elevation of catecholamines, for instance in pheochromocytoma, can cause cardiomyopathy, perhaps lesser elevation over a longer span of years could result in the myopathy that the authors suspected in at least one and perhaps three postoperative patients. I wonder how the mismatch in the peripheral vascular bed proximal and distal to the coarctation preoperatively might set the stage for abnormalities of control of the sympathetic nervous system vis-à-vis stimulation and response.

Clinical implications. Many questions come to mind. 1) If these abnormalities are found in such a selected and "benign" group of normotensive patients years after coarctation repair, should they be even more obvious and easy to detect in those with symptoms and recognized cardiovascular abnormality? 2) How does the left ventricle function preoperatively and at various age periods after operation? In this study, three of the four children studied within 4 years of surgery had an abnormal ejection fraction and two had cavity obliteration. 3) How do patients with operative relief of aortic stenosis fare if studied in this manner? In that condition there is not the disparity in the peripheral vascular bed that exists in coarctation above and below the obstruction. 4) If hyperkinesia is a forerunner of essential hypertension as has been suggested, would radionuclide cineangiocardiographic studies at rest and on exercise detect a population at risk for developing that condition? If hyperkinesia is important in essential hypertension, why were 28 of these 32 patients not hypertensive at rest or on exercise?

Insofar as coarctation of the aorta is concerned, we know at least four truths: 1) most patients with significant coarctation benefit from operation; 2) we do not yet know the ideal age for surgery; 3) surgery offers repair or relief but not total correction; and 4) these patients should continue under long-term follow-up so that we may continue to learn, advise and manage more wisely each year.

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