Myocardial Dysfunction in Active Rheumatic Carditis

The recent report by Essop et al. (1) from South Africa found no echocardiographic evidence of left ventricular myocardial dysfunction in 32 young patients (mean age 14 years) with active rheumatic carditis and congestive failure. All had isolated mitral regurgitation or combined mitral and aortic regurgitation. Echocardiographic studies were completed before and after prosthetic mitral or mitral and aortic valve replacement. Preoperatively, the percent fractional shortening was increased, and both left ventricular systolic and diastolic dimensions were increased. After surgical restoration of valvular competence, left ventricular measurements returned to normal, and the increased fractional shortening returned to control values.

This study is at marked variance with the time-honored concept taught for decades that myocarditis and not mitral regurgitation was the cause of cardiac enlargement and congestive heart failure in children with acute rheumatic carditis. This concept had considerable clinical support because those patients who slowly recovered from severe carditis were usually left with significant mitral regurgitation, which, in turn, was remarkably well tolerated for years. When patients did succumb from what was considered to be “toxic rheumatic myocarditis,” at autopsy the mitral valve appeared remarkably benign with minimal edema and some marginal vegetations. There was no contracture of the mitral leaflets or of the chordae that would have permitted significant mitral regurgitation. On the other hand, these patients had histologic confirmation of myocarditis with the presence of Aschoff bodies and infiltrates. Thus, the conclusion was well supported both by clinical experience and findings at autopsy.

When echocardiography became available, studies conducted in South Africa revealed a totally different cause of mitral regurgitation, which was found to be due to a combination of annular dilation and chordal elongation of the anterior leaflet (2). The chordal elongation caused the apposing edge of the anterior leaflet to prolapse into the left atrium, allowing mitral regurgitation with a characteristic jet striking the posterior wall of the left atrium. With increasing prolapse, the anterior leaflet could actually become flail, permitting massive mitral regurgitation. (Chordal elongation noted in earlier postmortem examination was misinterpreted as a compensatory response to left ventricular dilation [3].)

The decision to restore valvular competence, either by valvuloplasty or valve replacement in the face of active carditis was indeed a bold step forward. However, nearly all of these patients experienced a rapid and often dramatic improvement, seriously challenging the concept of myocarditis being responsible for cardiac failure (4). The well designed and adequately controlled study of Essop et al. (1) supports the extensive clinical experience in South Africa (5) as well as the limited experience in France (6) and the United States (7). The overall experience over the past 10 years quite emphatically disproves the concept that congestive failure in active rheumatic carditis is due to myocardial dysfunction.

The Editorial Comment by the highly respected Drs. Edwards (8) was extremely disappointing. Rather than truly addressing the question, “Congestive heart failure in rheumatic carditis: valvular or myocardial in origin?,” their comments were directed toward the difficulty determining whether the active carditis represented the initial or a recurrent episode. Begging the question in this manner is totally irrelevant because it makes little difference whether the active carditis is the first, second or third episode of rheumatic activity. The answer should have been, and clearly is, that mitral or mitral and aortic regurgitation, not myocarditis, is the cause of congestive failure in active rheumatic carditis in young patients.

Rheumatic fever is not a big problem in our country, and, fortunately, severe fulminating active rheumatic carditis is extremely rare. Even so, it is time to incorporate the extensive experience and new understanding provided by our South African colleagues in our consideration of this persistent problem.

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References

Reply

The comments of Veasy are of interest. It is indeed correct that in the presurgical era, some patients with severe carditis and congestive heart failure were able to make sufficient recovery to be able to tolerate underlying mitral regurgitation for many years. Thus does not, however, necessarily imply that the observed improvement reflected an amelioration of myocarditis with consequent improvement in left ventricular contractile function. An equally plausible explanation would relate clinical improvement to temporal changes in ventricular loading conditions, with transformation of acute mitral regurgitation to a subacute or chronic compensated form. The latter, whether of rheumatic or other etiology, may be tolerated for considerable periods of time. The patients included in our study comprised a group with such severe degrees of mitral regurgitation that despite cessation of physical activity and optimal pharmacologic therapy, heart failure remained refractory, necessitating surgical intervention.

As pointed out by Veasy, the relatively normal gross appearance of the mitral valve at surgery is beguiling. Inability to appreciate spatial relations and leaflet coaptation margins in the flaccid non-