potentially denying patients access to surgery that was feasible. The prognosis for unoperated severe symptomatic AS was very poor, reinforcing the role of intervention. Although testing is still preliminary, less invasive delivery of devices for the treatment of severe AS may find a role in patients with truly prohibitive operative risk.

It could be attractive to assume that these data are unique to a single institution. However, the surveyed institution has an active program in the treatment of valvular heart disease. The authors believe that these findings are likely reflective of general practice.

Letters to the Editor

Myocardial Late Enhancement in Duchenne and Becker Individuals in the Clinical Milieu

We read with interest the study by Silva et al. (1) of myocardial delayed enhancement (MDE) in patients with muscular dystrophy. This study focuses on 10 patients, 8 Duchenne muscular dystrophy (DMD) and 2 Becker muscular dystrophy (BMD) individuals age 7 to 18 years, 2 having dilated cardiomyopathy (DCM). It raises several issues.

The authors reported a significant correlation between MDE, as a marker of myocardial fibrosis (MF), and low left ventricular ejection fraction (EF) values.

Although only 2 patients had MF and abnormal echocardiography, their EF values were not shown. Similarly, the correlation between EF and MF in the 5 patients with MF and normal echocardiography should be focused on, as a wide variability in EF data (37.3% to 59.3%) was observed.

Correlation between contractile dysfunction and MF seems ambiguous, as 56.2% of dysfunctional segments had MF, whereas 43.8% did not.

Lack of muscle biopsy renders the diagnosis of 4 patients incomplete: Patients #2 and #3 diagnosed as DMD, carrying an in-frame (Becker) deletion. Patients #5 and #6 (siblings) were diagnosed as BMD, despite the absence of muscle biopsy data. In these patients without cardiomyopathy, the differential diagnosis with limb-girdle dystrophy should be considered.

Seven patients had no deletions of the dystrophin gene. Surprisingly, in these patients, screening for duplications or splicing mutations was not performed. Indeed, Patients #1, #5, and #6 (siblings) were diagnosed as BMD, despite the absence of muscle biopsy data. In these patients without cardiomyopathy, the differential diagnosis with limb-girdle dystrophy should be considered.

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The authors should be highly critical in their assumptions, emphasizing the uncertainties of the conclusions, specifically in view of the small study group.

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Reply

We appreciate the letter written in response to our recent article (1). We address the questions of Dr. Giglio and colleagues in the following text.

We demonstrated a clear correlation between amount of myocardial delayed enhancement and lower left ventricular ejection fraction (LVEF). Although only Patients #3 and #9 had low LVEF (47% and 42%) by echocardiography, they had even lower LVEF by CMR (36.7% and 33.6%). Additionally, cardiovascular magnetic resonance (CMR) detected 6 patients with mild to moderate LV dysfunction (4 patients) or low-normal LVEF (2 patients).

We disagree that the focus should be on the wide variability of ejection fraction. Despite the variability (small n), we noted a large and significant difference in LVEF by CMR in patients with versus without MF (48.3 ± 11.0% vs. 66.1 ± 0.6%, p = 0.02), whereas no difference was observed by echocardiography (52.4 ± 8.4% vs. 59.6 ± 5.5%, p = NS). Thus, CMR detected subtle LV function changes in patients with myocardial fibrosis (MF). Even in the range of normal to mild dysfunction, the correlation held true: the higher the amount of MF, the lower the LVEF.

We disagree that contractile dysfunction and MF seem ambiguous. Dysfunctional segments can influence adjacent segment contraction, even those without MF, by a complex relationship that depends critically on the amount of segmental MF, global LV function, and remodeling (2,3).

Lack of muscle biopsy did not turn the final diagnosis incomplete. Patients #2 and #3 have a typical clinical course of Duchenne muscular dystrophy, becoming wheelchair bound before 10 years old. Patients #5 and #6 were siblings and have a positive familial history of X-linked inheritance, excluding limb-girdle dystrophy hypothesis (4). All patients were screened for deletions. Seven cases had no deletions found, with 4 positive muscle biopsy (absence of dystrophin, Patients #7, #8, #9, and #10) confirming the diagnosis. We stated that, in only 3 cases (no exon deletions or biopsy), the diagnosis was based on the classical phenotype and typical familial history.

We apologize for not having cited the article by Giglio et al. (5), which has elegantly demonstrated the capability of ultrasonic tissue characterization in Duchenne patients.

Our conclusions are based solely on CMR’s capability of identifying MF in the myocardium of this small, but sufficient, group of patients. Our conclusion was that CMR is able to detect MF in muscular dystrophy patients earlier than routine cardiologic evaluation, not including any widely unavailable sophisticated technology.

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