The Child With Dilated Cardiomyopathy: Prognostic Considerations and Management Decisions*

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The published data on dilated cardiomyopathy in children are sparse and little is known about the long-term clinical course and the factors that may influence prognosis for better or worse. Such information is badly needed. As cardiac transplantation becomes more effective, the proper timing for heart replacement in a child with cardiomyopathy becomes critical. The death of a young patient with cardiomyopathy who could have survived with a transplanted heart is a tragedy. On the other hand, premature transplantation in a child who is destined to lead a relatively normal existence for a number of years presents risks, morbidity, life-style requirements and costs that are unnecessary and unacceptable.

Management of dilated cardiomyopathy in children. What do we know and what do we need to know to better manage children with dilated cardiomyopathy? Etiology is a primary consideration. Progress is being made, but the underlying cause for the vast majority of cases of dilated cardiomyopathy remains unknown. Undoubtedly there are multiple causes, often leading to a final pathway of clinically similar myopathies. Without basic knowledge preventive measures are not possible. On a clinical basis, physicians cannot look to prospective studies for guidance regarding treatment of children with dilated cardiomyopathy. Reliable predictive risk factors have not been identified by multivariate analyses. Thus, pediatric cardiologists must turn to retrospective studies as an adjunct to clinical judgment in considering management options. There have been a scattering of such reports (1-5) concerning dilated cardiomyopathy in children and only limited data are available.

Thus, as might be expected, only very basic conclusions have emerged that are generally accepted by the medical community: 1) Some children with established cardiac amyotrophy live a long time in a relatively stable clinical state; and 2) patients with progressive myocardial dysfunction associated with symptoms of congestive heart failure or serious rhythm disturbances, or both, have a poor prognosis. An important controversial question that remains unanswered for both adults and children with dilated cardiomyopathy is whether cardiac arrhythmias represent an independent risk factor. Most of the published discussion concerning rhythm disturbances in dilated cardiomyopathy has focused on ventricular arrhythmias, but a report (6) in this issue of the Journal has slightly shifted the emphasis in the pediatric age group to supraventricular arrhythmias—specifically, atrial fibrillation and flutter.

The present study. Friedman et al. (6) reviewed the data in 63 children with dilated cardiomyopathy. They present some important information on natural history. The 5 year survival rate was in the range of 80% and most deaths occurred in the first few years after diagnosis. Other series (1-5) have reported higher mortality rates. As emphasized in these published studies, symptomatic chronic congestive heart failure was associated with a poor prognosis. Some information regarding the effect of arrhythmias is presented in the report of Friedman et al. (6). However, as with other retrospective studies, there are few firm predictive data. Five of 29 patients in whom an arrhythmia was identified during exercise study or Holter ambulatory electrocardiographic (ECG) monitoring died. Among 34 patients with no documented arrhythmias, 5 patients died but none had had Holter monitor studies. Three of the 10 deaths were classified as sudden. It is very likely that the occurrence of sudden death is related to a rhythm disturbance whether or not mild (frequent premature ventricular complexes) or severe (ventricular tachycardia, atrial fibrillation/flutter) rhythm disturbances were noted earlier.

Ventricular versus atrial arrhythmias. The relation of ventricular and atrial arrhythmias to outcome has not been clear, although several publications (1-6) appear to confirm that patients who have cardiac arrhythmias in the presence of heart failure have a poor prognosis. In contrast to others, Friedman et al. (6) found that atrial arrhythmias were more common than ventricular rhythm disorders. Atrial fibrillation/flutter was documented in 10 patients and ventricular tachycardia was noted in 6 children during their clinical course. However, including these patients and others who had less significant ectopic activity, the authors (6) conclude that "... the presence or absence of any type of arrhythmia was not predictive of death, having an odds ratio of only 1.1:1." In contrast, the odds ratio for death with congestive heart failure was 11.7:1 and for persistent ST-T wave changes, 16.6:1. Despite the low odds ratio for arrhythmias, the authors strongly suggest that atrial flutter/fibrillation may be an important predictor of myocardial dysfunction and they advise vigorous management of children with cardio-myopathy who have these arrhythmias. Although these recommendations are not based on definitive information,
few would argue their merits. Undoubtedly, most physicians would also suppress ventricular tachycardia in children with dilated cardiomyopathy. The meaning of frequent single ventricular premature beats remains unclear.

Does treatment of arrhythmias improve prognosis? Documentation of whether "control" of arrhythmias improves prognosis is extremely difficult. In patients who are retrospectively analyzed with use of an occasional ECG and little or no long-term monitoring, this simply cannot be determined. In the present study (6) only 22 of 63 patients are reported to have had a Holter monitor study; 20 had two such studies. Judgments regarding the efficacy of antiarrhythmic treatment would require much more extensive monitoring and evaluation. The absence of a previously noted rhythm abnormality on a random ECG cannot be said to indicate that a rhythm disorder is controlled. It is possible that 1 or 2 h later, the ECG could once again show the arrhythmia to be present, and no different from or perhaps even more severe than the earlier arrhythmia. Even with extensive Holter monitoring, the findings may vary markedly in untreated patients. This study does not include information as to how often the patients were seen at follow-up study, how patients with arrhythmias were treated, whether patients were symptomatic because of rhythm disorders or if the symptoms disappeared with treatment. The efficacy of pharmacologic antiarrhythmic therapy in children with dilated cardiomyopathy remains unknown. There have been no published accounts of pediatric experience with an implanted defibrillator, but this approach may be useful in selected cases.

Management decisions. The question of whether cardiac arrhythmias are an independent predictor of death in children with dilated cardiomyopathy in the absence of confounding variables (such as congestive heart failure, ST-T wave changes and historical data relating to familial disease) remains unanswered. Sudden death implies the presence of a terminal arrhythmia, but the presence or absence of previous rhythm abnormalities may not be predictive. The effect of antiarrhythmic treatment is not known. Data have not been provided from the published retrospective analyses to guide the physician who must consider the timing for cardiac transplantation in a child with cardiomyopathy who appears to be in relatively stable condition. At present it is prudent to consider any child with persistent dilated cardiomyopathy to be a candidate for heart transplantation. Key factors that may affect judgment regarding urgency include progressive myocardial dysfunction, severe ventricular or atrial arrhythmias; syncope; elevated right atrial pressure; marked ECG changes (such as abnormal ST segments and left ventricular conduction delay); familial disease and systemic or pulmonary embolism, or both. Specific information as to which combination of these and other variables might provide reliable criteria for management decisions could emerge from experience with dilated cardiomyopathy in children; conclusions would not be contaminated by the presence of classic ischemic heart disease. However, the small number of pediatric cases available for study does not bode well for a clear-cut answer from a single institution. The argument for a prospective multicenter cooperative study is a strong one.

References