Editorial Comment

Evolving Concepts in the Management of Congenital Junctional Ectopic Tachycardia

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Junctional automatic ectopic tachycardias remain one of the most challenging dysrhythmias facing physicians who treat children. In this issue of Circulation, Villain et al report that despite advances in pharmacologic and device therapy, 35% of patients died of congenital junctional ectopic tachycardia. These data were collected over a prolonged period of time, and the prognosis today is probably somewhat better. The other form of junctional automatic ectopic tachycardia common to pediatric patients, postsurgical junctional ectopic tachycardia, has been equally devastating. These two entities have similarities and differences. They have been documented to have the same clinical electrophysiological mechanism, that is, enhanced automaticity in the conduction system. Enhanced automaticity is a difficult and uncommon mechanism in any clinical setting and in each part of the heart.

Automatic tachycardias are much more resistant to medical treatment than are reentrant tachycardias. They are virtually always chronic (except for surgical junctional ectopic tachycardia) and are resistant to overdrive pacing and cardioversion as well as to drugs. They often lead to a chronic cardiomyopathy and congestive heart failure (except for chronic automatic ventricular tachycardia, which has a rate only slightly greater than the sinus rate).

Recently, some patients with atrial automatic tachycardia have been found to respond to type IC antiarrhythmic drugs such as flecainide and encainide. Postsurgical junctional ectopic tachycardia has been shown to sometimes respond to propafenone, another IC agent. On the other hand, flecainide and encainide have been shown to have life-threatening proarhythmic effects in patients with damaged ventricles. Type IC drugs were not frequently used in the study of Villain et al. Thus, although further testing of these drugs is probably warranted in congenital junctional ectopic tachycardia, great caution must be used, including in-hospital monitoring and electrophysiological testing.

The Villain et al study points up another fact of modern-day cardiology. European cardiologists virtually always have new drugs before their counterparts in the United States. European patients with life-threatening dysrhythmias benefit from these drugs well before Americans. The Food and Drug Administration in the United States has taken a less strict policy on controlled testing of anti-AIDS drugs than many other drugs. Is AIDS any more life threatening than congenital junctional ectopic tachycardia? Shouldn’t a few U.S. cardiology centers be allowed to test potentially life-saving drugs earlier in their development? The problems found in the Cardiac Arrhythmia Suppression Trial (CAST) were not in critically ill patients tested in major cardiology centers but in asymptomatic patients.

An equally alarming problem is that few or no pediatric patients are tested in clinical trials of new antiarrhythmic drugs despite the fact that pediatric patients continue to die of dysrhythmias. These drugs are then available without formal testing for use by any physician on children. The pacemaker industry is to be commended for virtually always including a pediatric center in each clinical trial. Unfortunately, after a drug or pacemaker is released, there is little incentive for a company to do proper testing in children.

I believe that each drug and device should be tested in children very early in its development if there is any likelihood that it will be used in children. This should be required by the Food and Drug Administration. In addition, the Food and Drug Administration should help and encourage testing of each potential antiarrhythmic agent in the United States concomitant with testing in Europe so that critically ill patients who might otherwise die may receive treatment. Possible negative publicity from a child dying during testing of a new drug should not negate our concern for the children who will die without treatment or who will suffer when given inadequately tested drugs.

Another important point brought out by Villain et al is the need for increased information exchange between Europe and the United States. It seems

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possible that the “French” dose of amiodarone is more effective than the “American” dose, at least in the treatment of this dysrhythmia. In addition, a regimen of dose per square meter of body surface in infants yields a higher dose than does a per-kilogram regimen.

This disease entity calls into question our methods of treating patients with rare diseases. Must we have a double-blind multicenter trial before we can determine how to treat a patient for whom there is no standard therapy? No! It is the physician’s duty to treat a sick patient to the best of his or her ability. He or she must use experience with other similar disease processes. The physician should blend the principle of doing no harm with that of protecting the patient from harm. In the treatment of this disease, it is clear that catheter and surgical ablation should be reserved for the very ill patient for whom medical treatment has failed or who is not likely to survive long enough to determine whether medical treatment will work. Ablation treatment of this dysrhythmia is unproven and carries considerable short- and long-term risk, although DC or radiofrequency catheter ablation may prove life-saving in refractory patients. On the other hand, the implantation of a permanent pacemaker carries little risk and should prove effective in some patients based on both pathologic studies and clinical studies. Thus, a combination of initial permanent pacemaker implantation with the French dose of amiodarone is probably the conservative treatment of this dysrhythmia at the present time.

A final caveat is that patients with rare, life-threatening dysrhythmias are probably best treated in large quaternary referral centers. These centers should be encouraged to treat such patients under prearranged multicenter protocols. Because there is little financial incentive for industry to fund studies of rare diseases, government should do so either directly through the National Institutes of Health or indirectly through social programs. A rare cardiac dysrhythmia that requires expensive medical and drug treatment and familial travel is truly a disabling condition!

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(Circulation 1990;81:1713-1714)
Evolving concepts in the management of congenital junctional ectopic tachycardia.

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_Circulation_. 1990;81:1713-1714
doi: 10.1161/01.CIR.81.5.1713

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/81/5/1713.citation

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