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The Year in Congenital Heart Disease

The review by Graham (1) is an excellent and welcome summary of surgical, interventional, and medical progress in congenital heart disease. There are sufficient data to allow the reader to form an opinion, backed up by specific references, as well as useful critical remarks of consensus.

I was particularly interested in his comments about postoperative pulmonic insufficiency in patients with tetralogy who are now being recognized as a significant problem in a growing population of postoperative adults. Initially, we all had hoped that pulmonic insufficiency would not be a problem, and surgeons were urged to abolish completely any gradient across the right ventricular (RV) outflow tract because of concerns that if the RV pressure was still elevated it would create a risk for arrhythmias, and even sudden death. As these youngsters lived with their insufficiency, they developed large right ventricles, but were generally asymptomatic. This tolerance of volume overload is also characteristic of the left ventricle, but in their teens and twenties those young patients are beginning to need valve replacements. Unfortunately, we do not have the same reliable guidelines (ejection fraction) for the right ventricle as we have for the left.

Although we will have to face difficult decisions for our current generation of postoperative tetralogy patients with severe pulmonic insufficiency—considering the limited half-life for biological valve replacements, as Graham (1) mentioned—shouldn’t we revisit the degree of insufficiency being created in today’s infants and children? Many years ago one of the pioneers of surgical correction of tetralogies anticipated the problem: Frank Gerbode (personal communication, June 1965) warned that it would be better physiologically to leave a moderate degree of pulmonic stenosis rather than create severe pulmonic insufficiency. He reasoned that the less compliant right ventricle associated with moderate stenosis would resist the insufficiency, and that moderate stenosis would be better tolerated.

As a minimum, we should review the evidence as to whether we have unnecessarily accepted all the problems of severe pulmonic replacement absent a test of the alternate approach of moderation in disabling the valve at surgery.

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REPLY

It is a pleasure to respond to Dr. Guntheroth’s comments regarding my paper (1) and the current state of affairs for patients with tetralogy of Fallot and significant postoperative pulmonary insufficiency. As he clearly states, many of these adult patients have an enlarged right ventricle that can show a progressive decrease in ventricular function, which at some point becomes partially or completely irreversible. In addition, patients with a marked degree of right ventricular (RV) enlargement and decreased function appear to be potentially more susceptible to significant arrhythmias, which, on rare occasions, can be life-threatening. The optimal treatment for these patients is theoretically available with pulmonary valve replacement, which can be performed with low mortality and morbidity. When this procedure is done early enough in the course of inexorable decrease of RV function, there is a decrease in RV size (usually modest) and improvement in RV function and exercise ability. Unfortunately, valves that have been used in the pulmonary position have, under most circumstances, shown a half-life of only 10 to 12 years. The newer, larger bio-prosthetic pericardial valves have shown some promise for holding on longer, but the data are not complete on these valves to make that a clear-cut alternative with promise for a longer interval between reoperations.

The search for an optimal bio-prosthetic valve (one that can be grown from one’s blood cells outside the body and then reimplanted and possibly delivered with a catheter-interventional technique) is the hope of the future.

In the interim, I agree with Dr. Guntheroth that we should continue to revisit the issue of leaving mild pulmonary stenosis rather than create severe pulmonary insufficiency. This can be a difficult management decision in the operating room as the more severe tetralogy patients with severe outflow obstruction and small main pulmonary arteries are not amenable to successful repair without leaving them with significant pulmonary insufficiency. Moreover, when one has to do a significant ventriculotomy and leave moderate stenosis, right ventricle dysfunction can occur early in the postoperative period and be even more severe than that associated with moderate/severe pulmonary insufficiency.

During follow-up visits, postoperative tetralogy patients, most of whom have had an excellent overall outcome long term, continue to need attention to their RV size and function and to the potential for rhythm disorders; this should be done by physicians with expertise in both adult cardiology and congenital heart disease. Magnetic resonance imaging of the right ventricle with quantification of RV volumes and ejection fraction as well as exercise testing with maximum VO₂ measurement can be quite useful in terms of serial assessment and consideration for intervention. There is a growing need for training the next generation of physicians who can provide optimal care for this burgeoning group of adults with congenital heart disease.

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I read with interest the reports on defibrillation testing (DFT) during implantable cardioverter-defibrillator (ICD) implantation published recently in JACC (1,2). There is a significant, albeit low, risk to serious complications including death during DFT testing. If the implantation data as measured through the device are satisfactory, then should we induce ventricular fibrillation (VF) in a patient who has poor cardiac function? Should we crash a brand new car during its "test-drive" to see whether the airbags will deploy? The following cases, which are mere examples, demonstrate the professional and moral dilemma of DFT testing.

Two patients with severe cardiomyopathy underwent ICD implantation with excellent parameters. In the first case, during DFT testing, first shock to provide a 10-J safety margin and a subsequent maximum output shock failed to defibrillate. External defibrillation restored sinus rhythm but with severe electromechanical dysfunction requiring emergent placement of a ventricular assist device and subsequently a heart transplantation. Was cardiomyopathy in itself responsible for the ICD failure? Did the shocks cause electromechanical dysfunction? Could the patient have survived an out-of-hospital VF episode? Did DFT testing identify deficiencies at implant? In the second case, DFT testing was not performed because of the presence of atrial fibrillation, suboptimal anticoagulation profile, and evidence of sludge in the left atrial appendage by a transesophageal echocardiogram. Effective cardioversion and DFT testing after six weeks of anticoagulation was planned. Unfortunately, in the interim period, the patient met with an unnatural mode of death. Subsequently, the patient’s wife reported that “it may not mean much . . . but the defibrillator did go off . . . many times . . . it did work . . . when my husband died. . . .”

During automobile accidents the airbags drastically reduce morbidity and mortality, but there is also a spectrum of injuries associated with them (3,4). Taking the analogy of the airbags and the ICDs, both of which reduce fatalities, perhaps in the case of the first patient, the “airbag” in itself was not adequate to prevent the fatality and, if anything, perhaps caused “airbag”-associated injuries. In the case of the second patient, the “airbag” deployed appropriately but could not prevent a non-road-traffic-crash–related fatality.

Needless to say, until improvements in science and technology provide conclusive evidence that the ICDs effectively and predictably provide life-saving therapy without actually testing them at the time of implantation, the dilemma of DFT testing will remain unsolved.

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REPLY
I thank Dr. Kantharia for his interest in our recent paper (1). Identifying the optimal patient-specific implantable cardioverter-defibrillator (ICD) system and its programming, without the need for either ventricular fibrillation (VF) or shocks, is a major research goal. The patients reported by Dr. Kantharia both emphasize the importance of this goal and illustrate poignantly that it remains beyond our grasp.

The first case highlights the unmet need of developing effective treatment for life-threatening, post-VF electromechanical dysfunction (EMD) (2,3). Paradoxically, defibrillation testing may have saved this patient’s life: If VF had occurred as an outpatient, either defibrillation would have failed or the postshock rhythm would have been lethal EMD. To the best of my knowledge, fatal postshock EMD has not been reported after an inappropriate shock. Thus, postshock EMD probably is caused by a combination of VF and shocks, often prolonged VF and multiple shocks.

The second patient died from failed defibrillation with an untested ICD system. This case illuminates the need for a shockless method of assessing ventricular defibrillation efficacy, or at least a method that minimizes the risk of thromboembolism from atrial cardioversion. One consideration is continuous rapid atrial stimulation during ventricular defibrillation or vulnerability testing.

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