Editorial Comment

How Can Something So Small Cause So Much Grief? Some Thoughts About the Underdeveloped Right Ventricle In Pulmonary Atresia and Intact Ventricular Septum*

ROBERT FREEDOM, MD, FRCPC, FACC
Toronto, Ontario, Canada

Over the past 2 decades several investigators (1-4) have attempted to demonstrate with a variety of methodologies that the morphologically right ventricle in patients with either pulmonary atresia or critical pulmonary stenosis can grow after surgical intervention. That growth should occur in a morphologically right ventricle whose dimensions are nearly normal or in an occasional patient with a very small right ventricle is not surprising, provided that there is adequate relief of the severe obstruction to the right ventricular outlet tract. Indeed, perhaps in some patients with pulmonary atresia and intact ventricular septum, growth may seemingly result from resolution of massive ventricular hypertension and secondary attenuation of the functional cavity. (Most of my comments in this editorial will focus on patients with pulmonary atresia and intact ventricular septum because data from the Congenital Heart Surgeons Study [Kirklin JW. Unpublished observations] suggest that for many patients with pulmonary atresia and intact ventricular septum, the growth of the ventricle in these patients is not in any way normal. Most investigators would agree that volume-derived methodology based on right ventricular angiography in the patient with a small, hypertensive chamber has the potential for many intrinsic errors; although a decade ago we used similar methodology, today we urge caution in this approach (2). Nonetheless, we do not disagree with the observations of Schmidt and colleagues (10) or of others (1-4) that the right ventricle of some patients with pulmonary atresia and intact ventricular septum does in fact grow, occasionally remarkably (1-4), after surgical treatment of the right ventricular outflow tract. But the reality of right ventricular growth is not invariably predictive of a satisfactory long-term outcome. A profusely obstructive inlet may prevent a complete biventricular repair, with growth reflecting little more than pulmonary regurgitation into a poorly compliant chamber. Indeed, the initial battle (right ventricular growth) may be won, but the war (a successful biventricular repair, or even outcome) may be lost.

Surgical implications. The surgical success for patients with pulmonary atresia and intact ventricular septum does not in any way match the contemporary success of most institutions with the arterial switch operation for complete transposition of the great arteries (11-13). In large part this lack of surgical success with this lesion reflects the profound morphologic heterogeneity of hearts exhibiting pulmonary atresia and intact ventricular septum (14). In almost no other congenital heart malformation does the understanding of the unusual coronary artery circulation have the potential for define the limit of the endocardium in those patients with widespread ventriculocoronary connections and an extensive myocardial sinusoidal network (2). Others (3,5-7)eschewing any form of volume-derived methodology because of these difficulties, have utilized a variety of right ventricular inlet and outlet indexes with various degrees of success. Not only has determination of right ventricular size been the arbiter of surgical decision making, but also surgical algorithms based on the components of the right ventricle (inlet, outlet, and conduction zone) and how well endowed these elements are have been advocated, with various degrees of success (4,7-9).

The present study. Schmidt and colleagues (10) demonstrate in this issue of the Journal that echocardiographically derived volume information can be used to demonstrate right ventricular growth and functional alterations after surgery for pulmonary atresia or critical stenosis and intact ventricular septum. Furthermore, they provide correlations for the same patient between end-diastolic volume and stroke volume derived from their echocardiographic methodology and similar data for many of the same patients obtained from right ventricular angiography. This group validated the two-dimensional echocardiographically derived volume information in the fetal lamb. Yet the fetal lamb heart is not the heart of the newborn with pulmonary atresia and intact ventricular septum. The morphologically right ventricle of the patient with pulmonary atresia and intact ventricular septum is not in any way small. It seems likely that much of the early surgical experience with patients with pulmonary atresia and intact ventricular septum reflects the methodology of Schmidt and colleagues (10). This group has demonstrated that right ventricular angiography is less reliable than echocardiography for determination of right ventricular volume. Furthermore, this group has recently demonstrated that the right ventricle in patients with pulmonary atresia and intact ventricular septum does in fact grow, occasionally remarkably (1-4), after surgical treatment of the right ventricular outflow tract. But the reality of right ventricular growth is not invariably predictive of a satisfactory long-term outcome. A profusely obstructive inlet may prevent a complete biventricular repair, with growth reflecting little more than pulmonary regurgitation into a poorly compliant chamber. Indeed, the initial battle (right ventricular growth) may be won, but the war (a successful biventricular repair, or even outcome) may be lost.

Surgical implications. The surgical success for patients with pulmonary atresia and intact ventricular septum does not in any way match the contemporary success of most institutions with the arterial switch operation for complete transposition of the great arteries (11-13). In large part this lack of surgical success with this lesion reflects the profound morphologic heterogeneity of hearts exhibiting pulmonary atresia and intact ventricular septum (14). In almost no other congenital heart malformation does the understanding of the unusual coronary artery circulation have the potential for

*Editorial published in Journal of the American College of Cardiology reflect the views of the authors and do not necessarily represent the views of the American College of Cardiology.

From the University of Toronto Faculty of Medicine and the Hospital for Sick Children, Toronto, Ontario, Canada.

Address for reprints: Robert M. Freedman, MD, Division of Cardiology, The Hospital for Sick Children, 555 University Avenue, No. 182, Toronto, Ontario, ON MS5 1X8 Canada.

©1992 by the American College of Cardiology 0022-2822/92/1038-1015$3.00/0
altering and influencing both the initial and the subsequent surgical algorithms. In 60 years right ventricular-coronary artery communications have made the scientific pilgrimage from curiosity to incremental risk factor in this disorder (15–17). Today before one employs any of a wide range of surgical maneuvers that have the potential to promote right ventricular growth, one must define the presence or absence of ventriculo-coronary connections; and, if present, it is essential to determine if these connections promote a coronary artery circulation that is wholly or in large part dependant on the right ventricle (18–28). For those patients in whom most of the coronary circulation is so dependant, the surgical algorithm does not justify those procedures promoting either right ventricular growth or exclusion (29–35). Thus the construction of therapeutic algorithms for these patients requires a stepwise progression both asking and answering the following questions:

1. Does the patient have ventriculo-coronary connections? If so, how much of the coronary circulation is right ventricular-dependent, and can the right ventricle be safely decompressed? And, yes, cardiac catheterization with right ventricular-dependent, and can the right ventricle be safely

2. Is the patient a candidate for a biventricular or a univentricular repair? What morphologic variables are important in this determination:

3. Which factors best predict long-term outcome? What are the most appropriate initial surgical maneuvers? And subsequent surgical events? And the list goes on.

The reality of any single therapeutic maneuver providing therapeutic salvation is akin to a long-term cease-fire in those areas of the world devoured by genetic hatred: the cease fire lasts only slightly longer (perhaps shorter) than the headline. For pulmonary atresia and intact ventricular septum, there have been many headlines. But the promise of the morphologically right ventricle in pulmonary atresia and intact ventricular septum, we have all too frequently come to appreciate the question: How come something so small usually cause so much grief?

References

29. Billingham AM, Laos H, Boyce SW, George B, Sammill T, Williams RG.