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Editorial Comment

The Dinosaur and Banding of the Main Pulmonary Trunk in the Heart With Functionally One Ventricle and Transposition of the Great Arteries: A Saga of Evolution and Caution*

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The application of Fontan's ingenious approach of atrial partitioning with an atriopulmonary connection has been widely extended since its initial application to patients with the classic type of tricuspid atresia (1,2). Indeed, Fontan's operation is considered by many the operation of choice to afford long-term palliation to patients with most forms of single ventricle (including patients with right or left atrioventricular [AV] valve atresia and double or common inlet univentricular AV connection). The hemodynamic and angiocardiographic criteria for operability have been extensively evaluated and modified subsequent to the initial observations of Fontan and Choussat and their coworkers (3). This is not the appropriate forum to review these criteria. But there is a growing consensus that in assessing the suitability of any patient for this operation, there must be some determination of diastolic ventricular compliance. The methodology to define diastolic ventricular compliance is complex, and although such data have been derived from pressure-volume loops for a number of acquired disorders affecting the biventricular heart, there are considerably fewer data obtained from patients with a univentricular heart (4–8). Of the many variables that may directly or indirectly influence ventricular compliance, abnormal wall mass or disproportionate ventricular hypertrophy must certainly be considered.

Role of abnormal ventricular hypertrophy. There is increasing clinical evidence that "abnormal" ventricular hypertrophy is a risk factor for patients undergoing the Fontan operation (9–12). Such ventricular hypertrophy implies a stiffer, noncompliant chamber with impaired diastolic function. Obviously this will have an impact on patients who have undergone an atriopulmonary correction and may alter the mechanics and dynamics by which blood is "sucked" through the lungs (13).

Which factors seemingly contribute to abnormal ventricular hypertrophy in patients with a univentricular heart? Kirklin et al. (12) suggested that older age may be accompanied by ventricular hypertrophy and therefore proposed that the Fontan operation be performed at a young age (2 to 4 years). Furthermore, there is considerable evidence (9–12,14) that patients with a single ventricle and subaortic stenosis often do not fare well with Fontan's operation (or ventricular septation, for that matter). Clearly, subaortic stenosis, which usually results from a restrictive interventricular communication, will promote ventricular hypertrophy of a magnitude that is considered abnormal even for individuals with a univentricular heart.

Factors contributing to subaortic stenosis. Under which circumstances does subaortic stenosis occur in patients with a univentricular heart of left ventricular type, a rudimentary right ventricle and transposition of the great arteries? First, the interventricular communication may be restrictive at birth (or indeed completely sealed) (15). Second, a restrictive ventricular septal defect in this setting, and the resulting subaortic stenosis, occurs only uncommonly when there is significant naturally occurring pulmonary outflow tract obstruction (11). This is not terribly surprising considering the reciprocal relation between pulmonary and systemic blood flow in a one ventricle heart with obstruction to one arterial outlet (13,14). Finally, subaortic stenosis may develop insidiously, and its development may be determined in part by the natural history of restrictive *muscular* ventricular septal defect, namely, the tendency to spontaneous diminution in size. We have suggested elsewhere (9,11,15,16) that banding of the pulmonary trunk in this group of patients (those with a potentially or truly restrictive ventricular septal defect) could accelerate, by promoting myocardial hypertrophy, this natural predilection to spontaneous diminution in size or even closure. The presence of a band on the pulmonary trunk and a restrictive ventricular septal defect promote severe myocardial hypertrophy, and often rapidly. Thus, subaortic stenosis and its sequelae of myocardial hypertrophy and altered ventricular compliance should be anticipated in 1) patients whose systemic circulation is partly or wholly ductus dependent (implying inadequate forward aortic blood flow and by inference a restrictive ventricular septal defect); 2) patients whose initial two-dimensional echocardiographic and angiographic evaluations suggest a potentially or actually restrictive ventricular septal defect; or 3) patients subjected to banding of the pulmonary trunk.

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Early identification of patients at risk of developing subaortic stenosis. If an abnormally stiff and noncompliant ventricle bodes a poor outcome in patients undergoing Fontan's operation, then it is the mandate of the clinician to identify such patients at risk of developing subaortic stenosis early in their life and to provide surgical alternatives to procedures that are known to produce ventricular hypertrophy. The imaging of the ventricular septal defect by selective angiocardiography and two-dimensional echocardiographic modalities (with Doppler interrogation) is mandatory to this determination. The definition by Rothman et al. (17) in this issue of the Journal that subaortic obstruction is present where the defect is "small" angiographically (less than one-half the aortic root diameter) in the neonatal period probably excludes a substantial number of patients who will develop subaortic stenosis. Although this guideline provides a useful rule of thumb, the ventricular septal defect in these hearts is often elliptical, much like a "buttonhole," and, thus, some angiographically large defects may in fact be restrictive. Furthermore, I would argue that by the time a 30 mm Hg gradient is recorded across the ventricular septal defect (especially in patients who have had previous pulmonary artery banding) that subaortic obstruction is moderately advanced. This concern is bolstered by our observations and those of others (9,11,18) that, among some patients who have had banding and have the appropriate ventricular anatomy and a discordant ventriculoarterial connection, unequivocal anatomic evidence of subaortic stenoses may be present despite the absence of a pressure gradient at rest. The judicious use of parenteral isoproterenol in the catheterization or echocardiographic laboratory to provoke a pressure gradient and thus to unmask subaortic stenosis is strongly urged (9,11,19).

Surgical management. Rothman and colleagues (17) in the preceding paper describe the surgical management of subaortic stenosis in 24 patients with some form of single ventricle. Fourteen (58%) of their patients died and the mortality rate for those with a previously banded pulmonary trunk was slightly higher (63%; 10 of 16). No patient in their series has as yet undergone a Fontan procedure when subaortic stenosis was recognized preoperatively. The Boston Children's group, having used a variety of surgical approaches to palliate subaortic stenosis, currently advocate abandoning pulmonary artery banding as a form of palliation when subaortic stenosis is present or anticipated (17). Instead they suggest the construction of a proximal main pulmonary trunk to aortic anastomosis and in the neonate and young infant the performance of a controlled systemic to distal pulmonary artery shunt, an approach used to palliate babies with the hypoplastic left heart syndrome, with a Fontan procedure in early childhood (20). The experience from Toronto is similar to that recorded by Rothman and his colleagues. We, too, have applied all the procedures recited in this large clinical series, and with basically similar results (9,11,17). We, like others, have evolved a Norwood-type approach to palliate the neonate and young infant with plans for a later Fontan procedure (21,22).

Thus, one might ask: Is there still a role for banding of the pulmonary trunk? The answer is uncertain. Indeed, a rare patient with a very large ventricular septal defect (assuming the same basic anatomy) may clinically benefit from a banding procedure (without developing real or occult subaortic stenosis). But will this procedure have an impact on increased ventricular mass and stiffness? Thus, like the dinosaur, banding of the pulmonary trunk once had its place in the evolution of surgical procedures applied to this specific group of patients. But, unlike the dinosaur, pulmonary artery banding is not yet extinct, though its cardiac niche is becoming ever smaller.

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