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Editorial Comment

Dopper Color Flow Mapping and Prediction of Ventricular Septal Defect Outcome*

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Predicting the Outcome of Ventricular Septal Defects

Clinicians have placed great importance on predicting the outcome of ventricular septal defect. This allows the development of an efficient and effective management plan and aids in appropriate family education. For almost 3 decades clinical scientists have sought to understand the factors governing the probability of ventricular septal defect closure (1–4). Early clinical studies employed serial cardiac catheterization; some were models of study design and others utilized a mixed study group. All were hampered by a limited ability to localize the ventricular septal defect borders on anteroposterior and lateral cineangiographic views; also, hemodynamically insignificant defects were underrepresented by these studies. The highly refined techniques of Doppler echocardiography now make it possible to determine all borders of a ventricular septal defect, its size, the presence of associated lesions and the pressure decrease (across a restrictive defect) as well as the dynamics of flow and degree of shunting across the defect. It is now practical to document the very small ventricular septal defect and to follow its fate.

Present study. The study by Hornberger et al. (5) in this issue of the Journal demonstrates the ability of Doppler color flow mapping to accurately determine ventricular septal defect size. The study tracks the course of defect size in patient groups with and without congestive heart failure. In essence, the study demonstrates the ability of Doppler echocardiography to accurately reflect the status of a ventricular septal defect. By following serial measurements of size, ultimate course may be predicted. In this study design, the chief variable examined as a predictor of outcome was the presence or absence of congestive heart failure. In this regard, the study is similar to that of Hoffman and Rudolph (1) showing a more favorable outcome in patients without congestive heart failure. It is useful to look at this study in the context of all factors that may influence the course of a ventricular septal defect.

Variables Affecting Outcome of Ventricular Septal Defect

Defect size. The presence of overlying atrioventricular valve (AV) tissue impedes the quantification of the effective orifice size of a perimembranous ventricular septal defect. Doppler color mapping clearly predicted ventricular septal defect size when compared with angiographic and surgical measurements. In general, initial size correlated weakly with outcome, but all defects ≤4 mm closed over a 40 month observation period. Also, patients with an initial defect diameter ≥8.2 (±1.9) mm or defect/aortic root diameter ratio ≥0.67 (±0.18) were more likely to develop congestive heart failure (5).

Recent abstract presentations (6–8) have noted that technical features such as gain, transmission power, carrier frequency, pulse repetition frequency, wall filter display mode and packet size have an important effect on the apparent size of jets assessed by Doppler color flow mapping. Great care must be taken to standardize these variables when comparing serial ventricular septal defect sizes.

Ventricular septal defect position. In the study of Hornberger et al. (5), 75% of mid and posterior muscular defects underwent reduction in size or closure. This group of defects was far more heavily represented in the patients without congestive heart failure; hence, it is difficult to determine whether defect size or position was the main factor. It is generally believed that defects extending to the cardiac crux (known as AV canal, AV septal or inflow defects) do not undergo spontaneous closure. Because of wide variability in nomenclature, these defects may occasionally be grouped with others under the term "perimembranous" (9). Because no separate classification of this group of defects is found in the study by Hornberger et al. (5), it is possible that a substantial number of these are present in the patients with a large perimembranous defect and congestive heart failure, a circumstance that would account for the relatively poor outcome in these patients. The same could be said for perimembranous defects extending into the outflow septum, the so-called doubly committed subarterial defect. The true perimembranous septum is small, so by definition, a large perimembranous ventricular septal defect must extend into the midmuscular, AV septal or outflow (infundibular) septum. Because defects in the latter two locations are unlikely

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to close, only those with midmuscular extension potentially have a favorable outcome (10). A more exact definition of all borders of the large ventricular septal defect should improve the prediction of clinical course.

Presence of aneurysm of the membranous septum. Ramaciotti et al. (11) showed a high degree of association between formation of aneurysm of the membranous septum and defect closure or decrease in size. In their study of 247 patients, 77% of patients with an isolated perimembranous ventricular septal defect had associated overlying AV valve tissue. The incidence of the latter was greater with small defects (hence its role apart from defect size may be obscured). Over a mean observation period of 27 months, clinical improvement occurred in 44% of patients with overlying AV valve tissue and 18% of patients without it. Only 28% of patients with overlying AV valve tissue had surgical closure whereas 84% of patients without it required surgical intervention. Similar data were obtained in the study by Hornberger et al. (5). None of the patients without overlying AV valve tissue had spontaneous diminution of ventricular septal defect size.

From these data two points seem clear: 1) the absence of overlying AV valve tissue predicts that ventricular septal defect size will not significantly decrease, and 2) fewer than ½ of defects of all sizes with overlying AV valve tissue will undergo spontaneous improvement. Interestingly, the Hornberger study (5) demonstrates progressive (but probable hemodynamically insignificant) tricuspid insufficiency or diversion of a ventricular septal defect jet into the right atrium in patients who develop overlying AV valve tissue.

Associated lesions. Ventricular septal defects are very unlikely to close in the presence of straddling AV valves or malalignment between the muscular and outflow (infundibular) sections of the septum, but such variations are excluded by the term simple or isolated ventricular septal defect.

Presence of Down's syndrome. In Ramaciotti's series (11) only 1 of 15 patients with Down's syndrome and isolated perimembranous ventricular septal defect had spontaneous improvement. Thus, associated chromosomal abnormalities may also be an important factor in the natural history of these defects.

Length of observation. Patients without congestive heart failure are more likely to undergo a longer period of observation without intervention and thus have the opportunity to express the natural history of the defect. In both recent and older series, the patients with a large ventricular septal defect and congestive heart failure were more likely than others to undergo surgical intervention or death. In former years only patients who underwent pulmonary artery banding were able to demonstrate closure of a large ventricular septal defect. No studies have included this group, but the occasional patient who has spontaneous ventricular septal defect closure and requires only pulmonary artery band removal is well known. Although a very large ventricular septal defect can undergo spontaneous closure if given sufficient time, this point is moot in the modern day when either prolonged congestive heart failure or pulmonary artery banding is an unacceptable alternative in the face of early and effective surgical closure.

Other Methodologic Problems

The presence of congestive heart failure is an expression of the natural history of ventricular septal defect and therefore is not properly used as a predictor of natural history. Many studies suffer from this problem. Nevertheless, they demonstrate that one of the important determinants of the presence of congestive heart failure, defect size, is related to outcome.

There will always be a number of patients with a very small ventricular septal defect who are not referred to cardiologists because their murmur is classified as functional or unimportant by the primary care giver. Thus, we will not know the true "denominator" of patients with ventricular septal defect and most clinical studies will be biased toward the more significant lesion unless an unselected group of neonates is studied prospectively.

Finally, as a sign of age, I am compelled to point out that Doppler echocardiography has never been shown to be more sensitive than auscultation in determining closure of a small ventricular septal defect. Nevertheless, Hornberger et al. (5) have clearly demonstrated that Doppler color flow mapping is complementary to auscultation, the traditional tools of electrocardiography and chest X-ray and echocardiographic imaging in delineating the status and natural history of the isolated ventricular septal defect.

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