procedures used in our patients served to protect their hearts to a greater degree than was possible ten years ago, and that this factor contributed to the better functional result.

Another factor that should be considered is the relative lack of hypertrophy as compared to dilatation demonstrated by the five patients for whom we have preoperative mass data. These patients showed relatively little hypertrophy for the degree of volume overload to which their left ventricles were subjected. The LVEDV/LV mass ratio of 0.81 in our patients compares to a LVEDV/LV mass of 0.69 (± 0.13) in 14 patients less than two years of age with VSDs not requiring early closure. The same ratio was 0.49 ± 0.09 in ten normal children of the same age. This relative lack of hypertrophy may, in fact, be the underlying reason for failure of medical management in these patients. On the other hand, the same lack of severe hypertrophy may have spared these patients the relative myocardial ischemia that may accompany the hypertrophic process, and thus have left them with better myocardial function once the volume overload was relieved.

Although the underlying mechanism is not clear, these data indicate that closure of VSDs in infancy can be expected to produce a good result in regard to left ventricular size and function, at least in the short term. Many factors must be weighed before early surgical closure of a VSD is undertaken. Our patients were operated upon because of refractory congestive heart failure and not for reasons of prophylaxis in preserving cardiac function. However, when early operative intervention is required, it is encouraging that good myocardial functional results can be expected.

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The Normal Spectrum of Mitral and Aortic Valve Discontinuity

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SUMMARY The relationship of the base of the left and noncoronary sinuses of the aortic valve and the adjacent aortic leaflet of the mitral valve was studied in 106 normal heart specimens and 184 specimens with isolated VSD. The results show a spectrum of persistence of the tissue along the inner curvature of the heart. This may help settle the recent controversy in the interpretation of echocardiograms of this area because the recorded mitral-aortic discontinuity may be due to this spectrum rather than to variations in technique.

that they are separated by only a thin band of fibrous tissue. A wider separation of these valves (discontinuity) has been of recent interest in the diagnosis of malposition of the great arteries1-3 including double outlet right ventricle4-5 because the mitral and adjacent semilunar valves are usually separated by a band of muscle, believed to be either persistence of the aortic conus2-6 or bulboventricular flange.

In this paper we describe a variable relationship of the base of the left and noncoronary sinuses of the aortic valve to the adjacent anterior leaflet of the mitral valve in normal hearts and in specimens with isolated VSD, and suggest that this variable relationship is probably pertinent to recent echocardiographic evaluations of this region.7-10

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Materials and Methods

One hundred and six normal hearts and 184 specimens with isolated ventricular septal defect from the pathological collections of the Johns Hopkins University and the Armed Forces Institute of Pathology were selected for study. All specimens had been fixed in formal and transferred to Kaisering’s solution.

Each specimen had been opened so that the aortic and mitral valve regions could be studied as a unit. Mitral-aortic valve separation (MAS) was defined as the shortest distance between the base of either the noncoronary or left coronary cusps and the basal attachment of the anterior leaflet of the mitral valve (fig. 1). Since this basal attachment defines the boundary of the free wall of the left atrium and the anterior leaflet of the mitral valve,9,11 MAS corresponds to the aorto-mitral continuity as defined by previous investigators studying normal and pathological material.4,6,12 In each specimen the aortic root circumference was measured along the ridge at the distal margins of the sinus of Valsalva (fig. 1). In our experience this measurement is a reliable way of estimating the size of the left ventricular outflow tract, particularly when the left ventricle has been sectioned horizontally or cut into longitudinal strips. Although VSDs were of various types,13 (i.e., supracristal, membranous, atrioventricular canal type, or muscular) and bicuspid aortic valve was present in 14 specimens, there were no differences in the spectrum of measurements of hearts associated with these variables. Therefore the results are not reported separately.

Since individual measurements noted independently by two observers did not vary by more than 1.0 mm for MAS or 2.0 mm for circumference of the aortic root, they were averaged.

Results

In the 106 normal specimens, increase in the aortic root circumference (as occurs with growth during childhood) was not accompanied by an increase in MAS, which varied from 0 to 7 mm (figs. 2A, 3). Since similar values for MAS were noted in infants and adults, this suggests that even though the heart enlarges during childhood, MAS remains constant.

In the 184 specimens with VSD, an increase in aortic root circumference was also not accompanied by an increase in MAS, which like the normal specimens varied from 0 to 7 mm (figs. 2B, 4). Since similar values for MAS were noted in infants and adults, this again suggests that even though the hearts with VSD enlarge during childhood, MAS remains constant.

Discussion

During the early stages of heart development the inner curvature has the same potential as the outer curvature and thus consists of cells that could contribute to all segments of the embryonic heart.14 After formation of the primary loop, the musculature along the inner curvature, being less vital to cardiac function, gradually becomes less significant in relation to the rest of the heart, due either to atrophy of disuse or programmed cell death.16 In time, the inner curvature contributes to the free wall of the left atrium adjacent to the atrioventricular valves (a thin but distinct entity) and the fibrous band between mitral and aortic valves.
In the present study of normal human specimens, the width of this fibrous band (MAS) was found to vary from 0 to 7 mm (fig. 2A, B), thus comprising a spectrum of discontinuity between mitral and aortic valves. We are unable to explain why this spectrum has been overlooked in previous descriptions of the fibrous skeleton of the heart.11, 16, 18

Why should the MAS vary within each group, yet not increase progressively with heart size? One possibility is that the musculature along the inner curvature regressed more completely in some embryos than in others. Thus in hearts where MAS = 0, there was complete absorption of the bulbouarricular19, 20 or conoventricular1 flange. In others, however, the disappearance of the flange may have been delayed, possibly due to malalignment of the inflow and outflow portions of the heart.

Gessner21 demonstrated that wedging a wire platform along the inner curvature of chick embryo hearts resulted in malalignment of the conotruncus in relation to the atrio-ventricular canal. In such embryos the musculature along the inner curvature persisted and double outlet right ventricle resulted. Similar findings have been produced in rat embryos with X-irradiation.22

Should human specimens with an increased MAS be considered formes frustes of double outlet right ventricle? The prevailing concept of double outlet is that the aorta rotates into the right ventricle through a VSD, and the MAS that results consists of muscle.9 The present study shows that although specimens with an isolated VSD do not have an increased MAS over normal, there is nevertheless a spectrum of discontinuity present, and double outlet may merely be further along the spectrum. Furthermore, a muscular MAS may not be an important diagnostic criterion for double outlet right ventricle, since the leaflets of the aortic and mitral valve will be the same distance apart regardless of their tissue composition. This point is relevant to recent discussions of echocardiographic observations of aortic-mitral displacement8-10 in which it was assumed that apposition of aortic and mitral valve leaflets is always present in normal hearts. In these discussions the discontinuity was attributed to transducer position or motion. While technique must always be considered in interpretation of variant echocardiograms, our data suggest that the basic tenet to this controversy (mitral-aortic continuity in the normal heart) may be in error. Our results also emphasize that interpretation of noninvasive cardiac data must be firmly based upon carefully collected anatomical information. This will be of particular importance in the study of specimens with double outlet right ventricle and tetralogy of Fallot, since we would predict a similar spectrum of MAS in these disorders.

It is possible that our findings may also aid in the interpretation of several other congenital conditions that affect the left ventricular outflow tract. Thus, the discrete membrane or diffuse fibrous thickening between aortic and mitral valve leaflets noted in some forms of subaortic stenosis23-26 may be associated with an increased MAS. Other specimens that should be studied carefully are those in which the placement of a pulmonary artery band was followed by development of muscular subaortic stenosis. Although clinical reports suggest that this stenosis results from hypertrophy of a displaced conal septum,29 it is conceivable that in a few cases, muscle fibers that may normally persist in the MAS37 had hypertrophied. Finally, discontinuity of the aortic and
mitral valves may also explain the increased vulnerability of some patients to the development of aneurysms in the space between aortic and mitral valves.27

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