Ventricular Volume Characteristics in Infants and Children with Endocardial Cushion Defects

JAY M. JARMAKANI, M.D., BARBARA GEORGE, M.D., AND JOHN WEHLLER, M.D.

SUMMARY Ventricular volume parameters were determined in 14 patients with isolated atrioventricular (AV) canal (Group I), five patients with AV canal and pulmonary stenosis (Group II) and 17 patients with ostium primum defect (partial AV canal) (Group III). Right ventricular (RV) and left ventricular (LV) volume parameters were determined from biplane cineangiocardiograms according to Simpson's rule method and the area length method. Right ventricular end-diastolic volume (RVEDV) and right ventricular systolic index (RVSI) were significantly greater than normal in Group I (RVEDV = 174 ± 15% of predicted normal, RVSI = 6.84 ± 0.47 l/min/m²) and Group III (RVEDV = 365 ± 37% of normal, RVSI = 13.54 ± 1.39 l/min/m²). Left ventricular end-diastolic volume (LVEDV) and left ventricular systolic index (LVSI) in Group I (LVEDV = 247 ± 20% of normal, LVSI = 10.04 ± 0.91 l/min/m²) and Group III (LVEDV = 169 ± 15% of normal, LVSI = 7.61 ± 0.69 l/min/m²) were both significantly greater than normal. The LV and RV ejection fractions in all groups were not significantly different from normal.

These data indicate that LV and RV volumes and outputs are higher than normal in patients with AV canal or ostium primum defects. The number of patients studied, however, is small and it is possible that some very young infants with AV canal could have small ventricular chambers. Thus, ventricular volume determination should be a part of the presurgical evaluation of these patients.

SEVERAL INVESTIGATORS have suggested that small ventricular size in patients with complete atrioventricular (AV) canal predisposes such patients to congestive heart failure and an increase in the morbidity and mortality after complete correction.1-8 Documentation of a small ventricle was observed by Goor and associates,1 Bharati and Lev,2 and Mair and McGoon.8 Goor described a patient with hypoplastic left ventricle where the AV valve was completely committed to the right ventricle. Bharati and Lev2 also suggested that the hemodynamic factors are responsible for the hypoplasia of the left or right ventricle.

In previous studies we have shown that ventricular septal defect was associated with an increase in LV volume and output, and atrial septal defect was associated with an increase in right ventricular (RV) volume.7 Furthermore, Graham and associates6 have shown that large ventricular septal defect was associated with increased RV volume in addition to an increase in LV volume. These observations suggest that AV canal should be associated with volume overload of both the right and left ventricle. Presently, however, there are no data on in vivo ventricular volumes in infants and children with complete AV canal or ostium primum defects (partial AV canal). Therefore, this investigation was designed to determine RV and LV volume parameters in infants and children with complete AV canal and ostium primum defects.

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Methods

This study included 36 patients who underwent routine diagnostic cardiac catheterization (table 1). The mean and SEM for the three groups are listed. The clinical diagnosis was made by cardiac catheterization and cineangiocardiography. The LV cineogram showed gooseneck deformity in all patients. The diagnosis of AV canal was based on the presence of bidirectional interventricular shunt on cineangiogram and equal RV and LV pressures. The diagnosis of ostium primum defect was made in the absence of ventricular shunt, and the RV pressure was less than LV pressure in all patients. The clinical diagnosis was confirmed in all but seven patients by surgery or autopsy.

Group I included 14 patients with complete AV canal and consisted of eight infants less than 1 year of age and six children more than 1 year of age. The RV pressure in these patients was equal to LV pressure and the pulmonary/systemic flow (Qp/Qs) ratio ranged from 1.2-4 (mean = 2.54 ± 0.27). There were no other associated intracardiac or extracardiac defects.

Group II consisted of five patients with complete AV canal and valvular pulmonary stenosis. The size of the pulmonic valve annulus was equal to or larger than normal, and no patient had subvalvular stenosis. The RV pressure in all patients was at systemic level, and four of the five patients had a dominant right-to-left shunt.

Group III consisted of 17 patients with ostium primum defect. The age range in this group was 1.8-18 years. The peak systolic RV pressure ranged from 22-70 mm Hg, and there were no significant gradients across the pulmonic valve, and no other intracardiac or extracardiac defects. The Qp/Qs ratio ranged from 1.9-5.0 (mean = 3.3 ± 0.3). Ventricular volumes were calculated from biplane cineangiocardiograms filmed after injecting 1.25-1.5 ml/kg...
body weight of contrast media into the right or left ventricle. Ventricular dimensions were corrected for linear magnification. The details of these methods were described previously.\(^5\),\(^6\) RV volumes were calculated by Simpson's rule method,\(^9\) and LV volumes were calculated by both the area length method\(^10\) and Simpson’s rule method. All calculated volumes were corrected by previously derived\(^5\),\(^6\) regression equations to determine the actual volumes. Right and left ventricular volumes were expressed as a percentage of predicted normal values, and all volume parameters were compared with the normal values.

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<th>Table 1. Hemodynamic and Volume Data</th>
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Results

The volume variables for each patient are listed in table 1 and figures 1-4. RV volume parameters were obtained on only 16 of the 36 patients. In the remaining 20 patients, RV cineangiograms were associated with premature ventricular contractions, and therefore, RV volumes were not determined. LV cineangiography in one patient (X10) with AV canal was also associated with premature ventricular contractions, and LV volume was not calculated.

RVEDV in Group I was significantly larger than normal and averaged 174 ± 15% of normal (fig. 1,
The only normal value, 119% in patient 14, was observed in a 3-day-old infant. This variable in Group II varied from 65%—149% of normal (table I). In patients with ostium primum defect, RVEDV was significantly greater than normal and averaged 265 ± 37% of normal (table I, fig. 1).

RV ejection fraction in all groups did not significantly differ from normal. Right ventricular systolic index (RVSI) (fig. 2) was significantly greater than normal in Group I (RVSI = 6.84 ± 0.47 l/min/m²) and Group III (mean = 13.54 ± 1.39 l/min/m²), but it did not differ from normal in Group II (table I).

LVEDV (fig. 3) was significantly larger than normal in Group I (mean = 247 ± 20% of normal) and in Group III (mean = 169 ± 15% of normal) and it was slightly, but not significantly, larger than normal in Group II (mean = 127 ± 13% of normal).

LV ejection fraction in all groups did not differ from normal (table I). Left ventricular systolic index (LVSI) (fig. 4) was larger than normal in Group I (mean = 10.04 ± 0.91 l/min/m²) and Group III (mean = 7.61 ± 0.69 l/min/m²) but it did not differ from normal in Group II.

**Discussion**

We have shown previously that right and left ventricular volume could be accurately determined in patients with left and right ventricular volume overload, as well as patients with RV pressure overload. The gooseneck deformity in the left ventricle could affect the accuracy of LV volume determination by the area length method. For this reason, LV volume was calculated by the area length and Simpson's rule methods. Volumes calculated by the Simpson's rule method were 1—8% larger than volumes calculated by the area length method. We felt that this error was within the limitations of the Simpson's rule method. LV volumes shown in table I and figures 1–4 are values calculated by the area length method. These
volumes, however, minimally underestimate volumes compared to those calculated by the Simpson’s rule method.

It should be noted that patients with complete AV canal and ostium primum defects have various degrees of AV valve incompetence which would cause an enlargement of the corresponding ventricle. RV volume was significantly increased in all patients with increased pulmonary blood flow (Groups I, III). These data are consistent with a previous observation and should be expected, since these patients have large left-to-right shunt at the atrial level, as well as left ventricular-to-right atrial shunts. The normal RV volume in patients with AV canal and pulmonary stenosis is also consistent with a previously reported normal RV volume in patients with tetralogy of Fallot or patients with pulmonary stenosis and atrial septal defect. These findings indicate that significant pulmonary stenosis in the presence of a large atrial septal defect or a large ventricular septal defect will cause a significant decrease in RV size. The RV ejection fraction in all groups was normal, but this finding should not be extrapolated to mean that RV contractility is normal in these patients. In the presence of large ventricular septal defect and AV valve incompetence, the afterload in these patients is less than normal. Furthermore, RV end-diastolic volumes are increased (increased preload). Thus, these two factors might lead to a normal RV ejection fraction in a patient with slight depression in myocardial contractility. The increase in RVSI in Groups I and III might be related to an increase in pulmonary blood flow and left-to-right shunt, as well as the incompetence of the AV valve and left ventricular-to-right atrial shunt. The normal RVSI in Group II, however, suggests that valvular pulmonary stenosis in these patients might decrease RV compliance, resulting in a reduction of RV filling and output.

The increased LVEDV in Group I is consistent with the previous observation which showed that unrestricted ventricular septal defect is associated with an increase in LVEDV. The increased LVEDV with normal LV ejection fraction in patients with ostium primum must be related to the incompetence of the AV valve. In patients with AV canal or ostium primum, Rudolph observed left ventricular-to-right atrial "obligatory" shunt. This type of shunt results in left and right ventricular volume overload and a significant increase in pulmonary blood flow. The increased LVEDV and output in 12 of the 17 patients with ostium primum defect supports this concept and confirms Rudolph’s observations. LV ejection fraction in all groups was not significantly different from normal, and for the same reasons discussed previously, this does not necessarily indicate that the contractility of the left ventricle is normal in these patients. LVSI (which represents forward systemic flow, a significant portion of the left-to-right ventricular and right atrial shunting) was significantly increased in Groups I and III but was normal in Group II. These findings are expected. They are also consistent with previous observations showing that left ventricular volume decreases in patients with ventricular septal defect and large left-to-right shunt in the presence of significant pulmonary stenosis.

These data indicate that the volume of the left and right ventricle is normal or increased in patients with isolated AV canal and ostium primum defect. It is conceivable, however, that a small sick infant with AV canal, or a child with pulmonary stenosis and AV canal, could have small ventricles. Goor et al. and Bharati and Levy showed that the commitment of the AV valve to one ventricle could lead to hypoplasia of the opposite ventricle. The number of patients in this study is somewhat limited, and the relationship between the AV valve and the two ventricles could not be determined. The data, however, indicate that the incidence of hypoplasia of one of the two ventricles must be rare. Mair and McGoon also suggested that a small left ventricle caused congestive heart failure in patients with AV canal after complete correction. This is certainly quite possible and is consistent with other findings that indicate LV hypoplasia might lead to a significant complication after complete correction of patients with tetralogy of Fallot. It should be noted, however, that the significant hypoplasia of the left ventricle (LVEDV less than 55% of normal) could lead to a significant decrease in cardiac output postoperatively, but this finding could not necessarily be extrapolated to patients with a small right ventricle when the right ventricular peak systolic pressure is normal. In these patients, atrial contraction contributes to pulmonary blood flow, and these patients tolerate RV hypoplasia much better than patients with LV hypoplasia.

Our data indicate that significant hypoplasia of the left ventricle in patients with partial or complete AV canal is uncommon. Furthermore, RV volume and output are also normal or increased in size with very few exceptions. However, due to the small number of patients in this study, no extrapolations as to the frequency of small or normal-sized ventricles in a large group of patients can be made. It is indeed the desperately sick, small infant that might benefit from studies of this type.

Acknowledgment

The authors wish to express their thanks to Mrs. Cathy Heteniak for the typing and editing of this manuscript.

References


Hemodynamic Effects of Intravenous Phentolamine in Low Output Cardiac Failure
Dose-Response Relationships

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SUMMARY Nineteen patients with chronic low output cardiac failure were studied before, during and after infusion of phentolamine in doses of 10, 20, 30 and 40 µg/kg/min. Significant reduction of left- and right-sided pressures and increases in cardiac index and heart rate (HR) were present within 15 minutes of starting phentolamine at the 10 µg/kg/min dose. Minimal additional effect was observed at 30 minutes. Increased dose from 10 to 20 µg/kg/min resulted in small but significant (P < 0.05) additional reduction in pressures and increases in HR. No additional significant changes occurred at doses of 30 or 40 µg/kg/min. Significant hemodynamic changes persisted for at least an hour (53 ± 3 min) after the phentolamine infusion was discontinued. Near maximal hemodynamic effects occurred within 15 minutes of starting phentolamine infusion and can be achieved at doses of 10 to 20 µg/kg/min. Increased HR during phentolamine infusion may limit its usefulness in patients with ischemic heart disease.

AFTERLOAD REDUCTION using vasodilators is becoming a widely used and valuable adjunct in the management of patients with severe heart failure. Sodium nitroprusside has become one of the more popular intravenous vasodilators. Although the acute clinical and hemodynamic responses to nitroprusside are frequently dramatic, its extended use over several days may be associated with toxicity due to accumulation of thiocyanate. Additionally, some patients do not respond adequately to nitroprusside. For these reasons, the efficacy of alternative vasodilators should be assessed in patients with severe heart failure. Among such agents, the alpha blocker phentolamine has been shown to exert beneficial hemodynamic effects when given intravenously to patients with heart failure. Previous studies have usually assessed the effects of phentolamine infused at a single dose. This study evaluates the acute hemodynamic effects of intravenous phentolamine infused at increasing dose levels in 19 patients with stable, chronic low output cardiac failure.

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Supported in part by the Medical Research Service of the Veterans Administration and Grant #M1178V, Ciba-Geigy Corporation, Summit, New Jersey.

Presented in part at the American Heart Association Annual Scientific Session, Miami Beach, Florida, November, 1976.

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Received December 12, 1977; revision accepted February 10, 1978.
Ventricular volume characteristics in infants and children with endocardial cushion defects.
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doi: 10.1161/01.CIR.58.1.153

*Circulation* is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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