Increased Mitral-Aortic Separation in Discrete Subaortic Stenosis

GLEN C. ROSENQUIST, M.D., EDWARD B. CLARK, M.D., HUGH A. MCALLISTER, M.D.
SAROJA BHARATI, M.D., AND JESSE E. EDWARDS, M.D.

SUMMARY We recently speculated that mitral-aortic separation (MAS) might be increased in discrete subaortic stenosis (DSS). We have examined this hypothesis in 22 heart specimens in which the subaortic obstruction originated on the muscular ventricular septum below the right aortic sinus, either as a discrete band, an accumulation of several bands or a diffuse ridge, and extended posteriorly into the MAS or anterior leaflet of the mitral valve or both, with a variable relationship to the aortic cusps and sinuses. No specimen had ventricular septal defect, supravalvular aortic stenosis or other features of Shone syndrome. The mean MAS was nearly twice that of 80 normal specimens (4.9 vs 2.5 mm), the range of MAS was increased from normal (0–11 vs 0–7 mm) and the mean diameter of the aortic annulus was decreased compared with the normal specimens, data that will be of interest to echo- and angiocardiographers in the clinical description of DSS, and to the surgeon who must resect these lesions.

IN DISCRETE SUBAORTIC STENOSIS (DSS), the left ventricular outflow tract is obstructed by a fibroelastic band or ridge of variable length, thickness, and relationship to the aortic valve.1–12 The lesion may extend onto the anterior leaflet of the mitral valve.

Recently we described a spectrum of discontinuity between aortic and mitral valves in normal heart specimens as well as those with ventricular septal defect, and speculated that mitral-aortic separation (MAS) might be increased in subaortic stenosis.13 The present paper examines this hypothesis in 22 heart specimens with DSS.

Materials and Methods

Of the 22 heart specimens with DSS, 12 were selected from the combined pathological collections of the University of Nebraska Medical Center, the Armed Forces Institute of Pathology and the United-Miller Hospitals; 10 specimens were from the Congenital Heart Disease Research and Training Center.

All specimens were from children or young adults, a number of whom had died after surgery to relieve the obstruction in the left ventricular outflow tract. At autopsy each specimen had been opened so that the aortic and mitral valve region could be studied as a unit.

In all specimens the subaortic obstruction was caused by fibroelastic tissue that originated on the muscular ventricular septum below the right aortic sinus (figs. 1–5). This tissue ranged from being a discrete band (figs. 1, 3 and 5) or an accumulation of several bands (fig. 1) to a diffuse ridge (fig. 2).

Posteriorly, the extension of this band or ridge had a variable relationship to the noncoronary sinus of the aortic valve; when the band or ridge was located close to the noncoronary sinus, the valve leaflet was thicker than normal, and sometimes connected to the obstruction by membranous extensions of the band (figs. 3 and 4). The band or ridge terminated in the tissue between the aortic and mitral valves (figs. 3 and 4) or extended onto the anterior leaflet of the mitral valve (fig. 1). In one specimen considered to have a circumferential diaphragm, the membrane extended under the left coronary sinus as well. Several specimens could be considered transitional between DSS and tunnel-type stenosis, because of the width of the fibroelastic ridge (fig. 2) and/or the degree of thickening of the anterior leaflet of the mitral valve, most marked on the posterior portion, and thick, short chordae tendineae.

Although the point of greatest thickening of the anterior leaflet of the mitral valve often lay opposite the band or ridge on the muscular ventricular septum, there was no consistent relationship between the two.

None of the specimens had ventricular septal defect, supravalvular aortic stenosis or additional features of the Shone syndrome14 (coarctation of the aorta, parachute mitral valve, supravalvular mitral stenosis, bicuspid aortic valve). Although the aortic leaflets were often thickened, the commissures were normal.

As in our previous investigation, we defined MAS as the shortest distance between the base of either the noncoronary or left coronary cusps and the mitral annulus; in all but one specimen the shortest distance was beneath the noncoronary cusp. Since the mitral annulus is the fibrotic junction of the left atrium and the anterior leaflet of the mitral valve,15 MAS corresponds to the aortic-mitral fibrous continuity defined by previous investigators studying normal and pathological material.16–17 Other measurements in-
In this child's heart with a 1-mm mitral-aortic separation (black arrows), the discrete subaortic stenosis (black and white arrows) consists of a flattened fibroelastic plaque along the ventricular septum (lower arrows, left), which merges with a flap or membrane that extends from the right coronary cusp (R) (arrows, upper left) and continues as a band onto a thickened anterior leaflet of the mitral valve (ALMV). L = left coronary cusp of the aortic valve; N = noncoronary cusp of the aortic valve.

The discrete subaortic stenosis in this specimen consists of a diffuse fibroelastic ridge under the right coronary cusp (R) (between arrows, left). The straight pin inserted from the left atrium marks the mitral annulus; the mitral-aortic separation is 7 mm. L = left coronary cusp; N = noncoronary cusp; ALMV = anterior leaflet of the mitral valve.

In this specimen a thick fibrous ridge under the right (R) and noncoronary (N) sinuses extends into a 6-mm mitral-aortic separation, leaving a cup-like depression between the aortic valve and crest of the ventricular septum. L = left coronary cusp; ALMV = anterior leaflet of the mitral valve.

Included depth of the anterior leaflet of the mitral valve from the annulus to the center of its free margin, depth of the noncoronary sinus between the middle of its base and the aortic valve annulus, and the circumference of the aortic and mitral valves at the annulus (fig. 6). Independent measurements were made by two observers and the results averaged because they did not differ more than 1 mm for MAS and 2 mm for other measurements. Measurements were compared with those from 80 normal heart specimens (15 children, 65 adults). Statistical comparison of the two groups was made using the paired t test.

Results

The circumference of the mitral valve and depths of the noncoronary sinus of the aortic valve and anterior leaflet of the mitral valve were similar in normal specimens and those with DSS (table 1). The circumference of the aortic valve was significantly less in DSS specimens than in normal hearts (45.7 mm vs 59.2 mm, p < 0.001). The mean MAS in DSS specimens (4.9 mm) was twice that of normal specimens (2.4 mm, p < 0.0001). The range of MAS was greater in DSS (0–11 mm) than in the normal specimens (0–7 mm).

Discussion

In a previous paper, we speculated that the MAS was wider in some normal heart specimens than in others because the regression or absorption of the bulboauricular or conoventricular flange in early heart development had been delayed. In some specimens, the MAS was 0; in others, it equaled one-third the diameter of the aorta.
FIGURE 4. In this child's heart, the discrete subaortic stenosis (arrows, upper left) involves the base of the aortic valve sinuses and thickened leaflets, but not the 4-mm mitral-aortic separation or mitral valve apparatus. R = right coronary cusp; L = left coronary cusp; N = noncoronary cusp; ALMV = anterior leaflet of the mitral valve.

FIGURE 5. The discrete subaortic stenosis in this specimen extends so close to the aortic valve that the base of the right (R) and noncoronary (N) leaflets are part of the shelf or membrane that extends into the 7-mm mitral-aortic separation. The mitral valve is slightly thickened. L = left coronary cusp; ALMV = anterior leaflet of the mitral valve.
An increase in MAS could contribute to the etiology of DSS if this alters the angle at which blood is ejected from the left ventricle during a critical period of early heart development. This, in turn, could cause the embryonic cells near the crest of the ventricular septum to accumulate and eventually differentiate into a ridge or band of fibroelastic tissue. A similar hypothesis has been proposed as the etiology for systolic pockets in the left ventricular outflow tract of the mature heart. Conversely, if the DSS is primary and present before the cells on the heart's inner curvature regress, an increase in MAS could result if absorption of the bulboventricular flange is delayed by altered blood flow or function in the embryonic heart. Likewise, the increased depth of the anterior leaflet in DSS suggests associated alterations of mitral valve morphogenesis. On the other hand, if the teratogenic influence resulting in DSS occurs after regression of the bulboventricular flange, the MAS could be normal.

The alternative argument that MAS is increased as the result of a decrease in the depth of the noncoronary cusp is refuted by the increase in this measurement in DSS compared with normals.

The finding that the aortic annulus may be smaller than normal in DSS suggests that fixed obstruction in the left ventricular outflow tract is a spectrum that may involve several anatomical sites. The smaller annulus may have resulted from proximity of the subvalvular obstruction to the valve or alterations in flow patterns during crucial periods of development.

Our studies may be of value to echocardiographers in the clinical description of discrete subaortic obstruction and to the surgeon who must resect these lesions. We plan to extend these studies to include tunnel subaortic stenosis, and the left ventricular outflow tract obstruction associated with the Shone syndrome. We expect to find a higher incidence of increased MAS and small aortic annulus in these anomalies as well.

Acknowledgments

The authors thank Lauren J. Sweeney for helping with measurements and for figure 6, and Jerry Lintz for statistical analysis.

References


---

**Table 1. Comparison of Measurements of Normal Heart Specimens from Children and Adults with Specimens with Discrete Subaortic Stenosis (DSS)**

<table>
<thead>
<tr>
<th></th>
<th>Normal specimens (n = 80)</th>
<th>DSS specimens (n = 22)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Range (mm)</td>
<td>Mean ± sd</td>
</tr>
<tr>
<td>Depth, anterior leaflet of the mitral valve</td>
<td>8–26</td>
<td>17.0 ± 4.4</td>
</tr>
<tr>
<td>Depth, noncoronary sinus of the aortic valve</td>
<td>7–19</td>
<td>14.2 ± 3.3</td>
</tr>
<tr>
<td>Circumference, aortic valve annulus</td>
<td>27.5–87</td>
<td>59.2 ± 16.7</td>
</tr>
<tr>
<td>Circumference, mitral valve annulus</td>
<td>41–114</td>
<td>74.3 ± 16.9</td>
</tr>
<tr>
<td>Mitral-aortic separation</td>
<td>0–7</td>
<td>2.4 ± 1.6</td>
</tr>
</tbody>
</table>
cases, one associated with patent ductus arteriosus, the other complicated by bacterial endocarditis. Int Assoc Med
Museums Bull 31: 73, 1950
5. Roberts WC: Pathologic aspects of valvular and subvalvular (discrete and diffuse) aortic stenosis. In Natural History and
1971, p 221
subvalvular aortic stenosis. Surgical experience and long-term
follow-up in 20 pediatric patients. Br Heart J 35: 443, 1973
Discrete subvalvular aortic stenosis in childhood. Am J Cardiol
38: 53, 1976
Thorax 13: 201, 1958
9. Walsh BJ, Connerty HV, White PD: Congenital subaortic
stenosis, with deformity of the aortic valve. Am Heart J 25:
837, 1943
caused by anomalies of the atriocentricular valves. J Thorac
Cardiovasc Surg 48: 278, 1964
11. Lillehei CW, Bonnabeau RC, Sellers RD: Subaortic stenosis,
diagnostic criteria, surgical approach and late follow-up in 25
343, 1942
13. Rosenquist GC, Clark EB, Sweeney LJ, McAllister HA: The
normal spectrum of mitral and aortic valve discontinuity. Cir-
culation 54: 296, 1976
14. Shone JD, Sellers RD, Anderson RC, Adams P, Lillehei CW,
Edwards JE: The developmental complex of parachute mitral
valve, supravalvular ring of left atrium, subaortic stenosis and
coarctation of aorta. Am J Cardiol 11: 714, 1963
15. McAlpine VA: Heart and Coronary Arteries. New York,
Springer-Verlag, 1975
16. Anderson RH, Becker AE, Losekoot TG, Gerlis LM:
Anatomically corrected malposition of great arteries. Br Heart
J 37: 993, 1975
17. Van Praagh R, Perez-Trevino C, Reynolds JL, Moes CAF,
Keith JD, Roy DL, Belcourt C, Wineberg PM, Parisi LF: Double
outlet right ventricle (S,D,L) with subaortic ventricular sep-
tal defect and pulmonary stenosis. Am J Cardiol 35: 42, 1975
18. Edwards JE, Burchell HB: Endocardial and intimal lesions (jet
impact) as possible sites of origin of murmurs. Circulation 18:
946, 1958
19. Maron BJ, Redwood DR, Roberts WC, Henry WL, Morrow
AG. Epstein SE: Tunnel subaortic stenosis: left ventricular out-
flow tract obstruction produced by fibromuscular tubular
narrowing. Circulation 54: 404, 1976

Status of Patients 5 or More Years After Correction
of Coarctation of the Aorta Over Age 1 Year

MARIUS M. HUBBELL, JR., M.D., RALPH G. O'BRIEN, PH.D., L. JEROME KROVETZ, M.D., PH.D.,
H. PAGE MAUCK, M.D., AND DOROTHY G. TOMPKINS, M.D.

SUMMARY In this retrospective study, we reviewed the records of patients who had coarctectomies at the
University of Virginia Hospital after 1 year of age. Follow-up data for 5 years or more after surgery were
available for 52 patients. Data from 23 similar patients from the Medical College of Virginia brought the total
postoperative sample size to 75. The blood pressure of this group of patients did not differ significantly from
that of the population at large. We conclude that successful repair of coarctation of the aorta in childhood or
early adolescence does not lead to a higher-than-expected incidence of resting hypertension in childhood.

REDUCTION OF SYSTEMIC blood pressure to normal or near-normal levels is a major therapeutic
goal in surgical correction of coarctation of the aorta. The attainment of this goal is controversial, although
significant reductions in resting systemic blood pressure are generally achieved. However, recent
reports vary in their conclusions about long-term results on hypertension.1 4 In this paper we report the
statistical analysis of blood pressure on 75 patients who underwent surgical repair of a coarctation after
age 1 year, with a minimum follow-up of 5 years. Systolic and diastolic blood pressure distributions
from these patients were compared statistically with those of the normal population.7 *

Premature death from cardiovascular disease after apparently successful surgical repair of coarctation has also become of more concern. A recent report suggests that the presence of associated congenital heart disease is a factor,6 and this was an important point in our analysis.

Methods

Patients younger than 1 year of age at the time of the initial operation were not included in this study,
since they had a high incidence of restenosis (32%) compared with only 2.6% in those requiring surgery
after 1 year of age. We obtained most of the data from the records of 111 male and 46 female patients who
underwent surgery to repair coarctation of the aorta at the University of Virginia Hospital (UVH) from
1954–1976. Age at operation ranged from 1–49 years, with a median of 13.5 years (fig. 1). Most (85%) of the

From the Departments of Pediatrics and Psychology of the
University of Virginia, Charlottesville, and the Department of
Pediatrics, Medical College of Virginia, Richmond, Virginia.
Address for reprints: Marius M. Hubbell, Jr., M.D., Department
of Pediatrics, Box 386, University of Virginia Medical Center,
Charlottesville, Virginia 22901.
Received June 30, 1978; revision accepted January 17, 1979. 
Circulation 60, No. 1, 1979.
Increased mitral-aortic separation in discrete subaortic stenosis.
G C Rosenquist, E B Clark, H A McAllister, S Bharati and J E Edwards

_Circulation_. 1979;60:70-74
doi: 10.1161/01.CIR.60.1.70
_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1979 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/60/1/70