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

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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.7,8

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,8 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

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Circulation 60, No. 1, 1979.
patients were less than 26 years old. Thirteen patients are known to have died (table 1). Despite repeated efforts to obtain further follow-up information, there has been no recent (since 1971) contact with 51 of the 144 patients. Follow-up data for 5 or more years after surgery were available in 52 patients.

In order to increase sample size for the examination of long-term postoperative blood pressure and electrocardiographic data, we reviewed the records of 23 similar patients from the Medical College of Virginia (MCV). Although the types of surgical repair were not tabulated, no patient in the follow-up study from either institution had clinical evidence of residual coarctation (systolic pressure ≥10 mm Hg or more in the arms compared with the legs).

Resting preoperative and postoperative blood pressures were obtained using standard techniques. In general, the fifth Korotkoff sound was used for diastolic pressures. The data were standardized by comparing both systolic and diastolic pressures relative to normal values for age and sex. We transformed pressures from patients younger than 18 years of age to "Z scores," using the means and standard deviations estimated from the curves given by Blumenthal; pressures from patients over 18 years of age were transformed using the norms of Master et al. Z scores are calculated by dividing the difference between the measured value and the normal mean by the standard deviation. Thus, a Z score is the number of standard deviations by which a measured value differs from the mean value for the age and sex of that patient. They are directly interpretable and are comparable across ages. Assuming a standard normal distribution, a Z score of 1.65 corresponds to the ninety-fifth percentile. The average Z score (the fiftieth percentile) is 0.0 for any age. For example, the popula-

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age at operation (years)</th>
<th>Preoperative blood pressure (mm Hg)</th>
<th>Age at death (years)</th>
<th>Other cardiac anomaly</th>
<th>Cause of death</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>3</td>
<td>135/80</td>
<td>3</td>
<td>Valvular aortic stenosis</td>
<td>At aortic valvulotomy</td>
</tr>
<tr>
<td>2</td>
<td>5</td>
<td>125/85</td>
<td>5</td>
<td>Valvular aortic stenosis</td>
<td>At aortic valvulotomy and coarctation repair</td>
</tr>
<tr>
<td>3</td>
<td>12</td>
<td>190/85</td>
<td>21</td>
<td>Valvular aortic stenosis</td>
<td>Bacterial endocarditis (aortic valve)</td>
</tr>
<tr>
<td>4</td>
<td>37</td>
<td>230/130</td>
<td>53</td>
<td>Valvular aortic stenosis</td>
<td>Heart failure 5 years postoperatively</td>
</tr>
<tr>
<td>5</td>
<td>49</td>
<td>210/85</td>
<td>53</td>
<td>Valvular aortic stenosis</td>
<td>Ruptured thoracic aneurysm</td>
</tr>
<tr>
<td>6</td>
<td>5</td>
<td>138/70</td>
<td>6</td>
<td>Mitral stenosis</td>
<td>Suicide (details unknown)</td>
</tr>
<tr>
<td>7</td>
<td>9</td>
<td>185/90</td>
<td>13</td>
<td>—</td>
<td>At reoperation 4 years later</td>
</tr>
<tr>
<td>8</td>
<td>18</td>
<td>170/90</td>
<td>18</td>
<td>—</td>
<td>Hemorrhage 1 month postoperatively</td>
</tr>
<tr>
<td>9</td>
<td>18</td>
<td>160/100</td>
<td>20</td>
<td>—</td>
<td>Auto accident</td>
</tr>
<tr>
<td>10</td>
<td>24</td>
<td>160/80</td>
<td>32</td>
<td>—</td>
<td>Drowned</td>
</tr>
<tr>
<td>11</td>
<td>25</td>
<td>200/100</td>
<td>25</td>
<td>—</td>
<td>Hemorrhage 3 months postoperatively</td>
</tr>
<tr>
<td>12</td>
<td>42</td>
<td>152/106</td>
<td>42</td>
<td>—</td>
<td>Intraoperative</td>
</tr>
<tr>
<td>13</td>
<td>47</td>
<td>220/110</td>
<td>52</td>
<td>—</td>
<td>At reoperation 5 years later</td>
</tr>
</tbody>
</table>
tion of normal 6-year-old children had a mean of 64 and a standard deviation of 9.8 for diastolic pressure. Thus, a 6-year-old child with a diastolic pressure of 80 would be given a Z score of \((80 - 64)/9.8 = 1.63\). The same Z score and percentile correspond to diastolic pressures of 86 and 90 for 10- and 14-year-old children, respectively. Such transformations are necessary to compare data from patients of different sex and age. The Z-score distributions were compared using Hotelling's \(T^2\) test (a parametric test).*

### Results

Thirteen patients are known to have died (UVH data) (table 1). Five deaths (patients 7, 8, 11–13) resulted from complications arising from the coarctation itself or from the repair. There was only one death resulting from a complication related to a coarctation among the 95 patients who were 15 years of age or younger at the time of surgery, whereas there were four among the 62 patients who were 16 years old or older. Thus, patients who were operated on after 15 years of age appear more likely to die as a direct result of the coarctation or its repair. (Fisher’s exact test, \(p = 0.08\).) In addition, patients 1–5 died from associated cardiovascular defects.

Only 29 of the 75 patients followed for 5 years or longer had no additional heart disease (table 2). Aortic valve disease was the most commonly associated lesion and was directly related to the deaths of five patients.

Preoperative blood pressures were available on 154 UVH patients. These data are shown in both scattergram (figs. 2 and 3) and standardized form (figs. 4 and 5). The average systolic and diastolic Z scores were 3.47 and 1.83, which were clearly abnormally high (Hotelling’s \(T^2\), \(p = 0.001\)). Shifting from Blumenthal’s data to Master’s data caused the discontinuities in the curves for the normal population included in figures 2, 3, 6, and 7.

Blood pressures were available 5 or more years postoperatively for 52 of these 154 UVH patients. Average preoperative pressures did not differ significantly (t test) between patients with and without complete postoperative data, indicating that no selection bias existed in the follow-up sample. The average decreases between pre- and postoperative Z scores were 3.15 (systolic), and 2.33 (diastolic), clearly indicating a reduction in hypertension (Hotelling’s \(T^2\), \(p = 0.001\)). (Some of this reduction is attributable to a regression to the mean effect.)

Postoperative blood pressure data for the combined sample of 52 UVH and 23 MCV patients are shown in both standardized (figs. 4 and 5) and in scattergram form (figs. 6 and 7). The Z scores were slightly below

---

### Table 2: Cardiovascular Diagnoses in Addition to Coarctation in the Follow-up Population

<table>
<thead>
<tr>
<th>Category</th>
<th>(n)</th>
<th>Subtotals</th>
<th>Totals</th>
</tr>
</thead>
<tbody>
<tr>
<td>No other congenital heart disease</td>
<td>29 (39%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other heart disease</td>
<td>46 (61%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aortic valve disease</td>
<td>25 (33%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Valvular aortic stenosis</td>
<td>7 (9%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bicuspid aortic valve (without stenosis or insufficiency)</td>
<td>4 (5%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Congenital aortic insufficiency</td>
<td>5 (7%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Valvular aortic stenosis and congenital aortic insufficiency</td>
<td>5 (7%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Valvular aortic stenosis and ventricular septal defect</td>
<td>1 (1%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Valvular aortic stenosis and patent ductus arteriosus</td>
<td>1 (1%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bicuspid aortic valve (without stenosis or insufficiency) and ventricular septal defect</td>
<td>1 (1%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bicuspid aortic valve (without stenosis or insufficiency) and congenital mitral stenosis</td>
<td>1 (1%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>7 (9%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ventricular septal defect and patent ductus arteriosus</td>
<td>3 (4%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mitral valve disease</td>
<td>7 (9%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Congenital mitral stenosis</td>
<td>2 (3%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Congenital mitral stenosis and patent ductus arteriosus</td>
<td>1 (1%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Congenital mitral insufficiency</td>
<td>2 (3%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Congenital mitral insufficiency and patent ductus arteriosus</td>
<td>1 (1%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Congenital mitral stenosis and insufficiency</td>
<td>1 (1%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>1 (1%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>3 (4%)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
normal for both systolic (−0.17) and diastolic (−0.49) pressures. Separate univariate t tests revealed that only the diastolic pressure was significantly below normal (t = 4.06, df = 73, p = 0.001). This result is qualified by the fact that the low diastolic Z-score means were absent in the older age group (≥18 years), which was standardized using Master's norms, where the fifth Korotkoff sound was taken to be the diastolic blood pressure (as in most of our patients). These norms are low relative to the Blumenthal norms, and therefore the lower postoperative diastolic pressures appear to be more normal in the older group, i.e., their Z scores will be larger. Only seven (9%) of the 75 patients had Z scores higher than the ninetieth percentile value of 1.28; six had high systolic pressure and one had a high diastolic pressure.

There was no evidence in our data that age at operation was related to the change (before vs after surgery) in blood pressure. The correlations of age at operation with the changes in systolic and diastolic Z scores were only 0.16 and 0.11, respectively. We performed simple and multiple correlation analyses to investigate possible interrelationships between age at surgery, length of follow-up and the various blood pressure measurements, but found nothing remarkable.

Examination of the 157 UVH records produced several subsidiary findings of interest. Four patients required reoperation: two (ages 7 and 9 years at initial surgery) for revision of the coarctation and two others because of a breakdown at the anastomotic site, one of which grew out a Mucor species. Five patients had preoperative cerebral vascular accidents at ages 5, 24, 26, 27 and 40 years. All but one (age 40 years) were caused by subarachnoid hemorrhage. Three had no congenital heart disease other than the coarctation. No patients are known to have had cerebrovascular accidents postoperatively. Three patients had bacterial endocarditis, two on the aortic valve and one on the anastomotic site; one of these patients (number 3) died. One additional patient is currently under investigation for a possible subclavian steal syndrome.

Discussion

Aortic valve disease is frequently found in association with coarctation of the aorta.10 A bicuspid aortic valve has been reported to occur in 25–85% of these patients.10–14 Not all of our patients had diagnostic quality aortograms to rule out bicuspid aortic valve disease, so the actual incidence of this anomaly may be higher. In this series, the ultimate prognosis after successful, uncomplicated surgical repair of coarctation depended largely upon the presence and the severity of other congenital cardiac anomalies; this finding was also noted in another series.2
Elective surgical repair is generally done in patients 4–6 years old. Patients over the age of 25 years at the time of operation have an increased risk of premature death due to cardiovascular causes. \(^1\) We found that patients age 16 years or older at the time of surgery were more likely to die as a direct result of their coarctation or its repair than patients 1–15 years old.

As expected, there was significant preoperative elevation of systolic and diastolic blood pressures. Postoperative blood pressures did not differ significantly from those of the general population. This was true for the UVH and MCV groups, taken alone or in combination. The slightly lower-than-normal diastolic blood pressure distribution may be a result of recording the fifth Korotkoff sound rather than the fourth as the diastolic blood pressure.

Because our results differ from those reported by Maron et al.\(^1\) and Nanton and Olley,\(^2\) we reanalyzed our blood pressure data using the methods of these two earlier studies. Patients in the Maron study were at least 2 years of age at time of surgery and were at least 19 years of age at follow-up. The minimum follow-up period was 11 years. Maron et al. classified their patients as hypertensive if either the systolic or diastolic pressure exceeded the ninetieth percentile of the norms published in the 1960–1962 U.S. National Health Survey.\(^16\) In our sample, 17 patients had the same ages and follow-up periods as the Maron sample, and none were hypertensive as defined by Maron. This is significantly less than the 22 of 59 patients reported by Maron et al. (chi square = 8.92, \(p < 0.005\)). Only five of our 42 patients (12%) who were 18 years or older were hypertensive according to the health survey norms. Except for one borderline

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**Figure 4.** Preoperative and postoperative systolic blood pressure Z-score distributions. The preoperative Z scores are from the total University of Virginia Hospital (UVH) operative population. The postoperative Z scores are from the UVH and the Medical College of Virginia study groups.

**Figure 5.** Preoperative and postoperative diastolic blood pressure Z-score distributions. The preoperative Z scores are from the total University of Virginia Hospital (UVH) operative population. The postoperative Z scores are from the UVH and Medical College of Virginia study groups.
case, these same patients were classified as hypertensive using the Z scores based on the Master norms. Thus, the discrepancy between the Maron study and the present study appears to be based on the data and is not merely a result of different methods.

Patients in the Nanton and Olley study were also at least 1 year old at time of surgery, but had a minimum follow-up period of only 6 months. Blood pressures were classified as hypertensive if they exceeded the ninetieth percentile of the Londe\textsuperscript{14} norms for patients younger than 15 years of age or the norms from the Society of Actuaries Build and Blood Pressure Study\textsuperscript{19} for ages 16 years and older. All 75 patients in our follow-up sample conformed to these age and follow-up requirements. According to these norms, 25 of our patients had systolic or diastolic pressures that exceeded the ninetieth percentile. This rate (25 of 75) resembles that (45 of 190) observed by Nanton and Olley (chi square = 2.57, \( p > 0.05 \)). Closer examination reveals that the normal data of the Society of Actuaries are much lower than either Master’s or the health survey’s norms. For example, the ninetieth percentile for the systolic pressure for 25-year-old men is approximately 141 according to either Master’s or the health survey’s norms, but is only 130.5 according to the Society of Actuaries report. Master et al.\textsuperscript{8} also found this to be the case in blood pressure data from insurance company studies. They concluded that this information is not epidemiologically useful because it attempts to define an ideal low-risk blood pressure base for life insurance purposes rather than to establish representative data for the general population. The fact that the Society of Actuaries norms are generally more than one-half of a standard deviation lower than other norms explains the higher rates of hypertension reported by Nanton and Olley. In fact, their patients seem to have had generally lower blood pressures.

Two other recent reports\textsuperscript{3, 4} state that unexplained hypertension was not a significant problem in their postoperative patients on late follow-up. In other recent studies, James and Kaplan\textsuperscript{6} found an appropriately high systolic blood pressure during graded submaximal exercise in nine of 14 patients 6 months to 15 years after correction of coarctation of the aorta, and Šamánek et al.\textsuperscript{14} found differences in blood flow between the upper and lower extremities in their patients 7–16 years after successful surgical repair. These studies were not done on any of our patients.

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 adulthood, and that subsequent morbidity is related to the presence of additional or associated cardiac lesions. Furthermore, the postoperative systolic blood pressure distribution did not differ significantly from that of the normal population, and the diastolic distribution was slightly (but significantly) lower, probably because the fifth Korotkoff sound was generally used, while the fourth was used in the Blumenthal norms.

References

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**Figure 6.** Postoperative systolic blood pressures for the University of Virginia Hospital (UVH) and the Medical College of Virginia (MCV) study groups. The values for the normal population (mean ± 2 SD) are indicated by the solid lines.

**Figure 7.** Postoperative diastolic blood pressures for the University of Virginia Hospital (UVH) and the Medical College of Virginia (MCV) study groups. The values for the normal population (mean ± 2 SD) are indicated by the solid lines.
Subxiphoid Two-Dimensional Imaging of the Interaltrial Septum in Infants and Neonates with Congenital Heart Disease

FREDICK Z. BIERMAN, M.D., AND ROBERTA G. WILLIAMS, M.D.

SUMMARY The intertrial septum (IAS) was studied by subxiphoid two-dimensional echocardiography (S2DE) in 88 infants under 12 months of age who weighed 1.2–9.1 kg. The IAS was adequately displayed in 87 of 88 patients. The morphology, presence and localization of defects in the IAS were evaluated by S2DE and retrospectively related to the findings at cardiac catheterization. In seven patients with no interatrial communication at cardiac catheterization, the IAS was straight, with an area of central thinning corresponding to the veil-like cover of the septum primum over the foramen ovale. The morphology of the IAS with a stretched, patent foramen ovale (56 patients) indicated the coexistence of a right or left ventricular volume or pressure overload, and was readily distinguishable from the IAS with a secundum type communication (13 patients). In patients with a stretched, patent foramen ovale and left ventricular overload lesions, the IAS was a nearly homogenous, curvilinear structure bowing into the right atrium, with a small area of septal dropout at the superior rim of the septum primum. In the presence of right ventricular overload lesions, the central defect of the foramen ovale was associated with a redundant flap valve of the septum primum billowing into the left atrium. In secundum type communications, the centrally located defect represented a deficiency rather than a redundancy of the septum primum. Balloon atrial septostomy (BAS) in 17 patients produced a secundum-type defect bordered by the flail remnants of the torn septum primum. Blalock-Hanlon septectomy (two patients) resulted in a large, posterior, sinus venosus-type communication which incorporated the preexisting BAS. Ostium primum defects (seven patients) were distinguished from the secundum lesions by their eccentric position in the IAS adjacent to the atrioventricular ring.

ASSESSMENT OF ATRIAL ANATOMY in newborns and infants requires determining visceroastral situs, identifying interatrial communication and recognizing the interatrial septal response to right-sided obstructive and left-sided volume-overload lesions. Current echocardiographic assessment of the atrial structures and the interatrial septum (IAS) is based on indirect inferences of right ventricular volume overload by M-mode⁸ and two-dimensional display of the IAS from the precordial and apical transducer positions.⁸ The views are compromised because the echo beam is not perpendicular to the posterior portion of the IAS, causing artifactual dropout of echoes from this portion of the septum which cannot be distinguished from actual defects.⁸ Imaging the atrial structures from the precordial or...
Status of patients 5 or more years after correction of coarctation of the aorta over age 1 year.

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